Newborn and Infant Hearing Loss: Detection and Intervention

ABSTRACT. This statement endorses the implementation of universal newborn hearing screening. In addition, the statement reviews the primary objectives, important components, and recommended screening parameters that characterize an effective universal newborn hearing screening program.

ABBREVIATIONS. UNHSP, universal newborn hearing screening program; EOAE, evoked otoacoustic emissions; ABR, auditory brainstem response; CDC, Centers for Disease Control and Prevention.

Significant hearing loss is one of the most common major abnormalities present at birth and, if undetected, will impede speech, language, and cognitive development.1–5 Significant bilateral hearing loss is present in ~1 to 3 per 1000 newborn infants in the well-baby nursery population, and in ~2 to 4 per 100 infants in the intensive care unit population. Currently, the average age of detection of significant hearing loss is ~14 months. The American Academy of Pediatrics supports the statement of the Joint Committee on Infant Hearing (1994), which endorses the goal of universal detection of hearing loss in infants before 3 months of age, with appropriate intervention no later than 6 months of age.6 Universal detection of infant hearing loss requires universal screening of all infants. Screening by high-risk registry alone (eg, family history of deafness) can only identify ~50% of newborns with significant congenital hearing loss.7,8 Reliance on physician observation and/or parental recognition has not been successful in the past in detecting significant hearing loss in the first year of life.

To justify universal screening, at least five criteria must be met:

1. An easy-to-use test that possesses a high degree of sensitivity and specificity to minimize referral for additional assessment is available.
2. The condition being screened for is otherwise not detectable by clinical parameters.
3. Interventions are available to correct the conditions detected by screening.
4. Early screening, detection, and intervention result in improved outcome.

The following are guidelines for the screening element of a UNHSP:

• Universal screening has as its goal that 100% of the target population, consisting of all newborns, will be tested using physiologic measures in both ears. A minimum of 95% of newborns must be screened successfully for it to be considered effective.16,19

• The methodology should detect, at a minimum, all infants with significant bilateral hearing impairment, ie, those with hearing loss ≥35-decibel in the better ear.1,16,19

• The methodology used in screening should have a false-positive rate, ie, the proportion of infants without hearing loss who are labeled incorrectly by the screening process as having significant hearing loss, of ≤3%. The referral rate for formal audiologic testing after screening should not exceed 4%.16,17,19,21

• The methodology used in screening ideally should have a false-negative rate, ie, the proportion of infants with significant hearing loss missed by the screening program, of zero.21,22

• Until a specific screening methodology is proved to be superior, the Academy defers recommendation as to a preferred method. Currently, acceptable methodologies for physiologic screening include evoked otoacoustic emissions (EOAE) and auditory brainstem response (ABR), either alone or in combination. Both methodologies are noninvasive, quick (<5 minutes), and easy to perform, although each assesses hearing differently. EOAE

5. The screening program is documented to be in an acceptable cost-effective range.11,12

Although additional studies are necessary, review of both published and unpublished data indicates that all five of these criteria currently are achievable by effective universal newborn hearing screening programs (UNHSP).5,13,15–28 Therefore, this statement endorses the implementation of universal newborn hearing screening. In addition, this statement reviews the primary objectives, important components, and recommended screening parameters that characterize an effective UNHSP.

The Academy recognizes that there are five essential elements to an effective UNHSP: initial screening, tracking and follow-up, identification, intervention, and evaluation.23,14 The child’s physician and parents, working in partnership, make up the child’s medical home and play an important role in each of these elements of a UNHSP.20

SCREENING11,13,14

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measures sound waves generated in the inner ear (cochlea) in response to clicks or tone bursts emitted and recorded via miniature microphones placed in the external ear canals of the infant. Although EOAE screening is even quicker and easier to perform than ABR, EOAE may be affected by debris or fluid in the external and middle ear, resulting in referral rates of 5% to 20% when screening is performed during the first 24 hours after birth. ABR measures the electroencephalographic waves generated in response to clicks via three electrodes pasted to the infant’s scalp. ABR screening requires the infant to be in a quiet state, but it is not affected by middle or external ear debris. Referral rates <3% may be achieved when screening is performed during the first 24 to 48 hours after birth. Referral rates <4% are generally achievable with EOAE combined with automated ABR in a two-step screening system or with automated ABR alone. In a two-step system using EOAE as the first step, referral rates of 5% to 20% for repeat screening with ABR or EOAE may be expected. The second screening may be performed before discharge or on an outpatient basis within 1 month of age. Screening should be conducted before discharge from the hospital whenever possible.

Each birthing hospital should establish a UNHSP with a designated medical (physician) director and sufficient staff to perform the following:

1. Develop the screening protocol and select the screening method(s).
2. Provide appropriate training and monitoring of the performance of staff responsible for performing hearing screening.
3. Provide the parents or guardians information concerning the screening procedure, costs, potential risks of hearing loss, and the benefits of early detection and intervention.
4. Establish a system that ensures confidentiality and allows the parents or guardians the opportunity to decline hearing screening. In most institutions, general hospital consent obtained at time of admission is considered to be inclusive of routine care, such as newborn hearing screening.
5. Ensure that all individuals performing hearing screening are trained properly in the performance of the tests, the risks including psychological stress for the parents, infection control practices, and the general care and handling of infants in hospital settings according to established hospital policies and procedures.
6. Establish clear guidelines for responsibility of documenting the results of the screening procedure.
7. Develop mechanisms for communicating results of screening in a sensitive and timely manner to the parents and the child’s physician(s). If repeat screening is necessary after discharge from the hospital, ensure that appropriate follow-up is provided.
8. Work with local, state, and national monitoring systems to identify all cases of significant hearing loss occurring in infants designated initially as free of hearing impairment by the UNHSP (false-negatives).

9. Secure funding for the program. Funding through third-party reimbursement is essential to cover the costs of the UNHSP, including the initial screen(s), as well as of diagnostic and intervention services. The cost of complete screening in statewide programs ranges from ~$7 to $26 per infant screened. Additional studies (some of which are ongoing) are necessary to quantify costs of tracking, diagnostic, and intervention services.

10. Collect critical performance data to ensure that each UNHSP meets the criteria specified in this statement. These data should be reported in a regular and timely manner to a statewide central monitoring program.

**TRACKING AND FOLLOW-UP**

The following are guidelines for the tracking and follow-up elements of a UNHSP:

- Universal screening has as its goal that there will be 100% follow-up of all infants referred for formal audiologic assessment and for all infants not screened initially in the birthing hospital whose parents did not refuse screening. A minimum of 95% successful follow-up is required for a UNHSP to be considered an effective screening program.
- State departments of health, in coordination with programs mandated by Part C of the Individuals with Disabilities Education Act, should:

1. Establish and maintain a central monitoring system for all hearing screening programs within the state. Critical performance data, including number of infants born; the proportion of all infants screened; the referral rate; the follow-up rate; the false-positive rate; and the false-negative rate should be collected in a timely manner.
2. Establish and maintain a tracking program that monitors all referrals and misses. Monitoring should ensure that children with significant hearing loss are not missed, ie, all children designated as free of hearing loss by the UNHSP, but who are later detected to have significant hearing loss, are identified by the statewide tracking program.
3. Develop mechanisms for communicating results of follow-up activities with the parents/guardians and the child’s physician(s), audiologist, and speech language therapist.
4. Ensure that hearing screening is performed on all out-of-hospital births.
5. Report the screening performance parameters of individual hospital-based UNHSFs within the state in a timely manner.
6. Report critical performance data of each UNHSP (without personal identifiers) to a national Early Hearing Detection and Intervention monitoring program established by the Centers for Disease Control and Prevention (CDC).
IDENTIFICATION AND INTERVENTION\textsuperscript{13–15,24–28}

The following are guidelines for the identification and intervention element of a UNHSP:

- Universal screening has as its goal that 100\% of infants with significant congenital hearing loss shall be identified by 3 months of age and shall have appropriate and necessary intervention initiated by 6 months of age.\textsuperscript{5–7}
- Appropriate and necessary care for the infant with significant hearing loss should be directed and coordinated by the child’s physician within the medical home, with support from appropriate ancillary services.\textsuperscript{29}
- A regionalized approach to identification and intervention for infants with significant hearing loss is essential, ensuring access for all children with significant hearing loss to appropriate expert services. It is recognized that professionals with demonstrated competency to provide expert services in the identification and intervention of significant hearing loss in young infants are not available in every hospital or community. The child’s physician, within the medical home, working with the state department of health must ensure that every infant with significant hearing loss is referred to the appropriate professional(s) within the regionalized system.
- It is anticipated that there will be increased demand for qualified personnel to provide age-appropriate identification and intervention services for young infants with significant hearing loss. As a result, there will be a need for the training and education of additional expert care providers.

EVALUATION\textsuperscript{13–15,26–28}

The following are guidelines for the evaluation element of a UNHSP:\textsuperscript{5}

- The UNHSPs should be evaluated on an ongoing and regular basis by the state monitoring system for performance with regard to parameters enumerated in “Screening” above.
- Tracking and follow-up should be evaluated on an ongoing and regular basis by the state monitoring system, as well as through a national monitoring system to be established by the CDC.
- Intervention services should be evaluated on an ongoing and regular basis by the state department of health to ensure that sufficient expert services are available for children identified with significant hearing loss, that the services are accessible to the children in need, and that outcomes from interventions provided are effective.

OTHER RECOMMENDATIONS AND ISSUES

The following are additional recommendations of the Academy for developing a UNHSP:

- The Academy recommends that each American Academy of Pediatrics chapter assume a leadership role in state-based efforts to promote optimal implementation of UNHSPs. Effective state-wide programs require broad-based support and collaboration. Collaboration should include (but not be limited to) appropriate professional organizations, parent advocacy groups, deaf and hard-of-hearing adults, physicians, audiologists, speech and language therapists, nurses, administrators, payers, legislators, and state departments of health and special education.
- The Academy shall identify, develop, and disseminate educational materials regarding effective hearing screening programs.\textsuperscript{15}
- To promote additional research and the development of the needed infrastructure to provide universal newborn hearing screening, the Academy recommends the following:
  1. The National Institutes of Health support ongoing research to improve the efficacy of screening, identification, and intervention.
  2. The Health Resources and Services Administration promote the development of a state-based early hearing loss identification and intervention network.
  3. The CDC establish and maintain a national monitoring and evaluation program for early hearing loss identification and intervention.
- Physicians should provide recommended hearing screening, not only during early infancy but also through early childhood for those children at risk for hearing loss (eg, history of trauma, meningitis) and for those demonstrating clinical signs of possible hearing loss.\textsuperscript{9,14} Although most hearing loss in children is congenital (ie, present at birth), a significant portion of hearing loss is acquired after birth.\textsuperscript{2–4} Regardless of the age of onset, all children with hearing loss require prompt identification and intervention by appropriate professionals with pediatric training and expertise.

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\textsuperscript{AMERICAN ACADEMY OF PEDIATRICS 529}

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