

Importance of newborn hearing screening

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Abstract US Preventive Services Task Force recommends universal screening of all newborns for early detection of hearing impairment and early intervention to prevent its effects on normal development and acquisition of language skills. During comparison of universal screening of all newborns versus targeted screening of high risk cases to detect hearing impairment, it emphasized the importance of Universal newborn hearing screening (UNHS) among neonatologists, pediatricians as well as ENT specialists taking care of newborns and young children. Among those who had early versus late confirmation of hearing loss and those who had undergone universal newborn screening versus none, better language outcomes at school age were found than those not screened. Infants identified with hearing loss through universal newborn screening had earlier referral, diagnosis, and treatment than those not screened. Targeted screening of newborns with high risk criteria missed 50% of cases of hearing impairment, either because infants were hearing impaired but did not meet any of the high-risk criteria, or because they developed hearing loss after the newborn period.

Keywords Hearing impairment · Hearing screening · Targeted screening · Newborn screening.

Abbreviations UNHS—universal newborn hearing screening, PCHL—permanent congenital hearing loss, OAE—otoacoustic emission, ABR—auditory brainstem response.

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Introduction

In spite of different criteria used for defining hearing impairment, approximately 1-2 newborns/1000 live births have moderate to profound bilateral neural hearing loss. In addition 1-2/1000 may have milder or unilateral hearing impairments. The onset of hearing loss can occur at anytime during childhood. Compared with children with normal hearing, those with hearing loss have more difficulty learning vocabulary, grammar, word order, idiomatic expressions, and other aspects of verbal communication [1]. Hearing loss in children is also associated with delayed language, learning, and speech development and with low educational attainment [2]. Hearing disorders have also been associated with increased behavior problems, decreased psychosocial well-being, and poor adaptive skills [3, 4]. Once parents and teachers are aware of a valid reason for the child's poor attention or behavior, adjustments can be made.

The aim for Universal newborn hearing screening (UNHS) is to detect moderate-to-severe permanent, bilateral congenital hearing loss averaging 30 to 40 dB or more in the frequency region important for speech recognition (~500–4000 Hz). The focus of UNHS is on congenital as opposed to acquired or progressive hearing loss that may not be detected in the newborn period.

Targeted screening means selective screening of newborns based on the presence of risk factors or associated conditions. Newborns at high risk are those with risk factors known to be associated with PCHL and/or newborns admitted to the NICU. Risk factors associated with a higher incidence of PCHL include:

- NICU admission for ≥ 2 days
- Syndromes associated with hearing loss which include Waardenberg Syndrome, Pendred Syndrome, and Usher Syndrome
- Family history of hereditary childhood sensorineural hearing loss
- Craniofacial abnormalities

- Congenital infections such as cytomegalovirus, rubella, syphilis, herpes simplex and toxoplasmosis [2]
- Bacterial meningitis
- Birth weight less than 1500 grams
- Ototoxic medications use like amino glycosides, loop diuretics etc.
- Low Apgar scores of 0–4 at 1 min or 0–6 at 5 min
- Mechanical ventilation lasting ≥ 5 days
- However, $\sim 50\%$ of infants with PCHL do not have any known risk factors [5, 6]

Newborn hearing screening involves the use of objective physiologic measures. Currently, otoacoustic emissions (OAEs) and/or auditory brainstem responses (ABRs) are most often used to detect sensory or conductive hearing loss [7]. Both technologies are noninvasive recordings of physiologic activities that are quick, easily recorded in newborns, inexpensive and are highly correlated with the degree of peripheral hearing sensitivity. In UNHS programs, a 2-step process using OAEs followed by ABRs in those who fail the first test is often used to improve test performance. Under ideal conditions, instruments designed specifically for newborns can test and record findings on sleeping newborns in < 5 minutes. Screening is performed during postpartum hospitalization for most of the newborns or within one month for those born outside hospital. Screening tests like observing behavioral response to noise or Crib-o-gram are not recommended.

Infants not passing the newborn screening tests are referred for confirmatory testing for a diagnosis of PCHL. Confirmation requires an evaluation by an audiologist using behavioral, as well as technological, methods. It is recommended that early intervention (within six months of newborn screening) should be designed to meet the individualized needs of the infant and family, including acquisition of communication competence, social skills, emotional well-being, and positive self-esteem. Early intervention includes evaluation for amplification or sensory devices, surgical and medical evaluation, and communication assessment and therapy. Cochlear implants are often considered in infants with severe-to-profound hearing loss after inadequate response to hearing aids [8, 9]. Research in neurology and auditory cortical development suggests that early versus late implantation may be linked to more normal cortical auditory pathway development.

The focus of the report was on 3 key questions. Literature was searched to systematically identify articles relevant to the key questions [10]. Databases included the Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, Database of Abstracts of Reviews of Effects (through the fourth quarter of 2007), and Ovid Medline (2000 to November 2007 for key questions).

- (1) Among infants identified by universal screening who would not be identified by targeted screening, does initiating treatment before 6 months of age

improve language and communication outcomes? It was found that children with hearing impairment confirmed by ≤ 9 months of age had significantly better scores than those confirmed later on tests of receptive language and expressive language. Differences in higher scores for early versus late confirmation are equivalent to an increase of 10 to 12 points in the verbal compared with nonverbal IQ. Several outcome measures were examined using validated methods, including receptive and expressive language, receptive vocabulary, cognition, articulation, reading comprehension, intelligibility, and family functioning. Evaluators were blinded to the children's history and hearing status. Regression models were constructed that controlled for confounders, including nonverbal IQ, maternal education, paternal occupational prestige, and family functioning. All of these studies report better outcomes for children with hearing impairment identified and/or treated early versus late.

- (2) Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months of age for infants at average risk or for those at high risk? It was found that for children with unknown causes for hearing impairment, the median ages of confirmation for screened versus nonscreened children were 4.0 vs 25.0 months for mild or moderate impairment and 2.0 vs 15.0 months for severe or profound impairment. The median ages for hearing aid fitting for screened versus nonscreened children were 6.0 vs 30.5 months for mild or moderate impairment and 4.0 vs 16.0 months for severe or profound impairment.
- (3) What are the adverse effects of screening and early treatment? Parents expressed anxiety when their infants did not pass the initial screening test that resolved for most after a confirmatory test indicating normal results [11]. Most parents reported benefits of treatment, including improved communication, self-confidence, well-being, and social relationships.

Data from a large nonrandomized trial and descriptive studies indicate that infants at average and high risk with PCHL born in hospitals with UNHS have earlier referral and initiation of treatment than those born in hospitals without UNHS. The most recently published studies emphasize incorporation of UNHS as a routine practice. Studies of adverse effects of screening indicate that usual parental reactions to an initial nonpass include worry, questioning, and distress. Negative emotions resolve for most parents when a diagnostic test is provided with a normal result. Results of this review indicate that infants identified with PCHL through UNHS have significantly earlier referral, diagnosis, and treatment than those identified in other ways. Data also showed improved language outcomes at school age which strengthen the case for UNHS.

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