



SOUND IDEAS

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Some babies are born listeners . . . Others need your help.

U.S. Preventive Services Task Force report on newborn hearing screening

On October 23, 2001, The *Journal of the American Medical Association* (JAMA) published an article based on the recently released statement from the U.S. Preventive Services Task Force (USPSTF) about newborn hearing screening. This report and the JAMA article have been frequently cited in ways that have led to incorrect conclusions. As stated in the JAMA article, the goal of the USPSTF report was certainly worthwhile:

To identify strengths, weaknesses, and gaps in the evidence supporting UNHS [universal newborn hearing screening].

Unfortunately, several of the headlines and statements in recently released articles and commentaries are misleading or contribute to readers drawing conclusions that were never intended.

It is very important to understand what was really said in the USPSTF report and associated JAMA article. Unfortunately, several of the headlines and statements in recently released articles and commentaries are misleading or contribute to readers

drawing conclusions that were never intended. For example:

Expert panel won't endorse routine newborn hearing tests.
—American Hospital Association News

Researchers find hearing testing for newborns a wash.
—Oregonian Newspaper

U.S. Task Force takes no stand on neonatal screening for hearing loss.
—Reuters Health

Such headlines have led many people to conclude that the Task Force is opposing universal newborn hearing screening programs. However, the actual report and statements to the media by the authors of the reports and the chair of the USPSTF give a very different message. For example, Diane Thompson, the senior author for the JAMA article, told HealthScout.com on October 23, 2001:

We're concerned that the media will interpret our report to say we are not in favor of universal screening, and that is not true. It's just that the evidence to show whether universal screening can improve language skills for these children is not clear . . . Modern hearing tests make it possible to screen newborns with high specificity and sensitivity . . . They've gotten better and better in the last 5 or 6 years.

The next day, Mark Helfand, the senior author for the USPSTF Systematic Evidence Review Number 5: Newborn Hearing Screening, told *The Oregonian* newspaper:

Our summary of evidence did not find evidence anywhere that screening is either harmful or that it doesn't work . . . We just failed to find some proof that by detecting them and treating them earlier, you really make a difference in how well their language is in the long run . . . There is no indication in our work that parents should turn screening down, or that it doesn't work, or that a deaf child won't benefit.

Finally, Alfred O. Berg, the chair of the USPSTF, told *Reuters News* on October 24, 2001:

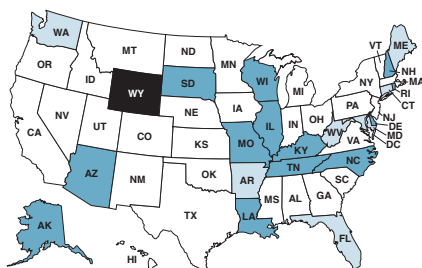
This is not a recommendation to stop screening, nor is it a recommendation to start screening. It's just an assessment of what evidence we have that supports the practice . . . If you're going to [conduct neonatal screening for hearing], you ought to do it in a systematic way so we can gather evidence necessary to tell whether it makes a difference. continued, page 5

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Federal funding for EHDI expands: A personal perspective from Arkansas

Thanks to funding made available as a result of legislation sponsored by Congressman James Walsh (R-NY), 23 more states and territories have been awarded new federal grants or cooperative agreements to expand and improve early hearing detection and intervention programs (see map). This brings the total number of states and



■ CDC
■ MCHB
■ CDC & MCHB

territories with federal funding specifically targeted on EHDI programs to 51. MCHB grants focus primarily on developing a statewide system of EHDI services, while CDC cooperative agreements focus primarily on developing better tracking and data management systems for EHDI programs and integrating those systems with other public health information systems, such as heelstick screening, vital statistics, and immunization registries. As a part of this funding, several of the states (i.e., CO, HI, RI, and UT) are also conducting targeted research about EHDI programs in cooperation with staff at CDC.

Most of these states were assisted in developing their funding applications by a workshop conducted by NCHAM and funded by MCHB last May. The following comments from the EHDI coordinator in Arkansas provide useful

insights about how people can work together cooperatively and systematically to develop a successful proposal for federal funding.

As coordinator of the Arkansas Infant Hearing Program, I participated in writing a grant to the Maternal and Child Health Bureau (MCHB) in December 2000 for establishing the Arkansas Early Hearing Detection and Intervention (EHDI) system. When the grant was not funded, I was disappointed. A few weeks later, though, I learned that MCHB was sponsoring a workshop where staff from the National Center for Hearing Assessment and Management (NCHAM) would help those states that had not yet received MCHB funding for its EHDI system to develop a successful proposal. Each state was to send several people, including representatives from audiology, the state agency for the Children With Special Health Care Needs (CSHCN) program, parents, early interventionists, and the infant hearing screening coordinator. Attending from Arkansas were the state coordinator for infant hearing screening, a pediatric audiologist with 12 years' experience at Arkansas Children's Hospital, the parent of a (adult) child with hearing loss who is also the director of the Disability Rights Center in Little Rock, the medical director of the CSHCN program for Arkansas, and me.

At the Salt Lake City meeting on April 30-May 1, 2001, attendees from each state were seated at the same table, along with a national EHDI

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THE WEB

www.infanthearing.org

- For answers to frequently asked questions of parents of deaf and hard of hearing infants and children, go to the Texas Connect home page under "Links" at www.infanthearing.org or go directly to www.callier.utdallas.edu/txc.html.
- Need information on issues related to Universal Newborn Hearing Screening? Refer to our *Abstracts and Citations* on the Web at www.infanthearing.org. Find abstracts on topics such as:
 - Family Issues
 - Etiology and Epidemiology of Hearing Loss
 - Policy Statements
 - Recommendations and Consequences of Hearing Loss



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WE'RE ELECTRONIC!

Future editions of this newsletter will primarily be distributed electronically on our Web site at <http://www.infanthearing.org>. To have a printed copy sent, register at www.infanthearing.org. Send us your E-mail address at nchamhelp@coe.usu.edu, and we will e-mail you each time the newsletter is published. If you would like to submit an article, contact the editor, Karen Ditty, at DittyKM@aol.com.

When do I refer for audiological and medical evaluation?

The second principle listed on the Joint Committee on Infant Hearing (JCIH): Year 2000 Position Statement, Principles and Guidelines for Early Hearing Detection and Intervention Programs states:

All infants who do not pass the birth admission screen and any subsequent rescreening begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age.

Achieving this goal is not always as easy as it sounds. The majority of infants pass their initial hearing screen; however, there will be some that “refer” from their initial hearing screen.

What diagnostic services are available in the community?

Depending on the size of the community and the parents’ financial resources, traveling to another city may be necessary for appropriate audiological and medical intervention. Hospital screening program staff should be aware of what resources are available in their community so they can advise parents about the alternatives.

Screening programs should also emphasize the need to keep referral rates as low as practicable. Some protocols provide for outpatient rescreening of infants who do not pass the birth screen within 1 month of hospital discharge. This mechanism for rescreening also minimizes the number of false-positive referrals for follow-up audiological and medical evaluation and is invaluable in communities where audiological personnel are limited.

is referred, follow up should occur as soon as possible, but no later than 90 days after birth. The hospital personnel should stress the importance of this follow-up evaluation to the parents as a precaution to rule out hearing loss and communicate this information to the baby’s primary care physician. The physician’s role as that infant’s “medical home” is critical to ensure the infant will return for follow-up

audiological and medical evaluations. If a hearing loss is determined, then habilitation should commence as soon as possible, but not later than the infant’s 6-month birthday. This time period is recommended by the JCIH position statement to initiate early intervention for speech and language development.

What is an appropriate diagnostic audiological evaluation?

Audiological evaluations of infants and young children need to be provided by an audiologist who specializes in pediatric audiology. The audiologist should be able to perform diagnostic ABR testing (including clicks, tone pips, and bone-conducted clicks when applicable), OAE, and Tympanometry procedures for infants less than 6 months of age. They should also be able to perform or refer the infant to a pediatric audiologist familiar with the protocols of fitting an infant with a hearing aid if required.

Medical evaluation should be included when hearing loss is confirmed to identify any anatomical or medical causes of the hearing loss and to provide appropriate genetic counseling.

Without committed personnel and persistent UNHS program managers, infants “referred” for subsequent audiological follow up can be lost among the sea of infants born each day. Having both the policies and procedures clearly spelled out for the hospital personnel and appropriate pediatric audiology sites identified for follow-up testing will make the referral process a friendlier and less-intimidating process for the family of the newborn.

TECHNIQUES AND TIDBITS

Looking for a family resource guide for EHDI programs?

The University of Texas at Dallas has developed some wonderful materials which are very useful for people providing services to families of infants and young children who are deaf or hard of hearing. They are called *Texas Connect Family Resource Guide* and *Texas Connect Topic Cards*. Although they were originally developed for programs in Texas, they are available to anyone at very reasonable prices. The materials include:

- A brochure for physicians and hospital personnel that provides information related to diagnostic assessment and intervention for infants referred through the newborn hearing screening program.
- A resource guide for families that provides basic information related to identification and early intervention for infants and young children who are deaf and hard of hearing (English or Spanish).
- Topic cards for families that provide information on topics of individual interest related to assessment and early intervention for infants and young children who are deaf and hard of hearing (series of 12 cards in English or Spanish). Topics include:
 - Tests Used to Assess Hearing
 - Types and Causes of Hearing Loss
 - Options for Communication
 - On Being Deaf—A Cultural View
 - Tips for Infant Hearing Aid Use
 - The How and When of Cochlear Implants
 - Encouraging Early Communication
 - How a Hearing Aid Works, and more

Karen Clark is the project director and had the benefit of numerous contributors to the finished product. The *Texas Connect Family Education Topics Cards* and portions of the *Family Resource Guide* are available at the UTD/Callier Center for Communication Disorders (www.callier.utdallas.edu).

The genetics of infant hearing loss

About 3 in every 1,000 newborns has a permanent hearing loss. About 50% of these losses are thought to be due to environmental factors, such as bacterial or viral infections like rubella or CMV or the use of ototoxic drugs, such as aminoglycosides. Other times, the cause is genetic and is due to changes in the genes involved in the hearing process. In about 30% of babies with a hearing loss, the loss is part of a syndrome, meaning that these babies have other medical problems. More than 400 syndromes have been identified which can cause hearing loss (e.g., Waardenburg, Usher, Pendred, Alport, etc.). The other 70% of cases are nonsyndromic, which means that the baby does not have other medical problems.

According to the Joint Committee on Infant Hearing, the families of all babies with hearing loss for whom there is not

Families of all babies with hearing loss for whom there is not a clear-cut etiology should be given the option of genetic evaluation and counseling by a medical geneticist.

a clear-cut etiology should be given the option of genetic evaluation and counseling by a medical geneticist. Many people think that the primary reason for such genetic evaluation and counseling is so the family can be informed about their chances of

having additional children with hearing loss. In reality, the genetic evaluation yields much more important information that can have a significant impact on how the infant is treated. For example, whether a baby's hearing loss will become worse can sometimes be predicted if the specific cause is known. Also, for a substantial number of infants, deafness is only one of a group of medical problems the baby may have, and genetic testing may indicate whether the baby is likely to have other problems with the heart, kidneys, or eyes. The following table indicates problems other than deafness associated with common forms of syndromic deafness, all of which can be identified through genetic evaluations.

Common Forms of Syndromic Deafness	
Syndrome	Main Features (besides deafness)
Alport	Kidney problems
Branchio-oto-renal	Neck cysts and kidney problems
Jervell and Lange-Nielsen	Heart problems
Neurofibromatosis Type 2	Nerve tumors near the ear
Pendred	Thyroid enlargement
Stickler	Unusual facial features, eye problems, arthritis
Usher	Progressive blindness
Waardenburg	Skin pigment changes

Recent discoveries about the genetics of hearing loss have substantially increased the information available through genetic testing. For example, recent discoveries about mutations

in the Connexin 26 gene have shown that this gene may be responsible for 20-30% of all congenital hearing loss. There are several different types of mutations in this gene which can contribute to hearing loss. One mutation called "35delG" is found in 2-3% of Caucasians of European descent. Another mutation of the Connexin 26 gene referred to as "167delT" is found in almost 5% of the Ashkenazi Jewish population. Usually mutations in the Connexin 26 gene are recessive, meaning that both the mother and father of the baby need to have the mutation for it to cause a hearing loss in the baby. Hearing loss due to mutations of the Connexin 26 gene is almost always congenital, severe profound, not progressive, and occurs without any other medical problems (nonsyndromic).

Making sure that all infants with hearing loss—where the etiology is not clear—receive competent genetic evaluation and counseling can have many benefits for the baby and family. Such procedures can:

- Avoid unnecessary and often costly clinical tests necessary to rule out conditions associated with syndromic deafness.
- Determine the cause of hearing loss which can dispel misinformation and allay parental guilt.
- Develop an individualized treatment strategy to ameliorate the complications of various syndromes (e.g., Vitamin A therapy may be beneficial to persons with Usher syndrome in slowing retinal degeneration, or treatment of children with Jervell and Lange-Nielsen syndrome can minimize cardiac complications).
- Anticipate potential associated health problems and offer appropriate referral for therapeutic options (e.g., in Stickler syndrome, doctors can watch for myopia and early retinal detachment; or in Branchio-oto-renal syndrome, doctors can watch for kidney complications).

The benefits of making sure genetic testing and evaluation is offered to parents of children with hearing loss are substantial. However, appropriate explanations of the genetics of

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hearing loss can be complicated and difficult to understand. An excellent pamphlet is available from the Harvard Medical School Center for Hereditary Deafness and can be viewed or downloaded at www.infanthearing.org/ehdi/index.html. Finding a place to do genetic testing can also be difficult for families who do not live near major population centers. An excellent Web site which lists many of the labs that do genetic testing is available at www.genetests.org under the "Laboratory Directory" section. Additional information is now being prepared by the American College of Medical Genetics and will be distributed at the first national EHDI meeting in Washington, DC (www.infanthearing.org/announcements/2002ehdi/index.html).

U.S. Preventive Services Task Force report . . . from page 1

So what are the major conclusions of the USPSTF report on Newborn Hearing Screening and the JAMA article? The following quotes come directly from the JAMA article:

- *Moderate, severe, and profound congenital PHL [permanent hearing loss] is associated with delayed language, learning, and speech development. This delay is measurable before age 3 years and has consequences throughout life. On average, deaf students graduate from high school with language and academic achievement levels below those of fourth-grade students with normal hearing (p. 2000).*
- *Diagnosis and treatment are often delayed until ages 1 or 2 years in children with congenital PHL, particularly among those at low risk for PHL. Current theory holds that auditory stimuli during the first 6 months of life are critical to the development of speech and language skills (p. 2000).*
- *In the only controlled trial, parents whose infants were screened had anxiety and attitudes similar to parents in the unscreened group (p. 2006).*
- *UNHS increases the chance that diagnosis and treatment will occur before age 6 months (p. 2008).*
- *Evidence is inconclusive . . . [that] identification and treatment prior to age 6 months improve[s] language and communication in infants who would not be diagnosed that early in a selective, high-risk screening program (p. 2008).*
- *Modern screening tests for hearing impairment can improve identification of newborns with PHL, but the efficacy of UNHS to improve long-term language outcomes remains uncertain (p. 2000).*

Taken together—especially in conjunction with the massive amounts of experiential and clinical evidence demonstrating the importance of identifying infants and young children with congenital hearing loss as early as possible—the USPSTF report and the JAMA article are really an endorsement of the universal newborn hearing screening programs now mandated by law in 36 states. Although the USPSTF appropriately notes that more research is needed on the long-term effects of language development, the report provides convincing data that delayed identification of hearing loss has lifelong negative consequences, newborn hearing screening has high specificity and sensitivity and leads to earlier identification, and researchers have not yet discovered negative side effects. Rather than worry about the negative impact of a few misleading media headlines, people advocating for young children should be using these

positive conclusions to continue the expansion and improvement of universal newborn hearing screening and intervention.

The full USPSTF report, the JAMA article, and responses from various groups and organizations can be viewed at www.infanthearing.org/announcements/taskforce/index.html.

Federal funding . . . from page 2

technical assistance advisor and/or NCHAM representative and the principal investigator from a successful MCHB EHDI grant. The opportunity to discuss various aspects of an EHDI program for Arkansas with participants in those areas was invaluable. In fact, each of the other participants contributed ideas of how particular facets of EHDI might be implemented in Arkansas. Collectively, we identified the most immediate needs for the Arkansas EHDI system, which led to some immediate changes upon our return to Little Rock. Additionally, workshop staff provided a sounding board for our ideas as well as input on writing the grant and implementing EHDI. Each state is unique and at various stages of EHDI implementation. What information is pertinent and how that information should be presented can vary among states. An open discussion of grant dos and don'ts was moderated by workshop staff, complete with questions for the MCHB representative, which proved to be beneficial later when writing the grant. In addition to other state representatives sharing their strategies in writing their successful grants, this discussion helped us understand how to fit the required information about the state EHDI system into the specific format for the grant. With a 35-page limit to the proposal, that information was essential!

It was very exciting when we were notified in September that Arkansas had been approved for funding. I know that the grant Arkansas submitted this year was more complete than the December 2000 grant. In large part, I can attribute that to the 2-day meeting sponsored by MCHB and hosted by NCHAM. Thanks!

—Laura Smith Olinde

EDITOR'S NOTE: For information or a summary of your state's EHDI program, go to www.infanthearing.org and click on "State EHDI Grants."

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primarily be distributed electronically
at www.infanthearing.org.

To have a printed copy sent,
register at www.infanthearing.org.
HEAR's to early hearing screening
for all babies!

UPCOMING EVENTS

February 11-13, 2002 • MCHB/CDC Annual Meeting on Successful Statewide Early Hearing Detection and Intervention Programs, 2002 National EHDI Meeting, Tysons Corner Marriott Hotel, Vienna, Virginia. **Contact:** Kathleen Watts, 435.797.3459, www.infanthearing.org/announcements/2002ehdi/index.html.

March 18, 2002 • RIHAP Eighth Annual Seminar, "Challenge 2002: Pathways for Families With Children Who Are Deaf or Hard of Hearing," Rhode Island Convention Center, Providence, Rhode Island. **Contact:** RIHAP, 401.274.1122, ext. 1853 or 1836, www.womenandinfants.com/pathways.html.

April 18-21, 2002 • American Academy of Audiology Annual Meeting, Philadelphia, Pennsylvania. **Contact:** American Academy of Audiology, 8201 Greensboro Drive, Suite 300, McLean, VA 22102, 703.610.9022, Fax: 703.610.9005.

May 30-June 1, 2002 • Second International Conference on Newborn Hearing Screening, Diagnosis, and Intervention, Milan (Villa Erbal/Como), Italy. **Europe Contact:** Dr. Ferdinando Grandori, Fax: +39.02.2399.3367, E-mail: nhs2002@biomed.polimi.it. **USA Contact:** Dr. Deborah Hayes, 303.861.6424, E-mail: hayes.deborah@tchden.org.