

Ototoxicity monitoring as part of risk monitoring in the EHDI system

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Services provided by St. Luke's

JCIH Position Statements



Services provided by St. Luke's

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 2000 Position Statement

- **Recommended ALL infants screened before hospital discharge**
- **Risk monitoring:**
 - **Audiological testing every 6 months until age 3 years.**



JCIH 2007 Position Statement

- Expanded definition of targeted hearing loss to include:
 - Neural hearing loss (Auditory Neuropathy/ Dysynchrony) in infants admitted to the NICU
- 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 at least one diagnostic evaluation by 24-30 months of age.”



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**



Extended NICU stay

National Perinatal Research Center (NPIC) (Quality Analytic Services (QAS) ~ made the recommendation regarding NICU stay for JCIH 2007

- 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)**



ECMO treatments

- **Extracorporeal Membrane Oxygenation (ECMO)- is an aggressive treatment that is used for the life support in infants with respiratory or cardiopulmonary failure**
- **Study found receiving aminoglycoside antibiotics cumulative of 14 days or more in the course of ECMO raised the risk of SNHL by 5.56 times**



Ototoxicity defined...

medications that can damage the ear, resulting in hearing loss, ringing in the ear, or balance disorders.



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)



Why concern about ototoxicity with infants?

Most frequently occurring risk factors

- **Ototoxic Medications (>70%)**
- **Severe Asphyxia (>50%)**
- **Mechanical Ventilation less than 5 days (>25%)**
- **Low birth weight (>20%)**
- **Parental/Physician concerns (>15%)**
- **ECMO (>10%)**

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



Least frequently occurring risk factors ($<10\%$)

- **Hyperbilirubinemia**
- **Craniofacial anomalies**
- **Family history**
- **Congenital infections**
- **Bacterial meningitis**
- **Substance abuse (maternal)**
- **Neurodegenerative disorders**

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



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)
- “...weekly or biweekly monitoring is recommended ideally.” “...follow-up testing should also be scheduled a few months after drug discontinuation.” (AAA Ototoxicity Monitoring, 2009)



Gentamicin

- **Introduced 1963**
- **Most common aminoglycoside used in NICU**
- **Low cost**
- **Effectiveness against most Gram-negative bacteria**



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**



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
- United Kingdom study (2002) found 1 in 206 newborns expressing the mutation
- Texas study (1999) only 1 in 1,161 newborn with mutation



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



Recently published aminoglycoside research

- **Designer aminoglycosides prevent cochlear hair cell loss and hearing loss (Huth et al, 2015)**
- **Aminoglycoside ototoxicity and hair cell ablation in the adult gerbil: A simple model to study hair cell loss and regeneration (Abbas et al, 2015)**



Risk Monitoring Program



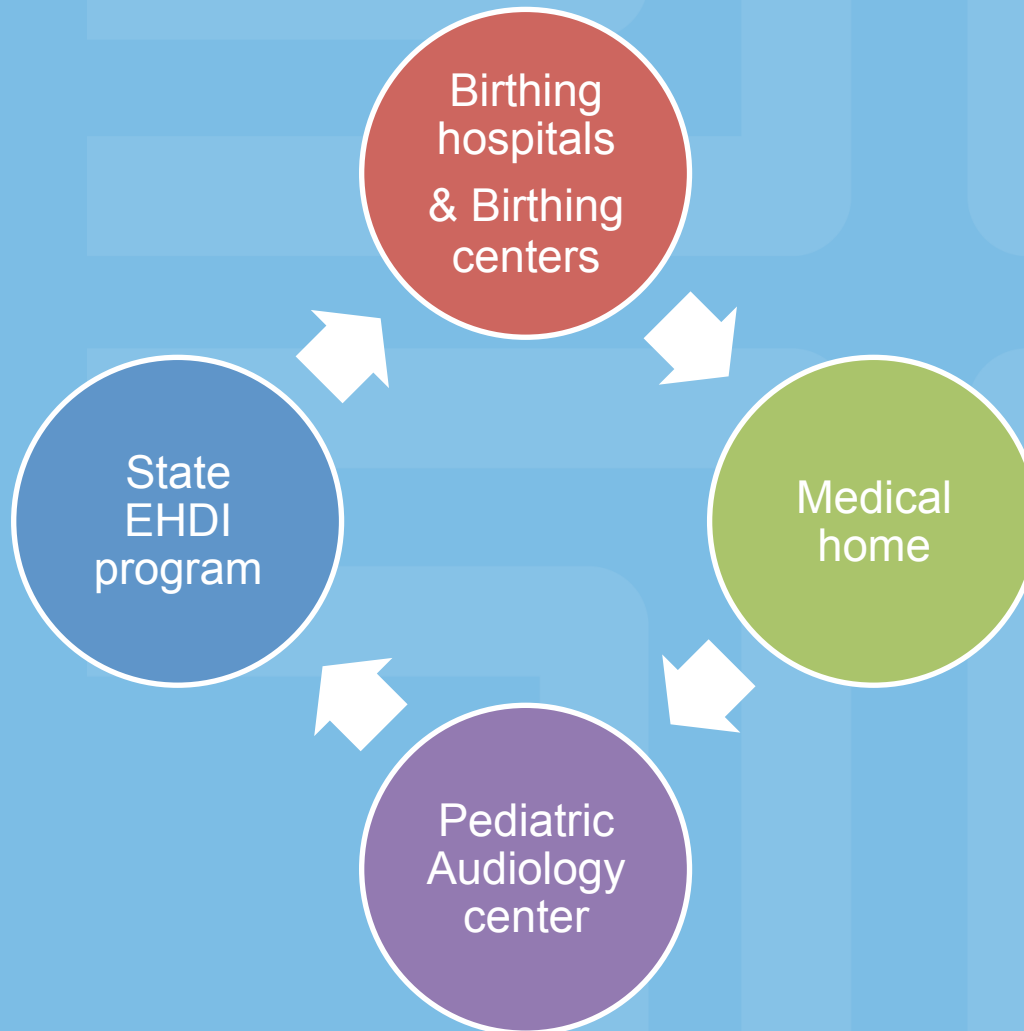
Services provided by St. Luke's

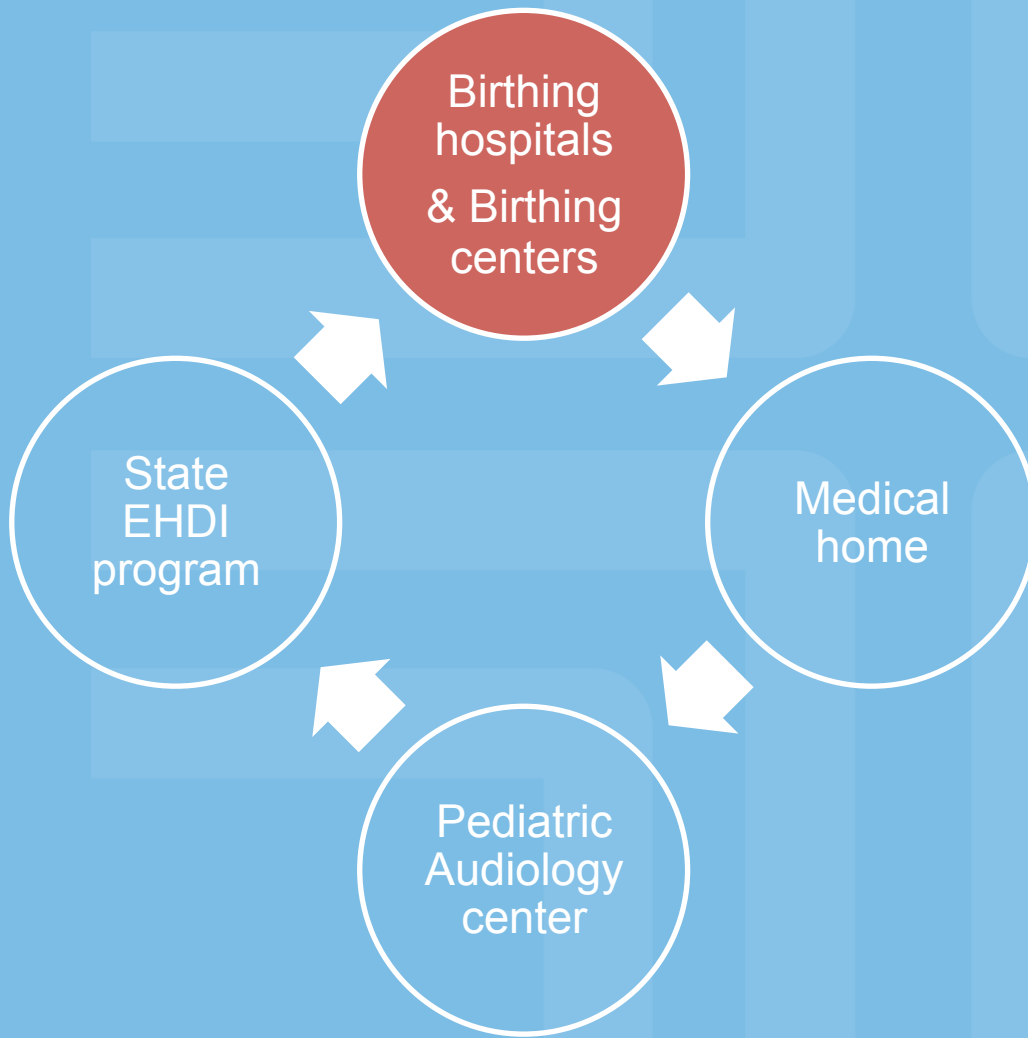
Goals of risk monitoring program

- 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



Risk Monitoring Program





Birthing Hospitals/Birthing Center roles:

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



Provide on-site hospital/birthing center training

- **Provide training annually**
 - **Physicians**
 - **Nurse Managers**
 - **Nurses (Screeners)**
 - **Midwives**



Script for hospital staff

“Your baby has been identified as having a high risk (_____) for a late-onset hearing loss. The recommended protocol for babies with high risk indicators is an audiological evaluation around 9 months of age. We will provide a copy of this referral form to the pediatric audiology center and they will contact you for an appointment.”



Idaho EHDI Referral forms

STEP 3: RISK ASSESSMENT:

RISK INDICATORS for LATE-ONSET CHILDHOOD HEARING LOSS:

- Family History of Permanent Hearing Loss < 18 yrs of age
 - NICU stay >5 days
 - Syndrome Associated with HL (e.g. Downs)
 - Congenital Infection (e.g. T-O-R-C-H)
 - Postnatal Infection (e.g. Meningitis)
 - Craniofacial Anomalies
 - Ototoxic Medications - any amount
 - Mechanical Ventilation - any amount
 - Head Trauma Other _____
- (monitoring through age 3 is recommended for most risk factors)

IDAHO SOUND BEGINNINGS (ISB)
 Early Hearing Detection and Intervention
 Department of Health and Welfare, Infant-Toddler Program

FAX TO (208) 332-7331
 Within 5 days

Complete Form for All: Refers Risks Transfers Missed or Incomplete

Birth Hospital: _____
 (*Transfers only) Receiving Hospital: _____ (Please Print Firmly)

Within 5 days of screening or discharge—Distribute copies to: Audiologist IDB Hospital Parent Physician
 White Gold Pink Green Yellow

Send to: Idaho Sound Beginnings-EHDI, PO Box 83726, Boise, ID 83725-9816 or Fax: (208) 332-7331

1. BABY'S INFORMATION:
 Baby's Med Record #: _____
 Baby's Name: Last _____ First _____
 DOB: ____/____/____ Gender: M F
 Nursery: Well Baby NICU/Special Care
 Baby's Primary Physician/Clinic: _____
 Mother's name: _____

2. CONTACT INFORMATION:
 Parent/Guardian: Last _____ First _____
 Address: _____
 City: _____ State: _____ Zip: _____
 Main Phone: _____ Text: _____
 Alternate Phone/Contact: _____
 Email/Other contact: _____

3. HEARING SCREEN RESULTS:
 First Screen: R Pass Refer No Result
 L Pass Refer No Result
 Sex Pass Refer No Result
 Second Screen: R Pass Refer No Result
 L Pass Refer No Result

4. RISK ASSESSMENT (check all that apply)
 For LATE-ONSET CHILDHOOD HEARING LOSS:
 Family History of Permanent Hearing Loss <18 yrs of age
 NICU stay >5 days
 Syndrome Associated with HL (e.g. Downs)
 Congenital Infection (e.g. T-O-R-C-H)
 Postnatal Infection (e.g. Meningitis)
 Craniofacial Anomalies
 Ototoxic Medications - any amount
 Mechanical Ventilation - any amount
 Parent or Physician Concern
 Head Trauma Other _____
 (Monitoring through age 3 is recommended for most risk factors)

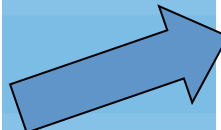
Nursing/screening staff will inform you of the final results of the baby's hearing screen and give you a copy of these results. If your baby **passes** the hearing screen, you will be given an appointment and/or follow-up information. If you have questions please contact Idaho's Early Hearing Program, Idaho Sound Beginnings, at (208) 332-8829. Financial Assistance for diagnostic testing may be available.

Your baby **did not pass** the hearing screen. Hearing testing should be completed before baby is 6 months old. If baby is not hearing, all the services necessary for speech and language development, early identification can minimize any communication delays.
 Your baby is **at risk** for late-onset childhood hearing loss. Hearing testing of approximately 18 months of age is recommended for most risk factors. A Pediatric Audiologist can advise on the appropriate monitoring schedule for your baby.

Audiologist: _____
 Phone: _____
 Address: _____
 App. date/time: _____

I have been informed of my baby's hearing screen results and of the need for diagnostic audiology (hearing testing before the age of 3 months if baby did not pass) to determine if a hearing loss is present. If baby passed the hearing screen, all risk factors are present (see above). Hearing testing is recommended at approximately 3 months of age. (American Academy of Pediatrics (AAP) Guidelines)
 I hereby give permission to the staff of the above-named hospital/screening site to release medical information necessary to complete an audiology evaluation for my child to the label audiologist(s) or the audiologist of my choice and physician. I also give permission to the hospital and audiologist/clinician, and Idaho Sound Beginnings to share the results of the hearing screening and diagnostic audiology evaluation with the above-named physician, the Idaho Infant-Toddler Program, Idaho School for the Deaf and Blind, and Idaho Needs 4 Voices. I understand that the information will only be used to ensure that appropriate and timely medical, educational, and audiology services are made available to my child.
 Hearing screening results are reported to Idaho Sound Beginnings-Idaho's Early Hearing Detection & Intervention Program and are not shared with the above listed entities or any other outside entities without parent/guardian consent.
 I have had the opportunity to read this clinic notice of Privacy Practices. I understand that this information will not be shared with unauthorized individuals. This authorization expires 36 months from the date signed.

PARENT/GUARDIAN: _____ DATE: _____ 8/2012





Guidelines for Risk Monitoring for Delayed Onset Hearing Loss

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.

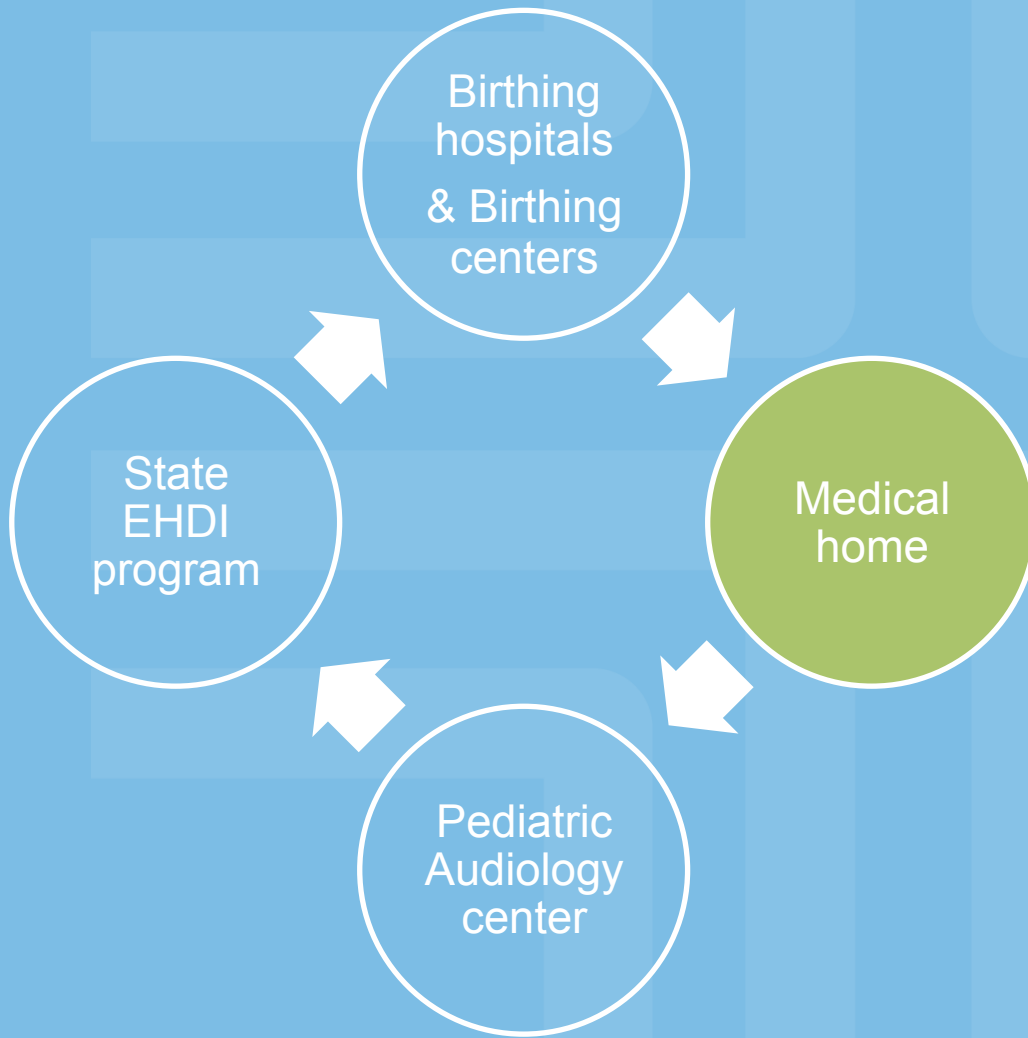
NOTE: If baby REFERS on the newborn hearing screening after two attempts – Recommendation for Diagnostic ABR evaluation to be completed by 3 months of age (JCIH 2007)

* Any parental/caregiver hearing concerns warrants a referral to a pediatric audiologist.

** Infants readmitted to the hospital within the first 30 days of life should be re-screened if any risk indicators are present.

References:

- Figor BJ, Neebult MW, Mullen CH, Feldman HA, Jones DT. Factors associated with sensorineural hearing loss among survivors of extracorporeal membrane oxygenation therapy. *Pediatrics* 2005; 115(6):1519-1528.
- Joint Committee on Infant Hearing, Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*. 2007; 120(4):898-921. doi: 10.1542/peds.2007-2353.
- Van Riper, Lori A.; Kilroy, Paul R. ABR Hearing Screening for High-Risk Infants. *American Journal of Otolaryngology*. 20(4):516-521, July 1999.



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**





Guidelines for Risk Monitoring for Delayed Onset Hearing Loss

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- *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.

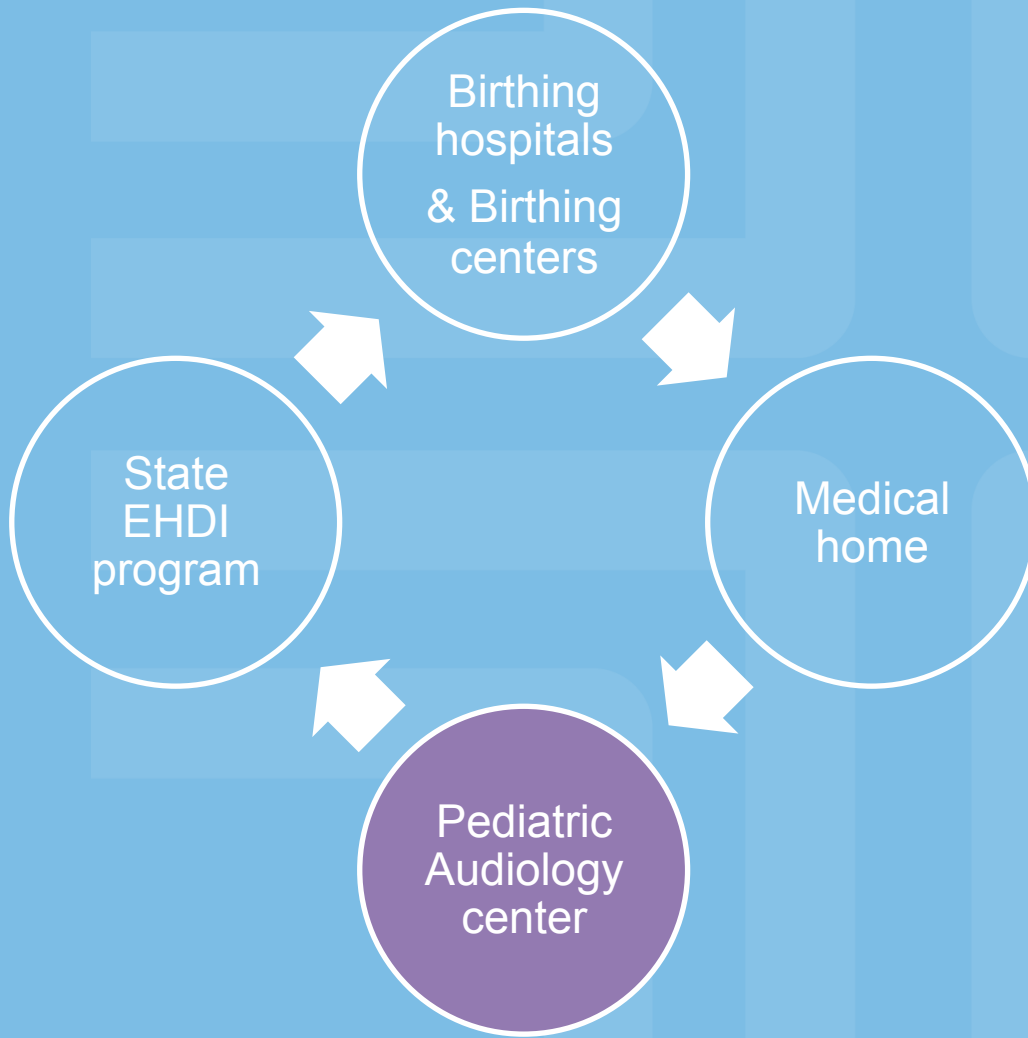
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- Van Riper, Lori A.; Kilroy, Paul R. ABR Hearing Screening for High-Risk Infants. *American Journal of Otolaryngology*. 20(4):516-521, July 1999.



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**



Monitoring in audiology clinic

- 5 audiology clinics
- Southwest Idaho and eastern Oregon
- 20 audiologists



Increasing # of hospital referring for risk indicators

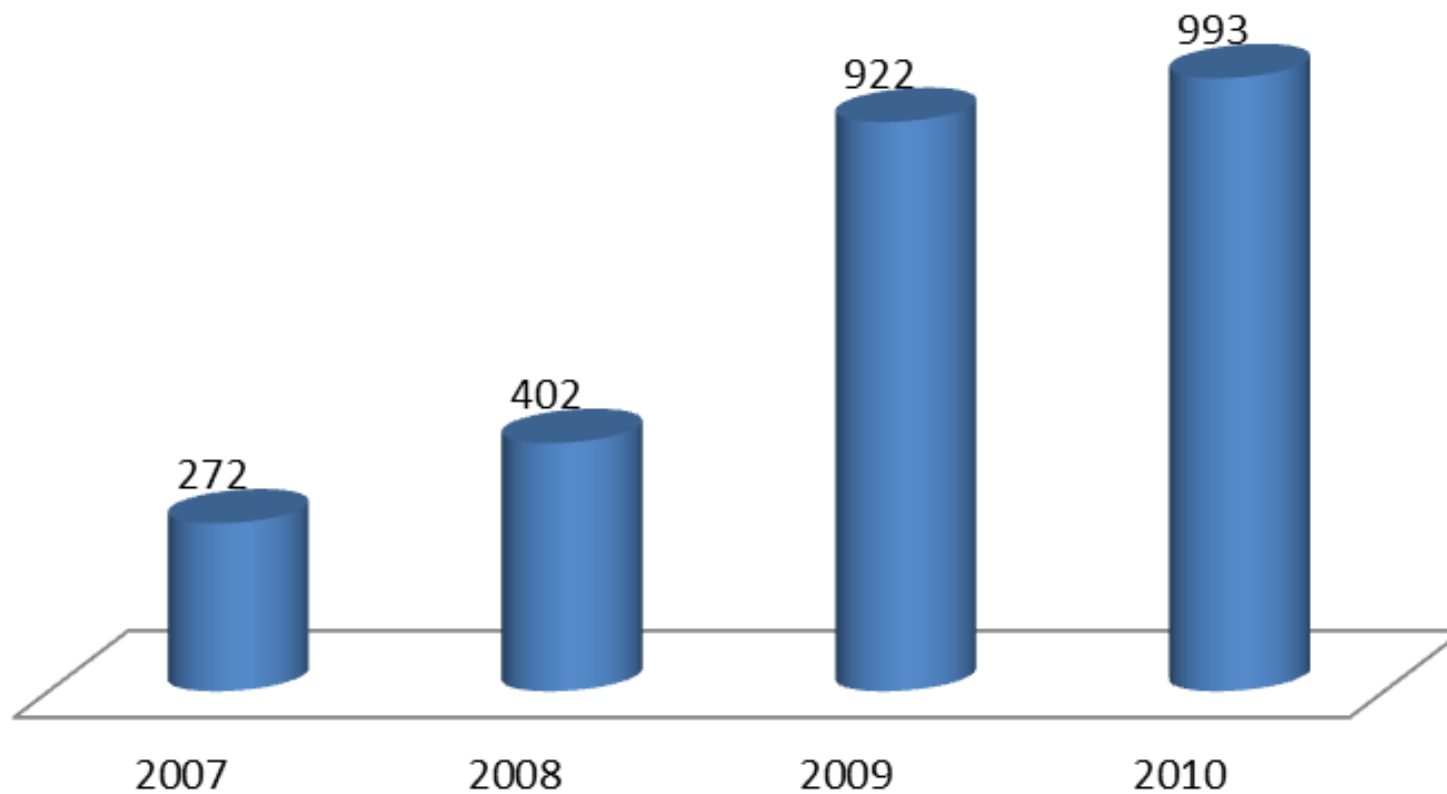
2007 & 2008: 2 HOSPITALS

2009 & 2010: 3 HOSPITALS

2011: 4 HOSPITALS



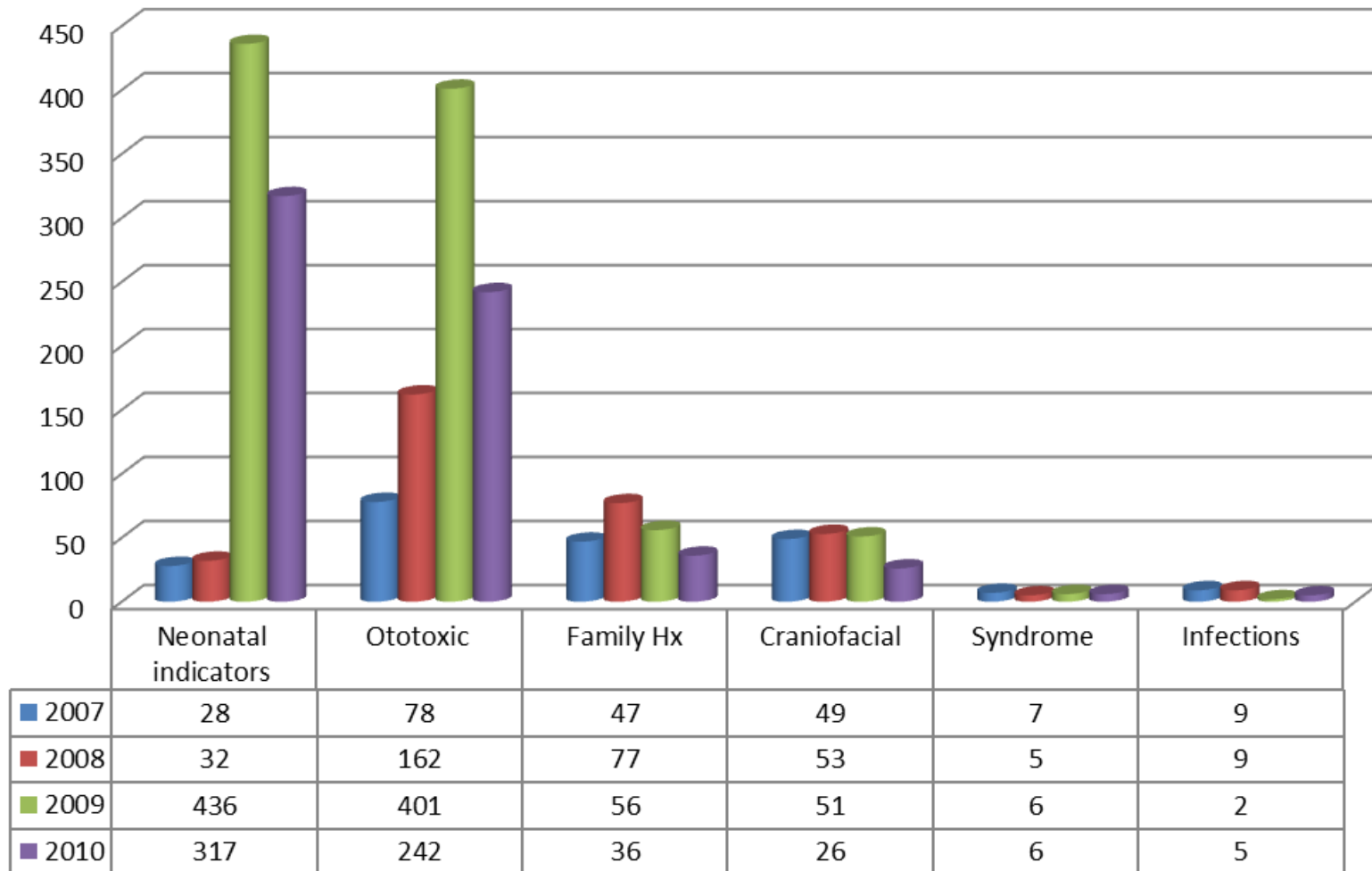
■ # babies referred for high risk monitoring



Elks HEARING & BALANCE
CENTER

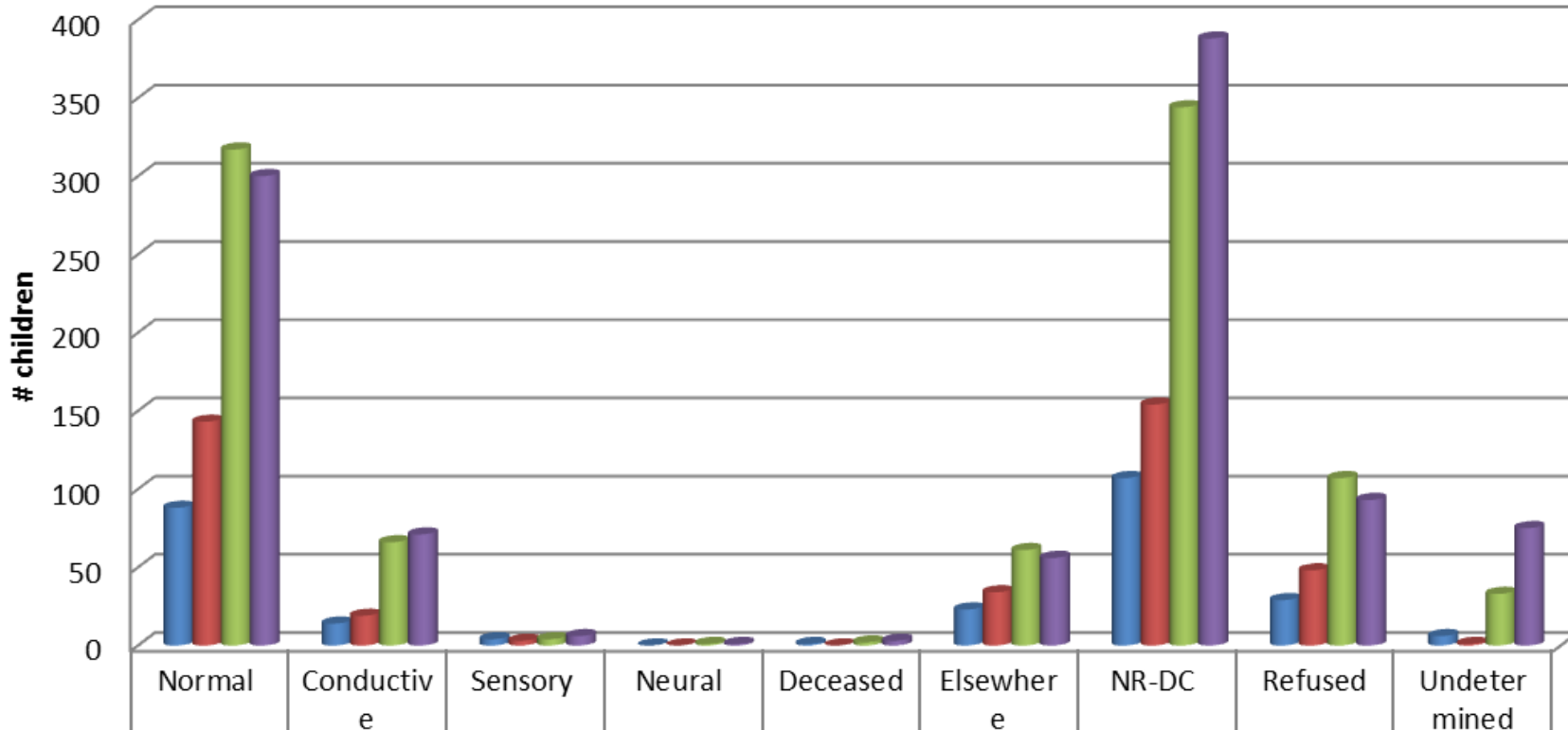


referrals per high risk indicator



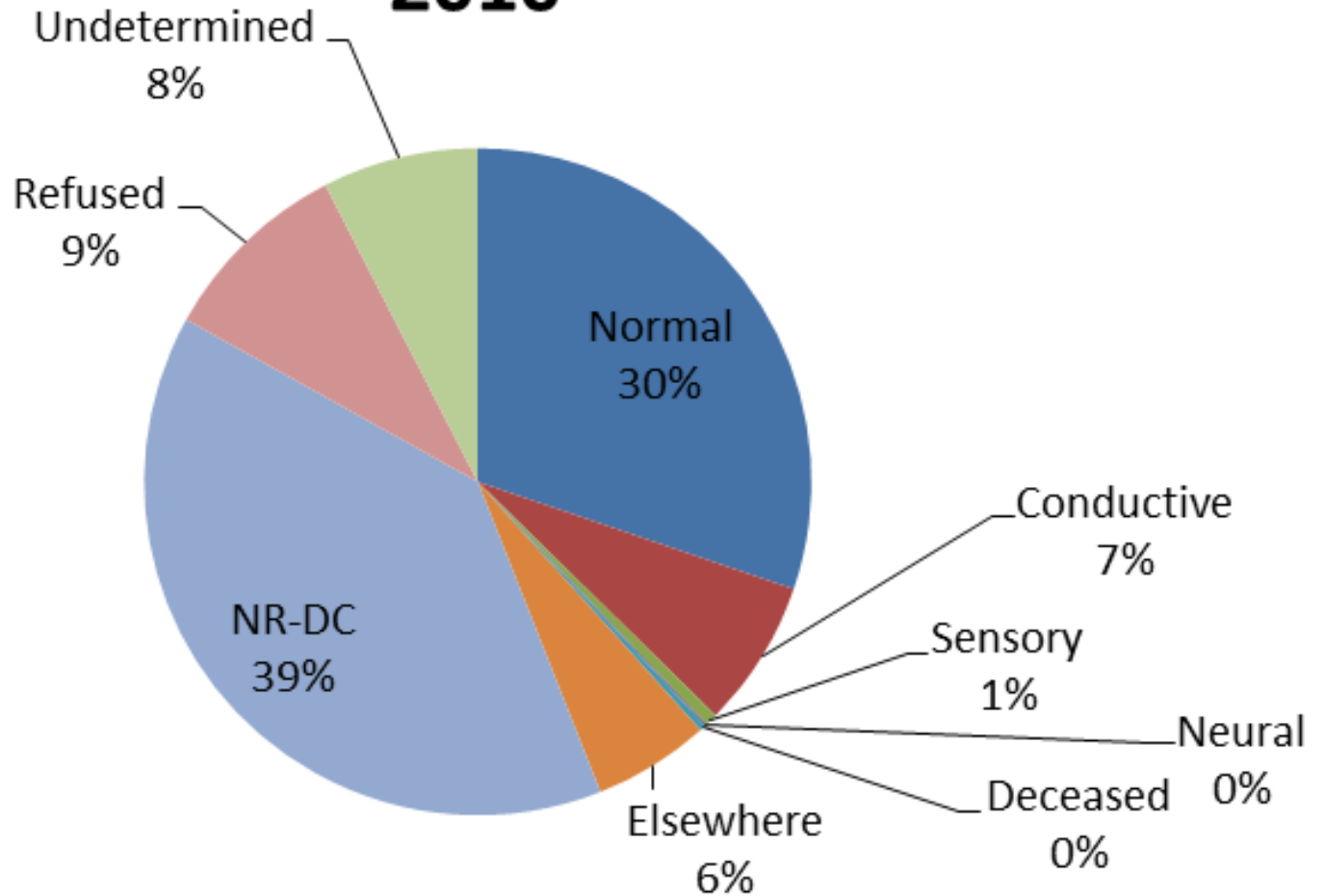
NAME	DOB	MR#	1st ATTEMPT	2nd	3rd	RESULTS	SENT TO EHD	Au.D.	FAM	NEONATAL	UTEROTOT	CRANIO	SYND	NEURO	HEAD	POST
NAME, #####	#####	#####	not good-sent letter			NR-DC	yes	Jess		1		1				
NAME, #####	#####	####	April Hold are	for 4/2	forms to	WNL	yes	Arpil W				1				
NAME, #####	#####	####	to Eagle.5/3/1	for 7/1	r 8/24	WNL	yes	Jessica E		1		1				
NAME, #####	#####	####	Jan Hold are	1/13/1	11 lv	NR-DC	yes	Jess				1				
NAME, #####	#####	####	1 ISB to Namp	07/6/11	w/Jes	WNL	yes	Jess	1							
NAME, #####	####	###	copy sent to Mem	in Boise	for care											
NAME, #####	#####	####	B in July hold.	11 LVM	1 sen	NR-DC	yes	Jess		1		1				
NAME, #####	#####	####	B in July hold.	11 LVM	1 ser	NR-DC	yes	Jess		1		1				
NAME, #####	####	###	h Oct Hold are	B to On	5/10	NR-DC	yes	Jess		1		1				
NAME, #####	#####	####	11 ISB to Eagl	Forms to Del		WNL	yes	April W.		1		1				
NAME, #####	#####	####	B in July hold.	11 LVM	7/08	NR-DC	yes	Jess		1						
NAME, #####	####	###	Jan Hold are	11 lvm	21/1	WNL	yes	Shannon		1						
NAME, #####	#####	####	1 ISB to Boise	good-s	7/13	SNHL	yes	Jess		1						
NAME, #####	#####	####				COND	yes	Jenna								
NAME, #####	#####	####				WNL	yes	Alison								
NAME, #####	#####	####	Boise	nt to sched		REFUSED	yes	Jess								
NAME, #####	#####	####	oise front offic	# sent letter		NR-DC	yes	Jess		1						
NAME, #####	#####	####	Feb Hold are	/11 LVM	1 lvm	NR-DC	yes	Jess				1				
NAME, #####	#####	####	March Hold are	rof town	sent	NR-DC	yes	Jess								
NAME, #####	#####	####	h Oct Hold are	B to Ontario.		WNL	yes			1		1				
NAME, #####	#####	####	h Oct Hold are	B to Ontario.		COND	yes	Larissa		1		1				
NAME, #####	#####	####	April Hold are	SB to Ontario		NR-DC	yes	Jess		1		1				
NAME, #####	#####	####	1 ISB to Boise	d 7/15	w/Jes	WNL	yes	Jess		1		1				
NAME, #####	####	###	1 ISB to Boise	d 7/15	w/Jes	WNL	yes	Jess		1		1				
NAME, #####	#####	####	h April Hold are	/11 LV	7-14	WNL	yes	Maria								
NAME, #####	#####	####	sent to Nici/Boise.	BD		COND	yes	Jess	1	1		1				
NAME, #####	#####	####	sent to Nici/Boise.	BD		WNL	yes	Jess	1	1		1				

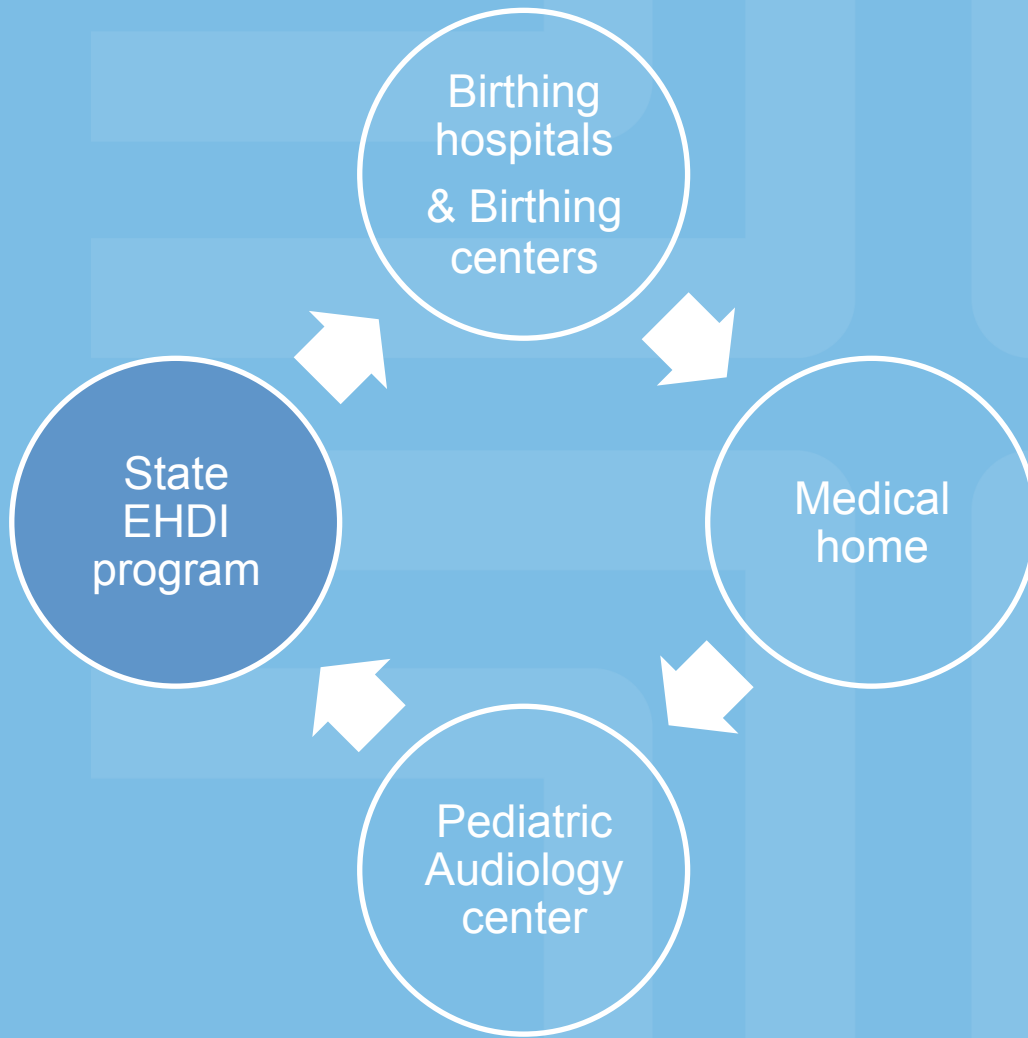
Tracking outcomes



	Normal	Conductive	Sensory	Neural	Deceased	Elsewhere	NR-DC	Refused	Undetermined
2007	88	14	4	0	1	23	107	29	6
2008	143	19	3	0	0	34	154	48	1
2009	317	66	4	1	2	61	344	107	33
2010	300	71	6	1	3	56	388	93	75

2010





State EHDI program roles:

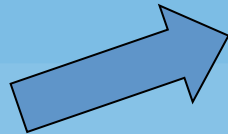
- Providing training and support for hospitals, birthing center, physicians, and pediatric audiologists on risk factor
- Providing a method for hospitals, birthing centers and pediatric audiologists to report information regarding infants with risk indicators to the state EHDI program
- Tracking and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



Idaho EHDI program



Data collected by referral forms



STEP 3: RISK ASSESSMENT:

RISK INDICATORS for LATE-ONSET CHILDHOOD HEARING LOSS:

- Family History of Permanent Hearing Loss < 18 yrs of age
 - NICU stay >5 days
 - Syndrome Associated with HL (e.g. Downs)
 - Congenital Infection (e.g. T-O-R-C-H)
 - Postnatal Infection (e.g. Meningitis)
 - Craniofacial Anomalies
 - Ototoxic Medications - any amount
 - Mechanical Ventilation - any amount
 - Head Trauma Other _____
- (monitoring through age 3 is recommended for most risk factors)

IDAHO SOUND BEGINNINGS (ISB)
 Early Hearing Detection and Intervention
 Department of Health and Welfare, Infant/Toddler Program
 FAX TO (208) 332-7331
 Within 5 days

Complete Form for All: Refers Risks Transfers Missed or Incomplete

Newborn Hearing Screening Referral Form
 Birth Hospital: _____
 (Transfers only) Receiving Hospital: _____ (Please Print Firmly)

Within 5 days of screening or discharge—Distribute copies to: Audiologist ISB Hospital Parent Physician
 White Gold Pink Green Yellow

Send to: Idaho Sound Beginnings-EHDI, PO Box 83726, Boise, ID 83725-9816 or Fax: (208) 332-7331

1. BABY'S INFORMATION:
 Baby's Med Record #: _____
 Baby's Name: Last _____ First _____
 DOB: ____/____/____ Gender: M F
 Nursery: Well Baby NICU/Special Care
 Baby's Primary Physician/Clinic: _____
 Mother's name: _____

2. CONTACT INFORMATION:
 Parent/Guardian: Last _____ First _____
 Address: _____
 City: _____ State: _____ Zip: _____
 Main Phone: _____ Text: _____
 Alternate Phone/Contact: _____
 Email/other contact: _____

3. HEARING SCREEN RESULTS:
 First Screen: R Pass Refer No Result
 L Pass Refer No Result
 Second Screen: R Pass Refer No Result
 L Pass Refer No Result

4. RISK ASSESSMENT (check all that apply)
 For LATE-ONSET CHILDHOOD HEARING LOSS:
 Family History of Permanent Hearing Loss < 18 yrs of age
 NICU stay > 5 days
 Syndrome Associated with HL (e.g. Downs)
 Congenital Infection (e.g. T-O-R-C-H)
 Postnatal Infection (e.g. Meningitis)
 Craniofacial Anomalies
 Ototoxic Medications - any amount
 Mechanical Ventilation - any amount
 Parent or Physician Concern
 Head Trauma Other _____
 (monitoring through age 3 is recommended for most risk factors)

Monitoring staff will inform you of the final results of the baby's hearing screen and give you a copy of these results. If you baby passes the screen or follow-up for risks, you will be given an appointment and/or follow-up information. If you have questions please contact Idaho's Early Hearing Program, Idaho Sound Beginnings, at (208) 334-8829. Financial Assistance for diagnostic testing may be available.

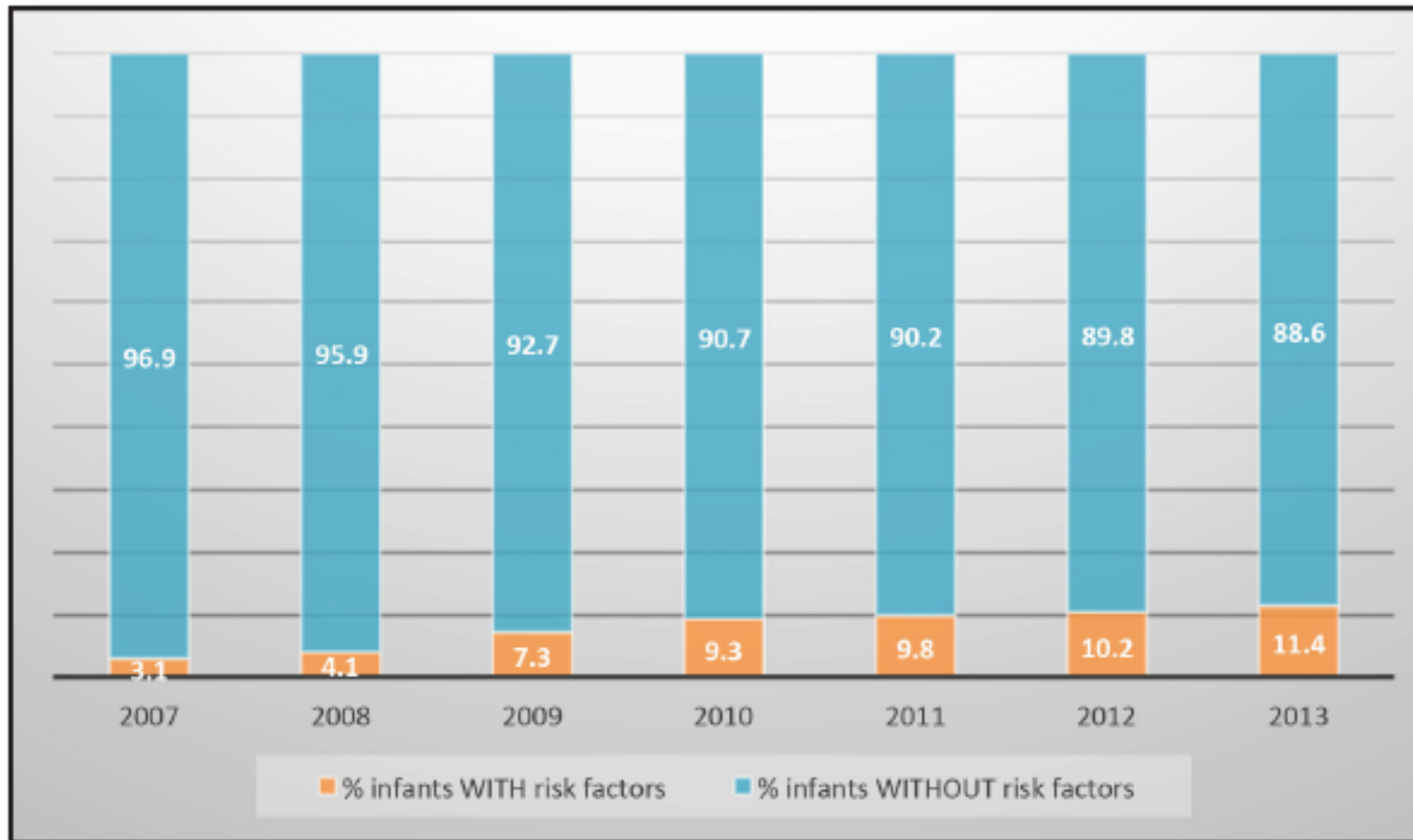
Your baby did not pass the hearing screen. Hearing testing should be completed before baby is 6 months old. If baby is not hearing, all the sources necessary for speech and language development, early identification can minimize any communication delays.
 Your baby is at risk for late-onset childhood hearing loss. Hearing testing of approximately 1 year of age is recommended for most risk factors. A Pediatric Audiologist can advise on the appropriate monitoring schedule for your baby.

Audiologist: _____
 Phone: _____
 Address: _____
 Appt. date/time: _____

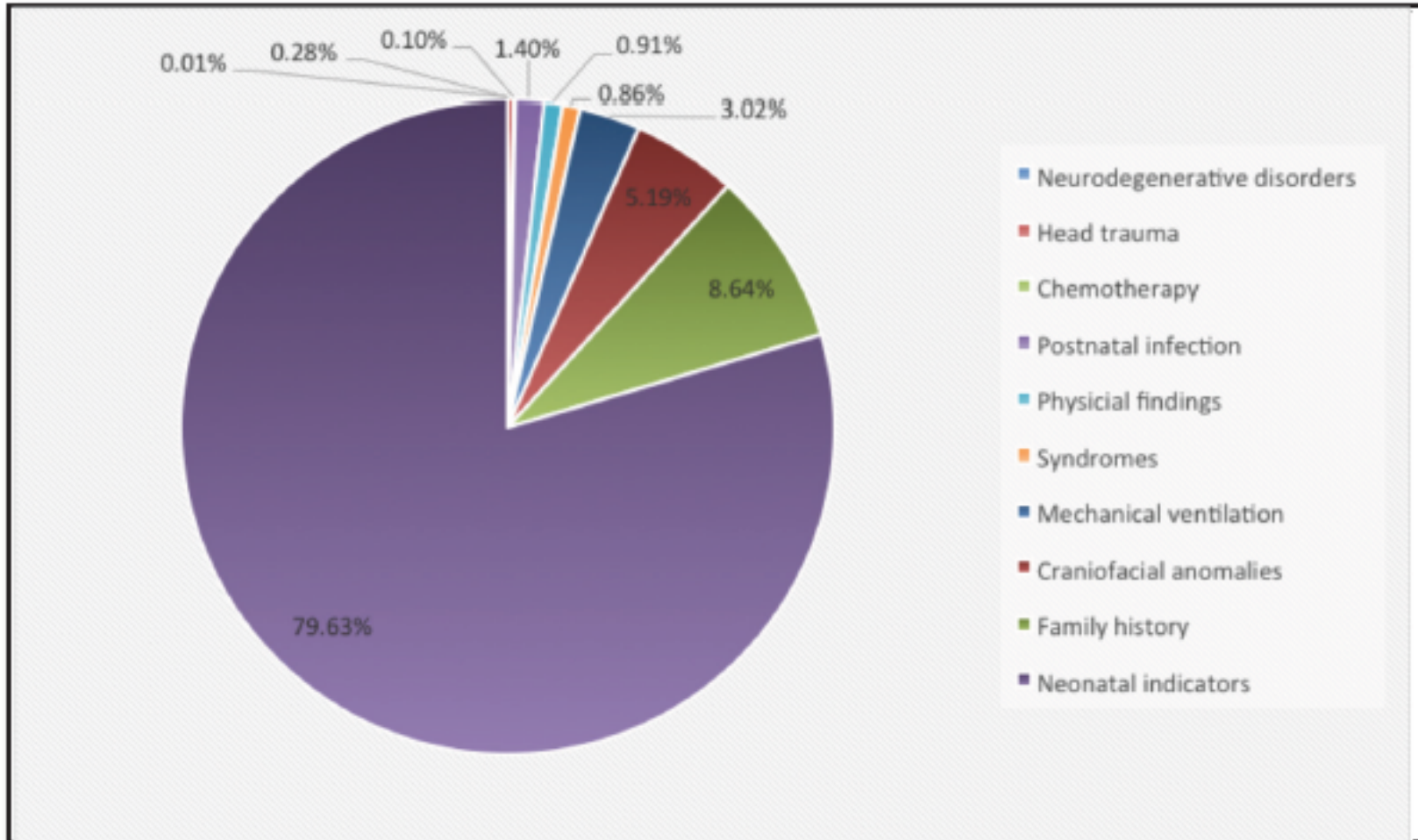
I have been informed of my baby's hearing screen results and of the need for diagnostic audiology (hearing testing before the age of 3 months if baby did not pass to determine if a hearing loss is present. If baby passed the hearing screen, all risk factors are present (see above). Hearing testing is recommended at approximately 3 months of age. (American Academy of Pediatrics (AAP) Guidelines).
 I hereby give permission to the staff of the above-named hospital/screening site to release medical information necessary to complete an audiology evaluation for my child to the lab(s) audiologist(s) or the audiologist of my choice and physician. I also give permission to the hospital and audiologist/clinician, and Idaho Sound Beginnings, to share the results of the hearing screening and diagnostic audiology evaluation with the above-named physician, the Idaho Infant/Toddler Program, Idaho School for the Deaf and Blind, and Idaho Needs 4 Voices. I understand that the information will only be used to ensure that appropriate and timely medical, educational, and audiology services are made available to my child.
 Hearing screening results are reported to Idaho Sound Beginnings-Idaho's Early Hearing Detection & Intervention Program and are not shared with the above listed entities or any other outside entities without parent/guardian consent.
 I have had the opportunity to read this clinic notice of Privacy Practices. I understand that this information will not be shared with unauthorized individuals. This authorization expires 36 months from the date signed.

PARENT/GUARDIAN: _____ DATE: _____ 8/2012

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



Number of Risk Indicators Reported in ISB 2007-2013 Data



Ototoxic medication



- January 2008-December 2014
- 4701 infants passed newborn hearing screening (Ototoxic medication only)
- 2 with diagnosed with delayed-onset hearing loss
 - 1 unilateral severe to profound
 - 1 bilateral mild to moderately severe



Idaho EHDI: Diagnostic testing recommendations for infants with risk indicators

Idaho Sound Beginnings Best Practice Protocol



Audiology Assessment for Risk Factor Follow-up

"The timing and number of hearing re-evaluations for children with risk factors should be **customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss.**"
(JCIH 2007 Position Statement)

Early and more frequent assessment may be indicated for children with: cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, culture-positive postnatal infections in association with sensorineural hearing loss; for children who have received ECMO or chemotherapy; and when there is a caregiver concern or a family history of hearing loss (JCIH 2008 clarification)

Recommended *Minimum* Standards:

Behavioral testing at 9 months of age**

All testing should be ear-specific

Tests included in this evaluation are:

- Family/child history
- Otoscopy
- Visual Reinforcement Audiometry for each ear:
 - Minimal Response levels for air conduction: 500, 2000 and 4000 Hz
 - Bone conduction as needed to rule out conductive pathology
 - Speech Awareness Thresholds (SAT)
- Limited Otoacoustic Emissions, DPOAE and/or TEOAE
- Immittance battery:
 - 226 Hz probe tone tympanometry-each ear.
 - Ipsilateral acoustic reflexes at 500, 1000 and 2000 Hz.(can also use broadband noise reflex – normal is less than 80 dB HL)
- ABR testing is indicated, if hearing loss is diagnosed, or if responses to behavioral audiometry are not reliable.

Based on: American Speech-Language-Hearing Association. (2004). *Guidelines for the Audiologic Assessment of Children from Birth to 5 Years of Age.* [Guideline]. www.asha.org/policy

**The recommendation for the initial risk factor evaluation to be done at 9 months of age is based on the following factors:

- The ease of testing using Visual Reinforcement Audiometry for the child and family. and
- The ability to gather the greatest amount of information quickly with minimal repeat visits, balanced with ...
- The ability to identify and address hearing losses and caregiver concerns early enough during the critical "language learning period" to maximize communication skills and minimize speech and language delays. Testing of a 2 year old can also be difficult, time consuming and delays identification.

Risk Indicators Associated with Permanent Congenital, Delayed-onset, or Progressive Hearing Loss in Childhood

1. **Caregiver concerns** regarding hearing, speech, language or developmental delay
2. **Family history** of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: **ECMO**, assisted ventilation, exposure to ototoxic medications (gentamycin/tobramycin) or loop diuretics (furosemide/Lasix) and hyperbilirubinemia requiring exchange transfusion.
4. In utero infections: **CMV**, herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical finding, such as a white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. **Syndromes associated with hearing loss** or progressive or late onset hearing loss such as neurofibromatosis, osteopetrosis and Usher syndrome; other frequently identified syndromes including Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. **Neurodegenerative disorders**, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. **Culture-positive postnatal infections associated with sensorineural hearing loss**, including confirmed bacterial and viral (especially herpes viruses and varicella meningitis).
10. Head trauma, especially basal skull/temporal bone fractures that requires hospitalization.
11. **Chemotherapy**

Risk factors **bolded** are considered to have a greater concern for delayed onset hearing loss and monitoring of those children should be **more frequent** than once following the neonatal period.

Idaho Sound Beginnings (EHDI)

Infant Toddler Program, 450 W. State St.FI-5, Boise, ID 83720-0036
(208) 334-0829 (208) 332-7331 FAX
Cynthia Carlin, EHDI Project Coordinator -- carlinc@dhw.idaho.gov

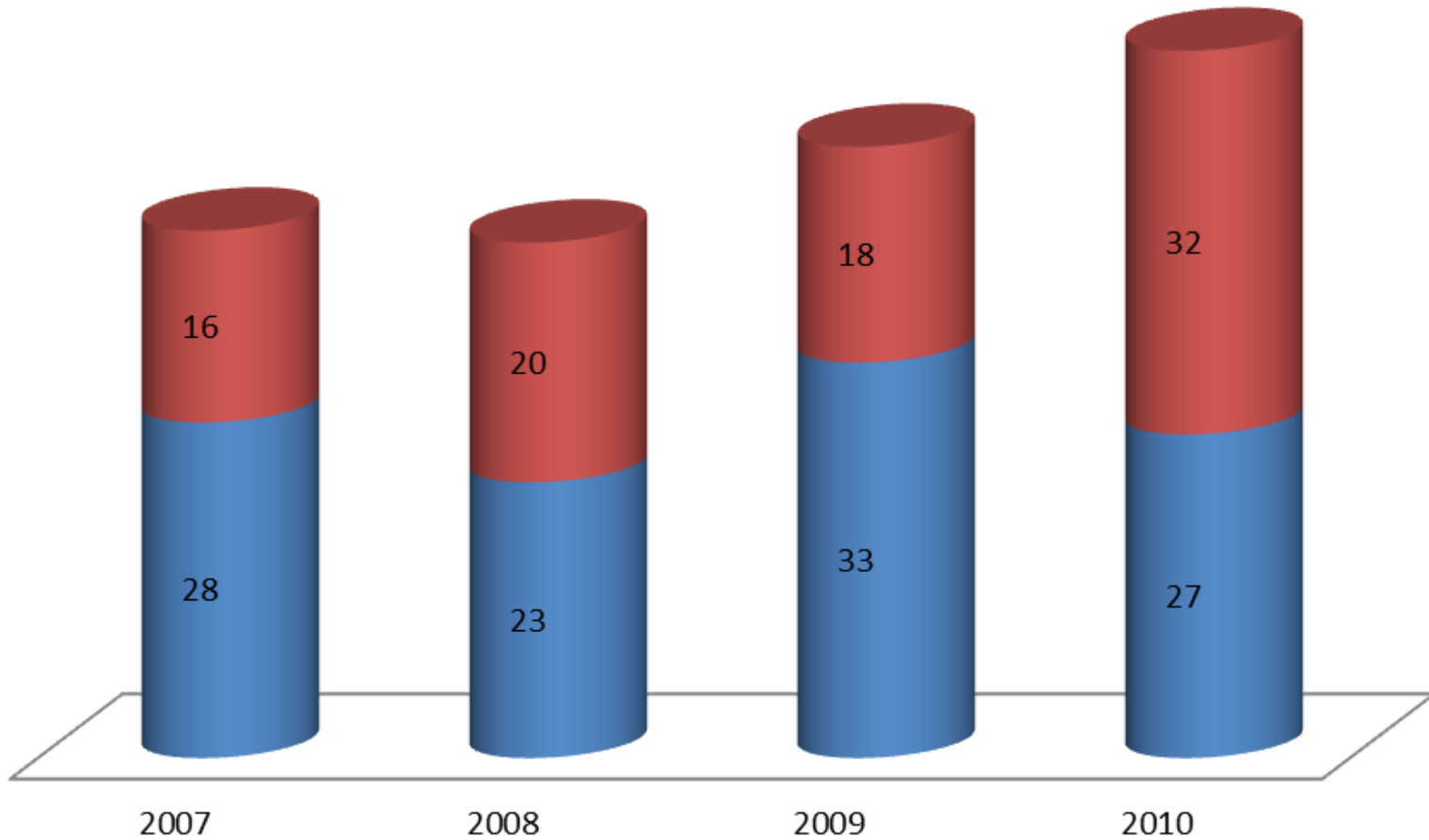


Principles and Guidelines for Early Hearing Detection and Intervention Programs:
Appendix 2. Joint Committee on Infant Hearing
2007 Position Statement (www.jcih.org)

Funding Provided By Maternal & Child Health Bureau (MCHB), Health Resources And Services Administration (HRSA)

Idaho EHDI: Diagnosed hearing loss

- # infants w/hearing loss & HIGH RISK INDICATORS
- # infants w/hearing loss & NO RISK INDICATORS



Idaho data (2007-2011)

Delayed onset hearing loss

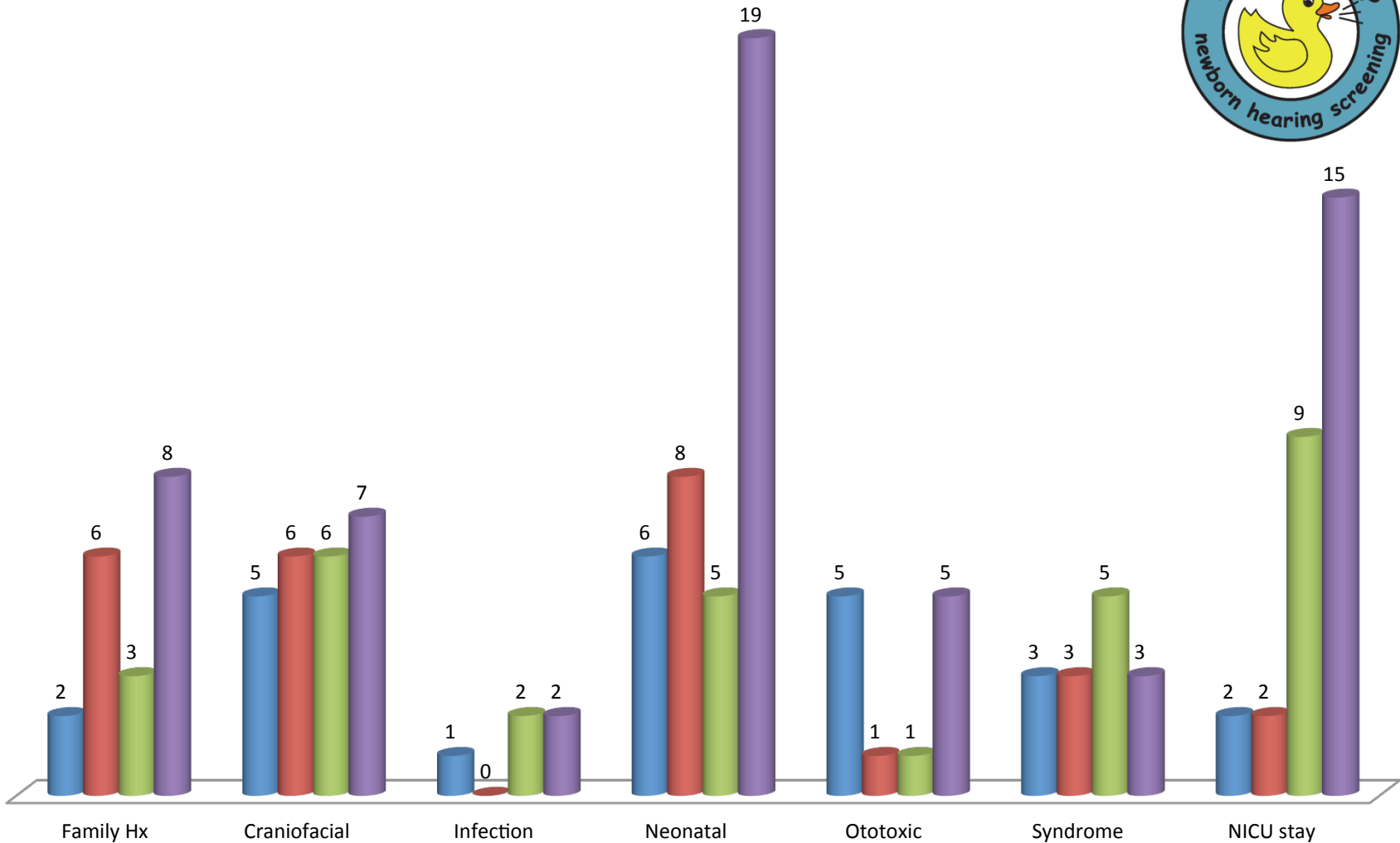
- **2.7 infants per 10,000 diagnosed with delayed onset hearing loss with risk indicators**
 - Those with hearing loss the most frequently reported risk indicators were NICU stay (15 infants), ototoxic medications (13 infants)



of risk indicators reported in infants with hearing loss



■ 2007 ■ 2008 ■ 2009 ■ 2010





Guidelines for Risk Monitoring for Delayed Onset Hearing Loss

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.

NOTE: If baby REFERS on the newborn hearing screening after two attempts – Recommendation for Diagnostic ABR evaluation to be completed by 3 months of age (JCIH 2007)

* Any parental/caregiver hearing concerns warrants a referral to a pediatric audiologist.

** Infants readmitted to the hospital within the first 30 days of life should be re-screened if any risk indicators are present.

References:

Fligor BJ, Newkirk MW, Mullen CH, Feldman HA, Jones DT. Factors associated with sensorineural hearing loss among survivors of extracorporeal membrane oxygenation therapy. *Pediatrics* 2005; 115(8):1519-1528.

Joint Committee on Infant Hearing. Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*. 2007; 120(4):898-921. doi: 10.1542/peds.2007-2333.

Van Riper, Lori A.; Kileny, Paul R. ABR Hearing Screening for High-Risk Infants. *American Journal of Otolaryngology*. 20(4):516-521, July 1999.

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.



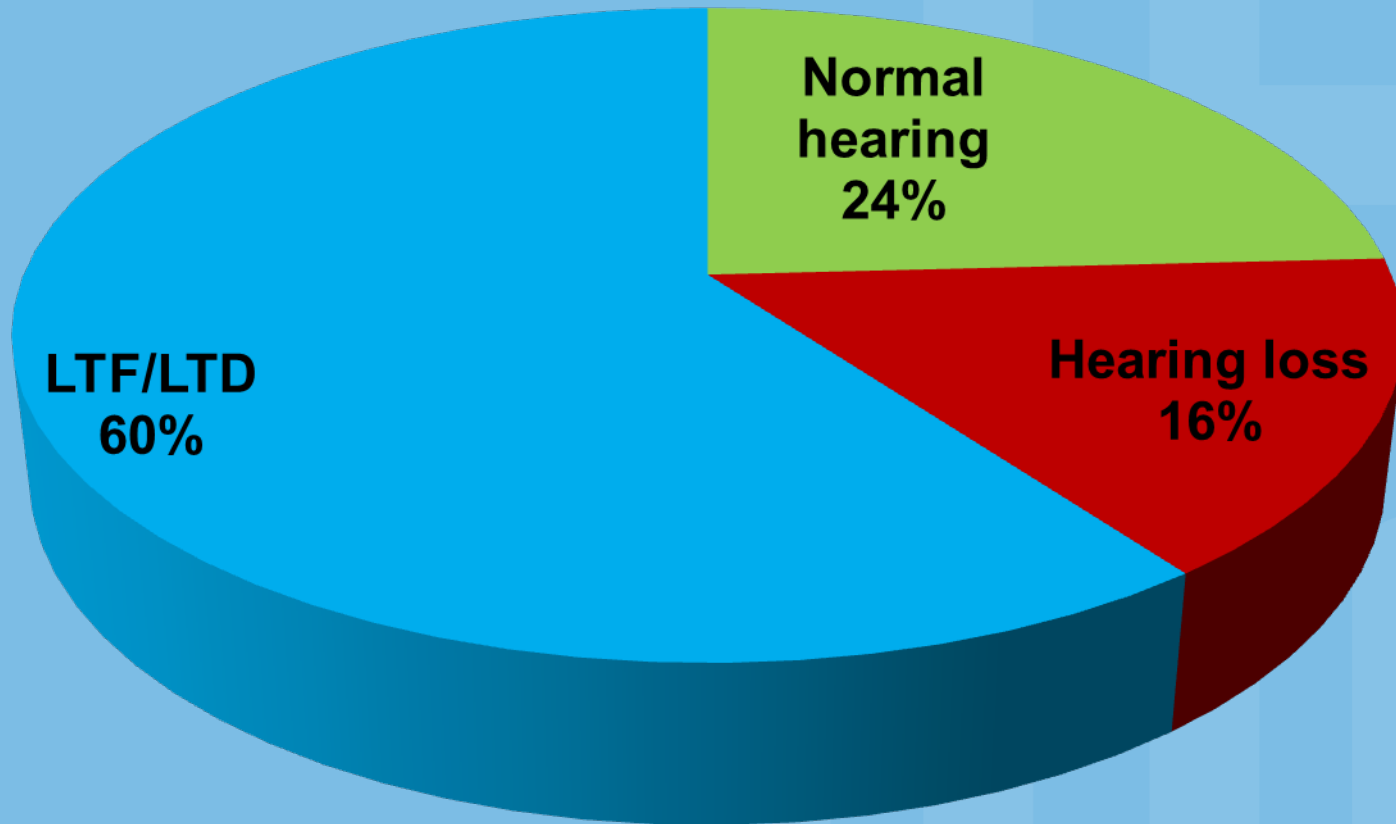
Class B: Risk indicators

- *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

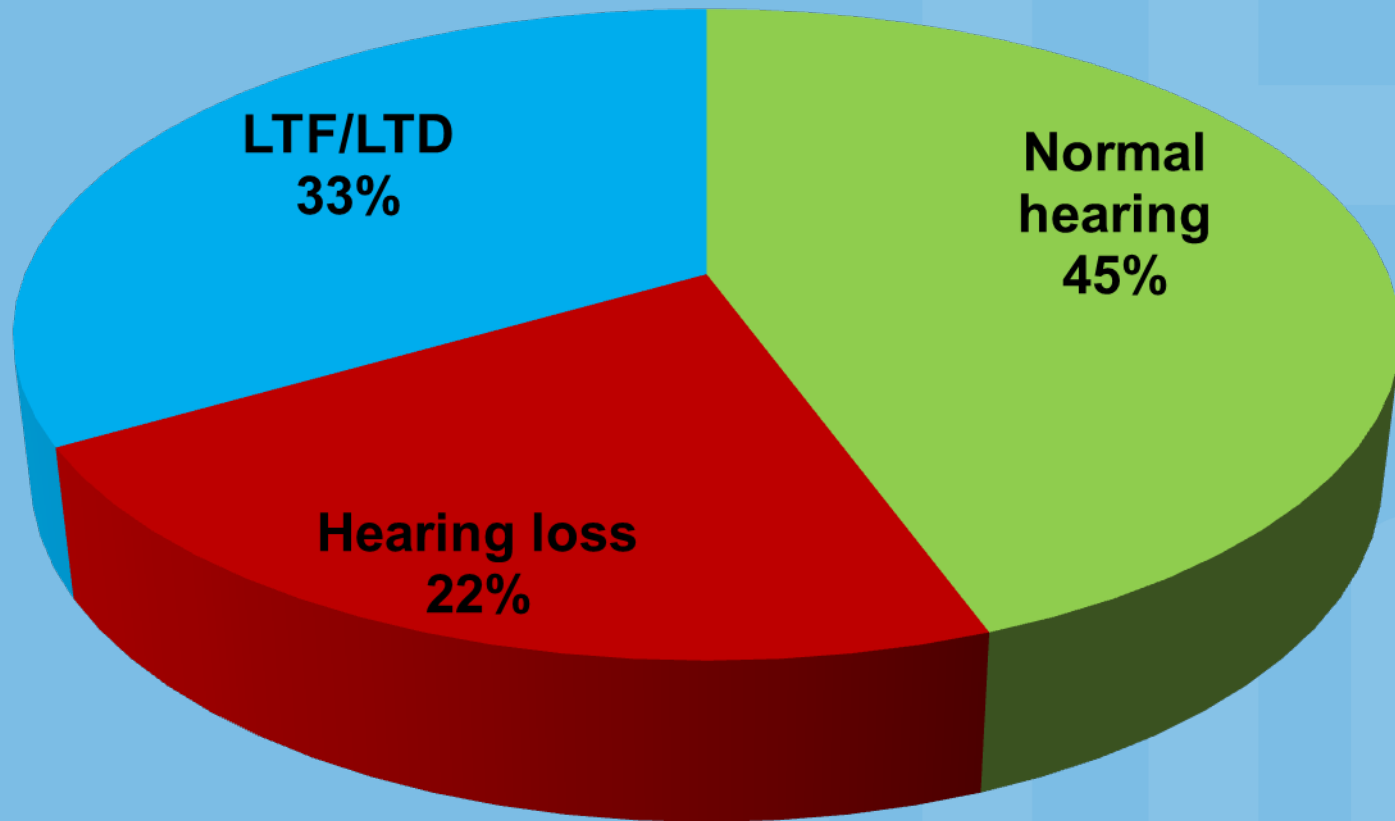
If baby passes the newborn hearing screening & has one or more CLASS B risk indicators = Recommendation for diagnostic pediatric hearing evaluation by 1 year of age.



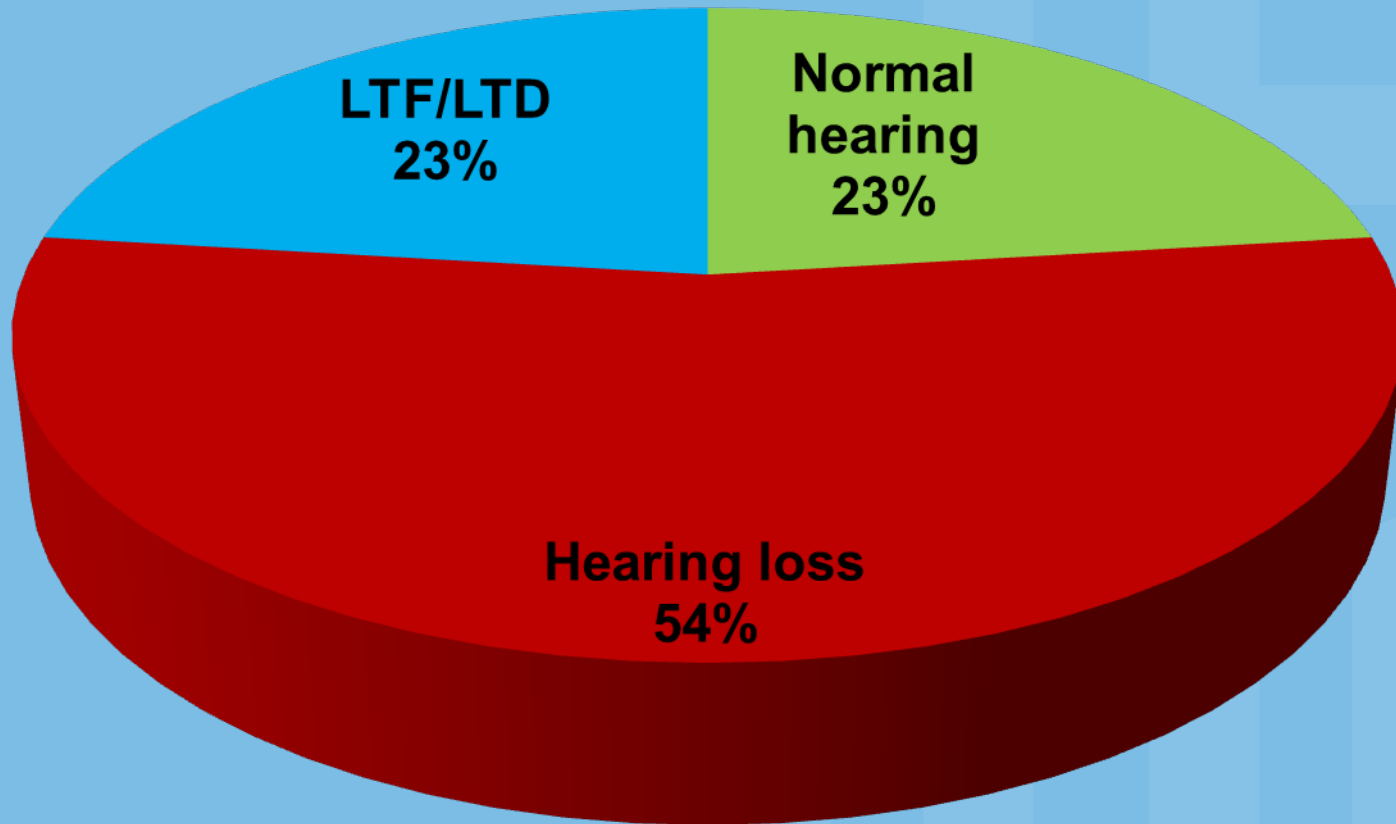
Class A risk indicators (n= 153)



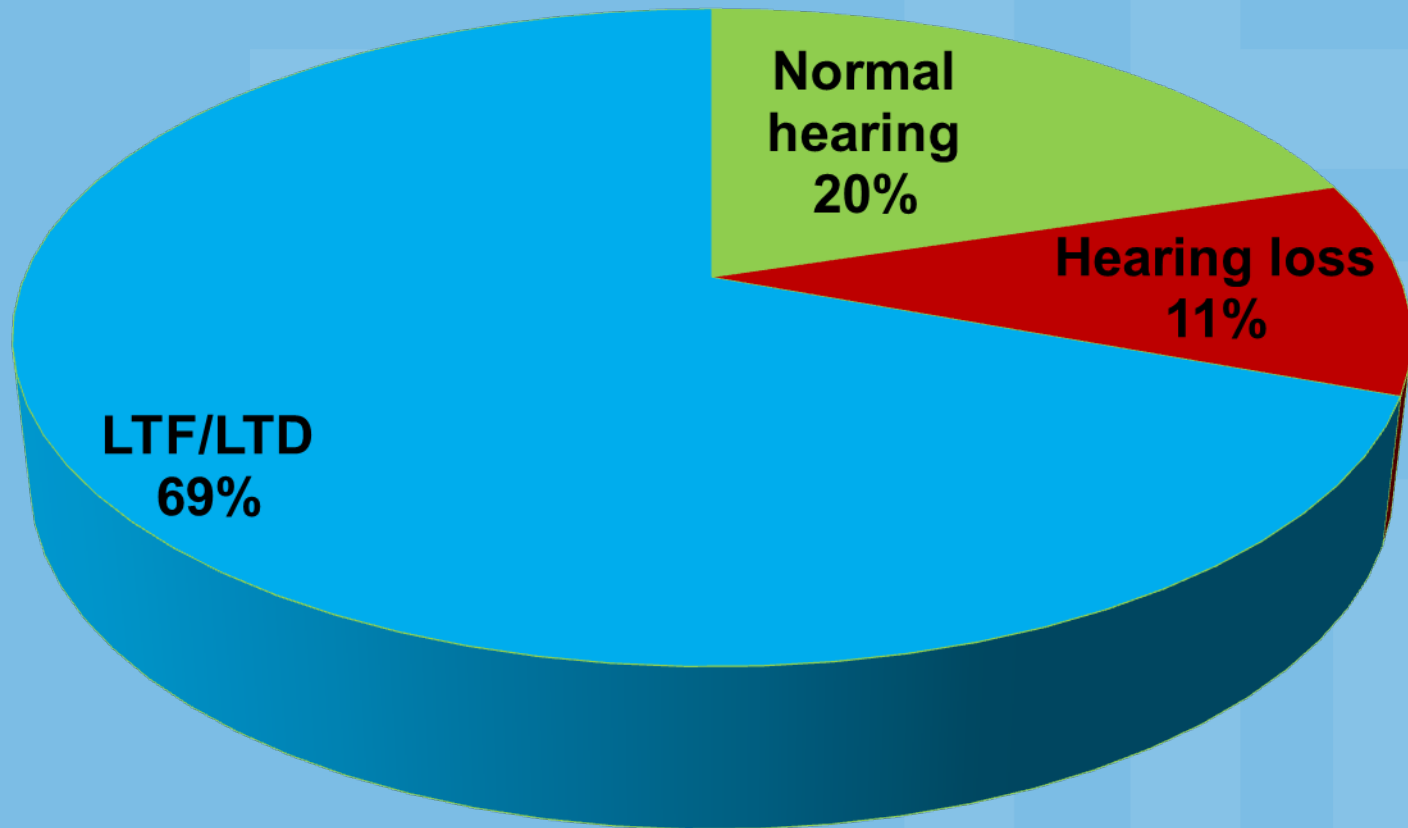
In utero & postnatal infections (n =18)



Syndromes (n=13)



Craniofacial anomalies (n=114)



Things to remember

- Risk monitoring programs need participation from hospital, birthing centers, medical homes, audiology centers, and state EHDI programs
- Training from state EHDI programs is important
- No “gold standard” protocol for risk indicator monitoring (ototoxic medications)
- Data collection is an important

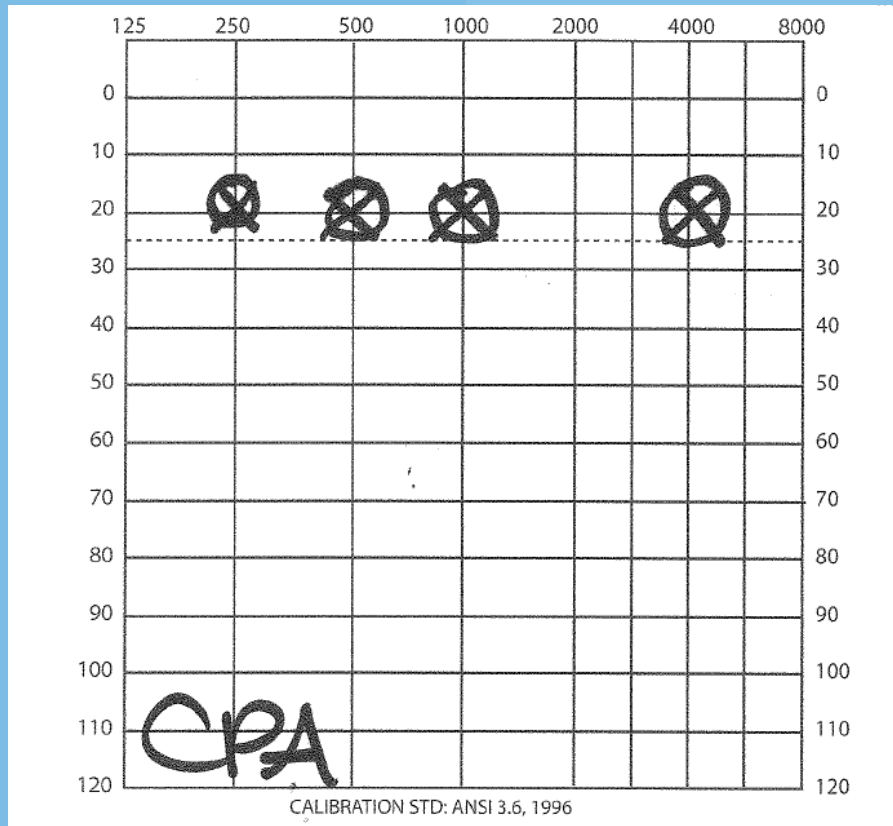


Case #1

- 4 yr old female
- Reason for referral: Speech delays
 - Speech therapy twice per week
- Birth history:
 - 32 weeks gestation (2lb 6oz)
 - NICU stay 1 month
 - Ototoxic medication (Gentamicin)
 - Passed AABR hearing screening
 - In 2004, Idaho did not have risk monitoring program



Audiometry



Speech audiometry

SRT at 10 dBHL in each ear

Tympanograms

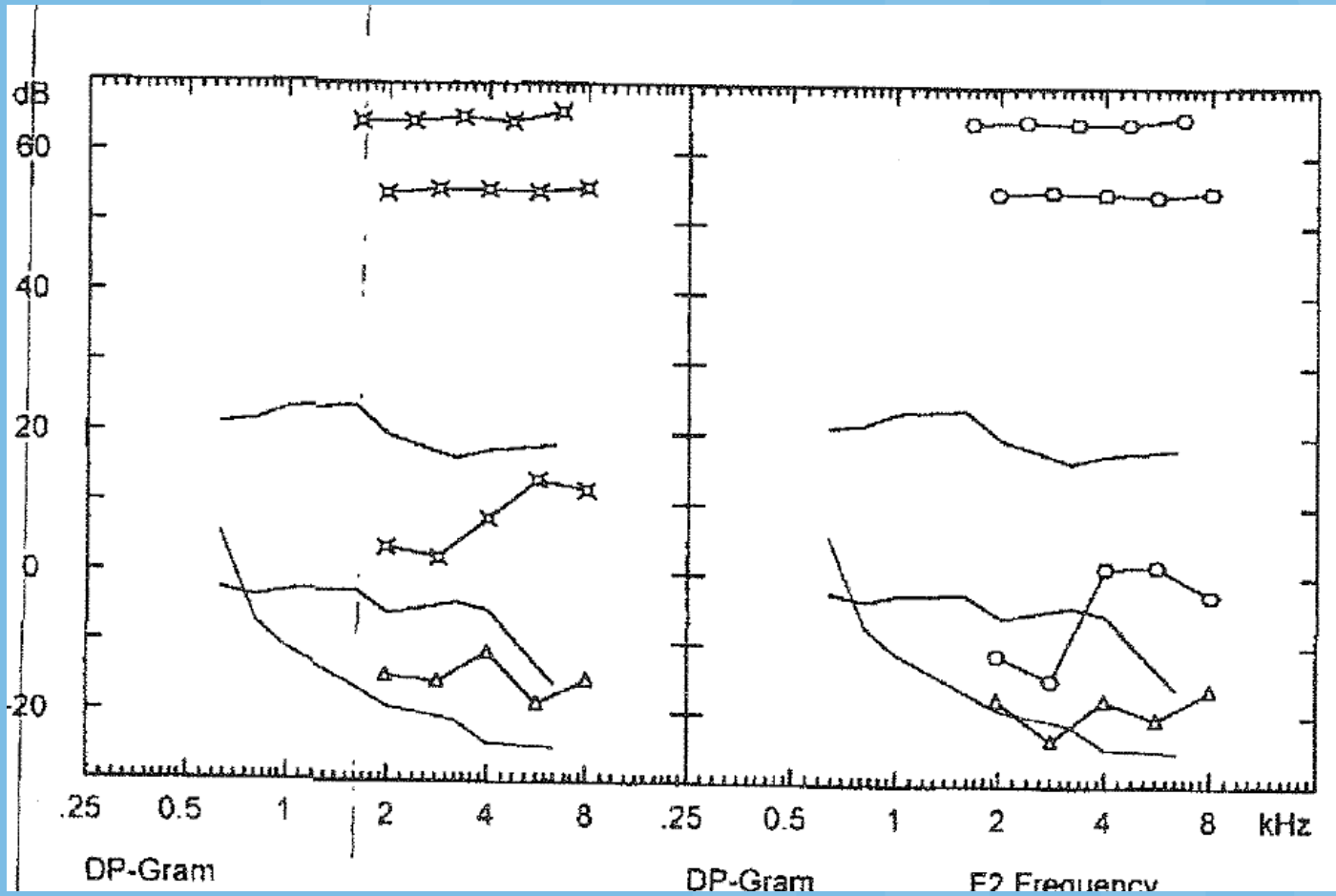
Type As bilaterally

Ipsilateral MEMR

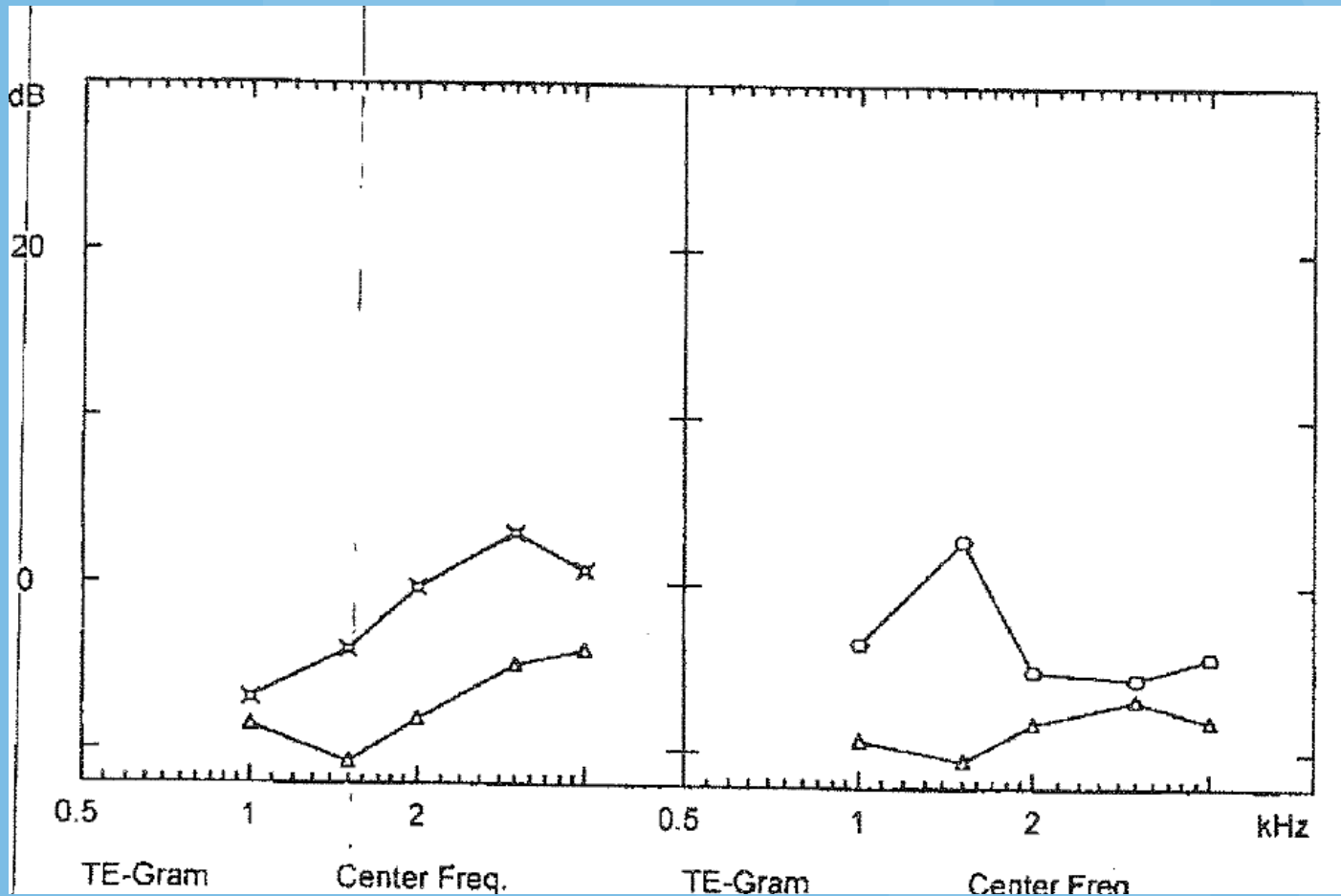
Absent bilaterally



DPOAE



TEOAE



3 years later...

- 8 years old
- Physician requested audiogram due to previous recommendations
- Mom has no significant hearing or speech concerns, but patient
 - Frequently asks for repetition
 - Listens to TV “very loud”



Audiometry

Speech Audiometry:

Word recognition @

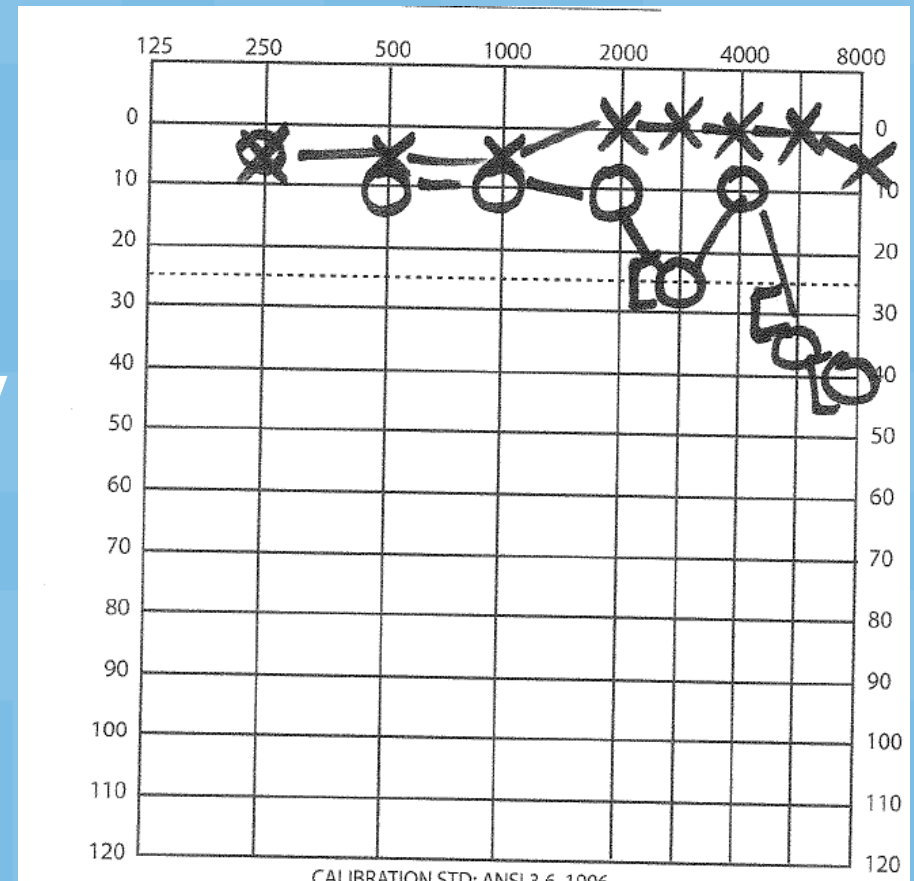
50 dBHL- 84% right, 100% left

SRT- 10 dBHL right, 0 dBHL left

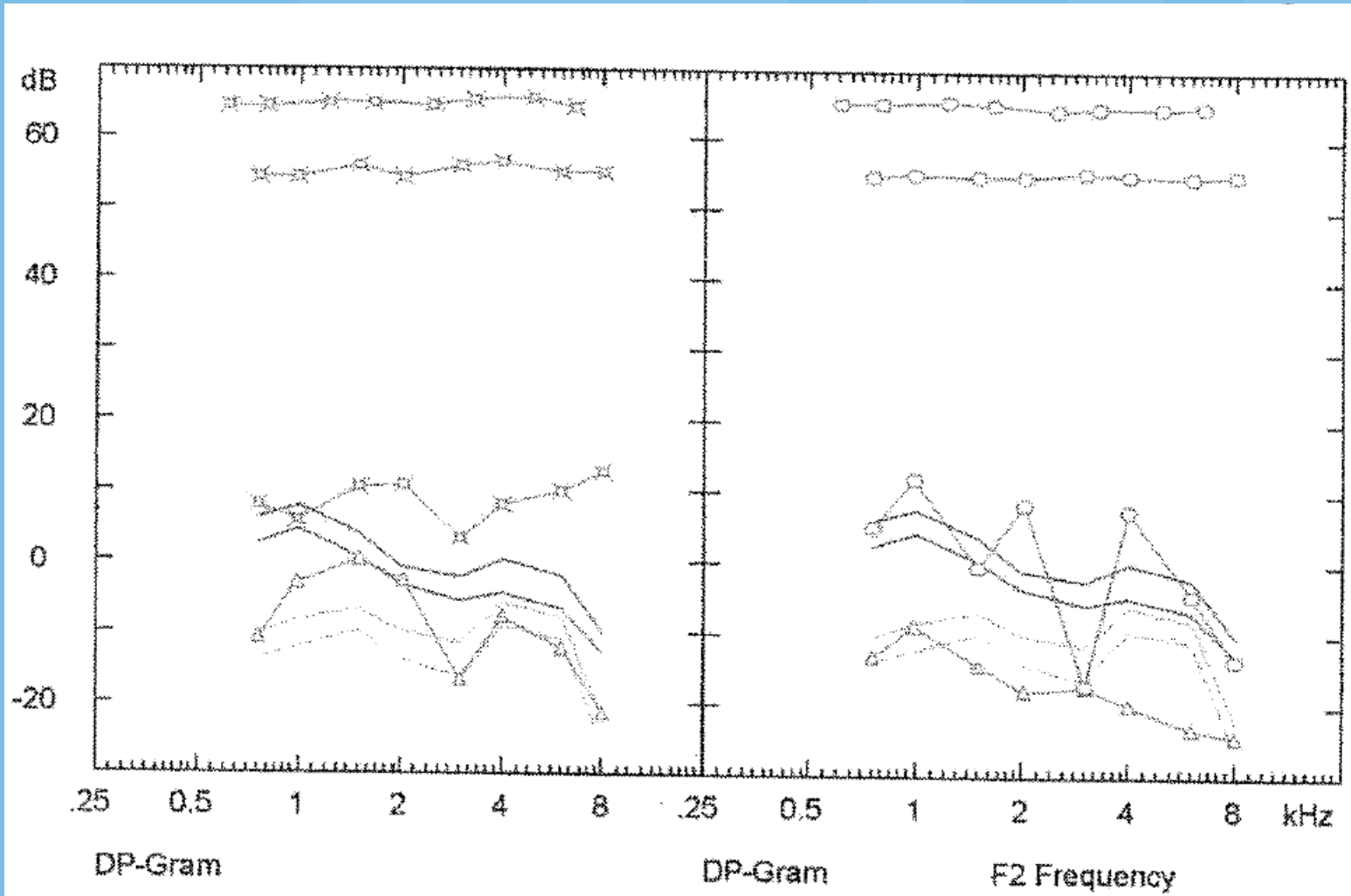
Tympanograms: Type A bilaterally

MEMR:

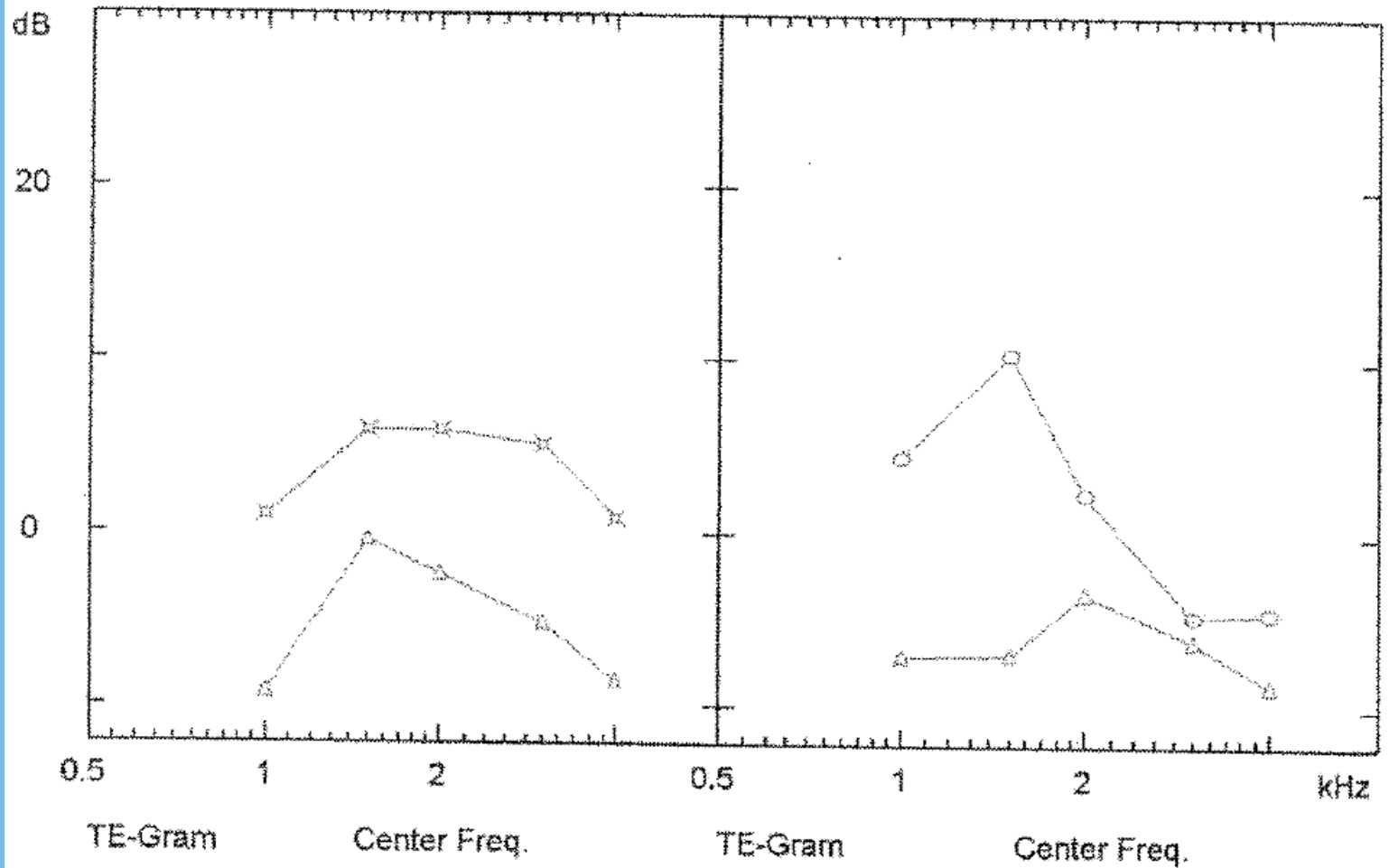
- Ipsilateral left/right present
- contralateral right present
- contralateral left absent



DPOAE



TEOAE



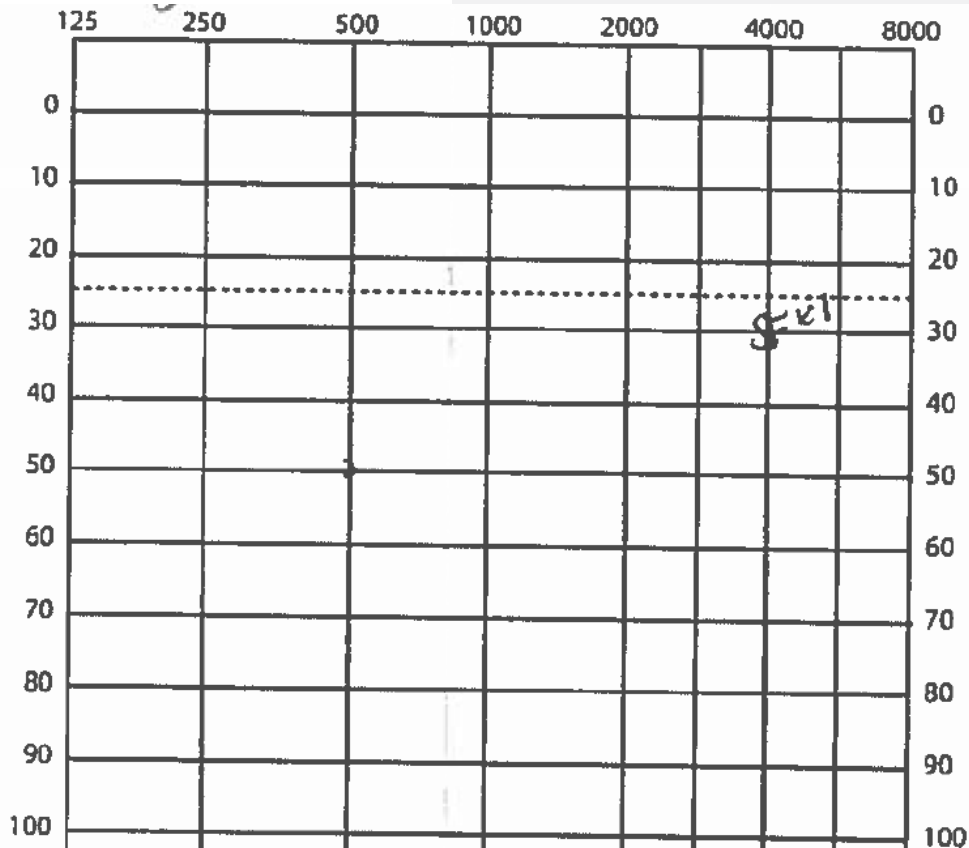
Case #2

- **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

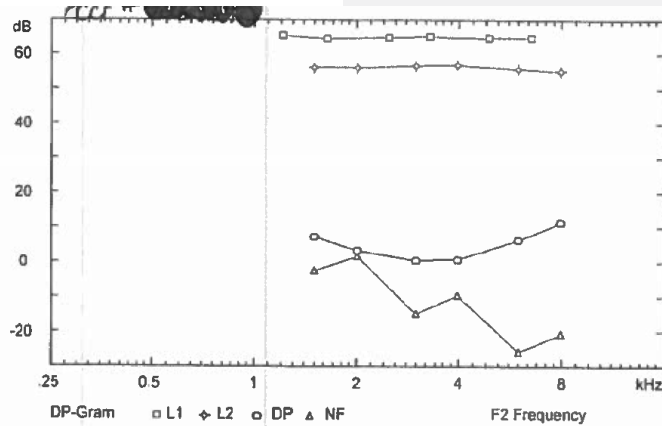


LEFT	KEY	RIGHT
X(□)	AIR CONDUCTION (MASKED)	O(△)
> (J)	BONE CONDUCTION (MASKED)	< (L)
SF	SOUND FIELD	SF
A	AIDED	A
CI	COCHLEAR IMPLANT	CI

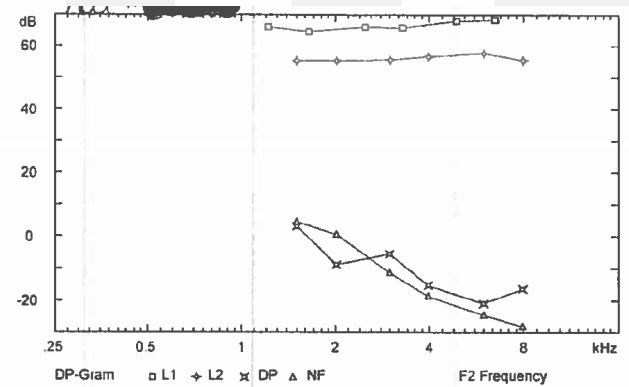
	PHONE RIGHT	PHONE LEFT	BINAURAL	UNAIDED	AIDED
PREDICTED SRT (PTA)	dB	dB	dB	dB	dB
OBTAINED SRT (SAT) (QUIET)	30 dB	40 dB	20 dB	dB	dB
MAXIMUM COMFORT LEVEL	dB	dB	dB	dB	dB
TOLERANCE THRESHOLD	dB	dB	dB	dB	dB
	dB	dB	dB	dB	dB
SDS ● _____ dB HTL QUIET	%	%	%	%	%
SDS ● _____ dB HTL QUIET	%	%	%	%	%
SDS					



OAE tracing



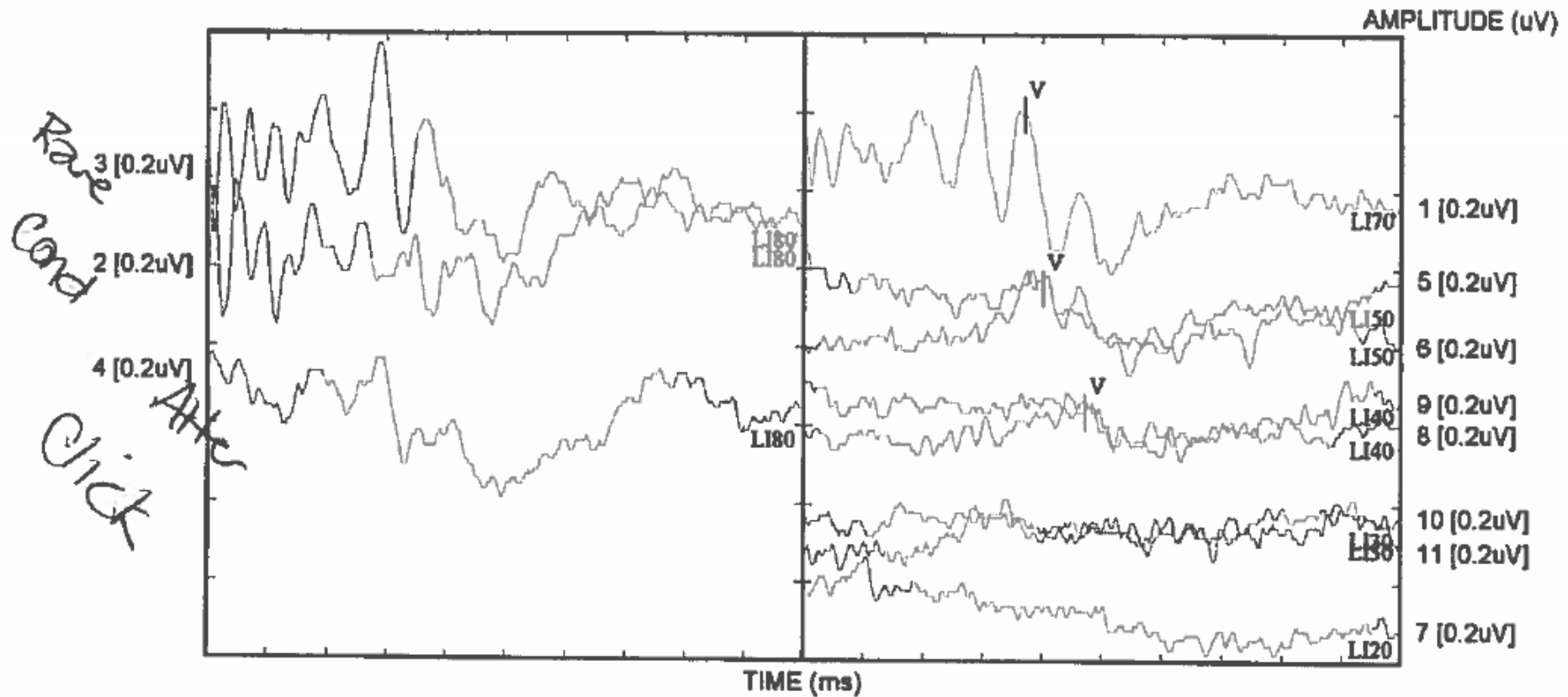
L1(dB)	L2(dB)	F1(Hz)	F2(Hz)	GM(Hz)	DP(dB)	NF(dB)	DP-NF(dB)
64.9	55.5	6516	7969	7206	11.5	-20.6	32.1
64.7	55.9	4922	6000	5434	6.4	-25.8	32.2
65.1	56.9	3281	3984	3616	0.7	-9.4	10.1
64.7	56.9	2484	3000	2730	0.5	-14.8	15.3
64.7	56.2	1641	2016	1818	3.2	1.6	1.6
65.3	56.2	1219	1500	1352	7.1	-2.7	9.8



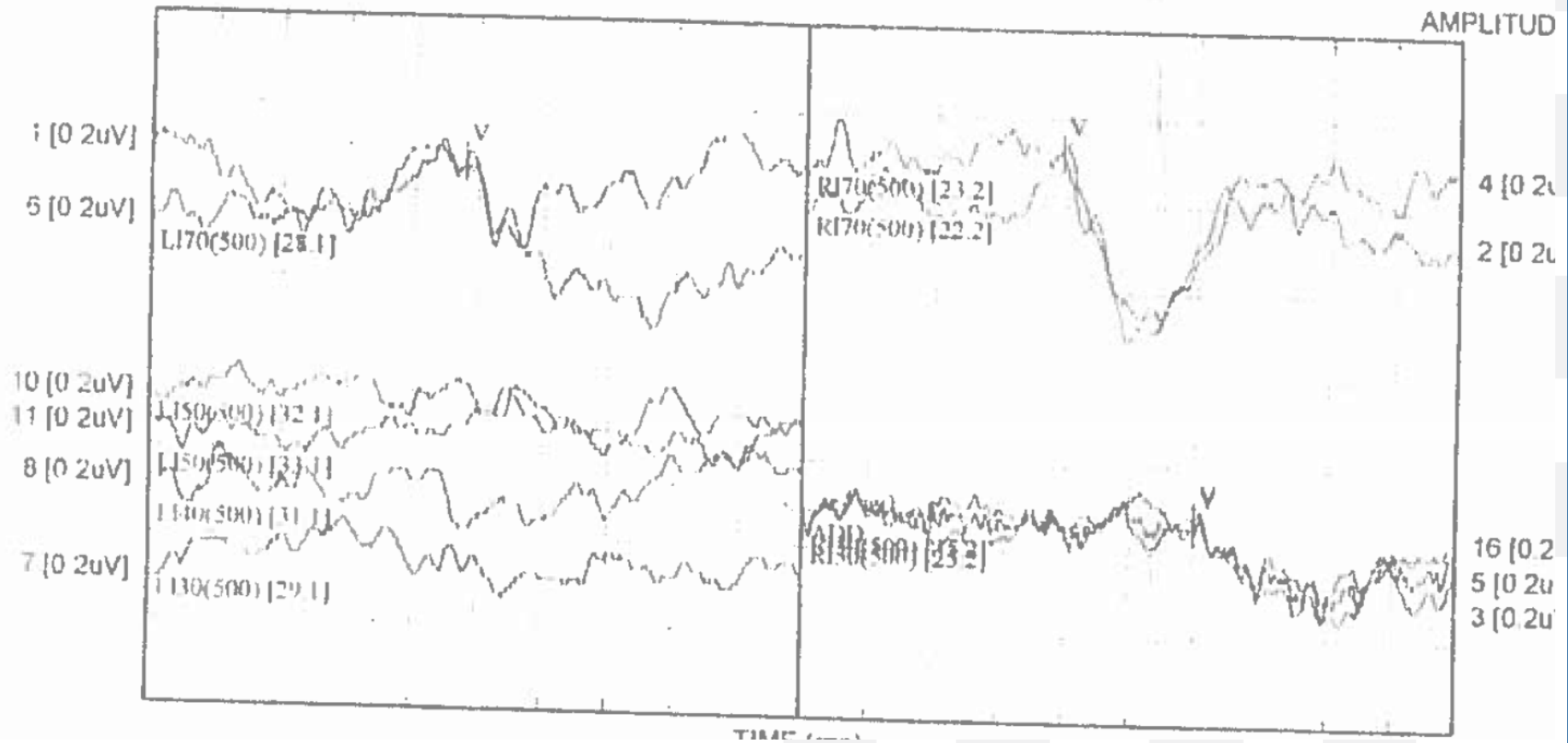
L1(dB)	L2(dB)	F1(Hz)	F2(Hz)	GM(Hz)	DP(dB)	NF(dB)	DP-NF(dB)
68.3	55.6	6516	7969	7206	-16.4	-28.1	11.7
68.0	57.8	4922	6000	5434	-20.9	-24.6	3.7
65.7	56.6	3281	3984	3616	-15.5	-18.8	3.3
66.1	55.7	2484	3000	2730	-5.7	-11.4	5.7
64.5	55.3	1641	2016	1818	-8.9	0.4	-9.3
65.9	55.3	1219	1500	1352	3.0	4.5	-1.5



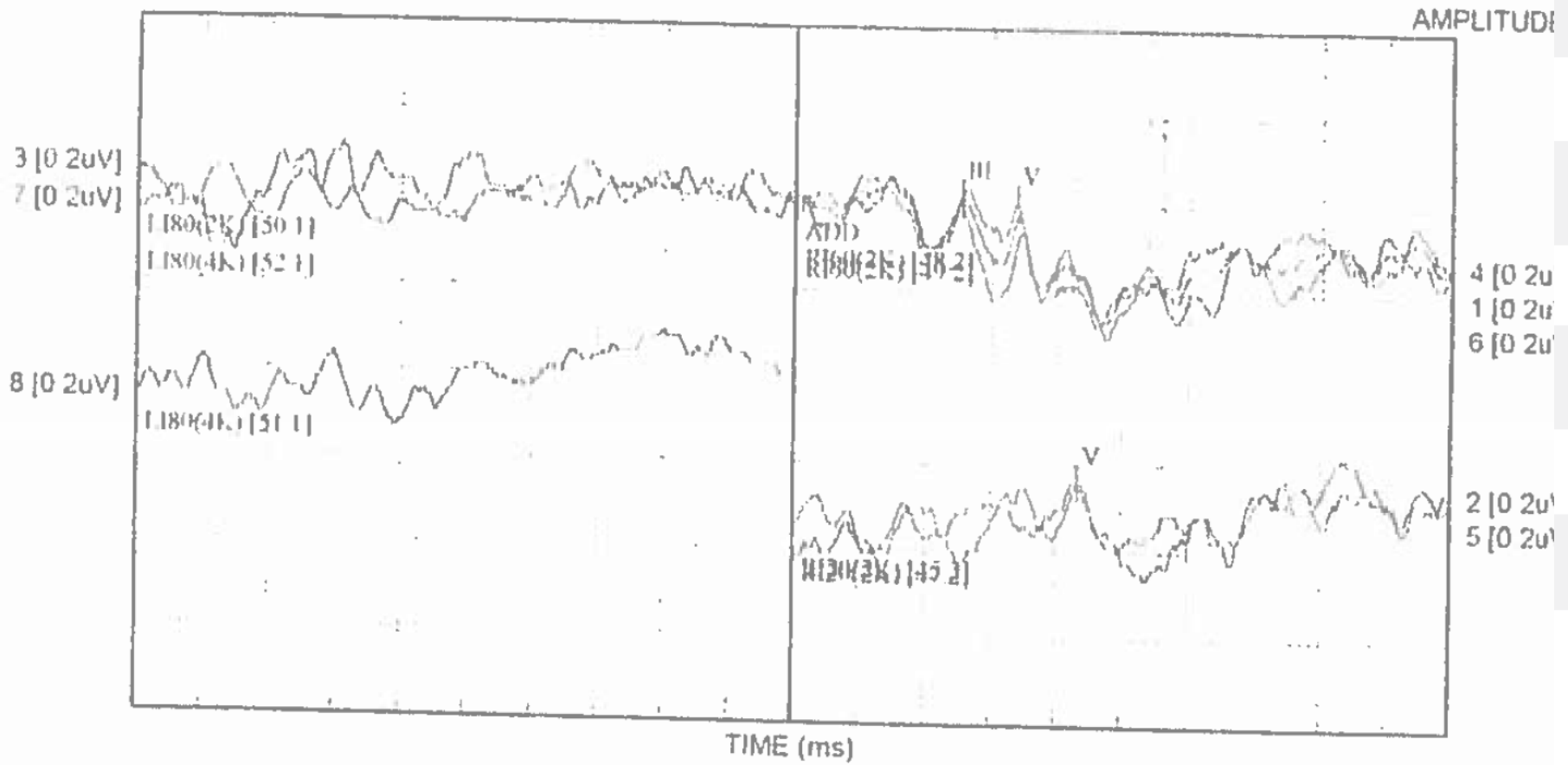
ABR evaluation 10 months old



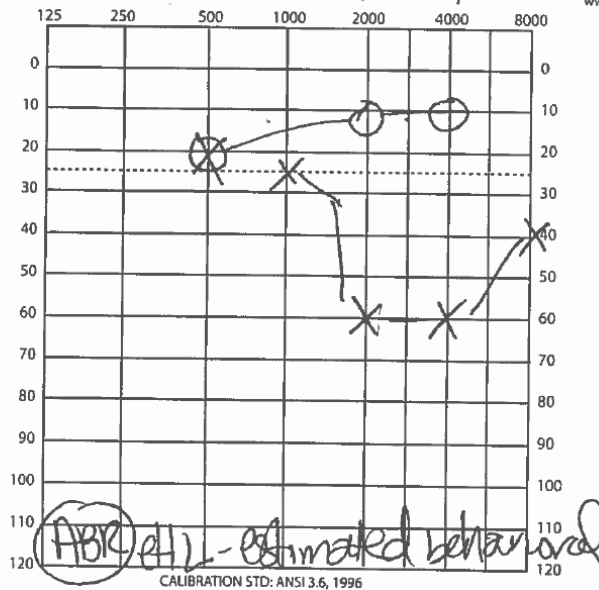
ABR tracing



ABR tracing



ABR eHL



LEFT	KEY	RIGHT
X (□)	AIR CONDUCTION (MASKED)	O (△)
> (□)	BONE CONDUCTION (MASKED)	< (□)
SF	SOUND FIELD	SF
A	AIDED	A
CI	COCHLEAR IMPLANT	CI

	PHONE RIGHT	PHONE LEFT	BROUHAUI	UNAIDED	AIDED
PREDICTED SRT (PTA)	dB	dB	dB	dB	dB
OBTAINED SRT/SAT (QUIET)	dB	dB	dB	dB	dB
MAXIMUM COMFORT LEVEL	dB	dB	dB	dB	dB
TOLERANCE THRESHOLD	dB	dB	dB	dB	dB
SDS	dB HTL QUIET	%	%	%	%
SDS	dB HTL QUIET	%	%	%	%
SDS	dB HTL QUIET	%	%	%	%

OAE Results		
TEOAE	Right	Left
DPOAE	Right	Left

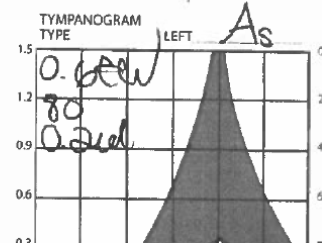
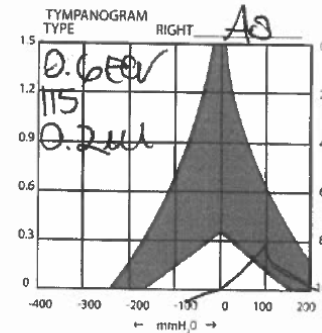
Handwritten: Absent Absent

ABR Click V Thresholds		
AIR	Right	Left
BONE	Right	Left

Handwritten: 40 40

ACOUSTIC REFLEXES		
Left Contralateral	HZ	Right Contralateral
_____	500	_____
_____	1000	_____
_____	2000	_____
_____	4000	_____
Right Ipsilateral	HZ	Left Ipsilateral
80	500	NR
_____	1000	NR
_____	2000	NR
_____	4000	NR

Handwritten: TS BB NR



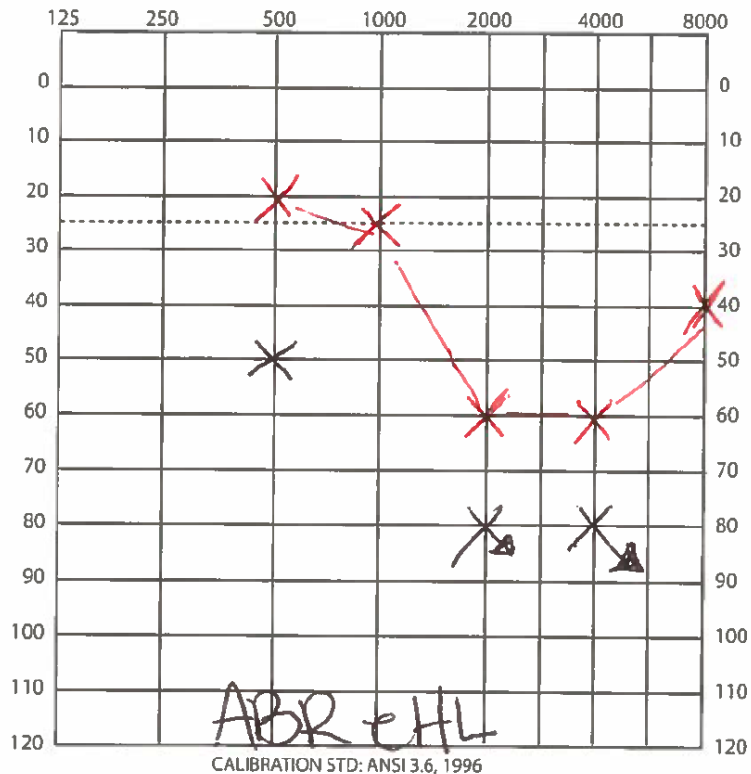
COMMENTS



10 months old, 18 months old

AUDIOLOGIST _____

Ontario 1182 SW 4th Ave. Ontario OR 97914 p. 541.881.0970
www.elkshearingandbalance.org

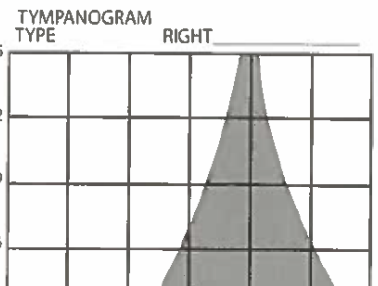


LEFT	KEY	RIGHT
X(□)	AIR CONDUCTION (MASKED)	O(△)
>(□)	BONE CONDUCTION (MASKED)	<(△)
SF	SOUND FIELD	SF
A	AIDED	A
CI	COCHLEAR IMPLANT	CI

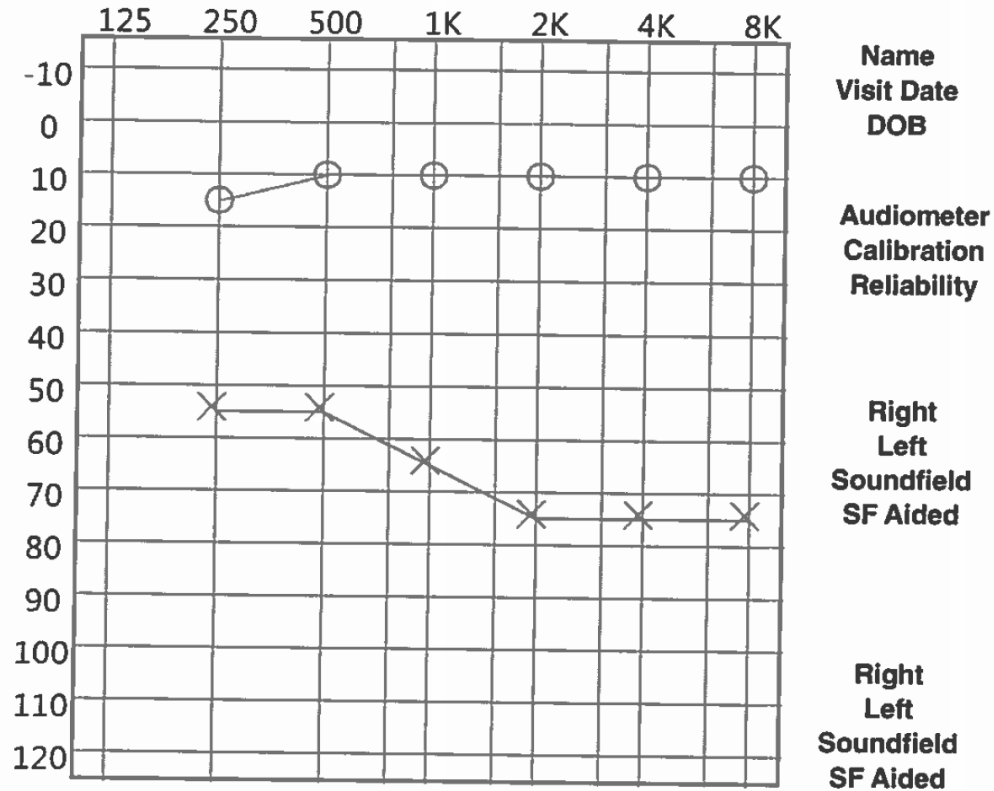
	PHONE RIGHT	PHONE LEFT	BINAURAL	UNAIDED	AIDED
PREDICTED SRT (PTA)	dB	dB	dB	dB	dB
OBTAINED SRT/SAT (QUIET)	dB	dB	dB	dB	dB
MAXIMUM COMFORT LEVEL	dB	dB	dB	dB	dB
TOLERANCE THRESHOLD	dB	dB	dB	dB	dB
	dB	dB	dB	dB	dB
SDS @ _____ dB HTL QUIET	%	%	%	%	%
SDS @ _____ dB HTL QUIET	%	%	%	%	%
SDS	%	%	%	%	%

OAE Results		
TEOAE	Right _____	Left _____
DPOAE	Right _____	Left _____

ACOUSTIC REFLEXES		
Left Contralateral	HZ	Right Contralateral
_____	500	_____
_____	1000	_____
_____	2000	_____



3 years old



Name
 Visit Date
 DOB

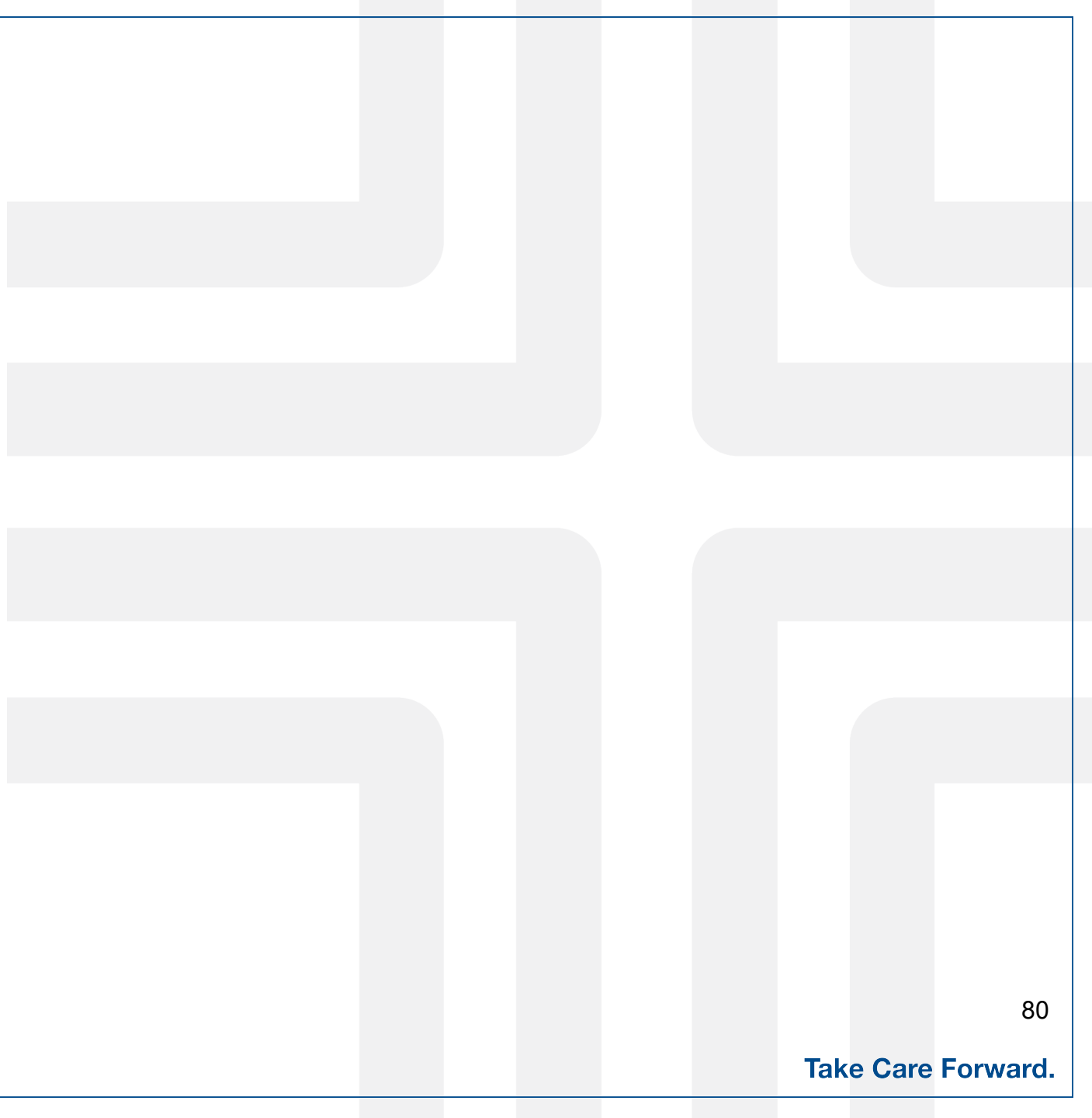
Audiometer
 Calibration
 Reliability

Right
 Left
 Soundfield
 SF Aided

Right
 Left
 Soundfield
 SF Aided

	R	L	B		R	L	B		R	L	B
AC Unmasked	○	×	⊂	MCL	M	M	M	No Response	↙	↘	↓





Questions and Answers



REFERENCES

- American Academy of Audiology Position Statement and Clinical Practice Guidelines: Ototoxicity Monitoring. October 2009.
- ASHA 2015. Ototoxic Medications.
- Antibiotics increase risk of hearing loss in patients with deadly bacterial infections. July 2015. Oregon Health Sciences University.
- Cone-Wesson et. al. (2000). Identification of neonatal hearing impairment: Infants with hearing impairment. *Ear and Hearing, 21, 488-507.*
- Fligor, B. (2008). Hearing outcomes in the most critically ill neonate population. *Audiology Today, 20 (5), 9-16.*
- <http://www.ncham.org>
- Hall (2007). New Handbook of Auditory Evoked Potentials.
- Hi-Track data from Idaho Sound Beginnings Program (2007-2015).
- Joint Committee on Infant Hearing (2000). Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Invention Programs.
- Joint Committee on Infant Hearing (2007). Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Invention Programs. *Pediatrics, 120, 898-921.*
- Kraft et. al. (2014). Risk indicators for Congenital and Delayed-onset hearing loss. *Otology and Neurootology.*
- *The NCHAM book, Chapter 10, Risk monitoring for Delayed-Onset hearing loss.*
- Van Riper & Kileny (2002). ABR hearing screening for high-risk infants. *Neonatal Intensive Care, 15, 47-54.*
- Zimmerman E, Lahav A (2012). Journal of Perinataology, Ototoxicity in preterm infants: effects of genetics, aminoglycosides, and loud environmental noise.

