Auditory Neuropathy/ Auditory Dys-synchrony - Cued Speech use

Complete spoken language through vision

Information Sheet 12

Auditory Neuropathy/Dys-synchrony (AN/AD)

There is growing interest in AN/AD and increasingly specialists are recommending Cued Speech. We are very grateful to Charles Berlin Ph.D., Professor of Hearing Science and Clinical Professor of Otolaryngology Head and Neck Surgery, LSU Health Services Center, USA, both for his permission to quote him in the following article and for checking the remaining text for accuracy.

Introduction

Auditory Neuropathy or Auditory Neuropathy/Dys-synchrony (AN/AD) was first recognised in the 1980s and is a comparatively new and a potentially confusing diagnosis. Children are described as having AN/AD when results of two so-called 'objective tests' conflict. Thus, AN/AD is diagnosed when tests of outer hair cells (Otoacoustic emissions) suggest normal hearing but tests of the inner hair cells and nerve fibers (ABR) suggest poor hearing and/or deafness (see overleaf for more information). In other words, the child's ears appear to be 'hearing' by the emissions test but brainstem tests to ascertain whether there is enough neural synchrony to allow speech to be 'heard' within the brain, is grossly abnormal.

Children diagnosed with AN/AD may function as totally deaf or (in about 7% of cases) may experience no problem and acquire language naturally and normally. Other children fit into this continuum anywhere between the two extremes.

For some AN/AD children their hearing may fluctuate from day to day or even hour to hour based on fluctuations in body temperature or background noise. A minority outgrow the disorder. Children tested in clinical conditions in a sound-proof booth may appear to have virtually normal hearing but when noise is added hearing for speech is described as 'universally very poor'.

It is now estimated that 10% of the deaf population may have AN/AD. It is widely recommended that Cued Speech is used with AN/AD babies and children for a number of reasons; one of which is that about 7% of the children with this problem ultimately need NO intervention. Therefore, one does not want to do harm with hearing aids or implants, but can ensure language development regardless of the outcome by using Cued Speech.

Cued Speech is a simple sound-based system comprising eight handshapes used in four positions near the mouth together with the lip patterns of normal speech so as to make all the sounds of spoken language fully comprehensible to deaf babies, children and adults

More information about Cued Speech?

Video, booklet and more information sheets available.

Learn to Cue

Regular courses (some residential) are arranged, with low cost or free tuition.

Call an experienced user now to discuss the benefits of Cued Speech

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Auditory Neuropathy/Dys-synchrony (AN/AD)

What is AN/AD?

It was originally thought that the auditory nerve was always to blame so the condition was described as Auditory Neuropathy.

It now appears that additionally the sound being received by the child's brain can be muddled or 'dys-sychronised' because of the absence or breakdown of a specialised cell in the inner ear, the Inner Hair Cell. It sounds as if the child is listening through a very poorly tuned radio. Sound may be distorted and sometimes sound like little more than static or white noise. This is one of the reasons why some professionals prefer to label the condition Auditory Neuropathy/Dys-synchrony, because often the nerve is perfectly intact.

Recommendations for management of AN/AD

One of the leading authorities on AN/AD is Charles Berlin Ph.D., who is Professor of Hearing Science and Clinical Professor of Otolaryngology Head and Neck Surgery, LSU Health Services Center and also retired Director of the Kresge Hearing Research Laboratory, New Orleans. He has managed over 300 cases of the disorder.

His data has shown that Auditory Verbal Therapy (AVT) prior to a cochlear implant has been the LEAST successful way to manage AN/AD, while AVT AFTER the implant is an excellent choice. He writes: *'Since we saw our first patient 20 years ago (and did not know what we were seeing) and because we have tried hearing aids on many of them (all unsuccessfully as a tool for learning language) we no longer recommend aids or try to correct the audiogram'.*

Professor Berlin makes the following recommendations for the early months:

'*If a child is newborn and has a history of prematurity or hyperbilrubinemia* (neonatal jaundice) *we recommend watchful waiting and regular assessment of normal auditory orienting, babbling and language comprehension. During this watchful waiting period we urge parents to learn to use Cued Speech (CS) as a way to supplement lip reading and teaching the phonology of the home language'*

How the diagnosis is made

Typically children, and now, with early diagnosis, babies, are tested with the following:

1 Otoacoustic Emissions (OAEs) tests whether the outer hair cells of the ears function normally. The OAE test uses small earphones to present clicks or a series of paired tones to the ear. A microphone then measures an echo response from the inner ear and this estimates how the outer hair cells respond to sound.

2 Auditory Brainstem Response (ABR) uses electrodes placed on the head. Like the OAE test, sounds are presented to the ear through small earphones, but in this test the electrodes pick up responses which indicate how the hearing nerve and parts of the brain are responding to sound.

With auditory neuropathy ABR responses are absent or abnormal but the OAEs are (or once were) normal. Early diagnosis is important because over time, or after hearing aid use, the OAEs often disappear and the patients become indistinguishable from those patients with less complicated forms of deafness.

There are other indicators (for example Middle Ear Muscle reflexes which are absent or seriously elevated) which are outside the scope of this article.

Note:

Dr Berlin writes:

'Children and adults with AN/AD can be very confusing. Because their audiograms either fluctuate markedly or range from total deafness to almost normal sensitivity, audiologists and otolaryngologists may mismanage them for long periods of time. This mismanagement may take the form of constant reliance on hearing aids and strict Auditory Verbal Therapy with no visual cues or diagnosing them as "normal" because they have normal otoacoustic emissions or mis-diagnosing them as having "Central Auditory Processing Disorders". Their hearing abilities lie along a continuum from virtually normal in clinical hearing performance in a sound booth (except when noise is added, in which case hearing is universally very poor) to total deafness, but they always have absent or grossly abnormal Brainstem Responses and elevated or absent middle ear muscle reflexes.'

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Language vs. speech

Dr Berlin writes that it is very important to distinguish between speech and language: '*The family must understand and appreciate the many differences between speech and language.*

'Our primary job is to teach children LANGUAGE by stimulating their brains with meaningful words, sounds, symbols and associations. Normal-hearing children learn language as a result of auditory stimulation, by eavesdropping, imitating, playing verbal games, singing songs, etc. Children with hearing disorders cannot learn language as easily by eavesdropping like their normal-hearing peers. They have to learn by eavesdropping VISUALLY. Thus, language can be learned and appreciated WITHOUT hearing speech clearly and without expecting speech to be produced. That is to say, abstract concepts and their representation can be signed, or Cued (eg put the book ON…under…along side of…the table; play nicely; show me a picture of ….; where is…; what day is today?; etc.) but until the child has grasped the nature of these abstractions, speech will not necessarily follow. Signs generally carry NO representation of the sounds of the language. Thus the signs for BABY and the sign for BOY have no /b/ sound coded in them. Those words, when properly Cued, will always have a representation of a /b/ sound in them.

'Using Cued Speech, families can also raise multilingual children who might understand Spanish, English and Dutch for example. I know of families where the children use three modes of communication, spoken English, ASL, and Cued English.

'The main point here is that if parents want speaking children, they first have to teach language comprehension and that is most easily accomplished by eavesdropping and imitating.'

Successful interventions

'For all AN/AD children, whether they outgrow the disorder or not, lip-reading and facial cue reading will nurture language growth, while hearing aids and COVERING the mouth has not in the past been very successful. Hence the unique value of Cued Speech as a tool to allow the child to eavesdrop on the spoken language of the home regardless of whether invasive interventions are used. Its major advantages are that the family does NOT have to learn a new sign for each concept or word, and the grammar, syntax and vocabulary match whatever the family uses for speech. Thus CS is ideally suited for bi-lingual families because Cued Speech can be used as a tool to teach virtually ANY language.

'Cued Speech complements baby signs, signing and speaking and cochlear implants ideally, and we have seen the fastest language growth and quickest transition to spoken language and literacy from children who have been implanted by one year of age and/or were exposed to CS and then had implants after age 2. CS remains a lifelong tool, useful when implants fail or when the implants are off during bathing, swimming, or for adults where ambiguity of spoken messages cannot be resolved by hearing alone, etc.''

What are the most common causes?

Dr Berlin writes:

'*The most common causes shown in our data base of 300 patients are:*

- *Genetic absence or compromise of inner hair cells secondary to otoferlin or other mutations (like Mohr-Tranebjaerg) affecting the senses. These sometimes spread to outer hair cells and obliterate otoacoustic emissions, complicating the diagnosis. Some of the rare forms bring with them late onset blindness and ataxia making visual forms of language support more complicated.*
- *Genetic diseases of the peripheral nerves (eg Charcot Marie Tooth Disease, etc.)*
- *Mitochondrial disorders which affect vision, motor function as well as hearing.*
- *Absence or compromise of inner hair cells secondary to anoxic or hypoxic episodes.*
- *Hyperbilirubinemia requiring transfusion, with or without concurrent kernicterus.*
- *Premature birth with low Apgar scores and respiratory distress.*
- *Ototoxic drugs which affect inner hair cells.*

Background on the Inner Ear

'The ear contains both hair cells and nerve fibers. Hair cells are divided into 2 types, outer hair cells and inner hair cells. The outer hair cells actually contract and expand and parts of the cell appear to be low level pre-amplifiers. That is to say they strengthen faint sounds so that the inner ear can detect and respond to them more readily. The parts of the inner ear that actually stimulate the hearing nerve fibers are the INNER hair cells. Thus, the outer hair cells (which are the source of otoacoustic emissions or sounds coming FROM the ear) amplify the sounds, and the inner hair cells activate the hearing nerve fibers for the brain to interpret. Auditory Neuropathy/dys-synchrony is a disorder which strikes anywhere from the inner hair cells to the nerve fibers. It affects the outer hair cells sometimes in later stages or after prolonged hearing aid use.'

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Early Cochlear Implantation

Given the fluid nature of AN/AD and the possibility of spontaneous recovery **early and almost immediate** cochlear implantation is only strongly recommended in the few cases where the cause of the AN/AD is clearly hereditary and when the history of other family members would indicate that this is the best route. It is however always the parents' choice and implantation can be very successful especially when preceded by and later supported by CS. Cued Speech is always a useful tool for teaching language and syntax and clarifying the new sounds that the child will hear through the implant.

Dr Berlin writes:

Management and Outcomes

'Auditory Neuropathy/Dys-synchrony as a diagnosis does NOT necessarily mean the nerve of hearing is irreparably damaged or that nothing can be done for it. In about 2/3rds of the cases there is no demonstrable peripheral nerve damage and in those who have such damage, palliation is still possible. (Starr et al.)

'In virtually all well-programmed cases we have seen so far, cochlear implants have helped synchronize the auditory nervous system and assist in speech and language development or clarity. (Shallop et al. House group etc.)

'Hearing aids have been recommended and we tried them for 20 years. We have yet to see patients learn language and show normal speech development with hearing aids and Auditory Verbal Therapy (with no visual cues) at the same rate as a child with simple outer hair cell loss. This conflicts in part with other people's reports in the literature (eg Rance et al.) but, had we published even our first 10 years of experience, we would have also thought that hearing aids were of value because they seemed to enhance sound field sensitivity and "improve the audiogram". Here we must repeat an important principle of patient management which I first heard from Dr. Nancy Young, a Chicago-based paediatric otolaryngologist: Treat the CHILD not the TEST RESULTS.

'As we said earlier, a few AN/AD children we have followed learn language with no intervention although they persist in having trouble hearing in noise. They are often mis-diagnosed later in life as having "Central Auditory Processing Disorders". The absence of middle ear muscle reflexes and an abnormal ABR test quickly identifies them as having a mild form of Auditory Neuropathy.'

Dr Berlin's recommendations & conclusions:

'Our goal is to help you raise a "literate taxpayer", a phrase I learned from one of my patient's fathers.

'Because the outcome with AN/AD children is not nearly as predictable as with simple outer hair cell hearing loss, we urge families with children who have this diagnosis to first learn and institute Cued Speech around the child so that language comprehension can be assured. Baby signs do NOT conflict with cues and can be used to communicate along with speech so that the baby can have an additional method of expression back to the parents. Then the family is prepared for any number of outcomes.

- *'If the child acts deaf and remains delayed in spoken language, Cued Speech and signs will help assure language acquisition. At that point the family has many options, including, at one extreme, adopting Deaf Culture and Signs for their family, or at the other end of the continuum opting for a cochlear implant and spoken language. Either way the use of cues will have facilitated language acquisition and done no harm.*

- *'If the child shows few if any signs of auditory problems, the Cued Speech will help in noisy situations but can be phased out if and when it is no longer needed.*

- *'If and when the child is implanted, Cued Speech will help disambiguate the new sounds and phonemes the child will learn with the implant, and can be retained as a tool for difficult listening situations or when the implant is off.*

- *'American Sign Language is NOT spoken English on the hands but has a spatial and visual syntax and grammar of its own. If the family opts to do nothing invasive other than use visual language, combining ASL with cues will allow the child to join Deaf Culture, while still having an additional language acquisition tool that supports literacy and English (or any other desired language) word order and usage. Thus the child can have the joys and benefits of belonging to the Deaf world, but have a tool that will make it unnecessary to learn English as a second language.'*

Further information?

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