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Managing Patients with Auditory Neuropathy / Auditory Dys-synchrony

Charles I. Berlin, PhD

If you are an audiologist who has to manage a family with Auditory Neuropathy, this may help. To better understand the disorder it perhaps should be re-named as AUDITORY DYS-SYNCHRONY.

Auditory Neuropathy is better defined as auditory dys-synchrony. It is seen when OAEs are present and ABRs are absent. Sometimes the ABR appears to be present but it is really Cochlear Microphonic or hair cell response rather than a neural response. Using one positive and one negative polarity click often resolves the problem and separates neural from hair cell responses because the hair cell responses reverse polarity when the neural responses do not. Middle ear reflexes are virtually always absent.

One of my parents started a list serve for parents and professionals dealing with AN at: <http://www.onelist.com/community/AuditoryNeuropathy>

We hold out great hope for the parents because Linda Hood and I (along with Arnie Starr, Terry Picton, Yvonne Sininger, Jon Shallop Laszlo Stein, Nina Kraus, Mike Gorga, Pierre Deltenre, and others) have seen and consulted on over 250 such patients. The newborn patients, especially those with surprisingly mild hyepbilirubinemia (under 14 ml/dl), fall into four categories:

1. Some actually get better, start to hear and speak within a year or two.
2. Some get worse, lose their emissions and cochlear microphonics.
3. Some stay the same.
4. Some develop peripheral neuropathies later in life. This latter category more commonly describes adult onset AN.

Their speech perception is seriously de-synchronized and in my experience auditory verbal therapy, where there are no visual cues, usually does NOT work. Nor do hearing aids except to improve sensitivity, but they do not lead to auditory verbal language comprehension. If there is any usable hearing, it fails rapidly with the slightest bit of simultaneous noise. Thus they sometimes appear to hear and sometimes don't. Please note that when parents of AN children say that their child sometimes hears without hearing aids, that this very well may be true and should not be ascribed to the parents' failure to accept their child's deafness.

Pouring visual language (I prefer Cued Speech {CS} but don't insist on it) into these children as early as possible postures the parents to handle this bizarre state of circumstances. If the child gets better, no harm is done and if CS is used it facilitates phonologic awareness and English Grammar. It is

also easier to learn than American Sign Language (ASL) or Signed Exact English (SEE) and a new sign is not needed for any word already in the parents' vocabulary. Using it around the house, especially as family members communicate to one another, allows the child to eavesdrop on the family interactions and learn language naturally. See <http://www.cuedspeech.org> for more information

The strategy of making language visible while we watch and wait is the most likely to maximize success. (Success here is defined as raising a "literate taxpayer".*) If the child's hearing deteriorates over time, he has language from the visual mode. And CS does NOT conflict with ASL or SEE. If the child's hearing improves, no harm has been done, whereas the output of most hearing aids can actually destroy normal inner ear tissues.

While hearing aids don't work well for any length of time, I know of 14 children in the US to date who have been implanted 12 successfully. I think because the electrical stimulation re-synchronizes the nerve fibers which have been disabled either because of poor mechanical coupling, poor biochemistry pre-synaptically or (less likely) because of axonic disease. As of today, I know of four AN children who are about to be implanted and I will update this page regularly as we get new data.

So what do we tell the parents? The child can go one of four ways (see Four Categories above). Pour visual language into the child as soon as you can, watch and wait for a year or two and if by age 18 months to two years you don't see much audition developing and if you want an oral auditory child, consider an implant. If you insist on a hearing aid, use JUST ONE and monitor the emissions and CM in BOTH ears to see if the child is progressively losing auditory function. If the child loses emissions and CM in both ears, consider the implant. If he keeps his emissions, also consider the implant. Such children can easily live in Deaf Culture as well,-- keep that open as an option if the parents so desire. AS LONG AS YOU GET VISUAL LANGUAGE INTO THE CHILD AS SOON AS POSSIBLE YOU ARE ON THE RIGHT TRACK.

If the parents want an implant and get one, CONTINUE TO USE VISUAL CUES and VISUAL LANGUAGE and gradually fade them out as the child learns to associate what he hears with what he has seen so far. Ultimately you want him to use his ears and brain but visual modes like CS virtually guarantee a literate taxpayer* at the end of this journey, WHETHER OR NOT THE IMPLANT IS SUCCESSFUL.

We also discourage parents from becoming zealots for a particular educational or social methodology. All of these things are just tools to be available and make life and communication easier for the AN child and his family.

There is lots more to tell. Call my voice mail at 1-800-497-4327 and tell me when you want me to call you back at our expense.

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P.S. If you work around or have access to a large school for the Deaf where people don't value hearing aids or speech, let us know. That is where we

find most of these AN patients and will be glad to come there and study the children in the school. We also find a surprising number among successfully implanted patients check their unoperated ear for emissions!!

* A concept taught to me by Dylan S. , the parent of one of my now-implanted patients.



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