Workshop Proceedings

National Workshop on Mild and Unilateral Hearing Loss

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Minimal is not inconsequential.

–Fred Bess, 2004[1]

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This report was developed with contributions from workshop participants and prepared by Jamie Elliott, with assistance from other members of the CDC Early Hearing Detection and Intervention program, under the leadership of John Eichwald.

Abstract

The Early Hearing Detection and Intervention (EHDI) Program at the Centers for Disease Control and Prevention (CDC), in collaboration with the Marion Downs Hearing Center, convened the National Workshop on Mild and Unilateral Hearing Loss on July 26–27, 2005, in Breckenridge, Colorado. More than 50 national and international experts representing the areas of research, clinical practice, early intervention, parent and national organizations, and state and federal agencies attended the workshop. The goal was to review and discuss information that would facilitate the development of a set of recommendations related to identification, assessment, and intervention appropriate for infants and children with mild and unilateral hearing loss.

Prior to the workshop, participants were given an overview of the relevant research related to prevalence, screening, diagnostics, amplification, early intervention eligibility, clinical practice, and outcomes. The workshop began with plenary presentations addressing relevant topics, such as the scientific evidence related to the prevalence and impact of mild and unilateral hearing loss. Discussion among four breakout groups addressed screening for hearing loss, diagnostic evaluation and follow-up, hearing technology, and early intervention. Each of these groups identified issues and barriers, as well as areas for future research, which are discussed in this report. Each breakout group also began discussing both short-term and long-term recommendations. Key recommendations, once finalized, are expected to be published in 2006.

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Acronyms and Abbreviations

Organizations / Agencies / Programs

AAA	American Academy of Audiology
AAP	American Academy of Pediatrics
ASHA	American Speech-Language-Hearing Association
CDC	Centers for Disease Control and Prevention
CDC EHDI	Centers for Disease Control and Prevention / Early Hearing Detection and Intervention (Program)
CHAC	Children's Hearing Assessment Centre (Nottingham, England)
CSDB	Colorado School for the Deaf and the Blind
DSHPSHWA	Directors of Speech and Hearing Programs in State Health and Welfare Agencies
HRSA	Health Resources and Services Administration
JCIH	Joint Commission on Infant Hearing
MCHAS	Modernising Children's Hearing Aid Service (University of Manchester, England)
MCHB	Maternal and Child Health Bureau
NCBDDD	National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention
NCHAM	National Center for Hearing Assessment and Management
NECTAC	National Early Childhood Technical Assistance Center
NHSP	Newborn Hearing Screening Programme (United Kingdom)
NIDCD	National Institute on Deafness and Other Communication Disorders, National Institutes of Health
NIH	National Institutes of Health

Other Abbreviations

504	Section 504 of the Rehabilitation Act of 1973
AABR	automated auditory brainstem response
ABR	auditory brainstem response
ASSR	auditory steady-state response
BAHA	bone-anchored hearing aid
CMV	Cytomegalovirus
CROS	Contralateral routing of signals
CT	computerized tomography
Cx26	Connexin 26 protein

dB HL	decibels hearing level
dB nHL	decibels normal hearing level
EHDI	early hearing detection and intervention
EVA	enlarged vestibular aqueduct
HISS	Hearing in Schools Study
HL	hearing level
IDEA	Individuals with Disabilities Education Act of 2004 (IDEA 2004)
IEP	Individual Education Plan
LVAS	large vestibular aqueduct syndrome
MHL	mild bilateral hearing loss
MLU	mean length of utterance
MRI	magnetic resonance imaging
MSHL	minimal sensorineural hearing loss
NHANES-III	National Health and Nutrition Examination Survey-III (1988–1994)
NHS	newborn hearing screening
NHSP	newborn hearing screening program
NICU	neonatal intensive care unit
NITS	noise-induced threshold shift
OAE	otoacoustic emissions
PCP	primary care provider
PTA	pure tone average
S/N	signal-to-noise (ratio)
SIFTER	Screening Inventory for Targeting Educational Risk
SLP	speech-language pathology
UHL	unilateral hearing loss
UNHS	universal newborn hearing screening
VRA	visual reinforcement audiometry
WISC-R	Wechsler Intelligence Scale for Children, Revised
WRAT-R	Wide Range Achievement Test, Revised

Introduction

The primary goal of early hearing detection and intervention (EHDI) programs are to ensure that all newborns are screened for hearing loss before one month of age, all infants who screen positive have a diagnostic audiologic evaluation before 3 months of age, and all infants identified with hearing loss begin receiving early intervention services before 6 months of age. The Children's Health Act of 2000[2] established the EHDI program in the National Center on Birth Defects and Developmental Disabilities (NCBDDD) at the Centers for Disease Control and Prevention (CDC). The vision of the CDC EHDI program is to promote communication from birth for all children. By supporting the development of tracking and surveillance systems and other related activities in states and territories, CDC EHDI works to ensure that all children with hearing loss achieve communication and social skills commensurate with their cognitive abilities. Under its Congressional authority to develop standardized procedures for data management and program effectiveness, CDC EHDI identified differences among state and territory EHDI programs in the reporting of the identification of infants with mild and/or unilateral hearing loss.

Universal newborn hearing screening has demonstrated success in identifying moderate to profound degrees of hearing loss in children, yet a substantial body of research indicates that mild and unilateral hearing loss also have a detrimental effect on children's development. For example, children with mild and unilateral hearing loss score lower on tests of speech, language, school achievement, and behavior than do children without hearing loss.[3,4,5,6,7,8,9,10,11,12,13]

The possible under-identification and impact of mild and unilateral hearing loss prompted CDC EHDI, in collaboration with the Marion Downs Hearing Center, to convene the National Workshop on Mild and Unilateral Hearing Loss as a forum to discuss issues related to the identification, assessment, and intervention appropriate for infants and children with mild and unilateral hearing loss. The workshop brought together more than 50 national and international experts with the goal of reviewing and discussing information to facilitate development of short-term and long-term recommendations. Participants included researchers, clinical practitioners, early interventionists, representatives from parent and national organizations, and representatives from state and federal agencies.

This report summarizes the presentations, breakout group discussions, and research needs identified at the workshop. Breakout groups engaged in preliminary discussions of short-term and long-term recommendations; however, these recommendations were not finalized during the workshop. Recommendations/proposals are expected to be published in 2006 after further refinement by a committee with representation from national experts, a family support organization, and state and federal agencies.

Overview

CDC EHDI, in collaboration with the Marion Downs Hearing Center, convened the National Workshop on Mild and Unilateral Hearing Loss on July 26–27, 2005, in Breckenridge, Colorado. The objectives of the workshop were as follows:

- Review and evaluate scientific research and other data related to mild and unilateral hearing loss.
- Develop recommendations related to identification of mild and unilateral hearing loss.
- Develop recommendations related to appropriate intervention(s) for mild and unilateral hearing loss.
- Identify potential areas for future research related to mild and unilateral hearing loss.

To accomplish these objectives participants were asked to

- Review relevant research.
- Participate in one of four breakout group discussions about a particular aspect of mild and unilateral hearing loss.
- Participate in the development of recommendations.

A Steering Committee—with representation from CDC EHDI, the Marion Downs Hearing Center, the National Institute on Deafness and Other Communication Disorders (NIDCD) at the National Institutes of Health, and the National Early Childhood Technical Assistance Center (NECTAC), as well as two identified independent experts, one state EHDI Coordinator, and a parent representative—provided subject matter expertise to help develop the objectives of the workshop. This Steering Committee also developed the agenda for the workshop, set the roster of speakers, and identified key stakeholders and other individuals to be invited to the workshop. The meeting was announced in the *Federal Register* on May 26, 2005 (Volume 70, Number 101).[14]

The workshop began with plenary presentations addressing relevant topics, such as the scientific evidence related to the prevalence and impact of mild and unilateral hearing loss (see Appendix A for Workshop Agenda). These presentations established the context for the remainder of the workshop agenda. On the second day of the workshop, four facilitated breakout groups were formed to discuss the following areas: screening for hearing loss, diagnostic evaluation and follow-up, hearing technology, and early intervention. The objectives of these breakout groups were as follows:

- Identify issues and barriers.
- Identify areas for future research.
- Develop realistic, short-term recommendations.
- Develop long-term recommendations.

For the purposes of the workshop, the following working definitions were used:

A **permanent mild bilateral hearing loss** exists when the diagnosis indicates there is, in both ears, a calculated or predicted average pure tone air conduction threshold at 0.5, 1, 2 kHz between 20 and 40 decibels hearing level (dB HL) or pure tone air conduction thresholds greater than 25 dB HL* at two or more frequencies above 2 kHz (i.e. 3, 4, 6, 8 kHz).

A **permanent unilateral hearing loss** exists when the diagnosis indicates there is a calculated or predicted average pure tone air conduction threshold at 0.5, 1, 2 kHz of any level greater than or equal to 20 dB HL or pure tone air conduction thresholds greater than 25 dB HL at two or more frequencies above 2 kHz in the affected ear with an average pure tone air conduction threshold in the good ear less than or equal to 15 dB.

In both of these definitions, when an air-bone gap of greater than 10 dB exists at any of the two of these same frequencies, medical intervention has been ruled out.

These definitions were adapted from: Bess FH, Dodd-Murphy J, Parker RA. Children with minimal sensorineural hearing loss: prevalence, educational performance, and functional status. Ear and Hear. 1998;19(5)339–54.[5]

*American National Standards Institute. Specifications for Audiometers. ANSI S3.6-2004. New York: American National Standards Institute, Inc.; 2004.[15]

Presentations **(listed in order according to the workshop agenda)**

PREVALENCE AND SCREENING

Prevalence and Screening in Newborns

Judy Gravel, PhD

Newborn hearing screening (NHS) programs have provided us with information about the prevalence of permanent forms of hearing loss in the neonatal period. When the degree of hearing loss is defined as permanent bilateral moderate degree (40 decibels hearing level [dB HL]) and greater, existing studies suggest that prevalence rates are about 1.13/1,000 overall, and differ according to nursery (e.g., well-baby: 0.49/1,000; neonatal intensive care unit [NICU]: 4.8/1,000). The prevalence rate for unilateral hearing loss (of moderate degree and greater in the impaired ear) are estimated at about 0.83/1,000, and similar to bilateral hearing loss, the rate differs substantially according to nursery (e.g., well-baby: 0.41/1,000; NICU: 3.2/1,000).[16] Given that a hearing loss is of moderate degree or worse, and of either congenital or of neonatal onset, the vast majority of children with bilateral and unilateral hearing loss will be detected by NHS efforts.

However, infants with lesser (milder than 40 dB HL) degrees of permanent bilateral and unilateral hearing loss, and those with later onset or acquired hearing loss (of similar or greater degree) will not be identified through present NHS programs. Several barriers to early identification of milder forms of hearing loss exist. First, current screening tools (otoacoustic emissions [OAE] and auditory brainstem response [ABR] technologies) cannot reliably distinguish between normal hearing and mild hearing loss.[17] Second, neonatal screening test outcomes ("pass-refer" status) of infants with later confirmed mild sensorineural hearing loss are variable among technologies.[18] Finally, a commonly used two-step, two-technology neonatal hearing screening protocol (fail OAE; pass automated-ABR) misses as many as 80% of infants with later confirmed cases of mild bilateral and unilateral hearing loss.[19] Given these caveats, a *conservative* estimate of the prevalence of mild permanent bilateral and unilateral hearing loss in the neonatal population based on Johnson et al. 2005 data is 0.55/1,000.[20] Remarkably, this rate is about a hundred fold less than the prevalence of mild bilateral and unilateral forms of hearing loss in school-aged children reported at 54/1,000 by Bess and colleagues.[5]

Key Points:

1. *The numbers of children with mild forms of hearing loss are significantly under-identified and/or significantly under-reported in the United States at the present time*. Based on aggregate data reported by states to the Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA) and shared with CDC (2003—Version E Report and based on about 1.6 million infants screened) prevalence rates are estimated at 0.19/1,000 for unilateral hearing loss and 0.09/1,000 for bilateral permanent mild hearing loss.[21] Based on the Johnson et al. 2005 data, a conservative estimate is a rate at least twice as large.[19]

2. *Existing prevalence estimates of mild and unilateral forms of permanent hearing loss in the newborn period are highly variable and considerably lower than rates reported for children at school age*.[5,22]
3. *Current screening test protocols and pass-refer criteria will likely not identify the majority of infants with congenital and early-onset forms of permanent bilateral and unilateral mild hearing loss*.
4. *Early identification of mild forms of hearing loss in infancy and in the preschool period presents multiple technological, logistic, and practical challenges*. These include:
    - Lack of uniform standards for the calibration of OAE and ABR technologies.
    - Insufficient manufacturer supporting evidence to allow for the determination of the validity of specific pass-fail criteria and automated screening algorithms.
    - Potential variability in screening results as a function of earphone type (ABR technologies).
5. *Research and demonstration programs are lacking.* To develop critical tools, research data and demonstration programs are needed to a) determine the sensitivity and specificity of existing screening tools and test protocols for the identification of mild forms of permanent hearing loss; b) develop screening tools specifically designed to differentially identify mild permanent hearing loss from losses of a greater degree, and from temporary conductive loss; and c) establish efficient, effective, and practical hearing screening programs for the identification of mild forms of permanent hearing loss beyond the newborn period and before entry into elementary school.

Are Early Hearing Detection and Intervention Systems Missing Children with Minimal Hearing Loss?

June Holstrum, PhD, and Marcus Gaffney, MPH

CDC EHDI collaborated with the Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA) to develop a core set of data items for state early hearing detection and intervention (EHDI) programs. This presentation describes the data items, their characteristics, and their limitations. The aggregate data is collected annually through a voluntary response survey and is available online at the CDC EHDI website (www.cdc.gov/ncbddd/ehdi/dips.htm).

DSHPSHWA EHDI data in 2003 indicated that 1.08/1,000 infants had a hearing loss,[21] which contrasts with reported prevalence rates among infants ranging from 2.00/1,000 to 6.00/1,000 [23,24,25] and prevalence of hearing loss in school children ranging from 5% to 15%.[26,27,28,29] The discrepancies in prevalence rates may be the result of progressive or late onset hearing loss, infections and illnesses, trauma, noise, and system misses. System misses include technology used, screeners, emphasis on lowering the referral rates, lack of interest in minimal hearing loss, infants lost to the system, lack of follow-up of high-risk infants, and lack of screening during the preschool years.

A subset of the 2003 DSHPSHWA data was used to provide supporting evidence that babies with hearing loss may be missed by EHDI systems. The following issues were discussed:

- Babies with mild and unilateral hearing loss are being under-identified at birth.
- Programs need to determine the minimum hearing level that should consistently be used when screening infants.
- Loss to follow-up must be reduced.
- There is currently no systematic way to screen preschool children.
- Screening needs to be on going to identify infants missed at birth and those with progressive, late onset, and acquired hearing loss.
- Studies are needed to determine the prevalence of hearing loss in preschool-age children.

Prevalence of Unilateral and Mild Hearing Loss in School-Age Children
Fred H. Bess, PhD

The prevalence of mild and unilateral and hearing loss among school-age children is not well understood. Prevalence rates vary on the basis of the criteria used to define hearing loss and the method used to measure hearing (e.g., survey versus audiometry). It is believed that the milder the hearing loss, the greater the prevalence. To date, three large-scale studies have been conducted in an effort to determine prevalence.[5,27,30] Niskar and colleagues analyzed data from the Third National Health and Nutrition Examination Survey (NHANES-III) that was conducted in the United States by the National Center for Health Statistics from 1988–1994.[27] NHANES-III is a national, population-based, cross-sectional survey consisting of a household interview, laboratory, and physical examinations, and cognitive testing. The NHANES-III protocol for audiometry included pure tone air conduction tests (0.5, 1, 2, 3, 4, 6 kHz) to 6,166 children ranging in age from six to nineteen. In their analyses Niskar et al. defined hearing loss as an average cut-off of $\geq$16 dB for low-frequency hearing loss (0.5, 1, and 2 kHz) or high-frequency hearing loss (3, 4, and 6 kHz). They found that 7.1% of children had low-frequency hearing loss, 12.7% had high-frequency hearing loss, and in total, 14.9% had any hearing loss.[27] In another study using the same NHANES-III dataset, Niskar and colleagues reported on the prevalence of noise-induced threshold shift (NITS) in a sample of 5,249 children. In this study NITS was defined as 1) thresholds at 0.5 and 1 kHz of <15 dB; 2) maximum threshold values at 3, 4, or 6 kHz at least 15 dB poorer than the poorest threshold for 0.5 and 1 kHz; and 3) thresholds at 8 kHz had to be at least 10 dB better than the poorest threshold for 3, 4, and 6 kHz.[30] Finally, Vanderbilt University conducted a large education-based study to determine the prevalence of hearing loss in grades 3, 6, and 9. A total of 1,218 children served as the subject sample. Minimal sensorineural hearing loss (MSHL) was defined using three different hearing loss categories: bilateral sensorineural hearing loss (average air conduction thresholds between 20 and 40 dB in both ears), high-frequency sensorineural hearing loss (mean air conduction thresholds >25 dB at two or more frequencies above 2 kHz in one or both ears), and unilateral sensorineural hearing loss (mean air conduction thresholds >20 dB in the impaired ear). Air conduction thresholds ranged from 0.5 to 8 kHz; 3 and 6 kHz were also included. Bone conduction thresholds were also obtained if the subject met the criteria for minimal

sensorineural hearing loss. Tympanograms were used as supplemental evidence for identifying conductive or sensorineural loss.[5]

The following general findings can be drawn from these three studies:

- When all forms of hearing loss are considered, the prevalence is somewhere between 11% and 15%—that is, between 5 million and 7 million school-age children.[5,27]
- Prevalence of NITS in one or both ears is 12.5%.[30]
- At least 5.4% of school-age children exhibit minimal sensorineural hearing loss—about 1 in 20 or 2,484,000 children.[5]
- Unilateral sensorineural hearing loss is the most prevalent form of loss, affecting at least 3% or 1,380,000 school-age children; the data from Niskar and colleagues suggest that the prevalence of unilateral hearing loss may be even higher.[27]

Presentations

DIAGNOSIS, AMPLIFICATION, AND OUTCOMES

Issues Associated with Conducting Diagnostic Audiologic Evaluations in Children with Suspected Mild and Unilateral Hearing Loss

Barbara Cone-Wesson, PhD; Yvonne Sininger, PhD; Judith Widen, PhD

One issue associated with conducting diagnostic audiologic evaluations in children with suspected mild and unilateral hearing loss relates to the accuracy of the methods used to estimate hearing thresholds in infants and young children. Are the methods sufficiently accurate, and the variability sufficiently limited to differentiate mild hearing loss from normal hearing sensitivity? This question was applied to two types of evoked potential measures: ABR and auditory steady-state response (ASSR). Examples of ABRs to 0.5 and 4 kHz tone burst stimuli at 60, 40 and 20 decibels normal hearing level (dB nHL) showed clearly detectable wave V as low as 20 dB nHL. Data from Stapells, Gravel, and Martin comparing infant and adult ABR thresholds to tone burst stimuli at 0.5, 1.5, 4, and 8 kHz showed that the mean for infants was 5–10 dB higher (poorer) than for adults, with comparable variability around the mean for both adult and infant groups.[31] Stapells et al. showed high correlations (r=0.94 to 0.97) between infant tone-ABR thresholds in dB nHL with pure tone behavioral thresholds in dB HL. With respect to ASSR, findings from Rance and Rickards showed that the correlation between behavioral and ASSR thresholds is not as good for mild loss as for more severe hearing loss.[32] In addition, regression coefficients for young infants differ from those of older children and adults. ASSR thresholds are elevated with respect to behavioral thresholds by a greater amount (up to 15 dB) compared with data from older children and adults, suggesting some effects of neurodevelopment on ASSR threshold. With respect to behavioral pure tone thresholds, Norton et al. demonstrated that a well-controlled visual reinforcement audiometry (VRA) protocol could be used to obtain minimum response levels at 1, 2 and 4 kHz and for speech for each ear using insert earphones.[17] It was not possible to obtain all thresholds on every ear (in the time allotted), although reliable results were obtained for more than 95% of infants.

For both ABR and VRA, accuracy in estimating threshold will be related to step size and stopping level. To detect a mild loss, a smaller step size (e.g., 5 dB rather than 10) and a lower stopping level (e.g., 15 dB HL rather than 25 dB HL) may be needed. Tone burst ABR threshold estimation requires careful attention to the details of response filtering (e.g., a 20–30 millisecond response window will be needed to detect low-frequency tone burst responses at near-threshold levels in infants). Another issue relates to whether clinical settings have the time, equipment, and expertise required to achieve the precision of threshold estimation shown in research studies.

Separating out transient middle ear problems, such as otitis media with effusion, may be a larger issue in detecting mild loss than in more severe losses. The inaccuracy of bone-conduction testing may also be a larger issue with mild losses than with more severe.

Beyond sensitivity assessment are issues of speech discrimination and determination of uncomfortable listening levels, and a child's functional use of hearing may be important in determining appropriate habilitative measures for mild or unilateral loss.

Convincing parents that a "mild" or unilateral hearing loss is a problem seems to be especially difficult in newborns and very young infants whose responses differ very little or in subtle ways from babies without hearing loss.

A final consideration is how the diagnostic issues may differ as a function of age across newborns, infants, toddlers, and older children.

Audiological Management and Family Factors for Children with Mild and Unilateral Hearing Impairment

Kirsti Reeve, PhD

Before considering amplification for children with minimal hearing loss, one must recognize that a large number of children with this type of loss are unidentified and are not receiving any kind of audiological management. Many others are identified late, and any intervention and aid provision is delayed with the resulting impact on language development. Research has shown that fitting of aids soon after identification can minimize language delay;[33] however, there can be a delay of up to 7 months in provision of amplification for children with mild and unilateral hearing loss.[23,34] There is also a low compliance in aid use among children with unilateral hearing loss.[35]

A questionnaire survey of 56 heads of audiology services in the United Kingdom (UK) revealed that children with minimal hearing loss form a small part of their caseload: children with bilateral mild loss comprise 8% and children with unilateral loss comprise only 4%.[36] The modal age of referral was between 4 and 6 years for both groups of children, implying either that they were identified through the school entry hearing screen or that starting school revealed previously unidentified problems resulting in the child being sent for a hearing test. The most frequent management options for both groups of children were to give advice to the parents and place the child on review. Children with mild bilateral loss were significantly more likely to receive amplification than were children with unilateral loss, although the questionnaire also documented professional uncertainty regarding the appropriateness of amplification provision for bilateral losses between 25 and 40 dB HL—that is, the range that includes the majority of children with mild bilateral loss.

The parents of 60 children with minimal hearing loss in the Nottingham, Trent region of the UK were also surveyed on the impact of their child's minimal hearing loss on quality of life for both the child and the family. Of the parents whose children had been fitted with hearing aids, more than half (53%) found it very or quite difficult to get their child to use the aids after initial fitting. When asked about current aid use, only 44% of children with mild bilateral loss and 26% of children with unilateral loss were wearing

their aids all day. One-quarter of children with mild bilateral loss and half of the children with unilateral loss never wore their aids despite parents reporting benefit in speech perception, both in quiet and in noise, when the aids were used.

Key Points:
In the UK, there is professional uncertainty as to whether amplification is appropriate for children with minimal bilateral hearing loss. If amplification is provided, age of fitting is late, and this late fitting is not necessarily related to a late age at identification. There is a low compliance with amplification when provided under current management practices, despite parents reporting some benefit in speech perception with aids.

Issues Related to Amplification for Children with Mild or Unilateral Hearing Loss
Sarah McKay, MEd

For children with mild hearing loss, we must take into account diagnostic factors when choosing amplification. A key consideration is whether we have enough information from ABR and OAE test results to confidently fit a child with amplification. Can we accurately determine the degree, type, and configuration of hearing loss in infants? Do we have accurate immittance measurements and bone conduction results? Since we are dealing with a small variation between normal hearing and a mild hearing loss, these issues are critical.

Additionally, one must explore options for amplification based on age. The possible advantages of fitting a child with mild hearing loss prior to 6 months of age are that the benefits of early intervention are well documented,[37] and it follows the 1-3-6 guidelines.[38] We have to consider, however, if we have enough diagnostic information to confidently fit an infant with mild hearing loss. Waiting to fit until after a child is 6 months of age allows the audiologist to obtain potentially more precise audiometric information via VRA. Other factors to consider are that, before 6 months, the proximity of parent to infant allows for a greater signal-to-noise (S/N) ratio. It is after 6 months, when a baby is sitting and crawling, that the S/N ratio may become less optimal. A further consideration of waiting until after a baby is 6 months is that it allows time to evaluate middle ear status and possibly treat middle ear pathology. With mild hearing loss, it is clearly important to determine if a conductive component exists.

For children with unilateral hearing loss (UHL), diagnostic factors are also an issue, and, as with mild bilateral hearing loss, we must be able to determine the type, degree, and configuration of hearing loss. Since the advent of newborn hearing screening, children with UHL are now often being identified within the first 3 months of life. One important concern for children with UHL is auditory deprivation. Auditory deprivation has been described as more marked if it has an early onset (1–2 years), in individuals with UHL and in children with bilateral hearing loss who are fit amplification monaurally.[39] The questions remain: What degree of UHL causes auditory deprivation? And what is the "critical time" period in which we should be providing amplification to children with UHL? *Possible advantages of fitting UHL prior to 18 months of age are as follows:* the

potential for auditory deprivation may be minimized, the impaired ear can have the opportunity to benefit from amplification in case the hearing loss is progressive or if it becomes bilateral, and there may be an easier adjustment for the child. *Possible advantages of waiting until after 18 months are as follows:* more reliable and precise audiometric information can be obtained via VRA and conditioned play audiometry, early word recognition abilities can be assessed, speech and language skills can be monitored, and increased mobility at that age and the less optimal S/N ratio may necessitate amplification. Also important to consider are social/emotional and acceptance issues of early vs. later fitting, the various amplification options based on the degree of UHL, and whether fitting rationales and hearing aid characteristics should be different with UHL than with bilateral hearing loss.

Further research is needed in the following areas:

- Evidence of the number of children missed by the newborn hearing screening who actually have mild hearing loss.
- Evidence of the benefit of fitting children with mild and UHL with amplification.
- Evidence of what age is best to fit children with mild and UHL.
- Factors, genetic and otherwise, that predispose children with mild or UHL to a higher risk for progressive hearing loss.

FM for Minimal Hearing Loss

Sandra Abbott Gabbard, PhD, CCC-A

Children with mild or UHL have consistently demonstrated a risk for decreased speech recognition in noise.[40,41,42,43] The use of an FM system with a remote environmental microphone and a receiver at the child's ear will result in an improved S/N ratio and will therefore improve many children's ability to understand speech in noise.[44,45,46,47,48] The signal from a remote microphone can be sent directly into the ear of a listener by using FM system technology and a variety of transmitters and receivers. This direct transmission from a remote microphone is commonly used in classrooms to reduce the interference between a child and the teacher;[49] however, the use of this technology on younger children has not been well studied. At the University of Colorado Hospital, we have had a loaner FM program for preschool children for many years. Parents of children with all degrees of hearing loss report successful use of this technology in noisy environments.

Minimal Hearing Loss and Cognitive Performance in Children: Brief Update

Danielle S. Ross, PhD, MSc; Susanna Visser, MS; June Holstrum, PhD; Aileen Kenneson, PhD, MS

This presentation summarizes preliminary analyses conducted at the National Center on Birth Defects and Developmental Disabilities at the CDC. There are two goals of these analyses. The first goal is to estimate the prevalence of unilateral and slight–mild

bilateral hearing loss among a nationally representative sample of school-age children in the United States. The second goal is to evaluate the relationship between unilateral and slight–mild bilateral hearing loss and performance on standardized intelligence and achievement tests in this population of children.

The data for this study are from the Third National Health and Nutrition Examination Survey (NHANES-III)[50] which was conducted in the United States by CDC's National Center for Health Statistics from 1988 to 1994. NHANES-III is a national, population-based, cross-sectional survey consisting of a household interview, laboratory testing, physical examinations, and cognitive testing.

The case definitions for unilateral and slight–mild bilateral hearing loss used in these preliminary analyses were based on pure tone averages (PTA) of 0.5, 1, 2, and 4K Hz. "No hearing loss" was defined as <15 dB HL in both ears; "slight" hearing loss as 15–25 dB HL; "mild" as 26–30 dB HL; and "moderate plus" as 31+ dB HL. Unilateral hearing loss was defined as <15 dB HL in the better ear and $\geq$15 dB HL in the worse ear. Slight-mild bilateral hearing loss was defined as 15–30 dB HL in the better ear. Data from a total of 5,305 children were included in these analyses (normal hearing: N = 4,922; slight–mild: N = 140; unilateral: N = 243). The majority of children in the hearing loss groups had slight losses (15–25 dB HL).

For the cognitive testing, NHANES-III includes two nonverbal subscales (Digit Span and Block Design) of the Wechsler Intelligence Scale for Children–Revised (WISC-R) and two subscales (reading and arithmetic) of the Wide Range Achievement Test–Revised (WRAT-R). The testers were not aware of the status of any of the children's hearing.[51]

Results show that there are approximately 6.4% of children (2.7 million) in the United States age 6–16 years with unilateral or slight–mild bilateral hearing loss (based on weighted proportions). Children aged 6–16 years with unilateral or slight–mild bilateral hearing loss are at least twice as likely to score two standard deviations below the norm on standardized arithmetic and reading tests. Children aged 6–16 years with unilateral or slight–mild bilateral hearing loss may also be at higher risk for scoring one standard deviation below the norm on the block design subtest of the WISC-III.

It is possible that some children in this sample have underlying deficits related to etiology. These analyses are in progress. Defining a profile of children with unilateral or slight–mild bilateral hearing loss who are at higher risk for failure on standardized tests than their peers with normal hearing is an important focus for future research.

The Hearing in Schools Study: Prevalence, Impact, and Genetics of Slight/Mild Sensorineural Hearing Loss in Elementary School Children

Barbara Cone-Wesson, PhD

The Hearing in Schools Study (HISS) was funded by the National Institute on Deafness and other Communication Disorders (NIDCD) within the National Institutes of Health

(NIH) by a grant (R01 DC 005662-03) to principal investigator, Dr. Melissa Wake, Director (Research and Public Health), Centre for Community Child Health at the University of Melbourne and Murdoch Children's Research Institute in Victoria, Australia.

The specific aims of the study were to 1) ascertain the prevalence of slight/mild sensorineural hearing loss in elementary school children; 2) evaluate the language, academic performance, social, and health-related quality of life outcomes in children with slight/mild sensorineural hearing loss; 3) describe the phenotype-genotype for slight/mild sensorineural hearing loss; 4) develop and trial informed consent procedures for DNA specimens; 5) describe the impact of genetic testing on children and families; and 6) develop education and counselling approaches to facilitate comprehension of genetic information.

Phase I of the study comprised a cross-sectional cluster survey of children in Melbourne, Australia (population 3.4 million). The target population was 7,784 first- and fifth-year school children in 89 schools. The survey had an 85% response rate. The parent questionnaire contained questions about risk factors for congenital hearing loss, current concerns about the child's hearing, family history of hearing impairment, any history of environmental exposure to loud noise, and any difficulties parents had with hearing. The questionnaire also contained age-specific questions about health-related quality of life and child behavior.

The entire cohort of children had otoscopy and pure tone screening audiometry (3,367 grade 1 students, mean age = 7.2 years; and 3,214 grade 5 students, mean age = 11.1 years). The screening test consisted of three presentations of pure tones at 15 dB HL at test frequencies of 0.5, 1, 2, 3, 4, 6, and 8 kHz. To pass the screen, the child had to respond correctly to two-thirds of presentations at each frequency. If one frequency was failed, a full audiometric evaluation took place, including determination of pure tone air and bone conduction thresholds, tympanometry, and acoustic reflex threshold tests. All audiometric evaluations were conducted in a sound-treated audiometric test booth by certified audiologists.

A low and high frequency pure tone average (PTA) was calculated using thresholds at 0.5, 1, and 2 kHz for the low frequency PTA and 3, 4, and 6 kHz for the high frequency PTA. Hearing loss was classified as normal (thresholds of ≤15 dB HL), slight (16–25 dB HL), mild (26–40 dB HL), and moderate or greater (>40 dB HL). Fifty-five children were identified with slight/mild bilateral sensorineural hearing loss for a prevalence of 0.88%; 40 were classified with a slight loss; and 15 had a mild loss. Eighteen children were in year 1, and 37 were in year 5.

Phase II of the study ascertained language, academic, social, and quality of life outcomes in 48 case children with slight/mild hearing loss and 96 control children with normal hearing, matched to case children on sex, school, age, and year level. Children with slight/mild sensorineural hearing loss had similar outcomes to children with normal hearing on measures of language, reading, behavior, and parent- and child-reported

health-related quality of life questionnaires. Children with slight/mild sensorineural hearing loss revealed poorer outcomes on a test of phonological short-term memory (e.g., nonsense word repetition task). The child's self-reported hearing-related problems on the emotional scale were significantly correlated to severity of hearing impairment.

It is known that mutations in the *GJB2* gene, which encodes the gap junction protein Connexin 26 (Cx26), are commonly found in people with a genetic hearing loss. A change in the gene resulting in the protein coding, labeled as V37I, can result in mild hearing loss. Four of 48 case children were found to have the V37I change in both genes, and two more cases had the V37I change in one gene. No controls were found to have the V37I variation.

A small pilot study regarding concerns about genetic testing for hearing loss revealed that parents were unfamiliar with genetic testing. Further, the focus of parental concern regarding genetic testing was on the immediate effects on the child (e.g., the potential for pain, distress to the child, and classroom disruption). The main issues identified were storage of and access to tissue and test results.

Key Points:
The prevalence of slight/mild bilateral sensorineural hearing loss among school-aged children in this study was 0.88%, and despite poorer phonological skills in children with slight/mild sensorineural hearing loss, language, reading, behavior, and health-related quality of life scores were similar to their peers with normal hearing. Four of 48 cases (8.3%) were homozygous for Cx26 mutations. Qualitative findings regarding the impact of genetic testing indicated that genetic testing and research were not inherently anxiety-provoking.

Academic, Social, and Behavioral Outcomes in Children with Minimal Hearing Loss
Anne Marie Tharpe, PhD

In 1998, Bess and colleagues found that 5.4% of school-age children whom they examined had minimal sensorineural hearing loss (MSHL), including unilateral loss. Of those children, about 37% had failed a grade in school and an additional 8% were judged not to be performing at grade level. This reflects a failure rate approximately 10 times that of the general school population in the Middle Tennessee region where these children were sampled. These academic data are remarkably similar to those obtained a decade earlier from a sample of school-age children with unilateral hearing loss.[52] Other investigators have also found high rates of academic problems in this population.[7,53,54,55]

Closer examination of this group of children with MSHL found that they exhibited greater dysfunction than their normal-hearing counterparts on domains of stress, self-esteem, behavior, energy, and social support. Furthermore, the children with MSHL consistently performed more poorly on the Screening Instrument for Targeting Educational Risk (SIFTER) than their peers with normal hearing.[56] The SIFTER is a

teacher questionnaire designed to provide a valid method by which children with hearing problems can be screened educationally.

Largely because of the findings of decreased energy in the children with MSHL in the Bess et al. study,[5] Bourland-Hicks and Tharpe[57] examined school-age children with mild and high-frequency hearing losses and their age- and grade-matched peers on a listening effort task. This study used a dual-task paradigm in which the children were instructed to perform a speech perception task in varying levels of background noise (e.g., the primary task). At the same time, they were asked to press a button when a light appeared on the table directly in front of them (e.g., the secondary task). The reaction time to the button press was the dependent variable with the theory being that, as more effort is expended on the listening task, more time will be required to react to and press the button for the secondary task. This method was used because it is suggestive of the multiple tasks that a child might be asked to perform while in a classroom. That is, children are asked to listen, take notes, and process information all at the same time. The children with hearing loss in this study had significantly longer reaction times on the secondary task than their peers with normal hearing, suggesting that they were exerting more listening effort.

Key Points: There is evidence to suggest that many children with minimal to mild hearing loss, including those with unilateral hearing loss, experience 1) reduced speech recognition ability under adverse listening conditions; 2) academic difficulties including poor performance on basic skills tests, low teacher ratings of communication and attention, grade failure, and need for resource assistance; 3) social emotional dysfunction including low self-esteem, low energy, high stress, and need for social support; and 4) increased listening effort.

Outcomes in Children with Mild and Unilateral Hearing Loss
Christie Yoshinaga-Itano, PhD, CCC-A, CED

Johnson reported that 0.16 in every 1,000 Colorado school-age children have a hearing loss that is not judged to be educationally significant, while 0.66 in every 1,000 school-age children in the state are identified as having an educationally significant hearing loss (Johnson C. Colorado statistics on children with hearing loss. Unpublished report. 2005.) Of these children, 51.4% have bilateral hearing loss,* 32.4% have unilateral hearing loss, 8.3% have a bilateral high-frequency hearing loss, and 8.0% have hearing loss caused by otitis media. If both educationally significant and non-educationally significant hearing loss are combined, there is an incidence of hearing loss among Colorado school-age children of 2.2%. In 2002–2003, 57.0% of the children with bilateral hearing loss had an individual education plan (IEP), 26.0% of the children with unilateral hearing loss had an IEP, 21.0% of the children with bilateral high-frequency hearing loss had an IEP, and 26.0% of children with otitis media and hearing loss had an IEP.

In 2002–2003, Johnson[58] also conducted a survey of 135 children from Colorado and 20 children from Michigan who had unilateral hearing loss. The survey gathered information about each child's grade, pure tone average, type of hearing loss (conductive,

sensorineural, other), type of services (IEP, covered under Section 504 of the Rehabilitation Act of 1973[59], other), primary disability, use and type of amplification, and SIFTER results. Data were available from children with unilateral hearing loss from preschool through high school. More than half (56.7%) had hearing loss in the right ear, while 43.2% had hearing loss in the left ear. The pure tone average ranged from 30 to 120 dB HL, with a mean of 71.8 dB HL.

SIFTER results for these children with unilateral hearing loss were as follows: In the area of academics, 71.0% had no service, 50.0% were covered under 504, and 31.0% had an IEP. In the area of attention, 69.0% of the children with no services passed, 70.0% covered under 504 passed, and 55.0% with IEPs passed. In the area of communication, 60.0% with no service passed, 40.0% covered under 504 passed, and 28.0% of the children with an IEP passed. In the area of classroom participation, 81.0% with no services passed, 100% of the children covered under 504 passed, and 55.0% of the children with an IEP passed. In the area of behavior, 94.0% of the children with no services passed, 90.0% covered under 504 passed, and 76.0% of the children with an IEP passed. *Children with an IEP and unilateral hearing loss in the right ear had the following results:* 7.3% failed behavior, 19.5% failed classroom participation, 39.0% failed communication, 24.4% failed attention, and 39.0% failed academics. *Children with an IEP and unilateral hearing loss in the left ear had the following results:* 12.1% failed behavior, 19.5% failed classroom participation, 45.5% failed communication, 24.2% failed attention, and 42.4% failed academics.

Key Points: Overall, data indicate that the greater the need, the more intense the type of service. Significant numbers of students who are <u>not</u> receiving services demonstrated difficulty on the SIFTER. SIFTER results did not vary significantly for children with unilateral hearing loss in the right ear versus those with unilateral hearing loss in the left ear. Also notable is that, unless a child is in special education or covered under 504, a school often does not know that the child has a hearing loss. Case managers or teachers of the deaf and hard of hearing are not always available to complete assessments, unless the child is in special education. In addition, general education teachers often are not motivated to complete SIFTERs because they do not recognize the problem or need.

Children with mild hearing loss have better receptive and expressive language, expressive vocabulary, personal-social development, emotional availability, speech intelligibility, more vowels and more consonants when they are identified early than when they are identified later. Children with mild hearing loss have significantly poorer language development than children with normal hearing, even when early-identified. The language development of early-identified children with mild hearing loss is similar to that of children with all other degrees of hearing loss (moderate, severe, and profound).

*This study looked at school-aged children with mild hearing loss and used the definition of hearing loss as stated in the Colorado Exceptional Children's Act , 2002, 2.02(3): "…A deficiency in hearing sensitivity shall be one of the following:

1. A three-frequency pure tone average hearing loss in the speech frequency of ≥ 20 dB HL in the better ear which is not reversible in a reasonable amount of time.
2. A high-frequency pure tone average hearing loss of ≥ 35 dB HL in the better ear for two or more of the following frequencies: 2, 3, 4 or 6 kHz.

3. A three-frequency pure tone average unilateral hearing loss of ≥ 35 dB HL which is not reversible within a reasonable period of time."[60]

Language Outcomes in Young Children with Unilateral Hearing Loss
Allison Sedey, PhD, CCC-SLP-A; Arlene Stredler-Brown, MA, CCC-SLP, CED; Karen Carpenter, MA

This presentation summarizes results of a study examining demographic characteristics and language outcomes of 26 children in Colorado with unilateral hearing loss. All but 2 of the children were screened for hearing loss at birth, and all but 2 were confirmed to have unilateral loss prior to 6 months of age. The sample of children was essentially equally divided in terms of both gender and ear impaired (right vs. left). All degrees of hearing loss were represented approximately equally except for mild hearing loss, which was present in only 8% of the children. Four of the children had disabilities in addition to their hearing loss, and thus, were not included in the analyses of language outcomes.

Language data were gathered from a videotaped parent-child interaction and two parent-report instruments, the Minnesota Child Development Inventory and the MacArthur Communicative Development Inventories. The majority of the children were assessed on at least two occasions that were 6 months or more apart, with many children receiving three or four assessments over a 2-year period. At the time of testing, the children ranged in age from 12 months to 5 years.

For each of the language measures, the percentage of children falling well below age expectations was determined. On the two parent-report instruments, 17% to 23% of the children (depending on the measure examined) fell below the 10^{th} percentile compared with normative data for hearing children of the same age. Based on the spontaneous language sample obtained during the parent-child interaction, 33% of the children had a mean length of utterance (MLU) that was below age expectations.

Language data were examined across time and measures for the 15 children who participated in the assessment on multiple occasions. A consistent pattern of language delay was demonstrated by 27% of the children, with an additional 7% presenting with a borderline delay. The demographic characteristics of the children with language delay were, in many respects, unremarkable in that they were similar to the other children in the sample. Specifically, all of the children with language delay were Caucasian, were identified by 2 months of age, had hearing loss that was congenital and of unknown etiology, and were from families with relatively high incomes and high levels of education. Half of the children had hearing loss in the right ear; the other half were impaired in the left ear. The one characteristic that did appear to set these children apart from the majority of the group was their degree of hearing loss. Although not all of the children with severe to profound hearing loss demonstrated delayed language, all of the children with language delays had a severe to profound loss in their impaired ear.

Differences in Outcomes for Right Ear vs. Left Ear Unilateral Hearing Losses
Marilyn Neault, PhD, CCC-A

The needs of children with unilateral hearing loss command less attention, awareness, and advocacy than the needs of children with bilateral hearing loss. Children with unilateral loss are not eligible for early intervention in some states. They are less likely than their peers with bilateral loss to know other children or adults with hearing loss similar to their own. There are few parent workshops or mentoring activities designed for their needs. They are less likely to undergo a medical and radiological workup for the cause of the hearing loss.

Determining whether children with right ear vs. left ear hearing loss are more at risk for negative outcomes may help in defining a target subpopulation for more intense intervention. Anatomical and physiological substrates should encourage pursuit of this question. Above the level of the cochlear nucleus, crossed pathways (bringing activity originating from each ear to the opposite side of the brain) predominate over uncrossed pathways. Contralateral dominance for cortical auditory processing,[61] coupled with the larger left planum temporale region of the posterior temporal lobe[62] yields a correspondence between asymmetries in anatomy and speech perception.[63] Schmithorst et al.[64] found evidence for functional reorganization dependent on the side of the hearing loss. Functional MRI (fMRI) showed that children with left ear loss had more activation in the right superior temporal gyrus and children with right ear loss had more activation in the left inferior frontal area, when children ages 7–12 years with right vs. left ear hearing loss listened to random tones. Unless early brain plasticity allows for adequate compensation in auditory neural pathway development, children with right ear hearing loss warrant examination to determine whether they are more at risk for difficulties in speech perception than children with left ear hearing loss.

Few studies of unilateral hearing loss outcomes have examined right vs. left ear differences. A trend toward poorer outcome in children with right ear loss was noted by Bess and Tharpe.[42] Oyler et al.[12] found right ear loss more common in those who repeated a grade or received special services. Hartvig Jensen et al. [55,65] studied 30 children age 10–16 years with unilateral hearing loss. Children with right ear loss had significantly poorer ability than children with left ear loss to hear interrupted speech in background noise. Children with right ear loss also scored poorer on intelligence scale subtests (similarities and digit span) that are sensitive to subtle differences in the processing of input. Survey of academic progress showed that 95% of children with left ear loss and 45.5% of those with right ear loss were making satisfactory academic progress, with the remainder requiring resource help.

Key Points:

- Anatomical and physiological asymmetry for speech perception in the brain provides a theoretical framework for risk of poorer outcomes in children with right ear hearing loss than in children with left ear hearing loss.

- Only limited published data exist to support the theory that children with right ear hearing loss are at greater risk for negative outcomes than those with left ear loss.
- Researchers with existing or prospective databases of unilateral loss outcomes should be encouraged to analyze their data to determine whether outcomes vary as a function of right vs. left ear hearing loss. Such data would help to determine whether children with right ear hearing loss should receive more aggressive intervention, such as more intensive early language therapy and/or amplification.

Progression from Unilateral to Bilateral Loss
Marilyn Neault, PhD, CCC-A

Because newborns with a bilateral "refer" result are more likely to have permanent hearing loss that unilateral refers, and because bilateral loss impacts language development more severely than unilateral loss, follow-up for infants with unilateral "refer" results on newborn screening tests often is not as aggressive as for bilateral referrals. However, children referred for unilateral loss may have an outcome of bilateral loss.

Two groups of newborns move from unilateral refer to bilateral hearing loss status:

1. Those who actually had bilateral hearing loss at the time of the screening.
    a. Mild hearing loss in the ear that passed the screening because the screening stimulus level may have exceeded the intended sound pressure level in the infant's ear.
    b. Hearing loss mild enough to pass at the intended screening intensity in the better hearing ear.
2. Those who had unilateral loss at the time of the screening but develop bilateral hearing loss later.
    a. Middle ear fluid in one ear at the time of the screening, but both ears affected by the time of the diagnostic evaluation.
    b. Unilateral permanent loss at the time of the screening, with development of bilateral permanent loss because of the underlying etiology.

Massachusetts newborn hearing screening data for 2003 and 2004 were analyzed to determine how many unilateral referrals showed bilateral hearing loss on diagnostic audiological evaluation. In 2003 and 2004 combined, 1,887 newborns were referred out of 158,523 screened, resulting in a refer rate of 1.2%. Of the 1,887 refers, 1,455 (77%) were unilateral and 432 (23%) were bilateral. Of the 1,455 unilateral refers, 101 (7%) had unilateral hearing loss, and 58 (4%) had bilateral hearing loss. Of the 159 unilateral refers who had hearing loss, 64% had unilateral hearing loss, and 36% had bilateral hearing loss. Thus, more than one-third of the unilateral refers who were found to have hearing loss on diagnostic audiological evaluation showed bilateral rather than unilateral loss. In fact, although the chance of having any hearing loss was only 11% for a unilateral refer vs. 39% for a bilateral refer, 28% of children who were found to have bilateral loss came to the diagnostic audiological evaluation as a unilateral referral (referenced from personal communication with Farrell J, Stone S, Lui CL; 2005).

Causes for true progression from unilateral to bilateral hearing loss in early infancy include hereditary progressive sensorineural hearing loss, cytomegalovirus (CMV), enlarged vestibular aqueduct, and other unknown etiologies. Barbi et al.[66] showed that children who passed the newborn hearing screen but were later diagnosed with sensorineural hearing loss had higher than the expected incidence of asymptomatic CMV in stored samples of neonatal blood. Licameli et al. (referenced from personal communication with Licameli G, Robson C, Kenna M; 2001) found that of 18 children with unilateral sensorineural hearing loss who underwent CT scans of the temporal bones, 8 (45%) had abnormal findings, and 5 of those 8 children had bilaterally abnormal cochlear anatomy in spite of having one normally hearing ear. The CT scan findings included bilaterally enlarged vestibular aqueduct and Mondini deformity, both associated with progressive hearing loss.

Key Points:

- Newborns who refer in one ear on the newborn hearing screening test are at risk for bilateral hearing loss, because of screening test variables or progression in hearing loss.
- Children with unilateral hearing loss need close monitoring of the better hearing ear to check for progressive hearing loss.

Presentations

EARLY INTERVENTION, ELIGIBILITY, AND CLINICAL PRACTICE

Current Federal Initiatives

Sharon Ringwalt, PhD, CCC-SLP

Reauthorization of IDEA[67] in 2004: On December 3, 2004, President Bush signed the reauthorized Individuals with Disabilities Education Act of 2004 (IDEA 2004)[68] into law (http://www.nectac.org/idea/idea2004.asp). Most provisions in the new law took effect on July 1, 2005. The proposed regulations for Part B of IDEA were released, for public comment, on June 10, 2005 (http://www.wrightslaw.com/idea/law/idea.regs.propose.pdf).[69] As of July 27, 2005, the Part C regulations have not yet been released.

The following implications for audiology have been identified by professional organizations: 1) direction was provided on the need to provide information about "the full range of options to families of deaf and hard of hearing children;" 2) "interpreting services" were identified as a related service; 3) "teachers of the deaf" were classified as special educators; 4) states were given flexibility to define personnel qualifications; 5) regional and State centers for children, such as Schools for the Deaf, were retained as sites for service, when this is considered the least restrictive environment for an individual child; and 6) both "cued language services" and "sign language [services]" were established as early intervention services, for children from birth to 3 years of age.

Reauthorization of Head Start Funding—Recent History and Current Status:

- On July 25, 2003 the House of Representatives passed H.R. 2210, "The School Readiness Act of 2003,"[70] a bill to reauthorize The Head Start Act. This bill did not have bipartisan support.
- The Senate Health Education Labor and Pensions Committee passed a Head Start reauthorization bill in October 2003 with bipartisan support. It has not seen action on the floor of the Senate. The 2003 Senate Committee bill is substantially different from the House-passed bill, H.R. 2210.
- Since then, Head Start has been extended with yearly appropriations.
- House bill H.R. 2123 (the Head Start School Readiness Act of 2005[71]) was approved by the House Committee on May 18th by a vote of 48-0.
- Senate bill S. 1107 (Head Start Improvements for School Readiness Act[72]) was ordered to be reported without amendment on May 25, 2005. The Senate Health Education Labor and Pensions Committee passed this bill in June 2005 by voice vote.
- The goals of the House bill, H.R. 2123, include closing the school readiness gap and strengthening the academic content of the program, addressing weaknesses in Head Start financial controls and improving accountability, strengthening early childhood services for disadvantaged children, increasing requirements for teachers in Head Start, and improving coordination among early education programs.
- The Senate bill, S. 1107, is similar to H.R. 2123.

- A vote by the full House of Representatives is expected to occur during Fall 2005. Similarly, the Senate bill is awaiting floor action.

For more information, please e-mail nectac@unc.edu or visit the NECTAC website at www.nectac.org[73]

Part C Eligibility for Infants and Toddlers with Hearing Loss
Karl White, PhD, and Jamie Elliott, MPA

Part C Early Intervention services are an integral part of early hearing detection and intervention systems, and eligibility for Part C services varies dramatically from one state to another. In 2002, the National Center for Hearing Assessment and Management (NCHAM) gathered information to analyze state and territory Part C eligibility for infants and toddlers with hearing loss.[74] NCHAM obtained Part C eligibility requirements as outlined in state plans or other official state documents submitted to the National Early Childhood Technical Assistance Center (NECTAC). In addition, NCHAM directly contacted some states and territories directly to clarify their eligibility requirements. An analysis of this information revealed that children with established risk conditions were eligible for Part C-funded services in all 50 states, Washington D.C., and the five surveyed U.S. territories*. Forty-three states, Washington D.C., and the 5 surveyed territories listed specific conditions for eligibility. Thirty-two states, Washington D.C., and 2 territories listed hearing loss as a specific risk condition. Four states, Washington D.C., and 1 territory (Puerto Rico) operationally defined hearing loss in their state plan. Thirteen states unofficially offered a definition of hearing loss. Thirty-three states and 4 territories did not report a definition of hearing loss in their state plan or other official documents in 2002.

*The five territories referenced are American Samoa, Guam, Northern Marianna Islands, Puerto Rico, and the Virgin Islands

UK Trial of Early Amplification in Children with Unilateral or Mild Hearing Impairments
Adrian Davis, PhD; Shirley DeVoe, MSc; Helen Robertson, BM MRCPCH MSc

The Medical Research Center's Hearing and Communication Group and Manchester University have worked for many years looking at the epidemiology of hearing in the adult and pediatric populations of the UK and its implications for public health. The National Health Service in England continues to be recognized on an international level for their exciting and challenging comprehensive Newborn Hearing Screening Programme (NHSP). Currently 75% of all newborns are tested through this program, and the goal is to screen all newborns in 2006.

The NHSP aims to identify children with permanent moderate to profound bilateral hearing impairment. The research study aims to provide the evidence base for the relevance of early identification of children with unilateral or mild–moderate bilateral

permanent hearing impairment. There are two arms to the study, each having a different design but running simultaneously.

The NHSP is identifying 0.7 children per l,000 with unilateral hearing loss. It is known that childhood unilateral loss is associated with a number of subtle perceptual and educational deficits. Through a series of focus groups with professionals and parents, there was clinical equipoise concerning whether these children should receive early intervention through amplification. The project design is a randomized control trial of age of amplification for children with permanent congenital unilateral hearing impairment (40–85 dB HL in the poorer hearing ear). Children will be randomized into one of three groups: amplification introduced at around 6 months of age, amplification introduced at around 17–19 months of age, and no amplification (all children receiving at least current best practice). The project will provide an assessment of the efficacy, appropriateness, and effectiveness of amplification strategies for children with early-identified congenital permanent unilateral hearing impairment.

The NHSP is identifying significant numbers of children with bilateral mild to moderate hearing loss. Little evidence exists to guide parents and clinicians in the very early management of these children (e.g., is amplification acceptable, beneficial, and cost effective if introduced at a very early age?). Studies show that these children are at risk of developing deficits in language, social, and communication skills, which can affect their education attainment. Ninety consecutively identified children with mild–moderate hearing loss will be enrolled in this study, at least 45 of whom are/were aided in the first year of life. One hundred twenty normal-hearing children will be recruited to match the "early aided" children.

Language, social, and communication development will be assessed using age-standardized tests where possible and long-term outcomes compared across intervention groups and normal-hearing children.[75,76,77,78,79,80,81,82,83,84]

Clinical Implications of Children with Minimal Hearing Loss
Arlene Stredler-Brown, MA, CCC-SLP, CED

Research supporting current trends: Based on the literature, there is strong evidence that many school-age children with minimal hearing loss are at an elevated risk for developmental delays.[3, 12,52,54] There is very little evidence, however, to indicate whether this delay begins during infancy and/or preschool and whether early intervention is effective in this population. Based on recent findings of clinical data in Colorado, the proportion of infants and toddlers with minimal hearing loss experiencing developmental delays appears to be comparable to that of school-age children. In response to this finding, Colorado's early intervention program for children with hearing loss has consistently identified and offered treatment to children with minimal hearing loss. Generally, children with mild, bilateral hearing loss are considered categorically eligible for services. Children with unilateral hearing loss are considered "at risk" for delays and are monitored regularly. When a developmental delay is diagnosed, treatment is offered.

Funding for treatment comes from a variety of sources, including the state's EHDI program, local Part C agencies, and the state school for the deaf.

Colorado's programs are outcome-based and driven by data.[85,86,87] An effort has been made to objectively evaluate, identify, and treat children with minimal hearing loss statewide. It has been effective to provide education to change current practices, which often see young children with minimal hearing loss as having a condition that is of negligible significance.

Impact of minimal hearing loss on a child's development: A minimal hearing loss may have implications for a child's ability to listen to and understand auditory information, a child's speech and language development, and/or a child's behavior. [12,88,89] While auditory information may be audible, even a minimal hearing loss can make sounds unintelligible.[53,88] Distance hearing is also affected by a minimal hearing loss, and this loss of distance hearing can impact incidental learning. Issues related to the development of speech and language include the ability to hear word-sound distinctions that form important morphological markers and difficulty with word recognition and spelling.[88] Additionally, as a result of the amount of effort they expend in all listening situations, children with hearing loss may be more fatigued than their normal-hearing peers.[5]

Treatment considerations: An individualized assessment of each child's skills includes audiological assessment and assessment of performance in all developmental domains. Audiological assessment also monitors for later-acquired loss, fluctuating thresholds due to middle ear fluid, and possible progression. For children with a delay in communication and/or language, treatment addresses the development of functional auditory skills, speech and language skills, and adjustments to the environment. Families of infants and toddlers benefit from up-to-date medical counseling and parent-to-parent support.[90,91]

Breakout Group Discussions

After the plenary presentations, workshop participants separated into four breakout groups with the charge of identifying issues, barriers, and areas for future research, and developing both short-term and long-term recommendations. Suggested group assignments had been determined by the Steering Committee prior to the workshop, and a facilitator had been recruited for each group. The groups convened for more than 5 hours on the second day of the workshop, after which each group facilitator reported the identified issues and barriers, areas of future research, and discussion of recommendations to the entire workshop.

Synopses of each breakout group discussion, highlighting key issues addressed and research needs identified, are provided here. Research needs have also been compiled in Appendix C.

NOTE: While breakout groups were asked to discuss recommendations related to mild and unilateral hearing loss, these recommendations have not been finalized. Recommendations/proposals are expected to be published in 2006 after further refinement by a committee with representation from national experts, a family support organization, and state and federal agencies.

Breakout Discussion – Screening for Hearing Loss

Facilitator: Judy Gravel, PhD

The discussion of screening for mild bilateral hearing loss (MHL) and UHL focused on the newborn period through early childhood. The group differentiated between three pediatric categories: newborn, preschool, and school-age children. A summary of the discussion specific to each of these pediatric categories is included below.

The group discussed the main concerns about differentiating minimal forms of hearing loss from normal hearing in infants and children. The group felt that screening programs for mild hearing loss may be compromised when definitions for normal hearing have not been established. There are few empirical data that can be used as the basis for delineating normal hearing across the frequency range and for various signal delivery methods (insert earphones and sound field) and test procedures (electrophysiologic) that could be applied to the screening of infants, preschool, and school-age children. Although this question could be examined through meta-analyses, there are likely insufficient published data are likely insufficient at present to be useful for completing such an analysis for newborn and preschool-age children. Finally, there was discussion of whether definitions of MHL and UHL should include subcategories such as "slight," "minimal," and "mild." Such terms may underemphasize the effects of lesser degrees of hearing loss on language development, academic achievement, and behavior.

Prevalence rates for MHL and UHL are currently not clear in the three pediatric age groups of interest. Among other issues, the group felt it would be helpful to determine a reasonable upper limit on the number of screening tests before referral for confirmatory audiologic testing. Repeated screenings until an infant finally passes a hearing screen could contribute to the number of infants with MHL being missed in the newborn period. There are no published data demonstrating the impact of MHL and UHL in children younger than school-age. The poor acoustic conditions of most daycare and educational environments and the paucity of educational audiologists nationwide were also recognized. Screening models that include follow-up for maximizing our ability to detect, assess, and manage children with MHL and UHL across the age range (birth to school-age) may help to address some of these issues. These models could include innovative language and functional screening tools in addition to audiometric hearing screening. The group acknowledged that significant cost-shifting would be necessary to develop innovative screening programs (as described below) from those currently existing.

The need for educating parents and professionals about MHL and UHL was discussed. It was suggested that medical schools could provide more training about pediatric hearing loss, including MHL and UHL. The group also felt that increasing public awareness about UHL and MHL, including information about risk indicators and developmental milestones for hearing, speech, and language, could be beneficial. Suggested communication channels included parent magazines, television, the Internet, American Association of Retired Persons, milk cartons, diaper and baby food manufacturers, car

seat companies, and MHL/UHL simulations for use with the public and parents of children with lesser degrees of hearing loss.

Newborn Screening

The group deemed identification of infants at risk for UHL and MHL in the newborn period an important public health issue, given the potential for interventions to prevent language and developmental delays. New data suggest that at 80% of infants with mild hearing loss are missed by a commonly used two-stage (OAE-AABR) screening protocol.[19] The group felt that it might be beneficial if manufacturers of screening equipment would provide clinicians with detailed specifications on how signal level was determined, as no calibration standards exist. It might be particularly helpful if the following information was included: 1) estimated signal ear canal level (automated-ABR equipment) for screening hearing loss; 2) signal-to-noise algorithm; 3) pass criteria; 4) additional filter or signal processing used to reduce noise; and 5) number of passes in "coupler" for 100,000 tests to determine the false-negative rate for the screening algorithm.

As more advanced newborn hearing screening technologies are developed, efforts directed at increasing the effectiveness of identifying MHL and UHL might also be beneficial. Follow-up testing of children who do not pass hearing screening could include testing in both ears as opposed to limiting testing to the "referred" ear. While the group discussed the fact that more children with MHL would be identified in the newborn period if the screening pass level on existing screening instruments were lowered (for example, 35–25 dB HL for AABR), it was recognized that such changes would increase the false-positive rate and that empirical data would be needed in order to determine the cost-benefit of such modifications.

Performing diagnostic testing prior to discharge for universal newborn hearing screening (UNHS) "refers" was discussed as a means of lowering the loss to follow-up rate, but the group noted short discharge times, lack of technology, and inadequate staff expertise as possible limitations to such a strategy. Studies examining the benefits of adding molecular screening (e.g., CMV, Cx26, Pendred's syndrome) of infants identified as at risk for late-onset hearing loss could enhance efforts to increase the sensitivity and specificity of early identification efforts.

Preschool Screening

The group felt that, following the neonatal screen, it might be beneficial for children to have their hearing screened annually prior to school entry. Acknowledging that such annual screening may be impractical, the group considered that at least one "universal" hearing screening to identify children with MHL and UHL should occur between ages 1 and 5 years. In a discussion of optimal timing for such a screen, it was recognized that a single time or locality for a "universal" screen in the preschool period may not be feasible. The group considered the possible benefits of targeted screening and discussed potential strategies for a preschool hearing screen, including during an immunization visit to the primary care provider, through Head Start, Child Find and state 3–5 programs (Part B, Section 619), and at daycare facilities. The group felt that having one additional

hearing screening before 3 years of age, ("one-before-three") could be beneficial in identifying mild and unilateral hearing loss that may have been missed during the newborn period. It was discussed that state EHDI programs and EHDI coordinators could assume a lead role in the preschool screening effort. The need for collaboration between state departments of health and departments of education in this effort was also discussed. Mirroring a school-age screening program, a preschool hearing screen program could incorporate behavioral and physiologic screening technologies. It was felt that enrollment into 0–3 (Part C) and 3–5 programs (Part B, Section 619) could be an optimal time for an audiological hearing screen for some children. Costs associated with such a targeted screening including follow-up were acknowledged.

School-age Screening

Existing data suggest that identifying both MHL and UHL in school-age children could be beneficial for some children. Current state and local policies and procedures for school-age screening may not necessarily identify all children with these forms of hearing loss. The group agreed that an annual screen for hearing loss might be ideal, and it was recognized that annual screening may not be practical or effective for the identification of MHL and UHL. Since approximately 98% of children enter the public school system at age 5–6 years, this event was discussed as a potential point at which a "universal" hearing screen could take place. Screening programs at school entry should consider using controlled, portable acoustic environments and behavioral methods (conditioned play), as well as physiologic screening technologies (tympanometry, acoustic middle ear muscle reflex, and evoked otoacoustic emissions).

It was felt that optimal times for hearing screening of school-age children could occur at school entry, middle school entry, and one point in between (possibly third grade). It might be beneficial for all children newly enrolled in school, identified by teachers or parents as having a possible hearing loss, and/or those who are academically challenged (including those with developmental delays) to have sensory screening at more regular intervals. The Medical Home was recognized as an important support and a mechanism for ensuring follow-up. The group discussed whether existing EHDI programs should consider expanding their efforts towards comprehensive school screening programs (including provisions for follow-up) for identification of all forms of hearing loss including MHL and UHL.

Research Needs Identified for Screening for Hearing Loss

- Establish normal hearing thresholds for infants, preschool, and school-age children through systematic meta-analyses of the current literature on this topic.
- Establish better estimates of the prevalence of mild and unilateral sensorineural hearing loss from birth through school-age. Data are needed from large cross-sectional and prospectively followed cohorts.
- Conduct a randomized controlled trial (longitudinal prospective study) that accounts for multiple demographic, socio-economic, familial, and auditory variables while comparing interventions for infants with unilateral hearing loss.

- Collect data on whether multiple (repeat) newborn hearing screenings increase the probability of a false-negative outcome. In so doing, determine a reasonable upper limit on the number of screening tests before referral for confirmatory audiologic testing.
- Study models (which include follow-up) of screening for mild hearing loss in school-age children. These can include evaluating test environments and existing test procedures/screening protocols that maximize the sensitivity and specificity of identifying mild and unilateral hearing loss and follow-up.
- Examine the Third National Health and Nutrition Examination Survey (NHANES-III) data using the workshop definition of permanent mild bilateral hearing loss and permanent unilateral hearing loss (definition adapted from Bess et al. 1998[5]). [Note: Dr. Danielle Ross and colleagues at CDC have undertaken this work; analyses should be complete in the near future.]
- Develop innovative and effective screening tests (technologies) and tools, including tests of speech and language proficiency, tests of functional hearing, academic achievement, and social/behavioral functioning.
- Develop protocols adaptable for various sites and environments in which infants and children receive hearing screenings.
- Develop improved behavioral screening technologies that include the use of automated algorithms for identifying mild hearing loss in infants.
- Examine the possible benefits and harms of adding molecular screening for mild and unilateral hearing loss in addition to a direct hearing screening for identifying late-onset, progressive hearing loss (e.g., CMV, Cx26, Pendred's/enlarged vestibular aqueduct [EVA]).

Other items noted:

- Virtually no data exist regarding the effectiveness of current screening devices for use in the detection of mild hearing loss. Gathering this data alone could increase our knowledge on this topic.
- We must increase public and professional awareness of mild and unilateral hearing loss.
    - Develop a mild hearing loss simulator that could be used to demonstrate mild and unilateral hearing loss to parents and the general public. Such a device could be used for research similar to Haggard and Primus.[92]
- Alternatives to the current adjectives or classifications of lesser degrees of hearing loss are needed, as terms such as "minimal" or "mild" may convey a lack of importance.

Breakout Discussion – Diagnostic Evaluation and Follow-up

Facilitator: Judith Widen, PhD

The discussion of Diagnostic Evaluation and Follow-up focused on: 1) diagnostic audiologic evaluation; 2) other evaluations; and 3) follow-up and transitions to early intervention.

Diagnostic Audiologic Evaluation

The group identified potential barriers to diagnostic evaluation and follow-up for infants with mild and unilateral hearing loss. Sensitivity of screening technology was identified as a possible barrier, as screening equipment typically does not detect hearing loss milder than 30–35 dB. Since present screening technology may not identify mild hearing loss in infants, it is unclear how these infants are identified and re-enter EHDI systems. For some infants identified with hearing loss, financial concerns and the need for insurance referrals may also create barriers to receiving diagnostic audiology examinations or needed follow-up care. Other possible barriers identified include lack of access to and/or availability of appropriate care from a qualified pediatric audiologist, lack of persistent follow-up, variability of ASSR in the low frequencies and low intensity levels, inconsistent use of correction factors in VRA and ABR testing, and issues in differentiating between conductive, mixed, and sensorineural hearing loss. The group also noted that labels such as "mild," "slight," and "less than mild" are used inconsistently and may de-emphasize the effects of lesser degrees of hearing loss on child development. The impact and importance of identification and intervention for mild and unilateral hearing loss was felt to be often lacking in parent and provider education.

The group discussed the best practices set forth in the ASHA Guidelines for Audiologic Assessment of Infants Birth to Five, 2004.[93] Due to the lack of national standards for ABR and VRA levels, the measurement of a threshold measured at 20–30 dB nHL (relative to normal adult thresholds) could result in mild losses being missed. In addition, many "normal" hearing infants have detectible ABRs at 0–10 dB nHL. The group discussed using a battery of diagnostic tests, including bone conduction, OAE, tympanometry, and acoustic reflexes, to distinguish between sensorineural and conductive hearing loss. The suggested timeframe and methods for obtaining ear-specific and accurate VRA information was also discussed. The group considered the Medical Home as one venue for identifying children at risk for developing hearing loss.

Other Evaluations

The group discussed a number of evaluations that could complement diagnostic audiological testing. For younger children, the discussion included etiological investigations such as genetic screening and counseling, electrocardiography (EKG), computed axial tomography (CAT), magnetic resonance imaging (MRI), renal ultrasound scan, and clinical lab testing such as for toxoplasmosis, rubella, CMV, herpes, syphilis, and thyroid stimulating hormone. For older children, the discussion included urinalysis and ophthalmologic examinations, visual evoked potentials, and electroretinography for

the identification of Usher syndrome. The group considered the timing of these evaluations and their potential for detecting hearing loss that often goes unidentified, particularly mild or unilateral hearing loss. The diagnostic yield for mild and unilateral hearing loss from these evaluations was also discussed. However, the possibility that specialists performing these evaluations may not understand the impact of hearing loss was also considered. The responsibility for ordering etiology investigations may fall on a primary care provider (PCP), an otolaryngologist, or a geneticist who may have limited knowledge of hearing loss. Finally, insurance coverage may create a barrier for accessing these evaluations if they are not included as a policy provision.

Follow-up and Transitions to Early Intervention

The group emphasized viewing a child in a holistic manner when assessing early intervention needs and involving a multidisciplinary team in assessment and early intervention. When transitioning to early intervention, the group noted that parents and providers may not understand the potential impact of mild and unilateral hearing loss on child development. Some interventionists may lack the knowledge, skills, and expertise in hearing loss (and particularly mild and unilateral hearing loss) needed to provide early intervention for these infants and children. The group discussed ways to educate parents and professional about referring children to appropriate Part C programs and early intervention.

Research Needs Identified for Diagnostic Evaluation and Follow-up

- Explore how diagnostic tests differ for unilateral and mild hearing loss.
- Measure actual differences in sound pressure levels in the ear canals of infants.
- Determine bone conduction thresholds in infants.
- Develop, with parents' and audiologists' input, recommendations on how to best inform parents about the potential outcomes of children with mild and unilateral hearing loss.
- Determine the diagnostic yield for other tests, such as genetic, computerized tomography (CT), and MRI while considering the possible harms.
- Collect additional data about the developmental outcomes of children using different types of hearing technology.
- Examine whether or not testing for other conditions (such as aminoglycides use and mitochondrial mutation) could yield results for children with mild and unilateral hearing loss.
- Determine the diagnostic yield for children with mild/unilateral hearing loss from positive CMV in urine versus blood.
- Determine the optimal time to do CT and MRI for enlarged vestibular aqueduct/large vestibular aqueduct syndrome (EVA/LVAS) in children with no other diagnosis.
- Consider when children should have an ophthalmologic evaluation to rule out Usher syndrome.
- Consider diagnostic evaluations at key transition stages (e.g., preschool to elementary school, and/or elementary school to high school).

Breakout Discussion: Hearing Technology

Facilitator: Anne Marie Tharpe, PhD

The group first discussed at what age slight hearing loss (16–25 dB HL) and mild hearing loss (26–40 dB HL) could be accurately diagnosed. Although there was not full agreement among the group members, it was decided that, under optimal conditions, an accurate diagnosis of slight loss could be made between 6 and 9 months developmental age and mild loss at 6 months or younger developmental age. It was acknowledged that there are potential limitations to current physiologic and behavioral diagnostic techniques for defining thresholds in this young age range. These limitations may include confounding factors such as the presence of otitis media with effusion (OME) and variability with, and lack of agreement between, auditory brainstem responses and behavioral threshold responses.

Once the degree, configuration, and type of hearing loss can be definitively determined, those with slight and mild degrees of long-standing hearing loss in the speech frequency range may be considered candidates for hearing aids. For those with UHL, contralateral routing of signal (CROS) hearing aids might not be recommended until the child is able to control his or her communication environment. Other technologies such as bone-anchored hearing aids (BAHA) or transcranial hearing aids may be considered with children 5 years of age and older. However, it was noted that there are no available data to support the use of either of these technologies with children. It was felt that data are also lacking in terms of support for traditional hearing aid use for children with UHL. However, this working group discussed the potential benefits of a trial usage of a hearing aid on an aidable ear with UHL at the time of diagnosis as a potential option.

Regardless of the decision of hearing aid fitting on a child with mild or UHL, given the published literature on this topic, group members felt that use of an FM system might enhance communication. For example, when parents are communicating with their child while in a car, a stroller, or other settings/situations that may compromise optimum communication due to background noise and lack of visual cues, an FM system might help the child discriminate the speech signal from background noise. Such fittings would require parent and, when appropriate, child counseling on the proper and selective use of an FM system.

In addition to hearing technology, the benefits of regularly monitoring the development of speech, language, and communication, as well as audiologic and otologic status, for children with MHL were discussed. Extensive counseling regarding the potential developmental risk factors associated with MHL, accommodations, and available services (state and local agencies) could be made available to families and, when appropriate, to the child.

The group identified several potential barriers to the use of hearing technology with this population, including a lack of parental education regarding the high-risk status of children with MHL. The group noted that some parents may misinterpret certain auditory

behaviors in their children; for example, parents may assume that if a child turns toward some sounds, he or she hears normally. Concern was also expressed that visible amplification devices may result in the labeling of children. Another possible barrier raised in the discussion was a lack of professional education regarding the high-risk status of children with MHL and UHL. In addition, the group agreed that there is often uncertainty in the diagnostic and treatment process, especially for infants and very young children. For example, difficulty in hearing aid coupling can be a barrier, especially in infancy. The group agreed that an evidence base for the optimum configuration of technology fitting (e.g., unilateral vs. bilateral, FM on one or both ears) would be beneficial.

Research Needs Identified for Hearing Technology

- Collect outcome data on amplification of unilateral hearing loss (early versus late; FM versus hearing aid).
- Explore what is "aidable" in terms of unilateral hearing loss.
- Compare the effects of early FM system use, hearing aid use, and no amplification on communication, educational, and social/emotional outcomes in children with unilateral hearing loss.
- Collect more data on children with slight hearing loss (i.e., 6–25 dB HL)
- Develop more speech perception measures for children birth–3 years of age.
- Determine the consequences of not aiding severe/profound unilateral hearing loss if there is potential for cochlear implantation in the future.
- Determine the effect of frequent/full-time FM use on the auditory skill development (listening in noise; localization) of children with mild or unilateral hearing loss.
- Determine the impact of OME on the development of children with mild hearing loss.
- Examine directional microphone use in children with mild hearing loss and unilateral hearing loss (in preschool and school-age children).
- Evaluate transcranial hearing aids, bone-anchored hearing aids, and fully implantable hearing aids in children.
- Explore coupling issues of FM and hearing aids in all groups (monaural versus binaural; FM only on one ear or both, etc.).
- Develop more sensitive and age-appropriate outcome measures/functional assessments (especially for birth–3 years of age).
- Determine whether verification/validation procedures should be different for unilateral hearing loss versus bilateral hearing loss.

Breakout Discussion: Early Intervention

Facilitator: Arlene Stredler-Brown, MA, CCC-SLP, CED

The early intervention breakout group discussed four discrete topics, addressing relevant issues and potential research needs for each:

1. Transition from identification to early intervention.
2. Models of early intervention.
3. Components of a developmental screening battery.
4. Components of a developmental diagnostic protocol.

Transition From Identification to Early Intervention

Some professionals may be disseminating previously published information that does not accurately describe the implications of minimal hearing loss. The group discussed actions that could improve this situation. One action step is to promote the dissemination of up-to-date information. Professional education could target physicians, audiologists, speech/language pathologists, teachers of the deaf/hard of hearing, members of Child Find teams, classroom teachers, and Part C service coordinators. It is more likely that parents will act on the information they receive if the information from various professionals is consistent.

Through their Part C programs, states have identified conditions for which individuals are eligible to receive services. Minimal hearing loss may or may not be included in a particular state's list of established conditions. If a state excludes minimal hearing loss from its eligibility list, it will be difficult for affected children to receive early intervention services. In addition, parents may be left with the impression that minimal hearing loss is not important. The current body of evidence suggests that minimal hearing loss can put young children at risk for developmental delay. A focus on the implications of minimal hearing loss might motivate each state to review its Part C eligibility criteria.

There are several early intervention data systems operating through state EHDI programs, state Part C programs, and state Child Find programs. The group discussed the feasibility for these programs to collect and integrate demographic data on children with minimal hearing loss. Ongoing data management for children entering school at 3 years of age was also discussed.

Some topics for further study include: 1) distributing information about minimal hearing loss to students in pre-service training programs (e.g., speech/language pathologists, audiologists, and teachers of the deaf and hard of hearing); 2) recognizing states with operational systems that identify, treat, and/or monitor outcomes of children with minimal hearing loss to serve as models for other states; and 3) conducting parent surveys to identify effective services parents have received and services they wish they had received.

Models of Early Intervention

Research to date suggests that mild bilateral hearing loss is associated with an elevated probability of speech and/or language delay in school-age children. However, as noted previously, not all children with mild bilateral hearing loss are eligible for state-funded services through Part C of IDEA. With this in mind, alternative funding sources to support treatment programs for these children were discussed. Possible alternative funding sources include state schools for the deaf and state Medicaid programs.

Studies indicate that 22% to 35% of school-age children with UHL fail at least one grade (review article by Lieu).[94] Based on this information, the group felt that implementation of a statewide monitoring program might be more appropriate than offering treatment to all children with UHL.

Group members discussed best practices for intervention/therapy for children with minimal hearing loss. The discussion of intervention/therapy included the following: 1) face-to-face contact with the family (in contrast to monitoring by phone and mail which makes the condition seem less important); 2) delivering information by a professional knowledgeable about hearing loss; 3) presentation of an unbiased list of treatment approaches that includes information about speech and language development, functional auditory skill development, and amplification; 4) parent-to-parent support; 5) support to child care providers; and 6) access to role models with mild and/or UHL. The group felt that services for children with minimal hearing loss who are fitted with amplification would need a more prescriptive treatment program.

Future research may address the impact of mild hearing loss and the likelihood of delay associated with this condition in infants and children birth–3 years of age. A prospective study to identify which children with UHL have a high likelihood of experiencing delays could be conducted. Randomized trials could identify outcomes for children who received intervention compared with those who did not. The randomization could be conducted through a natural selection process based on state programs' current eligibility criteria. This type of study would look at long-term outcomes for children with minimal hearing loss.

Components of a Developmental Screening Battery

The group acknowledged that a developmental screening process for children with minimal hearing loss would identify those who are experiencing delays. To this end, group members discussed a developmental screening battery to include tests that are standardized on hearing children. These tests could measure skills in multiple developmental domains including speech, language, and functional listening skills. It was felt that all children eligible for Part C services, as a result of any one of many developmental disabilities, could benefit from a screen for hearing loss.

Components of a Developmental Diagnostic Protocol

A diagnostic developmental assessment for children with minimal hearing loss could include norm-referenced tests that measure speech, language, and functional auditory

skills. In addition, tests could examine skills in all developmental domains, as a child with hearing loss is at increased risk for a secondary disability. Research could focus on the average performance of children with minimal hearing loss and factors predictive of successful outcomes.

Research Needs Identified for Early Intervention

- Determine whether professionals who work with children who have mild or unilateral hearing loss are adequately trained to do so. Identify their training needs. Investigate the implications for teacher preparation programs and speech-language pathology (SLP) training programs.
- Conduct descriptive studies of successful early intervention systems; successful state systems can serve as useful models for other state systems.
- Survey parents to identify the early intervention techniques that are successful, the services the parents felt they did not receive, and the services they did receive that were not useful.
- Research findings could be shared with state Part C coordinators for use in updating their eligibility criteria. Current evidence on mild hearing loss may not be conclusive. A larger body of evidence may be needed to establish Part C policy.
- Conduct prospective studies to identify which children with mild or unilateral hearing loss have a high likelihood of delay.
    - In a large sample, profile children with delays (e.g., percentage of children who show delay, at what age delay is exhibited, description of the delay).
    - Conduct randomized trials to compare outcomes for children who receive intervention with those for children who receive no intervention.
    - Investigate long-term outcomes to determine if children birth–3 years of age who experience delays are the same children experiencing delays in school.
    - Identify the hearing status of children who are high school dropouts. Investigate a correlation between hearing loss and high school dropout rates.
- For children with unilateral hearing loss, identify covariables such as laterality of hearing loss, the effects of amplification, personal amplification versus sound field, socioeconomic status, degree of loss, parent education, conductive versus sensorineural loss, etc.
    - Collect a larger body of evidence for such covariates for children birth–3 years of age.
    - Explore outcomes for children in preschool using amplification to determine if there are implications for amplification use with children under the age of 3.
- Investigate the assessment instruments commonly used to evaluate speech and language in children with normal hearing to determine if these instruments adequately measure delays for children with hearing loss:
    - Determine if commonly used instruments are culturally and linguistically appropriate.
    - Conduct validation studies on the correlation between parent-report and clinician-administered tests. Does parent reporting yield the same results as clinician-administered tests to determine eligibility for services?

- Provide evidence to justify early intervention for children with mild and unilateral hearing loss, such as evidence of critical periods for speech and language development.
- Conduct longitudinal studies to determine if early intervention prevents problems during the school years.
- Identify expectations for children with minimal hearing loss:
  - Identify factors that predict successful speech, language, and behavior outcomes.
  - Survey parent satisfaction with services.
  - Determine the average performance of children with minimal hearing loss. Look at outcomes longitudinally.
  - Address some of the issues requested by the US Preventive Services Task Force. This Task Force requested high-quality, large-scale longitudinal follow-up studies that:
    - Quantify the consequences of false-positive screens and false-negative screens to determine if there are clinically important harms that result from screening.
    - Identify speech, language, and scholastic achievement over time.
    - Promote comprehensive cost-benefit analyses that include the cost of tracking and follow-up for all children screened.

Next Steps

The National Workshop on Mild and Unilateral Hearing Loss was an important step in defining key issues related to mild and unilateral hearing loss. CDC EHDI, the Marion Downs Hearing Center, and workshop participants are now engaged in a variety of activities to increase the visibility of these issues. We anticipate that the momentum generated through the discussions, suggestions, and support at the workshop will lead to the development of proposals related to the early identification, assessment, and interventions appropriate for infants and children with permanent mild and unilateral hearing loss.

An extensive literature review of research related to mild and unilateral hearing loss was undertaken before to the workshop, and summary tables from this literature review are being prepared for release. Research needs identified at the workshop, particularly in the areas of prevalence and outcomes, are also being reviewed. In addition, federal agencies are exploring potential collaboration to advance research related to mild and unilateral hearing loss.

Presentations on mild and unilateral hearing loss were given at the American Speech-Language-Hearing Association Conference in November of 2005 (http://convention.asha.org/handouts.cfm), and are being coordinated for the National EHDI Conference in February of 2006 (http://www.cdc.gov/ncbddd/ehdi/meeting.htm), and the American Academy of Audiology Conference in April of 2006 (http://www.audiology.org/convention/2006/). An abstract is also being submitted for the Newborn Hearing Screening 2006 (http://nhs2006.isib.cnr.it/) international conference in Cernobio, Italy.

Meanwhile, publication opportunities related to mild and unilateral hearing loss are being pursued. Short-term and long-term recommendations discussed at the workshop continue to be refined and further developed; however, these recommendations were not finalized during the workshop. Recommendations/proposals are expected to be published in 2006 after further refinement by a committee with representation from national experts, a family support organization, and state and federal agencies.

Through these and other efforts, we aspire to bring this long standing issue the attention and action it deserves and to make a difference for children with mild and unilateral hearing loss and their families.

APPENDICES

APPENDIX A – Workshop Agenda

National Workshop on Mild and Unilateral Hearing Loss

Sponsored by the CDC Early Hearing Detection and Intervention (EHDI) Program
and the Marion Downs Hearing Center

Tuesday, July 26, 2005

(Peak 5 Conference Room - Beaver Run Resort)

12:30pm **Workshop Check-in**

Pick up materials

1:00pm **Opening Session**

Welcome – *John Eichwald*

Workshop overview and objectives – *Marcus Gaffney*
- Steering Committee comments

Overview of the research review and summary tables – *Danielle Ross*

1:30pm **Prevalence and Screening Session**

Prevalence and screening in newborns – *Judy Gravel*

Collection of data on infants with mild and unilateral hearing loss – *June Holstrum and Marcus Gaffney*
- Loss to follow-up

Prevalence in school-age children – *Fred Bess*

Break

2:15pm **Diagnosis, Amplification, and Outcomes Session**

Diagnostic evaluation – *Judith Widen, Barbara Cone-Wesson, and Yvonne Sininger*
- Audiologic management and family issues – *Kirsti Reeve*

Amplification – *Sarah McKay*
- FM systems – *Sandra Abbott Gabbard*

Outcomes in early childhood and school-age children[7] – *Anne Marie Tharpe and Danielle Ross*

Language, speech, and social-emotional outcomes of infants and children with mild and unilateral hearing loss and their families – *Christie Yoshinaga-Itano*

Language outcomes for young children with unilateral loss – *Allison Sedey*

Differences in outcomes for right ear vs. left ear unilateral hearing losses – *Marilyn W. Neault*

- Progression from unilateral to bilateral hearing loss

Break

3:30pm **Early Intervention, Eligibility, and Clinical Practice Session**

Early intervention services for children with mild or unilateral hearing loss – *Sharon Ringwalt*

- Overview of NECTAC and the relationship with EHDI
- State Part C eligibility requirements – *Karl White and Jamie Elliott*

Overview of U.K. HTA trial of early amplification in children with mild and unilateral hearing loss – *Shirley DeVoe*

Clinical implications of children with minimal hearing loss (mild bilateral and/or unilateral hearing loss) – *Arlene Stredler-Brown*

Break

4:45pm **Breakout Groups**

Review objectives:

- Identify issues and barriers.
- Identify areas for future research.
- Develop realistic, short-term recommendations.
- Develop long-term recommendations.

Breakout Group 1: Screening for Hearing Loss

Group Facilitator: Judy Gravel

Breakout Group 2: Diagnostic Evaluation and Follow-up

Group Facilitator: Judith Widen

Breakout Group 3: Hearing Technology

Group Facilitator: Anne Marie Tharpe

Breakout Group 4: Early Intervention

Group Facilitator: Arlene Stredler-Brown

5:30pm Adjourn

Wednesday, July 27, 2005

(Peak 5 Conference Room - Beaver Run Resort)

8:30am **Opening Session**

Review of proceedings from day one

9:00am **Reconvene All Breakout Groups**

Break

10:30am **Reconvene All Breakout Groups**

11:30pm – 1:00pm **Lunch (on your own)**

1:00pm **Breakout Group 1 Summary**

Summary of discussions by group facilitator
Group discussion

Breakout Group 2 Summary

Summary of discussions by group facilitator
Group discussion

Break

2:45pm **Breakout Group 3 Summary**

Summary of discussions by group facilitator
Group discussion

Breakout Group 4 Summary

Summary of discussions by group facilitator
Group discussion

4:15pm **Closing Session**

Review of key points
Open discussion

5:00pm Adjourn

APPENDIX B – List of Breakout Group Members

Breakout Group 1: Screening for Hearing Loss

Judy Gravel (Group Facilitator)
Janet Farrell (Recorder)

Karen Anderson
Barbara Cone-Wesson
Albert Mehl
Nancy Pajak
Karl White

Breakout Group 2: Diagnostic Evaluation and Follow-up

Judith Widen (Group Facilitator)
Vickie Thomson (Recorder)

Laura Jennings-Kepler
Pam Mason
Maryrose McInerney
Sarah McKay
Marilyn Neault
Helen Robertson
Shirley Russ
Yvonne Sininger

Breakout Group 3: Hearing Technology

Anne Marie Tharpe (Group Facilitator)

Fred Bess
Leisha Eiten
Sandra Abbott Gabbard
Cheryl Johnson
James Miller
Robert Oyler

Breakout Group 4: Early Intervention

Arlene Stredler-Brown (Group Facilitator)

Lyn Bopp
Sarah Borton
Karen Carpenter
Wendy Dudley
BJ James
Terry Keegan
Annette Landes
Delores Orfanakis
Susanne Reed
Kirsti Reeve
Julie Reichman
Sharon Ringwalt
Allison Sedey
Katherine Thomas
Lynn Wismann-Horthor
Christie Yoshinaga-Itano

Rotating

John Eichwald	June Holstrum
Jamie Elliott	Lynn Luethke
Irene Forsman	Danielle Ross
Marcus Gaffney	Elizabeth Sullivan

Breakout Group Objectives

- Identify major issues and barriers.
- Identify areas for future research.
- Develop realistic, short-term recommendations for identification and intervention.
- Develop long-term recommendations for identification and intervention.

APPENDIX C – Further Research Needs

The four breakout groups were asked to identify research needs related to their particular area of discussion. Breakout group facilitators reported the research needs identified by each group to all the participants in the final afternoon of the Workshop. Below are the research needs suggested by each group. (Note: Research needs identified by each breakout group are also included above, following each breakout group discussion.)

Research Needs Identified for Screening for Hearing Loss

- Establish normal hearing thresholds for infants, preschool, and school-age children through systematic meta-analyses of the current literature on this topic.
- Establish better estimates of the prevalence of mild and unilateral sensorineural hearing loss from birth through school-age. Data are needed from large cross-sectional and prospectively followed cohorts.
- Conduct a randomized controlled trial (longitudinal prospective study) that accounts for multiple demographic, socio-economic, familial, and auditory variables while comparing interventions for infants with unilateral hearing loss.
- Collect data on whether multiple (repeat) newborn hearing screenings increase the probability of a false-negative outcome. In so doing, determine a reasonable upper limit on the number of screening tests before referral for confirmatory audiologic testing.
- Study models (which include follow-up) of screening for mild hearing loss in school-age children. These can include evaluating test environments and existing test procedures/screening protocols that maximize the sensitivity and specificity of identifying mild and unilateral hearing loss and follow-up.
- Examine the Third National Health and Nutrition Examination Survey (NHANES-III) data using the workshop definition of permanent mild bilateral hearing loss and permanent unilateral hearing loss (definition adapted from Bess et al. 1998[5]). [Note: Dr. Danielle Ross and colleagues at CDC have undertaken this work; analyses should be complete in the near future.]
- Develop innovative and effective screening tests (technologies) and tools, including tests of speech and language proficiency, tests of functional hearing, academic achievement, and social/behavioral functioning.
- Develop protocols adaptable for various sites and environments in which infants and children receive hearing screenings.
- Develop improved behavioral screening technologies that include the use of automated algorithms for identifying mild hearing loss in infants.
- Examine the possible benefits and harms of adding molecular screening for mild and unilateral hearing loss in addition to a direct hearing screening for identifying late-onset, progressive hearing loss (e.g., CMV, Cx26, Pendred's/enlarged vestibular aqueduct [EVA]).

Other items noted by the Screening for Hearing Loss group:

- Virtually no data exist regarding the effectiveness of current screening devices for use in the detection of mild hearing loss. Gathering this data alone could increase our knowledge on this topic.
- We must increase public and professional awareness of mild and unilateral hearing loss.
    - Develop a mild hearing loss simulator that could be used to demonstrate mild and unilateral hearing loss to parents and the general public. Such a device could be used for research similar to Haggard and Primus.[92]
- Alternatives to the current adjectives or classifications of lesser degrees of hearing loss are needed, as terms such as "minimal" or "mild" may convey a lack of importance.

Research Needs Identified for Diagnostic Evaluation and Follow-up

- Explore how diagnostic tests differ for unilateral and mild hearing loss.
- Measure actual differences in sound pressure levels in the ear canals of infants.
- Determine bone conduction thresholds in infants.
- Develop, with parents' and audiologists' input, recommendations on how to best inform parents about the potential outcomes of children with mild and unilateral hearing loss.
- Determine the diagnostic yield for other tests, such as genetic, computerized tomography (CT), and MRI while considering the possible harms.
- Collect additional data about the developmental outcomes of children using different types of hearing technology.
- Examine whether or not testing for other conditions (such as aminoglycides use and mitochondrial mutation) could yield results for children with mild and unilateral hearing loss.
- Determine the diagnostic yield for children with mild/unilateral hearing loss from positive CMV in urine versus blood.
- Determine the optimal time to do CT and MRI for enlarged vestibular aqueduct/large vestibular aqueduct syndrome (EVA/LVAS) in children with no other diagnosis.
- Consider when children should have an ophthalmologic evaluation to rule out Usher syndrome.
- Consider diagnostic evaluations at key transition stages (e.g., preschool to elementary school, and/or elementary school to high school).

Research Needs Identified for Hearing Technology

- Collect outcome data on amplification of unilateral hearing loss (early versus late; FM versus hearing aid).
- Explore what is "aidable" in terms of unilateral hearing loss.
- Compare the effects of early FM system use, hearing aid use, and no amplification on communication, educational, and social/emotional outcomes in children with unilateral hearing loss.

- Collect more data on children with slight hearing loss (i.e., 6–25 dB HL)
- Develop more speech perception measures for children birth–3 years of age.
- Determine the consequences of not aiding severe/profound unilateral hearing loss if there is potential for cochlear implantation in the future.
- Determine the effect of frequent/full-time FM use on the auditory skill development (listening in noise; localization) of children with mild or unilateral hearing loss.
- Determine the impact of OME on the development of children with mild hearing loss.
- Examine directional microphone use in children with mild hearing loss and unilateral hearing loss (in preschool and school-age children).
- Evaluate transcranial hearing aids, bone-anchored hearing aids, and fully implantable hearing aids in children.
- Explore coupling issues of FM and hearing aids in all groups (monaural versus binaural; FM only on one ear or both, etc.).
- Develop more sensitive and age-appropriate outcome measures/functional assessments (especially for birth–3 years of age).
- Determine whether verification/validation procedures should be different for unilateral hearing loss versus bilateral hearing loss.

Research Needs Identified for Early Intervention

- Determine whether professionals who work with children who have mild or unilateral hearing loss are adequately trained to do so. Identify their training needs. Investigate the implications for teacher preparation programs and speech-language pathology (SLP) training programs.
- Conduct descriptive studies of successful early intervention systems; successful state systems can serve as useful models for other state systems.
- Survey parents to identify the early intervention techniques that are successful, the services the parents felt they did not receive, and the services they did receive that were not useful.
- Research findings could be shared with state Part C coordinators for use in updating their eligibility criteria. Current evidence on mild hearing loss may not be conclusive. A larger body of evidence may be needed to establish Part C policy.
- Conduct prospective studies to identify which children with mild or unilateral hearing loss have a high likelihood of delay.
    - In a large sample, profile children with delays (e.g., percentage of children who show delay, at what age delay is exhibited, description of the delay).
    - Conduct randomized trials to compare outcomes for children who receive intervention with those for children who receive no intervention.
    - Investigate long-term outcomes to determine if children birth–3 years of age who experience delays are the same children experiencing delays in school.
    - Identify the hearing status of children who are high school dropouts. Investigate a correlation between hearing loss and high school dropout rates.
- For children with unilateral hearing loss, identify covariables such as laterality of hearing loss, the effects of amplification, personal amplification versus sound field, socioeconomic status, degree of loss, parent education, conductive versus sensorineural loss, etc.

  - Collect a larger body of evidence for such covariates for children birth–3 years of age.
  - Explore outcomes for children in preschool using amplification to determine if there are implications for amplification use with children under the age of 3.
- Investigate the assessment instruments commonly used to evaluate speech and language in children with normal hearing to determine if these instruments adequately measure delays for children with hearing loss:
  - Determine if commonly used instruments are culturally and linguistically appropriate.
  - Conduct validation studies on the correlation between parent-report and clinician-administered tests. Does parent reporting yield the same results as clinician-administered tests to determine eligibility for services?
- Provide evidence to justify early intervention for children with mild and unilateral hearing loss, such as evidence of critical periods for speech and language development.
- Conduct longitudinal studies to determine if early intervention prevents problems during the school years.
- Identify expectations for children with minimal hearing loss:
  - Identify factors that predict successful speech, language, and behavior outcomes.
  - Survey parent satisfaction with services.
  - Determine the average performance of children with minimal hearing loss. Look at outcomes longitudinally.
  - Address some of the issues requested by the US Preventive Services Task Force. This Task Force requested high-quality, large-scale longitudinal follow-up studies that:
    - Quantify the consequences of false-positive screens and false-negative screens to determine if there are clinically important harms that result from screening.
    - Identify speech, language, and scholastic achievement over time.
    - Promote comprehensive cost-benefit analyses that include the cost of tracking and follow-up for all children screened.

APPENDIX D – Bibliography

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