This document compiles abstracts of papers that were presented at a 3-day conference of experts which developed a consensus statement on early identification of hearing impairment in infants and young children. Papers addressed taxonomy; epidemiology; developmental consequences of early hearing impairment; methodology, instrumentation, and personnel; and models for early identification and follow-up. Abstracts of the following papers are included: "Early Identification of Hearing Impairment in Infants and Young Children" (Robert J. Ruben); "Site of Lesion and Age of Onset" (Jerome O. Klein); "Causal Factors and Concomitant Impairment" (Michael A. Karchmer); "Incidence/Prevalence" (Patrick E. Brookhouser); "At-Risk Populations" (N. Wendell Todd); "Auditory Development" (Joseph W. Hall); "Language Acquisition" (Mary P. Moeller); "Effects of Hearing Impairment on the Development of Speech" (D. Kimbrough Oliver); "Social/Emotional and Academic Consequences of Hearing Loss" (Diane Brackett); "Benefits of Screening at Birth: Economic, Educational, and Functional Factors" (Marion P. Downs); "High-Risk Register" (Allan O. Diefendorf); "Behavioral Measures" (Judith S. Gravel); "Acoustic Immittance Measures" (Robert H. Margolis); "Developmental Screening in a Busy Clinic: A Luxury or a Necessity?" (Noel D. Matkin); "Recording the Auditory Brainstem Response in Infants" (Terence W. Picton); "The Auditory Brainstem Response in Infants: Basic Aspects" (Richard C. Folsom); "Auditory Brainstem Response Testing in the Neonatal Intensive Care Unit" (Bruce A. Weber); "Evaluation of Hearing in the Neonate Using the Auditory Brainstem Response" (Yvonne S. Sininger); "Hearing Screening of Infants with Auditory Brainstem Response Protocols, Personnel, and Price" (James W. Hall III); "Otoacoustic Emissions: Basic Aspects" (Theodore J. Glattke); "Utility of Distortion-Product Otoacoustic Emissions in Identifying Hearing Impairment in Infants and Young Children" (Brenda L. Lonsbury-Martin); "Characteristics of Transient Evoked Otoacoustic Emissions in Pediatric Populations" (Susan J. Norton); "Practicality, Validity, and Cost-Efficiency of Universal Newborn Hearing Screening Using Evoked Otoacoustic Emissions" (Karl R. White); "Models for Early Identification and Followup: An Overview" (Robert G. Turner); "U.S.A. Models" (Thomas M. Mahoney); "Canadian Models and Issues" (Martyn L. Hyde); and "The Public Health Perspective on Screening—U.K. Experience and Recommendations" (Mark P. Haggard). Most papers contain references. (DB)
EARLY IDENTIFICATION OF HEARING IMPAIRMENT IN INFANTS AND YOUNG CHILDREN

March 1-3, 1993
Mosur Auditorium, Clinical Center
National Institutes of Health
CONSENSUS DEVELOPMENT CONFERENCE ON
EARLY IDENTIFICATION OF HEARING IMPAIRMENT
IN INFANTS AND YOUNG CHILDREN

NIH Consensus Development Conference
March 1-3, 1993
Masur Auditorium
Warren Grant Magnuson Clinical Center
National Institutes of Health
Bethesda, Maryland

Sponsored by the Office of Medical Applications of Research and the National Institute on Deafness and Other Communication Disorders and cosponsored by the National Institute of Child Health and Human Development and the National Institute of Neurological Disorders and Stroke of the National Institutes of Health
CONTENTS

Introduction to the Consensus Development Conference on Early Identification of Hearing Impairment in Infants and Young Children ............................................. 1

Agenda ........................................................................................................... 5

Panel .............................................................................................................. 9

Speakers ...................................................................................................... 11

Planning Committee ..................................................................................... 13

Abstracts .................................................................................................... 15

I. Introduction and Overview

   Early Identification of Hearing Impairment in Infants and Young Children
      Robert J. Ruben, M.D., F.A.C.S., F.A.A.P. ................................................. 17

II. Taxonomy

   Site of Lesion and Age of Onset
      Jerome O. Klein, M.D. ............................................................................ 21

   Causal Factors and Concomitant Impairment
      Michael A. Karchmer, Ph.D. ................................................................. 25

III. Epidemiology

   Incidence/Prevalence
      Patrick E. Brookhouser, M.D., F.A.C.S. ............................................. 27

   At-Risk Populations
      N. Wendell Todd, M.D. ...................................................................... 37

IV. Developmental Consequences of Early Hearing Impairment

   Auditory Development
      Joseph W. Hall, Ph.D. ......................................................................... 43

   Language Acquisition
      Mary P. Moeller, M.S. ................................................................. 45
Effects of Hearing Impairment on the Development of Speech
D. Kimbrough Oller, Ph.D. ................................................................. 53

Social/Emotional and Academic Consequences of Hearing Loss
Diane Brackett, Ph.D. ................................................................. 57

Benefits of Screening at Birth: Economic, Educational, and Functional Factors
Marion P. Downs, M.A., D.H.S. ......................................................... 63

V. Methodology, Instrumentation, and Personnel

High-Risk Register
Allan O. Diefendorf, Ph.D. ............................................................. 67

Behavioral Measures
Judith S. Gravel, Ph.D. ................................................................. 71

Acoustic Immittance Measures
Robert H. Margolis, Ph.D. ............................................................ 75

Developmental Screening in a Busy Clinic: A Luxury or a Necessity?
Noel D. Matkin, Ph.D. ................................................................. 79

Recording the Auditory Brainstem Response in Infants
Terence W. Picton, M.D., Ph.D. ......................................................... 83

The Auditory Brainstem Response in Infants: Basic Aspects
Richard C. Folsom, Ph.D. ............................................................... 87

Auditory Brainstem Response Testing in the Neonatal Intensive Care Unit
Bruce A. Weber, Ph.D. ................................................................. 91

Evaluation of Hearing in the Neonate Using the Auditory Brainstem Response
Yvonne S. Sininger, Ph.D. .............................................................. 95

Hearing Screening of Infants With Auditory Brainstem Response: Protocols, Personnel, and Price
James W. Hall III, Ph.D. ............................................................... 99

Otoacoustic Emissions: Basic Aspects
Theodore J. Glatke, Ph.D. ............................................................. 103

Utility of Distortion-Product Otoacoustic Emissions in Identifying Hearing Impairment in Infants and Young Children
Brenda L. Lonsbury-Martin, Ph.D. ............................................... 107
Characteristics of Transient Evoked Otoacoustic Emissions in Pediatric Populations
Susan J. Norton, Ph.D., CCC-A .................................................. 111

Practicality, Validity, and Cost-Efficiency of Universal Newborn Hearing Screening
Using Evoked Otoacoustic Emissions
Karl R. White, Ph.D. ................................................................. 115

VI. Models for Early Identification and Followup

Models for Early Identification and Followup: An Overview
Robert G. Turner, Ph.D. ............................................................ 119

U.S.A. Models
Thomas M. Mahoney, Ph.D. .................................................... 123

Canadian Models and Issues
Martyn L. Hyde, Ph.D. .............................................................. 129

The Public Health Perspective on Screening—U.K. Experience and Recommendations
Mark P. Haggard, Ph.D. ............................................................. 133

v
INTRODUCTION TO THE CONSENSUS DEVELOPMENT CONFERENCE ON EARLY IDENTIFICATION OF HEARING IMPAIRMENT IN INFANTS AND YOUNG CHILDREN

There is a clear need in the United States for improved methods and models for the early identification of hearing impairment in infants and young children. Approximately 1 of every 1,000 children is born deaf. Many more are born with less severe degrees of hearing impairment, while others develop hearing impairment during childhood. Reduced hearing acuity during infancy and early childhood interferes with the development of speech and language skills. Although less well documented, reduced auditory input also adversely affects the developing auditory nervous system and can have harmful effects on social, emotional, cognitive, and academic development, as well as on a person's vocational and economic potential. Moreover, delayed identification and management of severe to profound hearing impairment may impede the hearing-impaired child's ability to adapt to life in a hearing world or to prepare for life in the hearing-impaired community.

The critical period for language and speech development is generally regarded as the first two years of life and, although there are several methods of identifying hearing impairment during the first year of life, the average age of identification in the United States remains close to 3 years. Lesser degrees of hearing loss may go undetected even longer. The result is that for many hearing-impaired infants and young children, much of the critical language and speech learning period is lost. There is general agreement that hearing impairment should be recognized as early in life as possible, so that the remediation process can take full advantage of the plasticity of the developing sensory systems.

Infant hearing screening has been attempted with a number of different test methods, including cardiac response audiometry, respiration audiometry, alteration of sucking patterns, movement or startle in response to auditory stimuli, various behavioral paradigms, measurement of acoustic reflexes, and more recently, auditory brainstem response (ABR) audiometry. In addition, attention has recently turned to the measurement of otoacoustic emissions, which shows promise as a fast, inexpensive, noninvasive test of cochlear function. Each method is effective in its own way, but technical or interpretative limitations have impeded widespread application. Moreover, these approaches vary widely in their sensitivity, specificity, and predictive efficiency in identifying hearing impairment.

Today, most neonatal screening programs are focused on infants who satisfy one or more of a number of criteria for inclusion in a "high-risk registry." However, the use of the high-risk registry to limit the population being screened excludes approximately 50 percent of infants with hearing impairment. The preferred screening test method has come to be ABR, combined with audiologic followup and/or diagnostic ABR for those infants who fail the screening protocols. Despite the relatively good predictive efficiency of ABR, its cost, time requirements, and technical difficulties have discouraged the general application of this method in screening the far larger newborn population not meeting the high-risk registry criteria. Consensus on a unified approach to early identification has also been delayed by the scarcity of data on the relative sensitivity, specificity, predictive efficiency, and cost effectiveness of the hearing screening techniques currently used to identify hearing impairments in infants and young children.
This consensus development conference will bring together specialists in audiology, otolaryngology, pediatrics, neonatology, hearing science, speech-language pathology, health care administration, epidemiology, counseling, and other health care areas, as well as representation from the public. The purpose of the conference is to reach agreement on which children should have their hearing screened or tested and at what age, which methods and models are preferred for identifying hearing impairment in infants and young children, and the key areas for future research.

Following 1½ days of presentations by medical experts and discussion by the audience, an independent consensus panel will weigh the scientific evidence and prepare a draft statement in response to the following key questions:

1. What are the advantages of early identification of hearing impairment and the consequences of late identification of hearing impairment?

2. Which children (birth through 5 years) should be screened for hearing impairment and when?

3. What are the advantages and disadvantages of current screening methods?

4. What is the preferred model for hearing screening and followup?

5. What are the important directions for future research?

On the final day of the meeting, the consensus panel chairperson, Dr. Gregory Matz, Professor and Chairman, Department of Otolaryngology, Loyola University of Chicago Medical Center, will read the draft statement to the conference audience and invite comments and questions. A press conference will follow to allow the panel and chairperson to respond to questions from media representatives.

General Information

Conference sessions will take place in Masur Auditorium, Warren Grant Magnuson Clinical Center. The telephone number for messages during the conference is (301) 496-2520 (voice)/(301) 496-6896 (TTY/TDD/TT). All messages will be posted on the board inside the auditorium. For outgoing calls, TTY/TDD/TT services are available at the following locations: (1) in Room 1N312, (2) at the Red Cross desk, and (3) at the public telephone.

Sign Language Interpreting

Sign language interpreting and other reasonable accommodation provided.

Cafeteria

The Clinical Center cafeteria on the B-1 level, one level below Masur Auditorium, is open from 7:30 a.m. to 2:00 p.m. daily. The cafeteria on the second floor of the ACRF wing is open 24 hours. To get to the ACRF cafeteria, take the escalator from the lobby level just past the elevators near the Clinical Center front entrance.
CME Credit

The Foundation for Advanced Education in the Sciences/National Institutes of Health is accredited by the Accreditation Council for Continuing Medical Education (CME) to sponsor medical education for physicians.

The Foundation for Advanced Education in the Sciences/National Institutes of Health designates this continuing medical education activity for 15 credit hours in Category 1 of the Physicians Recognition Award of the American Medical Association.

Sponsors

This conference is sponsored by the Office of Medical Applications of Research and the National Institute on Deafness and Other Communication Disorders and cosponsored by the National Institute of Child Health and Human Development and the National Institute of Neurological Disorders and Stroke of the National Institutes of Health.
AGENDA

Monday, March 1

8:30 a.m. Welcome and Introduction Ralph F. Naunton
           Director
           Division of Communication
           Sciences and Disorders
           National Institute on Deafness
           and Other Communication
           Disorders (NIDCD)
           James B. Snow
           Director
           NIDCD

8:35 a.m. Charge to the Panel John H. Ferguson
           Director
           Office of Medical Applications
           of Research

8:45 a.m. Conference Issues Gregory J. Matz
           Conference and Panel
           Chairperson

I. Introduction and Overview

9:00 a.m. Early Identification of Hearing Impairment
           in Infants and Young Children Robert J. Ruben

II. Taxonomy

9:30 a.m. Site of Lesion and Age of Onset Jerome O. Klein

9:50 a.m. Causal Factors and Concomitant
           Impairment Michael A. Karchmer

10:10 a.m. Discussion

III. Epidemiology

10:40 a.m. Incidence/Prevalence Patrick E. Brookhouser

11:00 a.m. At-Risk Populations N. Wendell Todd

11:20 a.m. Discussion
### Monday, March 1 (continued)

**IV. Developmental Consequences of Early Hearing Impairment**

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Speaker</th>
</tr>
</thead>
<tbody>
<tr>
<td>11:35 a.m.</td>
<td>Auditory Development</td>
<td>Joseph W. Hall</td>
</tr>
<tr>
<td>11:50 a.m.</td>
<td>Language Acquisition</td>
<td>Mary P. Moeller</td>
</tr>
<tr>
<td>12:05 p.m.</td>
<td>Discussion</td>
<td></td>
</tr>
<tr>
<td>12:20 p.m.</td>
<td>LUNCH</td>
<td></td>
</tr>
<tr>
<td>1:20 p.m.</td>
<td>Effects of Hearing Impairment on the Development of Speech</td>
<td>D. Kimbrough Oller</td>
</tr>
<tr>
<td>1:40 p.m.</td>
<td>Social/Emotional and Academic Consequences of Hearing Loss</td>
<td>Diane Brackett</td>
</tr>
<tr>
<td>2:00 p.m.</td>
<td>Benefits of Screening at Birth: Economic, Educational, and Functional Factors</td>
<td>Marion P. Downs</td>
</tr>
<tr>
<td>2:20 p.m.</td>
<td>Discussion</td>
<td></td>
</tr>
</tbody>
</table>

**V. Methodology, Instrumentation, and Personnel**

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Speaker</th>
</tr>
</thead>
<tbody>
<tr>
<td>2:50 p.m.</td>
<td>High-Risk Register</td>
<td>Allan O. Diefendorf</td>
</tr>
<tr>
<td>3:05 p.m.</td>
<td>Behavioral Measures</td>
<td>Judith S. Gravel</td>
</tr>
<tr>
<td>3:20 p.m.</td>
<td>Acoustic Immittance Measures</td>
<td>Robert H. Margolis</td>
</tr>
<tr>
<td>3:35 p.m.</td>
<td>Developmental Screening in a Busy Clinic: A Luxury or a Necessity?</td>
<td>Noel D. Matkin</td>
</tr>
<tr>
<td>3:50 p.m.</td>
<td>Discussion</td>
<td></td>
</tr>
<tr>
<td>4:20 p.m.</td>
<td>Recording the Auditory Brainstem Response in Infants</td>
<td>Terence W. Picton</td>
</tr>
<tr>
<td>4:35 p.m.</td>
<td>The Auditory Brainstem Response in Infants: Basic Aspects</td>
<td>Richard C. Folsom</td>
</tr>
<tr>
<td>4:50 p.m.</td>
<td>Discussion</td>
<td></td>
</tr>
<tr>
<td>5:20 p.m.</td>
<td>Adjournment</td>
<td></td>
</tr>
</tbody>
</table>
Tuesday, March 2

V. Methodology, Instrumentation, and Personnel (continued)

8:00 a.m. Auditory Brainstem Response Testing in the Neonatal Intensive Care Unit
Bruce A. Weber

8:15 a.m. Evaluation of Hearing in the Neonate Using the Auditory Brainstem Response
Yvonne S. Sininger

8:30 a.m. Hearing Screening of Infants with Auditory Brainstem Response: Protocols, Personnel, and Price
James W. Hall III

8:45 a.m. Discussion

9:05 a.m. Otoacoustic Emissions: Basic Aspects
Theodore J. Glattke

9:20 a.m. Utility of Distortion-Product Otoacoustic Emissions in Identifying Hearing Impairment in Infants and Young Children
Brenda L. Lonsbury-Martin

9:35 a.m. Characteristics of Transient Evoked Otoacoustic Emissions in Pediatric Populations
Susan J. Norton

9:50 a.m. Practicality, Validity, and Cost-Efficiency of Universal Newborn Hearing Screening Using Evoked Otoacoustic Emissions
Karl R. White

10:05 a.m. Discussion

VI. Models for Early Identification and Followup

10:35 a.m. Models for Early Identification and Followup: An Overview
Robert G. Turner

10:55 a.m. U.S.A. Models
Thomas M. Mahoney

11:15 a.m. Canadian Models and Issues
Martyn L. Hyde

11:35 a.m. The Public Health Perspective on Screening—U.K. Experience and Recommendations
Mark P. Haggard

11:55 a.m. General Discussion
Tuesday, March 2 (continued)

12:30 p.m.  Adjournment

Wednesday, March 3

9:00 a.m.  Presentation of Consensus Statement  Gregory J. Matz
           Conference and Panel Chairperson

9:30 a.m.  Discussion

11:00 a.m.  Panel Meets in Executive Session

1:00 p.m.  Press Conference

2:00 p.m.  Adjournment
Conference and Panel Chairperson:
Gregory J. Matz, M.D.
Professor and Chairman
Department of Otolaryngology
Loyola University of Chicago Medical Center
Maywood, Illinois

Agnes M. Aamodt, Ph.D., R.N.
Professor Emerita
College of Nursing
University of Arizona
Tucson, Arizona

Thomas G.R. Bower, Ph.D.
Founder's Professor of Human Development
University of Texas at Dallas
Richardson, Texas

Michael B. Bracken, Ph.D.
Professor and Vice Chairman
Department of Epidemiology and Public Health
Yale University School of Medicine
New Haven, Connecticut

Richard A. Chole, M.D., Ph.D.
Professor and Chair
Department of Otolaryngology
University of California at Davis
Davis, California

M. Jane Collins, Ph.D.
Associate Professor and Chair
Department of Communication Sciences and Disorders
Louisiana State University
Baton Rouge, Louisiana

Ronald S. Fischler, M.D., F.A.A.P.
President
North Scottsdale Pediatric Associates, P.C.
Scottsdale, Arizona

James F. Jerger, Ph.D.
Professor and Head
Division of Audiology and Speech Pathology
Department of Otorhinolaryngology and Communicative Sciences
Baylor College of Medicine
Houston, Texas

Richard H. Jones, Ph.D.
Professor of Biometrics
Department of Preventive Medicine and Biometrics
School of Medicine
University of Colorado Health Sciences Center
Denver, Colorado

Rodger Martinez, M.R.P.
Vice Chairman
Albuquerque Area Indian Health Board, Inc.
Ramah, New Mexico

Fred D. Minifie, Ph.D.
Professor
Department of Speech and Hearing Sciences
University of Washington
Seattle, Washington

Ruth Nass, M.D.
Associate Professor of Neurology
New York University Medical Center
New York, New York
Brenda M. Ryals, Ph.D.  
Associate Professor  
Department of Speech Pathology and Audiology  
James Madison University  
Harrisonburg, Virginia

Susan Sniderman, M.D.  
Professor of Pediatrics  
Department of Neonatology  
University of California at San Francisco  
San Francisco, California

Barry Zuckerman, M.D.  
Professor of Pediatrics  
Boston University School of Medicine  
Boston, Massachusetts
SPEAKERS

Diane Brackett, Ph.D.
Associate Professor
Communication Sciences
University of Connecticut
Storrs, Connecticut

Patrick E. Brookhouser, M.D., F.A.C.S.
Director
Boys Town National Research Hospital
Omaha, Nebraska

Allan O. Diefendorf, Ph.D.
Associate Professor
Department of Otolaryngology
Indiana University School of Medicine
Riley Hospital for Children
Indianapolis, Indiana

Marion P. Downs, M.A., D.H.S.
Professor Emerita
Department of Otolaryngology
University of Colorado Health Sciences Center
Denver, Colorado

Richard C. Folsom, Ph.D.
Associate Professor
Department of Speech and Hearing Sciences
University of Washington
Seattle, Washington

Theodore J. Glattke, Ph.D.
Professor
Department of Speech and Hearing Sciences
University of Arizona
Tucson, Arizona

Judith S. Gravel, Ph.D.
Director of Audiology
Associate Professor of Otolaryngology
Albert Einstein College of Medicine and The Montefiore Medical Center
Bronx, New York

Mark P. Haggard, Ph.D.
Director
MRC Institute of Hearing Research
Professor
University of Nottingham
Nottingham
United Kingdom

James W. Hall III, Ph.D.
Associate Professor and Director of Audiology
Director
Vanderbilt Balance and Hearing Center
Vanderbilt University
Nashville, Tennessee

Joseph W. Hall, Ph.D.
Professor
Division of Otolaryngology/Head and Neck Surgery
University of North Carolina School of Medicine
Chapel Hill, North Carolina

Martyn L. Hyde, Ph.D.
Director, Research and Development
Otologic Function Unit
Mount Sinai Hospital/The Toronto Hospital
Professor
Department of Otolaryngology/Preventive Medicine and Biostatistics/Speech Pathology
University of Toronto
Toronto, Ontario
Canada

Michael A. Karchmer, Ph.D.
Dean, Graduate Studies and Research
Gallaudet Research Institute
Gallaudet University
Washington, DC

Jerome O. Klein, M.D.
Professor of Pediatrics
Boston University School of Medicine
Boston, Massachusetts
Brenda L. Lonsbury-Martin, Ph.D.
Professor and Director of Research
Department of Otolaryngology
University of Miami Ear Institute
Miami, Florida

Thomas M. Mahoney, Ph.D.
Director
Bureau of Communicative Disorders
Utah State Department of Health
Salt Lake City, Utah

Robert H. Margolis, Ph.D.
Professor and Director of Audiology
Department of Otolaryngology
University of Minnesota
Minneapolis, Minnesota

Noel D. Matkin, Ph.D.
Professor
Department of Speech and Hearing Sciences
University of Arizona
Tucson, Arizona

Mary Pat Moeller, M.S.
Coordinator
Center for Childhood Deafness
Boys Town National Research Hospital
Omaha, Nebraska

Susan J. Norton, Ph.D., CCC-A
Director
Research and Clinical Audiology
Children's Hospital and Medical Center
Associate Professor
Department of Otolaryngology
University of Washington School of Medicine
Seattle, Washington

D. Kimbrough Oller, Ph.D.
Professor
Departments of Psychology, Pediatrics,
and Otolaryngology
University of Miami
Miami, Florida

Terence W. Picton, M.D., Ph.D.
Professor of Medicine
Division of Neurology
University of Ottawa
Ottawa, Ontario
Canada

Robert J. Ruben, M.D., F.A.C.S., F.A.A.P.
Professor and Chairman
Department of Otolaryngology
Professor of Pediatrics
Albert Einstein College of Medicine and
The Montefiore Medical Center
Bronx, New York

Yvonne S. Sinner, Ph.D.
Director
Children's Auditory Research and
Evaluation Center
House Ear Institute
Los Angeles, California

N. Wendell Todd, M.D.
Associate Professor
Department of Otolaryngology/Surgery
and Pediatrics
Emory University
Atlanta, Georgia

Robert G. Turner, Ph.D.
Department Head
Department of Communication Disorders
Louisiana State University Medical Center
New Orleans, Louisiana

Bruce A. Weber, Ph.D.
Associate Professor
Department of Otolaryngology
Duke University Medical Center
Durham, North Carolina

Karl R. White, Ph.D.
Professor
Departments of Psychology and
Special Education
Utah State University
Logan, Utah
PLANNING COMMITTEE

Chairperson: Ralph F. Naunton, M.D.
Director
Division of Communication Sciences and Disorders
National Institute on Deafness and Other Communication Disorders
National Institutes of Health
Bethesda, Maryland

Patricia Blessing
Public Affairs Specialist
National Institute on Deafness and Other Communication Disorders
National Institutes of Health
Bethesda, Maryland

Allan O. Diefendorf, Ph.D.
Associate Professor
Department of Otolaryngology
Indiana University School of Medicine
Riley Hospital for Children
Indianapolis, Indiana

Jerry M. Elliott
Program Analyst
Office of Medical Applications of Research
National Institutes of Health
Bethesda, Maryland

John H. Ferguson, M.D.
Director
Office of Medical Applications of Research
National Institutes of Health
Bethesda, Maryland

Judith S. Gravel, Ph.D.
Director of Audiology
Associate Professor of Otolaryngology
Albert Einstein College of Medicine and the Montefiore Medical Center
Bronx, New York

William H. Hall
Director of Communications
Office of Medical Applications of Research
National Institutes of Health
Bethesda, Maryland

Lynn E. Huerta, Ph.D.
Program Administrator
Division of Communication Sciences and Disorders
National Institute on Deafness and Other Communication Disorders
National Institutes of Health
Bethesda, Maryland

James F. Kavanagh, Ph.D.
Deputy Director
Center for Research for Mothers and Children
National Institute of Child Health and Human Development
National Institutes of Health
Bethesda, Maryland

Noel D. Matkin, Ph.D.
Professor
Department of Speech and Hearing Sciences
University of Arizona
Tucson, Arizona
The following are the abstracts of presentations to the Consensus Development Conference on Early Identification of Hearing Impairment in Infants and Young Children. They are designed for the use of panelists and participants in the conference and as a reference document pertinent to the conference for anyone interested in the conference deliberations. We are grateful to the authors who have summarized their materials and made them available in a timely fashion.

Ralph F. Naunton, M.D.
Director
Division of Communication Sciences and Disorders
National Institute on Deafness and Other Communication Disorders
National Institutes of Health

Jerry M. Elliott
Program Analyst
Office of Medical Applications of Research
National Institutes of Health
The transition from an industrial to a communication society has occurred during the 20th century. This changed economic and social basis of society has focused a new awareness of those diseases that interfere with the effective communicative functioning of an individual in the communication age. Communication disorders in general and hearing loss in particular, have become fundamental morbidities resulting in crippling impairments in a communication-based society.

Screening for hearing loss in school-age children was first undertaken in the 1920’s and 1930’s. The concept of early identification of younger children, however, needed to await the development of technology to determine accurately the extent of their hearing loss and the development of facilities and programs for effective interventions for these very young hearing impaired. The techniques for diagnosis and intervention evolved from the 1950’s to 1970’s. The Joint Commission on Infant Hearing published its first statement on neonatal hearing screening for hearing impairment in 1971. Since then, there have been further advances in the ability to diagnose hearing loss accurately and precisely from birth onward, and there has been progress in the implementation of interventions, many of which are effective.

Two concepts underlie the basis for early identification:

1. There is a critical period for the establishment of optimal language; the earlier an intervention can be instituted, the better the outcome will be.

2. Remediation of hearing defects result in improved communication.

If there is delay in detection and intervention during the critical period for development of language, then the child will become language impaired—data substantiate this at different levels of hearing loss. Additionally, a child with impaired hearing will have deficits in communication, and early remediation of the hearing impairment will restore the child’s ability to communicate.

Hearing impairments vary in their effect on the infant and child in regard to threshold, frequency range, and more complex functions such as signal detection. Other substantial variables that affect the outcome of hearing impairment in an infant or young child are age of onset; rate of progression; variability of impairment over time (e.g., conductive losses from otitis media with effusion); variable conductive loss associated with static, progressive, or unilateral sensory or neural losses; deficits in other sensory systems (e.g., visual impairments); cognitive skills (e.g., mental retardation); and the quality of sensory input (e.g., linguistic, sensory, and/or social deprivation). The total effect of a hearing impairment is a complex function of the type and extent of the impairment and the biological/social characteristics of the infant or child. The slight conductive loss in a child with a moderate sensorineural deficit will be more burdensome than the same conductive loss in a normal hearing infant. A moderate conductive hearing loss in a normal child whose parents provide an optimal linguistic environment will have little or no effect on language, whereas the same loss in a
normal child who is subjected to extremely poor linguistic input because of inadequate or absent parenting will adversely affect language.

The definition of hearing impairment must take into account all the contributing variables. As the criteria develop from the historical ones of a pure tone threshold to a more complex set of functions, the significant variables of the type of impairment interacting with a sensory context must be taken into account. Identification programs of the next decade need to be configured to look at those who are at risk for hearing impairment and those who are at risk for the effects of hearing impairment.

As identification programs are established, conflict will arise between need and available resources. Criteria for identification should be determined that will identify both those with the greatest need and those who will benefit most from early detection. It should be apparent to a communication-based society that one of the most critical areas of health delivery is the optimalization of the communicative skills. Early identification is the basis for potential effective interventions. Often with the knowledge that there are effective interventions, many children are identified, but access to the interventions is not available for all those identified. All identifications must be evaluated as to the following:

- The intervention resources available
- Effectiveness of case management of those identified
- Quality of access to the resources
- The measure of the outcome of the intervention—which is the communication of ability as evidenced by the child's language.

Since the 1970's, improvement has been made in one outcome measure of early detection of severely hearing-impaired infants and children, resulting in a decrease in the age of detection and intervention. For some of the most in need, even this limited outcome is unsatisfactory because of the delay of diagnosis and lack of access. This is especially true in our cities with large populations of medically underserved and in some rural communities.

The complexity of the variables for identification criteria of which we are now cognizant dictates a multidimensional approach for the identification of hearing impairments. Several strategies have evolved. The first is the use of the high-risk registry. These registries must be modified to include the risks intrinsic to the child and/or the child's environmental contacts. The second is the use of the various screening techniques for all newborns. This approach has been limited as to the threshold of identification and also in its incapacity to identify progressive losses. A third strategy is intensive education of health care deliverers, with regard to the sequelae of failure to intervene early in cases of hearing deprivation and the efficacy of intervention programs. Fourth, most recently, language is used as an identification strategy for communication disorders. Language must be the essential outcome measure of the interventions for hearing impairments in the infant and young child, and language can be used to identify those affected adversely by a hearing impairment. The use of a language screen has the additional advantages of using few resources; it can detect the effect without
requiring special measures of the hearing loss or of other background variables and may define other communication disorders, which are prevalent and adversely affect the language outcome of the infant or young child.

All four strategies will be needed to effect a comprehensive program of early identification so that the linguistic and communicative sequelae of hearing impairments can be prevented or cured.

A healthy society of the 21st century will be characterized by the communication effectiveness of its populace. We must accustom ourselves to the awareness that sound, when diminished either in quantity or quality, is no less a disease vector than a virus or bacteria; it, too, causes detrimental change in the organism and resultant morbidity. In the communication age, biology and culture interact in the creation and treatment of disease.

REFERENCES


SITE OF LESION AND AGE OF ONSET

Jerome O. Klein, M.D.

Hearing loss identified in children may have its onset in utero, during the first days of life, and in the infant and preschool child. The causes of hearing impairment are varied and include congenital and postnatal infections, genetic disorders, drugs and toxins, hemorrhage, hypoxia, and noise. Information about site of the lesion is available from experimental animal models, autopsy data, and selective diagnostic procedures. Recent and past monographs are valuable resources for information about the epidemiology and pathogenesis of hearing impairment in children.¹ ²

Hearing Loss in the Neonate

Congenital Infections

Congenital infections due to rubella virus, cytomegalovirus, and Treponema pallidum may be responsible for hearing loss. The etiologies and lesions of congenital infections are reviewed extensively in the literature.³ Congenital rubella may cause a necrotizing angiopathy of small blood vessels that results in tissue necrosis and inflammatory changes in various organs including the inner ear. The central nervous system lesions of congenital syphilis are also vascular, including an endarteritis leading to neuronal injury followed by fibrosis. The lesions of cytomegalovirus infection acquired in utero include focal encephalitis and invasion by viral inclusion-bearing cells and viral antigen-containing cells within structures of the inner ear including the organ of Corti and in epithelial cells of the striae vascularis.

Congenital Malformations

Congenital malformation of the auricle, external ear canal, and ossicles are rare causes of conductive hearing loss in the neonate.

Genetic Disorders

Approximately 50 percent of congenital sensorineural hearing loss is reported to be genetic. The hearing loss may be part of an identifiable syndrome, but many appear without any clear association. Among the syndromes that have been identified are Treacher Collins (mandibulofacial dysostosis), Goldenhar (eye, ear, spine deformities), osteogenesis imperfecta, Waardenburg, Crouzon, and Klippel-Feil.

Factors Associated With Pregnancy, Delivery, and the Nursery

Asphyxia in utero, at Delivery, or After Birth. Hearing loss associated with intrapartum asphyxia is due to damage to the cochlear nuclei as well as production of hemorrhage in the inner ear. Direct hypoxic injury to brainstem auditory nuclei may also occur.
Hyperbilirubinemia. Damage to the neural auditory pathways and to the cochlear nucleus due to bilirubin toxicity has been documented in experimental animal models.

Extension of Intracranial Hemorrhage into the Inner Ear.

Ototoxic Drugs Administered to the Pregnant Woman. Although streptomycin and other aminoglycoside antibiotics administered to the pregnant woman are potentially ototoxic for the fetus, documentation of such an effect is limited. Case reports of ototoxicity in infants whose mothers were treated with streptomycin during pregnancy are associated with risk factors including maternal renal failure or use of another ototoxic drug.

Ototoxic Drugs Administered to the Newborn Infant. The aminoglycoside antibiotics neomycin, streptomycin, kanamycin, and gentamicin have been implicated as a cause of sensorineural hearing losses in infants and children. Incrimination of the drug alone is often difficult because of other high-risk factors present in the infants including asphyxia, hyperbilirubinemia, and noise exposure.

Hearing Loss in the Infant and Preschool Child

Bacterial Meningitis

Hearing loss is the most frequent severe sequelae of bacterial meningitis. Sensorineural hearing loss has been documented in 6 to 15 percent of children with Hemophilus influenzae meningitis, 31 percent of children with pneumococcal meningitis, and in 10.5 percent of patients with meningococcal meningitis. Many children have hearing loss as the only neurologic deficit following bacterial meningitis.

The mechanisms responsible for hearing deficits following meningitis include the spread of infection along the auditory canal and cochlear aqueduct, and serous or purulent labyrinthitis and subsequent replacement of the membranous labyrinth with fibrous tissue and new bone.4

Viral Labyrinthitis

Mumps, measles, influenza, parainfluenza, adenovirus 3, varicella-zoster, cytomegaloviruses, and the arenavirus of Lassa fever have been associated with sudden sensorineural hearing loss. Khetarpal and colleagues reviewed temporal bone findings in patients with presumed viral labyrinthitis.5

Hearing Loss Associated with Otitis Media

Fluctuating or persisting loss of hearing is present in most children who have middle ear effusion. Conductive loss in the range of 15 to 40 dB occurs in children with middle ear effusion.6 The hearing loss is influenced by the volume of fluid in the middle ear and not by the quality of fluid. The hearing impairment is usually reversed with resolution of the effusion.

Permanent conductive hearing loss may occur because of irreversible changes resulting from recurrent acute or chronic inflammation, including adhesive otitis media or ossicular discontinuity. Cholesteatoma may be a destructive process in the middle ear that may damage the ossicles, tympanic
membrane, and the bony frame of the middle ear. Damage is believed to occur by enzymatic destruction and from pressure exerted by the expanding mass.

High negative pressure in the ear or atelectasis in the absence of effusion is another cause of conductive loss.

Sensorineural hearing loss is an uncommon sequela of acute otitis media or otitis media with effusion. A reversible hearing impairment is attributed to the effect of increased tension and stiffness of the round window membrane. A permanent sensorineural loss may occur as a result of spread of infection or productions of inflammation through the round window membrane, development of a perilymphatic fistula in the oval round window, or a suppurative complication such as meningitis or labyrinthitis.

Ototoxic Drugs

Salicylates, antimalarial drugs including quinine and chloroquine, aminoglycoside antibiotics, erythromycin, vancomycin, cisplatin, and loop diuretics such as furosemide have been associated with sensorineural hearing loss.

Ear drops containing aminoglycoside antibiotics are a common treatment for children who have chronic suppurative otitis media or for children who have a perforation of the tympanic membrane or have placement of tympanotomy tubes. Solutions containing aminoglycoside antibiotics (including the drug vehicles such as propylene glycol) instilled into the middle ear of experimental animals may cause severe cochlear damage and loss of hair cells. In general, clinical studies of patients receiving ototopical drugs have not corroborated the results of the animal studies.

Noise

Noise-induced hearing loss was discussed at a recent Consensus Development Conference held in January 1990. Liberman noted that if the exposure is sufficient, most structures of the inner ear can be damaged, including all cell types within the organ of Corti and most cell types in the lateral wall (stria vascularis, spiral ligament, spiral prominence, etc.).

Miscellaneous Causes of Conductive Hearing Losses

Obstruction of the external canal that leads to conductive hearing loss may occur as a result of cerumen impaction and presence of foreign bodies, stenosis of the external canal, development of cholesteatoma and masses such as aural polyps, and external otitis. Perforation of the tympanic membrane from various causes may result in hearing loss. Blood can enter the middle ear space as a result of head trauma or leukemia. Benign bone growth can present in the middle ear causing hearing loss.
REFERENCES


CAUSAL FACTORS AND CONCOMITANT IMPAIRMENT

Michael A. Karchmer, Ph.D.

This presentation discusses from a national perspective concomitant impairments occurring among deaf and hard-of-hearing children and youth in relation to other factors, including the reported cause of the hearing impairment. Specifically, the presentation describes the extent to which these children and youth in special education programs have disabilities in addition to hearing loss, and it characterizes the groups of students with specific conditions.

The information being presented comes from a continuing national study, the Annual Survey of Hearing-Impaired Children and Youth. This study, conducted since 1968 by the Gallaudet Research Institute’s Center for Assessment and Demographic Studies, includes information on individual children and youth across the country who receive special educational services in relation to their hearing impairment and other disabilities. This presentation focuses on data collected through the Annual Survey in the spring of 1992. Information on nearly 48,000 students nationwide was included in this database.

The Annual Survey gathers information about characteristics of the students and the educational services they receive. The information comes from official school records; the usefulness of the data for biomedical purposes is somewhat limited, and the conclusions based on analysis of this database need to be carefully interpreted. Nevertheless, the large number of cases in this database gives the opportunity to demonstrate meaningful relationships that can lead to other research.

Overall, as of the spring of 1992, about 30 percent of all hearing-impaired students (ages 3 to 21) in special education programs had concomitant conditions that significantly affected the educational process. The specific conditions varied widely, ranging from physical or health conditions (e.g., heart disorders, visual problems, etc.) to impairments of learning, cognitive, or intellectual function. About 10 percent of the total group was reported to have at least two conditions in addition to deafness. Cognitive and intellectual disabilities were reported about twice as often as physical conditions. The number of additional conditions and their type were found to be strongly related to such factors as age, ethnic/racial status, and degree of hearing loss. For example, African-American students were found to be much more likely than other students to have additional conditions.

The reported causes and the age at onset of hearing loss also were found to be strongly related to the nature of concomitant conditions. For example, students with reported hereditary causes were much less likely to have additional disabilities than students whose hearing losses were due to other causes. Also, causes associated with onset at birth (with the exception of hereditary causes) tend to be associated with higher rates of physical handicapping conditions than causes with onset after birth.
INCIDENCE/PREVALENCE

Patrick E. Brookhouser, M.D., F.A.C.S.

In basic terms, epidemiology is the study of the occurrence and distribution of physical and psychological conditions (e.g., acute or chronic disorders) with respect to time, place, and specified populations. The term incidence, as applied to a condition such as hearing loss, refers to the number of cases that had their onset during a defined time period (e.g., month, year) (NHIS, 1986). In contrast, the term prevalence refers to the number of persons in a population of interest who have the condition at a given point in time (NHIS, 1986). The term average annual point prevalence estimate is applied to the average of 52 individual weekly prevalence estimates obtained in such questionnaire-based studies as the National Health Interview Survey.

Other basic epidemiologic terminology includes the concept of impairment, which is defined as "a chronic or permanent defect, usually static in nature, that results from disease, injury or congenital malformation...(which) represents a decrease in or loss of ability to perform various functions" (NHIS, 1986) or alternatively as "any loss or abnormality of psychological, physiological or anatomical structure or function" (WHO, 1980). On the other hand, a disability entails "any long- or short-term reduction of a person's activity as a result of an acute or chronic condition" (NHIS, 1986) or "any restriction or lack of ability to perform an activity in the manner or within the range considered normal for a human" (WHO, 1980).

Epidemiologic studies of hearing loss can be broadly categorized as (1) interview/questionnaire surveys (IQS's), which do not involve direct evaluation of hearing, and (2) clinical examination studies (CES's), in which an individual subject's hearing is evaluated as one phase of the data collection process. An important example of the IQS format is the annual National Health Interview Survey (NHIS), which reports data regarding an array of acute and chronic conditions gathered by interviews/questionnaires from a sample of the civilian noninstitutionalized U.S. population. The NHIS is an ongoing survey whereby each week a probability sample of the U.S. population is interviewed by U.S. Bureau of the Census representatives. The entire population sample interviewed during 1990 consisted of 46,476 households containing 119,631 individuals, and annual average point prevalence estimates were calculated for hearing loss from the survey results.

An important strength of the NHIS is the statistical validity of the population sample selection methodology. Significant weaknesses include some respondents' failure to report hearing loss because of the condition's attendant social stigma and the survey's dependence on secondhand information regarding family members not accessible to the interviewer. Variations in phraseology and formulation of interviewers' questions can significantly influence completeness of reporting by participants. For example, the 1990 questionnaire included a set of special questions on hearing impairment that were not included in 1989; thus the reported overall prevalence for both hearing impairment and tinnitus in 1990 exceeded 1989 figures by 15.1 percent and 21.9 percent, respectively. Examples of standard NHIS questions on hearing loss are: Does anyone in the family now have deafness in one or both ears? Does anyone in the family now use a hearing aid? Special questions attempt to further quantify the hearing loss, such as, Without a hearing aid, can (Subject X) usually hear and understand what a person says without seeing his face if that person whispers to (Subject X)
from across a quiet room?" or, alternatively, if that person speaks loudly in (Subject X's) better ear?"

Additional information also was obtained regarding the age of onset of the reported hearing loss and the presence of tinnitus, as well as the use of hearing aids and assistive listening devices. Figure 1 shows trends in NHIS prevalence data from 1983 through 1990.

Another source of prevalence information for children and youth served by school programs is the Thirteenth Annual Report to Congress on the Implementation of the Individuals With Disabilities Education Act (IDEA). During the 1989-90 school year, 4,219,463 children, ages 6 to 21, received special education services under PL 94-142. Of this total population, 56,955 (1.35 percent) were classified as hearing impaired, with an additional 1,564 qualifying as deaf-blind. At present, the States are not required to report handicap-specific data for infants and preschoolers. The Annual Survey of Hearing-Impaired Children and Youth, a voluntary survey conducted by the Center for Assessment and Demographic Studies at Gallaudet University, provides additional demographic, audiological, and educational information about children receiving services provided by schools for the deaf and by local school districts across the United States (see Figure 2). The 46,666 children and youth included in the 1989-90 Annual Survey represent about 60 percent of the number of children receiving educational services as reported by States in compliance with the IDEA mandate. Inclusion criteria for these two surveys are disparate enough to preclude valid point-by-point interstudy comparison. Figures 3a and 3b compare Annual Survey data collected in 1989-90 with comparable information from 1984-85.

Although substantial prevalence information derived from CES's is available, inconsistencies of study design make cross-comparison and generalization of results problematic. Examples of parameters that must be specified carefully in any CES-type study include the following:

- Type of auditory stimuli utilized (e.g., pure tone, warble tone, click, tone burst, speech)
- Test conditions (e.g., audiometric test suite, neonatal ICU, quiet schoolroom)
- Type of response elicited and technique utilized (e.g., evoked brainstem response, evoked otoacoustic emission, conditioned response-VRA, BOA, or speech)
- Immitance results, if utilized
- Bilateral vs. unilateral results (e.g., better ear only, worse ear only)
- Inclusion criteria for the study population with regard to pure conductive vs. pure sensorineural vs. mixed hearing losses
- Inclusion criteria regarding progressive and fluctuating losses
- Criteria for identifying the existence of a hearing loss in a study participant (e.g., absence of evoked otoacoustic emission or auditory brainstem response, response worse than upper limits of normal range at any of the standard pure-tone test frequencies (0.25, 0.500, 1, 2, 3, 4, or 8 kHz), pure-tone average below normal range as defined by different authors, agencies, and professional organizations (0.5,1,2; 0.5,1,2,3; 0.5,1,2,3,4; 1,2,3; 1,3,4; 2,3,4; or 1,2,3,4 kHz)
• Upper limits of normal range (e.g., 15, 20, 25, or 40 dB) and whether age-specific and technique-specific norms were used with subjects such as high-risk neonates and infants

• Demographics of study populations (e.g., age, gender, racial/ethnic characteristics) and other special considerations (e.g., admission criteria for NICU, physiological state of NICU baby at time of test, acceptability of consanguineous marriages in certain ethnic/religious groups)

• Diagnostic criteria for various disorders if etiology-specific prevalence data are reported (e.g., was rubella diagnosed by rash only or was laboratory confirmation required?). Before etiology-specific results can be generalized across populations, information should be available about routine childhood immunization schedules for rubella, mumps, measles, and *Hemophilus influenzae* in the comparison groups.

Given the aforementioned caveats about interpreting results, several well-designed reviews report substantially similar prevalence information. Representative examples are depicted in Tables 1 and 2. Approximately 1.0-2.0/1,000 children in developed countries have a bilateral sensorineural hearing loss (SNHL) of 50 dB or worse, whereas the bilateral losses in 0.5-1.0/1,000 children exceed 75 dB. In Third World countries, the prevalence of SNHL is estimated to be at least twice as great, with infectious etiologies playing a greater role than in developed nations (Davidson, Hyde, and Albert, 1989). The value of reports from countries with government-sponsored health care systems, such as the United Kingdom, are enhanced by mandatory reporting requirements in some instances. In a recent multiyear study, Davis and Wood in Nottingham (UK) reported that 1 of 943 babies had an SNHL or mixed hearing loss; broken down by immediate postnatal care history, 1 of 174 NICU babies vs. 1 of 1,278 non-NICU infants had an SNHL or mixed hearing loss. Most etiology-specific studies are hampered by the difficulty of assigning a firm diagnosis in 30 to 40 percent or more cases, plus uncertainty about the accuracy of diagnosis in many cases with a presumptive etiology of recessively inherited, nonsyndromic hearing loss.

Well-designed, multicenter, etiology-specific epidemiologic studies of childhood hearing impairment are sorely needed to aid in planning allocation of resources for diagnosis, treatment, (re)habilitation, and education of these children, as well as targeting research support. Such studies would be extremely helpful in assessing the outcome of major public health initiatives, such as early childhood immunization against common infectious etiologies of hearing loss (e.g., *Hemophilus influenzae*, meningitis, mumps, measles).
Figure 1. National Health Interview Survey* Prevalence of Hearing Impaired per 1000 Persons by Age

![Bar chart showing prevalence of hearing impaired per 1000 persons by age from 1983 to 1990.](chart)


Figure 2. Hearing-Impaired Children and Youth in School Programs for Hearing Impaired Under 19 Years of Age*

![Bar chart showing number of hearing-impaired children and youth in school programs from 1976-77 to 1989-90.](chart)

*Adapted from Schildroth and Hatto, 1991 and from Schildroth, 1986.
Figure 3a. Annual Survey of Hearing-Impaired Children and Youth* in School Programs for Hearing Impaired.

Comparison by Severity of Loss

Figure 3b. Annual Survey of Hearing-Impaired Children and Youth* in School Programs for Hearing Impaired.

Comparison by Age

*Adapted from Schildroth and Hatto, 1991

3% 3% 11% 11% 21% 25% 24% 27% 26% 24% 15% 10%

Age in Years

< 3 3-5 6-9 10-13 14-17 > 17

1984-85 N=49,166 1989-90 N=46,045

*Adapted from Schildroth and Hatto, 1991
### Table 1. Prevalence of Childhood SNHL per 100,000 (Presumed Etiology)

<table>
<thead>
<tr>
<th>Authors</th>
<th>Prenatal</th>
<th>Perinatal</th>
<th>Postnatal</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parving (1983) 35 dB/2-12 y/o</td>
<td>73</td>
<td>20</td>
<td>7</td>
<td>38</td>
</tr>
<tr>
<td>Martin (1982) 50 dB/8 y/o</td>
<td>28</td>
<td>14</td>
<td>12</td>
<td>38</td>
</tr>
<tr>
<td>Feinmesser (1982) 55 dB/8 y/o</td>
<td>65</td>
<td>22</td>
<td>10</td>
<td>73</td>
</tr>
<tr>
<td>Schein (1974) Deaf/3 y/o</td>
<td>105</td>
<td>10</td>
<td>136</td>
<td>82</td>
</tr>
</tbody>
</table>

Adapted from Davidson et al, 1989

### Table 2. Incidence of Profound Childhood Deafness per Million Births

<table>
<thead>
<tr>
<th>Authors</th>
<th>Total Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chung et al (1959)</td>
<td>453</td>
</tr>
<tr>
<td>Sank (1963)</td>
<td>797</td>
</tr>
<tr>
<td>Fraser (1965)</td>
<td>500</td>
</tr>
<tr>
<td>Brown &amp; Chung (1971)</td>
<td>500</td>
</tr>
<tr>
<td>Furusho &amp; Yasuda (1973)</td>
<td>406</td>
</tr>
<tr>
<td>Nance et al (1977)</td>
<td>500</td>
</tr>
<tr>
<td>Newton (1985)</td>
<td>800</td>
</tr>
<tr>
<td>Majumder et al. (1989)</td>
<td>600</td>
</tr>
</tbody>
</table>

Adapted from Morton, 1991
SUGGESTED READINGS


Goldstein DP. Hearing impairment, hearing aids and audiology. ASHA, Sep 1984;24-38.


Hearing loss in children commonly occurs in two forms, conductive and sensorineural. Conductive hearing loss in children is attributable to two conditions: (1) otitis media, which typically involves mild to moderate hearing loss; and (2) congenital malformation of the middle and external ear, known as microtia, which typically involves moderate to severe hearing loss. Factors associated with an increased occurrence of otitis media are well known. Interestingly, cystic fibrosis patients seem resistant to having otitis media; studying these patients may yield insight into the occurrence of otitis media. The occurrence of microtia is quite different in various races: in New Mexico, the rate in Indians is about eight times that in Anglos, and the rate in Hispanics is nearly twice that in Anglos.

Sensorineural hearing loss, in contrast to conductive hearing loss, is less well defined as to “at-risk populations.” Worldwide, at least one-third of children with sensorineural hearing loss lack an etiologic explanation. The rate of childhood sensorineural hearing loss in developed countries is about half the rate in underdeveloped countries. My experience with Native Americans of the Southwest was that they too have bilateral severe-profound sensorineural loss about 2.5 times the rate of non-Indians of Arizona and New Mexico.

Recent reports of both prospectively and retrospectively acquired data suggest populations of children at increased risk for sensorineural hearing loss (see Tables 1 and 2). Generalizations based on these data are, of course, speculative. The case-control study of metropolitan Atlanta children, being done by Yeargin-Allsopp et al. of the Centers for Disease Control, may generate hypotheses about risk factors.

In 1972, the Joint Committee on Infant Hearing recommended a five-item high-risk register as a tool for the early identification of hearing loss. Additional items were added in 1982, and again in 1990. The inadequacy of our knowledge of sensorineural hearing loss in children is exemplified by experiences with the three versions of the high-risk register. In Maryland, about 10 to 12 percent of newborns meet at least one risk criterion to get on the register. Of these children, about 5 percent are eventually labeled as having a hearing loss. However, the high-risk register leads to the diagnosis of only about 50 percent of the children who are born with a hearing loss. “The statistics are within a range similar to that seen in other States performing infant hearing screening.”

My opinion of populations of children who are at increased risk for sensorineural hearing loss is summarized in Table 3.
<table>
<thead>
<tr>
<th>First Author</th>
<th>Year</th>
<th>Population</th>
<th>Factors&lt;sup&gt;a&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salamy&lt;sup&gt;8&lt;/sup&gt;</td>
<td>1988</td>
<td>252 newborns ≤1.500 grams followed through 3 years of age; U. California, San Francisco</td>
<td>bad &quot;neonatal status&quot;: many ICU days, many x-rays 14 (5%) SNHL 12 in sickest quartile 2 in next sickest 25%</td>
</tr>
<tr>
<td>Halpern&lt;sup&gt;9&lt;/sup&gt;</td>
<td>1987</td>
<td>799 newborns in intensive care followed through 4 years of age; Stanford, California</td>
<td>6.1% hearing loss four accurate predictors: length of stay in ICU gestational age TORCH infections craniofacial anomalies</td>
</tr>
</tbody>
</table>

<sup>a</sup>ICU, intensive care unit; TORCH is an acronym for complex congenital perinatal viral infections: toxoplasmosis, rubella, cytomegalovirus, and herpes.
<table>
<thead>
<tr>
<th>First Author</th>
<th>Year</th>
<th>Population</th>
<th>Factors (ratio or percent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pablal0</td>
<td>1991</td>
<td>Nottingham, United Kingdom 46 children with bilateral SNHL or mixed hearing loss ≥40 dB HL at 0.5, 1, 2, and 4 kHz in the better ear</td>
<td>1.2 such cases/1,000 live births male/female = 1.55 30 genetic 42 acquired 28 unknown</td>
</tr>
<tr>
<td>Dias11</td>
<td>1990</td>
<td>Portugal (N = 934) national clinic</td>
<td>21 hereditary 15 perinatal 10 rubella 6 meningitis 4 measles, mumps 2 ototoxicity 42 unknown, other</td>
</tr>
<tr>
<td>Bastos12</td>
<td>1990</td>
<td>Angola: its only ENT clinic 105 consecutive cases of moderate-profound SNHL</td>
<td>22 meningitis 12 measles, mumps 7 genetic 5 ototoxicity 64 unknown, other</td>
</tr>
<tr>
<td>Das13</td>
<td>1988</td>
<td>Greater Manchester County, United Kingdom studied 164 born 1981-84</td>
<td>1.14/1,000 births male/female = 1.48 36 unknown etiology 20 genetic 15 perinatal factors 10 congenital infections 6 meningitis 4 syndromic 4 chromosomal</td>
</tr>
<tr>
<td>Wolff14</td>
<td>1987</td>
<td>Gallaudet survey United States</td>
<td>9 due to meningitis</td>
</tr>
<tr>
<td>McPherson15</td>
<td>1985</td>
<td>Gambia (700,000 persons) national survey found 259 children with severe-profound SNHL</td>
<td>32 meningitis 8 familial 2 rubella 2 congenital measles 56 unknown, other</td>
</tr>
</tbody>
</table>
TABLE 3. Factors for Likelihood of Bilateral Sensorineural Hearing Deficit in Populations of Children Less Than 5 Years of Age

(in order of highest to least likelihood)

<table>
<thead>
<tr>
<th>Factor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parent/caregiver concern regarding child's hearing</td>
</tr>
<tr>
<td>Family members with hearing loss in childhood</td>
</tr>
<tr>
<td>Craniofacial anomaly</td>
</tr>
<tr>
<td>Congenital infections: syphilis, TORCH</td>
</tr>
<tr>
<td>Premature birth, with prolonged intensive care</td>
</tr>
<tr>
<td>Meningitis</td>
</tr>
<tr>
<td>Ototoxic medicine(s)</td>
</tr>
<tr>
<td>Living in an underdeveloped country</td>
</tr>
</tbody>
</table>

REFERENCES


AUDITORY DEVELOPMENT
Joseph W. Hall, Ph.D.

Hearing loss at an early age, whether due to conductive or to sensorineural impairment, may result in effects that can be construed as being related to sound deprivation. Most previous research in this area has been aimed at the question of whether a purely conductive hearing loss will result in deprivation effects. Unfortunately, lesion techniques intended to cause specifically conductive losses may also result in sensory damage. Therefore, it has not always been clear whether neural effects found following experimental manipulations have been due to the attenuation of sound (intended conductive loss effect), or to the (unintended) damage to sensory elements. Recent studies on the chick and on the ferret have indicated that purely conductive deficits in these animals apparently do not result in neural abnormalities at levels corresponding to the cochlear nucleus, whereas sensory lesions do lead to relatively rapid neural changes at this level. The picture is markedly different when neural elements that receive binaural enervation are considered. Here, it appears that even discretely conductive loss can result in abnormal neural connectivity at neural levels associated with binaural interaction. It is possible that this occurs only when there is some degree of binaural asymmetry in the conductive loss.

Relatively few psychoacoustical investigations have been performed of hearing in humans who have suffered conductive hearing losses during the years of auditory development. In fact, only recently have there been systematic studies on the development of basic auditory abilities in children with normal otological histories. A general finding of interest from the relatively sparse latter work is that although some aspects of auditory function appear to be adult-like in the infant listener, many basic aspects of hearing (such as the ability to use binaural cues to hear in noise, or the ability to resolve the temporal features of a dynamic sound) appear to continue to develop over at least the first few years of life. Some auditory abilities appear to continue development over the first decade of life. The fact that some aspects of auditory development are relatively protracted in the human listener probably has important implications for the existence of a "critical period" for auditory development.

Recent psychoacoustic studies in our and other laboratories have indicated that the masking level difference (MLD) is usually reduced in children who have a long-term history of hearing loss due to otitis media with effusion (OME). Furthermore, the MLD often remains abnormally small in these children even after normal hearing thresholds have been restored through medical intervention. Similar results have been reported for children having a history of congenital atresia. One possible interpretation of these results is that poor binaural hearing may be due in part to effects related to auditory deprivation. Interestingly, the MLD is reduced primarily because the NoSo threshold is elevated, indicating a problem at or central to a site of binaural interaction. The processing of the signal in noise when no binaural cues were available (NoSo threshold) was as good in the children with OME history as in the children with normal hearing histories. This result would indicate no effect of deprivation with regard to monaural processes that probably rely primarily upon relatively peripheral auditory functions.

Several important questions need to be addressed in future investigations of the effect of hearing loss on auditory development. The first is the relation between the basic psychoacoustical finding for
binaural detection in noise and the "real world" problems in understanding speech that a child with OME history may experience in a noisy background. Now that a case has been made that auditory processing problems exist using well-defined and easily controlled (but highly artificial) stimuli, research is called for that uses stimuli much like those encountered in real environments. Second, the sparse data available indicate no effect of deprivation for relatively simple monaural tasks. It is important to determine whether deprivation-like effects exist for monaural processes that must rely on higher degrees of auditory processing complexity. Third, whereas most of the research examining the effect of auditory deprivation in humans has centered on conductive hearing losses, it is an egregious oversight to ignore consideration of sensorineural lesions. Physiological data indicate that the neural changes resulting from sensory lesions in young animals can be remarkable (although it is not clear what the changes may mean for auditory function). It is also unclear what response the central auditory system may have by way of accommodating a sensory loss and whether such a response is limited by age. These questions are crucial in developing rational plans for the habilitation of hearing in developing children.
It has been well documented that the majority of children with sensorineural hearing losses, even those of mild degrees, experience significant delays in English language acquisition. Lack of acoustic salience of linguistic cues and late identification of hearing loss are known sources of linguistic delay in this population. Historically, research on deaf children has focused on their weaknesses in sentence-level English syntax. Current theories demand that we avoid such an isolated view of language and broaden models of language learning to include such influences as interactional contexts, discourse, social knowledge, and culture. Perhaps reductionistic views of language have contributed to our limited success in making changes in the language abilities of hearing-impaired children. This paper will address ecological and pedagogical influences on language learning in children with hearing loss. These areas have the potential to greatly influence child language learning, yet have received little attention in the literature.

In recent years, child language specialists have stressed the importance of viewing the language learner as inseparable from the ecological system of the family. There is an increased appreciation for the transactional nature of communication and for the strong influence of family and social contexts on language learning. In contemporary thinking, children are seen not as passive recipients of input, but as active constructors of meaning, who influence their communicative partners and the environment around them. Examination of the language-learning needs of children with hearing loss from an ecological perspective points to challenges and future needs.

Familial Influences: Stress

Familial stress can have an impact on the nature of parent/child interactions and the consequent quality of the language-learning environment. The presence of a hearing-impaired child in hearing families is known to contribute to parental stress. White found that maternal language repertoires with deaf children often reflected the mother’s emotional status. Greenstein reported that affective aspects of mother/infant interaction were more highly correlated with child language development than were technical features of the language input. These and other studies underscore the importance of intervention efforts focused on family support and adjustment. Greenberg found more developmentally mature communication and higher quality interaction in families who had received early intervention services, such as those mandated by PL 99-457. However, a survey of 134 instructional programs for hearing-impaired children revealed a lack of readiness for programs to provide the comprehensive family-focused services mandated by this law. Family mental health issues impact a child’s language acquisition in ways we do not fully understand and have likely underestimated. Research related to family systems approaches is needed. It has taken many years for parent/infant intervention to shift from child-centered to family-focused approaches. We still do not have an adequate foundation in research to support the complete implementation of effective family-centered models of early intervention.
Issues of Caregiver Style

The literature suggests that hearing parents of deaf children do not always adhere to communicative behaviors known to facilitate language acquisition. Some desirable parental language behaviors include use of semantically contingent responses, responsive interpretation of early communication signals, and maternal use of expansions. Interactive behaviors of hearing mothers and deaf children have repeatedly been described as intrusive, directive, and overly controlling, as compared to mothers of hearing children. Reports have also identified less contingent responding, decreased positive affect and social play, and infrequent use of expansions. It is not fully understood to what extent these differences result from interactive influences (e.g., passivity on the part of the child) or parental view of their role as a language "teacher." Connard and Kantor found that quality and development of communicative interaction between normal-hearing mothers and hearing-impaired children is central to the success of early education programs. These findings address the need for attention to the quality of parent/child interaction in early intervention programs (which in some cases may be intact from the outset and require support rather than "fixing"). Hearing parents could benefit from modeling from deaf parents, who have been observed to be highly responsive in their interactions with their deaf children. We may have historically underestimated the impact of caregiver interactions on child language acquisition. In our experience, more impact on language learning is realized from parent-centered intervention efforts. Investigations of efficacious ways to support adult learners would be of value.

Quality of Language Input

The impact of degraded language input on language acquisition is not fully understood. Studies suggest that unlike parents of hearing children, hearing parents of hearing-impaired children may not uniformly increase syntactic complexity or length of input as the child ages. For some parents, this may relate to limited manual communication proficiency. The literature has also documented that hearing parents and teachers have considerable difficulty accurately using sign systems designed to represent English (MCE). Inconsistencies in signed input may contribute to language deficiencies in deaf children. Clearly, in families where only one person (e.g., often the mother) signs, the child has limited exposure to dialogue exchanges, and may struggle with pragmatic aspects of language acquisition. In fact, signing deaf children have been found to use the heuristic function much less than hearing counterparts. Family values are often passed along through conversational exchanges overheard by children. When families limit the interactive use of signing in the home, children are at risk for missing important modeling that teaches them how to use language in social contexts. Family members often lack support for learning to sign. Early and aggressive training in sign communication for entire families is needed. Research investigations are also needed to determine efficacious methods for family sign training. Parents’ language needs have often been ignored, even though we recognize that parental input is fundamental to the child’s acquisition process.

Degraded signed input may also have unique consequences for the learner in terms of language accessibility. Recently, researchers have suggested that MCE systems may be incompatible with the way visual/spatial languages are organized, and suggest that MCE systems are theoretically unlearnable. Schick and Moeller found that students exposed to MCE consistently at home and at
school acquired considerable facility with broad aspects of English structure and content. However, few subjects gained facility with the morphological system of English. This led the authors to question the practice of representing morphemes in the manual code. Schick stresses that hearing signers often include signed morphemes at the expense of natural visual prosody. Sign prosody and fluency may enhance accessibility of language meaning for the learner. We need a better understanding of the effects of sign fluency and prosody on language learning and accessibility.

Language Selection Issues

Problems with MCE systems and cultural issues have led many to advocate the use of American Sign Language (ASL) in educational programs. It is argued that a bilingual-bicultural approach, where ASL is the child's first language and English the second, would better serve the linguistic and sociocultural needs of the child. There is evidence from studies of deaf children of deaf parents that a bilingual approach can be highly effective and lead to English literacy attainment and enhanced cultural participation. The bilingual-bicultural viewpoint is gaining increased momentum nationally. Most professionals in deafness see the advantages of including ASL and Deaf cultural perspectives in educational programs. However, unanswered practical problems with implementation have contributed to polarization of viewpoints (ASL vs. English). We need to focus on understanding how ASL and English can be effectively combined in schools and homes to benefit deaf children. How will the communicative needs of hearing family members be addressed? What curriculum modifications will lead to acquisition of ASL, English, and literate language use? Solutions that include exposure to the most proficient signers (e.g., deaf adults) need to be explored. These are a few of the many unresolved issues facing professionals and parents today.

Pedagogical Practices

Educational practices for deaf children have often failed to keep pace with theoretical advances in language. For example, it is known that children with hearing losses are better able to learn vocabulary when a rich context is provided and new information is tied into networks of existing information. Yet, programs for deaf and hard-of-hearing children often approach vocabulary instruction in a decontextualized fashion. Regular education has implemented considerable reform based on current models of language and literacy attainment. Similar reform is needed in the management of children with hearing loss. Thinking and language connections, and expansion of world knowledge networks, need to be stressed throughout intervention efforts. In addition, hard-of-hearing children continue to receive fragmented programming and isolated rather than integrated teaching strategies. This problem demands our attention.

Efficacy Research

Problems raised in this paper need to be addressed through early and intensive intervention efforts. Efficacy research with deaf children reveals (1) linguistic and social/emotional advantages of early intervention, with more pronounced advantages in early vs. late starters; (2) distinct advantages of early efforts with multiply handicapped children; (3) increased program cost-effectiveness related to early intervention; and (4) reduced parental stress and improved parental communication in intervention families compared to non-intervention parents. Two studies failed to show lasting gains associated with early intervention efforts. As Guralnick points out, however,
many early intervention studies were based on "first-generation" programs, which were often child-centered rather than family-focused. This may account for contradictory efficacy results. Clearly, what is now needed are studies of the efficacy of "second-generation," family-focused programs, and a clear examination of the successfulness of professionals in establishing partnerships with parents in intervention. Particular attention must be given to affective issues and to relationship, input, and visual signal characteristics. A broadened cultural perspective is needed when examining issues related to the management of signing children.

REFERENCES


EFFECTS OF HEARING IMPAIRMENT ON
THE DEVELOPMENT OF SPEECH

D. Kimbrough Oller, Ph.D.

Speech Production

It is well known that children with severe or profound congenital hearing impairments have
speech handicaps by the second or third year of life. What is much less well known is that the effects
of hearing impairment are clearly present in the first year of life. The reason for the lack of awareness
of the effects of deafness on vocal development is complex. Some influential writings from the
1960's, in particular Lenneberg, Rebelsky, and Nichols, and Mavilya, had asserted that deaf and
hearing infants were notably similar in vocalizations of the first year. But these claims were based on
theoretically primitive evaluations of the vocal patterns of a very few infants. Mavilya's study, for
example, focused primarily on phonetic transcriptions of vocalizations in three deaf infants. The
method failed to note the differences between deaf and hearing infants, presumably because phonetic
transcription is insensitive to those differences. In fact, phonetic transcription is unable to capture the
primary developments in vocalizations of the first year of life.

In order to evaluate infant speech-like sound development, a new framework of study has
emerged. We term the framework “infraphonological” because instead of focusing on the concrete
phonological units of mature systems of speech (for example, segments such as [b], [p], [a], [u], etc.),
it focuses on the infrastructural properties of speech systems, the deep features of vocalization that
characterize well-formed utterance in natural language. For instance, instead of noting whether infants
produce more [k]'s or more [l]'s in the first 3 months of life, we note whether they progress from
unarticulated to articulated vocalization. Instead of focusing on the number of [pa]'s and the number
of [ma]'s produced at 7 months of age, we note whether the infant progresses from producing ill-
formed “marginal” syllables (articulated too slowly for purposes of speech), to producing well-formed
“canonical” syllables.

Having specified these infraphonological descriptors of early vocalizations, we have found that
hearing infants normally begin the canonical stage of well-formed syllable production at 5 to 7 months
of age. No normally developing child in three longitudinal studies at the University of Miami and the
University of Washington has shown an onset of canonical babbling later than 10 months of age.
Such information is garnered by a combination of parent interviews conducted by phone and
laboratory evaluations to confirm the parental descriptions. The laboratory work includes tape-
recorded samples of vocalizations that are categorized infraphonologically to assess the occurrence of
canonical and precanonical utterances quantitatively.

While hearing infants begin canonical babbling by not later than 10 months of age, deaf infants
appear never to begin canonical babbling that soon. From the various longitudinal studies, we now
have compiled onset of canonical babbling data on more than 70 hearing infants and 21 deaf ones.
The youngest age of onset in the deaf group is 11 months and the oldest is well beyond 30 months.
The lack of overlap in the two distributions suggests that the vocal distinction between deaf and
hearing infants is virtually complete. Canonical babbling is perhaps the most salient vocal event of the
first year of life, yet previous researchers failed to note its occurrence. Why? Because phonetic transcription (the primary method of traditional research) imposes the assumption of well-formedness on infant utterances, whether or not the infant utterances actually meet the infraphonological requirements of well-formedness. If one transcribes a 3-month-old’s utterance as [ba], it is naturally assumed that the syllable is well-formed, yet prior to the onset of the canonical stage there are only rare occurrences of canonical syllables. The phonetic transcription method begs the question regarding the extent to which infant sounds resemble mature speech, because it imposes a categorization system that presumes similarity. The infraphonological approach on the other hand opens the door to more insightful description, because it monitors the emergence of the system of speech-like control that will determine the child’s ability to produce speech-like sounds in general.

The significance of the discovery that deaf and hearing infants vocalize in starkly different ways goes well beyond scientific curiosity. Since parents appear to be able to recognize the onset of canonical babbling, it appears that a very cost-effective method of screening for profound hearing impairments might be based upon evaluation of infant vocalizations through a parent interview lasting only a few minutes and administered by a nurse or technician. The test would take place after the infant passes the age of 11 months. The proposed method’s potential is supported by the fact that the onset of canonical babbling appears to be very robust, with regard to risk factors such as low socioeconomic status (SES) or prematurity. We have evaluated substantial numbers of infants both in lower middle class and extreme poverty circumstances and have found that canonical babbling onset appears undisturbed. Similarly, healthy preterms from 1,500 to 2,000 grams birthweight show no delay in the onset of canonical babbling at corrected age. The results we have seen in vocal development of deaf and hearing infants also provide a basis for improved methods of intervention, since they suggest clear guidelines about training, guidelines that focus on infrastructural improvement rather than the teaching of speech segments implied by phonetic transcription.

Speech Perception

There has been virtually no research on speech perception in the deaf infant in the first year of life. In fact, the methods of evaluation of speech perception in infancy are very delicate and subject to problems of misinterpretation and bias even when working with hearing infants. There are three available methods. The first studies in infant speech perception were based on the monitoring of heart rate (HR) deceleration. In essence, in the HR method heart-rate changes are monitored after a change in a sound presented in a repeating background. Studies show that infants’ heart rate decelerates to changes in syllables. The second group of studies were based on the high-amplitude sucking (HAS) paradigm. In this paradigm an infant sucks on a pacifier connected to a pressure transducer. Sound is presented contingent on the sucking; increases in sucking rate in response to change in sound are taken as indications of discrimination. The third paradigm is called visually reinforced infant speech discrimination (VRISD) and was developed by Eilers, Wilson, and Moore. In this paradigm an infants’ head turns in response to changes in a repeating background signal are monitored, as well as responses during control intervals when the sound does not change. A predominance of turning during change trials is taken to indicate discrimination.

The three paradigms have differing applications (HR and HAS are applicable in early infancy, whereas VRISD is applicable after 6 months of age) and differing constraints of interpretation. All are subject to important limitations. For example, the HR and HAS paradigms are, for practical purposes,
inapplicable in single-subject designs—one must compare groups of experimental subjects (those that are exposed to a change in sound) with groups of controls (those that hear no change). VRISD can be used with individual subjects, because multiple experimental and control trials are presented to each subject; however, this technique requires the child to be able to turn toward sound, which is uncommon prior to 6 months of age. All three paradigms are delicate to administer and can be affected by subtle biases of the experimenter. While it is hard to gauge the relative reliability of infant speech perception and adult speech perception tasks, it seems certain that the adult tasks yield much greater clarity of results and are much less vulnerable to experimental error. Given these facts, it appears that speech perception evaluation in the hearing-impaired infant will be slow in coming.

Acknowledgment

This research is supported by NIH NIDCD grant 5-R01-DC00484 to D.K. Oiler.

REFERENCES


The primary effect of hearing loss is that children are denied full access to their own speech and the speech of others, thus impeding the acquisition of spoken language. The severity and permanence of the hearing loss and resulting language delays determine the extent to which social/emotional and academic areas are affected. The consequences of hearing loss are most obvious for children with severe and profound hearing loss, who account for only a small percentage of hearing-impaired children. The majority of children with hearing impairments exhibit the less visible sequelae associated with mild/moderate, unilateral, or conductive hearing loss. These infants and children may display social and academic deficits if their hearing losses remain unidentifiable throughout the language-learning years.

Social Adjustment

Hearing-impaired children develop positive social behaviors through the same process as do normal hearing children. Their interactions are shaped in the early years through parental reaction and in the school years from peers and teachers. Although an auditory impairment makes the social development process substantially more difficult, special early intervention can result in normal social adjustment for the hearing-impaired child.

Severe/Profound Hearing Loss

There is nothing inherent in hearing impairment that causes social adjustment problems in severely hearing-impaired children. It is the reaction of the family and poor communication development that contribute to the reported self-esteem and social adjustment problems. Social immaturity begins to be apparent during the preschool years in the form of impulsivity, provocative behavior, physical aggressiveness, and frequent requests for adult approval. This immaturity appears to increase with age as greater communicative demands are made on the students. Without facility with spoken language, these children depend on the good will of adults, both teachers and parents, to interpret, rephrase, and verbally mediate when necessary.

Data suggest that children educated in self-contained residential programs evidence the highest levels of self-esteem. It is unclear, however, if this is due to the overprotectiveness of the educational environment. Reich, Hambleton, and Houldin in comparing a variety of mainstream settings found that being educated with normal hearing classmates exaggerated the student’s differences instead of diminishing them. When the mainstream experience is structured and supported, social adjustment issues abate. Mainstreamed students, in general, demonstrate earlier maturity and independence relative to their self-contained classmates.

Social adjustment and self-esteem have their roots in parent-child interaction. Most hearing-impaired children are born to hearing parents. These adults bring to the child-rearing process the expectation that parent-child interaction will rely on auditory language-based input. These uninformed
parents find themselves in the position of adapting to the non-auditory style required to comfort, stimulate, and calm the deaf infant. If the parents are unaware of the hearing loss, then they may misinterpret their infant's intent. These infants display blank affect while concentrating on parent's face or avert eye gaze from their interactive partner, toward the topic item, and then back to the communicator. On the surface, this behavior, which is the result of the infant visually ordering the world, appears to be rejection and social nonresponsiveness to the parent. Effective communication, i.e., accurately reading each other's signals, is the vehicle for transmitting positive feelings of self and worth.

Parental interaction style can contribute to mixed messages and negative self-esteem. Parents of deaf children exhibit a more controlling approach to verbal interaction than exists with normal hearing children. This control response is most obvious in (1) excessive and extensive adult turns in conversation (filling all gaps with narration); (2) increased use of imperatives and questions; and (3) adult-dominated topic initiations. Hearing mothers of deaf children often fail to follow their child's visual topic indicators and provide contingent input that is timed correctly, both factors found to facilitate language growth in normal infants. Regardless of the communication mode that the parents choose to employ with their infant, they enter a prolonged period of self-conscious interaction that alters the natural style that previously existed. Thus, the building blocks of self-esteem and social adjustment are disrupted by the self-conscious behavior.

Unilateral Hearing Loss

Children with unilateral hearing loss portray inconsistent behavior patterns due to the difficulty they have hearing in noisy environments. Teachers rated them as having greater difficulty in peer relationships, less social confidence, greater acting out or withdrawal, greater frustration, increased dependence on teacher, and greater distractibility in the learning environment.

Mild/Moderate Hearing Loss

There are little data on the social adjustment of children with mild/moderate hearing loss. Since they are frequently the only hearing-impaired student in the school, it is not unusual for them to feel totally isolated, not fitting into either the deaf or the hearing world. Often personal and classroom amplification have been recommended but are not used, as they try to eliminate any physical signs of "difference." These are children who never quite meet their parents' and teachers' expectations, because their normal appearance belies their actual performance.

Conductive Fluctuating Hearing Loss

Fluctuating conductive hearing loss results in inconsistent behavioral patterns that are often misunderstood by adults and teachers. This child is able to perform at expected levels one day but is unable to on the next due to the presence of a hearing loss.
Academic Consequences

While the majority of research into the academic consequences of hearing loss has focused on severe and profoundly hearing-impaired students, there is evidence that children with unilateral, mild/moderate, and conductive hearing loss experience educationally handicapping delays.

Severe/Profound Hearing Loss

For severe and profoundly impaired students, estimates of reading levels vary from third to seventh grade at the completion of high school, irrespective of communication system, SES, parents hearing status, and educational setting.4,5 The best predictor of educational attainment is English language knowledge, with vocabulary being a stronger influence than syntax. Nonlinguistic math attainments are higher and evidenced growth with age, but using present-day teaching techniques, do not reach levels of normal hearing peers.6 The highest achievements were noted for students who were educated with normal hearing classmates beginning from 3 to 6 years of age.7 However, the fact that these students had better hearing than students who were mainstreamed at a later time, contributed to their elevated achievement levels.

Unilateral Hearing Loss

Students with unilateral hearing loss may evidence learning deficits far in excess of what would be expected with one normal ear. In studying children with unilateral hearing loss of 45 dB or more in the affected ear, Bess found that 35 percent failed one or more grades, with 13 percent requiring support services.8 Factors such as early onset of the hearing loss, birth complications, loss of 61 dB or greater, and right ear impairment were characteristic of unilaterally impaired children who exhibited educational problems.

Mild/Moderate Hearing Loss

Mild/moderate bilateral permanent hearing loss produces many of the same education problems exhibited by the severely impaired children, but the degree of the problem is less. It is not unusual for these children to exhibit mild delays in reading and in language-based subjects resulting from communication deficits. Blair et al. reported on the educational attainments of a group of children whose better ear hearing losses were 40 dB or less and a control group of matched normal hearing peers.9 In reading and math, the mildly hearing impaired students performed at grade level for each of the 4 years tested, but always performed significantly below their matched peers, with the discrepancy enlarging over the 4 years. These children are described as underachieving relative to their potential.

Conductive Fluctuating Hearing Loss

Some children with early fluctuating conductive hearing loss demonstrate a deficit in attention for language-related tasks and selective listening factors related to success in the educational environment. Because the development of reading skills requires mastery of auditory-verbal code, it is not surprising that reading, which is the basis for language-based activities, is affected by this early fluctuating disruption in the auditory signal. When conductive hearing loss, which has increased rates of occurrence in lower SES areas, is coupled with the decreased levels of language stimulation in this
environment, the children are at greater risk for related academic delays. Kessler and Randolph found that 55 percent of a group of third-grade children with histories of middle ear disease before the age of 3, received speech/language, reading, or learning support, in contrast to 31 percent for a control group. The early onset of middle ear disease encouraged inefficient listening strategies which continued to affect school performance after the active ear disease had diminished.

Conclusion

Early identification and intervention can prevent the social/emotional and academic consequences of mild/moderate, unilateral, and fluctuating conductive hearing loss and reduce the enormity of the deficits in more severely hearing-impaired children. Parent education can replace detrimental interaction patterns with those that facilitate effective communication between parent and infant, resulting in positive self-esteem and social adjustment. By recognizing the existence of hearing loss during the early months of life, auditory skill development can progress at normal or near normal rates for the milder degrees of loss, thus reducing the communication deficits which translate into academic delays. For severely hearing-impaired children, early identification and aggressive intervention increase the possibility of age-appropriate language and academic achievement, and early mainstreaming.

REFERENCES


BENEFITS OF SCREENING AT BIRTH: ECONOMIC, EDUCATIONAL, AND FUNCTIONAL FACTORS

Marion P. Downs, M.A., D.H.S.

Economic Data

The 350,000 profoundly deaf in this country earn 30 percent less than the general population, resulting in an annual income loss of $2.5 billion (present value). In addition, there are 21,000,000 people with lesser hearing losses that result in some degree of reduced income. Exact figures are not available, but as little as a 10 or 15 percent reduction in the income of this larger group would cost society another $76.5 billion annually.

Deaf people who had normal hearing until 3 years of age earn 5 percent more than those born deaf, because of the language skills acquired in the first 3 years. Identification at birth and immediate intervention for the congenitally deaf would result in some approximation of the language skills of those with onset of deafness at age 3. The result would be a marked improvement in earned income, approaching $129 million per year for the profoundly deaf group.

The estimated total cost to society from deafness and hearing impairments stated above—$79 billion per year—might be reduced by 5 percent through newborn identification, that is, up to $3.9 billion per year.

The cost of educating the 350,000 deaf people in schools for the deaf is $29,000 per person per year, ($348,000 each for 12 years of schooling) bringing the total cost of educating all presently deaf people to $121.8 billion. No estimate is available on the considerable costs of educating the 59,312 hearing-impaired students currently enrolled in the special education programs of the public schools.

Northern and Downs have itemized the lifetime costs of deafness in an individual to be $1,021,170. Early identification resulting in improved language skills would probably reduce that figure by some part of the 5 percent cited above. Court judgments have been awarded for nearly $3 million for deafness incurred through hospital negligence.

Educational Data

A fourth grade equivalency language level still persists for the deaf population. Yet children with any degree of hearing loss can be expected to have reduced academic achievement. Levitt et al. found that children with milder losses down to 40 dB had reduced language skills similar to those of children with losses up to 110 dB—a nonlinear relationship of language to hearing loss. Levitt showed that a direct linear relationship exists between speech intelligibility and hearing loss, confirming that speech is an “overlaid function,” whereas language is a biobehavioral system with a period of major plasticity that comes to an end early in life.
Hearing losses as mild as 20 dB from otitis media (OM) impair language skills significantly, and young children do not "grow out" of such language delays. Accurate testing for even mild losses in the infant population is requisite.

Levitt et al. reported that the earlier the intervention, the better were the language skills and academic achievement. He found "strong quantitative evidence of the association between superior speech and language skills and early intervention." Similar conclusions were reported by the Utah SKI*HI project.

A Lexington School for the Deaf study compared infants admitted to the program before and after 16 months of age, finding that the earlier-admitted children were statistically superior in language and in parent-infant communication. Parental bonding was stronger for the earlier-admitted children.

These studies indicate that (1) early identification permits improved language skills to develop, (2) later identification results in poorer language skills and educational performance, and (3) bonding and effective interpersonal communication develop best when the hearing loss is identified as early as possible.

Theoretical Considerations

Ruben and Rapin state that input from the peripheral auditory system is necessary for the maturation and innervation of portions of the central auditory system (CANS). The ability to hear environmental sounds has the greatest effect in shaping auditory ability from the time the inner ear becomes functional (fifth gestational month) to the time when maturation of the CANS is completed (18 to 28 months).

Animal studies document the plasticity of the CNS, which can be modified by experiential deprivation of acoustic and other sensory stimuli. Webster and Webster's studies on the defects of the auditory nuclei in mice that were given conductive losses have been supplemented by their later studies on higher animals. These researchers found that in animals whose hearing is functional in utero (like humans), conductive losses do not affect the CANS. They thus demonstrated the powerful influence of early auditory stimulation beginning in fetal life, where hearing is fully developed by the fifth month of gestation.

Human research has been conducted with auditory brainstem response (ABR) techniques. Dobie and Berlin, using ABR to measure neural activity associated with hearing, reported absent binaural interaction in a child with early recurrent OM. Finitzo-Heber corroborated this study, finding no binaural interaction in 60 percent of children with early OM history, compared with 10 percent absence in children with no OM history. She also found a lack of clear middle latency potentials in the affected children.

Conclusions

Any alternative to universal newborn hearing screening is unacceptable, as very few physicians screen young children for hearing. A dependence on parental report has resulted in identification too late for adequate language skills to develop. Unless universal newborn hearing screening is
instituted, we will continue to see academic performances well below the hearing-impaired students’ potential for achievement and for the best possible quality of life.

Research Goals

1. Information on the number of deaf and hearing-impaired, on the costs of educating the hearing-impaired in the public schools, and on the psychosocial factors in these groups.

2. Basic research on the neurophysiology of auditory deprivations and of efferent development in the CANS.

REFERENCES


3. Center on Deafness, Denver. Personal communication.


In 1969, the Joint Committee on Infant Hearing (JCIH)—composed of representatives from otolaryngology, pediatrics, nursing, and audiology—was established and charged with responsibility for making recommendations concerning newborn screening programs. The JCIH, recognizing the problems associated with universal screening, did not support the concept of mass hearing screening for all newborns. The JCIH instead endorsed the concept of a high-risk register (HRR) for selecting infants who should receive hearing assessment. Initially, five factors were identified as placing an infant at increased risk for hearing loss. The JCIH revised and expanded the high-risk criteria in 1982, and more recently in 1990, to include the 10 risk factors shown in Table 1.

<table>
<thead>
<tr>
<th>Year</th>
<th>Risk Factors for Hearing Deficit</th>
</tr>
</thead>
</table>
| 1972 | 1. Family history  
      | 2. Hyperbilirubinemia requiring exchange  
      | 3. Congenital infections (TORCH)  
      | 4. Craniofacial anomalies (Defects)  
      | 5. Birth weight <1500 grams |
| 1982 | 6. Bacterial meningitis  
      | 7. Asphyxia  
      | Apgar score ≤3 at 5 minutes |
| 1990 | 8. Ototoxic medication  
      | 9. Mechanical ventilation ≥10 days  
      | 10. Syndromes that include hearing loss |

The HRR has been used for almost 20 years and continues to be recommended as an integral component in early identification protocols (EIdP). Despite the use of this tool in two different applications (hospital based and birth certificate based), there is general agreement that the HRR identifies only about 50 percent of infants with sensorineural hearing loss. If we are to continue to recommend the application of the HRR, we must address two fundamental questions: (1) How well do the individual risk factors predict hearing loss? and (2) How often do children with sensorineural hearing loss exhibit risk factors?
How Well Do the Individual Risk Factors Predict Hearing Loss?

Several studies have attempted to evaluate risk factors. Meaningful comparisons among these studies are difficult because of differences in the definitions of risk factors, as well as in the criteria for determining if an infant has a risk factor. In addition, many of these studies provide questionable data because of the manner in which infants were selected (i.e., sampling method, sample bias, sample size) and subsequently followed (see Turner and Cone-Wesson). Therefore, accurate information on rate of positive identification and false-positive and false-negative rates cannot be calculated. Unfortunately, even after 20 years of HRR use, the data are insufficient for adequately evaluating the individual risk factors.

How Often Do Children With Sensorineural Hearing Loss Exhibit Risk Factors?

In 1983, Stein, Clark, and Kraus, reported that nearly one-third of hearing-impaired infants residing in a well-baby nursery (WBN) would not have been detected through the 1982 HRR. Pappas found that only 46 percent of infants would have been identified using the 1982 HRR. In other words, 54 percent of the infants were not suspected of being at risk for hearing loss based on the 1982 criteria and thus were not referred for hearing evaluation during the neonatal period. Elssmann, Matkin, and Sabo also reported that of the subjects with congenital hearing impairment in their study, only 48 percent had high-risk factors for hearing loss.

The variable identification rate of the 1982 HRR in a WBN or intensive care unit (ICU) reveals an unacceptably high percentage of infants with hearing loss who are not identified and thus escape systematic followup for congenital or delayed-onset hearing loss. On the basis of the fundamental questions concerning HRR, two practical questions must be posed: (1) Does the HRR facilitate the recommendations of the 1990 Joint Committee on Infant Hearing Position Statement? and (2) Is there a need for a HRR or the “high-risk classification” protocol?

Does the HRR Facilitate the Recommendations of the 1990 Joint Committee on Infant Hearing Position Statement?

In 1982, and again in 1990, the JCIH recommended that, whenever possible, diagnostic testing should be completed and habilitation begun by the time an infant with a congenital impairment reaches the age of 6 months. Unfortunately, for nearly all hearing-impaired infants, the age of diagnosis and the resulting enrollment in a habilitation program far exceeds the Joint Committee’s recommendations. These findings suggest that individuals should be skeptical about the efficacy of the HRR in facilitating the goals of the JCIH. While aggressive followup is a major complicating factor in ELPD regardless of screening methodology, the first objective of screening must be to accurately identify those infants who must be followed. The HRR has not yet demonstrated the accuracy that is necessary if followup is to be carried out in accordance with the JCIH recommendations, since the false-positive and false-negative rates are high.

Is There a Need for a HRR or the “High-Risk Classification” Protocol?

There is some evidence that the ability to identify and diagnose hearing loss at an early age has been improved through the use of birth certificate-based high-risk registries.
and Behrens (1991) found that parents of high-risk children, on average, suspect a problem approximately 5 months earlier and obtain a hearing test approximately 7 months earlier than parents of children with no risk factors for hearing loss. These results provide evidence that a HRR can be of substantial assistance in the early identification of hearing loss.

The 1990 JCIH statement presents a two-tier scheme that may counter some of the problems associated with the 1982 single-tier scheme. When a two-tier scheme is used throughout a child's first 2 years of life, earlier identification of delayed-onset and acquired hearing loss may be facilitated. The two-tier system allows children to remain in a "guarded" classification through age 2, until a hearing loss has been ruled out. The application of the 1990 HRR would remind pediatricians, primary care providers, and public health nurses about the association of risk factors with hearing loss in infants, and the importance of early referral for hearing evaluation.

Neonatal hearing screening programs, however, that rely on the HRR as the first filter for identifying all newborns in need of further assessment will fall short of the objective of identifying all significant hearing losses before 6 months of age. This leads the author to conclude that the most effective way to comply with the JCIH goals is to implement valid, reliable, and cost-effective screening measures that can be used to evaluate newborns from both WBN's and ICU's before hospital discharge.

REFERENCES


BEHAVIORAL MEASURES

Judith S. Gravel, Ph.D.

Prior to the advent of the auditory brainstem response (ABR) screen, behavioral screening was the only option available to clinicians interested in identifying neonates at risk for hearing loss. Behavioral methods consisted of the use of behavioral observation techniques completed in the nursery. A calibrated stimulus source consisting of a suprathreshold signal, weighted in the higher frequency regions, was used. The neonate was screened in a quiet, light-sleep state. Generally, two or more observers (who were aware of the stimulus presentation) judged the presence of a response to the signal based on an observable change of state. Problems with such procedures included (1) maintaining the state of the infant, (2) biased observations, (3) high intensity of the screening stimulus (precluding the identification of mild to moderate degrees of hearing loss), (4) inability to obtain ear- or frequency-specific responses, (5) the need for more than one trained observer, and (6) the habituation of response behaviors to repeated stimulus trials, precluding the examination of response reliability.

The introduction of automated behavioral screeners was designed to overcome some of the limitations of earlier subjective observation techniques. Devices such as the Crib-O-Gram eliminated the need for trained observers and thus the bias imposed by the subjective nature of their observations. The infant was placed on a mattress under which was a motion-sensor monitoring device that recorded the infant's movement. According to preset criteria, when the neonate was sufficiently quiet, a suprathreshold stimulus was presented, and any motion made by the infant that occurred during or just after signal presentation was considered a response. The neonate was maintained in the screening device until a sufficient number of trials had been collected. Disadvantages of the automated procedures once again included the use of suprathreshold stimuli and the excessive length of time required for some infants to complete the procedure.

In 1985, Durieux-Smith and her colleagues examined the Crib-O-Gram and ABR screening methods in the same infants. More than 300 NICU babies were examined using both procedures. Briefly, approximately one-half of the infants who passed the ABR screen in at least one ear failed the Crib-O-Gram, while 30 percent of newborns with moderate hearing losses by ABR passed the Crib-O-Gram screening. Moreover, when the reliability of the automated behavioral procedure was examined (the same infants examined on two separate occasions), 32 percent changed categories (pass to fail or vice versa).

This report, as well as those of others, demonstrated that the Crib-O-Gram and other neonatal behavioral screening procedures had high false-positive as well as unacceptably high false-negative rates. Thus, behavioral techniques are not presently considered to be efficient or effective procedures for use in neonatal hearing screening programs.

Recently, Werner and her colleagues developed the Observer-Based Psychoacoustic Procedure (OPP). This method has provided a bias-free means for the examination of the development of auditory function (sensitivity; frequency and temporal resolution) in infants (as young as 2 to 5 weeks through 6 months of age). The computer-assisted method requires a highly trained observer who
judges whether or not a signal is present during a trial interval by observing the baby. The observer is unaware of whether the trial was a signal or control (no signal) trial. Using signal detection theory, the presence of a response is determined based on the percentage of correct judgments by the observer. Bernstein and Gravel have reported on the use of a similar bias-free observation procedure with neonates. Measures are completed in the nursery in an acoustically treated enclosure (the NEST). The procedure, however, was designed to examine an infant’s overall auditory responsiveness (peripheral as well as central function). Presently, these newer, observer-based procedures are not being promoted (nor are they currently well suited) for use as neonatal hearing screening methods.

With older infants and toddlers (beginning at approximately 5 to 6 months of age and through 2 to 2.5 years), visual reinforcement audiometry (VRA) is a reliable and valid technique for use in infant hearing screening programs and can be used to identify infants with various degrees and configurations of hearing loss. A response (a head-turn) is reinforced (by the activation of a highly animated toy) only when the behavior occurs in the presence of a test signal. VRA utilizes operant conditioning techniques that are developmentally appropriate for this age group. Recently, a computer-assisted screening procedure (the CAST; Classification of Audiograms by Sequential Testing) has been developed, which uses the VRA procedure. The CAST shows great promise as an efficient and effective screening tool.

While some form of the VRA procedure (either manual or automated) is available in most clinical audiology facilities, its application as a screening tool is currently restricted to “at-risk” infants referred to such sites for screening. Behavioral screening programs that use the conditioned head-turn procedure have thus far been limited, and currently, no equipment that is efficient and cost-effective is available for routine use in day-care facilities, preschool programs, or physicians’ offices. The operant head-turn procedure is seen as a potentially useful tool because it is efficient, reliable, pleasant, and provides information on auditory function even in the presence of middle ear pathology.

For preschoolers (3 to 5 years of age), conditioned play audiometry is the procedure of choice for use in hearing screening programs. Similar to techniques used with older children, the procedure requires that the young child respond to the presence of the signal with a manual (“play”) response, such as dropping a block or stacking a ring. The method can be used readily with this age range and requires only a portable audiometer and a trained examiner to complete the procedure. Screening levels (ASHA, 1985) applicable to older children have been successfully used with this population.

Although reliable and valid behavioral screening methods are available for use in infants and young children in the 6-month to 5-year age range, there are practical difficulties that have limited their widespread use with these groups. Briefly, the problems include (1) access to the populations requiring screening; (2) lack of inexpensive and highly reliable screening equipment that can be easily operated, maintained, and transported; (3) the training of personnel; and (4) the availability of and access to audiological/medical assessment and followup.
REFERENCES


ACOUSTIC IMMITTANCE MEASURES

Robert H. Margolis, Ph.D.

Acoustic immittance measures have three potential applications that may contribute to the success of an infant hearing screening program. First, they may be useful for detecting middle ear pathology, a condition important to detect because of its potential contribution to hearing loss or because the disease itself requires intervention. Second, they may be useful for detecting conditions of the external or middle ear that may affect the outcomes of other tests. These include material in the newborn middle ear (e.g., amniotic fluid, effusion, and unresolved mesenchyme), ear canal collapse, ear canal debris such as vernex caseosa, and middle ear air pressure. Third, acoustic immittance measures, specifically the acoustic stapedius reflex (ASR), may be useful for screening for hearing impairment.

Detection of Middle Ear Pathology

In older children and adults, acoustic immittance measures are used routinely in the evaluation of middle ear function. Although there are exceptions, middle ear disease has predictable effects on tympanometry and acoustic reflex measures, providing information that contributes to the diagnosis. In young infants the situation is less clear. Tympanograms recorded from healthy newborn ears differ in many important respects from those of older children and adults. In addition, there are reports of normal tympanograms in the presence of otoscopically confirmed otitis media in infants. To understand these findings, it is necessary to consider the tympanometric characteristics of normal adult ears, how tympanograms of normal infants differ from adults, and how these characteristics are altered by middle ear pathology.

Perhaps second only to Terkildsen and Thomsen's first use of tympanometry for the clinical evaluation of the middle ear, the most important contribution to understanding aural acoustic immittance measures is the Vanhuyse model (see Hunter and Margolis for a discussion). The model predicts an orderly sequence of tympanometric patterns as the frequency of the probe tone is altered and provides a framework for understanding the effects of pathology on middle ear mechanics. Using the conventional low-frequency probe tone (226 Hz), normal adult tympanograms are always single-peaked. With increasing frequency, patterns become increasingly complex.

The first reports of tympanograms obtained from infant ears produced evidence that the newborn middle ear is fundamentally different from those of older children and adults. Using a low probe frequency, tympanograms from newborns were frequently double-peaked. Using a more analytical technique, Himelfarb et al. demonstrated that the newborn ear is characterized by a high acoustic resistance and a low acoustic reactance, the inverse of the resistance/reactance relationship seen in older children and adults.

The most detailed study of the physical characteristics of the newborn ear was reported by Holte et al. They recorded multifrequency tympanograms in healthy infants longitudinally over the first 4 months of life. The newborn ear appeared to be characterized by two resonant frequencies, one in the vicinity of 450 Hz and another around 710 Hz. The presence of the low-frequency resonance is consistent with mass loading of the middle ear, perhaps due to amniotic fluid, effusion, or unresolved
middle ear mesenchyme, which have been observed in infant temporal bones. This may also account for the occurrence of double-peaked, low-frequency tympanograms in newborns. By 4 months of age, the infant tympanograms were similar to those of adults, conforming to the Vanhuyse model, although the resonant frequency is probably slightly higher than in adults.

If newborn tympanometric characteristics are dominated by the presence of material in the middle ear, it may be difficult to distinguish between this condition and middle ear disease. On the other hand, there is evidence that the use of probe frequencies higher than the conventional 226 Hz may be effective in detecting middle ear disease in newborns. Marchant et al., Hirsch et al., and Hunter and Margolis reported cases of infants with apparently normal 226 Hz tympanograms, flat tympanograms obtained with a higher probe frequency, and otoscopically confirmed middle ear effusion. A prospective study, with careful otoscopic examinations, is badly needed to determine the value of tympanometry for detecting middle ear disease in newborns.

Detection of Conditions That May Affect the Outcomes of Other Tests

Abnormal conditions of the external or middle ear may affect the outcomes of hearing tests either by producing a hearing loss or by affecting the ability to record a response. These conditions include material in the middle ear, ear canal collapse, ear canal debris, and middle ear pressure.

Since otoacoustic emission (OAE) techniques require transmission of the response through the middle ear, these methods may be influenced by material in the middle ear. The effects of unresolved middle ear mesenchyme or other material that may be in the newborn middle ear on OAE recordings are unknown. Since surgical exploration of the middle ear is seldom an option, the most feasible method for studying this may be the determination of the relationship between tympanometric recordings and OAE’s.

Because the osseous ear canal wall is incompletely developed in the newborn, ear canal collapse frequently occurs. This condition can be detected by tympanometry. Since the volume of air in the ear canal contributes to the measured acoustic admittance, a small volume is indicative of ear canal collapse. This can often be avoided through gentle distention of the ear canal by pulling backward in the pinna, and through the use of insert earphones.

The newborn ear canal usually contains vernix caseosa, which may influence the outcome of other tests, particularly OAE’s. This, along with material in the middle ear and ear canal collapse, may be largely responsible for the high failure rates being reported in neonatal OAE screening programs.

Middle ear pressure, a frequent occurrence in children, also influences OAE measurements, probably due to its effect on middle ear mechanics. Naeve et al. recorded transient-evoked OAE’s in normal adult ears with varying ear canal air pressures. OAE amplitudes were diminished and their spectra were altered by positive or negative ear canal air pressures. They suggested that middle ear pressure may have a similar effect of reducing OAE amplitudes. Trine et al. recorded transient-evoked OAE’s from patients with tympanometrically detected middle ear pressures. OAE’s were recorded with ambient pressure in the ear canal and the pressure necessary to compensate for the middle ear pressure. In all cases, compensating for middle ear pressure increased OAE amplitude. These studies
suggest that the presence of middle ear pressure may result in false-positive screening outcomes using OAE as the screening measure and that compensating for middle ear pressure may reduce the occurrence of false positives.

Detection of Sensorineural Hearing Loss

Acoustic immittance measures, specifically the acoustic stapedius reflex (ASR), may be useful as a screening method for sensorineural hearing loss (SNHL). Several methods have been reported for detecting hearing loss by ASR measurements. Most of these methods are based on the differential effect of SNHL on ASR thresholds elicited by tonal and noise stimuli. Although these methods and their outcomes are strongly influenced by recording technique, there is evidence that ASR may provide a basis for an effective, nonbehavioral screening test for hearing loss.18

REFERENCES


DEVELOPMENTAL SCREENING IN A BUSY CLINIC: 
A LUXURY OR A NECESSITY?

Noel D. Matkin, Ph.D.

A basic premise relative to the pediatric audiolologic assessment of young children at risk for hearing impairment is that each evaluation should be organized so that the findings not only lead to definitive classification of the child's hearing status but also facilitate development of an intervention plan. This premise implies that the assessment protocol should be based on a developmental, rather than a medical, model. Far too often, the focus of the assessment is on the function of the ear rather than on the function of the whole child. In keeping with the two Federal mandates, Public Laws 94-142 and 99-457, intervention should be based on knowledge of the child's present level of function in the major developmental domains, and the family should be viewed as partners during the evaluation. The validity of these concepts as embedded in the two Federal mandates is apparent, yet the time and cost constraints in a busy clinical practice are major obstacles to taking a developmental stance during assessment. My goal is to describe the conceptual framework and the mechanism for a holistic approach to the evaluation of young children at risk for hearing loss. This protocol was developed and refined in the pediatric hearing clinics at the University of Arizona.

In 1953, Myklebust proposed a very simple yet comprehensive developmental model, which has been easy to incorporate into a clinical practice. He posited that all children, including those with hearing loss, grow and develop in four key areas: cognition, language competence, motor abilities, and social/emotional skills. Furthermore, he recommended that each of these developmental areas be considered during the case history, during observation of the child's behavior, and during formal testing.

Regardless of the assessment protocol or the developmental model one utilizes, two key questions need to be answered when the presence of either conductive or sensorineural hearing loss in a young child is confirmed. First, to what extent has the hearing loss resulted in a lag in language acquisition and the development of functional communication skills? And, second, does the child's developmental profile of strengths and limitations suggest that the limitations are related directly to a hearing loss, or to the presence of additional developmental disabilities? Answers to both questions are of critical importance as a comprehensive plan for intervention is formulated.

Before describing a specific clinical protocol in which a developmental approach is taken, it is worthwhile to consider the implications embedded in each of the preceding two questions. First, to what extent has the hearing loss resulted in a delay in speech-language acquisition? Recognizing the magnitude of a child's speech-language delay at the outset fosters more appropriate decisions relative to the need for amplification and specialized preschool education. A number of studies over the past three decades have revealed a relationship between magnitude of the impairment and extent of the speech-language delay. As the magnitude of the hearing loss increases, the richness of daily auditory input is reduced and, as a result, both language comprehension and verbal expression increasingly are delayed. A careful analysis of these studies reveals a wide dispersion of findings among children with similar degrees of hearing loss. In other words, grouping children by magnitude of hearing loss does not assure homogeneity relative to their language competence and verbal communication skills. There
are numerous variables—in addition to hearing loss—that are associated with the rate of a child’s acquisition of speech and language. Thus one cannot validly predict a particular child’s language function on the basis of group data.

Another issue that merits consideration, and which is supported by a growing body of literature, is that educationally significant language delays are characteristic of many children with minimal hearing loss. The term minimal hearing loss is used in this context to include three types of loss: very mild bilateral sensorineural impairments limited to the higher test frequencies, permanent unilateral impairments, and relatively long-term conductive impairments associated with early recurrent otitis media. As an aside, these three groups of children account for at least 80 percent of the referrals to our children’s hearing clinics. In all three instances, the presence of a speech-language delay should be confirmed or ruled out at the time of initial identification. The traditional reactive stance reflected by such advice as “Wait and see if he outgrows the speech problem” or “Let’s see if the loss causes problems in school” is not defensible. Two relatively new visual aids effectively highlight the effect of different degrees of hearing loss on detection of the various components of the speech spectrum. With high-frequency hearing losses, the use of the dot matrix is revealing. In contrast, the use of the A, B, C contours with those cases having relatively flat hearing loss readily reveals the potential impact of so-called mild impairments.

The increasing prevalence of children with one or more developmental disabilities in addition to hearing loss is the second reason that developmental screening at the earliest possible time is imperative. A decade ago it was determined that on the average one of every three children with congenital sensorineural hearing impairment had one or more additional developmental disabilities. Identifying the presence of such multiple developmental disabilities facilitates appropriate referrals for additional assessment. Such multidisciplinary evaluations result in a remedial plan that not only considers the child’s hearing loss but also deals with major concomitant developmental problems.

At intake, the parents are asked to complete a medical and a developmental questionnaire. The primary caregiver’s ratings of the child’s development on these questionnaires serve to focus our inquiry during Phase 2—the case history. Those behaviors rated as fair or poor are thoroughly discussed with the parents in order to verify and expand upon their concerns. If two or more developmental areas are reportedly delayed, we then shift into Phase 3. During this third and final phase, the Minnesota Child Development Inventory (MCDI) is completed by the family or the legal guardian.

The MCDI screening instrument was selected for daily use in our clinics for several reasons. First, use of a developmental questionnaire signals that we value the parents’ input and view them as partners in the evaluation process. Second, the MCDI is most appropriate for young children between the ages of 18 months and 5 years—an age span in which the vast majority of referrals to our pediatric audioligic facility are initiated. Third, the visual profile plotted from the parents’ yes/no responses on 320 questions facilitates immediate feedback to the family relative to the child’s strengths and limitations; at the same time, the rationale for recommending additional assessments becomes readily apparent. Finally, investigations of this screening tool have revealed very acceptable concurrent and predictive validity. The best news is that a review of the intake questionnaire and the subsequent scoring of the MCDI takes no more than 10 additional minutes. From my perspective, it is an
excellent use of time, because our focus is effectively shifted from the child’s ears to his/her overall function.

In conclusion, developmental screening at the initial identification of hearing loss in a young child is a necessity, not a luxury, if a comprehensive intervention plan is to be implemented at the earliest possible age. Early identification without early intervention obviously has little meaning.

REFERENCES

Auditory Brainstem Responses

The auditory brainstem response (ABR) is the electrical activity generated in the brainstem in response to a brief auditory stimulus. This response is volume-conducted through the brain and can be recorded from the human scalp. The following waveforms were recorded between the vertex and the mastoid in response to 70 dB nHL clicks. Positivity of the vertex relative to the mastoid is plotted by an upward deflection. Two traces were superimposed for each recording in order to demonstrate the reliability of the responses. The response in the adult shows a sequence of seven positive waves of which the first, third, and fifth are indicated by arrows. The positive waves in the ABR are usually identified using Roman numerals (I-VII). The infant response differs significantly from the adult response. The amplitude is smaller, the latencies of the peaks are longer, and the waveform is simpler. The arrows indicate the waves I, III, and V in the neonatal recordings. In the infant, the other waves are often difficult to recognize. The typical response is elicited by a click formed by passing a 100 μs square wave through an earphone.

Averaging

In scalp recordings the ABR is mixed with electrical activity from many other generators. This activity, coming from other regions of the brain and from the scalp muscles, completely obscures the response. The best way to recognize the ABR signal in this background noise is to use averaging. With averaging, the noise decreases in amplitude by a factor equal to the square root of the number of trials averaged (√N), whereas the signal remains constant. The amount of averaging required to demonstrate the absence or presence of a response depends upon the amplitude of the response and the amplitude of the background noise. For recordings of the ABR in infants, typical protocols average
the responses between 1,000 and 2,000 stimuli. Prior to averaging, it is essential to reduce the background noise as much as possible in order to give the averaging process a headstart. Babies are most effectively tested while asleep, and responses to stimuli that occur when the baby is moving should be rejected from the averaging using an artifact-rejection protocol.

**Recording Techniques**

In the infant, electrodes are placed just in front of the anterior fontanelle and on the mastoid or earlobe ipsilateral to the ear of stimulation. Care must be taken to ensure that one is not using the opposite ear, since the brainstem response is more laterally oriented in infants than in adults. Electrodes are connected to the skin using isotonic saline jelly and attached using hypoallergenic tape. Most instruments provide a means to monitor the connectivity of the electrodes by measuring the interelectrode impedance. The signals recorded from the electrodes are amplified in order to bring them to levels that can be easily evaluated in an averaging computer and then digitized at a rate sufficient to portray the ABR waveform (10 kHz).

**Filtering**

Filtering is an additional way to enhance the signal-to-noise ratio of a recording. Optimally, one would pass frequencies according to the signal-to-noise ratio at each frequency. In this way the signal would pass through the filter, but the noise would not. The filter characteristics could be varied further with different latencies of the response (time-varying filter). Unfortunately, when evaluating the ABR to near-threshold stimuli, the noise and the signal have very similar frequency characteristics, and there is little change in these frequency characteristics with time. A simple bandpass filter is therefore probably as effective as a more accurately designed filter. It must be remembered that the infant ABR is different from the adult ABR, and the bandpass should allow more low frequencies in the recording. A recommended bandpass is 20-2,000 Hz.

**Recording Efficiency**

Time is a very important parameter when one is designing an effective way to assess hearing in infancy. At rapid rates, more responses can be averaged within a given period of time, and the signal-to-noise ratio can therefore be increased more quickly. However, physiological responses generally decrease in amplitude at rapid rates. There is thus a trade-off between the decrease in amplitude of the response and the greater decrease in the background noise at rapid rates. Waves I-IV of the ABR decrease quite rapidly as the stimulus rate is increased beyond 10/s. However, wave V decreases much less, and rates of about 50/s are recommended as the most efficient rates for recording this wave.

Because the infant ABR to near-threshold stimuli may last for 20 ms, presenting the stimuli at rates faster than 50/s would cause the responses to one stimulus to overlap with the responses to preceding stimuli. Presenting stimuli according to maximum length sequences (MLS) can allow the overlapped responses to be disentangled. Using MLS techniques, responses can be recorded at stimulus rates of several hundred per second. The amplitude of the response decreases at these rapid rates, and there is probably not much increase in averaging efficiency for responses to high-intensity stimuli. However, the near-threshold response probably does not decrease as much with increasing...
stimulus rates. MLS techniques may therefore increase the speed whereby these near-threshold responses can be recognized.

Because the response to stimulation of the left ear is almost completely independent of the response to stimulation of the right ear, the response to two ears can be recorded concurrently. In simple averaging, the stimuli can be alternated between the two ears. With MLS techniques, stimuli can be presented to the two ears using a separate MLS sequence for each ear. Two ears can thus be evaluated in the same time as it takes to evaluate one ear.

Response Detection

Once a recording is made, a decision is necessary as to whether a response is present or absent. Averaging never fully removes the background noise: \( \frac{1}{\sqrt{N}} \) never reaches 0, regardless of how large \( N \) becomes. The evaluator is therefore never sure that a recorded waveform represents a real response or just some residual unaveraged background noise. A relatively simple way to determine whether the response is present is to replicate the recording and to see how well the two tracings can be superimposed. The difference between the tracings provides some rough estimate of the amount of residual noise present in the recordings. More formal techniques exist to assess the signal-to-noise ratio, and these are readily available on several commercial instruments. The optimal way of using these signal-to-noise estimates is to continue averaging until a response becomes clearly recognizable or until the possibility of a response greater than some minimum amplitude can be ruled out.

The decision concerning whether a response is present or not is made even more efficient if the response is compared to a normal template. This is what the human interpreter does when looking at waveforms. There are sufficient normative data available at the present time to construct general templates of the ABR to a 30 dB nHL click in an infant. Some commercial instruments include such a template-comparison in the automatic recording process.

Validity of the Response

Given the presently available equipment, one should be able to record a click-ABR in an infant at near-threshold levels (30 dB nHL) without any technical difficulty if the cochlea responds to the stimulus and if the auditory nerve and brainstem are working normally. The validity of the response when used to detect a sensorineural hearing loss in infancy therefore depends upon two provisos:

1. A conductive loss would decrease the effective intensity of the stimulus reaching the cochlea and raise the threshold for the ABR. It is therefore important to reduce the incidence of transient conductive hearing losses in the population tested. Perhaps testing at age 1-3 months may be better than testing in the perinatal period because of the lower incidence of conductive hearing loss at the later age.

2. A neurological disorder may prevent the response from being recorded. Hydrocephalus and demyelinating disorders may interfere with the generation of the ABR but not impair the hearing threshold. If no clear brainstem wave V is present, the electrocochleographic response or ABR wave I should be evaluated in order to check the function of the auditory nerve.
SUGGESTED READINGS


THE AUDITORY BRAINSTEM RESPONSE IN INFANTS: BASIC ASPECTS

Richard C. Folsom, Ph.D.

Introduction

The auditory brainstem response (ABR) has been shown to be particularly well suited for estimation of hearing levels in infants because it approximates behavioral thresholds in the mid-to-high frequencies, is stable over time, and is unaltered by sleep or sedation. Responses from infants have been described extensively, providing a broad literature base in both the basic aspects and the clinical application of this tool.

Of the five to seven vertex-positive waves that make up the ABR, wave V is the largest and most robust wave across stimulus intensities. This wave is most often used to determine the ABR threshold, or more accurately, the ABR visual detection threshold, defined as the intensity level at which a replicated response can just be differentiated from background noise. The term ABR threshold should not be confused with hearing threshold as determined with pure-tone signals, because ABR's and the stimuli that generate them are substantially different from behavioral responses to sound.

Stimulus Type and Frequency Specificity

The ABR exists as a result of synchronous neural activity facilitated by short stimulus onset times. Click stimuli, having abrupt rise times, are therefore best for creating synchronous activity. This abrupt stimulus creates an excitation area along the basilar membrane that is wider than for pure-tone stimuli. Thus, the capacity to resolve frequency-specific information is reduced. At moderate intensities, the frequency specificity of click stimuli in normal-hearing individuals is dominated by the basal end of the cochlea (~2,000 to 8,000 Hz). Clicks are similarly efficient in generating responses in infant subjects, although there is evidence that, when compared with adults, the frequency region that dominates the infant response is shifted toward lower frequency contribution (i.e., ~1,000 to 4,000 Hz for infants).

Short-duration tones (tonepips or filtered clicks) have been used to mitigate the limited frequency specificity available from click stimuli. These stimuli have relatively narrow spectra with rise times that are sufficiently short to generate a synchronous neural response. Masking studies have shown that at low-to-moderate intensities, these signals are frequency specific. At high stimulus intensities, the basal (high-frequency) end of the cochlea dominates the response regardless of tonepip frequency. Basic research using masking profiles and tuning curves has shown that tonepip stimuli are frequency specific and valid for assessing discrete cochlear regions in both adult and infant subjects.

Stimulus Intensity and ABR Threshold

For clinical purposes, intensity levels of the short-duration signals commonly used in generating the ABR are often described in reference to behavioral thresholds for the same stimulus by a jury of normal-hearing adult listeners, and termed "dBnHL." The sound pressure level (SPL) of these stimuli can be measured by using a sound-level meter with peak-hold capability (dBpeakSPL or dBpSPL) or...
by displaying the amplitude of a click on an oscilloscope and matching its voltage with a pure-tone (dBpeak-equivalent SPL or dBpeSPL). Because of the very short duration of click stimuli, behavioral threshold, or 0 dBnHL, corresponds to approximately 30 dB or 36 dB for peak-equivalent or peak SPL measurements, respectively.

Under optimal conditions, ABR threshold approaches behavioral threshold for the stimulus (approximately 5 to 10 dBnHL). This holds true for both adult and infant subjects when one is using either clicks or tonepip stimuli of various frequencies. For premature infants who are tested at less than 33 weeks of gestational age, there is evidence that ABR threshold is elevated. Elevated thresholds at this age, however, may reflect the sensitivity of the ABR as a tool for threshold assessment in a very immature system rather than the actual sensitivity of the auditory system itself.

ABR Latency and Morphology

Maturation of the auditory system is not complete at birth, particularly the auditory neural pathway from which the ABR is generated. Consequently, ABR's from infants undergo significant changes early in life and are different from those observed in adults. The waveform undergoes changes in morphology and latency as a function of age. Latency is the most widely used measurement value and is defined as the elapsed time from the stimulus onset to the peak of the wave being measured (absolute latency) or as the difference in time between the absolute latencies of two wave peaks (interwave latency). For click stimuli, there is evidence that the latency of ABR wave I (reflecting synchronous activity in the VIIIth nerve) reaches adult values within the first few weeks of life. The later waves (III and V) do not reach adult values until 18 to 24 months of age. Since waves III and V result from activity in the brainstem portions of the auditory pathway, change in latency of the click-evoked ABR is consistent with maturation within the central portion of the pathway. Clinical application of ABR latencies requires age-specific normative values.

For preterm infants, latencies of all waveform components are prolonged compared with those for term infants. ABR's can be first recorded at about 27 to 29 weeks of gestational age. Latency decreases rapidly until around 35 weeks of gestational age, with more gradual decreases until term (38 to 40 weeks). In preterm infants, the interwave latency can be as much as 7 to 8 ms (30 weeks of gestational age) decreasing to around 5.2 ms at term. Interwave latency for the adult click-evoked ABR is approximately 4.0 ms.

When frequency-specific stimuli are used, maturational effects vary with frequency. In general, when ABR's are generated in the basal portion of the cochlea, as with high-frequency tonepips or unmasked clicks, the above-described age-dependent differences in ABR absolute latency are observed. When responses are restricted to the mid-to-apical portions of the cochlea, ABR latencies from infants and adults are the same.

Absolute latencies of the ABR have an inverse relationship with stimulus level. As intensity is decreased, latencies increase. This latency change is roughly equivalent for all waves of the response, suggesting that peripheral changes (such as stimulus level or peripheral hearing loss), will first affect the latency of wave I, with subsequent waves shifting accordingly.
ABR morphology is largely determined by the component amplitudes. ABR amplitude defines the magnitude of the response (in voltage) and is commonly a peak-to-peak measurement from the positive peak to the following negative deflection. ABR amplitude is not usually employed in laboratory norms because it is variable and subject to nonauditory (electrode impedance, myogenic activity, etc.) as well as auditory factors. In adults, using surface electrodes, ABR wave V is larger than wave I; the V/I amplitude ratio between these waves approaches 2 to 3 (i.e., wave V is 2 to 3 times as large as wave I). The ABR morphology is simpler in infants than in adults, with the three basic components (I, III, and V) assuming greater prominence in the infant response. As with adults, wave V is the largest of the wave components, although amplitudes of waves I and III are more similar to V in infants (V/I amplitude ratios in infants are around 1).
Once a high-risk baby has been discharged from the newborn nursery, there is a strong likelihood that he/she will fail to meet outpatient appointments for followup testing and care. One recent study repeatedly contacted the caregiver to maximize return rate and found that more than one-third of college-educated mothers failed to return their child regularly for scheduled appointments, and more than two-thirds of mothers with a ninth grade education failed to do so. Because of the very poor return rate for outpatient appointments, a logical time to attempt early identification of hearing loss is prior to the time the newborn is discharged from the hospital. Hearing screening within the newborn nurseries markedly reduces the number of babies who require followup appointments as an outpatient. This allows the clinician to focus attention on a much smaller group of newborns (6 to 10 percent) who have either failed the hearing screening or are at risk for a delayed-onset or progressive hearing loss.

The greatest concentration of babies at risk for hearing loss occurs within the neonatal intensive care unit (NICU), so it is appropriate to select this as the primary screening site. Hearing testing within the NICU, however, poses some significant challenges to the clinician. If a newborn is residing in the NICU, it can be safely assumed that significant health problems still exist. As a result, the baby is routinely housed in an incubator and is connected to one or more monitors and infusion pumps. The electrical interference generated by such apparatus can greatly contaminate responses to hearing testing stimuli.

The acoustic test environment in the NICU is also far from optimum. The ambient noise level is high (averaging around 65 dBA). An incubator generates its own acoustic noise, so its plastic walls provide little or no net improvement in the general background noise level.

From a technical standpoint it clearly would be preferable to defer all hearing screening until the newborns are very near full-term and are residing in open bassinets, free from monitor and pump interference. Unfortunately, this ideal is usually impossible to achieve. Fiscally, tertiary care nurseries are routinely deficit operations. In addition, there is considerable demand for the limited bed space. Both factors contribute to the need to transfer newborns to their home hospital as soon as they no longer require the specialized services of the NICU. At my own institution, Duke University Medical Center, babies frequently will be transferred out of the NICU while they are still in incubators and well before fully satisfactory auditory brainstem response (ABR) test conditions are achieved.

Given the goal of testing all at-risk newborns prior to their hospital discharge, the ability to conduct hearing screening within the NICU is of major importance. It is in this environment that ABR audiometry has no rival as a screening procedure. Numerous studies have documented the effectiveness of ABR hearing screening in the NICU. When an appropriate test protocol is used, premature infants as young as 30 weeks of gestational age, can be tested at low stimulus intensity levels. Although ABR response amplitude decreases as stimulus intensity is reduced, it is possible to regularly screen hearing in the NICU at levels of 30 to 35 dB nHL. The recent introduction of clinical
test equipment with digital amplification and digital filtering capabilities has further improved the clinician's ability to perform ABR screening at low-intensity levels in this environment.

An ABR to an air-conducted click stimulus provides information about the newborn's peripheral hearing status in the 2,000 to 4,000 Hz range. A hearing impairment in this region can be expected to elevate the threshold for ABR detection and result in a failure of the screening test. If a neonate fails to demonstrate a response at the hearing screening level, ABR testing permits the stimulus intensity to be manipulated to provide information about the severity of hearing loss. Latencies of the component waves of the ABR provide additional information about the nature and magnitude of a hearing loss.

In addition to air conduction (AC) testing with click stimuli, bone conduction (BC) clicks can also be presented to the newborn to help differentiate between a conductive and sensorineural hearing impairment. Differences in responses to air- and bone-conducted stimuli must be interpreted with caution, however, because the distribution of energy for AC and BC clicks are quite different.

In clinical ABR screening, the basic question is whether the test stimulus has elicited a detectable response at the screening intensity level. Thus, all component waves of the ABR need not be present for a response to be detected. The clinician need only have confidence that the click stimulus has elicited synchronous neural firings within the auditory system. For premature newborns, wave III, rather than wave V, may be the dominant component of the ABR. Routinely multiple runs are obtained with the same stimulus parameters. Good agreement between these replications increases the clinician's confidence that a response has occurred.

Although the ABR has been used within the NICU for more than 20 years, research frontiers still exist. An example of this is the recent application of a new ABR technique to hearing testing of full-term and premature newborns. With maximum length sequence (MLS) analysis, click stimuli can be presented at rates much faster than are used in conventional ABR testing. Rather than a typical click rate of 30 to 40/s, rates of over 900/s have been shown to elicit clearly defined responses in the NICU. When test conditions are favorable, faster stimulus rates may result in shorter test time. When test conditions are less favorable, as is usually the case in the NICU, faster rates may increase response clarity because more individual responses are combined in the composite ABR. This, and similar lines of investigation, warrant further attention.

REFERENCES


EVALUATION OF HEARING IN THE NEONATE USING THE AUDITORY BRAINSTEM RESPONSE

Yvonne S. Sininger, Ph.D.

One of the basic criteria for establishing a program to screen for any health-related disorder is the existence of a screening tool that validly differentiates the disease from the nondisease state. To determine whether programs should be established for early identification of hearing loss, we must first know if tools are available to accurately predict auditory sensitivity in neonates, infants, and young children. This presentation will give evidence that the auditory brainstem response (ABR) is such a tool when proper stimulus, recording, and analysis techniques are applied.

Measuring ABR does not measure hearing. The ABR is the synchronous neural activity generated in the auditory nerve and brainstem, which is initiated by sound presented to the ear, and which is recorded from surface electrodes. Hearing is the conscious perception of sound initiated by such activity. The relationship between hearing threshold and ABR threshold is determined primarily by how well the minute signals making up the ABR can be recorded and distinguished from background noise. When ABR recording parameters are selected to optimize the detection of the response, the threshold of the ABR and the threshold of hearing are very closely related. 1,2

Several factors influence the threshold of ABR. The most important is the sensitivity of the auditory mechanism. This mechanism is the basis for the relationship between ABR and hearing. Those peripheral (external, middle ear, and cochlear) factors that elevate hearing thresholds will also raise the ABR threshold by an equal amount. Other factors that influence the ABR threshold need to be carefully controlled when using this response to predict hearing. These factors influence ABR threshold as well as response amplitude and detectability when suprathreshold stimuli are used in screening protocols. Several of these factors will be discussed including recording factors (electrode placement, signal filtering, amount of averaging) and methods for distinguishing presence or absence of response.

ABR threshold is defined as the lowest stimulus level that produces a clear wave V, the largest and most robust of the waveform peaks. For purposes of predicting hearing sensitivity, it is important to use recording parameters that optimize detection of this peak. In adults, this peak is recorded with the largest amplitude when differential electrodes are placed in a vertical orientation, recording between the vertex and nape of the neck. Sininger and Don have shown that ABR detection threshold is reduced by more than 4 dB using this recording montage. Preliminary data from newborns indicate that the amplitude of the ABR recorded from the vertex to seventh cervical vertebra is generally larger than the ABR when recorded from a standard vertex-to-mastoid montage.

Response filtering can have an even more dramatic effect on wave V amplitude. Spectral analysis of the infant ABR shows that it has significant low-frequency energy. This information can be reduced by standard 100 or 150 Hz high-pass filtering during recording. We have demonstrated the adverse effects of high-pass filtering on wave V amplitude of the infant ABR by comparing two averages, one with high-pass filter of 100 Hz and another of 30 Hz. Individual sweeps were recorded from newborn infants in response to clicks of 15, 30, and 60 dB nHL and 500 Hz tone bursts with a
recording bandpass of 30 to 3,000 Hz and stored offline. These data were then refiltered digitally with a high-pass filter of 100 Hz. The individual sweeps in each condition were averaged to produce ABR's. The amplitudes of the response are significantly larger when the 30 Hz high-pass condition is used and the estimated signal-to-noise ratio ($F_p$) is greater.

The ABR consists of tiny signals (in the nanovolt range) when measured at the surface of the head. These neural potentials must be pulled out of background noise (muscle response, EEG, EKG, etc.) that may be 100 to 1,000 times this magnitude. Stimulus-triggered signal averaging is used to improve this signal-to-noise ratio by enhancing all stimulus-related signals and canceling random noise factors. The amount of signal averaging needed to detect the response will be determined by the amplitude of the neural response and the magnitude of the noise contaminants. These factors cannot be known in a clinical evaluation. The amount of averaging needed to achieve a given signal-to-noise ratio in the ABR is inversely related to the sensation level of the stimulus, but large amounts of intersubject variability are related to differences in noise levels. Determination of response presence or absence, in a screening paradigm, will require that adequate time be spent averaging. If this is not done, excessive background noise may be misconstrued as demonstrating that the subject has hearing impairment.

Finally, an adequate measure of the signal-to-noise ratio in the ABR recording will dramatically reduce the subjective component of response determination. An online measure known as $F_q$ is updated at regular intervals during the averaging process. Statistical properties of this measure allow it to be used to determine the confidence level that indicates a true ABR is present. Criterion $F_q$ values can be determined based on the level of confidence desired, and averaging can be halted when this criterion is reached. $F_q$ grows with averaging when a response is present but, when no ABR is present, $F_q$ values generally hover around 0.8. $F_q$ can be used to determine when averaging has sufficiently reduced background noise and can be used as an objective criterion for response detection.

When these principles are applied to ABR recording, this technique can be shown to be extremely accurate in predicting hearing levels. Sininger has shown that prediction of average pure-tone hearing levels is consistently within 10 dB and ABR threshold in infants and young children when midline recording montage is used and $F_q$ criteria are applied to response detection.

More relevant to screening concerns is the ability to distinguish between response and no response ABR's in newborns. ABR's were recorded from normal full-term neonates using a 30 to 3,000 Hz bandpass and a vertex to seventh cervical vertebra electrode montage. Responses to clicks of 30 dB nHL are compared to silent control runs. The distribution of $F_q$ values for each shows almost no overlap; receiver operating characteristics are excellent and, based on an $F_q$ criterion of 2.4, the false-positive detection is 1 percent, true-positive detection is 98.5 percent, and $A'$ is 0.994. Hyde et al. in an actual ABR screening of high-risk infants with followup audiometric evaluation found that the sensitivity of the test was 0.904 and the specificity 0.907.

In conclusion, when recording is optimized for infant hearing detection, the ABR has been shown to be an accurate predictor of hearing loss. This technique will allow us to implement successful and cost-efficient infant hearing screening programs.
REFERENCES


5. Sininger YS, Cone-Wesson BK. Unpublished data.


HEARING SCREENING OF INFANTS WITH AUDITORY BRAINSTEM RESPONSE: PROTOCOLS, PERSONNEL, AND PRICE

James W. Hall III, Ph.D.

In this review of hearing screening based on auditory brainstem response (ABR), the term infant includes children from birth to 2 years of age. Five assumptions about hearing screening with ABR are amply supported by clinical experience and research findings:

1. Numerous commercially available instruments are routinely used for infant hearing screening.
2. There are devices for conventional ABR screening and interpretation (by skilled operators) and devices for automated recording and interpretation (by persons without such skills).
3. ABR's can be reliably recorded from infants as young as 30 weeks of gestational age and in newborn intensive care nursery (NICU) settings.
4. ABR screening can identify infants who are likely to have a handicapping hearing impairment.

Instrumentation, Methodology, and Personnel

All ABR devices used in hearing screening have in common components for generating an acoustic stimulus, delivering the stimulus to the ear, detecting bioelectric activity at the scalp (which may include the ABR), processing the bioelectric activity to enhance the response and to minimize background activity (noise), and displaying and storing, or printing, the processed (averaged) activity for analysis. There are, however, no widely accepted guidelines or published standards for the performance of ABR instrumentation, or for the optimal equipment settings for infant hearing screening with ABR. Some ABR stimulus and acquisition parameters (e.g., transducer type, stimulus intensity level, filter settings, number of stimulus repetitions), which can markedly affect ABR screening outcome, vary widely among published studies of infant hearing screening, as well as in clinical practice. Also, important but unappreciated methodologic factors may affect screening outcome. For example, all commercially available ABR instruments have as an option the click stimulus, which is universally employed in ABR hearing screening. However, the acoustic characteristics of the click stimuli that actually reach the eardrum and evoke an infant's ABR may vary widely among ABR systems and clinics, depending on transducer type and integrity, specific stimulus parameters, and physical properties of the infant's ear.

Suboptimal test parameters, combined with other weaknesses in test protocol, can contribute to an excessive proportion of false alarm rate, i.e., a fail outcome in a normal-hearing infant. False failures add substantially to the cost of screening. The expenses associated with followup assessment (administrative and parental costs and fees for diagnostic testing) are needlessly incurred for these normal-hearing infants. On the average, screening failure rates of 12 to 15 percent, and higher, are reported in the literature. Since the actual rate of handicapping sensorineural hearing impairment in the NICU population is on the order of 4 or 5 percent, we can assume typical false failure rates of up to 7 to 10 percent, even though under ideal test conditions (e.g., screening term infants in a sound-treated environment) failure rate may be as low as 5 percent. The author's experience suggests that a failure rate as low as 8 percent is more reasonable under typical NICU conditions, assuming that otherwise optimal screening test parameters and policies are followed.
Qualifications or credentials of the tester are rarely addressed in the literature. Personnel trained and experienced in ABR measurement are required with conventional instrumentation. It is best if one person records the ABR and interprets the response. This person can then adjust the test approach and "trouble shoot" during recording, as indicated, to increase the quality of the data recorded and the confidence of the ABR interpretation. An automated ABR screening device, such as the ALGO-1 infant hearing screener,1.2,11 can be operated equally well by a volunteer or another person without special training in ABR principles or procedures. Both conventional and automated ABR hearing screenings should be followed immediately with a signed report of the findings, and recommendations for management. An audiologist is best suited for conventional ABR hearing screening, for reporting the results of screening (conventional or automated), and for assuming responsibility for followup hearing management.

Performance and Cost of Hearing-Screening Techniques

A precise definition of the performance (accuracy) of ABR hearing screening is needed, but it is not easily determined. There are two fundamental measures of ABR performance in hearing screening: hit rate and false alarm rate. Hit rate is the proportion of infants with hearing impairment who are identified with the ABR screening protocol, whereas false alarm rate is the proportion of normal-hearing infants who are incorrectly classified as hearing impaired. Both measures of ABR screening performance remain elusive for a variety of practical reasons.3-9 There is no "gold standard" for ABR screening accuracy that can be used at the time of the screening. The next best alternative is to evaluate the accuracy of screening outcome in the context of hearing status assessed many months, or even years, later by behavioral audiometry. The problems with this approach are readily apparent, even with an optimized ABR screening technique and protocol. Some infants will "fail" the screening in the newborn ICN, and later demonstrate normal auditory function, because of transient or reversible peripheral or central auditory disorders. The impact of this problem can be reduced by using an ABR latency analysis strategy to differentiate conductive versus sensorineural screening failures.12,6 Other infants will "pass" the screening but show subsequent behavioral evidence of delayed, progressive, or acquired hearing impairment, such as infants receiving ototoxic drugs in the ICN, with perinatal infections, or with familial hearing impairment. This is no minor problem. Our experience at Vanderbilt University Hospital with the 1990 Joint Committee risk criteria suggests that approximately one-third of the infants who are at risk for hearing loss meet one or more of the criteria associated with progressive loss.10

Definition of ABR screening accuracy, of course, also depends highly on the criteria used for a pass versus fail outcome and how hearing impairment is defined. Reported prevalence of hearing impairment in infants in the ICN differs among published studies (generally between 2 to 5 percent, but higher if conductive hearing impairment is included). Prevalence data are lacking for infants at risk for progressive hearing impairment. Knowledge of the prevalence of initial, and progressive, hearing impairment in the population to be screened is useful for selection of the optimal test protocol, and also for estimating the costs of the screening protocol.9 Another important factor contributing to the ultimate hit rate of screening, and also in decisions regarding the screening protocol to be used, is the percentage of infants who will be successively followed up for diagnostic testing after screening failure, and the cost of such followup assessment. This factor deserves more attention.
The cost of infant hearing screening has been addressed by several authors.\textsuperscript{7-13} Perhaps the most complete estimates on screening costs are those derived from models by Turner.\textsuperscript{7-9} The typical hospital protocol involves hearing screening of at-risk infants in the ICN, with followup diagnostic assessment at 3 to 6 months after birth of the failures, plus those infants who could not be tested in the ICN. For this type of protocol, Turner estimates a cost of $77 for each infant screened and $5,500 for each infant identified with a moderate or severe bilateral sensorineural hearing impairment. However, costs of actual hospital-based hearing-screening programs have not yet been calculated based on this logical model.

REFERENCES


OTOACOUSTIC EMISSIONS: BASIC ASPECTS

Theodore J. Glattke, Ph.D.

Otoacoustic emissions provide both the basic scientist and the clinician with noninvasive tools to examine the function and integrity of the cochlea. Emissions are believed to be the result of active biological processes that produce motion of the structures of the inner ear. The motion patterns are coupled to the stapes footplate through inner ear fluids. A portion of the energy associated with the resulting stapes displacement is reflected back into the cochlea, and a portion is transmitted through the middle ear ossicles to cause displacement of the tympanic membrane and, ultimately, low-intensity sounds that can be detected in the ear canal.

Emissions often are classified by characteristics of stimuli associated with their generation. The initial report of Kemp described one type of emission, a prolonged low-intensity acoustic response to abrupt stimulation of the ear. Called a “Kemp echo,” this type of emission is more generally known as an evoked or stimulated otoacoustic emission (EOAE) and specifically as a transient evoked otoacoustic emission (TEOAE). Other types of EOAE’s include those located at the frequency of a continuous tonal stimulus (stimulus frequency otoacoustic emission, or SFOAE) and distortion product otoacoustic emissions (DPOAE), which are located at frequencies other than those used to stimulate the ear. As Kemp and his colleagues recently discussed, virtually any recording of sound pressure in the ear canal of a human subject who is receiving a stimulus and hears normally will include the stimulus and cochlear emissions. Emissions also occur spontaneously, without any known stimulation, in approximately 40 percent of individuals with normal hearing.

Although there are several stimulus conditions that will produce emissions, they probably derive, at least in part, from some common mechanism. In addition, there is substantial circumstantial evidence linking them to micromechanical events in the cochlea and, perhaps, in the outer hair cells of the cochlea. Some of the characteristics of emissions are as follows:

- Emissions present in normal human ears tend to have a maximum amplitude that does not exceed 20 dB sound pressure level (SPL).
- Effective stimulus levels (measured in the ear canal) for EOAE’s extend to below the threshold of hearing. The signal intensity associated with a TEOAE may equal or exceed the magnitude of the eliciting stimulus. Emission amplitudes reach asymptotic values when external stimuli reach mid-intensities.
- A prevalent form of spontaneous emissions and some forms of stimulated emissions consist of narrowband signals, suggesting that they arise from very restricted regions of the cochlea.
- Emissions occur with a delay (even for continuous stimuli) that is inversely related to the frequency of the emission and that is consistent with delays anticipated by cochlear traveling wave effects.
Emissions are influenced by conditions that are known to be associated with vulnerability of the outer hair cells.

Emissions can be suppressed by competing stimuli presented ipsilaterally. The tuning of the suppression patterns is as precise as that found for electrical responses from single hair cells or single neurons in the auditory nerve.

Emissions can be altered by stimuli presented contralaterally at stimulus levels below those believed to be associated with activation of the acoustic reflex or simple acoustic crossover.

The examination of emission phenomena has occurred in concert with revolutions in thinking and in novel demonstrations about how the normal cochlea functions. Those events have helped to minimize apparent conflicts that lie in the pathway of auditory theory. Among the issues and findings have been the following:

- If one extrapolates linearly, from measurements of basilar membrane displacement for high-intensity stimuli to estimates of displacement at threshold, the result is a physiological impossibility on the order of fractions of an angstrom.

- Direct measurements of basilar membrane displacements to low-intensity stimulation reveal that the near-threshold response is highly nonlinear. That is to say, relatively robust displacements are preserved in the face of significant attenuation of the external stimulus.

- The nonlinear portion of the displacement pattern is sharply defined around a point of maximum displacement, consistent with the precision of tuning of single hair cells and single neurons.

- The nonlinearity is vulnerable to conditions that affect the health of the cochlea, in general, and the outer hair cells of the cochlea, in particular.

- Individual outer hair cells have been shown to have motile properties in response to chemical, electrical, or mechanical stimulation in vitro.

- Individual outer hair cells have been shown to preserve the essence of their tuning characteristics after being removed from the basilar membrane in vitro.

- The outer hair cells enjoy a rich efferent innervation that terminates directly on cell bodies. This innervation could provide the infrastructure for alteration of cochlear activity in response to contralateral stimulation.

Two modern views of cochlear function are in conflict regarding their assumptions about whether or not the cochlea (1) is a passive resonator system or (2) incorporates an active biological amplifier system to govern its near-threshold response. The threshold and sharp tuning patterns of individual neurons have been simulated in both types of models. Regardless of the outcome of the debate over the existence of a cochlear amplifier, the circumstantial evidence linking otoacoustic emissions and outer hair cells to normal, low-intensity nonlinear behavior of the ear is compelling.
On one hand, emissions have been demonstrated to be associated with operation of the ear at low intensities, to be highly nonlinear, to be vulnerable to cochlear insult, to provide evidence of precision in tuning, and to be influenced by contralateral stimuli that probably are mediated via the efferent neural supply to the cochlea. On the other hand, the near-threshold mechanical or electrical behavior of the ear is highly nonlinear, vulnerable to cochlear insult, precisely tuned, and influenced by contralateral stimulation.

The new investigative tools that we have available present us with challenges that are similar to those involved in the interpretation of cochlear electrical events. Nonetheless, the opportunity to examine the ear preneurally has many important implications for detection and isolation of cochlear disorders, disorders of the nervous system that may involve the efferent innervation of the ear, and for the study of normal cochlear processes.

SUGGESTED READINGS


Current methods of screening for hearing impairment in infants and young children have inherent shortcomings, including lengthy examination times, the need for highly trained testgivers and interpreters, and limited ability to evaluate significant aspects of the frequency range. With the discovery of otoacoustic emissions and their development into practical measurement techniques, a new objective method for examining the processing capability of the peripheral hearing apparatus was introduced to the audiology field. In particular, of the varieties of evoked otoacoustic emissions currently known, the two types being most intensely investigated for their practical applications are transiently evoked and distortion-product otoacoustic emissions (TEOAE’s and DPOAE’s). Here we discuss the potential advantages and limitations of using DPOAE’s to predict the healthiness of cochlear function for the ears of newborns, young infants, and children.

The type of equipment for eliciting and measuring DPOAE’s is essentially the same as for measuring TEOAE’s, that is, with the capability to generate an optimal stimulus and to record the emitted response. However, for measuring DPOAE’s, the requisite stimulus consists of two continuous, simultaneously applied pure tones, referred to as the f₁ and f₂ primary frequencies, which are separated by an appropriate number of hertz. When two tones are introduced to the ear simultaneously, a number of intermodulation distortion products are produced that can be accurately predicted by the arithmetic relationship of the f₁ and f₂ stimuli.

In humans, however, the most robust intermodulation frequency is at the 2f₁-f₂ frequency, and it is this DPOAE that is typically measured. To avoid introducing artifactual distortion into the measurement system, it is common to externalize the ear speakers with respect to the ear-canal probe, so that the two primaries can be generated separately and then mixed acoustically within the canal. Currently, there are three distortion-product emission instruments available commercially: the ILO92DP (Otodynamics Ltd.), the Model 330 (Virtual Corporation), and the CUBeDIS (Etymotic Research).

A great amount of research in animal models on the fundamental mechanisms governing the generation of DPOAE’s has been performed over the past decade. More recently, investigative study has focused on establishing the practicality of DPOAE testing in the audiology clinic. Although the promising future of applying evoked emissions to neonatal hearing screening was recognized soon after their initial discovery, the earliest efforts to investigate this feasibility were focused on the TEOAE. This early emphasis on using TEOAE’s rather than DPOAE’s as a method of screening newborn hearing is understandable because of the great similarity between TEOAE’s and the currently accepted standard procedure for objectively screening hearing based on auditory brainstem response (ABR) testing. That is, both TEOAE and ABR testing depend on the ability of a transient acoustic stimulus, such as a click, to elicit a broadly generated physiological response.
More recently, however, because of the recognized benefits of DPOAE testing and because this evaluation can be completed within a timeframe that is similar to obtaining TEOAE’s, researchers have begun to explore the applicability of DPOAE’s as a screening technique for newborns.1,15 Some of the advantages of DPOAE compared with TEOAE testing in this special subject population are its abilities to

- Examine a wide frequency range typically extending from 0.5 to 8 kHz
- Test important low frequencies (i.e., <1 kHz), owing to a combination of implementing data analysis procedures specially developed to retrieve small signals from noisy background activity, and prior knowledge of the response frequency, i.e., that the DPOAE is equivalent to the 2f₁-f₂ frequency
- Obtain more complete frequency-specific information based on the pure-tone features of the eliciting stimuli.

Although general details concerning the benefits and limitations of DPOAE’s as a screening method for neonatal hearing are presently being established, particular information about this application is lacking. For example, current procedures use stimulus features that are based on our knowledge about eliciting maximal responses from adults. Consequently, it is well established that the optimal frequency ratio of f₁/f₂ is 1.22,16,17 but systematic study of this stimulus property for eliciting maximal DPOAE’s from the ears of newborns, older infants, and young children has not been undertaken, to date. Similarly, the ideal range of stimulus levels for evoking DPOAE’s from young ears remains unknown. This latter information is critical, because of the necessity of limiting evaluation of outer hair cell function with DPOAE’s to the “active” rather than “passive” generators of cochlear biomechanical activity, although the existence of such discrete processes remains to be demonstrated in human ears.

REFERENCES


CHARACTERISTICS OF TRANSIENT EVOKED OTOACOUSTIC EMISSIONS IN PEDIATRIC POPULATIONS

Susan J. Norton, Ph.D., CCC-A

Evoked otoacoustic emissions (EOAE's) are sounds that are recorded in the external ear canal but that originate within the cochlea. Evoked otoacoustic emissions themselves are not necessary for hearing, but reflect processes within the cochlea that are necessary for hearing. In recent years it has become evident that physiologically vulnerable elements within the cochlea are responsible for the good sensitivity, sharp frequency selectivity, and wide dynamic range associated with normal hearing. Increasing evidence indicates that these elements are the outer hair cells (OHC's) and that they are responsible for the generation of EOAE's. Thus EOAE's are potentially a noninvasive tool for studying outer hair cell function and, indirectly, for detecting hearing impairment involving outer hair cell dysfunction. One of the most promising clinical applications of EOAE's is in pediatric audiology. Among the potential pediatric applications are screening for hearing impairment in neonates and infants and separating peripheral hearing loss and central auditory dysfunction in infants and young children, particularly those with multiple disabilities.

Several characteristics of EOAE's make them attractive tools for neonatal hearing screening: (1) In the absence of external or middle ear abnormalities, EOAE's can be recorded from all normal-functioning human cochleas; (2) EOAE's are frequency specific and in neonates can be measured over a broad range of frequencies important for speech and language (i.e., 500 to 6,000 Hz); (3) EOAE's are absent in young ears with cochlear hearing losses of 40 to 50 dB HL or greater involving outer hair cell damage; the outer hair cell is the predominant site of lesion in early onset and congenital sensorineural hearing loss and losses due to perinatal and neonatal trauma; (4) EOAE's are independent of neural activity and are hence neurological abnormalities; and (5) EOAE's can be measured rapidly and noninvasively without the need to attach electrodes.

Transient evoked otoacoustic emissions (TEOAE's) follow presentation of brief acoustic stimuli such as clicks and tone pips. TEOAE's are frequency dispersive, frequency specific, and nonlinear. Emission latency varies inversely with frequency, suggesting that an emission of a particular frequency is generated at the cochlear place appropriate to that frequency. Another indication that TEOAE's are frequency specific is that their spectra are determined by the spectra of the evoking stimulus and the audiometric configuration. TEOAE amplitude also increases nonlinearly as a function of stimulus level. At low stimulus levels, in the normal-hearing ear the emission amplitude increases as stimulus level is increased. As the stimulus approaches 50 to 60 dB peSPL, emission amplitude increases more slowly and finally saturates such that further increases in stimulus level result in no significant increases in OAE amplitude.

The relationship between TEOAE characteristics and audiometric status is somewhat unpredictable in adults. However, in infants and children the relationship appears to be quite systematic. In a study of 4- to 13-year-old normal-hearing and hearing-impaired children with known audiograms, we found that if the pure-tone average (PTA) for 1,000, 2,000, and 4,000 Hz was greater than 30 dB HL, a TEOAE was never observed in response to clicks presented at 74 dB peSPL or less. If the PTA was greater than 40 dB HL, a response was never observed to 86 dB peSPL clicks. In
addition, we observed a systematic decrease in TEOAE amplitude in response to a given stimulus as a function of audiometric PTA. Response spectra also tend to mirror the shape of the audiogram for moderate- to high-level stimuli in children. In addition, two aspects of TEOAE growth functions for impaired ears compared with those of normal ears are important. First, the total power in the TEOAE in response to high-level stimuli (76 to 80 dB peSPL) is within the range observed in normal-hearing adults. Second, in contrast to normal ears, as the stimulus level is decreased, TEOAE amplitude decreases rapidly.

We evaluated the accuracy of TEOAE characteristics in identifying sensorineural hearing loss using methods derived from signal detection theory. Receiver operating characteristic (ROC) curves for several degrees of hearing loss were constructed. Using a criterion PTA of 30 dB HL and varying TEOAE amplitude in 1 dB steps, we obtained ROC curves with P(A)=1.0 or perfect separation of those children with losses equal to or greater than 30 dB HL from those with PTA's better than 30 dB HL. Thus, in the absence of middle ear pathology, TEOAE's are intimately linked to those elements within the cochlea responsible for absolute sensitivity, such that their characteristics vary systematically with thresholds up to about 40 to 50 dB HL. For hearing loss greater than about 40 to 50 dB HL, that is, those ears in which there is probably damage to inner hair cells as well as to outer hair cells, TEOAE's are absent. In this case a 60 dB loss cannot be distinguished from a 90 dB loss, that is, TEOAE's would be absent in both cases.

Almost immediately following Kemp's (1978) original report of TEOAE's, it was suggested that they could be used as a screening tool for detection of hearing impairment in neonates. Early reports indicated that neonates had robust TEOAE's, which were on average 10 dB greater than in adults. We have found that at all stimulus levels normal neonates had significantly higher amplitude TEOAE's than adults (p < 0.001). Possible explanations include differences in the volume of the external ear canal, differences in the middle ear transfer function, differences in cochlear function, or a combination of all three. The work of Keefe and his colleagues indicates that there are few changes in middle ear mechanics, but significant changes in external ear canal characteristics.

Although several studies have been done of TEOAE's in neonates, it is not yet clear what the referral criteria should be for TEOAE screening. As in most studies of auditory brainstem response (ABR) in neonates, the "pass-fail" criteria are based on adult data, and only those infants who "fail" are followed. Essentially, as in most ABR screening programs, it is assumed that if neonates demonstrate TEOAE's at least as large as those observed in normal-hearing young adults, they have normal hearing! There are two problems with this approach. First, because neonates on average have significantly larger TEOAE's than do adults, it is not clear that the same pass-fail criteria are appropriate. Second, the true accuracy of TEOAE's as a screening tool for hearing loss in neonates cannot be determined unless followup behavioral confirmation of hearing status is obtained for all infants screened in close temporal proximity to the time of screening, that is, within 6 months. Unfortunately, this is generally not possible in a standard clinical setting. However, most studies have identified primarily infants having moderately severe to profound hearing losses and no measurable TEOAE. Based on our knowledge of TEOAE's in normal-hearing and hearing-impaired young children, it is theoretically possible that some neonates with mild to moderate hearing losses could have low-amplitude emissions, comparable to those observed in normal-hearing adults.
TEOAE's have several advantages over ABR for neonatal hearing screening. TEOAE's can be obtained more rapidly than ABR because no electrodes need be attached. TEOAE's also are sensitive to a broader frequency range than ABR. Finally, there is considerable evidence that evoked otoacoustic emission generation is preneural and independent of synaptic transmission. When both ABR and TEOAE's are obtained from infants and children who are neurologically at risk, a small percentage have robust TEOAE's and either absent ABR or significantly elevated ABR thresholds. Thus, in a population neurologically at risk, either tool alone may provide inadequate information about auditory system status. Only after we have identified and followed a sufficient number of these children over several years will we understand the implications of these findings. However, for normal full-term neonates, TEOAE's promise to be a rapid and reliable tool for screening neonates, infants, and young children.

REFERENCES


PRACTICALITY, VALIDITY, AND COST-EFFICIENCY OF UNIVERSE NEWBORN HEARING SCREENING USING EVOKED OTOACOUSTIC EMISSIONS

Karl R. White, Ph.D.

A recent, but rapidly expanding, body of research provides convincing evidence that the implementation of universal newborn hearing screening programs based on the measurement of evoked otoacoustic emissions (EOAE's) would result in much earlier identification of hearing loss. As shown in Table 1 and summarized below, published research from multiple, independent laboratories and clinical trials in the United States, England, Germany, and France provides compelling evidence of the practicality, validity, and cost-efficiency of newborn hearing screening using EOAE as compared with available alternatives.

Practicality

Published evaluations of EOAE for newborn hearing screening consistently conclude that the technique is simple, fast, and noninvasive. Those who have published results based on screening relatively large numbers of babies report valid completion of an EOAE screen for upwards of 95 percent of the infants selected for screening. Time required to do EOAE testing is very short, with reports ranging from 5 to 20 minutes for total testing time including preparation and necessary transportation. Screening can be done by trained technicians, and soundproofing of testing location is not required. The most convincing demonstration of the feasibility of EOAE-based screening comes from the Rhode Island Hearing Assessment Project (RIHAP), where all newborns at Women and Infants Hospital of Rhode Island (approximately 1,000 infants per month) are currently being screened. Although existing research demonstrates that EOAE screening can be feasibly implemented in hospital settings, continuing refinements in EOAE software and hardware will make such testing even easier. Thus, there is no longer any doubt that universal newborn hearing screening using EOAE is practical.

Validity

The sine qua non of a newborn hearing-screening technique is its ability to distinguish between those infants who have a hearing loss and those who do not. The overall conclusion from the published research cited in Table 1 is that EOAE screening is as valid as the auditory brainstem response (ABR). Bonfils et al., Stevens et al., and Plinkert et al. reported that EOAE identified from 92 to 97 percent of the same children identified by ABR, and Uziel and Piron reported that the same children were identified. By comparing screening results for both ABR and EOAE with behaviorally confirmed hearing loss at 6 to 12 months, Kennedy et al. and White et al. reported that EOAE identified all of the children identified by ABR, in addition to 8 to 10 children per 1,000 with mild sensorineural or conductive hearing losses who were missed by ABR.

EOAE screening within 3 days of birth does result in a number of infants being referred who have normal hearing. However, this number is similar to ABR used under similar conditions and results in more successful identification of infants with unilateral sensorineural, mild bilateral...
<table>
<thead>
<tr>
<th>Study</th>
<th>Description of Sample</th>
<th>Basis of Comparison</th>
<th>Results/Conclusions</th>
</tr>
</thead>
</table>
| Bonfils et al. (1988)<sup>1</sup> | 46 infants' ears; 30 with normal hearing, and 16 with sensorineural hearing loss | EOAE compared with ABR (≥30 dB) at mean age of 4.6 months | **EOAE's were present 97% of the time when ABR wave V threshold ≤30 dB; and never present when ABR thresholds ≥40 dB. ABR required average of 40 minutes, EOAE required 5 minutes per baby.**  
**"EOAE technique is simple, fast, and noninvasive, as well as totally objective and sensitive."** |
| Stevens et al. (1989, 1990)<sup>2</sup> | 723 NICU infants                                           | TEOAE testing at average of 1 month of age compared with ABR (≥43 dB in one ear and 53 dB in the other) testing at 8 months of age | **Initial EOAE had sensitivity of 93% and specificity of 84% with ABR at 3 or 8 months of age. Concluded that EOAE should be used as an initial hearing screen followed by ABR for infants who fail initial EOAE. ABR screening required twice as long.  
"[EOAE is] less invasive than the brainstem electric response test [and] would make a better screening test."** |
| Plinkert et al. (1990)<sup>3</sup> | 95 ears of 53 high-risk infants                            | EOAE compared with ABR at mean age of 9 weeks             | **EOAE compared with ABR (≥30 dB) had sensitivity of 90% and specificity of 91%.  
"EOAEs are a reliable and practical method for screening of auditory dysfunction in high-risk infants."** |
| Uziel & Piron (1991)<sup>4</sup> | 55 full-term infants with no risk factors and 40 NICU infants | EOAE compared with ABR (≥30 dB) both at 1 to 5 days of age | **All ears producing no emissions demonstrated ABR thresholds above 30 dB. Time required for screening was 5 minutes for EOAE vs. 35 minutes for ABR. Concluded that EOAE provides an objective tool for rapid and effective screening of cochlear impairment in neonates.** |
| Kennedy et al. (1991)<sup>5</sup>  | 370 infants (223 NICU, 61 from normal nursery with family history of hearing loss or craniofacial abnormalities, and 86 from normal nursery with no risk factors) | EOAE testing at 1 month of age compared with behaviorally confirmed hearing loss (≥40 dB) at 8 months of age | **EOAE and ABR identified the same 3 children with sensorineural hearing loss, but EOAE identified 7 of 10 children with conductive hearing losses, while ABR identified only 4 of 10. EOAE screening required half as long as automated ABR. Concluded that universal newborn hearing screening should be done with EOAE followed by ABR for those who fail the initial screen.** |
| White et al. (1993)<sup>6</sup>  | 1,850 infants representative of general population (1,546 regular nursery and 304 NICU) | EOAE testing at 1 to 4 days compared with behaviorally confirmed hearing loss at 6 to 12 months of age | **Universal newborn hearing screening using EOAE identified 5.95 infants per 1,000 with sensorineural loss and 20.0 infants per 1,000 with persistent fluctuating conductive loss. Sensitivity/specificity with behaviorally confirmed loss at 8 to 12 months was 100% and 82%.  
"TEOAE screening of all newborns...is fast, economical, noninvasive, and accurate in identifying more infants with...hearing loss than other available techniques."** |
sensorineural, and conductive losses. Furthermore, if the two-stage screening process recommended by previous investigators is used (i.e., those infants referred from the initial screen are rescreened later using either ABR or EOAE), the false-positive rate drops dramatically. Taken together, existing research provides clear evidence that screening using EOAE validly distinguishes between infants who are hearing impaired and those who are not.

Cost-Efficiency

Prohibitive costs have been the major obstacle preventing widespread implementation of newborn hearing screening. Unfortunately, most cost analyses of newborn hearing-screening programs are based on hypothetical situations. In contrast, an ingredients approach to cost analysis was used at RIHAP to document the actual costs of EOAE-based screening including equipment, supplies, overhead, and salaries and fringe benefits of all staff involved in the screening (whether contributed or directly billed). The resulting analysis demonstrated that an EOAE-based newborn hearing-screening program could be done for less than $25 per child, with a cost of $3,364 for every infant identified with a sensorineural hearing loss. This contrasts with the cost of identifying a child with PKU or hypothyroidism of almost $41,000 per child. The relatively low cost per child screened and per child identified in newborn hearing screening using EOAE, combined with the fact that third-party payors in several States are beginning to reimburse for newborn hearing screening, suggests that cost is no longer an obstacle to implementation of universal newborn hearing screening.

Important Directions for Future Research

Based on the dramatic progress over the past 5 years, it is clear that further research, including the following, will continue to improve the cost-efficiency of newborn hearing screening.

- Published reports of EOAE screening trials have all used transient evoked otoacoustic emissions. Research is needed on the applicability of distortion-product evoked otoacoustic emissions as a newborn hearing-screening technique.

- Limited data suggest that automated ABR techniques may become another viable alternative for newborn hearing screening. However, as is the case with all technological innovations, research about the practicality, validity, and cost-efficiency of automated ABR in comparison to other techniques, including EOAE, is needed before broader use is encouraged. Thus far, the one study reported in a clinical setting concluded that automated ABR was better than traditional ABR, but not as efficient as EOAE.

- Continued research focused on improvements in EOAE hardware and software (e.g., automated scoring routines; smaller, more portable equipment) will increase the probability of widespread implementation.

- The potential of EOAE for identifying risk of conductive hearing loss is promising, but needs further research.

- There is an urgent need for long-term followup of infants who pass initial EOAE screening as well as those who fail (this is also needed for other screening techniques).
REFERENCES


MODELS FOR EARLY IDENTIFICATION AND FOLLOWUP: AN OVERVIEW
Robert G. Turner, Ph.D.

Models of Early Identification Protocols

Most protocols for the early identification of hearing loss can be represented by one of four models.

M1: Obvious. In this strategy, parents and professionals wait until the hearing loss becomes obvious before diagnosis and habilitation. Thus, there is no effort at early identification. By default, this is probably the most common strategy in the United States at this time, and that is the problem.

M2: Birth Data. Some time after the infant is discharged from the nursery, birth records or birth certificate data are reviewed to determine which infants meet high-risk criteria. These infants are referred for audiological evaluation, usually at an age appropriate for behavioral testing. The State of Utah has had such a program for many years.

M3: In-Hospital Screening. Most efforts at early identification of hearing loss use some type of screening before the infant is discharged from the hospital. The purpose is to identify those infants at risk for hearing loss. A variety of techniques are used for the screening. Probably the most common is the high-risk register (HRR), which has the advantage of not requiring physical contact with the infant before discharge. Behavioral screening with high-level tones or noises have been used, but have fallen into disfavor. Computerized cradles (Crib-O-Gram and the Auditory Response Cradle) offered promise but were largely supplanted by auditory brainstem response (ABR) testing. ABR is the most popular physiological technique, but is now being challenged by otoacoustic emissions.

M4: Total Referral. All infants are referred for evaluation at 3 to 5 months of age; there is no in-hospital screening. This strategy is seldom used because of the expense of following and evaluating all newborns, but should be considered for two reasons. It serves as a reference for the other models, and it has legitimate use in certain situations.

Evaluation of Models

The models can be evaluated by four measures. These are (1) age of identification of hearing loss, (2) performance of the model, (3) cost of implementing the model, and (4) cost-effectiveness of the model.

Initially, we will compare all four models in terms of age of identification and cost of implementation. The Joint Committee on Infant Hearing has recommended that hearing loss be identified and habilitation begun by 6 months. This is the standard against which we will compare age of identification. Cost of implementation is the average cost of the testing and followup per newborn without regard to type of nursery. Table 1 provides this information for the four models. These estimates come from several sources, including the model developed by Turner.
TABLE 1

<table>
<thead>
<tr>
<th>Model</th>
<th>Age of Identification</th>
<th>Cost per Infant</th>
</tr>
</thead>
<tbody>
<tr>
<td>M1: Obvious</td>
<td>30 months</td>
<td>&lt;$1</td>
</tr>
<tr>
<td>M2: Birth data</td>
<td>12 months</td>
<td>$2</td>
</tr>
<tr>
<td>M3: In-hospital screening</td>
<td>6 months</td>
<td>$20</td>
</tr>
<tr>
<td>M4: Total referral</td>
<td>6 months</td>
<td>$90</td>
</tr>
</tbody>
</table>

M1: Obvious. There is no effort at early identification; thus there is no expense for screening or followup. The only expense would be for the eventual diagnostic evaluation, making the cost of this approach quite small. However, this approach results in an average age of identification of 2 1/2 years or older.  

M2: Birth Data. Experience in Utah indicates an age of identification of about 1 year at a cost of approximately $2 per infant. This strategy significantly reduces the age of identification relative to no identification program; however, this model does not meet the recommendation of the Joint Committee.

M3: In-Hospital Screening. This model identifies the at-risk infants before discharge. Infants return for evaluation with ABR at 3 to 5 months of age, making identification and initiation of habilitation possible by 6 months. The cost varies depending on technique, but cost can be as little as $20 per infant for certain screening strategies.

M4: Total Referral. This strategy is usually the most expensive, but does permit identification and habilitation by 6 months of age.

Performance and Cost-Effectiveness

Of the four models, only M3 and M4 have the potential to meet the recommendation of the Joint Committee for identification and habilitation by 6 months. We will consider these two in more detail, evaluating performance and cost-effectiveness using estimates from Turner.  

The measure of performance will be hit rate, which is the percentage of hearing-impaired infants identified by the screening. The measure of cost-effectiveness will be the cost of testing and followup per hearing-impaired infant identified.

For M3, we will consider screening strategies of HRR, ABR, and HRR+ABR (infants who fail the HRR receive ABR). It is necessary to consider the well-baby nursery (WBN) separately from the intensive care nursery (ICN).
It is evident from Table 2 that any of the screening strategies, including total referral, is appropriate in the ICN. There is little difference in performance or cost-effectiveness. Newer techniques like otacoustic emissions might provide some improvement in performance or cost-effectiveness, but this is not critical.

The real dilemma is in the WBN. Any strategy will be much less cost-effective in the WBN than in the ICN because of the lower prevalence of hearing loss. The HRR significantly reduces costs in the WBN but also significantly reduces performance. Screening all newborns with ABR would identify most hearing loss, but would be more expensive. One solution is a new screening procedure with performance comparable to ABR, but more cost-effective. Modeling has shown that the cost of administering the test (i.e., time required) is more important than false alarm rate at determining cost-effectiveness.

Conclusions

- Without an organized effort, age of identification is significantly delayed.
- To meet the recommendation of the Joint Committee, at-risk newborns must be identified soon after birth (M3 and M4).
- We know how to screen effectively in the ICN using ABR.
- The real problem is screening in the WBN.
- We need a screening test that has a high hit rate and that can be performed much more quickly than ABR.
REFERENCES


In the early 1970's, universal implementation of a hearing-screening register for high-risk newborns was recommended in the United States, and data of that time suggested a risk register might have the capacity to identify up to 75 percent of all severe to profound hearing-impaired newborns. In 1975 it was estimated that only 1 percent of the live births in the United States were screened by a hearing risk register, whereas by 1981, this coverage increased to an estimated 6 percent of the total newborn population. It was reported in 1984 that five States had active statewide high-risk screening programs, four had a regional program, and eight were planning future programs. By 1988, 26 States reported programs that varied from consultative recommendations only to universal high-risk screening. Fourteen States had legislative mandates, 10 had voluntarily implemented State-operated programs, and 2 had private projects. Five States reported in-hospital auditory brainstem response (ABR) neonatal screening, of which three screened only neonatal intensive care unit (NICU) babies. Seven reported using a high-risk registry that notifies parents of the infants' risk status at about 4 months postpartum, and the remaining States had some combination of a high-risk registry and in-hospital screening. Of 14 States with a legislative mandate, only 8 had implemented some type of screening program, mainly due to a lack of associated funding.

In 1992 another survey of States using hearing high-risk registers was published. Of 42 States responding, 18 reported using a high-risk register for hearing screening, and in 15 the goal of the register was to screen all live births. Connecticut, Florida, Kentucky, Maryland, Massachusetts, New Jersey, Oklahoma, and Virginia had a legislative mandate; Georgia, New Mexico, North Carolina, North Dakota, Oregon, and Utah had statewide nonmandated programs; and California, Arizona, and Nevada reported screening only NICU graduates. This report included a comprehensive table of high-risk register criteria utilized by each State's health and welfare agencies to obtain information about hearing impairment identification programs in their States. In a preliminary report, 16 reporting programs were legislatively mandated, 5 were statewide, and 21 were in individually operated birthing sites. This comprehensive survey had 52 questions that included items on funding, test protocols, followup protocols, equipment, costs, and habilitative options, among others. The American Speech-Language-Hearing Association is in the process of finishing the study.

Selected Model Programs

Five programs for early identification of hearing impairment are presented. These programs were selected because of their diversity, and it is hoped that they represent an adequate sample of major early identification efforts in the United States. No attempt is made to offer a comprehensive review of the many excellent early identification programs in the country.

Utah High-Risk Screening Program

Program Type: Statewide high-risk register by birth certificate with State followup, education, and testing; implemented by public health department; nonmandated.
This program is implemented by the Utah Bureau of Communicative Disorders and is funded through the Federal Maternal and Child Health block grant to the State. The Utah birth certificate has been used for hearing-risk register data since a question pertaining to family history of hearing loss was added in 1978. By computerized search of the birth certificate, most risk criteria associated with neonatal hearing loss are screened on almost every live birth in the State. Parents of at-risk infants are sent a computer-generated risk notice packet at about 4 months postpartum. This contains educational materials and a postage-paid response card, on which a hearing-screening appointment is requested after the infant is 6 months of age. Nonrespondents receive a second notice in 2 months. Statewide 1/2-hour screening appointments include visual reinforcement audiometry, acoustic immittance procedures, and parent counseling. Followup may include ABR testing and, recently, otoacoustic emissions.

A total of 489,256 live birth certificates were screened between 1978 and 1990, resulting in 42,744 (8.7 percent) parental notifications. Fifty-one percent responded, resulting in the screening of 7,439 infants. Of these, 2,049 (27.5 percent) infants were found with conductive hearing loss and 200 (2.7 percent) with sensorineural impairment. Fourteen percent of the 200 sensorineural cases had mild losses, 21 percent were moderate, 8 percent were moderate/severe, 27 percent were severe, and 24 percent were profound losses. The average age of identification in the program is under 11 months. Total program costs are approximately $1.50 per live birth, including audiological screening. The major advantage of the program is that nearly 100 percent of the States' newborn population can be efficiently screened for risk of hearing loss by computerized search of birth certificates. Followup is also effectively provided by extensive computer management, and a high degree of public and professional awareness is generated. The major weakness of a hearing risk register is that it may only be sensitive to about 50 percent of the newborn population with congenital hearing loss.9,10 When combined with the high attrition rate of most followup programs, the maximum potential for risk register programs to identify the majority of hearing-impaired infants in a large newborn population is severely limited.

Winter Park (Florida) Memorial Hospital Infant Hearing Program

Program Type: Hospital-based; screens all live births; well-baby nursery and NICU; volunteers used for screening; uses automated ABR equipment plus a high-risk register with followup services.

Winter Park Memorial is a private not-for-profit hospital with more than 1,700 nursery admissions per year. Although the hospital does not have in-house audiology; private-practicing audiologists offer services as allied health staff. In 1983, the Infant Hearing Program (IHP) began as a nonprofit service that offered ABR screening to all newborns, using appropriately trained volunteers. Since 1988, infants are screened under standing orders, 7 days a week, 365 days a year, using the Algo-1 Plus automated ABR instrument. By using this device, a relatively small number of volunteers can screen all hospital live births with consistent results. A high-risk register is also completed on every baby to identify those that require monitoring for delayed-onset hearing loss. Screening results are provided to the infant's physician, who shares the information with the parents. Infants who fail the procedure are referred by the physician to community audiologists outside the hospital. Ninety-seven percent of admitted infants are screened prior to discharge, and because of the increasing number of rapid discharges, it is necessary to screen well babies at 12 hours of age. Phone calls and written notices are sent to the parents of infants not screened before discharge. From 1988 to 1990,
3,943 (96.5 percent) of admitted infants were screened, with 340 (8.7 percent) referred for additional evaluation. Only 18 volunteers performed these screenings. Of 88 of these infants tested in one followup program, 48 (56 percent) were found with confirmed hearing impairment. Forty-one had conductive and 7 had sensorineural losses. This program offers universal newborn hearing screening to nearly all live hospital births, by using an efficient and cost-effective, volunteer-operated, automated ABR screening protocol. Universal coverage also promotes a high level of parent and provider education, and the use of volunteers promotes wide community support. Challenges include recruiting capable volunteers to make long-term time commitments, providing adequate volunteer training, and providing ongoing audiological supervision of volunteers and other hospital staff.

University of Virginia Health Science Center Newborn Hearing Screening Program

**Program Type:** Hospital-based screening of at-risk newborns; normal newborn nursery and NICU; conventional ABR screening by audiologists with followup services.

Approximately 500 newborns per year are found to be at-risk in this medical center, as determined by a Virginia health department hearing risk assessment form. This comprises about 7 percent of the normal newborn nursery (NBN) and 50 percent of the NICU population. NBN staff contacts audiology to provide ABR screening for their at-risk infants. Audiology reviews NICU risk forms and schedules ABR screening of at-risk infants who are ready to test (34 weeks of gestational age, free from aminoglycoside and loop diuretic therapy, those extubated and those close to discharge). Parents of infants who are missed are telephoned or mailed a notice that recommends followup procedures. From 88 to 92 percent of the at-risk infants pass the initial screen by showing repeatable ABR's at 30 dBnHL bilaterally. Followup is recommended for 25 percent of the passing infants. Of the 8 to 12 percent that fail, 40 percent pass followup testing, 40 percent fail, and 20 percent are lost to followup. Both the health department and the university contact parents by mail to provide information regarding the infants' fail status and followup recommendations. If the followup results suggest possible hearing loss, more extensive otologic and audiologic evaluations are performed. This program has the advantage of screening at-risk infants prior to hospital discharge, thus reducing the number of infants lost to followup. Another advantage is the multiagency cooperation demonstrated, which may include medical care and hearing aid provision to infants enrolled in the Virginia Children's Specialty Clinic. The program needs to develop a systematic protocol to follow infants who pass initial ABR and are at risk for possible progressive hearing loss. Also, as indicated earlier, up to 50 percent of newborns with hearing impairment may not be identified by a program based on the risk register.

The Rhode Island Hearing Assessment Project

**Program Type:** In hospital; mass newborn screening; NICU and well-baby nursery; uses transient evoked otoacoustic emissions screening by unsophisticated screeners.

In 1990 a project was initiated at Women & Infants Hospital, with joint funding from the Department of Education and the Bureau of Maternal and Child Health, to begin universal hearing screening of newborns by transient evoked otoacoustic emissions (TEOAE). In 1992 new legislation was passed in Rhode Island mandating universal newborn hearing screening and a reimbursable fee structure. Since April 1, 1992, all newborns have been screened with TEOAE, including all NICU and
normal nursery infants. The hospital has approximately 9,400 deliveries per year, of which 1,400 infants are cared for in the NICU. Approximately 26 newborns are screened per day. Infants who pass the initial TEOAE are discharged. Infants who do not pass return in 4 to 6 weeks for a second TEOAE, and if they do not pass this, they receive ABR screening. Infants who fail the TEOAE and ABR at <60 dB are referred for a behavioral audiological evaluation. Infants who fail the rescreen ABR at >60 dB are referred for a diagnostic ABR, and partial passes are referred for a behavioral assessment at 6 months. Between August 1990 and February 1991, 1,850 infants were screened, of which 304 (16.4 percent) were cared for in the NICU. Confirmed bilateral sensorineural hearing losses were identified in two of the normal nursery infants and four of the NICU graduates, while unilateral sensorineural losses were found in two normal nursery newborns and three NICU graduates. Approximately 50 percent of the infants identified did not have one of the 1992 Joint Committee on Infant Hearing risk factors.

TEOAE is quick (10 minutes per child), noninvasive, inexpensive ($25 per child), and easy to administer by staff who are not highly trained. It has lowered the age of identification and rehabilitation for infants with hearing impairment in the State and is considered successful. Program weaknesses include the fact that TEOAE does not discriminate between conductive and sensorineural hearing losses and does not identify hearing threshold. Also, there is a 30 percent TEOAE failure rate for infants under 24 hours of age, which improves to 18 percent 3 to 4 days following birth. This is important when one is considering the growing trend toward early hospital nursery releases, including monetary incentives from some insurance carriers.

Colorado Newborn Hearing Screening Project

Program Type: Universal statewide newborn screening and followup; hospital based; uses volunteers who usually screen with automated ABR.

This program is a unique attempt to accomplish statewide universal newborn hearing screening through a consortium of supporters. Coordinated by the Colorado Department of Health, the program has a goal to screen 80 percent of the State’s newborns by 1995, by placing newborn hearing-screening services in every birthing hospital in the State. During the first year, 5 hospitals covering almost one-third of Colorado births have joined the program and it is expected that by January 1993, 12 more will be in operation. Gaining access into hospitals is the first and most challenging step. This includes convincing hospital administrators and physicians of the need for the program and the fact that it can be cost-effective or even revenue generating. Instrument costs pose a problem for smaller hospitals with low newborn nursery populations, such as those in rural areas. Screening costs range from $25 to $60, and most private insurance carriers pay in full. Medicaid and HMO’s pay a standard rate for birthing costs and are often inflexible about paying for added procedures. The program uses volunteers who are supervised by staff or contract audiologists. An overriding experience is that each hospital has a different philosophy, needs, and resources, and that each requires an approach that accommodates these individual differences. Since March 1992, one hospital program screened 1,246 (88 percent) of 1,414 live births, losing only 12 percent to early discharge and parents waiving the procedure. Twenty-three (1.8 percent) infants were referred for further assessment, and 5 were found with confirmed sensorineural losses. Of the five, three had a known risk factor. Program strength lies in its diverse support from public health, academia, the hospitals, and the audiology community. The primary challenge is to coordinate the ongoing efforts of all of the participating
hospitals in areas of funding, volunteer training, data management and reporting, habilitative referrals, and patient tracking.

Summary

As seen in the above programs, newborn hearing screening in the United States encompasses a wide variety of models, initiatives, and protocols that reflect the diverse and, some would say, rather chaotic state of primary and preventive health care in the country. If you happen to be born in a certain State or in a large metropolitan hospital, or happen to be a graduate of an NICU, you might have your hearing screened before 6 months of age. Even then, the probability that your hearing will actually be screened depends on other extenuating factors, such as (1) your parents’ willingness and ability to bring you in for followup services; (2) if you have a primary care provider who is sensitive to the importance of early identification; (3) if the NICU you graduated from has standing hearing-screening orders; (4) if you have been identified as being at-risk for hearing loss; and (5) if your health insurance, your parents, or a public or private agency is willing to pay for the screening services.

Recent findings suggest that as many as 4 to 6 out of every 1,000 newborns in the United States will have sensorineural hearing impairment of some degree, in at least one ear. Unfortunately, it is also estimated that only 3 to 10 percent of the total U.S. newborn population is screened for hearing loss. With these data, it is little wonder that the average age of identification of hearing loss in the country is between 2 and 2½ years, and that this late identification has seemingly not improved over the past decade. It is doubtful that any single hearing-screening strategy can ameliorate this situation in the near future, before some form of standardized national health care is in place. Universal newborn and infant hearing screening may rightfully be the ultimate goal for the Nation. But until such is compatible with other national health priorities that encourage primary prevention services, the many diverse early-identification protocols that presently exist in the United States will need to continue at the local, regional, and State levels.

Acknowledgment

The author thanks Dr. Judy Marlowe (Florida), Dr. Betty Vohr (Rhode Island), Kathy Waters (Colorado), and Dr. Roger Ruth (Virginia) for submitting information for this presentation.

REFERENCES


CANADIAN MODELS AND ISSUES

Martyn L. Hyde, Ph.D.

This paper addresses the design of service programs for detection and measurement of hearing loss in the neonate and young infant. The results of an informal survey of some programs in several centers throughout Canada are outlined. Most programs are based on risk registers and the auditory brainstem response (ABR) test, and exist by virtue of local expertise and initiatives. There appears to be no well-developed, consistent set of Federal or Provincial policies relating to early identification, although the principle is widely endorsed.

One program surveyed is based at Mount Sinai Hospital, Toronto. It has operated for a decade and has assessed some 4,000 high-risk infants. The program goals are to screen all at-risk infants born at Mount Sinai, provide accurate initial audiometric assessment, and to initiate management of hearing loss at or before 6 months of age. Some key features and results of this program will be assessed critically, and used to illustrate some basic issues in program design and evaluation.

The Mount Sinai program is professionally staffed. It is based on a high-risk register compiled from infant and maternal medical chart review by a registered nurse. The register used since 1982 is similar to the 1990 Joint Committee recommendations. Infants at risk are evaluated by ABR in an audiology clinic at 3 to 4 months, corrected age. Compliance for attendance has stabilized at about 90 percent of the at-risk group, for the last 4 years. Factors contributing to the good compliance include aggressive and persistent telephone followup, strong support from most primary care physicians, and integration of hearing screening with other followup services wherever possible. The rationale for screening at 3 to 4 months, as distinct from neonatal testing, is primarily a low correlation between click ABR thresholds neonatally and at 3 to 4 months of age, observed in an early, research phase of the program.

ABR testing is done during natural sleep. Click and tonepip stimuli are used. The protocol is progressive, with an initial screening component and detailed assessment if warranted. Infants with substantial threshold abnormalities usually receive a repeat evaluation after 1 to 2 months, including ABR, middle latency responses, and click evoked otoacoustic emissions. Audiometric data and recommendations are sent to the physician primarily responsible for the infant’s care. Management occurs at several neighboring institutions. The options are varied, ranging from periodic monitoring through to an intensive, multidisciplinary team approach.

All program data are computerized and form a valuable resource for both program quality management and research projects. A recent followup study of more than 700 children aged 3 to 6 years indicated that the ABR techniques used were acceptably accurate.

The clinical and research experiences at Mount Sinai Hospital are relevant to many of the well-known, basic issues in early identification program design and evaluation. A conceptual and procedural framework for developing these issues is provided by clinical decision analysis, which focuses attention on the objectives, structure, and outcomes of the various alternative program strategies. Brief discussion follows on several of these issues.
Program Goals

Goals should be defined as precisely as possible, so that their achievement or the lack of it is verifiable quantitatively. An implicit goal such as to minimize language developmental delay should be set in the context of the many other societal factors that either cause delay per se or interact with the effects of hearing impairment. A more focused goal of detecting and measuring pure-tone hearing loss leads immediately to questions about what types, severities, frequency profiles, and temporal profiles of hearing loss are to be detected, and why. The exact functional relationships between hearing loss and language delay are important and poorly understood, yet these relationships must underlie hearing loss detection targets as well as costs of detection failure and audiometric error. The precise definition of the characteristics of the target impairments, especially lower limits of severity, has a significant effect on test protocol and test accuracy estimates, as well as on program yield and followup costs.

Risk Registers

It is useful to regard the risk register as a multicomponent test protocol in series with subsequent tests such as the ABR. The basic statistical properties of registers, such as their sensitivity and specificity, are not yet well understood, nor is there a clear rationale by which to optimize their performance. Areas of interest here include the alleged upper bound of about 50 percent for register sensitivity, the development of quantitative risk indices, as opposed to simply binary risk criteria, and risk quantification for progressive impairment that will escape neonatal screening.

When To Screen

Even though there is a prevalent opinion that neonatal, predischarge screening is desirable, on the grounds of access and likelihood of postdischarge loss of contact, there are several points in favor of screening at 3 to 4 months, if a risk register is used. These include transient threshold abnormalities, expression of progressive impairments, assessment errors associated with acute concurrent disorders in the neonate, and logistical and other factors. Geographic and cultural factors may affect followup compliance, but appropriate program design and effort can produce a good response rate. When there is no clear risk factor, obtaining compliance can be difficult.

Screening Tests

ABR testing is likely to be an important component of any early assessment program. Results of followup studies indicate, however, that the accuracy of the ABR may depend quite strongly on the exact definition of the target impairment. Problem areas include the detection of mild and frequency-specific impairments. The contribution of high-frequency tonepip ABR's to the early detection of progressive sensorineural impairment is an area of interest.

For any specific definition of target impairment, program yield and ABR test accuracy also depend strongly on ABR abnormality criteria. The most sensitive aspects are the lower limits of abnormality, and the manner in which tonepip results are incorporated in the test failure criteria. These points are illustrated using ABR outcomes in about 1,500 high-risk infants.
Methodologically, so that results are valid and generalizable, it is important to use a multiparametric approach when evaluating and reporting screening test performance. Sensitivity and specificity data for various values of test failure criteria can be incorporated into relative operating characteristics (ROC's). These ROC's will change as the target disease definitions change, leading to the concept of ROC families as a basic test accuracy descriptor. An alternative that is arguably more useful in some situations is a family of likelihood ratio functions, leading more directly to a Bayesian probability revision viewpoint for the risk register and subsequent test sequence. Disease prevalence and post-test probability are central to deciding the most cost-effective assessment strategy.

Outcome Values and Costs

Any full analysis of a decision tree for assessment protocol alternatives must include not only procedural resource costs, but more importantly, the assignment of monetary values to false-negative screening errors. It may be desirable to go beyond a simple, expected value approach when allocating such costs in decision analyses. One approach to this difficult but crucial problem of outcome valuation is through a “hearing capital” health-economic model. The methodology of such an approach may include attempts to quantify the multiattribute utilities and values placed on various possible outcome states of the affected child.

Sample Size—A Caveat

A final issue is that much of the quantification required to develop programs that are effective and cost-efficient is based on statistical point estimation of proportions, such as prevalence values and test or risk factor sensitivities and specificities. Careful consideration of the sample sizes needed to produce useful estimates with acceptable precision and decision analyses with useful strategic discriminative sensitivity reveals the importance of both statistical meta-analysis and multicenter, large-sample investigations.

SUGGESTED READINGS


As elsewhere, much research and development in the United Kingdom (U.K.) on the screening of children's hearing has been driven by the "push" of technological and behavioral/physiological possibility. However, the "pull" of public health need has also been recognized, because of the requirement for proposals for new hearing screening systems. These proposals ultimately compete for limited resources within the health care system as a whole, and, in the short term, compete for research and development (R&D) funds. Public health "need" is nowadays defined as an undesired health state for which an effective solution or treatment exists.

The chief difference in health care services between the U.K. and the U.S.A. lies in the absence of family pediatricians, with the preventive child health surveillance system in the U.K. being staffed largely by health visitors (HV's), which are public health pediatric nurses who work for general practitioners (GP's) or as part of the centralized community services for a health district (population 0.2 to 0.8 million). The latter affiliation is seen especially in deprived areas, where the GP service tends to be poor or over-structured. Despite constraints on supply, comprehensive health care is (still) freely available to all, and its real costs are very low (6 to 7 percent of GNP in the U.K. as opposed to 13 percent of GNP in the U.S.A.). Health beliefs and obstacles to access, rather than a family's resources, determine the incompleteness of coverage of hearing screens.

In the previous decade, there has been much R&D activity in screening for early identification of hearing impairments in children in the U.K., a fair proportion of it at the MRC Institute of Hearing Research. Five main strands can be noted:

1. In new screening techniques and associated technologies, there have been one major development (evoked otoacoustic emissions - EOAE) and several minor developments. Recently, a modified technique has been developed that offers a significant acceleration of test time with EOAE, making the test virtually instantaneous. In some applications (possibly not at-risk neonatal screening), this technique may materially tilt the balance toward feasibility or affordability of screening programs or will shift the logistics of assessment stages post-screen or surveillance.

2. Several health services research (HSR) projects have examined the effectiveness of the U.K.'s main existing screen at 8 months. Although some excellent implementations exist, so do some very poor ones.

3. Researchers, professional organizations, and voluntary (patients') organizations have produced several good-practice guidelines for pediatric audiology that include screening as well as assessment and treatment/habituation.

4. The Health Department commissioned a review entitled Screening Children's Hearing (HMSO, 1991). This review produced a clarification of concepts and terminology, a large
literature digest as a resource, and recommendations for how screening programs should subsequently evolve. Essential public health concepts such as coverage and actual incremental yield (in addition to the traditional measurement concepts of sensitivity and specificity) are now widely understood in U.K. audiology, as is the prevalence/severity distribution.

(5) There has been a high level of activity in the development of neonatal screens on at-risk populations using the auditory brainstem response (ABR) and EOAE. Some of this monitored service development has been programmatic rather than cumulative and generalizable. However, several centers have entered a coordinated trial or have maintained stable procedures for long enough to accumulate numbers from which conclusions can be drawn. The major project of this type will soon be producing its final report.

Consensus does not exist on the definition of the target group at any age. For screening neonates at risk for hearing impairment, many favor 50 dB HI bilaterally. Controlled trials are required to determine whether a lower criterion may be appropriate. As a consequence of activities (2) to (4) above, plus limited interim publication of results and an annual series of workshops with typically more than 100 attenders associated with (5) above, there is now wide agreement in the U.K. on the following eight conclusions.

(a) Neonatal screening of children at risk (defined as neonatal intensive care >48 hours, craniofacial abnormality, or a clear family history of early childhood deafness) is feasible, with high coverage and cost-effectiveness (an added cost of about $45-60 to the NICU overall cost per child). With high coverage, about 50 percent of target children can be identified using these three risk factors. However, implementation issues such as transducer coupling, ambient noise, and staff skills (especially line management) should not be underestimated.

(b) Screening by the ABR test alone, delayed for a period to allow CNS maturation, poses problems because many infants are not brought to outpatient appointments for followup testing. When screening is not widespread, the low coverage tends to disadvantage the social strata that are at increased risk.

(c) A prescreen with EOAE, followed by a sift with an abbreviated ABR at 50 dBnHL to reduce false-positives, appears to be a highly cost-effective option for neonatal screening.

(d) A clearly attractive proposal has yet to emerge for how best to cover children not at risk in addition to children in whom hearing impairment occurs postnatally. Three alternatives currently exist in the U.K., (i) universal neonatal screening, (ii) active universal surveillance with universal priming of parental awareness, and (iii) universal behavioral screening by HV's at about 8 months. All have practical implementation difficulties and/or theoretical drawbacks from the public health perspective in terms of costs, benefits, lack of benefit, or risks. Research and development would be required to produce a satisfactory arrangement for those not at risk.
(e) Whether for otitis media or postnatal sensorineural hearing impairment, the case for a universal screen between 1 and 5 years has not been convincingly made in terms of test accuracy, case yield, cost-effectiveness, or health gain (benefit) for a definable set of target individuals. Definition of an appropriate at-risk group plus new techniques could make such provision a partial solution to problem (d).

(f) Some postnatal hearing impairments arise from conditions for which children already receive medical attention and that, although rare, have greatly increased the risk of hearing impairment, the notable instance being meningitis. To recommend “screening” for such a specific group of children would undermine useful terminological distinctions for referrals. Also, the cost savings from not routing them directly to full assessment are small, because the group is small. “Mandatory cross-referral” is our currently recommended term for what is an important concept for a cost-effective process. Although malpractice litigation is much less common in the U.K. than in the U.S.A., it may have to constitute the ultimate underpinning of the word “mandatory,” so data to substantiate practice guidelines will be essential before any at-risk conditions may be added to the list after meningitis.

(g) The existing near-universal screen at school-entry involving the pure-tone sweep test has value educationally and as a safety net to catch any deficiencies of the earlier screening system in the overall public health provision.

(h) No convincing public health evidence exists favoring universal screening at any age after 5 to 6 years, until, perhaps, 50 years of age.
NOTICE

REPRODUCTION BASIS

☐ This document is covered by a signed “Reproduction Release (Blanket)” form (on file within the ERIC system), encompassing all or classes of documents from its source organization and, therefore, does not require a “Specific Document” Release form.

☑ This document is Federally-funded, or carries its own permission to reproduce, or is otherwise in the public domain and, therefore, may be reproduced by ERIC without a signed Reproduction Release form (either “Specific Document” or “Blanket”).