



EHDI Coordinator Track - Risk Factors

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Services provided by St. Luke's



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We do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation

Learning Objectives

1. Participants will be able to identify Joint Commission on Infant Hearing Risk Factors from the 2019 Position Statement.
2. Participants will be able to identify Class A/B risk factors from the Idaho EHDI Risk Factor tool.
3. Participants will be able to educate parents on follow-up procedures for each Risk Factor for late on-set or progressive hearing loss.

Joint Committee on Infant Hearing (JCIH)

JCIH established in 1969

Members from:

American Academy of Pediatrics

American Academy of Ophthalmology and Otolaryngology

American Speech & Hearing Association

Position statements: 1972, 1982, 1990, 1994, 2000, 2007, 2019

JCIH 2019

Appendix

2: RISK INDICATORS FOR HEARING LOSS

Table 1
Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen

	Risk Factor Classification	Recommended Diagnostic Follow-up	Monitoring Frequency
	Perinatal		
1	Family history* of early, progressive, or delayed onset permanent childhood hearing loss	by 9 months	Based on etiology of family hearing loss and caregiver concern
2	Neonatal intensive care of more than 5 days	by 9 months	As per concerns of on-going surveillance of hearing skills and speech milestones
3	Hyperbilirubinemia with exchange transfusion regardless of length of stay	by 9 months	
4	Aminoglycoside administration for more than 5 days**	by 9 months	
5	Asphyxia or Hypoxic Ischemic Encephalopathy	by 9 months	
6	Extracorporeal membrane oxygenation (ECMO)*	No later than 3 months after occurrence	Every 12 months to school age or at shorter intervals based on concerns of parent or provider
7	In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis	by 9 months	As per concerns of on-going surveillance
	In utero infection with cytomegalovirus (CMV)*	No later than 3 months after occurrence	Every 12 months to age 3 or at shorter intervals based on parent/provider concerns
	Mother + Zika and infant with <i>no</i> laboratory evidence & no clinical findings	standard	As per AAP (2017) Periodicity schedule
	Mother + Zika and infant with laboratory evidence of Zika + clinical findings	AABR by 1 month	ABR by 4-6 months or VRA by 9 months
	Mother + Zika and infant with laboratory evidence of Zika - clinical findings	AABR by 1 month	ABR by 4-6 months Monitor as per AAP (2017) Periodicity schedule (Adebajo et al., 2017)
8	Certain birth conditions or findings: • Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia • Congenital microcephaly, congenital or acquired hydrocephalus • Temporal bone abnormalities	by 9 months	As per concerns of on-going surveillance of hearing skills and speech milestones
9	Over 400 syndromes have been identified with atypical hearing thresholds***. For more information, visit the Hereditary Hearing Loss website (Van Camp & Smith, 2016)	by 9 months	According to natural history of syndrome or concerns
	Perinatal or Postnatal		
10	Culture-positive infections associated with sensorineural hearing loss***, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis	No later than 3 months after occurrence	Every 12 months to school age or at shorter intervals based on concerns of parent or provider
11	Events associated with hearing loss: • Significant head trauma especially basal skull/temporal bone fractures • Chemotherapy	No later than 3 months after occurrence	According to findings and or continued concerns
12	Caregiver concern**** regarding hearing, speech, language, developmental delay and or developmental regression	Immediate referral	According to findings and or continued concerns

Note. AAP = American Academy of Pediatrics; ABR = auditory brainstem response; AABR = automated auditory brainstem response.


* Infants at increased risk of delayed onset or progressive hearing loss

**Infants with toxic levels or with a known genetic susceptibility remain at risk

***Syndromes (Van Camp & Smith, 2016)

****Parental/caregiver concern should always prompt further evaluation.

Idaho Screening Results Form



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: **Failed** ☐ **Risks** ☐ **Transfers*** ☐ **Missed** ☐ **Refused** ☐

Birth Hospital: _____

(*Transfers only) Receiving Hospital: _____

Send to: Idaho Sound Beginnings-EHDI, 450 W State St 5th Fl Boise, ID 83702 or Fax: (208) 332-7331

1. BABY'S INFORMATION:

Baby Vital Record #: _____

Baby's Name: _____
Last First

DOB: ____/____/____ Gender: ☐ M ☐ F

Nursery: ☐ Well Baby _____ Number of days in NICU/PICU _____

Baby's Primary Physician/Clinic: _____

Mother's name: _____

2. CONTACT INFORMATION:

Parent/Guardian: _____
Last First

Mailing Address: _____

City: _____ State: _____ Zip: _____

Main Phone: _____ Text? Yes / No

Alternate Phone/Contact: _____ Text? Yes / No

Email/other contact: _____

3. RISK ASSESSMENT (check all that apply)
FOR DELAYED—ONSET CHILDHOOD HEARING LOSS:

Class A Class B

☐ In utero Infections (CMV, Zika)
☐ Postnatal Infection (Meningitis)
☐ Syndrome associated with hearing loss
☐ Cleft Palate
☐ Anotia / Microtia /Atrisia
☐ Agensis of Corpus Callosum
☐ White forelock
☐ ECMO assisted ventilation
☐ Head trauma involving basal skull/temporal fracture

☐ Family History
☐ In-utero infection
☐ Ear pits or ear tags
☐ Cleft lip
☐ Mechanical ventilation
☐ NICU stay of 5 days or more
☐ Ototoxic exposure

4. HEARING SCREEN RESULTS:

First Screen: R ☐ Pass
☐ Failed (Refer)
☐ No Result
 Date L ☐ Pass
☐ Failed (Refer)
☐ No Result

Second Screen: R ☐ Pass
☐ Failed (Refer)
☐ No Result
 Date L ☐ Pass
☐ Failed (Refer)
☐ No Result

COVID-19 Tracking - Did the mother test positive for COVID-19? ☐ YES ☐ NO

Please call the audiology clinic to set up or confirm an appointment.

☐ Your baby **FAILED** the hearing screen and/or has Class A risk indicators. Audiological testing needs to be completed before baby is **3 months** old.

☐ Your baby is **AT RISK** for delayed-onset childhood hearing loss. Audiological monitoring at approximately **9 – 12 months** of age is recommended for most risk factors. A Pediatric Audiologist can advise on the appropriate monitoring schedule for your baby.

Your Follow-Up Appointment:

Clinic: _____

Phone: _____

Appt Date: _____ Time: _____

For a listing of Pediatric Audiologists visit www.EHDI-PALS.org

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, but risk factors are present (see above), hearing testing is recommended at approximately 3-12 months of age (Joint Committee on Infant Hearing, 2019). If you have any questions about testing, or need information on financial assistance, please contact Idaho's Early Hearing Program, Idaho Sound Beginnings (ph. 208-334-0829).



Idaho

Most frequently occurring risk factors	Least frequently occurring risk factors (<10%)
<p>Ototoxic Medications (>70%)</p> <p>Severe Asphyxia (>50%)</p> <p>Mechanical Ventilation less than 5 days (>25%)</p> <p>Low birth weight (>20%)</p> <p>Parental/Physician concerns (>15%)</p> <p>ECMO (>10%)</p>	<p>Hyperbilirubinemia</p> <p>Craniofacial anomalies</p> <p>Family history</p> <p>Congenital infections</p> <p>Bacterial meningitis</p> <p>Substance abuse (maternal)</p> <p>Neurodegenerative disorders</p>
(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, 1999; Van Riper & Kileny, 2002; Hall, 2007)



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
- * Hyperbilirubinemia requiring exchange transfusion

If baby passes the newborn hearing screening & has one or more CLASS A risk indicators =
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)

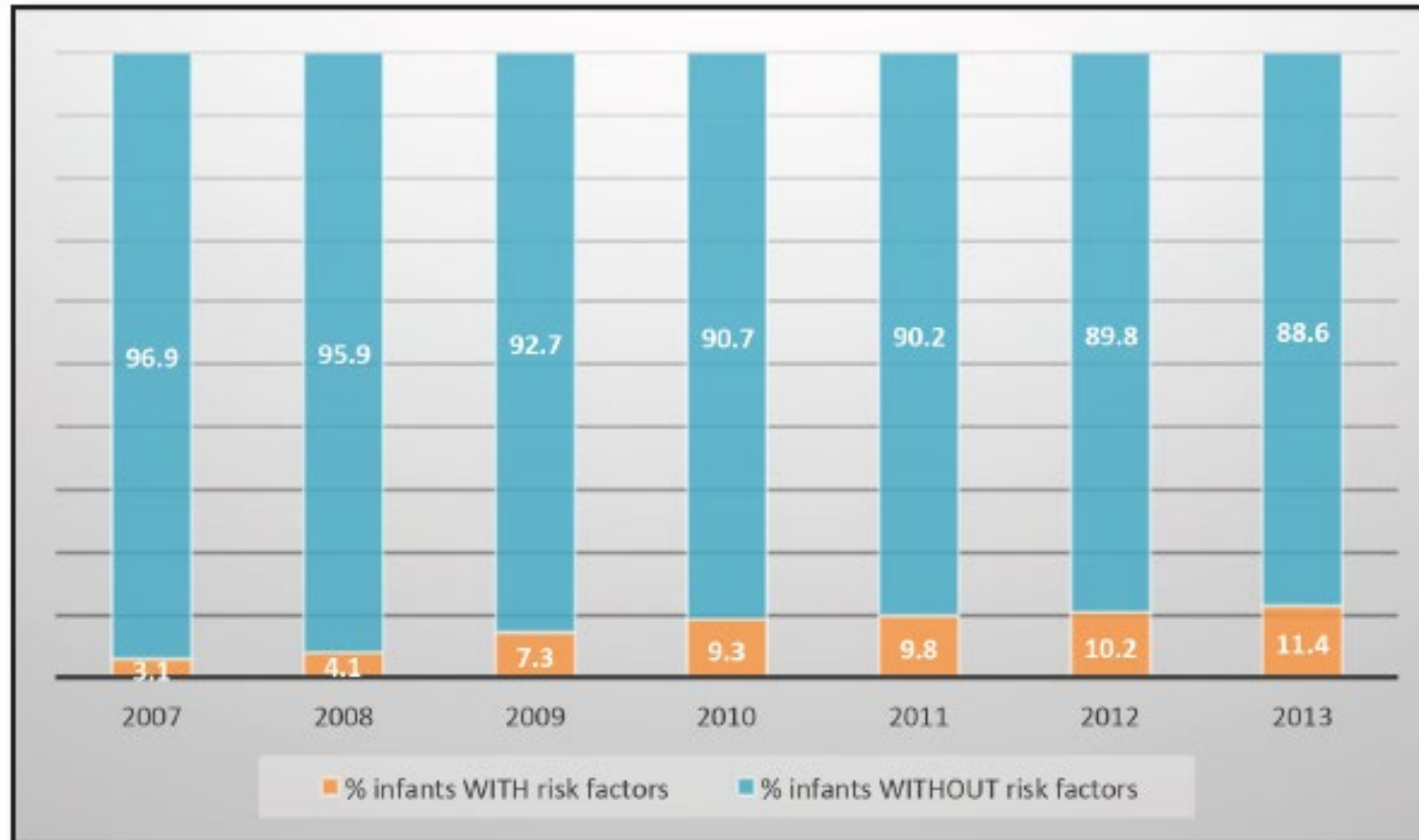
- * Any parental/caregiver hearing concerns warrants a referral to a pediatric audiologist.
- ** Infants readmitted to the hospital within the first 30 days of life should be re-screened if any risk indicators are present.

References:

Figon RI, Neuhoff MW, Mullen CH, Feldman HA, Jones DT. Factors associated with sensorineural hearing loss among survivors of extracorporeal membrane oxygenation therapy. *Pediatrics* 2005; 115(8):1019-1026.
Joint Committee on Infant Hearing. Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*. 2007; 120(4):998-1011. doi: 10.1542/peds.2007-2222.
Van Riper, Lori A.; Klenz, Paul R. ABR Hearing Screening for High-Risk Infants. *American Journal of Otolary*. 20(4):516-521, July 1999.



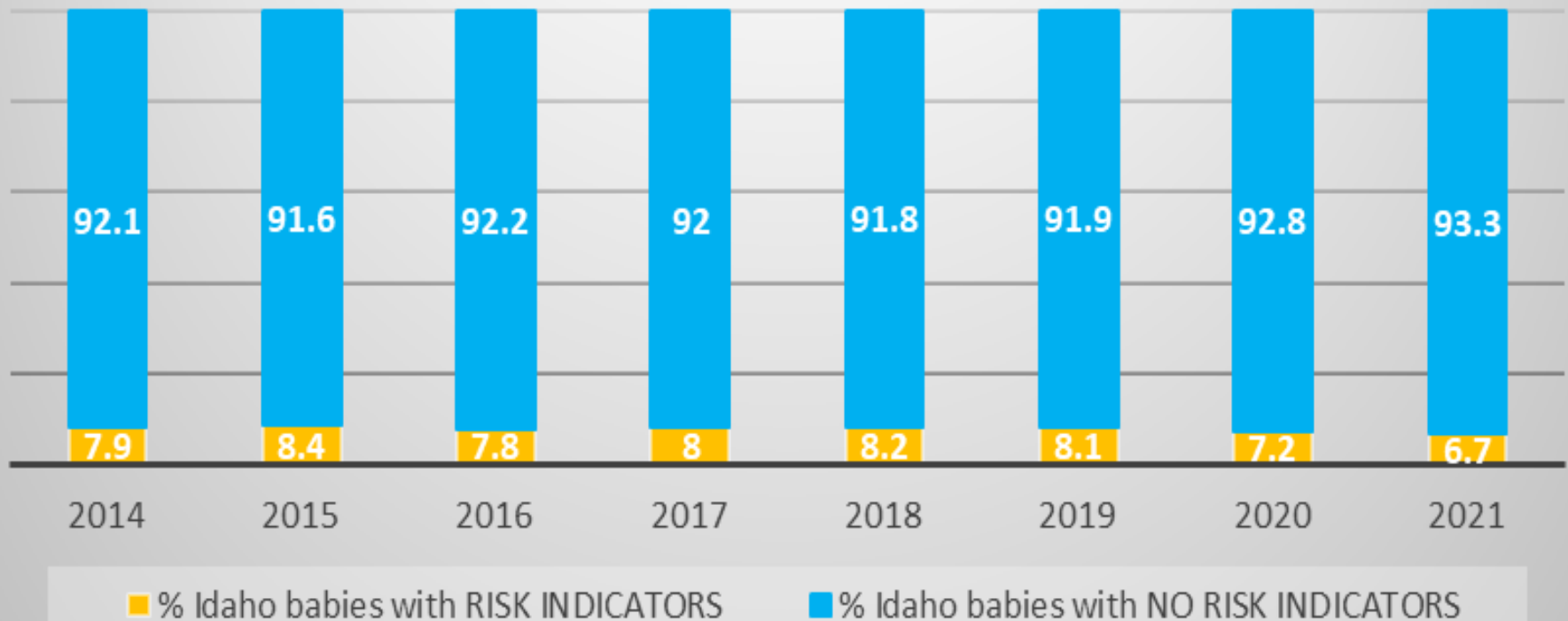
Prevalence of Infants with a Risk Indicator in ISB 2007-2013 Data



Stich-Hennen, J. R. & Bargen, G. A. (2017)



Prevalence of Idaho babies with risk indicators (2014-2021)





ISB Data

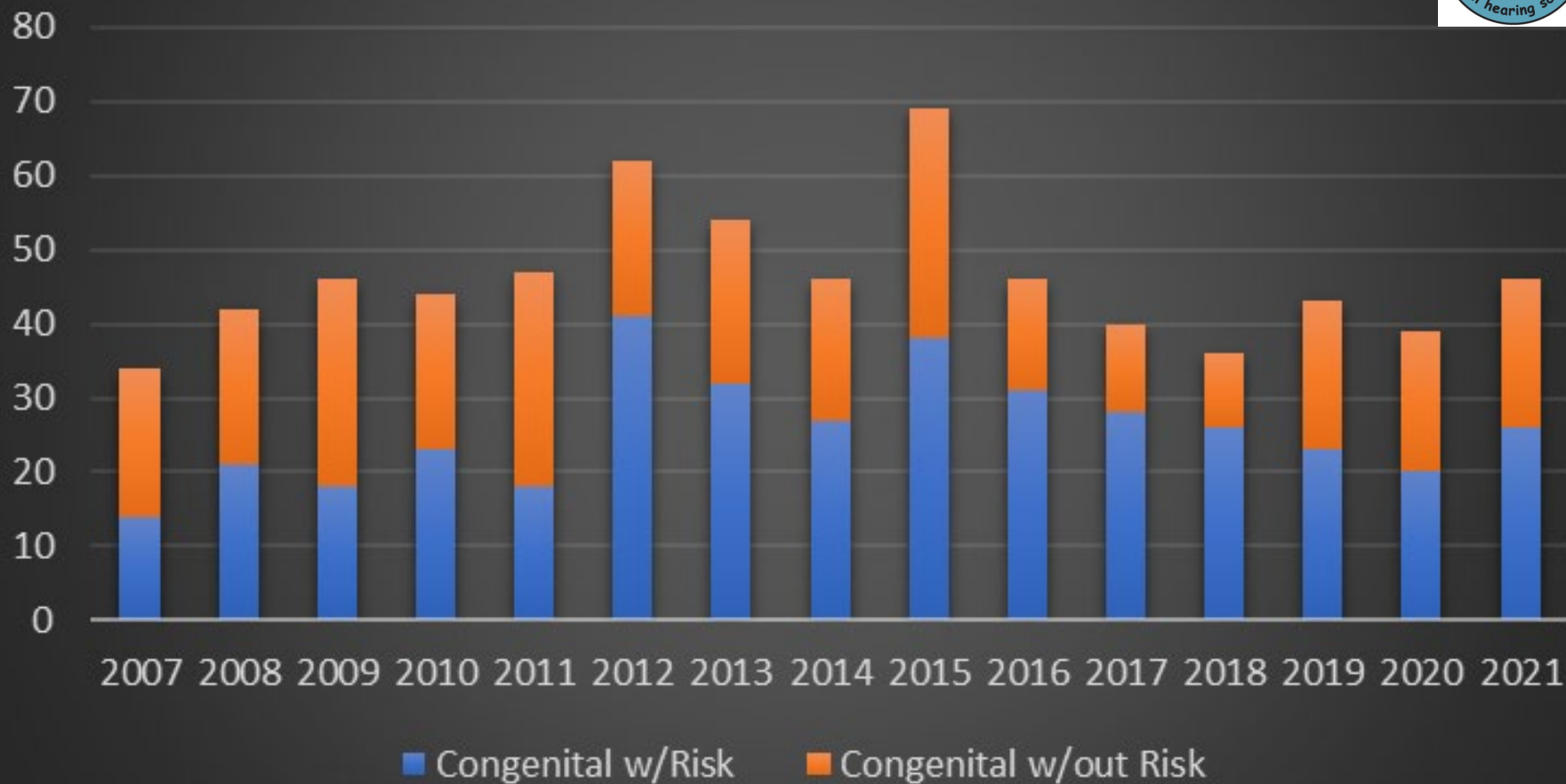
DOB
01/01/2014-
12/31/2021

Total births= 176,461

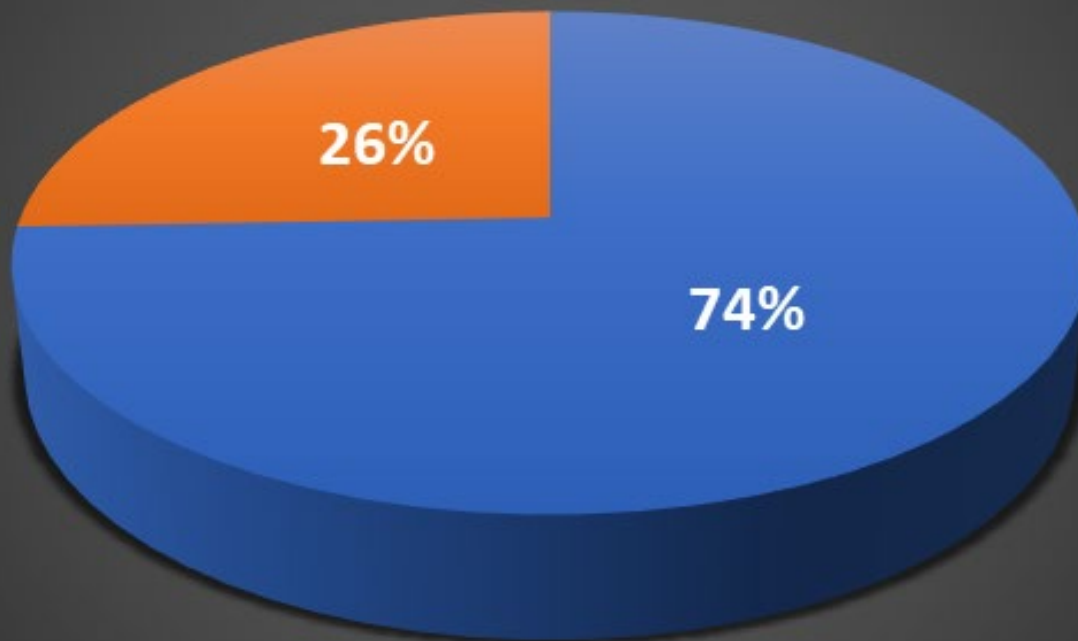
Congenital hearing
loss= 365 (2/1000)

Acquired/delayed
onset hearing loss= 86

Congenital hearing loss in Idaho



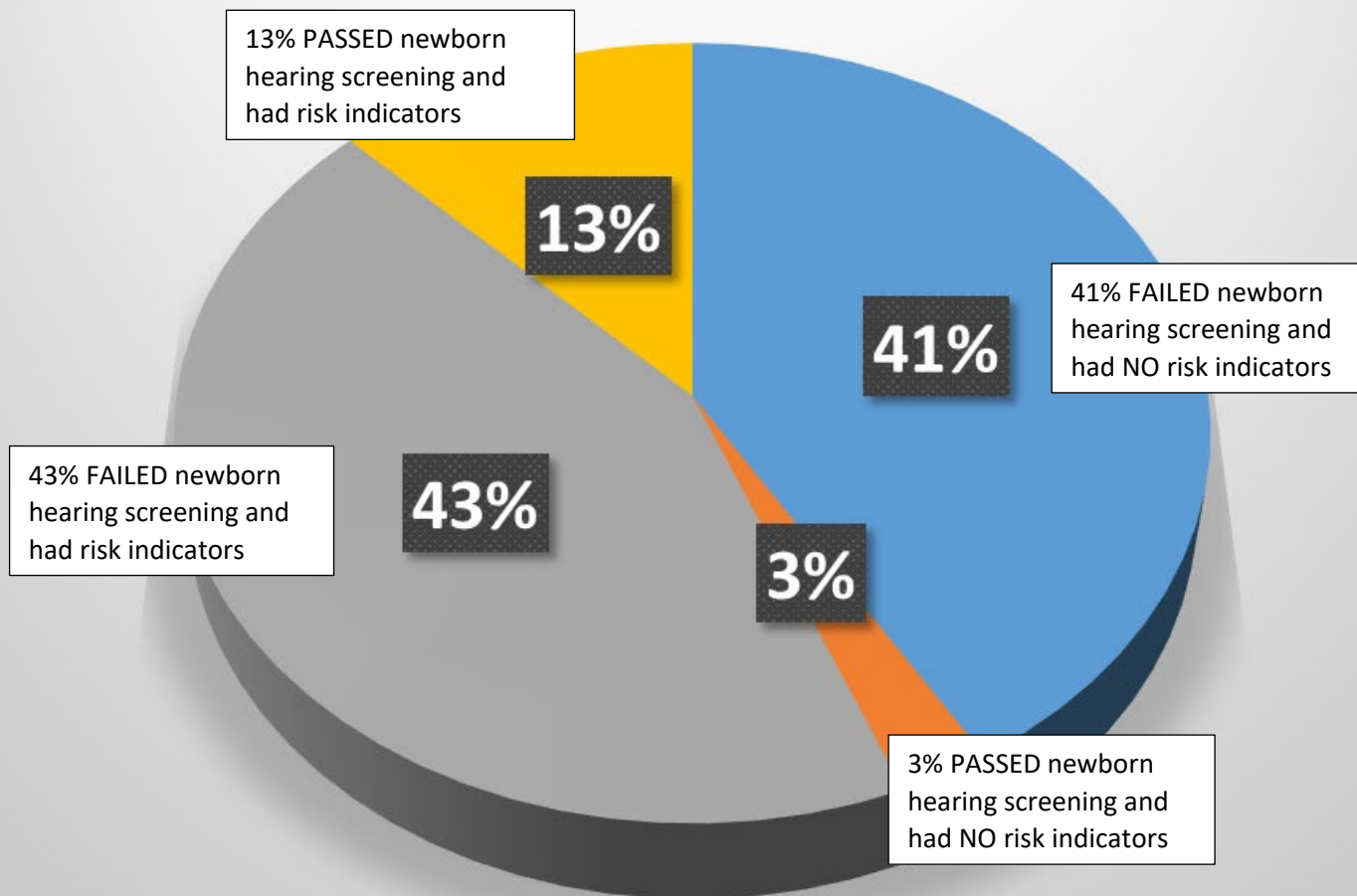
Acquired/delayed onset childhood hearing loss (Idaho 2014-2021)



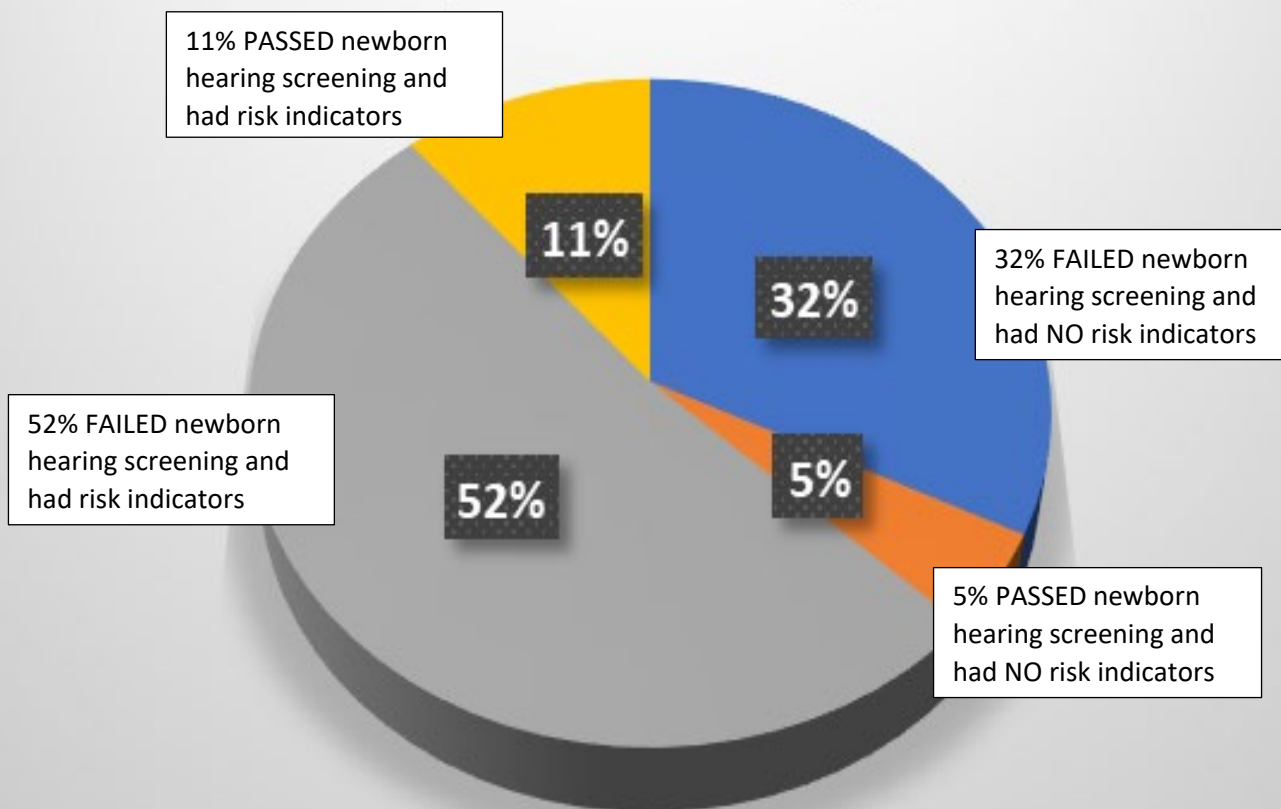
■ Acquired w/Risk

■ Acquired w/out Risk

Childhood hearing loss in Idaho (2007-2015)



Childhood hearing loss in Idaho (2016-2021)



REFERENCES

Cone-Wesson et al. (2000). Identification of neonatal hearing impairment: Infants with hearing impairment. *Ear and Hearing*, 21: 488-507.

Hi-Track data from Idaho Sound Beginnings Program (2007-2021).

Joint Committee on Infant Hearing (2007). Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*, 120: 898-921.

Joint Committee on Infant Hearing (2019). Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Journal of Early Hearing Detection and Intervention*, 4(2), 1-44. DOI: <https://doi.org/10.15142/fptk-b748>

Stich-Hennen, J. & Bargaen, G. (2017). *Implementing a Two-Class System for Monitoring Risk Factors for Delayed-Onset Hearing Loss*. The Journal of Early Hearing Detection and Intervention. 2017; 2(1): 55–63

Stich-Hennen, J. & Bargaen, G. (2015). Risk monitoring for delayed-onset hearing loss. In L. R. Schmeltz (Ed.) *The NCHAM book*, Chapter 10.



IDAHO DEPARTMENT OF HEALTH & WELFARE

CAMERON GILLILAND - Administrator
DIVISION OF FAMILY AND COMMUNITY SERVICES
450 West State Street, 5th Floor
P.O. Box 83700, Boise, ID 83720-0038
PHONE: 208-334-8100
FAX: 208-334-7100

Child's Name: Letter, Preview
Date of Birth: 9-12-2012
Hospital Letter Preview Hospital



4-1-2013

Dear Mr./Ms. Letter Contact:

Congratulations! We hope you are enjoying the exciting first year of your baby's life.

Your baby had their hearing screened shortly after birth. While your baby may have passed their screening there is a possibility that hearing loss could develop later on due to the risk factor(s) that were identified:

- Risk Indicator A, B, C

We recommend at least one diagnostic hearing test at approximately 9-12 months of age by a pediatric audiologist. Please talk to your baby's physician about obtaining a referral for a hearing evaluation.

If you need help finding a pediatric audiologist in your area, please visit www.EHDI-PALS.org or you can call us at (208) 334-0829 if you do not have internet access. We encourage you to make an appointment for your child as soon as possible. If you have any questions or concerns, please call (208) 334-0829 or email us at IdahoSoundBeginnings@dhw.idaho.gov.

Included is a brochure on Developmental Milestones, a free program to help you understand your child's development.

Warmest regards,

Brian Shakespeare
Idaho Sound Beginnings Program Coordinator

Andrea Amestoy, R.N.
Parent Outreach Consultant

Enclosure:

How to Prepare for Your Child's Hearing Test
Information on www.EHDI-PALS.org
Developmental Milestones Brochure

Idaho Sound Beginnings
Early Hearing Detection and
Intervention (EHDI)



Idaho - Parent Letter



BRAD LITTLE - GOVERNOR
DAVE JOHNSON - DIRECTOR

IDAHO DEPARTMENT OF HEALTH & WELFARE

CAMERON GILLILAND - Administrator
DIVISION OF FAMILY AND COMMUNITY SERVICES
450 West State Street, 5th Floor
P.O. Box 87750, Boise, ID 83725-0000
PHONE: 208-334-8700
FAX: 208-332-7330

Dear Preview-Doc,

Letter, Preview, Date of Birth: 9-12-2012 had a hearing screening at Letter Preview Hospital.

**Has Risk Indicators for Delayed or Late On-set Hearing Loss
Risk Indicator A, B, C**

A diagnostic hearing evaluation with a pediatric audiologist is strongly recommended prior to twelve (12) months of age. For a list of pediatric audiologists please visit www.EHDI.PALS.org, or you can contact Idaho Sound Beginnings at 208-334-0829. If your patient needs financial assistance, the parent of your patient can contact the Infant Toddler Program at 208-334-6530 prior to scheduling the appointment. Financial assistance through the Infant Toddler Program is not retroactive.

Please check a box and sign the form.

- ☐ I will speak/have spoken to the parent/guardian of this patient about scheduling a hearing evaluation.
- ☐ This is no longer my patient. The new physician is _____.

Physician Signature

Physician Printed Name

Date

Please return this form to Idaho Sound Beginnings by fax to 208-332-7331 or by secure email to IdahoSoundBeginnings@dhw.idaho.gov

Thank you,

Brian Shakespeare
Public Health Coordinator, Early Hearing Detection and Intervention

Idaho Sound Beginnings
Early Hearing Detection and
Intervention (EHDI)



Idaho - Physician Letter

Alabama's Listening!
UNIVERSAL NEWBORN HEARING SCREENING

**ALABAMA
PUBLIC
HEALTH**

**UNIVERSAL NEWBORN HEARING
SCREENING DEVELOPMENTAL
MILESTONES IN HEARING**

**USE THIS GUIDE TO SEE HOW YOUR
CHILD IS GROWING AND LEARNING**

Birth to three months:

- Jumps or blinks to loud sounds
- Wakes up to loud sounds
- Quiets when he or she hears mom's voice

Three months to six months:

- Turns eyes or head to search for the sound source
- Responds to your voice even when you cannot be seen
- Enjoys toys that make sounds
- Starts babbling

At six months:

- Responds to his or her name
- Turns head to the direction of the sound source
- Begins to imitate speech sounds

At ten to twelve months:

- Understands and follows simple directions
- Gives a block or toy to you when asked for it without pointing
- Imitates speech sounds of others

At thirteen to eighteen months:

- Follows simple one step directions
- Uses 3-20 single words
- Points to 1-3 body parts when asked

At nineteen to twenty-four months:

- Understands approximately 300 words
- Puts two words together ("eat cookie") by 24 months of age
- Points to five body parts
- Responds to "yes" or "no" questions

Alabama – Developmental Milestones

RISK FACTORS FOR DELAYED OR LATE-ONSET HEARING LOSS

Universal Newborn Hearing Screening, Diagnosis, and Intervention

Joint Committee on Infant Hearing 2019 Guidelines

Follow up recommended immediately if:

1. Caregiver concerns regarding hearing, speech, language, and developmental delays.

Follow up recommended by 3 months of age if:

1. Infections while pregnant: CMV (cytomegalovirus).
2. Extracorporeal membrane oxygenation (ECMO).
3. Serious head injury that required hospitalization.
4. Chemotherapy.
5. Culture-positive infections (bacterial and viral) like meningitis, encephalitis, chicken pox.

Follow up recommended by 9 months of age if:

1. Family history of hearing loss.
2. Baby admitted to the Neonatal Intensive Care Unit (NICU) for more than 5 days.
3. Exposure to infections like herpes, rubella, syphilis, toxoplasmosis, zika, and meningitis.
4. Hyperbilirubinemia with exchange transfusion.
5. Craniofacial abnormalities, microcephaly, hydrocephalus, and temporal bone abnormalities.
6. Asphyxia or Hypoxic Ischemic Encephalopathy.
7. Administration of certain IV (intravenous) medications: aminoglycoside, antibiotics, for greater than 5 days.
8. Any of the over 400 syndromes like: cleft palate, Usher syndrome and many more.

Alabama – Risk Factors

Alabama – Risk Factor Letter

<<DATE>>

<<mfnm>> <<mlnm>>
<<str1>>
<<cty>>, <<state>> <<zip>>

DOB: <<brthdt>>
Patient ID: <<intlbn>>

Dear Parent or Guardian:

Please refer to the newborn hearing screening risk factor card enclosed. A hearing risk factor does not mean a child will develop hearing loss, but it is recommended your child have additional hearing evaluation to ensure normal hearing and language development.

Notify your child's doctor if you have concerns regarding your child's hearing, speech, or language development. Please call 1-866-828-6755 if you have any questions or concerns about newborn hearing screening, or you may visit the newborn hearing website at www.alabamapublichealth/newborn-hearing-screening.

Sincerely,



Mary Ellen Whigham, R.N.
Alabama Early Hearing Detection and
Intervention Coordinator
Alabama Newborn Screening Program

Enclosure

Virginia EHDI- Information System (VISITS)

Identifying Children at Risk



Your Baby's Hearing Results

Congratulations on the birth of your baby! The facility where your baby was born conducted a newborn hearing screening.

Even though your baby passed the hearing screening, they are **AT RISK for developing late onset hearing loss.**

Risk indicator(s):
Schedule a diagnostic hearing evaluation with a pediatric audiologist **between 12 to 24 months of age.**

Language development begins at birth. It is recommended that you **monitor milestones** in your baby's hearing and speech development throughout the first year. If you have concerns please contact your primary care provider to discuss getting tested sooner.

Milestones to Monitor

<p>Birth to 3 months old:</p> <ul style="list-style-type: none"> ▶ Blinks or jumps when there is a sudden, loud sound ▶ Quiets or smiles when spoken to ▶ Makes sounds like "ooh" or "ahh" 	<p>3 to 6 Months Old:</p> <ul style="list-style-type: none"> ▶ Looks for sound with eyes ▶ Recognizes parent's voice ▶ Uses many sounds, squeals and chuckles
<p>6 to 9 Months Old:</p> <ul style="list-style-type: none"> ▶ Turns head toward sounds ▶ Understands "no-no" or "bye-bye" ▶ Makes babbling sounds ("baba," "mama," "gaga") 	<p>9 to 12 Months Old:</p> <ul style="list-style-type: none"> ▶ Repeats simple words and sounds ▶ Correctly uses "mama" or "dada" ▶ Responds to singing or music ▶ Points to favorite toys and objects when asked

EHDI VIRGINIA Virginia Early Hearing Detection & Intervention Program
VA_EHDI@vdh.virginia.gov

Birthing
Centers

Hospitals



Audiology
Facilities

National EHDI Conference Risk Factors 2023

Who has access to view Risk Indicators using EHDI-IS



Hospital
Staff

Audiologist

PCP's

EHDI

Children at Risk

Year	Number of Births	Number of Risk Factors Documented	Percent Born with Risk Factors
2017	99,444	7,512	7.55%
2018	99,116	6,782	6.84%
2019	97,400	6,554	6.73%
2020	94,846	5,878	6.20%
2021	96,120	5,849	6.09%

	2017	2018	2019	2020	2021
NICU	5,461	5,122	5,077	4,624	4,730
Exposure to Ototoxic Medication	2,941	2,495	2,610	2,465	2,277
ECMO	1,138	1,305	1,360	1,274	1,289
Family History	930	834	655	544	471
In Utero Infection	89	119	107	96	71
Craniofacial	65	95	81	83	71
Postnatal	20	18	24	12	7
Stigmata	22	18	19	16	16
Hyperbilirubinemia	13	23	20	17	18

Communicating Risk



ROBO CALLS



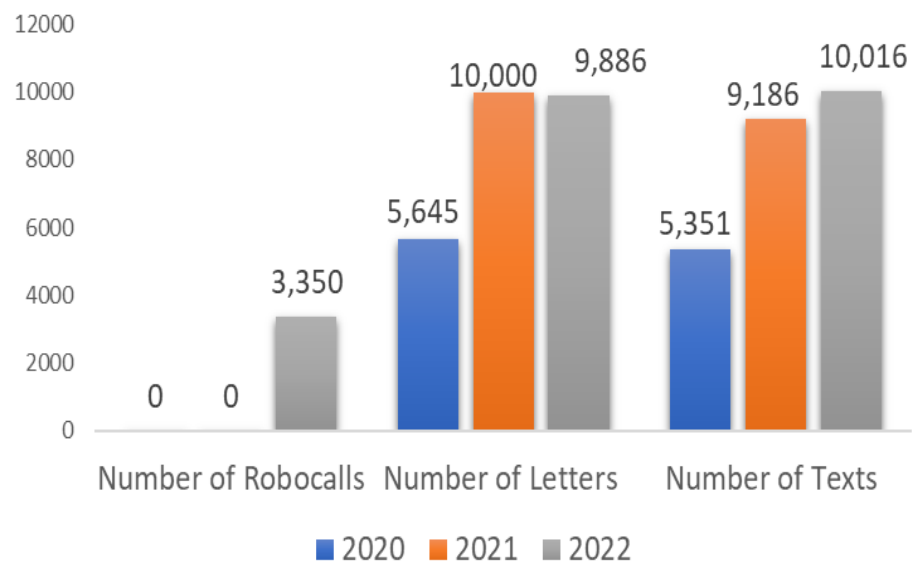
TEXT



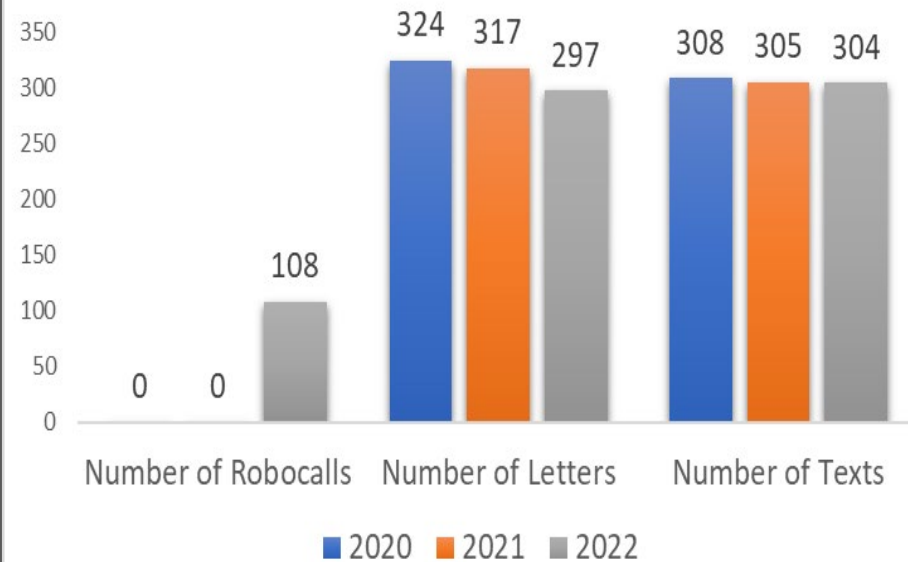
LETTERS

Follow-up for Children at Risk

Children Pass with Risk



Children Fail with Risk



Discussion Questions

How many of you currently track risk factors in your states?

Discussion Questions

What challenges are you experiencing
regarding risk factor
tracking/monitoring?

Discussion Questions

What has worked in your state to track risk factors?

Discussion Questions

Does your state experience push back from physicians or other professionals?

Discussion Questions

What are your communication methods of risk factors to parents, physicians, other stack holders?

Discussion Questions

What is something you could see implementing in your state next Tuesday, next month, next year to improve your state's risk factor monitoring?