



Auditory Neuropathy Spectrum Disorder




How Does Normal Hearing Work?

The ear is divided into three main parts:


Outer ear	Middle ear	Inner ear
<ul style="list-style-type: none">• Consists of the pinna and ear canal.	<ul style="list-style-type: none">• Consists of the eardrum and three tiny bones called ossicles.	<ul style="list-style-type: none">• Consists of the cochlea, as well as the vestibular (balance) system.

- Sound travels through the outer, middle, and inner ear. All the parts of the hearing system need to work properly in order for us to hear normally.

The pinna picks up sound waves from the environment and sends them through the ear canal to the eardrum.



The sound waves cause the eardrum to vibrate, which sets the ossicles in motion.



This motion transfers the vibrations to the fluid-filled cochlea, which causes the tiny sensory cells (called outer hair cells “OHCs” and inner hair cells “IHCs”) to bend:

OHCs make soft sounds louder.

IHCs convert the vibrations into neural signals that are sent to the hearing nerve .

The hearing nerve sends these signals through the neural hearing pathway to the hearing center in the brain, which interprets them as sound.

Hearing loss happens when any part of the hearing system does not work properly.



What are the Different Types of Hearing Loss?

There are generally three types of hearing loss:

- **Conductive hearing loss:**
 - Occurs when there is a problem in the outer and/or middle ear, which prevents the proper transmission of sound to the inner ear.
 - This type of hearing loss is usually temporary, but may be permanent in some cases.

- **Sensorineural hearing loss (SNHL):**
 - Occurs when there is a problem in the inner ear (cochlea) and/or the hearing nerve, which prevents the proper transmission of sound to the rest of the hearing pathway.
 - This type of hearing loss is usually permanent.
- **Mixed hearing loss:**
 - Occurs when there is a problem in the outer and/or middle ear as well as the inner ear. It is a combination of both conductive and sensorineural hearing loss.
 - The sensorineural component is usually permanent, while the conductive component may be temporary or permanent.



What is Auditory Neuropathy Spectrum Disorder (ANSD)?

ANSD is a less common type of SNHL, where there is a problem in the way sound is transmitted from the cochlea (in the inner ear) to the brain.

In ANSD, sound reaches the cochlea normally, but the timing or pattern of transmission becomes disrupted and does not travel normally along the hearing pathway to the hearing center in the brain.

It usually affects both ears, but may affect one ear only.



When Does ANSD Occur?

- **ANSD can occur at birth both in newborns with serious health problems who had to stay in the Neonatal Intensive Care Unit (NICU) for some time, as well as newborns who were born healthy (without complications).**
- **Although ANSD mostly occurs during childhood, it can occur at any other age.**



What Causes ANSD?

The reasons are often unknown, but there are several factors that may increase the risk for ANSD.

Risk Factors During Infancy or Early Childhood:

- Inherited (genetic) reasons, even if there is no history of hearing loss in the family
- Premature birth (<37 week of pregnancy); the lower the number of weeks the greater the risk
- Absent or underdeveloped hearing nerve
- Delayed development of the hearing nerve
- Very low birth weight
- Lack or insufficient supply of Oxygen at birth
- Severe jaundice (due to high levels of bilirubin in the blood)
- Certain infections/illnesses (e.g. cytomegalovirus, mumps, meningitis)
- Certain neurological disorders (e.g. cerebral palsy)
- Treatment with certain medications which may affect hearing

Risk Factors During Middle/Late Childhood or Adolescence:

- Some cases of head injury/trauma
- Certain syndromes and neurodegenerative diseases (e.g. Friedrich's ataxia, Charcot-Marie-Tooth (CMT) disease, Guillain-Barre syndrome (GBS), Deafness-Dystonia-Optic Neuropathy (DDON) syndrome (also known as Mohr-Tranebjaerg syndrome), Stevens-Johnson syndrome, Ehlers-Danlos syndrome, Leber's Hereditary Optic Neuropathy)



How is it Different from Common SNHL?

Common SNHL:

The problem typically occurs when the OHCs or both OHCs and IHCs are damaged or missing. When they do not work properly, the sound is not transmitted properly to the hearing nerve.

ANSD

The OHCs typically work properly, but there is a problem in one or more other parts of the hearing pathway after that point. When this happens, the sound is not transmitted properly to the brain.

The location of the problem may be in the IHCs, connection between the IHCs and the hearing nerve, hearing nerve (or its nerve cells), brainstem, or a combination of these parts.

Because of these potential different problem locations, ANSD can affect children in different ways, and its symptoms can vary from one child to another. That is why it is called a 'spectrum' disorder.



How Does ANSD Affect Hearing and Understanding?

- **Because the quality of transmitted sound may be poor, the information reaching the brain is not always easily understood.**
- **This affects the child's ability to hear and understand sounds, including speech, which can result in delayed speech and language development based on the child's age.**



Hearing Ability

- **Hearing levels may vary from normal hearing to profound hearing loss.**

- **The child's responses may be inconsistent (the child may sometimes appear to hear and understand, and sometimes not).**
- **Hearing may remain the same or may change over time (it may get better, worse, or fluctuate from day to day or even throughout a day).**



Speech Understanding Ability

- **The child may have no difficulties**
- **The child may have difficulty distinguishing sounds (cannot tell differences between them)**
- **The child may be able to hear speech, but in an unclear or distorted manner (e.g. like a**

poor phone reception or a fuzzy radio station with interference)

- **Abilities are highly variable; and cannot be predicted from the hearing level**
 - **May be similar to a child with common SNHL of the same hearing level (the difficulties are expected from the hearing level)**
 - **May be very poor compared to the hearing level (the difficulties are more than what is expected from the hearing level)**

Hearing and Understanding in Noise

- **The child may have difficulty in noisy situations only**

- **The child may have difficulty in both quiet and noisy situations**
- **Difficulties may range from mild to severe**



When is ANSD Identified?

- **ANSD can be identified at any age, even in newborns.**
- **Most children with ANSD are born with it and can be identified in the first few months of life.**
- **Depending on the type of test used during newborn hearing screening, some newborns with ANSD may pass the initial screening at birth.**
 - **This may delay identification of the disorder, as the hearing problem may**

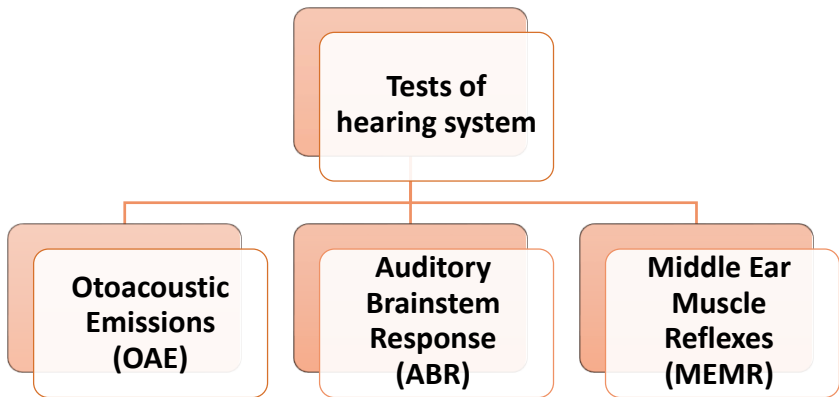
not be suspected until several months later based on the parents' observations of their child's responses to sounds.

- **Other children may develop symptoms at older ages or have a progressive onset and may not be identified until middle/late childhood or adolescence.**



How is ANSD Diagnosed?

- **The audiologist will perform different types of hearing tests to find out if the child has ANSD.**
- **These tests measure how well different parts of the hearing system are working, and they are harmless and painless.**



Otoacoustic Emissions (OAE):

- **Tests the response of the OHCs in the cochlea (in the inner ear).**
- **For children with ANSD, the results are usually present and normal, because the problem happens after the OHCs.**
- **In some cases, and over time, the OAEs may naturally become reduced or absent.**

Auditory Brainstem Response (ABR):

- **Tests the function of the hearing nerve and part of the hearing pathway to the brainstem, and provides an estimate of hearing level in cases of common SNHL.**
- **For children with ANSD, the results are usually absent or severely abnormal, even if the child shows responses to sounds, because the sound is not traveling through the hearing pathway in a coordinated way. Therefore, ABR cannot provide an estimate of hearing levels in cases of ANSD.**
- **Part of the ABR test also checks how the OHCs are working. This can be used to help diagnose ANSD in children whose OAEs are no longer present.**

Middle Ear Muscle Reflexes (MEMR):

- **Tests a tiny muscle in the middle ear (behind the eardrum) and the hearing nerve in response to loud sounds.**
- **For children with ANSD, the results are usually absent because the nerve cannot properly stimulate that muscle.**
 - **The combined results of these tests help diagnose ANSD, but they cannot predict the child's hearing levels or how the child will respond to sounds.**
 - **It is important to know that even if ANSD is identified early, it will take time, further testing, and frequent visits to measure the child's hearing levels.**



What Happens after Diagnosis?

- **When your child is first diagnosed with ANSD, it is usually not possible for the audiologist to say how severely the ANSD will affect your child's hearing.**
- **The audiologist will usually tell you that you need to wait until your child is old enough (around 6 or 7 months old) and developmentally able to undergo a behavioral hearing test to measure your child's hearing levels.**
- **Your child will then be referred to a speech-language pathologist (SLP) to assess their communication skills and provide you with**

counseling and training strategies that you can apply with your child.

- **Your child may also need to be referred to other specialties such as Genetics, Neurology, Ear-Nose-Throat (ENT), and/or Radiology for further investigations, if needed.**



How are Hearing Levels Measured?

Behavioral Hearing Test: is a test aimed to determine your child's hearing level (the softest level of sound they can hear), based on their behavioral responses to different sounds during the test, which the audiologist does by monitoring your child's reactions to these sounds.

- **Young children may not be able to tell us if they heard a sound, but they will show a reaction when they hear it, by looking at or turning to the sound source, smiling, or stopping what they are doing when they hear the sound.**
- **With older children, a conditioned play task can be used (e.g. putting a toy in a box when a sound is heard).**

Although it may be frustrating for you as a parent to wait during this period, it is important to understand that this is in the best interest of your child for several reasons:

- **This type of test is usually possible when your child is 6 or 7 months old, but for some**

children, it may not be possible to get reliable responses until later.

- **Multiple visits will be required using more than one testing technique in order to get all the information needed to confirm your child's hearing levels and monitor any changes over time.**
- **Even when your child is old enough, the obtained hearing levels give us little if any information about how well your child will receive and develop speech and language.**
 - **Speech understanding abilities cannot be predicted from hearing levels.**
 - **Even if your child's hearing level is normal, the sound reaching the brain may not be clear or may not make sense.**

- **Getting an idea of your child's speech understanding will likely not be possible until they are older.**

This means that we cannot predict the outcomes (how much your child will hear, whether their hearing will change over time, how well they will be able to understand or learn speech and language, how they will communicate in the future).



Because of this, your child's hearing needs to be tested regularly.



Can ANSD Improve Naturally?

- **In some children diagnosed with ANSD, especially those who are born prematurely,**

hearing may improve during the first year or two of life.

The reason for that is:

- **In these children, the hearing nerve pathway will gradually continue to develop with time, resulting in an improvement in hearing ability.**
- **The development of the hearing nerve pathway happens between 26 to 29 weeks of pregnancy, and some of these children are born before this process is complete.**
- **There is no way to tell whether or not this will happen with your child.**

- **It is therefore important to closely monitor your child's auditory responses and test their hearing on a regular basis.**
- **The child's hearing may recover fully, partially, or may not change at all.**



How is ANSD Managed?

- **There is no medical treatment for ANSD at the current time. However, there are ways to help manage the hearing difficulties and its effects on speech and language development.**
- **Management of hearing difficulties and speech and language development in children with ANSD is challenging because hearing levels can fluctuate, and speech understanding difficulties may persist even after management.**

- **The main goal of management is to provide good access to sounds to improve your child's hearing and help develop speech, language, and communication skills.**
- **The type of management will depend on the cause (if known), degree of hearing loss (once confirmed), and child's age at diagnosis; and may include the following:**

Hearing Aid

- **An electronic device that is typically worn behind the ear. It amplifies sounds and helps improve the child's ability to hear.**

Cochlear Implant

- **An electronic device that consists of two parts; an external part that is typically worn behind the ear, and an internal part that is surgically implanted under the skin (in the skull bone). It sends electrical stimulation directly to the hearing nerve to improve the child's ability to hear.**

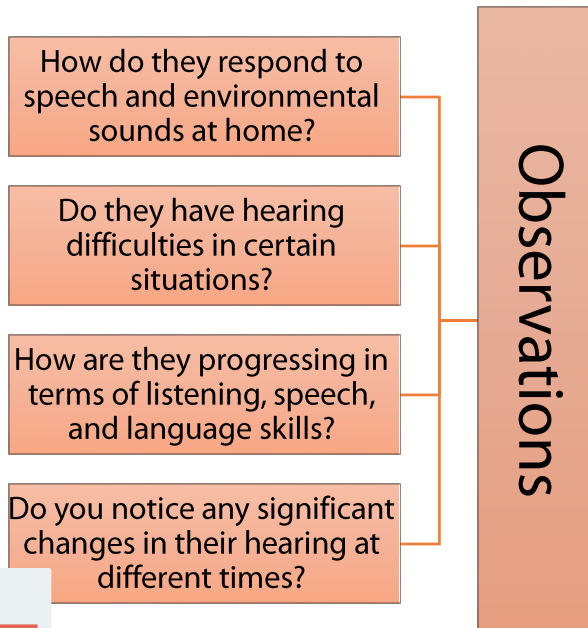
Assistive Listening Device:

- **An electronic device that can assist in hearing and understanding speech in noise. It may be recommended alone or in combination with a hearing aid or cochlear implant.**



Which Management Option is Best for My Child?

- **To provide the best possible management, the audiologist first needs to establish consistent and stable hearing levels.**
- **To help the audiologist choose the most suitable option for your child, it is important for you to share your observations about your child:**





When Will My Child Be Fit with Hearing Aids?

- **When reliable behavioral hearing test results are obtained and hearing loss is confirmed, hearing aids will be the first management option, regardless of the degree of hearing loss.**
- **It is recommended that your child be fit with hearing aids as early as possible. The hearing aids will be programmed based on the behavioral hearing test results.**
- **If your child was born prematurely, there is a possibility of spontaneous improvement in hearing; therefore, hearing aid fitting is often delayed further and/or monitored more closely.**



Will My Child Benefit from Hearing Aids?

- **Because we cannot predict how your child's hearing will develop, we have to proceed gradually and with caution. For this reason, the audiologist may set the hearing aids at lower settings than your child's hearing levels, and gradually increase the settings over time.**
- **There is no way to predict how your child will perform with hearing aids without a trial period. This is why you should make sure that your child wears the hearing aids during all waking hours.**
- **Some children may have good benefit, and some may have limited to none.**

- **Hearing aids might make sounds louder but may not solve the problem of making them clearer, and your child may still have difficulty understanding speech.**
- **After hearing aid fitting, your child's hearing levels will continue to be checked on a regular basis to assess hearing aid benefit.**
- **Your child's speech and language development will also be regularly monitored.**
- **Some children, even with well-programmed hearing aids, cannot benefit from the sounds they hear, and might continue to have difficulty discriminating and**

understanding speech, and therefore cannot learn to speak normally.

- **If your child makes little progress in speech and language development and understanding, even with appropriate hearing aids, then the option of cochlear implantation may be considered.**



Does My Child Need a Cochlear Implant?

- **If your child has a severe to profound hearing loss, has gone through a trial period of hearing aid use, and showed limited or no benefit in terms of hearing level, speech and language development, and understanding, the audiologist will discuss the option of cochlear implantation.**

- **Your child would then need to be evaluated by an ENT doctor and would need to undergo a CT scan and/or MRI to check if the hearing nerve is intact.**
- **In some cases, children with ANSD who have hearing losses which are better than severe to profound (moderately-severe for example), might be candidates for a cochlear implant, if they are not showing enough benefit from the hearing aids.**



Will My Child Benefit from a Cochlear Implant?

- **Children with ANSD may show similar benefit from cochlear implants as children with common severe to profound SNHL.**

- **This is because the direct electrical stimulation from the cochlear implant to the hearing nerve helps overcome the problem of sound transmission caused by ANSD.**
- **How well your child performs with cochlear implants will depend on the cause of ANSD (which is often unknown), your child's current age, speech and language skills, previous hearing aid use, age at implantation, compliance with device use, and aural (re)habilitation both before and after implantation.**



How Will My Child Develop Speech and Language Skills?

- **After hearing aid fitting, your child will be referred to a SLP specialized in aural**

rehabilitation to assess, facilitate, and closely monitor your child's development in speech and language, communication, and listening skills.

- Early intervention and regular follow-up are important to learn how your child hears and understands speech, and also to get the maximal benefit from hearing aids or cochlear implants.**
- The SLP will assist you in developing effective language environments for your child by advising you to talk to your child, teaching your child how to speak, and counseling you on when using gestures and/or visual cues might be needed.**

- **The SLP plays an important role in determining the benefit and progress from hearing aid or cochlear implant use by reassessing your child's communication skills after the hearing management, therapy sessions, and consistent home training.**



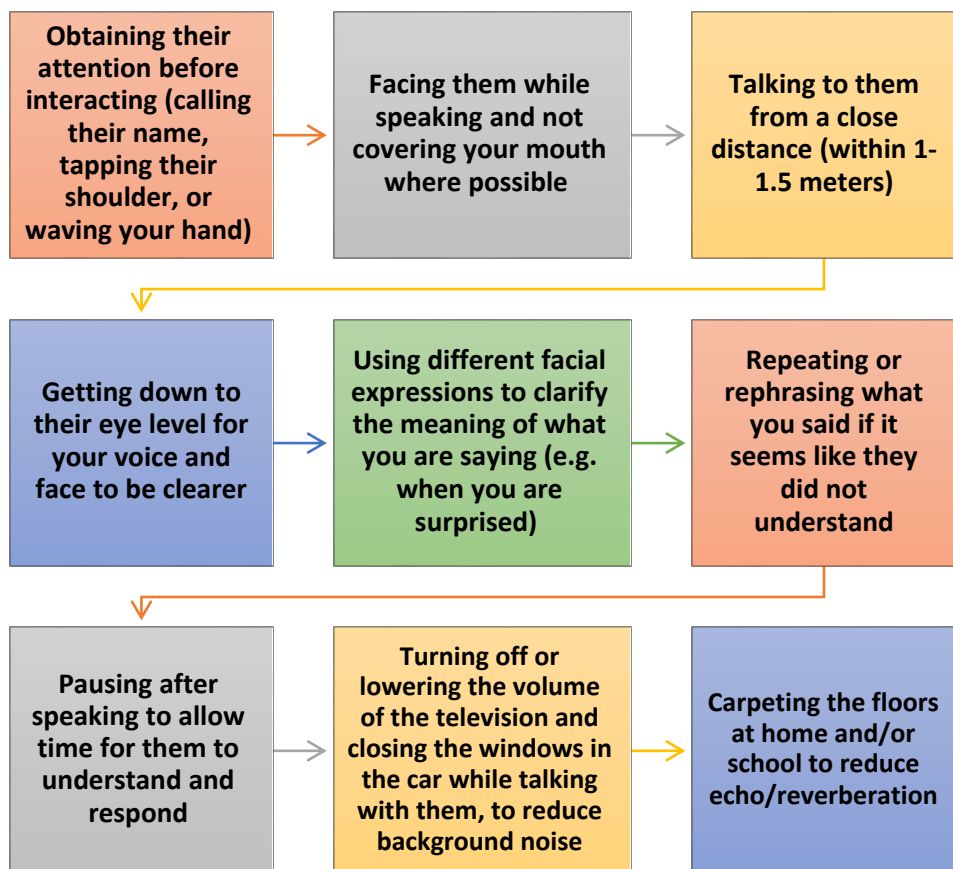
Will My Child Develop Normal Speech and Language Skills?

- **Many children develop good speech, language, and communication skills with hearing aids or cochlear implants, aural rehabilitation, and speech and language therapy.**
- **It is important that you play an active role in the rehabilitation process and apply the therapy techniques at home and**

throughout the day, as you will be spending much more time with the child than the SLP.

- **When interacting with your child, try to improve the listening environment in order to facilitate communication and help them make the most of the speech that they hear.**

Tips to improve the listening environment and facilitate communication with any child with hearing loss:





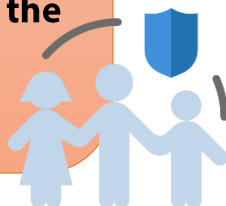
What if My Child Does Not Develop Normal Speech and Language Skills?

- **Not all children require the same approach to learn and improve their listening, speech, and understanding skills. If your child is having difficulties understanding spoken language, the SLP may need to try a different approach and evaluate its outcome over time.**
- **You might be advised to use other communication approaches, such as sign language (visual communication) or a combination of sign and speech (total communication), as needed.**
- **The use of a visual communication approach in addition to spoken language, might be beneficial to ease your child's**

frustration at not being able to effectively communicate, especially when they are very young.

- **Despite the challenges that you might face throughout your child's speech and language and hearing development, it is important to be flexible and involved throughout this process. The audiologist and SLP will work together to come up with the most suitable plan for your child and guide and support you throughout the process.**

For any child with hearing loss and/or speech and language issues, proper diagnosis and early intervention are essential, but family involvement in the rehabilitation journey is key!



لأن الوعي وقاية ..

إدارة التشخيص الصحي

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