

Guidelines for Aetiological Investigation into Auditory Neuropathy Spectrum Disorder in Children and Young Adults

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Background:

Definition of ANSD

Auditory neuropathy spectrum disorder (ANSD) describes a condition in which a patient’s outer hair cell function, as demonstrated by otoacoustic emissions (OAE) and/or cochlear microphonic (CM), are (or were at one time) present, and auditory brainstem responses (ABR) are abnormal or absent ^{1,2}.

ANSD is an electrophysiological finding, based on a pattern of results, which is due to a range of different pathologies affecting the auditory pathway, from the inner hair cell to the brain stem, leading to problems in the propagation and conduction of the auditory signal. Although ANSD is generally accepted as a term for use in neonates and young children, when the true nature of the problem is unknown, the term auditory neuropathy is more correct for late onset pathology of this type. It should be noted that some authorities favour the terms auditory neuropathy and auditory dysynchrony³ where the underlying defect is understood. In addition, cochlear nerve hypoplasia or aplasia (cochlear nerve deficiency) has a different clinical picture but presents initially as ANSD. These guidelines have considered evidence which includes all terms but use ANSD to cover the range of pathologies presenting in this way.

Age of Identification

ANSD can present at any age. Where ANSD is congenital or acquired during the perinatal period the pathology may be identified as a consequence of the newborn hearing screen, particularly if the child has been admitted to neonatal intensive care. It is, however, a pathology that can be missed in well babies when automated ABR is not used routinely as part of the screen in these babies as in the UK. ANSD may therefore present during early childhood ⁴ with delayed speech and language development, hearing problems or educational difficulties. Some genetic causes of ANSD can lead to the late onset of auditory symptoms ^{5,6,7,8,9} and acquired causes can present throughout life ^{10,11,12}.

These guidelines can be applied to adults depending on their clinical presentation, but detailed investigation of adult auditory neuropathy is outside the scope of this guideline and later acquired causes have not been considered.

Reasons for Investigation

There are several reasons why it is important to establish the cause of ANSD:

1. To answer the questions parents or patients may have, 'why has my child got ANSD?' 'Why have I got ANSD?'
2. To identify syndromes associated with ANSD in order to provide information of linked pathology.
3. To understand the prognosis of the hearing difficulties and the potential for recovery or progression.
4. To investigate whether there are associated conditions which may affect the general prognosis or require specific treatment.
5. To provide information for optimal rehabilitation e.g. cochlear implant ^{8,13,14,15,16}.
6. To identify genetic causes and to inform genetic counselling e.g. recurrence of ANSD in a future child.
7. To counsel patients and their families on the effects of balance disorder if the vestibular system is involved.
8. To provide information for epidemiological studies to better understand the pathology.

Aim and Scope:

The aim of these guidelines is to establish an evidence-based approach to the investigation of the cause of ANSD. These guidelines were produced in line with the procedure detailed in the BAAP manual for producing guidelines.

These guidelines are for use in the United Kingdom but could be applied world wide depending on local clinical expertise, test facilities and resources. The intended users of these guidelines are health practitioners with a special interest in Audiovestibular Medicine.

The guidelines:

- Provide up to date advice on effective clinical practice
- Support staff in improving and benchmarking Audiovestibular Medicine services
- Identify audit measures for performance and review
- Promote patient safety and implementation of clinical governance

These guidelines are evidence-based and link their concluding recommendations to the evidence identified through a literature search. They are not intended to restrict clinical freedom, but practitioners are expected to use the recommendations as a basis for their practice. Areas lacking in evidence may form the basis for future research.

Timing of investigations:

The timing of tests will depend on the clinical presentation, time window for the specific test, the individual or family's readiness to proceed and, in children, how well the child can cooperate with the tests. The process of aetiological investigations is an ongoing one and it is important to revisit this periodically because:

- New medical information and new tests may become available
- New symptoms may develop e.g. visual difficulties
- New information relating to family history may become available
- Individuals or parents may request investigation

Following the identification of ANSD after the newborn hearing screen: all children identified as having the findings of ANSD on initial ABR testing, which usually occurs at about 4 weeks corrected age, should be seen for aetiological investigation as soon as possible. While ABR testing may occur throughout the first year or two of life to determine whether ANSD is a permanent finding, it would be inappropriate to delay medical investigation because aetiological test results may indicate optimal audiological management. In addition, some tests are time limited.

Who can undertake aetiological investigations?

A medical practitioner with the appropriate knowledge and skills can undertake aetiological investigations. People should be referred appropriately when this service is not available locally. It is the responsibility of the doctor providing the aetiology service to provide accurate and unbiased information to individuals and parents (or carers), if applicable, about the investigations (pros/cons, outcomes and details of procedure etc). This should be done as soon as possible after ANSD is confirmed so that they can make a well-informed decision to have or not to have each investigation.

Subjects

These guidelines apply to children and adolescents with auditory neuropathy spectrum disorder in one or both ears. Acquired causes likely to be encountered during childhood and adolescence are covered as well as genetic causes. They also include 'delayed auditory maturation' or 'transient ANSD' as the initial clinical picture is that of ANSD with subsequent recovery¹⁷.

Search Methodology:

The literature search covered databases including Pubmed, Medline, Embase, AMED, BNI, CINAHL, HMIC, PsychINFO and Cochrane Library Database. The keywords detailed in Appendix 1 were used. The search was carried out by all the members of the guideline group [A-K]. All relevant articles including randomised control trials, systematic reviews, meta-analyses, observational studies, case reports and expert opinion were reviewed. Some

review articles were referenced but not included to support recommendations in the guidelines. Case reports and series were included as there was paucity of references with level of evidence 1 and 2. Articles not available in English or only available in abstract forms were excluded. Animal studies were also excluded. Relevant guidelines and standards from other national and international organisations were included in this review. The literature search covered a period up to 27/02/2018. The abstracts of the list of articles obtained following the literature review were scanned to produce a list of articles relevant to the guideline. This was done by all members of the guideline group [A-K]. Full texts of all these relevant articles were obtained with the help of librarians. Members of the guideline group [A-K] reviewed the full texts of the articles. The articles relevant to the guideline were graded for evidence level by members of the guideline group [A-C].

Keywords:

The keywords were guided by questions using the PICOT format:

- Population to which the question applies
- Intervention (e.g. or diagnostic test, exposure etc.) being considered in relation to this population
- Comparison(s) to be made between those receiving the intervention and those who do not receive the intervention
- Outcome(s) i.e. any effect caused by the intervention
- Timeframe

The keywords used are detailed in the Appendix 1:

Grade of evidence and recommendation

The evidence from the full text articles was graded according to the Scottish Intercollegiate Guideline Network [SIGN] grading system as follows:

Level of evidence	Definition
1++	High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+	Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1-	Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++	High quality systematic reviews of case control or cohort studies High quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well-conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
2-	Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal

3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

The strength of recommendations in this guideline is based on the SIGN grading of evidence as follows:

Recommendation A - This recommendation is based on evidence rated as 1++ or 1+ directly applicable to the target population and demonstrating overall consistency of results.

Recommendation B - This recommendation is based on evidence rated as 2++ or based on extrapolated evidence from studies rated as 1++ or 1+ directly applicable to the target population and demonstrating overall consistency of results.

Recommendation C - This recommendation is based on evidence rated as 2+ or based on extrapolated evidence from studies rated as 2++ directly applicable to the target population and demonstrating overall consistency of results.

Recommendation D - This recommendation is based on evidence rated as level 3 or 4 or based on extrapolated evidence from studies rated as 2+.

Guidelines for good practice

Aetiological investigations are based on the available evidence, expected yield and take into account the causes of ANSD in children and adolescents. ANSD presents throughout life and it is likely that different causes present in neonates and young children from those presenting later on in childhood or adolescence. The clinician needs to consider this when planning investigations.

The results of investigation should be interpreted in a clinical context. The clinician should be aware that it may sometimes be difficult to pinpoint the aetiology of ANSD despite investigations.

INVESTIGATIONS

1] Clinical History: Recommendation C ^{15,18,19,20,21,22,23}

It is important to recognise that several different pathologies can present as ANSD and it is appropriate to get a detailed history and perform a detailed examination on every child in order to identify the underlying aetiology. For details of the recommended history and examination please refer to BAAP Guidelines on PCHI, [severe to profound](#) and [mild to moderate](#) ^{24,25}.

There is a very clear association with neonatal hyperbilirubinaemia and a traumatic neonatal period giving rise to both permanent ANSD and delayed maturation or transient ANSD in neonates ^{18,19,20,22,23,26}.

For children with ANSD the following are of particular importance:

- History of presenting complaint
 - Onset, duration and progression of symptoms of hearing loss and of associated problems
 - Change in hearing with temperature fluctuation ^{27,28}.
- Antenatal History
- Birth history
- Postnatal history
 - Prematurity, jaundice, birth weight, hypoxia, ventilation, sepsis, NICU stay and ototoxic medication
- Developmental milestones
- Family history with three generation family tree
 - Deafness, ANSD or Inherited conditions including optic atrophy or neuropathy such as Hereditary Sensory Motor Neuropathy (Charcot-Marie-Tooth Syndrome)
- Medical history
 - Meningitis/ Infectious illness such as mumps ^{29,30,31}.
 - Encephalitis ⁸
 - Thiamine deficiency ³²
 - Visual problems (direct questioning needed)
 - Sensory/motor neuropathy
 - Balance and coordination
 - Drug/chemical toxicity ^{33,34}
 - Other illnesses

The history and examination are important not only for identifying aetiological factors in hearing loss but also for detection of conditions requiring medical management: e.g. visual or neurological problems. This is to be done with a problem-solving approach rather than as a tick box exercise.

Timing of assessment: When ANSD is confirmed, at first visit.

2) Clinical examination: [Recommendation D] ^{15,26,35,36}

Peripheral neuropathy has been reported as occurring in 40% of patients with ANSD, more commonly in those over 15 years of age ²⁶. Cerebellar signs and optic atrophy are hallmarks of several genetic conditions known to give rise to ANSD.

Examination should be as for any child with PCHI but concentrating on:

- Clinical examination of craniofacial region including head circumference measurement
- Full neurological assessment including gait, tendon reflexes, muscular strength and coordination, cerebellar signs, sensory system examination, as feasible
- Developmental assessment
- Clinical vestibular examination
- Eye examination

- Relevant examination of parents if indicated by history
- If no cause is identified the clinician should be highly suspicious of an evolving syndrome. Repeat neurological examination and eye examination is important.

Timing of assessment: When ANSD is confirmed, at first visit, as soon as opportunity provides. Repeat as indicated by change in symptoms or test findings.

3) Family audiograms: [Recommendation D]

ANSD can be inherited and the pattern of inheritance i.e. recessive, dominant, X-linked or mitochondrial can help with identification of aetiology. Parents and siblings should have their hearing checked. PTA may not be sufficient to exclude ANSD, hence, if there is the slightest suspicion of a hearing difficulty, the parent/sibling should be referred for a full evaluation to include stapedial reflexes and, if these are abnormal, diagnostic ABR.

- Request audiograms of parents and siblings.
- Consider more detailed testing, including ABR, if hearing problems are suspected.

Timing of assessment: Early, before the genetics referral.

4) MRI of Internal Auditory Meati and brain [Recommendation C]

Diagnostic radiological imaging is the highest yielding test for evaluating children with ANSD^{37,38,39,40,41}. The commonest abnormalities include cochlear nerve deficiency, brain abnormalities and prominent temporal horns^{42, 43}. Cochlear nerve deficiency is reported in 28% of ears⁴³ and is more common in unilateral as compared to bilateral ANSD^{38, 42}. It is therefore important to image the cochleo-vestibular nerve and its cochlear branch. Abnormalities have been found on brain imaging in up to 40% of children with ANSD⁴³. There is some evidence to link hydrocephalus to ANSD and this may be identified on MRI^{43, 44}. There have also been individual case reports of space occupying lesions^{45, 46} and of multiple sclerosis in children presenting with ANSD⁴⁷.

- MRI is the preferred investigation for ANSD because the cochlea-vestibular nerve and its cochlear branch can be identified and measured.
- It is important to obtain sagittal oblique views of the IAMs to estimate the size of the cochlear nerve.
- MRI of the brain should be performed in children with ANSD⁴³.

Timing of investigation: Soon after identification, best within 3 months of age in natural sleep if identification follows newborn hearing screen [to avoid the need for sedation].

5) Genetic tests for ANSD: Recommendation D

Several genetic mutations have been reported in children with ANSD^{48, 49}. Inheritance of non-syndromic ANSD has been shown to be dominant^{50, 51, 52, 53} or recessive^{27, 54, 55, 56} and syndromic forms may be dominant^{6, 9, 57, 58}, recessive^{8, 49, 59, 60}, X-linked^{61, 62, 63} or mitochondrial^{49, 64}. With a non-syndromic presentation by far the most papers relate to the gene OTOF^{27, 55, 65, 66, 67}. Several case reports detail other genetic mutations including Pejvakin^{56, 65}, AUNA1^{48, 50}, DIAPH3⁵³ and also Connexin 26^{68, 69}. ANSD associated with peripheral neuropathy, cerebellar features and optic atrophy has been linked with mutations giving rise to Friedreich's Ataxia^{7, 59}, Charcot-Marie-Tooth Syndrome/Hereditary Sensory Motor Neuropathy^{6, 70}, Leber's Optic Atrophy⁶⁴, Kjer's disease⁵⁸ and Mohr-Tranebjærg Syndrome⁶¹. Brown-Vialetto-van Laere Syndrome is recognised as a cause of ANSD⁷¹ and, in addition, ANSD has been reported in Gaucher Disease⁷², Refsum Disease⁷³, Niemann-Pick Disease⁷⁴ and in hypomyelinating leukodystrophy⁷⁵. CAPOS syndrome has recently been described as having progressive ANSD and episodes of aseptic encephalitis⁸.

The choice of genetic testing will depend on the clinical picture and family history.

- Informed consent should be taken from parents prior to genetic testing. Parents should be informed that DNA is stored in the laboratory after testing and that genetic testing can take a long time. Permission should be taken to share results with other family members/professionals (see [guidelines for consent for genetic testing](#)⁷⁶).
- Consider genetic testing for otoferlin, pejvakin etc. if non-syndromic and especially if the behavioural thresholds indicate a severe to profound hearing loss. The presence of a genetic cause of ANSD will indicate the need for early trial of amplification and cochlear implant.
- Consider blood test for GJB2 [Connexin 26] mutations in cases where the aetiology has not been determined. Connexin 26 mutations have been described in cases of ANSD^{68, 69}.

More widespread genetic testing for deafness will become available with the advent of Next Generation (Massively Parallel) sequencing where large numbers of genes can be sequenced rapidly and cost-effectively. In the case of nonsyndromic deafness many genes can be tested simultaneously, without regard to phenotype but this will make interpretation of multiple novel or rare genetic variants more difficult initially. Guidelines for further genetic testing are likely to evolve over the next few years.

Timing of investigation: For neonates - at identification of bilateral ANSD if significant perinatal trauma or hyperbilirubinaemia have been excluded. For all older children- at presentation of bilateral ANSD.

6) Ophthalmic assessment: [Recommendation D]

Examination of the eyes is important in children with ANSD because the visual pathway can also be affected in a number of syndromes involving auditory neuropathy, specifically in some genetic disorders e.g. OPA1^{13, 57}, Leber's Optic Neuropathy⁶⁴, Friedreich's Ataxia⁵⁹, Kjer Disease⁵⁸, Mohr-Tranebjærg Syndrome⁶¹, CAPOS Syndrome⁸. Charcot-Marie-Tooth

Disease and other Hereditary Sensory and Motor Neuropathies^{6,70}. There is also a case report that links delayed visual maturation with delayed auditory maturation⁷⁷.

The strength of evidence is sufficient to suggest detailed ophthalmological examination by an ophthalmologist with consideration given to electrophysiological testing (VEP) when indicated or if the cause of ANSD remains undetermined.

In addition, these children, in common with others with PCHI, can have ophthalmic abnormalities which can remain undetected and impact on the child's communication including speech reading, uptake of sign language and reading of facial expressions. The incidence of ophthalmic disorders in children with PCHI is estimated to be between 20 and 60%.⁷⁹.

- The child should be referred for a full ophthalmic assessment by an ophthalmologist following the identification of ANSD and at any time if parents or the education service have concerns.
- The ophthalmic examination should include formal testing and recording of:
 - **Fundoscopy**
 - **Visual acuity**
 - **Visual field assessment**
 - **Assessment of colour vision** (blue/yellow and red/green)
 - Functional assessment of vision
 - Refraction
 - Assessment of ocular alignment and eye movements
 - Assessment of binocular vision
 - depending on the feasibility and the age of the child.

The highlighted tests are of particular importance in ANSD as early indicators of optic atrophy.
- VEP (visual evoked potentials) should be performed if there is a possibility of optic atrophy.
- If the cause of ANSD is unknown: further assessment including VEP should be performed:
 - Prior to referral to a geneticist or neurologist
 - At referral for cochlear implantation and other complex interventions

Timing of investigation: Soon after identification of ANSD, repeated as needed.

7) Vestibular investigations: [Recommendation D]

Research has shown vestibular abnormalities in subjects with ANSD. Abnormal vestibular function has been reported in 43% of children with ANSD⁷⁹ using a variety of tests including rotational chair testing. There are also reports of abnormal VEMPs⁸⁰ and bithermal calorics⁸¹. The commonest vestibular test abnormality to be reported has been in VEMPs with 80 to

100% of adolescents and adults with ANSD showing abnormalities^{82, 83}. It is recommended that vestibular function is checked, especially in subjects with peripheral neuropathies^{84, 85}.

The temporal profile and progression of vestibular function in ANSD is not known, but reports in adults have indicated the need for monitoring⁸⁶.

- All children with ANSD should have a clinical vestibular examination as soon as possible after identification. This should be done within the first few weeks of life when ANSD is identified early.
- Consider further vestibular investigations if ANSD is permanent especially if imbalance or delayed motor milestones are part of the picture or if cochlear implant is considered.

Timing of investigation: Vestibular examination - when ANSD is confirmed, at first visit, as soon as opportunity provides. Repeat as indicated.

Formal vestibular investigations – as indicated clinically.

8) Chromosomal studies/CGH microarray:

Indicated if:

- History of developmental delay
- Dysmorphic features

Chromosome analysis has been replaced by more detailed CGH microarray which itself may be replaced in time by massive parallel sequencing techniques which can detect copy number changes. Laboratories may request parental bloods in order to fully interpret findings.

9) Serology: [Recommendation D]

For congenital infection

There is no evidence linking **congenital infection** to ANSD but testing should be considered in at risk children as a matter of course. In particular it is important to look for evidence of CMV infection as there is a limited window of opportunity for identification and treatment if indicated.

Mothers may be screened for these infections in pregnancy and some babies may have testing in NICU. As many of these babies can be asymptomatic at birth, if the testing or immune status of the mother is unknown it is best to investigate the neonate. These tests may also be done on maternal stored (booking) serum if available.

For acquired infection:

- Mumps: there are isolated case reports of mumps infection giving rise to acquired unilateral ANSD²⁹: serology may aid aetiological diagnosis although the MMR vaccine, if it has been given, may mean that serology is unhelpful.

Onward Referral

10) Referral to Clinical Geneticist:

Consider referral of children with ANSD for a genetic opinion:

- In those without a history of perinatal trauma, according to your local pathway, if, after completion of investigations, no cause has been identified
- On parental request
- For an interpretation of genetic mutation testing, if required
- If there is a family history of ANSD and no cause has been identified

11) Referral for a comprehensive paediatric neurological evaluation

ANSD can occur as part of a neurological condition therefore referral should be made:

- If there is any suggestion of a peripheral sensory or motor neuropathy or other neurological anomalies.

Testing children with conditions associated with ANSD

There are some specific neurological conditions where there is a high risk of a child or young adult having ANSD as part of the clinical picture although the individual may have normal pure tone thresholds on testing. A high prevalence of poor speech perception, particularly in the presence of background noise, has been reported in CMT, Friedreich's ataxia and Leber's optic atrophy^{6,7,60,65}, and in some cases ABRs are also abnormal. It is important to be aware of the possibility of ANSD in these individuals and plan the history and investigations accordingly.

References:

1. Berlin, C. I. *et al.* (2010) 'Multi-site diagnosis and management of 260 patients with auditory neuropathy/dys-synchrony (auditory neuropathy spectrum disorder).', *International journal of audiology*, 49(1), pp. 30–43. doi: 10.3109/14992020903160892.
2. Guidelines for identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder Guidelines Development Conference at NHS 2008 Como, Italy. <https://www.childrenscolorado.org/globalassets/departments/ear-nose-throat/ansd-monograph.pdf>
3. Starr and Rance (2015). 'Auditory Neuropathy' *Handbook of Clinical Neurology*, Vol. 129 (3rd series) pp 495-508, The Human Auditory System, G.G. Celesia and G. Hickok, Editors, Pub. Elsevier
4. Boudewyns, A. *et al.* (2016) 'Auditory neuropathy spectrum disorder (ANS) in referrals from neonatal hearing screening at a well-baby clinic', *European Journal of Pediatrics*, 175(7), pp. 993–1000. doi: 10.1007/s00431-016-2735-5.
5. Kaga, K. (2016) 'Auditory nerve disease and auditory neuropathy spectrum disorders', *Auris Nasus Larynx*, 43: pp 10–20. doi: 10.1016/j.anl.2015.06.008.
6. Rance, G. *et al.* (2012) 'Auditory function in children with Charcot-Marie-Tooth disease', *Brain*, 135(5), pp. 1412–1422. doi: 10.1093/brain/aws085.
7. Rance, G. *et al.* (2008) 'Speech perception ability in individuals with Friedreich ataxia', *Brain*, 131(8), pp. 2002–2012. doi: 10.1093/brain/awn104.
8. Tranebjærg *et al.* (2018). 'The CAPOS mutation in ATP1A3 alters Na/K-ATPase function and results in auditory neuropathy which has implications for management.' *Human Genetics*. 137(2):111-127. doi: 10.1007/s00439-017-1862-z.
9. Leruez, S. *et al.* (2013) 'Sensorineural hearing loss in OPA1-linked disorders', *Brain: A Journal of Neurology*, 136(7), pp. 1–6. doi: 10.1093/brain/aws340.
10. Rance, G. *et al.* (2014) 'Auditory neuropathy in individuals with Type 1 diabetes', *Journal of Neurology*, 261(8), pp. 1531–1536. doi: 10.1007/s00415-014-7371-2.
11. Norrix, L. W. and Velenovsky, D. S. (2014) 'Auditory Neuropathy Spectrum Disorder: A Review', *Journal of Speech Language and Hearing Research*, 57(4), p. 1564-76. doi: 10.1044/2014_JSLHR-H-13-0213.
12. Prabhu, P., Avilala, V. K. Y. and Manjula, P. (2012) 'Predisposing Factors in Individuals with Late-Onset Auditory Dys-Synchrony', *Asia Pacific Journal of Speech, Language and Hearing*, 15(1), pp. 41–50. doi: 10.1179/136132812805253758.

13. Santarelli (2015) 'OPA-1 related auditory neuropathy: site of lesion and outcome of cochlear implantation', *Brain*, 138(3), pp. 563–76. doi:10.1093/brain/awu378.
14. Han et al (2017) *ATP1A3* mutations can cause progressive auditory neuropathy: a new gene of auditory synaptopathy. *Scientific Reports* 7: 16504 | doi:10.1038/s41598-017-16676-9.
15. Vlastarakos, P. V. et al. (2008) 'Auditory neuropathy: Endocochlear lesion or temporal processing impairment? Implications for diagnosis and management', *International Journal of Pediatric Otorhinolaryngology*, 72(8), pp. 1135–1150. doi: 10.1016/j.ijporl.2008.04.004.
16. Wu, C., Hsu, C. and Huang, F. (2017) 'Timing of cochlear implantation in auditory neuropathy with OTOF mutations: our experience with 10 patients', *Clinical Otolaryngology* doi: 10.1111/coa.12949
17. Abdollahi, F. Z., Ahmadi, T., Manchaiah, V., Lotfi, Y. (2016) 'Auditory Brainstem Response Improvements in Hyperbilirubinemic Infants', *J Audiol Otol*, 20(1), pp. 13–16. doi: 10.7874/jao.2016.20.1.13.
18. Amin, S. B. et al. (2017) 'Auditory toxicity in late preterm and term neonates with severe jaundice', *Developmental Medicine and Child Neurology*, 59(3), pp. 297–303. doi: 10.1111/dmcn.13284.
19. Xoinis, K. et al. (2007) 'Extremely low birth weight infants are at high risk for auditory neuropathy.', *Journal of perinatology*, 27(11), pp. 718–23. doi: 10.1038/sj.jp.7211803.
20. Bielecki, I., Horbulewicz, A. and Wolan, T. (2012) 'Prevalence and risk factors for Auditory Neuropathy Spectrum Disorder in a screened newborn population at risk for hearing loss', *International Journal of Pediatric Otorhinolaryngology*, 76(11), pp. 1668–1670. doi: 10.1016/j.ijporl.2012.08.001.
21. Dowley, A. C. et al. (2009) 'Auditory neuropathy: Unexpectedly common in a screened newborn population', *Developmental Medicine and Child Neurology*, 51(8), pp. 642–646. doi: 10.1111/j.1469-8749.2009.03298.x.
22. Uhler, K. et al. (2012) 'A tutorial on auditory neuropathy/dyssynchrony for the speech-language pathologist and audiologist', *Seminars in Speech and Language*, 33(4), pp. 354–366. doi: 10.1055/s-0032-1326917.
23. Coenraad, S. et al. (2011) 'Risk factors for auditory neuropathy spectrum disorder in NICU infants compared to normal-hearing NICU controls', *Laryngoscope*, 121(4), pp. 852–855. doi: 10.1002/lary.21430.
24. BAAP (2015) [Guidelines for aetiological investigation into severe to profound bilateral childhood hearing impairment](http://www.baap.org.uk) www.baap.org.uk .

25. BAAP (2015) [Guidelines for aetiological investigation into mild to moderate bilateral permanent childhood hearing impairment](http://www.baap.org.uk) www.baap.org.uk
26. Starr, A., Sininger, Y. S. and Pratt, H. (2000) 'The varieties of auditory neuropathy.', *Journal of basic and clinical physiology and pharmacology*, 11(3), pp. 215–30. doi: 10.1515/JBCPP.2000.11.3.215.
27. Varga, R. *et al.* (2006) 'OTOF mutations revealed by genetic analysis of hearing loss families including a potential temperature sensitive auditory neuropathy allele.', *Journal of medical genetics*, 43(August), pp. 576–581. doi: 10.1136/jmg.2005.038612.
28. Zhang, Q. *et al.* (2016) 'Temperature sensitive auditory neuropathy', *Hearing Research*, 335, pp. 53–63. doi: 10.1016/j.heares.2016.01.008.
29. Salvinelli, F. *et al.* (2004) 'Preserved otoacoustic emissions in postparotitis profound unilateral hearing loss: a case report.', *The Annals of otology, rhinology, and laryngology*, 113(11), pp. 887–890. doi: 10.1177/000348940411301107
30. Prabhu, P. (2016) 'Acquired auditory neuropathy spectrum disorder after an attack of chikungunya: case study', *European Archives of Oto-Rhino-Laryngology*, 273(1), pp. 257–261. doi: 10.1007/s00405-015-3578-9.
31. Celis-Aguilar, E., Macias-Valle, L. and Coutinho-De Toledo, H. (2012) 'Auditory Neuropathy Secondary to Cryptococcal Central Nervous System Infection in 2 Immunocompromised Patients', *Otolaryngology-Head and Neck Surgery*, 147(3), pp. 597–598. doi: 10.1177/0194599812442488.
32. Attias, J, *et al.* (2012). 'Auditory system dysfunction due to infantile thiamine deficiency: Long-term auditory sequelae', *Audiology and Neurotology*, 17(5), pp. 309-320. doi: 10.1159/000339356.
33. Siram R; Botta R; Kashikunte C; Pal PK; Yadav R (2017) Chronic encephalopathy with ataxia, myoclonus, and auditory neuropathy: A case of bismuth poisoning *Neurology India*. 65(1):186-187. doi: 10.4103/0028-3886.198185
34. Draper, T. H. J. and Bamiou, D.-E. (2009) 'Auditory neuropathy in a patient exposed to xylene: case report.', *The Journal of laryngology and otology*, 123(4), pp. 462–465. doi: 10.1017/S0022215108002399.
35. Lepcha, A. *et al.* (2015) 'Neurological associations in auditory neuropathy spectrum disorder: Results from a tertiary hospital in South India.', *Annals of Indian Academy of Neurology*, 18(2), pp. 171–180. doi: 10.4103/0972-2327.150578.
36. Inscoe, J. and Bones, C. (2016) 'Additional difficulties associated with aetiologies of deafness: outcomes from a parent questionnaire of 540 children using cochlear

- implants', *Cochlear Implants International*, 171(1), pp. 21–30. doi: 10.1179/1754762815Y.0000000017.
37. Levi, J. *et al.* (2013) 'Clinical characteristics of children with cochlear nerve dysplasias', *Laryngoscope.*, 123(3), pp. 752–756. doi: 10.1002/lary.23636.
38. Kim, S. H. *et al.* (2016) 'Diverse etiologies manifesting auditory neuropathy characteristics from infants with profound hearing loss and clinical implications', *International Journal of Pediatric Otorhinolaryngology*, 86, pp. 63–67. doi: 10.1016/j.ijporl.2016.04.013.
39. Peng, L. *et al.* (2016) 'Evaluation of cochlear nerve diameter and cross-sectional area in ANSD patients by 3.0-Tesla MRI.', *Acta oto-laryngologica*, 136(8), pp. 792–9. doi: 10.3109/00016489.2016.1159329.
40. Huang, B. Y. *et al.* (2010) 'Brain stem and inner ear abnormalities in children with auditory neuropathy spectrum disorder and cochlear nerve deficiency', *American Journal of Neuroradiology*, 31(10), pp. 1972–1979. doi: 10.3174/1jnr.A2178
41. Mohammadi, A., Walker, P. and Gardner-Berry, K. (2015) 'Unilateral auditory neuropathy spectrum disorder: retrocochlear lesion in disguise?', *J Laryngol Otol*, 129 Suppl (November 2014), pp. S38-44. doi: 10.1017/s0022215114002734.
42. Liu, C. *et al.* (2012) 'Unilateral auditory neuropathy caused by cochlear nerve deficiency.', *International journal of otolaryngology*, 2012, Article ID 914986. doi:10.1155/2012/914986.
43. Roche, J. P. *et al.* (2010) 'Imaging Characteristics of Children with Auditory Neuropathy Spectrum Disorder', *Otology & Neurotology*, 31(5), pp. 780–788. doi: 10.1097/MAO.0b013e3181d8d528.
44. Berg, A. L. *et al.* (2005) 'Newborn Hearing Screening in the NICU: Profile of Failed Auditory Brainstem Response/Passed Otoacoustic Emission', *Pediatrics*, 116(4), pp. 933–938. doi: 10.1542/peds.2004-2806. doi: 10.1542/peds.2004-2806.
45. Boudewyns, A. N. *et al.* (2008) 'Case report: "Auditory neuropathy" in a newborn caused by a cerebellopontine angle arachnoid cyst', *International Journal of Pediatric Otorhinolaryngology*, 72(6), pp. 905–909. doi: 10.1016/j.ijporl.2008.02.003.
46. Kottapalli, S. and Madden, C. (2013) 'Acquired auditory neuropathy spectrum disorder in a child with a brain tumour', *Hearing, Balance and Communication*, 11(1), pp. 47–51. doi: 10.3109/21695717.2012.756217.
47. Mallur, P. S. and Lalwani, A. K. (2007) 'Fluctuating Corticosteroid-Responsive Auditory Neuropathy/Dyssynchrony Is Suggestive of Central Nervous System Pathology', *Otology & Neurotology*, 28(8), pp. 1002–1004. doi: 10.1097/MAO.0b013e318157f7f5.

48. Manchaiah, V. K. C. *et al.* (2011) 'The genetic basis of auditory neuropathy spectrum disorder (ANSO)', *International Journal of Pediatric Otorhinolaryngology*, 75, pp. 151–158. doi: 10.1016/j.ijporl.2010.11.023.
49. Cacace, A. T. and Pinheiro, J. M. B. (2011) 'The mitochondrial connection in auditory neuropathy', *Audiology and Neurotology*, 16, pp. 398–413. doi: 10.1159/000323276.
50. Kim, T. B. *et al.* (2004) 'A gene responsible for autosomal dominant auditory neuropathy (AUNA1) maps to 13q14-21.', *Journal of medical genetics*, 41(11), pp. 872–6. doi: 10.1136/jmg.2004.020628.
51. Xing, G. *et al.* (2007) 'Clinical and genetic features in a Chinese pedigree with autosomal dominant auditory neuropathy', *ORL*, 69(2), pp. 131–136. doi: 10.1159/000097981.
52. Starr, A. *et al.* (2004) 'a Dominantly Inherited Progressive Deafness Affecting Distal Auditory Nerve and Hair Cells', *JARO*, 5, pp. 411-426. doi: 10.1007/s10162-004-5014-5.
53. Schoen, C. J. *et al.* (2010) 'Increased activity of Diaphanous homolog 3 (DIAPH3)/diaphanous causes hearing defects in humans with auditory neuropathy and in *Drosophila*.', *Proceedings of the National Academy of Sciences of the United States of America*, 107(30), pp. 13396–401. doi: 10.1073/pnas.1003027107.
54. Varga, R. *et al.* (2003) Nonsyndromic recessive auditory neuropathy is the result of mutations in the otoferlin (OTOF) gene. *J. Med Genet* 40: pp. 45-50. doi: 10.1136/jmg.40.1.45
55. Rodríguez-Ballesteros, M. *et al.* (2008) 'A multicenter study on the prevalence and spectrum of mutations in the otoferlin gene (OTOF) in subjects with nonsyndromic hearing impairment and auditory neuropathy', *Human Mutation*, 29(6), pp. 823–831. doi: 10.1002/humu.20708.
56. Delmaghani, S. *et al.* (2006) 'Mutations in the gene encoding pejvakin, a newly identified protein of the afferent auditory pathway, cause DFNB59 auditory neuropathy.', *Nature genetics*, 38(7), pp. 770–8. doi: 10.1038/ng1829.
57. Huang, T., Santarelli, R. and Starr, A. (2009) 'Mutation of OPA1 gene causes deafness by affecting function of auditory nerve terminals', *Brain Research*, 1300, pp. 97–104. doi: 10.1016/j.brainres.2009.08.083.
58. Haaksma-Schaafsma, S. C., van Dijk, P. and Dikkers, F. G. (2012) 'Auditory and optic neuropathy in Kjer's disease: case report', *The Journal of Laryngology & Otology*, 126(3), pp. 309–312. doi: 10.1017/S0022215111002696.
59. Rance, G. *et al.* (2010) 'Auditory perception in individuals with Friedreich's ataxia', *Audiology and Neurotology*, 15(4), pp. 229–240. doi: 10.1159/000255341.

60. Alodaib, A. *et al.* (2016) 'Whole-exome sequencing identifies novel variants in PNPT1 causing oxidative phosphorylation defects and severe multisystem disease', *European Journal of Human Genetics*, 25(1), pp. 79–84. doi: 10.1038/ejhg.2016.128.
61. Tranebjærg, L., Jensen, P. K. and van Ghelue, M. (2000) 'X-linked recessive deafness-dystonia syndrome (Mohr-Tranebjærg syndrome).', *Advances in oto-rhino-laryngology*, 56, pp. 176–180.
62. Zong, L. *et al.* (2015) 'Mutations in apoptosis-inducing factor cause X-linked recessive auditory neuropathy spectrum disorder.', *Journal of medical genetics*, 52(8), pp. 523–31. doi: 10.1136/jmedgenet-2014-102961.
63. Wang, Q. J. *et al.* (2006) 'AUNX1, a novel locus responsible for X linked recessive auditory and peripheral neuropathy, maps to Xq23-27.3.', *Journal of medical genetics*, 43(7), p. e33. doi: 10.1136/jmg.2005.037929.
64. Rance, G. *et al.* (2012) 'Auditory function in individuals within Leber's hereditary optic neuropathy pedigrees', *Journal of Neurology*, 259(3), pp. 542–550. doi: 10.1007/s00415-011-6230-7\rE-pub 2011 Sep 2.
65. Wang, J. *et al.* (2011) 'Variants of OTOF and PJKV genes in Chinese patients with auditory neuropathy spectrum disorder', *PLoS ONE*, 6(9), p. e24000. doi: 10.1371/journal.pone.0024000.
66. Iwasa, Y. *et al.* (2013) 'OTOF mutation screening in Japanese severe to profound recessive hearing loss patients.', *BMC medical genetics*, 14(1), p. 95. doi: 10.1186/1471-2350-14-95.
67. Matsunaga, T. *et al.* (2012) 'A prevalent founder mutation and genotype – phenotype correlations of OTOF in Japanese patients with auditory neuropathy', *Clin Genetics*, 82(5), pp. 425–432. doi: 10.1111/j.1399-0004.2012.01897.x.
68. Cheng, X. *et al.* (2005) 'Connexin 26 variants and auditory neuropathy/dys-synchrony among children in schools for the deaf', *American Journal of Medical Genetics*, 139 A (1), pp. 13–18. doi: 10.1002/ajmg.a.30929.
69. de Carvalho, G. M. *et al.* (2016) 'Relationship between patients with clinical auditory neuropathy spectrum disorder and mutations in Gjb2 gene', *Open Neurology Journal*, 10, pp. 127–135. doi: 10.2174/1874205X01610010127.
70. Butinar, D., Starr, A and Vatovec, J. (2000) 'Brainstem auditory evoked potentials and cochlear microphonics in the HMSN family with auditory neuropathy.', *Pflugers Archiv : European journal of physiology*, 439, pp. R204-R205. doi: 10.1007/s004240000146.

71. Menezes, M. P. *et al.* (2016) 'Auditory neuropathy in Brown-Vialetto-Van Laere syndrome due to riboflavin transporter RFVT2 deficiency', *Developmental Medicine and Child Neurology*, 58(8), pp. 848–854. doi: 10.1111/dmcn.13084.
72. Campbell, P. E. *et al.* (2003) 'A model of neuronopathic Gaucher disease', *Journal of Inherited Metabolic Disease*, 26(7), pp. 629–639. doi: 10.1023/B:BOLI.0000005619.14180.5c.
73. Oysu, C. *et al.* (2001) 'The site of the hearing loss in Refsum's disease', *International Journal of Pediatric Otorhinolaryngology*, 61(2), pp. 129–134. doi: 10.1016/S0165-5876(01)00559-6.
74. King, K. A. *et al.* (2010) 'Auditory phenotype of Niemann-Pick disease, type C1.', *Ear and hearing*, 35(1), pp. 110–7. doi: 10.1097/AUD.0b013e3182a362b8.
75. Yuvaraj, P. *et al.* (2015) 'Auditory neuropathy spectrum disorder in hypomyelinating leukodystrophy--A case study', *International Journal of Pediatric Otorhinolaryngology*, 79(12), pp. 2479–83. Doi: 10.1015/j.ijporl.2015.10.053.
76. Consent and confidentiality in clinical genetic practice: Guidance on genetic testing and sharing genetic information A report of the Joint Committee on Clinical Genetics http://www.bsgm.org.uk/media/678746/consent_and_confidentiality_2011.pdf
77. Aldosari, M., Mabie, A. and Husain, A. M. (2003) 'Delayed visual maturation associated with auditory neuropathy/dyssynchrony', *J Child Neurol*, 18(5), pp. 358–361. doi: 10.1177/08830738030180052001.
78. Quality Standards in Vision Care for Deaf Children and Young People. Guidelines for professionals. [NDCS and SENSE 2009]. [Quality Standards Document] http://www.ndcs.org.uk/family_support/childhood_deafness/vision_care_for_your_deaf_child/index.html
79. Nash, R. *et al.* (2014) 'Vestibular function in children with auditory neuropathy spectrum disorder', *International Journal of Pediatric Otorhinolaryngology*, 78(8), pp. 1269–1273. doi: 10.1016/j.ijporl.2014.05.008.
80. Akdogan, O. *et al.* (2008) 'Vestibular nerve functions in children with auditory neuropathy', *International Journal of Pediatric Otorhinolaryngology*, 72(3), pp. 415–419. doi: 10.1016/j.ijporl.2007.11.004.
81. Sinha, K. S. *et al.* (2014) 'Cervical vestibular evoked myogenic potentials and caloric test results in individuals with auditory neuropathy spectrum disorders', *Journal of Vestibular Research: Equilibrium and Orientation*, 24(4), pp. 313–323. doi: 10.3233/VES-140510.
82. Singh, N. K., Sinha, S. K. and Barman, A. (2016) 'Assessment of otolith mediated neural reflexes through cervical and ocular vestibular evoked myogenic potentials in individuals

with auditory neuropathy spectrum disorders', *Hearing, Balance and Communication*, 14(2), pp. 77–90. doi: 10.3109/21695717.2016.1152047.

83. Sinha, S. K., Shankar, K. and Sharanya, R. (2013) 'Cervical and ocular vestibular evoked myogenic potentials test results in individuals with auditory neuropathy spectrum disorders', *Audiology Research*, 3, p. e4. p. 26-30. doi: 10.3109/21695717.2016.1152047.
84. Fujikawa, S. and Starr, a (2000) 'Vestibular neuropathy accompanying auditory and peripheral neuropathies.', *Archives of otolaryngology--head & neck surgery*, 126(12), pp. 1453–6. doi:10.1001/archotol.126.12.1453.
85. Buetti, B. and Luxon, L. M. (2014) 'Vestibular involvement in peripheral neuropathy: review', *International journal of audiology*, 53(6), pp. 353–9. doi: 10.3109/14992027.2014.885121.
86. Masuda, T. and Kaga, K. (2011) 'Influence of aging over 10 years on auditory and vestibular functions in three patients with auditory neuropathy', *Acta Otolaryngol*, 131(5), pp. 562–8. doi: 10.3109/00016489.2010.534112.

Appendix 1: Keywords

Auditory Neuropathy Spectrum Disorder/ANSD/Auditory Neuropathy/Auditory Dyssynchrony/Auditory Dysynchrony

- Unilateral / Bilateral
- Congenital/acquired
- History
 - Birth history – prematurity/preterm, low birth weight, anoxia, hypoxia, ventilation/CPAP intraventricular haemorrhage, ototoxicity, jaundice/hyperbilirubinaemia/kernicterus, maternal health, IVF, drugs, foetal alcohol syndrome/fetal alcohol syndrome/FAS,
 - Family History
- Examination
- Genetics – CAPOS, Otoferlin, Pejvakin, Pelizaeus-Merzbacher disease and leucodystrophies/leukodystrophies, consanguinity, mitochondrial mutations
- Neurological conditions – Charcot-Marie-Tooth syndrome, Friedreichs’ ataxia, hypoplastic cochlear nerve/aplasia, neuropathy, hereditary motor and sensory neuropathies, hydrocephalus, autonomic neuropathy, Refsum syndrome, delayed auditory maturation,
- Autoimmune disease
- Ototoxicity/neurotoxicity – Drugs e.g. cisplatin, Heavy metals e.g. lead
- Imaging
- Blood tests – haematology, biochemistry, immunology etc
- Congenital/acquired infection e.g. CMV, HIV, toxoplasma, rubella, herpes, Epstein Barr virus/EBV, meningitis
- Family audiograms
- Vestibular
- Ophthalmological – ERG, optic atrophy, visual maturation delay, central blindness
- Other investigations – renal, neurological, cardiological e.g. EMG, nerve conduction studies, somatosensory evoked responses,
- Other conditions: Paraneoplastic syndrome, temperature sensitivity, Vitamin deficiencies e.g. B1,6,12, diabetes

Appendix 2: Abbreviations and Acronyms:

ABR	Auditory brainstem evoked response audiometry
ANSD	Auditory neuropathy spectrum disorder, including, in this document, auditory neuropathy and auditory dyssynchrony
AUNA1	Auditory neuropathy, autosomal dominant 1 gene
AUNX1	Auditory neuropathy, X-linked recessive 1 gene
BAAP	British Association of Audiovestibular Physicians

CAPOS	<u>C</u> erebellar ataxia, <u>a</u> reflexia, <u>p</u> es cavus, <u>o</u> ptic atrophy, <u>s</u> ensorineural hearing loss,
CGH	Comparative genomic hybridization
CM	Cochlear microphonic
CMT	Charcot-Marie-Tooth syndrome
CMV	Cytomegalovirus
DIAPH3	Diaphanous related formin 3 gene
EMG	Electromyography
FA	Friedreich's ataxia
HSMN	Hereditary sensory motor neuropathy
IAMs	Internal auditory meati
MRI	Magnetic resonance imaging
NICU	Neonatal intensive care unit
NHSP	Newborn Hearing Screening Programme
OAE	Otoacoustic emissions
OPA1	Optic atrophy 1
OTOF	Otoferlin
PCHI	Permanent childhood hearing impairment
PTA	Pure tone audiogram
VEP	Visual evoked potential
VEMPs	Vestibular evoked myogenic potentials

Appendix 3: Useful resources for parents

- NDCS publications: 'Auditory Neuropathy Spectrum Disorder' and 'Auditory Neuropathy Spectrum Disorder: To Parents From Parents' - both available on NDCS website www.ndcs.org.uk
- <https://alicesears.com/> the website of a family with two children with ANSD
- https://www.youtube.com/watch?v=IY5Yliu_4t4 a simulation of what sound may be like for those with ANSD

Appendix 4: Audit Measures

The proforma of the BAAP national audit developed for this guideline can be used to benchmark practice. Available on www.baap.org.uk.

Appendix 5: Future Research

The evidence to support aetiological investigations is limited. Areas of research that could help to support an evidence base include:

- Yield of aetiological battery and individual aetiological tests/ assessments in children with various causes of ANSD
- Yield of history and clinical examination using a prospective study

- Systematic review of studies on aetiological investigations

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