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Beyond Newborn Hearing Screening:

Recognizing the Signs of Late Onset Hearing Loss in Infants and Young Children

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With 39 out of 50 states mandating universal newborn hearing screening (UNHS), and newborn hearing screening data being collected from all 40 states, it would be easy to assume that the identification of permanent childhood hearing loss is guaranteed. Yet, the fact is that some childhood hearing losses have a later onset and will not be identified through newborn screening methods. This article responds to some basic questions about late-onset hearing loss in infancy and childhood.

What is the prevalence of childhood hearing loss?

Current UNHS statistics indicate an overall hearing loss prevalence rate of 1-2 per 1000 at birth. These prevalence statistics are consistent across the US and are not dependent on the particular hearing screening method being used. Statistical information about the prevalence of hearing loss in older children is difficult to find and interpret for a number of reasons. Late onset or progressive hearing loss can be due to hereditary factors, infection, trauma, noise exposure or teratogens. Studies also vary in how "significant hearing loss" is defined. As a result, the prevalence of late onset hearing loss is not well defined. In general there is a trend toward increasing rates of hearing loss as children get older.

Can newborn hearing screening miss hearing loss that is present at birth?

It is possible for some children to have a mild or minimal hearing loss at birth and pass universal hearing screening. This is due, at least in part, on the underlying assumptions about newborn hearing screening. Any type of universal screening program needs to achieve a low false-alarm rate and a high "hit" rate. The goal for UNHS is that few children are referred for additional, more expensive testing who do not need it and those who are referred have a high likelihood of having hearing loss. To meet these requirements, current UNHS methods may not identify children with mild hearing losses. If no further audiological monitoring is being completed within the child's medical home, the result could be late identification of milder degrees of hearing loss.

In some instances, mild hearing loss that is present at birth may progress to more severe hearing loss after the child goes home from the hospital. Rapidly progressive hearing loss can be associated with several congenital conditions, including Cytomegalovirus (CMV) and Large Vestibular Aqueduct (LVA) as well as some genetically inherited losses.

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What are the most common causes of late-onset hearing loss?

The major categories of late-onset loss are acquired, structural, and genetic.

Acquired

Among acquired late-onset losses, congenital CMV (both symptomatic and non-symptomatic) is the most common and accounts for around 1/3 of all hearing loss in children. Hearing loss associated with CMV may be both late onset and progressive within the first years of life. Even asymptomatic congenital CMV infection carries an increased risk of hearing loss. Hearing loss prevalence rates of 7- 15% in asymptomatic cases have been reported. Congenital symptomatic CMV infection carries greater risk for hearing loss and a higher percentage of children with active CMV symptoms at birth have hearing loss identified through UNHS, with further progression reported within the first 2-3 years (Fowler, et al 1997; Barbi, et al 2003). Other childhood illnesses may also cause hearing loss. These include viral or bacterial meningitis, mumps and other viral infections that cause a high fever or central sequelae. Head trauma with skull fracture is one type of traumatic late-onset loss. Chemotherapeutic agents containing platinum, such as cisplatin, are among the best known ototoxic medications.

Structural

Structural causes of late-onset hearing loss may occur with a number of syndromes. Structural deformities of the cochlea such as LVA and Mondini malformation are congenital but not always related to a specific syndrome. Cochlear malformations affect hearing differently in different children. Some hearing losses may occur earlier and others may not present until later childhood. Structural malformations of the inner ear are associated with sudden and extreme progression and fluctuation of hearing.

Genetic

Genetic causes of late-onset hearing loss may be syndromic or non-syndromic. Full explanations of specific syndrome characteristics can be found online on **OMIM, the Online Mendelian Index in Man**.

Syndromic losses include:

- Pendred's Syndrome, which is associated with LVA
- Branchio-Oto-Renal Syndrome (BOR), associated with Mondini deformities
- Alports Syndrome with progressive renal failure and late occurring, progressive hearing loss
- Stickler Syndrome, a connective tissue syndrome with late occurring vision problems and hearing loss
- Usher Syndrome with progressive blindness and deafness. Usher Type I is associated with more severe hearing loss, lack of vestibular function and blindness. Types II and III typically show less severe hearing loss, less severe vestibular effects and more residual vision, with Type III occurring rarely
- Neurofibromatosis Type II with progressive hearing loss resulting from auditory nerve tumors
- Other neurodegenerative syndromes may be associated with late onset hearing loss, but are not as common as the syndromes listed above (e.g. Refsum Disease)

Non-syndromic losses include:

- Dominant-progressive hearing loss
- Family history of late-occurring hearing loss
- Connexin 26, which may have late-onset hearing loss in rare occurrences. A small number of studies have shown progressive hearing loss with Connexin 26

What are the main risk factors associated with late-onset loss?

- Congenital CMV infection
- Meningitis or mumps infections
- Family history of late-onset hearing loss
- Syndromes associated with late-onset hearing loss
- Head trauma, especially with basal or temporal bone fracture
- Chemotherapy, especially when administered in conjunction with radiation

How can Primary Care Providers monitor for late-onset loss?

Performing surveillance and screening within the medical home is the best way to monitor infants and young children for late-onset hearing loss. Primary Care Providers are the medical providers who see the child most often and are able to review auditory skill development and developmental milestones at well-child visits. An immediate referral for audiological evaluation is warranted if parents express concerns about a child's hearing responsiveness or speech and language development. For young children and infants under 3 years of age, typical in-office hearing screening methods are not effective and a referral to a pediatric audiologist is recommended. All children with an identified risk factor for late-onset hearing loss should receive a comprehensive audiological assessment as soon as behavioral testing can be completed. Even if that child has passed newborn screening and no parental concerns have been expressed, a comprehensive evaluation can identify subtle or progressive losses which require remediation and monitoring. A combination of electrophysiological and developmentally-appropriate behavioral tests can be used to test hearing at any age and any developmental level.

Early identification of hearing loss leads to better speech, language and learning outcomes for children. Knowledge of the risk factors for late-onset hearing loss and continued vigilance in screening, monitoring and referral are vital. The goal is to insure that the listening and learning needs of all children are met.

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Supported by the National Institute on Deafness and Other Communication Disorders (NIDCD R25 DC006460-03)