It is currently estimated that 7.5 million children 5 years old or younger have disabling hearing impairment (>35 dB HL) worldwide, the vast majority (80% or more) of whom reside in developing countries. Of the 70 million babies born yearly in Sub-Saharan Africa and South Asia, for example, about 420,000 or 6 per 1000 live births are likely to have permanent congenital or early-onset hearing loss (PCEHL) compared with less than 28,000 or 2 per 1000 live births in developed countries. Without timely detection and appropriate intervention, these children are unlikely to acquire optimal speech and language skills essential for their education and vocational attainment in today’s communication world.

Newborn hearing screening (NHS) in the first month of life is crucial for facilitating early hearing detection and intervention (EHDI) of significant PCEHL including neural loss (auditory neuropathy/dysynchrony). The goal of early detection is to allow as little time as possible to elapse between the onset of hearing loss and its detection and subsequent management with a view to minimizing auditory deprivation while maximally stimulating auditory development during the peak period for neural growth. Since early auditory stimulation is the foundation for optimal speech and language development in the first year of life, EHDI facilitates linguistic competence and literacy development for deaf and hard of hearing children.

Evaluating the performance of hearing screening tests

The process of screening should identify infants with PCEHL for whom further action is warranted (test-positives) and infants without PCEHL for whom no further action is warranted (test-negatives).

It is highly unlikely that any hearing screening test can accurately distinguish all infants with PCEHL from those without, due to the inherent differences in biomedical investigation and test algorithms.

Consequently, a hearing screening test usually results in four main outcomes:

1. Infants with PCEHL accurately identified (True-Positives)
2. Infants without PCEHL accurately identified (True-Negatives)
3. Infants with PCEHL not accurately identified and classified as having normal hearing (False-Negatives)
4. Infants without PCEHL not accurately identified and classified as having abnormal hearing (False-Positives)

The performance of an infant hearing screening test based on these four outcomes can be further evaluated on the basis of the following parameters:

- **Sensitivity**: Probability of a positive test in children with hearing loss or the percentage of children with hearing loss correctly detected
- **Specificity**: Probability of a negative test in children without hearing loss or the percentage of children without hearing loss correctly detected as having normal hearing
- **False Positive Rate (FPR)**: Probability of a child without hearing loss testing positive or the percentage of children without hearing loss who had positive test results
- **Positive Predictive Value (PPV)**: Probability of a child having hearing loss when the test is positive or the percentage of those with positive test results who actually have hearing loss
- **Positive Likelihood Ratio (PLR)**: Likelihood that a positive test result
will be found in patients with hearing loss compared to patients without hearing loss. In effect, PLR tells us how much more likely a positive test is to be found in patients with hearing loss as opposed to patients without hearing loss.

- **Negative Likelihood Ratio (NLR):** Likelihood that a negative test result will be found in patients without hearing loss compared to patients with hearing loss. In effect, NLR tells us how much more likely a negative test is to be found in patients without hearing loss as opposed to patients with hearing loss.

**An ideal hearing screening test** should be simple to apply, safe, reliable and valid. It’s reliability is dependent on providing consistent results while validity involves the accurate detection of the majority of children with hearing loss (high sensitivity) without designating most children without hearing loss as failing the test (high specificity). In considering, the population of infants with positive results, the percentage of infants without hearing loss will be very low (low FPR) while the percentage of those with hearing loss will be very high (high PPV).

**Like most screening tests, hearing screening cannot offer a guarantee of protection against PCEHL.** In any screening test, there is an irreducible minimum of false positive results (wrongly reported as having the condition) and false negative results (wrongly reported as not having the condition).

**Tools for hearing screening in infants**

**Otoacoustic emissions (OAE)**

Otoacoustic emissions are low intensity sounds generated from the outer hair cells of the cochlea in response to audible sounds. There are two main types of *automated otoacoustic emissions* (AOAEs) namely, *transient-evoked otoacoustic emissions (TEOAE)* and *distortion-product otoacoustic emissions (DPOAE)*. TEOAE, also known as cochlear echoes, are low intensity sounds originating from the active amplification of the outer hair cells and can be elicited in response to clicks or tone bursts presented to the ear through a light weight probe that houses both a transducer and microphone/receiver. The emissions are then matched through advanced digital processing technology with a standard template before giving a ‘pass’ or ‘refer’ result.

A typical TEOAE instrument is light, portable and powered by an inbuilt rechargeable battery that can last many hours of continuous use (Figure 1).
The recording often takes seconds and can be administered without audiological expertise. The sensitivity and specificity are greater than 90%. One disadvantage with this test in newborns is that it is sensitive to peripheral hearing impairment such as mild conductive hearing loss resulting from debris associated with vernix caseosa and amniotic fluid in the external ear canal, in the first day of life. The test is sensitive to excessive internal noise from patient or ambient noise in the test environment and will not detect any retrocochlear dysfunction of the inner hair cells and beyond such as auditory neuropathy/dyssynchrony.

DPOAE differs from TEOAE because they are generated by two continuous pure tones introduced to the ear simultaneously. Because DPOAEs are evoked by frequency-specific signals, it is possible to use the response to predict frequency-specific hearing sensitivity across the frequency range of 500 to 8,000 Hz. DPOAE amplitude and pure-tone audiograms are somewhat but imperfectly comparable in the frequency region above 1,500 Hz. However, this advantage is not critical for screening infants and young children which is perhaps why TEOAE is the most widely used in infant screening programmes. Nonetheless, initial refer rates above 10% are not uncommon with TEOAE when conducted in babies prior to hospital discharge which reduces with subsequent test over time.

**Automated Auditory Brainstem Response (AABR)**

The ABR is an electrophysiological measure of the function of the entire auditory pathway recorded by three scalp electrodes (Figure 2). This test is not state-dependent as recordings are best obtained when babies are sleeping or sedated. In general, the click-evoked threshold predicts behavioural audiometric threshold in the 1,000 to 4,000 Hz range within 10 to 15 dB HL. It is, therefore, valuable as a confirmatory test in infants.

Figure 2: AABR screening in hospital (a) and in a Community Centre (b)

The automated version of ABR (AABR) was designed for screening purposes. A typical AABR instrument is also powered by an inbuilt rechargeable battery and when activated delivers at least one thousand soft-click stimuli at 35 dBnHL to the newborn’s ears through disposable flexi-coupler earphones at a rate of 37 clicks per second. The responses to the auditory stimuli are recorded with three surface jelly tab sensors or electrodes placed over the vertex, nape and the shoulder or the cheek. A “pass” is displayed when the manufacturer’s internally programmed template-matching algorithm matches ongoing brain wave or auditory
brain stem response at a minimum of 1000 sweeps. The status of the ear is deemed as “refer” if the likelihood ratio is less than the manufacturer’s algorithm after 15,000 sweeps, either from reduced or absent auditory brainstem response or inability to discriminate interference from a response. In infants older than three months, it may be difficult to conduct this test without sedation due to restlessness during testing.

**Questionnaire-based instrument**

Questionnaire-based screening uses a set of questions to elicit symptoms or indicators of possible hearing loss indirectly through proxies like parents or caregivers. Its major attraction is that it is the cheapest form of mass screening tool especially in resource-poor settings. Like any health-related questionnaire, the value is usually enhanced if it is quick and simple to administer and has been properly validated against an appropriate gold standard in a comparable target population. However, the accuracy and reliability of parental questionnaires are rather inconsistent and highly variable across studies with reported sensitivities of 34-71% and specificities of 52-95% in a comparison of three studies from Nigeria, Brazil and Australia. Questionnaires may therefore be grossly cost-ineffective for mass screening programmes because of the potential burden of false-positives. The only consistent success story appears only to be among adults especially the elderly with whom questionnaires have been found effective in characterising the handicapping effects of hearing loss. In developing countries where objective screening tests are not immediately feasible or affordable, the merits of questionnaire-based screening for infants continue to be the subject of debate. However, individual judgement is required as to when and where such an alternative screening is desirable and more beneficial than “no-screen”.

**Choice of screening tests for a screening programme**

No screening protocol is perfect. A practical approach is to recognise *a priori* cases that are likely to be missed by the choice of a particular protocol and to set up a surveillance system that will minimise potential drop-out rate resulting from the protocol.

There is no hard-and-fast rule about the choice of technology or the protocol to be used. However, a 2-stage screening with an initial OAE followed by AABR for all OAE referrals has several advantages over a single or 2-stage screening protocol with either TEOAE or AABR. For example, while OAE is generally preferred for initial screening, referrals can be excessive especially in busy hospitals where the discharge policy is less than 48 hours. Introducing AABR will reduce the pre-discharge referral rates substantially thus minimising the burden on follow-up services. The combination of OAE and AABR also facilitates identification of infants with auditory neuropathy.

The conditions of the environment in which the screening instruments are used are important factors that should be considered before the commencing screening. It is important to be prepared to manage the effect of excessive ambient noise on OAE recordings such as prolonged testing time, the inability to obtain accurate recordings and high false-positive rates. In contrast, AABR tests are less susceptible to background noise. However, they are difficult to conduct when the baby to be tested is restless or irritable because of the resultant myogenic interference which prolongs the testing time. Some practical tips for successful OAE screening are summarised in *Box 1 (below)*.
Screening schedule, tips and environment

- Quiet room
- Post a sign to inform others that a hearing test is in progress
- In conjunction with nursing staff, establish test times that are optimal for nurses, parents, and babies' schedules
- Complete the test prior to discharge

State of the baby

- Quiet, sleeping baby
- Clean diaper
- Swaddle baby
- Ideally, conduct the test after the baby has been fed

Tips for proper probe fit

Proper probe fit is the key to obtaining a good OAE screening test. A tight seal ensures that your screening test will be quick and accurate. Some tips for obtaining a good probe fit are:

1. **Selection of probe tip:** Use the largest possible probe tip since one that is too small will allow too much environmental noise in to interfere with the result

2. **Check probe tip:**
   - **For good fit** - Gently tug on the probe once it is in the ear to make sure there is resistance. If it slides out easily, it is too loose
   - **For debris** - Make sure the probe tip is clean. Often, the probe tip may become clogged by vernix or debris. If you need to clean the probe tip, be sure to remove it from the probe prior to cleaning, so you don't inadvertently push the vernix or debris into the probe

3. **Newborns with collapsed ear canals:** This common condition in newborns can be problematic if the baby has just been lying on the ear you wish to test. To fully open the ear canal and obtain a good probe fit you have to:
   - Gently massage the area in front of the ear in a circular motion for 10-20 seconds before inserting the probe tip
   - Gently pull up and back on the pinna, or outer ear, and massage/rotate gently a few times before inserting the probe
   - When inserting the probe into the ear, pull up and out gently on the pinna, or outer ear to open the ear canal fully

4. **Unsettled baby:** Having the baby swaddled during testing will help to make sure that the probe is not knocked out of the ear during the test

**Poor Technique:** Don't hold the probe during testing. This can cause the probe to touch the wall of the ear canal and prevent signal from getting through. It can also cause noise that interferes with testing

**Box 1: Practical tips for successful OAE screening**

Choice of screening coverage for a screening programme

In view of the fact that more than half of infants in many developing countries are delivered outside hospitals, there are four generic options to consider in implementing an effective hearing screening programme (Figure 3). A screening programme can either be hospital-based or community-based and can either seek to reach all newborns (universal screening) or predefined group of newborns (targeted screening).

<table>
<thead>
<tr>
<th>Screening coverage</th>
<th>Hospital-based</th>
<th>Community-based</th>
</tr>
</thead>
<tbody>
<tr>
<td>Targeted</td>
<td>THB</td>
<td>TCB</td>
</tr>
<tr>
<td>Universal</td>
<td>UHB</td>
<td>UCB</td>
</tr>
</tbody>
</table>

**Figure 3: Infant screening models: THB: Targeted hospital-based; TCB: Targeted community-based; UHB: Universal hospital-based; UCB: Universal community-based**

**Hospital-based or Community-based screening**

In order to achieve good coverage among eligible infants at the population level, newborn screening must be implemented shortly after birth. While hospitals provide the most convenient location for screening, hospital-based screening cannot cater for infants born outside hospitals who are in the majority in many low-income countries. Considering that the purpose of screening is to facilitate early detection of hearing loss that can impede speech and language development, hospital-based programmes will miss a significant proportion of infants with acquired, delayed-onset or progressive hearing loss. One approach is to have a community-based programme to comple-
ment the hospital-based programme in such settings. Alternatively, community-based platform such as provided by routine immunisation in the first weeks of life can be considered for infant hearing screening as has been demonstrated in a number of countries in sub-Saharan Africa. A community-based programme also has disadvantages. For example, it will miss infants that are presented late or not presented at all for immunisation. Facilities for conducting newborn screening may not be readily available such as a suitable test site with tolerable ambient noise levels.

Universal versus Targeted screening

Hearing screening is universal when it is directed at the whole population, and it is targeted when a subgroup or high-risk fraction of the community is selected for screening. Universal screening is more ideal but may be forestalled by the lack of requisite resources much more in developing countries which account for a disproportionate burden of infant hearing loss. Like many other health conditions, selecting those to screen based on the presence of established risk factors for hearing loss helps to curtail the quantum of resources required and potentially provides a better alternative than no screening. However, the range of risk factors that are relevant for this selection may vary, and the magnitude of the known risk factors may be negligible in some countries. For example, most of the commonly used risk factors proposed by Joint Committee on Infant Hearing (JCIH) (Table 1) such as in-utero infections or even accurate history of family deafness are difficult to ascertain in either hospital-based or community-based programmes. Many hospitals do not have NICU but at best SCBU which is of limited value as they cannot manage at risk babies such as the very preterm or those at the fringes of viability. Rarely reported factors such as hypertensive disorders in pregnancy, infant’s undernutrition and lack of skilled attendants at birth have been found to be significantly associated with permanent hearing loss thus demonstrating the need to establish context-specific risk factors from pilot universal NHS programmes.

Pre-Screening Procedure

Infection Control and Decontamination of Screening Equipment

Strict adherence to local infection control practices to decrease transmission of infectious agents should comply with local regulations. Education and training of hearing screening personnel on infection control practices should be one of the core areas of training.

Parental Education and Informed Consent

All parents should be well informed of the consequences of detecting hearing loss late, and the benefits of early hearing detection and intervention verbally as well as written information leaflet (Figure 4). Ward or clinic nursing sisters can efficiently carry out this function because of their vast experience with educating mothers. The limitation of the screening procedure in discriminating a hearing loss due to debris or vernix plug in the ear canals from true hearing loss which might require subsequent follow-up visits for necessary confirmation should also be emphasised as well as the importance of completing the screening process.

Pre-Screening Questionnaire

A structured questionnaire should be administered by the screening team to elicit medical, socio-economic history and context-specific risk factors from mothers or carers.
<table>
<thead>
<tr>
<th>JCIH</th>
<th>Risk Factors for Hearing Loss</th>
</tr>
</thead>
</table>
In-utero infections such as rubella, cytomegalovirus, syphilis, toxoplasmosis and herpes  
Craniofacial anomalies  
Birth weight less than 1,500g (3.3lbs)  
Hyperbilirubinaemia at levels requiring exchange transfusion  
Birth asphyxia with Apgar score 0-4 at 1 minute or 0-6 at 5 minutes  
Mechanical ventilation lasting five days or more  
Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss |
| **2000**     | Family history of sensorineural hearing loss  
NICU admission greater than 48 hours  
Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss  
Craniofacial anomalies  
In-utero infections such as rubella, cytomegalovirus, syphilis, toxoplasmosis and herpes |
| **2000**     | Parental/caregiver’s concern regarding hearing, speech, language and or developmental delay  
Family history of sensorineural hearing loss  
Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss or Eustachian tube dysfunction  
Postnatal infections associated with sensorineural hearing loss including bacterial meningitis  
In utero infections such as cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis  
Neonatal indicators especially hyperbilirubinaemia at levels requiring exchange blood transfusion  
 Syndromes associated with progressive hearing loss, such as neurofibromatosis, osteopetrosis, and Usher’s syndrome  
Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich’s ataxia and Charcot-Marie-Tooth syndrome  
Head trauma  
Recurrent or persistent Otitis Media for at least 3 months |
| **2007**     | Expanded definition of targeted hearing loss from congenital bilateral and unilateral sensory or permanent conductive hearing loss to include neural hearing loss such as auditory neuropathy/dyssynchrony in infants admitted to NICU or SCBU for more than 5 days |
| **2013**     | Supplement to JCIH 2007 Position Statement on the Principles and guidelines for early intervention following confirmation that a child or ward is hard of hearing |

Table 1: JCIH Risk Factors for Hearing Loss
Screening Procedure

Screening Protocols

Typical screening protocols for low-risk infants and high-risk infants (admitted for special or intensive care, or with known risk factors for PECHL) are presented in *Figure 4*.

![Figure 4: A typical low- and high-risk screening protocol](image)

Low-risk or Well Babies

Well babies can first be screened with AOAE between 24 and 48 hours after delivery or during immunisation or post-natal visits in the outpatient department. Babies who fail AOAE tests in the hospital nursery can either be rescheduled for a repeat test or second-stage screen with AABR which usually is conducted before hospital discharge. Babies who refer in one or both ears or unable to complete the screening prior to discharge should be followed-up during routine post-natal visits to the hospital. Those failing the follow-up AABR or AOAE rescreen depending on the screening protocol should be referred for full diagnostic/confirmatory evaluation. Well babies who are first seen at immunisation or outpatient clinics should be screened in line with the appropriate low- or high-risk screening protocol.

High-risk babies with known risk factors or Babies in NICU/Special Care Units

The screening for high-risk babies is with both tests (AOAE and AABR). First screening is usually with AOAE followed by AABR, regardless of AOAE test result, just before hospital discharge. Mothers whose babies refer on AABR should also be referred for a follow-up assessment during the routine post-natal visits to the hospital. Persistent AABR referral qualifies the baby for diagnostic evaluation.

Pass/Referral Criteria

For the well babies, the first-stage pass should be documented when clear responses from both ears are recorded with the AOAE screen. For babies referred in one or both ears the test should be repeated immediately. A first-stage referral should be documented after a second referral with AOAE. Clear responses from both ears are indicative of pass criteria for the second stage screen with AABR. Referrals from one or both ears should attract a repeat outpatient test at a future date while a persistent outpatient refer outcome, for both well, and sick babies should qualify for a full diagnostic assessment.

Communication with Parents

The screeners should be well tutored through ‘role-play’ on how to communicate screening results to parents. “Refer” outcomes during the various stages of
screening should not be presented as evidence of hearing loss but rather as an indication for further tests to rule out any uncertainty regarding the hearing status of the child. The significance of the follow-up appointment should, therefore, be emphasised to parents to ensure that the screening procedure is completed.

**Post-Screening Services**

**Diagnostic Services**

The diagnostic services should be accessible and affordable consisting of tympanometry including high frequency (1000 Hz) probe tones for babies less than 4 months old, diagnostic tone pip ABR with insert earphones and/or Visual Reinforcement Audiometry (VRA) for babies older than 6 months. Follow-up counselling appointments may be scheduled for the parents of babies who were confirmed with any degree of bilateral or unilateral sensorineural hearing impairment.

**Communicating results to parents**

It is important that information about the hearing screening and the results are conveyed to the parents in a professional, thoughtful and sensitive manner. By saying, "Your baby didn't pass the hearing screen" you may have just changed this family's whole life.

1. **Points to remember when communicating results to parents:** Information should be given both verbally and in writing. It is important to remind them not to leave the leaflet in the hospital or screening site. Inappropriate communication of screening results can cause undue stress and anxiety for the parents. Proper terminology must be used. Avoid using the word: "fail". If a baby does not pass the hearing screening, the term "refer" should be used instead. This is an emotionally sensitive time for parents - information should be conveyed in a supportive, confidential environment and an unhurried manner, with plenty of time allowed for answering questions.

2. **If a baby is referred for further testing:** If a baby does not pass the hearing screening, it is crucial NOT to use the words "failed" or "did not pass". This terminology implies to the parents that their baby has a hearing loss, or is deaf. Instead, say, "We are referring your baby to a nearby Specialist Hearing Centre for further testing," or "We are referring your baby for a re-screening because the test results were inconclusive today." Parents must be made aware that newborn hearing screening is designed to catch babies who are at risk for hearing loss and need further testing. It is important to remind parents that hearing loss does not necessarily equal deafness. Hearing loss can range in severity from mild to severe-profound (deaf). Additional diagnostic testing is needed to confirm their baby's hearing status. If a baby is referred, the family should be informed that there could be several reasons why their baby is being referred for further testing. The most common reasons are:
   - Ear canal blocked with debris (most common)
   - Presence of middle ear fluid
   - Permanent hearing loss (approximately 3 in 1000 births)

When screeners discuss test results with families they should be careful not to downplay a refer result, while at the same time being careful not to cause the family to panic.
**Intervention Services**

Counselling sessions should be available for all the parents of children failing the diagnostic tests. The purpose is to educate the parents on the implications of the results, the nature of support that would be required for the children and the role expected of parents. Ongoing parental surveillance is recommended for babies that did not qualify or who pass diagnostic services. Information leaflets (*Figure 5*) should be handed to all parents prior to the first-stage screening and thereafter for parents whose infants refer.

![Image](image.png)

*Figure 5: Parent information and consent leaflets*

**Evaluating the performance of a screening programme**

The effectiveness of early detection is best measured by the improvement in outcomes in the domains of speech and language, cognitive and behavioural as well as educational achievement. Such an evaluation often requires long term follow-up of children with PCEHL up till school age. However, quality indicators that are commonly regarded as “intermediate” outcomes or surrogates of long term outcomes for evaluating the effectiveness of infant hearing screening programmes are screening coverage, screening effectiveness, return rate, for diagnostic evaluation, age of confirmation of hearing loss, effectiveness of the screening protocol (number of babies screened/day, referral rates for OAEs and AABR, screening cost per child and yield for PCEHL).

1. Screening coverage as a percentage of infants screened before hospital discharge or within 3 months of age among all those eligible for screening should be ≥95%.
2. Screening effectiveness as measured by the referral rate for diagnostic evaluation among those completing the protocol ≤4%.
3. Return rate for diagnostic evaluation of at least 70%.
4. Age of confirmation of hearing loss by 3 months of age.
5. Effectiveness of screening protocol is based on an acceptable level of sensitivity, specificity and the likelihood ratios of the screening protocol.

**Concluding remarks**

This short review is intended to highlight some of the relevant issues in conducting infant hearing screening successfully in a developing country setting. It is by no means exhaustive. Readers will benefit from the list of suggested readings below and other related contributions in this series. It is important to emphasize that the value of early detection can only be optimised through a thoughtful and painstaking commitment to follow-up of those who require confirmatory evaluation not later than the first year of life and enrolment of those with hearing loss in
appropriate family-oriented rehabilitation programme. The choice of assistive hearing devices and communication modalities must be carefully evaluated after consultation with the family and relevant professionals. The detection of hearing loss marks the beginning of a lifetime journey for the affected children and their families. All care givers and service providers must bring their experiences to bear in supporting the families on this essential and irreversible journey as much as lies within their powers.

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