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Pediatric Auditory Neuropathy/Dys-Synchrony

Universal newborn hearing screening (UNHS) is currently the standard of care in 42 states. In addition to lowering the average age of identification of permanent childhood hearing loss, advances in audiological assessment have resulted in greater specification of what portion of the auditory system might be affected in cases of permanent hearing loss. The ability to evaluate the function of the cochlea separately from the auditory nerve now exists. Children with auditory neuropathy or dys-synchrony have evidence of normal cochlear function, but show impairment in the function of the auditory nerve. Auditory neuropathy/dys-synchrony presents significant challenges related to identification and management of hearing loss during childhood.

What is Auditory Neuropathy/Dys-Synchrony (AN/AD)?

AN/AD is a type of sensorineural hearing loss that can be congenital or acquired. Unlike other types of sensorineural hearing loss where both Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR) tests are likely to be abnormal, AN/AD is characterized by normal OAE results and significantly abnormal ABR responses, even when measured with very loud sounds. The combination of normal OAE responses and severely impaired ABR responses is thought to reflect normal outer hair cell (OHC) function in the cochlea and abnormal auditory nerve function. The site of lesion for AN/AD is often unknown, but possibilities include cochlear inner hair cells, cochlear spiral ganglia, and the auditory nerve. Audiograms of children with AN/AD vary from hearing in the normal range with complaints of difficulty hearing in background noise to profound hearing loss.

Who is at Risk for AN/AD and How Many Children are Affected?

Multiple risk factors for AN/AD have been described in the research literature:

- Hyperbilirubinemia (> 20 mg/dL total serum bilirubin, Shapiro, 2005)
- Anoxia / hypoxia z Family history of childhood hearing loss
- Prenatal/neonatal infections
- Congenital neurological problems

The incidence of permanent childhood hearing loss is estimated to be between 2 and 3 of every 1,000 babies born in the United States. Children with AN/AD represent a subset of the overall number of children with hearing loss. Recent data suggest that the incidence of AN/AD in the general population may be 1 to 2 per 10,000 births (Foerst et al. 2006). However, with OAEs being widely used as the primary screening method, and the fact that infants with AN/AD will pass OAEs , the exact incidence could be higher.

Does UNHS detect AN/AD?

If AN/AD is present at birth, detection depends on the type of screening protocol utilized. As previously discussed, children with AN/AD often have normal OAE results with a significantly abnormal ABR response. If newborn hearing screening consists only of OAE testing or if OAE is the first stage in the screening process, most children with AN/AD will not be identified. Hearing screening programs that utilize ABR will appropriately refer infants with AN/AD.

Unfortunately, the cost of providing ABR screening to every infant born in the US would increase the costs of UNHS substantially. The 2007 Joint Committee on Infant Hearing Position Statement recently recommended that infants who spend more than 5 days in the NICU should receive an ABR screening to improve identification of AN/AD. Therefore, many newborn hearing screening programs have elected to perform ABR screening on all babies with risk factors for AN/AD. Physicians should familiarize themselves with the type of screening tests that are utilized in local hospitals. Infants with risk factors for AN/AD or a history of residence in the NICU should be referred for ABR evaluation if one was not completed as part of the hearing screening.

What Types of Assessments and Intervention Should Be Provided?

Just as in all cases of childhood hearing loss, referral for early intervention services is critical as soon as a diagnosis is confirmed. Pediatric audiologists can provide diagnostic confirmation of hearing sensitivity and ongoing monitoring. Management of referrals to specialists is best coordinated through the Medical Home. Evaluations by an otolaryngologist, neurologist, geneticist and ophthalmologist help to identify any additional medical conditions.

Childhood AN/AD can result from a wide variety of etiologies. Because of this variability, children with AN/AD also have a wide range of responsiveness to sound. Some have behavioral hearing thresholds in the normal range, while others may have limited or no responsiveness to sound. Unfortunately, results from diagnostic OAE and ABR results will not predict a child's behavioral auditory responsiveness, despite the importance of these measures in the diagnosis of AN/AD. Given the spectrum of possible behavioral outcomes, intervention strategies should be customized based on the needs of each child. No single type of intervention is likely be effective for every child with AN/AD. Behavioral audiological evaluation should be completed as soon as it is developmentally appropriate, typically around 6 months of age. Behavioral audiometric results may not be obtainable until 12 months or later in children with concomitant developmental delays. Because hearing may fluctuate in children with AN/AD, follow-up audiological monitoring should ideally occur every three months until the child is three years old and every six months between three and five years.

Hearing Aids

For children with AN/AD who have behavioral audiometric responses to sound in the mild to severe hearing loss range, a trial with appropriately fi t hearing aids is an essential part of the early intervention process. Hearing aids provide audibility for speech sounds and improve access to language. Even with hearing aids, some children with AN/AD will not show improved sound awareness or progress in speech and language development. Research has indicated that approximately 50% of children with AN/AD will show improvement in speech recognition scores with hearing aids, while the remaining 50% show limited benefit (Rance, 2005). Clinical tools to predict which children with AN/AD will be successful with hearing

aids are not currently available. Therefore, a trial with amplification is recommended as soon as reliable behavioral audiometric data can be obtained. Since the child may or may not derive benefit from hearing aids, loaner hearing aids or an extended trial period through the hearing aid manufacturer should be provided whenever possible. Parents should be advised that their child's responsiveness with hearing aids may or may not improve. Monitoring of speech and language development as well as auditory skills should be undertaken to help assess the impact of amplification.

Cochlear Implants

For children with AN/AD who do not show improved responsiveness with hearing aids, research has suggested that cochlear implants (CI) can improve auditory awareness and speech and language development for some children. Several studies have compared children with AN/AD who received cochlear implants to children with other types of sensorineural hearing loss who use cochlear implants and have not found significant differences between the two groups. However, in some cases of AN/AD where the auditory nerve is either small or absent, cochlear implantation has not been as effective, and the results are much more variable (Bradley, et al. 2008). Based on these findings, cochlear implantation should be considered with caution in cases where hearing aids do not improve auditory responsiveness or result in improvement of speech and language skills. Parents should participate in extensive discussions with the ENT surgeon and audiologist to evaluate the risks and benefits of cochlear implantation for their child.

Other Considerations

Children with AN/AD are a unique and heterogeneous group. While some can develop auditory and oral skills necessary for spoken language development, others may require sign language or other visual cues to support their progress, even in cases where hearing aid and cochlear implant use has been implemented. Supporting the child's development based on individual skills and abilities is critical. Because nearly all children with AN/AD demonstrate significant difficulty listening in background noise, additional hearing assistance technology may be appropriate in school or in specific listening situations.

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