

# **PEDIATRIC Sensorineural Hearing Loss**

**Clinical Diagnosis and Management**



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# Introduction

## WHY DO WE NEED A HANDBOOK ON PEDIATRIC HEARING LOSS?

Landmark changes in the past 30 years have altered the way we identify, diagnose, evaluate, and treat children with hearing loss. Newborn hearing screening, advocated by the National Institute on Deafness and Other Communication Disorders panel in 1993, has made infant detection of hearing loss the norm rather than the exception. Genetic research and advances in imaging have opened up a world of understanding of the etiology of hearing loss. Technologic advances in amplification devices—in the form of cochlear implants, bone-anchored and digital hearing aids—have allowed parents to expect that their children with hearing loss will function independently in a hearing society. Research in cognition has helped us to understand that we actually hear with our brains, and that hearing function can affect cognitive processes as well as speech and language. However, our ability to diagnose hearing loss in children outstrips our knowledge about what to do about it—when to intervene, how to intervene, how aggressive we should be, and what outcomes should be measured. Pediatric hearing loss is an incredibly complex topic replete with controversies, evolving research findings, and subtle differences in management and diagnosis with different types of hearing loss.

Thus, we have compiled the chapters in this book to help pediatricians, residents, audiologists, and others who deal with pediatric patients to distill the breadth of knowledge on this topic into

one that is manageable and easily comprehensible. We intend that this manual will address clinical questions that arise in daily practice of pediatricians, audiologists, or otolaryngologists, and assist residents in preparation for in-service training exams. In addition, it can serve as a valuable teaching tool for students and residents in the clinic and on the floors. All three editors are clinicians, teachers, and researchers who have watched and participated in the evolving debates about best practices, and appreciate that the available literature is overwhelming, conflicting, and difficult to synthesize.

We have arranged this manual into segments that progress from the identification of hearing loss and how to diagnose it (newborn hearing screening, audiology, and presentation), to the rationale for why to intervene (functional consequences of hearing loss), then to evaluating the etiology of the hearing loss (imaging, evaluation of hearing loss), how to manage pediatric hearing loss (management of hearing loss, sudden sensorineural hearing loss), and ending with what we have yet to learn (questions with no answers). Along the way, bits of history will illuminate how the current state of knowledge about pediatric hearing loss has evolved into its current iteration of options, preferences, debates, and questions.

We hope that this manual will serve as a great resource for clinicians who encounter and treat children with pediatric sensorineural hearing loss.

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# **PART I**

## *Diagnosis and Presentation*





## CHAPTER 1

# Functional Consequences of Hearing Loss: What's Down Can Come UP!

*Donald M. Goldberg*

**T**he landscape of hearing loss continues to change, with truly exciting and limitless potential for newborns, infants, toddlers, and children with hearing loss. With the adoption of Universal Newborn Hearing Screening (UNHS) programs, children with hearing loss are being identified at much younger ages.<sup>1</sup> Three major interventions have resulted in impressive outcomes for many of these children:

1. the prompt fitting of personal hearing aids<sup>2</sup> and frequency modulated (FM), infrared (IR), or remote microphone (RM) systems;
2. the implementation of early intervention programs that emphasize the parents' role as a child's first and most important teacher<sup>3</sup>; and
3. for children for whom personal hearing aids do not amplify the entire speech spectrum sufficiently or adequately, cochlear implants (CIs) have become an effective option, especially for the youngest patients we serve.<sup>4,5</sup>

The purpose of this chapter is to provide an overview of the consequences and the impact of hearing loss on the speech, language, educational, and other developmental aspects of a child with sensorineural hearing loss (SNHL). The author contends that no one communication opportunity<sup>6,7</sup> is the answer for all families, nor is one specific hearing sensory technology the key, but nonetheless, the possibility for typical speech and language function is truly reachable for more children

with SNHL. High expectations for highly intelligible and age-appropriate spoken language<sup>8</sup> and educational success for these children are more achievable than ever before. So after some introductory baseline facts, this chapter will discuss what difficulties children with SNHL encounter and briefly address how many of the negatives can be ameliorated.

## THE INVISIBLE DISABILITY

One of the greatest challenges for children with hearing loss is the invisible nature of the disability. With the exception of children with syndromic etiologies, most children with hearing loss physically appear completely typical to someone who is unfamiliar with them. In fact, they are *typical*—they are children first, and “Yes, by the way, they have a hearing loss in one or both of their ears!”

An important consideration about the newborns tested and identified as having permanent hearing loss is that approximately 95% of their parents have completely typical hearing.<sup>9</sup> Therefore, hearing loss is a foreign entity for most families. Although parents and the general public may assume all children with hearing loss must learn to use sign language, it is critical for the families to learn that a variety of technology options exist, and a team of providers will help support and serve their child and family. The team members include early-intervention specialists, physicians, audiologists, speech-language pathologists, teachers of the deaf/educators of the deaf and hearing impaired, social workers, and occupational and physical therapists.

At the same time, these children are at risk for a variety of communication, educational, and other challenges. Therefore, service providers working with these children and their families must be well

aware of the possible sequelae of childhood hearing loss. This knowledge must be balanced by the understanding that with early detection and identification, early intervention, and excellent comprehensive management/treatment and follow-up in the areas of communication, audiology, and medical and otologic care, these children can achieve well beyond what was expected in the past.

## VARIABLES ASSOCIATED WITH CHILDHOOD HEARING LOSS

Children with hearing loss are an incredibly heterogeneous population. Multitudes of variables make research studies challenging with regard to matching subjects and/or comparing groups of patients due to the range of factors present. Some notable variables to be aware of include the following:

- ◆ Age of onset of hearing loss: congenital (present at birth); prelingual hearing loss (birth to approximately age 3 years); or postlingual (after the development of speech and language has been established).
- ◆ Age of diagnosis/identification.
- ◆ Type, severity, and etiology of hearing loss.
- ◆ Unilateral versus bilateral involvement.
- ◆ Age and consistency of use of hearing sensory technology.
- ◆ Benefit of hearing aid(s), cochlear implant(s), etc..
- ◆ Family investment and degree/amount of stimulation/input to child.
- ◆ Type of intervention services available.

- ◆ Presence of other or multiple disabilities<sup>10</sup>

## NEUROPLASTICITY

Carol Flexer and others have noted, “we hear with our brain, not our ears.”<sup>11</sup> The critical period for language stimulation is the first few years of life, based on longitudinal studies and radiographic studies of infants. Some of the critical findings of this work include research in the following:

- ◆ the area of auditory development<sup>12</sup>;
- ◆ outcomes of children whose hearing losses are identified early, who receive appropriately fit hearing technology<sup>13</sup>; and
- ◆ the outcomes of children who receive auditory-based early intervention with rich auditory/listening experiences, which stimulate the growth of auditory and language centers of the brain.

Pollack, Goldberg, and Caleffe-Schenck<sup>14</sup> and Roberts<sup>15</sup> have suggested that children need access to fundamental prelinguistic skills, and parents need to learn how best to engage in meaningful, stimulating, and interactive exchanges with their child. What appears to be most important to keep in mind with these early identified children is that they must be bathed in rich spoken and/or signed language.<sup>16</sup>

## SPEECH CONSEQUENCES OF SENSORINEURAL HEARING LOSS (SNHL)

The description of “deaf speech” has a regrettably long history.<sup>17–20</sup> Deaf speech is often characterized by speech and voice characteristics including the following:

- ◆ Inadequate breath control.
- ◆ Excessive and inappropriate pausing.
- ◆ Inappropriate pitch register.
- ◆ Hypernasality.
- ◆ Excessive tenseness or harshness.
- ◆ Inappropriate duration.
- ◆ Inappropriate syllable stress.
- ◆ Inappropriate prosody.
- ◆ Voice/voiceless confusion.
- ◆ Vowel neutralization.
- ◆ Consonant sound substitutions/distortions and final consonant deletions.

For many children today, early identification and the timely provision of advanced hearing sensory technology with early intervention services have made *remedial* speech training infrequent. The potential for high speech intelligibility is common.

Assessment of the speech sound or phonetic repertoire of children with hearing loss is required along with evaluation of the speech intelligibility of the speaker. Screening for *oral motor function*, often described as an *oral peripheral examination or screening*, should be completed. The form and function of the primary speech sound articulator—the tongue—should be evaluated for structure and function, specifically, the ease of movement up, down, and to each side and around. With appropriate lighting, the palatal arch should be examined, along with movements of the lips (closure and lip rounding). Parents should be queried regarding the status of the child’s dentition, along with probes of the parents regarding the child’s history of nursing, sucking, feeding, excessive drooling, and swallowing status.

Historically, many speech and language measures were developed for children with hearing loss, but today, it is estimated that approximately 80% of children

**Table 1–1.** Speech Tests

Developed for children with hearing loss	Ling Phonetic Level Evaluation (Ling, 1976, 2002) <sup>22</sup>
	CID Phonetic Inventory (Moog, 1988) <sup>23</sup>
	CID Picture SPINE (Monsen, Moog, & Geers, 1983) <sup>24</sup>
	Identifying Early Phonological Needs in Children with Hearing Loss (Paden & Brown, 1992) <sup>25</sup>
For children/individuals with typical hearing status	Goldman-Fristoe Test of Articulation-3 (GFTA-3) (Goldman & Fristoe, 2015) <sup>26</sup>
	Structured Photographic Articulation Test-III featuring Dudsberry (SPAT-D III) (Tattersall & Dawson, 2016) <sup>27</sup>
	Arizona Articulation Proficiency Scale-3 (Fudala, 2000) <sup>28</sup>
	Clinical Assessment of Articulation & Phonology (Secord & Donohue, 2013) <sup>29</sup>

with hearing loss are mainstreamed or included in today’s public school programs.<sup>21</sup> Thus, tests for children with typical hearing are appropriate tools and protocols for assessment/evaluation. Table 1–1 provides a listing of some commonly administered “speech” tests in use today.

**LANGUAGE CONSEQUENCES OF SNHL**

Arguably the most important functional component of communication for these patients is their spoken or expressive language and in comprehension or receptive language skills and abilities. The seminal work of Bloom and Lahey<sup>30</sup> and their model of the three language components remain the foundation of our understanding of language. These authors described the interaction of form, content, and use. *Form* can be broken down into the aspects of phonology, morphology, and syntax. *Content* refers to the semantics or “meaning” of language, with attention to seman-

tic relationships, for example “Agent + Action” such as “Mommy eat,” along with receptive and expressive vocabulary. Finally, language *use* is the pragmatics of language. Our social language addresses communicative intent, conversational turn-taking, and other social aspects of our communication.

Common language-specific deficits of children with hearing loss have been well documented.<sup>31</sup> Areas of delay or disorder due to hearing loss include vocabulary concerns, commonly attributed to the lack of “bathing” children in rich language, or incidental learning. Many children with hearing loss have the ability to understand concrete vocabulary (e.g., visible or touchable nouns, active verbs), but as words become more abstract, difficulties may be demonstrated (e.g., define or describe “dream”). Higher-level vocabulary challenges can appear with antonyms, homonyms, words with multiple meanings, and jokes and riddles, along with figurative language (e.g., idioms and similes).

In the area of form, *morphology* is impacted for many children with hearing loss. Potentially influenced by acoustics

and the difficulty with hearing final consonant sounds, specific morphemes may be difficult to hear and therefore may be missing in their spoken language (e.g., the /s/ and /z/ sounds used for plurality, possession, tense). Articles or “function” words—“the,” “a,” “an”—along with conjunctions “and” and “because” may be similarly dropped in oral communication for some.

The final language component—pragmatics—may also be adversely affected. If one considers the nuances of communication interactions, such as entering and exiting a conversation, appropriate pausing, and taking conversational turns, one should not be surprised to see social language deficits in some of these children. Nonverbal aspects of communication may be missed, such as the nuances of conversational exchanges and concepts of sarcasm, which are somewhat dependent upon excellence in hearing and communicative

competence. A related topic in the general area of language pragmatics is theory of mind. Westby described theory of mind (ToM) as “the ability to attribute mental states to yourself and others, and to understand that others have beliefs, desires, intentions, emotions and perspectives that are different from your own” and this likely explains “some of the pragmatic and social-cognitive difficulties,” including for children with hearing loss.<sup>32</sup>

Tests to evaluate language can include those developed for persons with hearing loss, but often tests standardized on children with typical hearing are being administered. If the ultimate goal is placement of the child with a SNHL with his/her hearing peers, making use of test protocols for hearing students would seem appropriate. Table 1–2 lists a variety of language tests for possible administration.

**Table 1–2.** Language Tests

Developed for children with hearing loss	Teacher Assessment of Spoken Language (TASL) (Moog & Biedenstein, 2010) <sup>33</sup>
	Cottage Acquisition Scales for Listening, Language, and Speech (CASLLS) (5th ed.) (Wilkes, 2010) <sup>34</sup>
For children with typical hearing	Child Development Inventory (CDI/Minnesota) (Ireton, 2005) <sup>35</sup>
	MacArthur-Bates Communicative Development Inventories: Words and Gestures (Fenson, Marchman, Thal, Dale, Reznick, & Bates, 2007a) <sup>36</sup>
	MacArthur-Bates CDI: Words and Sentences (Fenson et al., 2007b) <sup>37</sup>
	Receptive-Expressive Emergent Language-3 (REEL-3) (Bzoch, League, & Brown, 2003) <sup>38</sup>
	Expressive Vocabulary Test-2 (Williams, 2007) <sup>39</sup>
	Peabody Picture Vocabulary Test-4 (Dunn & Dunn, 2007) <sup>40</sup>
	Structured Photographic Expressive Language Test-Preschool 2 (SPELT-P2) (Dawson, Stout, Eyer, Tattersall, Fonkalsrud, & Croley, 2005) <sup>41</sup>
Structured Photographic Expressive Language Test-3 (SPELT-3) (Dawson, Stout, & Eyer, 2003) <sup>42</sup>	

*continues*

Table 1–2. *continued*


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Boehm Test of Basic Concepts 3—Preschool Version (Boehm, 2001) <sup>43</sup>
Boehm Test of Basic Concepts-3 (Boehm-3) (Boehm, 2000) <sup>44</sup>
Bracken Basic Concept Scale-Revised (Expressive/Receptive) (Bracken, 2006) <sup>45</sup>
Clinical Evaluation of Language Fundamentals—Preschool Version 2 (CELF-P-2) (Wiig, Semel, & Secord, 2004) <sup>46</sup>
Clinical Evaluation of Language Fundamentals-5 (CELF-5) (Wiig, Semel, & Secord, 2013) <sup>47</sup>
Preschool-Language Scale-5 (PLS-5) (Zimmerman, Steiner, & Pond, 2011) <sup>48</sup>
Comprehensive Assessment of Spoken Language 2 (CASL-II) (Carrow-Woolfolk, 2017) <sup>49</sup>
Oral and Written Language Scales-II (OWLS-II) (Carrow-Woolfolk, 2011) <sup>50</sup>
Test of Pragmatic Language-2 (TOPL-2) (Phelps-Terasaki & Phelps-Gunn, 2007) <sup>51</sup>

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## EDUCATIONAL IMPACT OF SNHL: LITERACY AND ACADEMIC PERFORMANCE

The educational impact of hearing loss on children is a minefield of controversy and debate due to contemporary advances in hearing habilitation. Historically, yearly reports from the Gallaudet Research Institute (GRI) and published in the April reference issue of the *American Annals of the Deaf*, suggested the limited academic behaviors of their deaf and hard of hearing population. Data from the 2008 report indicated that “for the 17-year-olds and the 18-year-olds in the deaf and hard of hearing student norming sample, the median Reading Comprehension subtest scores corresponds to about a 4.0 grade level for hearing students.”<sup>52</sup> Since 2008, yearly reporting has been removed, however, due to concerns about the data reported. Although studies have reported difficulties with emergent literacy skills,<sup>53</sup>

recent studies have suggested much more promising outcomes, including some reports of reading levels comparable to age-matched peers.<sup>54,55</sup>

The challenge in interpreting the available reading studies is that each is confounded by a variety of factors, including degree of hearing loss, age of onset, hearing technology history, etiology of deafness, parental involvement, and the communication and educational journeys the students have experienced. This is similarly apparent as one evaluates studies addressing the academic outcomes of school-aged children with hearing loss. General trends found across the commonly assessed areas of mathematics, oral language, reading, and written language, based on standardized tests of academic achievement, indicate a lower proportion of average and above-average ranges for children with hearing loss, compared to what is expected for children with normal hearing.<sup>56</sup> Again, the range of vari-