Newborn Hearing Screening: Overview

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Background
Hearing is fundamental to speech and communication. Hearing loss in children can result in delayed speech and language development, social and emotional problems, and educational failure. Early detection of hearing loss, even as early as the newborn period, will lead to early intervention and reduction of hearing loss handicap.

Bilateral hearing loss occurs in approximately 1 to 3 per 1,000 newborns in the well-baby nursery population which is more common than other existing disease screening programs such as phenylketonuria (10 per 100,000 births) or hypothyroidism (25 per 100,000 births). Furthermore, the number of newborns with hearing loss identified in the newborn intensive care unit (NICU) is 10 – 20 times higher than in the general well-baby population.

In the past, only infants at risk for hearing loss were screened. Actually, about 50% of infants with permanent congenital hearing loss do not have any known risk factors. Therefore hearing screening among the high risk group missed approximately 50% of infants with hearing loss. Since 1994, the Joint Committee on Infant Hearing (JCIH) - formed by representatives from the American Speech-Language-Hearing Association, the American Academy of Otolaryngology-Head and Neck Surgery, the American Academy of Audiology, the American Academy of Pediatrics, and the Directors of Speech and Hearing Programs in State Health and Welfare Agencies) has endorsed universal newborn hearing screening programs to identify all infants with hearing loss before 3 months of age, and intervention to improve hearing should be started as early as 6 months of age.

The JCIH 2007 defines the targeted hearing loss for universal newborn hearing screening programs as: congenital permanent bilateral or unilateral, sensory or conductive hearing loss, with average hearing recognition at 30 to 40 dB or more in the speech range frequency (approximately 500 – 4000 Hz). Neural hearing loss (e.g. auditory neuropathy/dysynchrony) in infants admitted to the NICU is also included. Prior to the newborn hearing screening programs, children who had severe to profound hearing loss or multiple disabilities might be identified before age 2.5 years while children with mild to moderate hearing loss were often not identified until school age. Implementation of the universal newborn hearing screening programs has been lowering the age of hearing loss detection.

Many studies demonstrate the importance of early identification and intervention. In 1998, Yoshinaga-Itano and Sedey et al reported that children with hearing loss identified by age 6 months had a significantly higher receptive, expressive and total language development than those of children identified at 7-12, 13-18, 19-24, and 25-34 months. Also no significant differences in language ability were found among groups of children who were identified when they were older than 6 months. These support the importance of identification and intervention within the first 6 months of life.

The US Preventive Services Task Force (USPSTF) concludes that there is moderate certainty that the net benefit of screening all newborns for hearing loss is moderate (grade: B recommendation), while the Canadian Task Force on the Periodic Health Examination states that there is good evidence to support the recommendation that repeated examination of hearing, especially in the first year of life, should be specifically considered in a periodic health examination (A recommendation). Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood

According to the JCIH 2007 Position Statement, the risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood are as follows: (* are of greater concern for delayed-onset hearing loss)

1. Caregiver concern* regarding hearing, speech, language, or developmental delay.
2. Family history* of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO*, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as CMV*, herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone.
6. Physical findings that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss such as white forelock, etc.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss*, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.

8. Neurodegenerative disorders*, such as the Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.

9. Culture-positive postnatal infections associated with sensorineural hearing loss*, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.

10. Head trauma, especially basal skull/temporal bone fracture* that requires hospitalization.

11. Chemotherapy*. All newborns with risk factors should be closely monitored for any changes in hearing status as well.

Screening technologies

Although behavioral audiometry can be used for evaluating hearing in the newborn period up to 2 years of age, the test is not very reliable before age 8 to 9 months. Behavioral audiometry is not sensitive or specific enough for a screening program. Thus, objective physiologic tests are recommended for newborn hearing screening programs. Current physiologic measures used include otoacoustic emissions (OAEs), either transient-evoked (TEOAE) or distortion-product (DPOAE), and/or auditory brainstem response (ABR). Both technologies are noninvasive and easily recorded in neonates. Moreover, both OAE and ABR measures are highly correlated with the degree of peripheral hearing sensitivity.

Otoacoustic emissions (OAEs)

OAEs are physiologic measurements of the response of the cochlea’s outer hair cells to acoustic stimuli. OAE measurement is done in each ear by placing a probe in the ear canal, stimulating by clicks or tone pips, and then measuring within 60 seconds or less with a microphone. The presence of OAE responses indicates normal or near-normal hearing. Ear canal obstruction and middle ear effusion can eliminate the OAEs, causing a positive test result (a “refer” outcome) in a normal cochlea function.

As OAE responses are generated by the outer hair cells of the cochlea, OAE cannot detect neural hearing loss (e.g., auditory neuropathy/dysssynchrony). Accordingly, OAEs are not a sufficient screening test in newborns who are at risk for neural hearing loss and they should undergo an ABR screening.

Auditory brainstem response (ABR)

ABR is an electrophysiologic measurement to assess auditory function from the cochlea through the auditory brainstem pathway. Because the ABR is generated by auditory neural pathways, it will detect neural hearing loss in newborns.

Conventional or diagnostic ABR can be used to evaluate the degree and the nature of the hearing loss but a well-trained professional is needed to do the test. ABR also requires a long testing time up to 25-60 minutes per newborn. The automated ABR (AABR) system was later developed specifically for the mass hearing screening for neonates. AABR measurements are obtained by placing disposable surface electrodes on the forehead, on the mastoid, and on the nape of the neck. The click stimulus (usually set at 35 dB hearing level) is delivered to the ear via disposable earphones. AABR systems compare an infant's responses with normal template responses developed from normative ABR infant data and shows the result as “pass” or “fail”. The infant passes the AABR if reliable responses are present at the screening level of 35 dB HL or lower. AABR is practical because the machine can easily be operated by a trained nonprofessional and the screening time required to evaluate both ears is 3-10 minutes.

Principles of Newborn hearing screening

According to the JCIH 2007, effective early hearing loss detection and intervention is based on the following summarized principles:

- All infants should have access to hearing screening using a physiologic measure before 1 month of age.
- All infants who do not pass the initial hearing screen and the subsequent rescreening should have appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age.
- All infants with confirmed permanent hearing loss should receive intervention services before 6 months of age.
- The child and family should have immediate access to high-quality technology, including hearing aids, cochlear implants, and other assistive devices when appropriate.
- All infants and children should be monitored for hearing loss in the medical home. Continued assessment of communication development should be provided by appropriate providers to all children with or without risk indicators for hearing loss.

Screening protocols in the well-baby nursery

All infants should be provided with one hearing screening and, when necessary, a repeat screening before the time of discharge or rescreening as outpatients within 1 month of hospital discharge. Re-screening is important because there are transient conditions which may cause an initial screening failure such as vernix blocking the ear canal or middle ear fluid that may resolve later. If the infants were born outside the hospital, they should be screened for hearing loss as outpatients before 1 month of age. The screening measurements may be single technology either OAE or AABR. However, if AABR is used, neural hearing loss can be detected.

Some programs use OAE for the initial screening followed by AABR for rescreening (2-step protocol) to take advantage of the low cost and speed of the OAE screen, as well as the lower referral rate of the AABR screen. The 2-step protocol yielded a screening sensitivity of 0.92 and a specificity of 0.98. Infants tested by the 2-step protocol who do not pass an OAE screening, but pass an AABR later are considered a screening “fail”. On the contrary, infants in the well-baby nursery who fail AABR testing should not be rescreened by OAE because such infants are at risk of having a subsequent neural hearing loss.
Parents of well-infants who pass the newborn hearing screening should be provided with information about hearing, speech, and language milestones. At each pediatric periodic scheduled visit children should be monitored for auditory skills, middle ear status and developmental milestones. If the children develop middle ear diseases or the developmental delay was detected, they should be referred for further evaluation.

**Screening protocols in the NICU**

NICU infants should be screened close to discharge when they are medically stable. The JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU because those infants are at risk of having neural hearing loss.

Infants having a risk factor who pass the neonatal screening should have at least 1 diagnostic audiology assessment by 24 to 30 months of age and should be closely monitored for any changes in hearing status because they take a risk of having delayed-onset or progressive hearing loss.

**Management for infants who do not pass the newborn hearing screening**

All infants who do not pass the initial hearing screen and the subsequent rescreening should have appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age. Comprehensive assessment includes:

- Child and family history.
- Otoscopic examination.
- Audiological evaluation by OAE, diagnostic ABR, frequency-specific tone bursts ABR, air and bone conduction ABR. Behavioral observation is unreliable for this age group.
- Medical evaluation for identification of syndromes associated with early or late onset hearing loss and indicated radiological and laboratory studies (including genetic testing).
- Eye examination.

All infants with confirmed permanent hearing loss should receive intervention services before 6 months of age. The intervention consists of parent counseling, evaluation for amplification by hearing aid fitting (or cochlear implants after inadequate response to hearing aids), monitoring of amplification, speech rehabilitation, periodic communication assessment and therapy.

**Newborn hearing screening: Siriraj project**

In the Faculty of Medicine, Siriraj Hospital, the universal newborn hearing screening program has not been implemented yet. This is a challenging project that will require a large number of health care personnel and financial support because approximately 9,000-10,000 newborns will be screened annually. The Division of Neonatology, Department of Pediatrics has implemented the hearing screening for all infants with high risk factors before discharge since 2004. Srisuparp, et al reported in 2005 that the protocol using trained nursing staff to perform the screening yielded good results. Nowadays, the Department of Otorhinolaryngology takes part in comprehensive audiological evaluation and intervention, and looks forward to providing the universal newborn hearing screening program in the near future.

OAE = Otoacoustic emission; AABR = Automated auditory brainstem response; NICU = Neonatal intensive care unit; BOA = Behavioral observation audiometry; VRA = Visual reinforcement audiometry; w/ = with; w/o = without

### REFERENCES