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Audiology and Speech

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Women's and Children's Health

Fast Facts

Early Hearing Detection and Intervention

1924-2024

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The International Association of Communication Sciences and Disorders (IALP) is very proud to be associated with *Fast Facts: Early Hearing Detection and Intervention*. As an organization, we are committed to improving the quality of life of all individuals with disorders of communication, speech, language, voice, hearing and swallowing. This comprehensive resource highlights the importance of universal newborn hearing screening and early hearing detection and intervention services, providing invaluable information to help healthcare professionals improve the lives of children who are diagnosed as deaf or hard of hearing.

Fast Facts for Healthcare Professionals

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Early Hearing Detection and Intervention

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Fast Facts: Early Hearing Detection and Intervention
First published 2023

S. Karger Publishers Ltd, Elizabeth House, Queen Street, Abingdon,
Oxford OX14 3LN, UK
Tel: +44 (0)1235 523233

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A CIP record for this title is available from the British Library.

ISBN 978-3-318-06736-1

Yoshinaga-Itano C (Christine)
Fast Facts: Early Hearing Detection and Intervention/
Christine Yoshinaga-Itano

Medical illustrations by Graeme Chambers, Belfast, UK.
Typesetting by Amnet, Chennai, India.
Printed in the UK with Xpedient Print.

Audiology and Speech

Women's and Children's Health

13 Unilateral hearing loss and auditory neuropathy spectrum disorder

Allison L Sedey, Mallene Wiggin, Hsiu-wen Chang and Christine Yoshinaga-Itano

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Unilateral hearing loss

UHL is defined as the presence of any type and degree of hearing loss in one ear with normal hearing in the other ear. The potential impact of hearing loss in 'just' one ear is often minimized by both medical professionals and laypersons, with the common misconception that children with UHL will experience little to no listening and/or communication difficulties or delays because they 'have one good ear'. Consequently, this population may receive inadequate medical, audiological and/or educational management.

However, the significant negative impact of UHL is well documented,[1–20] with consequences including deficits and/or delays in speech perception,[1–3] neurological development,[4,5] cognitive function,[6–8] socioemotional skills[9] and language acquisition.[3,10,11] Additionally, children with UHL are at risk for bilateral hearing loss resulting from either recurrent or persistent MEF in the normal-hearing ear and/or the development of PHL in the normal-hearing ear over time.[21–23]

Prevalence. Approximately 1 in 1000 newborns is identified with UHL, with estimates ranging from 0.8 to 2.7 per 1000.[24–27] Differences in prevalence rates may, in part, be attributed to variations in testing protocols, the testing equipment employed and the definition of hearing loss, as well as to regional variations in genetic and environmental risk factors.

Etiologies and risk factors. Although the cause of congenital and early-onset UHL is often unknown, a variety of conditions are associated with it, and additional factors place a child at high risk for UHL (Table 13.1).

Auditory sequelae. The processes involved in hearing with two ears (for example, head shadow effect, spatial release from masking and binaural summation) confer a variety of benefits that are unavailable or diminished for a child with UHL. Thus, this population experiences difficulties with:

- localizing where sound is coming from (which creates significant safety concerns)[1]

- understanding speech in noisy and/or reverberant environments[1–3]
- hearing speech originating on the side of the affected ear[28]
- following conversations that involve multiple people.[28]

For children with UHL, listening is more effortful, and they experience listening fatigue similar to children with bilateral hearing

TABLE 13.1

Common causes of UHL in childhood and additional risk factors

Common causes of UHL
• Structural abnormalities of the outer or middle ear
– Atresia
– Ossicular malformations
• Inner ear malformations
– Cochlear dysplasia
– Enlarged vestibular aqueduct
– Auditory nerve absence
– Auditory nerve deficiencies
• Genetic
– Associated with a syndrome such as Waardenburg, CHARGE, Stickler
– Non-syndromic
• cCMV infection
• Meningitis
• Chronic middle ear effusion
• Ototoxic medication
Additional risk factors for UHL
• Admission to the NICU
• Family history of hearing loss
• Craniofacial anomalies
– Cleft palate
– Microcephaly
– Microtia
– Temporal bone anomaly

loss and significantly greater than for children with normal hearing.[29] Taken together, these wide-ranging listening challenges contribute to significantly poorer speech recognition in both quiet and noisy environments.[1,3]

Neurological sequelae. In addition to difficulties in auditory/speech perception, structural neurological differences – both in the auditory area and other brain structures – have been associated with UHL in both pediatric and adult populations.[4] These differences extend to both cortical growth and synaptic development. Additionally, multiple functional connectivity differences between brain networks involved with cognition, language comprehension and executive function have been identified in children with UHL.[5]

Language and other developmental outcomes. An increasing body of evidence supports the presence of early language delays in many children with UHL[10,11,16–18] that persist through adolescence.[13,14] Scores that fall below age expectations have been noted in a variety of language areas including vocabulary,[10,11,15] morphology and syntax,[15] narrative skills,[20] verbal reasoning[8,14] and language comprehension.[13]

Infancy to preschool. Language challenges in very young children with UHL have been documented, with 41% of a sample population demonstrating delays in preverbal vocalizations[16] and average delays of 5 months noted in producing two-word phrases.[17] In studies of toddlers and preschool-age children with UHL, 25–35% were consistently reported to exhibit significant delays in both receptive and expressive language, even when hearing loss was identified early.[10,11,18] These delays appear to be most pronounced from 24 months of age.[10]

School-age children. Studies of school-age children indicate that, even by adolescence, delays in language persist.[13,14] Similar to the percentage of young children with UHL who demonstrate language delays, 25–40% of school-age children present with academic difficulties as evidenced by being retained a grade in school and/or requiring special education support.[13,14,30–32] A recent study of children from third to tenth grade found that, even after the advent of newborn hearing screening, fewer than 50% of children with UHL

scored in the proficient range on a state-wide literacy test. This result was similar to that of children with mild-to-moderate bilateral hearing loss.[19]

Cognitive-linguistic skills and socioemotional wellbeing. In addition to delays in general expressive/receptive language skills and academic development, on average, children with UHL demonstrate significantly lower scores on verbal, full-scale and/or performance intelligence quotient tests.[3,7,8] Reduced accuracy and efficiency in phonological processing and verbal working memory have also been noted.[6] Especially concerning are reports that children with UHL are at risk for social and emotional problems and demonstrate lower quality-of-life scores than hearing peers and peers with bilateral hearing loss.[9]

Factors predictive of language and academic outcomes. Attempts to identify which children with UHL are most likely to exhibit language and/or academic delays have yielded conflicting results. In several studies, children with more significant degrees of hearing loss in the affected ear were more likely to have listening, language and/or educational difficulties.[2,3,11,31] More recent studies have found no association between language outcomes and degree of hearing loss in the affected ear.[10,18] Several older studies found right-ear involvement to be more detrimental to academic achievement than left-ear involvement;[31–33] however, several others more recently reported no ear-specific differences in language measures.[3,10,11,13] Lower levels of maternal education were predictive of poorer language outcomes in one study,[10] but not another.[18] Thus, it is difficult to determine what factors may put a child with UHL at increased risk for language and academic delays.

Treatment/intervention options. Although it is still unclear which intervention practices – in terms of both amplification technology and early intervention practices – are most effective in minimizing the negative sequelae of UHL, intervention in cases of early childhood UHL should include a combination of amplification technology, optimization of the listening environment and FCEI (see Chapter 12). For optimal outcomes, these interventions should be started as early as possible – ideally by or before 6 months of age.[34]

Depending on the type and degree of hearing loss, the following types of hearing technology should be considered:

- conventional hearing aid on the affected ear
- cochlear implant in the affected ear
- contralateral routing of signal amplification system in which a transmitter and microphone is worn on the affected ear and sound is routed to the normal-hearing ear
- bone conduction hearing device
- FM/DM system in which sound is transmitted wirelessly from a speaker's microphone directly to the listener's (typically, normal-hearing) ear.

The listening environment for infants and toddlers with UHL can be optimized by minimizing background noise, reducing the distance from the speaker and directing speech to the affected ear.

FCEI should include encouraging people in the child's environment to use language-enhancing communication strategies such as increasing the quantity and quality of the language input to the child, using communication strategies such as imitation and expansion of the child's utterances and narrating the child's and adult's actions.

Providing optimal care. The first step in creating an effective system of care for children with UHL is acknowledging the significant challenges they face. A multidisciplinary team approach is critical given the wide-ranging issues (medical, hearing, cognitive, language, academic and socioemotional) faced by many children with UHL.[1–20] Prompt medical management of middle ear effusion is important to avoid UHL becoming a temporary, but possibly long-term, bilateral loss. Additionally, physicians can provide information and possibly treatment of outer and middle ear anomalies. As the anchor point of a child's medical care, pediatricians are in an excellent position to make referrals to other members of the care team including audiologists, ophthalmologists (given that vision pathologies commonly co-occur with hearing loss),[35,36] speech pathologists and early interventionists.

Audiological care should include regular hearing tests to monitor transient hearing loss due to middle ear effusion and/or permanent progression of hearing loss in the normal-hearing ear, something that has been reported to occur in 7–17% of children with UHL.[21–23] Early consideration and timely fitting of an amplification device, if

appropriate, may minimize some of the negative consequences of UHL.[37–39] Given reports of progressive hearing loss in the affected ear in 21–38% of cases,[21,22] audiological monitoring is critical in determining potentially shifting amplification needs.

Early intervention that is pre-emptive (rather than initiated only after a child demonstrates delays) should begin by 6 months of age to maximize language outcomes.[34] The intervention program should include deafness professionals, parent-to-parent support and involvement of adults whose lived experience includes UHL. For families who opt out of intervention or in areas where children with UHL are not automatically eligible for early intervention services, regular (every 6 months) monitoring of language and other developmental skills using standardized, norm-referenced assessments is critical to identify and address delays in a timely fashion.

Auditory neuropathy spectrum disorder

Auditory neuropathy is a clinical diagnosis used to describe individuals with auditory disorders resulting from dysfunction of the synapse of the inner hair cells and auditory nerve and/or the auditory nerve itself. Children with ANSD can be very different from one another because of the multiple potential sites where lesions can arise. As such, they require specialized procedures for identification and for treatment as habilitation/rehabilitation strategies can be successful but may differ according to the location of the damage to the hearing system.[40] The percentage of the population of children with ANSD that have successful developmental outcomes is as yet unknown.

Prevalence of ANSD ranges from 1–10% of children diagnosed with hearing loss dependent upon the population sampled and the heterogeneity of clinical profiles.[40–45] A higher prevalence of SNHL and ANSD is seen in infants in NICUs,[41–45] and ANSD accounts for up to 30% of SNHL in NICUs. The incidence of ANSD is higher in NICUs than in well-baby clinics.[40,45]

Characteristics of the hearing loss. Bilateral ANSD is found in about 75–79% of affected infants and unilateral ANSD is found in the remaining 21–25%.[45–47] Hearing thresholds can range from

thresholds in the normal range to total hearing loss.[44–50] Hearing thresholds frequently fluctuate – there can be over 40 dB variability – and fluctuations are more common in children than in adults.[44–50] Slow deterioration over time at high and mid frequencies has also been reported. Spontaneous improvement may occur when ANSD is associated with anoxia and hyperbilirubinemia in the newborn period.[44–50]

Children with ANSD typically have very poor speech discrimination, even with preserved hearing thresholds, and background noise can further deteriorate residual speech discrimination in children with ANSD.[44–50]

Etiology. The pattern of normal outer hair cell function combined with abnormal neural responses shown by ABR testing places the site of auditory neuropathy to the area of the ear including the inner hair cells, the connections between the inner hair cells and the cochlear branch of the eighth cranial nerve, the eighth cranial nerve itself, and potentially the auditory pathways of the brainstem.[51–53] Neural problems may be axonal or demyelinating and afferent as well as efferent pathways may be involved.

A range of risk factors for ANSD has been identified (Table 13.2).[40]

TABLE 13.2

Risk factors for ANSD

Risk factor	% of cases identified in
Presence of hyperbilirubinemia associated with severe jaundice in the newborn period	50–73%
Prematurity	30–46%
Ototoxic drug exposure	41–80%
Family history of hearing loss	36–38%
Mechanical ventilation	36%
Cerebral palsy	9–15%

Up to 40% of ANSD cases are reported to have a genetic etiology (Table 13.3).[45,54] The problem might also be related to a biochemical abnormality involving neurotransmitter release. The percentages of the population that have specific etiologic causes, whether genetic or environmental factors in utero or after birth, has not been determined.

Structural abnormalities seen in ANSD. Gardner-Barry[55] reported structural abnormalities in 43% of 142 children younger than 10 years seen at the Sydney Cochlear Implant Center in Australia. Of these, 16% had abnormalities on their CT scans, including Mondini deformities, wide internal auditory meatus, dysplastic apical turn, and abnormal vestibule and lateral semicircular canals. Compromised auditory nerves were seen in 20% of the bilateral and 6% of the unilateral ANSD cases.

TABLE 13.3

Genetic causes of ANSD

Non-syndromic
• Non-syndromic autosomal dominant (*AUNA1*, *PCDH9*)
• Non-syndromic autosomal recessive (*OTOF/DFNB9*, Pejvakin [*DFNB59*], *GJB2*)
• X-linked (*AUNX1*)
Syndromic
• Hereditary sensory-motor neuropathy-Lom (autosomal dominant, autosomal recessive, X-linked)
• Leber's hereditary optic neuropathy (mitochondrial)
• Autosomal dominant optic atrophy
• Autosomal recessive optic atrophy
• Mohr–Tranebjaerg syndrome (X-linked recessive)
• Fredreich's ataxia (autosomal recessive)
• Refsum's disease (autosomal recessive)

Teagle[56] reported the preimplant imaging results of 48 children with ANSD and found 23 different abnormalities in 38% of those assessed, the most common of which were:

- periventricular leukomalacia (15%)
- cochlear nerve deficiency in at least one ear (19%)
- Dandy–Walker malformation (4%)
- severe inner ear malformation, including cochlear hypoplasia (6%) and Arnold Chiari type II malformation (2%)
- optoinfundibular dysplasia (2%).

Diagnosis. Most newborn hearing screening programs use both OAE and aABR to test a baby's hearing. If infants are screened with OAE technology first, or if programs use only OAE screening, children with ANSD will pass the hearing screen and remain unidentified.[40]

To ensure identification of infants with ANSD, aABR screening should always be used in NICUs. Screening with aABR should also be carried out in well-baby clinics.

Children with ANSD are characterized by:

- an absent or abnormal middle ear muscle reflex
- present OAEs
- absent or abnormal ABR thresholds
- presence of a polarity-reversing cochlear microphonic followed by typically absent or significantly aberrant wave forms.

Middle ear muscle reflex tests how well the ear responds to loud sounds. In a healthy ear, loud sounds trigger a reflex and cause the muscles in the middle ear to contract. Infants with ANSD require much louder sounds to trigger a reflex.

Otoacoustic emissions testing measures how well the outer hair cells in the cochlea function. With ANSD, the OAE response is normal.

Auditory brainstem response testing measures whether the auditory nerve transmits sound from the inner ear to the lower part of the brain and how loud sounds have to be for the brain to detect them. With ANSD, there is an absent or abnormal response. Diagnostic ABR should include a search for a cochlear microphonic.

If an ANSD diagnosis is made, additional tests are required, including:[40]

- otologic evaluation with imaging of the cochlea and auditory nerve (CT and MRI) to see if the auditory nerve is present in both ears and if there are any inner ear abnormalities
- genetic testing to determine cause and, if necessary, provide appropriate treatments
- neurological testing by a neurologist to assess peripheral and cranial nerve function
- ongoing communication assessment to monitor speech and language development, evaluate the effectiveness of treatment and modify it when appropriate
- ophthalmology assessment to determine if there is any associated vision loss.

Auditory neuropathy and comorbidities. Due to the high incidence of ANSD in the NICU, ANSD may coexist or be associated with a multitude of other disorders that may impact communication development, including cognitive, neurological and perceptual disorders, speech-motor disabilities or visual disabilities. Socioemotional disorders, such as autism, are also seen.

The prevalence of these comorbidities in children with ANSD has not been well studied but Uhler et al.[46] reported that 57% of children with ANSD had hearing loss with additional disabilities. Significant cognitive delays were about twice to three times more common in children with bilateral ANSD than in children with bilateral SNHL.

Treatment/intervention options. The therapeutic interventions that are most effective for children with ANSD with specific characteristics are not yet clear, but some general therapeutic approaches include:[40,46,49]

- providing signal-to-noise maximization with FM listening devices
- amplification via hearing aids and cochlear implants
- speech/language intervention, which may include both auditory and visual communication strategies (for example, cued speech, sign language).

Key points – unilateral hearing loss and auditory nerve spectrum disorder

- UHL affects approximately 1 in 1000 newborns and has a significant negative impact on speech perception, neurological development, cognitive function, socioemotional skills and language acquisition.
- There is conflicting evidence on which factors predict delays in language and academic development in children with UHL.
- Early intervention, ideally by or before 6 months of age, is important for optimal outcomes in children with UHL.
- ANSD is a result of damage to the inner row of hair cells in the ear, or to the synapses between the hair cells and the auditory nerve, or damage to the auditory nerve itself.
- Infants with ANSD can be missed if only OAE testing is used for screening.
- ANSD can be associated with multiple comorbidities, including cognitive, neurological and perceptual disorders, speech-motor disabilities, visual disabilities and socioemotional disorders.

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