Risk Factors for Late-onset or Progressive Hearing Loss:

Newborn Screening is just the beginning, not the end

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Conflict of Interest

- I have no conflicts of interest to report

Learning Objectives

- Identify risk factors for late onset or progressive hearing loss
- With the goal of having teams consider this information when developing strategies to identify and track children with risk factors within primary care

2013 National CDC EHDI data

Total Births	**3,904,742**	**%**
Number screened	**3,478,986**	**97**
Not passing	**61,419**	**1.6**
Diagnosed	**31,318**	**58.7**
Normal hearing	**26,022**	**48.8**
Hearing Loss	**5,296**	**9.9**
Rate of hearing loss	**1.5/1000**	
Referred to EI	**4,817**	**87.4**
Risk Factors	**NO CONSISTENT POPULATION DATA**	

http://www.cdc.gov/ncbddd/hearingloss/2013-data/2013_ehdi_hsfs_summary_a.pdf

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Risk Factors for SNHL Martinez-Cruz et al Arch Med Res 2008

Risk Factor	SNHL(146) Yes/No	Control (272) Yes/No	OR	95% CI		*p*
Prenatal steroids	20/126	77/195	0.377	0.194	0.732	<0.001
Surfactant	27/119	110/162	0.331	0.176	0.579	<0.001
Asphyxia	41/105	67/205	1.166	0.738	1.841	0.55
Exchange	40/106	12/260	8.176	4.128	16.195	<0.000
Meningitis	15/131	7/265	4.368	1.738	10.976	<0.002
Amikacin	134/12	229/43	2.097	1.068	4.116	<0.033
Furosemide	114/32	131/141	3.834	2.424	6.066	<0.000
Persistent fetal circulation	12/134	14/258	1.650	0.742	3.668	0.288
IVH	71/75	32/240	7.100	4.344	11.605	<0.000
BPD	75/71	50/222	4.690	3.001	7.331	<0.000

Risk Indicators for permanent congenital, delayed onset or progressive hearing loss *noted at birth*

- Family history*
  - of permanent childhood hearing loss

- NICU stay > 5 days or any of following (regardless of length of stay):
  - ECMO assisted ventilation*
  - Ototoxic medications (such as gentamycin, tobramycin)
  - Loop diuretics (furosemide, Lasix)
  - Hyperbilirubinemia requiring exchange transfusion

JCIH, 2007

* = greater risk for **delayed** onset HL

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Risk Indicators for permanent congenital, delayed onset or progressive hearing loss

noted at birth

- In Utero infections
  - CMV*, herpes, rubella, syphilis, toxoplasmosis, Zika

- Craniofacial anomalies
  - especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
  - Children with cleft lip and palate are at higher risk for conductive hearing loss requiring monitoring and aggressive treatment of middle ear status

JCIH, 2007

* = greater risk for **delayed** onset HL

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Risk Indicators for permanent congenital, delayed onset or progressive hearing loss

- Physical findings associated with a syndrome known to include permanent HL (e.g. white forelock)

- Syndromes* involving hearing loss (such as)
  - Down Syndrome,
  - Neurofibromatosis,
  - Osteopetrosis,
  - Usher Syndrome,
  - Waardenburg Syndrome
  - Alport Syndrome
  - Pendred Syndrome
  - Jervell & Lange-Nielson

* = greater risk for **delayed** onset HL

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Risk Indicators for permanent congenital, delayed onset or progressive hearing *post-natal*

- Neurodegenerative disorders
  - Hunter syndrome
  - Sensory motor neuropathies (Frieidrich ataxia, Charcot-Marie-Tooth)
- Culture positive postnatal infections associated with HL*
  - Herpes, varicella, meningitis
- Head trauma (basal skull, temporal bone)*
- Chemotherapy*

* = greater risk for **delayed** onset HL

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Current JCIH monitoring interval statements

- The timing and number of hearing reevaluations for children with risk factors should be **customized and individualized** depending on the **relative likelihood** of a subsequent delayed-onset hearing loss.

- Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age.

Current JCIH monitoring interval statements

- **Early and more frequent assessment** may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture positive postnatal infections associated with sensorineural hearing loss; for children who have received extracorporeal membrane oxygenation (ECMO) or chemotherapy;

- **And** when there is caregiver concern or a family history of hearing loss.

Follow-up of Infant with Perinatal Risk Factor for Hearing Loss

Risk Factor	JCIH 2007	JCIH 2015	Monitoring; Parent concerns always prompt evaluation
Caregiver concern regarding hearing, speech, language delays	Immediate referral	Immediate referral	According to findings & concerns
Family History of hearing loss	24-30 m	By 7-9 m	Based on etiology
NICU care> 5 days, & regardless of LOS:	24-30 m	By 7-9 m	According to findings & concerns
ECMO	24-30 m	**≤ 3 m**	Every 12 m to school
hyperbilirubinemia with exchange transfusion.	24-30 m	By 7-9 m	According to findings & concerns
assisted ventilation	24-30 m	>5d By 7-9 m	
ototoxic medications	24-30 m	**>5d By 7-9 m**	
loop diuretics (Lasix),	24-30 m	>5d By 7-9 m	
Asphyxia or HIE	NA	By 7-9 m	

Follow-up of Infant with Perinatal Risk Factor for Hearing Loss

Risk Factor	JCIH 2007	JCIH 2015	Monitoring; Parent concerns always prompt evaluation
In-utero Torch infections	24-30m	7-9m	As per concerns of hearing & speech
CMV infection	24-30m	By 3m	Q 12 m to school age

Follow-up of Infant with Perinatal Risk Factor for Hearing Loss

Risk Factor	JCIH 2007	JCIH 2015	Monitoring; Parent concerns always prompt evaluation
Craniofacial anomalies, especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies	24-30	7-9m	As per concerns of on-going surveillance
Physical findings associated with a syndrome known to include permanent HL	24-30	7-9m	As per concerns of on-going surveillance
Syndromes associated with progressive HL such as NF, osteopetrosis, Usher's syndrome	24-30	7-9m	According to natural history of syndrome or concerns
Neurodegenerative disorders*, such as Hunter syndrome	24-30	7-9m	As per concerns of on-going surveillance

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Follow-up of Infant with Risk Factor(Perinatal or Postnatal) for Hearing Loss

Risk Factor	JCIH 2007	JCIH 2015	Monitoring; Parent concerns always prompt evaluation
Head Trauma	Follow-up Not specifically stated	≤ 3months	Surveillance/concerns
Chemo therapy*		≤ 3months	Surveillance/concerns
Culture-positive infections associated with sensorineural hearing loss*, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis		≤ 3months	Q 12 m or per concerns
Caregiver concern* regarding hearing, speech, language developmental delay and or developmental regression	immediate	immediate	immediate

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Bob's list of strategies

- **Discuss results** of the newborn hearing screening with parents
- **Identify factors** that place a baby **at risk** of hearing loss in the future
- **Develop, discuss, and document an individualized plan** for following babies who have risk factors. At a minimum, this should include a follow-up audiological evaluation within the first year of life. Some risk factors, however, may demand an earlier evaluation

Bob's list of strategies

- **Modify your electronic medical record** or paper chart as a way to prompt a discussion of risk factors and give a place to document that it occurred

- **Develop a diagnostic code** for those patients identified "at risk" for hearing loss
  - This will allow a way to track those patients who require follow up
  - The responsibility for tracking should also be assigned to a single office staff person
- **Act on parental concern** about hearing loss since it is a risk factor that should result in a prompt referral for evaluation. Never ignore a parental concern or dissuade a family from receiving an evaluation because the baby passed the newborn screen and therefore "could not have a problem with their hearing"

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- Maintain a high level of suspicion for **late-onset hearing loss**
  - Remember, **a passed** newborn **hearing screen does not protect** an individual **from hearing loss forever**
  - Send for diagnostic evaluation at **any point** families have concerns or there is speech/language delay

- It is helpful to **document risk factors** for delayed onset or progressive hearing loss and **monitor hearing and speech/language milestones** in children experiencing these risk factors