Hearing Loss in Medically Fragile Infants and Young Children

EHDI Conference 2009
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.
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

http://www.iurc.montp.inserm.fr/cric/audition/english/start2.htm
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.

JCIH 2007
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
- DSHPHWA
- 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
  - NCBDDDD
  - AAO-HNS

- Intervention
  - ASHA
  - AAA
  - JCIH
  - AAP
  - CED
  - PINES
  - SLP
  - ASDC
  - AGBell
  - AVI
  - HA
  - ALD
  - DSL
  - AGC
  - ALD
  - DSP
  - BTE
  - ITE
  - ASL
  - TC
<table>
<thead>
<tr>
<th>NICU Acronyms</th>
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<tbody>
<tr>
<td>AGA</td>
<td>GA</td>
</tr>
<tr>
<td>SGA</td>
<td>PO</td>
</tr>
<tr>
<td>LGA</td>
<td>CPAP</td>
</tr>
<tr>
<td>IUGR</td>
<td>NC</td>
</tr>
<tr>
<td>IV</td>
<td>NBN</td>
</tr>
<tr>
<td>RDS</td>
<td>WBN</td>
</tr>
<tr>
<td>PDA</td>
<td>NICU</td>
</tr>
<tr>
<td>ABDs</td>
<td>NEC</td>
</tr>
<tr>
<td></td>
<td>IVH</td>
</tr>
<tr>
<td></td>
<td>ROP</td>
</tr>
<tr>
<td></td>
<td>NG tube</td>
</tr>
<tr>
<td></td>
<td>APGAR</td>
</tr>
<tr>
<td></td>
<td>PROM</td>
</tr>
<tr>
<td></td>
<td>LBW</td>
</tr>
<tr>
<td></td>
<td>VLBW</td>
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<tr>
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<td>ELBW</td>
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</tbody>
</table>
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

<table>
<thead>
<tr>
<th>Sign</th>
<th>0 points</th>
<th>1 point</th>
<th>2 points</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A</strong></td>
<td>Activity</td>
<td>Absent</td>
<td>Arms &amp; Legs flexed</td>
</tr>
<tr>
<td></td>
<td>(muscle tone)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>P</strong></td>
<td>Pulse</td>
<td>Absent</td>
<td>Below 100 bpm</td>
</tr>
<tr>
<td><strong>G</strong></td>
<td>Grimace</td>
<td>No response</td>
<td>Grimace</td>
</tr>
<tr>
<td></td>
<td>(reflex</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>irritability)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>A</strong></td>
<td>Appearance</td>
<td>Blue-gray, pale all over</td>
<td>Normal except for extremities</td>
</tr>
<tr>
<td></td>
<td>(skin color)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>R</strong></td>
<td>Respiration</td>
<td>Absent</td>
<td>Slow, irregular</td>
</tr>
</tbody>
</table>

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
<table>
<thead>
<tr>
<th>Birth Weight</th>
<th>Extremely low birth weight (ELBW)</th>
<th>Very low birth weight (VLBW)</th>
<th>Low birth weight</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;1000 grams</td>
<td>&lt;1500 grams</td>
<td>&lt;2500 grams</td>
</tr>
<tr>
<td></td>
<td>– 2.2 pounds</td>
<td>– 3.3 pounds</td>
<td>– 5.5 pounds</td>
</tr>
</tbody>
</table>
Prevalence of Prematurity

- ~12.5% of births in the US are preterm
  

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

  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: ≤27 +6 weeks GA
- Very premature: 28 to 31+6 weeks GA
- Premature: 32 to 36+6 weeks GA
- Full Term: 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:
  - \( \leq 27 \) weeks gestational age
  - \( \leq 1000 \) grams birth weight
  - biliruben \( \geq 20 \) 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
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
  - Special Care Nursery
- Full Term
  - 25 wk GA
## Comparison: WBN vs. SCN

<table>
<thead>
<tr>
<th>Zachary</th>
<th>Zoe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Born at 39 weeks GA</td>
<td>Born triplet C at 25 wk GA</td>
</tr>
<tr>
<td>Birth weight 4860 gms</td>
<td>Birth weight 726 grams</td>
</tr>
<tr>
<td>(10 lbs. 11 oz.)</td>
<td>(1 lb 7 oz.)</td>
</tr>
<tr>
<td>APGARs 9 and 9</td>
<td>APGARs 5 and 8</td>
</tr>
<tr>
<td>C-section</td>
<td>C-section</td>
</tr>
<tr>
<td>Mild jaundice</td>
<td>Multiple complications and</td>
</tr>
<tr>
<td></td>
<td>risk factors for hearing loss</td>
</tr>
<tr>
<td>Discharged at day 4 of</td>
<td>Discharged at 1 year, 3</td>
</tr>
<tr>
<td>life</td>
<td>months</td>
</tr>
</tbody>
</table>
## Comparison WBN vs. SCN

<table>
<thead>
<tr>
<th>Zachary</th>
<th>Zoe</th>
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</thead>
<tbody>
<tr>
<td>IP Screening on day 3</td>
<td>Day 3 – intubated, bilious pneumonia, phototherapy</td>
</tr>
<tr>
<td>IP Rescreen on day 4</td>
<td>Day 4 – intubated, photo</td>
</tr>
<tr>
<td>OP Screening on day 11</td>
<td>Day 11 – intubated, phototherapy, small PDA, minimal grade 1 IVH</td>
</tr>
<tr>
<td>Diagnostic Evaluation on day 22 (week 3)</td>
<td>Day 22 – intubated, paralytics, diuretics, antibiotics, cardiac meds</td>
</tr>
</tbody>
</table>

**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

<table>
<thead>
<tr>
<th>Zack</th>
<th>Zoe</th>
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</thead>
<tbody>
<tr>
<td>ENT week 5</td>
<td>No IP screen – Dx eval</td>
</tr>
<tr>
<td></td>
<td>Concerns raised by OT/PT</td>
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<tr>
<td></td>
<td>Still on NC, acuity 5</td>
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<tr>
<td></td>
<td>Dx Eval on week 22</td>
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<tr>
<td></td>
<td>Chronological age (5 months), week 7</td>
</tr>
<tr>
<td></td>
<td>Corrected Age</td>
</tr>
<tr>
<td></td>
<td>ENT as IP 23 weeks</td>
</tr>
<tr>
<td></td>
<td>Parents to ENT 24 weeks</td>
</tr>
<tr>
<td>Comparison: WBN vs. SCN</td>
<td></td>
</tr>
<tr>
<td>-------------------------</td>
<td></td>
</tr>
<tr>
<td><strong>Zack</strong></td>
<td></td>
</tr>
<tr>
<td>- GA PINES week 4</td>
<td></td>
</tr>
<tr>
<td>- ENT visualized fluid week 5</td>
<td></td>
</tr>
<tr>
<td>- Tubes week 7</td>
<td></td>
</tr>
<tr>
<td>- Repeat ABR – severe to profound SNHL</td>
<td></td>
</tr>
<tr>
<td>- Hearing aids week 12</td>
<td></td>
</tr>
<tr>
<td>- CI workup 8 months</td>
<td></td>
</tr>
<tr>
<td>- CI bilateral 10 months</td>
<td></td>
</tr>
<tr>
<td><strong>Zoe</strong></td>
<td></td>
</tr>
<tr>
<td>- GA PINES week 37 (still an IP)</td>
<td></td>
</tr>
<tr>
<td>- Repeat ABR – sedated 41 weeks</td>
<td></td>
</tr>
<tr>
<td>- Hearing Aids – week 45</td>
<td></td>
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<tr>
<td>- Discharge from hospital 63 weeks 1 day</td>
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GA PINES = Georgia Parent Infant Network for Educational Services
## Implications for 1-3-6 EHDI Goals

<table>
<thead>
<tr>
<th>Well Babies</th>
<th>SCN</th>
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<tbody>
<tr>
<td>Should be able to meet outlined goals</td>
<td>Many preterm and/or medically fragile babies are ‘not ready’ for audiologic services until after their chronological age has passed the EHDI goals</td>
</tr>
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