Deaf and Hard of Hearing

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Topic

Reduced hearing sensitivity cannot be perceived as a singular disorder. It does not result in a particular set of factors that impact functioning. Rather, the influence of reduced hearing sensitivity depends on its etiology, characteristics, timing, and the role that these all play in an individual's development. For example, a child with congenital profound deafness that limits access to spoken language will be largely shaped by the condition, with impact on educational, social, and familial functioning. An older adult with an age-related progressive hearing loss will not have been influenced by hearing status throughout development but may experience emotional, social, and familial effects associated with more limited ability to communicate.

In summary, physiological, developmental, and environmental factors are significantly shaped by a person's hearing status. Reduced hearing sensitivity alone may, but not necessarily, impact intellectual, neuropsychological, emotional, social, or behavioral functioning. Appropriate supports and accommodations that maximize access to language and communication can substantially mitigate the negative consequences that are sometimes associated with reduced hearing.

The role that hearing status plays for the patient, and understanding of that on the part of their health-care providers, will influence the interactions between them in significant ways. Knowledge of key concepts can help in the understanding of the nature of reduced hearing sensitivity, hearing loss, being deaf or hard of hearing, Deaf culture (defined below), and the role of hearing status:

A. ***Physiology***

Several physiological characteristics of hearing influence an individual's functioning and also inform specific types of needed interventions and accommodations. These include degree of hearing, site of hearing loss, time of onset, and benefit from assistive devices [1]:

1. **Degree of hearing loss**

Normal	0–20 dB
Mild	21–40 dB
Moderate	41–55 dB
Moderately severe	56–70 dB
Severe	71–90 dB
Profound	>90 dB

a. ***Mild to moderate range***. Most individuals who experience reduced hearing in the mild and moderate ranges are able to

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access sufficient auditory information so that their hearing does not significantly impact their ability to develop linguistic competence or to perform academically. Yet, even a mild hearing loss can make it difficult for individuals to attend, to alert to their surroundings, and to appreciate what is happening around them. A mild to moderate hearing loss can influence one's communication abilities and impact social relationships.

b. ***Severe and profound range***. Hearing status in the severe and profound ranges often limits access to spoken language and influences educational and communication options. Individuals with hearing in this range may communicate using a visual language (e.g., American Sign Language (ASL) as used in the USA and Canada or another formal signed language used elsewhere in the world). Alternatively, a person may have varying degrees of ability to use spoken language with the aid of hearing aids or cochlear implants.

2. **Cause of hearing loss**

Conductive hearing loss	Disruption of sound waves caused by a physical blockage, typically in the middle ear, that limits hearing. Most frequently, conductive hearing loss is temporary and can be caused by otitis media (ear infections), "fluid in the ears" as a result of a sinus infection, or excessive cerumen (earwax). In such cases, removal of the blockage can restore hearing. Anatomical anomalies can also result in conductive hearing loss, which may be more permanent
Sensorineural hearing loss	Structural alterations to the nerves in the inner ear, most commonly in the hair cells of the cochlea or the auditory nerve (i.e., VIII cranial nerve) resulting in reduced hearing. Sensorineural hearing loss is permanent and can be progressive (i.e., it can worsen over time). The majority of individuals who are considered deaf or hard of hearing have this type of hearing loss
Mixed hearing loss	Hearing loss that has both conductive (blockage) and sensorineural (nervous system) components is referred to as a mixed hearing loss
Auditory neuropathy/ auditory dyssynchrony	This type of hearing loss, often referred to as "AN," is a result of improper transmission of sound from the inner ear to the auditory nerve or to the brainstem. This type of hearing loss often results in inconsistent hearing abilities, with periods of normal or near-normal hearing and periods of significant loss, making access to sound highly unpredictable

3. **Time of onset**

The timing of the onset of reduced hearing has significant implications for the individual's functioning and life experience. Typically, onset is characterized as *congenital*, *prelingual*, or *postlingual*, referring to whether the reduced hearing is present at birth, before a child has developed spoken language or after spoken language has been established. Congenital and prelingually acquired hearing loss may interfere with an individual's ability to develop spoken language abilities. Hearing loss that is acquired postlingually is less likely to have as significant of an impact of the development of spoken language skills. Once a child has developed a solid foundation in spoken language, experiencing a reduction in hearing abilities will not necessarily inhibit further development of speech and language.

4. **Benefit from assistive devices** [2]:

a. ***Hearing aids***. Many individuals with mild and moderate levels of hearing are able to access auditory information with the use of hearing aids. Typically, these amplify sound, but do not necessarily clarify the sounds that are heard. For many people, hearing aids allow for some, but not perfect, understanding of what is happening in their environment and can reduce the social isolation that can accompany progressive hearing loss.

b. ***Cochlear implants***. Individuals with more profound levels of reduced hearing may qualify for a cochlear implant. This is a device with electrodes that

are inserted directly into the cochlea that sends an electrical impulse to the auditory nerve. In the USA, candidacy for cochlear implant surgery is largely determined by the Federal Drug Administration (FDA). New technologies, such as hybrid cochlear implants, which function as a cochlear implant in the frequency ranges in which a person has a profound hearing loss and function as a hearing aid in the ranges in which a person hears better, helping to preserve some "natural hearing," are now available.

c. ***Hearing assistive technologies (HAT)***. Many deaf and hard of hearing individuals benefit from additional supports to promote their auditory access. Personalized listening frequency modulation (FM) systems are like individualized radio stations that operate on special frequencies. For example, personal FM systems can be used to target a speaker's voice directly to the microphones of an individual's hearing aids. Small, wireless, personalized microphones are another example of technology for amplification. These look similar to a writing pen and can be used discretely to improve hearing in loud environments or over a distance (such as across a large room). In group settings, these "smart devices" can detect the direction from which speech is coming and enhance the listener's access to that sound, over the background noises in the room.

d. ***Visual technologies***. For individuals who cannot hear certain environmental sounds, visual supports can be employed to ensure they are aware of their surroundings. Doorbells can be connected to a lamp, for example, which will flicker when a guest has arrived; fire alarms can be linked with flashing lights, and alarm clocks can be attached to vibrating devices that can wake a person who is deaf or hard of hearing from a deep sleep.

B. ***Terminology***

The terms used to describe and understand individuals with reduced hearing vary depending on the context of the hearing status. Individuals with reduced hearing sensitivity are typically referred to as hard of hearing, deaf, or Deaf [3]:

1. **Hearing loss**

   The phrase "individuals with hearing loss" has been commonly used, yet increasingly this has changed to "individuals with reduced hearing" to reflect the understanding that not all individuals with limited auditory access have experienced a "loss" of hearing (e.g., when an infant is born with reduced hearing, she may not have ever had a full range of hearing).

2. **Hard of hearing**

   Typically, an individual who identifies as *hard of hearing* has some degree of reduced hearing yet can still access sound and spoken language. Audiologically, this term often refers to a person whose hearing loss is in the mild or moderate range.

3. **deaf**

   When the hearing status is further reduced, and a person's auditory input is minimal such that there is limited functional access to spoken language, the term *deaf* may be used. From an audiological perspective, the person may have reduced hearing sensitivity in the severe to profound or profound range.

4. **Deaf**

   When an individual has significantly reduced hearing sensitivity, he may identify as Deaf and belong to the Deaf community, a recognized linguistic and cultural group. For those who self-identify as members of the Deaf community, the use of a uniform signed language and the incorporation of Deaf cultural norms are common. For "capital D (Deaf)" individuals, reduced hearing sensitivity is not perceived as a loss but rather as Deaf gain, the recognition by

members of the Deaf community that being Deaf has added numerous benefits to their lives.

5. **Hearing impairment**

   This a term that has historically been applied to individuals with reduced hearing. However, this term is not accepted by members of the Deaf community and is perceived as disrespectful.

Importance

A. ***Incidence* and *prevalence*:**

1. **At birth**

   The incidence of children born with profound hearing loss is 1 in 1000 births; the number of children born with reduced hearing sensitivity of any degree is 3 in 1000 [4].

2. **Children 12 years of age and younger**

   Recurrent otitis media (ear infection) is the leading cause of mild hearing loss for children. One in eight children under the age of 12 experiences some degree of hearing loss [5].

3. **Adults over the age of 18**

   A study conducted in the USA revealed that, in adults over age 18, 15 % experience reduced hearing [6]. The prevalence of reduced hearing increases substantially in older members of the population.

4. **Adults 70 years plus**

   Nearly two-third of adults age 70 and older experience significant hearing loss that impacts their functioning, particularly in the social realm [5].

5. **Worldwide**

   Five percent of people, or 360 million individuals, experience a "disabling hearing loss" (defined by the WHO as hearing loss greater than 40 dB in the better ear for adults and greater than 30 dB loss for children) [7]. The majority of people who have disabling hearing loss live in low- and middle-income countries. Reduced access to primary healthcare, as well as follow-up specialty services, has a negative effect on the overall health of deaf and hard of hearing individuals, in resource-rich countries as well as those with fewer resources [8].

B. ***Etiologies of reduced hearing*:**

1. **Hereditary conditions**

   Some etiologies of hearing loss are a result of genetics [9]. These can present as particular syndromes that include reduced hearing along with other physical conditions. Common syndromic conditions include Waardenburg syndrome, Usher syndrome, Pendred syndrome, and mitochondrial DNA mutations. With the exception of the mitochondrial mutations, many syndromes do not necessarily result in implications for cognitive, psychiatric, or neurological functioning. Non-syndromic etiologies of hearing loss, as the phrase implies, involve reduced hearing in the absence of other symptoms. A person with non-syndromic hearing loss is considered to be "just D/deaf." Examples include otosclerosis (more common in older adults and results in conductive hearing loss) and the GJB2 gene mutation (also known as Connexin 26), the most common genetic cause of deafness, accounting for up to 50 % of all non-syndromic sensorineural hearing loss.

2. **Non-hereditary conditions**

   Nonhereditary causes of reduced hearing are numerous [9]. The most common congenital cause of nonhereditary deafness is cytomegalovirus (CMV). Additional congenital causes include in utero exposure to rubella, toxoplasmosis, syphilis, and herpes simplex virus. Hearing loss that occurs after birth can be caused by a multitude of factors including exposure to bacterial meningitis, measles, mumps, hypoxia, and ototoxic medications. Hearing loss in older adults often results from extended exposure to noise and presbycusis (age-related hearing loss). The dif-

ferential impacts of the nonhereditary etiologies of hearing loss make it difficult to succinctly state the cognitive, psychiatric, and neurological impacts of each.

Practical Applications

A. ***Cultural vs. medical perspectives***

Working in rehabilitation, the aim of many professionals is to help patients to improve and "get better." From the medical perspective, it follows that restoration of hearing could, or even should, be the goal. Yet, this is in conflict with cultural perspectives on what it means to be a person who is Deaf. Practitioners are encouraged to consider the cultural competence that might be required in working with deaf and hard of hearing individuals, as they might with other minority groups [3]. Resources and information about how to create hospital-based [10] and psychotherapeutic services that support the cultural perspectives of being Deaf [11, 12] are available.

B. ***Health literacy***

Owing to reduced access to health-related information and barriers in accessing health-related services, deaf and hard of hearing individuals are at risk for marginalization in health-care systems [13]. Rehabilitation specialists should be cognizant of possible gaps in global health knowledge and check to ensure that information imparted to deaf and hard of hearing patients is understood. Professionals should also be aware that gaps in knowledge of health-related content is not suggestive of reduced cognitive ability; it is more likely attributable to reduced exposure to health content.

C. ***Psychological functioning and quality of life***

Outcomes and daily functioning for a patient who is deaf or hard of hearing are significantly influenced by attitudes toward hearing held by the patient and attitudes held by the important people in the patient's life. The role of communication with family members and significant others is likely to impact a deaf or hard of hearing person's social-emotional functioning, identity formation, and quality of life [14].

The type and degree of hearing loss, and the benefit from assistive listening devices, will influence the role that reduced hearing plays on social-emotional functioning at the individual level [15]. In broad terms, we will highlight particular challenges that frequently occur for individuals across four categories:

1. **Progressive hearing loss**

Progressive hearing loss requires frequent readaptation to changing levels of hearing. This can result in extended periods of grieving over lost abilities and fear of further loss. As a result, many people with progressively worsening hearing, primarily older adults who have age-related hearing loss, experience negative impacts on their social interactions and relationships with loved ones. They are at risk for feelings of social isolation, frustration, and depression.

2. **Hard of hearing**

Many persons who have moderate hearing levels feel that they are neither hearing nor Deaf. This experience of not fitting into either group can negatively impact identity formation as well as quality of life; indeed, studies of the latter show that hard of hearing individuals struggle more than individuals with typical hearing or those with much more significant hearing loss. Further, because a person who is hard of hearing can "sometimes hear things and sometimes not," the role that hearing plays in their social relationships can be confusing.

3. **Cochlear implants**

The goal for many parents of young children who receive cochlear implants is to be able to verbally communicate with their child. When parents and their children can use similar communication modes (e.g., the same spoken language or the same signed language), perceived family quality of life is improved [12, 14]. Some cochlear

implant users adapt relatively well to the hearing world and view their "ear gear" similar to eyeglasses, i.e., with the appropriate supports, they are able to function without limitations. In fact, for cochlear implant users who have good auditory access and strong language-based skills, quality of life is comparable with those in the general population. Yet, individuals who benefit from cochlear implants vary widely in their ability to use and understand spoken language. Many cochlear implant users struggle to "fit in" and, similar to their hard of hearing counterparts, may feel that they are not entirely *hearing* and yet not truly *deaf* [14].

4. **Deaf sign language users**

Reduced social opportunities and lack of understanding of the experience of being Deaf by members of society can lead to Deaf individuals feeling marginalized, left out, or lonely [15]. This can increase the rates of depression and anxiety in this population. However, Deaf individuals who have adequate social networks, as are often fostered through the Deaf community, report quality of life comparable with individuals in the hearing population [14].

D. ***Cognitive and neuropsychological functioning***

Overall cognitive function in deaf and hard of hearing individuals is distributed similarly to that of hearing individuals, with some differences in specific areas [16]. There are many factors that influence measurement and development of cognitive and neuropsychological functioning including etiology, timing and degree of hearing loss, access to early language, and educational opportunities. Children with neurological risk factors beyond hearing loss tend to have greater difficulties, while children without additional risk factors perform similar to their hearing peers.

1. **Attention and executive functioning**

Studies of attention and executive function offer mixed and task-dependent results: children with hearing loss perform like their hearing peers on tests of planning, impulse control, and cognitive flexibility when tasks are appropriate and accessible for both groups [17]. Language ability seems to be significantly positively correlated with executive functioning in both hearing and deaf children; this may have important implications, particularly in understanding the executive functioning skills of deaf or hard of hearing individuals who have had reduced access to language [18].

2. **Visual processing**

While there are subtle differences in visual processing skills for deaf individuals who sign, these are not typically observable in neuropsychological evaluation [19]. Studies of perceptual abilities in deaf individuals have documented both a deficiency of skills, as well as supranormal visual processing abilities [20]. The field continues to struggle with understanding the role of cross-plasticity of the brain in the presence of reduced sensory input.

3. **Working and short-term memory**

Memory and working memory vary in deaf and hard of hearing individuals; deaf signers have been shown to have an advantage on visual working memory tasks [21] but a disadvantage for linguistic working memory tasks [22]. Some of the differences documented in working memory between deaf and hearing individuals can be attributed to the types of information presented (e.g., recall for numbers is more "automatic" in deaf signers than is recall for letters) [23]. Yet, there do seem to be some consistent differences in span for serial recall, even in conditions that are known to "maximize span" for deaf individuals [24].

4. **Academic achievement**

Among deaf and hard of hearing individuals, the ability to attain a high level of academic achievement is influenced in large part by the extent to which educational information is accessible and appropriate educational opportunities are

provided. The belief that the use of a signed language will "stunt" a deaf person's ability to learn to read has been disproved [25], although, unfortunately, this false belief continues to be held by many professionals in Deaf education.

5. **Motor functioning**

Depending on the etiology of reduced hearing, motor functioning in deaf and hard of hearing individuals can be, although it is not necessarily, negatively impacted in individuals who are deaf or hard of hearing [26]. Although "motor deficits" cannot be generalized in this population, given the vital role of the inner ear structures in aiding with balance, it is perhaps not surprising that balance is reduced in many deaf and hard of hearing people. Interestingly, cochlear implants may enhance balance by granting greater access to auditory information that is used by the brain to foster balance; alternatively, the cochlear implantation surgery may cause trauma to the vestibular system, thereby increasing balance issues. At present, the jury is still out regarding the longitudinal impact that cochlear implants will have on balance and motor functioning.

E. ***Communication***

Communication and access to information is a primary concern for individuals with all degrees of hearing loss. Many children with mild hearing loss or unilateral hearing loss readily gain spoken language skills, althoughit is still possible that their language levels may be below their hearing peers. Functional imaging studies show that brain organization for language is differently distributed for deaf individuals who sign [27] and that deaf children exposed to early sign language can develop strong language skills. The availability of cochlear implants has increased the potential for developing spoken language in profoundly deaf children, though language outcomes even after early implantation are variable; many children who received implantation early show age-appropriate language skills, although a much smaller group of children show little development of spoken language despite access to sound [28]. Explaining the remaining variability in outcomes is an important area of current research.

Tips

- **Inquire about the individual's perspective on his hearing loss**. For any given patient, reduced hearing may be "detrimental" or "no big deal." Some young people may exhibit pride at being members of the Deaf community, while others may have never met another deaf or hard of hearing person and may feel painfully isolated. Older adults may see age-related progressive hearing loss as a natural aspect of aging that must be accepted, while others may resent the impact that changes in hearing status have had on their relationships. Knowing what being Deaf or hard of hearing means to a particular patient will allow health-care providers to best meet that individual's needs.
- **Know that reduced hearing has differential impacts**. Depending on etiology, some individuals are "just deaf," whereas others experience reduced hearing combined with additional physical limitations, reduced cognitive abilities, or psychiatric syndromes. Deaf individuals may have had exposure to Deaf culture, full of rich opportunities to communicate and interact with others similar to themselves, or they may have experienced painful loneliness and been cut off from interactions with others, or been perceived as being "less than smart," because of communication challenges. You cannot know the impact that reduced hearing has had on a particular patient without exploring some of these issues.
- **Recognize that for Deaf persons, spoken/written language may not be their primary language**. Many people assume that, if a patient is unable to talk, written exchanges are a valid substitute. For some this is true. Yet, for many, written language is a second language. Written exchanges of information with a patient who is deaf or hard of hearing will not be sufficient in many cases.

- **Ensure access to communication and information**. Interpreters should be secured if communicating with a patient who uses a signed language. In the USA, the Americans with Disabilities Act (ADA) necessitates that healthcare providers utilize interpreter services in order to promote communication with patients [29]. All patients, regardless of hearing status, should be granted access to information that is pertinent to their health. Involving family members as interpreters is not adequate and is discouraged. Using interpreters in psychological testing is also not advised.

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