UK National Screening Committee

UK National Screening Committee (UK NSC)

Screening for Newborn Hearing Screening Programme modification – ANSD

Date: 28 October 2020

Contents

Aim

1. To ask the UK National Screening Committee (UK NSC) to make a recommendation, based on the evidence presented in this document, on whether screening for Auditory Neuropathy Spectrum Disorder (ANSD) should be included in the well-baby newborn hearing screening protocol through the addition of the automated Auditory Brainstem Response (AABR) test.

Current Recommendation

2. ANSD is not currently screened for in the Newborn Hearing Screening Programme (NHSP). The addition of screening for ANSD as an extension to the current NHSP protocol was proposed in the 2017/18 annual call for topics. The submission was submitted by the National Deaf Children's Society (NDCS) and proposed that an Auditory Brainstem Response (ABR) test be carried out in addition to the Otoacoustic Emissions (OAE) test performed in the NHSP as the primary screen. It was agreed that the submission was within the remit of the UK NSC and that the submission be treated as a major programme modification. An evidence summary by Bazian Ltd. was commissioned in 2019.

3. The current well-baby protocol for the NHSP in England uses the automated otoacoustic emission (AOAE) test as the initial screening test with AABR performed as the second step only if the baby fails the AOAE in one or both ears. As ANSD is characterised by an 'AOAE pass/AABR fail' screening response it is possible that well-babies with ANSD would not be identified in the current well-baby protocol.

a. Concurrent AABR and AOAE screening is only performed in the neonatal intensive care unit (NICU) or special care baby unit (SCBU) protocols, as babies in these settings are known to be at higher risk of hearing problems, including ANSD.

b. The screening programme in Northern Ireland follows the same protocols as England.

c. Wales differs slightly in that well-babies who fail the initial OAE may be referred either directly for diagnostic ABR or for repeat OAE. Babies who spend >48 hours in NICU/SCBU receive initial AABR screening. However, as only high-risk babies receive initial AABR screening, the Welsh well-baby protocol would similarly not detect ANSD.

d. In Scotland, 2 different protocols are currently in operation in both well-baby and NICU/SCBU populations. In half of the 14 NHS boards, well-babies are screened using OAE only (with AABR only if the baby fails initial and repeat OAEs), while in the other half well-babies are screened using AABR (with repeat AABR for a fail response). Therefore, it is where this latter protocol is applied in Scotland that well-babies with the ANSD test profile could be screen-detected in the UK. NICU/SCBU babies in Scotland may be screened using AABR only (like Wales) or using both AOAE and AABR (like NHSP).

Evidence Summary

4. This review was undertaken by Bazian in accordance to the UK NSC evidence review process.

5. The review considered 3 questions:

   a. What is the incidence of ANSD in newborn babies in the UK? What proportion present in NICU/SCBU and what proportion present in the otherwise well-baby population?

   b. What is the test performance of AABR screening to identify ANSD among the well-baby population?

   c. What are the practical implications of including AABR screening for well-babies?

6. The review concluded that ANSD should not be added to the current well-baby protocol:

   a. There is uncertainty about the number of babies with ANSD not detected through the current protocol. The published literature on the incidence of ANSD among the NICU and well-baby populations in the UK was limited in volume and conclusions could not be drawn.

Unpublished data submitted from the NHSP in England (2006 to 2017) and UNHS in Scotland (2014 to 2017) provided the best information. However, the conclusions on the number of cases that might be missed and whether these would predominantly be bilateral or unilateral cases are uncertain and based on extrapolation from a small number of cases detected during 3 years of AABR screening in 7 health boards in Scotland.

- **Criterion 1 not met**

b. Only one small cohort trialled AABR screening in the well-baby population in one US region, where OAE was performed sequentially for those who failed the AABR. Sensitivity, specificity or NPV could not be assessed in this study due to the lack of audiology follow-up of babies who passed AABR, either at screening or repeat testing. There were also applicability concerns. Overall, the current evidence provides an unreliable indication of the test performance of AABR screening of well-babies.

- **Criterion 4 not met**

c. No UK evidence was identified to inform the practical implications of including AABR in newborn hearing screening for well-babies. The 2 studies included, both demonstrated that AABR screening takes longer to perform, permits screening of fewer newborns, and is associated with higher costs. There was also uncertainty on the inputs used to estimate cost and timing and inconsistency between the studies. Neither accounted for changes to hospital infrastructure, personnel requirements or training. There was uncertainty whether the devices used were applicable to the UK. The studies also considered only single stage screening, rather than a programme where both screening tests are performed concurrently. There was no information on the acceptability of well-baby AABR screening to either health professionals or parents, or whether this may affect screening uptake by parents.

- **Criterion 6, 14 and 18 not met**

Consultation

7. A three-month consultation, ending on the 19 October 2020, was hosted on the UK NSC website. 22 direct emails were sent to stakeholders, including 13 organisations and several individuals. Some organisations received duplicate emails. Stakeholders are listed in *Annex A*. Comments were received from the following 3 stakeholders (See *Annex B* for comments):

a. xxxx xxxx, NHSP Programme Manager (xxxx Hospitals NHS Foundation Trust)

b. xxxx xxxx, Consultant Audiovestibular Physician (British Association of Audiovestibular Physicians)

c. Royal College of Paediatrics and Child Health: Comments received on behalf of xxxx xxxx (British Association of Paediatricians in Audiology)

8. Overall the 3 stakeholders did not disagree with the conclusions of the review and broadly acknowledged the potential issues and challenges related to AAB R.

9. One stakeholder acknowledged the particular challenges of introducing AABR into the well-baby protocol where newborn screening is undertaken by maternity support workers in the community who are not trained in using AAB R as well as the possible practical, time and cost implications.

10. There were several comments from one stakeholder on the plain English summary.

Response: This is intended to provide a simplified overview for the general public and other non-specialists so would not be expected to fully capture the technicalities and intricacies assessed in the main review. However, one change was made on page 5 when referring to the number of babies missed. This was amended to “4 out of 6 per 100,000 babies” for accuracy and consistency.

11. Two possible inaccuracies on page 7 were highlighted, suggesting “respiratory distress” should be changed to “significant hypoxia” and “developmental delay” should be changed to “speech and language delay.”

Response: Respiratory distress was the term referred to in the British Society of Audiology guidance from the Sininger et al. reference. The second has been changed to “speech and language developmental delay.”

12. There were questions and comments on the data provided by the English and Scottish screening programmes. The relevant data was summarised in the question level synthesis and all the data made available to the reviewer was included in the appendix. There was also a comment on the aetiology data and whether this was routinely inputted into the national database.

Response: The NHSP responded that the aetiology module for the national NHS P IT system is under-utilised and that the data provided by them came from the audiology module where the audiologist has set the type of hearing loss as ANS D.

13. One comment referred to the uncertainty on the devices used in the studies included for question 3.

Response: The devices used in the US study are not manufactured in the UK and the devices in the Iranian study were not specified.

14. There was also a question on whether other countries were contacted regarding incidence data and cost, time, resource etc. implications.

Response: The methodology for the UK NSC rapid review process usually includes published literature only. We included the data from the English and Scottish Programmes as it was made available from established programmes.

15. Two stakeholders asked for clarification on the reference to the use of cochlear implants from the British Society of Audiology document on page 15.

Response: The reviewer has amended a sentence for accuracy to address this. The sentence “however, due to the variable outlook, cochlear implants are usually only considered at a later stage when audiological assessments are stable and it is clear that there is permanent profound hearing loss” has been changed to; “cochlear implants are usually only considered at a later stage when audiological test results are stable and demonstrate that the child has permanent profound hearing loss; and/or if the child shows limited speech discrimination with conventional hearing aids.”

16. Two stakeholders commented on the nature of ANSD and the uncertainties of its management. One comment asked whether AN was also included and stated that a definition of ANSD should have been included.

Response: The request from the annual call was for ANSD and a definition was provided in the introduction and approach on page 13.

Recommendation

17. The Committee is asked to approve the following recommendation:

Screening for ANSD through the addition of AABR in to the newborn hearing screening well-baby protocol is not recommended.

UK NSC members were given an additional 2 weeks to review, comment on and agree this recommendation following the public consultation and amendments made following the October UK NSC meeting.

This recommendation was approved by Chair’s action on 4 December 2020.

Criteria (only include criteria included in the review)	Met/Not Met
Criteria for appraising the viability, effectiveness and appropriateness of a screening programme	
The Condition	
1. The condition should be an important health problem as judged by its frequency and/or severity. The epidemiology, incidence, prevalence and natural history of the condition should be understood, including development from latent to declared disease and/or there should be robust evidence about the association between the risk or disease marker and serious or treatable disease.	**Not Met**
The Test	
4. There should be a simple safe precise and validated screening test.	**Not Met**
The Test, Screening Programme, Implementation	
6. The test, from sample collection to delivery of results, should be acceptable to the target population	**Not Met**
14. The opportunity cost of the screening programme (including testing, diagnosis and treatment, administration, training and quality assurance) should be economically balanced in relation to expenditure on medical care as a whole (ie. value for money). Assessment against this criteria should have regard to evidence from cost benefit and/or cost effectiveness analyses and have regard to the effective use of available resource.	
18. Adequate staffing and facilities for testing, diagnosis, treatment and programme management should be available prior to the commencement of the screening programme.	

Annex A

List of organisations contacted

1. xxxx xxxx
2. xxxx xxxx
3. xxxx xxxx
4. National Deaf Children's Society
5. Action on Hearing Loss (formerly RNID)
6. British Academy of Audiology
7. The Communication Trust
8. Royal College of Paediatrics and Child Health
9. The British Association of Perinatal Medicine
10. The Institute of Child Health
11. The Royal College of Midwives
12. The Faculty of Public Health
13. Royal College of General Practitioners
14. Royal College of Physicians
15. Royal College of Physicians of Edinburgh
16. Royal College of Physicians and Surgeons of Glasgow

UK National Screening Committee

Annex B

1. xxxx xxxx, NHSP Programme Manager (xxxx Hospitals NHS Foundation Trust)

Name:	xxxx xxxx	**Email address:**	xxxx xxxx
Organisation (if appropriate):	xxxx xxxx		
Role:	**NHSP Manager**		
Do you consent to your name being published on the UK NSC website alongside your response? **No**			

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
	General comments	The national programme determines whether a newborn screening programme model is 'hospital' or 'community' according to the age of the baby at the initial screen, with a hospital model referring to babies screened at less than 10 days old and a community model for babies over 10 days of age. Whilst there is not enough information at present to support a change to the newborn hearing screening programme, it should be noted that community models and some hospital models exist where care is delivered in community settings. This is where the hearing screener or health visitor deliver other aspects of the postnatal care. Some screening sites employ maternity support workers who complete the hearing screening, newborn bloodspot and other baby checks at day 5.

		The paper outlines that further evidence is needed on the practical implications of AABR screening. The inclusion of the AABR screen on these visits would be time consuming and may mean splitting aspects of the postnatal care with more appointments for the parent to attend.
		There may also be cost implications regarding training/diploma courses for these screening sites as not all screeners perform the AABR screen. Further costs may also result from acquiring more equipment and consumables for service coverage.

Please return to the Evidence Team at screening.evidence@nhs.net **by Monday 19 October 2020.**

UK National Screening Committee

2. xxxx xxxx, xxxx xxxx (British Association of Audiovestibular Physicians)

Name:	xxxx xxxx	**Email address:**	xxxx xxxx
Organisation (if appropriate):	BAAP		
Role:	xxxx xxxx		

Do you consent to your name being published on the UK NSC website alongside your response?

Yes No

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
Page 5 ,2nd para	ANSD means that there is a problem with the hearing nerve	It could also be a neurotransmitter issue involving the inner hair cell e.g. Otoferlin
Page 5, 2nd para	AOAE test	This checks the outer hair cell and therefore misses ANSD
Page 5, 3rd para	Some healthy babies … might have ANSD	Have you found out what proportion of well babies have ANS D? There are other countries where AOAE and AABR are used for NHSP. They will have the answer.
Page 5, 4th para	ANSD may actually affect 6 out of 100,000 well-babies	Have these babies had AOAE at the same time?
Page 5, 4th para	Miss 4 out of 6 well-babies	4 out of 6 per 100,000 babies
Page 5, 4th para	Might have ANSD that only affects one ear and so is less likely to affect their development	Do you have numbers i.e. unilateral vs bilateral and how much unilateral ANSD affects hearing and development?
Page 5, 5th para	This happens sometimes because it can take a few weeks for hearing to develop properly.	Hmmm... delayed maturation causing ABR abnormalities .... do you want to use the term ANSD here or use something like "transient ANSD"? I think you need to exclude delayed maturation before using the term ANSD but again ANSD electrophysiology and symptomatology could change over the years

		- Also we need to exclude absent VIII nerve before using the term ANSD I think.
Page 6, 1st para	No UK studies were found that could provide an estimate of resource use and costs in the UK.	surely this is should be available from other countries? Have you contacted them?
Page 6, 4th para	AABR performed as the second step only if the baby fails the AOAE in one or both ears.	The problem you will face is that AABR is geared to detect moderate or greater hearing loss. I think most AABR equipment deliver 35dB clicks so there is a downside to it. You will miss mild SNHL that could be beginning of a progressive HL!
Page 6, 5th para	The AOAE test tends to have a high false positive rate for any hearing loss, with subsequently higher referral rates than AABR	Doing AOAE on Day 2 (age 3 days) minimises this. It is true that AABR leads to reduced referral rate but at what expense - please see my earlier comment
Page 7, 2bd para	Respiratory distress	Should this be significant hypoxia?
Page 7, 2nd para	Developmental delay	Speech and language delay
Page 8, 1st para	Rather, the review solely focuses on the issue that AABR should be included in the first-stage of the protocol because AOAE would not detect ANSD among the well-baby population as ANSD is characterised by an AOAE test pass	Agree
Page 8, 3rd para	The focus of this rapid review was on ANSD	Did you include AN as initial publications were on this and not on ANSD?

Page 9, 2nd para	Incidence of ANSD	how did you define this? True ANSD excluding transient ANSD or overall ANSD diagnosed? If tha5 is the case what proportion recovered?
Page9, 3rd para	lower incidence of ANSD of 2 in 100,000.	I think you need to include the definition of the term ANSD - when did you use the term? what were the exclusions?
Page 10, 1st para	Further study of AABR screening in a larger sample of well-babies would be needed to confirm whether the incidence of 6 in 100,000 is correct,	Have you contacted other countries e.g. USA, Aus, NZ, Europe etc?
Page 10, 1st para	clinical diagnosis of bilateral ANSD affects	this is important - have people been inputting this data (aetiology) on the national database?
Page 10, 1st para	management and outcomes compared with screening detection	completely agree - we need to find out the causes, progression etc. I hope BAAP Aetiology course have highlighted the importance of collecting the aetiology
Page 10, 2nd para	These cases likely reflect transient ANSD at birth due to delayed neural maturation	How many were these preterm babies (not severe enough to end up in NICU? When did they do repeat testing?
Page 10, 4th para	A UK study is needed to assess the performance of AABR screening among well-babies using AABR and A OAE devices as used in the NHSP.	Agree
Page 11, 1st para	AABR and AOAE devices used are applicable to the UK.	Did you not find out?

Page 15, 1st para	performing ABR at this age	although this is the only way for confirmation of persistence of ANSD
Page 15, 2nd para	considered at a later stage when audiological assessments are stable and it is clear that there is permanent profound hearing loss.	Not quite sure about this. I think this is based on behavioural hearing level and speech discrimination. Those who have poor speech discrimination through hearing can be considered and have been implanted with significant benefit. Using electrical ABR might help looking at improvement of dysynchrony that will give an indication of benefit of CI.

Please return to the Evidence Team at screening.evidence@nhs.net **by Monday 19 October 2020.**

3. Royal College of Paediatrics and Child Health: Comments received on behalf of xxxx xxxx (British Association of Paediatricians in Audiology)

UK National Screening Committee

Name:	Royal College of Paediatrics and Child Health	**Email address:**	xxxx xxxx
Organisation (if appropriate):	Royal College of Paediatrics and Child Health: Comments received on behalf of Anne Marsden (BAPA)		
Role:			

Do you consent to your name being published on the UK NSC website alongside your response?

Yes

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
Page 6-7	Nature of ANSD	ANSD a label for a set of audiological results/ 'condition' that is poorly understood for long term outcomes for the child. There would need to be a longitudinal study to look at outcomes etc. The original HTA by Bamford and Davis in 1997 summarising the evidence for universal screening was about intervening with early rehabilitation etc. to improve outcomes. Without some evidence we would be screening for something (likely unilateral ANSD), we really weren't sure how to manage without evidence, if in fact it does need managing.
Page 15	Cochlear implants are used when it is clear that there is a profound hearing loss isn't correct.	This is referenced to the BSA document (ref 1). The BSA document states 'Behavioural pure tone thresholds are not a good guide to determine CI candidacy. Children with ANSD who have even relatively mild hearing losses on behavioural testing may be CI candidates if they do not show good progress with other interventions.'