

# Early Hearing Detection and Intervention (EHDI) Program

## 2006 Annual Report



### New AAP EHDI Chapter Champion appointed.

Congratulations to Dr. Brenda Balch!

In 2001, the American Academy of Pediatrics (AAP) implemented a program, "Improving the Effectiveness of Newborn Hearing Screening, Diagnosis and Intervention through the Medical Home," which focuses on increasing the involvement of pediatricians, primary care physicians, and other child health care providers in the early hearing detection and intervention (EHDI) process, by linking follow-up services more closely to the newborn's medical home.

The program represents a National AAP and Chapter partnership and is funded by the Maternal and Child Health Bureau, Health Resources and Services Administration and the Centers for Disease Control and Prevention.

Dr. Brenda Balch, a pediatrician from New London, CT, was appointed by the Connecticut AAP Chapter to be the state's Early Hearing Detection and Intervention (EHDI) Chapter Champion, and to serve as the point person on EHDI issues at the state and local level.

*Hats off to Dr. Balch!*



### History of Hearing Screening in Connecticut

**1985** - Connecticut legislation required screening "at risk" infants for hearing loss.

**1999** - Connecticut passed legislation for universal screening of all newborns.

**July 1, 2000** - Universal hearing screening was implemented statewide.

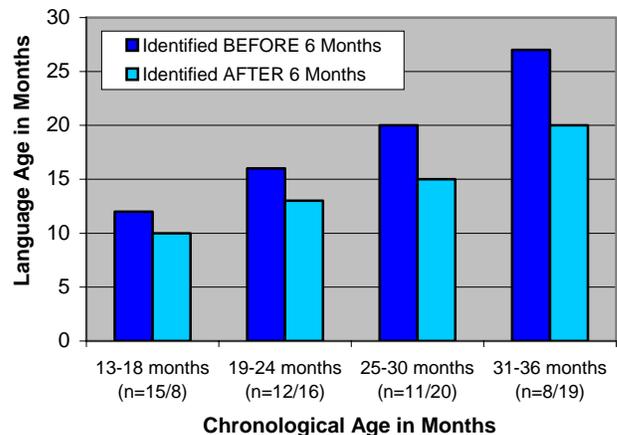
**2006** - To date, a total of **208,485** babies have been screened and **398** babies have been diagnosed with hearing loss, since Connecticut began universal newborn hearing screening.

### Benefits of Early Hearing Detection & Intervention

Research has shown that children born with a hearing loss who are identified early and given appropriate intervention before six months of age, demonstrate significantly better speech and reading and language comprehension than children identified after six months of age.

(Yoshinaga-Itano et al, Joint Committee on Infant Hearing Committee Meeting, 1996)

Language Comprehension Scores for Hearing Impaired Children Identified Before and After 6 Months of Age



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# NEWBORN HEARING SCREENING

## Screening Equipment

There are two objective physiologic measures used to detect targeted hearing loss in newborns and infants. Both technologies are noninvasive recordings of physiologic activity that underlie normal auditory function and that are easily recorded in neonates.



### Otoacoustic emissions (OAE):

Either transient evoked (TEOAE) or distortion-product (DPOAE). OAEs are sensitive to outer hair cell dysfunction and measures responses generated by the cochlea. OAE measures response to stimuli in the frequency range greater than 1500 Hz. OAE will not detect neural dysfunction.



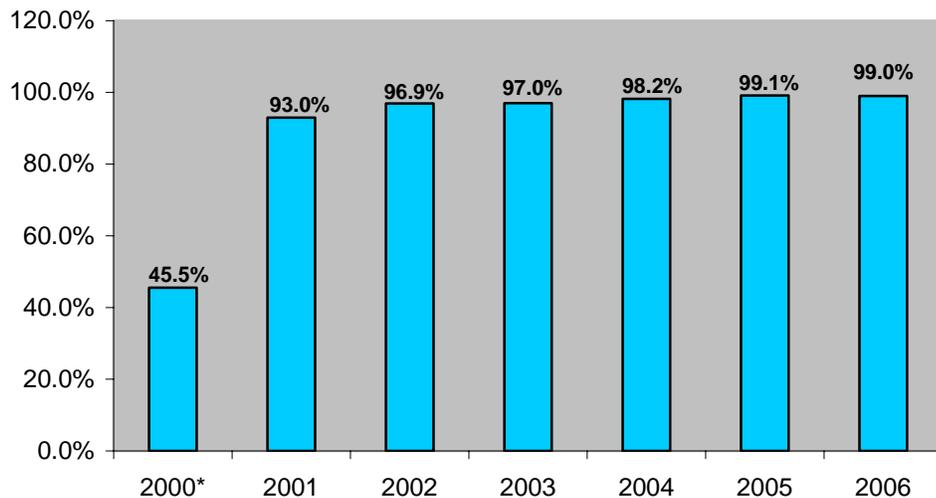
### Automatic Brainstem (ABR):

The ABR reflects activity of the cochlea, auditory nerve, and auditory brainstem pathways. When used as a threshold measure, the click-evoked ABR is highly correlated with hearing sensitivity in the frequency range from 1000 Hz to 8000 Hz.

## Connecticut Hospital Screening Guidelines (in part)

- NICU babies: Must have an ABR screen before discharge.
  - Well babies: Initial screen may be OAE or ABR.
  - All babies: If the baby does not pass the first screen it must be repeated before discharge using ABR.
- DO NOT OVERSCREEN, as it increases the likelihood of a false negative result.

## Connecticut Annual Hearing Screening Rates



\* Universal screening began 7/1/2000

Source: DPH EHDl Program database

## DIAGNOSTIC EVALUATION & FOLLOW-UP



Approximately 1-2% of all infants screened will be referred for diagnostic hearing testing. The initial diagnostic hearing evaluation should be scheduled promptly, prior to or upon discharge from the hospital, and should be conducted by the time the child is 3 months of age.

The child should be referred to an audiologist with the skills, equipment and expertise necessary to evaluate newborns and young infants. The audiologist should also be capable of providing audiological diagnostic testing and auditory habilitation services (amplification, selection and fitting).

The audiologist must decide on the **battery of tests** that is appropriate for each child based on: screening results, the medical history of the child, risk factors (if present), and the type, number, and timing of screenings carried out prior to the referral.

At least, an ABR is recommended as part of a complete audiological diagnostic evaluation for children under age 3 for confirmation of a permanent hearing loss.

All NICU infants who do not pass the ABR screen in the hospital should be referred to an audiologist for a comprehensive evaluation, including an ABR.

All re-screenings done as part of the diagnostic testing process should include both ears, even if only one ear did not pass the initial screen.

⇒ For a complete listing of diagnostic testing centers in CT, visit our website:  
[http://www.dph.state.ct.us/BCH/ehdi/dx\\_testing.htm](http://www.dph.state.ct.us/BCH/ehdi/dx_testing.htm)

### Connecticut Average Age at Diagnosis

National Goal = 3 months

Year	Average Age at Diagnosis (in months)
2000	1.65
2001	3.06
2002	1.78
2003	1.38
2004	2.63
2005	2.79
2006	2.42

2006 (n=67)	
Minimum Age:	0.2 months
Median Age:	1.8 months
Maximum Age:	9.0 months

### Types of Hearing Loss Identified in 2006

(n=67)

	Sensorineural	Conductive	Mixed	Undetermined	Auditory Neuropathy
<b>Bilateral n=36</b>	14	6	1	14	1
<b>Unilateral N=31</b>	7	9	0	15	0

## THE MEDICAL HOME & ROLE OF THE PRIMARY CARE PROVIDER

Pediatricians, family physicians, and other professionals, working in partnership with parents and other professionals, make up the infant's "medical home" (American Academy of Pediatrics, 2002). A *medical home* is defined as an approach to providing health care services where care is accessible, family-centered, continuous, comprehensive, coordinated, compassionate, and culturally competent.

The primary care provider acts in partnership with parents or other caregivers in a medical home to identify and access appropriate audiology, intervention, and consultative services needed in developing an appropriate plan for the habilitative care for infants identified with hearing loss, and infants with risk factors for hearing loss.

Responsibilities of the primary care provider include:

- Verifying that the hearing screening was conducted at birth
- Ensuring that an audiological assessment is conducted on all babies who do not pass the screening
- Initiating referrals for medical specialty evaluations necessary to determine the etiology of the hearing loss
- Confirming that a referral has been made to Birth to Three (*Visit: <http://www.birth23.org> for more information.*)
- Partnering with other specialists, including the otolaryngologist to facilitate coordinated care
- Monitoring middle ear status for the presence of effusion
- Providing ongoing surveillance of **all** children for speech, language and hearing, auditory skills and developmental milestones
- Reviewing each child's medical record and family history for the presence of risk indicators that require monitoring for delayed onset or progressive hearing loss and ensuring that an audiological assessment is conducted every 6 months up to age three.

**\*\* Remember:**

Not all hearing loss occurs at birth!

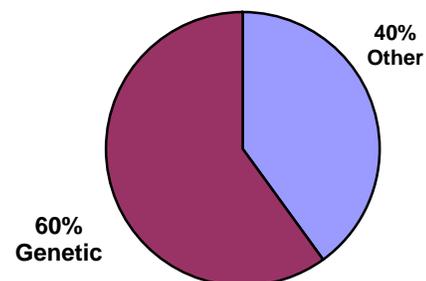
Any child with speech or language delays, or caregiver concern, should promptly be referred for an audiological evaluation.

### Genetics and Hearing Loss

Genetic factors are associated with 70% of congenital cases of hearing loss.

Of this group...

- ⇒ **40%** are syndromic, cases in which deafness can be diagnosed because of associated clinical findings or syndromes.
- ⇒ **60%** are classified as nonsyndromic and may be caused by a single gene mutation.



**The PCP should refer any child with a permanent congenital hearing loss to a geneticist for an evaluation.**

Genetic testing can assist in:

- ♦ Establishing an etiologic diagnosis for the hearing loss.
- ♦ Deciding how to best treat the hearing loss
- ♦ Determining whether certain medications should be avoided

The GJB2 gene (Connexin defects) are responsible for more than half of the genetic causes of profound deafness in the United States. Testing for GJB2 first may eliminate the need for more extensive testing.

Early identification of patients with certain forms of syndromic deafness can dramatically alter the risk of serious complications for the affected infant and other family members.

# EARLY INTERVENTION

## Early Intervention Services for Children with Hearing Loss

Research demonstrates that the sooner children with hearing loss are enrolled in an early intervention program, the more positive their developmental outcomes .

All children diagnosed with a hearing loss should be referred **to Birth to Three**, Connecticut's early intervention program. The referrals are made by calling the Child Development Infoline, **1-800-505-7000**. Children are normally referred by the audiologist at the time of diagnosis, but anyone, including the child's primary care provider, hospital staff or family member can refer a child.

Birth-to-Three has the following three specialty centers that work with infants and children who are deaf and hearing impaired:

- American School for the Deaf
- CREC/Soundbridge
- New England Center for Hearing and Rehabilitation

Each of the three specialty centers offer services to assist with family support and education, audiological monitoring and enhancing children's communication skills to promote optimal learning and development.

### Connecticut Average Age at Enrollment in Birth to Three

National Goal = 6 months

Year	Age at Enrollment (in months)
2000	2.13
2001	3.44
2002	2.64
2003	3.23
2004	3.83
2005	4.09
2006	3.10



<u>2006 (n=67)</u>	
Minimum Age:	0.6 months
Median Age:	3.2 months
Maximum Age:	8.5 months

### The Birth to Three Referral

All children with **any** type and degree of hearing loss should be promptly referred to Birth to Three.

This includes children with a:  
 undetermined type hearing loss,  
 unilateral hearing loss,  
 bilateral hearing loss, and/or  
 a mild hearing loss.

Birth to Three referrals are made by calling Child Development Infoline, **1-800-505-7000**.

## 2006 EHDI Program Highlights

The Connecticut Department of Public Health Early Hearing Detection & Intervention program, in conjunction with the University of Connecticut Health Center and UConn School of Medicine, released ***Newborn Screening in Connecticut***, a new web-based training for health care providers who care for infants and children.

The training is free and offers up to 4.5 continuing medical education credits and 3.7 nursing contact hours. Providers and health professionals can access the training by visiting [www.genetrain.org](http://www.genetrain.org).

The *Newborn Screening in Connecticut* web-based training includes the state's experts on newborn screening disorders from the University of Connecticut Health Center, the Yale School of Medicine and the Connecticut Department of Public Health. The training sessions include information on:

- Latest newborn screening technologies
- Categories of disorders for which babies are screened
- Actions required when a baby has a positive screen
- Communication between the medical home, DPH and clinical specialists
- Appropriate medical care for children with a diagnosed condition

*The Connecticut Department of Public Health is the state's leader in public health policy and advocacy with a mission to protect and promote the health and safety of the people of our state. For more information about the web-based training, please visit the DPH website at <http://www.dph.state.ct.us/BCH/EHDI.htm> or call (860) 509-8081.*

### What's New?

- All NICU babies now receive an automatic brainstem response screen before hospital discharge.
- *The Listen & Learn* Program was developed to provide caregiver education and follow-up speech, language and audiological services for infants with hearing loss who are not eligible for Birth to Three services.
- A draft appropriations bill was released that proposes expanding Birth to Three eligibility for babies with hearing loss to include those with unilateral and/or bilateral mild hearing loss.
- On May 2, 2007, the Department of Public Health's Early Hearing Detection & Intervention Program and the University of Connecticut's Department of Communication Sciences sponsored a conference for audiologists titled, "*Sound Foundations for Infants and Children with Hearing Loss.*"
- The State Medicaid Program added digital hearing aids to their fee schedule.
- Pediatric Grand Round presentations will be conducted at area hospitals to educate providers on newborn screening, testing, tracking & treatment, and the benefits of early intervention.

The Early Hearing Detection and Intervention Program 2006 Annual Report was prepared by the Connecticut Department of Public Health.

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Screening  
before  
**1**  
month

Diagnosis  
before  
**3**  
months

Early  
Intervention  
before  
**6**  
months



Keeping Connecticut Healthy