

Hearing Loss in Medically Fragile Infants and Young Children

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Addision, TX



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Healthy People 2010: Objective 28.11

Increase the proportion of newborns who:

Are screened for hearing loss by age 1 month

Have audiologic evaluation by age 3 months

Are enrolled in the appropriate intervention services by age 6 months.

“The 1-3-6 EHDI Plan”

National EHDI Goals

- Developed in collaboration with state EHDI programs, federal and national agencies, CDC developed EHDI program objectives and performance indicators
- Goals
 - Program Objectives
 - Performance Indicators

National EHDI Goals

- **Goal 1:**

- All newborns will be screened for hearing loss before 1 month of age, preferably before hospital discharge.

- **Goal 2:**

- All infants who screen positive will have a diagnostic audiologic evaluation before 3 months of age.

(Fit with amplification when appropriate within 4 weeks of identification)

- **Goal 3:**

- All infants identified with hearing loss will receive appropriate early intervention services before 6 months of age (medical, audiologic, and early intervention)

The 1-3-6 EHDI Plan

National EHDI Goals, cont.

■ Goal 4:

- All infants and children with late onset, progressive or acquired hearing loss will be identified at the earliest possible time

■ Goal 5:

- All infants with hearing loss will have a medical home as defined by the American Academy of Pediatrics

National EHDI Goals, cont.

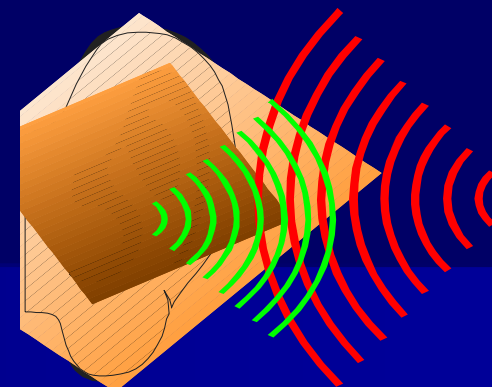
- **Goal 6:**

- Every state will have a complete EHDI tracking and surveillance system that will minimize loss to follow-up

- **Goal 7:**

- Every state will have a comprehensive system that monitors and evaluates the progress towards the EHDI Goals and Objectives

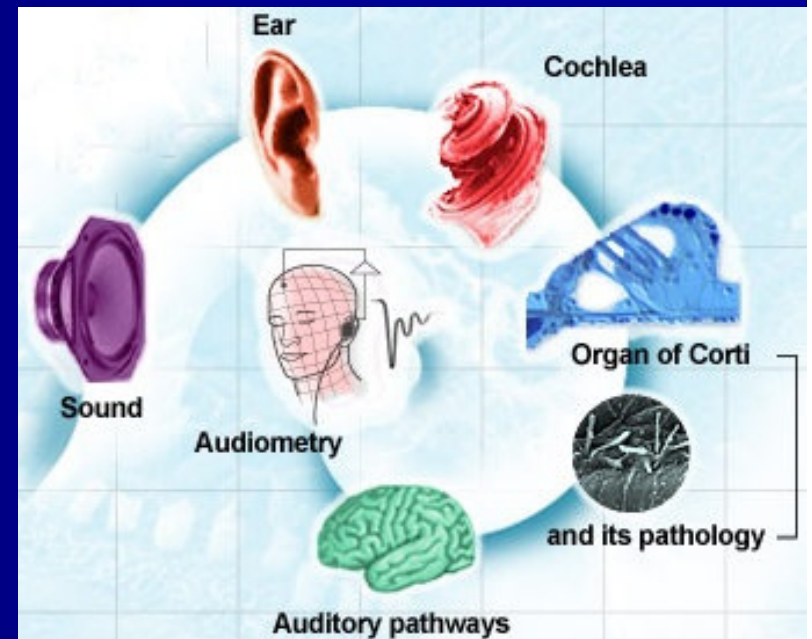
Prevalence of PCHL



- 1 / 1000 children born deaf (severe to profound bilateral SNHL)
- 2-4 / 1000 children with permanent childhood hearing loss 30 dB HL or greater
- 95% of children with substantial bilateral hearing loss are born to hearing parents
- Up to 60% of congenital hearing loss is genetic

Well Baby vs. Special Care Nursery (SCN) at Northside Hospital

- About 10% of all newborns will be special care babies (16,000-18,000 annual births)
- About 1 /100 from SCN will have PCHL



Definition of Targeted Hearing Loss

Expanded from congenital bilateral and unilateral sensory or permanent conductive HL to include

neural hearing loss (auditory neuropathy/dyssynchrony) in infants admitted to the NICU > 5 days.

Hearing Screening Protocols

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

Communication

- Information at all stages of the EHDI process is to be communicated to the family in a culturally sensitive and understandable format.
- Hearing screen information, audiology diagnostic and habilitation information should be transmitted to the medical home and the state EHDI coordinator.

JCIH 2007 Abbreviations

- JCIH
- EHDI
- ABR
- CMV
- ECMO
- AAP
- MCHB
- HRSA
- NIDCD
- CDC
- UNHS
- OAE
- IFSP
- OME
- FM
- DSHPSHWA
- GPRA
- OMB

Communication ?

■ Auditory Brainstem Response

- ABR
- BAER
- BSER
- BSERA
- EAP
- BEAP
- BERA
- AABR
- ABAER
- SABR

■ Otoacoustic Emissions

- OAE
- EOAE
- SFOAE
- TEOAE
- DPOAE
- COAE
- TOAE

COMMUNICATION?

Acronyms (cont.)

■ Related to Behavioral Assessment

- BOA
- VRA
- VROCA
- TROCA
- COR
- CPA
- DA
- OPP
- SRT
- SDT
- SAT
- MTS
- MRL
- NBN
- WT
- SF
- WRS
- AC
- BC

COMMUNICATION?

Acronyms

■ Professional Programs

- EHDI
- UNHS
- NHS
- HRSA
- MCHB
- NIDCD
- NIH
- CDC
- NCBDDD
- AAO-HNS
- ASHA
- AAA
- JCIH
- AAP
- CED
- PINES
- SLP
- ASDC
- AGBell
- AVI

■ Intervention

- HA
- ALD
- DSL
- AGC
- ALD
- DSP
- BTE
- ITE
- ASL
- TC

NICU Acronyms



- AGA
- SGA
- LGA
- IUGR
- IV
- RDS
- PDA
- ABDs
- GA
- PO
- CPAP
- NC
- NBN
- WBN
- NICU
- NEC
- IVH
- ROP
- NG tube
- APGAR
- PROM
- LBW
- VLBW
- ELBW

Screening: NICU

- 10-15% of the newborn population
 - Level I: basic care, well-infant nurseries
 - Level II: specialty care by a neonatologist for infants at moderate risk of serious complications
 - Level III: a unit that provides both specialty and subspecialty care including the provision of life support (mechanical ventilation)

Real World NICU



Major indicators in NICU

- Gestational Age
 - Term = 40 weeks
- Birthweight
- APGAR



APGAR Scoring

	Sign	0 points	1 point	2 points
A	Activity (muscle tone)	Absent	Arms & Legs flexed	Active movement
P	Pulse	Absent	Below 100 bpm	Above 100 bpm
G	Grimace (reflex irritability)	No response	Grimace	Sneeze, cough, pulls away
A	Appearance (skin color)	Blue-gray, pale all over	Normal except for extremities	Normal over entire body
R	Respiration	Absent	Slow, irregular	Good, crying

Normal 7-10; Some resuscitative measures anticipated 4-7;
 ≤ 3 – immediate resuscitation

Recommended Age Terminology

- **Gestational age (GA)**
 - Time from the first day of the last menstrual period to the date of birth, expressed as complete weeks
- **Chronological age**
 - Time elapsed after birth
- **Postmenstrual age**
 - Time in number of weeks after the day of menstruation
- **Corrected age**
 - Calculated by subtracting the number of weeks born before 40 weeks gestation from the chronological age. (for children up to 3 years)

Birth Weight Terminology

- AGA Average for gestational age
- LGA Large for gestational age
- SGA Small for gestational age
- IUGR Intrauterine growth restricted



Birth Weight

- Extremely low birth weight (ELBW)
 - <1000 grams
 - 2.2 pounds
- Very low birth weight (VLBW)
 - <1500 grams
 - 3.3 pounds
- Low birth weight
 - <2500 grams
 - 5.5 pounds

Prevalence of Prematurity

- ~12.5% of births in the US are preterm

Martin, J.A., et al. Births: Final Data for 2004. National Vital Statistics Reports, volume 55, number 1, September 29, 2006.

- Since 1981, the number of preterm babies born has increased by 36%

March of dimes – accessed Feb. 2009
<http://www.marchofdimes.com/peristats/>

Preemies

- at increased risk for:
 - newborn health complications
 - Chronic disabilities
 - Developmental delay
 - Cerebral palsy
 - lung and gastrointestinal problems
 - vision and hearing loss
 - death



Prematurity

- Extremely preterm
 - Very premature
 - Premature
 - Full Term
- $\leq 27 + 6$ weeks GA
 - 28 to 31+6 weeks GA
 - 32 to 36+6 weeks GA
 - 37 to 42 weeks GA



Distribution of prematurity

- 71.2 % 34 - 36 weeks
- 13 % 32 - 33 weeks
- 10 % 28 - 31 weeks
- 6 % < 28 weeks



Survival Rate

- About 80 % of GA>26 weeks survive to one year
- about 90 % at 27 weeks
- about 25 % develop serious lasting disabilities
- up to half may have milder problems, such as learning and behavioral problems

Common Problems in the NICU

- Related to breathing:

- Respiratory Distress syndrome (difficulties breathing)
- Apnea (breathing stops)
- Bronchopulmonary dysplasia (lungs not formed appropriately – chronic lung problems)

Respiratory Assistance

- ECMO
- Oscillator
- Mechanical ventilation
- CPAP (Continuous positive airway pressure)
- High Flow Nasal Cannula
- Nasal Cannula

Common Problems in the NICU

- Intraventricular Hemorrhage (IVH)
 - (bleeding in the brain)
- Patent Ductus Arteriosus (PDA) (heart)
- Necrotizing enterocolitis (NEC) (intestines)
- Retinopathy of prematurity (ROP) (Vision)
- Anemia (blood)
- Hyperbilirubinemia (blood)
- Infections

Northside Hospital - Atlanta

- 16,000-18,000 babies annually
- 125 bed level III NICU (~1900 annually)
- Staffing
 - 4 full time audiology technicians
 - 1 full time audiologist
 - 3 part time audiologists
 - 6 PRNs (weekends and holidays)

Northside Hospital Hearing Screening Protocol

- Well Baby Nursery
 - TEOAE
 - Wait 24 hours for vaginal delivery
 - Wait 48 hours for C-sections
 - Rescreen refers prior to discharge if time permits
 - NICU
 - TEOAE for all babies
 - AABR when in NICU >5 days

NICU Addition to JCIH

- Diagnostic Evaluations prior to hospital discharge for babies:
 - ≤ 27 weeks gestational age
 - ≤ 1000 grams birth weight
 - bilirubin ≥ 20 or exchange transfusion
 - syndrome with associated hearing loss
 - Audiologist/neonatologist discretion

Implications for Audiology

- All NICU babies on monitors
 - Oxygen saturation
 - Cardiac function
 - Respiratory rates
 - Temperature
- Many need additional assistance
 - Respiratory
 - Nutritional

Challenges to Screenings in the NICU

- **Electrical Noise (AABR)**
 - Monitors, leads to babies
- **Baby Noise (OAE and AABR)**
 - Stridor, grunting, congestion
 - Breathing, sucking
 - Myogenic artifact
- **Environmental Noise (OAE)**
 - Monitors, people, phones

Screening Challenges cont.

- Finding each baby's nurse
 - Permission to screen needed for every baby
- Co-bedded newborns
 - Too much help from siblings!
- Space
 - For instrumentation
 - For electrodes

Co-Bedding



Deciding to co-bed multiples is one of the few decisions families get
To make for their infants in the NICU

Case Studies

- Comparison of 2 babies

Newborn Nursery

Full Term

Special Care Nursery

25 wk GA



Comparison: WBN vs. SCN

Zachary

- Born at 39 weeks GA
- Birth weight 4860 gms (10 lbs. 11 oz.)
- APGARs 9 and 9
- C-section
- Mild jaundice
- Discharged at day 4 of life

Zoe

- Born triplet C at 25 wk GA
- Birth weight 726 grams (1 lb 7 oz.)
- APGARs 5 and 8
- C-section
- Multiple complications and risk factors for hearing loss
- Discharged at 1 year, 3 months

Comparison WBN vs. SCN

Zachary

- IP Screening on day 3
- IP Rescreen on day 4
- OP Screening on day 11

- Diagnostic Evaluation on day 22 (week 3)

Zoe

- Day 3 – intubated, bilious pneumonia, phototherapy
- Day 4 – intubated, photo
- Day 11 – intubated, phototherapy, small PDA, minimal grade 1 IVH
- Day 22 – intubated, paralytics, diuretics, antibiotics, cardiac meds
- Day 51 – weaned to CPAP

Comparison: WBN vs. SCN

Zack

- ENT week 5

Zoe

- No IP screen – Dx eval
- Concerns raised by OT/PT
- Still on NC, acuity 5
- Dx Eval on week 22
Chronological age (5 months), week 7
Corrected Age
- ENT as IP 23 weeks
- Parents to ENT 24 weeks

Comparison: WBN vs. SCN

Zack

- GA PINES week 4
- ENT visualized fluid week 5
- Tubes week 7
- Repeat ABR – severe to profound SNHL
- Hearing aids week 12
- CI workup 8 months
- CI bilateral 10 months

Zoe

- GA PINES week 37 (still an IP)
- Repeat ABR – sedated 41 weeks
- Hearing Aids – week 45
- Discharge from hospital 63 weeks 1 day

GA PINES = Georgia Parent Infant Network for Educational Services

Implications for 1-3-6 EHDI Goals

Well Babies

- Should be able to meet outlined goals

SCN

- Many preterm and/or medically fragile babies are 'not ready' for audiologic services until after their chronological age has passed the EHDI goals