

A Potpourri of EHDI issues

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www.jcih.org



Representatives: American Academy of Audiology, American Academy of Otolaryngology–Head and Neck Surgery, American Academy of Pediatrics, American Speech-language-hearing Association, Council on Education of the Deaf, Directors of Speech and Hearing Programs in State Health and Welfare Agencies.

The Agenda for this Session

- A little about the JCIH and its statements
- 2007 Position Statement –
 - Highlights and Clarifications
- Issues/Challenges (selected)
 - Screening
 - Diagnosis
 - Intervention



In the Beginning

- JCIH established in 1969
- Composed of representatives of professional organizations with interest in children with hearing loss
- Original charge: Make recommendations concerning early identification of children with HL, and newborn screening.

First Position Statement

- The first one page Position Statement was published in *Pediatrics* in 1971.
- It concluded that data at the time were inconsistent and misleading and therefore universal screening of newborn infants could not be recommended.
- [JCIH1971.pdf](#)

Second Position Statement: 1973

- This was the 1st recommendation for screening of infants with risk factors and
- 2nd a recommendation for continued surveillance, i.e. if the hearing of these infants is normal, they should continue to have regular assessments.
- [JCIH1973.pdf](#)

Third Position Statement: 1982

Fourth Position Statement: 1990

Fiddled around with risk factors
and

Divided them into 2 groups:

Birth to 28 days

29 days to 2 years

Fifth JCIH Position Statement: 1994

Pivotal Position Statement: things were happening:

- RI Hearing Assessment Program(RIHAP) demonstration project had been implemented
- NIH 1993 Consensus Conference endorsed universal detection of hearing loss.
- Colorado data of Yoshinago-Itano supported improved outcomes of infants identified < 6m of age.
- Endorsed universal detection of all infants before 3 months of age and intervention by 6 months.
- It did not endorse universal screening.

Sixth JCIH Position Statement: 2000

- 1st Endorsement of direct physiologic screening of all infants regardless of risk status.
- Expanded the concept of screening to EHDI – Early Hearing Detection and Intervention.
- The need for quality EHDI programs resulted from the mounting evidence of the importance of EI services in the 1st year of life to maximize language and communicative development.

Sixth JCIH Position Statement: 2000

- If universal screening, why risk indicators ?
- Risk indicators were to be used in regions where universal screening had not been implemented.
- Data were emerging indicating that a significant % of permanent HL is not identified by NB screening.
- Therefore risk factors for delayed hearing loss (JCIH 2000) gained importance.

Sixth JCIH Position Statement: 2000

- Continued with the recommendation:
Audiologic monitoring every 6 months
until 3 years for all infants
with 1 or more risk factors
for late onset HL.
- [jcih2000.pdf](#)

What happened between 2000-2007?

- > 95% of newborns in the United States and territories have their hearing screened.
- Current EHDI Programs
 - Screen \leq 1 month
 - Diagnosis \leq 3 months
 - Intervention \leq 6 months

JCIH 2007 Statement

- [JCIH 2007 Statement Pediatrics 898\[1\].pdf](#)
- [ExecSummFINAL\[1\].pdf](#)

Joint Committee on Infant Hearing

- Not a Commission, a Committee
- Authority?
- Politics?
- Writing skills?
- [Clarification Year 2007 statement\[1\].pdf](#)

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 Screen Protocols

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

Clarification

- Rationale for different protocols
- Where did the 5 days in the NICU come from?
- One of the considerations was the cost of making the change for all NICU babies which represent 10% of all newborns.
- All others can be screened with either OAE or ABR.
- JW comment: *automated ABR?*

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, evaluation of both ears is recommended, even if only 1 ear failed the initial screen.
- Rescreen with the same test you did before
JW comment: exceptions?

Rescreen of NICU Refers

- For infants who do not pass automated ABR in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation including ABR.

Diagnostic Audiology Evaluation

- Audiologists with skills and expertise in evaluating infants with hearing loss should provide audiology diagnostic and habilitation services.
- At least one ABR is recommended as part of a complete diagnostic audiology evaluation for children under 3 years of age for confirmation of permanent HL, in conjunction with other measures for validation of HL.

Diagnostic Audiology Evaluation

- Infants with a risk factor for HL should have at least one diagnostic audiology assessment by 30 m of age. Infants with risk factors associated with late onset or progressive loss (e.g. CMV or ECMO) are followed more frequently.
- For families who elect amplification, infants diagnosed with permanent hearing loss should be fitted with amplification within one month of diagnosis

Medical Evaluation

- All families should be offered a Genetics consultation.
- Every infant with a confirmed HL should have at least one exam by an ophthalmologist experienced in evaluating infants. Other specialty consultations may be indicated.
- The list of risk factors has been reorganized to a single list to focus on both early and late onset and/or progressive HL.

Risk Factors for Hearing Loss: 2007

- Caregiver concern* regarding hearing, speech, language
- Family history* of permanent childhood HL
- NICU care of >5 days, e.g. assisted ventilation ≥ 10 d, ototoxic medications ≥ 7 days, exchange transfusion, and ECMO*
- Intra-uterine TORCH infections, particularly CMV*
- Craniofacial anomalies, especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies

* greater concern for delayed-onset hearing loss

Risk Factors for Hearing Loss , cont

- Physical findings associated with a syndrome known to include permanent HL
- Syndromes associated with progressive HL* such as NF, osteopetrosis, Usher's syndrome
- Neurodegenerative disorders*, such as Hunter syndrome
- Postnatal infections associated with SNHL especially bacterial meningitis*
- Head trauma requiring hospitalization
- Chemotherapy*

* greater concern for delayed-onset hearing loss

Clarification

- Previous recommendation for every 6 months
 - too great a burden on system
 - infants with “unknown risk factors” develop delayed-onset HL

Thus responsibility for surveillance shifted to PCP with referral to audiologist > Risk factor list

Low risk – another assessment by 24-30 months

New concern – assess immediately

* **risk for delayed onset** - earlier & more frequent re-assessment

Clarification

- Recommendations regarding ototoxic medications

To be consistent with the intent of simplifying the referral process to NICU > 5 days, the recommendation has been reworded:

All infants with or without risk factors requiring NICU care of >5 days, including any of the following: ECMO,* assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion.

Surveillance and screening in the Medical Home

- All infants should have **regular** surveillance consistent with the pediatric periodicity schedule. **Auditory skills, milestones, parent concerns, and MEE** should be monitored.
- All infants should have an objective standardized screen of global development with a validated tool at 9, 18, and 30 months of age.
- Children who do not pass a medical home global screen or if there is concern regarding hearing or language should be referred for speech-language evaluation and audiology assessment.

Early Intervention

- All families of infants with all degrees of HL should be offered Early Intervention.
- The recognized point of entry for EI for infants with a confirmed HL should be linked to EHDI, and be provided by professionals with expertise in HL, including educators of the deaf and speech language professionals.
- Both home-based and center-based options should be offered as appropriate interventions.

Modes of Communication for children with permanent HL

- Families should be made aware of all communication options and available hearing technologies.
- Family choice guides the decision making process.

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.

Information Infrastructure

- States must develop adequate data management and tracking systems as part of an integrated child health information system in order to monitor the quality of EHDI services and provide recommendations for improving systems of care.
- A linkage between health and education is recommended to determine outcomes of children with hearing loss at school age.
- This is needed for planning and establishing public health and education policy.

Challenges Remain in 2007

- Follow-up rates of screen fails remain ~ 50%
- There is **failure to communicate information** to families in a culturally sensitive and understandable format
- Lack of **integrated state data tracking systems**
- A **shortage of facilities and personnel** with the experience and expertise needed to provide follow-up
- A **significant number of children** who need further assessment do not receive appropriate evaluations.

The EDHI Challenge ...

...continues in 2008 with continued efforts to achieve success needed on multiple levels:

- hospital,
- community,
- state and
- federal
- We all need to take the “EHDI Challenge”

Potpourri of EHDI Issues



Potpourri of EHDI Issues

- **Screening**

Who are we missing?

Why are we missing them?

(What hearing loss are we trying to detect?)

- **Diagnostic**

Who is available to do the audiologic dx?

Are infants getting a genetic evaluation?

- **Intervention**

Proposal for Guidelines

Screening Issues

- Who are we missing?
- Lost to follow-up/Lost to documentation (LTF/LTD)
 - NIHQ
 - CDC Contractual Agreements
 - ASHA Technical Paper
- Later onset hearing loss
 - Risk Factors
 - Surveillance

Are we missing mild hearing loss?

- British studies (Davis et al., 1997; Lutman et al., 1997)
- Identification of Neonatal Hearing Impairment
Norton, Gorga, Widen, Folsom, Sininger, Cone-Wesson, Vohr et al., *Ear & Hearing*, 2000 [link](#) here for this study
- A Multi-Center Evaluation of How Many Infants with Permanent Hearing Loss Pass a Two-Stage OAE/A-ABR Newborn Hearing Screening Protocol”
Johnson, White, Widen, Gravel, James, Kennalley, Maxon, Spivak, Sullivan-Mahoney, Vohr, Weirather, & Holstrum , *Pediatrics* 2005 [link](#)
- Massachusetts Loss to Follow-up on Use of Audiologic Evaluation Services (AES): (2002-2003 Births: 158,243)
Liu et al. *Pediatrics* 2008 [link](#)

Screening Issues

- Why are we missing mild hearing loss?
- Targeted hearing loss – 35 dBnHL click?
- < 30-40 dB, unusual configurations
- Calibration – scenario from KUMC
- Standards for calibration, or the lack of them
- Variability among screening devices, levels, pass-fail criteria

Screening Issues

- JCIH Stakeholders meeting
- Hosted by ASHA at its National Office in Rockville, MD
- September 17, 2008

- JCIH members & representatives of companies who manufacture or sell hearing screen devices

JCIH Stakeholder's Meeting

- Tone: friendly, healthy discussion of issues
- Speakers:
 - Judy Gravel
 - John Eichwald
 - Panel of JCIH members from the trenches
 - John Durrant
 - Bob Burkard

SPECIAL ARTICLE

Universal Screening for Hearing Loss in Newborns: US Preventive Services Task Force Recommendation Statement

US Preventive Services Task Force

ABSTRACT

DESCRIPTION. This is the 2008 update of the 2001 US Preventive Services Task Force recommendation on universal newborn hearing screening.

METHODS. The US Preventive Services Task Force weighed the benefits and harms of universal newborn hearing screening, incorporating new evidence addressing gaps identified in the 2001 US Preventive Services Task Force recommendation statement. Published literature on this topic was identified (by using Medline and Cochrane databases) and systematically reviewed.

RECOMMENDATION. Screen for hearing loss in all newborn infants (B recommendation).
Pediatrics 2008;122:143–148

Gravel:

Statement of the Challenges:

Where We Have Been,
Where We Are,
Where We'd Like to Go

Gravel et al.

A multisite study to examine the efficiency of the
OAE/AABR newborn hearing screening protocol:
Recommendations for Policy, Practice, and Research
American Journal of Audiology 14: S217-228, Dec 2005

Eichwald:

- Are we missing a substantial number of cases of mild or moderate hearing loss in the newborn period?
- Discrepancy between prevalence of hearing loss in infants versus school age children?

Prevalence Summary

RATE / 1000	NEWBORNS	SCHOOL AGE	INCREASE
MILD 21 – 40 dB	0.34 [†]	9.1	x 26.8
MODERATE 41 – 70 dB	0.53 [†]	1.8	x 3.4

[†] Assumed rates

Panel of JCIH members:

Stories from the trenches

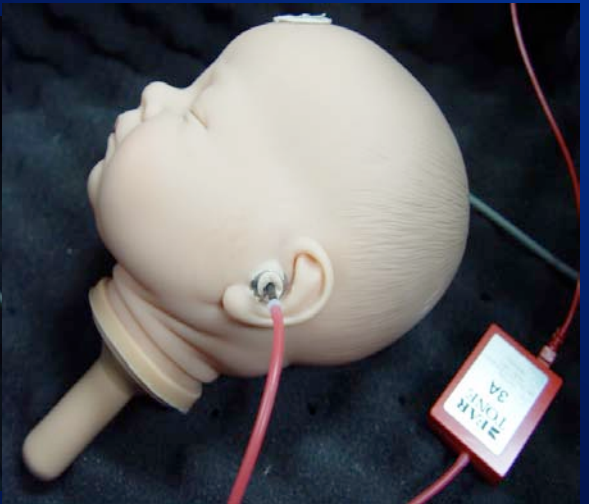
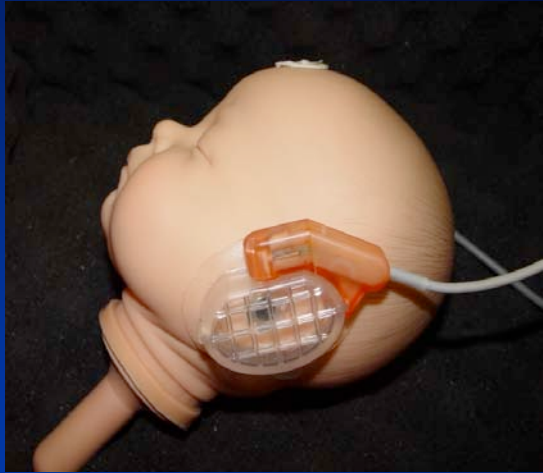
Colorado – pass-refer rates and prevalence rates
changed with change in equipment

UCLA – test with the “machine” that passes everyone

JD Durrant, D Sabo, R Delgado

Call for Standard of Calibration for Newborn Screening

- [Durrant Sabo Delgado.pdf](#)



Bob Burkard:

- Acoustic Calibration of Transients
Tutorial on terminology, limitations of SLMs,
how do we accurately measure the sound
pressure level of a transient?
- A Description of the ANSI Standards Process
[link](#)

Discussion JCIH Stakeholder's Meeting

- **Manufacturers:**

- What hearing loss do you want to screen for?
- You all are asking for things most of our customers don't use

- **JCIH:**

- Transparency and disclosure [link](#)
- Way to do at least weekly calibrations
- Data/information provided to user beyond pass/refer, i.e. wave forms, noise (for administrative coordinator of use of system)-
- Develop quality standards of performance – i.e. no false passes in noise, will run 1000 times and not give a false response [Brit link](#)

Diagnostic Issues - Audiologic

- There's not enough of us
- Efforts to train audiologists for pediatric practice
(Watching is not enough)
 - NCHAM
 - DOE
 - LEND

Diagnostic Issues - Medical

- Why is genetic testing recommended?
- Who's getting genetic testing?
- Why not?

DNA testing Issues

Mutation Detection Rate vs. Accuracy

[Link](#)

Accuracy:

All U.S. clinical tests regulated by CLIA
([Clinical Laboratory Improvement Amendments](#)),
Centers for Medicare & Medicaid Services (CMS),
[Department of Health & Human Services](#)

DNA testing Issues

Interpretation of results

- Ambiguous - “Uncertain variant” “unknown significance”
- Negative - family mutation vs. unknown (false reassurance)
- Positive – management issues
- Premutation

- Autonomy vs. coercion from other family members
- Nonpaternity
- Family relationship changes

PEDIATRI^X™

SCREENING

90 EMERSON LANE
BRIDGEVILLE, PA 15017



SoundGene™))))

The SoundGene™ Screening Panel detects the most common causes of hearing loss

The SoundGene™ Screening Panel

Connexin 26 (Cx26) GJB2 mutations¹:

35delG	167delT
235delC	M34T

Connexin 30 (Cx30) GJB6 large deletion

309kb large deletion

Mitochondrial mutations:

7445A>C (A7445C)	961T>C (T961C)
7445A>G (A7445G)	961T>G (T961G)
7444G>A (G7444A)	961delT+C(n)ins

Pendred SLC26A4 mutations:

L236P	1001+1G>A
E384G	T416P

Cytomegalovirus (CMV) - DNA

¹Under sublicense with Athena Diagnostics, Inc.: United States Patent Numbers 5,998,147 and 6,485,908 and patents pending

Intervention Issues

- JCIH 2nd focus – beginning now
- Early Intervention Position Paper
- CYI [link](#)

Conclusions

- We've got our work cut out for us!

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