Management of Children with Unilateral Hearing Loss

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KEYWORDS

• Unilateral hearing loss • Speech-language development • Children • Amplification • Quality of life

KEY POINTS

- Lack of binaural input results in difficulties with sound localization and understanding speech in noisy backgrounds.
- Children with unilateral hearing loss (UHL) are at risk for speech-language delay, poor academic performance, and decreased quality of life compared with children with normal hearing.
- As yet, no study has shown any specific intervention that can definitely mitigate the negative effects of UHL.
- Because evidence-based guidelines are lacking, individual patient and family needs and preferences must be considered to recommend interventions for children with UHL.
- Interventions may include preferential seating in class, individualized education program (IEP) or 504(c) plan, frequency-modulated (FM) systems, amplification devices (hearing aids or contralateral routing of signal [CROS] devices), osseointegrated bone conduction hearing devices, or cochlear implantation, tailored to fit the needs of the child and to set the child up for success in academic and social settings.

INTRODUCTION AND BACKGROUND

Epidemiology

The prevalence of UHL is estimated at 1 per 1000 children at birth,[1] increasing with age due to delayed-onset congenital hearing loss and acquired hearing loss. Because the prevalence of UHL can vary significantly according to the definition applied, it is useful to consider that at least one-third of all children born with hearing loss have UHL; epidemiologically, an estimated 3% to 6% of school-aged children in the United States have UHL.[2] By adolescence, the prevalence of UHL is as high as 14.0% if

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Abbreviations

BHL	Bilateral hearing loss
CI	Cochlear implantation
CT	Computed tomography
FM	Frequency modulated
IEP	Individualized educational program
QOL	Quality of life
UHL	Unilateral hearing loss

thresholds greater than 15 dB are considered and 2.7% if only thresholds 25 dB or higher are considered.[3] Furthermore, approximately 10% of children born with UHL eventually develop bilateral hearing loss (BHL).[4,5]

Etiology and Evaluation

The incidence of temporal bone anomalies in congenital UHL is high compared with congenital BHL. Enlarged vestibular aqueduct (EVA) and cochlear nerve aplasia or hypoplasia are increasingly identified using high-resolution computed tomography (CT) and MRI. Among children with severe to profound UHL, the prevalence of cochlear nerve aplasia or hypoplasia approaches 50%.[6,7] Other temporal bone abnormalities commonly reported include enlarged vestibular aqueduct, Mondini deformity, cochlear and vestibular malformations, and common cavity malformation, although the likelihood of identifying an abnormality may depend on the severity of UHL, with more severe losses associated with a greater percentage of anatomic abnormalities.[5,8,9] Although genetic causes are predominant for BHL, this is not true for UHL. Investigators have identified variants of genes associated with BHL, including Pendred syndrome (SLC26A4) associated with EVA, but have not found them to be major determinants of UHL.[8,10,11] Although families with sensorineural UHL have been reported, the genetic mutations associated with them have not been identified.[12–14] Other syndromic causes of childhood hearing loss may initially present, or simply be associated with, a unilateral loss, for example, branchio-otorenal syndrome and Waardenburg syndrome.

Other important causes of sensorineural UHL include congenital cytomegalovirus (CMV) infections, meningitis, and trauma. Although children with symptomatic congenital CMV infection are more likely to have BHL, children with asymptomatic CMV infection are more likely to have UHL.[15] Congenital mumps and measles are infrequent in the developed world because of childhood immunization schedules but should be considered for families who choose not to immunize their children or for children adopted without clear immunization history. Temporal bone trauma, as a result of motor vehicle accidents, falls, or other head trauma, is a common cause of acquired postlingual UHL.

Important causes of conductive UHL include unilateral aural atresia, cholesteatoma, chronic otitis media, otosclerosis, ossicular discontinuity, and congenital ossicular malformations. Otitis media, labyrinthitis, and cholesteatoma are possible causes for UHL, usually mild to moderate in severity, diagnosed on physical examination. Hearing loss associated with these entities must be evaluated by audiogram. Congenital ossicular malformations can be identified on high-resolution temporal bone CT scans, and otosclerosis or ossicular discontinuity can be verified at the time of a middle ear exploration or planned stapedectomy or ossicular reconstruction.

As noted in the diagnostic algorithm suggested by Preciado and colleagues,[16] temporal bone imaging is the most likely test that reveals a cause in children with UHL.