
Understanding Hearing Loss



HEARING SCREENING AND EVALUATION

We cannot overemphasize the importance of early detection when it comes to determining if a child is deaf/HH. As the United States Department of Health and Human Services (HSS, 1990) report states,

“If hearing impaired children are not identified early, it is difficult, if not impossible, for many of them to acquire the fundamental language, social, and cognitive skills that provide the foundation for later schooling and success in society.”

Early detection and intervention, along with early amplification and/or use of sign language, helps to prevent significant delays in communication development. Thus, it is critical to detect hearing loss in children as soon as possible, preferably before the child is one month of age. If a child is born prematurely, this would be calculated by the child’s corrected age. The Joint Commission on Infant Hearing recommends a 1-3-6 plan: newborn screening by 1 month of age, identification before 3 months of age, and early intervention before a child reaches 6 months in age. This plan is highlighted in the following diagram, ‘A Family’s Checklist- Infant Hearing’ which will be discussed further in this resource guide.

Please note: Although it is recommended that the child receive intervention services before 6 months of age (as indicated in this checklist), it is highly suggested that these services are sought simultaneously with the child's fitting for an amplification device, so that the early intervention specialist can provide instruction and support for using this device in the home setting.

A Family's Checklist – Infant Hearing

Child's Name: _____

Child's Date of Birth: ____/____/____

	Before 1 Month	Before 3 Months	Before 6 Months
<p>Normal Milestones</p> <p>Many babies meet normal milestones even if they have a hearing loss in one or both ears. You should only use these milestones to monitor your baby's hearing and development if your baby has PASSED the hearing screening or been evaluated by a Pediatric Audiologist...</p> <p>Months of Age</p> <p>2 Quiets when hearing a familiar voice. Makes vowel sounds like ahh, ohh</p> <p>4 Looks for sounds with his eyes. Uses sounds such as squeals, whimpers, chuckles</p> <p>6 Turns head toward sound. Babbles ba-ba, ma-ma, da-da</p> <p>9 Imitates speech sounds of others. Understands no-no or bye-bye. Turns head toward soft sounds.</p> <p>12 Correctly uses ma-ma or da-da.</p>	<p>Hospital Inpatient Screen</p> <p>Place: _____ Screen Date: ____/____/____</p> <p>Results: <u>Left Ear</u> <input type="checkbox"/> <u>Right Ear</u> <input type="checkbox"/></p> <p style="text-align: center;"> <input type="checkbox"/> Pass <input type="checkbox"/> Refer </p> <p style="text-align: center;"> <input type="checkbox"/> Repeat Screen Screen Date: ____/____/____ </p> <p>Results: <u>Left Ear</u> <input type="checkbox"/> <u>Right Ear</u> <input type="checkbox"/></p> <p style="text-align: center;"> <input type="checkbox"/> Pass <input type="checkbox"/> Refer </p> <p style="text-align: center;"> If your baby does not pass the screening in one or both ears, talk to your doctor about seeing a Pediatric Audiologist as soon as possible. </p> <p>If your baby passes, testing is done. Watch for normal milestones. Be sure your doctor gets the results. www.AZnewborn.com</p>	<p>Evaluation by Pediatric Audiologist. Be sure your doctor gets the results.</p> <p>Place: _____ Date: ____/____/____</p> <p>Results: <u>Right Ear</u> <input type="checkbox"/> <u>Left Ear</u> <input type="checkbox"/></p> <p style="text-align: center;"> <input type="checkbox"/> Normal <input type="checkbox"/> Hearing Loss </p> <p>If your baby has a HEARING LOSS, the next steps are:</p> <p>Evaluation by an ENT (Ear, Nose, and Throat) doctor</p> <p>Place: _____ Date: ____/____/____</p> <p>Hearing aid fitting (if appropriate) of loaner or permanent hearing aids by a Pediatric Audiologist.</p> <p>Contact Hands & Voices for family support: by phone at 866-685-1050 or www.AZHV.org</p>	<p>Enroll in Early Intervention program, if hearing loss in both ears</p> <p>Program: _____ Date: ____/____/____</p> <p>Learn about communication Options</p> <p>Learn about cochlear implants, if applicable</p> <p>Regular visits to a Pediatric Audiologist</p> <p>Evaluations:</p> <p><input type="checkbox"/> Ophthalmologist (eye doctor)</p> <p>Place: _____ Date: ____/____/____</p> <p><input type="checkbox"/> Genetic Specialist</p> <p>Place: _____ Date: ____/____/____</p> <p style="text-align: center;">You may need a referral from your doctor to see these specialists.</p>

TAKE THIS TO YOUR BABY'S DOCTOR

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Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

Every state and territory in the United States has established an Early Hearing Detection and Intervention (EHDI) program to ensure that all babies are screened for hearing loss before they leave the hospital or birthing center. Appropriate follow-up is given for babies who do not pass the screening, and intervention services are provided before the child reaches 6 months in age. The National Center for Hearing Assessment and Management serves as the national resource center for the implementation and improvement of comprehensive and effective EHDI services. Their website provides links to information about the EHDI programs in every state and territory, and also contains a wealth of information and resources related to early detection and intervention.

National Center for Hearing Assessment and Management

www.infanthearing.org


435-797-3584

The initial screen conducted at the hospital is called the “Universal Newborn Hearing Screening” (see following brochure). The procedures used in this screening are typically Otoacoustic Emissions (OAEs) and/or Automated Auditory Brainstem Response (AABR), both of which will be described later in this section of the guidebook. Both of these screens can be performed quickly, typically within 10-15 minutes if the baby is quiet or asleep. The Arizona Department of Health Services Office of Newborn Screening provides information on the newborn screening program, including specific information about the hearing portion of the screening.

Arizona Department of Health Services Office of Newborn Screening

www.aznewborn.com

602-364-1409



CAN MY BABY HEAR?

Your Baby's First Hearing Test Arizona EHDI

Many babies with hearing loss will react to loud sounds. The only way to know if your baby's hearing is normal is to have the screening done with special equipment.

Before you leave the hospital, be sure you know the results of your baby's hearing screening and what the next steps are.

Talk to your baby's doctor if you have any concerns or questions.

Why is hearing screening important?

- Hearing loss is one of the most common newborn conditions.
- Kids with hearing loss have no signs or symptoms.
- The first three years are the most important for learning language and how to communicate.

You need to know if your newborn baby has a hearing loss so you can get the help your baby needs. If hearing is found early, your child will have the best chance to learn the skills needed to be ready for school.

How is hearing screening done?

- Your baby will be screened for hearing loss in the hospital soon after birth.
- The screening takes 10-15 minutes if the baby is quiet or asleep.
- The screening is done with special equipment and is safe and does not hurt.

If your baby PASSES the screening...

It means that your baby's hearing was in the normal range during the screening. Hearing loss can develop after you leave the hospital. It is important to check your baby's hearing and communication development during the first year.

If your baby DOES NOT PASS the screening...

- By one month of age:** Make an appointment to come back to the hospital for an outpatient hearing screen within one to two weeks after birth. If your baby was in the neonatal intensive care unit (NICU) for more than 5 days or has a risk factor, talk to your baby's doctor. Make the outpatient appointment with a pediatric audiologist, as soon as possible.
- By three months of age:** Make sure that your baby has been tested by a pediatric audiologist by three months of age.
- By six months of age:** If a hearing loss is confirmed, your baby should be enrolled in an early intervention program by six months of age.

Is my baby at risk for hearing loss?

Yes, if your baby has one or more of the following risk factors or conditions at birth:

- Is older than 5 days
- Had an infection before or after birth such as CMV, herpes, rubella or meningitis.
- Has a family member with hearing loss from birth or childhood

These babies at risk can pass a hearing screening at birth but will need more testing later. Talk with your baby's doctor and make an appointment with a pediatric audiologist or hearing specialist for further testing.

Use this milestones chart to help you check your baby's hearing, speech and language development.

- By 2 months of age** a baby with normal hearing should be able to:
 - Quiet when hearing a familiar voice
 - Make sounds like ah and oh
- By 4 months of age** a baby with normal hearing should be able to:
 - Look for sounds with his eyes
 - Make sounds like squeals, whimpers or chuckles
- By 6 months of age** a baby with normal hearing should be able to:
 - Turn his head toward a sound
 - Make sounds like ba-ba, ma-ma, da-da
- By 9 months of age** a baby with normal hearing should be able to:
 - Imitate speech sounds made by others
 - Understand no-no or bye-bye
 - Turn his head toward a soft sound
- By 12 months of age** a baby with normal hearing should be able to:
 - Correctly use ma-ma or da-da
 - Respond to singing or music

A baby with mild hearing loss may also be able to do these things.

Arizona Department of Health Services

For more information about the Newborn Hearing Screening Program, please contact:
 Arizona Department of Health Services
 Office of Newborn Screening
 Arizona Early Hearing Detection and Intervention Program (EHDI)

250 N 17th Ave, 1st floor
 Phoenix, AZ 85007-3231
 Phone: (602) 364-1009
 or
 (800) 548-8382 (within Arizona)

Visit our website at:
www.AZnewborn.com

Deaf and Hard of Hearing call 711 for AZ Relay Service

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If your baby passes the newborn screening performed at the hospital or birthing center, it means that the sensory cells of the ear were found to be working properly during that screen. No further follow-up is typically needed unless there are known risk factors for hearing loss, or if there are further concerns. There is no single screening procedure that rules out all types and degrees of hearing loss. If you have concerns about your child's hearing, discuss this with your child's primary care doctor so that a plan of care can be implemented.

Some children pass the newborn hearing screening and develop a hearing loss after they leave the hospital. In this case, it is important for parents to be aware of potential signs that the child might have a hearing loss, and also be aware of any factors or conditions that may contribute to hearing loss. Please refer to the two previous documents for a description of the typical milestones in child development and a list of some of the conditions that might contribute to hearing loss developing later in a child's life.

If you are told that your baby did not pass the initial screen or is referred for further testing after the hearing screening, it may not necessarily mean that your child has a hearing loss. Rather, this initial screen is an indicator that the child needs to have his or her hearing checked again before one month of age. Babies may be referred for further testing after the newborn hearing screening because there is fluid in their ears or because the child was not asleep or quiet during the testing. A child may also be referred for further testing due to equipment problems, technical errors, or due to a true hearing loss. Based on statistics from the Arizona Department of Health for the year 2006, four percent of all babies who were screened in a hospital were referred for a second screen. Of these 3,922 babies, 35 babies were diagnosed with a confirmed hearing loss. Thus, the only way to confirm that the initial screen was accurate is to re-screen the baby within the recommended time frame.

The Arizona Department of Health Services Office of Newborn Screening is notified by the hospital when a child is referred for further testing after the newborn hearing screening. This office follows up with families and provides education and information to parents. However, you do not need to wait to hear from this office to get your child's hearing rescreened. This follow up, which is an outpatient procedure, is performed at many locations throughout Arizona. Simply contact one of the locations listed on the following link to make an appointment. You may want to contact your insurance plan member services representative to determine which

locations/providers are covered by your plan and if a referral from your primary care provider is needed.

Local Resource for Parents:

Available at: <http://azdhs.gov/lab/aznewborn/parents/hearing-screening.htm>

There are several hearing tests that an audiologist can perform to detect if sound is being transmitted efficiently through the auditory system and true hearing levels. The most commonly used tests are briefly described below.

- Otoacoustic Emission Test (OAE)- A small probe is inserted into the ear canal of a calm or sleeping child and presents soft sounds to measure echoes from the peripheral portion of the ear, as a means of determining if the cochlea is stimulated with audible sound. This test does not measure what the child hears, but simply tells whether the outer hair cells in the cochlea are functioning. If the cochlea does not respond to sound, there might be a hearing loss and further testing is needed.
- Auditory Brainstem Response (ABR)/Brainstem Auditory Evoked Response (BAER)- Sensors (small electrodes) are placed on the child's head and clicks or tones are presented through ear phones and/or a bone conduction headband to record brain activity in response to sound. This test can be performed while a small baby is asleep. It is often recommended that a baby older than four months be sedated because the test requires the child to remain still to rule out any other muscle reaction that could be interpreted as hearing. The ABR is considered to be highly accurate in detecting hearing loss when combined with other audiological test results.
- Auditory Steady-State Response- Like the previous test, electrodes are placed on the head during a state of sleeping or sedation, yet a continuous tone is emitted to elicit a response. This test is beneficial in determining the degree of hearing loss in the severe-to-profound range; however, it is not widely used clinically. However, it may be used in conjunction with an ABR to further confirm a diagnosis.
- Immittance testing- Immittance testing is comprised of tympanometry and acoustic reflex testing. Tympanometry is a measure of eardrum movement as a function of pressure and is used to determine if the middle ear is functioning properly. Abnormal tympanograms are indicative of possible middle ear pathology such as fluid, infection, or problems with the bones

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

of the middle ear (ossicles). Acoustic reflex testing is a measure of the stapedial reflex in the middle ear which contracts the stapedius muscle and ossicles in response to intensely loud sound. Acoustic reflexes may be absent in persons with conductive hearing loss, significant sensorineural hearing loss, acoustic nerve tumors, or facial nerve injuries. A patient must remain very still and quiet for accurate testing.

- Behavioral Observational Audiometry (BOA)- An audiologist watches an infant's face and body language to determine if he/she is responding to sound (such as changing sucking pattern, widening eyes, or searching for the source of the sound). This test is primarily used with infants and children with developmental delays.
- Visual Reinforcement Audiometry (VRA)- A sound is emitted through a loud speaker and the audiologist observes the child's behavior. If the child turns toward the speaker, a toy lights up or moves. This test is commonly conducted with children between 6 months and two years of age.
- Conditioned Play Audiometry (CPA)- Frequency (tone)- specific signals are delivered to an older child's earphone and the child is asked to respond with some action whenever a sound is heard (i.e. throwing blocks in the bucket or adding a piece to a puzzle). A speech test often accompanies this test. Words are spoken into headphones and the child is asked to repeat the words or point to pictures of the objects mentioned. Audiologists use these tests to determine the softest level at which the child can detect sounds and recognize speech.
- Bone conduction response- The audiologist places a bone conduction oscillator on the child's mastoid bone to bypass the outer ear and middle ear and stimulate the cochlea directly.
- Masking- If there is a suspicion that one cochlea hears better than the other, or is doing the work for them both, a noise from a headphone occludes one ear while the other ear is tested.

If your baby passes the second screening test, you still need to be aware of indicators that your child might have a hearing loss. Many babies with undetected hearing loss have a delay in language development. If you are concerned about your child's hearing and/or language development, you should contact your child's pediatrician and discuss your concerns. However, if you are not satisfied with the pediatrician's response, you can take your child to an Ear, Nose and Throat doctor (ENT or otolaryngologist), who will most likely have your child's hearing

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

evaluated by an audiologist specializing in pediatric care. Your insurance plan member representative can help you find an ENT if a referral from your primary care physician is needed.

If your baby does not pass the second screening, you should talk to your child's pediatrician about requesting a referral to an audiologist who specializes in pediatric care as soon as possible. An audiologist is a health care professional with a master's degree (M.A.) or doctorate (Au.D. or Ph.D.) from an accredited university program. Audiologists are trained to know the functions of the ear, how to determine if a person has a hearing loss, and what to do when a hearing loss is diagnosed.

When choosing an audiologist, or talking to your child's pediatrician about the audiologist he or she recommends, you will want to ensure that the audiologist you choose works primarily with children. The American Academy of Audiology is the world's largest organization of, by, and for audiologists with over 10,000 members. Their mission is to promote quality hearing and balance care by advancing the profession of audiology.

American Academy of Audiology

www.audiology.org

800-222-2336

If a hearing loss is confirmed by the child's audiologist, an evaluation by an ENT is the needed. The sooner the child visits the ENT, the sooner services can be administered. In some cases, an ENT and an audiologist work together at the same location, as many ENTs have on-site audiologists. For children who are diagnosed at an older age, pediatricians will often refer the child directly to an ENT.

An ENT specializes in ear, nose, throat, and head and neck disorders. As with audiologists, it is recommended that you find an ENT who works with children. The American Academy of Otolaryngology- Head and Neck Surgery, is the world's largest organization representing these specialists. Although their website is primarily for these doctors, there is a section for parents that gives health information on hearing-related topics.

American Academy of Otolaryngology - Head and Neck Surgery

www.ent.org

703-836-4444

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

A visit to the ENT is necessary to find out if there is a biological or medical reason for the hearing loss and if there is a medical treatment. It is often difficult to determine if a child has fluid in his or her ears that may be preventing the child from hearing within the normal ranges. If the child is over 12 months of age, an ENT might recommend a quick out-patient procedure to place tubes in the child's ear drums to drain any fluid that can interfere with hearing. Often this process may be frustrating for parents who are anxious to know if their child's hearing loss is temporary or permanent. Other setbacks like ear infections or sickness might also prevent a timely and accurate diagnosis of the child's hearing. This is another reason why it is essential to have a complete diagnostic hearing evaluation by the time the child is 3 months old in cases when the outpatient hearing rescreen is failed. A link to the list of Arizona providers for completing this diagnostic evaluation follows.

Early Hearing Detection & Intervention- Pediatric Audiology Links to Services (EHDI-PALS)

www.ehdipals.org

Children with a diagnosed hearing loss should have their hearing tested frequently, typically every 3-4 months for babies up to 2 years old and every six months for 3-5 year olds. Once children reach the age of 6, they need to have their hearing tested annually. After diagnosis, you will most likely be contacted by an Early Intervention specialist who will be assigned to work with your child from birth to three years of age. The goal of the early intervention program is to provide support and instruction to families within the natural learning environments that facilitate the child's successful engagement in relationships, activities, routines, and events of everyday life. This occurs in the context of the family's typical routines and activities so that information is meaningful and directly relevant to supporting the child in meeting the expectations of his or her environment. Early intervention is discussed in greater detail in the third section of this guide.

Parents may find the time of waiting and wondering during the screenings and evaluations to be very unsettling. Many parents who have children with hearing loss have typical hearing themselves, so it can be difficult to understand the unique experience of raising a child who is deaf or hard of hearing. Many parents say that it is helpful to connect with other parents who have already been through a similar experience. Arizona Hands & Voices (AZHV) is the state

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

chapter for the national Hands & Voices organization, which is a non-profit and parent-driven organization dedicated to supporting families and their children who are deaf or hard of hearing. AZHV also participates in the Guide By Your Side (GBYS) program, which provides emotional support and unbiased information to families by pairing them with trained Parent Guides who are or have been in similar situations, all free of charge.

Arizona Hands & Voices

www.azhv.org

866-685-1050

The American Society for Deaf Children (ASDC) was founded as a parent-helping-parents network and is now a national independent non-profit organization whose purpose is providing support, encouragement and information to families of children who are deaf or hard of hearing. ASDC provides resources, publications, and national family conferences.

The American Society for Deaf Children

www.deafchildren.org

800-842-2732

An online resource that can be helpful is My Baby's Hearing, developed by a team of professionals at Boys Town National Research Hospital. This website is divided into two sections: 'First steps' for newborn screening information, and 'Next steps' for when the child has been diagnosed with a hearing loss. The 'Next steps' portion has a 'parent to parent' section in which you can hear the views of parents with children who are deaf or hard of hearing.

My Baby's Hearing

www.babyhearing.org

402-498-6511



A Message from the Office of Newborn Screening

[Hearing Loss: Online Directory for Parents and Providers](#)



What is EHDI PALS (Early Hearing Detection & Intervention Pediatric Audiology Links to Services)?

An easy-to-use online directory matching hearing services to children's needs. For hearing tests and other hearing related services, go to: <http://www.ehdipals.org>

What information does EHDI PALS provide?

This site has information about hearing (audiology) services for children of all ages. The services and staff listed have the right equipment and skills to serve children.

How can I access it?

EHDI PALS is easy to use. Click "[Find a Facility](#)" to answer a few questions that take you to the services and staff closest to you.

What are some of the resources the website provides?

Click on "[Parent Resources](#)" to find questions to ask about your child's appointment. It also tells you about hearing programs in Arizona.

Click on "[Other Helpful Websites](#)" for national and state parent support groups and other resources related to childhood hearing loss and testing.

Each year in the United States, more than 12,000 babies are born deaf or hard of hearing; most have two hearing parents. Hearing loss can affect a child's ability to develop communication, language, and social skills. The earlier children with hearing loss start getting services, the more likely they will reach their full potential*.

*Adapted from the Centers for Disease Control and Prevention website – www.cdc.gov

TYPES AND DEGREES OF HEARING LOSS

Results from the hearing testing methods mentioned in the previous section may be presented on an audiogram. An audiogram is a graphic illustration of the responses obtained during a hearing test. Audiograms are essential for understanding the degree of a child's hearing loss, monitoring for any changes in auditory status over time, and determining whether the child will benefit from hearing aids or cochlear implants. However, an audiogram is not an accurate predictor of how a child perceives the speech sounds that he or she hears. Two children with the same audiogram may learn language, and develop speech and auditory skills, in very different manners.

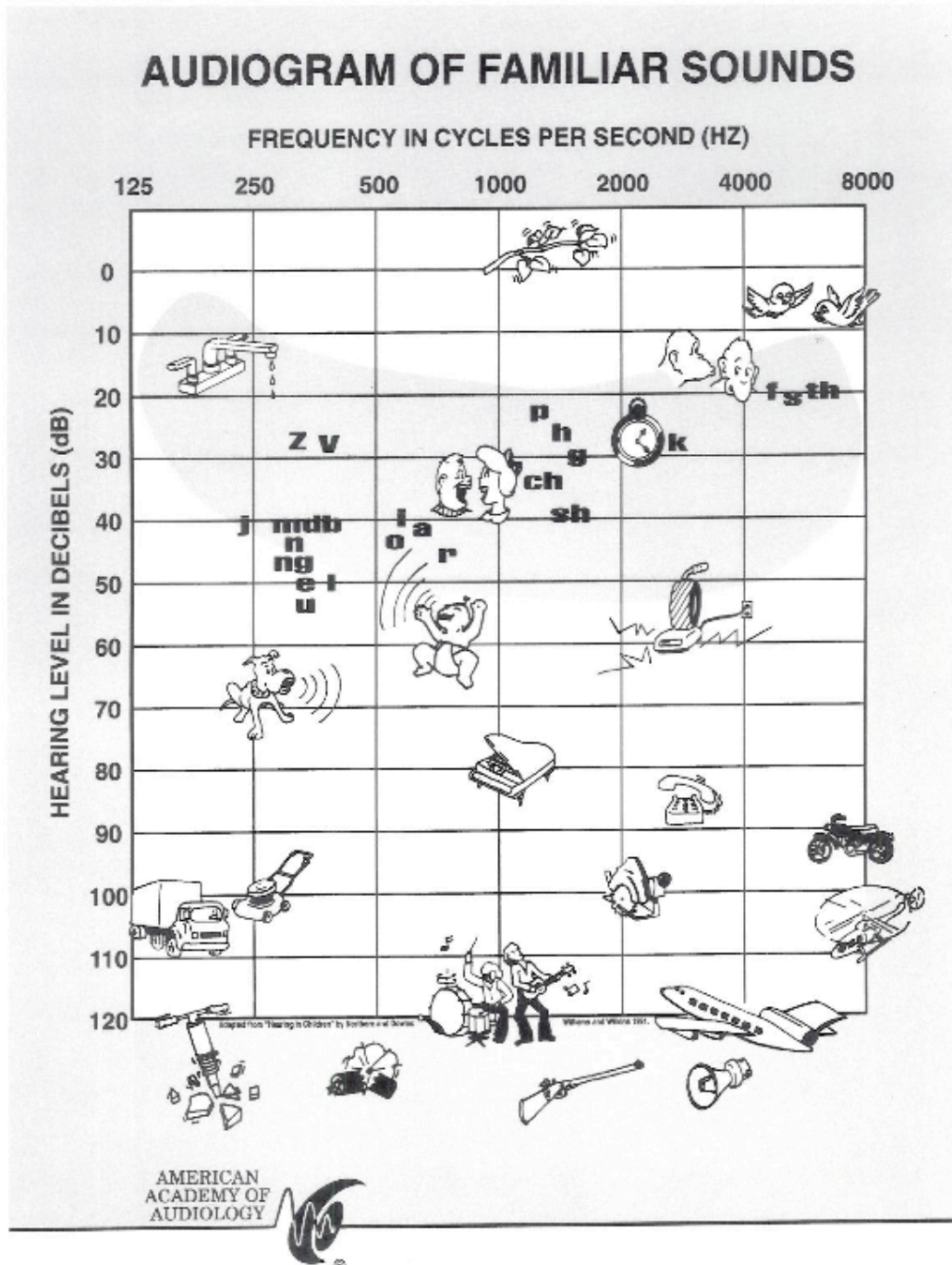
On an audiogram, the frequency of cycles per second (Hz) are shown horizontally. To the layperson, this can be described as pitch, or notes on a musical scale. Frequency is measured by the number of waves or cycles that a sound makes in a single second. Frequency increases on an audiogram from left to right, or from low to high frequency. Children with typical hearing can detect very low frequency of sound (i.e. a fog horn at 20 Hz) to a very high frequency (i.e. a whistle at 20,000 Hz). An audiogram shows test results in the range of frequencies that must be heard for development of speech and language, and thus its range is limited to frequencies between 250-8000 Hz.

The intensity, or loudness, of a sound (measured in decibels or dB) is shown on the vertical scale of an audiogram. The scale moves from soft on the top of the audiogram down to loud on the bottom of the scale. This measures how loud or soft a sound is heard. Children with typical hearing can hear each frequency at a level of 15-20 dB or less. Many audiograms that are presented to children have pictures representing sounds that can be heard by a person with normal hearing. For example, the following are sounds that can be heard at the corresponding decibels (dB):

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

Sound	dB Level
Whisper	about 20 dB
Refrigerator humming	about 40 dB
Normal conversation	about 60 dB
City noise	about 80 dB
Lawnmower	about 90 dB
Chainsaw	about 110 dB
Firecracker	about 140 dB

Audiograms for children also usually include a shaded area (that looks like a banana) that depicts the approximate frequency at which a specific sound is typically heard. Many speech sounds are produced in frequencies between 250 and 5000 Hz, and spoken between 20 to 60 dB. The softest sound is the ‘**th**’ as in the word ‘**with**,’ and the loudest speech sound is ‘**aw**’ as in the word ‘**saw**.’ The speech area on the audiogram is used to illustrate the child’s hearing thresholds (or what they are capable of consistently hearing). The child’s thresholds are marked with an ‘x’ or blue pen for the left ear, and with an ‘o’ or red pen for the right ear. An example of a child’s audiogram (without the threshold markings) follows:



<http://www.audiology.org>

11730 Plaza America Dr., Ste. 300, Reston VA 20190 • 703-790-8466 • 800-AAA-2336 • Fax: 703-790-8631

Source: Adapted from the Northern and Downs textbook, *Hearing in Children* (2002).
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The degree of an individual's hearing loss is determined by his or her hearing thresholds. In general, the following terms are used to categorize the levels of hearing loss:

- Minimal (16 to 25 dB)- This degree of hearing loss can be compared to people with normal hearing lightly plugging their ears with their fingers. A child with this degree of hearing loss typically has few difficulties with communication, but may have difficulty hearing soft speech or speech from a distance. It will most likely be difficult for them to hear speech in loud environments. A child with a hearing threshold of 16 dB can miss up to 10% of speech at a 3-foot distance. A hearing loss that is a few decibels greater can cause the child to hear inconsistent or distorted parts of speech, particularly with word endings like -s and -ed. If this degree of hearing loss is undetected, the child's behavior might be confused for immaturity or inattention.
- Mild (26 to 40 dB)- With this degree of hearing loss, it is often difficult to hear faint or distant speech, and a child cannot hear a whispered conversation even if it is in a quiet environment. A child with this type of hearing loss may hear speech, but tends to not be able to hear word fragments, endings, and indistinct word sounds. With a 30 dB hearing loss, a child can miss 25-40% of the speech signals, and often experiences difficulty learning early reading skills such as letter and sound association. The child may be more fatigued due to the extra effort needed to listen. Hearing instruments are usually recommended for children with this degree of hearing loss.
- Moderate (41 to 55 dB)- A child with this degree of hearing loss has difficulty hearing at conversational levels, and speech must be loud to be understood. The child's speech may also be affected, as some have unclear pronunciation and a flat quality to their voice due to their inability to monitor their own voice. A child with 50 dB hearing loss may miss up to 80% of all speech. In addition to hearing aids, an FM system is often recommended for children to overcome classroom noise and distance. Sign language or visual communication may help to improve language development.
- Moderately Severe (56 to 70 dB)- This degree of hearing loss inhibits a child from hearing conversational speech at conversational levels, as he or she may be able to hear a loud voice from about one foot from the ear. A child's speech and language will most likely be affected, as it is difficult to detect many speech sounds, especially vowels. If this degree of hearing loss is

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

present before the child is one year of age, speech and language will not develop spontaneously without amplification and intervention. Even with the aid of a hearing device, children with this degree of hearing loss may be aware of conversation around them but miss parts of words. A 55 dB hearing loss can result in missing up to 100% of speech without amplification. For children with this degree of hearing loss, hearing aids and an FM system are recommended, along with intense speech and language therapy and/or sign language or visual communication and early language development.

- Severe (71 to 90 dB)- A child with this degree of hearing loss cannot hear conversational speech or loud speech, and may be more aware of vibrations than tonal patterns. Even with hearing aids, the child may be unable to perceive high pitch speech sounds sufficiently. At this level of hearing loss, a child may be a candidate for a cochlear implant and can use sign language to communicate and promote early language development.
- Profound (91 dB or higher)- With this degree of hearing loss, a child cannot hear loud speech and will have difficulty hearing loud sounds. Twenty percent of infants diagnosed with hearing loss are considered to have a profound loss. For most children with this degree of hearing loss, they will not be able to perceive most speech sounds even with the use of traditional hearing aids. A child with profound hearing loss may be a candidate for a cochlear implant or need to rely upon sign language to communicate.

There is not a technical cut off for when a child with hearing loss is considered to be “deaf” or “hard of hearing,” but often a child is considered to be “deaf” if there is a hearing loss from severe to profound in which speech is not understood even with the use of hearing aids. A child is considered to be “hard of hearing” if the degree of hearing loss is in the range where speech is commonly used. Children considered to be “hard of hearing” typically can benefit from the use of hearing aids. As children get older, they may feel more comfortable identifying themselves as being deaf or hard of hearing, based on their own feelings, the language they use, and the groups of people they tend to socialize with. Often the word Deaf is capitalized to signify identification with others who are Deaf and share similar language, experiences, and a sense of common history and “culture.”

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

Even with the assistance of hearing aids and/or cochlear implants, a child who is deaf/HH will not have hearing equivalent to their peers with typical hearing. It is also important to remember that many children who use hearing aids or cochlear implants cannot hear when their hearing device is not being used (i.e. at bedtime or in the pool).

There are several websites that will allow you to experience the sounds with various degrees of hearing loss. For example, the website for the Phonak brand of hearing aids has demonstrations of how everyday sounds would be heard by a person with normal hearing, mild hearing loss, and moderate hearing loss.

www.phonak.com/us/b2c/en/hearing/understanding_hearingloss/how_hearing_loss_sounds.html

Each child's hearing is unique. Different things will impact how a child will use their available hearing. Below are some of the various factors that can impact a child's hearing, and some common terms used to discuss hearing loss.

- **“Age of onset”** is the age in which the hearing loss occurred. A loss that occurred before the child understands and uses language is called “pre-lingual,” while a loss that occurred after the child has some understanding of and is already using language (usually around the age of 2 years) is called a “post-lingual” loss.
- **Bilateral hearing loss** is a hearing loss in both ears.
- **Unilateral hearing loss** affects only one ear. These children typically have difficulty locating the source of sounds. Childhood illnesses, such as mumps, are often the cause of this type of loss.
- **Sensorineural hearing loss** is usually present when there is damage in the inner ear or cochlea and the hair cells are not functioning properly. There is often some distortion of nerve sound signals processed by the brain. This damage may occur during pregnancy or delivery, or it can be caused by genetics, exposure to excessive loud or sudden impulse sounds, ototoxic medications or infections such as measles, mumps, and meningitis. Often permanent hearing loss is classified as sensorineural loss because the way the brain and cochlea are contributing to

the loss of hearing cannot be established. This type of hearing loss cannot be treated medically because the hair cells are not functioning properly. Cochlear implants might be a choice for children with severe to profound sensorineural hearing loss, as those devices bypass the missing or damaged hair cells.

- **Conductive hearing loss** is caused by blocked or reduced movement of sound waves along the route from the outer or middle ear to the inner ear, preventing sound from being conducted normally. A conductive loss could be caused by blockage in the ear or damaged anatomical structures in the ear or ear canal. The causes of a conductive loss include earwax (cerumen) or other debris in the ear canal, fluid behind the ear drum, a middle ear infection (which could lead to a ruptured eardrum if not treated), trauma to the ear drum, a growth (cholesteatoma) on the ear drum, or a problem with the ossicles (middle ear bones). Conductive hearing loss can also result from birth defects in the head and neck, genetics, low birth weight, exposure to loud noises, head injury, and/or repeated middle ear problems.
- **Otitis Media** is an inflammation of the middle ear, commonly referred to as an ear infection. If the fluid is infected, it is often accompanied by a fever and earache. Middle ear fluid without infection is called “otitis media with effusion” (OME) and is usually treated with antibiotics. OME often has no other symptoms other than hearing loss due to a blockage created by the fluid in the middle ear. It can take up to six weeks for this fluid to drain, or tubes may be placed in the ears. The tubes are inserted during a short surgery, and they usually fall out on their own.
- **Mixed hearing loss** is a combination of conductive and sensorineural hearing loss. In this case, sound is not transmitted normally to the inner ear and there are also problems with the inner ear and/or neural parts of the auditory system.
- **Central hearing impairment** is caused by injury, disease, tumor, or unknown problems affecting the auditory centers of the brain. With a central hearing impairment, loudness of sound is not always affected, but the understanding of speech (also called clarity) often is impacted.
- **Central Auditory Processing Disorder (CAPD)** can also be present. This is not a hearing loss, but a problem in the brain that interferes with the ability to interpret sounds correctly.

Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

With a CAPD, it is difficult to localize the origin of sound or distinguish between two similar sounds. This condition is worse in a noisy environment and/or when a child is listening to complex information.

- **Auditory Neuropathy/Auditory Dyssynchrony (ANAD)** is a specific type of hearing loss in which the cochlea appears to be functioning normally, but sound cannot travel to the hearing center of the brain because the auditory nerve is not working properly. This is sometimes referred to as Auditory Dyssynchrony. Children with this condition may be able to hear sounds but cannot understand what those sounds mean, leading to difficulties in understanding speech clearly. To this child, sounds may fade in and out or seem out of sync. Children with this type of hearing loss can hear better on some days and not others, for no discernible reason. There is an increased risk of this condition if the child is born prematurely, has RH incompatibility, or had severe jaundice as a newborn. The following pages describe this type of hearing loss in greater detail and give specific resources for this condition.

Auditory Neuropathy/Auditory Dyssynchrony (ANAD)

What is ANAD?

Auditory Neuropathy/Auditory Dyssynchrony (ANAD) is a hearing disorder in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. Although ANAD is not yet fully understood, scientists believe the condition probably has more than one cause. In some cases, it may involve damage to the inner hair cells located within the cochlea—specialized sensory cells in the inner ear that transmit information about sounds through the nervous system to the brain. Other causes may include faulty connections between the inner hair cells and the nerve leading from the inner ear to the brain, or damage to the nerve itself. A combination of these problems may occur in some cases.

Although outer hair cells—hair cells adjacent to and more numerous than the inner hair cells—are generally more prone to damage than inner hair cells, outer hair cells seem to function normally in people with auditory neuropathy. Outer hair cells help amplify sound vibrations entering the inner ear from the middle ear. When hearing is working normally, the inner hair cells convert these vibrations into electrical signals that travel as nerve impulses to the brain, where the impulses are interpreted as sound.

Risk Factors

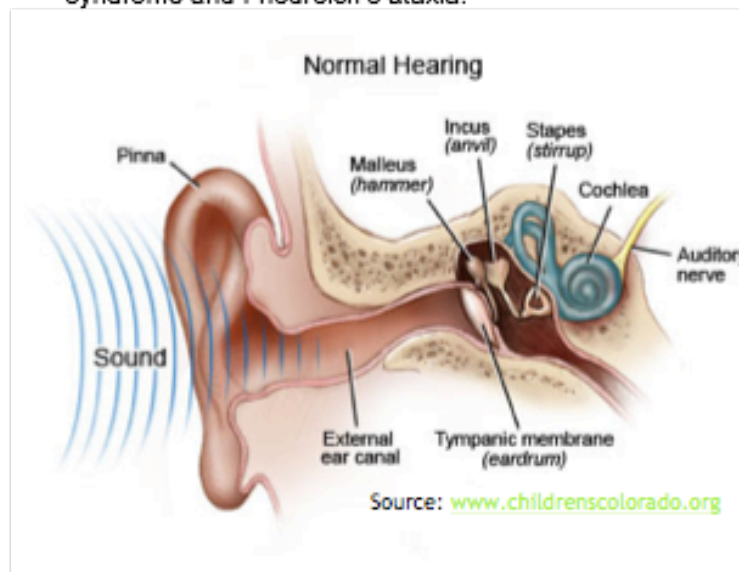
A variety of risk factors and etiologies have been linked to ANAD in children including:

- Anoxia (lack of oxygen) at birth
- Hyperbilirubinemia requiring blood transfusion (associated with severe jaundice during the newborn period)
- Infectious diseases (e.g. mumps)
- Immune disorders (Guillain-Barré syndrome)
- Hereditary (OTOF and DFNB59 gene mutations, Charcot-Marie-Tooth Syndrome, Friedreich's Ataxia, hereditary sensorimotor neuropathy, mitochondrial defects)
- Unknown

There are large individual differences among children with this hearing disorder. For some, hearing may improve over time. This is most common when the cause of the disorder is hyperbilirubinemia.

Genetic Link

The prevalence of ANAD is estimated to be 7-10% of sensorineural hearing loss and 10-14% of severe-profound sensorineural loss. ANAD runs in some families, which suggests that genetic factors may be involved in some cases. Some people with ANAD may have neurological disorders that also cause problems outside of the hearing system. Examples of such disorders are Charcot-Marie-Tooth syndrome and Friedreich's ataxia.



Clinical Presentation

Children with ANAD present with:

- Normal or near normal cochlear hair cell function indicated by present otoacoustic emissions, (OAE's)
- Absent or abnormal auditory nerve function as indicated by an auditory brainstem response (ABR) test
- Present cochlear microphonic response in the ABR where its direction reverses with stimulus polarity changes
- Absent or elevated middle ear muscle reflexes
- Audiometric hearing thresholds that may range from normal to a severe hearing loss and/or may fluctuate
- Difficulty understanding speech, especially in noise
- Speech perception worse than would be predicted by the degree of hearing loss.

Symptoms

Children with ANAD may have normal hearing, or hearing loss ranging from mild to severe. These individuals typically have difficulty understanding speech, especially in noise. Often, speech perception is worse than would be predicted by the degree of hearing loss. For example, a person with ANAD may be able to hear sounds, but would still have difficulty recognizing spoken words. Sounds may fade in and out for these individuals and seem out of sync and hearing may appear to fluctuate from day-to-day or even hour-to-hour. Additionally, other neuropathies may be present that can affect coordination for activities like writing, running, or talking.

Diagnosis

Audiologists use a combination of methods to diagnose ANAD. These include tests of auditory brainstem response (ABR) and otoacoustic emissions (OAE). The hallmark of ANAD is a negligible or very abnormal ABR reading together with a normal OAE reading. A normal OAE reading is a sign that the outer hair cells are working normally.

An ABR test monitors brain wave activity in response to sound using electrodes that are placed on the person's head and ears. An OAE test uses a small, very sensitive microphone inserted into the ear canal to monitor the faint sounds produced by the cochlea's outer hair cells in response to stimuli. Both tests are painless. Other tests may also be recommended as part of a more comprehensive evaluation of an individual's hearing and speech-

Genetic Testing

The genetic inheritance pattern of ANAD is autosomal recessive. This means that the parents of an affected child are unaffected, but carry a single mutation in the gene. Detection of pathogenic mutations in both copies of a child's OTOF (Otoferlin) or DFNB59 (Pejvakin) gene is considered a positive test result for ANAD.

Management

Management of ANAD is often complex due to the varied presentation of the disorder. Some patients benefit from hearing aids. However, many patients get limited hearing aid benefit or no benefit at all. At this time, there is no reliable way to predict who will and will not benefit. Some professionals recommend the use of Frequency Modulated (FM) systems. FM systems can improve listening in noisy backgrounds.

Some patients benefit more from a cochlear implant than from hearing aids. Those with some types of genetically inherited ANAD may benefit more from cochlear implants than patients with other causes of the disorder. Further research in this area is needed.

It is safe to say that no single teaching approach fits all patients with ANAD. It is helpful to work closely with a team of professionals. Some patients benefit from teaching that focuses only on learning to listen and speak. Others benefit from visual communication approaches (i.e., sign language or cued speech).

Resources For Parents/Patients:

1. **Boystown National Research Hospital:**
<http://www.babyhearing.org/HearingAmplification/Causes/Neuropathy.asp>
2. **National Institute on Deafness and other Communication Disorders:**
<http://www.nidcd.nih.gov/health/hearing/neuropathy.asp>
3. **Hands and Voices:**
http://www.handsandvoices.org/comcon/articles/aud_neuropathy.htm
4. **Cued Speech.org:** <http://www.cuedspeech.org.uk/auditory-neuropathy-auditory-dys-synchrony>
5. **Como, Italy Conference ANAD Guidelines:**
<http://www.childrenscolorado.org/conditions/speech/danielscenter/ANS-D-Guidelines.aspx>

Information adapted from:

1. National Institute on Deafness and Other Communication Disorders. *Auditory Neuropathy*. (2011). <http://www.nidcd.nih.gov/health/hearing/neuropathy.asp>
2. Boystown National Research Hospital. *My Baby's Hearing*. (2011). <http://www.babyhearing.org/HearingAmplification/Causes/Neuropathy.asp>
3. The Children's Hospital Guidelines Development Conference. (2008). *Guidelines for Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder*.
4. Partners Healthcare Center for Personalized Genetic Medicine and Harvard Medical School. *Auditory Neuropathy Spectrum Disorder*. (2011). <http://pcpgm.partners.org/Imm/tests/hearing-loss/ANS-D>

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Arizona Parent Resource Guide for Children who are Deaf or Hard of Hearing

Two other hearing related conditions are cochlear dysplasia and enlarged vestibular aqueducts (EVA) syndrome or large vestibular aqueducts (LVA) syndrome. Cochlear dysplasia is a condition in which the cochlea is malformed or has not developed normally. There are several common versions of cochlear dysplasia or malformation: Mondini malformation is the most common and results in only 1.5 turns in the cochlea, instead of the normal 2.5, as well as incomplete development of the membranous and bony labyrinths. Common cavity is the second most common version and occurs when the cochlea and vestibule are merged together forming a single cavity. Michel dysplasia/aplasia is the complete absence of inner ear anatomy and is the most severe form of congenital inner ear malformation, but also the least common. Cochlear implantation in malformed cochleas comes with its' own set of challenges, as does mapping post-implantation, and should be discussed at length with an ENT and audiologist familiar with difficult cases.

Vestibular aqueducts are narrow and bony canals that travel from the inner ear to deep inside the skull. Running through the vestibular aqueduct is a fluid-filled tube called the endolymphatic duct, connecting the inner ear to a balloon-shaped structure called the endolymphatic sac. A child's vestibular aqueducts are considered to be enlarged if they are greater than 1.5 millimeters in diameter. If this is the case, the duct and sac usually grow large as well. Although the function of the vestibular aqueduct and sac is not yet fully understood, the theory is that they ensure the fluid in the inner ear has the correct amount of ions. Most children with EVA develop some degree of hearing loss, and 5-15% of children with sensorineural hearing loss have this condition. Sometimes a diuretic is prescribed in an effort to reduce hearing loss associated with EVA. However, surgery is not considered to be a viable option as it can destroy the child's hearing. Steroids are sometimes given to treat sudden hearing loss, like if a child suffers a head injury and loses his or her hearing. However, no studies currently support the effectiveness of steroid treatments for children with EVA. You can minimize the effects of EVA by having your child avoid contact sports, wear head protection, and avoid extreme changes in pressure (like submerging in very deep water or engaging in any free-fall activities).