

# RISK MONITORING FOR DELAYED-ONSET HEARING LOSS IN YOUNG CHILDREN

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# **Learning Objectives**

- Identify risk indicators which require monitoring for delayed-onset hearing loss
- List risk indicators which require more frequent audiological monitoring
- Explain options for risk monitoring protocols



## Joint Committee on Infant Hearing (JCIH)

**JCIH was established in 1969** 

#### **Comprised of:**

- American Academy of Pediatrics
- American Academy of Ophthalmology and Otolaryngology
- American Speech & Hearing Association



# **JCIH** position statements



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# **JCIH 1973 Position Statement**

#### High risk criteria

- Family history of childhood hearing loss
- Intrauterine fetal infection (Rubella)
- Defects of ear, nose or throat (atresia, cleft lip/palate)
- Low birth weight (<1500 grams)</p>
- High bilirubin levels



# **JCIH 1982 Position Statement**

#### -High risk criteria

»Bacterial meningitis, severe asphyxia (i.e. low APGAR) were added

Screening recommendations

 »Ideally performed by 3 months (no later than 6 months)
 »Preferably under the supervision of an audiologist
 »Observation of behavioral or electrophysiologic response to sound



# **JCIH 1990 Position Statement**

#### High risk criteria additions:

»Ototoxic medications »Prolonged mechanical ventilation »Physical findings of syndromes »Parent/caregiver concerns »Head trauma »Neurodegenerative disorders »Infectious diseases associated with hearing loss Screening recommendation changes: »Auditory Brainstem Response measurement, not behavioral testing



## **JCIH 1994 Position Statement**

Studies showed that only 50% of all hearing loss were being identified using the High Risk Register

- Pappas, 1983
- Elssman, Matkin, Sabo 1987
- Mauk, white, mortensen, Behrens 1991



## **JCIH 2000 Position Statement**

#### Risk monitoring: –Audiological testing every 6 months until age 3 years.

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## Joint Committee on Infant Hearing (JCIH)

YEAR 2007 POSITION STATEMENT: Principles and Guidelines for Early Hearing Detection and Intervention Programs



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#### **JCIH 2007 Position Statement**

Expanded definition of targeted hearing loss to include:

 <u>Neural hearing loss (Auditory</u> <u>Neuropathy/Dysynchrony) in infants admitted to</u> <u>the NICU</u>

Separate protocols for NICU and well baby nurseries:

 NICU babies (>5 days) are to have ABR screening so that neural hearing loss will not be missed



#### **JCIH 2007 Position Statement**

#### Re-admissions

 Infant readmitted in the first month of life and present with conditions, which are associated with potential hearing loss, need a repeat hearing screen prior to discharge.

#### Monitoring of high risk indicators

 – "Infants with risk factors for hearing loss should have <u>at least one diagnostic evaluation by 24-30</u> <u>months of age."</u>



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#### JCIH 2007 Appendix 2: RISK INDICATORS FOR HEARING LOSS

- □ Caregiver concerns (re: hearing, speech, language, or developmental delay)
- □ Family history of permanent childhood hearing loss
- Neonatal Intensive Care (NICU) of more than 5 days or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide, Lasix), and hyperbilirubinemia that requires exchange transfusion.
- □ In-utero infections
- □ Craniofacial anomalies
- □ Known physical findings associated with a syndrome
- □ Syndromes associated with hearing loss, progressive hearing loss or late-onset hearing loss neurodegenerative disorders
- □ Culture-positive postnatal infections associated with hearing loss
- Head trauma, especially basal skull/temporal bone, requiring hospitalization
- □ Chemotherapy



# Risk indicators for delayed-onset hearing loss

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## Incidence of risk factors for hearing loss

## Epstein and Reilly (1989) reported 10-12% of all babies had at least one risk factor



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Most frequently occurring risk factors	Least frequently occurring risk factors (<10%)
Ototoxic Medications (>70%)	Hyperbilirubinemia
Severe Asphyxia (>50%)	Craniofacial anomalies
Mechanical Ventilation less	Family history
than 5 days (>25%)	Congenital infections
Low birth weight (>20%)	Bacterial meningitis
Parental/Physician concerns	Substance abuse (maternal)
(>15%)	Neurodegenerative
ECMO (>10%)	disorders

(Cone-Wesson, et al., 2000; Van Riper & Kileny, 1999; Van Riper & Kileny, 2002; Hall, 2007)

Frequency of hearing loss among high risk indicators

Craniofacial anomalies (>50%) ECMO treatments (>20%) Severe Asphyxia/ Mechanical ventilation (>15%) Congenital infections (>15%) Family History (>15%) Bacterial meningitis (>10%) Other risk indicators (<10%)

(Cone-Wesson, et al., 2000; Van Riper & Kileny, 2002; Hall, 2007; Fligor, 2008)

# **Ototoxic Medications**

 Over 200 known ototoxic medications (prescriptions and OTC)

Used to treat serious infections, cancer, heart disease

Damage may be temporary or permanent

 Aspirin (temporary)
 Cisplatin (permanent)



# Aminoglycosides

- Introduced in 1940s
- Used to treat serious infections due to multi-drug resistant Gram negative bacteria
- May remain in hair cells for months after application (Aran et al, 1999)
- "...<u>weekly or biweekly monitoring is recommended</u> ideally." "...follow-up testing should also be scheduled a <u>few months</u> after drug discontinuation." (AAA Ototoxicity Monitoring, 2009)

## Gentamicin

- Introduced 1963
- Most common aminoglycoside used in NICU
- Low cost
- Effectiveness against most Gramnegative bacteria



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### ASHA 2010- Evidence Based Systematic Review: Drug-Induced Hearing Loss-Gentamicin

- Systematic literature review (20 studies)
- Reported hearing loss from gentamicin induced cochleototoxicity ranging from 0-58%

 Studies varied in dosing, patient populations, diagnostic testing, diagnostic criteria for hearing loss



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### ASHA 2010- Evidence Based Systematic Review: Drug-Induced Hearing Loss-Gentamicin

- Trends noted in the studies:
  - Frequency of administration did not influence the likelihood of hearing loss
  - Dosing amount did not influence the likelihood of hearing loss



# A1555G genetic mutation

 Prezant et al (1993) reported on the genetic mutation A1555G, associated with aminoglycoside deafness

 Estivill et al (1998) reported profound hearing loss without aminoglycoside treatments



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# A1555G genetic mutation

 United Kingdom study (2002) found 1 in 206 newborns expressing the mutation

 Texas study (1999) only 1 in 1,161 newborn with mutation

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### Ototoxicity in preterm infants (Zimmerman E, Lahav A, 2012)

Effects of genetics – Iowa Children's Hospital (Ealy et al 2011) - N=703 (1.8% with mtDNA variant) – No hearing loss Loud noise exposure Animal studies have found potentiating effect between noise and aminoglycosides

# **Neonatal Intensive Care (NICU)**

National Perinatal Research Center (NPIC) (Quality Analytic Services (QAS)

- Approximately 25% of NICU infants are considered "LOW" risk and discharged by 5 days old.
- The remaining approximately 75% of NICU infants, who are hospitalized for greater than 5 days, are considered the "TARGET" population to rule out neural hearing loss.

\*\*NICU stay of greater than 5 days and exposure to loop diuretics were not associated with increased risk of hearing loss (Kraft et al, 2014)



# **Mechanical Ventilation/Hypoxia**

Cone-Wesson et al. (2000) estimates 1/56 children with permanent hearing loss at age 1

Robertson et al. (2002) found greater than 50% of severe neonatal respiratory survivors had sensorineural hearing loss at 4 years old

Beswick et al (2013) study found a correlation between postnatal hearing loss and prolonged ventilation (≥5 days)



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# Expracorporeal Membrane Oxygenation (ECMO)

 Expracorporeal Membrane Oxygenation (ECMO)- is an aggressive treatment that is used for the life support in infants with respiratory or cardiopulmonary failure

# **ECMO (Fligor, 2008)**

- N = 111 neonates
- Congenital Diaphragmatic Hernia raised risk of SNHL 2.6 times
- Aminoglycoside antibiotics cumulative of 14 days or more in the course of ECMO raised the risk of SNHL by 5.56 times
- ECMO 160 hours raised risk of SNHL 7.18 times



# Physical findings/Syndromes associated with hearing loss

Waardenburg syndrome Branchio-Oto-Renal (BOR) syndrome Stickler syndrome CHARGE syndrome NeurofibromatosisType II Downs syndrome Treacher Collins syndrome Usher syndrome Pendred syndrome Alport syndrome Jervell Lange-Nielsen



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# Infections

# **Congenital Infections**

- Cytomegalovirus (CMV)
- Rubella
- Herpes
- Syphilis
- Toxoplasmosis

# Postnatal infections

- Bacterial or viral meningitis
- Varicella
- Herpes viruses



# **Craniofacial anomalies**

- Head trauma
- Recurrent OME
- Cleft palate
- Abnormal pinna
- Abnormal ear canal
- Ear tags and pits
- Malformed eyes
- Choanal atresia
- Craniosynostosis
- Hemifacial microsomia

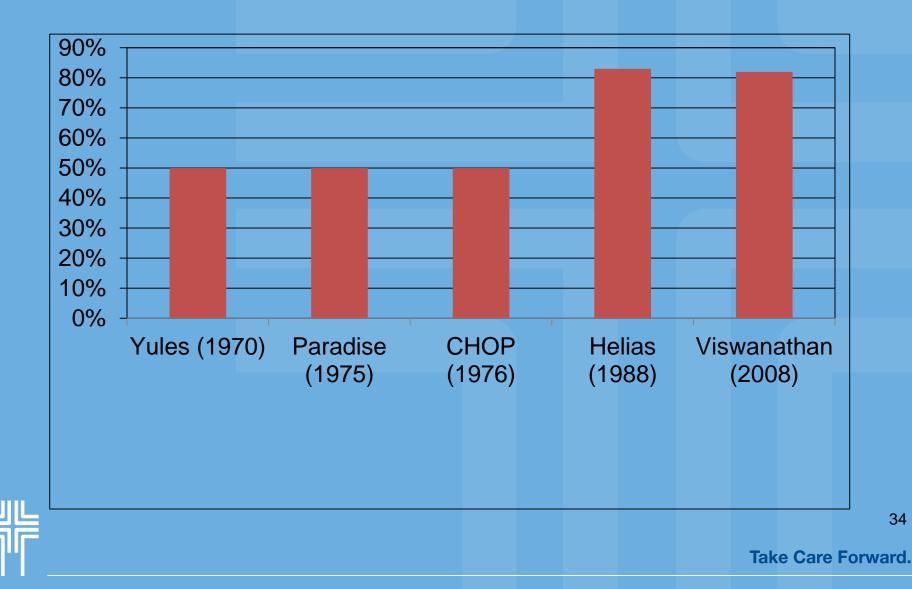




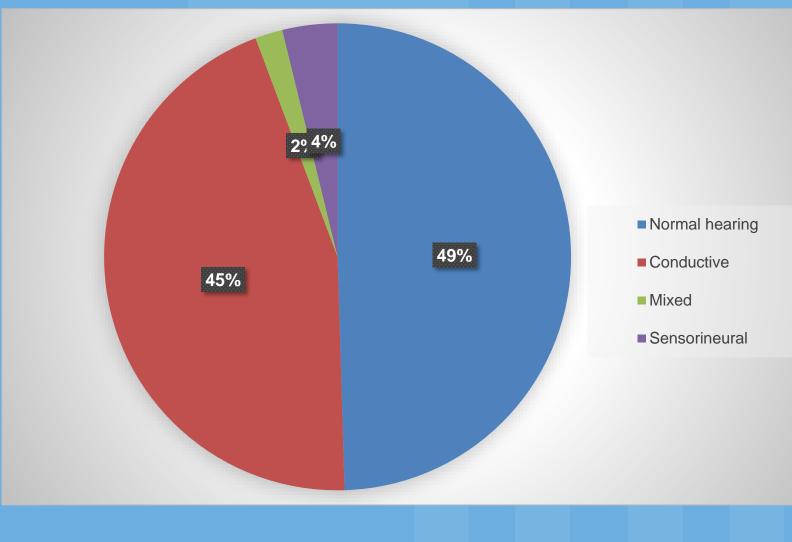


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## % of hearing loss in cleft palate patients



### Idaho Cleft Palate and Craniofacial Deformities team (Oct 2007- Feb 2010)



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# **Family History**

 Positive family history of congenital hearing loss or hearing loss acquired during childhood



 Australia study (Beswick, et al. 2013) showed that the risk factor of family history did predict the occurrence of postnatal hearing loss



### **Head trauma**

### Involving basal skull/temporal fracture that requires hospitalization

#### • May result in:

- Facial nerve paralysis (partial, complete)
- -Hearing loss (conductive, sensorineural, mixed)
- -Vertigo
- -Tympanic membrane perforations



## Neurodegenerative disorders/Sensory motor neuropathies

Hunter syndrome

Charcot Marie Tooth disease

Friedreich ataxia



# Recent publications looking at risk indicators for delayed-onset hearing loss



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### Beswick et al (2012)

Literature Review (40 articles)

 CMV, ECMO, congenital diaphragmatic hernia, persistent pulmonary hypertension associated with postnatal hearing loss



### Beswick et al (2013)

### N = 2107 children

• 2.7% with postnatal hearing loss

### Findings:

 Family history and craniofacial anomalies (monitored throughout childhood)

 Syndromes and prolonged ventilation (favorable results for monitoring)

Low Birth Weight (no monitoring)



### Kraft et al (2014)

- Prospective study
- Evaluate risk indicators for childhood hearing loss
- Estimate cost burden of monitoring imposed by some risk indicators
- Findings: NICU stay and exposure to loop diuretics are not associated with increased risk for delayed onset hearing loss



### Vos et al (2015)

- Literature review
- Findings
  - Family History of HL, consanguinity in (grand)parents, malformation syndromes, fetal alcohol syndrome (HIGH)
  - Hyperbilirubinemia (MODERATE)
  - Low birth weight, low APGAR, NICU stay, ototoxic medications (LOW/VERY LOW)





### **Risk monitoring programs**



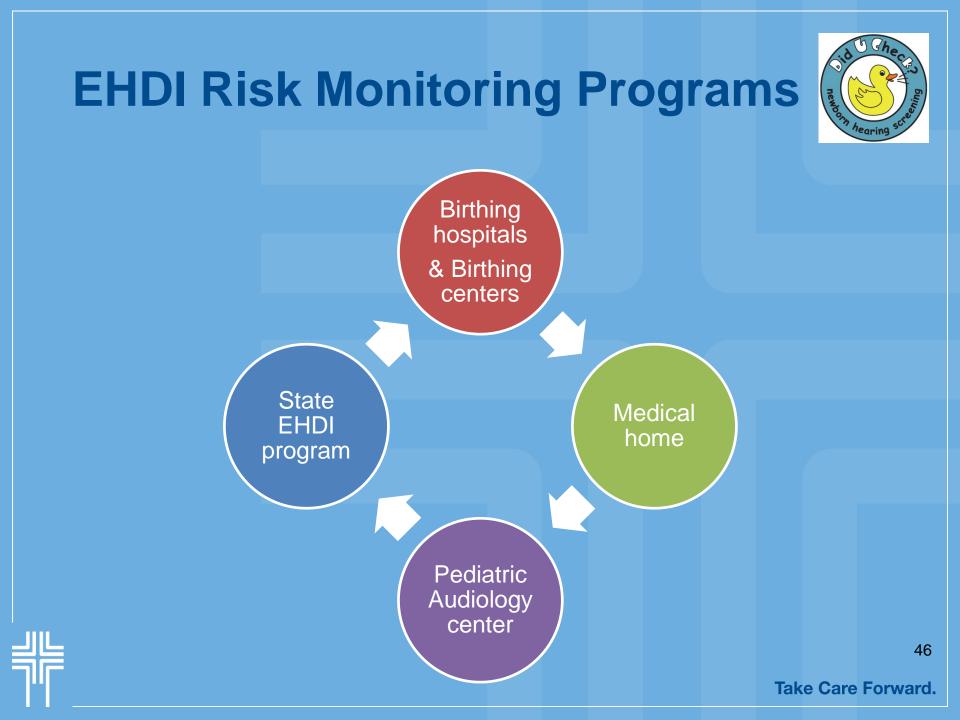
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### Goals of risk monitoring programs



- Identify infants and children at risk for delayed onset or progressive hearing loss
- Timely diagnostic assessments from a pediatric audiologist
- Maintain a monitoring and tracking system in the state EHDI data management system





### **Hospitals/Birthing Center roles:**



- Identify infants who have 1 or more risk indicators
- Provide family with referral to pediatric audiology clinic
- Provide family with information about risk indicators
- Provide medical home information regarding risk indicator referral
- Report infants with risk indicators to state EHDI program



### **Script for hearing screeners**



"Your baby has been identified as having a risk indicator for (\_\_\_\_) for delayedonset hearing loss. The recommendation for babies with risk indicators is an audiological evaluation around 9 months of age. You will receive a reminder letter when your baby is 8 months old along with a list of pediatric audiologists who can test infants and toddlers."



### **Medical home roles:**



- Being familiar with risk factors for delayed onset hearing loss
- Explaining screening results and answer questions for the family
- Encourage risk monitoring follow-up
- Providing family with referral to pediatric audiology clinic



### Pediatric audiology center roles:



- Providing appropriate comprehensive diagnostic testing for children with risk factors
- Knowledge of risk factors that have high prevalence of delayed onset hearing loss and require early and more frequent assessments
- Providing documentation regarding evaluation outcomes to state EHDI program



### **State EHDI program roles:**



- Providing training and support for hospitals, birthing center, physicians, and pediatric audiologists on risk factor
- Providing a method for hospitals, birthing centers and pediatric audiologists to report information regarding infants with risk indicators to the state EHDI program
- Tracking and surveillance of infants with risk factors



### Idaho EHDI program





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### Data collected by referral forms



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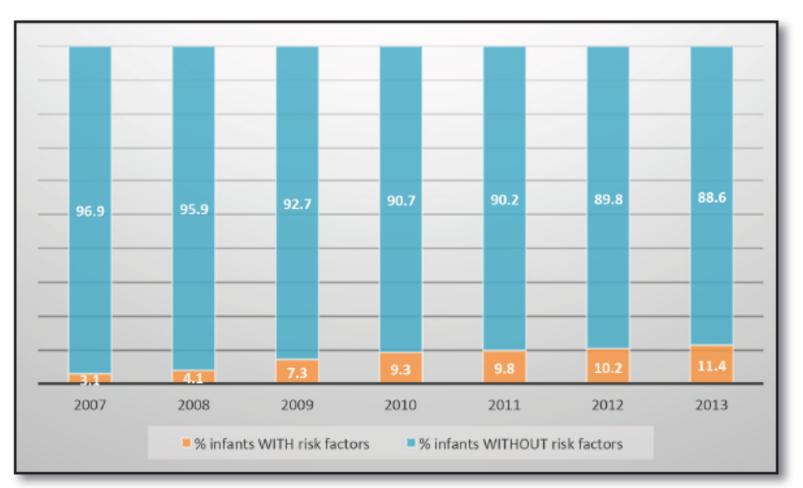
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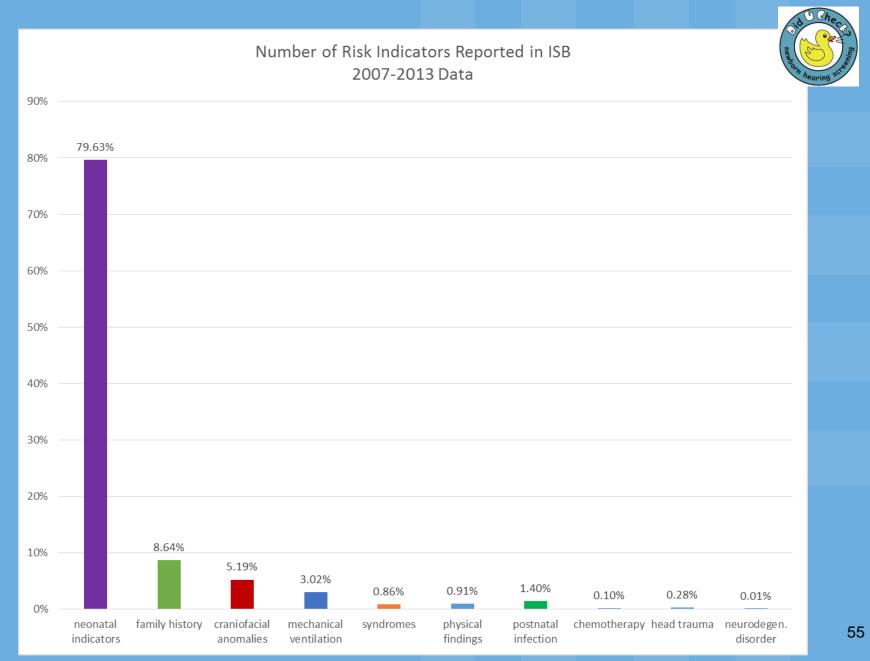
PARENT/GUARDIAN :

### Prevalence of Infants with a Risk Indicator in ISB 2007-2013 Data





54





### Idaho's classification system for risk monitoring



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### **Class A & Class B**

Pediatric audiologist and **NICU** physicians began discussions - May 2011 Developed guidelines - May-October 2011 Two hospitals implemented - October 2011





#### Class A: Risk indicators

 In-utero infections (congenital CMV) \*Culture Positive postnatal infection (Bacterial and viral meningitis) \*Syndromes associated with progressive or delayed onset hearing loss (Neurofibromatosis, Osteopetrosis, Usher Syndrome, Townes-Brock) \*Syndromes associated with hearing loss (Down syndrome and Sticklers) \*Cleft Lip/Palate \*ECMO assisted ventilation \*Head Trauma involving basal skull/temporal fracture that requires hospitalization Chemotherapy treatments \*Neurodegenerative disorders or sensory motor neuropathies

If baby passes the newborn hearing screening & has one or more CLASS A risk indicator = Recommendation for diagnostic ABR evaluation with pediatric audiologists by 3 months of age.

#### Class B: Risk indicators



\*Family history of childhood hearing loss \*In-Utero Infection (Herpes, Rubella, Syphilis, Toxoplasmosis) \*NICU stay of greater than 5 days \*Any amount of ototoxic exposure (aminoglycosides) \*Any amount of mechanical ventilation \*Craniofacial anomalies involving pinna, ear canal, ear pits and temporal bone anomalies

If baby passes the newborn hearing screening & has one or more CLASS B risk indicators = Recommendation for diagnostic pediatric hearing evaluation by 1 year of age.

### Data collected January 2012- December 2013

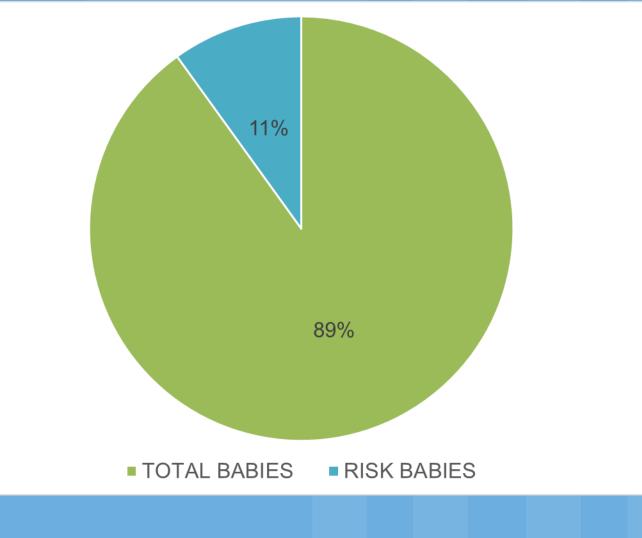


- HiTrack data management system
- Reviewed data November 2015
- N= 10,634 babies
- = 1.6% 175 babies with CLASS A risk indicator
- =11.04% 1175 babies with any risk indicator (CLASS A and/or CLASS B)



# % babies with reported risk indicators (2012-2013)

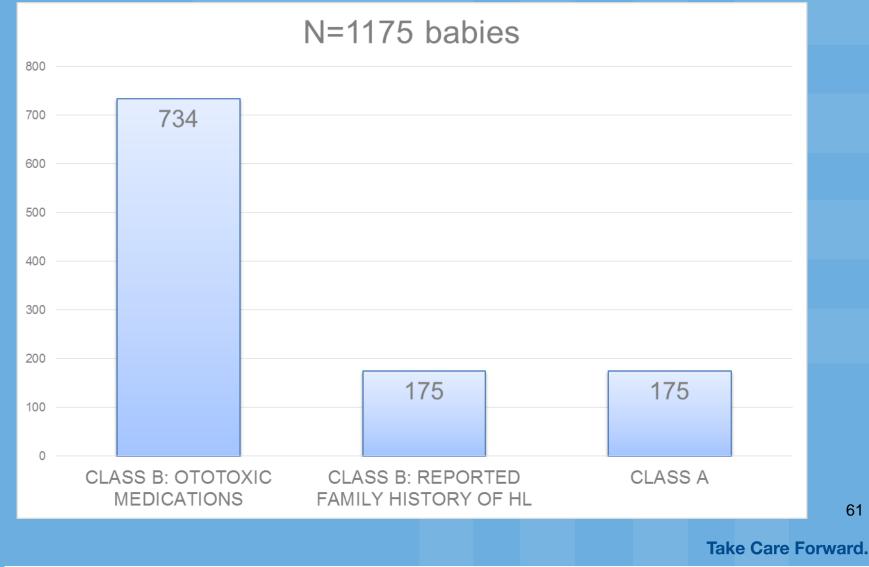


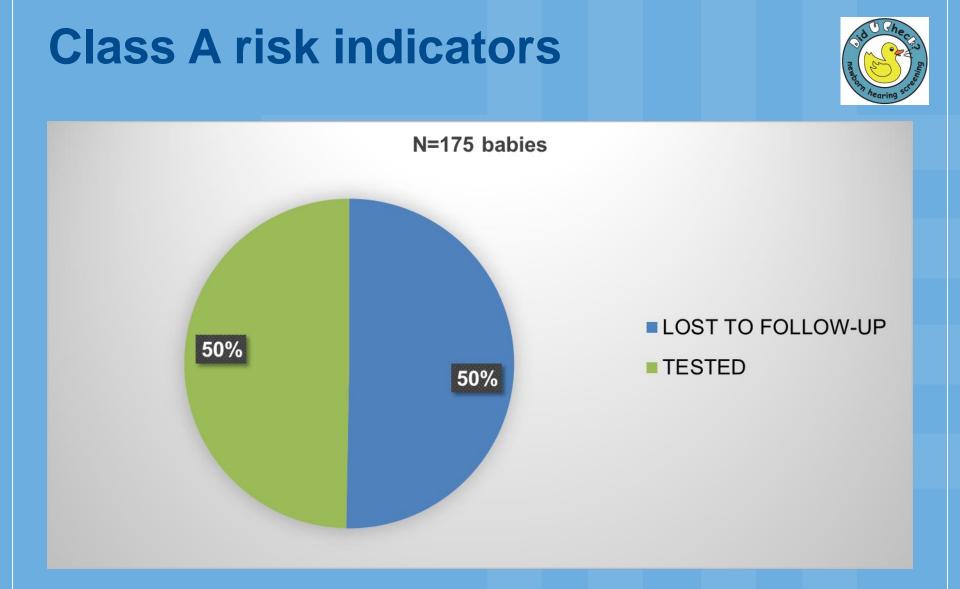


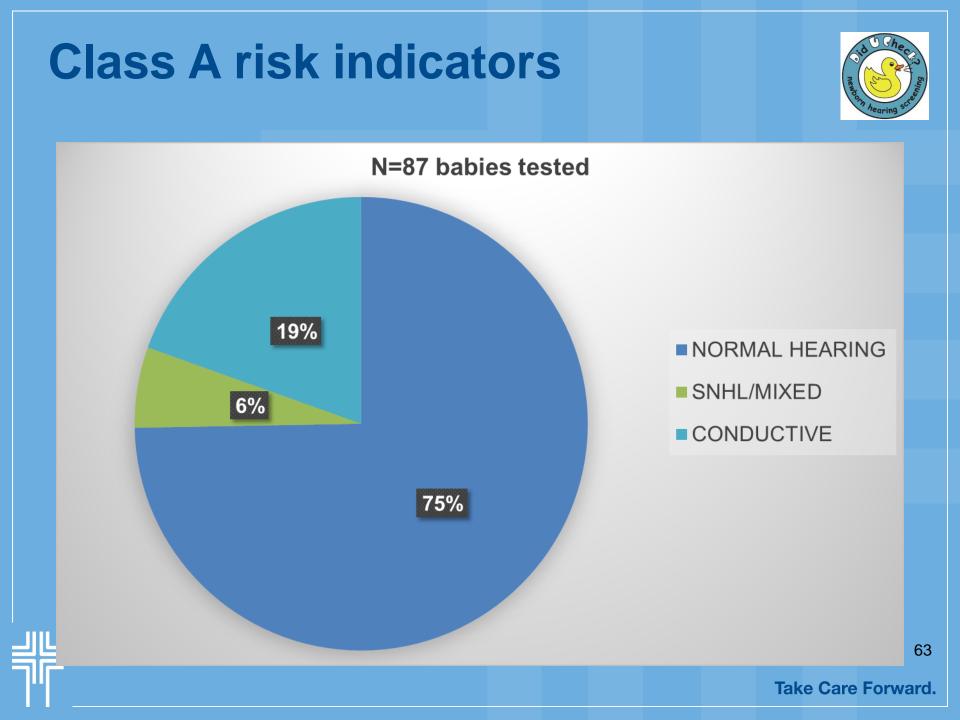
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### **Risk indicator occurrence**









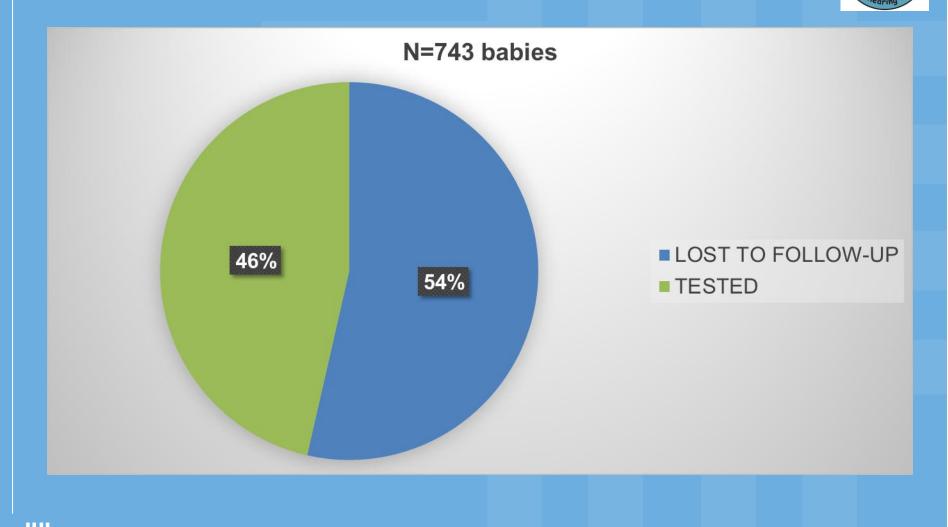
### Children with Class A risk indicators & permanent hearing loss

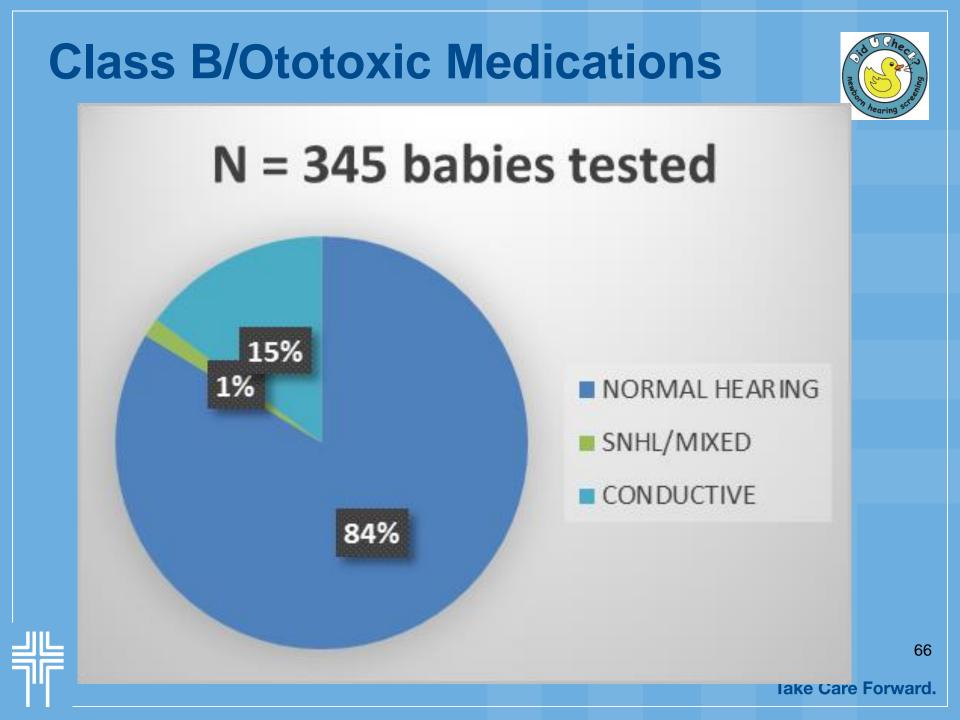


- 5/87 sensorineural/mixed hearing loss
  - 2 children with cleft palate
  - 1 child with Townes Brock syndrome
  - -1 child with Acrofacial Dysostosis
  - -1 child with congenital CMV



### **Class B/Ototoxic Medications**



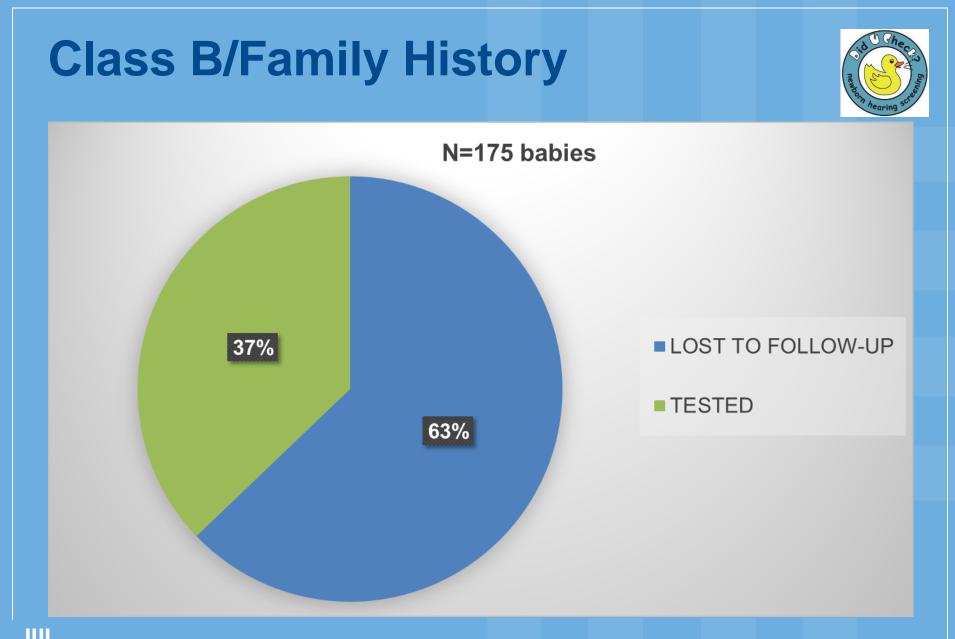


Children with history of Ototoxic medications & permanent hearing loss



- 5/345 children with sensorineural/mixed hearing loss
  - 1 child with ototoxic medications and extended NICU stay
  - 1 child with ototoxic medications, mechanical ventilation, extended NICU stay
  - 3 children ototoxic medication plus CLASS A risk indicator

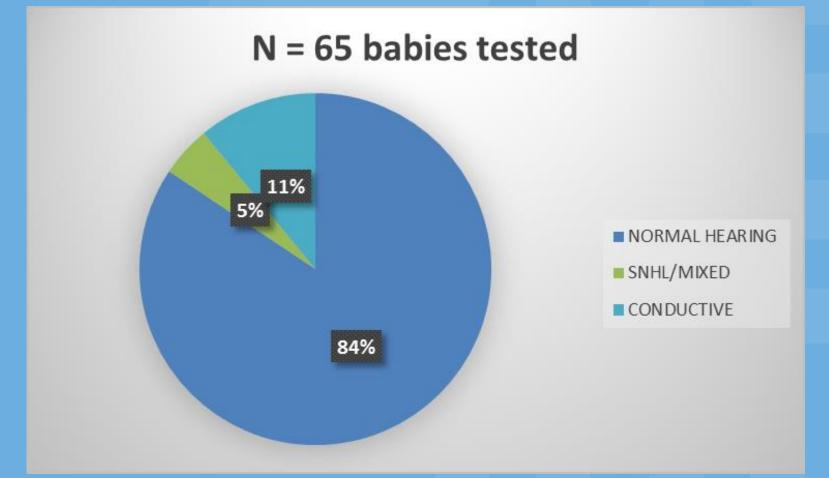




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### **Class B/Family History**







69

# Children with positive family history & permanent hearing loss



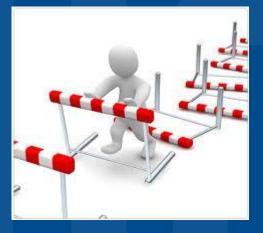
- 3/65 sensorineural/mixed hearing loss
  - 2 children with only risk indicator Family History (siblings)
  - 1 child with multiple risk indicators (syndrome, family history, ototoxic medications, mechanical ventilation, extended NICU stay)



### Barriers to monitoring risk indicators for delayed-onset hearing loss



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### **Barriers**

- Accurate reporting by hospital staff
- Accurate reporting by families (i.e. family history)
- Accurate and timely reporting by audiologists
- Shortage of pediatric audiologists
- High lost-to follow-up rates
- Lack of support by medical homes
- No standard protocol for audiological monitoring of risk indicators
  - What age to start/stop monitoring
  - What tests to use for evaluation



### Why do we monitor children with risk indicators for delayed-onset hearing loss...

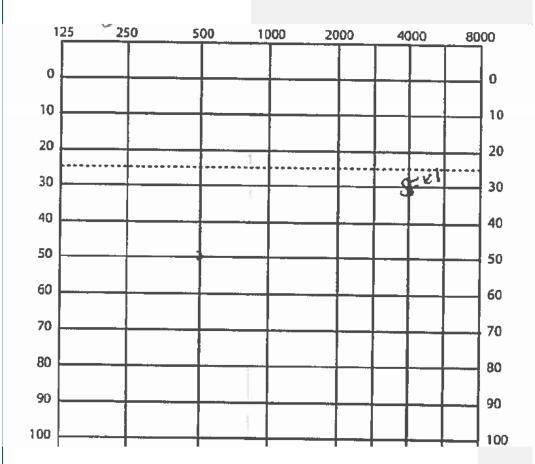


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### Case #1

- Passed AABR hearing screening
- Born at 35 weeks 6/7 days
- NICU stay less than 5 days
- Referred to audiology for risk indicator monitoring (Ototoxic medications)
- No family history of childhood hearing loss
- No history of otitis media

### Audiology Evaluation 9 months old



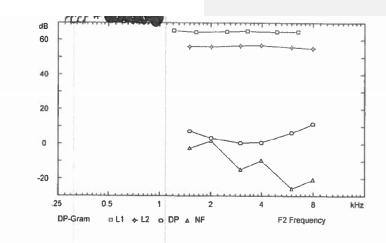
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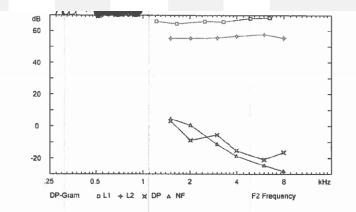
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### **Otoacoustic Emissions**





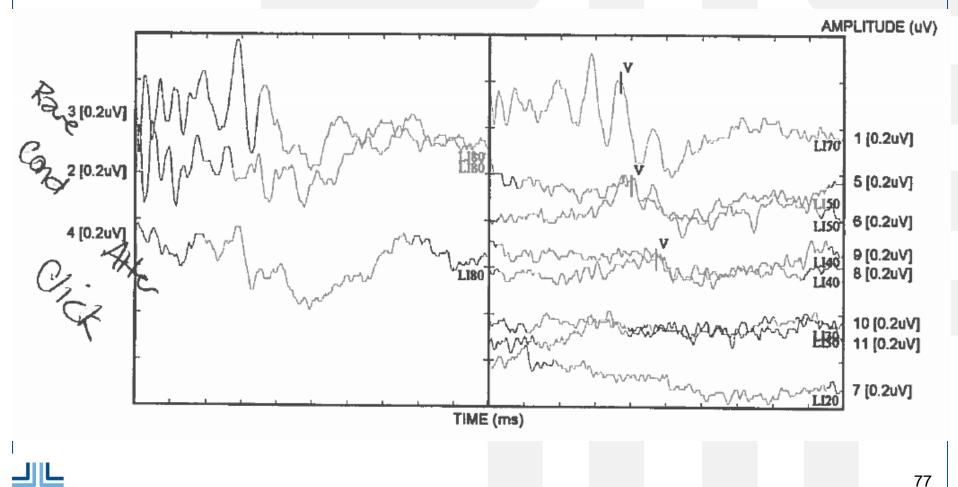
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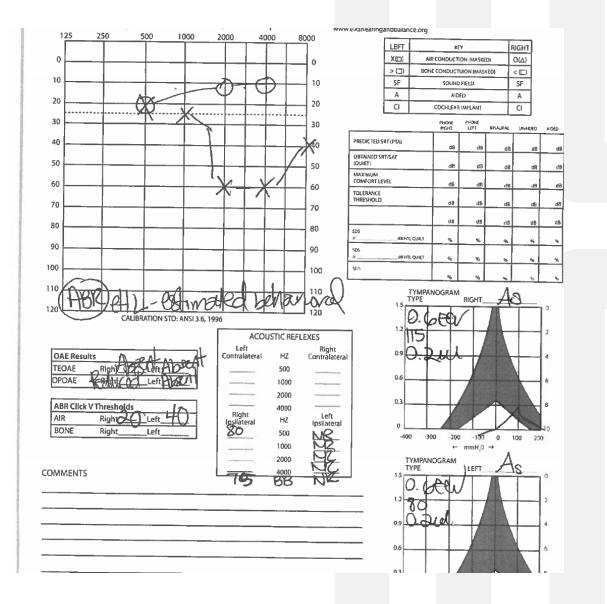
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65.7	56.6	3281	3984	3616	-15.5	-18.8	3.3
66.1	55.7	2484	3000	2730	-5.7	-11.4	5.7
64.5	55.3	1641	2016	1818	-8.9	0.4	-9.3
65.9	55.3	1219	1500	1352	3.0	4.5	-1.5

### ABR evaluation 10 months old

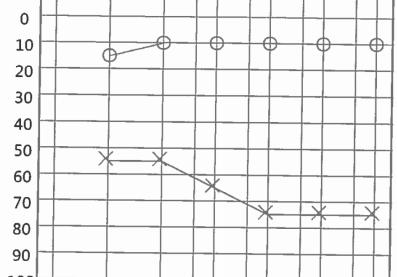


### **ABR eHL**



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79



### 3 years old

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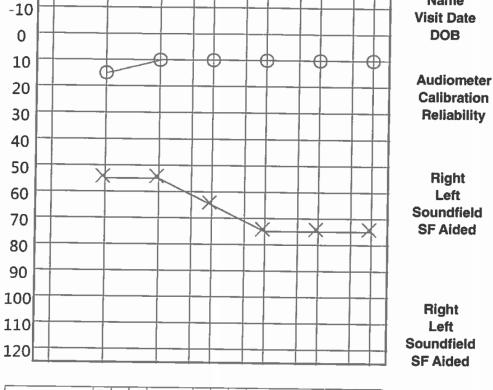
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Soundfield



### **Future research**



80



### **Questions and Answers**



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