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Newborn hearing screening in Hong Kong

Introduction

Significant hearing impairment (HI) is an important and common birth defect that occurs in 1 to 3 per 1000 live births and 2 to 4 per 100 neonatal intensive care infants.1-4 It occurs more frequently than many other newborn conditions for which screening is routinely performed, for example congenital hypothyroidism. Based on an annual birth rate of 55 000 in Hong Kong, about 165 infants were born with significant HI.3

Children with HI are deprived of an important source of sensory input. The
auditory stimuli that a child perceives in the first few months of life form the basis of speech, language, and cognitive development. If lacking, social, emotional, comprehensive, and motor development can be adversely affected. One third of children with even minimal hearing loss fail at least one grade and exhibit social and/or emotional problems by the time they reach the fourth grade. Despite advances in hearing aid technology, improved educational techniques, and intensive intervention services available to children with HI, there has been little advancement in their language development and academic performance. This may be due to late diagnosis: several prospective studies have consistently demonstrated that early diagnosis of HI and intervention can improve intellectual, language, and speech development.

One study reported that the only significant variable to affect development of language skills was the age at which HI was diagnosed. Children in whom hearing loss was identified by 6 months of age demonstrated significantly better language scores than those in whom it was diagnosed later. Landmark studies reveal that universal newborn hearing screening (UNHS) enables early identification and intervention prior to 6 months of age, such that near-age appropriate language skills and academic performance can be achieved. Recent advances in the area of hearing screening have facilitated the availability of more sensitive and easy-to-use screening tools that can effectively and reliably test hearing soon after birth. Thus, this issue has recently been hotly debated and many Asian countries have begun to develop and report studies of different models of newborn hearing screening and rehabilitation programmes.

Past experience and limitations of the distraction test as a screening tool

Hearing screening is an integral component of the comprehensive observation scheme and was implemented in Maternal and Child Health Centres (MCHC) in 1978. The behavioural distraction test is the principal means by which infant hearing is tested and is performed between the ages of 6 and 9 months on all children who attend the MCHC. One of the major limitations of this screening method is the validity of the test: it has poor sensitivity and low specificity. In the United Kingdom, the distraction test performed at 8 months has a reported failure rate of up to 16.3%, and is difficult to perform and unreliable in high-risk infants. Such infants may have physical or mental handicaps that interfere with their ability to respond to the test. Distraction tests are also time-consuming and must be performed by trained and experienced staff. In addition, the criteria for determining presence or absence of a reaction are somewhat subjective. In an infant who fails the first test, a re-test several weeks later is generally recommended because of the possible presence of temporary middle ear problems. Despite this, many hearing impaired children are either not diagnosed or are diagnosed late. Data collected by the MCHC in Hong Kong reveal that the usual age at diagnosis, especially for children without risk factors, is 18 months to 2 years, with most infants receiving treatment and rehabilitation after 2 years of age. The uptake of hearing screening for newborns registered at the centre was 81.7%, far below the required rate of more than 95% needed for any form of universal health screening. Nonetheless with this programme, the yield of babies with moderate or severe HI bilaterally is 0.2 per 1000, which is far less than expected.

High-risk newborn hearing screening

The 1994 Position Statement of the American Academy of Pediatrics Joint Committee on Infant Hearing recommended the maintenance of a role for high-risk indicators associated with sensorineural and/or conductive hearing loss in newborns and infants and modified the list of indicators described in the 1990 Position Statement. The Committee recommends a specific hearing protocol for high-risk infants when universal screening is unavailable. The indicators associated with hearing loss for use with neonates are:

1. family history of hereditary childhood sensorineural hearing loss;
2. in-utero infection, such as cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis;
3. craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal;
4. birth weight of less than 1500 g (3.3 lb);
5. hyperbilirubinaemia at a serum level requiring exchange transfusion;
6. ototoxic medications, including but not limited to aminoglycosides used in multiple courses or in combination with loop diuretics;
7. bacterial meningitis;
8. Apgar scores of 0 to 4 at 1 minute or 0 to 6 at 5 minutes;
9. mechanical ventilation lasting 5 days or longer; and
10. stigmata or other findings associated with a syndrome known to include sensorineural and/or conductive hearing loss.

In Hong Kong, most Hospital Authority (HA) birthing hospitals perform some form of hearing tests for at-risk infants, usually by brainstem auditory evoked potential (BAEP) or by otoacoustic emission (OAE) tests. These screening programmes nevertheless do not form part of a territory-wide protocol and are often uncoordinated with no formal tracking or follow-up system.

The first high-risk newborn hearing screening programme in Hong Kong in 1998 identified 5% of babies born at the hospital to be at risk, of whom 3.8% were confirmed to have hearing loss using conventional auditory brainstem response tests. Five years ago, a large-scale high-risk hearing screening project was conducted in Hong Kong and involved a birth cohort of 19 922 babies from five major public hospitals. Risk factors were based on the high-risk indicators modified from the American Joint Committee on Infant Hearing 1990 Position Statement. Screening was performed using a two-stage distortion product OAE test.
identification, intervention, and evaluation.\textsuperscript{27,28} UNHS programme: screening, tracking and follow-up, also recommended five essential elements of an effective programme to be cost-effective. Current available evidence confirms that a newborn hearing screening programme fulfills most of these criteria. The American Academy of Pediatrics (AAP) recommended that a newborn hearing screening programme fulfills at least five criteria before universal screening or parental recognition has not been very successful. This implies that the high-risk indicators are unreliable as a means of determining the individual need for newborn hearing screening. In these at-risk infants, the overall rate of HI in general was 4.5\% and of moderate-to-profound HI was 2.4\%. Hence, the overall prevalence of moderate-to-profound HI was 0.6 per 1000 births; much lower than the reported prevalence of 1 to 3 per 1000.\textsuperscript{1-4} Previous studies also confirm that high-risk screening is ineffective because it may miss at least 50\% of children with congenital HI.\textsuperscript{24,25} A UNHS may therefore offer a more rigorous approach that can achieve the aim of identifying most, if not all, babies with congenital hearing loss.

**Universal newborn hearing screening**

The Joint Committee on Infant Hearing 2000 Position Statement\textsuperscript{23} endorsed early detection of, and intervention for infants with hearing loss through integrated and interdisciplinary state, and national systems of UNHS, and family-centred intervention. The purpose of early detection of hearing problems and intervention is to maximise linguistic and communicative competence and liberal development for children who are hearing impaired.\textsuperscript{23} The American Academy of Pediatrics Task Force on Newborn and Infant Hearing recommended that universal detection of infant hearing loss requires universal screening of all infants.\textsuperscript{26} Reliance on a physician’s observation or parental recognition has not been very successful. At least five criteria must be fulfilled before universal screening is justified: (1) the availability of an easy-to-use test that possesses a high degree of sensitivity and specificity to minimise referral for additional assessment; (2) the condition being screened for is otherwise undetectable by clinical parameters; (3) there are interventions available to correct the conditions detected by screening; (4) early screening, detection, and intervention result in improved outcome; and (5) the screening programme is documented to be cost-effective. Current available evidence confirms that a newborn hearing screening programme fulfills most of these criteria. The American Academy of Pediatrics also recommended five essential elements of an effective UNHS programme: screening, tracking and follow-up, identification, intervention, and evaluation.\textsuperscript{27,28}

**Common devices used for universal newborn hearing screening**

**Brainstem auditory evoked potential**

The conventional BAEP is the most reliable method and the gold standard for evaluating peripheral auditory function of a newborn.\textsuperscript{29,30} It is based on a click stimulus to elicit an electrical response that is measured by surface electrodes. It should be performed and the results interpreted by a skilled technician, usually an audiologist. It also requires sedation of the baby for the procedure. Hence, due to the relatively high cost, conventional BAEP is not recommended as a screening test, but more commonly used as a diagnostic or confirmatory test.

**Otoacoustic emission**

At present, the choice of device for newborn hearing screening is between OAE and automated brainstem response (ABR), or a combination of the two. Two forms of OAE technology have been used: ‘transient or click-evoked otoacoustic emissions’ (TEOAE) and DPOAE. The differences stem from the choice of stimulus and the technology subsequently used to extract the response. Both are based on low-intensity sounds generated by the motile activity of the outer hair cells in the cochlea and detected by a microphone applied to the external meatus. The TEOAE employs click stimulation and averaging similar to screening by ABR where the response follows the stimulus. The DPOAE employs a series of tonal stimuli and the response occurs during the presentation of stimulus. Both OAE forms have the advantage of being simple and quick to perform with minimal disturbance to the babies. Commercially available devices have built-in full automation making interpretation easy. Appropriately trained nurses or health care assistants can perform the test so that, combined with the low-cost consumables, the cost per test is affordable. Nonetheless, if the ear probes are not carefully fixed or acoustically shielded, which can be difficult to achieve in small infants, these tests may be affected by ambient noise. The noisy environment of postnatal wards or out-patient clinics where the test is frequently performed, produces a high number of false-positive results because the noise interferes the delivery of stimulus. The false-positive rate is also high when the test is performed in the first few days of life (due to the presence of debris in the external ear canal and middle ear).\textsuperscript{31,32} The early discharge policy of most birthing hospitals further limits its use as a hospital-based screening tool. Repeating the test later (making it a two-stage OAE) may overcome some of these problems. Moreover, the device can only test up to the level of cochlea; it does not provide information on retrocochlear pathology in the auditory nerve, brainstem, and auditory cortex. Hence babies with risk factors for neurological disorders may show a normal OAE response and need additional screening by ABR. Despite this, OAE is still considered to be of value for screening large numbers of healthy infants because a very high proportion of congenital HI is related to pathology in the middle ear or the sensory cells of the inner ear. Depending on the above factors, the overall referral rate following OAE for diagnostic tests varies from 2.4\% to 10\%.\textsuperscript{33,34}

**Automated auditory brainstem response**

The automated auditory brainstem response (AABR)
test, a simplified version of ABR, comprises an electroencephalography system, a stimulus-generating system, ambient noise and myogenic activity detection systems, and the ABR detection algorithm with automation in result interpretation. It is tested by delivering a soft click to the baby’s ear through ear probes. The electrical response is picked up by disposable electrodes attached to the baby’s skin over the head and neck region. Signals from the skin electrodes are then transmitted to the computer for averaging and automatic analysis. This is a quick and simple test that can be performed reliably even in newborns under 24 hours old, and is thus ideally suited as a hospital-based screening programme. It has a lower referral rate, ranging from 4% to 0.82%. The two-stage AABR can further reduce the referral rate to between 2.5% and 0.12%. The currently available AABR machine and its consumables are nonetheless more expensive than the OAE device, which inevitably increases the cost of a screening.

The recent introduction of equipment that incorporates both OAE and ABR functions provides an additional option for screening. The former measures the health of the cochlear and the hair cells, and the latter tests the health of the neurons within the auditory brainstem structures. A two-stage OAE-ABR enables a complete evaluation of the baby’s auditory function and achieves a low referral rate for further diagnostic evaluation.

Regardless of the device and method chosen, a successful hearing screening programme requires an accurate, rapid, and simple-to-conduct test that can, preferably, be fully automated and provided at an affordable cost. A feasibility study based on the local setting and experience is essential before a screening programme is widely implemented.

**Pilot studies of hospital-based universal newborn hearing screening in Hong Kong**

The first pilot study of a UNHS programme was performed in a university hospital (Tsan Yuk Hospital) over a 4-month period in 1999. The study was undertaken to investigate the prevalence of HI, to assess the feasibility of implementing a UNHS programme in a maternity hospital, and to explore parental understanding of the detection of hearing loss in young infants and parental acceptability of newborn hearing screening. The screening was performed using a two-stage DPOAE (Otodynamics ILO 292 Echoprint System). Results demonstrated a high coverage rate of 99.3% and a prevalence of permanent bilateral HI of 0.28%. It was noted that the referral rate for diagnostic testing was unacceptably high when the test was performed within the first 3 days of life before hospital discharge. This confirmed previous experience that OAE may have high false-positive rates because of ear debris when performed within a few days of birth. The questionnaire study also showed that most parents had little understanding of hearing developmental milestones. Parental surveillance cannot therefore be considered a reliable means to detect hearing problem in newborns. Most (91%) mothers agreed neonatal screening for hearing defects was desirable and most (82%) preferred such screening to be carried out before discharge from the maternity unit. This study demonstrated that the implementation of hospital-based UNHS in Hong Kong is feasible and acceptable to parents. Further study is necessary to explore the use of other screening devices that may have a lower referral rate for diagnostic screening before hospital discharge.

In 2000, the HA set up a multidisciplinary working group that included paediatricians, audiologists, and otolaryngologists to consider the justification and the logistics of establishing a UNHS programme in Hong Kong. Another pilot study was conducted in three major public hospitals and involved a birth cohort of 4314 over a 5-month period in 2001. The study aimed to assess the referral rates of different screening protocols, namely (1) two-stage AABR using ALGO2 (Natus Medical, CA, US), (2) two-stage OAE-AABR using Bio-logic 2-in-1 screener (Mundelein, IL, US), and (3) two-stage AABR using Bio-logic 2-in-1 screener. The tests were completed before the babies were discharged from hospital with an overall coverage rate of 89.6%. In two hospitals, where the projects were coordinated by paediatricians, a higher coverage rate of over 95% was achieved. The referral rates of AABR-AABR (ALGO2) protocol were consistently shown across the three hospitals to be the lowest compared with OAE-AABR (Bio-logic) and AABR-AABR (Bio-logic). This study confirmed that UNHS is feasible. A well-coordinated hospital-based screening programme can achieve a high coverage rate and the two-stage AABR screening with ALGO2 yields the lowest referral rates. An earlier study also showed that an AABR programme has the lowest referral rate. The referral rate of 0.6% at the second stage in this study was much lower than the reported referral rate of 3.2%. Although the AABR programme has more expensive consumables, the lower referral rate means that the cost per infant screened is similar among all three programmes. If dedicated technicians are used, as in this study, referral rates and overall costs should also drop significantly.

**Pilot study of community-based universal newborn hearing screening in the Maternal and Child Health Centres in Hong Kong**

The MCHC of the Department of Health investigated the use of two-stage OAE to replace the distraction test. As not all births are registered at the regional MCHC, the coverage rate for the total birth cohort was unknown. This study showed a coverage rate of 72.5% of all babies registered at the four study centres. The most common reason for babies not receiving the test was that they were older than 2 months at registration. The test repeat rate was 9.2% and screen referral rate was 3.8%. The yield for
moderate or severe HI was 0.76 per 1000 babies screened. This study confirmed that screening performed in the MCHC setting after discharge from birthing hospitals cannot achieve a standard coverage rate of 95% as laid down in the Joint Committee on Infant Hearing 2000 Position Statement. The referral rate of the OAE, although lower than in the hospital screening, was much higher than the hospital-based two-stage AABR screening protocol.

Cost-effectiveness and parental stress of a universal newborn hearing screening programme

One of the major concerns among the health care administrators and opponents of UNHS is the cost-effectiveness and lack of solid evidence to confirm the long-term benefit to affected children. A meta-analysis of the effects of screening, early identification, and treatment on language outcome cautioned that there were several gaps in information about UNHS. Among 19 studies identified, only one was a controlled trial. Besides, when screening ensued before the age of 6 months, most studies demonstrating improved language and communication skills by the age of 2 to 5 years were based on ‘fair’ to ‘poor’ quality cohort studies. Based on a theoretical mathematical model devised from this literature review, extending screening to low-risk infants would detect one additional case before the age of 10 months for every 1441 low-risk infants (number needed to screen). In addition, 254 newborns would need further audiological evaluation because of false-positive screening results. The meta-analysis cautioned about the potential harm associated with false-positive screening including misdiagnosis, parental misunderstanding and anxiety, and unfavourable labelling. Nonetheless, using the latest technology, data from our local and other international studies using the AABR demonstrated sensitivity in the range of 80% to 90%, with a false-positive rate of less than 2%. With such a low referral rate, UNHS is both efficient and cost-effective. The proposed costs are much lower than the costs of the infant distraction screening test and the cost per child diagnosed with bilateral permanent HI is considerably less. There is an increasing body of evidence to confirm that early identification of HI and intervention by 6 months of age is associated with better expressive and receptive language, speech as well as social and emotional development in the first 5 years of life. Recent evidence also confirms that UNHS can reduce the age of confirmation of congenital HI and enable earlier intervention and hearing aid fitting, especially in infants without risk factors. With respect to parental stress, a study by Weichbold et al showed that if parents were well informed about the hearing test, the vast majority (84% of mothers) would respond positively to the question of “should UNHS be performed despite the possibility that parents become worried by false-positive test results?” Similar to our experience in the first hospital-based pilot UNHS study using the OAE, the general attitude among parents was very positive, despite the relatively high false-positive rate in the first stage of screening. Another study has also shown that the anxiety arising from false-positives and re-testing can be minimised by using screening tools with lower referral rates, improved information, and rapid and effective follow-up. Hence with a well-coordinated and well-informed UNHS programme, there is little to worry that screening all newborns causes unnecessary anxiety among mothers.

Collaborative model for a universal newborn hearing screening programme

It is important to emphasise that screening is only the starting point of an effective hearing screening and intervention programme. The Joint Committee on Infant Hearing 2000 Position Statement sets down the other important parameters of a UNHS programme: a tracking and follow-up programme should aim to achieve a minimum of 95% successful follow-up of all infants referred for formal audiological assessment and for all infants not screened initially at the birthing hospital. It is essential to establish and maintain a central monitoring system for early identification and intervention, as well as to provide ongoing and regular evaluation of the performance of UNHS programmes.

A collaborative programme that incorporates the specific competencies of the hospital, the MCHC service, and the education authority is the best collaborative model. In Hong Kong, almost all babies are born in hospitals. To achieve a high coverage rate, the test is best performed when the population is still captive, ie before hospital discharge. Hospital-based universal screening can achieve a much higher coverage than the distraction test and OAE screening programme at MCHC. The birthing hospital, apart from providing the screening service, can offer specialist support including audiologists, otolaryngologists for counselling, and further audiological assessment and genetic workup for abnormal cases. Paediatricians should coordinate and ensure access to appropriate expert services for all affected children. The MCHC should establish and maintain a central registry and monitoring system. They should also provide a ’mop-up’ service for missed cases and ongoing monitoring for late or acquired HI. Some infants may pass the initial hearing screening but require periodic monitoring to detect delayed onset sensorineural and/or conductive hearing loss. Even inherited causes of HI may manifest postnatally as late progressive HI. It is well documented that screening newborn babies with congenital cytomegalovirus infection may detect less than half of those with sensorineural hearing loss. Hence the MCHC should maintain a well-documented screening history and provide ongoing surveillance for those who pass the test at birth or have mild HI, especially those considered at risk for whom regular audiological monitoring should continue until 3 years of age. The Joint Committee on Infant Hearing 1994 Position Statement produced a list of health indicators associated with delayed sensorineural/conductive hearing loss that warrant re-screening:
Screening should be followed by evaluation, diagnostic workup, and a multidisciplinary intervention and rehabilitation programme. Upon confirmation by diagnostic test and assessment by audiologists and otolaryngologists, children with HI should be referred for prompt intervention and rehabilitation. In Hong Kong, The Audiological Services Section of the Education Department provides support to preschool and school-age children with HI. In addition to the provision of hearing devices, parental guidance, planning of an individualised habilitation programme, and review of school placement are important and ensure that these children with HI benefit maximally from early diagnosis. This undoubtedly presents a challenge to those involved and requires gradual accumulation and refinement of expertise relevant to the implementation of universal screening. Any UNHS is a multidisciplinary programme and will involve paediatricians, audiologists, otolaryngologists, nurses, speech therapists, community health care workers, and education specialists.

Conclusions

A comprehensive universal hearing screening and intervention programme is undoubtedly costly. To ensure an equitable service, a territory-wide protocol has to be developed. It is important to ensure that all the involved professions are committed to a ‘seamless’ collaboration that is responsive to families’ real needs. Good communication between all involved parties and the provision of appropriate information for parents to enable informed choices is vital. There should be a mechanism that ensures accessibility to high-quality audiological and rehabilitation services. The health authority should maintain quality, territory-wide data for service evaluation and be able to integrate the same with other child health services. This will facilitate monitoring for later development of permanent HI and the effectiveness of the screening programme. The implementation of a UNHS and rehabilitation programme in Hong Kong is feasible. Nonetheless, with current financial restraints and the compartmentalised health care and education system, it may be considered an ambitious endeavour that requires a substantial commitment of health care resources, energy, and coordination. Paediatricians, as child health advocates, should work towards the development and implementation of a well-coordinated, collaborative, multidisciplinary, cost-effective, and sustainable territory-wide UNHS and early intervention programme for our next generation of hearing impaired children.

References

23. Joint Committee on Infant Hearing; American Academy of Audiology; American Academy of Pediatrics; American Speech-Language-Hearing Association; Directors of Speech and Hearing Programs in State Health and Welfare Agencies. Year 2000 position


