NEWBORN HEARING SCREENING AND ASSESSMENT

Assessment and Management of Auditory Neuropathy / Auditory Dys-synchrony.

A Recommended Protocol

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SUMMARY
This document outlines the recommendations of the UK Newborn Hearing Screening Programme for the investigation and management of infants suspected of having Auditory Neuropathy (AN) or Auditory Dys-synchrony (AD). It draws extensively, with permission, from a draft Australian document[1].

1. BACKGROUND
1.1 Definitions and Terminology
We are in this document addressing the practical issues of management where we see the following pattern of test findings at the initial audiological assessment after the newborn screen:

- **Auditory Brainstem Response (ABR) absent or with severely abnormal morphology at high stimulus levels[a];**
- **Otoacoustic emissions (OAEs) and/or cochlear microphonic (CM) present;**

We thus have objective tests that demonstrate the presence of pre-neural responses but absent or abnormal neural responses.

This suggests activity in the outer haircells and perhaps some firing of the auditory nerve, but that the neural firing is desynchronised or that there is some other disruption or dys-synchrony in the transmission along the neural pathway so that no clear ABR is recordable. However the pattern of results could also be the result of delayed maturation or myelinisation of the auditory pathway, particularly in ex-SCBU / low birthweight babies.

The term 'Auditory Neuropathy' was coined by Starr and colleagues in 1996[3]. However some feel this implies a particular pathology and may be misleading[4] and have therefore preferred terms such as 'Auditory Dys-synchrony'[5] (AD), Auditory De-synchrony'[6] or 'Auditory mismatch'[7], which attempt to merely describe, or reflect better what is happening in the auditory system, without ascribing a specific locus of pathology. The term AD would therefore include both true AN (*i.e*. a true neural abnormality) and other possible underlying mechanisms resulting in neural dys-synchrony, as well as delayed maturation of the lower level auditory pathway. However all such children need to be reviewed and monitored in the same way and our management is unaltered by the label attached. We have therefore adopted the term AN/AD in this document.

AN/AD is a disorder that affects neural processing of auditory stimuli that will reduce a child's ability to understand speech and may affect ability to detect sound to various degrees.

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[a] As a definition of "severely abnormal ABR morphology" we suggest that of Sininger (2002)[2]:"The neural response (ABR) will be poor or completely absent but will occasionally show a small wave V response [at high stimulus intensities]. The majority of cases of AN/AD have a poor ABR preceded by a large inverting CM that can last up to 5 or 6 ms."

1.2 Prevalence

Sininger[2] estimates that AN/AD occurs in about 1 in 10 children who have hearing loss *and* severely abnormal ABR *i.e.* one in ten of those who, because of absent ABR, might at first sight be thought to have severe/profound sensorineural (cochlear) hearing loss (SNHL). The true prevalence of AN/AD in the paediatric population with hearing loss, however, is unknown. Moreover, the prevalence of indicators of AN/AD in the special care and well-baby populations has not been determined in large, prospective multi-centre investigations.

1.3 Natural History

It appears that AN/AD may stay the same, worsen or improve.

- The abnormal ABR may recover so that it is consistent with the behavioural threshold and with normal morphology. If the problem is due to maturation, recovery would normally be complete by 12-18 months. If not another cause of AN/AD should be suspected.
- In some cases perceptual ability seems to improve although ABR remains abnormal.
- OAEs which are present at initial assessment may disappear, whether or not the child is aided[2],

Children with AN/AD should be monitored carefully but we should guard against giving false hope that the condition will recover. Equally we should be careful to avoid assigning the diagnosis prematurely.

The impact of AN/AD on a child's hearing ability varies amongst individuals. It is not possible to predict either a degree of hearing loss or a prognosis for speech and language development based on the diagnosis of AN/AD.

1.4 Presentation

A baby with AN/AD may present with

- Absent or severely abnormal ABR at maximum stimulus levels (up to 100dBnHL)
- Evidence of normal outer haircell functioning (OAE or CM present)
- Absent or elevated stapedial reflexes (SRs)[b]

When older, children with AN/AD may exhibit some or all of the following features:

- Behavioural thresholds anywhere in the range from normal to profound
- Poorer thresholds in the low frequencies
- Variable responses from test to test, but generally reliable within a single test
- Auropalpebral reflex may be absent regardless of the degree of hearing loss.
- Speech discrimination poorer than the behavioural audiogram would suggest

[b] Stapedial reflexes are invariably absent or elevated[8] in cases of AN/AD (Hood, personal communication, reports some elevated reflexes in just 5 of their 225 AN/AD cases). However for infants under about 4 months where tympanometry and SRs must be recorded with invalid, high frequency probe tones[9] (1000Hz) there is no published evidence to demonstrate that they are practicably recordable or as useful in identifying AN/AD in young babies as in older children. Hence at present SRs are not part of the standard test battery at this age: this may change when further evidence accrues.

- Hearing aids may be of less benefit than the behavioural audiogram would suggest.
- Greater difficulties hearing in competing noise than expected from the behavioural audiogram and other features indicative of auditory processing difficulties.

Thresholds bear little relationship to speech discrimination ability and therefore management decisions are based on functional communication development rather than behavioural or ABR thresholds, unlike children with sensorineural hearing loss.

Children who have AN/AD will present with many different audiological and communication profiles, and will not fit a particular pattern of communication development or auditory responsiveness.

Although most AN/AD cases are thought to occur among special care/neonatal intensive care babies (see risk factors[b]), they can occur in the well-baby population and may of course pass the OAE screen. Currently AN/AD is not a target condition for NHSP, but by default will be detected by the protocol in SCBU/NICU babies[c]. Cases of AN/AD may thus be referred at a later stage and will need to be investigated, identified and managed then.

2. INITIAL ASSESSMENT

In the NHSP protocol, click ABR assessment will usually be the first investigation for babies referred following the screen. Where this shows no ABR response or a severely abnormal response at maximum presentation levels (80dBnHL or above), investigations to differentiate between AN/AD and SNHL (cochlear hearing loss) must be performed.

In an infant, abnormal or absent ABR may be due to:

1. Hearing loss - sensorineural, conductive or mixed
or
2. AN/AD due to delayed neural maturation
or

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[c] **Risk Factors for Auditory Neuropathy**

- Hyperbilirubinaemia
- Anoxia
- Hypoxia/Prolonged assisted ventilation[10]
- Extreme prematurity (<28/40)[10]
- Congenital brain anomalies[10]
- Demyelinating conditions such as Multiple Sclerosis
- Syndromes associated with other peripheral neuropathies (Charcot-Marie-Tooth, Friedrich's Ataxia)
- Genetic factors[2,11]

Auditory Neuropathy has also been reported in cases where it is related to a viral infection[12] or exacerbated by fever[13]

[c] In this document 'SCBU/NICU' means those classified as such by the NHSP screening protocol – *i.e.* those who are admitted to special care / neonatal intensive care for over 48 hours.

3. AN/AD due to other causes.

A diagnosis of AN/AD must always be excluded before proceeding to hearing aids on the basis of objective test results. ABR alone is not sufficient to do this[d]. As defined above the AN/AD test protocol should be followed as part of the assessment of every suspected case of permanent hearing loss. The NHSP protocols for the specific tests should be referred to[9].

In summary the assessment should include:

- **ABR - CM** response[e]. If ABR is absent or severely abnormal at 80dBnHL (or higher level), perform click CM with separate runs of condensation and rarefaction at 80dBnHL. This is not necessary if a robust OAE has already been shown to be clearly present and repeatable, but it may be useful as confirmation. If a cochlear microphonic (inverting with click polarity) is present and there is still no later true neural ABR response (*i.e.* not inverting with click polarity), assume AN/AD may be present. [See Appendix 2 for detailed notes on CM recording].
- **OAEs - either transient evoked (TEOAE) or distortion product (DPOAE).**

At assessments when infants are aged above about 4 months (see [b] above) also include if possible,

- **Stapedial reflexes (SRs) / Tympanometry.** Measurement should be done with 1000 and 2000 Hz stimuli and measurements should be contralateral if possible, otherwise ipsilateral.

Interpretation:

ABR (AC and BC) absent]
CM or OAEs present] implies AN/AD
(SRs absent / elevated)]
(Note OAEs may be absent due to conductive loss)

[d] Note that hydrocephalus may also present with wave I only on ABR testing. One should therefore be cautious before labelling as AN/AD.

[e] In the study by Rance et al[14] all subjects had evidence of outer hair cell function in the form of the Cochlear Microphonic but only about half had OAEs: this is presumably because the CM is less affected by middle ear factors. ***Therefore, all children with abnormal morphology or absent ABR tracings should be tested for a Cochlear Microphonic, even if OAE is absent*.**

ABR (AC and BC) absent / elevated	**]**	
CM and OAEs absent	**]**	**implies SNHL or CHL**

A key issue within AN/AD is **distinguishing true AN from delayed maturation** particularly in ex-SCBU/ very low birthweight babies. Care should be taken when interpreting ABR results for babies born prematurely or for those who have delays in other aspects of development, as the ABR response may still be maturing. To differentiate neural maturation changes from other causes of AN/AD, whenever possible ABR should be repeated before a definitive diagnosis is made, particularly in "at risk" babies: this should be preferably around 8-10 weeks corrected age and again at 9-15 months.

Note that in AN/AD, ABR thresholds do not predict behavioural thresholds

3. MANAGEMENT

The key issues in management are information and support for the family (particularly in view of the uncertainties around prognosis), the need for repeated audiological and communication assessment, and decisions about hearing aids or other intervention.

Children with AN/AD need reassessment at frequent intervals to determine whether the problem is maturational or not, and because of potential fluctuations in auditory function.

Families of children with AN/AD should be offered referral for early support. Children with AN/AD are at risk for communication difficulties and need to be monitored accordingly. The overall goal is to begin intervention as soon as the parents/carers feel ready to proceed and to establish a communication method for use by the child and family, and put in place a plan for continuing assessment of hearing and communication.

The decision on aiding will usually be delayed until behavioural thresholds are available

The management of the child with AN/AD requires a multidisciplinary team approach which may involve some or all of the following: audiology; audiological medicine; paediatrics; paediatric neurology; speech-language therapy; early education support; ENT/otolaryngology; genetics; neonatology; and families. One member of team should be designated to take ultimate responsibility for the management of the case. The timing of involvement of these professionals will depend on the individual case and the wishes of the family. All the members should be familiar with and knowledgeable about the condition. As a minimum we suggest the team should include a Paediatric Audiologist, a medical person (Audiological Physician, ENT consultant or Paediatrician), a Speech-Language Therapist, a Teacher of the Deaf, and a Neurologist.

As the number of AN/AD cases in any one area are very small and their management presents great practical challenges, we would like to encourage a network of teams with

expertise in this area, for tertiary referral. Such teams could offer support and guidance in diagnosis and management, ensure that families get the best information and advice, and build confidence in the local staff. Such teams should meet regularly to discuss cases, learn from experience and coordinate research.

3.1 Information and Support

The term AN/AD is a label for a pattern of test results; it is not a diagnosis and it is not immediately possible to predict the impact or even the most successful form of treatment. **Support and encouragement for parents is vital at this time.**

The confusion that parents are likely to feel may have a negative impact on the relationship between parent and professional. Ongoing communication, support, and information for parents is critical to successful management. As such, it is important to provide written information regarding the condition to families as well as to other professionals involved with the child, such as Health Visitors and GPs. AN/AD should be described specifically, including what is known and not known about the condition. A good information pack for parents would be invaluable and should be produced centrally in conjunction with parent groups. A national support group should also be considered.

It may be helpful to cover the following points:

- The term AN/AD is a label for a pattern of test results; it is not a label for a child.
- An absent ABR tracing does not necessarily imply a profound hearing loss.
- A diagnosis of AN/AD raises a warning flag for the audiologist and parents. We need to watch this child closely, as the child may not respond to sound in a typical way.
- Many children with AN/AD are able to make good use of their hearing.
- While we cannot predict the impact of AN/AD on the child at this early stage, by pooling test results and observations from parents, audiologists and other professionals we will be able to do as much for the child as possible.
- We cannot predict the impact on the child

3.2. Ongoing Audiological Assessment

Although maturation as a cause of AN/AD may be ruled out by the age of 12-18 months the audiological profile of children with AN/AD may not be stable. Therefore (with parental consent) it is critical that there is, at a minimum, ongoing and regular monitoring of auditory status (behavioural, electrophysiological and middle ear) and hearing, speech, language and general development.

a) Behavioural thresholds in each ear determined by an age-appropriate method (conventional Pure Tone Audiometry, Play Audiometry, VRA with insert earphones). These can be acquired when an infant reaches a developmental age of around 6 months and guide decisions on hearing aids. Any Behavioural Observational Audiometry (BOA) should be carried out and interpreted with extreme care[9]. If reliable results cannot be obtained because of significant developmental delay, BOA and informal observation may be useful in guiding management.

b) Electrophysiology - repeat ABR, CM and OAE recording at 8-10 weeks and 9-15 months.

c) Middle ear measurements/SRs. Monitoring of middle ear status is important as the presence of fluid in the middle ear will affect other tests, and children with AN/AD are as likely as any other to develop middle ear effusion. This should be done in conjunction with other testing. SRs should be measured also - this becomes more reliable and important beyond the early months

3.3 Monitoring and Assessment of Communication development

This may be done by the speech-language therapist, teacher of the deaf and families. This is the key determinant of management options. It requires standardised tests and functional observations.

Standardised tools (for example from the Early Support Programme[15]) should be used to determine communication development in infants and young children.

3.4 Intervention /Aids to Communication

a) Conventional Hearing Aids

There is some evidence that a significant number of children with AN/AD derive benefit from hearing aid fitting if there is a significant behavioural hearing loss[14] and about 50% of the children in this study gained significant benefit. Therefore, following the diagnosis of AN/AD, a trial of amplification should be undertaken. However because of doubt as to the benefit in children where behavioural thresholds are near-normal, **we would only recommend aiding for a child who has reliable elevated behavioural thresholds.** A number of other considerations and complications apply - for further details and discussion please see reference[1].

The decision on whether to aid can only be based on behavioural results and observations from families regarding the child's responses to sound. If reliable behavioural hearing thresholds cannot be obtained because of significant developmental delay, hearing aid fitting can begin based on parental concern regarding hearing loss and behavioural observation audiometry (unaided and aided) in the test situation.

Fitting hearing aids to children with AN/AD should be based on a prescriptive method specifically developed for infants and young children[16] (*e.g.* DSL i/o). The behavioural thresholds (not ABR/electrophysiological thresholds) should be used to establish amplification targets.

If a prescriptive method cannot be followed (based on lack of reliable behavioural thresholds), a conservative approach should be adopted beginning with a low gain aid and increasing the gain gradually if no responses from the child are observed.

Hearing aid benefit should be determined. Benefit is not determined based only on aided detection thresholds, but on the development of speech perception skills (see below). Monitoring the child's hearing aid fitting is important - refer to the appropriate MCHAS Guidelines[16].

Within reason, DSP hearing instruments that incorporate WDRC, AGC-I and output compression limiting should be used, in keeping with MCHAS protocols[16]

Recent studies indicate that the late cortical evoked potentials may help to differentiate those who are able to use hearing aids effectively to understand speech[17].

b) FM Systems

FM systems (with or without personal hearing aids) have been found to be beneficial for children with AN/AD[18]. A trial with an FM system should be undertaken as part of the hearing aid fitting process, particularly when the child is involved in a day care or educational situation in which poor acoustic conditions restrict access to spoken language.

c) Cochlear Implants

The literature is showing increasing numbers of children with AN/AD who benefit from cochlear implants[5,13,19], and this option should be considered when children are not making progress with hearing aids (*i.e.* they show no or very limited speech discrimination abilities). A trial of conventional amplification is important prior to cochlear implantation (this is an area where we need more research). Behavioural thresholds are not a good guide to candidacy.

Cochlear implant programmes in the UK will need to consider their candidacy criteria and their approach to cases referred for AN/AD based on the accumulating evidence from around the world.

d) Modes of Communication

This should be determined by the needs and desires of the family, taking into account the observed progress of the child. Evidence suggests communication approaches that use visual input such as total communication, signed English or cued speech are effective options for children with AN/AD. In infants and children who appear to make use of auditory information, cued speech[20] may be a particularly useful approach as both speech-reading and auditory cues are available simultaneously for the understanding of spoken English. On the other hand, an auditory-only approach (such as auditory-verbal therapy) is very unlikely to be successful and cannot be recommended unless the child has had a cochlear implant.

Regardless of communication method, it is important that parents become proficient in the method and use it regularly in the home.

3.5 Medical Referral

Ongoing Medical/Neurological Assessment is important.

As with any other child with a hearing problem, ENT involvement may be required to manage any conductive element identified.

AN/AD can be associated with other peripheral neuropathies, including some neuro-degenerative diseases. Now we are screening hearing neonatally, in such cases the AN/AD is often the first neuropathy to become apparent. It is therefore recommended that all children who are diagnosed with AN/AD are referred to a paediatric neurologist.

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APPENDIX 1 TIMELINE / CARE PATHWAY

APPENDIX 2 TESTING FOR COCHLEAR MICROPHONICS (CM)