

# Childhood Hearing Impairment

N. Wendell Todd MD MPH  
Adrienne M. Laury MD  
Emory University

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*slide courtesy of Arthur L Kellermann, MD, MPH*

## Rational for Auditory Screening of Newborns

- Communicative development is most rapid in early infancy, preparing the person for a lifetime of human communication
- 80% of language ability is established by age 18 months

## Occurrence of Hearing Loss

- 3 per 1000 infants are born with a permanent sensorineural hearing loss (~1 Georgia baby/day)
  - 1/1000 from the well baby nursery
  - 10/1000 from the NICU
- Rate increases to approximately 6/1000 by school age

*slide from Krista Biernath & John Eichwald, CDC EHQI*

## Rationale for Monitoring Language Development

Half of 1<sup>st</sup> graders manifest bilateral sensorineural hearing impairment after the newborn period

## Congenital Hearing Impairment in the United States: Etiologies

- Conductive -- 0.1/1000 births -- aural atresia
- Sensory -- 1-2/1000 -- most are genetic
- Neural -- <1/1000 -- ~1/2 genetic  
~1/2 hyperbilirubinemia

## Etiologies of Congenital Hearing Impairment (United States)

- Most are genetically determined:
- 70-80% autosomal recessive
  - 15-20% dominant
  - ~2% X-linked
  - <1% mitochondrial
- ~15% are syndromic:  
>400 distinct syndromes

## Potentially Treatable Causes of Sensory Hearing Impairment

- Infections: e.g., syphilis, toxoplasmosis, CMV
- Hypothyroidism
- Perilymphatic fistula
- Leukemic infiltration, usually granulocytic sarcoma of acute myelogenous leukemia
- Cochleotoxics: e.g., aspirin, quinine, aminoglycosides
- Autoimmune
- Vasculitis: e.g., Cogan's syndrome

## Potentially Treatable Causes of Neural Hearing Impairment

- Cerebello-pontine angle cysts and tumors
- Inflammation, e.g., syphilis, multiple sclerosis

## Children with Sensorineural Hearing Loss have more Otitis Media

- Brookhouser PE et al. (1993) Laryngoscope
- Vartiainen E (2000) J Otolaryngol
- Westerberg BD et al. (2005) J Laryngol

## Assess Hearing in Tympanostomy-Tube Patients

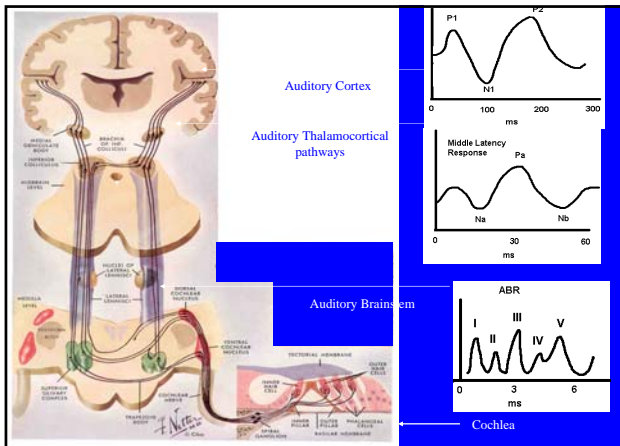
- makes sense
- Rosenfeld RM et al. Clinical practice guideline: Otitis media with effusion. Otol Head Neck Surg (2004)
- guideline American Academy of Otolaryngology Head Neck Surgery

## Screening Methods

- Automated Oto-Acoustic Emissions (A-OAE)
- Automated Auditory Brainstem Response (A-ABR)

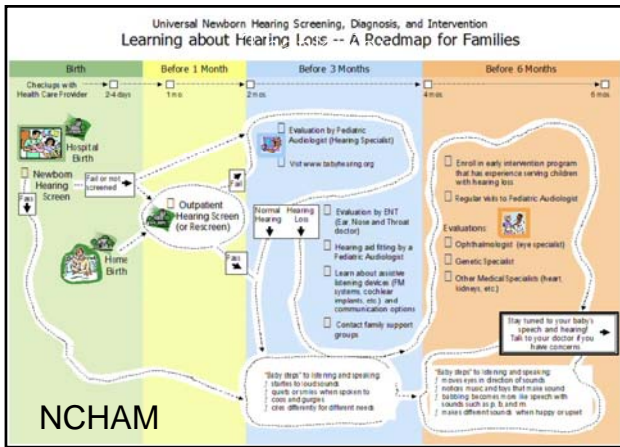
Each measures auditory physiology at an unconscious level.

Each has advantages & disadvantages.



## Newborn Hearing Screening in Georgia

- Legislation endorsed heavily by audiologists, and the Georgia Chapter of the American Academy of Pediatrics.
- Legislative mandate, effective July 2001, to educate about hearing and language development.
- Legislative intent: screen newborns.
- Established Advisory Committee, that "shall include at least one audiologist, one pediatrician, one hospital administrator...." *No otolaryngologist!*



## What is the Georgia otolaryngologist to do with these babies?

- Work with the child's "medical home".
- Little consensus about what to do.
- FDA requires medical clearance before hearing aid(s) are dispensed for a child.
- Georgia Medicaid requires that an otolaryngologist render "medical clearance".

- Verify
- Amplify
- Typify
- Investigate
- Consult
- Initiate
- No harm
- Assure
- Test
- Educate

VATICINATE, used here an acronym mnemonic, is a real word: to chant, fortell, sing, prophesy about things to come

## VERIFY

Use a battery of tests:

- Behavioral Audiometry
- Oto-acoustic emissions
- Auditory evoked potentials: clicks, tonebursts
- Tympanometry [with stapedius reflexes]

Does the compilation make sense? With what family thinks? With child's phonations/speech?

## Recommended infant diagnostic test battery (Joint Committee in Infant Hearing [JCIH], 2007)

- High frequency tympanometry
- Otoacoustic emissions
- Auditory Brainstem Response (ABR) click with ability to identify cochlear microphonic
- Tone burst ABR for a low and a high frequency
- Bone conduction ABR
- Behavioral observation audiometry

From

Karen Munoz' presentation, CDC EHDI teleconference 11/17/09

## Hearing Test Battery Differences (n= 284)

Facilities performing:

- JCIH recommended diagnostic battery: 5%
- Testing in addition to the recommended battery: 5%
- Diagnostic battery with missing components: 47%
  - Risk of misdiagnosis
- Partial battery with significant missing diagnostic detail: 39%
  - Insufficient detail to initiate intervention
- Diagnostic testing not available: 4%

Karen Munoz' presentation, CDC EHDI teleconference 11/17/09

## Challenges in completing testing before 3 months of age

Challenge	Frequency
Parent compliance in scheduling appointment	65%
Presence of middle ear fluid	56%
Infant has other medical/health issues	42%
Noisy results, repeat testing needed	32%
Parents live far from testing facility	31%
Parents have transportation problems	30%
Appointments are booked out several weeks	29%
Lack of timely referrals from screening	27%
"Other" reasons	13%
Testing only available under sedation	2%

Karen Munoz' presentation, CDC EHDI teleconference 11/17/09

## Important ABR Parameters

Delivery mode:

head phone; insert tube; bone oscillator

Stimulus type: click; tone pip

Click delivery:

refraction; condensation; alternating

Stimulus intensity: dBnHL vs dB SPL

Stimulus rate: need the numbers

Number of samples

## AMPLIFY

- If the data indicate bilateral impairment, amplify.
- Do not wait for the definitive battery of tests to get started.
- Use binaural amplification as an assessment tool of the infant's hearing.
- If cannot use earmolds (e.g., atresia, or canals too small), then bone oscillator aid. Bone-anchored-cochlear-stimulator.

## TYPIFY THE HEARING IMPAIRMENT

This guides the search for etiology and therapy:

Conductive -- fix surgically, hearing aids

Sensory -- hearing aids, cochlear implant

Neural -- total communication, cochlear implant

Mixed

## INVESTIGATE ETIOLOGY I

### History

What brings you here? What is the concern? Ask about 1st & 2nd degree relatives?

### Examine the patient:

General physical examination.

Height, weight, head circumference.

Birth marks.

Facial asymmetry.

Microscopic exam of tympanic membranes.

## INVESTIGATE ETIOLOGY II

### Laboratory studies:

Check that maternal and newborn screening tests were done, and are normal (syphilis, thyroid...)

Additional tests not rewarding unless clue from history, physical, or consultant.

EKG for long QT of Jervell & Lange-Nielsen

But, if history of "spell", or if a relative had unexplained sudden death, cardiology consult.

## CONSULT

### Ophthalmologist

to optimize patient's vision

to search for an explanation for hearing loss

### Radiologist

high resolution MRI (or CT) of ears (& brain)

timing: about 9 months of age

### Geneticist

## CONSULT GENETICIST IF:

### ANY OF THESE:

Parents considering another child.

A 1st or 2nd degree relative has same type and pattern of hearing impairment.

Patient has a syndrome, e.g. Waardenburg.

Patient has a genetic pattern of sensory hearing loss, e.g. "cookie-bite".

RE-CONSULT BEFORE PLANS OWN FAMILY.



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## INITIATE DISCUSSION OF PSYCHIATRIC ASPECTS

- Anticipatory guidance.
- Shock, denial, sadness & anger, equilibrium, re-organization.
- Guilt, over-compensation, rejection.
- Behavioral issues, sleep disturbances.
- Mis-match of chronologic and communicative age.

## NO HARM -- anticipate, prevent

- Cerumen management.
- Addressible risk factors for otitis media.
- Immunize.
- Cochleotoxic drugs.
- Noise toxicity.
- Degrading effect of noise on communication.
- Safety, including localization of source of sound.
- Sports: aquatics, gymnastics, SCUBA, bungee.

## ASSURE

Assure that the patient can/will communicate:

- Listening-speaking mode, or
- With lip-reading, or
- Total Communication, or
- Sign Language.

## TEST REPEATEDLY

- To identify changes in hearing, to continue to search for an accurate etiologic diagnosis, to verify the type of hearing impairment, and to assure amplification is appropriate.
- Each 3 months in 1st year, q 6 months until about 8 years, then annually.
- Genetics and ophthalmology, as indicated.
- Diagnostic audiometry in 1st degree relatives.

## EDUCATE

Opportunities for the child and family:

Georgia Parent Infant Network Educational Services  
Babies Can't Wait (early intervention program)  
Auditory-Verbal Center of Atlanta  
Atlanta Speech School (auditory-oral program)  
Atlanta Area School for the Deaf  
Children's Healthcare of Atlanta  
Internet sites: e.g., NCHAM

Parents' scrapbook of all tests and assessments, as patients are invariably seen in many formats.

With each patient encounter for hearing impairment, think VATICINATE

- |                      |                   |
|----------------------|-------------------|
| • <b>Verify</b>      | • <b>Initiate</b> |
| • <b>Amplify</b>     | • <b>No harm</b>  |
| • <b>Typify</b>      | • <b>Assure</b>   |
| • <b>Investigate</b> | • <b>Test</b>     |
| • <b>Consult</b>     | • <b>Educate</b>  |

## **GOALS:**

**by ages 1-3-6 months**  
(from Healthy People 2010)

- All newborns will be screened for hearing loss before 1 month of age.
- All infants who screen positive will have a diagnostic audiologic evaluation before 3 months of age.
- All infants identified with a hearing loss will receive appropriate early intervention services before 6 months of age.

## Four Noteworthy Realizations

- 3/1000 in newborns, but 6/1000 in 6-year-olds
- Newborn hearing screening advocates assumed that not passing screening would itself cultivate follow-up
- We need better measures of verbal expressive language development: **2 words at 2 years**
- About 15% of newborns with SNHL have "auditory neuropathy"

## THE CLOCK IS TICKING



# THANK YOU.

Resource:  
search internet for eBook + NCHAM

Rauschecker JP, Shannon RV: Science 295:1025-9, 2002

