

NEWBORN HEARING SCREENING AND ASSESSMENT

**Guidelines for surveillance and audiological referral
of infants & children following the newborn hearing screen**

Version 5.1

June 2012

NHSP Clinical Group

Graham Sutton¹ (Editor), Sally Wood¹, Rachel Feirn², Sally Minchom³, Glynnis Parker⁴, Tony Sirimanna⁵

¹ Newborn Hearing Screening Programme Centre, Royal Free Hospital, London, UK

² (nee Foley), formerly of Bristol Royal Hospital for Children, UK

³ North Wales NHS Trust, UK

⁴ Sheffield Children's Hospital, Sheffield, UK

⁵ Great Ormond St Hospital for Sick Children NHS Trust, London, UK

With thanks to the following for contributions to earlier versions; Roger Green (Windsor), Clare Robertson (Oxford), Kai Uus (Manchester), John Bamford (Manchester), Adrian Davis (London), Jane Grant (Milton Keynes), Sally Hind (Nottingham), Helen Robertson (Birmingham), Yogi Thakker (Milton Keynes), Peter Watkin (Whipps Cross), Tim Williamson (Bath)

Correspondence to Graham Sutton, c/o Newborn Hearing Screening Programme Centre, 344-354 Grays Inn Rd, London WC1X 8BP, UK.

Amendment History:

Version	Date	Amendment History
1	May 2003	Drawn up by group of professionals, then consultation through website including members of the NHSP Executive Group and NDCS. Approved by NHSP Executive and Steering Group.
2	June 2005	Edited after discussion with group of professionals, then consultation on website. Approved by NHSP Executive and Steering Group.
3	June 2006	Complete revision with NHSP Clinical Group. v3.0 agreed by Group 5/6/06 and submitted to Programme Director. Further amendments and queries by Director reviewed/ incorporated GS as v3.1 13/6/06.
4	July 2009	Complete revision with NHSP Clinical group. Draft v4.0 agreed May 2009. Further revisions made following website consultation, v4.1 approved July 2009 by Clinical Group and Director
4.2	November 2010	Minor amendments (see Appendix B). Approved after consultation with Clinical Group and Director
5	June 2012	Revised version v5.1 including major changes to targeted follow up categories as agreed by Clinical Group and Director after open consultation. Meningitis guidance revised and added as Appendix A

Forecast Changes:

Anticipated Change	When
Further Review	2015

CONTENTS

1. OVERVIEW

2. GENERAL COMMENTS

- 2.1 Other screens
- 2.2 Age at testing
- 2.3 Audiology teams and services
- 2.4 Information and data

3. CATEGORIES OF BABIES & RECOMMENDED FOLLOW-UP

A. RELATED TO SCREEN

- 3.1 Babies excluded from screen – Refer for ABR
- 3.2 Screen incomplete
 - Declined screen, missed screen, incomplete screen – No routine follow up
 - Moved into the country before 3 months of age – Screen
 - Moved into the country after the age of 3 months – Refer.
- 3.3 Referred on screen but missed audiology follow up – Follow up
- 3.4 Passed screen or audiology follow up, but with specific neonatal risk factors – Refer

B. REFERRAL ARRANGED LATER (NOT RELATED TO SCREEN)

- 3.5 Specific risk factor or concern occurring later (irrespective of newborn screen result) - Refer
- 3.6 Ototoxic drugs – Refer only at discretion of Paediatrician

SUMMARY OF RECOMMENDATIONS

APPENDIX A

Guidelines for audiological follow up of babies diagnosed with bacterial meningitis and/or meningococcal septicaemia

APPENDIX B

Development of these guidelines
Main Changes made in version 5
Main changes made in versions 4.1 and 4.2
Main Changes made in version 3

APPENDIX C

Requirements for audiological data entry in eSP

REFERENCES

Glossary

AABR	Automated Auditory brainstem response (screening test)
ABR	Auditory brainstem response (full assessment/ diagnostic test)
CFA	Cranio-facial abnormality
CMV	Cytomegalovirus
Corrected age	Age adjusted for prematurity (based on 40 week term)
ECMO	Extra-corporeal membrane oxygenation
eSP	e-Screener Plus (Electronic record system for NHSP)
MEE	Middle ear effusion
NHSP	Newborn Hearing Screening Programme (England)
NICU/SCBU	Neonatal intensive care unit / Special care baby unit (terms used interchangeably for NHSP). The NHSP NICU/SCBU screen protocol is for those on the unit for 48 hours or more.
PCHI	Permanent Childhood Hearing Impairment - defined here as ≥ 40 dBHL averaged over 0.5, 1, 2 & 4 kHz pure tone audiometry thresholds. It includes both sensorineural and permanent conductive impairments.
OAE	Otoacoustic emission
SNHL	Sensorineural hearing loss
VRA	Visual Reinforcement Audiometry

1. OVERVIEW

The aim of the Newborn Hearing Screening Programme in England (NSHP) is the early identification of all children who have a significant permanent childhood hearing loss^a (PCHI) in the neonatal period. However even with a very sensitive newborn hearing screen, some children will develop a hearing loss later or will have missed screening or follow up. Therefore a wider system is needed to identify these children. See Appendix B for a brief discussion of the prevalence of later onset loss.

This document identifies which children should be followed up and monitored, how and when this should be done, and by whom. It covers surveillance, referral and audiological monitoring following newborn hearing screening. It does not cover all the conditions requiring hearing assessment which may occur in older children. Training should be provided to primary care and other professionals to ensure early referral of children who have a high risk of late-onset or acquired hearing loss, and written care pathways should be developed at local service level to cover these.

The main changes made to this document over time are listed in Appendix B.

This version follows a major review of the evidence on yield in NHSP (see Wood et al 2011, Davis et al 2012). We now have more confidence that for many of the previous and commonly used risk factors (e.g. JCIH 2007) there is no good evidence of a strong link to progressive or late-onset hearing loss, and thus no need for routine follow-up after a clear pass on the newborn screen. We therefore believe this guidance represents current best practice. Any site considering diverging from this guidance should be able to clearly justify this to commissioners based on firm documented evidence. Finally the specific meningitis guidance, previously in a separate document, has been updated and revised, and is incorporated as Appendix A.

2. GENERAL COMMENTS

2.1 Other screens

A general principle is that children whose newborn hearing screen shows clear responses on both ears should not be subject to repeated screens, tests or follow-up unless they meet one or more of the criteria specified here. Parents will be given appropriate checklists after their newborn screen to refer to in the first instance if there is concern.

A child referred for audiological assessment should not be discharged until testing clearly and definitively shows that they have satisfactory responses^b.

Current advice is still that the School Entry hearing screen and follow-up tests be maintained (Bamford et al 2007) with staff appropriately trained, and clear protocols for both screen and follow-up. A system for recording screening activity and audit (to give coverage, referral and yield) of the School Entry Screen should be in place.

2.2 Age at testing

All ages in this document refer to corrected ages (from expected date of delivery), and are indicative – in practice ‘4 weeks’ may be 2 to 8 weeks, and ‘8 months’ may be 7 to 9 months (and in exceptional circumstances up to 12 months). While ABR testing is technically possible at any age, in practice it becomes increasingly difficult over 12 weeks as the required sleep state becomes less predictable. Reliable behavioural assessment by VRA is increasingly possible from 5-6 months of age, but developmental delay will affect the ability to test behaviourally, as may some serious medical conditions.

^a NHSP defines this as a bilateral permanent hearing loss averaging ≥ 40 dBnHL across 0.5 to 4 kHz

^b “clear and definitive” is a matter for clinical judgement. Normally it would mean with ear-specific hearing thresholds within normal limits across the frequency range. (see also NHSP assessment guidelines)

2.3 Audiology teams and services

55 Hearing testing should always be carried out by appropriately-trained members of the
audiology team (see NDCS 2000, DH 2008). Where this document refers to this team or
'Audiology', this may be in a hospital or community base, according to local arrangements.
In many areas initial assessment for older infants would be at an 'intermediate' clinic
before referral on to a main centre. For ABR / electrophysiological testing, referral should
60 be to an audiology department with the ability to perform definitive testing - not to
intermediate clinics or staff. The NDCS Quality Standards document (NDCS 2000)
contains detailed guidelines as to the requirements within the Audiology team. Testing
should be carried out in accordance with the published protocols (NHSP 2001-8).

65 We strongly recommend the use of VRA rather than distraction testing for behavioural
testing in infants. Ear-specific testing should be routinely available (see NHSP VRA
guidance for details).

2.4 Information and data

70 Details of data required to be entered onto eSP for those undergoing surveillance and
others are specified in the document 'Requirements for audiological data entry in eSP'
(see Appendix C).

75 Screening outcome and referral information of all babies should be entered on the national
screening database (eSP) as soon as possible and at least by the time the child is 3
months old.

Any baby having targeted follow-up should have at least the date, duration and outcome of
the appointment noted.

80 Any child with PCHI (of any degree in either ear) identified outside the newborn screening
process should have information entered on eSP, and it is the responsibility of the
Audiology service to do this. Some such cases (who may be 'false negatives' for the
screen) also need to be notified to the Programme Centre using the form 'NHSP Report of
85 case of PCHI not identified by the newborn hearing screen' which can be found at
<http://hearing.screening.nhs.uk/surveillanceguidelines>

For all PCHI cases data are required for each appointment from referral to confirmation of
PCHI and thereafter a regular update as per guidance

90

3. CATEGORIES OF BABIES & RECOMMENDED FOLLOW-UP

Those identified as having a permanent bilateral childhood hearing impairment must be
followed up on an indefinite basis.

95 A. RELATED TO SCREEN

3.1 Babies excluded from screen – Refer for ABR

The 2 groups who should not undergo the newborn screen are babies with

- 100 • **Microtia / external ear canal atresia** - where there is no patent ear canal in one
or both ears
- **Neonatal bacterial meningitis or meningococcal septicaemia^c** – Confirmed or
strongly suspected bacterial meningitis^d (any organism), or meningococcal
septicaemia

^c It is the responsibility of the paediatrician or neonatologist to make the judgement regarding 'strongly suspected', and their responsibility to notify audiology. Possible bacterial infection is commonly treated in SCBU by prophylactic antibiotics - screening may be completed unless advised to the contrary by the responsible clinician.

105 These exclusions are made because a) those with microtia / external ear canal atresia will always have a degree of loss, and b) for bacterial meningitis the risk of SNHL is very high (see Appendix B).

110 On recovery from the acute episode of meningitis these children should be referred by the Paediatrician to Audiology and given an early hearing assessment (ABR) within 4 weeks of discharge from hospital. The urgency for post-meningitis children is related to the risk of ossification of the cochlea and that urgent cochlear implant referral may be required. Note that viral meningitis is not considered to be a specific risk to hearing and where this is confirmed the screen can proceed as normal, with no need for extra follow up. More specific guidance on hearing assessment after meningitis is given in
115 Appendix A.

120 Referral is the responsibility of the medical team caring for the child. Screening teams should treat these babies as screen referrals and continue to log them as such in eSP using the screening outcome of 'incomplete-screening contraindicated' and expedite and monitor their referral to audiology in conjunction with the medical team.

Responsible for identifying child and referral to Audiology - Paediatrician
Responsible for arranging appointment and follow-up - Audiology (copy to NHSP screening manager)

125

3.2 Screen incomplete

- **Declined screen, missed screen, incomplete screen - No routine follow up**

130 Routine referral for targeted follow up is no longer recommended for this group. Screening teams should make vigorous efforts to maximise newborn screen coverage by 3 months of age, including for those who moved in before this age.

135 Programmes should consider the following mechanisms to maximise coverage: telephone / text / email reminders, outreach clinics, home visits, liaison with trust antenatal and newborn screening coordinators, midwifery and health visiting teams, contact with paediatric wards and intensive care units to identify readmitted unscreened babies. HVs and GPs must be informed of babies that have not completed screening. Screening teams will need to ensure that parents are provided with information about how to seek assessment in the event of future
140 concern.

The HV/GP should discuss with parents the implications of not having (or not completing) the screen. Parents and HVs/GPs should be made aware that they can request an audiological assessment at any time.

145 *Responsible for identifying child and alerting HV/GP – NHSP Screening manager*

- **Moved into the country before 3 months of age - Screen**

150 Programmes need to have mechanisms in place to identify these babies and offer the screen. Child health and HVs are the best source of information

Responsible for identifying child and alerting the Newborn hearing screening team –HV/Child Health Department/other local arrangement

Responsible for arranging newborn hearing screen appointment - NHSP screening coordinator

155

^d In practice it is not always simple to separate bacterial meningitis from viral cases, and many are treated before diagnosis. It may be simpler and more effective to allow all cases to be referred.

- **Moved into the country after the age of 3 months – Refer**

These babies are not eligible for newborn screening but the HV should discuss with the parents as soon as feasible and offer referral for age-appropriate testing.

160 *Responsible for identifying child – HV/Child Health Department/other local arrangement*
Responsible for arranging Audiology referral– HV

165 **3.3 Referred on screen but missed audiology follow-up – Follow up**

Audiology must make strenuous efforts to secure attendance of these babies for ABR or (if necessary) behavioural follow up including discussion with parents and liaison with the family health visitor and GP to facilitate attendance. Audiology and screening teams should liaise closely about these children to ensure that audiology staff are aware of the likelihood of PCHI in this group.

170
175 In the event of inability to secure attendance the HV and GP should be notified and advised about how to make a referral should the family indicate a willingness to attend in future. Completion of follow up for these children should be locally audited with responsibility for audit devolved to a named individual within screening or audiology.

Responsible for identifying child (as not having attended) – Audiology
Responsible for arranging further appointment – Audiology.

180 **3.4 Passed screen or audiology follow up, but with specific neonatal risk factors – Targeted Follow up**

These children to continue to be referred for targeted follow up (behavioural testing around 8 months), or sooner if local protocol in place

- **Syndromes associated with Hearing loss (including Down’s)**
- **Cranio-facial abnormalities including cleft palate**
- **Confirmed congenital infection^e (toxoplasmosis, rubella or CMV)**
- **SCBU/NICU over 48hr with no clear response OAE both ears but clear response on AABR**

190 Note that although family history (of permanent SNHL from childhood in parents/siblings) has been removed as a risk factor requiring routine targeted follow up (see Appendix B) any parent who still expresses concern about hearing despite the screen should always be directly referred to Audiology. (If concerns arise later, see under parental concern, section 3.5).

195 *Responsible for identifying child– Screening team*
Responsible for arranging appointment– Audiology.

200 **B. REFERRAL ARRANGED LATER (NOT RELATED TO SCREEN)**

3.5 Specific risk factor or concern occurring later (irrespective of newborn screen result) - Refer

205 *Immediate referral to Audiology for age-appropriate assessment as soon as possible carried out by an appropriately trained team.*

^e Note that screening teams may not always be aware of these children particularly where screening is completed early with a clear response before other test results are confirmed. It is assumed that other professionals involved with these families will be aware of the need for ongoing audiological assessment

- Parental or professional concern
- Confirmed or strongly suspected bacterial meningitis, or meningococcal septicaemia^{c, d}
- Temporal bone fracture
- Severe unconjugated hyperbilirubinaemia

210

Parental concern about an infant's hearing, or development of auditory or vocal behaviour should always be taken seriously. All professionals who may be in contact with a child should always feel able to refer to Audiology if there is parental concern, or if they themselves are concerned.

215

Responsible for identifying/referral to Audiology - whichever professional discovers or becomes aware of concern (may be HV /GP /Paediatrician /Speech-Language Therapist).

220

Responsible for arranging appointment – Audiology

The medical conditions named above can cause sensorineural hearing loss in a significant proportion of affected children (meningitis - Fortnum 1992, Fortnum & Davis 1993; temporal bone fracture - Zimmerman et al 1993, Lee et al 1998; hyperbilirubinaemia - Boo et al 1994, Shapiro 2003). If they occur at any point in infancy or childhood after the screen, then immediate referral should be made to Audiology for an age-appropriate audiological assessment on recovery and within 4 weeks of discharge from hospital.

225

230

*Responsible for identifying child and referral to Audiology - Paediatrician
Responsible for arranging appointment and follow-up - Audiology*

3.6 Ototoxic drugs – Refer only at discretion of Paediatrician

235

Various drugs are potentially ototoxic. The main group is aminoglycosides and these are very commonly used prophylactically in babies. Unless a baby is suspected or known to have the A1555G mitochondrial mutation^f (see below), the baby should be screened in the normal way and followed up if required as per standard screening protocol.

240

The responsibility for monitoring of children receiving ototoxic drugs and appropriate referral for audiological assessment lies with the Paediatrician and medical team. In deciding whether to make a referral for follow up beyond the screen one factor will be whether the monitored aminoglycoside levels have exceeded the therapeutic_range: see also national guidance on use of gentamicin for neonates (NPSA 2010).

245

However, any baby that is suspected or known to have the A1555G mitochondrial mutation and has received aminoglycosides (irrespective of whether blood levels are within the therapeutic range) should be referred for immediate follow-up and audiological monitoring irrespective of screen outcome.

250

*Responsibility for making the referral and communication with family - Paediatrician
Responsibility for making appointment - Audiology*

There is a brief discussion of the issues in Appendix B.

^f Babies with A1555G mitochondrial mutation may have a family history of sensorineural deafness from middle age in the affected individuals and the transmission is maternally inherited. It is now possible to test for this mutation.

255

SUMMARY OF RECOMMENDATIONS

A. RELATED TO SCREEN

260

3.1 Babies excluded from screen - Refer for ABR

- Microtia / external ear canal atresia
- Neonatal bacterial meningitis or meningococcal septicaemia

265

3.2 Screen incomplete

- Declined screen, missed screen, incomplete screen – No routine referral
- Moved in before 3m – Screen
- Moved in after 3m – Refer

270

3.3 Referred on screen but missed audiology follow up - Follow up

275

3.4 Passed screen or audiology follow up, but with specific neonatal risk factors – Refer For Targeted follow up

- Syndromes associated with Hearing loss (including Down's)
- Cranio-facial abnormalities including cleft palate
- Confirmed congenital infection (toxoplasmosis, rubella or CMV)
- SCBU/NICU over 48hr with no clear response OAE both ears but clear response on AABR

280

285

B. REFERRAL ARRANGED LATER (NOT RELATED TO SCREEN)

3.5 Specific risk factor or concern occurring later (irrespective of newborn screen result) - Refer

290

- Parental or professional concern
- Confirmed or strongly suspected bacterial meningitis or meningococcal septicaemia
- Temporal bone fracture
- Severe unconjugated hyperbilirubinaemia

295

3.6 Ototoxic drugs – Refer only at discretion of Paediatrician

300

APPENDIX A

Guidelines for audiological follow up of babies diagnosed with bacterial meningitis and/or meningococcal septicaemia

June 2012

Scope

310 These guidelines have been produced to help clinicians develop local protocols for hearing assessment in babies up to the age of one year who have been diagnosed with bacterial meningitis and/or meningococcal septicaemia.

General information

315 The responsibility for ensuring referral for hearing testing in this group of babies resides with the responsible Paediatrician. Protocols need to be in place to ensure referral from the paediatric wards or NICU/SCBU to the responsible clinician in Audiology. Hearing assessment needs to be carried out within four weeks of the child being well enough to be tested. **Urgent assessment is required to identify severe/profound hearing loss which may require cochlear implant(s) before any cochlear ossification takes place. The timing of tests needs to be practical and flexible. The aim should be to determine ear-specific and frequency-specific auditory thresholds as soon as possible, to identify hearing loss of any degree or configuration.** Children can also have complex developmental problems following meningitis.

Under 12 weeks corrected age:

325 The baby should be referred for assessment irrespective of whether or not they have been screened and irrespective of the screen result. as they are very high risk for having a hearing loss. Testing would normally be by ABR under natural sleep, preferably using both high and low frequency stimuli. If this is not possible, a diagnostic OAE test would be helpful but in this case further assessment (for ABR or behavioural testing) should be arranged.

Between 12 weeks and 7 months corrected age:

335 Options should be discussed by the audiologist with parents and include one or more of

- ABR under natural sleep (especially if the baby is still quite young), preferably with both high and low frequency stimuli;
- Diagnostic OAE test ;
- ABR under sedation (for older infants, if there is considerable parental/professional concern or if it has not been possible to obtain a reliable test without sedation);
- Waiting until behavioural testing around 7 months (if the baby is close to this age), bearing in mind the importance of urgent assessment as discussed above).

Over 7 months corrected age:

345 The baby should be referred to Audiology on discharge from hospital and seen within 4 weeks. Testing would normally be by VRA using ear-specific and frequency specific stimuli. A significant hearing loss should be excluded. If ear- and frequency-specific information cannot be obtained for whatever reason, the child should be further reviewed to rule out any milder degrees of loss.

350 For all ages, there appears to be no good evidence supporting the need for further follow up if the hearing is found to be satisfactory following meningitis (see note in Appendix B), so we do not make recommendations. However this is a matter for local policy.

355

APPENDIX B

1. Development of these guidelines

360 These guidelines have gone through numerous versions since the first issue in 2003: the main changes are detailed below.

365 The relative contribution of later acquired PCHI to the total population of PCHI in children has been difficult to ascertain. In the 1997 HTA review Davis et al reviewed the data and estimated the prevalence of bilateral congenital PCHI to be 1.12 per 1000 (95% CI 1.01 - 1.23) with an additional 0.21 per 1000 (95% CI 0.17-0.26) subsequently acquired, i.e. 16% of total; of these the proportion with progressive and late onset PCHI was about 10%. Fortnum et al (2002) estimated that the prevalence of bilateral PCHI rises to 1.65/1000 by 9 years of age. More recently Watkin and Baldwin (2010) have shown a prevalence of 1.51 per 1000 (95% CI 1.11-1.92) at the end of the first year in primary school (i.e. at age 370 6 years); some of the children in this study had moved in from countries outside the UK without newborn hearing screening and four had acquired a PCHI following meningitis.

2. Main changes made in version 5.1

375 Following a major review of the evidence on yield, a set of proposals were consulted on, and then finalised (see Wood et al 2011, Davis et al 2012). Major changes were made to the categories requiring follow up with many removed. Previously some categories which had little evidence supporting the need for follow-up after the screen had been included as advisory, on a precautionary basis. It was stated in earlier versions that we would review the policy when there was more evidence: we now have more confidence that for many of 380 these commonly used risk factors there is no good evidence of a strong link to progressive or late-onset hearing loss, and thus no need for routine follow-up after a clear pass on the newborn screen. Note this does not mean there is no risk associated with these factors, and where there is parental or professional concern about hearing, a referral to Audiology should always be made.

385 The following categories were removed from the list for targeted follow up:

- Family history of permanent SNHL from childhood (in parents or siblings)
- Severe jaundice / hyperbilirubinaemia (exchange transfusion level)
- Mechanical ventilation over 5 days, or who have undergone ECMO
- 390 • Neuro-degenerative or neuro-developmental disorders

395 Congenital infections (toxoplasmosis, rubella or CMV) are still in the list requiring targeted follow up but the wording has changed from 'confirmed or suspected' to 'confirmed congenital infection'

400 ECMO is only offered to babies in two (currently) screening sites in England and the number is very small. Although ECMO has now been removed from the list of risk factors requiring targeted follow, there is still some evidence of a higher risk of progressive and late onset hearing loss in survivors (Fligor et al 2005, Murray et al 2011), but any further hearing assessment will be arranged through the local ECMO follow up protocols.

We have removed the distinction between 'mandatory' and 'non mandatory' sections

405 Specific meningitis guidance, which was previously in a separate document, has been updated and revised, and incorporated as Appendix A above. We reviewed the evidence that there might be late onset hearing loss requiring further follow up after a satisfactory hearing test. The only support found for this was a single case in 59 in Woolley et al (1999), against other studies showing no late onset (e.g. Berlow et al 1980, Richardson et al 1997). Therefore no recommendation for further follow up is made provided the hearing 410 test is clearly normal.

Ototoxics

415 a) Unless a baby is suspected or known to have the A1555G mitochondrial mutation⁹ (see below), the baby should be screened in the normal way and followed up (if required) as per the standard screening protocol. Referral to Audiology beyond this is now only at the discretion of the responsible Paediatrician.

420 To summarise, the main drugs likely to be of concern in the newborn are aminoglycoside antibiotics (e.g. gentamicin) and frusemide (see Rybak 1996, Griffin 1988, Matz 1993, NPSA 2010). Although most of the babies are on NICU/SCBU, gentamicin is also widely used with well babies in some units. High trough blood levels of gentamicin have been associated with hearing loss (Lerner et al 1986). However risks appear low for most babies so advice is that the newborn hearing screen should be carried out as per normal, and a clear result is usually sufficient. In general hearing loss attributable to ototoxicity is likely to be mild and in the higher frequencies and this is more accurately assessed by frequency-specific behavioural testing around 8 months.

430 The Paediatrician should decide whether further audiological assessment is required beyond the newborn screen, either immediately or around 8 months and to make any referral to Audiology, Clearly the monitored blood levels will be one factor in such a decision.

Also note that some drugs may act synergistically (e.g. frusemide and gentamicin together will lead to higher chance of ototoxicity than if given individually).

435 b) A small number of babies are abnormally susceptible to aminoglycoside ototoxicity. One such group is those with A1555G mitochondrial mutation who may have a family history of hearing loss from middle age. If there is such a history we urge paediatricians to test for this, be cautious and consider alternative antibiotics, otherwise there is a risk of significant hearing damage (despite satisfactory blood levels) by the time the child is seen in audiology.

3. Main changes made in versions 4.1 and 4.2

Severe unconjugated hyperbilirubinaemia occurring after the screen

445 (v4.1) The above was added as a reason for referral by the Paediatrician for Audiological assessment.

(v4.2) We clarified the distinction between the above and the same condition occurring around the time of the screen.

Syndromes associated with hearing loss .

450 (v4.1) This category was added. There are many such syndromes although many are not recognised at birth or until after a hearing loss has been found – if in doubt seek expert advice (from an Audiological Physician or Paediatrician)

Congenital infections.

455 (v4.1) Herpes has been removed as a risk factor owing to lack of evidence relating it to hearing loss.

ECMO

460 (v4.1) ECMO was added explicitly as a risk for progressive and late onset SNHL. Although the risk had been known for some time and it is a very small group of children, the data of Fligor (2005) suggest 26% of survivors might develop hearing loss.

⁹ Babies with A1555G mitochondrial mutation may have a family history of sensorineural deafness from middle age in the affected individuals and the transmission is maternally inherited. It is possible to test for this mutation.

Ototoxic drugs

465 (v4.1)

This section was revised and expanded for clarity. We made recommendations separately for babies with and without evidence of the A1555G mitochondrial mutation.

470 We advised that provided monitored blood levels remained within the defined therapeutic range, risks appeared low for most babies and a clear screen was sufficient. The suggested delay in any follow up assessment to 8 months was pragmatic, not because we believed there is a high risk of drug-related hearing loss developing.

(v4.2)

475 Further amendments were made to simplify the advice and clarify responsibility for referral. With high levels of ototoxics, referral was entirely the responsibility of the Paediatrician not the screening team. This also removed the recommendation for further follow up if the early audiology assessment was satisfactory.

480 Other

(v4.1)

The risk categories were reorganised by age at which follow-up is required.

Discussion of role of the Health Visitor distraction test screen was removed.

485 We clarified that babies with cleft lip only (normal palate) do not require audiological follow-up, but if the screener is unsure whether the palate is affected or not, follow-up should be arranged.

490 Various references were updated and other cosmetic and minor changes made.

(v4.2)

Minor typos were corrected and line numbers added

495 4. Main changes made in version 3

Bacterial Meningitis.

This was added as a reason to exclude the baby from the newborn screen, with ABR follow-up for all cases recommended. The main ground for this is that babies who have bacterial meningitis have a very high risk of having a permanent hearing loss, much higher than for any other babies on NICU/SCBU - around 1/10 (Fortnum 1992) compared to about 1/100 for other NICU/SCBU babies, and 1/1000 for well babies so that both the screen performance and positive predictive value are totally different from that for the general population. There would be a high risk of missing resulting mild/moderate or high frequency loss in a screen. If a screen were to be carried out the information given to parents would have to very different - management is best handled by Audiology. It is also the case that any testing should be after recovery and not within 48hr of diagnosis, as 10% of cases may have a reversible SNHL in this time (Richardson et al 1997). Performing a screen might also lead to a high false positive rate due to higher incidence of middle ear effusion following meningitis (Fortnum & Davis 1993). Hence we concluded a screen is not appropriate for this population, and that full assessment by ABR is essential.

Ototoxic drugs.

515 We relaxed the advice to refer for ABR assessment all children who have had ototoxic drugs where blood levels exceeded the therapeutic range, on grounds of lack of strong evidence of related late onset or progressive loss. However we left open the option for local variance at the clinical judgement of the Paediatrician, and highlighted where there is increased susceptibility to ototoxic effects. An alternative view pointed to evidence that genetic susceptibility to aminoglycoside ototoxicity might be more common than previously suspected (Tang et al 2002, Arnos et al 2003) and that the association with serum levels

520 was not that well established. We stated this area might thus need to be reconsidered in the light of any future evidence.

Chloramphenicol and ampicillin were removed from the list of ototoxic drugs.

525 **Other**

The follow-up arrangements for babies who decline the newborn screen were changed so that no follow-up was needed unless parents request it. We gave more detailed explanations and more references where appropriate.

530 **APPENDIX C**
Requirements for audiological data entry in eSP

<http://hearing.screening.nhs.uk/espreferenceguide#fileid18492>

535 **(SEE SEPARATE PDF FILE)**

REFERENCES

- Arnos KS, Xia XJ, Norris G, Landa B (2003). *Relative frequencies of the mitochondrial A1555G and 961 del T mutations in the 12SrRNA gene in a large sample of deaf probands from the United States*. Am J Hum Genet 73, S543.
- Bamford J, Fortnum H, Bristow K, Smith J, Vamvakas G, Davies L, Taylor R, Watkin P, Fonseca S, Davis A, Hind S (2007). *Current practice, accuracy, effectiveness and cost-effectiveness of the school entry hearing screen*. Health Technol Assess 11(32):1–188
- Berlow SJ, Caldarelli DD, Matz GJ, Meyer DH, Harsch GG. *Bacterial meningitis and sensorineural hearing loss: a prospective investigation*. Laryngoscope. 1980 Sep;90(9):1445-52.
- Boo NY, Oakes M, Lye MS and Said H (1994). *Risk Factors Associated with Hearing Loss in term neonates with hyperbilirubinaemia*. Journal of Tropical Paediatrics, 40(4):194-7
- Borradori C et al (1997). *Risk factors of sensorineural hearing loss in preterm infants*. Biology of the neonate 71(1), 1-10.
- CSAG (Clinical Standards Advisory Group) - Cleft Lip and/or Palate. (1998). HMSO ISBN 0 11 322 103 7. 1-119
- Davis AC, Bamford JM, Wilson I, Ramkalawan T, Forshaw M, Wright S (1997). *A Critical Review of the role of neonatal hearing screening in the detection of congenital hearing impairment*. Health Technol Assess 1(10).
- Davis AC, Wood SA, Sutton GJ (2012). *Newborn Hearing Screening Programme (NHSP): Guidance for screening providers, commissioners and public health for changes in targeted follow up procedures*. <http://hearing.screening.nhs.uk>
- Wood SA, Davis AC, Sutton GJ (2011). *Newborn Hearing Screening Programme (NHSP): recommendations for changes in targeted follow up procedures*. <http://hearing.screening.nhs.uk/audiology>
- DH (2008). Department of Health *Transforming Services for Children with Hearing Difficulty and their Families. A Good Practice Guide*. www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_088106
- Down's Syndrome Medical Interest Group (2000). *Basic medical surveillance essentials for people with Down's syndrome. Hearing impairment* www.dsmig.org.uk/library/articles/guideline-hear-8.pdf
- Fligor BJ, Neault MW, Mullen CH, Feldman HA, Jones DT (2005). *Factors associated with sensorineural hearing loss among survivors of extracorporeal membrane oxygenation therapy*. Pediatr.115(6):1519-28.
- Fortnum HM (1992). *Hearing impairment after bacterial meningitis: a review*. Arch Dis Child 67(9),1128-33.
- Fortnum HM & Davis AC (1993). *Hearing impairment in children after bacterial meningitis: incidence and resource implications*. Brit J Audiol 27, 43-52
- Fortnum HM, Summerfield AQ, Marshall DH, Davis AC, Bamford JM (2001). *Prevalence of permanent childhood hearing impairment in the United Kingdom and implications for*

universal neonatal hearing screening: questionnaire based ascertainment study. Brit Med J 323(7312), 536-40

Griffin JP (1988). *Drug Induced ototoxicity (Review).* Brit J Audiol 22, 195-210.

JCIH (US Joint Committee on Infant Hearing) (2007). *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs.* Pediatrics 120, 898-921.

Konkle DF et al (1993). *Delayed onset hearing loss in respiratory distress syndrome: case reports.* J Am Acad Audiology 4, 351-354

Lee D, Honrado C, Har-El G, Goldsmith A. (1998). *Pediatric temporal bone fractures.* Laryngoscope. 108(6), 816-21.

Lerner SA, Schmitt BA, Seligsohn R and Matz GJ. *Comparative study of ototoxicity and nephrotoxicity in patients randomly assigned to treatment with amikacin or gentamicin.* Am J Med. 1986; 80: 98-104

Mann T, Cuttler K, Campbell C (2001). *Newborn hearing screens may give a false sense of security.* J Am Acad Audiol 12, 215-219.

Matz GJ (1993). *Aminoglycoside cochlear ototoxicity.* Otolaryngol Clin North Am. Oct;26(5),705-12.

Murray M, Nield T, Larson-Tuttle C, Seri I, Friedlich P (2011). *Sensorineural hearing loss at 9-13 years of age in children with a history of neonatal extracorporeal membrane oxygenation.* Arch Dis Child Fetal Neonatal Ed. 2011 Mar;96(2):F128-32..

National Patient Safety Agency (2010). *Safer use of intravenous gentamicin for neonates.* (Alert No.1085) DH Gateway reference 13534.
www.nrls.npsa.nhs.uk/alerts/?entryid45=66271

NDCS (2000). *Quality Standards in Paediatric Audiology Volume 4.* National Deaf Children's Society, London.

NHSP (2000-2012). Test guidelines and protocols including early assessment, ABR, tympanometry, BOA, VRA and distraction test.
<http://hearing.screening.nhs.uk/audiologyprotocols>

NHSP Clinical Group (2012). *Guidelines for audiological follow up of babies diagnosed with bacterial meningitis and/or meningococcal septicaemia.* June 2012

Parker G (1999, 2006). *Children with permanent hearing impairment who pass neonatal screening,* MSC thesis, University of Manchester 1999; Unpublished data 2006

Richardson MP, Reid A, Tarlow MJ, Rudd PT (1997). *Hearing loss during bacterial meningitis.* Arch Dis Child. 76(2) 134-8

Robertson CM et al (2002). *Late onset, progressive sensorineural hearing loss after severe neonatal respiratory failure.* Otology & Neurotology 23(3), 353-6.

Rybak LP (1996). *Ototoxicity.* Current Opinion in Otolaryngology & Head and Neck Surgery.

Shapiro SM (2003). *Bilirubin toxicity in the developing nervous system. Review.* Pediatr Neurol 29(5):410-21.

Tang HY, Hutcheson E, Neill S, Drummond-Borg M, Speer M, Alford RL (2002) *Genetic susceptibility to aminoglycoside ototoxicity: how many are at risk?* Genet Med 4,336-45

Watkin PM and Baldwin M (2010). *Identifying deafness in early childhood: requirements after the newborn hearing screen.* Arch Dis Child 96 (1), 62-66.

Woolley AL, Kirk KA, Neumann AM Jr, McWilliams SM, Murray J, Freind D, Wiatrak BJ. *Risk factors for hearing loss from meningitis in children: the Children's Hospital experience.* Arch Otolaryngol Head Neck Surg. 1999 May;125(5):509-14.

Zimmerman WD, Ganzel TM, Windmill IM, Nazar GB, Phillips M (1993). *Peripheral hearing loss following head trauma in children.* Laryngoscope.103, 87-91.