MY CHILD
WITH
AUDITORY NEUROPATHY SPECTRUM DISORDER

A Handbook for Parents

THE UNIVERSITY OF MEMPHIS

MY CHILD WITH Auditory Neuropathy Spectrum Disorder

A Handbook for Parents

Section

THE UNIVERSITY OF MEMPHIS

I. LETTER TO PARENTS

With good audiological management, children with hearing problems are able to function well with their hearing peers.

Dear Parents,

Your child has had their hearing tested. The audiologist who conducted your child's audiological testing is a Doctor of Audiology who is a trained professional with expertise in the function of the auditory system. The audiologist has determined that your child may have a hearing problem called auditory neuropathy spectrum disorder (ANSD).

We have created this handbook for you, the parent, to provide useful information to help you become more familiar about hearing and hearing disorders. It is a brief introduction to the parts and functions of the auditory system and problems that can occur within it. This is intended to guide you through the process of understanding your child's diagnosis of ANSD.

As we approach the management of your child's hearing loss, it is important for you to understand what ANSD is and how we come to distinguish it apart from other types of hearing loss. Upon reading the information presented, our goal is to help you:

- ☑ Understand your child's hearing.
- ☑ Become aware of all the management options available for ANSD.
- ☑ Have access to available resources.
- ☑ Become and remain involved in your child's audiological and educational management.

The team at The University of Memphis is here to help you in the process of managing your child's hearing and to provide you with information and choices you have. It is your decision to choose what is right for you, your family, and your child. We are here to work together in the process of helping your child to maximize their abilities and to communicate effectively.

This handbook will also help you understand the testing procedure we use to diagnose ANSD. It includes information on treatment of ANSD, and communication options. Although the team stands ready to assist you at any time, this handbook will help you to understand

your child's needs. We encourage you to contact anyone of us with any questions and/or concerns that you may have.

Here are team members you might want to contact with any questions:

Jennifer P. Taylor, AuD,CCC-A ... Audiologist.....jptaylr2@memphis.edu
Hannah Beth Scott, AuD,CCC-A.....Audiologist.....hannah.beth.scott@memphis.edu
Jordan Alyse Coffelt, AuD,CCC-A . Audiologist.....j.coffelt@memphis.edu
Casandra Banks, AuD,CCC-A ..... Audiologist.....cbbanks@memphis.edu
Matthew Hollis, AuD,CCC-A ..... Audiologist.....mhollis@memphis.edu

---

We look forward to helping you make decisions regarding your child in order to maximize and realize their potential.

Thank you,
The Auditory Neuropathy Spectrum Disorder Team
The University of Memphis Pediatric Auditory Research Laboratory

❖ Terms in bold print can be found in the glossary at the end of the handbook. ❖

II. HOW WE HEAR

The 3 parts of the ear

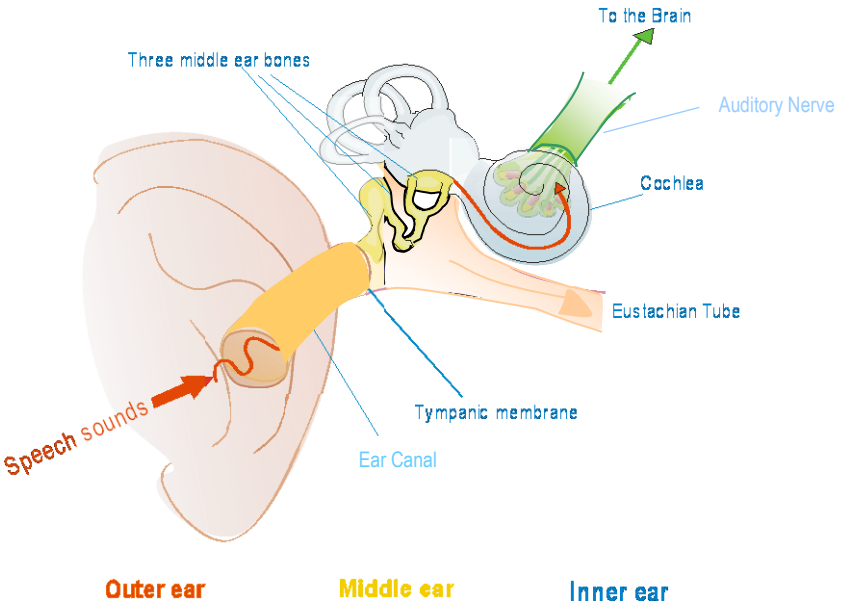
The ear consists of three main parts:

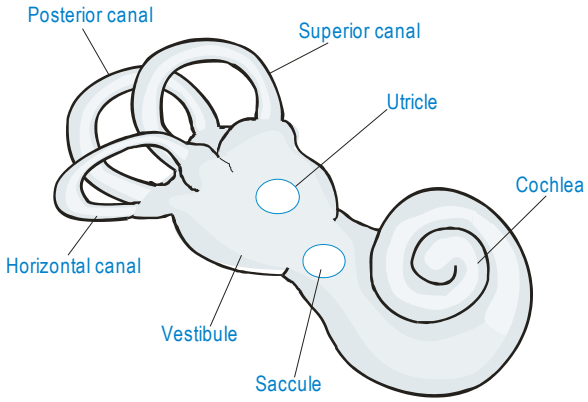
1. The outer ear	The part you see, which is called the auricle (**ohr**-a-kal).
2. The middle ear	The middle ear is a sealed area, with the eardrum on one end and the cochlea (**ko**-klee-uh) on the other. Inside this area are the three bones of the middle ear, also called the ossicles (**ah**-sih-kulz), and the muscles attached to them.
3. The inner ear	The part that is deep within the ear, incased in the temporal bone of the skull, is called the cochlea. The cochlea contains the tiny hair cells that trigger a signal to the auditory nerve, which takes the information to the brain. The inner ear also contains the vestibular (ve-**sti**b-ew-lar) system, which helps us maintain our sense of balance.

The Ear in Action

The Outer and Middle Ears

The outer and middle ears serve to capture sound and send it to the cochlea, where the mechanical vibrations are turned into electrical impulses that travel along the hearing nerve to the brain. The most common problems to occur here are outer or middle ear infections, both of which are easy to treat with antibiotics or minor surgery and rarely cause permanent hearing loss.





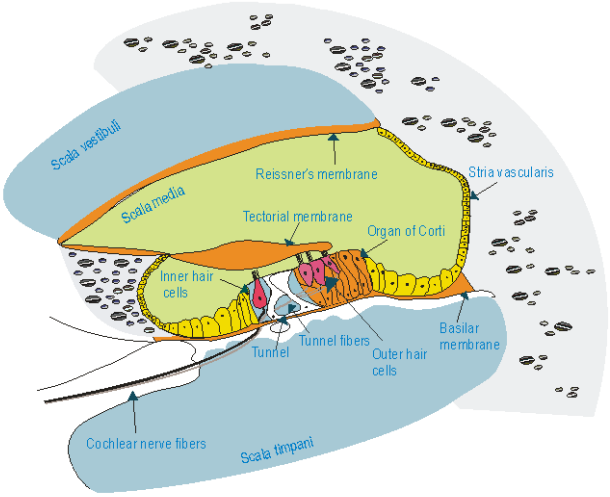
The Cochlear and Vestibular Systems

The Inner Ear

The inner ear contains the cochlea and the vestibule. The cochlea is the organ of hearing. The vestibular system is attached and controls our sense of balance. These two are connected and affect each other. When one is experiencing difficulty, it is not uncommon for the other to experience difficulty.

The cochlea

The cochlea has two types of hair cells. Each plays a role in hearing. The outer hair cells serve to help make sounds that come into the ear louder. They are a natural amplifier for the ear. The inner hair cells' job is to trigger impulses to send this signal to the auditory nerve.



Cross section of the cochlea

The ***outer hair cells*** are known as the "natural amplifier" of the ear. They amplify soft sounds so that they can be picked up by the ***inner hair cells***. Sometimes the ***outer hair cells*** may not

work, which results in a hearing loss. Outer hair cell loss results in the inability to hear soft sounds. This type of hearing problem is managed well with hearing aids and is often referred to in the medical community as sensorineural hearing loss.

The ***inner hair cells*** are known as the sensory transducers. Loud sounds or soft sounds that have been turned up by the ***outer hair cells*** activate the ***inner hair cells*** which send a signal to the hearing portion of the vestibulo-cochlear, or VIIIth nerve. That signal travels through the auditory system to the brain, where it is interpreted.

In normal hearing individuals these two types of hair cells work together to create a signal that is picked up by the auditory nerve and carried to the brain for interpretation. In a typical case of ANSD, the outer hair cells work well. There is a problem, however, with either the inner hair cells, the connection between the inner hair cells and the auditory nerve (the synapses), or the auditory nerve itself.

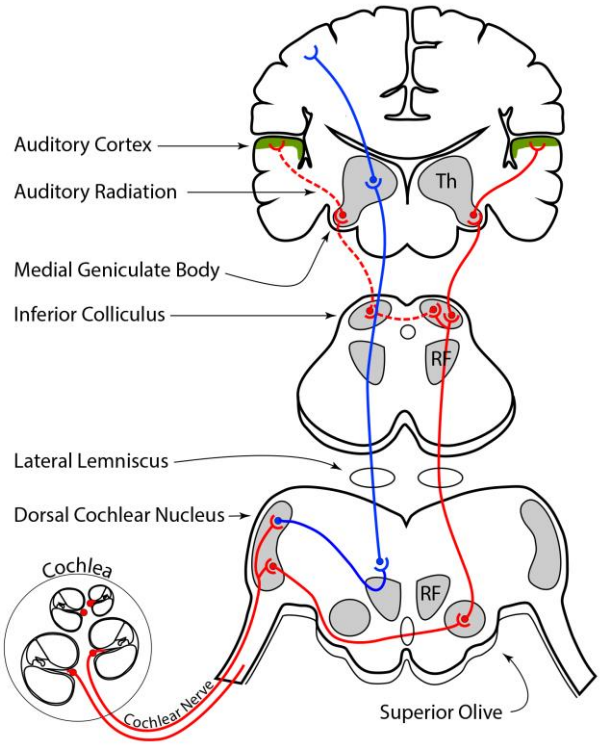
Because of this problem, the organ of hearing is unable to send a clear signal to the brain for interpretation. The signal that arrives at the brain for interpretation is not received in an organized fashion. Parents often report that their child appears to hear at times, but not at other times. The child may alert to sounds at times, but not consistently.

In some cases, the sound is not received at all. In severe cases of ANSD, all sounds seem the same to the brain. They all sound like static or simply noise. Such children can sometimes detect sounds but cannot tell what the sound is. A voice may sound the same as water running, a dog barking may sound the same as a car horn, a bird chirping may sound the same as a pan banging. Because of ANSD, the child is unable to make sense of sounds in thier world.

Some children with ANSD do develop some speech and language without intervention. However, as the child gets older a speech and language delay often becomes apparent. Listening in a noisy environment is always very difficult for people with ANSD.

The Auditory Nerve and Beyond

The Auditory Nerve transmits sounds from the inner ear to the brain. It starts in the cochlea in the inner ear and travels to the auditory cortex. The frequency, timing, and intensity of the signal is kept throughout the system so when the signal reaches the auditory cortex it can be understood.



The majority of children with ANSD need intervention in order to develop speech and language through the auditory channel without any delay. Some children with ANSD will need intervention to be able to develop speech and language at all. There is a critical period for language development between the ages of 0 and 3 years. It is important to pay close attention to your child's progress, or lack of progress, during this time since their auditory system needs the correct stimulation in order to develop spoken language.

The ANSD Team at The University of Memphis Pediatric Auditory Research Laboratory will be able to help you manage your child's hearing.

III. AUDIOLOGICAL TESTING

LISTEN…….to your Audiologist!!

Your audiologist is trained to measure your child's hearing thresholds. They can determine the type and degree of hearing problem and will make recommendations based on the results of the audiological testing. There are several audiological tests used to make decisions on how to move forward with treatment of your child's hearing loss.

Testing the Outer and Middle Ear

The outer ear is visualized by using an otoscope. ***Otoscopy*** reveals the ear canal and eardrum. ***Tympanometry*** is a type of test used to measure the flexibility of the eardrum. This can determine if there is fluid behind the ear drum or if other problems within the middle ear space exist. The muscles within the middle ear protect the ear from loud sounds. ***Middle ear muscle reflex*** testing is used to measure how loud a sound must be for those muscles to contract. This is often used as a screening tool for auditory neuropathy spectrum disorder (ANSD). ***Tympanometry*** and ***middle ear muscle reflexes*** are obtained by placing a small tip at the opening of the ear to apply light pressure and measure small pressure differences within the canal. All three tests are routinely used to assess the ear canal, eardrum, and middle ear and take only a few minutes to perform.

Testing the Inner Ear

Otoacoustic Emissions (OAEs)

One of the most powerful tools we have to test the outer hair cells are ***otoacoustic emissions*** (OAEs). OAEs test the function of the outer hair cells, or the "natural amplifiers" which lie inside the cochlea. For this test the audiologist places an instrument into the child's ear and presents sound. Then the instrument notes the echo that is created by the inner ear. In individuals with normal hearing this echo is robust. In cases of sensorineural hearing loss, the OAEs will be reduced or absent altogether. In case of ANSD the OAEs are usually present. This test is the primary test used for newborn hearing screenings.

Testing the Brain

Auditory Brainstem Response (ABR)

This test is also often part of the newborn screening process but can also be used as a full diagnostic test. If your child needs further testing beyond the hearing screen, he will be referred for an ABR. He will need to be asleep or resting quietly during this test. Sedation is often used to help older babies sleep through the procedure. The audiologist will place electrodes on your child's head and present sounds into your child's ear. The electrodes will pick up brain activity. In this way the audiologist can determine how loud the sounds have to be in order for the brain to detect them, and if the lower brainstem is receiving the information in a clear manner.

Cortical Auditory Evoked Potential Testing

Cortical Auditory Evoked Potentials (CAEPs) are used to tell if your child is processing sounds in their brain (at the level of the auditory cortex). Testing will be completed with your child awake. Electrodes will be placed on your child's forehead and ears to record their brain response while your child is listening to speech sounds at various loudness levels. This test can be done at any age. Because young children can't reliably tell us how they are hearing this test can be useful to figure it out for them. It can also tell us what level of sound they are able to process within the auditory portion of their brain. This can help you come up with the best intervention plan possible.

Other Important Tests

Genetic Testing

ANSD could be caused by many different factors, but genetics is one area that could be a possible cause. Even if ANSD is not part of your family history, it could still be caused by a genetic mutation. A genetic mutation exists when there is something wrong with a gene. Several genetic mutations are known to cause ANSD. Genetic testing is an important part of the diagnosis process to help pinpoint the cause of ANSD and help find the best intervention plan for your child.

MRI

An MRI will also be important for the diagnosis of ANSD. The MRI will take pictures of the inner ear and will evaluate if its shape is normal and if the auditory nerve is present or absent or very small.

THE AUDIOGRAM

The Audiogram

An audiogram is recorded through behavioral testing in a sound-proof booth. Children must be old enough to take this type of test in order for the results to be considered reliable. The audiologist presents sounds to the child through speakers or earphones and records the responses. A very young infant (6-24 months) will be tested using a conditional response called visual reinforcement audiometry (VRA), where as an older child can drop a block in a bucket when he or she hears a sound. The audiologist presents sounds of varying pitch, either high or low sounds, and then sees how loud the sound must be for the child to detect it.

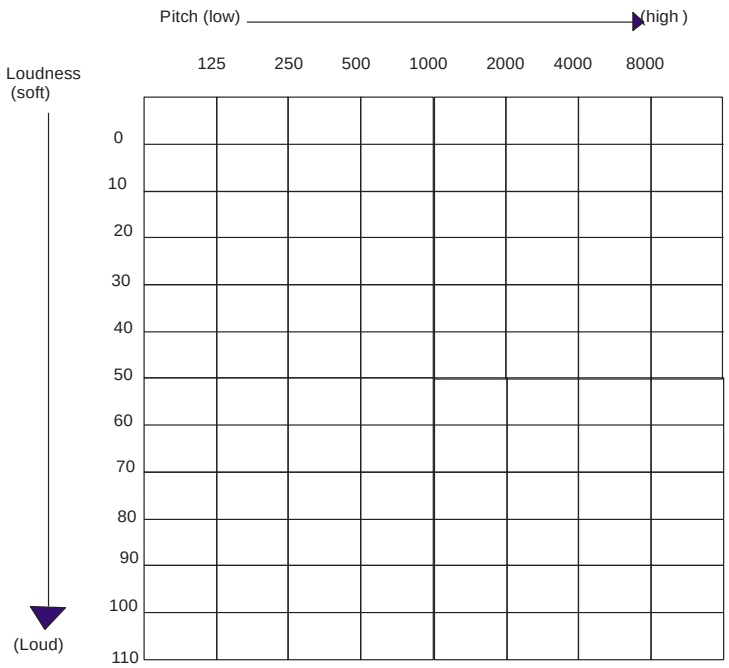


Chart Describing Hearing Loss

Less than 25 dB (decibels) for children	Normal
25 dB for children	Normal to borderline
30-40 dB	Mild hearing loss
45-60 dB	Moderate hearing loss
65-80 dB	Severe hearing loss
85 dB or higher	Profound hearing loss

The audiogram is useful in determining how much access to sounds a child have. It does not however give any information about how much speech is being understood by the child.

Other tests may be done in the sound-proof booth to see if your child is able to attach meaning to sounds. For example, the audiologist may present familiar words to an older child and ask him or her to identify the word by pointing to a picture.

We look forward to working together with you to help your child reach thier highest hearing potential.

Don't hesitate to call our department if you have questions.
Sincerely,
Audiology Department
The University of Memphis Pediatric Auditory Research Laboratory

IV. ASSISTIVE LISTENING DEVICES

Assistive Listening Devices for Your Child with auditory neuropathy spectrum disorder (ANSD)

Your child's audiologist will examine the audiological test results and discuss them with other team members. Together they will determine a recommendation regarding listening assistance for your child.

Hearing Aids

The team may recommend hearing aids for your child. However, ANSD is usually a problem of clarity of speech, not access to sounds. In other words, the hearing aids cannot really help with the speech clarity. While we know some children may benefit from amplification to learn language, we also know that this is the case for less than 10% of children.

If you and the team decide to try hearing aids, please keep in mind that it will not be necessary to try the hearing aids for very long before seeing if they provide benefit for language development. In other words, the use of loaner hearing aids might be more appropriate for a few weeks before investing in them. During this time, it is important to evaluate if your child progresses in understanding speech and not only if he/she reacts more to sounds. Having your child tested by an audiologist with the hearing aids on will be important. As important will be having regular speech and language evaluations.

Because hearing aids amplify all sounds including air conditioning fans and traffic noise, in cases of ANSD they should almost always be worn with a remote microphone (next section) so that the speech signal does not become lost.

Hearing aids allow sounds to become louder but do not improve the clarity. In more moderate or severe cases of ANSD it is suspected that sounds are quite unclear to the listener, so that making them louder may not help the child understand them. Again, hearing aids alone have not proven to help many children with ANSD to develop speech and language. In fact, too much amplification can damage the ears. That is why we recommend that the hearing aids not be set too loud and the trial period to be short. Hearing Aids may or may not be the best option for your child.

If your child is trying hearing aids, impressions of your child's outer ear and ear canal will be made. This is a quick and painless procedure. The audiologist will send this in to a laboratory where they will make ear molds that are specifically meant to fit your child's ear.

The audiologist will then order hearing aids or provide loaner hearing aids that are appropriate for your child's hearing loss. At your child's initial hearing aid fitting appointment, the audiologist will counsel you regarding the maintenance and use of the new hearing aids. There will be several hearing aid checks to address any questions or problems you may have and to chart your child's progress with the instruments.

Cochlear Implants

It may become clear that a combination of listening therapy with hearing aids will not be enough to help your child learn to hear and speak. At that time the team will discuss the option of cochlear implantation with you. A cochlear implant is a prosthetic hearing device that has internal and external components and is designed to replace non-functioning portions of the ear. The internal components are placed in the child's head during a surgical procedure lasting 2-3 hours. The child receives the external component about 3-4 weeks after the surgery. At that time the child's hearing is "born". The cochlear implant helps many people who do not benefit from hearing aids to hear and understand speech. Training and therapy are needed to be a successful implant user and not everyone is a candidate for the surgery. Cochlear implantation has been very successful for many children with ANSD, but again, expectations must remain reasonable, and results are sometimes not as good as predicted. We do recommend that you get in touch with other parents of children with ANSD who have had a child receive a cochlear implant.

The Remote Microphone

The team may recommend a remote microphone. The remote microphone system is similar to a personal radio system. The remote microphone system does two things:

1. It will reduce background noise
2. It will reduce the distance from the speaker's voice.

It can be used with hearing aids and cochlear implants and will facilitate speech understanding in noisy environments such as a daycare or a classroom.

Together with the ANSD team you will chart the course of your child's progress. With effective and timely intervention, we can work together to help your child develop speech and language skills.

The team at The University of Memphis Pediatric Auditory Research Laboratory is here to help you in the process of managing your child's disorder and to provide you with information and choices. However, it is up to you to make the decision that is right for you, your family, and your child. We are here to work together in the process of helping your child to maximize thier abilities and to communicate effectively.

V. AUDITORY NEUROPATHY SPECTRUM DISORDER: A CLOSER LOOK

Auditory Neuropathy Spectrum Disorder (ANSD)
This is diagnosed when the audiologist finds:

- Normal outer hair cell function.
- Abnormal neural function at the level of the VIIIth (vestibulo-cochlear) nerve and lower auditory brainstem.
- A problem with auditory timing.
- Hearing levels on the audiogram ranging from normal hearing to a profound hearing loss.

This condition was recently discovered and was not named until 1996. Doctors, researchers and audiologists are still in the process of learning about it. For example, the exact site in the auditory system where the timing break down occurs cannot always be identified. What we do know is that the ***outer hair cells*** usually work well, but there is a problem with either the ***inner hair cells***, the connection between the ***inner hair cells*** and the auditory nerve (the synapse), or the auditory nerve itself. It may be different for different people and may often be a combination of one or more of these things. Often times, the result is a disorder of timing, that is that the listener with ANSD may be able to hear soft sounds but cannot detect some of the very subtle differences in timing needed to discriminate many speech sounds.

There are some risk factors that seem to be associated with ANSD; they will prompt the doctor or audiologist to order specific tests to determine if ANSD exists in a patient. Some of these risk factors are prematurity, a history of neonatal illness such as hyperbilirubinemia (jaundice), or a history of ANSD within the family. However, there are many cases of ANSD with no known risk factors at all.

It is important to remember that every child with ANSD is unique. Some may have other disorders in association with ANSD and others may not. Some may have little to no sound awareness while others appear to have normal hearing with the sole complaint of difficulty hearing in noisy environments. It is possible to have ANSD in only one ear, but most commonly it affects both ears. ANSD can be caused by head injury or other trauma, but most commonly it is present at birth. Some infants, especially those who are born premature, may outgrow the

disorder by the first year of age. Because each case of ANSD is unique we will have to work together with you as a team in order to provide the best management possible for your child.

How Often Does It Occur?

Once thought to be very rare, it is now believed that ANSD is responsible for 10-15% of all permanent, inner-ear hearing loss.

Why Such a Long Name?

Auditory neuropathy spectrum disorder (ANSD) was first suspected in the late 1980s after **otoacoustic emissions** (OAEs) became widely used and it was discovered that some patients with hearing loss who were expected to have no **outer hair cell** function had normal OAEs. Over the next several years the disorder continued to be described in the scientific literature and was eventually named auditory neuropathy in 1996 by a neurology team composed of Arnie Starr, Terry Picton, Yvonne Sininger, Linda Hood, and Charles Berlin. As the professional community continued to learn more about this "new" hearing problem, it became evident that it was not a true neuropathy because in many cases the auditory nerve itself is just fine. Rather the problem seems to be more one of a dys-synchronous firing of the auditory nerve, hence the name auditory neuropathy spectrum disorder or ANSD.

Cochlear nerve deficiency which is a small or absent cochlear nerve is another way ANSD can be present. Cochlear nerve aplasia is when there is no hearing nerve present, while cochlear nerve hypoplasia is when there is a smaller than normal auditory nerve.

ANSD could be presynaptic or postsynaptic. Presynaptic ANSD would affect the inner hair cell or the space between where the signal leaves the IHC and gets picked up by the auditory nerve. Post synaptic ANSD would affect the auditory nerve right where it is picking up the signal or further down on the auditory nerve.

Genetic mutations can be a cause for ANSD. There are several known genetic mutations for this disorder. A genetic counselor will go over the results of genetic testing with you and discuss if there are any mutations that could cause ANSD.

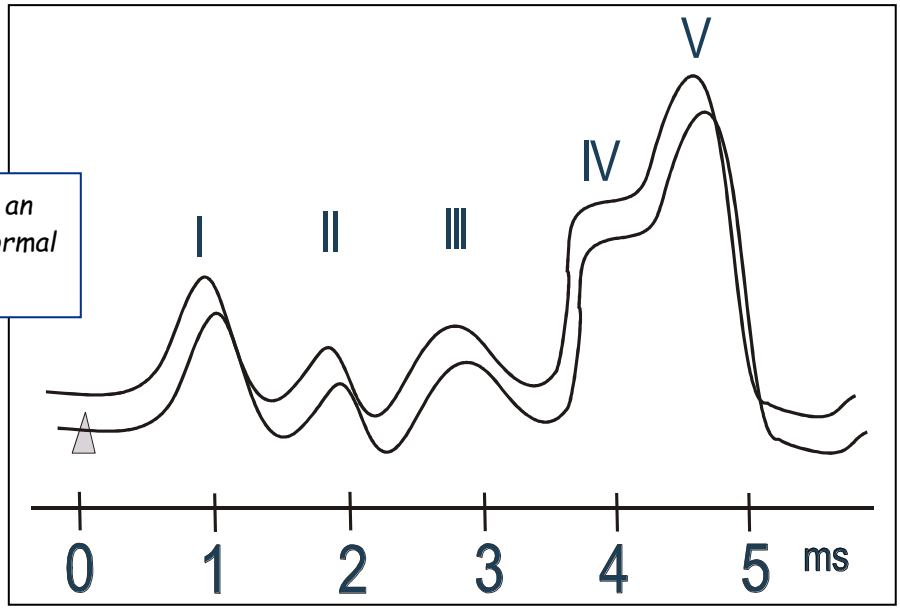
How Do We Diagnosis It?

Many of the tests described above are used in combination to arrive at the diagnosis of ANSD. The battery of tests that are the most important consist of ***otoacoustic emissions*** (OAEs), ***middle ear muscle reflexes*** (MEMR) and ***auditory brainstem responses*** (ABR). Since ANSD is

characterized by normal ***outer hair cell*** function and abnormal function in the region of the ***inner hair cells*** and/or auditory nerve, in order to differentiate ANSD from more common forms of cochlear hearing loss, the test battery must be sensitive to both cochlear and auditory nerve function. Most individuals with ***ANSD*** have intact ***OAEs*** because the outer hair cells function normally. The ***middle ear muscle reflexes*** (MEMR) are typically abnormal or absent because good neural synchrony is necessary for a normal MEMR.

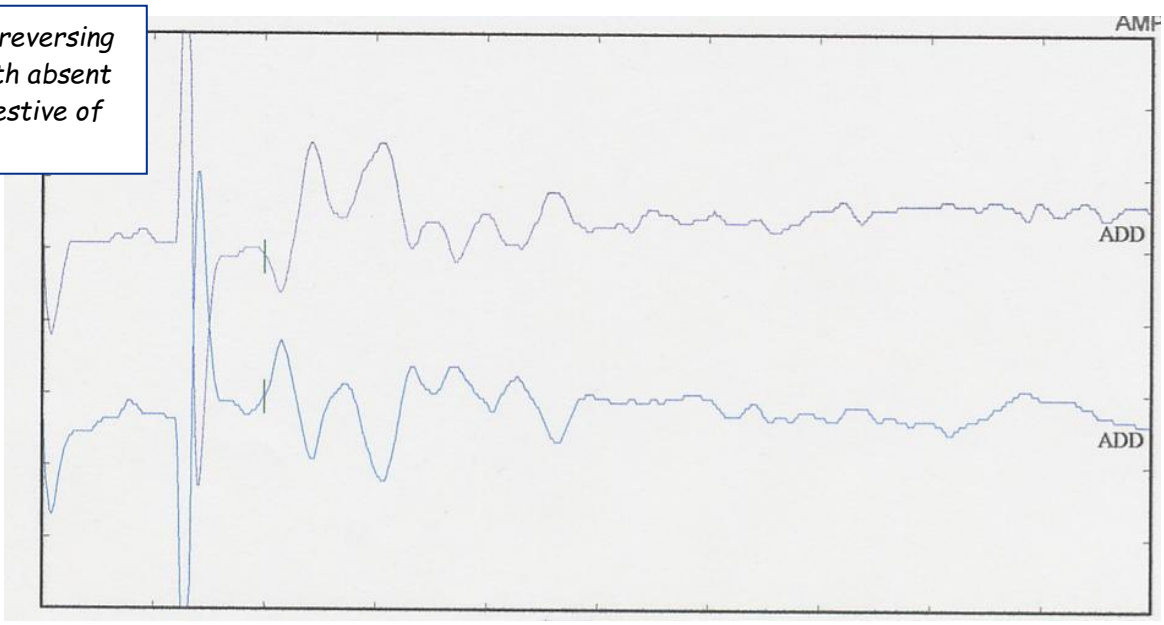
The ABR of individuals with ANSD is characteristic and unlike the ABR seen with other types of hearing problems. While most people with ANSD have normal OAEs, for reasons that are unknown, not all of them do. This is why it is very important that the ABR is done in a specific way that separates the cochlear from the neural response. The way to separate out these responses is to use two types of clicks as the stimulus: one positive polarity and one negative polarity click. The cochlear, outer hair cell part of the response is called the ***cochlear microphonic*** (CM). It occurs early in the recording and reverses depending on the type of click stimulus, whereas the neural response occurs later and does not reverse regardless of the stimulus type. This is the only way that an ABR test result can properly be used to differentiate ANSD from profound sensorineural deafness. As we've mentioned before, not everyone with ANSD is completely deaf. Many have quite a bit of sound awareness and it would be a mistake to manage them in the same way as a deaf child.

It is important that parents of children with ANSD understand a lot about their child's disorder, because unfortunately, being that it's a recently discovered condition, not all clinicians are fully aware of all the issues involved.



Expected ABR in an individual with normal hearing.

Cochlear Microphonic reversing in polarity changes with absent neural response suggestive of ANSD



AUDIOLOGICAL TESTING RESULTS

Expected test results in individuals with ANSD	
Test	**Results**
Pure tone thresholds	Variable – normal to profound hearing loss; any pattern; can be in only one ear
Speech reception in quiet	Variable – excellent to poor
Speech reception in noise	Generally poor
Middle ear muscle reflexes	Absent or elevated – both ipsilateral and contralateral
Otoacoustic Emissions	Usually present and robust
Cochlear Microphonic	Present – inverts with stimulus polarity reversal
Auditory brainstem response	Severely abnormal to absent
Masking level difference	Absent (0 dB)
Efferent suppression of OAEs	No suppression

Unlike with the more common form of permanent hearing loss that is due to ***outer hair cell*** loss, in cases of ANSD the ABR and ***audiogram*** do not provide us with information as to the severity of hearing loss. Initially, once the diagnosis has been made, we will learn the most about your child's sound awareness and ability to understand speech in different environments by listening to you. Other than your child, of course, you will be the most important team member in managing your child's hearing problem.

VI. COMMUNICATION OPTIONS

Facilitating Language and Cognitive Development

Because auditory input is distorted for children with ANSD, supplementing the auditory signal with visual information can dramatically aid an infant or child in learning language. If your goal for your child is to learn to hear and speak, we recommend a form of ***Visual communication*** that compliments your spoken language. American Sign Language, or ASL is the most common form of ***Visual communication,*** but it is a distinct language and therefore does not compliment English as well as some other forms of ***Visual communication.*** We recommend using ***Cued Speech*** or ***Signed Exact English*** which can both be used to represent exactly what is being said while you are saying each word. Speech reading permits the child to see the sounds that are produced on the lips. ***Cued Speech*** is a system that takes advantage of the 30% of speech sounds that are visible on the lips. The other 70% is conveyed with a system of hand movements for each vowel sound and hand shapes for each consonant. Cueing can be used to convey any language, not just English, and can typically be learning in a weekend course. Fluency, of course, takes a bit more practice. Signed Exact English has a sign for each word spoken and is considerably more complicated to learn. The idea behind using either system is that it will be phased out once your child no longer needs it. It may become evident at one point that your child has a mild case of ANSD and will not need a lot of intervention in order to develop normal speech and language. Or your child may receive a cochlear implant and be a successful user who does not need visual aids to understand what's being said. Regardless, infancy is such an important time for language learning that we do not want your child to fall behind during the initial period of watchful waiting.

Regardless of all these factors, it is important to remember that management is always individualized and what works for one family may not always be practical for another. Parents have the ultimate decision as to what is best for their child.

Further Questions?

Jennifer P. Taylor, AuD,CCC-A .....Audiologist.....901-678-5822
Hannah Beth Scott, AuD,CCC-A.....Audiologist.....901-678-5844
Jordan Alyse Coffelt, AuD,CCC-A .....Audiologist.....901-678-4619
Casandra Banks, AuD,CCC-A .....Audiologist.....901-678-5830
Matthew Hollis, AuD,CCC-A .....Audiologist.....901-678-4401

VII. Resources and Glossary of Terms

Resources:

American Academy of Audiology (AAA)
11730 Plaza America Drive, Suite 300
Reston, VA 20190
Tel/TTY: (800) AAA-2336
Email: info@audiology.org
Website: http://www.audiology.org

American Speech-language Hearing Association (ASHA)
10801 Rockville Pike
Rockville, Maryland 20852
Website: http://www.asha.org

Alexander Graham Bell Association for the Deaf and Hard of Hearing
3417 Volta Place, NW
Washington, DC 20007
Tel: 202/337-5220
TTY: 202/337-5221
Email: info@agbell.org
Website: www.agbell.org

Hearing Loss Association of America
7910 Woodmont Avenue, Suite 1200
Bethesda, MD 20814
Tel/TTY: (301) 657-2248
Website: http://www.shhh.org/

KidsHealth
http://kidshealth.org/

National Cued Speech Association
23970 Hermitage Road
Cleveland, OH 44122
Tel/TTY: 800-459-3529
Website: http://cuedspeech.org

Simulation of Auditory Neuropathy
http://faculty.sites.uci.edu/hesplab/simulations

Supporting Success for Children with Hearing Loss:
www.Successforkidswithhearingloss.com

We encourage you to listen to these simulations and to make copies to explain to your relatives and to your child's caregiver or teacher what ANSD sounds like.

GLOSSARY

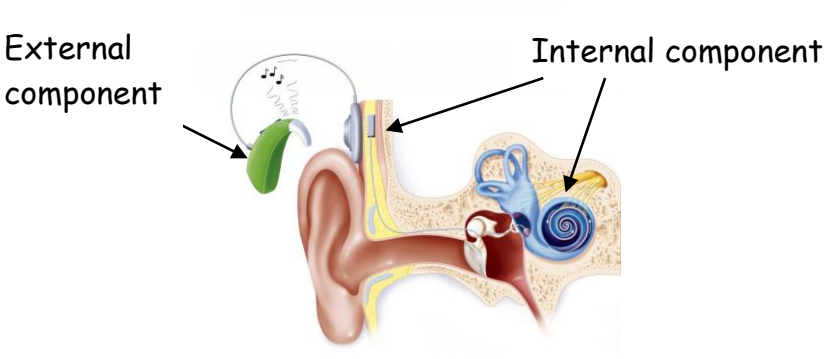
Audiogram – A graph with pitch from low to high across the top (in frequency) and loudness along the side (in decibels). The softest sound a person can detect (hearing threshold) for various pitches and for speech are recorded on this graph.

Auditory Brainstem Response (ABR) – a.k.a. Brainstem Auditory Evoked Response (BAER) and Auditory Evoked Response (AER). Test of neural synchrony throughout the auditory nerve and brainstem. It is used to evaluate the ability of neurons in the lower part of the auditory system to maintain precise timing and respond to external stimuli at the same time (synchronously). Electrical signals from the brain are picked up by small electrodes placed on the head.

Auditory Neuropathy Spectrum Disorder (ANSD) – A hearing disorder that is categorized by present otoacoustic emissions (normal cochlear outer hair cell function) and abnormal or absent auditory brainstem responses. With this type of hearing disorder the auditory nerve is unable to transmit sound clearly to the brain.

Cochlear Implant

A Cochlear Implant is a prosthetic device used to aid hearing. It has two components: the internal component and the external component.



The internal component is placed into the child's head in a surgical procedure.
The external component looks like a hearing aid.

Cochlear Microphonic (CM) – reflects the presence of an electrical field surrounding both inner and outer hair cells. In humans, its presence primarily suggests normal outer hair cell activity in the cochlea. The CM is known to follow the characteristics of the stimulus. That is, it will reverse with changes in the polarity of the click signal.

Conductive Hearing Loss – caused by pathology of the outer or middle ear which does not allow all the sound to reach the inner ear.

Efferent Suppression of OAE – test that involves the reduction in amplitude of otoacoustic emissions resulting from the activation of efferent neural pathways by a sound.

Hearing Aids – Amplification devices which amplify sounds with softer sounds receiving the most amplification. They were designed to manage hearing loss due to damaged outer hair cells.

Inner Hair Cells (IHC) – Sensory transducers of the auditory system. The IHCs are responsible for the activation of the auditory nerve.

Middle Ear Muscle Reflexes (MEMR) – a.k.a. acoustic reflexes, acoustic reflex thresholds, stapedial reflex. A brain stem mediated reflex in which a high intensity sound causes the stapedius muscle in the middle ear to contract. These are absent or abnormal in those with ANSD.

Outer Hair Cells (OHC) – Located in the cochlea, they amplify sounds reaching the ear before their transduction by the inner hair cells. They are sometimes referred to as the cochlear amplifier. OHC function is usually preserved in cases of ANSD.

Otoacoustic Emissions (OAE) – These are sounds produced by the contractile properties of the outer hair cells. Their recording allows for the assessment of the outer hair cell function in the cochlea. Absence of OAEs usually means that amplification is required. OAEs are usually normal in people with ANSD.

Pure-tone Audiometry – standard audiology testing that records a person's responses to various sounds. The results of the test are reported on a graph called the audiogram.

Remote Microphone – an amplification device. It consists of a microphone worn by the speaker and special hearing aids which pick up the signal from the microphone via FM waves. The voice of the person wearing the microphone sounds very close to the wearer's ear even when he or she is far away from the speaker. This device doesn't amplify as much as regular hearing aids but it is very useful when there is background noise. It increases the signal-to-noise ratio.

Sensorineural Hearing Loss – This type of hearing loss arises from the cochlear, either the outer hair cells, inner hair cells or a combination of the two. A sensorineural hearing loss will yield a synchronous neural response upon ABR testing at high intensities whereas the neural response in ANSD is absent or grossly abnormal due to a lack of neural synchrony.

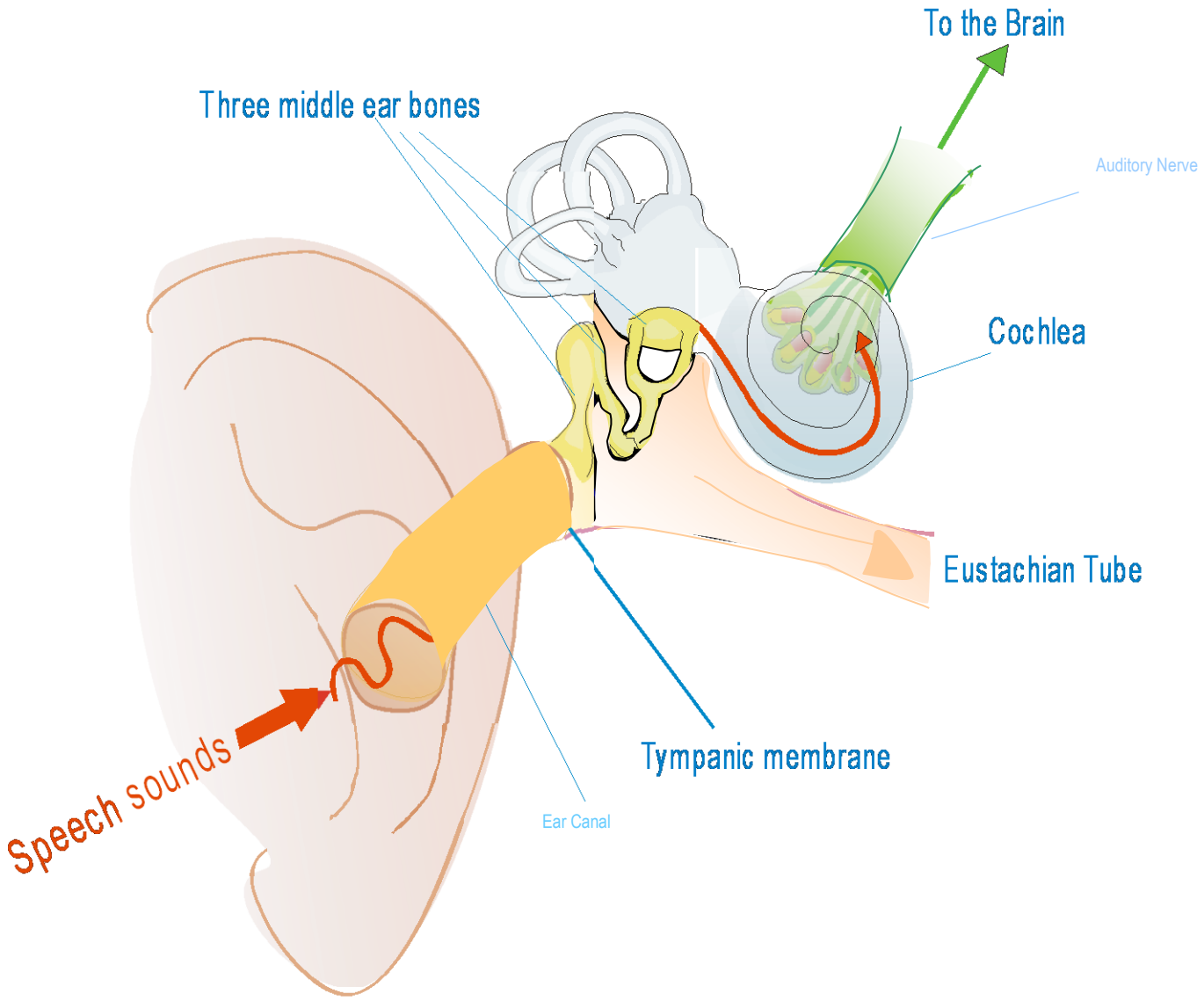
Speech Audiometry – standard audiology testing to find the behavioral threshold of speech stimuli and understanding of single words and/or sentences in quiet and in noise.

Tympanometry – testing used to measure the ability of the middle ear to conduct sound to the inner ear. It can determine if there is fluid behind the eardrum or other problems in the middle ear.

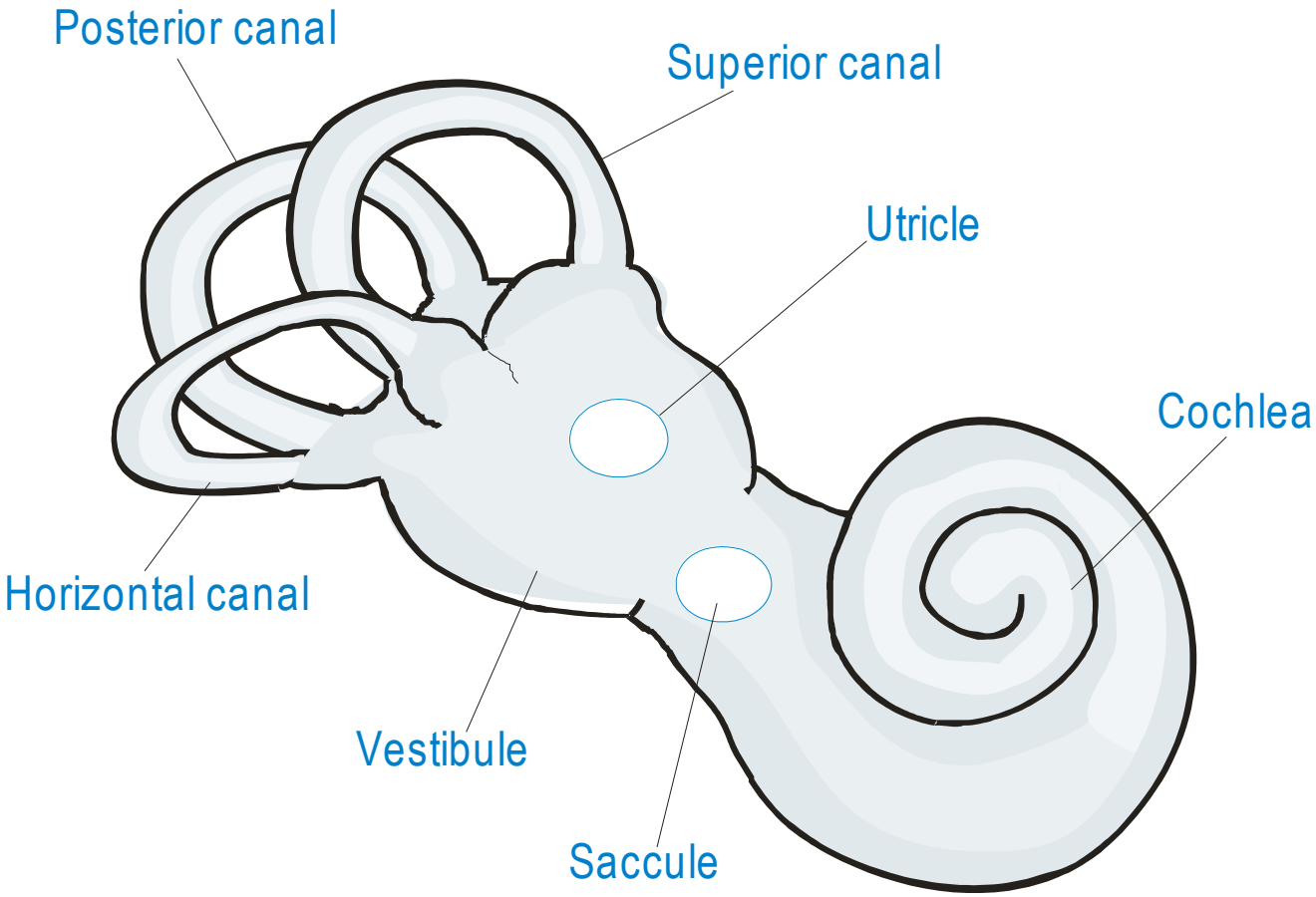
Visual Reinforcement Audiometry (VRA) – A method of behavioral testing used for infants and toddlers which uses puppets that light up to visually reinforce the child to turn thier head in response to various sounds presented in a sound booth. The results are recorded on an audiogram.

Auditory neuropathy spectrum disorder

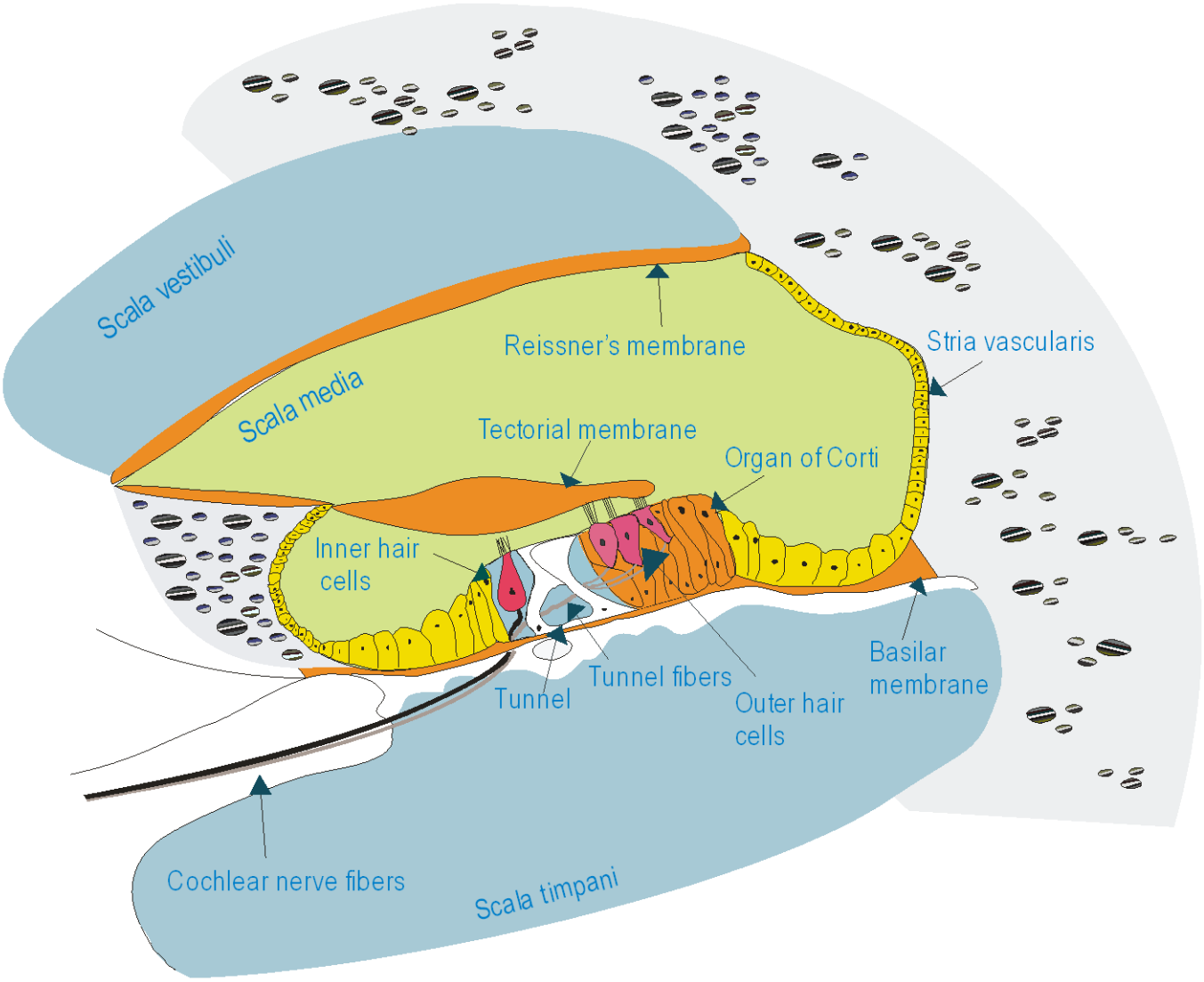
Graphics of the Ear



PERIPHERAL AUDITORY SYSTEM



The Cochlear and Vestibular Systems



Cross section of the cochlea

Panel of Tests For Patients with Auditory Neuropathy Spectrum Disorder

Purpose / Prescription	Test
The patient requires a complete ABR to rule out auditory neuropathy spectrum disorder.	Auditory Brainstem Response (ABR)
The patient requires an evoked otoacoustic emissions test in order to rule out auditory neuropathy spectrum disorder.	Otoacoustic Emissions (OAE)
The patient requires a test of the Auditory Reflex to determine threshold of response.	Middle Ear Muscle Reflex (MEMR)
The patient requires an unaided ear-specific audiogram.	Ear-specific measurement of hearing loss.
The patient requires a complete audiological test battery and a hearing aid fitting.	Audiological Battery: Comprehensive audiogram
The patient requires a CT Scan of the temporal bones with no contrast, both axial and coronal views.	CT Scan
The patient has a profound hearing loss with a possible diagnosis of auditory neuropathy, and requires an MRI of Internal Auditory Canals to determine whether or not there is a viable cochlear nerve.	MRI
The patient has a hearing loss with a possible diagnosis of auditory neuropathy spectrum disorder and requires a speech and language evaluation and follow-up treatment.	Speech / Language Evaluation

The patient has a profound sensorineural hearing loss with a possible diagnosis of auditory neuropathy spectrum disorder, and requires a genetic work-up to rule out genetic causes.	Genetics Study - Consult
The patient has a profound sensorineural hearing loss with a possible diagnosis of auditory neuropathy spectrum disorder, and requires a vision screen to rule out optical neuropathy and/or Usher's syndrome.	Opthalmology – Vision Screen Consult
The patient has a profound sensorineural hearing loss with possible diagnosis of auditory neuropathy. A neurological screening is necessary to determine if other neuropathies are present.	Neurology Consult

What Does the Student with Auditory Neuropathy Spectrum Disorder (ANSD) Need for Academic Success?

- A Quiet Environment

  Students with ANSD struggle to isolate and focus on the signal of the teacher's voice. A room that is noisy makes this much more difficult for them. This student will benefit from a room that is as quiet as possible. This can be facilitated by using carpet in the room, having the walls covered with sound-absorbing materials such as paper, using some shades over the windows, and putting tennis balls on noisy chair and desk legs.

- An Assistive Listening Device

  Students with ANSD often benefit from the use of a remote microphone device. This device acts like a personal radio that transmits directly from the teacher's voice to the student's ear. The student will still be able to hear thier own voice while using a remote microphone. Students may also use hearing aids with the remote microphone, or they may have a cochlear implant.

- Preferential Seating

  Students with ANSD need to sit away from noise producers such as heating systems or water fountains. Additionally, they need to sit in the front and they need to be able to see the teacher at all times.

- Support for language and vocabulary acquisition

  Students with ANSD often have a language delay secondary to the hearing problem. This does not mean that they are cognitively affected, but that their language is behind that of their normally hearing peers. They do not easily acquire vocabulary through casual listening as typical children do. These students need individual teaching sessions from a certified specialist to enable them to maintain a level of vocabulary acquisition that will allow them to participate fully in the classroom setting. This pre-teaching will allow the student with ANSD to tune into targeted vocabulary words, and benefit from teacher instruction in the classroom.

- Individual Speech and language therapy

  The student with ANSD will present with speech and language needs that are quite different from a typical child's needs. This student will need help developing speech through the auditory channel and will need to address articulation errors through refined listening activities.

- A Knowledgeable Team

  The student with ANSD needs an educational team that is aware of the special needs that accompany this diagnosis. Please read the packet of information and feel free to contact the ANSD Team at The University of Memphis Pediatric Auditory Research Laboratory with any further questions.

Your Child's Developing Speech

A child's speech begins with early vocalizations including playing with their voice, learning to start and stop their voice, make variations in pitch and intensity, and vowel cooing. This stage is followed by babbling, which is a playful, vocal sound. Babbling begins when children begin to start and stop their voice with emerging consonants. What we term as jargon, strings of sounds or syllables produced with a variety of stress and intonation, begins when children begin to string multiple vowel and consonant combinations together in a variety of pitch and intonation patterns. This jargon sounds very much like spoken language and the listener can often tell what the child is attempting to communicate. Eventually, a child will start to string the sounds together to form "true" words. Usually these first words will be embedded in a string of jargon, and as the child becomes adept at using spoken language, more and more true words will emerge.

Individual speech sounds are developed gradually. Many studies have reported the ages at which children typically produce each speech sound in the first or final position in words. A child may produce a sound correctly 50% of the time for months and sometimes a year before the sound is consistently correct. The following sequence of sound development is considered normal for a child:

STAGE ONE Vowel sounds such as ooooo, eeeee, and ahhhh.

STAGE TWO Suprasegmental voicing sounds such as rising inflection, pitch differences, starting and stopping the voice, playing with breath control.

STAGE THREE Bi-labial consonants, those that use two lips, begin to emerge such as mmmm, b, p, w paired with emerging vowel sounds, such as buh-buh-buh.

STAGE FOUR Sounds that involve the tongue and the roof of the mouth such as d, t, k,g, n, s,z begin to be paired with vowels, such as guh-guh-guh, da-da, ma-ma. As these sounds are reinforced, the child associates meaning with the sound, and real words begin to emerge such as momma, dadda, bye-bye.

STAGE FIVE We begin to hear f, v, h, y, l paired with vowels such as fah-fah, yuh. The child is producing strings of words along with jargon.

STAGE SIX Most consonants are heard and produced appropriately in words, including ch, sh, and j.

Are you concerned about your child's developing speech?

You can help your child speak more clearly if you do the following:

- Use short sentences and model correct speech for your child.
- Pronounce words slowly and correctly for the child to hear and imitate.
- Look at your child when you both are talking.

- Repeat new words and sounds over and over and use them often in your conversation with the child.
- Make a scrapbook with your child; cut out and paste pictures of objects the child learns to say or recognize.
- Praise your child when sounds are correct, especially if the sounds were previously difficult for the child.

You should have your child's hearing checked if:

- Your child does not seem to be making appropriate progress in the language levels described above.
- Strangers have difficulty understanding your child's speech.
- Your child shows frustration with thier speech.

Questions? You are encouraged to contact any of our audiologist at

The Memphis Speech and Hearing Center, or Dr. Thierry Morlet,

Director of the Pediatric Auditory Research LAB,

tmorlet@memphis.edu

We are happy to answer your questions!

Frequently Asked Questions

About Auditory Neuropathy Spectrum Disorder (ANSD)

Q. If my child has a hearing problem, why doesn't my audiologist recommend hearing aids?

A. Hearing aids only help in cases of damage to the outer hair cells in the inner ear (cochlea). In ANSD the outer hair cells are often working fine. The problem is with the inner hair cells. It may be the synapse (connection) between the inner hair cell and the auditory nerve, or it may be the nerve itself.

Q. Why does my child appear to hear me at times and not at others?

A. Children with ANSD are sometimes able to hear in quiet environments. It is more difficult for them when there is conflicting noise. If you call your child's name in a quiet room, he or she may look at you. In a noisier environment, your child may appear not to hear you at all.

Q. Are there degrees of ANSD?

A. Yes, a child can have mild, moderate, severe, or profound ANSD.

Q. Does the testing tell us how serious the ANSD is?

A. Unfortunately, no, not until a child is old enough to complete audiometric speech testing. The ABR can tell us whether or not a child has ANSD, but it does not tell us the degree, or the site of the damage. The damage may be in the inner hair cells, it may be the synapse between the inner hair cell and the auditory nerve, or it may be the nerve itself. The loss can be mild, moderate, severe or profound, but is always worse in noisy environments.

Q. With ANSD what does my child hear?

A. This depends on the severity of the problem. Some children, in mild cases, hear fairly well, and develop speech and language at near-normal levels. Other children cannot distinguish a person's voice from other sounds in the environment.

Q. What type of assistive listening device is recommended for children with ANSD?

A. In mild cases the child may do fine without an assistive listening device. With more severe problems a remote microphone (see section on assistive listening devices) may help to minimize background noise. In very severe cases a cochlear implant may be recommended. This type of hearing assistance will bypass the damaged part of the ear.

ANSD TEAM

AT

The University of Memphis Pediatric Auditory Research Laboratory and
The Memphis Speech and Hearing Center

TEAM MEMBER	POSITION	EMAIL
Thierry Morlet, PhD	PARL Director	tmorlet@memphis.edu
Jennifer P. Taylor, AuD,CCC-A	Audiologist	jptaylr2@memphis.edu
Hannah Beth Scott, AuD,CCC-A	Audiologist	hannah.beth.scott@memphis.edu
Jordan Alyse Coffelt, AuD,CCC-A	Audiologist	j.coffelt@memphis.edu
Casandra Banks, AuD,CCC-A	Audiologist	cbbanks@memphis.edu
Matthew Hollis, AuD,CCC-A	Audiologist	mhollis@memphis.edu