

Establishing a Universal Newborn Hearing Screening Programme

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Abstract

As congenital hearing impairment has a worldwide incidence of 4 to 5 per 1000 babies and is thus one of the most common congenital problems seen today, universal newborn screening has a crucial role to play in its early detection and intervention. It provides the opportunity for better outcomes and normal language development. Prior to embarking on a screening programme, the newborn population and the current health care system should be analysed to select the best method of coverage. The screening tool and protocol, communication of results, as well as the follow-up measures should be clearly determined and tested. The multidisciplinary team required should be provided with the necessary information. Parents need to be educated about the importance of early hearing screening. Data management and surveillance should be established in a systematic manner. The costs of the programme should be carefully anticipated and funding sources determined. Finally, support for the programme should be sought from governmental or public health bodies, to ensure the success of the programme. Legislation can be considered if necessary.

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Congenital hearing impairment is one of the most common congenital problems seen today, with a worldwide incidence of 4 to 5 per 1000 babies.^{1,2} Out of 1000 babies, 1.7 have severe or profound hearing loss.² By comparison, congenital hypothyroidism, which has a longer history of newborn screening, has a lower incidence of 1 in 3000 babies.³ Targeted hearing screening of babies with risk factors for hearing impairment failed to detect up to 50% of affected babies without any identifiable risk factors.⁴ More than 90% of affected children have parents with normal hearing.⁴ Without universal newborn screening, hearing impairment was often diagnosed very late. In Singapore, students in special schools for hearing-impaired children were diagnosed at a mean age of 20.8 months (range, 0 to 86) and received intervention at a mean age of 42.2 months (range, 1 to 120).⁵ Late diagnosis of congenital hearing impairment can result in significant delays in language and reading. In the United States, children diagnosed with severe to profound hearing impairment in the pre-UNHS era, completed 12th grade with a 3rd to 4th grade reading level and a language level corresponding to an 8 or 9 year old child.⁶ Children who were diagnosed and provided with intervention before 6 months had significantly better

outcomes and the opportunity for normal language development at 5 years.^{7,8} Although babies with significant bilateral hearing loss are those for whom early intervention is most urgent, it is also important to detect those with unilateral hearing loss, as they are 10 times more likely to be retained at least 1 grade compared to their unaffected peers.⁹ Even children with mild hearing loss had poorer communication scores than their peers at 3rd grade.¹⁰

Universal Newborn Hearing Screening (UNHS) is the first crucial phase of a 3-prong approach for Early Hearing Detection and Intervention (EHDI). The other 2 phases are the diagnostic audiological services and the interventional services for infants with confirmed hearing loss. In order to maximise the outcome of infants who are deaf or significantly hearing impaired, the 2007 position statement by the Joint Committee on Infant Hearing¹¹ recommended that

- i) all infants should be screened for hearing loss no later than 1 month of age,
- ii) infants who do not pass the hearing screen should undergo comprehensive audiologic evaluation no later than 3 months of age, and

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- iii) infants with confirmed hearing loss should receive appropriate intervention no later than 6 months after birth.

Several areas need to be carefully considered before embarking on a UNHS programme:

- i) *The newborn population.* The size, nature and geographical distribution of the population to be screened will make a huge impact on the programme. The larger the population to be screened, the larger will be the network of screeners who need to be trained, the greater the role of the coordinating office and the need for good data management systems and the greater the economics of scale. A wide geographic distribution may create difficulties with recall of patients to a central area. This can affect the choice of the screening protocol or create the need for screeners in a community or rural setting.
- ii) *The existing healthcare system.* Newborns are best screened in the birth hospital, prior to discharge, thus ensuring the best coverage. Current workflow and discharge practices need to be studied to determine the most appropriate interval between birth and screening. A protocol or mechanism should be developed for all infants who are born outside of a hospital or if hospital screening is missed. Working together with the current screening or immunisation programmes can also be a useful strategy, especially for out-of-hospital births. Screening for newborn hearing can be performed simultaneously with screening for congenital hypothyroidism/inborn errors of metabolism or with neonatal immunisation and reported together, as long as it is carried out by 1 month of age. If a second screen after discharge needs to be done in widely scattered communities, this can be timed together with the mother's or infant's postnatal visit.
- iii) *Education.* Before the screening is carried out, parents need to be educated about the need, importance and ease of hearing screening, the implications of the results and the impact of undiagnosed hearing loss. Common concerns in the individual community should be addressed. This information should also be made available to all medical staff involved in infant care to ensure consistency. They should understand the implications of a failed screen and the need to be evaluated audiological to exclude or confirm hearing loss. This information could be provided during routine maternal antenatal care.
- iv) *The screening tool.* Both the automated auditory brain stem response (ABR) and the otoacoustic emission (OAE) have been successfully used for the UNHS.¹⁰ The OAE is the simpler and shorter of the

2 tests, requiring approximately 10 minutes to perform. However it requires a fairly quiet room, is affected by debris in the ear and only reflects the status of the peripheral auditory system up to the outer cochlear hair cells.¹⁰ The ABR takes approximately 15 to 20 minutes per test, is more tolerant of environmental noise and is less affected by debris in the ears. Unlike the OAE, it reflects the status of the peripheral auditory system, the 8th cranial or vestibulocochlear nerve and the brainstem auditory pathway. Thus while both can detect sensorineural hearing loss, the OAE will not detect auditory neuropathy or dyssynchrony (AN/D). As the infants who received intensive care are particularly at risk of AN/D, automated ABR is the only screening test that should be used in this population.¹⁰ In terms of costs, the equipment for the AABR is generally more expensive than the OAE. As a result, pre-discharge screening costs vary depending on the equipment used (ABR, US\$32.81, 2-step ABR/AOE US\$33.05, OAE US\$26.89).¹¹

- v) *The screening and follow-up protocol.* Many programmes use a 2-step protocol using either OAE or ABR. An infant who does not pass the first screen can be rescreened with the same test either before hospital discharge or soon after discharge. A combination of OAE followed by automated ABR for rescreening can also be used. However, the infant who does not pass the automated ABR should not be rescreened with the OAE, which may miss an AN/S. Referral rates for audiological assessment differ significantly among the programmes (ABR, 3.2%, 2-step OAE/ABR 4.6%, OAE 6.4%: $P < 0.01$).¹¹ Thus, limited audiological services or access may require the use of costlier equipment with a lower false positive rate (0.2%¹² to 0.9%¹³ for the 2-step ABR, 1.7%¹⁴ for the 2-step OAE). A mechanism for onward referral for audiological investigations in infants who do not pass the screening should be established. The protocol should be tested in a pilot project, which can reveal barriers to a smooth screening process before the full programme is established. Infants who are readmitted in the first month of life for conditions associated with hearing loss should be re-screened before hospital discharge.¹⁵ As hearing loss can develop later in life, infants who pass the newborn hearing screen should have regular surveillance of developmental milestones, auditory skills and parental concerns. Those with risk factors for hearing loss should be referred for an audiological assessment at least once by 24 to 36 months of age.¹⁵
- vi) *Staff.* A multidisciplinary team should be involved in the establishment of the UNHS, including

neonatologists/paediatricians, otolaryngologists, audiologists, nursing staff and screeners. A formal training programme should be established for hearing screeners, as well as regular recertification. There should be a designated coordinator or coordinating office to be responsible for the direction of the programme, maintenance of equipment, adherence to established standards, follow-up of infants who either need rescreening, audiological confirmation of hearing loss or intervention services, data collection, communication with parents, other staff members and members of the infant's medical home, family support, etc.

- vii) *Communication of results.* Communicating and recording screening results needs to be conducted in a systematic manner, preferably verbally to parents after the screen, together with a written record of the result in both hospital and child's medical records. Parents should also receive either verbal or written education about the importance of follow up, if rescreening or audiological investigations are required. The follow up medical home care team should also be informed of the results, so that appropriate follow-up can be ensured.
- viii) *Data management, follow-up and surveillance.* Commercial data management systems are available to manage the large amounts of data that will emerge from the screening programme. They can also be used to identify those who require or have missed screening, rescreening, or audiological investigations and for quality control.
- ix) *Cost of the programme.* The factors that affect cost include the screening protocol and equipment, the number of infants who need to be screened, the number of staff required and the cost of their services etc. These costs must be balanced against the societal cost of hearing loss, which include the cost of special education and training and the loss of potential and lifetime earnings of an hearing impaired individual. Options for payment include insurance, self-pay, public sponsorship (full or partial), etc.
- x) *Support for the programme.* Public support from governmental and/or public health bodies would assist greatly in both the establishment, execution and funding of the UNHS. Some countries have used legislation to ensure that every infant has a hearing screen.

Despite the obvious challenges in the establishment of the UNHS, there is no doubt of its critical role in reducing the devastating effect of significant congenital, pre-lingual bilateral hearing loss. However, because of the need of sound audiological confirmatory testing and long-term intervention, it should be developed in tandem with these services.

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