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Review article

Management of unilateral hearing loss

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ABSTRACT

Objective: A representative sample of literature regarding unilateral hearing loss (UHL) was reviewed to provide evidence of the effects of UHL and the intervention options available for children with UHL. Considerations during the assessment and management of children with UHL are illustrated using case illustrations.
Method: Research articles published from 2013 to 2015 were searched in the PubMed database using the keywords "unilateral hearing loss". Articles from 1950 to 2013 were included from a previous literature review on minimal hearing loss [1]. A retrospective review of charts of 14 children with UHL was also conducted.
Results: The evidence indicates that children with UHL are more likely to have structural anomalies of the inner ear; may face challenges in six different domains, and have six intervention options available. Evidence also indicates that although some children appear to exhibit no delays or difficulties, others have significant challenges, some of which continue into adulthood.
Conclusions: Children with UHL have to be treated on a case-by-case basis. Parent education regarding UHL, its effects, and all available management options is critical so they can make informed decisions. Close monitoring and good communication between professionals in different domains is crucial in order to minimize the potential negative effects of UHL.

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1. Introduction

Unilateral hearing loss (UHL) is defined as hearing loss in one ear. The most widely used definition of pediatric UHL is when the air conduction pure tone average is ≥20 dB HL in the impaired ear, with no significant air-bone gap, and hearing thresholds in the normal-hearing (NH) ear are ≤15 dB HL [2], and was used in most of the literature as well as in the case illustrations. The conventional view regarding UHL was that it did not have a significant negative impact, and the child would develop typically with no particular challenges [3]. However, interest in UHL peaked in the 1980s with reports indicating that children with UHL do demonstrate academic, social, and behavioral problems more frequently than their peers [4–10].

Prevalence rates of UHL estimated in school-aged children range from 30 to 56 per 1000 [2,11]. Incidence rates of UHL among newborns are reported to be 0.83 to 2.7 per 1000 births [12,13]. The most recent nationwide data available in the USA indicate that 36% of infants identified with hearing loss in 2013 had UHL [14]. Although incidence and prevalence rates can vary significantly based on many factors [15], approximately 22–36% of infants identified with hearing loss each year from 2005 to 2013 in the USA have UHL [14].

In the 1980s and 1990s, most children with UHL were identified when they were school-aged. In one of the earliest reports of children with UHL in 1986, only 23% of children with UHL were identified before the age of 5 years [16]. The average age of identification of UHL has been decreasing over the years and has been reported to be 8.78 years in 1991 [17], 6.9 years in 1998 [18] and 4:11 years in 2003 [19]. The mean age of identification of UHL further decreased after the advent of universal newborn hearing screening (UNHS) programs: from 4.4 years to 2.6 years [20], and for mild bilateral and UHL from 5 years to 0.8 years [21] by the year 2014. Thus, in current clinical practice, infants are being identified with UHL at very young ages. However, despite early identification, there are no clear guidelines regarding the management of UHL in infants and young children and clinicians continue to be uncertain when making decisions regarding management options for this population.

The etiology of UHL varies, and as with bilateral hearing losses, is not always determined. Although the largest proportion of subjects has UHL of unknown etiology, causes include heredity, head trauma, viral causes (mumps, rubella), meningitis, and fetal alcohol syndrome [17,22]. A specific investigation of etiology of UHL found that 59% of children with UHL had a family history of hearing loss, with 26% specifically reporting a family history of UHL. Although less than half the subject sample had temporal bone imaging, 30% of those that did had structural anomalies including enlarged vestibular aqueducts, Mondini dysplasia and common cavity deformity [23].

Although the evidence regarding UHL has evolved considerably over the past three decades, children with UHL have been prevented from being referred to early intervention services due to a variety of misconceptions including: “one ear is good enough” and “there is not enough known about what to do; therefore, it is better to do nothing” [24]. Further, parents of children with UHL (42% of the subject sample) reported that they had received professional advice that hearing aids and assistive devices would not help, although a far smaller proportion of the sample (8%) indicated that the hearing loss was too severe for hearing aids [25].

Review articles on UHL have been published in recent years; however, they have been specific to certain aspects of UHL such as academic performance [26], language disorders [27], cochlear implantation [28–30], bone-anchored hearing systems [31], or geared to other professionals [32]. The purpose of this article is to provide a broad review of representative literature on UHL including 1) the effects of UHL on various domains of child development; 2) considerations during the assessment of children with UHL using case illustrations; and 3) intervention options for children with UHL, with the goal of reducing uncertainty and providing a resource to help clinicians make appropriate clinical decisions in the management of children with UHL.

2. Method

Articles for this review were selected based on a PubMed database search using the key words “unilateral hearing loss”. All articles published between 2013 and 2015 were reviewed for relevance. Articles published prior to 2013 were included from a previous literature review conducted on minimal hearing loss that included a search from 1950 to 2013 [1]. Content from the articles was broadly divided into two main categories: impact of UHL and intervention options for UHL, with smaller sections on the etiology (imaging studies) and functional assessment measures. Additionally, charts of 14 children with UHL evaluated by the authors were reviewed and data were gathered regarding age of identification of UHL, assessment procedures and age at hearing aid fitting in order to include case examples to illustrate considerations during the assessment of and intervention for children with UHL.

3. Results

3.1. Imaging

Imaging is used to identify the etiology of hearing loss due to a structural anomaly and is recommended by the Joint Committee on Infant Hearing (JCIH) in the absence of medical or genetic causes [33]. While the etiology of hearing loss may not be determined in 30–40% of infants with hearing loss [33], structural anomalies of the inner ear are much more common in children with UHL (50%) than bilateral HL (5%) [34]. Recent studies have found that 32–67% of UHL ears have aberrant inner ear anatomy [35–40]. The bony cochlear nerve canal (BCNC) width has been found to be significantly smaller in children with UHL [40–42], and smaller BCNCs are