Identification of Hearing Loss Post Newborn Hearing Screening in Children with Known or Suspected Neurodevelopmental Involvement

Savanah DiPasquantonio, M.S., Holly Duncan, Au.D., Amy Gaskin, Au.D., Dorothy Shiffler, Au.D.
The Kennedy Krieger Institute

Introduction

- Approximately 1.3 million American children under the age of three have hearing loss.
- Infants who passed their NBHS may develop, or show evidence of, infant or childhood HL post NBHS. This may reflect delayed-onset hearing loss associated with certain diagnoses/atypical neurodevelopment, as well as missed conductive, sensory, or neural hearing loss at the time of NBHS.
- In a report (Watkin & Baldwin, 2012), the prevalence of children confirmed as deaf or hard of hearing by school age was 3.65/1000 compared to a neonatal yield of 1.79/1000.
- Congenital, early-onset, or late-onset hearing loss may be the result of genetic, neurologic, anatomical, and environmental factors, or a combination of them.
- A primary goal of pediatric audiology is to ensure that children have adequate hearing to meet their communication and learning needs. When a specific population is identified to have a high risk for hearing loss, close monitoring and management is necessary.
- Early detection of hearing loss, and effective intervention can decrease the impact that hearing loss has on a child's development.

Learning Objectives

- Gain a better understanding of the prevalence of comorbid hearing loss in children with known or suspected neurodevelopmental involvement beyond what is addressed in JCIH
- Be aware of and use guidelines for referrals to audiology from other disciplines and guidelines for audiologic monitoring

Methods

- A literature review to establish the current evidence regarding the prevalence of hearing loss in pediatric patients with common neurodevelopmental disorders in which hearing loss is often comorbid and may be of postnatal onset or identification. Other factors, such as hearing loss characteristics, time of onset, and follow-up recommendations were also reviewed.
- A resource was created for professionals and parents to reference regarding hearing loss and common neurodevelopmental disorders, as well as the recommendations for audiologic referrals and monitoring.

	Type of HL	Etiology/Prevalence	Audiologic Recommendations *
Autism Spectrum Disorder (ASD)	CHL SNHL MHL *Progressive*	-Increased prevalence of RSOM (23.5%) -Increased rates of audiological dysfunction	-Evaluation in all cases where ASD is suspected or diagnosed
Cerebral palsy (CP)	CHL SNHL MHL	-Damage to inner ear due to numerous neonatal risk factors -Children with dyskinetic or hypotonic forms of CP who are more severely affected are more likely to have HL -HL prevalence (12%)	-Evaluation in all cases where CP is suspected or diagnosed
Charcot Marie Tooth syndrome	SNHL Neural Central *Sudden* *Progressive*	-Auditory neural function may worsen with disease progression -Electrophysiological evidence of auditory neuropathy with delayed or low amplitude auditory brainstem responses -Impaired processing of auditory temporal cues and/or abnormal speech understanding in everyday listening conditions	-Annual evaluations- important to include auditory evoked potentials and speech perception (in noise) assessment
Congenital Cytomegalovirus (cCMV)	SNHL Sudden *Late-onset* *Progressive*	-Damage to the neural cells and/or secondary to host-derived inflammatory responses to CMV in the inner ear -Leading cause of SNHL in children (33.3%)	-Initial diagnostic evaluation no later than 3 months of age -Re-evaluations every 6 months until 8 years old, or more often if fluctuations/progressive loss is noted
Down syndrome	CHL SNHL MHL *Progressive*	-Structural abnormalities in the outer, middle, and inner ear -RSOM -HL prevalence (36%)	-Evaluations every six months until school age, then annually through childhood
Fetal Alcohol syndrome	CHL SNHL MHL Neural Central	-RSOM due to an immune deficiency syndrome -Malformations of the Eustachian tube -Increased cell death in the embryonic inner ear, resulting in reduced numbers of sensory receptor cells and auditory nerve fibers -Malformations of various brainstem and forebrain structures involved in auditory processing	-ABR for NBHS, instead of OAE -Annual evaluations
Fragile X syndrome	CHL	-RSOM	-Evaluation in all cases where Fragile X is suspected or diagnosed
Friedreichs Ataxia	SNHL Neural Central *Progressive*	-Progressive hearing loss can start as early as birth -Disordered neural conduction in the central auditory pathways and synchrony in auditory nerve fiber discharge	-Annual evaluations- important to include auditory evoked potentials and speech perception (in noise) assessment
Hunter syndrome	CHL SNHL MHL *Progressive*	-RSOM, tympanomastoid abnormalities -Progressive HL can start as early as 2 years old -Early in the disease course, CHL is most common -As the disease progresses, patients with CHL may develop SNHL, resulting in a MHL	-Annual evaluations
Hydrocephalus	SNHL *Permanent* *Reversible*	-Auditory dysfunction may arise in the development of hydrocephalus, because of shunt operations, or as a result of neurosurgical intervention, including puncture and drainage of CSF out of the subdural space -CSF shunting for hydrocephalus has been associated with loss of hearing in some cases, and resolution of hearing loss in others	-Evaluation in all cases where hydrocephalus is diagnosed -Evaluation 6 weeks after any shunt placement or revision
Mitochondrial disorders	SNHL *Sudden* *Progressive* *Fluctuating*	-End-organ dysfunction due to deficient energy release within the stria vascularis or hair cells and/or the accumulation of mutated mtDNA within the cochlea	-Evaluation in all cases where mitochondrial disorders are diagnosed
Sickle cell disease (SCD)	CHL SNHL MHL *Progressive*	-Histopathological and degenerative changes have been observed in the temporal bone and organ of Corti -HL Prevalence (12-26%)	-Evaluation as part of the ongoing assessment of children with SCD is recommended

*Evaluations should occur more frequently dependent on the nature of the HL or if concerns for hearing arise
KEY: HL: hearing loss, CHL: conductive hearing loss, SNHL: sensorineural hearing loss, MHL: mixed hearing loss, RSOM: recurrent serous otitis media, ABR: auditory brainstem response, OAE: otoacoustic emissions, CSF: cerebrospinal fluid, NBHS: newborn hearing screening
References available upon request

Discussion

- Because of the diverse nature of neurodevelopmental disorders, the characteristics and prevalence of comorbid hearing loss is variable.
- The combination of undetected or untreated hearing loss and neurodevelopmental involvement puts young children at risk for language and communication challenges.
- In cases where neurodevelopmental disorders and hearing loss co-exist, diagnosis of one condition may lead to a delay in diagnosing the other.
- This variability highlights the need for parents, caregivers, primary care physicians, other healthcare providers and teachers to be informed about the potential for hearing loss among individuals with neurodevelopmental disorders, to advocate for hearing evaluations, and to assure immediate interventions once hearing loss is identified.