

## Auditory Neuropathy

Auditory neuropathy is a unique disorder of hearing in which the inner ear itself may be perfectly normal but speech cannot be heard clearly enough for a child to develop normal speech and language. The problem is in the cochlear nerve itself and its inability to clearly transmit sounds received by the inner ear to the brain where hearing and interpreting of speech actually occurs. If the auditory nerve has neuropathy, a child may be aware of sounds but the signal will be very poor like a very poor and unintelligible telephone connection. The result is that a child will react to sound and give no evidence to the parents that there is a hearing problem until it becomes clear at two years of age or older that despite hearing sounds, no speech has developed. An appointment with a hearing specialist is usually arranged and when routine hearing testing is performed it is found to be normal. This is the unique character of auditory neuropathy that is the most difficult to understand. A hearing test (behavioral or play audiometry) which records the child's responses to sounds may be normal and special testing of the hair cells of the inner ear are usually normal. Only the doctor and audiologists suspicions that a problem may be present lead to further testing which can confirm that sound transmitted through the hearing nerve becomes desynchronized. A standardized form of testing called brainstem evoked response audiometry (BERA) or auditory brainstem response (ABR) will show that the characteristic waves associated with sound conduction down the cochlear nerve are not well formed. The result is that the child with auditory neuropathy hears so poorly that they may never learn to speak intelligibly. Recently, it has become apparent that simple acoustic reflex testing which is often done routinely in adults but not in children, and can be done by most audiologists, can be helpful because these normal reflexes are uniformly absent in children with auditory neuropathy.

Auditory neuropathy may improve spontaneously but the chances of significant improvement after the first two years of life are small. This is very important because so much of early speech and language development happens early in life and incomplete learning cannot easily be corrected later. There is no medication currently known that can improve or speed the improvement of the health of the auditory nerve which has auditory neuropathy. Unfortunately, simple amplification of incoming sounds with hearing aids does not seem to help anymore than turning up a broken loud speaker. Fortunately there is a way to overdrive the poorly functioning nerve with sounds which are clear enough to support the development of speech and language during critical period for speech and language learning early in the child's life. The strong clear stimulus necessary to overdrive the distortion in the hearing nerve can successfully be given by a cochlear implant which is currently the best form of hearing aid for a child with auditory neuropathy. It may be difficult for parents of a child who hears sounds to accept that their child may require a cochlear implant but it is really just a hearing aid for a specific kind of hearing impairment. The goal of allowing speech and language development is the same and the expertise of pediatric implant doctors, audiologists and therapists together is required to deliver the best possible treatment for the child with auditory neuropathy. Even though auditory neuropathy has only been recognized as a common clinical problem in the last few years cochlear implantation has been proven to be a safe, effective and reliable treatment.

Auditory neuropathy may and often does accompany real hearing loss caused by inner ear problems. A cochlear implant is uniquely able to treat both problems. There is currently no way to program a conventional hearing aide to deliver a simplified implant-like signal that can be understood despite the sound blurring caused by the neuropathy.

This brief discussion should help you to better understand auditory neuropathy and the decision making surrounding your child's care. The members of your child's treatment team will help to clarify any remaining questions you may have.