Strategies for Identifying Risk Factors for Hearing Loss

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Financial Disclosure

- None relevant to this presentation

The Hearing Screening Gap

Over 98% of newborns are screened for hearing loss at birth.

2019 Summary of National CDC EHDI Data

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About 98% of newborns will pass their hearing screen…

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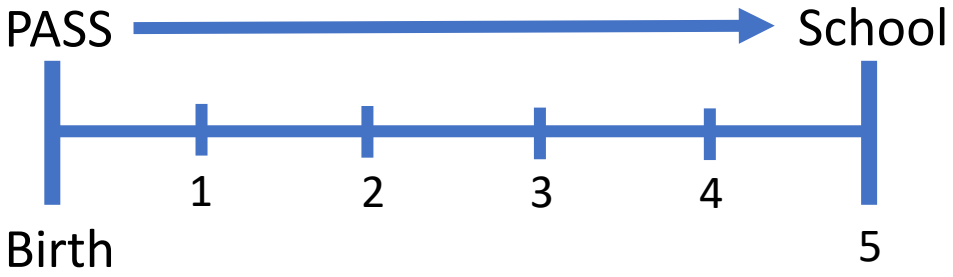

They may not be screened again until starting school.

The Hearing Screening Gap

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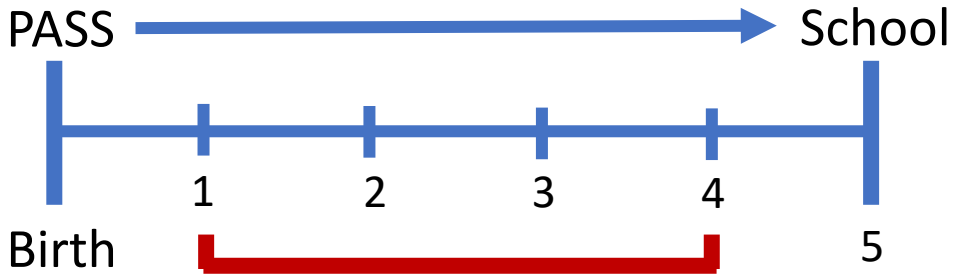
About 98% of newborns will pass their hearing screen…



Critical Period for Language Development

The Hearing Screening Gap

Who is at risk?

- Minimal hearing loss
- Frequency specific
- Late-onset
- Progressive

How do we decide which children to monitor during the “gap” period?

JCIH Risk Indicators for Hearing Loss

- Family History of HL
- NICU stay >5 days
- Hyperbilirubinemia requiring exchange transfusion
- Aminoglycoside administration >5 days
- Significant neonatal hypoxia
- ECMO
- In-utero infections (TORCH)
- Congenital CMV
- Craniofacial anomalies
- Neonatal meningitis

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DON'T FORGET:
Parent/Caregiver Concern

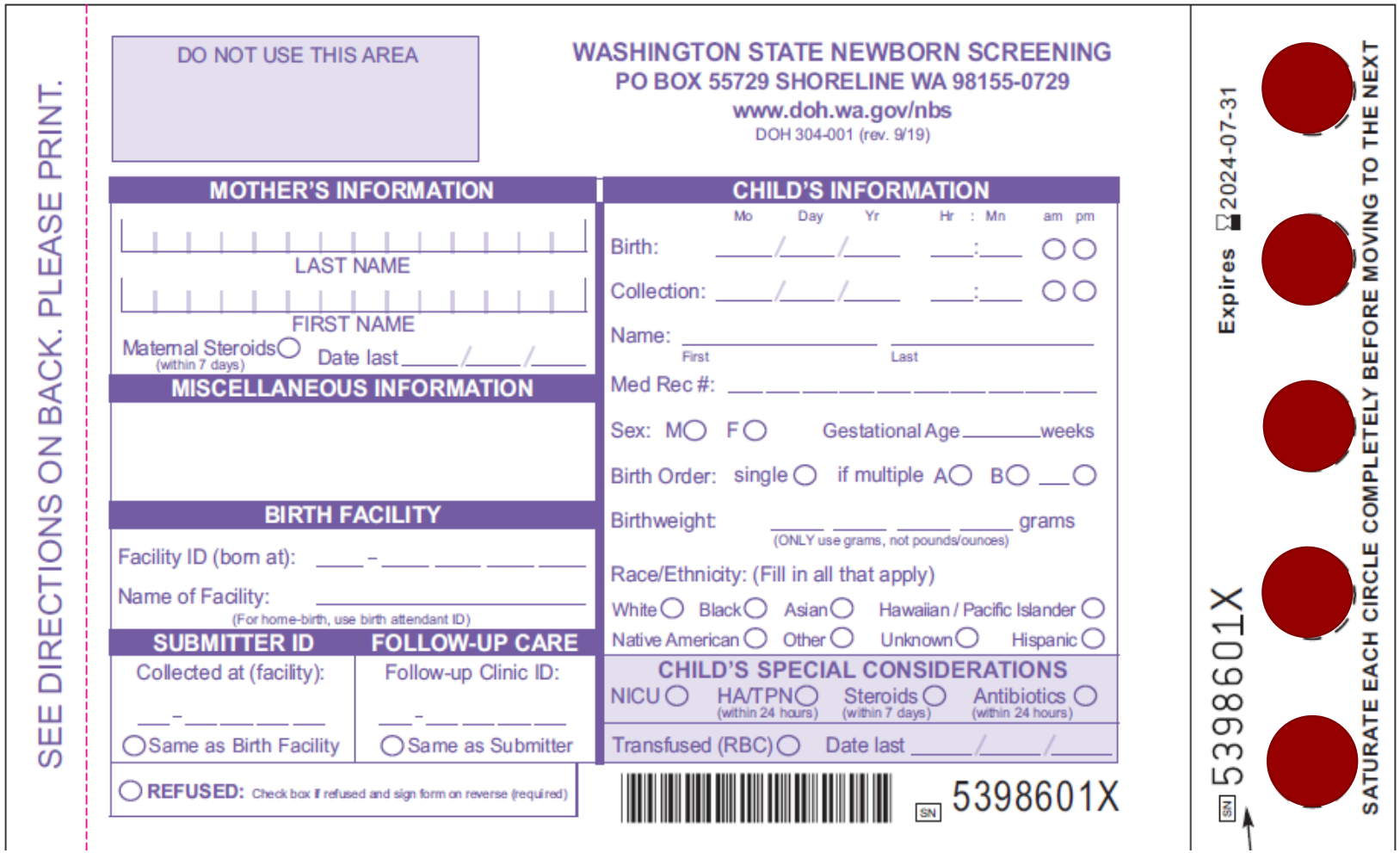
Strategies for Risk Factor Identification

- Karin Neidt: Washington EHDDI Database
- Katie Kuboushek: U of Michigan Electronic Medical Record
- Dylan Chan: Parent/Caregiver Concern

Washington State EHDDI Profile

- 84,000 births
- No mandate for screening or reporting
- System is linked with the Newborn Screening program
- Collects hearing screening and risk factor information on hearing screening card attached to blood spot card

Hearing screening/risk factor information data collection



SEE DIRECTIONS ON BACK. PLEASE PRINT.

DO NOT USE THIS AREA

WASHINGTON STATE NEWBORN SCREENING
PO BOX 55729 SHORELINE WA 98155-0729
www.doh.wa.gov/nbs
DOH 304-001 (rev. 9/19)

MOTHER'S INFORMATION

LAST NAME

FIRST NAME

Maternal Steroids ○ (within 7 days) Date last ____/____/____

MISCELLANEOUS INFORMATION

BIRTH FACILITY

Facility ID (born at): ____-____ ____ ____ ____

Name of Facility: _____
(For home-birth, use birth attendant ID)

SUBMITTER ID

Collected at (facility): ___-___ ___ ___ ___

○ Same as Birth Facility

FOLLOW-UP CARE

Follow-up Clinic ID: ___-___ ___ ___ ___

○ Same as Submitter

○ **REFUSED:** Check box if refused and sign form on reverse (required)

CHILD'S INFORMATION

Mo Day Yr Hr : Mn am pm

Birth: ____/____/____ ____:____ ○ ○

Collection: ____/____/____ ____:____ ○ ○

Name: _____ _____
First Last

Med Rec #: ___ ___ ___ ___ ___ ___ ___ ___ ___ ___

Sex: M ○ F ○ Gestational Age _____ weeks

Birth Order: single ○ if multiple A ○ B ○ ___ ○

Birthweight: ____ ____ ____ ____ grams
(ONLY use grams, not pounds/ounces)

Race/Ethnicity: (Fill in all that apply)

White ○ Black ○ Asian ○ Hawaiian / Pacific Islander ○
Native American ○ Other ○ Unknown ○ Hispanic ○

CHILD'S SPECIAL CONSIDERATIONS

NICU ○ HA/TPN ○ (within 24 hours) Steroids ○ (within 7 days) Antibiotics ○ (within 24 hours)

Transfused (RBC) ○ Date last ____/____/____

SN 5398601X

Expires 2024-07-31

SATURATE EACH CIRCLE COMPLETELY BEFORE MOVING TO THE NEXT

SN 5398601X

Risk Factor Information Collected

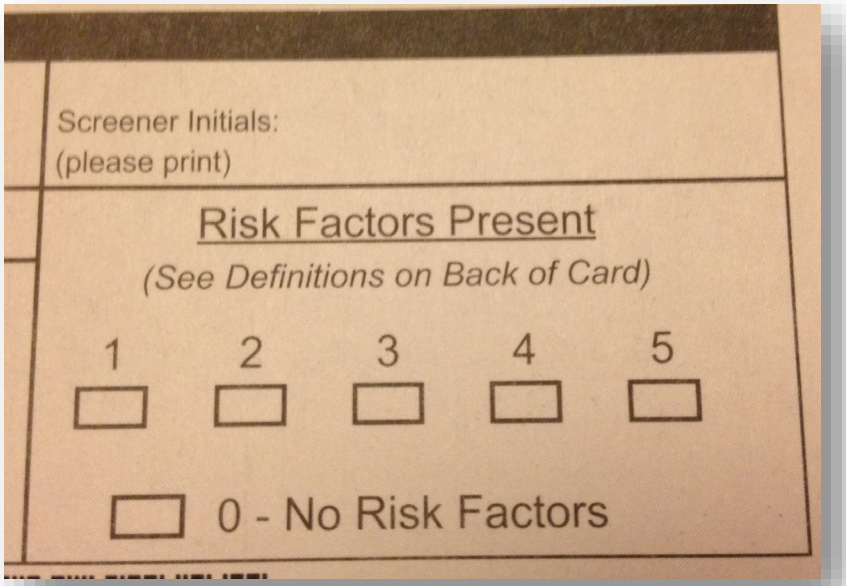
1 – NICU stay > 5 days

2 – Syndromic stigmata

3 – Family history

4 – Craniofacial anomalies

5 – In-utero infection



EHDDI follow-up for risk factors

Risk factor indicated	Child's age when provider is faxed	Follow-up recommendations
NICU	Provider is not faxed	-
Syndrome Family history Craniofacial anomaly	150 days	Diagnostic evaluation before 9 months of age
In-utero infection	30 days	Diagnostic evaluation before: • 3 months of age for CMV and • 9 months for other infections

Reporting by audiologists

- Audiologists can report more detailed risk factor information related to:
  - Caregiver concern
  - Family history
  - Maternal history (infections)
  - Patient history
  - Neonatal indicators
  - Craniofacial anomalies
  - Syndromes

Challenges

- Difficult for hospital screening staff to ascertain risk factor status
- Risk factor information is not always accurately reported by screening staff
  - Family history often over reported
  - Children with oral clefts are often not reported
- Only able to share broad risk factor information with providers (1-5)
- Lack of resources for EHDDI program to ensure infants with risk factors receive audiological evaluation

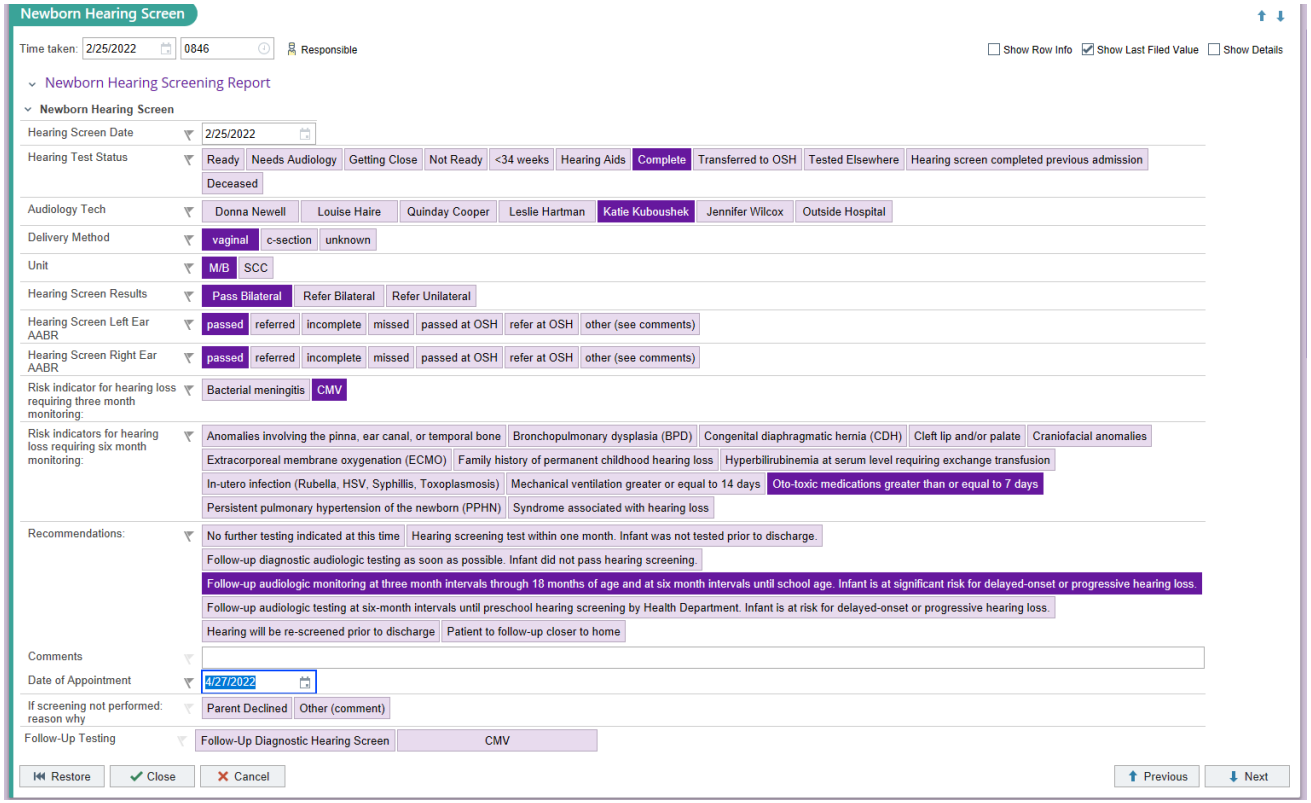
U of Michigan Health EHDI Tracking Methods

2000-2016 present

2016-2020

2020-

The great flowsheet build of 2020



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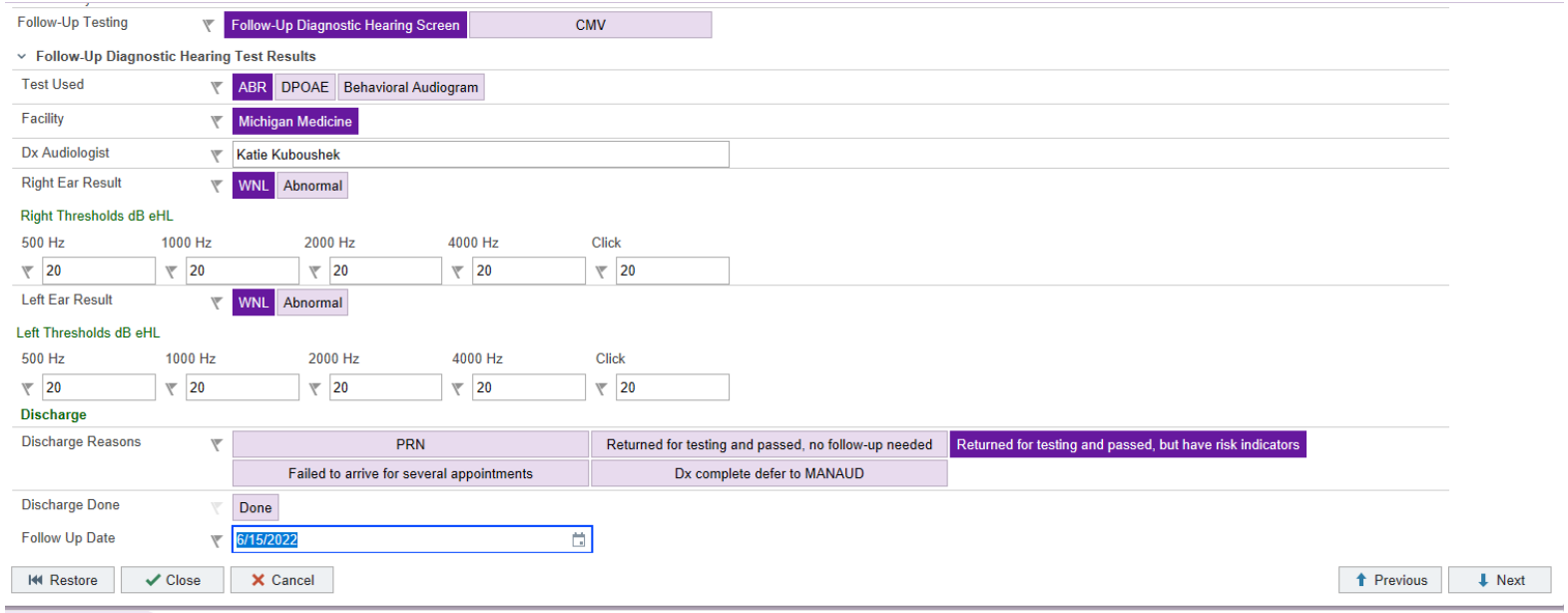


Current utilization

- State reporting (outpatient and inpatient)
- Risk indicators
- Tracking new id’s
- Lost to follow-up tracking
- CMV tracking

Future utilization

Early phase of transferring existing data into artificial intelligence system.

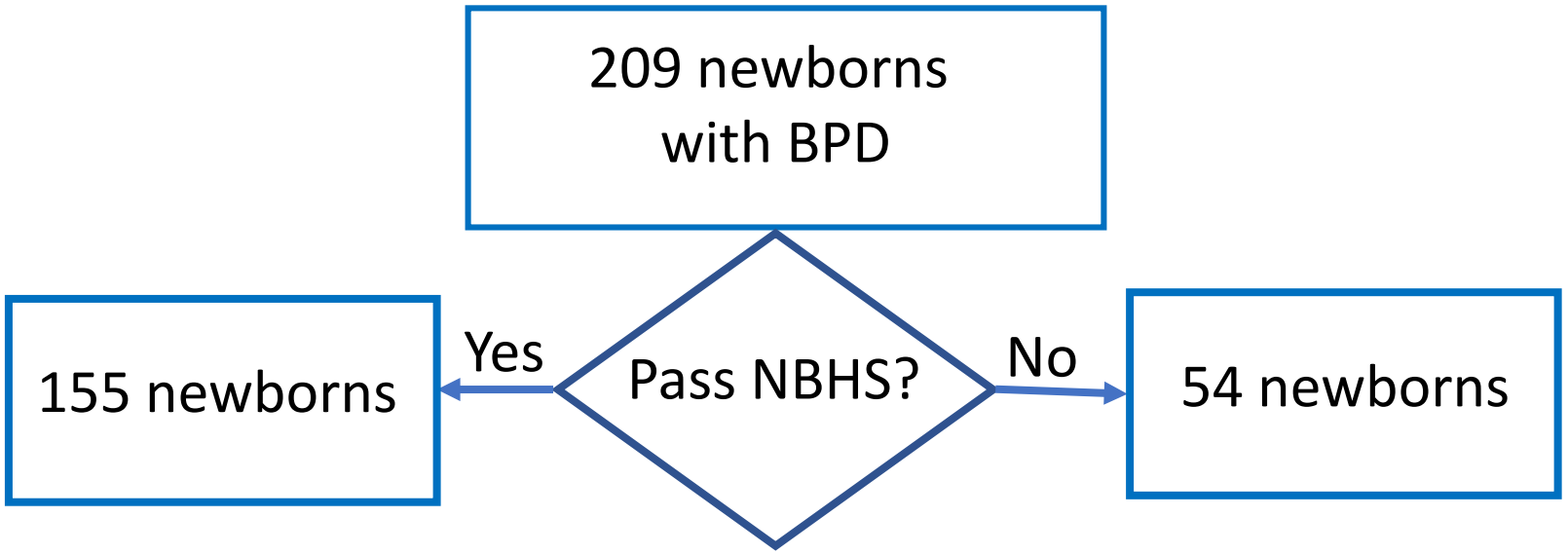
- Informed decision making
- Better treatment outcomes
- Improve lost to follow-up rates
- Identify unknown risk indicators

Example: Data in action

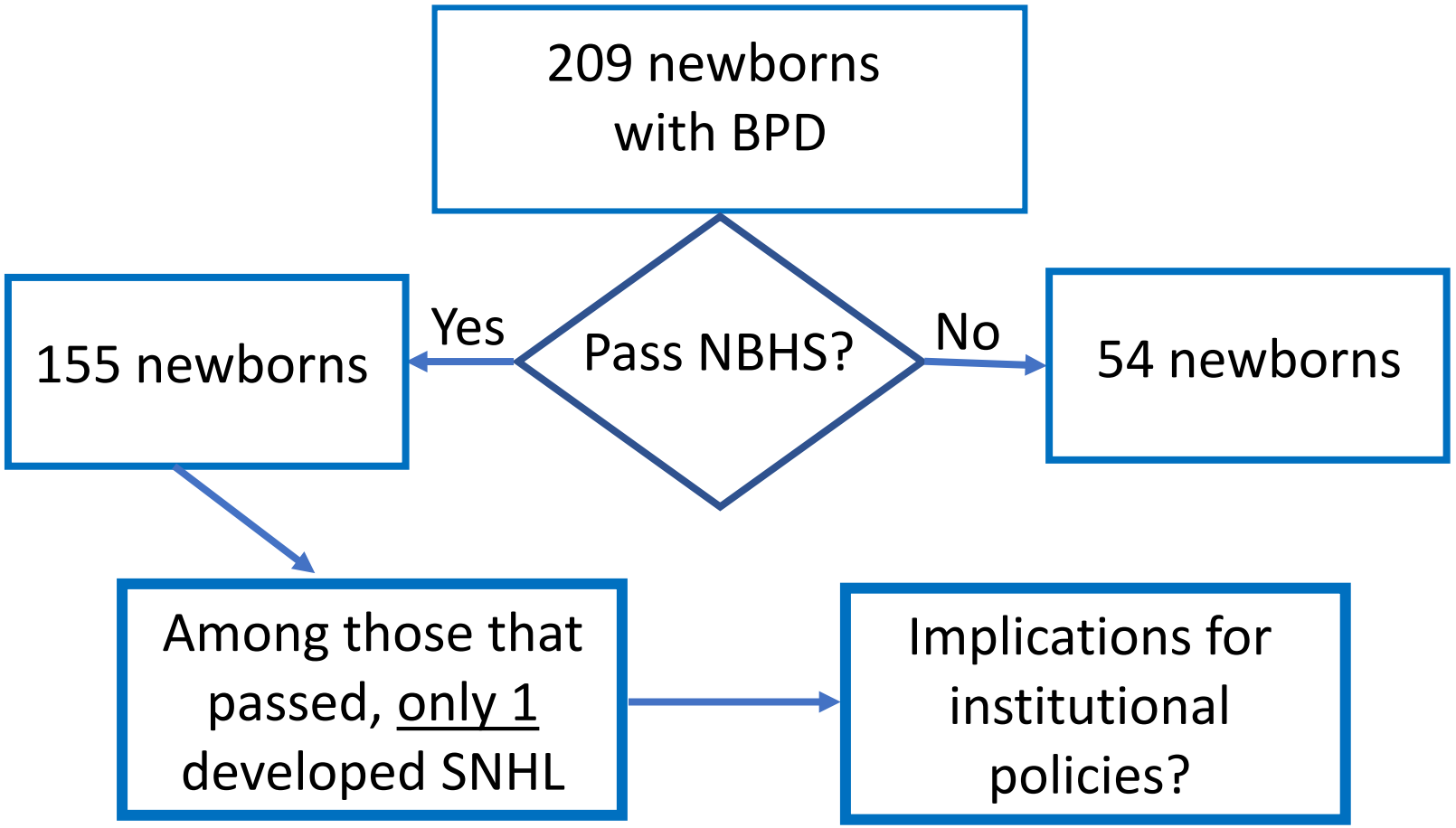
What is the rate of permanent hearing loss among premature newborns with lung disease (bronchopulmonary dysplasia)?

- Babies born 2013-2019
- Identify all with bronchopulmonary dysplasia (BPD)
- Check newborn hearing screening results
- Review all available audiograms

BPD: Newborn Hearing Screening



BPD: Hearing Outcomes



Later childhood hearing loss JCIH risk factors

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 no laboratory evidence & no clinical findings	standard	As per AAP (2017) Periodicity schedule
	Mother + Zika and infant with laboratory evidence of Zika + clinical findings Mother + Zika and infant with laboratory evidence of Zika - clinical findings	AABR by 1 month AABR by 1 month	ABR by 4-6 months or VRA by 9 months ABR by 4-6 months Monitor as per AAP (2017) Periodicity schedule (Adebanjo 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.

Journal of Early Hearing Detection and Intervention, 4(2), 1-44. DOI: 10.15142/fptk-b748
https://digitalcommons.usu.edu/jehdi/vol4/iss2/1/

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11	Events associated with hearing [illegible] • Significant head trauma especially basilar skull fractures • Chemotherapy	[illegible] occurrence	[illegible] and or continued concerns
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Parent or caregiver concern regarding hearing, speech, language or developmental delay.

Increasing volume on TV
Speaking louder
"Not listening"
Poor attention
Speech delay

Immediate Referral!

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How reliable is parent/caregiver concern?

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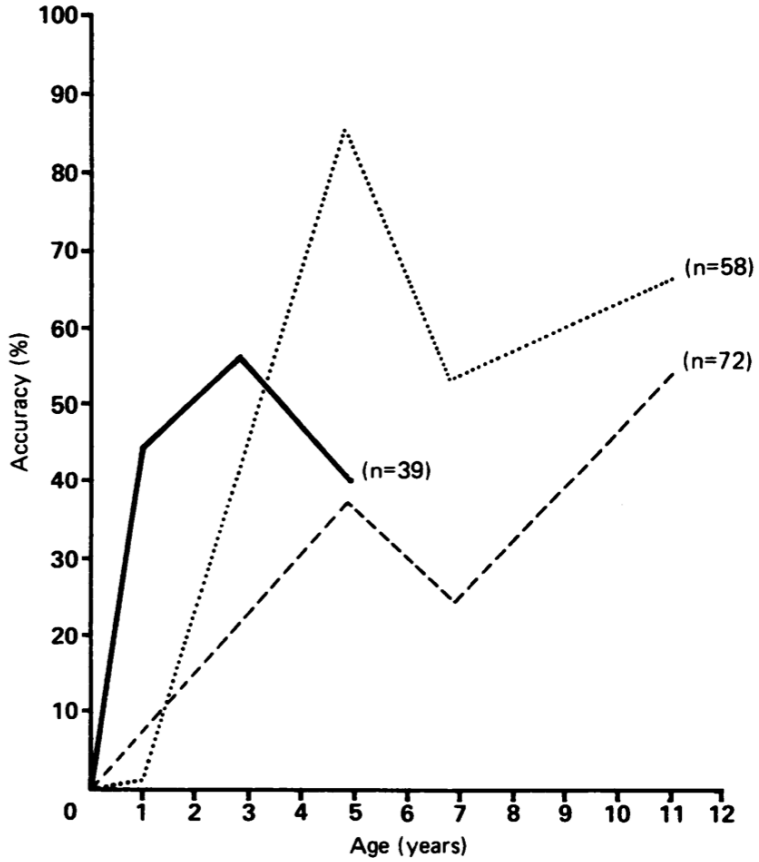
Parent concern SNHL



Sensitivity of parental suspicion preceding diagnosis of permanent hearing loss in childhood. Mild or moderate hearing loss (– – –), severe or profound hearing loss (—), and unilateral hearing loss (· · ·).

- 169 children with SNHL
- Parental suspicion highly insensitive to identifying hearing loss

Watkin *et al.* (1990), Arch Dis Child

Parent concern Otitis media & hearing loss

	Children with average PTA Thresholds >25 dB (n=17)	Children with average PTA Thresholds ≤25 dB (n=259)	
Parental suspicion of hearing loss (n=28)	2	26	PPV 7.1%
Parental perception of no hearing loss (n=248)	15	233	NPV 94.0%
	Sensitivity 11.8%	Specificity 90.0%	

- 276 children with concern for middle-ear effusion and hearing loss
- Parental concern for hearing loss:

  Sensitivity = 11.8%

Sensitivity:

What percentage of children WITH hearing loss had hearing loss suspected by the parents?

Lo *et al.* (2006), Eur J Pediatr

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  Specificity = 90%

Specificity:

What percentage of children WITHOUT hearing loss were accurately thought to not have hearing loss by their parents?

Lo *et al.* (2006), Eur J Pediatr

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- Parental concern for hearing loss:

  Sensitivity = 11.8%
  Specificity = 90%
  PPV = 7%

Positive predictive value:

What percentage of parents who thought their children had hearing loss were correct?

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- Parental concern for hearing loss:

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  Specificity = 90%
  PPV = 7%
  NPV = 94%

Negative predictive value:

What percentage of parents who thought their children did NOT have hearing loss were correct?

Lo *et al.* (2006), Eur J Pediatr

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- 276 children with concern for middle-ear effusion and hearing loss
- Parental concern for hearing loss:

  Sensitivity = 11.8%
  Specificity = 90%
  PPV = 7%
  NPV = 94%
  Odds ratio = 1.2

Odds ratio:

If a parent thinks their child had hearing loss, what is the chance that they actually have hearing loss, compared to kids whose parents DON'T think they have hearing loss?

Lo *et al.* (2006), Eur J Pediatr

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Parental suspicion very poorly correlated with actual hearing status

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Lo *et al.* (2006), Eur J Pediatr

Caregiver concern Speech/Hearing/Language

Table 4. Outcomes of Teacher Concerns for Speech, Hearing, and Language

Variable	Referral	No referral	OR (95% CI)	*P* value[a]
Referred screening				
Speech concern				
Yes	36	64	9.7 (6.4-14.8)	<.001
No	367	6353		
Hearing concern				
Yes	28	32	14.1 (8.9-25.0)	<.001
No	375	6385		
Language concern				
Yes	27	35	13.1 (7.8-21.9)	<.001
No	376	6382		
CHL	CHL	No CHL	NA	NA
Speech concern				
Yes	21	100	8.1 (5.0-13.3)	<.001
No	174	6720		
Hearing concern				
Yes	15	45	12.2 (6.7-22.3)	<.001
No	180	6580		
Language concern				
Yes	15	47	11.7 (6.4-21.3)	<.001
No	180	6578		
SNHL	SNHL	No SNHL	NA	NA
Speech concern				
Yes	1	99	5.7 (0.7-43.8)	.062
No	12	6708		
Hearing concern				
Yes	2	58	21.2 (4.6-97.6)	<.001
No	11	6749		
Language concern				
Yes	1	61	9.2 (1.2-72.0)	<.001
No	12	6746		

- Study of 6820 low-income preschoolers undergoing two-stage, single-visit pure tone audiometry/OAE hearing screening
- 99.6% screening success; 86% follow-up rate
- Teacher concern for language delay:

  Sensitivity = 8.2%
  Specificity = 99.3%
  PPV = 28.3%
  NPV = 97.1%
  Odds ratio = 13.4

Brodie and David *et al.* (2022), JAMA-Otolaryngology-Head and Neck Surgery

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Teacher concern associated with 13x greater rate of hearing loss

Listen to teachers/caregivers!

Brodie and David *et al.* (2022), JAMA-Otolaryngology-Head and Neck Surgery

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Hearing concern				
Yes	15	45	12.2 (6.7-22.3)	<.001
No	180	6580		
Language concern				
Yes	15	47	11.7 (6.4-21.3)	<.001
No	180	6578		
SNHL	SNHL	No SNHL	NA	NA
Speech concern				
Yes	1	99	5.7 (0.7-43.8)	.062
No	12	6708		
Hearing concern				
Yes	2	58	21.2 (4.6-97.6)	<.001
No	11	6749		
Language concern				
Yes	1	61	9.2 (1.2-72.0)	<.001
No	12	6746		

Teacher concern associated with 13x greater rate of hearing loss

Listen to teachers/caregivers!

(But, also listen to parents)

Brodie and David *et al.* (2022), JAMA-Otolaryngology-Head and Neck Surgery

In Summary

The hearing screening gap occurs during the critical period for language development.

Risk indicators can help us to identify which children to monitor during the gap period.

There are state-level and institutional-level models for risk indicator identification and monitoring.

Don’t forget the importance of parent/caregiver concern!