Universal Newborn Hearing Screening

A. STEVENS WRIGHTSON, MD, *University of Kentucky, Lexington, Kentucky*

Congenital hearing loss is estimated to affect one in every 1,000 newborns. Causes of hearing loss can be conductive, sensorineural, mixed, or central. Known risk factors for congenital hearing loss include cytomegalovirus infection and premature birth necessitating a stay in the neonatal intensive care unit. However, up to 42 percent of profoundly hearing-impaired children will be missed using only risk-based screening. Universal newborn hearing screening is a way to identify hearing-impaired newborns with or without risk factors. Newborns with positive screening tests should be referred for definitive testing and intervention services. Whether early intervention in hearing-impaired children identified with universal screening improves language and communication skills has not been established by good-quality studies. However, universal screening has been endorsed by most national children's health organizations because of the ease of administering the screening tests and the ability to identify children who may need early intervention. (Am Fam Physician 2007;75:1349-1352. Copyright © 2007 American Academy of Family Physicians.)

During the 2002-2003 school year, nearly 72,000 children in the United States received special services for hearing impairment.[1] The incidence of congenital hearing loss (i.e., hearing loss present at birth) is estimated to be one out of every 1,000 live births; however, emerging data from states with universal screening indicate that the incidence may be closer to two or three per 1,000 live births.[2]

If congenital hearing loss is not recognized and managed, a child's speech, language, and cognitive development are often severely delayed.[3]

Etiology of Hearing Loss

There are four types of hearing loss: conductive, sensorineural, mixed, and central.[2] Conductive losses are caused by a problem with the outer or middle ear and usually affect all frequencies to the same degree. Sensorineural loss is caused by problems in the inner ear or auditory nerve. Mixed losses involve both conductive and sensorineural etiologies. Central hearing losses are rare and are caused by problems along the auditory pathway or in the brain itself.[2]

More than 50 percent of hearing impairment in children is thought to be genetic and not related to infectious, anatomic, or other noninherited causes.[4] Cytomegalovirus infects about 1 percent of all newborns in the United States. Annually, between 6,000 and 8,000 infected newborns will have clinical manifestations, and 75 percent of these infants will develop sensorineural hearing loss.[4] *Table 1*[4] lists various causes of congenital hearing loss.

Risk Factor–Based Screening Is Not Enough

An admission to the neonatal intensive care unit (NICU) for more than two days increases the likelihood of the presence of hearing impairment 10-fold.[5] Early screening programs focused on children with known risks.[6] *Table 2* lists risk factors for hearing impairment in newborns.[7,8] However, recent studies indicate that 19 to 42 percent of profoundly hearing-impaired children will be missed with targeted, risk factor–based screening.[9]

Rationale for Universal Screening

Screening for hearing loss in newborns is based on two concepts. First, a critical period exists for optimal language skills to develop, and earlier intervention produces better outcomes.[10,11] Second, treatment of hearing defects has been shown to improve communication.[12]

Data from cohort studies indicate that diagnosis and intervention before six months of age can improve language and speech acquisition in hearing-impaired

SORT: KEY RECOMMENDATIONS FOR PRACTICE

Clinical recommendation	*Evidence rating*	*References*
Universal newborn hearing screening should be used to accurately diagnose moderate to severe sensorineural hearing loss.	C	7
Children with risk factors for hearing loss who have a negative hearing screen at birth should undergo audiologic testing every six months until three years of age.	C	7, 17, 19
Identification of hearing loss before six months of age improves language development and communication skills.	B	9, 15

A = consistent, good-quality patient-oriented evidence; B = inconsistent or limited-quality patient-oriented evidence; C = consensus, disease-oriented evidence, usual practice, expert opinion, or case series. For information about the SORT evidence rating system, see page 1289 or http://www.aafp.org/afpsort.xml.

children.[9-11] However, in children who are not screened at birth, diagnosis of hearing loss may be delayed by as long as three years.[13] Lowering the age at which screening and intervention services are initiated may improve language, cognitive, and social outcomes in hearing-impaired children.[14]

No prospective studies have compared outcomes of children who received screening through a universal newborn hearing screening program and those who were managed using a risk factor–based approach. Consequently, the U.S. Preventive Services Task Force found insufficient evidence to recommend for or against universal newborn hearing screening in the immediate newborn period.[15]

Despite the lack of high-quality evidence, in 1993 the National Institutes of Health Consensus Development Conference on Early Identification of Hearing Impairment in Infants and Children recommended universal newborn screening.[16] The Joint Committee on Infant Hearing issued similar guidelines in 1995 and again in 2000.[7,17] Universal newborn hearing screening is also recommended by the American Academy of Pediatrics, by the Centers for Disease Control and Prevention, and in Healthy People 2010.[7,18]

Children with risk factors should be screened not only at birth but also throughout childhood. The Joint Committee on Infant Hearing recommends continued surveillance of these children because they may be at risk of progressive hearing loss.[7,17] This recommendation includes audiologic testing every six months until three years of age.[19] In low-risk children, a repeat hearing screening is recommended before entry into kindergarten.

Table 1. Congenital Causes of Hearing Loss in Infants and Children

Central (rare)
Hyperbilirubinemia/kernicterus
Hypoxia
Intraventricular hemorrhage
Conductive
Anomalies of the pinna, external ear canal, tympanic membrane, or ossicles
Congenital cholesteatoma
Sensorineural
Anatomic
Genetic*: isolated impairment or syndromes (e.g., Waardenburg's syndrome [autosomal dominant], Usher's syndrome [autosomal recessive], Alport's syndrome [sex-linked disorder], Turner's syndrome [chromosomal abnormality])
Idiopathic
Infectious (e.g., cytomegalovirus, syphilis, herpesvirus, rubella, toxoplasmosis, group B streptococcal sepsis, streptococcal virus)

**—Causes 50 percent of sensorineural newborn hearing loss.*

Information from reference 4.

Screening Tests

Evaluation of hearing should include a demonstration of a behavior in response to a measured stimulus; this cannot be reliably performed on a child younger than eight to nine months.[9] Tests for infants include automated auditory brainstem response (AABR) and the transient evoked otoacoustic emissions (TEOAE) test.

AUTOMATED AUDITORY BRAINSTEM RESPONSE

AABR tests the auditory pathway from the external ear to the lower brainstem. The newborn's ears are covered with earphones that emit a series of soft clicks. Electrodes on

Table 2. Risk Factors for Congenital Hearing Loss

Craniofacial anomalies, including abnormalities of the pinna and ear canal
Family history of hereditary childhood sensorineural hearing loss
Neonatal intensive care unit admission for more than two days
Rubella or other fetal infection (e.g., herpes, cytomegalovirus)
Syndromes associated with hearing loss (e.g., Usher's syndrome, Waardenburg's syndrome)

Information from reference 7.

the infant's forehead and neck measure brain wave activity in response to the clicks. A computer then compares the brain wave activity with normal response templates and provides a pass or refer (i.e., fail) report.[5]

TRANSIENT EVOKED OTOACOUSTIC EMISSIONS

The TEOAE test evaluates the function of the peripheral auditory system, primarily the cochlea, which is the area most often involved in sensorineural hearing loss. Normally the cochlear hair cells generate "echoes" when presented with sound waves. The TEOAE test measures these echoes by placing a small microphone in the external ear canal and testing the response to a series of clicks. As with AABR, a computer-generated report compares the newborn's response to standardized emission norms and provides a pass or refer report.[5]

AABR and TEOAE have been shown to accurately diagnose moderate to profound sensorineural hearing loss in newborns. The sensitivity and specificity measured against an independent benchmark were 84 and 90 percent, respectively.[9] Children with a positive result should be referred for definitive testing and evaluation, including a detailed family history for genetic causes.[7] Using these devices, a universal newborn hearing screening program has been shown to reduce by eight months the average age at which hearing-impaired children receive hearing aids (i.e., to about six months of age).[14] There currently is no evidence to support either AABR or TEOAE as the preferred method of screening newborns.[7]

Drawbacks of Universal Newborn Hearing Screening

Universal newborn hearing screening produces a large number of false-positive test results. Both AABR and TEOAE can be influenced by motion artifact and therefore are more specific if performed on a sleeping child in a quiet room. The rate of false positives ranges from more than 30 percent for one-step programs using TEOAE[20] to less than 1 percent with a two-step process, such as retesting a child before discharge if the initial test is positive.[21]

Increased parental anxiety may result from a false-positive test, although this finding has not been demonstrated consistently in all studies.[22,23] Qualitative studies indicate that negative parental emotions may be addressed with more systematic education before and after screening.[23]

Despite these concerns, the consensus of multiple organizations that develop children's health guidelines is that the potential benefits of universal newborn hearing screening outweigh its adverse effects.[7] Currently, 37 states and the District of Columbia have enacted legislation requiring that hearing screening be performed on all newborns in hospitals and birthing centers *(Table 3)*.[24] Additional resources for physicians and patients are provided in *Table 4*.

Table 3. States That Have Legislation Requiring Universal Newborn Hearing Screening

Arkansas	Maine	Oklahoma
California	Maryland*	Oregon
Colorado	Massachusetts	Pennsylvania
Connecticut	Mississippi	Rhode Island
Florida	Missouri	South Carolina
Georgia	Montana	Texas
Hawaii	Nebraska	Utah
Illinois	Nevada	Virginia
Indiana	New Hampshire	West Virginia
Iowa	New Jersey	Wisconsin
Kansas	New York	Wyoming
Kentucky	North Carolina	
Louisiana	Ohio	

**—Including the District of Columbia.*

Information from reference 24.

Table 4. Additional Resources on Hearing Loss

National Center on Birth Defects and Developmental Disabilities
Web site: http://www.cdc.gov/ncbddd/

National Dissemination Center for Children with Disabilities
Web site: http://www.nichcy.org

National Institute on Deafness and Other Communication Disorders
Web site: http://www.nidcd.nih.gov/health/hearing

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The Author

A. STEVENS WRIGHTSON, MD, is an assistant professor in the Department of Family and Community Medicine at the University of Kentucky in Lexington. Dr. Wrightson received his medical degree from the University of Kentucky College of Medicine and completed his residency at the University of Kentucky Chandler Medical Center in Lexington.

Address correspondence to A. Stevens Wrightson, M.D., Dept. of Family and Community Medicine, K342 Kentucky Clinic, 800 South Limestone, Lexington, KY 40536 (e-mail: aswrig2@email.uky.edu). Reprints are not available from the author.

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