

# **Sound Beginnings:**

**Changing the World for Children with Hearing Loss**



## **Disclosures: Karl White, PhD**

**I have no relevant financial relationships with the manufacturer/s of any commercial product/s and/or provider of commercial services discussed in this CME activity.**

**I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.**

**Disclosures:**  
**Susan Wiley, MD**

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**Learning Objectives**

- Explain how to screen for hearing loss, describe the different degrees of hearing loss, and identify the risk factors of hearing loss
- Apply the strategies identified to screen by 1 month, identify by 3 months, and enroll into early intervention by 6 months children who are deaf or hard of hearing
- Identify follow-up strategies and procedures for children who didn't 'pass' their NBHS

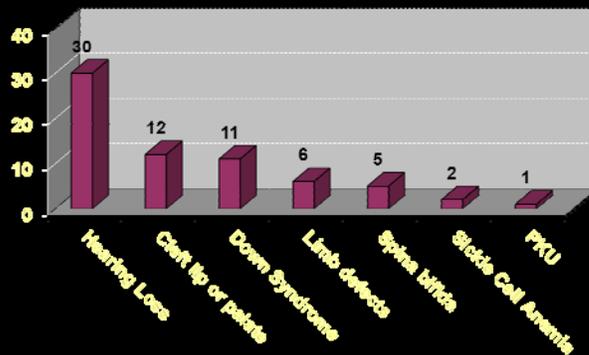


## Key Points

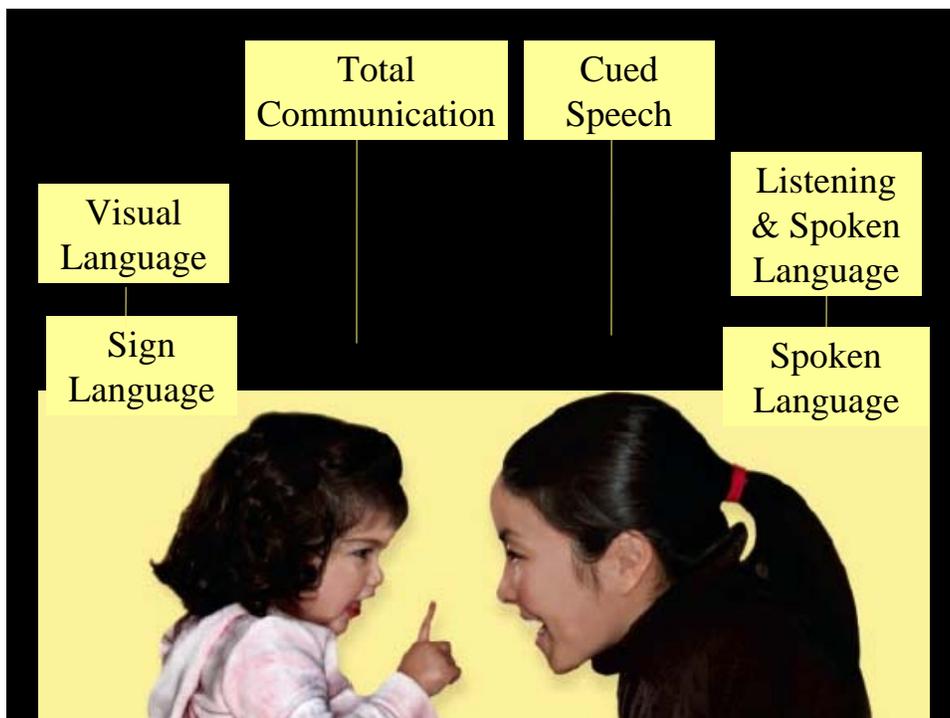
1. Most parents with a newly identified deaf child are completely surprised
2. Many of the professionals from whom parents seek help are not up-to-date
3. The most important thing to parents is to be able to COMMUNICATE with their child

## Little Known Facts About Congenital Hearing Loss

- Permanent hearing loss is the most frequent birth defect in the United States.



- If hearing loss is not identified and treated early, deaf children will require an additional \$400,000 per child in educational costs.

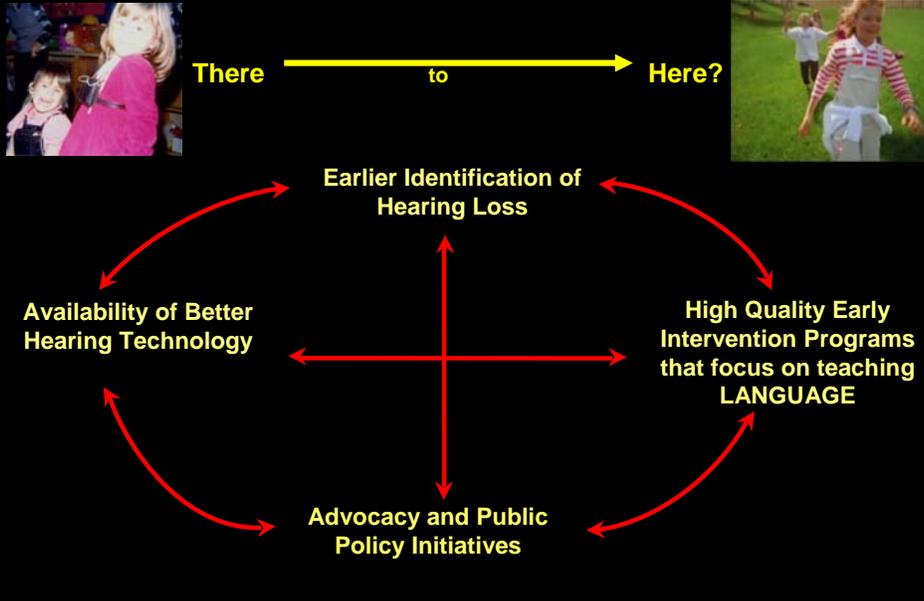




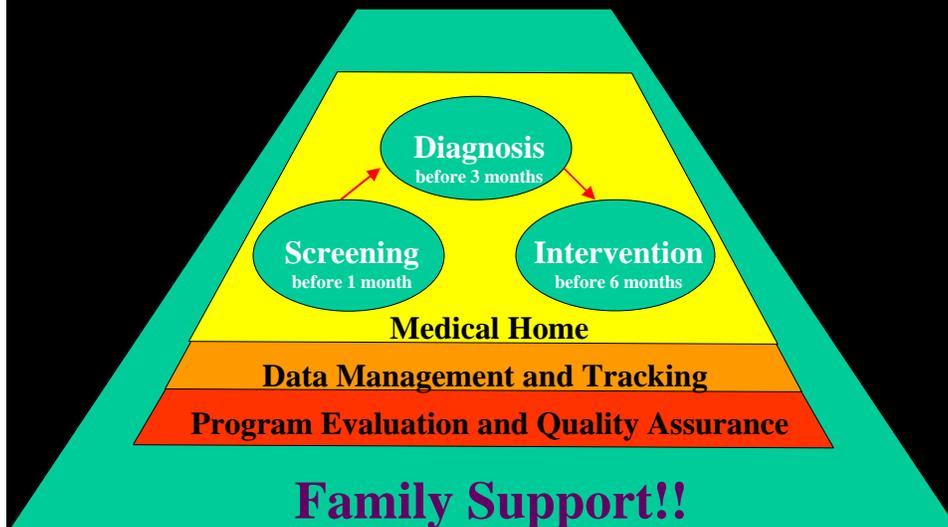




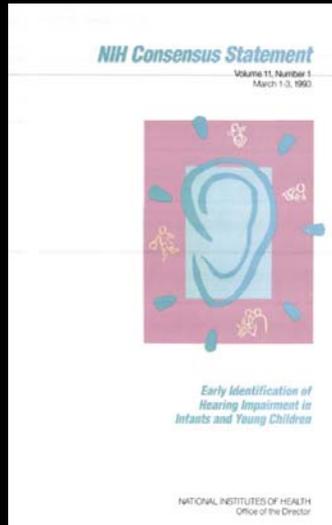
## What enabled us to move from ....



## Components of an Effective Early Hearing Detection and Intervention (EHDI) Program



**In March 1993, a Consensus Development Panel convened by the National Institutes of Health concluded that:**



- *“The average diagnosis of hearing impairment remains constant at about 2 ½ years of age.*
- *All infants should be screened for hearing impairment...this will be accomplished most efficiently by screening prior to discharge from the well-baby nursery.*
- *Identification of hearing impairment must be seen as imperative for all infants.”*



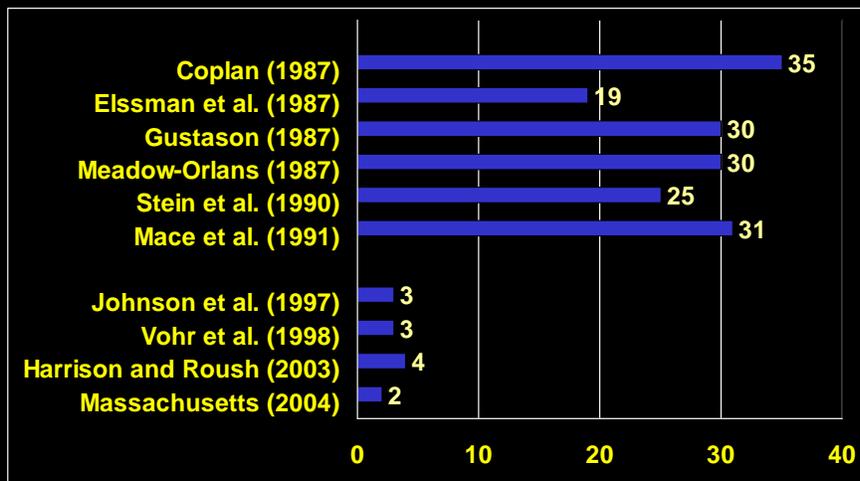
## Why is it important?

- **It meets the prerequisite for screening**
  - Condition sufficiently frequent in screened population
  - Condition serious or fatal without intervention
  - Condition must be treatable or preventable
  - Effective follow-up program possible
- **Early intervention for hearing loss improves language outcomes**

## Rate Per 1,000 of Permanent Childhood Hearing Loss in EHDI Programs

Site	Sample Size	Prevalence Per 1000
Texas (Finitzo et al 1998) (1/94 to 6/97)	54,228	2.15
Colorado (Mehl & Thomson, 1998) (1/92 - 12/96)	41,976	2.56
New Jersey (Barsky-Firsker & Sun) 1/93-12/95)	15,749	3.30
Hawaii (Johnson et al 1997) 1/96 - 12/96)	9,605	4.15
Massachusetts (2004) (1/04 – 12/04)	78,515	2.87

## Age in Months at Which Permanent Hearing Loss Was Diagnosed



## Characteristics of a good screening program

- Refer rate of 1.5-5.0% in well baby nursery and slightly lower in the NICU (resulting from 2-stage screening in the hospital)
  - 5.0% = 400 babies per 8000 births
- Ongoing training and monitoring program for personnel
- Structured plan for follow up
- Ability to track program performance (important for quality assurance and for JCAHO requirements)

## **Hearing Screen Protocols**

- **Screening results should be conveyed immediately to families so they understand the outcome and the importance of follow-up when indicated**
- **For rescreening, a complete evaluation of both ears is recommended, even if only 1 ear failed the initial screen**

## **Hearing Screen Protocols**

- **Separate protocols are recommended for NICU and well baby nurseries.**
- **NICU babies >5 days are to have ABR included as part of their screen so that neural HL will not be missed**

## 2007 JCIH Position on Screening

- NICU
  - >5 days in NICU
  - ABR should be included to screen for neural loss
  - Rescreen BOTH ears, even if only one ear fails
  - Non pass – refer to Audiologist
  - Readmission – rescreen before discharge
- Well baby nursery
  - Screen with OAE or ABR
  - Repeat screen when necessary before discharge
  - When using 2 step protocol test order should be OAE then ABR
  - Rescreen BOTH ears, even if only one ear fails

## Audiograms

Bilateral Profound Sensorineural Hearing Loss

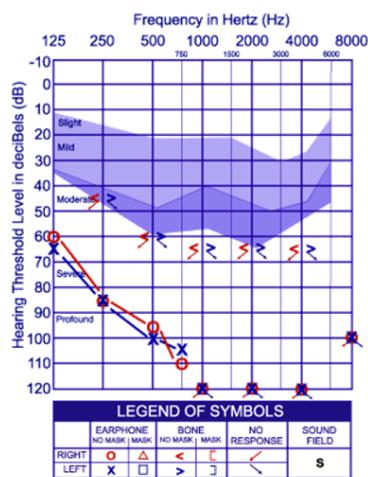


Image courtesy of EDEN - The Electronic Deaf Education Network

### Bilateral Mild to Moderate Sensorineural Hearing Loss

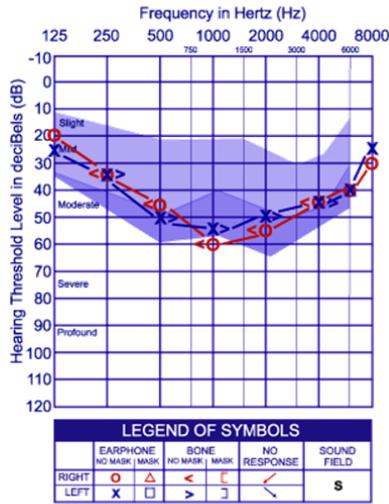
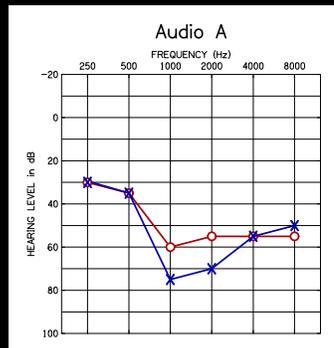
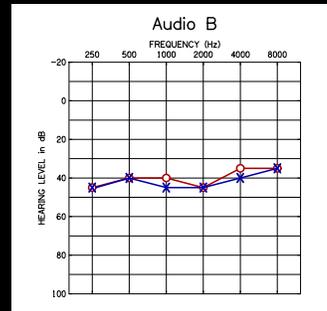


Image courtesy of EDEN - The Electronic Deaf Education Network

### 3 year old with moderate-severe loss: Inconsistent Intervention

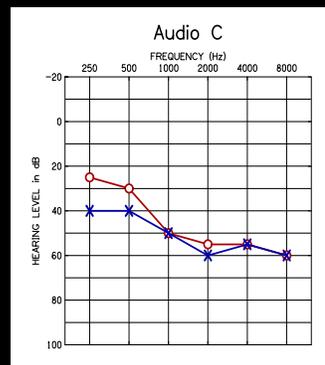


## 3 year old with moderate-severe loss: Consistent early intervention



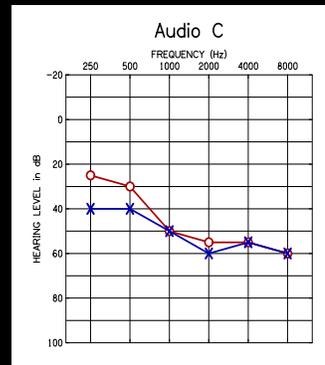
## 3 year old with mild-moderate loss: Identified at 3 years, 3 months

Pre-intervention sample



## 5 year old with mild-moderate loss: Identified at 3 years, 3 months

Post-intervention sample



### Risk Indicators for permanent congenital, delayed onset, or progressive hearing loss

- **Caregiver concerns\***
  - about hearing, speech, language, development
- **Family history\***
  - of permanent childhood hearing loss
- **NICU stay > 5 days or any of following** (regardless of length of stay):
  - ECMO assisted ventilation\*
  - Ototoxic medications (gentimycin, tobramycin)
  - Loop diuretics (furosemide, Lasix)
  - Hyperbilirubinemia requiring exchange transfusion

JCIH, 2007      \* = greater risk for **delayed** onset HL

## **Risk Indicators for permanent congenital, delayed onset, or progressive hearing loss**

- **In Utero infections**
  - CMV\*, herpes, rubella, syphilis, toxoplasmosis
- **Craniofacial anomalies**
  - especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- **Physical findings associated with a syndrome known to include permanent HL (e.g. white forelock)**
- **Syndromes\* involving hearing loss**
  - Neurofibromatosis, osteopetrosis, Usher, Waardenburg, Alport, Pendred, Jervell & Lange-Nielson

\* = greater risk for **delayed** onset HL

## **Risk Indicators for permanent congenital, delayed onset or progressive hearing loss**

- **Neurodegenerative disorders**
  - Hunter syndrome
  - Sensory motor neuropathies (Friedrich ataxia, Charcot-Marie-Tooth)
- **Culture positive postnatal infections associated with HL\***
  - Herpes, varicella, meningitis
- **Head trauma (basal skull, temporal bone)\***
- **Chemotherapy\***

\* = greater risk for **delayed** onset HL

## What Contributes to “Loss to Follow-up”?

- **Referral rates in the hospital are too high** (because of poorly trained screeners, poorly maintained equipment, lack of commitment, etc)
- **Ineffective information for parents** (about initial results, need for follow-up, what to do next, etc)
- **Accurate data isn’t shared quickly with the right stakeholders** (hospitals, state EHDl program, medical home, audiologists, early interventionists, etc)
- **Shortage of pediatric audiologists** (because of not enough training programs, poor reimbursement rates, rural/remote residences, etc)
- **Lack of knowledge about current “effective practices”** (among program managers, health care providers, early interventionists, etc).
- **Not enough public awareness about importance of issue** (taxpayers, administrators, extended family, etc)
- **Lack of resources** (for screening, follow-up diagnosis, early intervention, case management, etc)

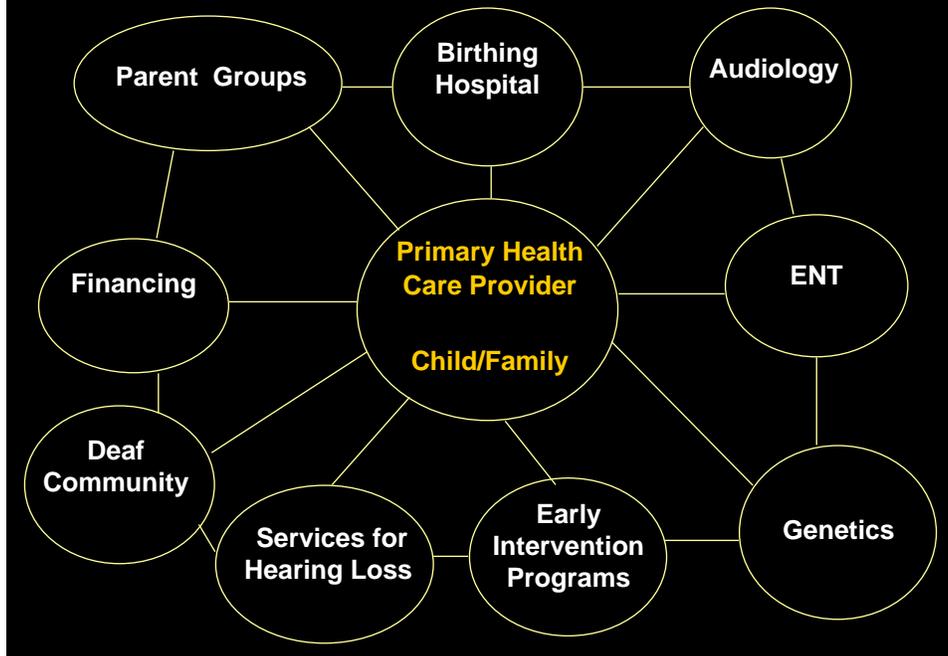
## The Hearing Head Start Project

- Feasibility study from 2001-2004
- 69 programs in 3 states with 3,000+ children screened
- Identified 2 per 1,000 with permanent hearing loss and 20 per 1,000 with unidentified transient losses
- **Currently in 21 of 50 states—expanding to others by 2015**



Eiserman WD, Hartel DM, Shisler L, Buhrmann J, White KR, and Foust T. (2008). Using otoacoustic emissions to screen for hearing loss in early childhood care settings. *International Journal of Pediatric Otorhinolaryngology*, 72, 475-482.

## EHDI and the Medical Home



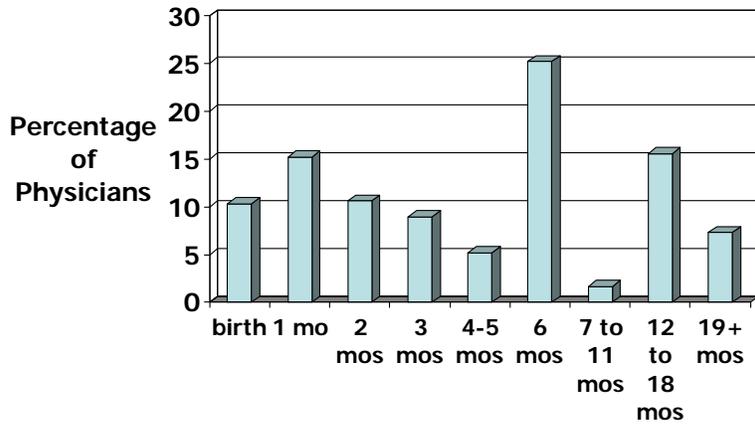
## Educating Primary Health Care Providers About Early Identification of Hearing Loss

Assume a newborn for whom you are caring is diagnosed with a moderate to profound bilateral hearing loss. If no other indications are present, to which specialists would you refer the baby?:

	Always or Often
Ophthalmological evaluation	0.6%
Genetic evaluation	8.9%
Otolaryngological evaluation	75.6%

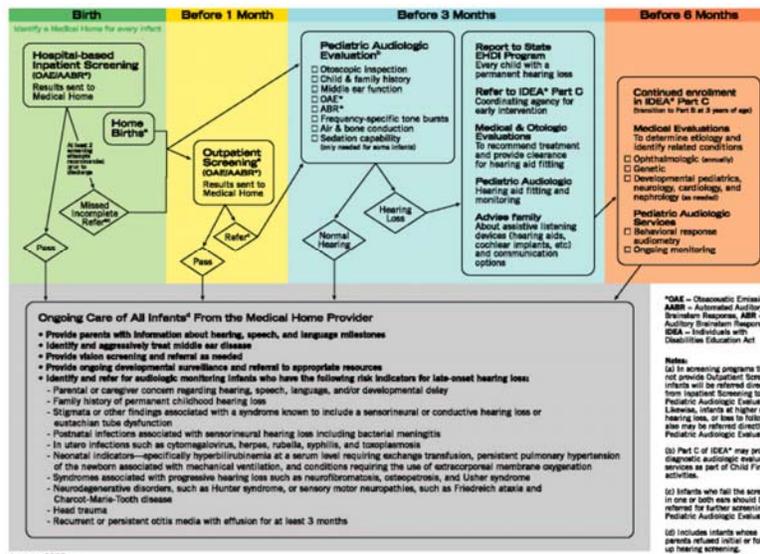
Moeller MP, White KR, & Shisler L (2006). Primary care physicians' knowledge, attitudes and practices related to newborn hearing screening. *Pediatrics*. 118, 1357-1370.

# When can an infant be fit with hearing aids?

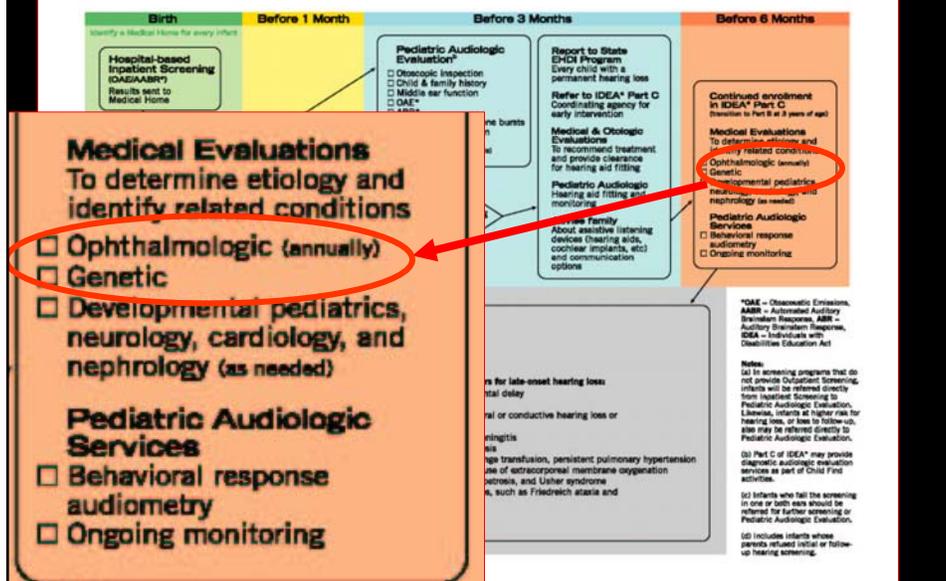


## American Academy of Pediatrics

### Universal Newborn Hearing Screening, Diagnosis, and Intervention Guidelines for Pediatric Medical Home Providers



## Universal Newborn Hearing Screening, Diagnosis, and Intervention Guidelines for Pediatric Medical Home Providers



## Medical Home: Strategies to Promote Follow Up

- At prenatal visit, encourage families to identify you as follow-up care location
- Inform hospital to facilitate communication of results (or set up system to receive birth information)
- Add checkbox on newborn well child form/patient chart/EMR for hearing screening results and risk factors
- At 1 week visit, make sure you have the screening results
- In children who did not pass their hearing screen, Confirm audiologist appointment with parents at time of PCP visit

## Medical Home: Strategies to Promote Follow Up

- Set up tracking system for infants who do not pass hearing screening
- Set up tracking system for children with identified risk factors for HL
- Encourage communication between providers (audiology, ENT, speech pathology, Early Intervention, etc)

National Initiative for Children's Healthcare Quality



### AAP: online module on Hearing Loss

[www.pedialink.org](http://www.pedialink.org)

- Web-based learning:
  - medical home in screening, follow-up, management, referral, and family support of children with hearing loss
  - the rates of hearing loss and understand the importance of early recognition and the ongoing screening/surveillance process
  - the differences between screening methods available for hearing loss and the audiologic evaluation process in infants and children
  - different types of hearing loss including the medical work-up of a child with an identified hearing loss
  - understand the risk factors of hearing loss, including the differences between medical and genetic risk factors

BOYS TOWN National Research Hospital

my baby's hearing

NIDCD

Versión en español

first steps  
newborn screening

my baby's hearing

Hearing & Amplification

Language & Learning

Parent to Parent

next steps  
my baby has a hearing loss

newborn screening

all about hearing loss

hearing aid choices

cochlear implants

causes of hearing loss

glossary

search

getting started: what is early intervention?

building our support team

decisions...decisions

building conversations

building concepts

positive parenting

learning from my family

learning through play

read with me

getting ready for school

search

coping with the diagnosis: parent views

how do we decide?

successes...great & small

what does the future hold?

working with professionals

parent talk

search

www.babyhearing.org/

CDC Department of Health and Human Services  
Centers for Disease Control and Prevention

Early Hearing Detection & Intervention (EHDI) Program

- **Educational Materials:**
  - What to ask your.... series
  - A parent's guide to HL
  - Guide to Early Intervention
  - Guide to Genetics and HL

www.cdc.gov/ncbddd/ehdi/

Topic Contents

- > [EHDI Home](#)
- > [EHDI Information](#)
- > [Research](#)
- > [Resources](#)
- > [State Information & Profile](#)
- > [What's New](#)

Quick Links

- [FAQ's About Hearing Loss](#)
- [State & Territory EHDI Contacts](#)
- [Mild & Unilateral](#)
- [2006 EHDI Data](#)
- [Abbreviations & Acronyms](#)
- [Contact Us](#)

Programs & Campaigns

- [Public Service Announcements](#)

Click here for new Educational Materials

EHDI Pubs Search

# What to ask your....

Audiologist, ENT, Medical Provider, SLP, Geneticist

**Questions You May Want to Ask Your Child's Early Interventionist**

Name of Early Interventionist: \_\_\_\_\_  
Phone/Contact Information: \_\_\_\_\_  
Appointment Date: \_\_\_\_\_  
Next Appointment Date: \_\_\_\_\_

Early intervention is a program for children from birth to 3 years of age who have a developmental delay. Some states also provide services for children who are "at risk" for developmental delay. Children with hearing loss typically need early intervention services. An early interventionist, a specialist who works with infants and toddlers, will help identify your child's needs and create an Individualized Family Service Plan (IFSP). This plan will be used to provide your child with the services he or she needs.

Early intervention services support families to help their children reach their full potential. These services are offered through public or private agencies. Your child may receive services at your home, a clinic, a daycare center, a hospital, or the local health department. States decide which children can receive services, but they follow rules under a federal law called "Part C" of the Individuals with Disabilities Education Improvement Act (IDEA). These services might be very helpful to your child.

Each state has an agency that serves infants and toddlers with hearing loss or other special needs. Once your child has been diagnosed with hearing loss, an early interventionist or someone with the state agency should call you. If you do not receive a call, or would like to know more about intervention services in your state, you can call the state office and ask to speak with the agency that serves children with special needs. The state number can be found in the blue colored pages of your local phone directory, under "State Government."

It is important that children with hearing loss begin early intervention services as soon as possible. The goal for every child with hearing loss identified at birth is to start early intervention no later than 6 months of age or within 3 months of diagnosis. This will help the child develop communication and language skills that will last a lifetime.

Department of Health and Human Services

**Early Interventionist Questions**

Questions about my/our child's early intervention services:

1. What is early intervention? What can you do for my child? What services do you provide?
2. Why is it so important for my child to start intervention this early?
3. How much will early intervention services cost?
4. How can my child learn to communicate? Can you tell me about sign language? Can you tell me about the different ways my child can learn to talk?
5. How are hearing aids and cochlear implants different?
6. How will I know if my child should get hearing aids or a cochlear implant?\*

\* This question can also be asked of your audiologist, ear nose and throat doctor (ENT), or other medical professionals.

After talking with the interventionist, I/we learned:

## Mild and Unilateral Loss

[www.cdc.gov/ncbddd/ehdi/unilateralhi.htm](http://www.cdc.gov/ncbddd/ehdi/unilateralhi.htm)

Early Hearing Detection & Intervention (EHDI) Program

EHDI > Mild and Unilateral Hearing Loss

### Mild and Unilateral Hearing Loss



- [Research Announcements](#)
- [National Workshop on Mild and Unilateral Hearing Loss \(2006\)](#)
  - [Agenda \(PDF\)](#)
  - [Proceedings \(PDF\)](#)
- [Mild and Unilateral Hearing Loss: Summaries of Research Articles](#)
- [Frequently Asked Questions about all types of hearing loss](#)
  - [What is the type and severity of hearing loss in infants throughout the United States?](#)
- [Helpful Links](#)
- [Recent Articles](#)

Downloadable pdf of the entire workshop

Tables of literature (evidence tables)

Here's the research





Search [infanthearing.org](http://infanthearing.org)

Home | Components | Resources | States | Legislation | About Us

**News and Events**

We have a new look! All of our information and resources are still available.







(((NCHAM serves as the National Resource Center for the Implementation and Improvement of comprehensive and effective Early Hearing Detection and Intervention (EHDI) systems. As a multidisciplinary Center, our goal is to ensure that all infants and toddlers with hearing loss are identified as early as possible and provided with timely and appropriate audiological, educational, and medical intervention.

**EHDI Components**

- Newborn Hearing Screening
- Early Childhood Hearing Screening
- Diagnostic Audiology
- Early Intervention
- Family Support
- Medical Home
- Data Management
- Financing & Reimbursements
- Program Evaluation



**State EHDI Information**

- Status of the United States
- State Profiles
- Web Sites & Guidelines
- EHDI Contacts
- 2004 State EHDI Survey
- State Coordinator Toolbox



**EHDI/UNHS Resources**

- UNHS Implementation Guide
- Addressing Privacy Regulations
- Position Statements
- EHDI/UNHS FAQ
- Slideshow Presentations
- Educational and Training Videos
- Fact Sheet [PDF]
- NCHAM Materials
- EHDI Implementation in Latin America
- EHDI E-Book
- More EHDI/ UNHS Resources...



**EHDI Legislation**

- State Legislation
- Rules & Regulations
- Legislative Summaries
  - By State: Table | Text
  - By Provisions



**EHDI E-Book**

The EHDI E-Book is Now Available to Download.



**NHSTC DVD**

Our Newborn Hearing Screening training curriculum DVD is now available.





# HANDS & VOICES

"What Works for your Child is what makes the Choice right"

How To Help

Home | About Us | Resources | Services | Chapters | Events | Contact Us

<b>Communication Considerations A to Z™</b>	Communication Considerations A to Z™ is a series from Hands & Voices that's designed to help families and the professionals working with them access information and further resources to assist them in raising and educating children who are deaf or hard of hearing.
<b>"What Parents Need to Know" Series</b>	The "What Parents Need to Know" Series discuss topics that parents often ask about.
<b>In a Perfect World</b>	In this series of articles, you'll hear from the Executive Director of Hands & Voices, LeeAnne Seaver on the issues of the day, and reflections about our kids.
<b>Early Intervention</b>	Support for parents that are going through the beginning stages of learning about the world of deaf / hh issues.
<b>Education</b>	Inclusion, Deaf Ed Reform, The Law, Advocacy - issues related to Schools and Education in general.
<b>Parent / Professional Collaboration</b>	As we make decisions about our kids future we need to remember that we will be collaborating with caring professionals whose job it is to guide and educate us about the many choices we make for our kids.
<b>Family Perspectives</b>	The views of Parents who've been there.
<b>Technology</b>	The latest articles on the latest research and the changing technology and how it affects our kids.
<b>Research</b>	Research oriented information on current deaf / hh issues.
<b>Deaf People &amp; Perspectives on Deafness</b>	Other issues that are of interest to families of children who are deaf / hh.

[www.handsandvoices.org/](http://www.handsandvoices.org/)

For tips on talking to your baby, read our page on [communicating with your baby](#).

#### [Communicating with your child](#)

Once your child is a bit older, there are 5 main ways he can learn to communicate:

- **Learn to use his hearing and speak.**
  - This is called the [auditory-oral method](#).
  - This method teaches children to listen, read lips and speak.
- **Learn to use the hearing he has left.**
  - This is called the [auditory-verbal therapy](#).
  - This method helps children use whatever hearing they have left.
  - This method does not encourage lipreading.
- **Learn American Sign Language**
  - This is called [bilingualism/biculturalism](#).
  - With this method, children learn American Sign Language and become part of Deaf Culture.
- **Learn to use handshapes that stand for sounds.**
  - This is called [cued speech](#).
  - This method gives people a way to "see" English.
  - It uses handshapes to "show" the sounds you can't see by reading lips.
- **Learn to use some of these methods together.**
  - This is called [total communication](#).
  - This approach uses different ways of communicating together, like signing and talking at the same time.

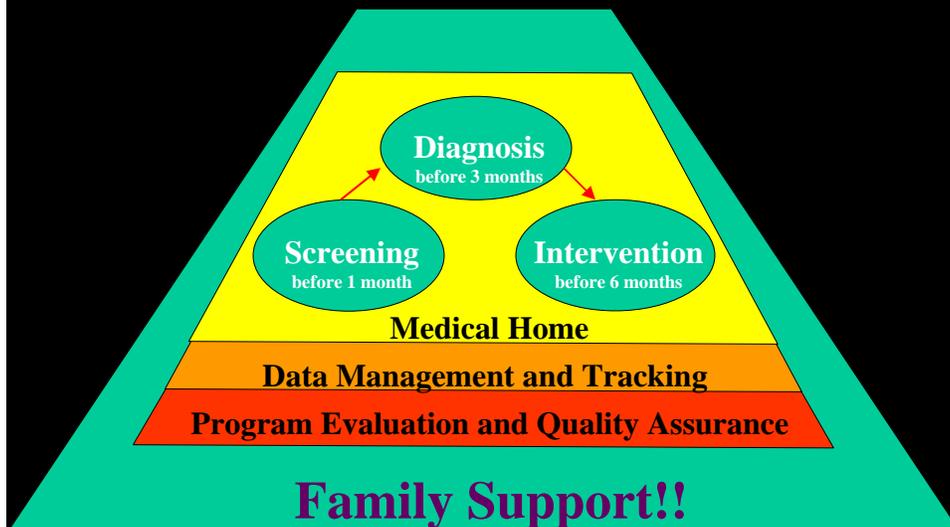
[How to choose a way to communicate](#)

**Questions?**

## Question #1

**What are specific, evidence-based strategies for strengthening the communication and reporting between the state newborn hearing screening program and the medical home?**

## Components of an Effective Early Hearing Detection and Intervention (EHDI) Program



### Answer #1

- State EHDI programs recognize the importance of and do their best to notify the medical home of a child's hearing screening results.
- This works best when the primary care pediatrician is identified prior to a child's hearing screen, or prior to the child's discharge from the hospital.
- Primary care pediatricians can help by verifying that their practice contact information is in the birthing hospital's/center's record, and that they have been identified as the primary health care provider for that child.
- With various policies and programs across the country, there isn't a "one-size fits all" system at this time.
- We have a long way to go in improving the link between the newborn hearing screening programs and the medical home, and we need YOUR help!

### Question #2

**What early intervention services are recommended for children with hearing loss, and when is the optimal time to begin utilizing these services?**

## Answer #2

- As soon as hearing loss is identified, children should be enrolled in early intervention (EI). Even very young children (infants at 2 months of age) can benefit from early intervention. This is usually based primarily on parent training in early infancy on what to expect and how to communicate with your child.
- States and counties' EI programs vary greatly from region to region. Preferably, early intervention providers would be knowledgeable in educational strategies for children who are deaf or hard of hearing. The Ski\*Hi curriculum has been used by many states to evaluate their programs:
  - Who are the providers?
  - How often are the service visits?
  - Are families feeling supported?
  - Are the children making progress?
- Additional services (such as occupational therapists, physical therapists, and/or behavioral specialists) should be provided when concerns arise. Having a screening system in place for children enrolled in Early Intervention for additional needs is appropriate.

## Question #3

**What are the best data tracking systems used by hospital systems and/or state programs?**

### **Answer #3**

- **The Centers for Disease Control and Prevention has given grants to almost all states to assist in developing data tracking systems.**
- **Types of tracking systems:**
  - Commercial products
  - State system by state IT departments
  - Built-in component of the medical record
- **Screening, Diagnosis, and Intervention referral records need to be transmitted to the entire care team: the medical home, audiologist, and the early intervention program.**
- **The most progressive systems have now automated this process. The primary care provider will receive an email or letter with the records/results and the next steps that must be taken.**

### **Question #4**

**How do we screen preschoolers for hearing loss in a pediatric practice?**

**What equipment is needed?**

## **Answer #4**

- **The age (1 – 5 years old) of the preschooler is a primary factor in determining the best type of screen to provide.**
- **The Administration on Children and Families funded a national project to provide early intervention programs with auto-acoustic emission equipment to screen children's hearing.**
  - **2 / 1,000 children were found to have permanent hearing loss that would not have otherwise been identified**
  - **20 / 1,000 children were found to have fluctuating conductive hearing loss that would not have otherwise been identified**
- **Some pediatricians offer in-office screening as part of the well-child visit.**
- **Day care and other early childhood programs often use behavioral screening (and sometimes auto-acoustic emission).**
- **All caretakers in a child's life need to consider hearing screening if a child's language development is lagging behind norms.**