

# Timely Detection of Late-Onset Hearing Loss in Taiwan: Risk Factor Follow-up



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# OUTLINE

01

Background

02

Late-onset  
hearing loss (HL) &  
its impacts

03

Risk Factors

04

Follow-up timeline

05

Conclusion



# Background : In Taiwan



## UNHS since 2012

2022: 98.7% screening rate



## Preschool Hearing Screening

2021:

59.3% screening rate

10.3% referral rate



# Late-onset Hearing Loss

## Definition

- Diagnosed after **3 months** of age in children who have passed UNHS



## Prevalence

- 7.5 in 10,000 children who passed UNHS
- Accounts for 22% of children with bilateral HL

## Challenges

- Parental difficulty in detection
- A pass in UNHS may foster the misconception that a child's hearing will always be normal



# Aspects of Impact Due to Late-onset Hearing Loss in Children

- Ability to perceive sounds, words, and language rules
- Speech and language development
- Communication skills
- Academic performance
- Cognitive development
- Social-emotional development

# Timely intervention is the key to reducing the impact of late-onset hearing loss



**Language  
performance**

**Late-  
onset HL**

**=  
N.S.**

**Early-  
onset HL**

When intervention was provided, on average, 1 month  
after identification for both groups

# Risk Factor Monitoring:

## A Pivotal Approach to Timely Detection

- Regular monitoring of children who pass UNHS but exhibit risk factors for HL enables timely and effective detection of potential late-onset HL (Weichbold et al., 2006).



# Risk Factors (1/2)

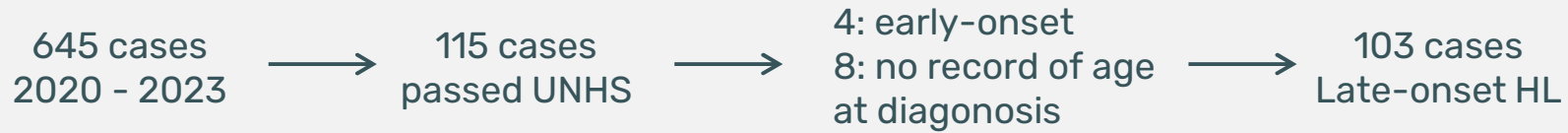
- **Perinatal factors**
  - Birth weight < 1500g
  - Apgar score 0-4 at 1min, 0-6 at 5min
  - NICU > 5 days
  - In utero infections: CMV, herpes, rubella, syphilis, toxoplasmosis
- **Medical treatments**
  - ECMO, assisted or mechanical ventilation, hyperbilirubinemia that requires exchange transfusion
  - Exposure to ototoxic medications (e.g., gentamicin, tobramycin) or loop diuretics (e.g., furosemide/Lasix)
  - Chemotherapy
- **Infections**
  - Culture-positive postnatal infections: bacterial and viral meningitis (especially herpes viruses and varicella)
  - Recurrent or persistent otitis media with effusion > 3 months



# Risk Factors (2/2)

- **Family history & concerns**
  - Family history of permanent childhood hearing loss
  - Caregiver concern regarding hearing, speech, language, or developmental delay
- **Disorders**
  - Syndromes associated with hearing loss: neurofibromatosis type II, osteopetrosis, Usher, Waardenburg, Treacher Collins, Alport, Pendred, Jervell and Lange Nielson
  - Neurodegenerative disorders: Hunter syndrome, Friedreich ataxia, Charcot-Marie-Tooth syndrome
- **Craniofacial anomalies**
  - Anomalies including cleft lip / palate, microtia, aural atresia, choanal atresia
  - Physical findings associated with syndromes known to include hearing loss (e.g., white forelock)
- **Head trauma**
  - especially basal skull/temporal bone fracture that requires hospitalization

# Risk factor investigation for CHF's children



Medical history  
review



Analysis of risk  
factors



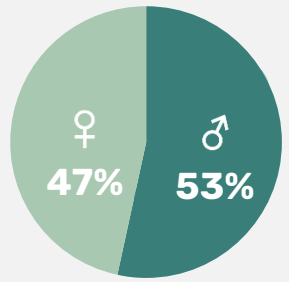
Descriptive  
statistics

may have had congenital  
HL undetected by UNHS



# Demographics

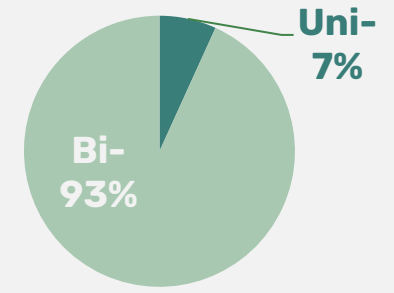
Sex



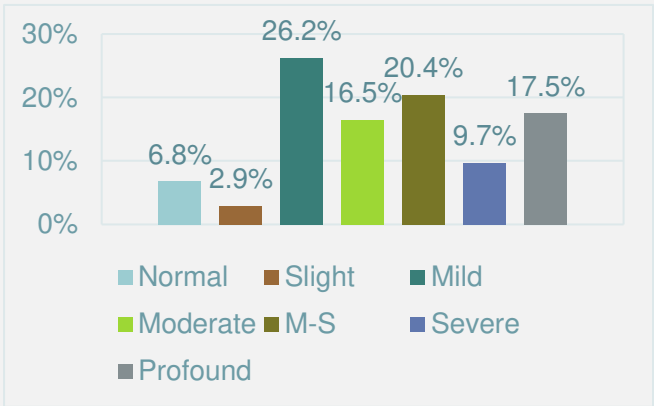
Age at diagnosis (mo.)

Mean: 39  
SD = 26  
Range: 4-127

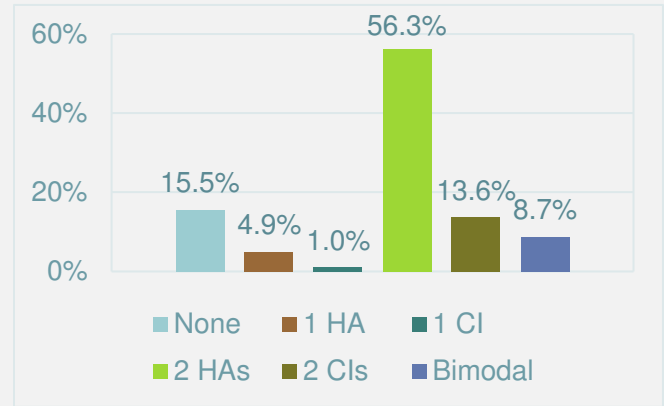
Uni- vs. Bi-lateral HL



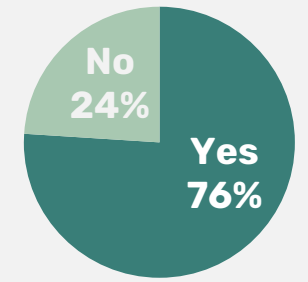
Degree of HL (better ear)



Technology



Intervention enrollment



# Risk factors exhibited in CHF's children (1/2)

- **Perinatal factors**

- Birth weight < 1500g
- Apgar score 0-4 at 1min, 0-6 at 5min
- NICU > 5 days
- In utero infections: CMV, herpes, rubella, syphilis, toxoplasmosis

Gestational diabetes /  
pre-eclampsia

- **Medical treatments**

- ECMO, assisted or mechanical ventilation, hyperbilirubinemia that requires exchange transfusion
- Exposure to ototoxic medications (e.g., gentamicin, tobramycin) or loop diuretics (e.g., furosemide/Lasix)
- Chemotherapy

- **Infections**

- Culture-positive postnatal infections: bacterial and viral meningitis (especially herpes viruses and varicella)
- Recurrent or persistent otitis media with effusion > 3 months

(Dumanch et al., 2017; JCIH, 1990, 2000, 2007, 2019)

# Risk factors exhibited in CHF's children (2/2)

- **Family history & concerns**

- Family history of permanent childhood hearing loss
- Caregiver concern regarding hearing, speech, language, or developmental delay

Identified HL  
related gene(s)

- **Disorders**

- Syndromes associated with hearing loss: neurofibromatosis type II, osteopetrosis, Usher, Waardenburg, Treacher Collins, Alport, Pendred, Jervell and Lange Nielson
- Neurodegenerative disorders: Hunter ataxia, Friedreich ataxia, Huntington's disease, Tooth syndrome

Branchio-Oto-Renal  
(BOR) Syndrome

Large vestibular  
aqueduct syndrome  
(LVAS) Syndrome

- **Craniofacial anomalies**

- Anomalies including cleft lip / palate, microtia, aural atresia, choanal atresia
- Physical findings associated with syndromes known to include hearing loss (e.g., white forelock)

- **Head trauma**

- especially basal skull/temporal bone fracture that requires hospitalization

# True examples of caregiver concern regarding hearing, speech, language, or developmental delay

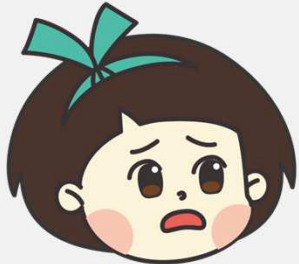
At age 2, he remains non-verbal, sporadically producing meaningless sounds, and rarely vocalizing.

When she was younger, she responded well to calls, but later showed no response to calls from behind.

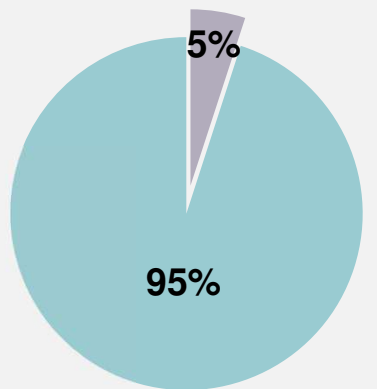
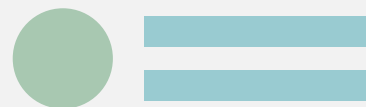
Currently, she only produces the sounds 'ma1' and 'ma4,' and has not yet produced 'ba4.'

Unable to form complete sentences, mostly using single words.

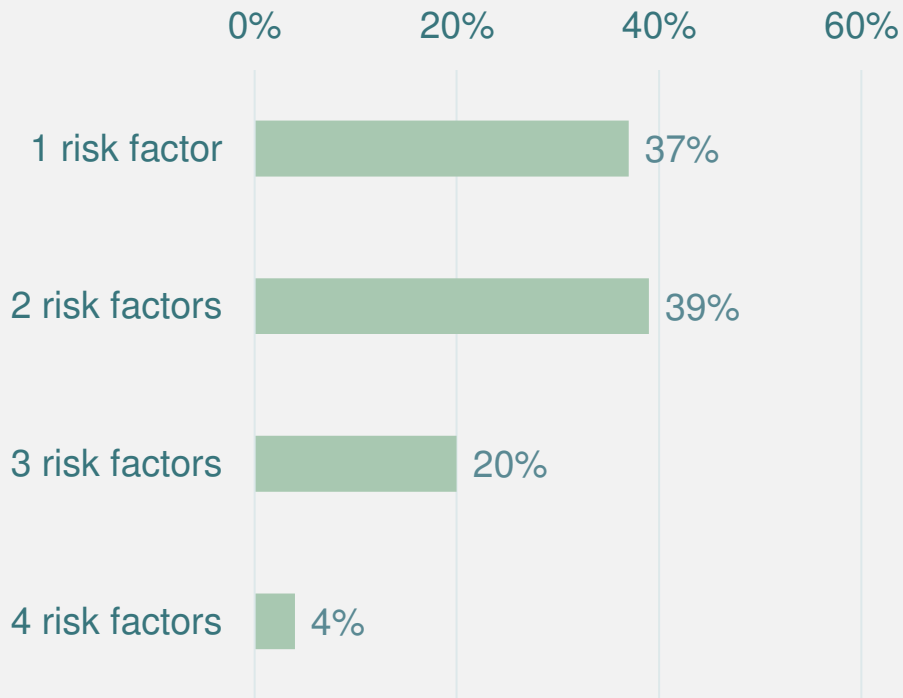
Displays poor response to sounds; slowly turns head to loud noises or rarely shows startle response even when being called loudly.



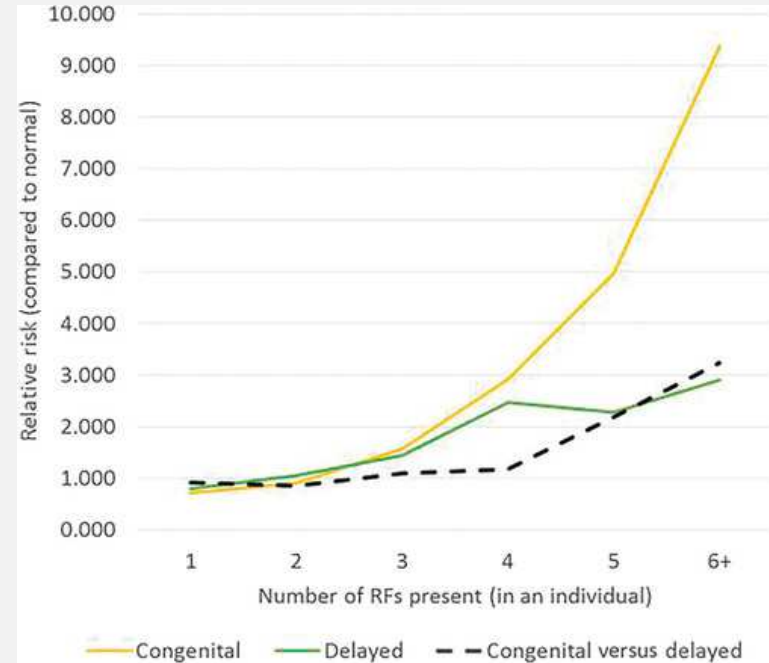
# The occurrence of risk factors is **variable and complex**



■ No risk factors  
■ One or more risk factors



The greater the number of risk factors exhibited, the higher the likelihood of both congenital and late-onset hearing loss



(Dumanchet al., 2017)



# Follow-up Timeline

## Babies who passed UNHS

### No risk factors

Ongoing surveillance of communicative development

### Certain risk factors

- Caregiver concern
- Family history
- NICU > 5 days
- ECMO
- CMV
- Syndromes associated with HL
- Neurodegenerative disorders
- Meningitis
- Head trauma

Hearing evaluation by 6 months of age

### Other risk factors

- Low birth weight
- Hyperbilirubinemia
- Ototoxic medications
- Congenital herpes, rubella, syphilis, toxoplasmosis
- Craniofacial anomalies
- Recurring or persistent otitis media with effusion > 3 mo.
- Others

Audiologic follow-up at 24-30 months of age

# Supporting children with late-onset hearing loss

## Risk factor follow-up

- Analysis based on CHF's database
- Analysis based on Taiwan Universal Health Insurance Database
- Public awareness promotion

## Hearing performance checklist

- For children between ages of 3 and 6
- Parents and/or preschool/kindergarten teachers
- Under development

## Preschool hearing screening

- Public health nurse training for preschool hearing screening
- Advocacy for local government implementation of preschool hearing screening



**2024**  
AG BELL GLOBAL  
**LISTENING AND  
SPOKEN LANGUAGE**  
VIRTUAL SYMPOSIUM  
**JUNE 26-27, 2024**

# Take home messages

- Reviewing and documenting the presence of **risk factors** for hearing loss is crucial in identifying the potential for late-onset HL.
- As per JCIH guidelines, children who pass UNHS but present risk factors for hearing loss should undergo hearing evaluation by **6 months of age**, or **between 24 and 30 months of age**, depending on the specific risk factors.
- Even with negative results of UNHS, hearing and speech professionals must NOT overlook **caregiver concerns** regarding their child's hearing, speech, and language development.
- It is important NOT to disregard the possibility of hearing loss even in children **without identified risk factors**.





# Acknowledgements



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# THANK YOU

Any questions?

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Helping Children with Hearing Loss Learn to Listen and Speak

