Healthy Hearing Program
Universal Newborn Hearing Screening

2016

Audiology Diagnostic Assessment Protocol

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Queensland Government

The Healthy Hearing Program

Audiology Diagnostic Assessment Protocol

2016

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FOREWORD

The following diagnostic protocols represent information supported by relevant and most recent scientific literature. The broader aim is to establish a gold standard of evidence-based clinical practice in Queensland for paediatric diagnostic audiologists.

This document is the second version of the Audiology Diagnostic Assessment Protocols, and has been reviewed by the members of the Healthy Hearing Program's Audiology Working Group.

This protocol governs the diagnostic audiology services provided to children referred to audiology sites across Queensland via the Healthy Hearing Program. Specifically, the audiology services receiving Healthy Hearing referrals are:

- Attune Hearing Mackay
- Attune Hearing Rockhampton
- Caboolture Hospital
- Cairns Audiology Group
- Gold Coast University Hospital
- Healthy Hearing TeleAudiology Service
- Hear and Say
- Ipswich Hospital
- Queensland Children's Hospital
- Logan Hospital
- Mater Hospital South Brisbane
- Sunshine Coast University Hospital
- Townsville Hospital
- Toowoomba Hospital

Format:
This protocol document will now exist in an electronic form, and will be updated as needed based on decisions made at the Audiology Working Group meetings.

Review Timeframes:
This protocol document will continue to be reviewed in its entirety on a five-yearly basis by the Audiology Working Group.

ACKNOWLEDGEMENTS

The editor of this document wishes to acknowledge that the United Kingdom's NHSP's Neonatal Hearing Assessment Protocols were used, with permission, as the basis for sections of this protocol document, with amendment for local practices.

The following NHSP documents were used in the development of this protocol:

1. Stevens, J., Sutton, G. & Wood, S. (eds). (2013). Guidelines for the Early Audiological Assessment and Management of Babies Referred from the Newborn Hearing Screening Programme Version 3.1. http://www.thebsa.org.uk/resources/guidelines-early-audiological-assessment-management-babies-referred-newborn-hearing-screening-programme/
2. Sutton, G. & Lightfoot, G. (eds). (2013). Guidance for Auditory Brainstem Response Testing in Babies Version 2.1. http://www.thebsa.org.uk/wp-content/uploads/2014/08/NHSP_ABRneonate_2014.pdf
3. Stevens, J., Brennan, S. & Lightfoot, G. (eds). (2009). Provisional Guidelines for Using Auditory Steady State Responses (ASSR) in Babies. Suggested parameter values for recording frequency-specific ASSR using Natus Biologic MASTER / GSI Audera / ICS Chartr EP 200 / Interacoustics Eclipse Systems. http://abrpeerreview.co.uk/onewebmedia/NHSP%20ASSR_guidance_version_030709.pdf
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Lastly, appreciation is also extended to the staff of the Healthy Hearing Team, who have contributed to the development of this document.

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GENERAL INFORMATION

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THE HEALTHY HEARING PROGRAM AIMS AND OBJECTIVES

AIM OF THE HEALTHY HEARING PROGRAM

Early detection of permanent childhood hearing loss (PCHL), supported by early intervention, is the primary aim of the Healthy Hearing (HH) Program. Ultimately, the goal is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing and this should be measured by the proportion of these children achieving linguistic skills equivalent to their non-verbal performance IQ at 3 years of age.

PROGRAM OBJECTIVES AND PERFORMANCE INDICATORS

The HH Program operates under agreed Performance Indicators for Newborn Screening. The indicators relevant to audiology are included in the table below, with comparison to the National Performance Indicators. Please note that the HH Program Performance Indicators over-ride the National Performance Indicators where they differ.

HH Performance Indicators	**National Performance Indicators**
>95% of babies with a bilateral refer result have been offered a diagnostic assessment appointment that will occur within 2 weeks of the referral date. If the child has not reached "term" by two weeks after referral date, the assessment should occur between 38 and 40 weeks gestation.	>97% of infants with a positive screen have commenced diagnostic assessment by three months corrected age.
>95% of babies with a unilateral refer result or ETS referral have been offered a diagnostic assessment appointment that will occur within 6 weeks of referral date (but not prior to the child reaching "term").	
>95% of babies referred for diagnostic audiology assessment attend audiology.	
>95% of babies who attend for diagnostic assessment have a confirmed diagnosis by 3 months of age.	
>97% of babies diagnosed with PCHL are referred to Australian Hearing within 7 days of confirmation of the diagnosis.	>97% of babies diagnosed with a permanent hearing loss are referred to Australian Hearing.

	>85% of children diagnosed with bilateral hearing loss >40dBHL are fitted with amplification by 6 months of age.
	>95% of children diagnosed with bilateral hearing loss >40dBHL are fitted with amplification by 12 months of age.
>97% of children with a confirmed 3FAHL of ≥90dBHL are offered referral for cochlear implant candidacy assessment (upon confirmation of hearing loss).	>97% of children with 3FAHL of ≥90dBHL at the initial diagnostic audiology appointment are offered referral for cochlear implant candidacy.

Note 1: Australian Hearing aim to see babies referred for PCHL within 2 weeks of receipt of referral.

Note 2: For audiological purposes, "term" is defined as 38 weeks gestation, and assessment is not to be scheduled prior to a child reaching their "term" date, in order to allow maturity of auditory nerve function. It is recognised that "term" may be defined differently in medical / midwifery areas.

AIMS OF THE HEALTHY HEARING PROGRAM DIAGNOSTIC ASSESSMENT PROTOCOLS FOR AUDIOLOGICAL PRACTICE

1. To ensure effective, efficient and accurate diagnosis of Permanent Childhood Hearing Loss (PCHL) for children referred from the HH Program, either at birth or during surveillance at later ages.
2. To ensure consistency and equity of access to high quality diagnostic audiology services throughout the state.
3. To ensure diagnostic procedures are evidence-based and reflect best practice for diagnostic audiological assessment for neonates and infants.
4. To provide management guidelines for neonates and infants with a range of audiological profiles, including sensorineural hearing loss, transient conductive hearing loss and auditory neuropathy spectrum disorder.
5. To provide the basis for:
   - Consistent and accurate information gathering for program reporting and evaluation purposes;
   - Service Level Agreements with districts;
   - Contracting with private paediatric diagnostic audiology services;
   - Training and credentialing of all Paediatric Diagnostic Audiologists.

At present, there are no training or credentialing schemes in place to train Audiologists in the area of Paediatric Diagnostic Audiology. It is recognised that inexperienced Audiologists are unable to

practice competently in the area of neonatal / infant diagnostic audiology without significant training and clinical supervision. For this reason, Audiologists wishing to practice in the area of Paediatric Diagnostic Audiology under the auspices of the HH Program must have demonstrated a significant period (2 years) of general paediatric diagnostic audiology experience. Should an Audiologist not meet this period of practice, then they will be required to undergo a 12 month training and mentorship program with an experienced Paediatric Audiologist.

Audiology Australia is currently developing a national training course for paediatric diagnostic clinicians. It will be called a Professional Practice Certificate in Infant Diagnostic Assessment. All paediatric audiologists in Queensland undertaking a HH caseload will be required to complete this course, once available, to continue to assess HH babies/infants.

THE HEALTHY HEARING PROGRAM STATE-WIDE TEAM ROLES AND RESPONSIBILITIES

The HH state-wide team employs Audiologist/s to oversee the Audiology component of the HH Program. The role of the Audiologist/s includes:

- Protocol development, revision and implementation.
- Quality control for audiology services provided for HH babies.
- Advocating for supervision and training requirements for audiologists.
- Advice and provision of statistical data relevant to the HH caseload.
- Representing the Qld HH Program and Paediatric Diagnostic Audiology at a state-wide and national level.
- Competency tracking and monitoring.
- Auditing of Audiology sites undertaking HH diagnostic work, and making recommendations for performance/quality improvement based on audit results. This includes the reporting of the findings to the Director of Audiology / Audiologist in Charge and the Executive Director of Allied Health.
- Providing relevant information for, and assisting with, the performance management of audiologists, in conjunction with the Director of Audiology / Audiologist in Charge for the relevant site.
- Reporting and management of clinical issues identified at individual sites &/or across the state in conjunction with the Director of Audiology / Audiologist in Charge and the Executive Director of Allied Health as required.
- Provision of Tele-Audiology Services for eligible HH babies.

AUDIOLOGY CLINICAL AUDITS - QUALITY ASSURANCE

The HH state-wide team currently performs clinical audits on a 3 yearly cycle, as part of the program's commitment to providing quality audiology services across the state. Each audiology site who provides services under the HH Program participates in the audit process. The audit results are used to highlight areas for improvement, and provide the basis for further protocol development and education opportunities for diagnostic audiologists. The results also provide clinics/clinicians with data regarding their performance against the Queensland HH Program Key Performance Indicators and the National Benchmarks.

The process involves the random selection of clinical cases by the state-wide HH Team. Each site is required to collate all clinical data (including reports, raw data, progress notes) relating to the selected cases for the auditor/s to review. A standardised audit tool is used across all audiology sites based on the protocol document. Results are collated, analysed and distributed to The Director of Audiology or Clinic Manager at each site in a report format, with any recommendations for improvements also included. For Queensland Health sites, the Executive Director of Allied Health will also be provided with a copy of the audit report, as they are required to sign off the report. The sites are provided the opportunity to respond to any feedback or recommendations provided in the report.

In the interests of ensuring a fair and unbiased audit, an auditor not linked to the site being audited will be appointed by the state-wide HH team.

In addition to this regular audit cycle, HH can provide information for ad-hoc audit requests as needed by audiology clinics undertaking a HH caseload.

AUDIOLOGY REFERRAL PATHWAYS

There are two referral streams for the HH Program: Diagnostic and Targeted Surveillance.

DIAGNOSTIC REFERRALS

The aim of this referral stream is to identify hearing loss which may impact on a child's speech and language development before 3 months of age. There are two types of Diagnostic referrals – Direct Refers and Early Targeted Surveillance.

A **Direct Refer (DR)** is a referral to Audiology for a neonate who has had their hearing screened using AABR and obtained a refer result (either unilaterally or bilaterally), or have been referred directly due to factors that make them unsuitable for screening (e.g. medical exclusion for atresia/microtia, craniofacial abnormalities).

The AABR2 result is deemed to be the final screening outcome for the child. If the child refers in both ears on AABR2 (regardless of AABR1 result), the referral is classed as a bilateral refer. If the child refers in only one ear on AABR2 (regardless of AABR1 result), the referral is classed as a unilateral refer.

The only exception to this rule is a Flip-Flop result. A flip-flop occurs when a child has a pass result in one ear and a refer result in the other ear on AABR1, and then on AABR2, the initial pass ear refers and the initial refer ear passes. In this instance, the child is screened for a third time, and the AABR3 result is deemed to be the final screening outcome as indicated above.

The aim is for all ***bilateral refers*** (AABR2 = refer result bilaterally) and ***medical exclusions*** from screening to be seen for diagnostic audiological assessment **within 2 weeks** from the date of referral. If the child has not reached term (38 weeks gestation) by two weeks after referral date, the assessment should occur between 38 and 40 weeks gestation. This is provided that the infant is medically stable.

The aim is for all ***unilateral refers*** (AABR2 = refer result unilaterally) from screening to be seen for diagnostic audiological assessment **within 6 weeks** from the date of referral (but not prior to the child reaching 38 weeks gestation), provided the infant is medically stable.

An **Early Targeted Surveillance (ETS)** referral is a referral to Audiology for a neonate who has had their hearing screened using AABR and passed bilaterally (on AABR1 or AABR2) but has a craniofacial risk factor (e.g. cleft palate) or syndrome associated with hearing loss (e.g. Down Syndrome, Pierre Robin Syndrome, Goldenhar Syndrome). These children are referred for diagnostic audiological assessment due to their increased risk of having a Permanent Childhood

Hearing Loss (PCHL). The aim is for all ETS referrals to be seen for diagnostic audiological assessment **within 6 weeks** from the date of referral (but not prior to the child reaching 38 weeks gestation), provided the infant is medically stable.

DIAGNOSTIC REFERRALS: Minimum Test Battery and Pass / Discharge Criteria for Functionally Normal Hearing Bilaterally

The minimum requirements to establish functionally normal hearing in BOTH ears, and to subsequently discharge a baby from the Healthy Hearing Program are as follows:

1. Click-evoked ABR (assesses broad spectrum, but is most sensitive in the 2-4 kHz region):
   - Pass level for both ears: Wave V present at **25dBeHL**** at normal latency for age.
   - An assessment at suprathreshold level **(65dBeHL**** recommended). Wave I, III and V must be clearly identifiable with good repeatability and morphology, absolute and interpeak latencies to be within normal limits for age, with interaural latency differences of ≤0.30msec.
   - Testing at a level between 25 and 65dBeHL must also be performed (e.g. **45dBeHL**** recommended) to allow for the plotting of a wave V latency-intensity function, and to enable the audiologist to track wave V through at least 3 intensity levels to ensure confident wave V identification.

** Refer to the table below for pass levels in dBnHL "dial" dependent upon the age of the baby and transducer used for assessment.

HH Pass Criteria for AC Click ABR			
Transducer (corrected age)	**Pass Level in dBnHL "dial"**	**Correction Factor (conversion from dBnHL "dial" to dBeHL)**	**Pass Level in dBeHL**
▪ Insert Earphones (age <24 wks)	20	+5	25
▪ Insert Earphones (age >24 wks) ▪ Headphones (any age)	30	-5	25

NOTE: Different crrections must be applied to the intensity level tested before comparison to the wave V latency normative data if insert earphones have been used to assess a baby <12 weeks corrected age (refer to page 82 for further information).

2. Assessment of hearing at 1 and 4 kHz must include one of the following:

Option 1: TEOAE testing (1-1.5 and 4 kHz):

- TEOAE activity must be present at ≥6dB signal to noise ratio, with minimum TE response amplitude of 0dBSPL, with reproducibility ≥80%, in 3 or more half octave bands centred at 1, 1.5, 2, 3 and 4kHz. It is essential for the response to be present in at least the 1 or

1.5kHz band, and the 4kHz band. The overall wave reproducibility and stimulus stability must also be ≥80%), or

Option 2: Tone Burst ABR or Tone Chirp ABR:

Pass level for both ears: Wave V present at 1kHz and 4kHz at **20dBeHL** (refer to the table below for pass levels in dBnHL "dial" dependent upon transducer used).

** **Please note**: Normal TEOAE activity must be recorded from both ears before a baby can be discharged from the HH Program with functionally normal hearing. It is therefore preferable that TEOAE testing is performed as a priority over TB AC ABR testing to meet the HH pass criteria wherever possible. Babies with absent TEOAEs in the presence of peaked tympanograms are considered to be at risk for late onset/progressive hearing losses, and will require ongoing surveillance even if hearing is determined to be functionally normal on Tone Burst ABR testing.

HH Pass Criteria for AC TB ABR				
Transducer (corrected age)	**Frequency Tested**	**Pass Level in dBnHL "dial"**	**Correction Factor (conversion from dBnHL "dial" to dBeHL)**	**Pass Level in dBeHL**
▪ Insert Earphones (age <24 wks)	1kHz	30	-10	20
	4kHz	20	0	20
▪ Insert Earphones (age >24 wks) ▪ Headphones (any age)	1kHz	35	-15	20
	4kHz	30	-10	20

3. High Frequency Tympanometry (1000Hz probe tone) must also be performed.

If the above pass criteria are met, but the baby had an absent TEOAE in the presence of a peaked tympanogram in one or both ears (i.e. the baby required frequency specific testing using TB AC ABR due to the absent TEOAEs), the baby should be considered at risk for late onset or progressive hearing loss. The absence of a TEOAE in a neonate is seen as significant, and may indicate sub-clinical outer hair cell problems that are not yet evident on ABR threshold testing. The baby is deemed at the time of the assessment to have functionally normal hearing bilaterally (hearing sufficient for early speech and language development based on tone burst ABR results) BUT is to be placed on review for Targeted Surveillance testing at 9 months corrected age (refer to Targeted Surveillance Referrals on page 20).

If the above pass criteria are met, and the baby has no High Risk Indicators for late onset or progressive hearing loss (refer to page 20), the baby is deemed to have functionally normal hearing bilaterally (hearing sufficient for early speech and language development) and is discharged from the Healthy Hearing Program.

If the above pass criteria are met, but the baby has High Risk Indicators for late onset or progressive hearing loss, the baby is deemed to have functionally normal hearing bilaterally (hearing sufficient for early speech and language development), BUT is to be placed on review for Targeted Surveillance testing at 9 months corrected age (refer to Targeted Surveillance Referrals on page 20).

DIAGNOSTIC REFERRALS: Pass / Discharge Criteria Not Met (Either Unilaterally or Bilaterally)

If the pass criteria are not met, the baby must be suspected as having a hearing impairment. Further diagnostic assessment is required, along with repeat confirmation of results to reach a diagnostic outcome for the baby. Where permanent hearing loss is suspected (or has not yet been excluded), repeat testing is recommended in most cases within 2 weeks of initial assessment.

Further assessment required for the ear/s where hearing loss is suspected:

1. Click-evoked AC ABR:
   - Testing continued to determine wave V threshold.
   - Assessment at higher intensities to enable clear identification of early wave components (where possible), and to track wave V to allow for the plotting of a wave V latency-intensity function, in order to assist in site of lesion determination.
2. Cochlear Microphonic:
   - CM testing is required for an ear when an absent or grossly abnormal click ABR has been obtained at high stimulus levels, in order to exclude the possibility of ANSD.
   - Should be performed at the same level as the click ABR with the same transducer (i.e. both performed with inserts) to remove doubt regarding the stimulus level in the baby's ear canal for valid interpretation.
3. Tone Burst AC ABR at 0.5, 1, 2 and 4kHz (where previous testing indicates the degree of potential hearing loss to be mild to moderately-severe):
   - Wave V threshold to be determined for each frequency, with testing at higher intensities to allow for confident wave identification.

   Or:

   ASSR at 0.5, 1, 2 and 4kHz (where previous testing indicates the degree of potential hearing loss to be severe to profound):
   - Threshold at each frequency to be determined.
4. Tone Burst BC ABR – at frequencies required dependent upon the audiological profile obtained for the baby:
   - Wave V threshold to be determined at frequencies where hearing loss is indicated on tone burst air conduction ABR testing, to determine conductive versus sensorineural involvement.

5. Further assessment as deemed appropriate for the baby's age, including but not limited to:
    - Acoustic Reflex Testing.
    - Distortion Product Otoacoustic Emissions (DPOAEs).
    - Behavioural Observation Audiometry (BOA) – recommended to cross-check the electro-physiological profile / provide information on behavioural responses to sound.
    - Visual Reinforcement Orientation Audiometry (VROA).

Timely repeat assessment is required for confirmation of the audiological profile for the baby (i.e. replication of tests to demonstrate stability and repeatability of results over time). Confirmation of results is imperative to establish that the degree and type of loss for each ear is replicable over time, as certain conditions exist where thresholds can improve over time, for example, delayed neuromaturation and Hypoxic Ischaemic Encephalopathy.

No child should be referred on or discharged from the program until a diagnosis has been confirmed. A loss should never be confirmed on the same day as the initial appointment, as it is necessary to demonstrate that results have not changed (improved or deteriorated) over time (see Diagnostic Referral Pathway on page 19 for suggested time frames). Children with atresia or atresia/microtia who have a purely conductive hearing loss in the affected ear only do not require a confirmation appointment to be conducted, if all required testing has been completed at the first appointment.

For HH purposes, **confirmation** has been defined as the repetition of at least 2 different thresholds for <u>each ear</u> demonstrating a hearing loss. The repetition of this testing must occur on a separate test occasion, to prove repeatability & stability of the data obtained over time. The thresholds that the audiologist chooses to repeat must be thresholds that were elevated on initial testing (i.e. not considered a functionally normal result on previous testing). Examples of this may include:

- Repetition of click AC ABR and TB AC ABR at one frequency (e.g. AC Click and 1kHz tone burst).
- Repetition of TB AC ABR at two frequencies (e.g. AC TB ABR at 1 and 4kHz).

Where hearing loss is only evident at one frequency, testing must be sufficient to confirm the type and degree of hearing loss at that frequency (e.g. repetition of AC and BC TB ABR at the relevant frequency).

Adequate confirmation can also be considered to have occurred where completion of TB BC ABR testing at a subsequent appointment confirms the TB AC ABR thresholds obtained at a previous appointment (i.e. sensorineural hearing loss confirmed). Please note that given the limitations in

BC ABR intensity test levels, this would only be possible for losses of no greater than a moderate degree.

The audiologist should note that repetition of TEOAE and Tympanometry testing is required at every test appointment.

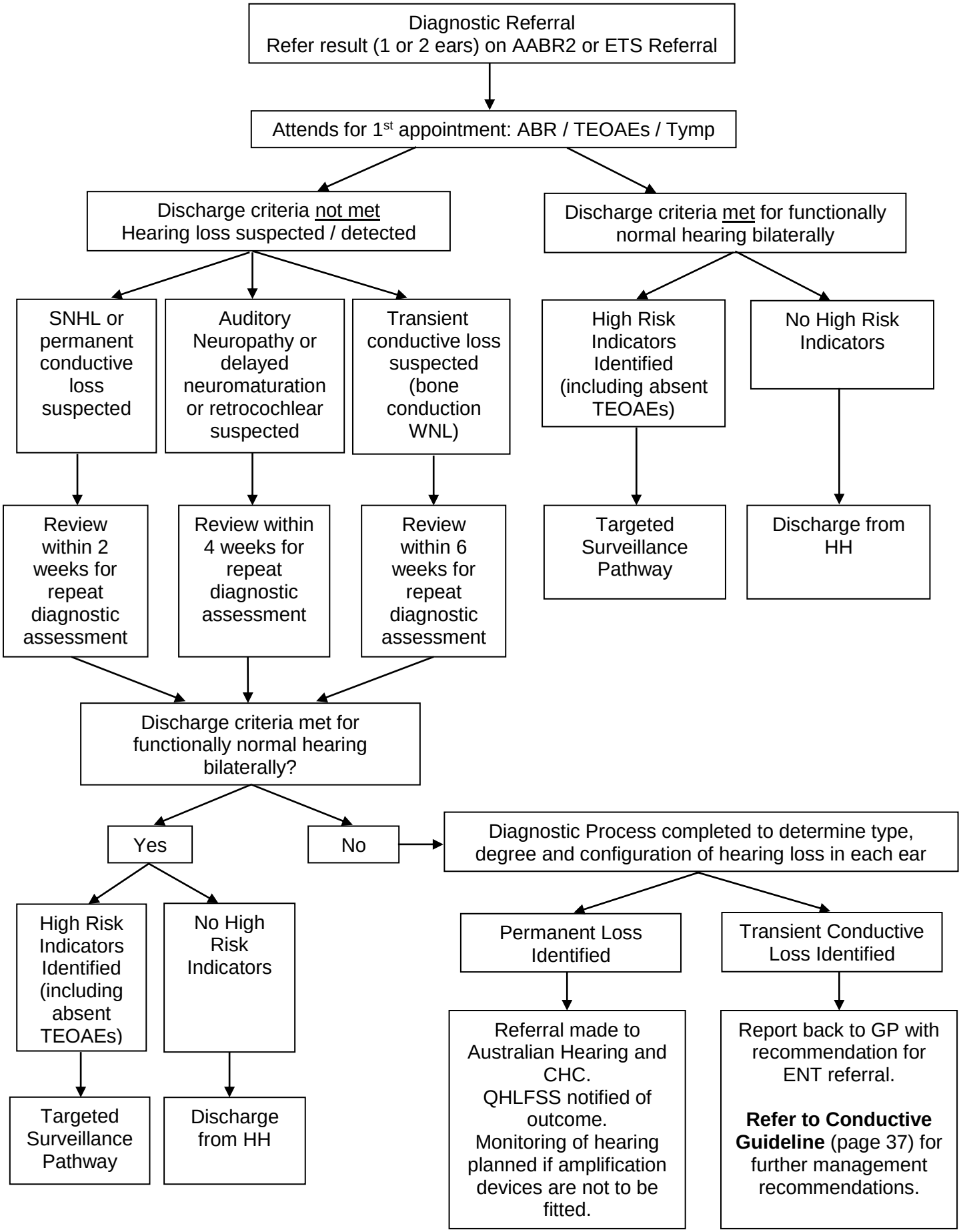
Frequency specific information is critical, as it is required prior to referral to Australian Hearing (AH) for appropriate device fitting and habilitation. While every attempt should be made to have testing completed prior to the referral to AH, it is recognised that some testing may not be able to be completed in a time frame that would not significantly delay hearing aid fitting. Some conditions negate the possibility of obtaining information across the full frequency spectrum (e.g. a poorly sleeping baby). Additional appointments should be made until as much frequency specific information can be gained to assist hearing aid fitting. If this is not possible for any reason, the clinician must document why deviation from the protocol has occurred. Where all results cannot be obtained immediately, the aim is to provide as much information as feasible to the AH audiologist to allow hearing aid fitting to occur as soon as possible.

In obtaining frequency specific information, 1 and 4kHz are considered priority, followed by 0.5kHz and finally 2kHz. These frequencies are in the most important order required for the Australian Hearing Audiologists to guide the fitting of hearing aids in neonates and young babies.

If ASSR testing is performed, the Audiologists at AH require both the “Estimated Audiogram” as well as the “ASSR Thresholds Audiogram” for the purposes of hearing aid fitting. It is the responsibility of the diagnostic audiologist testing the child to ensure that AH has the ASSR results in both formats.

All care should be undertaken to adhere to the test protocols. In extraordinary circumstances, deviation may occur. In this instance, the clinician is required to use appropriate clinical decision making before doing so, document the circumstances, and keep records regarding the deviation. Any deviation from the protocol should be included in the child’s audiology reports (not just in progress notes or internal tracking systems) so that the information is readily available to anyone accessing the results for the baby.

DIAGNOSTIC REFERRAL PATHWAY



TARGETED SURVEILLANCE REFERRALS

Targeted Surveillance referrals are made for neonates who have passed screening bilaterally, but have ***High Risk Indicators*** identified in their medical history. These children are deemed to be at risk for progressive or late onset hearing loss. The aim is for all targeted surveillance refers from screening to be seen for assessment at 9 to 12 months of age, and again at 3.5 years of age. Children referred as a Direct Refer who are found to have functionally normal hearing during diagnostic assessment, but have High Risk Indicators in their history, are also fed into the Targeted Surveillance pathway.

High Risk Indicators for Targeted Surveillance Referrals:

The following are the High Risk Indicators used by the HH Program:

- Family history of permanent childhood hearing loss (mother / father / siblings of baby only), excluding grommets, ear infection / trauma
- Syndromes associated with hearing loss
- Prolonged ventilation ≥120 hours (IPPV – Intermittent Positive Pressure Ventilation / CPAP – Continuous Positive Airway Pressure / HHFNCT – Humidified High Flow Nasal Cannula Therapy)
- Bacterial meningitis
- Severe asphyxia at birth (convulsions / HIE – hypoxic ischaemic encephalopathy / PPHN – Persistent Pulmonary Hypertension of the Newborn)
- Craniofacial anomalies e.g. Cleft palate (excludes cleft lip only and skin tags)
- Hyperbilirubinaemia levels ≥450µmol/l (Term) or ≥340µmol/l (Preterm)
- Proven/suspected congenital infection of the baby – Toxoplasmosis, Rubella, CMV, Herpes, Syphilis
- Professional / other major medical concerns
- Absent TEOAEs in the presence of peaked tympanograms during diagnostic audiology testing.

TARGETED SURVEILLANCE REFERRALS: Minimum Test Battery and Pass Criteria at 9 – 12 months

The minimum requirements for assessment of BOTH ears of an infant referred for targeted surveillance at 9-12 months are as follows:

1. TEOAE testing (1-1.5 and 4 kHz):
    - TEOAE activity must be present in both ears at ≥6dB signal to noise ratio, with a minimum TE response amplitude of 0dBSPL, with reproducibility ≥80%, in 3 or more half octave bands centred at 1, 1.5, 2, 3 and 4kHz. It is essential for the response to be present in at

least the 1 or 1.5kHz band, and the 4kHz band. The overall wave reproducibility and stimulus stability must also be ≥80%.

2. Tympanometry (226Hz probe tone) must also be performed:
   - The results must indicate no significant middle ear dysfunction at the time of testing (i.e. Type A, Ad, As or C1 are obtained for both ears).

If the above pass criteria are met, the infant is deemed to have passed this stage of surveillance testing and is to be recalled for final assessment at 3.5 years of age. If the only risk factor for the child was absent TEOAEs identified during diagnostic testing, the child can be discharged from the HH Program, based on the normal TEOAEs recorded bilaterally at this appointment.

If TEOAEs are present in conjunction with abnormal tympanometry findings, further assessment is not required until the child is 3.5 years of age, however the GP must be notified of the middle ear dysfunction identified during the assessment. Again, if the only risk factor for the child was absent TEOAEs identified during diagnostic testing, the child can be discharged from the HH Program, based on the normal TEOAEs recorded bilaterally at this appointment.

If the TEOAEs are absent (do not meet HH pass criteria), the baby must be further assessed and managed as per the Targeted Surveillance Referral Pathway outlined on page 25. The Clinical Decision-Making Matrix on page 32 can also be used to guide the initial stage of the assessment process.

Repeat assessment and/or further age appropriate diagnostic assessment must be undertaken to determine the type, degree and configuration of any loss identified in either ear:

- If further diagnostic assessment shows hearing thresholds to be within normal limits (at least at 1 and 4kHz bilaterally) with TEOAEs present on repeat assessment, then the child is to be recalled for final surveillance assessment at 3.5 years of age.
- If the TEOAEs were absent in the presence of normal hearing thresholds (at least at 1 and 4kHz bilaterally), the child's hearing is to be reviewed annually until 3.5 years of age. If TEOAEs are then present at subsequent review appointments, the child's hearing no longer requires annual review, and they are required to be recalled for final surveillance assessment at 3.5 years of age.
- If TEOAEs were unable to be obtained in the presence of normal hearing thresholds (at least at 1 and 4kHz bilaterally) e.g. child not co-operative or too noisy, and the TEOAEs were absent on previous assessment in the presence of aerated middle ears, the child's hearing is to be reviewed annually until 3.5 years of age. If TEOAEs are then present at subsequent review appointments, the child's hearing no longer requires annual review, and they are required to be recalled for final surveillance assessment at 3.5 years of age.

- If permanent hearing loss is confirmed in one or both ears, management and onward referral is as per the Diagnostic Referral Pathway on page 19.
- If transient conductive hearing loss is identified, the child is to be managed as per the Transient Conductive Hearing Loss Management Guideline outlined on pages 37-47.

TARGETED SURVEILLANCE REFERRALS: Minimum Test Battery and Pass Criteria at 3.5 years

The minimum requirements for assessment of BOTH ears of a child referred for surveillance at 3.5 years of age are as follows:

1. Threshold testing – PTA should be used in all instances where the child is developmentally able to complete testing reliably. VROA may be used for those children unable to be conditioned to respond reliably to PTA testing, so long as thresholds for both ears are obtained (i.e. with the use of inserts or headphones):
   - Thresholds of ≤20dBHL at 0.5, 1, 2 and 4kHz must be obtained for both ears.
2. Tympanometry (226Hz probe tone) must also be performed:
   - The results must indicate no *significant* middle ear dysfunction at the time of testing (i.e. Type A, Ad, As are obtained for both ears).
3. TEOAE testing (1-1.5 and 4 kHz):
   - TEOAE activity must be present in both ears at ≥6dB signal to noise ratio, with a minimum TE response amplitude of 0dBSPL, with reproducibility ≥80%, in 3 or more half octave bands centred at 1, 1.5, 2, 3 and 4kHz. It is essential for the response to be present in at least the 1 or 1.5kHz band, and the 4kHz band. The overall wave reproducibility and stimulus stability must also be ≥80%.

If the above pass criteria are met, the child can be discharged from the HH Program with normal hearing bilaterally.

If the child has normal hearing thresholds bilaterally, with middle ear dysfunction noted on tympanometry testing (that has had minimal impact on hearing thresholds given the normal hearing bilaterally), they can be discharged from the HH Program into the care of their GP for ongoing management of the middle ear dysfunction.

If the pass criteria are not met (i.e. hearing loss evident on PTA/VROA), repeat assessment and / or further age appropriate diagnostic assessment must be undertaken to determine the type, degree and configuration of any loss identified in either ear:

- If permanent hearing loss is confirmed in one or both ears, management and onward referral is as per the Diagnostic Referral Pathway on page 19.

- If transient conductive hearing loss is identified, it is to be reported back to the GP, with clinical review as deemed appropriate by the clinic. ENT referral should be recommended if the conductive dysfunction persists for more than 3 months.
- If the child has absent TEOAEs, with normal hearing thresholds bilaterally and aerated middle ears, they can be discharged from the HH Program. It is recommended that annual audiology review is offered to these children until they are at least 6 years of age, as the absence of the TEOAEs may indicate sub-clinical changes to the outer hair cells that are not yet evident using pure tone testing.

Difficult to Test Children

It is recognised that there will be some children that will not be capable of performing behavioural threshold testing (e.g. significant intellectual or physical impairment, autism or global developmental delay). Deviation from the standard protocol will therefore be required. The reason for deviation from the standard protocol above must be clearly documented in the child's medical notes.

For children unable to complete threshold testing (after at least 2 attempts at VROA +/- PTA methods) the minimum requirements for assessment of BOTH ears are as follows:

1. TEOAE testing (1-1.5 and 4 kHz):
    - TEOAE activity must be present in both ears at ≥6dB signal to noise ratio, with a minimum TE response amplitude of 0dBSPL, with reproducibility ≥80%, in 3 or more half octave bands centred at 1, 1.5, 2, 3 and 4kHz. It is essential for the response to be present in at least the 1 or 1.5kHz band, and the 4kHz band. The overall wave reproducibility and stimulus stability must also be ≥80%.
2. Tympanometry (226Hz probe tone) must also be performed:
    - The results must indicate no significant middle ear dysfunction at the time of testing (i.e. Type A, Ad, As or C1 are obtained for both ears).

If the above pass criteria are met, the child can be regarded as having functionally normal hearing up to the level of the outer hair cells bilaterally, and can be discharged from the HH Program.

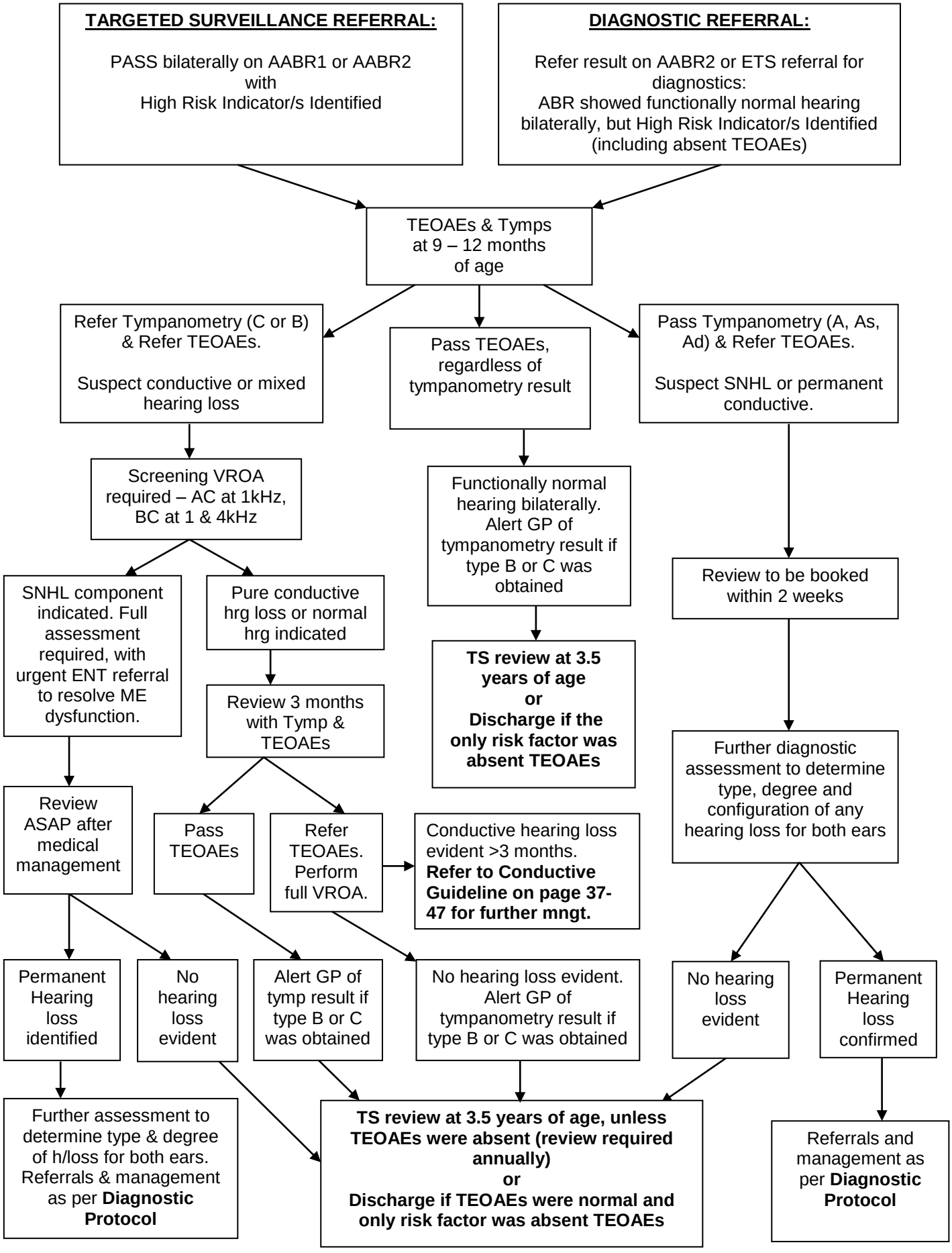
If TEOAEs are present in conjunction with abnormal tympanometry findings, further assessment is not required, however the GP must be notified of the middle ear dysfunction identified during the assessment. The child can be discharged from the HH Program.

If TEOAEs are absent, further diagnostic testing will be required to exclude the possibility of the child having a hearing loss. How this testing is to be performed needs to be decided with the

parents once the options have been discussed, including risks and benefits if testing under sedation or general anaesthetic is being considered (highly likely for this group of children).
If TEOAE testing is unsuccessful (i.e. child will not tolerate probe), and the child cannot complete VROA testing, discussions must be held with the parents regarding further testing options for their child (which may include reattempting at a later stage, objective testing under sedation or general anaesthetic, or no further assessment depending on the child's circumstances). Further testing should be carried out when parents are expressing concerns about their child's hearing, or when an audiologist suspects a hearing loss from the case history and/or behaviour of the child during attempts at testing.

All discussions with parents, including options provided and decisions made, regarding further testing must be clearly documented in the child's medical notes.

TARGETED SURVEILLANCE REFERRAL PATHWAY



QUEENSLAND HEARING LOSS FAMILY SUPPORT SERVICE

Parents of all children diagnosed with a permanent childhood hearing loss have access to the Queensland Hearing Loss Family Support Service (QHLFSS). The service consists of a state-wide team of family support facilitators who provide family-centred counselling and support to families of children diagnosed with a permanent hearing loss, until the child is six years of age.

The following services are provided:

- Support the family through the time of initial diagnosis of the hearing loss by providing counselling support, assessing the families support networks and, if required, making referrals to community agencies for ongoing support and/or assistance.
- Ensure families gain information about their child's hearing loss and the full range of habilitation options available to support their child's communication, development and health needs.
- Facilitate a process to support family decision making regarding habilitation methods and services.
- Assist in identifying the needs of children with a permanent hearing loss and their families, and advocate on their behalf within relevant services and systems.
- Contribute to the development of research and best practice in the delivery of family support services of individuals with a hearing loss.

The service is offered to all children diagnosed with a permanent hearing loss who access public or private audiology, medical and habilitation services from birth through to their 6th birthday.

An automatic referral is generated to QHLFSS for all babies who refer directly to audiology from screening (i.e. the child has obtained a 'refer' result for one or both ears). The audiologist must provide QHLFSS with copies of reports for all babies who refer directly from screening. QHLFSS will be aware of those diagnosed with Permanent Hearing Loss via this process, and will commence work with those families upon diagnosis.

There is no automatic referral generated to QHLFSS for babies who refer to audiology via Early Targeted Surveillance or Targeted Surveillance pathways, or for children who pass screening with no risk factors. If a child is diagnosed with a permanent hearing loss before 6 years of age, and was not a direct refer to audiology from screening (i.e. pass result bilaterally at birth), the audiologist must discuss referral to QHLFSS with the family at the time of diagnosis, and if the family consents, generate a manual referral to the QHLFSS. The link for the form is http://qheps.health.qld.gov.au/childrenshealth/resources/clinforms/docs/658924.pdf, and a copy can also be found in Part D of this document.

CHILDHOOD HEARING CLINIC

The Childhood Hearing Clinic (CHC) is a multi-disciplinary clinic for children diagnosed with a permanent hearing loss. The clinic provides the initial medical investigations, developmental assessment, hearing aid clearance, hearing aid fitting and opportunities for early intervention from Allied Health and other external agencies. CHCs are located at the Lady Cilento Children's Hospital (South Brisbane) and The Townsville Hospital.

These services are provided in a series of three to four appointments for babies less than 12 months of age.

The core clinical team consists of Diagnostic Audiologists, Paediatricians, ENT Specialists, Australian Hearing Audiologists, Speech Pathologists, and Queensland Hearing Loss Family Support Facilitators.

To access this service, the diagnosing audiologist must request a referral to CHC from the child's GP or other medical officer involved in the child's care. The GP is notified of the audiology results and the need for referral to CHC via fax, and the parents are also provided with an information sheet outlining the need for them to see their GP to arrange the referrals.

RESOURCES

Please refer to Part D of this document for resources and forms to assist in the diagnostic process:

REPORTING REQUIREMENTS

Audiologists should have assessment reports be completed and distributed within 7 days of the assessment. The following are the minimum reporting requirements for the HH Program. Please note that reports to the screening site and QHLFSS should only be sent until a HH Outcome has been reached.

DIRECT REFERS:

A copy of each audiology report is to be sent to:

- Screening site
- Family GP
- QHLFSS
- Parents
- Other professionals / services as requested by parents (e.g. Paediatrician)

When permanent hearing loss is suspected / confirmed, copies of all reports are to be sent to:

- CHC
- Australian Hearing

EARLY TARGETED SURVEILLANCE REFERRALS:

A copy of each audiology report is to be sent to:

- Screening site
- Family GP
- Parents
- Other professionals / services as requested by parents (e.g. Paediatrician)

When permanent hearing loss is suspected / confirmed, copies of all reports are to be sent to:

- QHLFSS**
- CHC
- Australian Hearing

TARGETED SURVEILLANCE REFERRALS:

A copy of each audiology report is to be sent to:

- Family GP
- Parents
- Other professionals / services as requested by parents (e.g. Paediatrician)

When permanent hearing loss is suspected / confirmed, copies of all reports are to be sent to:

- Screening site (only when a diagnosis has been confirmed)
- QHLFSS**
- CHC

- Australian Hearing

** Please remember that a manual referral is required to access QHLFSS if permanent hearing loss is diagnosed for children referred under the Early Targeted Surveillance or Targeted Surveillance Referral Pathways (refer to page 157 for the QHLFSS referral form).

NOTE 1:

At the completion of any test session where ABR testing was performed, all traces MUST be reviewed by a peer and co-signed regarding the agreement of interpretation. The co-signer should have a minimum of three years of current paediatric ABR experience. This process MUST occur regardless of the institutional setting and the degree of experience of the Audiologist. Should an Audiologist work as a sole practitioner, the Healthy Hearing Program will ensure that the Audiologist is linked to a peer reviewer to verify all ABR traces. Traces MUST be peer-reviewed and co-signed prior to the completion and distribution of the diagnostic report. Any issues with the traces identified by the co-signer must be discussed with the audiologist to ensure that the feedback, or any concern noted, is understood by both parties prior to interpretation of the results, to ensure accurate reporting of results is obtained.

NOTE 2:

Please be aware that Australian Hearing may apply their own correction factors to convert ABR thresholds to estimated hearing thresholds (dBeHL) suitable to be used for hearing aid fitting. It is acknowledged that correction factors may change over time as further research / data becomes available. As a result, ABR results should always be reported in dBnHL “dial” reading, with the transducer used for testing clearly reported. Any correction or calibration factors applied to the thresholds must be clearly documented in the report, including any additional correction factors not covered in this document.

NOTE 3:

It is not appropriate for audiologists to send reports to early intervention services or Advisory Visiting Teachers (AVTs) during the diagnostic and confirmation period. Audiology reports will be distributed to early intervention services by CHC, QHLFSS or Australian Hearing once the family have had an opportunity to explore all early intervention options available to them and make an informed decision for their child regarding the service they wish to access.

REPORTING TEMPLATES

There is capacity for the QChild database to generate audiology reports and letters. Please refer to the QChild 2 training manual for instructions on how to generate these.

QCHILD AND DATA ENTRY – KEY PERFORMANCE INDICATORS

The Healthy Hearing Program uses a state-wide data management system called QChild. This system is used by audiologists to record test results and audiological outcomes for every child seen for assessment via the Healthy Hearing Program. Please refer to the QChild manual for specific information relating to this database.

Key Performance Indicators for QChild:

Objective	Performance Indicator
Referrals to diagnostic audiology clinic are received and accepted within 7 days of referral date.	>95% of referrals are accepted on QChild within 7 days of referral date.
Data entry of audiology test results into QChild to occur within 7 days after each audiology appointment attended by the child.	>95% of audiology data is entered within 7 days of the appointment date.
Appointments not attended by the child (i.e. failed to attend) are recorded in QChild on the day of the scheduled appointment.	>95% of non-attended appointments are recorded in QChild on the day of the appointment.
Appointments cancelled by the parents/carer are to be recorded in QChild by the date of the scheduled appointment.	>95% of appointments cancelled by parents are recorded in QChild by the date of the scheduled appointment.

**** Note: All ABR thresholds are to be recorded in QChild in dBnHL "dial" reading (with the correct transducer selected), and in dBeHL once the correction factors have been applied in line with this protocol document.**

PART B

MANAGEMENT GUIDELINES

CLINICAL DECISION-MAKING MATRIX FOR DIAGNOSTIC TESTING OF BABIES / INFANTS

This matrix has been designed based on HF Tympanometry and TEOAE testing that is routinely conducted during the initial stages of an appointment when a baby arrives awake.

TEOAE & Tymp Result	Possible Outcomes	Testing Required
HF Tymp = Peaked TEOAE = Present 1 to 4kHz	1. Functionally normal hearing 2. Mild sensorineural hearing loss 3. Retrocochlear hearing loss / ANSD	1. Click AC ABR. 2. If AC click ABR abnormal, full diagnostic assessment to determine frequency specific thresholds (TB ABR – AC & BC). 3. CM testing not required (TEOAEs are present).
HF Tymp = Peaked TEOAE = Absent 1 to 4kHz	1. Functionally normal hearing 2. Sensorineural hearing loss (including ANSD) 3. Permanent conductive hearing loss (e.g. ossicular)	1. Click AC ABR and TB AC ABR at 1 & 4kHz. 2. If ABR not normal, perform BC ABR at appropriate frequencies. 3. Complete AC & BC testing at 0.5 & 2kHz. 4. If sev/prof SNHL loss indicated, perform CM to exclude ANSD.
HF Tymp = No peak TEOAE = Present 1 to 4kHz	1. Functionally normal hearing 2. Mild conductive dysfunction 3. Retrocochlear hearing loss / ANSD	1. Click AC ABR. 2. If AC click ABR abnormal, full diagnostic assessment to determine frequency specific thresholds (TB ABR – AC & BC). 3. CM testing not required (TEOAEs are present).
HF Tymp = No Peak TEOAE = Absent 1 to 4kHz	1. Functionally normal hearing 2. Conductive hearing loss (transient or permanent) 3. Sensorineural hearing loss (including ANSD) 4. Mixed hearing loss	1. Click AC ABR and TB AC ABR at 1 & 4kHz. 2. If AC ABR not normal, perform BC ABR at appropriate frequencies. 3. Complete AC & BC testing at 0.5 & 2kHz. 4. If sev/prof SNHL loss indicated, perform CM to exclude ANSD (when middle ear has peaked HF tymp).

CLINICAL GUIDELINE FOR THE DIAGNOSIS OF A SIGNIFICANT HEARING LOSS (PERMANENT / CHRONIC HEARING LOSS)

Aims

1. To ensure consistent information and clinical pathways are offered to parents of children diagnosed with a significant permanent hearing loss.
2. To provide effective, consistent and timely informational counselling to parents of children diagnosed with a significant permanent hearing loss.
3. To establish networks with other allied health professionals in order to best identify and meet parent and child need.

If a hearing loss is suspected following the initial appointment, follow up appointments are made to further investigate and confirm the type, degree and configuration of the hearing loss, and to obtain necessary site of lesion information.

Prior to the confirmation appointment, the Audiologist is to:

- Contact the QHLFSS and inform them of the impending diagnosis and of the need for them to contact the family.
- Plan the test requirements for the confirmation appointment to ensure the most important missing information is obtained first, and that all required testing is completed as efficiently as possible.

At confirmation of hearing loss:

At the time of diagnosis and confirmation of the hearing loss, the following information is to be consistently provided by the audiologist to the parents:

- Information about the degree of hearing loss and the implications of the hearing impairment.
- Information regarding the nature/type of hearing loss and best possible indications as to whether it is permanent.
- Information about the need for a series of events to be planned in the short term in order to assist the child to access sound for speech and language development.
- Acknowledgment to parents that it is a stressful time and that it is normal for families at such times of crisis to not remember all the information presented. Assure them that there will be other opportunities provided to talk, and that written information will be provided.
- Parents are to be advised that an appointment to attend CHC will be arranged, for their child to see an Ear, Nose and Throat Specialist (ENT), Australian Hearing and a Paediatrician. The audiologist will clarify whether the family wishes to use public ENT services or access

ENT services privately. The audiologist should remain in contact with the family to ensure progression to required medical services occurs.

- Continued contact with the family (in person or by phone) will occur on the same day or subsequent to the ENT appointment, **if appropriate,** to further discuss issues associated with the hearing loss and address any other questions that may have arisen. Wherever possible this appointment will be with the same audiologist who made the original diagnosis.
- The diagnosing audiologist will also contact the QHLFSS to assist families (should they have previously provided consent) through this stage of diagnosis and to guide the family through decision making regarding communication options and Australian Hearing appointments. Families not already receiving support from the QHLFSS service will be referred, should they consent.
- Consistent written information is to be provided to the families. In addition, the "Possibilities and Pathways" pack and Healthy Hearing DVDs should be provided to the family by the QHLFSS if they are involved. If the QHLFSS is not involved, the audiologist will need to contact QHLFSS to obtain these resources and provide them to the family directly.
- Parents are to be encouraged to write down questions to ask the Audiologist prior to the next appointment.
- Parents are to be encouraged to phone the Audiology clinic with any urgent concerns or questions.
- NOTE: Siblings of children diagnosed with PCHL without a specifically identifiable and known cause should also have their hearing assessed. It may also be worthwhile assessing the hearing of the parents of the newly diagnosed child. Referrals for these assessments will be arranged by the paediatrician during the child's CHC appointment following diagnosis. The audiologist should discuss the importance and rationale for this recommendation with the family.

Following confirmation of the hearing loss, the Audiologist is to:

- Notify the CHC co-ordinator and send the reports. The infant will see Australian Hearing and an ENT specialist as part of this clinic, unless the parents wish for the child to be seen by a private ENT specialist. QHLFSS will also see the child during CHC clinic as needed.
- Notify QHLFSS and send report.
- Notify Australian Hearing and send report to the child's closest infant fitting centre (Brisbane, Mt Gravatt or Townsville).
- Ensure both CHC and QHLFSS are copied into any correspondence regarding the arranging of appointments for the child.
- Ensure that adequate audiological information has been obtained and distributed prior to the CHC, Australian Hearing and ENT appointments, to ensure appropriate amplification and medical management occurs for the child.

- Make phone contact with the parents within 5 days following the diagnosis, to monitor the response of the parents and confirm appointment times, and document the contact in the patient's medical notes.
- Where a permanent hearing loss has been identified, and the child remains unaided (e.g. unilateral hearing loss, mild hearing loss, ANSD), the child needs to be placed on an annual review schedule for monitoring purposes until they reach 5 years of age. Annual monitoring is the responsibility of the diagnostic audiology service, unless alternative arrangements have been reached with Australian Hearing for that particular child. Australian Hearing is required to formally notify the diagnostic audiology site that amplification devices have not been fitted and that the child requires on-going monitoring (see Notification of Unaided Child Requiring Annual Monitoring Form on page 159). Where amplification devices have been fitted, Australian Hearing assumes responsibility for the clinical care of the child, including the monitoring of hearing thresholds.

RECOMMENDED PRACTICES FOR BREAKING NEWS TO PARENTS

Reference: "Working with Children with Development Disorders – A Life Course Perspective" (Presented by: Child Development Program, 15th November 2002)

Best practice in breaking news to parents of children with impairments involves consideration of the setting and structural procedures of the appointment, affective aspects of the Audiologist in breaking news and the content necessary in any information provided. The following are imperative in breaking news effectively to families:

Setting or Structural Procedures

- Give news soon after the completion of the assessment.
- Give news in person.
- Both parents are preferably present or alternatively a support person is present.
- Remember that the family have a right to privacy. Use a quiet room free from interruptions, be unhurried, allow sufficient time, and limit the number of professionals within the room.
- Have a colleague present to interpret information differently and to provide support.
- Facilitate a follow-up interview within a short space of time following the diagnosis (e.g. within 2 days).
- Provide written information.
- Be available for parents that may require repeated opportunities to discuss child/information.
- Offer contacts with support groups.

Affective Aspects

- Communication of hope is important.
- Characteristics of the Audiologist should include warmth, interest, tolerance of expression of emotion, patience, tolerance of non-acceptance of information, and a respectful attitude.

Content of Information

- Information should be clear and direct – what is known, what is not known.
- Language is simple and non-technical.
- Labels should be applied thoughtfully.
- Information should be given at the parents' pace. Ask them what they want to know.
- Acknowledgment and discussion of parental reaction should occur e.g. "How does that make you feel?"
- Consider the direction of the interview and evaluate the appointment afterwards.
- Continue to provide a supportive relationship.
- Check with parents, "Have I explained this well enough?"

Role of the Multi-Disciplinary Team

Although there will be some overlap in the communication that occurs with the family by different professionals, it is important that professionals involved only provide information directly relevant to their own area of expertise. Given the level of stress facing families in this situation, consistency and clarity is important in the messages they receive.

It is critical that any questions asked of a professional that are outside their area of expertise are referred back to the relevant professional. This can be achieved in two ways:

- By suggesting the parent ask the relevant professional directly and;
- By professionals informing the relevant other professional of the questions they were asked, so that direct follow up with the family can occur by the correct professional.

CLINICAL GUIDELINE FOR THE MANAGEMENT OF TRANSIENT CONDUCTIVE HEARING LOSS

It is recognised that the target condition of the HH Program is not transient conductive hearing loss. However, children identified with transient conductive hearing loss through the diagnostic and surveillance streams of the program need to be managed appropriately. The aim of this guideline is to provide audiologists with a clinical framework for this group of children.

There are no other existing guidelines for NHS programs to benchmark against, either internationally or within the individual states of Australia. The guideline has been developed by the HH Audiology Working Group. It is expected that the guideline outlined here will provide a framework for managing children with transient conductive dysfunction in the overall context of a Newborn Hearing Screening Program.

To be managed under this guideline, children who refer directly from screening must have had sensorineural hearing loss excluded for both ears (i.e. ear specific, frequency specific masked bone conduction results that fall within the normal range must have been established for the child).

- A child with conductive overlay who has not had underlying sensorineural hearing loss excluded must be treated as urgent until such time that functionally normal bone conduction thresholds have been established (see Diagnostic Pathway on page 19).
- A child with normal bone conduction thresholds (masked as required) in the presence of a hearing loss with peaked tympanograms (1000Hz probe tone), or Type A, As, Ad tympanograms (226Hz probe for older children), should be treated as a likely permanent conductive hearing loss until proven otherwise. The Diagnostic Pathway must be followed.
- A child with normal bone conduction thresholds (masked as required) in the presence of a hearing loss with no peaks (1000Hz probe tone), or Type B or C (226Hz probe tone for older children), can be considered to have a likely transient conductive hearing loss, unless there is evidence from prior or current testing that suggests otherwise. Caution must be exercised however when applying this label (e.g. if the child has a syndrome with possible permanent conductive hearing loss), as any transient middle ear dysfunction may mask any permanent component until it is resolved. Reassessment after medical management is necessary.
- Any moderately-severe conductive hearing losses must be considered as a likely permanent conductive hearing loss until proven otherwise, regardless of tympanometry results.

Aims:

1. To ensure appropriate and consistent medical management is sought, at appropriate time frames, for children with middle ear dysfunction and transient conductive hearing loss.

2. To minimise the risk of auditory deprivation and speech/language delays/disorders for children with ongoing (non-resolving) conductive hearing losses (similar to children with permanent hearing loss).
3. To minimise the risk of permanent hearing loss developing as a result of ongoing (non-resolving) middle ear dysfunction.
4. To provide appropriate audiological assessment while children are awaiting medical management to address the middle ear dysfunction, and after they have received medical management.
5. To ensure that permanent hearing losses are not missed, by ensuring all children referred for diagnostic assessment are not discharged until either normal hearing is established (i.e. conductive dysfunction has resolved or been managed) or a permanent hearing loss has been confirmed.
6. To ensure Australian Hearing (AH) resources are used where clinically indicated and in line with AH protocols, and not used as a solution to manage public health waiting lists for ENT services.

The management for children with transient conductive hearing loss will differ depending on their HH referral type i.e. Diagnostic referrals (Direct Refers & Early Targeted Surveillance) vs Targeted Surveillance referrals. Regardless of the referral type, it is recommended that the TEOAE results obtained for the child are used to guide the review process in the case of transient conductive hearing loss.

MANAGEMENT GUIDELINE – DIAGNOSTIC REFERRALS (DIRECT REFERS AND ETS)

When middle ear dysfunction is indicated in the presence of functionally normal hearing bilaterally (i.e. pass criteria met for functionally normal hearing on ABR and TEOAE testing), this does not necessitate the need for review. The presence of TEOAEs indicates that any middle ear dysfunction would be considered mild, despite a "no peak" result being obtained on High Frequency Tympanometry. As functionally normal hearing for both ears has been demonstrated, the child can be discharged, unless there are risk factors for surveillance. The GP, however, must be made aware of the middle ear dysfunction and the parents should be counselled accordingly.

Where hearing loss is indicated, and/or TEOAEs are absent, in the presence of middle ear dysfunction, the need for review arises. The aim is for the audiologist to build an audiological profile for the baby that will distinguish between temporary versus ongoing / non-resolving middle ear dysfunction. There needs to be a balance between monitoring of hearing and middle ear status, without needless over-testing.

The Conductive Guideline outlines management once the baby has had their hearing levels confirmed (over at least two ABR appointments), and normal bone conduction thresholds have been established and confirmed for both ears, following the referral from screening. The time frame between appointments during the initial diagnostic phase should not exceed 2 weeks, until such time that normal bone conduction thresholds have been established for both ears (i.e. until sensorineural involvement has been excluded bilaterally).

At the end of the diagnostic process, any infant with conductive hearing loss, in association with cleft lip/palate or craniofacial syndrome (with known association with middle ear dysfunction, such as Down Syndrome) should be referred to ENT as part of routine practice.

The first review, after initial diagnostic testing has been completed, is recommended at **6 months** of age. The child is assessed using TEOAEs and High Frequency Tympanometry. If middle ear dysfunction remains evident, ENT referral should be recommended to the GP.

Further audiological review at **9 months of age** is recommended, and again at **12-15 months of age** if conductive dysfunction persists at the 6 month review. Please refer to the flow chart on page 46 for the recommended management based on the results obtained at each assessment.

It should be noted that the results of the assessment at 9 months of age, and 12-15 months of age, have been deemed the critical points in determining further actions, which may include further audiology review, ENT referral, advocating for semi-urgent ENT services on behalf of the child, and referral to AH for fitting of amplification devices. The recommended actions are outlined below.

Diagnostic Referrals - Management at 9 month review

By this stage, the audiological profile will have demonstrated ongoing conductive dysfunction of up to 9 months duration. For all children at this point, ENT referral should be strongly recommended if it has not already been arranged by the child's GP (should have been recommended at the 6 month review). Phone follow up to the child's GP may be required to ensure that the ENT referral has been arranged.

Management from this point is determined based upon the child's audiological profile, and how rapidly ENT management is occurring for the child concerned. Children with moderate or greater bilateral conductive hearing loss are managed more aggressively than those children with mild or unilateral conductive hearing losses (see below). If ENT intervention (i.e. surgery, not just an ENT Consultation) is imminent, the audiologist does not need to do anything further other than keep the GP and ENT informed of the latest test results.

Moderate or greater (≥45dBHL) bilateral conductive hearing loss

The audiologist needs to:

- Strongly recommend referral to ENT for the following reasons (phone call to GP may be necessary to discuss importance):
    - For consideration of medical management of the conductive hearing loss.
    - For consideration of amplification devices.
- Provide Information to the parents regarding the following topics:
    - Conductive hearing loss and otitis media.
    - Reasons for ENT referral.
    - Prevention strategies.
    - Compensatory strategies that may be used to assist the child to hear better while awaiting medical management.
- Consider referral to AH in cases where the hearing loss is expected to be long standing for known clinical reasons (e.g. Cleft palate, Down syndrome).

If the GP has already referred the child to ENT, the audiologist should find out from the parent at this assessment which ENT service the child will be attending. If the child has been referred privately, they are not likely to have to wait for a long time for surgical intervention, where deemed appropriate. However, for children accessing the public system, the wait for an ENT appointment can be up to 12 months, if the child is assigned as a category 3 patient. It is the responsibility of the referring audiologist (or a nominated person in the workplace) to approach ENT, provide them with the child's audiology results to date, and advocate for semi-urgent ENT services (category 2) for the child, based on the degree of hearing loss and known consequences. When successful advocation occurs, upgrade to a category 2 means the child will be seen by ENT within 3 months. It is recognised that in some instances, advocation for upgrade may be unsuccessful, but it must be attempted on behalf of the child based on clinical grounds.

If the child is accessing public ENT services at a facility that the referring audiologist is not employed, it is the responsibility of the referring audiologist to contact the Audiologist in Charge at the ENT site and provide them with all clinical results and information for the child concerned. Ideally, the Audiologist in Charge at the ENT site will make arrangements for the ENT Service to be made aware of the child's audiological profile, to advocate for the child's ENT needs and assist with triage of the child's referral where appropriate.

Mild / Unilateral Conductive hearing loss:

The audiologist needs to:

- Recommend referral to ENT for consideration of medical management of the conductive hearing loss (phone call to GP may be necessary to discuss importance).

- Provide Information to the parents regarding the following topics:
    - Conductive hearing loss and otitis media.
    - Reasons for ENT referral.
    - Prevention strategies.
    - Compensatory strategies that may be used to assist the child to hear better while awaiting medical management.
- Explain to parents that further review to monitor hearing and middle ear dysfunction will occur while awaiting medical opinion.

All children with ongoing conductive hearing loss at the 9 month assessment, regardless of degree of loss, need to be recalled for review within 3 to 6 months, to ensure they are not lost to follow up whilst awaiting medical management.

Diagnostic Referrals - Management at 12-15 month review

Management at this review is not vastly different from the management at 9 months of age. For children with mild or unilateral conductive hearing losses, the management is the same as above.

For moderate bilateral conductive hearing losses, the responsibilities of the audiologist are the same. However, if ENT services were not upgraded to category 2 at the 9 month review, this should be reattempted, given further recorded evidence of significant hearing loss and middle ear dysfunction for a period in excess of 12 months. If still unsuccessful and ENT management is not imminent, referral to AH and referral for speech pathology assessment (if deemed appropriate) should be considered at this time.

Audiological review from this point should continue to occur at 6 monthly intervals, until such time that ENT management has occurred. The audiologist should make arrangements to assess the child's hearing following medical management (i.e. without overlying transient middle ear dysfunction) to determine the final diagnosis for the child. AH referral should be considered for amplification as clinically indicated.

MANAGEMENT GUIDELINES – TARGETED SURVEILLANCE REFERRALS

The first assessment under the Targeted Surveillance guidelines occurs at 9 - 12 months of age. The tests used for this assessment are Tympanometry and TEOAEs.

When middle ear dysfunction is indicated with present TEOAEs (as judged by HH pass criteria), there is no need for ongoing review of the middle ear dysfunction. The presence of

TEOAEs indicates that any middle ear dysfunction would be considered mild, despite the Tympanometry findings. The child should be placed on review for their final Targeted Surveillance Assessment at age 3.5 years. The GP, however, must be made aware of the middle ear dysfunction for them to manage as they deem appropriate, and the parents should be counselled accordingly.

When TEOAEs are absent in the presence of middle ear dysfunction, no inference can be made regarding cochlear function without further assessment. When this situation arises, some information using VROA needs to be obtained. It is recognised at the first TS appointment that time has not been allocated for this assessment to occur, hence a screening VROA is recommended. The minimum information required from the screening VROA is:

- unmasked bone conduction thresholds at 1 and 4kHz, and
- air conduction threshold at 1kHz (free-field), with additional frequencies tested if time permits.

This information will assist in determining if there is any suspicion of bilateral SNHL versus conductive hearing loss being indicated in at least one ear. If unmasked bone conduction is elevated, SNHL should be suspected bilaterally and the child would be further assessed as per the Diagnostic guidelines. Where unmasked bone conduction results are WNL, the child should be booked for review in 3 months (12-15 months of age), with time allowed for full VROA testing if needed at the review appointment. Please refer to the flow chart on page 46 for the recommended management based on the results obtained at each assessment.

Surveillance Referrals - Management at 12-15 month review

Testing at this review again commences with Tympanometry and TEOAE testing, as per the HH Targeted Surveillance protocol.

If TEOAEs are present for both ears (as judged by HH pass criteria):

- The child is deemed to have adequate hearing for ongoing development of speech and language, with no further testing required at this appointment. The child is to be placed on review for their final Targeted Surveillance Assessment at age 3.5 years.
- If middle ear dysfunction is indicated on tympanometry testing (i.e. Type B or C), the presence of TEOAEs indicates that any middle ear dysfunction would be considered mild. In this instance, the GP must be made aware of the middle ear dysfunction for them to manage as they deem appropriate, and the parents should be counselled accordingly.

If TEOAEs are absent, the audiologist must proceed to full VROA testing, to obtain as much information as possible regarding the hearing thresholds for both ears (including unmasked bone conduction results).

- Where functionally normal hearing is indicated on VROA testing for both ears, the child is deemed to have adequate hearing for ongoing development of speech and language, with no further testing required at this appointment. The child is to be placed on review for their final Targeted Surveillance Assessment at age 3.5 years. Again, the GP must be made aware of the middle ear dysfunction for them to manage as they deem appropriate, and the parents should be counselled accordingly.
- Where hearing loss is measured in the presence of type A, As or Ad tympanograms, it should be suspected as permanent (sensorineural or conductive), and the child should be further assessed as a matter of urgency as per the Diagnostic guidelines.
- Where unmasked bone conduction results are elevated, irrespective of the tympanometry results, SNHL must be suspected and the child should be further assessed as a matter of urgency as per the Diagnostic guidelines.
- Where hearing loss is indicated and unmasked bone conduction results are WNL, in the presence of Type C or B tympanograms, the child should be booked for further review in 3 months (i.e. at 15-18 months), again with time allowed for full VROA testing if needed. The consideration of ENT referral should be recommended to the GP.

Please refer to the flow chart on page 47 for the recommended management based on the results obtained at this assessment.

Surveillance Referrals - Management at 15-18 month review

Testing at this review again commences with Tympanometry and TEOAE testing, as per the HH Targeted Surveillance protocol (as for the 12-15 month review), with full VROA testing indicated if TEAOEs are absent.

The management pathway is exactly the same as outlined for the 12-15 month review, with the exception of the children where hearing loss is again indicated in the presence of normal unmasked bone conduction results and Type C or B tympanograms. For these children, non-resolving middle ear dysfunction, in the presence of hearing loss, has been demonstrated over 3 appointments spanning a period of 6 months. Management from this point is determined based upon the child's audiological profile. Children with moderate or greater bilateral conductive hearing loss are managed more aggressively than those children with mild or unilateral conductive hearing losses (see below).

Moderate or greater (≥45dBHL) bilateral conductive hearing loss

The audiologist needs to:

- Strongly recommend referral to ENT for the following reasons (phone call to GP may be necessary to discuss importance):
    - Medical management of the conductive hearing loss.
    - For consideration of amplification devices.
- Provide Information to the parents regarding the following topics:
    - Conductive hearing loss and otitis media.
    - Reasons for ENT referral.
    - Prevention strategies.
    - Compensatory strategies that may be used to assist the child to hear better while awaiting medical management.
- Consider referral to AH in cases where the hearing loss is expected to be long standing for known medical reasons (e.g. Cleft palate, Down syndrome).

If the GP has already referred the child to ENT, the audiologist should find out from the parent at this assessment which ENT service the child will be attending. If the child has been referred privately, they are not likely to have to wait for a long time for surgical intervention, where deemed appropriate. However, for children accessing the public system, the wait for an ENT appointment can be up to 12 months, and any surgical intervention another 12 months, if the child is assigned as a category 3 patient. It is the responsibility of the referring audiologist to approach ENT, provide them with the child's audiology results to date, and advocate for semi-urgent ENT services (category 2) for the child, based on the degree of hearing loss and known consequences. When successful advocation occurs, upgrade to a category 2 means the child will be seen by ENT within 3 months. It is recognised that in some instances, advocation for upgrade may be unsuccessful, but it must be attempted on behalf of the child based on clinical grounds.

If the child is accessing public ENT services at a facility that the referring audiologist is not employed, it is the responsibility of the referring audiologist to contact the Audiologist in Charge at the ENT site and provide them with all clinical results and information for the child concerned. An audiologist at the ENT site will be assigned by the Audiologist in Charge to advocate for the child's ENT services on behalf of the referring audiologist.

Mild / Unilateral Conductive hearing loss:

The audiologist needs to:

- Recommend referral to ENT for medical management of the conductive hearing loss (phone call to GP may be necessary to discuss importance).
- Provide information to the parents regarding the following topics:
    - Conductive hearing loss and otitis media.

- Reasons for ENT referral.
- Prevention strategies.
- Compensatory strategies that may be used to assist the child to hear better while awaiting medical management.

- Explain to parents that further review to monitor hearing and middle ear dysfunction will occur while awaiting medical management.

Audiological review from this point should continue to occur at 6 monthly intervals, until such time that ENT management has occurred, and/or the audiologist is able to assess hearing without overlying transient middle ear dysfunction to determine the final diagnosis for the child. AH referral should be considered for amplification as clinically indicated, especially where moderate or greater bilateral hearing loss is indicated for a period of greater than 12 months.

Please refer to the flow chart on page 46 for the recommended management based on the results obtained at this assessment.

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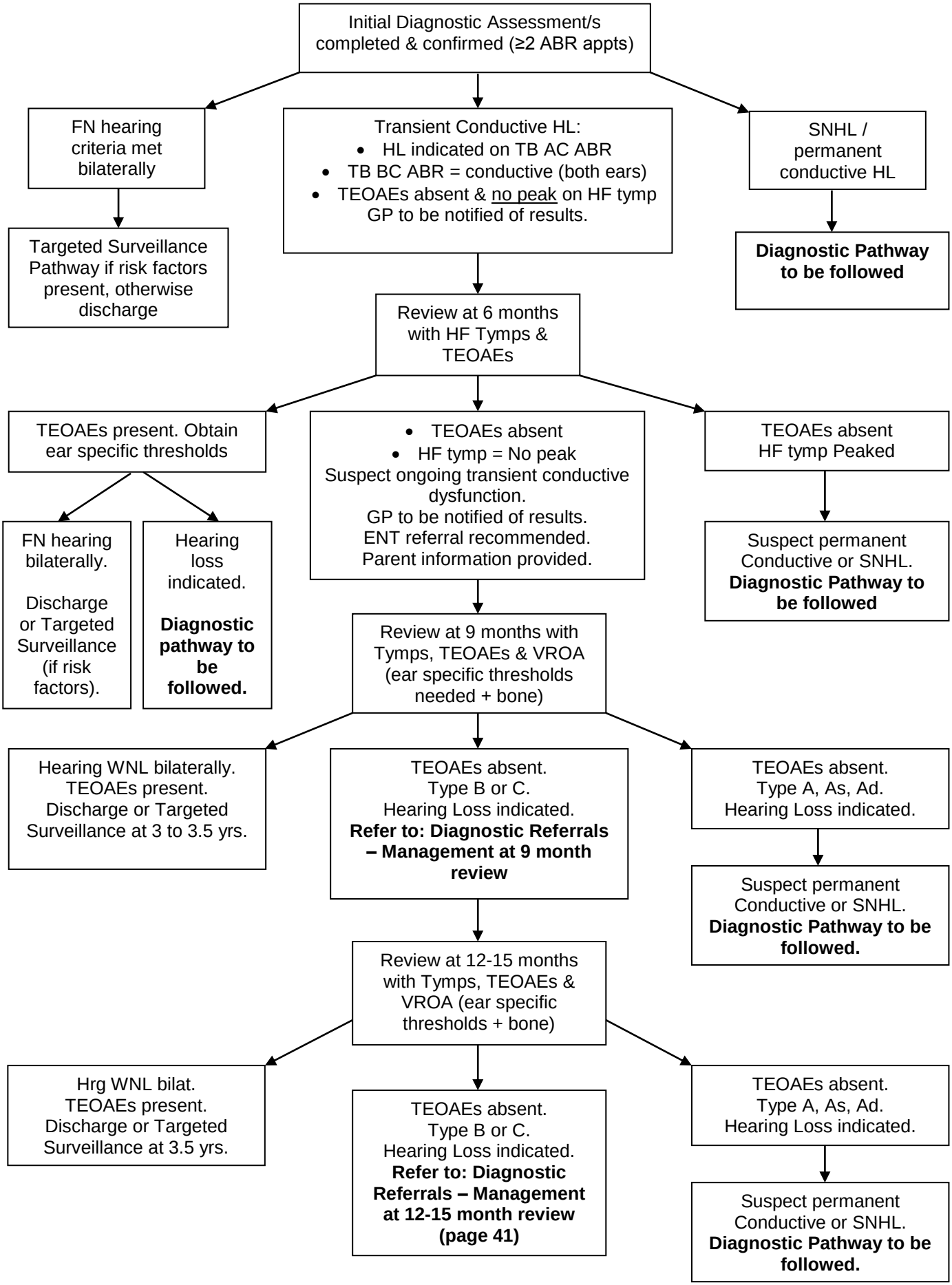
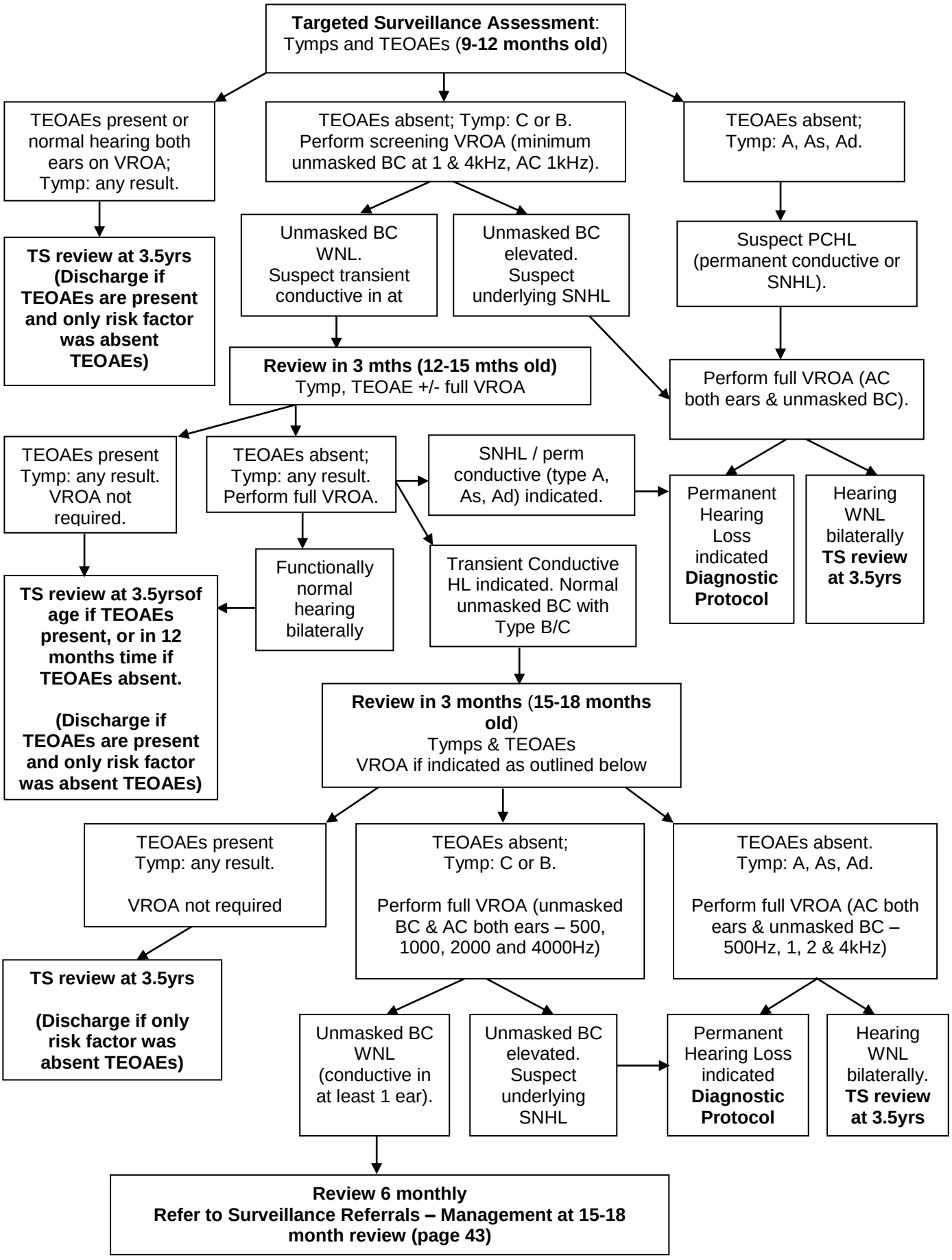
CONDUCTIVE MANAGEMENT GUIDELINE – DIAGNOSTIC REFERRALS



CONDUCTIVE MANAGEMENT GUIDELINE – TARGETED SURVEILLANCE REFERRALS



CLINICAL GUIDELINE FOR THE MANAGEMENT OF AUDITORY NEUROPATHY SPECTRUM DISORDER

This guideline specifically addresses the practical issues in the identification, assessment, diagnosis and management of infants presenting with the following pattern of results:

- Auditory Brainstem Response (ABR) absent or with severely abnormal morphology at high stimulus levels (at or above 80dBnHL "dial"), with
- Otoacoustic emissions (OAEs) and/or cochlear microphonic (CM) present.

This indicates objectively the presence of pre-neural responses but absent or abnormal neural responses.

This suggests relatively normal activity in the outer hair cells, but disruption to transmission at some point from the inner hair cells along the neural pathway to the brainstem. In some cases, the underlying reason for this pattern of test results will become evident, whereas in others the underlying reason may not be found. In some cases, neural firing may be occurring but with a lack of synchrony, so that no clear ABR is recordable. In some cases, dys-synchrony may also arise due to delayed maturation or myelination of the auditory pathway.

At the International Guidelines Development Conference (Como, Italy, 2008), consensus was reached to adopt the term 'Auditory Neuropathy Spectrum Disorder' (ANSD), to include both true auditory neuropathy (i.e. a true neural abnormality) and other possible underlying mechanisms resulting in neural dys-synchrony, as well as delayed maturation of the lower level auditory pathway (Northern, 2008). The term ANSD expresses the wide range of presentations, prognoses, and underlying aetiologies associated with the disorder.

ANSD may affect neural processing of auditory stimuli, which may reduce a child's ability to understand speech and may affect the ability to detect sound to various degrees. All such children need to be reviewed and monitored in a similar way, and their management differs from that of children with 'conventional' sensorineural or conductive hearing loss in important ways.

Risk Factors and Aetiology

ANSD is a label for a pattern of test results as defined above. It is not a diagnosis and further investigation is needed to ascertain this. ANSD may arise from a range of aetiologies. Infants with ANSD therefore require assessment, investigation and monitoring of neurodevelopmental progress by a physician with appropriate skills and an understanding of the condition. Diagnosis of the underlying aetiology may help to determine the most appropriate further management, including specific intervention if indicated.

Risk factors for ANSD from neonatal history include (but are not limited to):

- Extreme prematurity <28 weeks gestation
- Low birth weight / intrauterine growth restriction
- Hyperbilirubinaemia reaching exchange transfusion levels
- Hypoxic ischaemic encephalopathy / intraventricular haemorrhage (as is likely to occur in infants with prolonged assisted ventilation / severe sepsis)
  (Sininger, 2002; Berg et al, 2005; Madden et al, 2002; and Rance et al, 1999).

Genetic conditions that may give rise to the ANSD pattern of results include (but are not limited to):

- Otoferlin mutations (DFNB9 – autosomal recessive)
- Pejvakin mutations (DFNB59 – autosomal recessive)
- Familial delayed auditory maturation
- Neurodegenerative conditions e.g. Charcot Marie Tooth, Friedreich's Ataxia
- Metabolic conditions e.g. Maple syrup urine disease
- Mitochondrial disorders
  (Varga et al, 2003; Delmaghani et al, 2006; Aldosari et al, 2004; Starr et al, 1996; Spankovich & Lustig, 2007; Corley & Crabbe, 1999).

Some anatomical anomalies may also give rise to the ANSD pattern of results. In these instances, the case should be defined by the abnormality identified, rather than continuing to use the label ANSD. Examples include (but are not limited to):

- Hydrocephalus
- Brainstem anomalies
- Auditory nerve hypoplasia or aplasia
- Other anatomical brain anomalies e.g. Microcephaly, space-occupying lesions such as cerebellar tumours.
  (Sininger, 2002; Huang et al, 2010; Buchman et al, 2006).

Clinical Presentation and Prognosis

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

- Both ABR and behavioural thresholds are poor predictors of speech discrimination ability.
- The ABR may recover so that it is consistent with the behavioural threshold and has normal morphology (Psaromattis et al, 2006; Attias & Raveh, 2007). If the problem is due to delayed maturation, recovery would normally be complete by 12-18 months of age. It may not be helpful to continue to use the label of ANSD for these cases once maturation of the ABR has been confirmed.

- Behavioural thresholds may improve over the first 1-2 years of life (Madden et al, 2002).
- In some cases, the behavioural thresholds may appear to be satisfactory, with age-appropriate speech development, but the child may exhibit features consistent with auditory processing difficulties. There should be a local protocol for the ongoing monitoring of such cases.
- OAEs which are present at the initial assessment may disappear over time, whether or not the child is aided (Sininger, 2002; Deltenre et al, 1999; Starr et al, 2000).

Children with ANSD should be monitored carefully. We should guard against giving false hope that the condition will recover, but equally we should be careful to avoid assigning a long-term diagnosis prematurely.

When older, children with ANSD may exhibit some or all of the following features:

- Absent or elevated stapedial reflexes.
- Behavioural thresholds anywhere in the range from normal to profound, and any configuration.
- Variable responses from one test session to another, but generally reliable within a single session.
- Speech discrimination poorer than the behavioural audiogram would suggest.
- Hearing aids may be of less benefit than the behavioural audiogram would suggest.
- Greater difficulties hearing in competing noise than expected from the behavioural audiogram, and other features indicative of auditory processing difficulties.

As thresholds usually bear little relationship to speech discrimination ability, management decisions for these children should be guided much more by functional communication development rather than behavioural or ABR thresholds.

Remember that in ANSD, ABR thresholds do not predict behavioural thresholds or functional hearing ability.

Initial Assessment

As part of the assessment protocol, the click ABR will usually be the first investigation for babies with a refer result from screening. Note that with babies born prematurely, the initial ABR assessment should not be performed until the baby has reached term to allow adequate time for neural maturation. Where there is no ABR response at the normal maximum recommended stimulus levels, or a severely abnormal response at 75dBeHL or above, investigations to differentiate between ANSD and sensory (cochlear) hearing loss must be performed.

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

- Conventional hearing loss – cochlear, conductive or mixed
- ANSD due to delayed neural maturation
- ANSD due to other causes.

ANSD must always be excluded before proceeding to hearing aids on the basis of objective test results. The ANSD test protocol should be followed as part of the assessment of every suspected case of permanent hearing loss with absent / severely abnormal ABR, whether or not there are known risk factors for ANSD.

The assessment should include:

1. **ABR**: Click ABR should be performed, ideally using insert earphones, at 80-85dBnHL "dial".

2. **Tests of outer hair cell function**: If click ABR is absent or severely abnormal at 80dBnHL "dial", perform at least one of the following as appropriate:
   a. **Diagnostic TEOAEs**: If a robust TEOAE is found to be clearly present and repeatable, assume ANSD may be present. Note that TEOAEs may be absent for varying reasons, so the absence of a recordable TEOAE cannot be taken as evidence for the absence of ANSD.
   b. **Cochlear Microphonic (CM)**: Perform click CM with separate runs of condensation and rarefaction at the same stimulus level (not exceeding the recommended normal maximum stimulus level), as was used for the click ABR test, using insert earphones (80-85dBnHL "dial"). If a CM (inverting with click polarity) is present and there is still no later true neural ABR response (i.e. not inverting with click polarity), assume ANSD may be present. Testing using a "tube clamped" condition (as outlined in CM Protocol) must have been performed to confirm a true CM response.

   Note that significant overlying conductive dysfunction may prevent the CM from being detected. When abnormal tympanograms are present, it is not possible to exclude the possibility of ANSD; however in cases where there is no evidence for ANSD this should not delay the management of the child's hearing loss.

   If a robust diagnostic TEOAE has already been recorded, CM testing is not necessary (although it may be useful as confirmation). However, studies show that a substantial proportion of patients with ANSD and present CMs do not have recordable TEOAEs (Rance et al, 1999). <u>Therefore, all children with absent or severely abnormal ABR and absent TEOAE should be tested for a CM.</u>

3. **Stapedial reflexes (SR)**: For infants aged above 6 months, include if possible. SRs are invariably absent or elevated in cases of ANSD (Berlin et al, 2005).

Interpretation of Results

ABR (AC and BC) absent / severely abnormal CM or TEOAEs present (SRs absent / elevated)	⇨	Implies ANSD / Retrocochlear
ABR (AC and BC) absent / severely abnormal CM and TEOAEs not recordable (in absence of conductive dysfunction)**	⇨	Implies SNHL
ABR (AC and BC) present but elevated	⇨	Implies SNHL (or conductive or mixed) CM test not indicated

** The absence of TEOAEs and CM does not categorically rule out ANSD, but when both are absent it is reasonable to assume conventional hearing loss and proceed to manage the hearing loss on this basis. However, if robust TEOAEs and/or CM have been found to be present in one diagnostic test occasion and are not recordable at a future date, the label of ANSD should be maintained unless the ABR morphology also improves to become consistent with the behavioural thresholds. There are anecdotal reports that in some cases of ANSD, the TEOAE and/or CM can "burn out" with time. These cases are still likely to have ANSD, although they may additionally have hair cell damage, and should be managed accordingly.

ANSD vs Delayed Maturation

A key issue with ANSD is distinguishing long-term ANSD from delayed maturation, particularly in babies who have been in neonatal intensive care. Care should be taken when interpreting ABR results for babies born prematurely or for those who have delays in other aspects of development, as the ABR response may still be maturing (hence the rationale for HH protocols stating that initial ABR testing should not occur until baby has reached at least 38 weeks gestation). To help differentiate neural maturation changes from other causes of ANSD, whenever possible the ABR should be repeated before a definitive initial diagnosis is made. This should preferably occur 2 months after the first ABR.

As improvements in ABR and in behavioural thresholds over the early months of life have been reported in some infants (Madden et al, 2002; Psaromattis et al, 2006; Attias & Raveh, 2007), a further repeat ABR at a later age may be helpful in order to confirm the diagnosis. If this is felt to

be helpful for the management of the individual case, then a retest at around 12-18 months of age should be considered.

Management

This section refers primarily to the management of bilateral ANSD. However, some of this guidance will also be relevant to the management of unilateral ANSD.

The management of the child with ANSD requires a multi-disciplinary team approach, working in close partnership with the family. As a minimum, we suggest the team should include a paediatric audiologist, an ENT specialist, a paediatrician, speech pathologist, an Advisory Visiting Teacher for the Deaf / Teacher of the Deaf, and a neurologist. The timing of involvement of each of these professionals will depend on the individual case and the wishes of the family. All professionals working with a child with ANSD should be familiar with, and knowledgeable about, ANSD.

The management of ANSD presents great practical challenges. While it is feasible for all audiology clinics that perform ABR to carry out CM and OAE testing and raise the initial suspicion of ANSD, we recommend that those with little or no experience of these cases should seek advice from centres with high levels of expertise and more experience, to obtain a firm diagnosis and start the management process. Such centres need to be able to offer support and guidance in diagnosis and management, ensure that families get the best information and advice, and build confidence in the local staff.

(i) Information and support

The lack of certainty around prognosis can make ANSD a particularly difficult diagnosis for parents and families to deal with. In addition, many infants with ANSD will have other medical issues and/or have had a stormy neonatal course, meaning that hearing may not initially present as a high priority for parents (Uus et al, 2012).

The confusion and uncertainty that parents are likely to feel may have a negative impact on their relationship with professionals. Ongoing communication, support, encouragement, and information for parents are critical to successful management. It is important to provide written information regarding the condition to families, as well as to other professionals involved with the child, such as GPs and teachers. ANSD should be described specifically, including what is known and not known about the condition.

The following points should be covered with the family:

- The term ANSD is a label for a pattern of test results; it is not a label for a child.

- It is not immediately possible to predict the impact on the child, or the most successful form of treatment.
- An absent ABR does not necessarily imply a profound hearing loss.
- We need to monitor closely, as the child may not respond to sound in a typical way.
- Many children with ANSD are able to make good use of their hearing.
- However, the majority of children with this pattern of test results do turn out to have hearing problems of some degree.
- While we cannot predict the impact of ANSD on the child at this early stage, by pooling test results and observation from parents, audiologists and other professionals, we will be able to do as much for the child as possible.
- Establishing the child's early communication and language skills is important, and the use of visual cues is advisable until the child's true hearing ability is known.

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

(ii) Ongoing Audiological Assessment

The audiological profile of children with ANSD may not be stable. Therefore ongoing and regular monitoring of auditory status (behavioural, electrophysiological and middle ear) and hearing, speech, language and general development is required.

Audiological assessments must include:

- **Behavioural thresholds** in each ear determined by an age-appropriate method (VROA with inserts, Play Audiometry or conventional Pure Tone Audiometry). These can be acquired from a developmental age of around 6 months. Some children may have complex medical and developmental factors which present a challenge for behavioural testing. Such children must be assessed by suitably experienced and skilled professionals and results should be viewed in the context of the child's developmental status. If reliable results cannot be obtained because of significant developmental delay, Behavioural Observation Audiometry (BOA) and informal observation may be useful in guiding management. Any BOA should be carried out and interpreted with extreme care.

  <u>It is the responsibility of the diagnosing clinic to ensure that the infant is reviewed until initial behavioural thresholds have been established, and to continue to monitor behavioural thresholds if the child remains unaided.</u>

- **Electrophysiology:** As previously discussed, repeat ABR, CM, and OAE recording at 8-10 weeks corrected age. Consider a further repeat at 12-18 months, depending on the clinical situation of the child. A repeat ABR is strongly recommended between 12 and 18 months of age if behavioural testing consistently indicates hearing thresholds that are within normal limits in the ANSD ear/s.

- **Tympanometry / Stapedial Reflexes:** Monitoring of middle ear status is important as the presence of fluid in the middle ear will affect other tests, and children with ANSD are as likely as any other to develop middle ear effusion. This should be done in conjunction with other testing. Stapedial reflexes (SRs), using both tonal and broadband stimuli, should also be measured, particularly in infants over about 6 months of age.

The following assessments may also be valuable in the ongoing assessment of infants and children with ANSD:

- **Speech discrimination testing:** This should be attempted at as young an age as possible, including testing in noise. In addition to testing in the clinic, speech discrimination ability should also be evaluated in the child's natural environment (e.g. home, day care). This may be done in conjunction with education and/or speech pathology colleagues. Further work is needed on developing age-appropriate tools that are sophisticated enough to look in sufficient detail at early speech / language / communication development in order to inform the management of young infants with ANSD.

- **Advanced electrophysiological techniques:**

  <u>Electrocochleography (ECoG) and Electrically-Evoked ABR (eABR)</u>

  Recent studies have indicated that these tests may have a role in narrowing down site of lesion and helping determine cochlear implant candidacy. Studies using trans-tympanic ECoG have identified two abnormal potentials in some ANSD patients: an "abnormal positive potential" (enlarged summating potential) which has been interpreted as indicating a pre-synaptic disorder and may give a good CI prognosis, and a "dendritic potential" which has been interpreted as indicating a post-synaptic disorder and therefore likely to indicate a poorer CI outcome. At implantation, ANSD patients with a normal eABR have gone on to have good post-implant speech perception ability, whilst those with an abnormal eABR have had poor post-implant speech perception ability. However, these are invasive techniques which are still at a research stage and further work is needed.

  <u>Cortical Auditory Evoked Potentials (CAEPs)</u>

  There is emerging evidence that the presence or absence of auditory cortical responses in patients with ANSD might provide an insight into the degree of an individual's dys-synchrony

and whether hearing aids are likely to be helpful. The mechanism of this is unclear. Sharma et al (2011) suggest abnormal, or dys-synchronous, patterns of sub-cortical transmission, which occur in children with more disabling degrees of ANSD, have the potential to disrupt normal cortical development, and it is this abnormal cortical development that explains the failure to evoke cortical responses. Neary & Lightfoot (2012) propose that the lack of recordable cortical responses in some patients with ANSD occurs because only a modest degree of temporal dys-synchrony is required to "smear" the relatively short latency responses of the ABR but it takes a correspondingly greater degree of dys-synchrony to abolish the longer latency cortical responses. So if both the ABR and CAEPs are absent then it is reasonable to conclude that the dys-synchrony is profound and the prognosis with amplification may be poor; if the ABR is absent but the cortical responses are present the degree of dys-synchrony is likely to be modest and therefore the prognosis for amplification is better.

Thus cortical testing may have potential in informing patient management and could guide the expectations of patients and their parents. The recording of cortical responses to auditory stimuli is practical in older children and adults but the routine testing of infants is currently not yet standard clinical practice.

(iii) Monitoring and Assessment of Communication Development

Monitoring and assessment of language and communication development is the key determinant of management options. Close monitoring of communicative and developmental progress by parents and professionals together should be undertaken using appropriate monitoring tools. In addition, regular standardised assessments of language and communication should be undertaken by qualified Teachers of the Deaf, or specialist Speech / Language Therapists.

(iv) Communication Modes and Intervention

This should be determined by the needs and desires of the family, taking into account the observed progress of the child. For most children with ANSD, the use of a combination of communication systems that incorporates visual support is appropriate (e.g. Auditory/aural with lip-reading and natural gesture, total communication, sign language).

(v) Assistive Devices for Communication

Hearing aids, FM systems and Cochlear Implantation may be considered for children with ANSD:

- **Conventional Hearing Aids**

There is increasing evidence that a substantial number of children with ANSD derive benefit from hearing aid fitting if there is a significant behavioural loss. About 50% of the children in

one study gained significant benefit (Rance et al, 1999), although some clinics report much lower success rates (Berlin et al, 2010). Therefore a trial of amplification should be undertaken. However, due to doubt as to the benefit in children where behavioural thresholds are near-normal, **we would only recommend aiding for a child whose behavioural thresholds are reliably elevated**.

The decision on whether to aid must be based on behavioural results and observations from families and early interventionists regarding the child's responses to sound and early communication development. If reliable behavioural hearing thresholds are not yet available and there is significant concern from the family and early interventionist, hearing aid fitting can begin based on these concerns and BOA results (unaided and aided).

The fitting of hearing aids to children with ANSD should be based on a prescriptive method specifically developed for infants and young children. The behavioural thresholds (not ABR / electrophysiological thresholds) should be used to establish amplification targets. In order to provide the best chance of benefit, it is important that optimal audibility of speech sounds above threshold is achieved. Therefore, provided reliable behavioural thresholds are available, aids should be fitted to target based on these thresholds, rather than adopting a "conservative" approach. It is important to note that if thresholds fluctuate from test to test, the "best" threshold obtained should be used to avoid the risk of over-amplification.

Where there is a lack of reliable behavioural thresholds on which to base the prescription, a conservative approach should be adopted, beginning with low hearing aid gain and increasing the gain gradually if no responses from the child are observed.

Hearing aid benefit should be assessed. Benefit is determined primarily based on the development of speech perception skills, not on aided detection thresholds. Monitoring of the child's hearing aid fitting is essential.

Recent studies indicate the CAEPs may help to differentiate those who are able to use hearing aids effectively to understand speech (Rance et al, 2002; Cone-Wesson, 2004; Sharma et al, 2011; Cardon & Sharma, 2011). This technique has been outlined above (in section (ii) Ongoing Audiological Assessment).

- **FM Systems**

FM systems (with or without hearing aids) may be beneficial for children with ANSD who have residual speech recognition in quiet but experience difficulty in noise (Hood et al, 2004). A trial with an FM system should be considered as part of the hearing aid fitting process, particularly

when the child is involved in a day care or educational setting in which poor acoustic conditions restrict access to spoken language.

- **Cochlear Implants (CIs)**

The literature has shown increasing numbers of children with ANSD who benefit from CIs (Berlin et al, 2002; Northern, 2008; Berlin et al, 2010; Hood et al, 2004), and this option should be considered when behavioural responses indicate the child is behaving like a child with a severe/profound hearing loss, and/or when the child is not making progress with hearing aids (i.e. they show no or very limited speech discrimination abilities). A trial of conventional amplification is important prior to cochlear implantation. Behavioural thresholds are not a good guide to candidacy, and ANSD patients with even relatively mild hearing losses on behavioural testing may be CI candidates if they do not show good progress with other interventions.

Infants identified with ANSD can be referred to a CI centre for assessment as soon as there are significant concerns about behavioural responses to sound and/or concerns about communication / speech / language development. It is not appropriate to refer infants with ANSD for cochlear implantation based purely on ABR results, and generally behavioural measures will need to have been obtained. However, in view of the reports of significant improvement in auditory function in some infants with ANSD over time, the final decision to implant should not be made until audiological test results are stable and demonstrate unequivocal evidence of permanent ANSD (Northern, 2008). The exception to this is where there is a strong suspicion of a genetic cause for the ANSD known to be associated with profound deafness and good CI outcome; such cases can be referred and implanted in a similar timescale to infants with profound SNHL.

Local centres should be able to discuss cases of ANSD with a CI Team, and it is important that the CI team are able to accept referrals on the basis of assessment and advice, rather than on the assumption that they will be candidates for surgery. The approach and timescale for assessing such cases will need to be different from that for cases of conventional SNHL (except in the event of a known genetic cause as defined above), and assessment for possible delayed maturation must be carried out as part of the process of CI assessment. It is important to make clear to parents and involved professionals that the referral at this stage is for assessment and that it is not yet clear whether the child will turn out to be a suitable candidate for implantation. Responsibility for the ongoing monitoring of hearing and communication development, with appropriate modification of management strategies, also needs to be clearly agreed between the CI team and the local service.

(vi) Medical Referral

Ongoing medical / neurological assessment is essential. Some infants may already have an established neurological diagnosis. In others, ANSD identified by newborn screening may be the first indicator of an underlying neurological condition. It is therefore recommended that all children who are diagnosed with ANSD are assessed and investigated by an appropriately skilled and experienced otolaryngologist, paediatrician and/or paediatric neurologist.

Key elements that should be included in the assessment are:

- Detailed history, including family history and neonatal factors (e.g. gestation, hypoxia, assisted ventilation, hyperbilirubinaemia, intraventricular haemorrhage, hydrocephalus, convulsions).
- Examination, particularly focused on neurological and developmental assessment.
- Imaging – MRI brain and internal auditory meati to assess integrity of VIIth and VIIIth nerves.
- Ophthalmology assessment – particularly looking for evidence of visual cortical dysfunction, optic disc pathology.
- Peripheral nerve conduction studies – if generalised neuropathy is suspected.
- Referral to a geneticist, particularly if neurodegenerative condition is suspected.
- Metabolic studies as indicated by relevant clinical features (e.g. urine amino acids).

Acquired ANSD is particularly associated with neurodegenerative or metabolic aetiology and these children especially require detailed neurological assessment and investigation. Neurodevelopmental history, speech and language development, and neurological abnormalities should be noted in detail.

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

(vii) Management of Unilateral ANSD

- **Neonatal tests indicate unilateral ANSD with SNHL in the contralateral ear**

  Cases where the contralateral ear has a severe/profound hearing loss (i.e. absent/severely abnormal ABR with no evidence of good outer hair cell function) should be managed with caution, as it is possible that they may in fact be cases of bilateral ANSD. We would advise aiding of the “non-ANSD” ear, but decisions about cochlear implantation of either ear should not be made until there is unequivocal evidence of permanent profound hearing loss or ANSD on behavioural testing.

- **Unilateral ANSD with normal hearing in the contralateral ear**

  There is little consensus about the management of unilateral ANSD in young infants. The effects of unilateral SNHL are fairly well understood, but the impact on speech/language and educational progress varies between individuals. As discussed above, monitoring of hearing, communication and speech/language development are important. Children with unilateral ANSD should also receive a medical referral for aetiological investigation.

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CLINICAL GUIDELINE FOR THE USE OF SEDATION IN THE NEWBORN AND INFANT POPULATION FOR AUDIOLOGY TESTING

This guideline has been developed to assist audiologists in making clinically appropriate and considered recommendations to sedate babies and young infants for the purposes of audiological testing. The guideline is not intended to provide medical information, or replace medical guidelines, regarding the safe use of sedation in clinical settings. An appropriately trained paediatric anaesthetist should be consulted for specific medical guidelines for your individual test facility. Hospital procedures should be followed at all times when performing ABR under sedation.

The assessment of hearing in the newborn/infant population presents unique challenges, as the audiologist relies heavily on objective test measurements to estimate the child's ability to hear. While these tests do not require the co-operation of the infant, the need for the infant to sleep is critical. It is widely accepted that testing babies using ABR up until approximately 4 months (and sometimes 6 months) of age can usually be performed successfully in natural sleep state. When faced with an infant who does not sleep sufficiently for testing to proceed, the audiologist must decide whether to re-attempt in natural sleep, or make the recommendation to sedate the infant for testing. Factors to be considered include:

- the age of the child and expected sleep patterns.
- the compliance of the parents with preparation instructions for ABR appointments.
- the audiological information that has already been obtained for the baby.

The audiologist must fully understand the risks associated with sedation, particularly in the newborn / infant population, before making this recommendation to a parent.

HEALTHY HEARING SEDATION RECOMMENDATIONS

Sedation should only be considered when an infant will not sleep for electrophysiological testing. Clinical efficiency should never be a reason to sedate a child who is able to sleep naturally for testing, given the significant risks and possible adverse reactions associated with sedation. There should be strong clinical grounds for sedation to be recommended for an infant.

Given that testing under natural sleep conditions is usually possible only until the baby is around 4 months of age (sometimes older), it is critical that both initial and review ABR appointments for neonates be given priority. This gives the audiologist the best possible chance to complete testing in natural sleep conditions and avoid the need for sedation in this vulnerable population.

It is recommended that sedation not be used routinely in babies under 12 months of age for the purposes of audiological assessment. The use of sedation for infants under 12 months of age

carries significant risks (as outlined in the following Background Information section), and should therefore be considered for use in this age group in exceptional circumstances only.

The audiologist must consider what is already known about the infant's hearing in both ears, and the likely implications of delaying the testing until the child is developmentally ready to perform behavioural testing, or until the child has reached 12 months of age (when it is deemed less of a risk to sedate a child). Delaying the hearing test may be more appropriate than placing the infant under sedation, given the associated risks, in certain circumstances.

Where the delay in completion of testing is unlikely to have a detrimental effect on the child's early speech, language or communicative development, the decision to avoid sedation and delay testing should be made. Examples of this may include a unilateral hearing loss, or a confirmed hearing loss where testing at most, but not all, frequencies was completed, with sufficient information already obtained to confirm the hearing loss and allow initial hearing aid fitting. Behavioural testing in this case should be attempted as early as possible.

In cases where the audiological information is needed as a matter of clinical urgency, the recommendation to sedate may need to be made, despite the age of the child. An example of this may be an infant with a permanent bilateral hearing loss that is not sleeping sufficiently to allow full diagnostic assessment to be completed to determine site-of-lesion, or to obtain enough frequency specific information for appropriate hearing aid fitting. Parents need to be fully informed regarding the information the audiologist requires and the rationale for recommending sedation. The audiologist should also explain that the procedure carries some risk, and that this will be covered in full by the medical personnel / team admitting the child on the day. Some general information about the risks may be sought by the parents in order for them to agree to the sedation, so that the procedure can be arranged for their child.

Criteria for Recommending Testing Under Sedation for HH Infants

1. A minimum of 3 unsuccessful attempts at testing in natural sleep is recommended before making the recommendation for sedation for babies/infants.
2. A significant <u>permanent</u> (SN, retrocochlear or conductive) bilateral hearing loss, or unilateral retrocochlear hearing loss, must be indicated or strongly suspected (if unilateral or not retrocochlear, testing can be delayed).
3. If the child is less than 12 months of age and meets criteria 1 and 2 above, sedation can be recommended <u>only</u> if it can be demonstrated that a delay in testing would likely lead to detrimental outcomes for the child's speech, language or communicative development.
4. If the child is more than 12 months of age, sedation can be recommended where results have not been able to be obtained through a minimum of 2 attempts at behavioural testing.

BACKGROUND INFORMATION

Sedatives work by modulating signals within the person's central nervous system, and are typically used, for audiological purposes, to allow the completion of procedures that require little or no movement from the patient, such as ABR testing.

Sleep Patterns of Neonates and Infants

The number of hours spent sleeping per day diminishes as an infant ages. Per 24 hour period, the typical newborn sleeps 16 to 17 hours per day. This reduces to 14 to 15 hours sleep per day by the time the infant is 16 weeks of age, and to 13 to 14 hours per day by 6 months of age (Parmelee & Stern, 1972). Given this, it is critical that assessments for infants referred from newborn hearing screening are commenced as soon as practical, and follow-up assessments to confirm suspected hearing losses are treated as a priority, before this early window of opportunity for natural sleep is lost. Most audiologists report success testing babies using ABR in natural sleep conditions until about 4 months of age.

Risks and Adverse Reactions to Sedation

The risks and possible adverse reactions to sedation may differ slightly for different types of sedatives, but include the following:

- Over-sedation (to an unintended level)
- Oxygen desaturation
- Airway obstruction
- Apnoea
- Cardiovascular events (decrease in blood pressure, cardiac arrest and arrhythmias)
- Adverse drug reactions
- Paradoxical reactions and vomiting
- Escalation of required care (e.g. hospital admission, resuscitation)
- Permanent neurological injury / disability
- Death

(Cote & Wilson, 2006; Malviya et al, 1997).

Infants and children are at a higher risk for sedation related complications than adults. Several factors contribute to this, including the need for a deeper level of sedation to complete procedures, and the physiological and anatomical differences that makes them more vulnerable to respiratory depression and hypoxemia (Couloures et al, 2011; Cote et al, 2000). Age younger than 12 months and/or ASA (American Society of Anesthesiologists) physical status class III or IV (see table over page) were identified as the two factors contributing to an increased risk of respiratory arrest and sedation-related adverse events (Couloures et al, 2011; Malviya et al, 1997).

American Society of Anesthesiologists Physical Status Classification	
Class I	A normally healthy patient
Class II	A patient with mild systemic disease (e.g. controlled reactive airway disease)
Class III	A patient with severe systemic disease (e.g. a child who is actively wheezing)
Class IV	A patient with severe systemic disease that is a constant threat to life (e.g. a child with status asthmaticus)
Class V	A moribund patient who is not expected to survive without the operation (e.g. a patient with severe cardiomyopathy requiring heart transplantation)

Source: Cote & Wilson, 2006

With respect to children, the younger the child, the higher the likelihood of adverse respiratory events, especially if the child is under 12 months of age. In addition, children under 6 years of age are particularly vulnerable to the sedating medication's effects on respiratory drive, patency of the airway, and protective reflexes.

Tonsillar hypertrophy, adenoidal hypertrophy and obstructive sleep apnoea (OSA), which are all common in children, are also associated with a significantly increased risk of an adverse reaction to sedation (Cote et al, 2000). When awake, children with adenoidal hypertrophy and OSA experience no severe airway obstruction, due to better muscle tone responsible for patency of the airway, and due to compensatory mechanisms e.g. jaw opening and rocking, forward tongue position and change in head position. When sedated, children lose the compensatory strategies they use when they are awake, and have decreased muscle tone, to maintain airway patency (Biban et al, 1993). The shorter length and smaller diameter of the airway, and the smaller more collapsible bronchi may predispose children to the rapid development of respiratory problems while sedated. In addition, the shorter neck, softer larynx, proportionately larger tongue and immature intercostal and abdominal muscles in children may lead to additional difficulties if respiratory problems arise (Egelhoff et al, 1997). It is critical that the head is positioned in slight hyperextension or a neutral position (not in flexion) during sedation to prevent airway compromise in children (Egelhoff et al, 1997).

No sedation procedure is "risk-free", and this needs to be considered when making recommendations to sedate a child for audiological testing.

Avoiding Sedation – Appropriate Patient Preparation

Appropriate preparation prior to the baby arriving at the appointment is critical to the success of testing neonates in natural sleep conditions. The ideal preparation is to have a baby arrive at the appointment hungry and tired, ready for a feed and a sleep. Time needs to be allocated to allow

the baby to feed and fall asleep naturally, ready for testing to commence. It is imperative that parents understand these instructions. This may involve a phone call to the family, or having instructions translated for the family if they are of a non-English speaking background.

Please refer to the ABR Preparation Instructions for Parents included in Part D on page154.

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ADMINISTRATIVE GUIDELINE – MANAGEMENT OF 'UNABLE TO CONTACT', 'LOST TO FOLLOW-UP' AND 'FAILURE TO ATTEND'

UNABLE TO CONTACT / LOST TO FOLLOW UP

The HH Screening and Referral form includes at least 4 different contact options for each baby referred to audiology. Each different method of contact should be attempted to make contact with the family to minimise the lost-to-follow-up rate for children requiring diagnostic audiology services. When having difficulty reaching a family to schedule an appointment, the audiology site must attempt to contact the family in the following ways:

- Contact parents/carers via phone (home and mobile numbers as supplied).
- Send letter to parents/carers by mail to the last known address requesting them to contact the audiology site (not just the address listed on the S&R form – check local databases for most recent address).
- Contact the "alternative contact/s" listed on the HH referral form using phone numbers supplied.
- Contact the GP to check if new contact details are known.
- Child Safety must also be contacted if their involvement with the child is documented on the S&R form, or known by the audiology site.

Each attempted contact should be clearly documented in the child's record.

Once all avenues of contact have been exhausted and the family cannot be contacted, the baby can be discharged from the Healthy Hearing Program. A letter advising of the discharge needs to be sent to:

- The parents / carers (even though it may be returned to sender, it should still be attempted)
- The family's General Practitioner
- The Healthy Hearing screening site
- Child Safety if applicable (if their involvement with the child is documented on the S&R form, or known by the audiology site).

The letter should advise of the failure to attend or unable to contact / lost-to-follow-up status. The letter should contain information advising that the baby has been discharged from the Healthy Hearing Program, the implications of an undiagnosed hearing impairment, any results obtained to date, and the need for a GP referral for any further hearing assessment at a later date.

The 'Audiology Status' and 'Healthy Hearing Program Outcome' needs to be updated in the QChild database upon discharge.

FAILURE TO ATTEND INITIAL AUDIOLOGY APPOINTMENT

If a family fails to attend their initial diagnostic audiology appointment/s, every effort should be made to contact the family to discuss the urgency of the appointment and to encourage them to reschedule the appointment. The impact / implications of an undiagnosed hearing impairment on speech and language development should be thoroughly discussed.

The family may choose (and has the right to choose), to decline the appointment, despite the refer result from screening and subsequent discussions with an Audiologist. If a family declines audiology services, a letter advising the decline of the service should be sent to:

- The family
- The family's General Practitioner
- The Healthy Hearing screening site
- Child Safety (only if they are listed on the S&R form as a contact for the child).

The letter should contain information advising that audiology testing has been declined, the baby has been discharged from the Healthy Hearing Program, the implications of an undiagnosed hearing impairment, and that the baby will require a GP referral for any further hearing assessment at a later date.

The 'Audiology Status' and 'Healthy Hearing Program Outcome' needs to be updated in the QChild database upon discharge.

FAILURE TO ATTEND SUBSEQUENT AUDIOLOGY APPOINTMENTS WHERE PCHL IS SUSPECTED

If a family attends a diagnostic appointment where a permanent hearing loss is suspected, and the family then fails to attend subsequent appointments, every effort should be made to contact the family to discuss the urgency of the appointment and to encourage them to reschedule the appointment. The impact / implications of a permanent hearing impairment on speech and language development should be thoroughly discussed.

Unilateral PCHL suspected

If the family declines further audiology services and the loss is **unilateral** (and normal speech and language development is likely), a letter should be sent to:

- The family
- The family's General Practitioner
- The Healthy Hearing screening site
- Child Safety (only if they are listed on the S&R form as a contact for the child).

The letter should advise of the decline of further audiology appointments despite a suspected permanent unilateral hearing impairment. The letter should contain information advising that the baby has been discharged from the Healthy Hearing Program, the implications of a suspected unilateral hearing impairment, and that the baby will require a GP referral for any further hearing assessment at a later date.

The 'Audiology Status' and 'Healthy Hearing Program Outcome' needs to be updated in the QChild database upon discharge.

Bilateral PCHL suspected

If the family declines further audiology services and the baby is suspected of having a **permanent bilateral** hearing impairment, every effort should be made to contact the family to discuss the urgency of the appointment and to encourage them to reschedule the appointment. The impact / implications of a permanent hearing impairment on speech and language development should be thoroughly discussed. The GP and the QHLFSF should be contacted in person to discuss the possibility of encouraging and supporting the family to attend the appointment.

If the family declines further audiology despite these attempts, a letter needs to be sent to:

- The family
- The family's General Practitioner
- The Healthy Hearing screening site
- Child Safety (only if they are listed on the S&R form as a contact for the child).

The letter should advise of the decline of further audiology appointments despite a suspected permanent bilateral hearing impairment. The letter should contain information advising that the baby has been discharged from the Healthy Hearing Program, the implications of a permanent bilateral hearing impairment, and that the baby will require a GP referral for any further hearing assessment at a later date.

In addition, the Child Advocacy Service (CAS) should be contacted to discuss the case, and to lodge a Child Safety Notification (if advised to do so by CAS). A suspected permanent bilateral hearing loss has the potential to significantly affect the child's speech and language development, and access to communication. Every child has the right to access a form of communication, and as such, discussion of the case with the Child Advocacy Service, and notification to the Department of Child Safety if advised, is imperative.

All conversations with the family, GP, QHLFSF and Child Advocacy Service need to be clearly documented in the child's record. The outcome / decisions / management plans made as a result of these discussions should also be clearly recorded.

The 'Audiology Status' and 'Healthy Hearing Program Outcome' needs to be updated in the QChild database upon discharge.

PART C

GENERAL TESTING PRECAUTIONS AND CONSIDERATIONS

TEST SPECIFIC PROTOCOLS

GENERAL TESTING PRECAUTIONS AND CONSIDERATIONS

INFECTION CONTROL

At all times, local procedures should be adhered to, including the handling of infections such as MRSA. Testers should wash their hands thoroughly before and after handling the baby and make use of alcohol gel as prescribed by local infection control procedures.

ABR / ASSR Equipment

Suggested minimum practice is that all equipment other than the electrodes, preamplifier and earphone should remain outside the baby's cot or, when the baby is not in a cot, should be placed at a distance from the baby. Any item in the cot that is likely to come into contact with the baby should be cleaned, using a suitable method, before being used on another baby.

The earphones, bone conductor and electrode leads should be cleaned with soap and water or swabbed down with alcohol gel. It is recommended that local advice is sought regarding infection control methods.

Tympanometers

It is recommended that local advice be sought regarding best practices for prevention of cross-infection.

Electrodes

Disposable electrode tabs should be used at all times, as this minimises contact of the electrode leads with the baby and minimises cross-infection risks.

CONSIDERATIONS IN ELECTROPHYSIOLOGICAL TESTING

Test Environment

Ideally, electrophysiological testing should be performed in a sound proofed room or environment which meets the same standards as used in pure tone audiometry. In addition, levels of electrical interference (e.g. 50Hz mains) should be sufficiently low in the test environment such that the signal baseline is not adversely affected. Clinics should not be sited close to potential sources of interference such as high powered mains equipment, transformers, or plant equipment.

In some circumstances ABR testing is performed outside the designated clinic area, for example, on the ward or operating room. In this situation, levels of acoustical and electrical interference must be sufficiently low so as not to influence the results of the test. Careful selection of the local test area or room may be necessary in order to achieve satisfactory environmental conditions.

Electrode Impedances

The amplitude of any artefact resulting from induced electrical interference is proportional to the difference in the electrode impedances. A low value for the difference in impedances is most easily obtained by ensuring all electrodes have low impedances. The impedance, as measured between each electrode pair, should be in the range 1000 to 5000ohms. Although in good recording conditions and in a screened room higher electrode impedances can be tolerated, it has been found that in a Special Care Baby Unit, values below 5000ohms are normally necessary to reduce mains (50Hz) interference to an acceptable level. At high stimulus levels, a high electrode impedance would also give an unacceptably large stimulus artefact, which would obscure the initial part of the response and may activate the data rejection facility so that no data are averaged.

Transducers

Earphones should be of sufficient quality to deliver a stimulus up to 140dBSPL peak (about 107dBnHL for a click stimulus) without distortion. TDH39/49 headphones or insert earphones (e.g. type EAR-3A) are suitable. Headphones should be centred over the ear canal ensuring that the ear canal is not collapsed due to excess pressure. Insert earphones may be used, however the actual stimulus level is more uncertain due to greater variation in the enclosed volume of a baby's ear canal. The use of insert earphones reduces the need for masking. If insert earphones are used, care should be exercised that wax is not compacted by the probe to avoid blocking the sound pathway. Care should also be taken to ensure that the method of insertion is standardised (e.g. 3mm marked on the insert tubing).

Bone conductors (e.g. Radioear B-71) should be able to deliver a stimulus up to 60dBnHL (50dBnHL at 500Hz) without obvious waveform distortion. Stimulus levels should not exceed these values unless the bone conductor has been passed in calibration as being able to deliver higher levels without distortion.

Insert Earphones and Correction Factors (Intensity)

It is recognised that evidence exists to suggest that insert earphones can deliver effective stimulus levels up to 10dB to 20dB higher in the neonatal ear. As a result, the HH Program has adopted different "pass" levels dependent upon whether the infant has been assessed using headphones or insert earphones. Due to the differing pass levels for different transducers, there is no longer a need to correct for intensity in determining if a baby has reached pass levels for functionally normal hearing (this has been accounted for in the new "pass" levels). Correction factors for converting dBnHL "dial" to dBeHL for reporting and interpretation purposes have been adopted by the HH Program, and these correction factors take into account the possible intensity differences delivered by different transducers.

Correction Factors for Converting dBnHL "dial" to dBeHL

There is much debate in the literature as to how best estimate hearing thresholds of neonates from ABR data. It has been decided for HH purposes that ABR thresholds will be reported in two formats – dBnHL "dial" with the transducer used clearly indicated (i.e. the raw data with no correction factors applied for any purpose), and dBeHL (estimated hearing level). The correction factors to convert from dBnHL "dial" to dBeHL have been adopted from the NHS protocols, with some adjustment for local preference.

A decision was made to use only one set of correction factors for insert earphones and bone conduction testing for babies under 24 weeks of age. The strictest correction factors from the NHS data have therefore been adopted to ensure that babies with hearing losses are not missed as a result of this decision. These corrections factors are consistent with those used by other Newborn Hearing Screening programs around the world.

Correction Factors for Conversion from dBnHL "dial" to dBeHL (to be added to the dBnHL "dial" reading)					
Transducer (corrected age)	**Click**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Insert earphones (<24 weeks)	+5	-15	-10	-5	0
Insert earphones (>24 weeks)	-5	-20	-15	-10	-10
Headphones (any age)					
Bone Conductor (<24 weeks)		+5	+5	-5	0
Bone Conductor (24 weeks to 2 years)		-5	-5	-10	-10

Comparison of ABR Thresholds Obtained Using Insert Earphones to ABR Normative Data & Plotting of Wave V Latency Intensity Functions

At present, the HH Program compares click ABR wave latencies to normative data published by Cox (1985). Cox's data applies to ABR testing performed using headphones. There is a lack of normative data for ABR testing using insert earphones in the literature. As a result, correction factors are required to convert the test level using inserts to a comparable "estimate" of intensity for headphone testing, so that a comparison to normative data can be performed. It is recognised that it would be preferable to have separate normative data for headphone and insert earphone testing, but at this stage, that is not possible.

Where insert earphones have been used to assess a baby aged less than 12 weeks of age, a **10dB correction factor** for INTENSITY must be applied (added) to the dBnHL "dial" reading to correct for the effect of the smaller ear canal volume in babies of this age. Hence, if performing a click-evoked ABR and a threshold of 20dBnHL "dial" is obtained using insert earphones, that threshold is to be corrected to 30dBnHL (20 + 10 = 30) for comparison to normative data only.

Where insert earphones have been used to assess a baby older than 12 weeks of age, no correction of the dBnHL "dial" reading is necessary (as the effect is considered negligible by this age), and direct comparison with the Cox (1985) ABR normative data can be made.

No correction for LATENCY needs to be made for the use of insert earphones, as the delay in wave latencies (0.8msec) is already compensated for by the ABR equipment in commercial use.

When plotting the wave V latency-intensity function for babies tested using inserts less than 12 weeks of age, the +10dB correction needs to be added for each intensity level assessed.

Safe Testing Levels for Insert Earphones

If insert earphones are used for testing babies, the recommendation is that they should not be used above 85dBnHL "dial" unless they include a microphone to automatically adjust the stimulus level for ear canal volume. The reasons for this are that a baby has a much smaller ear canal which can give up to a 20dB rise in the stimulus level compared to the same insert earphone used in an adult (Voss & Herrman, 2005; Rance & Tomlin, 2006; Sininger et al, 1997), and the depth of insertion is also uncertain. This figure is thought to diminish over the early months of life as the ear canal grows.

There are occasions when testing at higher levels are either desirable or necessary (i.e. a child presenting with a severe to profound sensorineural hearing loss). The audiologist is permitted to test above 85dBnHL "dial", only after outer hair cell function has been conclusively established, using TEOAEs and CM testing if needed. **If TEOAEs and/or a CM are present, testing must not exceed 85dBnHL "dial" under any circumstances for babies less than 3 months of age**. Only when TEOAEs and CM are **both** absent, can testing at higher levels proceed (i.e. the audiologist has conclusively determined that there is no outer hair cell activity present).

Masking

As with pure tone audiometry, masking of the non-test ear is required in certain circumstances where the stimulus level is high enough to cross to the other cochlea and produce a response. If masking is not used a cross shadow response may mislead as to the true threshold. A masking calculator spreadsheet has been designed by Guy Lightfoot to decide when masking is needed and to calculate the level of masking noise required. It is available to download using the following link:http://webarchive.nationalarchives.gov.uk/20150408175925/http://hearing.screening.nhs.uk/audiology. It is located in the Audiological Assessment Guidelines section on this NHS webpage.

General masking rules are outlined below for air and bone conduction testing. Testing above the levels mentioned does not mean that masking will be needed, but indicates that the masking calculator should be used to check if masking is or is not required.

Air conduction testing (Clicks and Tone Burst/Pips): As a general rule and assuming the non-test ear cochlea is normal, then masking should be considered for stimulus levels above 65dBnHL (both headphones and insert earphones).

Bone conduction testing: As a general rule and assuming the non-test ear cochlea is normal, then masking should be considered for stimulus levels above 15dBeHL.

Reporting of Degree of Hearing Loss

To ensure consistent reporting of the degree of hearing loss across all diagnostic sites performing HH assessments, hearing losses measured on ABR testing are to be reported in line with the following criteria, based on the estimated hearing level (dBeHL).

Reporting of Degree of Hearing Loss for HH Purposes (TB AC ABR)	
Pass Level (20dBeHL)	Functionally normal
25-40dBeHL	Mild
45-55dBeHL	Moderate
60-70dBeHL	Moderately-severe
75-90dBeHL	Severe
≥95dBeHL	Profound

DEFINITION OF ABR THRESHOLD FOR HEALTHY HEARING

The ABR threshold has been defined as the lowest level at which a clear wave V response is present, with the absence of a recordable response at a level 5 or 10dB below the threshold, obtained under good recording conditions. Independent auditing of the results should not give thresholds that are more than 10dB different from those originally recorded.

For click ABR, wave latencies will be compared to the normative data established by Cox (1985). Wave latencies that fall outside the normative data should be reported as abnormal. Furthermore, the inter-aural latency difference should be no greater than 0.3ms.

Criteria for accepting the presence of a response and establishing the ABR threshold

The primary method of establishing the presence or absence of a response is visual interpretation. Replication of responses (as defined below) is therefore essential if a correct visual interpretation is to be made. Single recordings at one stimulus level are only acceptable at high suprathreshold

levels and providing that they form part of an intensity series of recordings. The only other exception to this should be where the result is not used in the determination of threshold. For example, if the first stimulus level is 45dBeHL and a flat trace is obtained then the best use of time may be not to replicate until a response is observed at higher stimulus levels and it is clear which levels need to be replicated to determine threshold.

Threshold of the response must be determined from an intensity series of traces, using stimulus levels both above and below the estimated threshold. This enables a reliable and robust interpretation of response threshold. Where hearing loss is suspected / indicated, it is recommended that replicated responses are obtained using increments of 10dB, at minimum stimulus levels 10dB below threshold, at threshold, and at 10dB and 20dB above threshold.

The exception to this is where a clear response is obtained at the recommended Healthy Hearing pass level for air conduction testing only. In this situation, testing below 25dBeHL for click AC ABR and 20dBeHL for TB AC ABR is not required.

Results should be clearly marked as to whether they indicate a threshold or the lowest level tested. It should also be made clear when a threshold measurement was not possible due to the limits of maximum stimulus output for the equipment used.

For each stimulus level the result should be marked / reported in one of three ways:

(a) Clear response present (i.e. there must be a high degree of correlation between the replications, the waveform should show the expected characteristics in terms of amplitude, latency and morphology, and the waveforms should be compared with those at other stimulus levels to confirm that they follow the expected changes with stimulus level).
(b) Response absent - good recording conditions but no response was recordable (i.e. the replications must be low in amplitude (peak to peak below about 0.05μV) with no significant correlation).
(c) Inconclusive - recording conditions too poor to establish a result (i.e. the replications will have amplitudes above about 0.05μV peak to peak without a high degree of correlation).

Where there is evidence of a possible response after two replications at a stimulus level, further replication may assist in determining whether a true response is present or not. For example, two further replications could be carried out and the replications could be added in pairs to produce two summed traces, each the average of a higher number of sweeps. In any decision, all recorded traces at each stimulus level should be considered and only those rejected where there is a good technical reason or where the noise levels (e.g. electro-myogenic activity) were high. Summed traces should be labelled appropriately.

A third replication should be carried out if there is any uncertainty in the result. If the results at this level are not absolutely unambiguous, a further pair of recordings should be obtained at a level 10dB higher to limit any possible error to 10dB.

Each threshold measurement should continue until there is a very high degree of confidence, with any inconclusive results being resolved.

It is important to note that since the tone burst ABR is generally less well-defined than the click ABR, it is essential to have good recording conditions with a quiet signal baseline. Even with these conditions met, it may necessary to use 3 or even 4 replications at one stimulus level in order to reach a definitive and reliable interpretation, particularly with stimulus levels close to threshold of the response.

Results should clearly be marked as to whether they are threshold or the lowest level tested. It should also be made clear in the results when threshold measurement was not possible due to the maximum stimulus limits of the equipment. A useful convention is to use '=' for threshold, '≤' for lowest level tested (where the threshold may be lower) and '>' for threshold above maximum level tested. For example, '≤35dBnHL' would mean a clear response at 35dBnHL but not tested below this level and '>80dBnHL' would mean no response at 80dBnHL but not tested above this level.

Independent auditing of the results should not give thresholds that are more than 10dB different from those originally recorded. The method of measuring threshold should be stated i.e. whether it is the lowest clear response or an interpolated value.

RECORDING SYSTEM CHECKS

The equipment should be checked at regular intervals (monthly is recommended) for system artefacts. This applies to all protocols for auditory evoked potentials. Using the normal protocol for testing, connect the electrodes together. Run the normal protocol twice and check that a flat trace is obtained with a minimal level of residual system noise (peak-to-peak below 0.05μV) and no correlation between repetitions.

At regular intervals (monthly is recommended), when testing a baby, carry out an additional control recording to check that there are no artefacts in the recording. Do this on a baby where a clear ABR response has been obtained. Block the sound from reaching the ear and repeat the test with the stimulus still on. This can be achieved by covering the earphone or by sealing the insert earphone tube (ensure that the sound is not audible). A flat trace should be obtained with a minimal level of residual EEG (peak-to-peak below 0.05μV for a quiet baby) and no correlation between repetitions.

CALIBRATION

A 'Stage A' check should be carried out on the ERA equipment prior to use. Details of this are available below. ISO 389-6 (2007) provides reference equivalent threshold sound pressure levels (RETSPL) for click and tone burst stimuli used for AC ABR for certain types of transducer. It also provides a standard for reference equivalent threshold force levels (RETFL) for use for click BC ABR for the B71 type bone vibrator. However there are no RETFLs for tone burst BC ABR. A procedure for calibration is given in IEC 60645-3 (2007). In July 2005, a provisional set of reference levels for AC and BC ABR was agreed for the NHSP: and the latest version of these figures is published on the NHSP website with the ISO389-6 values used where appropriate (NHSP ABR calibration specification and ABR calibration data. Newborn Hearing Screening Programme (England). http://hearing.screening.nhs.uk/protocols_calibration). These values should be used to calibrate equipment used for hearing assessment. It is important to note that the stimulus rate will affect the psycho-acoustic threshold but not the ABR threshold (Lightfoot et al, 2007). When performing a subjective (Stage A) listening check therefore, the use of the recommended stimulus rate near 50 per second will make the clicks sound about 3dB too loud. Bear this in mind or perform the listening check at a stimulus rate of 20 per second. It is important that the physical intensity of the stimulus does not change as the stimulus repetition rate is changed since this can introduce errors. This is difficult to detect subjectively and NHSP can advise which instruments require users to apply a correction for this problem.

CHECKLIST FOR DAILY & MONTHLY FUNCTION CHECK OF ABR SYSTEMS (Stage A Check)

Based on B700/701, BS EN ISO 8253-1:1998

1. Clean equipment and examine for damage or wear. Check headphones, bone conductor, insert earphones and leads for signs of damage.
2. * Switch on & adjust according to handbook.
3. * Earphone serial numbers or marking match with equipment.
4. * Check battery state, if appropriate.
5. * Electrode impedance test correct with dummy load.
6. * # Threshold levels of stimuli to be used are subjectively correct for:
   a) Air conduction (for all transducers used).
   b) Bone conduction.
7. * High level (max 80dBnHL) listening test with stimuli to be used satisfactory by:-
   a) Air conduction (for all transducers used).
   b) Bone conduction.
   c) Masking (including insert).
8. Attenuator sweep subjectively satisfactory.
9. Noise, hum and break-through levels are adequately low.

10. Radiated noise from instrument is acceptable at the patient's position.
11. Headbands are in good condition and tensions subjectively correct.
12. Amplifier: select calibration mode (or loop test mode), run test and check averaged waveform is of expected amplitude and morphology.
13. Connect amplifier inputs together, run test and check that the noise floor meets equipment specifications.
14. Check test parameters against the relevant departmental / NHSP protocol.
15. * Reset all controls to normal operating positions for commencement of patient testing.

** Tests marked with an asterisk are recommended for checking at the start of a session when the equipment is used; other checks may be performed at monthly intervals. Additionally, it is vital that all checks are conducted prior to and following objective calibration and whenever the user has reason to question the correct function or adjustment of the system.*

 # Threshold levels may be tested at the rate employed in the ABR test but note that in theory, these levels are correct only when a rate of 20/s is used in a subjective listening check. If the stimuli appear too loud, repeat the check at a rate of 20/s.

Ongoing vigilance: Whenever an elevated ABR threshold is recorded, check that the stimulus is being delivered at the expected level. Monitor waveforms recorded in babies with normal ABR thresholds and report / investigate any unexpected artefacts.

1. CLICK-EVOKED AIR CONDUCTION ABR TEST PROTOCOL

SCOPE

This protocol aims to provide guidelines for testing babies in the first few months of life by ABR using air conducted (AC) click stimuli for the purposes of assessing hearing sensitivity. The document covers the technical procedure of carrying out a click-evoked air conduction ABR test and the reporting of the results. This protocol refers to diagnostic ABR (i.e. non-automated) ABR. Any changes in this protocol should be performed with careful consideration and with appropriate clinical justification.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR CLICK AC ABR

Electrode Location:	One Channel: • Positive electrode: High forehead/Vertex • Negative electrode: Ipsilateral mastoid • Common electrode: Contralateral mastoid Two Channel: • Negative electrodes: Ipsilateral mastoid and contralateral mastoid. • Positive electrode: High forehead as near to Cz as possible, avoiding the fontanelle. • Common electrode: Forehead. Contralateral electrodes will be used in the two channel recording.
Stimulus:	Alternating polarity click Rate: 11.1/s, 22.1/s, up to a maximum of 33.1/s+ Stimulus presentation level: 2 traces required at 65, 45 and 25dBeHL as a minimum for normal discharge. Clinical threshold seeking required if loss is suspected.
Transducer:	TDH39/49 preferably EAR-3A insert earphones may be used
Coupler value for 0dBnHL:	Refer to NHSP calibration data
Amplifier reject levels:	±5 to ±10μV
Filters:	Low frequency: 30Hz High frequency: 1500Hz
Window length:	15ms
Number sweeps averaged per replication :	Minimum 1500 sweeps Typically 2000 sweeps
Display:	0.1μV on amplitude axis = 1 – 2ms on time axis
HEALTHY HEARING PASS CRITERIA:	Wave V present down to 25dBeHL

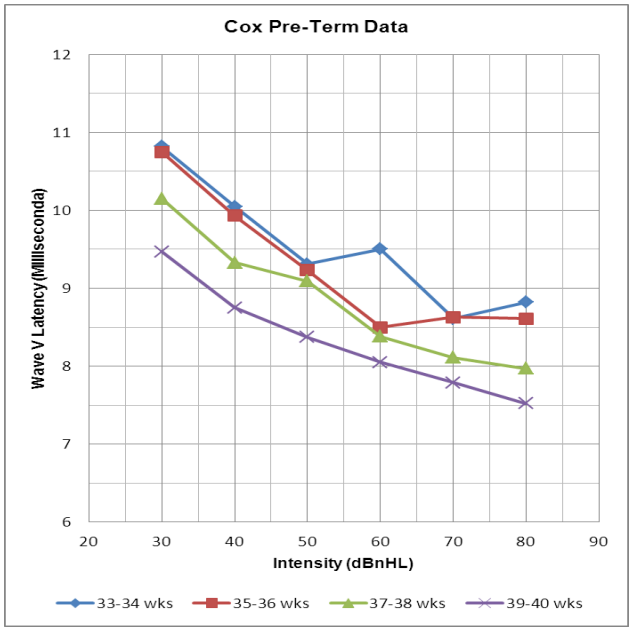
+ Refer to section on stimulus and stimulus rate for further information.

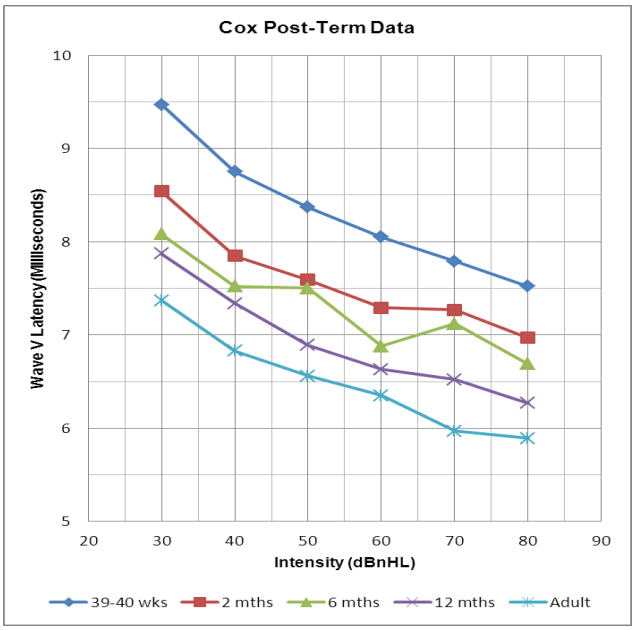
HEALTHY HEARING CLICK-EVOKED AC ABR QUICK SUMMARY

- Both left and right ears must be tested, regardless of whether the infant has a bilateral or unilateral refer result from screening.
- Testing is to be carried out at 65, 45 and 25dBeHL, with at least 2 traces at each test level required, as a minimum for both ears. Clinical judgement is required if waves are not identifiable at these levels. Should a hearing impairment be suspected, then clinical threshold seeking is required, with a minimum of two replications required at threshold.
- A child is deemed to have passed this test if a repeatable wave V is seen down to 25dBeHL at normal latency for age.
- Major wave components (I, III and V) should be present at supra-threshold levels (65dBeHL or higher as necessary).
- A latency-intensity function for the absolute latency of wave V is to be plotted and compared to age appropriate norms (derived from Cox, 1985). **Please note that if inserts have been used to test a baby less than 12 weeks of age, a 10dB correction factor must be added to the dBnHL "dial" reading for comparison to ABR normative data and for plotting of the wave V latency-intensity function (to correct for the increase in intensity in the neonatal ear associated with insert use).**
- The central transmission time at supra-threshold level (i.e. at 65dBeHL) should be compared to age appropriate norms, and between ears for symmetry (significant asymmetry = inter-aural latency difference of > 0.3ms).

NORMATIVE DATA

The paediatric norms below represent upper limits of normal for wave V in each population (i.e. mean latency + 2SD). Data was derived from Cox (1985).





33 – 34 Weeks CA – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	3.88	6.49	8.82	6.47
70	4.27	6.80	8.61	6.40
60	4.45	7.06	9.50	6.51
50	4.97	7.07	9.31	6.44
40	5.53	7.43	10.05	6.48
30	6.43	8.52	10.82	6.67

35 – 36 Weeks CA – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	3.51	5.89	8.15	6.18
70	3.69	6.07	8.63	6.36
60	4.15	6.59	8.50	5.99
50	4.20	7.40	9.23	6.47
40	5.35	7.42	9.71	6.40
30	5.96	8.22	10.75	6.46

37 – 38 Weeks CA – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	3.08	5.55	7.97	5.95
70	3.58	5.57	8.11	6.01
60	4.25	5.92	8.38	6.12
50	4.22	6.89	9.09	6.07
40	4.78	6.93	9.33	6.15
30	5.86	7.87	10.15	6.41

39 – 40 Weeks CA – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	2.97	5.50	7.52	6.11
70	2.96	5.45	7.79	5.81
60	3.54	5.95	8.05	5.71
50	4.09	6.19	8.37	5.86
40	4.80	6.84	8.75	5.91
30	5.21	7.47	9.47	5.82

2 months – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	2.50	4.82	6.97	5.47
70	2.41	5.09	7.27	5.36
60	2.98	5.34	7.29	5.23
50	3.23	5.59	7.59	5.30
40	3.54	6.08	7.85	5.43
30	4.35	6.42	8.54	5.51

6 months – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	2.12	4.72	6.69	5.23
70	2.43	4.94	7.12	5.23
60	2.55	5.26	6.88	5.01
50	3.10	5.49	7.50	5.12
40	3.45	5.67	7.52	5.23
30	4.08	6.08	8.08	5.18

12 months – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	2.10	4.15	6.27	4.69
70	2.27	4.30	6.52	4.79
60	2.49	4.45	6.63	4.90
50	2.70	5.07	6.89	4.83
40	3.23	5.17	7.34	4.93
30	3.78	6.13	7.87	5.13

Adult – Mean + 2SDs				
dBnHL	I	III	V	I-V
80	2.07	3.98	5.89	4.48
70	2.17	3.32	5.97	4.36
60	2.42	4.51	6.35	4.37
50	2.91	4.85	6.56	4.37
40	3.08	5.07	6.83	4.13
30	3.55	5.62	7.37	4.46

COMPREHENSIVE PROTOCOL INFORMATION

Electrode Location

One channel or two channel recording can be used.

In *single channel recording*, electrodes are located as follows:

- Negative electrode: ipsilateral mastoid. Use a low mastoid position unless wishing to record the post-auricular myogenic response.
- Positive electrode: high forehead as near as possible to Cz[a] and midline. The fontanelle should be avoided but the electrode should be placed as close as possible to this otherwise the ABR response will be reduced in size.
- Common electrode: contralateral mastoid.

The above electrode configuration should result in wave V being plotted upwards on the display. If this is not the case then the positive and negative electrode connections should be reversed.

In *two channel recording*, electrodes are located as follows:

- Negative electrodes: ipsilateral mastoid and contralateral mastoid.
- Positive electrode: high forehead as near to Cz as possible, avoiding the fontanelle.
- Common electrode: forehead.
- Contralateral electrodes will be used in the two channel recording.

Stimulus and Stimulus Rate

An electrical click of 100µs at a stimulus rate of around 22.1 per second is recommended, with alternating polarity to minimise stimulus artefact. Stimulus rates between 11.1 and 33.1 per second have been deemed acceptable for Healthy Hearing purposes, however it is recommended that the click rate of 33.1 per second (where it is used as default) is abandoned in favour of the slower suggested rates (22.1 or 11.1 per second) when wave morphology is poor. A figure such as 22.1/s, which does not have a common factor with mains frequency, should be chosen to minimise any mains artefact. A check should be made to ensure that the stimulus rate allows the data collection to be completed prior to the next stimulus being given. This can be done by using the following equation: Maximum stimulus rate = 1000 ÷ time window. Conversely, the maximum time window = 1000 ÷ stimulus rate.

Stimulus Intensity Scale

Stimulus levels should be recorded in dBnHL “dial”. The “nHL” can be taken to imply the use of either ISO389-6 (2007), NHSP-recommended calibration values (refer to NHSP Guidance for Auditory Brainstem Response testing in babies), or other calibration datum that is referenced to the average psycho-acoustic threshold of a group of normally hearing young adults. The level of stimulus output should be monitored by listening to the earphone at critical points during the test.

Stimulus Polarity

The majority view, based on current practice, was that for newborn hearing screening and threshold measurement, alternating polarity should be used to minimise the stimulus artefact. Note that the polarity of the stimulus can affect the response waveform and in cases of ANSD, it may be useful to record responses to both rarefaction and condensation clicks.

Transducers

These should be of sufficient quality to deliver a stimulus up to 140dBSPL peak (about 107dBnHL) without distortion. TDH39/49 headphones are recommended. Earphones should be centred over the ear canal ensuring that the ear canal is not collapsed due to excess pressure. Insert earphones may also be used (e.g. type EAR-3A).

Insert Earphones and Stimulus Intensity

If insert phones are used to test babies, the recommendation is that they should not be used above 85dBnHL "dial" unless they include a microphone to automatically adjust the stimulus level for ear canal volume. The reason for this is that a baby has a much smaller ear canal which can give up to a 20dB rise in the stimulus level compared to the same insert earphone used in an adult (Voss & Herrman, 2005; Rance & Tomlin, 2006; Sininger et al, 1997). This figure is thought to diminish over the early months of life as the ear canal grows.

Evidence suggests that insert earphones give higher stimulus levels in babies less than 12 weeks of age. As such, a 10dB correction factor has previously been added to the click stimulus level in dBnHL "dial", if inserts have been used, to correct for the effect of the smaller ear canal volume. While this is no longer required for the reporting of thresholds (the dBeHL correction factors take this effect into account), it is required for the purposes of comparing click latencies to the ABR latency normative data, and for the plotting of wave V latency intensity functions.

As an example, if performing a click-evoked ABR and a threshold of 20dBnHL "dial" is obtained with inserts, that threshold is to be corrected to 30dBnHL for comparison to the normative data, and for plotting of the wave V latency-intensity function only.

There is no need to correct for the increase in intensity for HH reporting purposes, as the reporting method adopted by the HH program, which includes the conversion of results to dBeHL, takes this effect into account for babies less than 12 weeks of age.

HH Pass Levels and Reporting of Thresholds / Correction Factors for dBeHL

The pass level for Click AC ABR testing is regarded as 25dBeHL for HH purposes. Therefore the dBnHL "dial" reading to achieve 25dBeHL will differ depending on the age of the infant and the

transducer used for testing. Please refer to the table below for the HH Pass Levels in dBnHL "dial" for click, based on the transducer used and the age of infant at the time of testing.

Reporting of thresholds should be done in dBnHL "dial" with the transducer used for testing clearly indicated. The thresholds also need to be reported in dBeHL, using the correction factors below (based on the transducer used and age of the infant).

HH Pass Criteria & Correction Factors for AC Click ABR			
Transducer Used (Corrected Age)	**Pass Level in dBnHL "dial"**	**Correction Factor (conversion from dBnHL "dial" to dBeHL)**	**Pass Level in dBeHL**
▪ Insert Earphones (age <24 wks)	20	+5	25
▪ Insert Earphones (age >24 wks) ▪ Headphones (any age)	30	-5	25

Amplifier and Artefact Rejection Levels

The sensitivity/gain should be set such that any electrical activity greater than about ± 10μV is rejected. Some users have found that lower rejection levels down to ± 5μV are better. A value of between ± 5μV and ± 10μV is therefore recommended. The equipment manuals should be checked to determine what value of amplifier sensitivity is required to achieve these rejection levels. The use of higher rejection levels is not recommended when recording conditions are difficult (e.g. high muscle activity). The result is likely to be a poorer signal to noise ratio in the averaged signal. Babies need to be sleeping or in a very settled state when testing.

Filters

Low frequency (high pass): A value of 30Hz is recommended. A value between 20 to 30Hz has been found to give the best signal to noise ratio of wave V near threshold.

High frequency (low pass): A value around 1500Hz is recommended. There is little response energy above this frequency. A higher value generally adds more electronic noise from the amplifier.

Notch Filter

Under normal recording conditions this will not be required, as 50Hz mains artefact should be absent or minimal with good electrode practice using current equipment. If mains artefact levels are high, it is better to identify and remove the source of the problem rather than use a notch filter which can introduce distortion.

Window Length and Averaging

The longest response acquisition window consistent with the chosen stimulus rate should be used. This will normally be less than 15ms (see section on stimulus rate). The number of sweeps per replication should be varied depending on both the size of the response and the level of background activity. It will normally vary between about 1500 and 2000 sweeps, although higher numbers will be required when the responses are small or the background noise is high. Typically a figure of 2000 sweeps is recommended, with a minimum of 1500 required for Healthy Hearing purposes.

The number of rejected sweeps will depend on the level of background activity and the rejection level that has been chosen. Whenever the electrical activity from the baby exceeds the rejection levels for long periods of time, data collection must be halted until the baby has settled.

Data rejection facilities on current equipment are effective in rejecting most unwanted sweeps. However, when there is a long period of high background activity and the data collection is not interrupted there will be a greater proportion of sweeps containing higher levels of noise added to the averaged response. Recordings made where there is a high number of rejected sweeps must be treated with caution. Do not widen the reject levels in an attempt to complete testing on an unsettled baby.

Display

The convention of plotting wave V upwards is proposed as it is the most common in current practice. It is recommended that the display be always set at a fixed number of μV/division. The amplitude (y) and time (x) scales should be such to ensure that small waveforms near threshold are visible. It is recommended that 0.1μV on the response amplitude (y) axis is the same size on the display as 1 – 2ms on the time (x) axis. It is also recommended that the same display scales are used for all tests. An automatic display gain must not be used, as an inappropriate gain for assessment of the response may be set.

Control Recordings

It is also recommended that a control recording is carried out with the stimulus acoustically blocked whenever the ABR response is marginal and/or is of the form that could be an artefactual response (e.g. in tone burst ABR it is often the case that only wave V and SN_{10} are observed). Mains artefact could produce a similar response. If the response is artefactual and is not an electro-physiological response to the sound stimulus, it will still be present when the stimulus is acoustically blocked. Note that marginal responses, that do not take the form of a potential artefact, may not need to be checked if well supported by clear repeatable responses at stimulus levels 5 or 10dB higher.

If artefactual responses are observed, then it is essential to determine their source and remove them from the recording process. Advice should be sought (including contact with equipment manufacturers where necessary) so that the source of the artefacts can be eliminated.

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2. TONE BURST / PIP / CHIRP AIR CONDUCTION ABR TEST PROTOCOL

SCOPE

This protocol aims to provide guidelines for testing babies in the first few months of life using the ABR evoked by tone burst, pip or chirp stimuli for the purposes of assessing frequency-specific hearing sensitivity. Please note the term 'tone burst' will be used for ease, but applies to tone burst, pip or chirp stimuli throughout this protocol. Implementation and application of the tone burst ABR requires a greater level of knowledge and interpretation skills when compared to the click ABR, so it is recommended that testers are familiar and confident with the click ABR before using tone burst stimuli. This protocol covers the technical procedure of carrying out a tone burst ABR test and its use primarily as an audiological threshold tool at the diagnostic stage following referral from the neonatal hearing screen.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR TONE BURST AC ABR

Electrode Location:	One Channel: • Positive electrode: High forehead/Vertex • Negative electrode: Ipsilateral mastoid • Common electrode: Contralateral mastoid
Stimulus:	Tone burst/pip/chirp with alternating polarity. Stimulus presentation level: 2 traces required at 50, 30 & 20dBeHL as a minimum for normal discharge (1 & 4kHz). Clinical threshold seeking is required if loss is suspected.
Stimulus timing:	Tone burst/pip: Linear rise-plateau-fall typically Blackman 2-1-2 cycles
Stimulus rate:	27.1 to 27.7 per second
Earphone:	TDH39/49 preferably EAR-3A (insert earphones)
Coupler value for 0dBnHL:	Depends on exact stimulus and measurement parameters (refer to Calibration information)
Amplifier reject levels:	±3 to ± 10μV
Filters:	Low frequency: 20 to 30Hz High frequency: 1500Hz (3000Hz for CE-Chirp)
Window length:	20ms to 25ms*
Number sweeps averaged per replication :	Minimum 2000 sweeps
Display:	0.05μV or 0.1μV on amplitude axis = 1 to 2ms on time axis
HEALTHY HEARING PASS CRITERIA for functionally normal hearing at any frequency (0.5 to 4kHz):	Wave V present down to 20dBeHL

* The longer time window of 25ms must be considered, especially when testing the lower frequencies when conductive dysfunction is evident.

HEALTHY HEARING TONE BURST AC ABR QUICK SUMMARY

Tone burst testing is used in two ways for testing of infants referred by the HH program:

1. Tone burst ABR can be used in the determination of functionally normal hearing where click ABR has been obtained down to 25dBeHL, but TEOAEs have been unable to be obtained at the desired pass level for frequency specific information at 1 and 4kHz. In this instance:
   - Each ear that has not met pass criteria on TEOAE must be tested.
   - Testing is routinely carried out at 50, 30 and 20dBeHL.
   - At least 2 traces at each test level are required.
   - The presence of wave V at 20dBeHL is considered "functionally normal" at any given frequency, and can exclude all but a very mild hearing loss.
   - Latency and morphology of responses must be considered in determining presence versus absence of a response.

2. Tone burst ABR is also used in determining the degree and configuration of hearing loss, in a child who does not meet the pass criteria for functionally normal hearing. In this instance, testing should be conducted at all 4 major frequencies (500, 1000, 2000 and 4000Hz) down to threshold for each ear where a hearing loss is indicated.

COMPREHENSIVE PROTOCOL INFORMATION

Many aspects of the methodology for the tone burst ABR (TB ABR) are similar to those already outlined in the click ABR protocol. This protocol will address in more detail the issues which are specifically relevant to the use of tone burst stimuli. Please note that this protocol applies to tone burst ABR testing when used as a measure of hearing threshold. In this document the term "tone burst" is considered equivalent to the term "tone pip" and "tone chirp", unless specifically indicated.

Tone burst ABR waveforms are generally smaller, broader and longer latency than the click ABR, particularly with mid to low tonal frequencies. It often consists of a slow wave V component followed by a negative trough, sometimes called the SN10. The data collection parameters are adjusted in order to capture this type of response. It is important to note that the response obtained to a CE-chirp stimulus is usually of a shorter latency than a tone burst stimulus, especially to low frequencies. Larger amplitudes are also found (across all test frequencies) with the use of narrow-band chirp stimulus, except at high intensity levels, when compared to tone burst stimulus (Rodrigues et al, 2013).

Electrode Location

One channel or two channel recordings can be used.

In *single channel recording*, electrodes are located as follows:

- Negative electrode: ipsilateral mastoid. Use a low mastoid position unless wishing to record the post-auricular myogenic response.
- Positive electrode: high forehead as near as possible to Cz[a] and midline. The fontanelle should be avoided but the electrode should be placed as close as possible to this otherwise the ABR response will be reduced in size.
- Common electrode: contralateral mastoid.

The above electrode configuration should result in wave V being plotted upwards on the display. If this is not the case then the positive and negative electrode connections should be reversed.

In *two channel recording*, electrodes are located as follows:

- Negative electrodes: ipsilateral mastoid and contralateral mastoid.
- Positive electrode: high forehead as near to Cz as possible, avoiding the fontanelle.
- Common electrode: forehead.

Contralateral electrodes will be used in the two channel recording.

Type of Stimulus

For tone burst stimuli, a brief tonal stimulus with a rise and fall period in the range of 1 to 2 cycles each and a plateau of 1 to 2 cycles is recommended. A tone burst with 2 cycle rise and fall times and 1 cycle plateau is widely used, often referred to as a 2-1-2 cycle tone burst. The ramp for the rise and fall of the stimulus can be either linear or Blackman type. The tonal frequency is typically in the range of 500Hz to 4kHz. There is some evidence that ipsilateral masking noise (high pass or notched noise) can improve the frequency specificity of the test.

The patented narrow-band CE-Chirps (Claus-Elberling Chirp) are used for frequency-specific chirp testing. CE-chirp testing generally results in larger wave V response amplitudes (particularly at lower intensity levels) and shorter latencies (particularly for lower test frequencies such as 500 and 1000Hz), when compared to tone burst testing.

The polarity of the tone burst / chirp stimulus has minimal effect on the response waveform. It is recommended therefore that an alternating stimulus polarity is used for all testing. This also has the advantage of minimising the effects of stimulus artefact in the final averaged waveform. The level of stimulus output should be monitored by listening to the earphone at critical points during the test. Stimulus levels for each tone burst frequency should normally be recorded in dBnHL (i.e. as referenced to the average psycho-acoustic threshold of a group of normally hearing young adults).

Stimulation Rate

A stimulus presentation rate of 27.1 to 27.7 per second is recommended, with alternating polarity to minimise stimulus artefact. A value such as 27.1/s, which does not have a common factor with mains frequency, should be chosen to minimise any time-locking with mains interference. A check should be made to ensure that this stimulus rate allows data collection of each individual sweep to be completed prior to presentation of the next stimulus. This can be done by using the following equation: Maximum stimulus rate = 1000 ÷ time window. Conversely, the maximum time window = 1000 ÷ stimulus rate. A stimulus rate of 27.1/s is compatible with the recommended sweep time of 20 to 25ms for collection of tone burst ABR data.

Transducer

The requirements of the transducer for tone burst stimuli are similar to those for the click. They should be of sufficient quality to deliver a stimulus of up to around 135 dB SPL peak (about 110dBnHL) without distortion. A TDH39/49 headphone or insert earphone (e.g. type EAR-3A) are suitable. Calibration may be more difficult with insert earphones due to greater variation in enclosed volume. However insert earphones reduce the need for contralateral masking. If insert earphones are used then care should be taken to ensure that wax is not compacted by the probe to avoid blocking the sound pathway. Earphones should be centred over the ear canal ensuring that the ear canal is not occluded by any excess pressure.

Insert Earphones and Stimulus Intensity

If insert phones are used to test babies, the recommendation is that they should not be used above 85dBnHL “dial” unless they include a microphone to automatically adjust the stimulus level for ear canal volume. The reason for this is that a baby has a much smaller ear canal which can give up to a 20dB rise in the stimulus level compared to the same insert earphone used in an adult (Voss & Herrman, 2005; Rance & Tomlin, 2006; Sininger et al, 1997). This figure is thought to diminish over the early months of life as the ear canal grows.

There is no need to correct for the increase in intensity for HH reporting purposes however, as the reporting method adopted by the HH program, which includes the conversion of results to dBeHL, takes this effect into account for babies less than 12 weeks of age.

HH Pass Levels, Reporting of Thresholds & Correction Factors for dBeHL

The pass level for functionally normal hearing for TB AC ABR testing is regarded as 20dBeHL (regardless of the frequency) for HH purposes. Therefore the dBnHL “dial” reading to achieve 20dBeHL will differ depending on the frequency tested, the age of the infant and the transducer used. Please refer to the following table for the HH Pass Levels in dBnHL “dial” for each frequency, based on the transducer used and the age of infant at the time of testing.

TB AC ABR – HH Pass Levels – In dBnHL "dial"					
Transducer	**Age**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Insert earphones	<24 weeks	35	30	25	20
Insert earphones	>24 weeks	40	35	30	30
Headphone	Any age				

Reporting of thresholds should be done in dBnHL "dial" with the transducer used for testing clearly indicated. The thresholds also need to be reported in dBeHL, using the correction factors below (based on the transducer used and age of the infant).

HH Correction Factors – conversion from dBnHL "dial" to dBeHL (to be added to dial reading)					
Transducer	**Age**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Insert earphones	<24 weeks	-15	-10	-5	0
Insert earphones	>24 weeks	-20	-15	-10	-10
Headphone	Any age				

Amplifier Artefact Rejection

An artefact rejection limit in the range of +/-3 to +/-10μV is recommended in order to reject a high proportion of noisy signals. This will improve the signal to noise ratio and assist the detection of small responses close to threshold. Levels above +/-10μV should not be used. The level of stimulus artefact rejection is often controlled by the sensitivity/gain selection on the signal amplifier.

Filters

The recommended filter bandwidth on the signal amplifier is a low frequency cut-off (high pass) of 20Hz or 30Hz, and a high frequency cut-off (low pass) of 1500Hz. These settings are the same as for the click ABR and will enable the slower waves of the ABR to be recorded (wave V and SN10).

Notch Filter

A 50Hz mains notch filter should not be used as this may distort or attenuate the slower components of the response (wave V and SN10).

Window Length and Averaging

The recommended window length is in the range 20 to 25ms in order to be sure of acquiring the complete SN10 component of the ABR (see section on stimulus rate above). The number of sweeps per replication accepted should be varied depending on both the size of the response and the level of background activity. Since the tone burst ABR is generally less well-defined than the click ABR, a minimum of 2000 sweeps is recommended. Increasing this number to 3000 or 4000

may be advisable for identification of small responses close to threshold or if the signal baseline is noisy. The number of rejected sweeps will depend on the level of background activity and the reject level that has been chosen. Whenever the electrical activity from the baby exceeds the reject levels for long periods of time it is recommended that data collection is halted until the baby has settled.

Recordings made where there is a high number of rejected sweeps should be treated with caution. Data rejection facilities on current equipment are effective in rejecting most unwanted sweeps. However, when there is a long period of high background activity and the data collection is not interrupted there will be a greater proportion of sweeps containing noise which might distort the averaged response. Replication of the waveform is an excellent technique for differentiating between genuine response and noise.

Display

The convention of plotting wave V upwards (SN10 down) is proposed as it is the most common approach in current audiological practice. It is recommended that the display be always set at a fixed number of μV/division. The amplitude (y) and time (x) scales should be such to ensure that small waveforms near threshold are visible. It is recommended that 0.05μV on the response amplitude (y) axis (or 0.1μV for larger responses) has the same size on the display as 1 to 2ms on the time (x) axis. An automatic display gain is not recommended, as an inappropriate gain for assessment of the response may be set.

Calibration

There are no international standards on the RETSPL values for tone burst used for ABR. A procedure for calibration is given in IEC 645-3 (1994). It is important to note that the stimulus rate will affect the sound level of the psycho-acoustic threshold of the tone burst to a much greater extent than the threshold of the ABR. In the IEC 645-3 (1994) standard, a stimulus rate of 20/s is recommended or the rate that is used when testing clinically. These recommendations are open to question due to the effect of stimulus rate noted above. If a local biological calibration is carried out, it is recommended that this is performed using a relatively slow stimulus rate of 10 per second.

Samples of tone burst data from four centres are presented (in the following table) which describe the peak-to peak equivalent sound pressure level (dBppeSPL) for 0dBnHL (RETSPL). The exact measurement parameters for each centre differ slightly with respect to stimulus rate, coupler and tone burst characteristics but serve as a useful guide to the approximate values expected.

Site	Reference	Earphone	500Hz	1kHz	2kHz	4kHz
Vancouver	Stapells & Oates (1997)	TDH 49	24.6	23.2	26.1	29.0
Nottingham	Mason (1999)	TDH 39	26.5	22.0	29.0	29.5
Los Angeles	Sininger et al (1997)	Insert	23.2	21.8 (1.5 kHz)		17.8
Sheffield	Stevens (2001)	TDH 39	24.1	21.3	24.9	29.0

It is important to be aware that calibration values will be different for insert type earphones when compared to the TDH39/49. Also existing calibration values on commercial equipment need to be checked as there is a likelihood that these may relate to a long duration burst of pure tone rather than the shorter tone burst. For a more detailed explanation please consult the appropriate reference. You are strongly advised to seek expert help if you are not familiar with the measurement of peak to peak equivalent SPL. It is informative to compare these values of the ppeSPL for the tone burst with the recommended value for the click (33dBppeSPL for a TDH39 earphone – see Stevens et al, 1999).

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3. BONE CONDUCTION (BC) TONE BURST ABR TEST PROTOCOL

SCOPE

This protocol sets out to provide guidelines for testing babies in the first few months of life by bone conduction (BC) ABR using tone burst stimuli for the purposes of assessing hearing sensitivity. The protocol covers the technical procedure of carrying out a bone conducted ABR test and the reporting of the results.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR TONE BURST / PIP / CHIRP BC ABR

Electrode Location:	As per Click Air Conduction ABR
Stimulus:	Alternating tone burst. Rate 27.1 – 27.7/s
Stimulus Timing:	Rise-plateau-fall typically Blackman 2-1-2 cycles
Bone Conductor:	Radioear B71 or B70
Coupler value for 0dBnHL:	Refer to NHSP calibration data
Amplifier reject levels:	±10 to ± 25µV
Filters:	Low frequency: 30Hz High frequency: 1500Hz
Window length:	20ms minimum
Number sweeps averaged per replication :	Minimum 2000 sweeps Typically 3000 sweeps
Display:	0.1µV on amplitude axis = 1 – 2ms on time axis

HEALTHY HEARING TONE BURST BONE CONDUCTION ABR QUICK SUMMARY

Bone conduction ABR testing is performed to assist in determining the type of hearing loss present. As it is a test used for differential diagnosis, bone conduction ABR testing should always be carried out down to threshold (i.e. until a 'no response' result is obtained), and not stopped at a pre-determined "pass" level. The frequencies requiring assessment via bone conduction should be guided by the TB AC thresholds already obtained for the child i.e. BC testing should be performed at frequencies where hearing loss is evident on TB AC ABR assessment.

The use of tone burst bone conduction ABR is essential in accurately determining frequency specific cochlear reserve. For HH purposes, click BC ABR testing is not to be performed, as the use of click stimuli provides an overall indicator only, and can lead to misdiagnoses where hearing loss is sloping or evident at specific frequencies only.

Reporting of Thresholds and Correction Factors for dBeHL

Reporting of thresholds should be done in dBnHL "dial". The thresholds also need to be reported in dBeHL, using the correction factors below (based on the age of the infant).

HH Correction Factors for Bone Conduction – conversion from dBnHL "dial" to dBeHL (to be added to dial reading)					
Transducer	**Age**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Bone Conductor	<24 weeks	+5	+5	-5	0
Bone Conductor	24 weeks – 2 years	-5	-5	-10	-10

COMPREHENSIVE PROTOCOL INFORMATION: TONE BURST BONE CONDUCTION ABR

Electrode Location

A one or two channel recording can be performed. The purpose of two channel recording is to record the contralateral response to aid in the determination of which cochlea is being stimulated.

One channel recording: (electrodes as used for air conduction click ABR)

- Negative electrode: ipsilateral mastoid.
- Positive electrode: high forehead as near to Cz as possible, avoiding the fontanelle.
- Common electrode: contralateral mastoid.

Two channel recording:

- Negative electrodes: ipsilateral mastoid and contralateral mastoid.
- Positive electrode: high forehead as near to Cz as possible, avoiding the fontanelle.
- Common electrode: forehead.

Contralateral electrodes will be used in the two channel recording.

The above electrode configurations should result in wave V being plotted upwards on the display. If this is not the case then the positive and negative electrode connections should be reversed.

Bone Conductor

This should be of sufficient quality to deliver a stimulus up to 50dBnHL without distortion. A suitable bone conductor is the Radioear type B-71. The B70 may offer an alternative. A check should be made that the impedance of the bone conductor is correct for the equipment being used. The bone conductor should be frequently checked to make sure that the sound output is as expected. A listening test near threshold, and at maximum output level, should be carried out. The sound output can become distorted at high sound levels. The output of the bone conductor measured on an artificial ear should be checked at high levels for waveform distortion. The level of the output should also be checked to see that it follows the value of the attenuator.

Location of Bone Conductor

The bone conductor should be placed on the mastoid, avoiding the mastoid electrode. The bone conductor lead should be kept away from the electrode and electrode lead. The mastoid site gives

a higher stimulus level when compared to a forehead placement. A mastoid location also takes full advantage of the inter-aural attenuation which is at least 20dB for clicks in babies under 12 weeks corrected age.

Pressure to Apply

A moderate force should be used to apply the bone conductor. Tests on an artificial mastoid have demonstrated an error of no more than 2dB over a wide range of applied forces.

Electrode Impedances

Bone conductors can generate large artefacts, particularly at high stimulus levels. Low electrode impedances are of particular importance for BC ABR.

Stimulus and Stimulus Rate

For a single channel recording, a stimulus rate of around 27.1 to 27.7 per second is recommended using **alternating** polarity tone bursts/pips/chirps to minimise stimulus artefact. A maximum rate of 27.7/s is recommended. A figure such as 27.7/s, which does not have a common factor with mains frequency, should be chosen to minimise any mains artefact. The justification for the nature and rate of the stimulus can be found in the tone burst AC ABR protocol.

A check should be made to ensure that the stimulus rate allows the data collection to be completed prior to the next stimulus being given. This can be done by using the following equation: Maximum stimulus rate = 1000 ÷ time window. Conversely, the maximum time window = 1000 ÷ stimulus rate.

The audiologist should consider lowering the rate further if waveform morphology is poor and waveform peaks cannot be identified. Stimulus rates as low as 7.1 or 5.1/s can be used if required.

Stimulus artefact will be a particular problem given the combination of the high levels of electromagnetic field radiated from the bone conductor and the long length of the tone burst stimuli. Alternating polarity should be used to minimise any stimulus artefact. Low electrode impedances are therefore particularly important. Note that the polarity of the stimulus can affect the response waveform, and in cases of auditory neuropathy, it may be useful to record responses to both rarefaction and condensation polarities independently.

Obtaining BC ABR thresholds at 500Hz can be difficult due to significant stimulus artefact. It is preferable that bone conduction testing be attempted at this frequency when clinically indicated, however it is recognised that testing at 500Hz may need to be abandoned if artefact cannot be

overcome satisfactorily for testing to proceed. When this occurs, a notation should be made that testing at this frequency was attempted but unsuccessful.

The level of stimulus output should be monitored by listening to the earphone at critical points during the test. Stimulus levels should normally be recorded in dBnHL (i.e. as referenced to the average psycho-acoustic threshold of a group of normally hearing young adults).

Effect of Age on Stimulus

There is a change in the effectiveness of the stimulus (actual intensity delivered to the cochlea) for bone conduction testing in the neonatal period. The bone conducted stimulus is effectively stronger when applied to a baby, owing to the smaller mass that the bone conductor needs to stimulate. Corrections for effective stimulus level do not need to be applied for HH purposes, as the correction factors used to convert the thresholds from dBnHL "dial" to dBeHL take this effect into account (see below).

Masking

Masking of the non-test ear is required where the stimulus level is high enough to cross to the other cochlea and produce a response. If masking is not used a cross shadow response may mislead as to the true threshold. A masking calculator spreadsheet has been designed by Guy Lightfoot to decide when masking is needed and to calculate the level of masking noise required. It is available to download using the following link: http://webarchive.nationalarchives.gov.uk/20150408175925/http://hearing.screening.nhs.uk/audiology.

The audiologist should err on the side of caution and assume an interaural attenuation of 0dB for all bone conduction testing, regardless of the age of the baby. As a general rule for bone conduction testing, and assuming the non-test ear cochlea is normal, then masking should be considered for bone conduction stimulus levels above 15dBeHL. The masking calculator should, however, always be used to determine if masking is required (even at 15dBeHL or lower), as masking requirements differ, dependent upon the age of the infant.

Amplifier and Artefact Rejection Level

The values used should be the same as for AC ABR. The sensitivity/gain should be set such that any electrical activity greater than about +/-25µV is rejected. Some users have found that lower rejection levels down to +/-10µV are better. A value between +/-10 and +/-25µV is therefore recommended. The equipment manuals should be checked to determine what value of amplifier sensitivity is required to achieve these rejection levels. The use of higher rejection levels is not

recommended when recording conditions are difficult (e.g. high muscle activity). The result is likely to be a poorer signal to noise ratio in the averaged signal.

Filters

The filters should be the same as those used for AC ABR. For the low frequency (high pass) filter, a value between 20 to 100Hz is recommended. A value between 20 to 30Hz has been found to give the best signal to noise ratio of wave V near threshold. For the high frequency (low pass) filter, a value of around 1500Hz is recommended. There is little response energy above this frequency. A higher value generally adds more electronic noise from the amplifier. The order of the filters in different machines varies. Check the manual to determine what the order of the filter is. A higher order filter has a greater effect but may introduce more distortion of the waveform. If the order is greater than two or the manual is unclear, refer to expert help to see if filter values should be altered.

Notch filter

Under normal recording conditions this will not be required as 50Hz mains artefact should be absent or minimal with good electrode practice using current equipment. If mains artefact levels are high it is better to identify and remove the source of the problem rather than use a notch filter which can introduce distortion.

Window Length and Averaging

The values for the parameters are the same as for AC ABR. The longest response acquisition window consistent with the chosen stimulus rate should be used. This will normally be between 18 and 20ms. The number of sweeps per replication accepted should be varied depending on both the size of the response and the level of background activity. It will normally vary between about 1500 and 2500 sweeps, although higher numbers will be required when the responses are small or the background noise is high. Typically 2000 sweeps is recommended with a minimum of 1500. Exceptionally there may be such a large ABR response or low background activity that fewer sweeps can be used.

The number of rejected sweeps will depend on the level of background activity and the rejection level that has been chosen. Whenever the electrical activity from the baby exceeds the rejection levels for long periods of time, it is recommended that data collection is halted until the baby has settled. Data rejection facilities on current equipment are effective in rejecting most unwanted sweeps. However, when there is a long period of high background activity and the data collection is not interrupted, there will be a greater proportion of sweeps containing higher levels of noise added to the averaged response. Recordings made where there is a high number of rejected sweeps should therefore be treated with caution.

Display

The convention of plotting wave V upwards is proposed as it is the most common in current practice. It is recommended that the display be always set at a fixed number of μV/division. The amplitude (y) and time (x) scales should be such to ensure that small waveforms near threshold are visible. It is recommended that 0.1μV on the response amplitude (y) axis has the same amplitude on the display as 1 to 2ms on the time (x) axis. An automatic display gain is not recommended as an inappropriate gain for assessment of the response may be set.

CALIBRATION

There are no international standards on the RETFL values for tone bursts used for BC ABR. A procedure for calibration is given in IEC 60645-3 (2007). It is important to note that the stimulus rate will affect the sound level at psycho-acoustic threshold. In the IEC 60645-3 (2007) standard a stimulus rate of 20 per second is recommended, or the rate that is used when testing clinically. These recommendations are open to question due to the effect of stimulus rate noted above. If a local biological calibration is carried out, it is provisionally recommended that this is carried out at a stimulus rate of 10 per second.

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4. AUDITORY STEADY STATE RESPONSE (ASSR) TEST PROTOCOL

SCOPE

ASSR is an objective test that may be used in the estimating of hearing thresholds in neonates and infants. However, the consensus at present is that it should not be used alone for the assessment of hearing in babies who have elevated thresholds (Stapells, 2010).

For HH purposes, it is recommended that ASSR may be used as a frequency-specific cross-check of ABR thresholds as part of the overall test battery, where AC ABR results suggest hearing thresholds in the severe to profound range (i.e. for losses ≥70dB). This recommendation is based on the study by Rance et al (2005) on the estimation of hearing thresholds using ASSR testing in infants, which clearly demonstrated a higher predictive value of behavioural thresholds for infants with greater degrees of hearing loss.

Bone conduction testing using ASSR is not encouraged at this stage, due to the limited research available in this area for neonates and infants. Bone conducted tone burst ABR testing is to be used in preference as an indicator of bone conduction thresholds for HH purposes.

The following information is provided for the ASSR systems currently in use in Queensland, and is not intended to imply any relative merit of these systems over any others that are commercially available. The protocol will cover the suggested testing parameters for the Natus Biologic MASTER, the GSI Audera, Interacoustics Eclipse and Vivosonic Integrity V500 systems.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR ASSR

Filter Settings:	Fixed by system.
EEG Reject Levels:	Closest equipment setting to ±10µV. A lower value than this can be used if the baby has very low EEG down to about ±5µV.
Recommended maximum recording time:	4 minutes for each run at one stimulus level
Stop criteria for presence of a response:	A stop criteria of $p<0.02$ (98% confidence) is recommended to accept the response. Times to reach $p<0.02$ vary between test frequencies (e.g. It can often take longer to reach this criteria for 500Hz). • MASTER: set $p<0.02$ (beware default may be set to $p<0.05$ for green light – needs to be amended from default protocol. • Audera: Fixed at >97% ($p<0.03$) • Eclipse: Set to accuracy option $p<0.01$ • Vivosonic: $p<0.01$
Noise floor for good recording condition:	Where this data is available, the provisional recommended value is <10nV root mean square to accept that good recording conditions were achieved. Where noise is higher than this, a result needs to be considered as inconclusive.

Minimum recordings:	Threshold Threshold - 5dB Threshold + 5dB
Amplitude Modulation Depth:	100%
Frequency Modulation Depth:	20% (±10%)
FM to AM Phase:	–90° (+270°)
Modulation Type:	Enable exponential modulation (if available) Enable FM modulation
Carrier frequencies:	500, 1000, 2000 & 4000Hz
Modulation rates:	• MASTER: can be variable and are set by user. Consult manufacturer's information. Very important to consider if carrying out multi-frequency testing. • Audera: 74Hz for 500Hz, 81Hz for 1kHz , 88Hz for 2kHz, & 95Hz for 4kHz. • Eclipse: Select 90Hz range • Vivosonic: use 80Hz
Stimulus levels:	Normally record responses in 10dB steps. However, the use of a 5dB step either side of threshold will improve (decrease) the predicted threshold range on the estimated audiogram.
Initial intensity level:	Be guided by ABR thresholds – start 10dB above predicted / expected threshold at each frequency to be assessed.

HEALTHY HEARING AUDITORY STEADY STATE RESPONSE QUICK SUMMARY

The provisional Healthy Hearing definition for an ASSR threshold is equivalent to the definition for ABR threshold (which is defined as the lowest level at which a clear response is present, with the absence of a recordable response at a level 5 or 10dB below the threshold, obtained under good recording conditions).

For each frequency tested using ASSR, the definition of threshold is the lowest level at which the target response criteria value of $p<0.02$ is obtained, with the absence of a recordable response at a level 5 or 10dB below this threshold, obtained under good recording conditions (with "absence of a response" defined as $p>0.02$ with the noise floor <10nV). There should also be a response meeting the $p<0.02$ criteria at 5 or 10dB above threshold. If the threshold is obtained at the maximum stimulus output of the system, there should be a further run at the maximum stimulus level meeting the $p<0.02$ criteria instead of the run at 5 or 10dB above threshold to confirm the result as present.

For ASSR testing, both the "Thresholds Audiogram" and the "Estimated Audiogram" should be provided when results are reported.

Although simultaneous two-ear testing is possible with some ASSR systems, this is generally not recommended for HH purposes.

COMPREHENSIVE PROTOCOL INFORMATION: ASSR

Transducers

For air conduction, insert earphones or supra-aural headphones can be used. However at high stimulus levels, inserts are recommended as they are less likely to produce stimulus artefact. The increased stimulus intensity due to the smaller ear canal volume in babies must be considered if inserts are being used. Refer to Stevens et al (2007) for more detail.

Maximum Stimulus Levels

Care should be exercised when using high levels of stimuli. This is particularly the case for multi-frequency stimuli where the overall intensity will be higher than the level set for each single frequency. Some systems will warn the user when a single frequency should be used rather than multiple frequencies (e.g. MASTER and Eclipse Systems).

Electrode Location

For single ear recordings:

Positive electrode:	High forehead (as high as possible but avoiding the fontanelle)
Negative electrode:	Ipsilateral mastoid
Common electrode:	Contralateral mastoid

CALIBRATION

Contact a calibration specialist in your area for further information if required.

Headphones

These can be calibrated in dBHL using pure tones as follows. The method is similar to the peak-to-peak calibration of transient tone burst or tone burst stimuli.

1. The transducer is placed on a suitable coupler connected to a sound level meter. The output of the sound level meter is displayed on an oscilloscope.
2. An amplitude-modulated tone from the ASSR system (with no exponential or frequency modulation) is fed to the transducer at a known stimulus level and displayed on the oscilloscope.
3. The peak-to-peak amplitude of the modulated tone is equated (on the oscilloscope) with a pure tone from a suitable source. The frequency of this reference tone is not critical but should be within the recording bandwidth of the sound level meter. A 1kHz pure tone is a suitable choice or the frequency of the ASSR carrier.
4. The pure tone is then calibrated in dBHL using RETSPL data for pure tones (ISO 389-1).
5. This value in dBHL will be the output of the ASSR system at the set stimulus level. The ASSR system should be adjusted until they are the same value.

NB. The Eclipse uses a chirp based stimuli and requires a slightly different procedure.

Insert Earphones

These can be calibrated in dBHL in the same way as earphones, using a suitable coupler for the insert and the appropriate reference levels (RETSPL in ISO389-2). Although this will calibrate the equipment for use in adults, allowance needs to be made for smaller ear canal volumes in babies, which will increase the stimulus level. Refer to Stevens et al (2007) for more details on this.

Result Formats

If ASSR testing is performed, the Audiologists at AH require both the "Estimated Audiogram" as well as the "ASSR Thresholds Audiogram" for the purposes of hearing aid fitting. It is the responsibility of the diagnostic audiologist testing the child to ensure that AH has the ASSR results in both formats.

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5. COCHLEAR MICROPHONIC (CM) TEST PROTOCOL

SCOPE

This protocol aims to provide guidelines for CM testing for babies in the first few months, in order to differentially diagnose SNHL from ANSD. This protocol covers the technical procedure of carrying out the CM test and interpretation of the results.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR CM TESTING

Electrode Location:	• Positive electrode: High forehead (as close to vertex as possible, avoiding fontanelle) • Negative electrode: Ipsilateral mastoid • Common electrode: Contralateral mastoid
Stimulus:	Separate recordings of Rarefaction and Condensation clicks Rate: 22.1/s, up to a maximum of 87.1/s Stimulus presentation level: 80-85dBnHL "dial"
Transducer:	EAR-3A insert earphones
Coupler value for 0dBnHL:	IEC60126 coupler = 26.5dBppeSPL IEC318-4 coupler = 35.5dBppeSPL
Amplifier reject levels:	±3 to ± 10μV (±3μV recommended)
Filters:	Low (high pass): 100 – 300Hz High (low pass): 3000 – 5000Hz
Window length:	10ms
Number sweeps averaged per replication :	Minimum 1500 sweeps Typically 2000 sweeps
Display:	Default: 0.05 – 0.1μV amplitude axis = 1ms time axis Small or absent CMs: 0.025 – 0.05μV = 1ms Large CMs: 0.1 – 0.2μV = 1ms

HEALTHY HEARING COCHLEAR MICROPHONIC TESTING QUICK SUMMARY

- CM testing is indicated for an ear when an absent or grossly abnormal ABR has been obtained, in order to exclude the possibility of ANSD.
- If a TEOAE is reliably present, it provides evidence of outer hair cell function, and CM testing is not usually necessary. The absence of a TEOAE does not exclude ANSD, and CM testing is required in this instance.
- Testing is carried out at 80dBnHL or 85dBnHL "dial".
- Insert earphones must be used.
- Minimum of 1500 sweeps are required using condensation polarity clicks (replicated at least twice).
- Minimum of 1500 sweeps are required using rarefaction polarity clicks (replicated at least twice).
- A control recording required with insert earphone tube clamped (transducer position not to be moved), allowing a possible CM responses to be validated or rejected as stimulus artefact (replicated at least twice).

COMPREHENSIVE PROTOCOL INFORMATION

Background

The cochlear microphonic (CM) is a pre-neural response from the cochlear outer hair cells which is thought to follow the waveform of the stimulus – it is as though the cochlea is acting as a microphone, hence the term. Like the otoacoustic emission, when reliably present, it can be taken as evidence of hair cell function, but cannot be used to estimate hearing threshold.

When present, the CM is usually easy to record from babies using the same surface electrodes and methods for recording the more familiar ABR. The protocol guidelines for ABR tests should therefore be followed, although some important differences are required if the CM is to be successfully recorded.

Electrode Location

- Negative electrode: ipsilateral mastoid (cannot use nape of neck to record a CM).
- Positive electrode: high forehead (as close to vertex as possible, avoiding fontanelle).
- Common electrode: contralateral mastoid.

The mastoid electrodes should ideally be sited as close as possible to the meatus (and therefore cochlea) as practicable. The ABR guidelines for babies recommend a low mastoid position to allow room for a mastoid placement of the BC transducer and to maximise the ABR response. Placement of two electrodes, one for ABR and one for CM, is not practical and so it is recommended that the guidance for ABR testing is followed but that the 'low mastoid' position is interpreted as no more than 1cm lower than the meatal level of the ear.

It is recommended that the electrode leads are gathered together (or twisted together) and physically separated from the transducer cables and transducer to minimise the extent of stimulus or other electromagnetic artefact.

Stimulus Rate

A stimulus rate of 22.1/s is typically used, however a maximum up to 87.1/s can be used. Being a pre-neural response, the CM is not subject to neural fatigue and may be recorded as fast as the time window allows. A faster rate reduces the acquisition time required to measure the response.

Stimulus Polarity

The recommended method is to use separate replicated runs of condensation and rarefaction polarity clicks. Many ABR systems have a facility whereby the responses evoked by rarefaction and condensation stimuli using an alternating polarity stimulus can be displayed simultaneously. This alternative approch is acceptable.

Stimulus Intensity

The recommended test level is 80dBnHL “dial”. Testing can be performed at 85dBnHL “dial”, but the higher the stimulus level, the larger any stimulus artefact will be. Stimulus levels above 85dBnHL must not be used in neonates for CM testing. As with all insert measurements, if a clear recording is not obtained, check that the sound has been delivered to the ear canal at the desired level (i.e. that the insert or tubing has not become blocked).

Masking

Since the CM is a ‘near field response’, there is no requirement to mask the non-test ear during CM testing, even if masking is needed when recording the ABR.

Transducers

Tubal insert earphones MUST be used. These have a remote transducer coupled by an acoustic tube (e.g. ER-3A). This introduces a time delay (about 0.9ms) between the electrical signal at the transducer and the acoustic stimulus at the ear canal, enabling separation in time of the electromagnetic stimulus artefact from the CM. If conventional supra-aural earphones were to be used, the CM and stimulus artefact would occur almost simultaneously and would therefore be difficult to distinguish. Tubal insert earphones have a further important advantage: the acoustic stimulus can be easily blocked during a control run by clamping the tube between the transducer and the ear tip. This forms an important element of the test procedure, as in this condition, the electrical artefact remains whilst the stimulus is effectively withdrawn, thus allowing a possible CM response to be validated or rejected as artefact.

When clamping the insert tube, care must be taken not to move the transducer or its lead, since this would change any stimulus artefact, introducing uncertainty into the interpretation of the presence of a CM. The initial positioning of the insert transducer therefore needs to allow the tubing to be clamped. Neverless, the transducer should not be placed close to the mastoid electrode or its lead.

Artefact Rejection Levels

A value of ±3μV is recommended where possible. A value of ±10μV should not be exceeded. The recommended filters allow a strict artefact rejection to be used.

Filters

- Low cut (high pass) filter: 300Hz (if not available, use the highest value available between 100 and 300Hz). This minimises recorded background myogenic and EEG activity.
- High cut (low pass) filter: 3000 to 5000Hz.

Window Length and Averaging

A time window of 8 to 10ms is recommended. The CM will end long before the 10ms and this short time window allows a rapid stimulus rate to be used and allows the region of interest to be examined in greater detail. The number of sweeps per replication is typically 2000, with a recommmended minimum of 1500. If alternating polarity with simultaneous collection of responses to condensation and rarefaction stimuli is used, then typically 4000 sweeps should be averaged.

Display

Because of the large range of CM amplitudes, the aspect ratio used for the display scale may need to be modified beyond that normally used for recording ABR responses. Use the normal click AC ABR scale as the initial default, as the more sensitive ABR scale may assist in the interpretation of small or absent CMs. A less sensitive scale may be appropriate for large CMs. The scale should be chosen on the basis of most clearly demonstrating the presence or absence of a CM.

Calibration

The international standard ISO 389-6 (2007) gives the reference equivalent SPL for clicks and tone bursts/pips. The reference levels are outlined in the Quick Guide: Recommended Parameters for CM Testing at the beginning of this protocol.

DISPLAYING AND INTERPRETING CM TRACES

The replicated waveforms should be superimposed and the separate polarities displayed immediately above and below each other without overlapping (see below for an example) to look for the following characteristic features of the CM:

- A sinusoidal segment that has mirror image (inverts) in the two stimulus polarity waveforms, beginning within 1ms of stimulus and possibly lasting up to 5 or 6ms. If the polarity of the measured potential reverses with click polarity, this is consistent with cochlear microphonic basis for the potential. Note that tests using alternating clicks would yield a flat line if the potential was a genuine CM (due to cancellation of the response). If the response polarity does not reverse with changes in stimulus polarity and the response persists using alternating clicks, this is consistent with a neural (ABR) basis for the potential. For a CM to be regarded as “clear”, its size should be substantially greater than the residual noise (as judged from the average gap between the replicates), preferably with a signal to noise ≥3:1.
- If a CM potential appears present, it is important to verify that it is not a stimulus artefact. Perform replicated additional runs (of either polarity – it is not necessary to obtain both) at the same stimulus level but with the tubing clamped. If the potential is clearly eliminated, it is a true biological potential. If the measured potential remains, it is due to stimulus artefact. In this case, separate the transducer and electrodes as much as possible and repeat the entire test

sequence. Carry out further replications of any test where there is any doubt over the presence of a cochlear microphonic or artefact.

- If no potential CM is evident it is not necessary to perform a control run with the tubing clamped.

Please see below traces to assist in interpetation of present vs small vs absent CM repsonses.

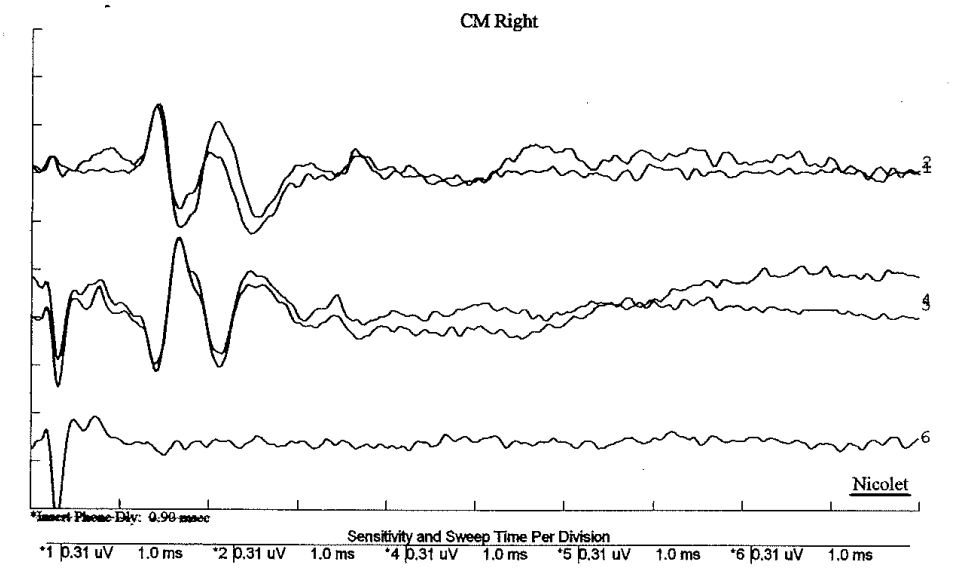

Figure 1: Clear and large (about 0.8µV) CM present. Top: Rarefaction click. Centre: Condensation click. Bottom: Condensation click with tube clamped. Note that the initial deflection in the condensation waveforms is stimulus artefact. The CM is not present in the clamped waveform. Ideally the clamped waveform should have been replicated but in such an obvious case the lack of replication does not introduce uncertainty. Because of the large size of this CM, a display scale beyond that recommended for ABR has been used.

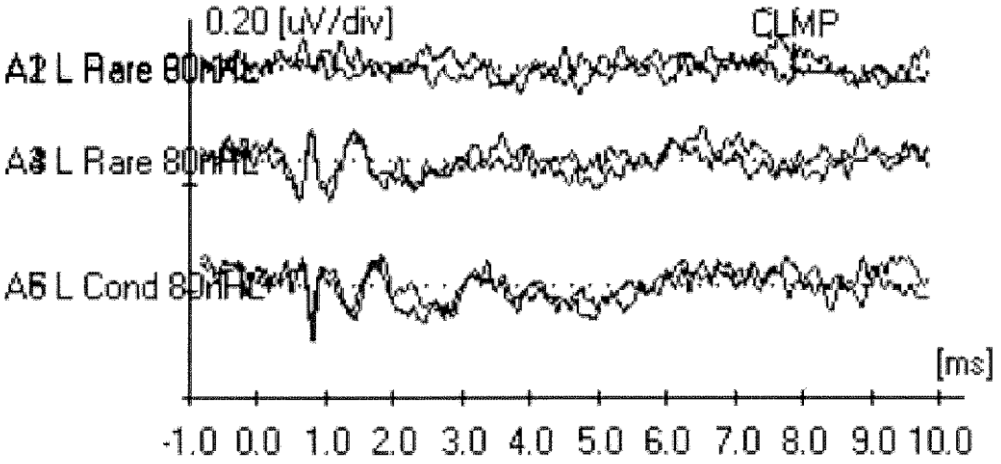

Figure 2: A small but clear CM is present in the centre and bottom waveforms, which is not evident in the clamped waveforms at the top. The superimposition of replicated waveforms provides an estimation of the residual noise thus allowing the significance of waveform features to be assessed. The display scale falls within the range normally used for ABR.

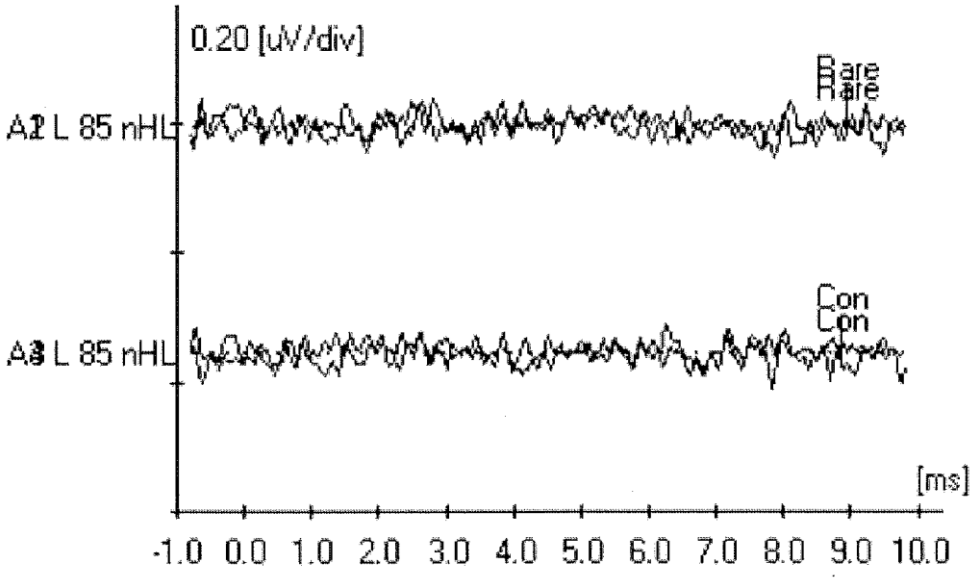

Figure 3: Absent CM. The low level of residual noise (as judged from the average gap between replicates) confirms that the recording conditions were good. There is no feature in the waveforms with the characteristics of a CM, so no clamped waveforms are necessary.

NOTES

- The CM threshold level is not a useful predictor of behavioural threshold, as even in normally hearing infants it cannot be reliably measured at levels below 50 – 60dBnHL.
- Figure 1 shows a very large CM. This is sometimes seen in ANSD and may be associated with abnormal efferent suppression of hair cell activity. There have been anecdotal reports of similarly large OAE responses in some cases of ANSD. However large CM and OAE responses are not always seen in ANSD.
- Although not relevant to distinguishing between ANSD and SNHL, as with ABR testing, make a note of any behavioural response to the CM stimulus.

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6. TRANSIENT EVOKED OTOACOUSTIC EMISSION (TEOAE) TEST PROTOCOL

SCOPE

This protocol describes recording parameters to be used in the recording of TEOAEs for babies and children. In addition, it includes pass/refer criteria to be used for Healthy Hearing purposes.

TEOAE has been selected as the method to be used in the Healthy Hearing Diagnostic Audiology Protocols due to its strengths in hearing loss identification and noise rejection. However, an Audiologist may use their discretion regarding when to use DPOAEs in addition to (not substituted for) TEOAEs to enhance the test battery. As a result, the remainder of this protocol will cover TEOAEs only.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR TEOAE TESTING

Probe fitting:	Well fitted probe with no significant change in fit over recording interval. Minimal stimulus "ringing". Ear canal response is relatively flat.
Stimulus:	Click between 75 and 100pps
Stimulus level (ppe):	80 ± 3dBpeSPL into neonatal ear canal or equivalent volume cavity
Variation of stimulus level between probes:	± 2dB
Data reject level:	At or below 55dBpeSPL
High pass filter to remove low frequency noise:	Around 1.2kHz
Bandwidth:	Able to record between 1000 and 5000Hz
Data collection / analysis window :	Start - 4ms; End - 10 to 12.5ms
Minimum number of responses:	Averaged 50 sweeps at low stimulus level equivalent
Maximum recording time:	6 minutes
Present response criteria:	• ≥ 6dB signal to noise ratio in 3 or more of half octave bands from half octave bands centred at 1/1.5, 2, 3, and 4 kHz, with a minimum TE response amplitude of 0dBSPL and minimum reproducibility of 80%. • It is essential for a response to be present in the 1/1.5 and 4 kHz bands to ensure normal hearing across the frequency range. • Minimum overall reproducibility of 80%. • Minimum stimulus stability of 80%.

HEALTHY HEARING TRANSIENT EVOKED OTOACOUSTIC EMISSIONS QUICK SUMMARY

- Stimulus of 80dBpeSPL ± 3dB is required for testing.
- Good "check-fit" with minimal stimulus ringing, with broad, relatively flat ear canal response.

- Pass criteria for child to be deemed to have functionally normal hearing in the presence of a normal click evoked ABR:
    - Minimum 6dB SNR in at least 3 frequency bands (including at least 1 or 1.5kHz, and 4kHz), with a minimum TE response amplitude of 0dBSPL, and minimum reproducibility in each band of 80%.
    - Minimum overall reproducibility of 80%.
    - Minimum stimulus stability of 80%.

COMPREHENSIVE PROTOCOL INFORMATION

For the purposes of diagnostic assessment for the HH Program, operator controlled testing rather than automated testing is required (i.e. diagnostic protocols are to be used instead of automated TEOAE screening protocols).

Environment

OAE screening in noisy environments is time consuming and inefficient. Every effort should be made to screen in a room without continuous background noise such as air-conditioning, ventilation or road traffic noise. Occasional voices and other noises are less of a problem since they are rejected by the instruments' artefact rejection system. Noise can also be minimised by ensuring the probe cord is not rubbing against clothing.

Probe Checks

It is advised that the probe is checked for sound output and microphone sensitivity daily, or in the case of heavy usage, after every 50 test runs. Sound output should also be checked immediately after any changes have been made to the probe. To check the sound output, an acoustic loop-back test with the probe in a test cavity should be completed first, followed by a 'biological' TEOAE check.

Probe Fitting

Good probe fitting is the single most important aspect of TEOAE testing. Probes need to be fitted well in the ear canal to collect the most sound and to exclude the most external noise. Problems are typically caused by debris in the ear canal, TEOAE probe blocked by debris, fluid immobilising the eardrum or by the ear canal itself being collapsed between the probe and the eardrum. The most successful OAEs are recorded and obtained from a well-sealed ear canal (Kemp, 2008).

Fitting the probe need not disturb the baby who should be quiet during the test. It is essential that good probe fitting is achieved BEFORE in-the-ear calibration, since is impossible to overcome the effect of poor fitting by correcting the stimulation drive. Instruments must therefore provide a clear

indication of the insertion and coupling achieved by the operator, who should confirm probe fitting is optimised BEFORE they activate any in-the-ear calibration.

A good probe fit can be verified by viewing the click stimulus waveform. The ideal is a clean, clear, positive and negative deflection lasting no longer than 1ms, followed by a straight line indicating no or very limited 'ringing', or oscillation, of the waveform. The ear canal response should be appropriately broad and relatively flat.

Stimulation and In-The-Ear Calibration

The recommended stimulus level for the TEOAE click stimulus is 80 ± 3dB peak equivalent sound pressure level (dBpeSPL) as measured in the neonatal ear canal or an equivalent sized cavity. Instruments should provide a means of checking that the required stimulation levels in the ear have been achieved.

Data Rejection Level

The data rejection level should be set as low as possible and not above 55dBpeakSPL.

Filtering

Filtering to remove noise below 1000Hz is highly desirable particularly when viewing the result in the time domain. High pass filters at around 1200Hz and falling at 12 or more dB/octave have little effect on the TEOAE response from infants and greatly improve response quality.

Signal Processing

TEOAE systems use signal averaging and frequency analysis to enhance and display the response. The instrument must use a numerical assessment of the confidence that a true OAE response has been observed. The instrument must also use a numerical assessment of the level of noise contamination present in that band, since only by knowing this can it be decided if an OAE was not seen because it was too small, or because it was probably obscured by noise. TEOAE instruments may additionally give an overall wide band signal to noise ratio of response reproducibility index. While useful in assessing the quality of the test environment – overall wide band quality indicators should not dominate the assessment over frequency specific measurements.

Analysis Window

To ensure a high probability that the 'response' seen is a true cochlear response and not due to artefact, TEOAE systems use non-linear (saturated) component extraction and a delayed analysis window to excludes any expected stimulus artefact. To minimise stimulus artefact from contaminating the waveform, the analysis window for data collection should start at 2.5 to 4

milliseconds after delivery of the stimulus. The recommended start time is 4ms. The effective start time will depend on the gate function used in the analysis window. The start time should be 4ms taking this into account. The length of the data collection time, following the stimulus, varies dependent upon the stimulus rate used. The recommended end of the data collection and analysis window is between 10 and 12.5ms.

Data Averaging

There should be a minimum amount of good data (below the reject level) of 50 sweeps at the low (i.e. quiet) stimulus level in the non-linear mode. Stimuli are often presented in packets, for example, in groups of 8 (2 stimuli at the high level to 6 at the low level). Where possible, collecting a larger number of sweeps (i.e. 260) at the low or quiet level is encouraged.

Maximum Test Time

The recommended maximum test time is 6 minutes. If more than 6 minutes actual testing time is required, it indicates that the testing conditions are unsatisfactory for successful testing.

CRITERIA FOR DIAGNOSTIC TESTING

For the purposes of this protocol, it is necessary to set pass-criteria for testing such that there is a negligible probability that moderate or greater bilateral hearing impairment, of cochlear origin present at birth, will be missed consistent with an acceptable pass rate.

For an ear to be judged to have “passed” a TEOAE test, there must be a high probability that a response-like signal is present at the frequencies expected. This is usually determined by the degree of reproducibility or the signal to noise ratio in each frequency band, although other statistical methods can be applied. With TEOAEs this assessment is typically made in each of several half octave frequency bands. Using the proposed time window given above, the proposed criteria is ≥ 6dB signal to noise ratio, with a minimum reproducibility of 80%, in 3 or more of half octave bands from half octave bands centred at 1/1.5, 2, 3, and 4kHz. This is a minimum requirement and, where equipment permits and there is no significant increase in test time, the test may be continued to higher pass criteria (e.g.10dB signal to noise ratio). Where different bandwidths and/or centre frequencies are used the pass criteria should be at least as stringent as one of these options.

The response obtained must also be large enough to be within the normal physiological range for TEOAEs. The proposed minimum TE response amplitude level is 0dBSPL.

A response should therefore be reported as present within a particular half octave band if the signal to noise is ≥ 6dB, the minimum TE response amplitude exceeds 0dBSPL, and reproducibility is at least 80%.

When TEAOE testing is used (in conjunction with other test results) to discharge a baby or child from the Healthy Hearing Program with functionally normal hearing, it is essential for a response to be present in at least 3 frequency bands, 2 of which must include the 1 or 1.5kHz band and the 4kHz band. The response should also have an overall minimum reproducibility of 80%, and stimulus stability should also be at least 80%.

Failure to observe an OAE

A recordable TEOAE indicates the presence of a normal outer hair cell function at or near the frequencies present in the emission, as well as a high degree of normality in the function of the middle ear.

Absence of TEOAEs could be for one of many reasons (e.g. poor recording conditions, bad probe fitting, blocked probe, the presence of outer or middle ear disease, absent cochlear response, or an amplitude too small to record). It is the responsibility of the audiologist to ensure that poor testing conditions have been adequately addressed when recording TEOAEs.

Normally hearing ears produce a wide range of TEOAE intensity and waveforms. Some healthy ears may only produce emissions strong enough to be visible above the infant and background noise in only a narrow range of emission frequencies whilst others will produce a broad range of emission frequencies. Where TEOAEs are unexpectedly absent or below the minimum amplitude requirement (e.g. in cases of rising or sloping hearing losses), TB ABR (or ASSR if appropriate) should be used to establish whether hearing at that frequency is normal or abnormal.

TEOAE amplitudes are largest in newborns and infants, than older children and adults. The amplitude of responses diminishes most noticeably in the first few months after birth (thought to be due to the increase in ear canal size of the neonate as they grow). Despite this drop in amplitude in early life, there are no aging effect on TEOAEs (i.e. studies have failed to demonstrate any further reduction in TEOAE amplitudes with increasing age, when hearing levels are equivalent to those of younger adults). When hearing is normal, TEOAEs will be normal in 99% of the population. The absence of a TEOAE is regarded as a significant clinical finding (where middle ear dysfunction and poor recording conditions have been eliminated as possible causes).

Babies and children with absent TEOAEs in one or both ears in the presence of aerated middle ears and normal thresholds (on tone burst ABR testing or behavioural audiometry), should be

placed on review (for HH purposes) for ongoing monitoring of hearing, as the absent TEOAEs may be indicative of outer hair cell dysfunction that may not yet be detectable on ABR testing or the audiogram.

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7. HIGH FREQUENCY (HF) TYMPANOMETRY (1000 Hz) TEST PROTOCOL

SCOPE

This protocol aims to provide recommendations for accurately determining the middle ear status of young infants who have been referred for diagnostic audiological assessment. The accurate determination of middle ear function is required for both audiological and medical management, and tympanometry is important in assisting this determination.

The aims of this protocol are to summarise the evidence base for high-frequency tympanometry, and to provide guidance as to the interpretation of test results. It assumes an understanding of conventional tympanometry (typically carried out using a probe tone of 226 Hz) and only attempts to cover those aspects that may differ for the assessment of middle ear function in young infants.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR TYMPANOMETRY

Probe tone:	If infant < 6 months (corrected for prematurity) – use 1000Hz probe tone. If infant > 6 months & < 9 months (corrected for prematurity) – test using both 226Hz & 1000Hz probe tone – if discrepancy, take 1000Hz result as correct. If infant > 9 months (corrected for prematurity) – use 226Hz probe tone.
Pressure Range:	The direction of pressure change should be from positive to negative. The range should be at least from +200daPa to -400daPa.
Pump Speed:	A fast screening mode speed of up to 400daPa/sec should be used.

HEALTHY HEARING HIGH FREQUENCY (1000Hz) TYMPANOMETRY QUICK SUMMARY

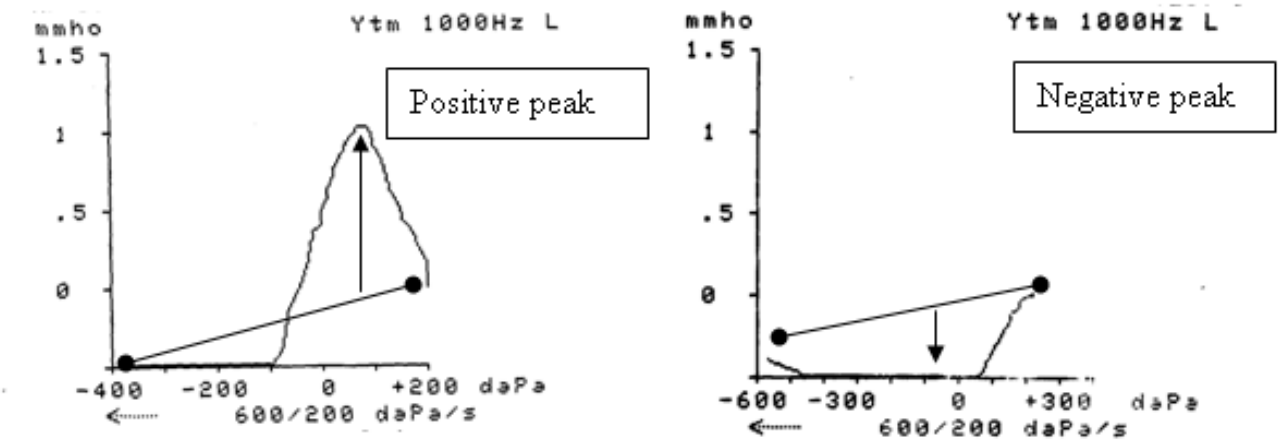
The classification system proposed by Baldwin (2006) and adapted from Marchant et al (1986) is used for the interpretation of HF tympanograms for Healthy Hearing Program purposes. This system proposes only two possible clinical outcomes – normal or abnormal. Traces with positive peaks indicate an aerated middle ear system even when the positive peak is at a negative middle ear pressure (MEP). Flat traces (no peak) or traces with negative peaks (troughs) indicate middle ear dysfunction is present. This is based on a reliably recorded, repeatable trace.

Procedure:

- Perform otoscopy first
- Use a 1000Hz probe tone and admittance (Y) measurement.
- Draw a baseline between points on the trace at pressure extremes (+200 to -400/-600daPa). If the trace disappears below the *x* axis the baseline should be drawn to the *x* axis as shown in Figure 1.

- Identify the main peak which can occur at any middle ear pressure.
- Draw vertical line from this baseline to the peak of the trace. If the peak is above the baseline it is a positive peak (normal); if it is below the baseline it is a negative peak (abnormal). Refer to Figure 1 below. Also refer to Figure 2 and 3 on pages 122-123 for additional examples of normal and abnormal HF tympanograms for neonates.
- If there is a positive and a negative peak the trace should be classified as positive.
- If the conditions are good and the outcome is clear, repetition is not always necessary to draw a conclusion. However traces should usually be repeated if possible to check for reliability. Repeated traces should be classified in the same category of positive or negative. If the outcome is not clear the trace should always be repeated.

Figure 1. Example of how to determine a positive and negative peak (adapted from a method used by Marchant et al 1986).



COMPREHENSIVE PROTOCOL INFORMATION

Low frequency probe tones are invalid in early infancy. The choice between 678Hz and 1000Hz is based on sensitivity and ease of interpretation. McKinley et al (1997) reported that 1000Hz results were less straightforward to interpret; however if the data are re-classified into 'flat' versus 'all other' (which seems more appropriate), then the relationship to OAE results is better for 1000Hz tympanometry than for 678Hz. Baldwin (2006) reported fewer difficult to classify traces when using the 1000Hz tone compared to 678Hz. Using the 1000Hz tone, 5% of traces were 'difficult to classify' in the "normal" group and only 0.6% in the "MEE" group. The use of 1000Hz is therefore recommended in early infancy.

The use of 1000Hz probe tone tympanometry is recommended to test babies less than 6 months corrected age. Tympanometry using 226Hz should not be used. A simple classification scheme is proposed in which there are only two possible clinical outcomes – normal or abnormal, indicative of middle ear dysfunction absent or present respectively. This is based on a reliably recorded trace.

Traces with positive peaks are suggestive of an aerated middle ear, even when the positive peak is at a negative middle ear pressure (MEP). Flat traces (no peak) or traces with negative peaks (troughs) indicate middle ear dysfunction is present.

The sensitivity and specificity of the method is good but not perfect as a small number of traces are not easily classified. Therefore in determining the management of an individual infant, tympanometry needs to be considered in the context of other audiological test results. This protocol is consistent with the recommendations of the US Joint Committee on Infant Hearing (JCIH) 2007 Position Statement for the audiological evaluation of infants aged from birth to 6 months of age.

Up to what age is it necessary to use higher frequency probe tones?

Meyer et al (1997) predicted that valid low frequency tympanometry would only be possible when the adult middle ear resonant frequency was reached. This occurred at around 4 months in one longitudinal case study (Meyer et al 1997). However, low frequency tympanometry can remain invalid up to 5 months of age and variability is currently unknown (Baldwin, 2006). Alaerts et al (2007) recommended 1000Hz below 3 months and 226Hz above 9 months but there was uncertainty about the appropriate choice in the intermediate period. Differences between the neonatal and adult ear in energy transmission (Keefe & Bulen, 1993) and admittance phase measurements (Holte, 1991) which were evident at 4 months had disappeared by the age of 6 months. Tympanometry using the 226Hz probe tone is used clinically to differentiate pathological ears in babies over 6 months old and further longitudinal studies would be necessary to establish the variability in the age at which it became valid. Until this is known we advise that high frequency tympanometry should be used and 226Hz tympanometry be avoided up to the corrected age of 6 months. Between 6 and 9 months of age it is advisable to consider using both 226Hz and 1000Hz. If results are discrepant, the audiologist should err on the side of caution and consider the 1000Hz result more accurate in this age group, and the 226Hz result should be disregarded.

Calibration

Equipment should be calibrated regularly according to IEC 61027:1991. Before use, a simple check on functioning should be carried out by testing one's own ear or in a cavity.

Practical Considerations in Testing

- Otoscopy should be carried out before tympanometry, as well as a general inspection of the external ear for obvious signs of disease, blockage or malformation such as atresia or stenosis. Care should be taken not to insert the otoscopic speculum deep into the ear canal of young babies.

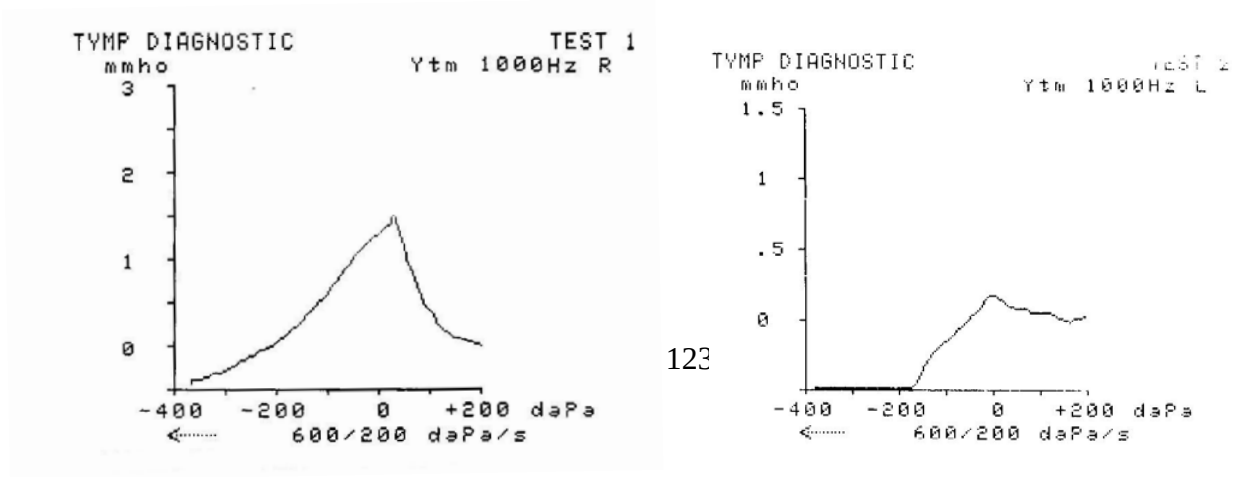
- Conical tips, or those with a flange designed to seal against the entrance to the external auditory canal, may be used for the tympanogram measurement. The pinna should be pulled gently to straighten the ear canal and position the tip.
- Movement of the infant and crying can result in a false peak on the tympanogram. The baby does not need to be asleep but should definitely be resting quietly during the test.
- Equipment should be set to measure admittance (not impedance).
- Traces should usually be repeated, if possible, to check that the result is replicable and not due to artefacts such as baby movement. It is especially important to retest any ear with an abnormal or difficult-to-interpret tympanogram.
- The direction of pressure change should be from positive to negative. The range should be at least from +200daPa to –400daPa. A fast screening mode speed of up to 400daPa/sec should be used.
- Clinicians should be mindful when interpreting the ECV measurements for 1000Hz tympanometry that most tympanometers in clinical use do not usually reflect the true ear canal volume of the infant. The ECV may need to be disregarded as a true ECV measure when high frequency probe tones are used, although it can still be useful to indicate a blocked ear canal.

HF (1000Hz PROBE TONE) TYMPANOGRAM INTERPRETATION

The system recommended for interpretation is that of Baldwin (2006), adapted from Marchant et al (1986) which improved intra-tester agreement significantly. Admittance is used in this scheme and one should check that the equipment is set to measure this. Middle ear pressure (MEP) is not considered in this interpretation scheme. A positive peak at negative or positive MEP was more likely to occur in the "normal" group and was not a feature of the middle ear effusion (MEE) group. The MEE group typically had no discernible peak and the tympanograms were characteristically "trough shaped" (refer to Figure 1 on page 120).

Hence, a positive peak obtained on high frequency tympanometry should be reported as indicative of an aerated middle ear system. A result with no peak or a negative peak should be reported as indicative of middle ear dysfunction.

Figure 2: Examples of tympanograms with positive peaks classified as "normal".



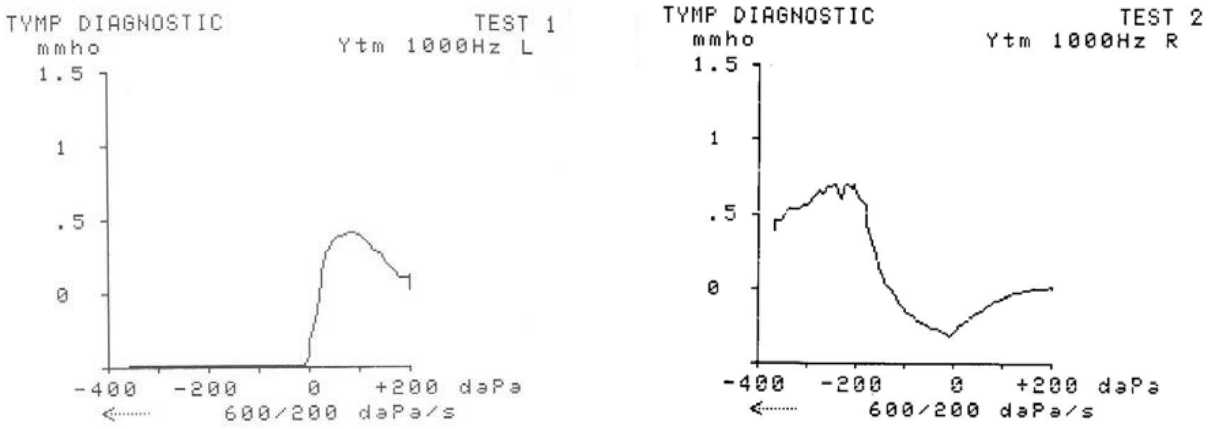
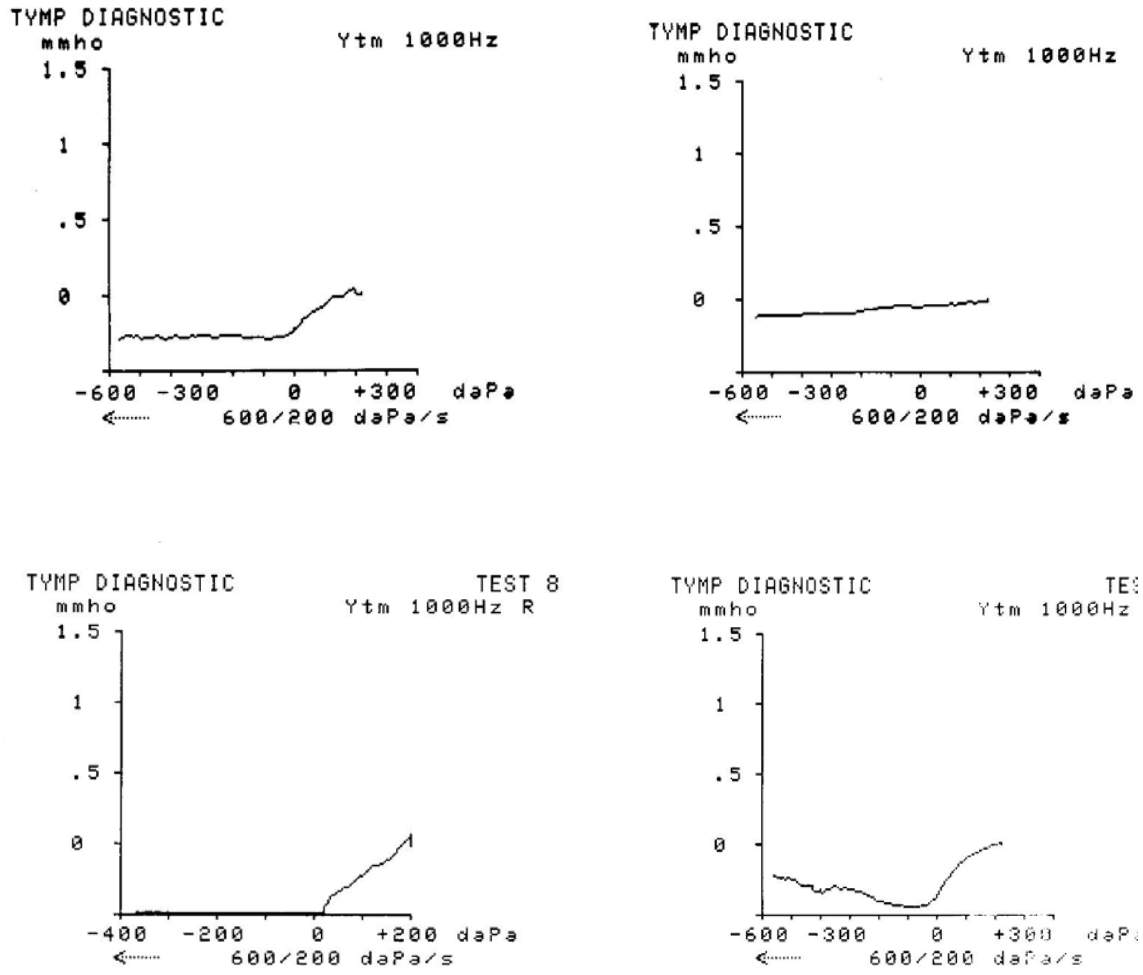


Figure 3: Examples of tympanograms classified as “abnormal” (i.e. no peak).



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8. TYMPANOMETRY (226Hz) TEST PROTOCOL

SCOPE

This protocol aims to provide recommendations for conducting 226Hz tympanometry testing for the assessment of middle ear status in young infants and children. It also aims to provide guidance regarding the interpretation and reporting of tympanometry results. Acoustic reflex measurement and Eustachian tube function testing are not covered, and are beyond the scope of this document.

The accurate determination of middle ear status is required for both the audiological and medical management of the child with hearing loss.

QUICK GUIDE: RECOMMENDED PARAMETERS & SETTINGS FOR TYMPANOMETRY

Probe tone:	For infants > 9 months of age (corrected for prematurity), the 226Hz probe tone is used. If infant is aged between 6 & 9 months of age (corrected for prematurity), they should be tested using both 226Hz & 1000Hz probe tones – if discrepancy exists, take 1000Hz result as correct. 226Hz probe tone should not be used for infants less than 6 months of age (corrected for prematurity).
Pressure Range:	The direction of pressure change should be from positive to negative. The range should be at least from +200daPa to -400daPa.
Pump Speed:	A fast screening mode speed of up to 400daPa/sec should be used.

HEALTHY HEARING 226Hz TYMPANOMETRY QUICK SUMMARY

Procedure:

- Obtain case history and perform otoscopy first to check for contraindications to testing.
- Obtain an air tight seal, and perform the test using a 226Hz probe tone and admittance (compliance) measurement. Ensure that the tympanometer is set to run from positive to negative pressure to avoid collapsing the ear canal (+200 to -400daPa).
- The diagnostic tympanometer will provide the values for ear canal volume, static compliance and peak middle ear pressure, and these values should be recorded and compared to the normative data to determine the tympanogram type using the Jerger (1970) classification system.
- Ensure the tympanogram is of good quality, with minimal client movement / artefact. Repeat testing may be required to obtain a tympanogram of sufficient quality to make a valid judgement regarding the child's middle ear status.

COMPREHENSIVE PROTOCOL INFORMATION

Tympanometry is used as an objective test of middle ear function. It assesses the compliance of the entire middle ear system as a function of changes in air pressure in the ear canal (ANSI S3.39-

1987). When performed in isolation (i.e. without acoustic reflexes also being assessed), tympanometry cannot be used as a complete test of middle ear function, and results need to be interpreted and reported accordingly. It is important that tympanometry results are interpreted in the context of the entire test battery and otoscopic findings.

Tympanogram procedures have remained virtually unchanged since 1970, with the most commonly used tympanometry typing procedure proposed by Jerger in 1970 (outlined below). Tympanograms are classified as Type A, B or C, based on the compliance and middle ear pressure results obtained. The ear canal volume should also be considered in determining patent middle ear systems (i.e. tympanic membrane perforations or patent grommets in situ).

Normative Data for 226Hz Tympanometry

The normative data in use for the HH program are outlined below:

Static compliance (SC):	0.3 to 1.6 cubic centimetres (cc or mmho)
Middle ear pressure (MEP:	+50 to -100 (or -150) decaPascals (daPa) *
Ear canal volume (ECV):	Varies by age of child **
	For children 6 months to 6.7 years: ECV = 0.3 to 0.9cc (Shanks et al, 1992)
	For children > 6.7 years, adult values: ECV = 0.6 to 1.5cc

* Middle ear pressures between -100daPa and -150daPa can be interpreted as normal, if hearing is normal and there are no air-bone gaps or conductive involvement evident on the audiogram or elsewhere in the test battery.

** Ear canal volumes above 1.5cc are sometimes recorded for a multitude of reasons (e.g. previous mastoid surgery, normal individual variation), and must be reported cautiously, especially when otological history is complicated.

Jerger (1970) Tympanogram Classification System

There are 3 main types of tympanograms in this classification system:

- A **Type A** tympanogram (A, As and Ad) is based on a result that has middle ear pressure falling within the normal range.
- A **Type C** tympanogram is based on a result that has middle ear pressure falling below the normal range (i.e. less than -100 or -150daPa).
- A **Type B** tympanogram is based on no measurable static compliance with change in pressure.

Tympanogram Classification			
Type	**SC (cc)**	**MEP (daPa)**	**ECV (cc)**
A	0.3 to 1.5	+50 to -100 (-150)	WNL
As	<0.3	+50 to -100 (-150)	WNL
Ad	>1.5	+50 to -100 (-150)	WNL
C1	>0.1	Between -100 (-150) and -250	WNL
C2	>0.1	<-250	WNL
B	No peak	No peak	WNL
B (high volume)	No peak	No peak	Larger ECV than expected for age (or pre-grommet insertion)

Contraindications to Performing Tympanometry

Tympanometry testing should be attempted on every child seen where medically appropriate. Tympanometry testing should never be performed:

- on discharging or bleeding ears.
- on ears with microtia and/or atresia.
- when foreign bodies are evident in the ear canal (e.g. insects, cotton buds, beads). Note that grommets are not considered to be a foreign body.
- when excessive wax is present, where there is a risk that insertion of the probe tip may push against the impacted wax, risking damage to the eardrum.

Tympanometry should also not be performed on an ear recovering from outer / middle ear reconstructive surgery, until clearance / permission has been given by the treating ENT surgeon.

Limitations of Tympanometry:

It must be acknowledged that there are limitations in tympanometry testing:

- A unique pattern does not exist for every middle ear disorder. The middle ear system is complex with multiple elements contributing to admittance. There is not a direct 1:1 correspondence between specific middle ear abnormalities and specific tympanogram types. The same pathology can produce different tympanogram types.
- The more lateral pathology will dominate, and each individual pathology will not be individually reflected. For example, a perforation can co-occur with a cholesteatoma or stapes fixation. The more lateral pathology is recorded.
- Variability occurs in values with overlap between normal and diseased middle ear systems. Many pathologies occur on a continuum rather than as discrete steps.

Interpretation and Reporting

For the above reasons, tympanometry findings should always be interpreted in the context of all known audiological information (case history, otoscopic findings, and the degree, type and configuration of the hearing loss indicated on other test results), as for all other audiological assessments. Tympanometry results should never be interpreted in isolation as a stand-alone test.

For abnormal results, the term “pathology” should not to be used in reporting as it implies a disease process, which is outside the professional boundaries of an audiologist to diagnose. When reporting abnormal tympanometry findings, the term “dysfunction” should be used.

Included in the table below are examples of how each tympanogram type can be reported.

Tymp Type	Reporting	Comments
A	Tympanometry showed normal middle ear pressure and compliance at the time of testing.	Can occur in otosclerosis, cholesteatoma.
As (shallow)	Tympanometry showed normal middle ear pressure with reduced middle ear compliance, consistent with a hypomobile middle ear system.	Does not solely indicate otosclerosis e.g. scarred TM, recovering OM, normal variation.
Ad (deep)	Tympanometry showed normal middle ear pressure with increased compliance, consistent with a hypermobile middle ear system.	Does not solely indicate ossicular discontinuity e.g. scarred TM, normal variation.
B (flat) **ECV = normal**	Tympanometry showed nil compliance with change in pressure at the time of testing, consistent with middle ear dysfunction.	Can occur in OM, otosclerosis, cholesteatoma.
B (flat) **ECV = large** (no grommet visualised)	Tympanometry showed nil compliance with change in pressure, with large cavity volume, consistent with tympanic membrane perforation/patent middle ear system.	Only reported like this if no history of mastoid surgery and grommet in-situ not able to be visualised.
B (flat) **ECV = large** (grommet visualised)	Tympanometry showed nil compliance with change in pressure, with large cavity volume, consistent with patent grommet in-situ/patent middle ear system.	Only reported like this if grommet can be visualised.
C (negative)	Tympanometry showed significant negative middle ear pressure with normal compliance, consistent with Eustachian tube dysfunction.	Middle ear pressures between -100 and -150 can be interpreted as normal (i.e. Type A) if hearing is normal and there are no air-bone gaps on the audiogram.

Interpretation Notes:

- Type A, As or Ad tympanograms are typically not indicative of transient middle ear dysfunction (as middle ear pressure is in the normal range). When a child has a conductive

hearing loss in the presence of a type A, As or Ad tympanogram, a permanent conductive hearing loss must be suspected until proven otherwise.

- Type C or B tympanograms may indicate transient conductive dysfunction. In labelling a conductive hearing loss as transient, the audiologist must have considered all audiological results to date for the child, including the degree and configuration of the hearing loss. Testing after resolution or medical management must be performed, to ensure a permanent conductive hearing loss has not been missed.

Calibration

The calibration of the machine should be checked daily using the test cavity supplied by the equipment manufacturer. Volumes in the range of 0.5 to 5.0cm^3 are recommended. A calibration check in the test cavity should produce a horizontal line, and the volume measured must be within the tolerance levels specified by the manufacturer (most specify a 5% tolerance for cavity volume). Based on a 5% tolerance, the acceptable volumes for cavity volumes are outlined below:

Test Cavity	Acceptable Value
0.5cm^3	0.5cm^3 (0.48 to 0.52cm^3 if recorded to 2 decimal places)
2.0cm^3	1.9 to 2.1cm^3
5.0cm^3	4.8 to 5.2cm^3

Equipment operating outside of specifications /tolerances should not be used to test patients.

In addition, a daily “biological” check should be performed on an ear with a known peaked tympanogram to check overall machine function.

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9. BEHAVIOURAL OBSERVATION AUDIOMETRY (BOA) TEST PROTOCOL

INTRODUCTION

Behavioural Observation Audiometry (BOA) involves presenting sounds to a baby and observing their responses. In the context of this paper, it is restricted to the responses before the developmental age at which babies can localise sound to the side.

BOA is known to be a test of limited reliability. Some babies will appear to respond even though they may not have heard, and others will have heard but not demonstrated any observable response. Even in favourable circumstances, very young babies do not respond to the quietest sounds that they can hear. They only demonstrate responses to supra-threshold levels of stimuli. It follows that babies with cochlea deafness with recruitment may demonstrate responses to supra-threshold levels while being unable to hear quieter sounds.

The battery of physiological tests of hearing such as ABR, ASSR, OAEs, tympanometry, and acoustic reflex testing are considerably more objective and reliable, and are the preferred method of assessment for neonates referred for diagnostic testing through the HH Program. However, there may be instances where BOA may be useful as a demonstrative tool for parents whose children are diagnosed with hearing impairment and/or ANSD. Some parents may have difficulty acknowledging the results of the physiological objective tests of hearing. It might be expected that most parents would be carrying out their own hearing tests at home when their baby is suspected of being hearing impaired or deaf.

A variety of types of stimuli, responses, and test protocols have been employed (Wharrad, 1988). The expected behavioural responses range from internal autonomic (e.g. heart rate) to body movements (e.g. the face and eyes), from excitatory or reflexive to inhibitory or attentive.

USE OF BOA IN THE CONTEXT OF THE HH PROGRAM

The overall aims of BOA testing in general are:

- to determine age appropriateness of response to sound.
- to confirm existence of behavioural responses to sound.
- to allow comparison of aided versus unaided responses to sound.

BOA may therefore, in the context of the HH Program, have a place in contributing to:

a) demonstrating a baby's responses, or lack of responses, to sound.
b) demonstrating responses to sound where ANSD has been diagnosed.

c) demonstrating the benefit of hearing aids, and giving indications of uncomfortable loudness levels.
d) providing more information about the hearing of neurologically immature babies where there may be doubts about the accuracy of the ABR.

BOA is not to be used to predict the type or degree of hearing loss in neonates referred as part of the HH Program. When BOA is used, parents should receive a full explanation of the difficulties and pitfalls when observing a small baby's responses to sound.

The following recommendations aim to provide general guidelines on the performance of BOA testing only, in order to allow reasonable individual practice and to exclude inappropriate practice. Sticking rigidly to protocols should be avoided as the state of the baby will change within the test (Wharrad, 1988).

TEST REQUIREMENTS AND PROCEDURE

Test Environment

The ideal test environment will be a sound-proof room, with minimal ambient noise and minimal visual distractions. The lighting should be dim, and the audiologist needs to be aware of reflections and shadows.

Noise-maker and Stimuli Suggestions

- Maracas
- Castanets
- Cellophane
- Woodblock
- Tambour
- Sleigh bells
- Small bell
- Chime bar
- Spoon in cup
- Horn
- Popcorn/batteries in jar
- Rattle
- Manchester rattle
- Drums
- Voice / speech
- Narrowband noise and broadband noise
- Warble tones

Ideal Test State of the Infant

- The state of arousal is critical to obtaining responses.
- For younger babies (maybe under 3 months), testers should aim to catch the ideal state between light sleep and quiet awake (Bench, 1975; Bench, 1976). The moment when the eyelids are half-closed can be particularly sensitive.
- For older babies, "stilling" or attentive responses can be observed when they are more awake, but care must be taken to avoid states of strong internalised attention, or strong visual fixation. (Sonksen, 1983).
- Maintain complete quiet for few minutes prior to commencing the test.
- Advise parents to have baby fed, bathed & changed.
- It is best for the baby to be reclining in bassinet/bouncer, not in caregiver's arms (avoids any potential cueing).

Tester / Observer

- The tester presents the noisemaker and observes the sound level meter.
- The observer watches for the infant responses and record details (noisemaker used, stimulus level & response observed).
- The tester can also comment on response observation, if possible.
- Observer bias is reduced if the infant is in a light sleep state.
- The observer can be "blind" to the presence of the stimuli. This may be achieved by video recording, or observing from an adjoining sound-treated room, or viewing the behaviour in the presence of masking through earphones (Gans, 1987).

Stimulus Presentation

- The behaviour of the baby in quiet must be observed for a short period prior to the presentation of any stimuli (Bench, 1975).
- The duration of the stimulus needs to last several seconds, as the alerting and response time is longer in younger babies (Sonksen, 1983).
- It is recommended to start with softer high frequency noisemakers, and then move to lower frequency soft sounds.
- If no response to sound is observed, check that the infant is not too deeply asleep and gradually increase the intensity of the noisemakers.
- Present noisemakers at 20^{O} behind head, 30-45cm away from ear and out of sight, for 2 to 5 seconds.
- Leave at least 10 second inter-stimulus intervals between presentations to avoid habituation.
- Habituation occurs rapidly when stimuli are supra-threshold. The setting of stimulus levels should move quickly to identify the lowest level at which responses can be observed.

- Choose the presentation period carefully, and wait until baby is settled and in the required state for noisemaker presentation.
- Be aware of tactile cues (e.g. air from voice or horn).
- Use non-stimulus trials to check for observer bias. Stimuli below the level of responsiveness could act as the equivalent of "catch" or no-sound-trials.
- Ideally require two agreed responses per stimulus per ear/side of presentation.
- Do not present the same stimulus more than twice in a row, as habituation will likely occur.
- Rapid rise time will generally produce better responses.
- Use the startle response noisemaker last (i.e. hooter).
- Test stimuli can be generated by loudspeakers, to reduce the possibility of baby responding to other non-auditory stimuli.

Determining Responses to Sound

- The wider the frequency band of the test stimuli, the more likely the baby will be to respond (Bench, 1975; Bench, 1976; Trinder, 1990). Broad and narrow band noise must be available. Narrow band noise at low and high frequencies should be included in the range of stimuli.
- Responses are most likely to be elicited to speech and broadband noise, then low frequencies, then warble and pure tones, then high frequencies.
- There is usually little interest in high frequencies before 3 months of age.
- The probability of eliciting response increases with increasing stimulus intensity, bandwidth and duration (up to 3 seconds), decreasing fundamental frequency and decreasing rise time.
- Response must be observed no more than 2 to 3 seconds after stimulus presentation.
- Acceptable responses:
    - Arousal from sleep state: eye blink, whole body shudder, eye opening, head turn, marked limb movement, or any combination the above.
    - Changes in behaviour: cessation of movement, ceasing/commencing crying or vocalization, altered sucking rate, eyelid activity, Moro's response, breathing changes, eye widening, grimacing, auropalpebral reflex.

INTERPRETATION OF BOA RESULTS

As BOA is used, for HH purposes, as a demonstration tool rather than an assessment tool, it is important to report BOA results within this context.

- Results should be reported as either consistent or inconsistent with the baby's known audiological profile from electrophysiological testing. The degree or type of loss cannot be determined using BOA (i.e. be careful not to over-interpret results).
- Always consider the infant's activity state during test when considering their responses.
- Consider whether developmental age is commensurate with chronological age.

REPORTING SUGGESTIONS

Record during testing:

- Ear/side of presentation.
- Stimulus type: name (be specific) and frequency composition (broadband, low/mid/high frequency).
- Stimulus intensity level.
- Observed response (in detail) or lack of response (NR) per presentation of each stimulus.
- Additional comments regarding general infant state or test conditions.

Specific BOA Report Considerations:

- Include state of child during test.
- Frequency range of stimulus and response levels.
- Examples of types of responses observed.
- Comment on reliability of test results.
- What can/cannot be excluded in terms of hearing impairment.
- Avoid labelling the type or degree of hearing loss based on responses to BOA, or using the term *'normal hearing',* as BOA testing cannot determine this.

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10. VISUAL REINFORCEMENT ORIENTATION AUDIOMETRY (VROA) TEST PROTOCOL

SCOPE

The document sets out to provide guidelines for testing babies with a minimum developmental age of 5 to 7 months. The test is suitable for infants who are able to sit unsupported or with minimal support and who have good head control. It is assumed that the reader of this document is familiar will the principles of VROA. This protocol covers only VROA using one-sided reinforcement (rather than two-sided reinforcement of the conditioned orientation response test).

The document covers the technical procedure of carrying out a manual VROA test, equipment/environment considerations, basic interpretation of the results, reporting and patient handling procedures relevant to the test.

For some aspects of VROA testing, it is recognised that different approaches exist. However, for the purposes of standardisation, a decision to adopt the one described approach has been made for HH purposes. It is advised that only experienced Audiologists (or those led by such colleagues) should consider deviating from this protocol.

QUICK GUIDE: RECOMMENDED PARAMETERS AND SETTINGS FOR VROA

Position of Reinforcers:	90° azimuth
Positioning of Loudspeaker:	90° azimuth At least 1 metre from the test position and approximately level with the child's head.
Stimulus type:	Free Field: Warble tones and/or narrow band noise Headphones / Inserts / Bone conductor: Warble or pure tones
Suggested order of stimulus delivery:	1 kHz → 4 kHz → 500 Hz → 2 kHz
Pass levels:	Sound-field: 25dBHL Insert earphones / headphones: 20dBHL

HEALTHY HEARING VISUAL REINFORCEMENT ORIENTATION AUDIOMETRY QUICK SUMMARY

Procedure:

- Perform otoscopy and tympanometry testing first where possible.
- Decide what information is required (sound-field vs individual ear measures) and choose the appropriate transducer for the testing. The over-arching aim is to determine hearing thresholds for each ear as early as possible.

- Condition the child in order to establish consistent responses to warble tone presentation using behavioural conditioning techniques with visual rewards. This is most commonly performed using a 1kHz warble tone.
- Commence testing once a consistent conditioned response has been established.
- At a minimum, the major speech frequencies (500, 1000, 2000 & 4000Hz) are to be tested.
- Testing at other frequencies may be performed if required and the child's attention allows for testing to continue.
- Bone conduction testing should be performed as clinically indicated whenever a hearing loss is identified. Masked bone conduction results are desirable when indicated, but it is recognised that this is usually not possible to obtain with children under 3 years of age.
- A minimum response level of 25dBHL for sound-field testing is considered functionally normal for HH purposes. When testing has been performed using insert earphones or headphones, the pass level for normal hearing is 20dBHL.

COMPREHENSIVE PROTOCOL INFORMATION

Visual Reinforcement Orientation Audiometry (VROA) is a key behavioural test for young children. It is central to completion of the diagnostic process for those hearing-impaired infants identified by newborn screening. Furthermore, contemporary paediatric amplification fitting methods rely on solid foundations of measurement to ensure the validity and reliability of hearing aid fitting. Also, it is hoped that the standardised VROA technique described here will facilitate training of paediatric Audiologists. This protocol aims to include practical guidance on key issues, particularly around utilisation of the test and interpretation of results.

Throughout this protocol, the audiologist presenting the test tones and visual rewards will be referred to as the Presenter (in the Control Room), and the audiologist in with the child will be referred to as the Observer (in the Test Room). It is recognised that both audiologists play vital roles in testing the child and determining the validity of responses from the child.

TEST FACILITIES & REQUIREMENTS

Rooms

The Test Room should be of adequate size to accommodate the child, parents, and an audiologist comfortably. The test room should offer minimal visual distraction to the child. It is advised that room lights should be capable of being dimmed, in order to permit enhancement of illuminated visual rewards. This is particularly useful for children who have a visual impairment or who are just distractible.

The preferred arrangement is for control of stimuli and rewards to be operated by the Presenter from a separate (control) room. Such an arrangement allows for discreet communication to the

Observer controlling the child's attention, reduces the potential for distraction, and allows for optimum (frontal) observation of the child's behavioural responses by the Observer. The Test and Control rooms should be separated by a one-way window (or alternative arrangements provided, e.g. image on a monitor screen) such that the child is not distracted, yet allowing both the Presenter and the Observer a clear view of the child. There should also be the facility for the Presenter to hear sounds made in the room, to ensure appropriate timing of stimulus presentations.

Calibration

Stimuli presented through 'closed circuit' transducers (headphones, bone conductor or insert-earphones) should be calibrated in accordance with the relevant ISO standards in dBHL. For consistency between HH diagnostic clinics, it is recommended that sound-field rooms are calibrated in dBHL also. Calibration of stimuli presented in the sound field is less straightforward. Most test environments do not provide the ideal anechoic condition and a number of measures have to be taken to ensure that the sound delivered to the patient's ear is accurate and stable. Soundfield calibration requires a considerable knowledge of the use and limitations of sound level meters and soundfield acoustics, and expert help should be sought for the calibration of sound field test rooms and equipment.

A visual examination and listening test should be carried out weekly (Stage A check). Such checks are particularly important for VROA given the variety of stimuli and transducers typically employed and routing of signals between rooms often via additional cable connections. Full calibration of the VROA room is required at initial set-up, annually thereafter, and intermediately following any major changes to the room (e.g. a change to the room layout) or changes in external noise levels. The test environment should be clearly documented with a defined layout of furniture, furnishings, equipment and positions for people in the room during testing. It also recommends that marks be provided to floors and ceilings to ensure that layout and positions remain consistent, as any deviation may compromise calibration.

Transducers and Stimulus Delivery

There are advantages and disadvantages/limitations related to each method of stimulus delivery. However, a full range of transducers should be available for use to ensure complete and accurate assessment of a child's hearing. The transducers available should include speakers for soundfield presentation, supra-aural headphones (e.g. TDH39/49), insert earphones (e.g. EAR-3A coupled with foam tip or to child's own ear mould), and a bone conductor.

The selection of transducer is determined by the audiologists testing the child, and will depend upon what information is required, and what information has already been obtained for the child.

However, use of insert earphones is strongly preferred for those suspected or known to have a permanent hearing loss, in order to obtain reliable ear-specific information while minimising the risk of cross-hearing and the need for masking.

Stimulus Type

For sound-field testing through the speaker, frequency-modulated (warble) tones should be used in the first instance. If the child is unresponsive to warble tones, narrow-band noise can also be used to condition the child to the task, and to gain an estimation of hearing levels. Please note that if testing is conducted using narrow band noise, the child will require repeat testing in a timely manner using warble / pure tones to confirm results.

For testing using headphones or insert earphones, both warble tones and pure tones are acoustically acceptable as a stimulus. For inserts used with a child's ear moulds, actual levels will differ from standard calibration.

Test Equipment and Set-up

It is recognised that a variety of test room arrangements can be employed for VROA. However, the preferred test equipment and set-up using a single speaker and single reinforcement unit arrangement is the basis of the test procedure presented below. The arrangement should allow the Presenter to present live speech to the infant through the sound-field speaker, via a microphone with presentation level controlled by the audiometer intensity attenuator.

Visual Reinforcement – Toys and Illumination

The toys used for reinforcement (e.g. puppets) should be presented within a cabinet. The cabinet should have a smoked perspex or glass front such that the toys are not visually attractive without illumination. A switch in the Control Room should control the illumination of the cabinet (and animation of the toys if the set-up allows). Ideally, a variety of toys or puppets should be provided for reinforcement. Alternatively, a variety of equivalent video images may also be used as reinforcers.

Positioning of Reinforcers

The child should be positioned at a 90° azimuth (or as close to 90° as possible), with the ability to re-position them to reduce the angle if required. A 90° azimuth is used in order to elicit the clearest head turn, while a lesser angle may be appropriate if the child is not developmentally ready for full head turn. The reinforcers should be located approximately level with the child's head at a distance of 1 to 2 metres from the child.

Positioning of Loudspeakers

Loudspeakers should be positioned at a 90° azimuth (RETSPLs are only available for this angle of presentation). The speakers should be positioned at least one metre from the test position (at the same distance to the testing position where calibration was carried out). Speakers should be positioned adjacent to reinforcement apparatus and approximately level with the child's head.

Positioning of Child and Observer

Ideally, all infants tested using the VROA technique should be seated independently, in a secure high chair or appropriate child-sized chair. Very young or distressed infants may be seated on a parent's knee, gently supported at the waist and facing forward (although this is not the preferred option due to potential of parental cueing to sound, and should be used as a last resort). Parents should be seated on the opposite side to the reinforcement (i.e. not between the child and the speaker/reinforcer) and behind the child's line of vision, to minimise potential cueing. The child should be at a point determined and marked during calibration of the sound field. A low table is placed in front of the child to provide a surface for the distracting activity. The Observer is either seated on a low chair on the other side of the table, or kneels in front of the table, facing the child.

Position of Presenter in Control Room

The Presenter should have a clear view of the child's face, the Observer and the parents in the room.

Communication Between Presenter and Observer

Good two-way communication is an essential requirement for the test to be performed efficiently and accurately. Communication from Observer to Presenter should be direct and discreet so as to avoid auditory distractions for the child being assessed.

Hearing Protection for Observer and Parents

The Control of Noise at Work Regulations (2005) stipulate the daily personal noise exposure levels beyond which hearing protection should be used. If daily noise exposure is above the first action level of 80dB(A) but below the second action level of 85dB(A), hearing protection should be available to the Observer. If daily noise exposure is beyond the second action level, or if any peak levels exceed 137dBSPL, then hearing protection must be used.

Calculations for a VROA system with a maximum output of 115dB(A) indicate that the second action level could be exceeded when testing a child with severe or profound hearing loss. In addition, some of the sound levels used may be uncomfortably loud. Hence hearing protection (earmuffs and / or earplugs) should also be available for parents, the Observer and the Presenter. The maximum output at each frequency should be measured and this information used to calculate

likely noise exposure levels according to Queensland Legislative requirements. This information can be used to specify local hearing protection policy.

TEST PROCEDURE

Test Set Up

Following equipment checks, the parent(s) and child are brought into the room and introductions made. History taking provides an opportunity for the child to settle in an unfamiliar environment and for the audiologist to observe the child. If the child is becoming restless, it may be appropriate to cut the history short and begin testing. At an absolute minimum, information should be obtained about the child's developmental and visual status before starting the test.

The test procedure is explained to the parent with suitable cautions about cueing the child to the presence of an auditory stimulus, and the need to minimize distracting noise. A VROA technique using two trained professionals is recommended at all times, unless in extraordinary circumstances. If a trained Observer is not being used, particular attention must be paid to instructions given to the parents remaining in the test room with the child.

If there is any doubt about the child's ability to respond in the desired manner (i.e. with a head-turn) this can be discussed with the parent. If necessary, head control and turning can be checked by having the child visually track an object of interest through an arc of 180°.

Any other adults or children present at the appointment are best invited to sit in the Control room to observe, or can be asked to wait outside if appropriate. If others must remain in the Test Room, they are to be seated directly behind the child being assessed, to minimise distraction and cueing.

The child will be seated in the test position, and care should be taken when positioning parent and child to ensure that soundfield calibration (if relevant) is not compromised. If headphones, inserts or bone conductor are being used, they should be fitted to the child as required. For the bone conductor, an elasticised headband may be used to position the bone conductor in place as a more comfortable alternative to a conventional 'Alice Band'. If the child is resistant to wearing the headphones or bone conductor using conventional fitting methods, they can be hand-held in place by the parent. The Observer needs to ensure that the position of the transducer/s remains optimal throughout testing if the parent is holding them in position by hand.

The Observer will choose a suitable table-top activity (e.g. playing with small quiet toys). The toys selected and manner employed will be the minimum necessary to encourage the child to adopt a midline forward position and maintain alertness. Most importantly, the Observer should provide no change in activity linked to stimulus presentation which could serve as a cue for signal presentation

(e.g. distinct and rhythmical phasing should be avoided). The Observer will also avoid noisy play, and refrain from engaging with the child too fully, except for praising a correct response.

Conditioning

Before testing begins, it is essential that the child understands the task, as demonstrated by consistent responses to sound, though the use of conditioning techniques. Some children will give a clear and repeatable head-turn to an auditory stimulus without any formal conditioning, while others will require a number of conditioning trials.

To commence conditioning, a 1kHz stimulus (without any visual cues) is presented at a level judged adequately supra-threshold. Consideration should be given to the type and degree of hearing impairment anticipated for the child. As a guide, 60 to 65dBHL is a suitable conditioning starting level for routine purposes where no more than a mild loss is suspected. Another frequency may be selected if it is judged that the child is likely to be more responsive at that frequency (e.g. a lower initial frequency would be appropriate if there is suspicion that the child has a severe high frequency hearing loss). If the child gives a clear head turn within 2 to 3 seconds, visual reinforcement is then provided in combination with the test tone, for a further 2 to 3 seconds. This should be replicated for a second time to ensure adequate conditioning prior to the commencement of testing. If the child responds in the same manner to the second tone at the same intensity level, the child can be considered as conditioned and the test sequence can begin.

If the child does not respond spontaneously with a head turn in response to the initial tone, a more formal conditioning procedure is needed. The child may simply require their attention to be directed to the sound and then the visual reward presented shortly after. Additional trials without visual reward may then produce an independent head turn to the test tone alone, with visual reinforcement presented after the head turn response.

If this is still not adequate, both the stimulus and visual reward can be presented simultaneously, and the child's attention directed towards the visual reward. A number of such paired presentations may be required. When a head turn response is reliably elicited to the combined stimulus, conditioning is checked by presenting the auditory signal alone and presenting the visual stimulus as a reward after the head turn response. Once the child is responding reliably to sound alone, they can be deemed to be adequately conditioned and testing can begin.

If the child responds to the combined stimulus/reward but fails to demonstrate a response to the auditory stimulus alone, it may be that the stimulus is insufficiently interesting or is not audible. This can be checked by changing the stimulus, for example, changing the frequency and/or intensity of the stimulus used for conditioning, use narrow band noise, or ultimately attempt

reconditioning using a vibro-tactile stimulus delivered at around 50dBHL at 500Hz (generated via the bone conductor held in the palm of the child's hand) using the paired presentation technique (i.e. both the vibro-tactile stimulus and the visual reinforcement are presented simultaneously).

If the child is not responding to the stimulus/reward combination it may be that the reward is insufficiently visible or interesting. This may be remedied by lowering the room lighting, changing the reward, using two or more rewards in combination. Alternatively it may be that the child is not developmentally ready for the procedure or is not sufficiently motivated by the reward.

If reliable conditioning cannot be established after all techniques outlined above have been exhausted, testing cannot proceed. Reliably conditioned responses to sound are essential in order to obtain accurate and meaningful results from the VROA technique. In this instance, other test procedures will be required to assess the child's hearing.

Testing

When reliable conditioning has been established (at least two consecutive correct unprompted responses), the Presenter will proceed to the test trials proper. Here sound only will be presented for 2-3 seconds. If the Presenter and Observer judge that the child has turned in response to the sound, then visual reinforcement will be presented for 1-2 seconds. The desired response is a clear head-turn to view the reinforcer. Eye glances or small movements should be interpreted with more caution and be reported as such.

False 'checking' responses will be managed by using variable inter-trial intervals, some of long duration. Additionally, the use of deliberate control trials may be employed. Withholding the visual reinforcer for a moment or two after the child turns also may help to distinguish checking glances, which are often short-lived, from real responses.

The '10dB down, 5dB up rule' for stimulus presentation should not be rigidly applied through threshold measurement. Once responses have been established to the initial high level, the level should be dropped as rapidly as possible (perhaps 20dB steps) as long as responses are still observed. The Presenter should determine the presentation level based upon age of the child, attention state, and other factors concerned with time. However, around the estimated threshold, the '10dB down, 5dB up rule' should be adopted. The criteria for threshold will be 2 out of 3 responses at any level. The minimum response level at one frequency should be defined before moving to another frequency where possible.

The initial and subsequent test frequency will vary for each patient according to information obtained by previous methods and the need for further information. When changing stimulus

frequency, present the initial stimulus at a level judged to be above threshold. It may also be helpful to present clear supra-threshold stimuli or re-condition a child who has become distracted. For a child who is restless or bored, it may be possible to maintain/restore interest by using a combination of warble tones and narrow-band noise, randomly changing frequency, and increasing or varying the visual reward (e.g. changing toys or multiple toys).

It may be useful to measure speech detection thresholds using live speech, in order to provide some validation of the information obtained from electronically generated stimuli. The Presenter should talk to the child through the sound-field speakers, using their name frequently, while slowly raising the level from around 20dB(A) (using the audiometer intensity attenuator), until a response is seen. The recorded level in dB(A) can be compared with the average minimum response level for the child.

Tips for Effective VROA Testing

- The procedure relies on the continued cooperation of the child, in particular their ability to stay in the required test position. Time to complete the testing will therefore be limited. To avoid delay/disruptions, ensure that all required equipment is checked in advance prior to bringing the child into the test room (stage A calibration checks are completed, reward system operating and communications equipment ready for immediate use).
- Some children may be upset by certain animated toys. If so, reward through simple illumination rather than animation or switch to alternative toys.
- To extend interest in responding, switch reward toys and/or use in combination. Also be prepared to take a break from testing and return to complete the assessment, or switch testers. The interest of older children in particular may be extended by praise/encouragement of correct head turn, provided by the Observer.
- Towards the end of the test procedure, return to the first frequency tested and present at MRL (or 5dB above that dial level) to check if the child still responds. This information will help the Presenter judge the validity of the later responses.

The Most Common Pitfalls of VROA Testing

- Inadequate test set-up and communication between Presenter and Observer.
- Attempting conditioning to sub-threshold stimuli.
- Not establishing clear responses at supra-threshold levels before descending to threshold.
- Incorrect scoring as true responses (i.e. scoring of movement other than a clear head-turn, or false positive (checking) responses).
- Distinct and/or rhythmical phasing of attention by the Observer such that response cues are given to the patient.

- Use of toys or behaviour by the Observer (or parent), that are too distracting for the child and so inhibit responses to sound.
- Over-emphasis on quantity of results (number of thresholds obtained) rather than quality (reliability) of those thresholds obtained.
- Not using time efficiently, often spending too long at high intensities.
- Inaccurate interpretation and reporting of results due to inadequate consideration of differences in infant MRLs compared to adult normative (threshold) values (see below).

Testing Children with Vision Disorders, other Disabilities, or at an Early Age

Visual disability may interfere with conditioning and responses. Consider bringing the reward closer to the child, or consider using more visually contrasting rewards (e.g. bright flashing light). Removal of the smoked perspex cover on the reinforcer unit should be considered. Dimming the room lights will also increase the contrast. For the more severely visually impaired, use of other sensory reinforcement such as air puffs, vibratory stimulation or music may be needed to bring children under stimulus control.

General developmental delay may not interfere with VROA. However, motor difficulties may obscure head-turn responses. A more flexible approach to response reward and interpretation may be appropriate. However, any deviations from the standard approach should be documented and described when reporting.

Although VROA is generally reliable in the assessment of normally developing children from 30 weeks corrected age, some infants may be testable at younger ages, from approximately 20-26 weeks corrected age. Testing at this age may be required because of parental or professional concern, or may be of particular value to early diagnosis and habilitation. However, for younger children it should be recognised that a sequence of test appointments may be required to incrementally gain the information required (e.g. a series of frequency and ear specific MRLs).

For children with disabilities or where VROA is used speculatively at an early age, a realistic appraisal of the likelihood of test success should be presented to parents/carers before testing. Presenters should also seek the advice of parents/carers in advance of assessment to determine the appropriate test strategy. For more detailed information on conducting VROA for children with disabilities, clinicians are advised to refer to Coninx & Lancioni (1995).

Sequence and Objectives of Assessment

The sequence of assessment should be adapted depending on the objectives of the Audiologist and the status of the child. However, the testers must be aware that the co-operation and interest of the child may fail at any time and this should be reflected in the sequence of assessment. The

clinically more important information should be obtained first. As a guide the following sequence of frequencies are suggested for an initial formal behavioural assessment, commencing with stimuli presented in the sound-field: 1 kHz → 4 kHz → 500 Hz → 2 kHz.

Sound-field testing could be followed by delivery of stimuli through insert earphones where ear specific information is required, or where results will be used to guide amplification. Bone conduction testing may also be required, albeit with awareness of increased likelihood of vibro-tactile responses at the lower frequencies compared to adults. If ear-specific thresholds are desired and the use of insert earphones is contra-indicated (e.g. due to wax, microtia), the use of a hand-held, single TDH earphone could be considered.

The timescale for acquisition of MRLs should be considered carefully. On the basis that quality of results takes prominence over quantity (of MRLs) consideration should be given to arranging a sequence of appointments, particularly where a large quantity of information is required, where the child is only just at sufficient developmental age for VROA testing, or has relevant disabilities that may impact upon testing. An appointment duration of 30 to 45 minutes would be typical to obtain soundfield VROA results.

INTERPRETATION OF RESULTS

The process described previously provides calibration to adult norms for a conventional audiometric technique. There are no specific international standards on the RETSPL values for stimuli used for VROA. Audiologists should be mindful of the influence of age of the child and the test method employed when interpreting and reporting results.

There are numerous factors contributing to the known difference between infant VROA MRLs and adult normative thresholds. Normally hearing infants performing VROA require a higher intensity stimulus to induce a response (e.g. a head turn) than that required for normally hearing adults performing pure tone audiometry. Although some studies have investigated and reported on the difference between MRLs obtained by VROA in infants and adult threshold normative data, the data set (relating to test frequency, age of subject and type of transducer) is far from complete. Further studies are required to confirm and build upon this knowledge base before we can endorse a series of specific correction factors for VROA. With due consideration of the above, the information presented below represents current information on the scale of infant-adult correction factors based upon the mode of stimulus delivery, stimulus type and age. Consequently, the correction values indicated are provisional at this time.

Sound-field

Information available on the relationship between adult thresholds and MRLs for soundfield VROA test indicates that normally hearing infants (aged 7-12 months) present mean thresholds at approximately +10dB relative to the adult normative RETSPLs (from 0.5kHz to 4kHz). This means that a correction of -10dB needs to be added to the infant's MRL to convert to an "adult dBHL" value. It needs to be recognised however that children older than 12 months are assessed using this technique, and any "pass" level needs to be appropriate for their MRLs.

Therefore, it is suggested that when testing in the soundfield, hearing should be tested down to at least 25dBHL (equivalent to adult 15dBHL). Responses at this level are accepted as indicative of functionally normal hearing for the better hearing ear. Such guidance should not discourage testing down to lower levels where possible. Those interpreting and reporting results should be mindful that sound-field assessment only indicates the hearing status of the better hearing ear at each test frequency. Testing of individual ears should be pursued to ensure that infants with mild and unilateral hearing losses are detected.

Insert Earphones

Information available on the relationship between adult thresholds and MRLs obtained using insert earphones indicates that normally hearing infants (aged 7-12 months) present mean thresholds higher than adult normative RETSPLs (from 0.5kHz to 4kHz). The difference appears to vary as a function of frequency, and the differences are outlined in table below.

Comparison of infant MRLs for insert earphone VROA studies (normal hearing infants).

Reported Studies	**Age Range**	**Mean MRLs in dBHL (standard deviation)**			
		500Hz	**1000Hz**	**2000Hz**	**4000Hz**
Nozza (1995)	6-11 months		14.1		
Nozza & Henson (1999)	6-11 months	17.2		6.8	
Parry et al (2002)	8-12 months	16.4 (5.9)	13.3 (6.1)	7.1 (5.5)	6.4 (6.2)
Recommended Provisional Corrections (subtracted from MRL to obtain "adult dBHL")		**-15**	**-15**	**-5**	**-5**

The recommended correction values presented may be used to convert infant MRLs into values that are more comparable to an adult audiogram. Correction of raw MRLs achieved by insert earphone VROA may be appropriate when estimating the audiometric profile, which is required when using prescription formulae to fit hearing aids.

It should be noted that studies have not been performed in infants / children with sensorineural hearing loss, or in children older than 12 months of age.

Bone Conduction

No studies have been identified relating to bone conduction VROA MRLs.

REPORTING OF RESULTS

Reporting of results should be clearly accompanied by a description of the type of transducer, the dB scale used, and the reliability of assessment. Where recorded results represent the MRLs (dial settings) this should be indicated. Similarly, the nature of any MRL threshold corrections applied (to guide interpretation or to provide an estimated audiogram for the purpose of prescription for amplification) should be indicated. The Audiologist leading the VROA procedure should be responsible for ensuring that results are appropriately documented and reported upon.

Finally, guidance on the matter of MRL to adult threshold corrections may change with the outcome of further research. The use of this (standardised) VROA protocol in further research studies around VROA MRLs is encouraged in order to facilitate future transferability of findings.

STAFF TRAINING AND EXPERTISE

Staff engaged in performing VROA testing should have received specific training and documented assessment to demonstrate their competency to perform this specialised test. The level of competency should be at least adequate for the role performed (whether supporting or leading the assessment). Those leading the assessment should be competent in briefing and debriefing carers, use of the equipment, and the correct interpretation of results to ensure appropriate recording and reporting of results for use by oneself and others. To ensure correct reporting of results for interpretation by others, it is particularly important that the difference between MRLs obtained by VROA and adult normative thresholds are recognised and understood.

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11. PURE TONE AUDIOMETRY (PTA) TEST PROTOCOL

SCOPE

This protocol aims to provide recommendations for conducting Pure Tone Audiometry (PTA) testing for the determination of behavioural hearing thresholds in young children. It also aims to provide guidelines for the consistent reporting of degree of hearing loss. The accurate determination of hearing thresholds is critical to ensure appropriate management of the child with hearing loss.

PTA testing using play audiometry techniques is typically able to be performed on children from a developmental age of 2.5 to 3 years.

QUICK GUIDE: RECOMMENDED PARAMETERS FOR PURE TONE AUDIOMETRY

Stimulus Type:	Pure tones should be attempted first. If child not responding consistently, consider using pulsed tones, and warble tones as a last resort.
Minimum Frequencies:	Octave frequencies between 500Hz and 4000Hz. 250Hz and 8000Hz desirable if child's attention allows. Inter-octave frequencies should be tested where hearing thresholds differ by 20dB or more between adjacent octaves.
Ears:	Both ears to be tested.
Transducers:	Headphones or insert earphones may be used for air conduction testing. Bone conduction testing to be performed as indicated.
Pass criteria for 3.5 year Targeted Surveillance appt	Hearing thresholds of 20dBHL or better in both ears at 500, 1000, 2000 and 4000Hz.

HEALTHY HEARING PURE TONE AUDIOMETRY QUICK SUMMARY

Procedure:

- Perform otoscopy, tympanometry and TEOAE (if applicable) testing first where possible.
- Condition the child in order to establish consistent responses to tone presentation using behavioural conditioning techniques (play audiometry).
- Commence testing once a conditioned response has been established. Testing is performed using the Hughson-Westlake ascending test technique.
- Air conduction testing should be performed for both ears. At a minimum, the major speech frequencies (500, 1000, 2000 & 4000Hz) are to be tested. Testing at 250 and 8000Hz should be considered if the child's attention allows for testing to continue.
- Bone conduction testing should be performed as clinically indicated whenever a hearing loss is identified. Masked bone conduction results are desirable when indicated, but it is recognised that it is not always possible to obtain, particularly with children under 3-4 years of age.
- Where screening is necessary, a screening level of 20dBHL or lower should be selected. Screening using intensities at and above 25dBHL is not recommended, in the interests of

ensuring that mild hearing losses are not missed. Where screening has been performed (rather than threshold seeking), this should be documented on the audiogram.

COMPREHENSIVE PROTOCOL INFORMATION

For paediatric testing, it is recommended that otoscopy, tympanometry and TEOAE testing (where applicable) is performed prior to commencing PTA testing. The results of these tests should be used to guide the audiologist in determining which ear to test first (the ear that is most likely to be the better hearing ear), and to determine an appropriate intensity level to commence the procedure at, in order to maximise the chances of the child conditioning appropriately for the test.

Transducers

Air conduction testing can be performed using either headphones (TDH 39/49) or insert earphones (EAR-3A). In instances where there are unexplained high frequency air bone gaps using headphones, the audiologist must be alert to the possibility of a collapsed ear canal producing an artefactual conductive hearing loss. Retesting of air conduction thresholds should be performed using either an insert earphone (preferably) or a hand-held headphone to rule out collapsed canals as the cause of the hearing loss measured.

Stimulus

Testing should always be attempted using pure tones in the first instance. However children (particularly younger children) may be less responsive to pure tones than other tones such as pulsed tones or warble tones. When a child will not respond reliably to pure tones, pulsed tones and warble tones can be utilised to attempt to achieve more consistent responses to sound. Where pure tones have not been used for testing, it should be clearly indicated on the child's audiogram.

Conditioning

The child should be conditioned at clearly audible levels using play audiometry techniques. Conditioning should be continued until such time that consistent responses to tone presentation have been established. For a child with suspected normal hearing, an appropriate starting level for conditioning would be 40-50dBHL at 1kHz. Conditioning at higher levels is often required for children with a hearing loss.

Verbal instructions should not be given for play audiometry for children under 5-6 years of age. Rather, the child should be conditioned using behavioural conditioning techniques, as this means that children with underlying language disorders or language processing difficulties can be accurately and reliably assessed without misunderstanding the test procedure.

If the child is not conditioning to the task, the audiologist must consider the possible reasons for the lack of conditioning, including hearing loss, intellectual impairment and developmental delay. Where hearing loss is suspected, the audiologist should try a number of things with the aim of producing a consistent response to the test tones:

- Swap ears and re-attempt conditioning (in case the poorer ear has been chosen to commence conditioning, or child has a unilateral hearing loss).
- Change the frequency of the tone used for conditioning (e.g. try a lower or higher frequency in case child has a sloping loss in either direction).
- Use a pulsed or warble tone instead of a pure tone.
- Increase the intensity of the test tone used for conditioning (within safe levels).
- Attempt conditioning with the bone conductor (better ear always captured).
- Attempt conditioning with the bone conductor held in the child's hand at high intensity levels, to produce a vibrotactile sensation. If vibrotactile conditioning is successful, this indicates that the child cognitively grasps the concept of responding behaviourally to a stimulus. In this instance, conditioning should be re-attempted using auditory stimulation, with the suspicion that the child has a hearing loss and the tones used for conditioning previously were likely below the child's hearing thresholds.

For children who cannot be reliably conditioned after all of the above have been attempted, PTA testing (play audiometry) should be abandoned, and the audiologist should attempt testing using a VROA technique (refer to VROA protocol).

Testing

Testing can commence once the child has been conditioned, and reliable consistent responses to sound are being observed. Testing is performed using the internationally accepted Hughson-Westlake procedure. At a minimum, both ears should be tested at 500, 1000, 2000 & 4000Hz. The inter-octave frequencies should be tested where there is a difference of 20dB or greater between adjacent octave frequencies.

Testing should also continue to 250 and 8000Hz if the child's attention allows. However this should not be the priority if a hearing loss is indicated, as bone conduction testing would be deemed more of a priority to establish the type of hearing loss present, before the child loses attention/interest in the task.

Bone conduction testing should always be performed when a hearing loss is indicated on the audiogram in order to determine the type of hearing loss measured.

Masking for both air conduction and bone conduction thresholds should be used where clinically indicated, when the child is capable of performing masked testing reliably. It is acknowledged that masking with younger children is not always possible. If masking has been attempted but was unsuccessful, it should be documented clearly on the child's audiogram.

Reporting of Hearing Thresholds

Hearing thresholds of 20dBHL or better are considered to be within the normal range.

For consistent reporting of the degree of hearing loss for the Healthy Hearing Program, the descriptors in the following table should be used.

Description of hearing levels	Threshold Range in dBHL
Normal	-10 to 20dBHL
Mild	21 to 40dBHL
Moderate	41 to 55dBHL
Moderately-severe	56 to 70dBHL
Severe	71 to 90dBHL
Profound	>90dBHL

PART D

FORMS / RESOURCES

ABR Preparation Instructions for Parents

Audiology Diagnostic Checklist

ABR Testing Quick Guide

QHLFSS Referral Form

Notification of Unexpected Hearing Outcome

Notification of Unaided Child Requiring Annual Monitoring

CMV PCR Pathology Request Form

ATTENTION PARENTS

PLEASE READ BEFORE THE DAY OF YOUR BABY'S HEARING TEST APPOINTMENT

Preparation Instructions for Babies for ABR Testing

Your baby has been referred for a hearing test. The test your child is having is called an ABR (Auditory Brainstem Response) test. The test uses electrodes (sticky tabs) placed on your baby's head to record tiny responses from your baby's hearing nerves. This testing requires your baby to be asleep. Testing cannot be completed unless your baby is asleep for the test.

The appointment takes approximately 2 to 3 hours.

It is important that your baby arrives at the appointment tired and hungry. We do not want your baby arriving at the appointment already asleep, as this will reduce the amount of time the audiologist may have to get results for you on the day.

To assist us in getting your baby ready for testing, please:

- Keep your baby awake for the 2 to 3 hours leading up to the appointment.
- Do not let your baby fall asleep in the car on the way to the appointment. Even a short nap may mean that your baby will not sleep for the ABR appointment. Bringing another adult with you to keep the baby awake during the trip can be very helpful.
- Delay your baby's feed until you arrive for the appointment. Time is allocated during the appointment for you to feed your baby and for your baby to fall asleep.
- Avoid using moisturisers or oils on your baby's skin on the day of the appointment, to make sure that the electrodes will stick on your baby's skin properly for testing.

Tired babies with nice full tummies are very likely to fall asleep for testing to start.

It is essential that you do not bring other children to the appointment, as it is a very long appointment and the test needs to be performed in quiet conditions. Child minding facilities are not available, so it is best to plan for your other children to be left with a suitable carer.

HEALTHY HEARING CHECKLIST – DIRECT REFER

NAME: _____ DOB: _____ DATE: _____

CLINICAL:		**ADMIN:**	
Click ABR: wave V threshold obtained (25dBeHL)		ABR traces co-signed	
Click ABR: wave I, III, V clearly identified (65dBeHL)		QChild data entered	
Click ABR: intermediate intensity tested (45dBeHL)		Review appointment booked	
1 & 4k pass criteria met (TEs or TB/pip/chirp)		Surveillance referral completed	
HF tymp performed		Release of Info Form signed if needed	
FN hrg indicated no risk factors = Discharge		Unexpected Outcome Form completed if applicable	
FN hrg indicated with risk factors = Surveillance		Results documented in patient medical record	
Hearing loss indicated (see below): continue testing to obtain as much info as possible.		Results documented in baby's Personal Health Record (red book)	
HEARING LOSS INDICATED:		**REPORTS – INFORMATION TO BE INCLUDED:**	
Freq-spec AC ABR thresholds obtained (0.5-4k)		Child's Name, Address and DOB	
Freq-spec BC ABR thresholds obtained		Mother's Full Name	
TEOAEs repeated		Screening Hospital & UR Number	
Tymp / Reflexes		Date of Assessment	
Cochlear Microphonic (sev to prof SNHL)		Corrected Age at Assessment	
ASSR (sev to profound losses if needed)		Screening Results	
Behavioural testing as indicated		Risk Factors	
Click repeated if required		Gestational Age at Birth	
Written info provided to parents		Relevant case history information	
		Transducer & correction factors noted	
MANAGEMENT / REFERRALS:		**REPORTS – TO BE SENT TO:**	
Normal hearing (no risk factors) = Discharged		Parents	
Normal hearing with risk factors = Surveillance		GP	
Hearing loss indicated = Needs review appt		Screening Hospital Site Co-ordinator	
		QHLFSS	
HEARING LOSS CONFIRMED:		Medical Chart / Patient Record	
Refer to CHC <12 mths (Paed, ENT, AH, SP)		CHC (if appropriate)	
Refer to AH		ENT (if appropriate)	
Refer to ENT		Paediatrician (if appropriate)	
Notify QHLFSS – manual referral if ETS baby		Australian Hearing (if appropriate)	
Notify CI Program (sev to prof SNHL indicated)		Cochlear Implant Program (if appropriate)	
Reviews arranged if not being aided by AH		Other:	
Sibling hearing tests recommended		Other:	

ABR TESTING QUICK GUIDE

Click ABR Test Levels to Meet Discharge Criteria – in dBnHL "dial"		
Test Level Required	**Headphones / Inserts > 24 weeks**	**Inserts < 24 weeks**
25dBeHL	30dBnHL "dial"	20dBnHL "dial"
45dBeHL	50dBnHL "dial"	40dBnHL "dial"
65dBeHL	70dBnHL "dial	60dBnHL "dial"

HH Pass Levels – In dBnHL "dial"						
Transducer	**Age**	**Click**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Insert earphones	<24 weeks	20	35	30	25	20
Insert earphones	>24 weeks	30	40	35	30	30
Headphone	Any age					

HH Correction Factors – conversion from dBnHL "dial" to dBeHL (to be added to dial reading)						
Transducer	**Age**	**Click**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
Insert earphones	<24 weeks	+5	-15	-10	-5	0
Insert earphones	>24 weeks	-5	-20	-15	-10	-10
Headphone	Any age					
Bone conductor	<24 weeks		+5	+5	-5	0
Bone conductor	24 weeks – 2 years		-5	-5	-10	-10

<u>* TESTING REMINDER *** :</u> If click ABR is abnormal at or above 75dBeHL, CM testing must be completed to exclude ANSD.**

Click ABR - Correction Factors for Comparison to Latency Normative Data (for comparison to age appropriate norms– to be added to dBnHL "dial")		
Transducer	**Age**	**Correction Factor**
Insert earphones	<12 weeks	+10
Insert earphones	>12 weeks	0
Headphone	Any age	0

Reporting of Degree of Hearing Loss for HH Purposes (TB AC ABR)	
Pass Level (20dBeHL)	Functionally normal
25-40dBeHL	Mild
45-55dBeHL	Moderate
60-70dBeHL	Moderately-severe
75-90dBeHL	Severe
≥95dBeHL	Profound

Children's Health Queensland Hospital and Health Service

Queensland Government

Queensland Hearing Loss Family Support Service

Referral

(Affix patient identification label here)

URN:

Family Name:

Given Names:

Address:

Date of Birth: Sex: ☐ M ☐ F ☐ I

Please use this form for children who have NOT been referred via the Healthy Hearing Newborn Screening Program to the Queensland Hearing Loss Family Support Service. This will include children with:

1. Permanent hearing loss identified through the Healthy Hearing Targeted Surveillance program
2. A diagnosis of 'later onset' or acquired permanent hearing loss

NAME OF REFERRER:

☐ Audiology ☐ Paed/ENT ☐ EQ AVT ☐ GP ☐ Other:

Referrer address:

Referrer phone: **Email:**

Audiologist and clinic attending (if different from referrer):

Child's full name: **Child's DOB:**

Mother's name: **Phone:**

Father's name: **Phone:**

Address:

✶ Please attach audiology report, previous referral information, and any other relevant documentation ✶

HEARING INFORMATION

*Reason for original referral to referrer, dates of assessment, **type / degree of hearing loss**, medical / developmental issues e.g. syndromes, pregnancy and birth complications, aetiology, family history of HL*

PSYCHOSOCIAL FACTORS

Family history, relationships, medical, mental health, financial / housing, support, other current stressors

Is the family aware of the referral **AND** have they given consent for QHLFSS contact? ☐ Yes ☐ No

NON HEALTHY HEARING CHILDREN ONLY: Consent to Healthy Hearing Data Collection (Qchild)? ☐ Yes ☐ No

Please email or fax the completed referral form to:

Brisbane
Ph: (07) 3310 7809
Fax: **(07) 3310 7808**

Townsville
Ph: 1800 352 075
Fax: **(07) 4724 1480**

Email: **QHLFSS@health.qld.gov.au**

Referrer's signature:

Date:

DO NOT WRITE IN THIS BINDING MARGIN

QHLFSS REFERRAL

v1.00 - 07/2016

00007:658924

Notification of Unexpected Hearing Outcome – Healthy Hearing Program

Use this form to report PCHL of any degree not identified by Newborn Hearing Screening.

To be completed by Audiologist and returned to HH Team by: fax 3310 6233, or by email healthy_hearing@health.qld.gov.au. Parental consent is required before returning this form.

CHILD DETAILS			
Surname:		**First Name:**	
DOB:		**Birth Hospital:**	
Mother's full name (at time of birth of child):		**Birth Hospital UR (if known):**	

NEWBORN SCREENING RESULTS						
Right ear:	□ **Pass**	□ **Refer**	□ **Medical Exclusion**	□ **Missed**	□ **Declined**	□ **Unknown**
Left ear:	□ **Pass**	□ **Refer**	□ **Medical Exclusion**	□ **Missed**	□ **Declined**	□ **Unknown**

High Risk Indicators Present? □ **No** □ **Yes (please provide details)**

HEARING LOSS INFORMATION (please provide copies of all assessment results and reports with this form)**				
Date referred for assessment				
Referral source				
Referral reason				
Date hearing loss confirmed				
Type of hearing loss	**Right ear:**		**Left ear:**	
Degree of hearing loss	**Right ear:**		**Left ear:**	
Date referred to Aust Hearing				
Date aided	**Right ear:**		**Left ear:**	
Likely onset (please circle):	**Right ear:**	□ **Congenital** □ **Acquired – post meningitis** □ **Acquired – other** □ **Possible/probable late onset** □ **Possible/probable progressive** □ **Unknown** □ **Other** _____ _____	**Left ear:**	□ **Congenital** □ **Acquired – post meningitis** □ **Acquired – other** □ **Possible/probable late onset** □ **Possible/probable progressive** □ **Unknown** □ **Other** _____ _____

RELEVANT CLINICAL HISTORY

Please provide any relevant history, including medical conditions, illnesses, aetiological investigations. Include information regarding parents' opinion regarding duration / onset of hearing loss.

REPORTING CLINICIAN

Notification of Unaided Child Requiring Annual Monitoring – Healthy Hearing Program

Use this form to notify a diagnostic clinic of an unaided child with diagnosed hearing impairment requiring on-going monitoring of hearing status.

To be completed by the Australian Hearing Audiologist, and returned to HH Team by email to healthy_hearing@health.qld.gov.au.

CHILD DETAILS

Surname:		**First Name:**	
DOB:		**Birth Hospital:**	
Mother's full name (at time of birth of child):		**Birth Hospital UR (if known):**	
Address:			
Phone Numbers:			
Diagnostic Audiology Site (where child's hearing loss was detected):			

REQUEST DETAILS – PLEASE PLACE AN 'X' IN THE APPROPRIATE BOX

The above child requires annual audiological monitoring until 5 years of age for the following reason:

☐	Child has a permanent unilateral hearing loss and has not been fitted with an amplification device
☐	Child has a permanent mild bilateral hearing loss and has not been fitted with an amplification device
☐	Child has been identified with unilateral ANSD and has not been fitted with an amplification device
☐	Child has been identified with bilateral ANSD and has not been fitted with an amplification device
☐	Child has a confirmed permanent hearing loss (not ANSD, unilateral or mild) and has not been fitted with an amplification device

Diagnostic Site requested to perform monitoring:	

ADDITIONAL INFORMATION

Please provide any relevant additional information for the diagnostic audiology site:

AUSTRALIAN HEARING AUDIOLOGIST

Audiologist:			
AH Centre:		**Email:**	
Phone:		**Date:**	

Pathology Queensland

Queensland Government

Patient Details

FACILITY

GENDER UR PREFIX UR NO DATE OF BIRTH

☐ M ☐ F

PATIENT SURNAME (Please print or place sticker on this area) PATIENT FIRST NAME

PATIENT ADDRESS

Medicare Details

Patient status at the time of the service or when specimen collected (please tick) Yes

- ☐ Private patient in a private hospital or approved day hospital facility
- ☐ Private patient in a recognised hospital
- ☐ Public patient in a recognised hospital
- ☐ Outpatient in a recognised hospital

PHLEBOTOMY USE ONLY

MEDICARE NUMBER

HEALTH FUND NAME

VETERANS AFFAIRS IRN

Indigenous status
Aboriginal ☐ Non
TSI ☐ Indigenous ☐
Both ☐ Not stated ☐

MEDICARE ASSIGNMENT FORM (Section 28A of the Health Insurance Act 1973) I offer to assign my rights to benefits to the approved pathology practitioner who will render the requested pathology service(s), and any eligible pathologist determinable service(s) established as necessary by the practitioner

Patient Signature X Date / /

PRACTITIONERS USE ONLY (Reason patient cannot sign)

Birth Hospital Info

HOSPITAL REGISTRATION DETAILS Hospital of Birth

Surname Child Date Of Birth

Given Names Child Gender

UR Numberof Child

Surname Mother

Given Names Mother

Privacy Note: The information provided will be used to assess any Medicare benefit payable for the services rendered and to facilitate the proper administration of government health programs, and may be used to update enrolment records. Its collection is authorised by the provisions of the *Health Insurance Act 1973*. The information may be disclosed to the Department if Health and Ageing or to a person in the medical practice associated with the claim, or as authorised/required by law.

REC'D TIME INITIALS

☐ I have signed the above assignment to elect to have my pathology services bulk billed to Medicare. Code CONVM Billing Cat PA

Your doctor has recommended that you use Pathology Queensland. You are free to choose your own pathology provider. However, if your doctor has specified a particular pathologist on clinical grounds a Medicare rebate will only be payable if that pathologist performs the service. You should discuss this with your doctor.

DOCTORS: Please complete ALL relevant areas in the red section

LAB NO

WARD/ CLINICAL UNIT

LAB USE ONLY

Request Details

TEST REQUESTED

CMV PCR testing from Newborn Screening Card

CMVPCR

No other testing can be ordered on this form

CLINICAL NOTES/MEDICATIONS GESTATIONAL AGE K=

Permanent Hearing Loss? ☐ Yes ☐ No

Diagnostic Audiology Service*:

*Queensland Health Healthy Hearing program will only fund testing that is within program criteria. If this information is not indicated, the patient may be privately billed.

Address of Requesting Practitioner*:

CONSULTANT/SENIOR MEDICAL OFFICER SURNAME (Please print) INITIALS

Doctor

SURNAME OF REQUESTING DOCTOR (Please print) AUSLAB CODE

FIRST NAME PROVIDER NUMBER

Requesting Doctor's Signature X Date Requested / / Self Determine ☐

URGENT ☐ TEL ☐ PAGE ☐ FAX ☐ CONTACT NO

Copy

COPY REPORT TO: SURNAME (Please print) INITIALS

COPY REPORT TO ADDRESS

Parental / Guardian Consent:

I, (mother/father/guardian) give permission for a blood spot from the Newborn Screening Card of the above mentioned child, which was collected on the birth of the abovementioned child and is currently held by the Newborn Screening Unit of Pathology Queensland, to be released to the Molecular Diagnostic Unit, Pathology Queensland and to be tested for Cytomegalovirus nucleic acid. I understand that the Healthy Hearing Program will be notified of the results and I may be contacted by staff associated with the Healthy Hearing Program. I also understand that de-identified data may be used for statistical reporting and research purposes. In signing this form, I confirm that I am the next of kin or the legal guardian of the child and the child has been diagnosed with a permanent hearing loss. I also acknowledge that the Molecular Diagnostic Unit will return the Newborn Screening Card and any unused specimens to the Newborn Screening Unit on completion of the test and that the specimen will not be used for any other testing without the written permission of the child's next of kin or legal guardian. I understand that although the identification of Cytomegalovirus nucleic acid on this test may indicate a possible cause of the child's permanent hearing loss it does not exclude other possible causes of the permanent hearing loss and that other testing may also be recommended for the child. I may be contacted if the Newborn Screening Card is not located using the information above

Signature (mother/father/guardian) ..... **Date** ..... /..... /..... **Contact Number**.....

The Healthy Hearing program will only accept the expenses of CMV PCR testing from Newborn Screening Card for children who are: 1) Diagnosed with permanent hearing loss and 2) 5 years or younger. If outside these criteria please invoice privately.

Signature (mother/father/guardian) ..... **Date** ..... /..... /..... **Billing Address**.....

Service	Funding Currently Available (subject to eligibility)	Cost if Unfunded
CMV PCR Testing	Healthy Hearing Program	$46.44
Screening Card Retrieval	Healthy Hearing Program	$40.50

Requesting Practitioner's Statement:

- I have explained to the mother/father/guardian the testing procedure, its purpose and its limitations
- I have given the mother/father/guardian an opportunity to ask questions about any of the above matters and raise any other concerns which I have answered as fully as possible. I am of the opinion that the mother/father/guardian understood the information provided.

Signature (Requesting Practitioner) ..... **Date** ..... /..... /..... **Contact Number**.....

Was an interpreter required? Yes/No If yes, was an interpreter present? Yes/No

Please complete and return via email to:
Healthy_Hearing@health.qld.gov.au

To contact Healthy Hearing: Phone 07 3310 6220

Healthy Hearing Criteria Met? ☐ No ☐ Yes - Healthy Hearing Billing HHP