# A Provider's Handbook on Culturally Competent Care

## PEOPLE WITH HEARING LOSS
## 1ST EDITION

*Kaiser Permanente National Diversity Council*
*and the Kaiser Permanente National Diversity Office*

KAISER PERMANENTE®

# A PROVIDER'S HANDBOOK ON CULTURALLY COMPETENT CARE

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# INTRODUCTION

Kaiser Permanente is committed to providing linguistically and culturally competent care to its increasingly diverse membership, aimed at eliminating disparities in health status and health care delivery. Culturally competent care refers to the delivery of health care services in a manner that acknowledges and understands cultural diversity in the clinical setting, respects patients' health beliefs and practices, and values cross-cultural communication. Linguistically competent care is the ability to communicate effectively in the patient's language, recognize that language and culture are interconnected, and understand how language reflects culture and simultaneously shapes it.

Culture shapes our thinking; furthermore, our thinking shapes language. This powerful interrelationship affects all human interactions. Considering and respecting a patient's cultural and linguistic needs are key elements to patient-centered care. Ensuring effective communication in the clinician-patient encounter is pivotal to quality of care, patient satisfaction, and patient safety. Our dedication to linguistically and culturally competent care and services is aligned with Kaiser Permanente's mission to provide personalized care and to improve the overall health of the communities we serve.

The goal of the Provider's Handbook series is to provide Kaiser Permanente's health care clinicians with an overview of the cultural and epidemiological differences that characterize major cultural groups represented within our membership. In no way is our intention to stereotype patients by groups. Rather, the handbooks focus on common characteristics of each group that may have implications for health care organizations and practitioners. The handbooks present health care research, practical information, and tools that can help our providers become more aware and knowledgeable about the health care needs of our diverse membership. This is the first edition of the Provider's Handbook on Culturally Competent Care: People with Hearing Loss. It is part of a series of educational resources.

This handbook evolved from a series of initiatives to help providers and staff increase their cultural and clinical understandings of people with hearing loss. It begins with general overviews of cultural and epidemiological aspects of this diverse population. Subsequent chapters highlight hearing loss risk factors and diseases, present better ways to understand hearing loss, and summarize improved interventions for children and adults. Also outlined are legal issues, guidelines from the Americans with Disabilities Act, and resources found throughout Kaiser Permanente to support members with hearing loss and their care providers.

The information provided in this handbook comes primarily from four sources: articles published in medical and health care journals, state and federal publications, information from government and community agencies, and Kaiser Permanente experts. An editorial board composed of Kaiser Permanente physicians, staff, and community experts reviewed and contributed to this handbook.

# DEFINITIONS AND DEMOGRAPHICS

## Who Are People with Hearing Loss?

It is difficult to summarize in one handbook all the important aspects relevant to providing care to patients with hearing loss. This is particularly true because of the complex sociocultural and clinical factors that contribute to hearing loss. For example, great diversity exists among people with hearing loss, with ranges of hearing ability and differing cultural identities. Those who identify themselves as part of the **Deaf** community, for instance, see themselves as having a distinct cultural identity, whereas the majority of people with hearing loss may still identify with the hearing mainstream. The reasons for this phenomenon are complex but could be related to the fact that the overwhelming majority of people with hearing loss once had normal hearing capabilities. Within this group, however, some of these individuals may see their hearing loss as a distinct identity. They may commonly identify as **hard of hearing** or as **people with hearing loss**.

People with hearing loss are from many diverse groups reflecting different generations, races, ethnicities, immigration statuses, socioeconomic statuses, religions, and sexual orientations. In many instances, individuals may not identify themselves as people with hearing loss nor see their hearing loss as a particular illness or defining characteristic.

It is important to note that individuals with hearing loss may also exhibit other common simultaneous health concerns such as visual impairments, loss of natural teeth, depression, or ear infections. These may have existed prior to or concurrently developed with the hearing loss. Be aware, as concerns like these can be easily overlooked when the hearing condition becomes the clinical focus.

### Prevalence

Although statistics vary, hearing loss affects a large portion of the general population. According to the "Summary Health Statistics for U.S. Adults, National Health Interview Survey, 2005 Report" there are 36.5 million non-institutionalized adults with hearing troubles. This represents about 16.7% (1 out of 6) of U.S. adults, 18 years of age or older. Within this group, an estimated 2.2% of individuals (803,000 persons) are deaf. The remaining individuals have hearing loss which ranges from mild to profound.

The report "MarkeTrak VII: Hearing Loss Population Tops 31 Million People" by the Better Hearing Institute (BHI) published in July, 2005 estimates that there are more than 31.5 million people in the U.S. who find it difficult to hear without the use of a hearing aid.

As of 1995, hearing loss ranked as the third most common chronic condition in the U.S, according to the Centers for Disease Control and Prevention (CDC), after arthritis and high blood pressure. Hearing loss is one of the top three most common birth defects in the United States.

### What Do We Know?

The sociocultural and clinical information regarding hearing loss is well summarized in "Hearing Loss: Terminology and Classification" (1997), developed by the Joint Committee of American Speech-Language Hearing Association (ASHA) and the Council on Education of the Deaf (CED). Their position statement is below.

- Individuals who are deaf or hard of hearing constitute a heterogeneous group.
- Hearing level and the ability to develop language skills vary among individuals.

- A variety of factors, like other disabilities, motor function, and/or cognitive function, affect the communication function of individuals with hearing loss.

- The age of the individual when the hearing loss occurred, when it was identified, the type of services received, and when those services began influence communication choices.

- One's family, cultural values, and community support can strongly impact the individual. Examples include, but are not limited to, access to interpreters, communication techniques, use of remaining hearing, and the use of spoken or signed languages.

- Individuals may interact differently with others depending on their work, education, and social environments. The presence or absence of and access to interpreters, appropriate technology, and one's communication partners influence communication.

### People Who Are Deaf

While this handbook does not focus on those who were born deaf, it is essential to recognize the distinct needs and cultural aspects of both hard of hearing and deaf patients. People who are deaf represent about 2.2 percent of those with a hearing loss, according to the National Health Interview Survey in 1990-1991. However, deaf people come to the clinical encounter with unique needs and present specific concerns that differ from those with other types of hearing loss, including age- and noise-related losses.

Because of the wealth of information that exists about the deaf population, both within Kaiser Permanente and in the community, a separate handbook would be appropriate. When it is fitting, information regarding the deaf is presented here as it relates to people with hearing loss.

## Definitions and Terminology: Past & Present

### Common Terms

Since the 1970s, there have been unclear definitions and differing social views regarding hearing loss. Commonly, hearing loss was misunderstood as a normal part of aging that did not require treatment. Discrimination against those with disabilities, including those with significant hearing loss, was commonplace. Hearing loss was viewed as an impairment or handicap in one's life—highlighting what one could not do. For example, the term **hearing-impaired** is very controversial in the Deaf community. Most Deaf people do not want to be referred to as hearing-impaired because the term may imply that deafness indicates the lack of the ability to live a full or happy life.

#### The World Health Organization (WHO) Definitions

Terms such as impairment, disorder, and disability were defined by the World Health Organization (WHO) in 1980. The WHO classification of functioning and disability defines:

- **Impairment** as an abnormality of a structure or function; therefore, a hearing impairment could be an abnormality of any part of the ear or of the auditory system

- **Disability** as the functional consequence of an impairment such as the inability to hear certain frequencies or any sound at all

- **Handicap** as the social consequence of having the impairment. Many times it is not a disability that is limiting but how people negatively treat or value a person with a disability. With hearing loss, people may become isolated, lose their jobs, or make significant changes in their lives in order to communicate

These antiquated terms may still be heard today. Although an impairment perspective still persists regarding hearing loss, newer definitions and ways of describing the experience transcend this outmoded view. The following section includes current terms that are respectful and not rooted in an impairment perspective.

## Current Language and Issues

### A Framework Shift at the WHO

The WHO's current framing of disabilities now focuses on levels of functioning and health status. This focus is a part of the "International Classification of Functioning, Disability and Health" (ICF). ICF is a universal classification of health and disabilities. As WHO states, "Previously, disability began where health ended; once you were disabled, you were in a separate category."

Instead of emphasizing a person's hearing loss, the focus is on the health, quality of life, and well-being of the individual. This framework recognizes that disabilities and hearing loss can happen to anyone at any age, and thus makes hearing loss a common human experience, without stigma. The ICF, in particular, reinforces a shift in perspective by moving the focus from the cause of disability to the impact of disability.

The ICF is an example of a cultural shift in the language of how we describe people with disabilities like hearing loss. One outcome of this shift has been the idea that people have every right to choose what they wish to be called as a group or as an individual. Person-centered language acknowledges the person and describes the condition, e.g., a person with hearing loss or a person who is hard of hearing.

### Deaf

The National Association of the Deaf (NAD) defines a person who is **deaf** as someone who is "unable to hear well enough to rely on their hearing and use it as a means of processing information." This can include the complete loss of hearing in one ear as well. According to the CDC, the term "deaf" describes "someone who has approximately 90 dB [see "Degrees of Hearing Loss" below for an explanation of the decibel scale] or greater hearing loss or who cannot use hearing to process speech and language information, even with the use of hearing aids." Adults who are "late-deafened" have become deaf after acquiring hearing and language abilities. Those who are prelingually deaf typically use sign language because verbal language has not and cannot be developed.

A cultural definition of deafness also exists. The term Deaf (with a capital 'D'), as defined by Carol Padden and Tom Humphries in 1988, refers "to a particular group of deaf people who share a language—[in the U.S.] American Sign Language (ASL)—and a culture." According to these authors, in *Deaf in America: Voices from a Culture*, "[the] members of this group have inherited their sign language, use it as a primary means of communication amongst themselves, and hold a set of beliefs about themselves and their connection to the larger society." To some Deaf people, suggesting the use of assistive hearing devices has negative cultural, social, and emotional ramifications to their culture, families, and identity.

### Hard of Hearing

The term **hard of hearing** is sometimes used to describe people who have a less severe hearing loss than deafness. These individuals usually can understand conversational speech either with or without hearing aids. The International Federation of Hard of Hearing People states that the term "hard of hearing" means all people who have a hearing loss and whose usual means of communication is by speech. This definition includes those who have become totally deaf after acquisition of speech. NAD has a different definition for the term. It defines "hard of hearing" to mean those who have some hearing, are able to use it for communication purposes, and who feel reasonably comfortable doing so.

## Technical Definitions

There are other terms commonly used to describe the various types of hearing loss. Many hearing loss definitions are listed in a publication called *A Parent's Guide to Hearing Loss* by the CDC's Early Hearing Detection and Intervention Program. Some of these important terms are listed below.

- **Bilateral hearing loss**: a hearing loss that exists in both ears
- **Prelingual hearing loss**: a hearing loss presented before a child learns to talk
    - § Sometimes, if the individual is deaf, the term *prelingually deaf* is used
- **Postlingual hearing loss**: hearing loss that happens after a child learns to talk
    - § If the individual is deaf, the term *postlingually deaf* is sometimes used
- **Unilateral hearing loss**: hearing loss that affects only one ear

Besides having problems hearing speech clearly, people with hearing loss can also have the following experiences as manifestations of their hearing loss.

- **Impaired sensitivity**: the inability to hear soft sounds
- **Altered loudness perception**: a reduced tolerance for loud sounds and abnormal perception causing certain sounds to be perceived as louder than they are
- **Impaired frequency selectivity**: an inability to detect subtle differences in words having similar pitch patterns
- **Reduced temporal integration**: longer periods of time needed to fully detect sounds
- **Altered pitch perception**: perceived pitch distortions
- **Impaired localization ability**: a difficulty in determining the location of sound
- **Increased susceptibility to noise interference**: having greater difficulty understanding language in areas or locations with background noise

[See the Bibliography for more print and online resources.]

Despite these common terms and definitions, the way people identify or label themselves is a very personal and individual decision. Health care providers can be supportive by encouraging individuals to self-identify. Be aware, however, that not all people prefer to use person-centered language, as when a person says, "I am Deaf."

## How We Hear

When we listen, the outer ear takes in sound waves like a funnel and moves them to the external ear canal. At the end of the canal is the tympanic membrane (eardrum). The tympanic membrane vibrates when it is hit by the sound waves. These vibrations then pass through the ossicles (small bones) in the middle ear. The vibrations then pass to the cochlea, which contains a fluid that transmits pressure changes. The tiny hair cells that line the cochlea pick up the vibrations from the fluid and create nerve

impulses that go to the auditory (VIII) nerve. The nerve then carries this message to the brain where it is interpreted as a sound. Problems in any of these mechanisms can lead to hearing loss.

## Causes of Hearing Loss

The etiology of hearing loss can vary widely. This section highlights the majority of causes of hearing loss. Most people experience a gradual onset of hearing loss, called presbycusis, caused by the aging process. Presbycusis is a common type of hearing loss seen in primary care. There are various conditions and risk factors that can result in a sudden hearing loss. The following table shows reported causes of hearing loss and their distribution in the general population.

**Table 1: Percent Distribution and Number of Reported Causes of Hearing Trouble of Persons 3 Years of Age and Over by Type of Cause, U.S., 1990-1991.**

| Cause of Hearing Loss | Distribution due to Cause |
|---|---|
| At birth | 4.4% |
| Ear infection | 12.2% |
| Ear injury | 4.9% |
| Loud brief noise | 10.3% |
| Other noise | 23.4% |
| Getting older | 2.8% |
| Other causes | 16.9% |
| **Total** | **100%** |

*Source: National Center for Health Statistics, Data from the National Health Interview Survey, Series 10, Number 188, Table 16, 1994.*

### Physical Blockage

One simple cause of hearing loss is a build-up of cerumen (ear wax) in the ear canal. People who wear hearing aids may experience increased wax in the ear canals, since the hearing aid or earmold tends to reduce the amount of natural ventilation to the ear. Another cause is foreign objects that are pushed too far into the ear canal. Individuals may use hairpins, match sticks, or cotton swabs to clean their ears but can make matters worse by causing injury or pushing wax further into the ear canal.

### Acute Illness or Infection

A cold or sinus congestion can cause some hearing loss. When there is a difference in pressure between the air outside and the air in the ear, it is not uncommon to experience some temporary hearing loss. An infection in the middle ear causing a build-up of fluid, a discharge from the ear, or perforations of the tympanic membrane can also lead to conductive hearing loss.

### Noise

Noise is one of the leading contributors to premature hearing loss. Exposure to loud noise even for a few minutes can cause a loss of hearing. Overexposure to quieter, more sustained noise can also cause permanent damage. For example, continual exposure to noise exceeding 85 dB can harm and damage hearing. Commonly, people experience tinnitus (ringing in the ears) due to noise.

Unlike many of the causes of hearing loss, exposure to noise is one of the few that can be controlled and prevented. Urging individuals to take simple steps like turning down noise levels, limiting the time

exposure to noise, and wearing hearing protection can safeguard one from hearing loss. More information can be found in the "Frequently Asked Questions" (FAQ) section of the American Tinnitus Association website. [See the Bibliography for print and online resources.]

## Aging

Aging is the number one cause of hearing loss, typically called presbycusis. Over the last few decades, there has been a significant increase in hearing loss due to the growing aging U.S. population. As baby boomers age, the prevalence of hearing loss will continue to rise.

## Ototoxic Drugs

The book *Ototoxic Drugs Exposed: Prescription Drugs and Other Chemicals That Can (and Do) Damage Our Ears*, by Neil Bauman, is the result of four years of research into approximately 1,000 ototoxic drugs. Some of these medicines are powerful and are used in extreme cases, but others are commonly used. For instance, high doses of aspirin could alter one's hearing, and the antibiotic gentamycin can cause inner ear damage if given incorrectly. Some ototoxic drugs can cause tinnitus, while others can temporarily cause hearing loss or permanent damage. Below are some of the medications Neil Bauman researched. This is not an exhaustive list.

**Table 2: Some Ototoxic Medications and Examples.**

| Medication Types | Medicine Names |
|---|---|
| ACE Inhibitors | Enalapril (*Vasotec*), Moexipril (*Univasc*), Ramipril (*Altace*) |
| Acetic Acids | Diclofenac (*Voltaren*), Etodolac (*Lodine*), Indomethacin (*Indocin*), Ketorolac (*Toradol*) |
| Alpha Blockers | Doxazosin (*Cardura*) |
| Aminoglycosides | Amikacin (*Amikin*), Gentamicin (*Garamycin*), Kanamycin (*Kantrex*), Neomycin (*Neosporin*), Netilmicin (*Netromycin*), Streptomycin, Tobramycin (*Tobradex*) |
| Angiotensin-2-Receptor Antagonists | Eprosartan (*Teveten*), Irbesartan (*Avapro*) |
| Anti-Arrhythmic Drugs | Flecainide (*Tambocor*), Propafenone (*Rythmol*), Quinidine (*Cardioquin*), Tocainide (*Tonocard*) |
| Anti-Cancer Drugs | Buserelin (*Suprefact*), Carboplatin (*Paraplatin*), Cisplatin (*Platinol*), Vinblastine (*Velban*), Vincristine (*Oncovin*) |
| Anti-Convulsant Drugs | Carbamazepine (*Tegretol*), Divalproex (*Depakote*), Gabapentin (*Neurontin*), Tiagabine (*Gabitril*), Valproic acid (*Depakene*) |

| | |
|---|---|
| Anti-Malarial Drugs | Chloroquine (*Aralen*), Mefloquine (*Lariam*), Quinine (*Legatrin*) |
| Anti-Retroviral Protease Inhibitors | Cidofovir (*Vistide*), Ganciclovir (*Cytovene*), Ritonavir (*Norvir*) |
| Benzodiazepines | Diazepam (*Valium*), Estazolam (*ProSom*), Midazolam (*Versed*) |
| Beta-Blockers | Atenolol (*Tenormin*), Betaxolol (*Betoptic*), Metoprolol (*Lopressor*) |
| Bicyclic Anti-Depressants | Venlafaxine (*Effexor*) |
| Calcium-Channel-Blockers | Diltiazem (*Cardizem*), Nifedipine (*Adalat*), Nisoldipine (*Sular*) |
| COX-2 Inhibitors | Celecoxib (*Celebrex*), Rofecoxib (*Vioxx*) |
| H1-Blockers | Cetirizine (*Zyrtec*), Fexofenadine (*Allegra*) |
| Immunosuppressant Drugs | Cyclosporine (*Neoral*), Muromonab-CD3 (*Orthoclone OKT3*), Tacrolimus (*Prograf*) |
| Loop Diuretics | Ethacrynic acid (*Edecrin*), Furosemide (*Lasix*), Torsemide (*Demadex*) |
| Macrolide Antibiotics | Clarithromycin (*Biaxin*), Erythromycin (*Eryc*) |
| Opiate Agonist Drugs | Codeine (*Codeine Contin*), Hydrocodone (*Vicodin*), Tramadol (*Ultram*) |
| Propionic Acids | Flurbiprofen (*Ansaid*), Ibuprofen (*Motrin*), Naproxen (*Anaprox*) |
| Proton Pump Inhibitors | Esomeprazole (*Nexium*), Lansoprazole (*Prevacid*), Rabeprazole (*Aciphex*) |
| Quinolones | Ciprofloxacin (*Cipro*), Ofloxacin (*Floxin*), Trovafloxacin (*Trovan*) |
| Salicylates | Aspirin, Mesalamine (*Asacol*), Olanzapine (*Zyprexa*) |
| Selective Serotonin Reuptake Inhibitors (SSRIs) | Fluoxetine (*Prozac*), Fluvoxamine (*Luvox*), Sertraline (*Zoloft*) |
| Serotonin-Receptor Agonists | Almotriptan (*Axert*), Naratriptan (*Amerge*), Sumatriptan (*Imitrex*) |
| Thiazides | Bendroflumethiazide (*Corzide*), Indapamide (*Lozol*) |
| Tricyclic Anti-Depressants | Amitriptyline (Elavil), Clomipramine (Anafranil) |

*Source: Hearing Loss Help, May 15, 2007*
*http://www.hearinglosshelp.com/articles/ototoxicupheaval.htm*

### Head Injuries

A blow to the head, concussion, or injury caused by a blast may result in damage to the auditory nerves or the auditory cortex. A large or sudden barometric change, as with diving or an explosion, can rupture the eardrum. It is not uncommon for severe blows to the head to result in a decrease in hearing ability.

## Types of Hearing Loss

Hearing loss is sometimes referred to as the invisible disability because of the lack of obvious physical signs, unless one uses a hearing aid. Hearing loss is usually grouped into two major categories: sensorineural hearing loss and conductive hearing loss. However, a mixed hearing loss (a combination of the two) can also occur.

### Sensorineural Hearing Loss

The majority of hearing loss is the result of sensorineural damage to the functionality of the inner ear or hearing nerve. For example, this type of damage is characterized by injuries made to the 30,000 to 50,000 hair cells in the inner ear that are responsible for transmitting sound information to the auditory nerve. This damage can be the result of a wide variety of factors such as environmental exposures, the aging process, illness, trauma, and ototoxic chemicals. Sensorineural damage is the most prevalent cause of hearing loss. Hearing aids, cochlear implants, and other devices may assist individuals with this type of hearing loss.

### Conductive Hearing Loss

Conductive hearing loss is caused by physical barriers that stop sounds from getting through the outer or middle ear. This hearing loss can be caused from structural or mechanical damage to the outer and/or middle ear(s), including the ossicles (small bones in the middle ear). Patients can receive medical or surgical treatment for a conductive hearing loss, which can restore hearing partially or completely.

### Mixed Hearing Loss

Mixed hearing loss is a combination of conductive and sensorineural hearing losses involving both the middle and inner ear.

## Degrees of Hearing Loss

A loss in hearing is characterized by frequency, intensity, or both. Hearing loss severity is based on how well a person can hear frequencies and their intensity, most often associated with speech. Taking frequency and intensity into account, hearing loss can be described as mild, moderate, severe, or profound and is measured in decibels (dB). Intensity of hearing loss is measured in the range of 0 to 120 dB (usually in the better hearing ear) and in the frequencies of 500, 1000, 2000, and 4000 Hz. The table below summarizes the corresponding measure, effect on the person, and possible intervention for each descriptor.

**Table 3: Hearing Loss Description, Measure, Effect, and Possible Interventions**

| Description | Measure (dB) | Effects | Possible Actions |
|---|---|---|---|
| Normal | 0-25 dB | No or very slight hearing problems. Able to hear whispers. | Prevention and education. |
| Mild | 25-40 dB | Able to hear and repeat words spoken in normal voice at 1 meter. | Prevention and education. Hearing aids may be needed. Consider other social supports. |
| Moderate | 41-60 dB | Able to hear and repeat words spoken in raised voice at 1 meter. | Prevention and education. Hearing aids usually recommended. |
| Severe | 61-80 dB | Able to hear some words when shouted into better ear. | Prevention and education. Hearing aids needed. If no hearing aids available, speechreading and signing should be taught. |
| Profound (including deafness) | 81 dB or greater | Unable to hear and understand even a shouted voice. | Hearing aids may help in understanding words. Additional social supports or rehabilitation needed. Speechreading, sometimes signing, and visual aides required. Possible cochlear implant evaluation. |

*Source: World Health Organization, December 31, 2006*
*http://www.who.int/pbd/deafness/hearing_impairment_grades/en/index.html*

## Functional Aspects of Hearing Loss

Most individuals, even with normal hearing or mild hearing loss, cannot hear everything in a noisy setting such as a busy restaurant or on the street with traffic. Those who have a moderate hearing loss may need the help of an assistive hearing device under the same conditions. Hearing aids may be of limited or no help to individuals with a profound hearing loss. These individuals may need to rely on their vision to facilitate communication using written notes, speechreading (formerly known as lip reading), gestures, computers, email, sign language, finger spelling, text telephones, faxes, and interpreters.

Gradual hearing loss is the most common form. Because of this, many individuals and their loved ones do not notice changes until significant signs become evident. Common signs and symptoms of hearing loss can include:

- Asking to have things repeated often
- Reacting to loud noise
- Playing the television or radio too loud
- Not hearing the telephone ring
- Looking at people when they talk, making it easier to understand what is said

- Experiencing ringing in the ears
- Speaking in a nasal, high-pitched, or monotone voice
- Articulating words less
- Ignoring sounds coming from behind
- Misunderstanding conversations or what is said
- Speaking too softly or too loudly
- Needing to stand close to a person speaking
- Having trouble hearing when the speaker's face is not visible
- Having trouble hearing when it is too noisy
- Complaining that people are mumbling
- Straining to listen but not understanding or comprehending
- Not always responding when spoken to, or responding inappropriately

Many of these behaviors, such as speaking softly, may be culturally determined and may not reflect a hearing loss if evaluated out of context.

# Demographics and Trends

## Age and Gender

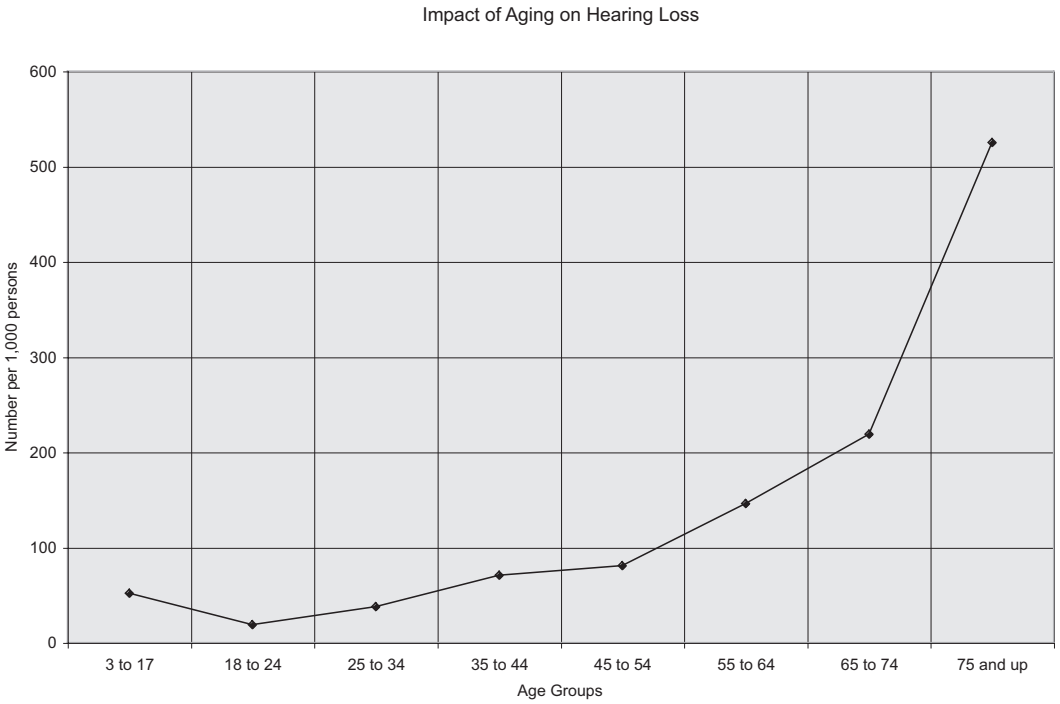
It is projected that by 2050, the number of people with hearing loss will increase by 102%, doubling the number of people with hearing loss. The prevalence of hearing loss in younger populations is increasing due to more noise exposure. This rate also is partially attributed to the growing aging population, which has a higher prevalence of hearing loss compared to younger adults.

Current prevalence rates are listed below:

- **Newborns**: although studies vary in the prevalence rates for hearing loss in newborns, general estimates range between one and six per 1,000 newborns
- **Children**: according to the "Third National Health and Nutrition Examination Survey, 1988 to 1994," 14.9% of U.S. children aged 6 to 19 have a low-frequency or high-frequency hearing loss of at least 16 dB hearing level in one or both ears
- **General population**: one in six people (16.7% of the general population) have some degree of hearing loss
- **Ages 65 to 74**: for the population aged 65 to 74, one in three people (30.4% of this group) have a hearing loss
- **Over 75 years old**: For those over the age of 75, one in two people (48.1% of this group) have a hearing loss

Hearing loss is the third most common chronic health condition affecting the older population. The number of affected individuals is likely to rise with the large number of aging baby boomers. The figure below highlights the impact that advancing age has on hearing ability.

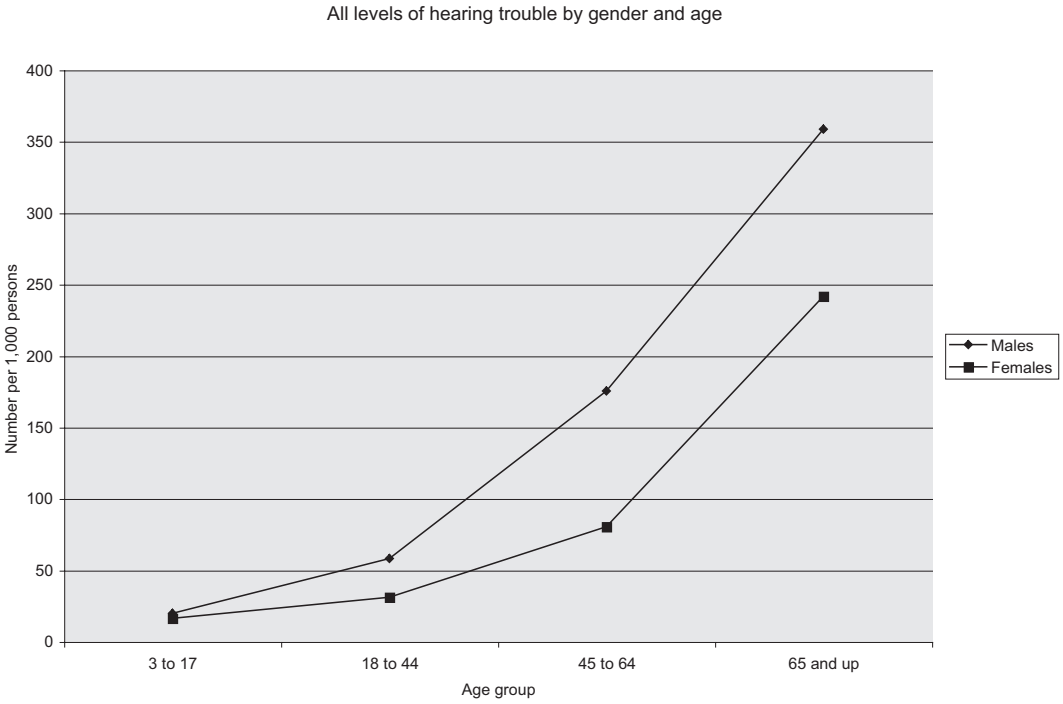
**Figure 1. Average Annual Age-specific Number of Persons, 3 Years of Age and Over, Who Cannot Hear and Understand Normal Speech per 1,000 Persons: U.S., 1990-1991.**




*Source: Vital and Health Statistics. Prevalence and Characteristics of Persons with Hearing Trouble: United States, 1990-91, Series 10: Data From the National Health Survey No. 188*

The prevalence of hearing loss differs according to gender. Hearing loss rates are higher for men than women. For persons 3 years and older, 59.1% of males and 40.9% of females have some degree of trouble hearing. Under the age of 18 years, there is not a pronounced hearing loss difference between genders. The following graph shows the number of people with hearing loss, by gender, per 1,000 persons.

**Figure 2. Average Annual Crude Number of Persons by Sex with Hearing Trouble per 1,000 Persons: U.S., 1990-1991.**




*Source: Vital and Health Statistics. Prevalence and Characteristics of Persons with Hearing Trouble: United States, 1990-91, Series 10: Data from the National Health Survey No. 188*

Despite the fact that woman have a higher life expectancy than men, men still have a higher prevalence of hearing loss than women.

## Race and Ethnicity

There is limited information regarding hearing loss within racial and ethnic groups. The "Summary Health Statistics for U.S. Adults: National Health Interview Survey 1997" data highlight the prevalence of hearing trouble among adults in different racial and ethnic groups:

- Twenty percent (20%) of non-Hispanic whites
- Eleven percent (11%) of non-Hispanic blacks
- Nine percent (9%) of Hispanics
- Eleven percent (11%) of other non-Hispanics

Below is the prevalence of hearing trouble among adults aged 65 years and older in different racial and ethnic groups:

- Fifty-three percent (53%) of non-Hispanic white men compared to 35% of non-Hispanic white women

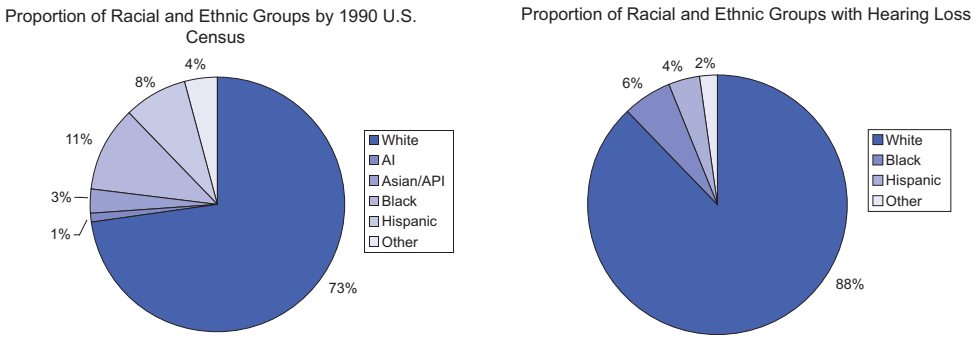
- Thirty-one percent (31%) of non-Hispanic black men and 24% of non-Hispanic black women
- Thirty-six percent (36%) of Hispanic men and 29% of Hispanic women

Although 70% of the U.S. population is white, 92% of those who report having hearing trouble are white individuals. At first glance, hearing troubles appear to disproportionately affect the white population. However, the white population tends to be older than other racial and ethnic groups, and this may partly account for the disproportionate distribution of hearing troubles within this population. After adjusting for age, of those reporting hearing trouble, the percentage of white individuals drops to 90.2%. Hispanics account for 7.9% of those with hearing troubles. Data for Asian-American, Asian-Pacific Islander, and Native-American populations are based on small sample sizes, which makes it difficult to make generalizations about these particular groups. The fact that there is a significant African-American population in the United States, and yet a surprising lack of data on the causes and effects of hearing loss within that population, indicates the need for further research on hearing loss prevention and treatment for black patients.

The underrepresentation of people of color in hearing loss data, both now and in the past, may reflect health care institutional barriers such as not having an interpreter present, lack of insurance, lack of transportation, and lack of culturally sensitive care, thereby rendering this data less reliable. The following table shows available hearing loss data by age groups, race, and ethnicity in comparison to the 1990 nationwide census. Although this data is from 1994, it is the one of the most comprehensive studies to date of people reporting hearing loss. The data indicate a lack of information in regards to how issues of race and ethnicity affect and are affected by hearing loss.

With this understanding, it is recommended that each individual take steps to continue one's education on the topic by keeping apprised of recent research in medical and academic journals. The data presented here are meant to provide the informational foundation for an understanding of what has been measured to date, rather than provide practitioners with analysis that is specific enough to guide care.

**Figures 3 & 4. 1990 Racial and Ethnic Groups U.S. Census in comparison to Proportion of Racial and Ethnic Groups with Hearing Loss. Average Annual Crude Number of Persons by Sex with Hearing Trouble per 1,000 Persons: U.S., 1990-1991.**




*Source: U.S. Census, 1990*

However, in recent years, there has been an increase in research on hearing loss particularly as measured in children. The Gallaudet Research Institute, part of Gallaudet University, a university serving the deaf and hard of hearing population, produces an Annual Survey of Deaf and Hard of Hearing Children and Youth. Some findings from the Institute's "2006-2007 National Survey" are depicted below. The discrepancy between the census data above from 1990 and the 2006-2007 data below could be caused by several factors, one of which is the different types of hearing loss most commonly acquired by children

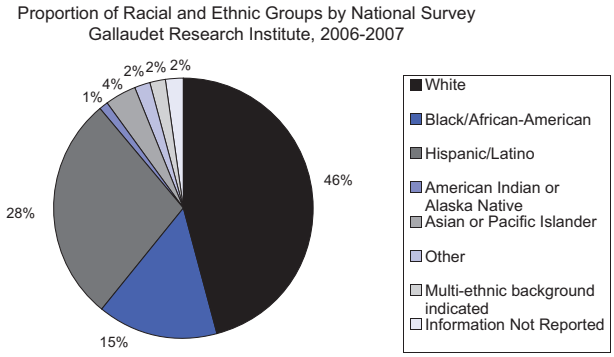
vis-à-vis adults. Access may also be a cause. While many children are required by public school systems to undergo hearing screenings, the treatment of hearing issues in adults often depends upon whether or not the adults have health insurance.

It is also important to note that in the measurement of hearing loss in students, the prevalence rates as measured by the United States Department of Education under the Individuals with Disabilities Education Act (IDEA) do not reflect the racial and ethnic population trends. As Diane M. Scott explains in Hearing Research: Children From Culturally and Linguistically Diverse Populations:

"The racial/ethnic breakdown of children between the ages of 6 and 21 years served under IDEA, Part B for 1998-99 showed some differences as compared to the general population. For example, African-American children represented 14.8% of the general population and 16.8% of children with hearing impairment. Hispanic children accounted for 14.2% of the general population and 16.3% of children with hearing impairment. White children represented 66.3% of the general population and 63.0% of students classified as hearing impaired."

The lack of data on the prevalence and causes of hearing loss among different racial, cultural, and ethnic groups persists, but an in-depth analysis of annual surveys such as that of the Gallaudet Research Institute, over time, could indicate target areas in the American social and cultural landscapes as major areas for potential systematic improvement.

**Figure 5. National Summary of Breakdown of Individuals with Hearing Loss by Racial and Ethnic Group: U.S., 2006-2007.**




*Source: Gallaudet Research Institute Annual Survey of Deaf and Hard of Hearing Children and Youth. http://gri.gallaudet.edu/Demographics/*

## Education and Socioeconomic Status

Individuals with an annual family income of $10,000 or less have a higher prevalence of hearing loss and constitute about 14.4% of all individuals reporting difficulty hearing among all income levels. Persons with a family income of less than $20,000 are twice as likely to have a hearing loss as compared to people with incomes greater than $50,000. Thus, hearing loss prevalence decreases as family income increases.

Hearing loss is more common for those who do not have a high school diploma compared to high school graduates. This relationship is consistent across all age groups. This may be explained by the fact that children with an undiagnosed hearing loss may exhibit learning difficulties, and other consequences may result, such as dropping out of school.

## Mental Health

There have been several studies indicating that the lack of treatment of hearing loss leads to negative social and emotional consequences for individuals. These studies establish that lack of intervention is associated with reductions in social functioning, psychological wellness, self-esteem, and general quality of life. Issues around mental health and well-being are common, not only for older adults but most likely for anyone who has experienced hearing loss. Social isolation is one effect of hearing loss that can result in a wide range of psychological conditions, especially depression. The presence of depression can complicate physical problems but may go unrecognized because of the multiple physical problems presented in the health care encounter.

### Older Adults

The report "The Consequences of Untreated Hearing Loss in Older Persons" (1999), by the National Council on the Aging, outlines many mental health concerns for older populations. Those with untreated hearing loss were more likely to report depression, anxiety, and paranoia and less likely to participate in organized activities in comparison to those who wore hearing aids. In addition, although hearing aids can reduce depression, the remaining communication struggles can contribute to depression. Suicide is highly correlated with major depression. The link among hearing loss, depression, and suicide in this population behooves the health care provider to become aware of the multiple educational, prevention, and intervention opportunities to address these issues with this population.

According to the National Strategy for Suicide Prevention (NSSP), people who are 65 years and older have the highest suicide rates of any age group. In addition, older persons frequently use highly lethal methods, make fewer attempts per completed suicide, have a higher male-to-female ratio than other groups, have often visited a health care provider before their suicide, and have more physical illnesses than other age groups. Since suicide is preventable and providers have frequent contact with older persons, there are opportunities to intervene and save lives. NSSP estimates that:

- Twenty percent (20%) of people over 65 years old who commit suicide visited a physician within 24 hours of their act
- Forty-one percent (41%) visited within a week of their suicide
- Seventy-five percent (75%) have been seen by a physician within one month of their suicide

### Clinical and Behavioral Signs

Awareness of a variety of clinical and behavioral clues is useful in the identification of depression in patients. Persistent and vague complaints such as pain, headaches, fatigue, insomnia, GI symptoms, arthritis, multiple diffuse symptoms, and weight loss are well-known primary presentations of depression in the elderly. Further, for older individuals, depression is often characterized by memory problems, confusion, social withdrawal, loss of appetite, inability to sleep, irritability, and in some cases, delusions and hallucinations. Depression in older adults may be mistaken for dementia. Depression symptoms may be so disabling that individuals cannot articulate their distress or reach out for help. On the other hand, demanding behaviors and frequent phone calls may be a sign of reaching out for help.

### Creating a Safe Environment

To make patients feel more comfortable, it is important to provide a safe climate to allow patients to express their feelings related to hearing loss. Offering emotional support, engaging patients in conversation, and listening carefully to the words they choose are essential to creating a safe environment. It is important not to minimize the person's feelings, but to point out realities and offer hope. Patients' remarks about death ideation or suicide must be taken seriously by the health care provider and the patient should be referred to a mental health specialist immediately.

## Health Care Provider Tips

- Hearing loss is often an invisible disability. As of 1995, hearing loss was the third leading chronic health problem in the U.S., after arthritis and hypertension. Regular screening is important.

- There are 36.5 million non-institutionalized adults with hearing troubles. This represents about 16.7% of U.S. adults, 18 years of age or older. This group is growing.

- The etiology of hearing loss can vary widely. Nevertheless, most people experience a gradual onset of hearing loss, called presbycusis, caused by the aging process. Hearing loss from noise exposure is common as well. Knowing these key exposures, providers can help people retain their remaining hearing abilities through interventions.

- Hearing loss is the most common sensory deficit in children, with an incidence of approximately one in every one-thousand live births. Limited English proficient (LEP) parents and families may require an interpreter when discussing the health of their children. Please prepare ahead of time to arrange this service.

- Hearing loss is usually grouped into two major categories: conductive hearing loss and sensorineural hearing loss. However, a mixed hearing loss, a combination of the two, can also occur.

- Losses in hearing can be characterized by frequency, intensity, or both. Hearing loss severity is based on how well a person can hear the frequencies or intensities most often associated with speech. Taking frequency and intensity into account, hearing loss can be described as mild, moderate, severe, or profound. People do not typically recognize when their hearing is deteriorating.

- As with most people, how individuals identify or labels themselves is personal and sometimes difficult to understand. Nevertheless, health care providers can be supportive by encouraging individuals to self-identify. Identifying individuals as hard of hearing before they are ready to accept their hearing loss can negatively impact their health care experience.

# CULTURAL PERSPECTIVES, BELIEFS, AND BEHAVIORS

As with the Deaf population and most cultural groups, people's perspectives and beliefs of self-identity are typically learned in early childhood. The vast majority of people with hearing loss participate in and identify with the hearing world. Therefore, they do not characteristically adopt an established identity centered on their hearing loss. Cultural and personal beliefs about hearing loss can be strong determinants in people's readiness to take steps to address their hearing loss.

## The Costs of Hearing Loss

Hearing loss is costly and causes other hardships for individuals and for society over time. A CDC article summary states that the estimated lifetime cost for a person with a hearing loss in 2003 was $417,000 more than those without hearing loss. The estimated lifetime costs for all people with hearing loss born in 2000 will total $2.1 billion. Of these hearing loss costs, 6% are direct medical costs, 30% are non-medical expenses, and 63% are for lost wages or work. There are other indirect costs associated with untreated hearing loss, such as negative social, psychological, emotional and quality of life consequences for the individual. Some of these issues are discussed later in this section.

## Stigma and Barriers to Treatment

With the exception of those born with hearing loss and/or those who are Deaf-identified, people with hearing loss typically do not embrace hearing loss as part of their personal identity. This may be due to different cultural attitudes, negative stereotypes, or stigma associated with hearing loss.

### Myths

Some people are informed by cultural myths about hearing loss. The variety of popular myths shapes people's understandings of the abilities and characteristics of people with hearing loss and the deaf. Some of these erroneous myths include:

- Everyone who is deaf knows sign language
- Everyone can speechread
- Speech reading gives 100 percent understanding
- People with hearing loss can hear when they want to
- People who are deaf are mentally deficient
- Hearing loss is a normal part of aging and does not need medical intervention
- Hearing loss only happens with the elderly
- Having a hearing loss only requires louder volumes to hear better
- Hearing aids cannot help, do not work very well, or are cosmetically undesirable

### Individual Challenges

Many people who have lost or are losing their hearing are concerned about maintaining their independence. In *Missing Words: The Family Handbook on Adult Hearing Loss*, Dr. Donald Holden writes, "One of the most crippling fears that people with a hearing impairment face is insecurity." There may be some people who are well informed but may hide their condition or disguise their behaviors out of embarrassment and shame. Not dealing with these feelings may be a detriment to one's self-esteem, self-worth, and may delay the individual in seeking help. At times, individuals as they age or experience gradual hearing loss may endure feelings of loss or grief, resort to different coping strategies, and may be resistant to change depending on awareness and motivation. Nevertheless, in "Hearing Loss," an online public course, Eileen Seigel states that acceptance comes after depression and involves the individual's realization that "there is something wrong with my hearing, not with me."

## Loss and Grief

People can react with intense feelings to loss. As Helen Keller, who was prelingually deaf and blind, stated, "The problems of deafness are deeper and more complex, if not more important, than those of blindness. Deafness is a much worse misfortune. For it means the loss of the most vital stimulus—the sound of the voice that brings language, sets thoughts astir and keeps us in the intellectual company of man." It is this loss of connection with others that can lead to feelings of helplessness, anger, resentment, and/or depression.

The intense feelings of loss may reflect different stages of grief. Adults who have early-onset hearing loss often report that despite the challenges, they incorporate the hearing loss into their identity. This lends an opportunity for individuals to learn ways to cope, as people with hearing loss, as they grow and develop.

This may not be the case with people who have acquired hearing loss later in their lives. Within this group, for those who have experienced a sudden or rapid loss of hearing, it can be quite disorienting. Many people state that hearing loss robs them of their understanding of their identity and can cause an identity crisis. Many will mourn the loss of their hearing as they go through Kubler-Ross' **stages of grief.** The stages are typically not a linear, sequential process. For example, it is not uncommon for someone to move from the depression stage back to the anger stage.

#### Stages of Grief

**Shock stage**: after hearing information, a cognitive and behavioral paralysis

**Denial stage**: ignoring or discounting the information

**Anger stage**: frustration expressed and "why me?" fixations

**Bargaining stage**: negotiating for an escape from what is presently true

**Depression stage**: final realization of the information to be true

**Acceptance stage**: finding new ways to cope and move forward

Understanding the different stages of grief and the patient's corresponding stage can help to inform appropriate interventions by health providers. A patient's migration through the various stages of grief are to be expected. Consider them as normal reactions to the experience of loss. It is difficult to generalize this process because linguistic or cultural norms may shape a different process of grief for an individual, one that is more culturally relevant. For example, in some cultural groups, the expression of anger and blaming others may not be considered an appropriate response. Accordingly, the anger stage may be expressed as a period of silence, withdrawal, deep contemplation, or woefulness.

## Coping

Coping with hearing loss is a personal and culture-bound experience. **Coping** can be defined as an emotional, cognitive, or behavioral process to address a stressful circumstance. The experience of losing one's hearing, suddenly or over time, can be difficult to manage. Different cultural groups may have their own sets of beliefs and norms about hearing loss, thus shaping how stressful the experience might be. Needing to circumvent new barriers, interpersonal stress, and miscommunication are some demands placed on a person with hearing loss who has not been treated. A person's ability to cope with these demands depends on available resources such as one's stamina, health, social support, self-esteem, level of optimism, financial power, abilities for self-care, and ability to access health care.

The coping strategies people employ can reflect a person's stage in the grief process. Some of these coping strategies can be idiosyncratic or harmful. There are few alternatives for people who either deny a hearing loss or do not want to seek help for hearing difficulties. These individuals may find themselves experiencing a greater level of stress as they try to cope with their hearing loss in the following ways:

- They may ask people to repeat themselves frequently or ask many clarifying questions, which can be stressful or exhausting for individuals with hearing loss. It may also lower their self-esteem, add confusion to the communication process, or frustrate the speaker. During a health care encounter, these individuals may require extra time for evaluations, counseling, or other interventions.

- They may give non-verbal cues indicating they understand when in fact they do not, such as nodding, smiling, saying "ok," and thereby encouraging the speaker to continue. In this case, provider recommendations may be missed unless there is a family member or friend present at the office visit to recall the information.

- Some individuals may dominate conversations by doing most of the talking to lessen the risk of missing information. They may change the subject if they get lost in the conversation or are unable to hear the key content of the conversation.

- Some people stop socializing because they cannot hear. They may try to avoid social interactions because they may feel unsafe, embarrassed, or depressed due to their inability to hear conversations. Mental health comorbidities such as anxiety and affect disorders (depression in particular) should be assessed.

- People with hearing loss may blame other people or situations for their hearing problems. For example, they may say that they cannot hear because others mumble, have an accent, or have a speech impairment.

- People with hearing loss may continue to deny a hearing loss by saying it is not a serious problem. They may refuse to seek assistance when suggested, as they may not realize that their hearing loss is causing problems in their daily interactions.

- Some may be self-conscious of their appearance when wearing a hearing aid or feel it would make them appear old or disabled. These individuals generally will not buy hearing aids, but if they do, they might hide them away in a dresser drawer.

- They may have a fatalistic perspective and feel powerless to change their circumstance. Some people may perceive their hearing loss to be a punishment, curse, or religious message. Some may decide to give up. They may not make any changes or learn new skills, and may rely on others' help with tasks.

As with most chronic conditions, it is important for the patient to acquire new behaviors to prevent further hearing loss and improve quality of life. Foregoing these coping strategies can be an important step

in inciting the individual to be more involved in making healthy choices. Since this can be particularly difficult to do, individuals will be at different stages in their readiness to change. Health care providers can help individuals take more action regarding their hearing loss by employing different interventions to jump-start change. The diversity of a patient needs to be considered when assessing his or her readiness for change. The expression of willingness or readiness will vary and is culturally influenced.

## Beliefs

Beliefs are rooted in cultural information that attempts to explain our external and internal worlds. The effectiveness of a health care provider is determined by his or her responsiveness to the different cultural beliefs held by patients with hearing loss. For example, the U.S. culture tends to emphasize the value of independence of the individual. Thus, beliefs around hearing loss and its treatment are strongly rooted in this orientation. Some individuals therefore may reject the idea of needing to rely on hearing aids. Others with different cultural orientations may not think of hearing loss as something that is treatable. Some may need to consult with family members, elders, or community resources before delivering bad news to someone. In some cultures, disclosing possible risks of the treatment to the patient is perceived as harmful or counter-productive.

Cultural explanations and ways of understanding and treating health problems become very pertinent when a person's beliefs or belief system contradict or opposes a biomedical orientation of the health care provider. If not handled carefully, the patient's health, trust, and satisfaction can be compromised.

One way to help think about a patient's beliefs, as they pertain to implementing treatment, is the **Health Beliefs Model**. The Model is made up of six constructs that explain the types of personal beliefs that influence health behaviors. These constructs influence decisions to prevent, screen for, and control illness. The following list, found in a National Cancer Institute publication, "Theory at a Glance: A Guide for Health Promotion" explains that people are willing to take an action or make a change if they:

- Believe they are susceptible to the condition (perceived susceptibility)
- Believe the condition has serious consequences (perceived severity)
- Believe taking action would reduce their susceptibility to the condition or its severity (perceived benefits)
- Believe costs of taking action (perceived barriers) are outweighed by the benefits
- Are exposed to factors that prompt action (e.g., a television ad or a reminder from one's physician to get a mammogram) (cue to action)
- Are confident in their ability to successfully perform an action (self-efficacy)

Helping a patient to understand the cultural orientation of the health care provider may not be enough to encourage the person to accept treatment. Framing medical opinions and recommendations in cultural terms that are relevant to the patient is an effective way to engage the individual in treatment.

## Adherence

Helping patients change unhealthy behaviors is important for providers. For example, a patient may be reluctant to wear a hearing aid. Change interventions are especially critical with patients needing to make lifestyle modifications for disease prevention, long-term disease management, mental health, and general quality of life.

A provider may have patients who seem unable or unwilling to make changes upon which they have previously agreed. This behavior is considered non-adherence, or as traditionally called, noncompliance. The concept of patient non-adherence focuses on behavioral change outcomes that may ignore the needed supports, motivation, and appropriate steps to adopt a new behavior. The concept of non-adherence may assign blame to the patient when, in fact, there may be cultural or language barriers that hinder change. Language barriers can create confusion or even complete misunderstandings. Change is infrequently a quick, single event. When one labels a patient non-adherent, it highlights patient failure rather than acknowledging where the patient is in the stages of change. Listening closely, providing support and furnishing information in a nonjudgmental manner are key. [See the Bibliography for more print and online resources.]

## Stages of Change

Prochaska and colleagues, in the 1990s, developed a paradigm to approach this complex idea of behavioral change. This paradigm was coined the "transtheoretical model of behavioral change." A wide range of health behaviors have been investigated using this paradigm showing successful results. This approach can be applied cross-culturally, if it is linguistically and culturally appropriate.

The transtheoretical model of change is a helpful tool to understand patient readiness for change and overcome barriers to change. It has been shown to improve patient satisfaction and lower provider frustration during the change process.

The **stages of change** framework, summarized by five stages, helps us understand how shifts of behavior occur. This framework has been successfully used for a variety of problem behaviors. It is important to remember that this is not a linear model and individuals may move in different directions toward different stages:

- **Precontemplation** is when an individual has to change a behavior but is unwilling or even unaware of the need for change. Denial is a common way for people to remain in this stage.

- **Contemplation** is when an individual recognizes the need for change; at this stage the individual is only pondering change. Depending on cultural norms of expression of grief, this is not an uncommon place for people to grieve the reality of their hearing loss.

- **Preparation** is a stage that combines intention and planning. Individuals in this stage are intending to take action in the next month, or so, and have not taken action in the past year.

- **Action** is the stage where a patient begins to adjust his or her behavior or surroundings. This is a time of exploration and testing. It is important to remember to support changes a person chooses to make. Recommending a particular behavior may go against certain cultural norms. Some individuals may not reach the next stage.

- **Maintenance** is when a person has consistently employed the new behavior learned in the action stage.

It should be highlighted that these phases do not follow a simple, linear progression, but usually involves moving between stages in any direction. This is described as a spiral pathway of change and suggests that individuals will move forward, and then regress to previous stages. The person advances by learning from mistakes and effective strategies.

Although this is one approach to motivating patients, it is important to reflect upon one's style and how one engages patients. Consider the techniques that would best help the patient at a given stage of change. Refer to the book *Motivational Interviewing: Preparing People for Change, 2nd Edition* by William R. Miller and Stephen Rollnick for exhaustive information on this topic.

## Health Care Provider Tips

- Many people are informed by cultural attitudes, negative stereotypes, or stigma associated with hearing loss. Some people are informed only by cultural myths about hearing loss. Patient education and prevention strategies should be discussed with the patient and her or his social support.

- Consider a person's language abilities and literacy levels when giving oral or written instructions or advice. If the patient's record indicates an interpreter is needed, prepare for this service ahead of time. Please refer to the next chapter for more on communication issues.

- Adults with hearing loss often have emotional reactions that reflect different stages of grief or responses to their loss. The expression of these responses varies culture to culture, person to person. Exploring these responses with the patient, or consulting with someone from a similar cultural background may be helpful.

- It is important for the person with hearing loss to acquire new behaviors to prevent further hearing loss and improve quality of life. A wide array of personal and cultural-bound coping strategies and rationales need to be addressed in order for individuals to make healthy choices.

- Helping a patient to understand the cultural orientation of the health care provider may not be enough to encourage the person to accept treatment. Framing medical opinions and recommendations in cultural terms that are relevant to the patient is an effective way to engage the individual in treatment.

- Change interventions are tools that are especially critical to assist patients to make lifestyle modifications for disease prevention, long-term disease management, mental health, and general quality of life. Consider reading some of the recommended resources or consulting with a social worker or psychologist for more information.

# COMMUNICATING WITH PEOPLE WITH HEARING LOSS

## Effective Communication

At a very basic level, communication is the process of transmitting information among people. Because language is part of our daily interactions, we may not realize that it is a convoluted process fraught with potential sources of error that can cause the meaning of a message to become skewed. This chapter presents strategies, legal requirements, and tips for effective communication with people with hearing loss.

When hearing loss is considered to be the problem, there is a fairly limited range of solutions. When hearing loss is considered to be the cause of the broader problem of a loss of communication, a broader range of solutions is available. The process of communication involves the signal source, the listening environment, and the receiver. In addition, visual and tactile senses may also be used for more effective communication.

Verbal communication can be a challenge for an individual with a hearing loss. Words or sentences may be misunderstood and wrong messages received. Clear communication is crucial in a medical setting when the information being communicated has an effect on patients' health and well-being, and influences their trust in providers and the health care system. The challenge of clear communication does not rest completely upon the patient.

The varying degrees and configuration of hearing loss are not obvious. The loss may be mild, and patients may communicate well under optimal conditions. When operating under less-than-optimal conditions, hearing loss presents a challenge to the patient and health care provider. For instance, patients with hearing loss may not be aware that their names have been called in the waiting room. A patient may miss part of what is being said during an examination if the provider is not directly in front of him or her.

Talking louder to a person with a hearing loss may not increase understanding. In fact, when voices are raised, the voice clarity becomes distorted. In dominant U.S. culture, when a person talks too loud it can be perceived as anger, and thus elicit other emotional reactions in patients. One of the symptoms of sensorineural hearing loss is called recruitment. Recruitment is an abnormal sensitivity to loudness or things that get too loud too fast. People with recruitment can be bothered by sounds that are too loud even though they may not be too loud for a person with normal hearing. Talking louder to a person with this symptom complicates comprehension for the individual with hearing loss.

Below are some tips and suggestions to improve communication between the patient and health care provider. Some of these tips may be helpful to improve communication between the patient and the patient's friends, family, and significant others.

- It is useful to say the person's name before speaking. Many persons with hearing loss complain that they were not aware they were being spoken to and missed beginnings of sentences.
- Try to situate yourself at the person's eye level.
- Stay out of the shadows by keeping light on the face, especially the mouth, while talking. Communication is better facilitated when people are facing each other. The prime of the outer ear is at an angle to catch the airborne message.

- Avoid chewing gum, drinking, or eating while talking. This can make it easier for people to speechread, if they are able. A third of phonemes (speech sounds) are visible; seeing this can give the listener clues to what is being said; but facial expressions can also communicate whether the speaker is excited, concerned, angry or happy.

- By law, for a deaf or limited English proficient (LEP) person, an interpreter must be present if necessary.

## Behaviors in Communication

There is often a mutual sense of frustration between the patient and the provider when there are communication issues due to hearing loss. Less than five percent of those with a hearing loss use badges or stickers to identify themselves as hard of hearing for emergencies. Individuals with a hearing loss may appear aloof, confused, or appear disoriented to their hearing counterparts. They may speak too loudly or softly, they may respond inappropriately to questions, or may respond not at all. At times people with hearing loss may appear to be anxious, nervous, or even withdrawn in some cases. Because these behaviors can look like symptoms of other problems, the risk of misdiagnosis is high in this population.

If a unilateral loss is present, patients may turn their heads to one side in order to listen. Screening for hearing loss should be commonplace. Often patients, staff, and providers may be unaware of assistive technologies available for use.

When individuals from the Deaf community come into a medical center, the surroundings may breed anxiety, since many of our warning signals (such as fire alarms) are based on sound technology. The patient may feel a sense of helplessness as few of the staff are familiar with the Deaf culture. Staff often quickly give up trying to communicate with the Deaf patient. A common stereotype that people may have of the Deaf is that they are mentally deficient. This may lead staff to totally ignore a Deaf person.

### Speechreading (Lip reading)

Often speechreading is not an effective form of communication for individuals with hearing loss. Some people with hearing loss do, however, rely on speechreading for communication. Forty to sixty percent of English sounds look alike when spoken. On average, even the best speechreaders only understand 25 percent of what is said to them, and many other individuals understand far less. Speechreading may be particularly difficult in the medical setting where complex medical terminology is often used. Only 30 to 40 percent of the English language is visible on the lips, so listening for fragments of speech is necessary for patients with hearing loss.

People's ability to speechread improves when they have some hearing capability already in place, and their hearing loss occurs gradually over time. Since spoken language is already known, speechreading and other skills generally improve over time. Thus, generally speaking, speechreading is not acquired by congenitally deaf persons and only 10 percent of people with hearing loss rely exclusively on speechreading. If a speaker wears a moustache, has an accent or speech impediment, is far away from the listener, or if there is more than one speaker, the speechreader will have additional difficulty.

The speechreader also studies facial expressions and body language and combines all the information to reach a logical conclusion. Additionally, people with hearing loss base their communication efforts on the primary language spoken during their childhood which may not be English. Limited English proficient (LEP) individuals attempting to speechread an English speaker can have great difficulty comprehending what is said.

## Sign Languages

In the U.S., if someone is prelingually deaf, this individual most likely communicates through **American Sign Language (ASL)**. Most conservative estimates in *A Journey into the Deaf-World* ranks ASL as the sixth most commonly used language in the U.S., if English is excluded. It is estimated that there are 500,000 to 2 million ASL users. It is a complete and natural language, not a word-for-word translation of English.

ASL differs in grammar and syntax from English. The signed message, gestures, and facial expressions create the vocabulary, grammar, and punctuation. The same gesture may have different meanings with hearing individuals compared to ASL. Take, for example, nodding one's head. To a hearing individual it may mean agreement, whereas to one using ASL, it may mean that the audience understands what is signed or the signer may continue. Words may also hold a different meaning. For example, "positive" in ASL means "good." Thus, a positive HIV test may be interpreted as being a desired outcome unless care is taken to clarify the meaning of the result. Medical terms, such as nausea or allergy, may not easily translate to an ASL user. Family members may not be fluent in ASL and should not be presumed to know how to sign.

ASL is not the only sign language used in the United States or internationally. There are countless different sign languages and dialects throughout the world. For instance, in Mexico, a predominant sign language is Mexican Sign Language *(Lenguaje de Señas de México)*. In addition, there is a Mayan Sign Language also known as Nohya Sign Language or Yucatec Maya Sign Language (*Lenguaje Mímico Maya*).

### Qualifications of Interpreters for Sign Language

As stated in the Americans with Disabilities Act, a qualified "interpreter is one who is able to interpret effectively, accurately, and impartially, using any specialized vocabulary necessary." Health care interpreters, for example, are trained in medical terminology and vocabulary. Qualified interpreters have appropriate credentials indicating their qualifications. These interpreters have been assessed and tested using criteria reflecting the interpreter standards of practice. If you'd like more information about interpreters for sign languages and their standards of practice, please visit the website of the Registry of Interpreters for the Deaf.

There are a variety of types of interpreter, and not all deaf or individuals with a hearing loss use the same kind of interpreter. The health care provider should ascertain the particular language needs of the patient before requesting an interpreter. Some individuals may require interpreters who are fluent in American Sign Language (ASL), a language that has a grammar and syntax that is different from English. Others may require interpreters who use Signed Exact English (SEE), a form of signing that uses the same word order and syntax as English. Others who do not know any sign language may require oral interpreters who take special care to articulate words to hard of hearing individuals. Some may need cued speech interpreters who give visual cues to assist in speechreading.

## Utilizing Telephone and Technology

Delivering information to a patient with a hearing loss may occur through several telephone-based devices. The following is a list of these devices and helpful tips to providers using such devices:

- Answering machines/Voicemail: the caller should repeat the telephone number of the medical facility twice. Also, rephrase the information, or spell out words and names. Fax machines can also be used.

- Compatible telephone: this telephone generates a magnetic field that can be picked up by turning on a "T" switch to activate the telecoil in a hearing aid.

- Assistive listening devices (ALD): these are tools that bring the speaker's voice directly to the ear with or without a hearing aid. They also help to overcome the problems of distance and surrounding noise.

- FM transmitter and Infrared (IR) Systems: an FM transmitter broadcasts a signal by radio waves from the sound source to a receiver worn by the listener. The waves can cover several hundred feet and pass through physical obstructions. An IR receiver transmits sound via IR waves to a receiver worn by the listener. With IR, however, the signal stays within the room and so is used in settings that require confidentiality, such as courtrooms. Infrared devices are often used in theaters.

- TTY/TDD (teletype/telephone device for the deaf): this is a device used by those who cannot use the telephone because of hearing or speech loss. Similar to a typewriter, the unit shows the conversation on a screen so that it can be read.

- Telecommunication Relay Services: this allows text telephone users to communicate with a non-text telephone user by way of a relay service communications assistant (a nationwide relay service that has a toll-free telephone number). The assistant types into the TTY what the provider says and then also reads what is typed by the patient through the TTY.

- The most common tool that deaf people use to communicate with hearing people is email or texting on personal digital assistants (PDAs) or mobile phones. This may be helpful to others with hearing loss.

- The newest of recent technology is Video Relay Services (VRS). This service enables deaf and people with hearing loss who use ASL to conduct video calling with anyone, through a sign language interpreter. The person with hearing loss sees an interpreter on the television or computer and signs to them. Then the interpreter uses a standard telephone and relays the conversation.

Health care providers that routinely provide telephone services for patients must make these services available to individuals with hearing loss. It is less common for these individuals to use TTYs for telephone communication than in the past, but in a hospital setting TTYs should still be available. Health care providers can receive incoming calls from TTY/VRS users through relay systems. Relay services enable individuals who use TTY/VRS to communicate by telephone with individuals who are hearing. In a relay system, a third person, called a "communications assistant," acts as an interpreter for the verbal, signed, or written communication used by the communicators. Health care providers are not charged for use of the relay center. Rather, the costs of providing relay services are spread among all telephone users. While using these systems, the language diversity and the literacy level of patients should be considered.

Not all individuals need TTYs to communicate. Some individuals have enough hearing to enable them to use telephones that are compatible with hearing aids, or telephones with amplifiers. Health care providers should make these auxiliary aids available for outgoing calls from their facilities if they offer outgoing telephone services to the general public.

Where patients in hospitals and nursing homes are able to watch television, deaf patients must be able to see the captions on closed-captioned programs. All televisions manufactured after July 1, 1993 with screens that are 13 inches or larger have built-in decoder ability. On older televisions, a separate decoder can be connected to the receiver. At times, health care providers offer information to clients and patients in the form of videotapes. The ADA requires that all public accommodations, including health care providers, make aurally delivered information available to any individual with hearing loss. One very effective way of making videotapes accessible to these individuals is to caption the tapes.

Health care providers that offer training sessions, health education, or conferences to the general public must make these events accessible to the deaf and individuals with hearing loss. In addition to interpreters and real-time transcribers, there are a variety of assistive listening systems that may be appropriate to eliminate problems with distance and background noise for individuals who use hearing aids.

### Other Options for Communication

Cued-speech uses hand signals and spoken English (which is speechread). **Total communication** is a philosophy of using any means possible to communicate the message effectively. It may involve spoken language, auditory training, sign language, miming, or gestures. Writing is used as well, and can be time-consuming for both the patient and the provider. Often, important issues may be left out in the interest of time. Since many deaf people consider American Sign Language (ASL) to be their first language, they may have difficulty understanding English since it is a second language for them.

Written notes may provide adequate communication with the patient, but their efficacy depends on the patient's reading level. On average, the reading level of the deaf community is lower than that of hearing people. Because the grammar and syntax of ASL differ considerably from English, writing back and forth may not provide effective communication between the deaf patient and the health care provider. Moreover, information that would otherwise be spoken may not be written. If a health care provider is providing less information in writing than he or she would provide when speaking to a hearing patient, this is an indication that writing is not an effective communication in this context.

## Legal, Regulatory, and Accreditation Requirements

The **Rehabilitation Act of 1973 (Section 504)** and the **Americans with Disabilities Act (ADA) of 1990, Titles II and III**, mandate the provision of qualified interpreters. Several states have legislated certification requirements for sign language interpreting. Within these guidelines, several mandates exist, many of which require assistance for people with hearing loss and the deaf. Some of these mandates are highlighted below:

- Identify those with a hearing loss using audiometric screening
- A qualified ASL interpreter must be provided for routine medical services and for medical emergencies 24 hours a day whenever requested
- Communication devices must be available, such as assistive listening devices, TTY, strobe light alerts, and telephone amplifiers
- Protocols and procedures should be in place to prevent psychological and social isolation of people with hearing loss

**Title VI of the Civil Rights Act of 1964** is a federal law prohibiting discrimination by recipients of federal financial assistance on the basis of race, color, or national origin. Persons with limited English proficiency (LEP) are protected under the Act and are entitled to equal access and participation in federally funded programs through the provision of language services. **The National Standards on Culturally and Linguistically Appropriate Services (CLAS)** standards, produced by the Office of Minority Health, are a series of recommended and required practices for health care organizations to make their practices more culturally and linguistically accessible. In particular, CLAS standards 4 to 7 are current Federal requirements for all recipients of Federal funds based on Title VI of the Civil Rights Act of 1964.

In California, **MediCal Managed Care Program** requirements mandate developing, implementing, and monitoring a system to provide cultural and linguistic services. MediCal managed care requires 24-hour access to interpreter services for all members, at all provider sites. In 2003, the State of California passed **Senate Bill 853** (SB 853), requiring that health plans ensure access to services among members who may be limited English proficient. This law is enforced through regulations issued by the Department of Managed Health Care (DMHC). These regulations were finalized in 2006 and all California health care systems need to be fully compliant by January 2009. The regulation requirements are:

- Translation of vital documents into the two most common languages preferred by patients
- Capture, track, and report the language preferences of patients to the DMHC
- Communication to patients in their preferred language(s)
- Ensure 24-hour, 7 days per week access to language assistance services, free of cost
- Train employees who have routine contact with members on cultural diversity and language services
- Assess, qualify, and train all bilingual staff serving as interpreters
- Ensure that contracted providers are compliant with SB 853
- Monitor and regularly report compliance with SB 853

Additionally, accreditation agencies such as the **National Council of Quality Assurance (NCQA)** and the **Joint Commission on Accredited Healthcare Organizations (JCAHO)** have cultural and linguistic requirements.

# Physical Settings

## Reception Area

Patients with hearing loss may experience anxiety waiting for their names to be called in the reception area. Visual barriers, such as posts or aquariums, may interfere with their line of sight and may not allow them to see staff come into the room. This is further complicated because the acoustics of the waiting room may produce significant background noise, especially if children or music are present. The caretaker of the individual may also be a person with a hearing loss. The raising of voices or shouting by staff distorts sound, thus making it harder for the person to understand. This also may embarrass the person or violate rules of confidentiality. Digital hearing aids may shut off automatically when a patient walks through a sliding, motion-sensor door; this may not be immediately recognized by the patient.

## Examination Rooms and Offices

Communication is an extra challenge in a medical examination room. To be heard and understood, a health care provider should not turn away from the patient to enter information into a computer. Avoid describing details of the patient's health status while out of the patient's direct line of sight. Wearing a mask may inhibit communication; face shields or transparent masks improve visualization of face and lips. Missed information may require the provider to repeat the same information. This may add time to the examination. If the patient understands what the provider says and recommends, then adherence to advice may increase significantly.

Ideally, accommodations for interpreters or visual electronic devices should be prepared at the time the appointment is made. If communication devices are not readily available, some patients may benefit from the careful use of a stethoscope as an audio amplifier. The provider speaks into the diaphragm as the patient listens with the ear piece. Exam rooms may have a great deal of ambient noise, such as computers, paper gowns, suction machines and water running during hand washing. Background noise should be limited as much as possible. Sound loses half of its intensity for every three feet of distance. For example, "your daughter is pretty," can become "your daughter is pregnant," at a distance greater than three feet. However, reducing the distance between the provider and patient may have cultural implications.

Ultimately, it is the responsibility of the provider and staff to ensure patient comprehension. Tape recording the discussion for a hearing relative may be helpful. Repeating each other's sentences is another way to check for mutual understanding. Speaking slowly or in short phrases forces individuals to enunciate all the syllables and gives the listener time to comprehend what is being said. Short sentences are easier to understand than long ones, which may contain more pieces of information.

One symptom of sensorineural hearing loss can be loss of clarity (speech recognition or speech discrimination) and hearing aids may not be able to totally compensate for poor clarity. In addition, the person's hearing loss may have progressed since being fitted with the hearing aid.

### Hospital Setting

Upon registration, all staff and security personnel should be tactfully informed of the name and location of the patient with a hearing loss to help him or her in case of emergency situations, such as a fire within the facility. Providing private rooms can be helpful if acoustics in the facility are poor. When relying on the written word to relay information, the patient's ability to read should be verified. Hearing aids may receive interference in a hospital setting; using closed caption video can facilitate understanding. It may also be helpful to allow family and friends to remain with the patient to avoid loss of information. This is especially important when obtaining an informed consent.

### Procedure Rooms and Operating Room Settings

Before a procedure, printed literature explaining the goals, risks, steps of the procedure, and potential side-effects may ensure patient comprehension. However, the literacy level and language needs of the patient must be taken into account.

Many of the procedures are performed with the provider not visible to the patient, such as with the lumbar puncture. It is helpful to plan a series of visual cues or hand signals that can be used during the procedure for communication.

Patients should be instructed to take their hearing aids with them on the day of the procedure. Although the aid must be removed for certain procedures or surgeries, the patient will need the hearing aid in position to help with orientation following an anesthetic procedure. What is often thought as a post-anesthetic disorientation is often a problem with a hearing aid that is not in place or not functioning. Therefore, the staff needs to know how to put hearing aids on and adjust the volume.

## Health Care Provider Tips

### Assuring Successful Communication

Below are some tips and suggestions intended to help health care providers remove communication impediments and improve communication with patients.

#### General Guidelines

- Providers can familiarize themselves with the Deaf culture, the experiences, and psychology of people with hearing loss.

- Recognize that patients with hearing loss may have different emotional reactions and coping strategies. Stages of grief, denial, social isolation, depression, talking obsessively, and giving cues that they understand when they do not are all common behaviors.

- Be aware of accents or speech impediments that can make it difficult for people with hearing loss to understand, especially when they are unfamiliar with particular speech inflections or accents.

- Ensure that patients, especially the aging, do not also have a visual impairment which could interfere with speechreading and reading written notes.
- It is important to determine the patient's hearing abilities to gauge which communication supports are appropriate.
- Assess the best method for communication by asking patients about their language or communication preferences, ideally before their appointments.
- Recognize that patients with hearing loss may also be limited English proficient. This may be true for Deaf patients whose first language can be American Sign Language (ASL) or another sign language.
- Ask patients how you can best communicate with them.

### Communication Guidelines

- If the patient relies on sign language or is limited English proficient, arrange for a certified/qualified interpreter to be present. While working with an interpreter, keep these guidelines in mind:
  - § Speak directly to your patient, not the interpreter
  - § The interpreter will communicate everything that is said by both parties
  - § The interpreter relays meaning for meaning; not word for word
  - § Avoid medical jargon, slang, and acronyms
  - § Speak at an even pace; ask one question at a time
  - § Avoid changing ideas in the middle of a sentence
- If the patient prefers to communicate with you and the staff by speechreading, ensure the following:
  - § Gain your patient's attention; use visual alerts or gestures
  - § Maintain eye contact with your patient
  - § Do not use exaggerated lip or mouth movements
  - § Speak in a regular voice volume
  - § Avoid turning your head, taking notes, or entering information in the patient's medical record which obscures the view of your face
  - § Avoid having your back to a light or window
- If the patient prefers to communicate by writing notes, keep these tips in mind:
  - § English is a second language for most deaf people
  - § Be aware that American Sign Language has its own syntax and does not follow the syntax of English

- Keep your writing at a low reading level and accessible by minimizing jargon and complicated terms

- Writing notes is time-consuming, resulting in communication that may be incomplete. Use short, precise phrases, pictures, or diagrams when appropriate

# RISK FACTORS

## Perinatal Issues

The first trimester of pregnancy is important for the healthy formation of the fetus' membranous labyrinth. Abnormalities in this structure lead to cochlea and otic capsule malformations. The second and third trimesters are important times for maturation of the auditory hair cells and auditory nerve. Although the developing sensory systems have a great deal of plasticity, the auditory nervous system development is dependent on receiving input, or stimuli, in order to grow its neural structures. Part of this growth during the first three years of life is crucial in the development of speech and language. Early identification can help target treatment. Universal hearing screening is recommended within the first three months of life.

One to four newborns out of every thousand are born with severe to profound hearing loss. Without newborn hearing screening, the average age of hearing loss detection is between 14 and 30 months of age. Even if all infants who fit high risk criteria are screened, it is estimated that half of the infants with hearing loss will not be detected. Even a normal newborn screen can miss a delayed-onset, progressive hearing loss. Continued monitoring of hearing abilities is recommended every three to six months following the initial screen for those infants identified with high risk factors. Often it is parents who identify 70 percent of these late-diagnosed children. If appropriate language milestones are not met by 18 months of age or if the use of words suddenly stops, further testing is warranted.

In 1994, the Joint Committee on Infant Hearing developed a list of 10 items that place infants in a high risk category for hearing loss. These risks include:

1. Family history of hearing loss
2. Congenital/neonatal infections
3. Craniofacial anomalies
4. Low birth weight (<1,500 grams)
5. Hyperbilirubinemia
6. Ototoxic medication treatment, like aminoglycosides
7. Bacterial meningitis
8. Low Apgar scores, such as zero to four in one minute, and zero to six at five minutes
9. Mechanical ventilation for five or more days
10. Facial features consistent with or the presence of a syndrome that is usually associated with congenital hearing loss

Other identified risk factors include:

1. Admission to the NICU
2. Length of stay in the NICU
3. Prematurity
4. Hypoxemia
5. Respiratory distress syndrome
6. Intracranial hemorrhage
7. Retrolental fibroplasias
8. Maternal drug use, including alcohol
9. Neonatal blood transfusions

It is hypothesized that hypoxia leads to progressive degeneration of the auditory hair cells. Mechanical ventilation is strongly associated with late onset progressive hearing loss, persistent pulmonary hypertension, and prematurity. Prematurity is also associated with intracranial hemorrhage and sepsis.

# Congenital Hearing Loss and Associated Syndromic Losses

Sensorineural hearing loss is the most common form of congenital hearing loss. The greatest risk of its onset occurs when there is both a history of chromosomal anomalies combined with congenital structural anomalies. According to Bluestone, Stool, et al., an anomaly is an isolated malformation that results in a structural change. A syndrome can be considered a pattern of multiple anomalies, thought to be pathogenetically related, found in an individual. Craniofacial anomalies may be associated with more than one syndrome. The sites of abnormalities affecting hearing are in the region of the cochlea, the auditory (VIII) nerve, or the central nervous system. There are more than one hundred genes associated with hearing loss and more than 350 genetic conditions causing genetic deafness.

The hearing loss of a majority of newborns will not be identified as part of a syndromic loss. In those infants who have a non-syndromic sensorineural hearing loss, more than half will have mutations in two genes: GJB2 and GJB6. Mutations in these two genes are responsible for the most common cause of hereditary hearing loss. These chromosome-13 genes produce the cochlear protein connexin 26. Connexin 26 is responsible for maintaining potassium levels in the endolymph of the cochlea. Infants will present a non-progressive moderate to profound bilateral sensorineural loss. Many institutes recommend connexin 26 mutation screening if a bilateral sensorineural loss greater than 40 dB is found. Several endocrine abnormalities tend to be present in many genetic syndromes. The most common of these is Pendred's Syndrome, in which there is an error in thyroxin production. Down's syndrome usually demonstrates a mixed loss, with sensorineural and conductive components.

Craniofacial abnormalities are associated with an increased risk of hearing loss. Usually, loss associated with anomalies of the pinna or middle ear will be conductive. Aural atresia, including external auditory canal atresia, has an incidence of one in 10,000 to 20,000 births. The more common syndromes include Treacher-Collins Syndrome (mandibulofacial dysostosis), Goldenhar's Syndrome (oculoauriculovertebral dysplasia), Crouzon's Syndrome (craniofacial dysostosis), and Apert's Syndrome (acrocephalysyndactyly). CT scanning may be helpful in identifying abnormalities of inner ear structures, but it is not usually performed until the age of four when the development of the temporal bone and mastoid allow for better identification of these structures.

Ocular abnormalities (retinitis pigmentosa and choroidoretinits) are found in a few inherited sensorineural hearing losses. These include Usher's Syndrome, Alstrom's syndrome, and Refsum's Disease.

Hearing loss associated with cardiac defects is most commonly seen in Jervell and Lange-Nielson Syndrome. There may be a family history of fainting spells with hearing loss to alert the provider of this possibility.

Renal disease associated with hearing loss is most commonly seen in Alport's Syndrome. Approximately half of the patients with Alport's Syndrome have a high-frequency sensorineural hearing loss. Males demonstrate a stronger and greater manifestation of the disease.

Waardenburg Syndrome is the most common depigmentation syndrome. This is also the most commonly diagnosed inheritable hearing disorder, with about 20 percent of Waardenburg patients having a hearing loss. This syndrome has an incidence of about two newborns per 100,000 births.

Skeletal dysplasias or dysostoses may be associated with either conductive or sensorineural hearing losses due to abnormal temporal bone growth. Otosclerosis and osteogenesis imperfecta are the most common examples of this abnormal bone formation. Otosclerosis is twice as common in females compared to males, and is the most common cause of conductive hearing loss in patients over 15 years of age. [See also "Pediatric Hearing Loss, Non-Otitis Related"]

## Infections

Perinatal infections include in-utero infections due to cytomegalovirus, measles, rubella, syphilis, toxoplasmosis, and herpes. With widespread immunization more commonly practiced, deafness-causing fetal rubella and infant meningitis have dramatically declined. However, meningitis remains the leading cause of acquired hearing loss in infants and children. The Hib vaccine has dropped the overall incidence of Hemophilus Influenza-induced hearing loss to about 11 percent. Streptococcal pneumonia now leads these infections with an incidence of 23 percent. Of patients surviving meningitis, up to 35 percent will have some amount of hearing loss. About seven percent of children surviving bacterial meningitis will have a sensorineural hearing loss, but all children should be referred for audiometric evaluation following this illness.

Additionally, children with cleft palates have an increased incidence of recurrent middle ear infections. Conductive hearing loss occurs in approximately 90 percent of these children.

## Drug Exposure

Ototoxicity occurs in approximately 10 percent of those treated with aminoglycosides. After early exposure to an aminoglycoside, the outer hair cells (in the basal turn of the cochlea) are affected, leading to a high-frequency sensorineural hearing loss. In later stages, the inner hair cells are involved. At this point, otoacoustic emissions can be used to measure the degree of hearing loss. After treatment of the aminoglycoside is discontinued, the organ of Corti continues to be affected. Vestibular toxicity also occurs, and positional nystagmus may be the first abnormality noted in cases of ototoxicity. The loss may be unilateral and the ototoxicity may be reversible in 10 to 15 percent of patients. Periodic audiometric testing is recommended in the following high risk groups:

- Patients with impaired renal function
- Those with elevated peak and trough serum levels
- Those with preexisting hearing loss
- Patients with a prior history of ototoxic drug treatment or a history of taking more than one ototoxic drug (especially loop-diuretics)
- Patients exposed to treatment longer than 14 days
- Patients over 65 years of age

Diuretics may potentate the effects of aminoglycosides. When loop diuretics are used alone, they also appear to have a temporary ototoxic effect. Salycylates enter the perilymphatic system and appear to have cochlear effects. This is possibly metabolic, as their effect is typically reversible. Cisplatinum can be both ototoxic and vestibulotoxic. Toxicity is also suggested by otalgia and tinnitus.

## Noise Exposure

Occupational noise exposure is the primary risk factor for noise-induced hearing loss. Approximately 30 to 40 million people in the U.S. work in environments in which harmful noise levels exist. About 50 million people use firearms in the U.S. With these exposures considered, an estimated 11 million people show signs of permanent noise-induced hearing loss. Of these, 5.2 million are between the ages of 6 and 19, representing 12 percent of U.S. children and teenagers. Exposure to loud music in the 1960s accounts for the majority of patients, currently in their sixth decade of life, diagnosed with hearing loss. The

increasing popularity of portable compact disc players and other personal music players, especially with earphones that fit inside the ear canal, has increased the number of affected younger individuals. Noise-induced hearing loss is characterized by a decrease in hearing sensitivity between 3,000 and 6,000 Hz, with a characteristic notch at 4,000 Hz.

The amount of trauma varies with the intensity and length of noise exposure. Noise above 85 dB is harmful, and for every five dB increase in noise, the safe exposure time is cut in half. For example, at 85 dB, eight hours of maximum listening time is recommended by the National Institute of Occupational Safety and Health, but only one hour for 110 dB is recommended. Above 116 dB, noise is unsafe for any length of time. Most personal music devices can generate 91 to 121 dB of sound; using the in-the-ear earphones can increase this by as much as seven to nine decibels. Radios, music players, and other devices should be turned down if one cannot hear someone shouting from three feet away. Personal music devices, with earphones or earpieces, should be turned down if someone else can hear the music.

There have been many educational gains in the last few decades about the risks of noise exposure and in the development of hearing conservation programs. Yet the number of workers with limited English proficiency experiencing noise-induced hearing loss is rising. This may be due to the difficulty in understanding the educational/training programs because of poor access to interpreters or translated information. It is also possible that the use of hearing protectors impedes communication on the job, making it even more difficult for limited English proficient workers to communicate effectively.

## Head Trauma

Trauma to the inner ear may occur with rapid barometric pressure changes, as with scuba diving or flying in a poorly pressurized aircraft. Activities that increase the risk of this type of trauma, including contact sports, should be avoided for children with inner ear anomalies because sudden hearing loss and/or CSF leaks can occur. Head trauma may be blunt or penetrating, and may be peripheral or central. When blunt trauma occurs to the parietal region of the skull and creates fractures in the temporal bone, the fracture will run longitudinally in the bone. Longitudinal fractures account for 75 percent of temporal bone fractures. Minor, concussive sensorineural losses in the high frequencies may occur. Occipital or frontal trauma leads to a transverse fracture of the temporal bone, making up 25 percent of temporal bone fractures. This type of damaging injury can result in severe sensorineural hearing loss and may also cause severe loss of vestibular function. Facial nerve paralysis occurs in 50 percent of these cases. In most cases of traumatic losses, the hearing loss is temporary, but some may progress. Older patients and those with poor initial pure tone averages have a higher risk of these progressive losses.

Concussive effects can occur without fractures. Rapid deceleration-impact injuries can result in ossicular dislocations and fracture the stapes crura. A penetrating trauma may lacerate the external auditory canal, tympanic membrane, or damage the ossicular chain, auditory, or vestibular systems. Welders are at risk for hot spark slag burns of the tympanic membrane, which do not spontaneously heal, and thus may result in a conductive loss. A blunt or penetrating trauma can result in later complications, like stenosis or atresia of the external auditory canal. These can lead to a delayed conductive hearing loss.

## Age-Related Hearing Loss

Presbycusis is the most common sensory deficit in the elderly. Aging of the auditory system can be divided into three categories:

- Age-related degeneration, called presbycusis
- Socioacusis, which is secondary to environmental causes
- Nosocusis, which is due to co-morbid disease, diet, or personal habits such as smoking and drinking alcohol

Nosocusis is most commonly seen in males as a high-frequency, symmetric sensorineural loss in the frequencies above 2,000 Hz. About one-third of people over the age of 65 are affected. There may be a degeneration of hair cells resulting in a high-frequency nerve loss, but normal speech discrimination remains. Degeneration of the stria vascularis within the middle and apical turns of the cochlear can result in an equal loss in all tones. If neurons in the spiral ganglion degenerate, this can lead to poor speech discrimination, but this is uncommon. Lastly, degeneration of the cochlear duct and basilar membrane can lead to a sloping hearing loss with an increase in hearing above 1,000 Hz (also uncommon). Besides the amounts of hearing loss expected from aging, certain factors can increase hearing loss like ototoxic trauma, noise trauma, and family history. Those with cardiovascular disease are eight times more likely to have a hearing loss compared to those of the same age group without heart disease. Cigarette smokers are two times more likely to have a hearing loss compared to nonsmokers.

## Health Care Provider Tips

- Provide newborn hearing screening to all infants rather than just those that meet high risk criteria. Outreach to limited English proficient parents and families. Utilize interpreters for these populations, if needed.
- Early identification is an important first step towards treatment. Continued monitoring of hearing is recommended every three to six months following the initial screen for those infants with identified high risk factors.
- There are many contributors to hearing loss including genetic syndromes, family history, associated disease processes, infections, drug exposure, noise exposure, head trauma, and age. There are different treatment regimes for the different causes of hearing loss.
- Limited English proficient persons may be experiencing noise-induced hearing loss because of their difficulty understanding hearing protection practices discussed in educational/training programs. Furthermore, it is also possible that the use of hearing protectors impedes on-the-job communication, making it more difficult for limited English proficient workers to understand others.

# OCCUPATIONAL EXPOSURE, DISABILITY, AND LEGAL ISSUES

There are approximately 30 to 40 million workers exposed to hazardous, job-related noise each year, according to the National Institute for Occupational Safety and Health (NIOSH). Nine million people are at risk for hearing loss caused by occupational agents, like solvents and metals. Noise-induced hearing loss is the second most commonly reported occupational injury or illness. As common as this occupational injury is in patients, it is important to remember the impact that hearing loss has on the patient's job and personal life.

## Types of Hearing Loss

In certain occupations and industries, acoustic trauma or noise exposures are common. These industries are closely regulated by the Occupational Safety and Health Administration (OSHA). The risk of hearing loss from occupational exposures varies from individual to individual. Occupational risk factors include noise intensity, duration, and frequency exposures. Individual risk factors may include genetic or pre-existing hearing loss caused by medical or environmental factors. Occupational hearing loss typically manifests as sensorineural symmetrical hearing loss with a characteristic notch in assessments. Industries involving welding or solvents make people more prone to middle ear and slag injuries that can result in conductive hearing losses.

## Evaluation and Assessment

Baseline audiometric exams and evaluations by an otolaryngologist are critical to establishing the type of hearing loss. This can also determine whether or not the patient has other underlying etiologies. A thorough history is equally as important and should include questions regarding these considerations:

- Ear injury
- Ear surgery
- Head injury
- Noise exposure outside of existing occupation (such as with hobbies, like hunting)
- Ototoxic drugs
- Pre-existing hearing problems prior to employment
- Other accompanying symptoms such as vertigo and tinnitus
- History of military service with accompanying noise exposure

Most employers with significant rates of occupational noise exposure require yearly screening, usually at their facilities, and are required to have hearing protection devices available for their employees. If possible, take time to inquire about the employer's existing guidelines on hearing protection and encourage the patient to use hearing protection regularly. Encourage patients to find out about their employers' existing hearing conservation program. Provide patients copies of their audiogram so they can present it to their employers' health division.

## Referrals

Appropriate and timely utilization of the otolaryngologist, audiologist, and occupational medicine physician is needed to facilitate accurate documentation for disability and worker's compensation.

If available in your region, occupational medicine is a critical resource after occupational noise-induced hearing loss has been identified. Members of this profession can organize and document the precise information needed once patients raise concern about their hearing loss. If existing guidelines are not enforced, occupational medicine can help advocate for the patient. Audiology assessments and audiometric testing provide critical, objective information which can delineate the type of hearing loss and whether hearing aids are needed.

An otolaryngology referral must be provided to the patient; an otolaryngologist can review and document the comprehensive history. The otolaryngologist can perform a head and neck examination to find the etiology of the hearing loss. From there, the physician can make further recommendations.

## Disability and Impairment

Occupational noise exposure can cause tinnitus and temporary or permanent hearing loss. The American Medical Association's (AMA) 5th edition of *Guides to the Evaluation of Permanent Impairment* is currently the standard used to rate the range of occupational hearing loss. It is critical to thoroughly evaluate and document occupationally induced hearing loss to determine what immediate supports can be provided to the patient. However, as explained in Guides to the Evaluation of Permanent Impairment, "When a physician is asked to evaluate work-related disability, it is appropriate for a physician knowledgeable about the work activities of the patient to discuss the specific activities the worker can and cannot do, given the impairment." Special tables and formulas have been created to determine the level of hearing impairment in each ear separately and together-termed as a binaural impairment. These numbers are used to create the individual's percent score of the impairment.

## Management and Recommendations

When it comes to protecting patients' existing hearing and preventing further loss, care must be taken to ensure that they feel comfortable accessing the health care system. This can be either within Kaiser Permanente facilities or through the employer's health division. Patients should be encouraged to see a health care provider when they notice a change in hearing or an increase in tinnitus. The primary care physician, mid-level providers, and specialists all have a role in emphasizing the importance of hearing protection, on and off the job. This includes:

- The consistent use of properly fitted earplugs or muffs, either provided by the employer (when on the job) or self-provided (when off the job) when exposed to loud noise
- The prevention of further hearing deterioration by avoiding loud noises whenever possible (e.g., loud music through headphones, firearms use, and power tools use)
- Regular audiometric evaluations to determine if hearing aids are required

A comprehensive team approach in addressing the issue of occupational hearing loss is of utmost importance. This approach includes an awareness of and responsibility by the employer and patient to adhere to OSHA standards. An integrated approach with the primary care provider, otolaryngologist, audiologist, and occupational health physician is essential in mitigating the prevalent problem of hearing loss.

## Compliance and Legal Issues

In-patient, out-patient, and emergency health care provided by hospitals and nursing homes that receive federal financial assistance are required to comply with Section 504 of the Rehabilitation Act. Public health care facilities are regulated under Title II and private health care facilities are regulated under Title III of the Americans with Disabilities Act (ADA). The regulations of Title III of the ADA apply to most doctors and other private health care providers. These require health care facilities to provide patients equal opportunities to participate in and benefit from all services. In other words, people cannot be denied any service because they are deaf or have a hearing loss.

Private health care providers' obligations explained under Title III of the ADA are summarized in the following information. Title III of the Americans with Disabilities Act prohibits discrimination against individuals with disabilities in places of public accommodation. Private health care offices and facilities are considered places of public accommodation. The Department of Justice has issued regulations for the obligations of public accommodation under Title III at 28 C.F.R. Part 36. The Department's analysis of this regulation is at 56 Fed. Reg. 35544 et seq. (1991).

Title III of the ADA applies to all private health care providers, regardless of the size of the office or the number of employees. It applies to providers of both physical and mental health care. Hospitals, nursing homes, psychiatric and psychological services, offices of private physicians and dentists, health maintenance organizations (HMOs), and health clinics are required to adhere to Title III of the ADA. If an office of a doctor, dentist, or psychologist, for example, is located in a private home, the portion of the home used for public purposes, including the entrance, falls under the ADA definition of a place of public accommodation.

A health care provider must communicate effectively with all patients with hearing loss seeking or receiving services. Such individuals may not always be patients of the health care provider. For example, if prenatal classes are offered as a service to both fathers and mothers, a father with a hearing loss must be given auxiliary aids or services to avail him the benefits of the class as other fathers with no hearing loss. Similarly, a deaf parent of a hearing child may require an auxiliary aid or service to participate in the child's health care or to give informed consent for the child's medical treatment. Classes, support groups, and other services that are open to the general public must be accessible for participants with hearing loss. For example, Kaiser Permanente members with a hearing loss have Pocketalkers, TTYs, and interpreter services available at medical centers region-wide.

## Health Care Provider Tips

- Noise-induced hearing loss is the second most commonly reported occupational injury. Nine million people are at risk for hearing loss caused by solvents and metal agents.
- Occupational risk factors for hearing loss include the noise intensity, duration, and frequency of exposures. Asking your patients about their work environments and safety practices can alert you to possible hearing loss.
- Baseline audiometric exams and an evaluation by an otolaryngologist are needed to establish the type of hearing loss and to examine the possibility of other underlying etiologies. Completing a thorough history is equally important.
- Most employers with significant occupational noise exposure require yearly screenings, usually at their facilities, and must make hearing protection devices available to their employees.

- Facilitate accurate documentation for disability and worker's compensation. Appropriate and timely utilization of the otolaryngologist, audiologist, and occupational medicine providers is recommended.
    - § If available in your region, occupational medicine is a critical resource once a potential hearing loss has been identified.
    - § The audiology assessment and audiometric testing provide objective information to delineate the type of hearing loss and whether hearing aids are needed.
    - § An otolaryngology referral needs to be given so the physician can review and document the process. The provider reviews the comprehensive history, performs the head and neck examination, and makes further recommendations.
- Emphasize to patients the importance of hearing protection, on and off the job.
- Advocate for a comprehensive team approach to address a patient's occupational hearing loss.

# IDIOPATHIC SUDDEN SENSORINEURAL HEARING LOSS

## Introduction

Idiopathic sudden sensorineural hearing loss (ISSNHL) can be defined as greater than 20dB of hearing loss in at least three audiometric frequencies and occurring within 3 days or less. The reported incidence is five to 20 people per 100,000 every year. The reported recovery rate, without treatment, ranges from 32% to 65%.

There are two theories behind the etiology of ISSNHL. The first suggests that a loss of blood supply to the inner ear through thrombosis, hemorrhage, or vasospasm results in ISSNHL. Some evidence exists to support this hypothesis. For instance, in one study, oxygen concentration in the inner ear of patients with ISSNHL was 30% lower than in controls. Disturbances in the microcirculation of the inner ear in patients with ISSNHL have also been identified. Viral inflammation of the cochlea and/or inner ear, possibly secondary to a recent upper respiratory infection, is the second theory behind the etiology of ISSNHL. Patients with ISSNHL have seroconverted to several viruses after infection. Histopathology performed on temporal bones from patients, who had a history of ISSNHL, have results consistent with viral infections.

At this time, the only treatment for ISSNHL shown efficacious in rigid clinical trials is systemic corticosteroid therapy. However, the reported success rate of this treatment varies from 49% to 79%. There is evidence to suggest that intratympanic dexamethasone (IT-Dex) therapy improves treatment success by increasing intra-cochlear corticosteroid concentration via the round window membrane. Success rates in pilot studies range from 66% to 80%. IT-Dex therapy can also reduce the incidence of toxic side effects associated with systemic corticosteroid therapy. It may also be an alternative therapy in treatment of those with diabetes.

## Patient History

The evaluation and treatment of patients with sudden hearing loss should ideally be performed within 7 days. The first priority is to rule out other potential causes of hearing loss, like autoimmune hearing loss or perilymph fistula, which have proven treatments. A thorough history is essential to determine if there is pre-existing hearing loss or if the current hearing loss was sudden or progressive over several days. Important considerations for correct treatment include: an upper respiratory infection in the previous 1-2 weeks, a history of hearing fluctuation, a recent ear infection, surgery, hospitalization, exposure to ototoxins (aminoglycosides), trauma, drainage, vertigo, tinnitus, pain, or a family history of hearing loss. Medical conditions associated with sudden hearing loss include diabetes, syphilis, chronic renal disease, and cardiovascular disease.

## Physical Examination

The physical examination of patients with ISSNHL is unremarkable. The examination of the tympanic membranes in cases of ISSNHL appears normal and the Weber fork exam lateralizes to the unaffected side. Neurological exams are also normal.

## Work-up

If the history and physical exam are consistent with ISSNHL, an audiogram is ordered to assess the type of hearing loss. FTA-abs is ordered to rule out syphilis and an MRI of the internal acoustic canal (IAC) is

ordered to rule out acoustic neuroma. Acoustic neuromas can be associated with sudden hearing loss up to 25% of the time.

## Prognosis without Therapy

With ISSNHL cases, mild low-frequency losses have the best prognosis. Patients with a flat profound loss have the worst prognosis. Spontaneous recovery to functional levels is thought to occur 32% to 65% of the time without treatment. This recovery takes place over a period of 2 to 3 weeks, after which recovery rates steadily decline. Minimal improvements are observed after 1 to 2 months.

## Standard Treatment

Patients within 14 days of the onset of ISSNHL are treated on a prednisone taper beginning with 60mgs/day for 7 days, followed by a 7 day taper. The prednisone is intended to counter the inflammatory effects of a viral inner ear infection, as opposed to being immunosuppressive. Patients are counseled on the side effects of prednisone, especially if patients have a history of diabetes, ulcers, are pregnant, or breast-feeding. Audiograms obtained within 4 weeks after the taper determine if the therapy was effective. Hearing aids are offered to those patients as appropriate. Follow-up is on an as-needed basis.

## Failed Therapies

Many treatments for ISSNHL have been tested but have failed. These include hyperbaric oxygen, free radical scavenging vitamins, gingko biloba, magnesium treatment, agents that decrease blood viscosity (dextran, pentoxifylline, procaine, heparin), and vasodilatory agents (histamine, papverine, verapamil, carbogen). Carbogen (95% oxygen and 5% carbon dioxide) inhalation therapy is thought to lead to increased cochlear blood flow by overriding intracranial vascular autoregulation, but it has shown mixed results and its use remains controversial.

The efficacy of anti-viral medications has been evaluated in prospective, randomized, double-blind, placebo-controlled, multi-center clinical trials. Anti-viral medication did not provide more benefit to patients than steroid treatment alone in the treatment of ISSNHL in two trials. However, earlier animal studies showed faster hearing recovery and less extensive cochlear destruction when acyclovir was added to steroids in the treatment of HSV-1 induced labyrinthitis.

## Intratympanic Steroids as New Therapy

In 1980, Wilson et al. published a double-blind, prospective study examining the efficacy of oral steroid treatment for ISSNHL. Sixty-seven patients enrolled in the study; of these patients, 33 received steroids and 34 received a placebo. Another 52 patients refused treatment and were counted as control subjects. After matching for age and vertigo, patients that presented a mid-frequency hearing loss and received oral steroids were found to have significantly better hearing, especially those younger than 40 years old. Wilson also reported that 78% of patients that presented moderate hearing loss improved on steroids compared to 38% in the placebo group.

Since then, very few rigid follow-up studies have been performed confirming the efficacy of oral steroid treatment for ISSNHL. Moskowitz et al. performed a prospective, randomized trial comparing a steroid treated group with a placebo group. Eighty-nine percent of the patients in the steroid group recovered functional hearing compared to 44% in the placebo group. The study, however, was not blinded.

Most of the research since Wilson's study has been retrospective. Some have indicated that ISSNHL is responsive to steroid therapy, while others have not. Fetterman reviewed the treatment of 837 patients with ISSNHL and his data suggested that the best recovery took place in patients receiving steroids,

though statistical significance was not achieved when comparing this group to control subjects. In 1984, Byl published an eight-year study questioning the efficacy of steroid treatment for ISSNHL

Some studies exist which suggest that steroids are not efficacious in treating ISSNHL. Kanzaki et al. prospectively followed 183 patients with ISSNHL from 1980 to 1985 and found no significant difference between patients treated with steroids and those in the control group. Cinamon et al. also published a prospective, double-blind, placebo controlled study on 41 patients and found no improvement in the hearing results of the steroid treated group. Thus, controversy continues regarding the efficacy of oral steroids in the treatment of ISSNHL.

Intratympanic dexamethasone (IT-Dex) provides a higher intra-cochlear concentration of medication than oral administration, suggesting the possibility that intratympanic delivery is more effective in treating ISSNHL than oral steroids alone. Preliminary studies using IT-Dex for ISSNHL show promising results. In a study by Chandrasekhar, 10 patients with ISSNHL were treated sequentially with IT-Dex. Eight out of 10 patients experienced improved hearing. Gianoli et al. showed that 44% of patients with ISSNHL who failed oral steroid therapy responded to intratympanic corticosteroid therapy. Kopke et al. confirmed this result by showing that 4 out 6 patients who had failed oral steroids responded to intratympanic steroid therapy for up to 6 weeks after the initial hearing loss. These initial studies indicate that intratympanic steroid therapy may not only be more effective than oral steroid therapy but may have a longer therapeutic window. The therapeutic window is 6 weeks for intratympanic steroids, but only 10 to 14 days for oral steroids. Complications from intratympanic steroid therapy were not noted in these initial studies.

The Head and Neck Surgery Department at Kaiser Permanente San Diego has been conducting a prospective, randomized, double-blinded, multi-centered trial comparing the hearing results of ISSNHL patients who have received IT-Dex and/or systemic corticosteroid therapy. Patients with less than a 6-week history of ISSNHL are randomized to one of three arms. One group of patients is given IT-Dex therapy, another is given standard therapy with systemic corticosteroids, and the remaining patients are given IT-Dex plus systemic corticosteroids. Patients are categorized as responsive to therapy if their post-treatment audiogram improves to within 50% or more of their pre-hearing loss speech reception score or averaged pure-tone score. Fifty patients have enrolled in the study to date and hearing results for all three treatment arms are regularly evaluated for statistical significance.

## Method of Intratympanic Steroid Injection

IT-Dex can be administered up to 6 weeks after the initial event, thus providing a longer therapeutic window than oral steroid therapy which is traditionally said to fail if not given within 10-14 days. Typically, a 12 mg/ml solution of dexamethasone is used in a buffered solution. For its application, the patient is placed in a supine position, and the ear canal is then irrigated once with alcohol and completely dried. A small needle stick is then made in the anterosuperior quadrant of the tympanic membrane (as a blowhole) and then 0.5 mls of dexamethasone is injected at body temperature into the posterior/inferior quadrant. Topical anesthetic is not used because it is more painful than the needle stick alone and puts the patient at risk for perforation through necrosis of the tympanic membrane at the site of use. The patient is left supine, with the head turned so the injected ear is facing upward, and the head is lowered for 20 minutes. The patient will return after one and two weeks for re-test audiograms and repeated injections. Residual perforations can be treated with a routine paper patch, though probably uncommon. A final audiogram is performed 4 weeks after the final injection.

## Health Care Provider Tips

- ISSNHL traditionally requires treatment within 7 days of the onset of hearing loss.
- ISSNHL is a diagnosis of exclusion and needs evaluating by an otologist or neurotologist. Do not independently treat ISSNHL with a prednisone taper.
- For the work-up, make sure to order an audiogram and an MRI to rule out an acoustic neuroma.
- Standard treatment for ISSNHL, which may be given within 14 days of diagnosis, is a prednisone taper over two weeks once diagnosis is confirmed.
- Many therapies have been tried but have failed, except for intratympanic dexamethasone, a promising new therapy that is currently being evaluated in Kaiser San Diego's Head and Neck Surgery Department. This is a prospective, randomized, double-blinded, multi-centered trial.

# AUTOIMMUNE HEARING LOSS

The concept of immune-mediated hearing loss, or Autoimmune Inner Ear Disease (AIED), was first described and published by Dr. Brian McCabe of the University of Iowa in 1979. Since then, research aimed at identifying the underlying processes of AIED continues to grow, but understanding of the problem continues to be inadequate. Due to the lack of an identifiable pathologic mechanism for this type of hearing loss, the ability to successfully treat AIED is hampered.

By definition, AIED refers to a rapidly progressive (over a course of weeks to months) sensorineural hearing loss that responds to the administration of corticosteroids. There are several theories to explain the cause of AIED. The cross-reaction theory is currently the most compelling causation. It proposes that the inner ear shares common antigens with a foreign agent such as a virus, bacteria, or chemical substance that the body's immune system is combating. These shared antigens in the inner ear may then become targeted by antibodies or rogue T-cells causing damage to the inner ear. The bystander theory suggests that damage to the inner ear releases cytokines, which then provoke delayed, additional immunologic reactions. This can create a cyclical set of symptoms with attacks and remissions. Some variants of Meniere's syndrome may be related to such a mechanism. The theory of intolerance, currently out of favor, states that the ear, similar to the eye, has partially privileged immune status. This means that the body's immune system has an incomplete knowledge of the normal inner ear antigens. Therefore, after some event (i.e., surgery, infection, trauma), these antigens may be released causing the body to incorrectly identify them as foreign and then mount an immunologic attack. Finally, there may be genetic factors that increase an individual or family's susceptibility to immunologic damage to the inner ear.

AIED may be associated with traditional autoimmune diseases such as systemic lupus erythematosis, Söjgren's syndrome, ankylosing spondylitis, Cogan's syndrome, ulcerative colitis, Wegener's granulomatosis, rheumatoid arthritis, Bechet's disease, and scleroderma. About 80 percent of AIED cases have no identifiable systemic autoimmune disease. The hearing loss in AIED is reported to be bilateral in 80 percent of cases. Associated symptoms such as tinnitus (83 percent) and vestibular dysfunction (79 percent) are common. Some researchers feel that 15 to 20 percent of bilateral Meniere's syndrome cases may be due to immunologic dysfunction. Some forms of AIED may have predominantly vestibular symptoms with little or no hearing loss. There does not appear to be a gender predilection in the disease. Positive response to the administration of corticosteroids remains the most useful diagnostic criteria.

Autoimmune inner ear disease as a cause of hearing loss is a diagnosis of exclusion, since the condition is rare. It causes less than one percent of all cases of hearing loss or dizziness. A patient presenting with documented progressive sensorineural hearing loss should have blood tests performed that check for possible metabolic and infectious causes, and autoimmune screening. The following tests may be ordered at the physician's discretion based on clinical suspicion: anti-nuclear antibodies (for lupus), erythrocyte sedimentation rate (for Cogan's), anti-gliadin antibodies (for Celiac disease), thyroid stimulating hormone (for thyroid disease), fasting blood glucose (for diabetes), syphilis screening, and Lyme titer. Broader screening for autoimmune disease may be done with Raji-Cell assay, rheumatoid factor, and anti-smooth muscle antibodies. If two or more of these latter tests are positive or the hearing loss rapidly worsens, perform a Western Blot Assay. This is to check for antibodies directed against 68 kD (heat shock protein-70) and P0 inner ear antigens. The reliability of these anti-cochlear antibody tests remains controversial. Some authors feel their specificity and sensitivity are poor, and that the tests may be directed at the wrong immunologic targets.

An MRI, with and without gadolinium, focusing on the internal auditory canals and cerebellopontine angles should be performed to check for possible tumors, pachymeningitis, or meningeal carcinomatosis. In addition to the contrast enhanced images, the T-2 axial images should be carefully reviewed to identify any inner ear malformations such as an enlarged vestibular aqueduct or Mondini variants.

Treatments for AIED remain limited. The hallmark of positive corticosteroid response for diagnosis remains the mainstay of treatment. A high dose trial of corticosteroids, usually prednisone or decadron, is taken for four weeks. The patient must be warned of the risks of high dose corticosteroid use such as cataract formation, avascular necrosis of the hips, etc. If effective, then a very gradual taper to the lowest dose possible, to maintain stable hearing, is undertaken. Long term treatment is fraught with side effects of prolonged steroid use. Cytotoxic agents, such as methotrexate and cyclophosphamide, have been shown to be ineffective in AIED. Anti-tumor necrosis factor drugs, such as etanercept, are being currently studied. The initial optimism generated by animal studies of the reduction of inflammation and hearing loss in acute immune-mediated labyrinthitis does not appear to bear out in recently published human pilot studies. Finally, should patients progress to the point that they do not benefit from conventional amplification, cochlear implantation has been demonstrated to be successful in these individuals.

## Health Care Provider Tips

- Autoimmune Inner Ear Disease (AIED) refers to a rapid and progressive sensorineural hearing loss that responds to the administration of corticosteroids. It may be caused by a variety of theories including (1) cross-reaction, (2) bystander, and (3) intolerance.

- Genetic factors may increase an individual's or family's susceptibility to immunologic damage of the inner ear.

- AIED may be associated with traditional autoimmune diseases.

- Note that a patient presenting with documented progressive sensorineural hearing loss should have blood tests that check for possible metabolic and infectious causes, and autoimmune screening. Autoimmune inner ear disease as the cause of hearing loss is a diagnosis of exclusion, since the condition is rare.

- Although positive corticosteroid response for diagnosis remains the mainstay, treatment of AIED remains limited. The patient must be warned about the risks and possible side effects of high dose corticosteroid use.

- Cochlear implantation has been demonstrated to be successful with patients that progress to the point that they do not benefit from conventional amplification.

# PEDIATRIC POPULATION

## Education

Providers in the medical community have the opportunity and professional responsibility to help families connect and collaborate with the appropriate school personnel who provide services to students with hearing loss. This chapter outlines present policies addressing children with disabilities and hearing loss. The final section summarizes steps to identify children with hearing loss in the educational system.

### Policies and Legislation

Federal legislation mandates services for children with disabilities and those who have a hearing loss meeting specific criteria. The Individuals with Disabilities Education Act (IDEA) is the most recent law, reauthorized in 2004, pertaining to children with disabilities. IDEA requires access to a free and appropriate education (FAPE) for all children with disabilities. Part B of IDEA, the Education of Children with Disabilities program, pertains to preschool and school-age children, ages 3 to 21.

Section 504 of the Rehabilitation Act of 1973 is commonly referred to as the civil rights legislation for the disabled. This was the first law specifically protecting the rights of disabled persons by prohibiting recipients of federal funds from discriminating against "otherwise qualified individuals." Section 504 and IDEA have many similarities that together provide comprehensive protection to all children, whether or not they are identified as disabled under special education statutes.

The Americans with Disabilities Act (ADA), modeled after the Rehabilitation Act of 1973, replaces the word "handicap" with "disability," and pertains to all employers, not just those receiving federal funds. In the educational setting, the ADA helps assure that information is accessible and useable by students with communication disabilities, whether or not they qualify for special education services.

### Identification

IDEA requires states to identify children with disabilities, including hearing loss, residing in each state. The process of identifying these students, however, varies from state to state. Most states have mandates or regulations, and "in all states with mandates, legislation directs the agency responsible for overseeing school-based hearing screening activities." According to Penn, the most common authoritative agencies responsible for state hearing screening programs are State Departments of Health and Education, either independently or cooperatively. "Appendix A" summarizes school-based hearing screening programs for some states Kaiser Permanente serves. It also includes information on target populations, test frequencies, screening levels, and referral criteria.

While federal and state government requires hearing screening to be available for all children, it is important to remember that home schooled students and students at private schools may not receive recommended hearing screenings.

Besides federal and state regulations, there are professional guidelines that provide a framework for hearing screening programs and professional codes of conduct. The Guidelines for Audiology Services in the Schools specifies an ongoing identification program: "Each year the identification program should provide screening for all children at specified ages or grade levels; all children referred for or placed in special education programs; all children referred by parents, teachers, and concerned third parties; and all children considered 'at-risk' for a hearing loss including students with a history of exposure to noise."

Furthermore, the guidelines cited above indicate, "to be effective, the identification program must develop expedient lines of communication and referrals between educators, families, and the medical

community." Collaboration with service providers for students results in more efficient and cost-effective service delivery, a smoother referral process for medical, audiological, and educational follow-up, and informed families who will have improved access to appropriate services for their children.

Although private sector audiologists play an important role in the evaluation and management of childhood hearing loss, there are advantages the educational audiologist can provide to families and students, or any child whose hearing is questionable. Some advantages are listed below.

- Testing is provided free of charge.
- The audiologist can spend as much time and as many sessions needed to obtain thorough audiometric information. Some children need several sessions to be conditioned to respond to sounds consistently or to establish a comfort level with the examiner; this takes time and repeated visits.
- For some children, the office of an educational audiologist is not as intimidating as a medical office.
- The audiologist readily has access to information on the student's auditory functioning in the classroom and on the playground.
- The audiologist coordinates the fitting of assistive technology, such as the appropriate personal FM equipment provided by the school district, to interface with hearing aids fit by private dispensers.

The audiologist working in a school setting is aware of the educational effects of hearing loss and this can help identify co-existing conditions that may affect children's academic progress. "It is not unusual for parents and school personnel, who are generally not aware of the effects of hearing losses, to assume that all of children's deficits are related to their hearing losses, when in fact there may be other conditions that need to be addressed."

## Placement Process for IDEA and 504

The process of the assessment and placement component for IDEA or 504 begins with a concern about a child identified by the parent, teacher, school nurse, physician, or other individual or agency. The first step is a hearing screening or audiological assessment to confirm or rule out hearing loss as the problem. The assessment can help to determine the appropriateness of a special education referral.

For the Individual Education Program (IEP) process, a pre-referral meeting (known by different names in each state) with the teacher and special education provider is held prior to making the formal special education referral. The purpose of the meeting is to discuss the child's problems, difficulties, and needs. This informs the group of possible strategies and supports to assist the child prior to a referral and multidisciplinary assessment. If the team recommends a special education referral, they may determine what assessments will be needed, who will conduct them, who will obtain parent permission, and when the IEP meeting will occur.

This meeting is not necessary for children with obvious special education needs, like those with significant sensory disabilities. When a multidisciplinary assessment is recommended, obtain parental permission and notify the parents of their rights before the assessment is commenced. The assessment obtains information on the child's functioning to determine eligibility for special education.

Eligibility and disability are somewhat confusing terms. Completed assessments indicating the needs of the child determine the presence of a disability. Once a disability is documented, eligibility for special education and related services is considered by ascertaining if the disability adversely affects educational

performance. For instance, the child may still obtain reasonable benefit from regular education alone. For children, the question of how much loss constitutes a disability cannot be determined by audiometrics alone. Many factors affect the relationship of hearing impairment and the ability to compensate for it. Age of onset, age of intervention, intellectual capacity, neurological function, central auditory processing ability, environment, other health factors, and effects of otitis media are all variables which affect the level and intensity of a hearing loss. For some children, a minimal loss can have significant implications, whereas for others, the same impairment may present no consequences.

According to Johnson et al, for a child to be considered disabled due to a hearing loss, one of the following audiometric criteria should be met:

- A bilateral hearing loss of at least 20 dB PTA exists in the better ear
- An unilateral hearing loss of at least 35 dB PTA is in the affected ear
- A bilateral high-frequency hearing loss exists, averaging at least 35 dB at any two frequencies for 2,000 Hz, 4,000 Hz, or 6,000 Hz
- Or a fluctuating conductive hearing loss meets one of the above criteria for at least three months (cumulative) during the school year or for four months annually

## IEP (Special Education) or 504 plan (Regular Education)

To determine whether a child may be eligible for IEP or a 504 Plan, the IEP team or eligibility committee must review all the assessment results, determine the child's strengths and needs, and then ask the following questions.

1. *Have sufficient assessments been completed, documented, and considered to determine the student's current level of functioning, achievement, and performance; and to determine the student's educational needs?* If "NO," the meeting must be rescheduled so that all necessary assessments are completed.
2. *Can the student receive reasonable educational benefit from regular education alone?* If "YES," the child does not meet special education criteria and the meeting is terminated; the child may then be referred for 504 considerations, if appropriate.
3. *Does the child have a disability which adversely affects educational performance (as defined by state Rules to Administer the Individuals with Disabilities Education Act)?* If "NO," the special education meeting is terminated; the child may then be referred for 504 considerations. If "Yes," proceed with the development of goals and objectives for the IEP.

The eligibility for a 504 plan is determined when a disability substantially limits one or more major life activities. If the student meets the disability criteria, then 504 needs are identified and a plan is developed. It is also possible that children could receive services under both IDEA and 504, if they are eligible as "disabled" under IDEA.

### Individual Education Program (IEP)

An Individual Education Program (IEP) addresses the academic and support services needed by the student with hearing loss as identified by a multidisciplinary team. Communication skills, cognitive abilities, motor functioning, social-emotional development, adaptive behavior, health history, and academic status must all be considered when creating the IEP.

The *Guidelines for Audiology Services in the Schools* states:

> "Educational services may be provided through a number of delivery options, including, but not limited to, home intervention, consultation/collaboration, itinerant instruction, team teaching, resource special education, self-contained special education classes, and residential placement."

Regarding students' goals and needs in these programs, some of the most important aspects of habilitation are:

- Medical treatment, when indicated
- Selection of appropriate amplification such as a hearing aid, cochlear implant, and/or FM system at the earliest possible age
- Auditory skill development training
- Training in the use of hearing aids in various settings, including use of amplification in noisy classrooms and social situations
- Structuring a successful learning environment including teacher selection, optimal room acoustics, accessibility to information, and peer and teacher in-services
- Development and remediation of communication, including pragmatic skills
- Training in the use of visual information to supplement auditory input
- Academic tutoring or specialized instruction
- Facilitated transitions between programs, agencies, and vocational settings
- Counseling

The habilitative needs of children with hearing loss may be met using a variety of service delivery models and a diverse group of service providers. This group can include audiologists, speech-language pathologists, teachers, psychologists, counselors, social workers, physical therapists, occupational therapists, nurses, and physicians. The service delivery model used will depend on the administrative philosophy of the individual state, local school systems, and available resources. However, "all states must ensure that local education agencies provide the essential service components necessary to meet state and federal education and civil rights statutes and regulations."

Least restrictive environment (LRE) is an IDEA concept that has been controversial when applied to the education of children meeting disability criteria. The pendulum for education of deaf students has swung from placement in separate classrooms and facilities, to mainstreaming (in the 1970s), to the inclusion movement of the 1980s, and most recently, to a growing movement back to center-based and multi-district shared programs.

For children, the primary issue in the LRE discussion continues to be: what roles do Deaf culture and having a peer-group with hearing loss play in their education experience? We know that the socialization aspects of education have significant consequences for academic as well as non-academic development. Each child should have access to a continuum of placement options including home school, regular education, center-based programs, and regional and state schools for the deaf.

### 504 Plan

Section 504 is a regular education program; under its authority, schools must make adequate provisions so that students with disabilities are not excluded from, denied benefits of, or subjected to discrimination under any program or activity because facilities are inaccessible or unusable. Johnson et al points out that most children with hearing loss (determined by specific audiometric criteria) should meet 504 disability requirements since they have a sensory impairment that impacts education by "substantially limiting major life functioning in the areas of hearing, speaking, and learning." Typical services and accommodations that might be provided to these students under a 504 plan include amplification (personal FM or sound field systems), note takers or interpreters, classroom and environmental accommodations, instructional modifications, and assistive technology (such as captioning, or TDD).

While legislation and policies are crucial to assuring that all students with hearing loss receive appropriate education services, developing and maintaining partnerships with families is equally important to empower them to advocate for their children's best interests.

## Auditory Neuropathy

Auditory neuropathy is a unique hearing disorder in which the inner ear, or cochlea, is normal but the transmission from the cochlea to the auditory brainstem is impaired. Some scientists believe the disorder is caused by damage to the inner hair cells of the cochlea that transmit information to the auditory cortex. Others believe the disorder lies in the brain. It can affect people of all ages, from infancy through adulthood. Although the condition is rare, it is unknown how many people are actually affected by this disorder. Auditory neuropathy is also referred to as auditory dys-synchrony.

People with auditory neuropathy can exhibit any type of audiogram, from normal to severe. The characteristic symptom of auditory neuropathy is poor speech perception abilities, or the inability to understand speech clearly. Speech discrimination is typically worse than predicted by the degree of hearing loss depicted on the audiogram; the person can hear sounds but cannot understand the meaning of the words. Hearing in a noisy environment is often worse. Auditory neuropathy is also characterized by fluctuations; for example, understanding appears to be better on one day than another. Research has shown that some infants who have been diagnosed with auditory neuropathy improve and start to hear and speak within a year or two. Other infants stay at the same level or become worse. Adults with auditory neuropathy can have hearing that remains stable, fluctuates up and down, or progressively worsens depending on the underlying cause.

The cause of auditory neuropathy is not clear. Some children who have been diagnosed with auditory neuropathy had medical difficulties as a newborn such as jaundice (hyperbilirubinemia), premature birth, low birth weight, or inadequate oxygen supply. Auditory neuropathy also has a genetic component, noted by its presence in generations of some families. Some patients with neurological disorders, such as Charcot-Marie-Tooth Syndrome and Friedriech's ataxia, have been shown to have auditory neuropathy.

Auditory neuropathy is diagnosed using auditory brainstem response (ABR) tests and otoacoustic emissions (OAE) tests. The classic sign of auditory neuropathy is a normal OAE test result and an abnormal ABR test result. OAE measures the outer hair cells of the inner ear, which are not affected by auditory neuropathy. In newborn hearing screening, automated versions of both the ABR and OAE tests are used. Since many of the infants diagnosed with auditory neuropathy are in the Neonatal Intensive Care Unit (NICU), many states recommend that only automated ABR should be used for screening this population.

Researchers are looking for effective treatments and there remains much controversy in the profession about treatment options. Hearing aids, cochlear implants, listening devices (such as FM systems), and sign language techniques have been tried and reported to have success in some patients and not for others. The diagnosis of auditory neuropathy is very challenging for families. Each child is unique and may respond to

the various treatments and communication options differently. This requires a team approach with the family and other caregivers. Families should seek out professionals with an expertise in auditory neuropathy who can provide unbiased information regarding treatment and communication options.

There are two main philosophies of how to teach infants and children with auditory neuropathy to communicate. One philosophy promotes using sign language to help the child develop language since spoken language is often difficult for the child to understand. The second philosophy encourages the use of listening skills and spoken language together with technologies such as hearing aids and cochlear implants. A combination of these two approaches can also be used. Adults with auditory neuropathy and older children who have already developed spoken language may benefit from learning how to speechread.

Researchers are working to understand the causes of auditory neuropathy and are searching for genes that may be involved in causing this condition. Researchers are also continuing to investigate the potential benefits of cochlear implants for children with auditory neuropathy and are examining why cochlear implants may benefit some people with the condition but not others.

## Recurrent Otitis Media and Hearing Loss

Recurrent otitis media is one of the most common medical disorders in children and is the most common cause of hearing loss in children. Children who are at high risk for recurrent otitis media include those with a cleft palate or any craniofacial abnormalities, a family history of otitis media with effusion, and a pre-existing sensorineural hearing loss. A careful history obtained from the parents, the physical examination, and the diagnostic evaluations are critical in determining the impact of otitis media on the child's hearing. Often, the signs and symptoms are not ear pain and drainage alone. Pulling on the ears, complaint of pressure in the ears, turning the television volume up loud, poor pronunciation of words, sitting too close to the television, or seeming to ignore one's parents when called, are all signs that a child with recurrent ear infections may have a fluctuating or permanent hearing loss. Whether it fluctuates or is persistent, hearing loss is still perhaps the most common complication of otitis media.

All children with recurrent otitis media or chronic otitis media with effusion should have their hearing assessed. The purpose of this is to determine the presence and type of hearing loss (e.g., conductive, sensorineural, or mixed hearing loss) and the degree of hearing loss through further audiometric testing and an otolaryngology referral. The usual finding on an audiometric examination is a mild to moderate conductive hearing loss which is frequently reversible with resolution of the effusion. However, causes of permanent conductive hearing loss may be related to recurrent or chronic inflammation within the middle ear, such as adhesive otitis, tympanosclerosis, or discontinuity of the ossicles. Permanent sensorineural hearing loss can also occur when an infection spreads through the round or oval window membranes. Essentially all children should have their hearing tested when there is a history of recurrent infections, chronic infections, or the presence of a chronic effusion.

Behavioral testing, tympanometry, otoacoustic emissions, and brain stem auditory evoked response are all methods of testing with the reliability of some being determined by the cooperativeness of the child. In the child who already has a known pre-existing hearing loss and recurrent otitis media, or chronic otitis media with effusion, the threshold for referral to the otolaryngologist should be much higher, given the higher risk of further damage to the child's hearing. The timing of these infections occurring in the child's life can impact speech and language development and performance in school; thusly, a heightened awareness of otitis media by the practitioner and the parents is important. A child who is severely or profoundly deaf and has otitis media may not show clear signs of infection. This may prove to be more of a challenge for parents to pick up on cues prior to an actual examination.

Nonsurgical management of hearing loss resulting from otitis media can include: valsalva maneuver, antimicrobials, corticosteroids, decongestants, antihistamines, allergy controls, and careful monitoring.

Surgical interventions may include myringotomy with or without ventilation tubes, adenoidectomy with or without tonsillectomy, tympanoplasty, or tympanomastoidectomy.

In conclusion, it is important that health care providers:

- Obtain a thorough history from the parents of the child with recurrent otitis media
- Look for signs of hearing loss
- Utilize audiometric testing to identify hearing loss in children with recurrent or chronic otitis media
- Have a heightened awareness of children with pre-existing hearing loss—with these children one should have a lower decision threshold to refer patients to further otolaryngology evaluation
- Recognize high risk groups and risk factors
- Consider the impact of recurrent otitis media and hearing loss on a child's behavioral, speech, language, and educational development
- Be aware of surgical and nonsurgical therapeutic options

## Newborn Hearing Screening and Speech Development

Newborn hearing screening, prior to hospital discharge, is rapidly becoming the standard of care in the United States. Newborn hearing screening identifies two infants out of every 1,000 births with hearing loss. Until the advent of newborn hearing screening, most children were not identified until the age of two. Hearing loss in infants can negatively impact speech and language acquisition, academic achievement, and social and emotional development. If detected, however, these negative impacts can be diminished and even eliminated through early intervention. Research has demonstrated that infants who are screened, diagnosed, and receive early intervention by six months of age have near normal language outcomes by age three. Those infants whose hearing loss is detected later than six months often demonstrate delays the rest of their lives. Most states have implemented Early Hearing Detection and Intervention (EHDI) programs through their state health departments. The Joint Committee on Infant Hearing, the National Institutes of Health, and the Maternal Child and Health Bureau have endorsed EHDI programs. [See the Bibliography for print and online resources.]

Hearing screening is performed using objective technology. There are two technologies used primarily in the newborn nursery. An otoacoustic emission (OAE) measures the integrity of the hair cells in the cochlea (inner ear) by emitting a sound into the ear via a probe. In a normal hearing ear, the hair cells echo the sound and a computer determines if the response is normal. Auditory Brainstem Response (ABR) measures the brainwaves in response to sound. Both technologies are automated and can be used by a variety of trained personnel. If a baby does not pass the hearing screen before hospital discharge, the parents are then instructed to return to the hospital for a rescreen or referred directly to an audiologist. An audiologist with equipment and expertise in assessing infants can determine what type and degree of hearing loss the infant may have.

Infants below six months of age should have a complete battery of tests that include diagnostic ABR, OAE, high-frequency tympanometry, and behavioral observation audiometry. Behavioral testing alone is not appropriate for this age. Even a mild hearing loss can cause speech and language delays. Infants who have a permanent sensorineural hearing loss can be fitted with hearing aids immediately. Infants are then enrolled in an early intervention program to assist with the development of normal speech and language.

Newborns born in alternative birthing facilities, including home births, should have access to screening before one month of age.

Even if infants pass a newborn hearing screening, they could develop hearing loss later. If an infant has a high risk factor, such as a family history of childhood hearing loss, the baby should be monitored with ongoing audiologic testing. Traditionally, children with unilateral hearing loss are expected to develop normal speech and language. However, one-third of these children must repeat a grade in school. Close monitoring of speech and language outcomes for these children is important. Infants with a unilateral hearing loss are at a high risk for developing hearing loss in the other ear. These infants should have audiologic monitoring every three months until the age of three. Whenever parents express concern about their child's hearing or speech, they should be referred immediately to Child Find or an audiologist for screening and diagnosis. [See the Bibliography for print and online resources.]

## Speech and Language Development

Speech is defined as the production of sounds of a language organized into words or word groups. Language is the organized system of symbols that individuals use to communicate. Language can be spoken, written, or signed. Receptive language is the ability to comprehend language and expressive language is to communicate appropriately.

The ear is fully developed in the young fetus at four months gestation. Language begins developing in utero as soon as the fetus hears its mother's voice. Young children communicate before they are able to talk, using eye contact, crying, vocalizing, gestures, speech, and sign language. Each child is unique and develops speech and language at her or his own rate. Below is a chart showing a timeline for speech and language development. If a child does not meet the majority of the milestones in one age range, the patient should be referred to an audiologist (to rule out hearing loss) and to a speech/language pathologist. Remember that hearing can be tested at any age.

**Table 4: Normal Pediatric Speech and Language Development**

| Hearing and Understanding | Talking |
|---|---|
| **Birth–3 Months**<br>• Startles to loud sounds<br>• Quiets or smiles when spoken to<br>• Seems to recognize a familiar voice and quiets if crying<br>• Increases or decreases sucking behavior in response to sound | **Birth–3 Months** |
| **4–6 Months**<br>• Moves eyes in direction of sounds<br>• Responds to changes in tone of your voice<br>• Notices toys that make sounds<br>• Pays attention to music | **4-6 Months**<br>• Babbling sounds more speech-like with many different sounds, including p, b and m<br>• Vocalizes excitement and displeasure<br>• Makes gurgling sounds when left alone or when playing |
| **Hearing and Understanding** | **Talking** |
| **7 Months-1 Year**<br>• Enjoys games like peek-a-boo and pat-a-cake<br>• Turns and looks in direction of sounds<br>• Listens when spoken to<br>• Recognizes words for common items like "cup," "shoe," and "juice"<br>• Begins to respond to requests ("Come here," "Want more?") | **7 Months-1 Year**<br>• Babbling has both long and short groups of sounds such as "tata upup bibibibi"<br>• Uses speech or non-crying sounds to get and keep attention<br>• Imitates different speech sounds<br>• Has one or two words (bye-bye, dada, mama) although they may not be clear |
| **1-2 Years**<br>• Points to a few body parts when asked<br>• Follows simple commands and understands simple questions ("Roll the ball," "Kiss the baby," and "Where's your shoe?")<br>• Listens to simple stories, songs, and rhymes<br>• Points to pictures in a book when named | **1-2 Years**<br>• Says more words every month<br>• Uses some 1-2 word questions ("Where kitty?" "Go bye-bye?" "What's that?")<br>• Puts 2 words together ("more cookie," "no juice," and "mommy book")<br>• Uses many different consonant sounds of the beginning of words |
| **2-3 Years**<br>• Understands differences in meaning ("go-stop," "in-on," "big-little,", and "up-down")<br>• Follows two requests ("Get the book and put it on the table.") | **2-3 Years**<br>• Has a word for almost everything<br>• Uses 2 to 3 word "sentences" to talk about and ask for things<br>• Speech is understood by familiar listeners most of the time<br>• Often asks for or directs attention to objects by naming them |

| | |
|---|---|
| 3-4 Years<br>• Hears when called from another room<br>• Hears television or radio at the same loudness level as other family members<br>• Understands simple, "who?," "what?," "where?," and "why?" questions | 3-4 Years<br>• Talks about activities at school or at friends' homes<br>• People outside family usually understand child's speech<br>• Uses many sentences that have four or more words<br>• Usually talks easily without repeating syllables or words |
| **Hearing and Understanding** | **Talking** |
| 4-5 Years<br>• Pays attention to a short story and answers simple questions about it<br>• Hears and understands most of what is said at home and in school | 4-5 years<br>• Voice sounds clear and similar to other children<br>• Uses sentences that give lots of details (e.g., "I like to read my books")<br>• Tells stories that stick to topic<br>• Communicates easily with other children and adults<br>• Says most sounds correctly except a few like *l, s, r, v, z, ch, sh,* and *th*<br>• Uses the same grammar as the rest of the family |

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### Education

The success of a child with hearing loss in school depends on the support from family and teachers. Audiologic services are mandated in the public schools. Public schools provide diagnostic capabilities for older children and rehabilitative support. Educational audiologists are skilled at recommending modifications to the educational setting to ensure the success of the child. The educational audiologist monitors the child's personal hearing aids and selects, evaluates, and fits classroom amplification such as individual or group FM systems, assistive listening devices, and sound-field systems. The educational audiologist works with the teachers and parents to develop an Individualized Educational Plan (IEP) or Individualized Family Service Plan (IFSP). These plans identify goals and objectives, and designate appropriate educational placement, service, and equipment.

## Pediatric Hearing Loss (Non-Otitis Related)

Non-otitis related pediatric hearing loss is divided into sensorineural (SNHL) and conductive (CHL) categories. Congenital, or present since birth, sensorineural hearing loss (unilateral or bilateral) occurs with a frequency of approximately 1 out of 1,000 to 2,000 live births. The vast majority of cases (80 percent) have a genetic etiology caused by gene mutation. An autosomal recessive inheritance pattern accounts for over 75 percent of these cases. Inherited forms of SNHL may be further divided into Syndromic and Non-syndromic types. Congenital CHL occurs less frequently, at a rate of approximately 1 in 10,000 births. Most of these cases are related to auricular/EAC atresia or hypoplasia. Congenital isolated ossicular fixation or absence is a rare form, accounting for 0.1 to 0.5 percent of pediatric CHL.

## Congenital Sensorineural Hearing Loss (SNHL)

Syndromic SHNL is associated with recognized genetic syndromes. While the list of syndromes with associated SNHL is fairly long, the three most commonly encountered in practice are: Treacher-Collins, Goldenhar's, and Branchio-Oto-Renal.

**Treacher-Collins Syndrome** may result in both SNHL and CHL. Typical clinical findings of this syndrome include:

- Coloboma of the lower eyelid
- Micrognathia
- Hypoplasia of the zygomatic arches
- Microtia
- Macrostomia (fish mouth)
- Inferior displacement of the lateral canthi, relative to the medial canthi (anti-mongoloid canthal slant)

**Goldenhar's Syndrome** has diverse etiologies and is not related to a single gene locus. Its clinical features include:

- Hemifacial microsomia
- Microtia
- Otomandibular dysostosis
- Coloboma of the upper eyelid
- Epibulbar lipodermoids
- Vertebral anomalies

**Branchio-Oto-Renal Syndrome** is characterized by:

- Cataracts
- Branchial cleft fistulae
- Pre-auricular pits
- SNHL
- Mondini and other inner ear malformations may be present

**Other sensorineural hearing losses:**

- Alport Syndrome (renal failure and progressive SNHL)
- Jervell and Lange-Neilsen Syndrome (prolonged QT interval and Itorsade de Pointe arrhythmia, sudden syncopal episodes, and severe to profound SNHL)
- Pendred Syndrome (thyroid goiter and SNHL)
- Usher Syndrome (retinitis pigmentosa and inner ear dysfunction, Type I has SNHL and vestibular symptoms, Type II has SNHL alone, Type III has SNHL and variable SNHL)
- X-linked Charcot Marie Tooth Syndrome (peripheral neuropathy, "champagne bottle" calves and variable SNHL)
- Waardenburg Syndrome (lateral dystopia of the medial canthi/lachrymal punctae, heterochromia iridis, white forelock)
- Mohr-Tranebjaerg Syndrome (postlingual SNHL associated with dystonia, spasticity, dysphagia, and optic atrophy with an X-linked inheritance pattern)
- Fredreich's ataxia mimics Mohr-Tranebjaerg Syndrome but has cardiomyopathy
- Stickler Syndrome (hypoplasia of the midface, SNHL, arthropathy, and progressive myopia in the first year of life)
- Norrie Disease (pseudotumor of the retina, retinal hyperplasia, necrosis of the retinal inner layer, cataracts, progressive SNHL, and mental disturbance)

While syndromic forms of SNHL make up only 20 percent of congenital SNHL, non-syndromic congenital SNHL is responsible for the remaining 80 percent. Previously, the latter was not recognized as genetically caused, but in the past 15 years or so, this error has been rectified. Population analysis studies have suggested that in all likelihood, there are more than 100 genes involved in non-syndromic hearing loss. In the past 13 years, more than 40 loci for autosomal dominant non-syndromic SNHL have been mapped and a dozen genes cloned. Although heterogeneous in etiology, roughly half of non-syndromic SNHL patients have a mutation in the connexin-26 molecule. The nomenclature for different inheritance patterns are: DFNA—autosomal dominant, DFNB—autosomal recessive, and DFN—X-linked. Autosomal dominant inheritance means that if the person possesses the gene, he or she will manifest the disease. In autosomal recessive inheritance, a faulty gene is required from each of the parents. Therefore, if an individual has a single copy of the faulty gene, the condition will not manifest. Finally, X-linked inheritance affects males, almost exclusively, as the faulty gene on the X chromosome is not counteracted by the Y chromosome (unless a female has the faulty gene on both her X chromosomes).

Non-inherited congenital SNHL makes up about 20 percent of congenital SNHL. The most common causes are Mondini dysplasia to varying degrees, CHARGE association/syndrome, and enlarged vestibular aqueducts.

## Congenital Conductive Hearing Loss

Atresia of the external auditory canal is the most common cause of congenital, non-otitis related and CHL. Originally not thought to be genetically mediated, recent studies suggest a possible role for a deletion in the long arm of chromosome 18 as a possible genetic etiology. Occurring in 1 of 10,000 births, seventy

percent of atresia cases are unilateral. There is a strong gender predisposition, with a male to female ratio of 3 to 2. The right ear (61 percent) is more commonly affected that the left ear (39 percent). Significant advances in the surgical correction of EAC atresia have been made in the past 25 years.

Traditionally, EAC atresia reconstruction was a procedure reserved for patients with bilateral atresia. This was due to fears of facial nerve injury and the relatively poor surgical results achieved in that era. Since the advent of CT scanning, the anatomy of the mastoid and middle ear is readily assessed preoperatively. Based on a variety of factors, suitability for reconstruction may also be determined preoperatively. The factors considered important for operative success include: adequate middle ear/mastoid aeration, ossicle formation, oval window formation, and position of the facial nerve. With modern imaging and surgical techniques, unilateral EAC atresia reconstruction is commonly done by experienced hands, with excellent results and few complications. Following EAC atresia reconstruction, unilateral cases achieve pure tone averages within 30 dB of the normal ear, in 65 to 70 percent of cases. A residual air/bone gap of 25 dB or less is reported in 75 to 80 percent of cases. With proper attention to regular cleaning of the reconstructed ear canal, long-term follow up shows no deterioration of hearing results.

An alternative to EAC reconstruction in the setting of EAC atresia, particularly for those patients whose anatomy make them poor reconstruction candidates, is the implantation of a Bone Anchored Hearing Aid (BAHA®). The development of osteointegration using titanium implants has allowed for reliable fixation of an abutment to the bone behind the ear. The titanium screw protrudes through the skin and the abutment allows a vibrating hearing device to be attached resulting in bone conduction hearing. Recent refinements in the surgery technique of implanting the titanium screw have reduced complications. There is, however, resistance from insurance companies regarding coverage for this device, as it is regarded as an implantable hearing aid. Selective approval for the use of this device in patients with bilateral CHL, for whom no other reasonable alternative exists, will likely be the norm.

Congenital fixation or absence of the auditory ossicles, while uncommon, is readily corrected with surgery. Middle ear exploration with ossicular reconstruction or in the case of stapes fixation, stapedectomy results in a residual air/bone gap of less than 15 dB in more than 90 percent of cases. In the case of agenesis of the oval window, a rare malformation, surgical correction does not provide as reliable or complete correction of the conductive hearing loss. Defects of the middle ear and ossicles occur in many of the syndromes that are associated with syndromic SNHL. Roughly one-quarter of the reported cases of congenital ossicular malformation are associated with Branchio-Oto-Renal, Treacher-Collins, and Kippel-Feil syndromes. Other reported syndromes with ossicular fixation/absence are: Crouzon, Alpert, CHARGE, Goldenhar's, and X-linked stapes fixation.

## Auditory Processing Disorders

Auditory processing refers to what the brain does with what is heard. Although the ear itself picks up sound and directs it to the auditory system, auditory processing skills allow individuals to understand and act upon the information received through the ears.

For many years, the issues related to the perceptual processing of auditory information have been considered as "central auditory processing," but recently the terminology has been refined to simply "auditory processing." The terms Auditory Processing Disorders (APDs) and Central Auditory Processing Disorders (CAPDs) are considered synonymous.

The hearing mechanism has two primary parts:

1. The *peripheral system* is composed of the outer, middle, inner ears, and the auditory nerve
2. The pathways to the brain and the brain itself are referred to as the *Central Auditory Nervous System* (CANS)

Those individuals who may ultimately be diagnosed with an APD frequently have hearing abilities within the normal range, but are not as efficient or effective in acting upon the auditory information they receive as one would predict based upon their hearing acuity. Some of the symptoms of APDs may mimic a hearing loss; it is critical to assure that normal hearing is present prior to proceeding with an evaluation of auditory processing skills. Furthermore, people with hearing loss may have difficulties processing auditory information accurately. In such an instance, hearing loss and APD may co-exist, both as separate but related entities. Management strategies may overlap for these conditions.

## Definitions of APD

Broadly stated, auditory processing (AP) refers to the efficiency and effectiveness by which the central nervous system (CNS) utilizes auditory information. Narrowly defined, AP refers to the perceptual processing of auditory information in the CNS and the neurobiological activity that underlies that processing giving rise to electro-physiologic auditory potentials.

APDs are a multi-faceted set of problems that occur in different listening situations like being able to receive, analyze, organize, store, and retrieve auditory information. These processes may become more difficult in unfavorable listening conditions such as classrooms, large meeting spaces, parties, shopping malls, and sporting events.

Types of skills that may be affected by auditory processing include the following:

1. *Sound localization and lateralization:* the ability of the listener to know where sound occurs in space
2. *Auditory discrimination:* the ability to distinguish one sound from another, such as in the words "cat" and "hat"
3. *Auditory pattern recognition:* the ability to determine similarities and differences in patterns of sounds
4. *Temporal aspects of audition:* generally refers to the abilities to sequence sounds into meaningful combinations such as words and phrases, and to distinguish separateness when sounds are rapidly following each other
5. *Auditory performance with competing acoustic signals:* refers to the ability to understand the primary message when other noise is present in the environment such as other people talking, the television, or other ambient noises from heaters/air conditioner fans, the buzzing of electric lights, and lawn mowers
6. *Auditory performance with degraded acoustic signals:* refers to the ability to understand speech or other sounds when some of the information is degraded or missing (this can occur with fast rates of speech, significant unfamiliar accents of the speaker, or any activity that may block or mask parts of the primary signal)

Individuals who have APDs are diverse and may present a wide variety of symptoms or signs. Many of these difficulties manifest as learning difficulties, poorly constructed spoken language or limited understanding of language, poor academic performance, or attention difficulties, which appear similar to hyperactivity. An adult with auditory processing difficulties will describe difficult academic experiences and perhaps poor school performance during childhood.

## Behavioral Characteristics Present in Individuals with APD

According to the "ASHA 2005 Report," individuals suspected of having APDs frequently present one or more of the following behavioral characteristics:

- Difficulty understanding spoken language in competing messages, noisy backgrounds, or in reverberant environments
- Misunderstanding messages
- Inconsistent or inappropriate responses
- Frequent requests for repetitions by saying "what" or "huh" often
- Taking longer to respond in oral communication situations
- Difficulty paying attention
- Being easily distracted
- Difficulty following complex auditory directions or commands (sometimes simple auditory directions can be difficult to remember or follow)
- Difficulty localizing sound
- Difficulty learning songs or nursery rhymes
- Poor musical and singing skills
- Associated problems in reading, spelling, and learning

Other sources note that an unusual sensitivity to sound is another characteristic that is frequently observed.

It is important to note that this is not an exhaustive list of symptoms, and that not all behaviors may be observed in a single individual. In fact, some children diagnosed with APDs have good musical abilities and readily process pitch patterns, songs, and rhymes.

APDs may be diagnosed in early childhood, although many children are not identified until they are in an academic situation. Some parents report that children later identified as having an APD did not readily pay attention to voices or other sounds in the crib, while other parents describe children as hypersensitive to noise. Even as preschoolers, these particular children often avoid noisy environments such as birthday parties, child-oriented restaurants, play areas, and playgrounds. Other children with APDs may appear to be daydreaming or tuned out, and may not respond to having their names called or when someone is trying to get their attention.

Frequently, initial concerns are not raised regarding the child's abilities until the child is in kindergarten or first grade. Difficulties noted at this time often include concerns regarding phonics skills, pre-reading, and comprehension of the sounds associated with letters. Children with APDs may have difficulty following directions and may often have high visual attention, having quickly learned to copy the actions of fellow students. Symptoms are particularly noticeable when directions must be listened to and implemented independently of others.

As children progress through elementary school, increased reading difficulties are often noted, especially as new vocabulary and more complex tasks are undertaken. Children with APDs often fail to develop sound-symbol relationships in reading and language decoding. These children are viewed as immature, as they appear easily frustrated or overwhelmed in a typical classroom environment. If not detected, APDs can lead to poor self-esteem, frustration, and a dislike of school, particularly in the subjects of reading, spelling, and writing. Many children with APDs report that mathematics is one of the subjects they enjoy and in which they feel accomplished, until word/story problems are introduced.

Through early elementary school, many children (later diagnosed with APDs) appear to be functioning adequately because they do not call attention to themselves through disruptive behavior. Specific clues to their increasing difficulties are observable. For instance, they are often the children in the classroom who "know everything about everybody." They are highly visual so they can often report who has entered the classroom, taken a bathroom break, or walked past their classroom door. Additionally, they perform significantly better in one-to-one interactions than in the classroom or with a small group. One of the greatest difficulties in identifying children at this stage is that assessments for special education/support services are completed on an individual basis - generally in quiet environments which will not reflect their actual classroom performance. There is often a significant discrepancy between non-verbal performance measures, verbal-skills based scores, IQ scores, and academic testing results.

As academic expectations increase, usually noted from approximately third and fourth grade upward, children with APDs appear to have increasing difficulties in following classroom directions, understanding and analyzing new classroom vocabulary, and can lack the ability to translate classroom work into homework and individual assignments. Often, they appear to have very weak auditory memory skills, and they may be better able to function when notes are provided or when the teacher utilizes pre-written material on the board. They may be able to identify what homework assignments are, but may be completely unable to complete the actual tasks. Many children are described as doing better in classrooms where they do or see new information rather than hear about it. Parents of children of this age and older, often describe that they are re-teaching or providing 1-on-1 tutoring for the child to complete homework.

Children with APDs are often viewed by teachers and parents as having substantial inconsistencies in their learning abilities. They appear to have poor listening habits, but it is more often a reflection of the environment than the child. Some children are described as lazy because they appear not to pay attention or because adults incorrectly assume that the child deliberately chooses not to respond to auditory information. Some children with APDs are described as hyperactive, and subsequently misdiagnosed with attention deficit hyperactivity disorder (ADHD), because they have a hypervigilance to understand and recognize the multitude of auditory input in an environment. For example, most individuals with APDs can consciously hear the on/off cycling of a heating/air-conditioning fan. Children with APDs must consciously note, find, and determine the source of the fan noise prior to proceeding with their current activity, making them look both highly distractible and potentially hyperactive.

## Etiologies of Auditory Processing Disorders

Although the full range of etiologic factors involved in APDs is not well understood, several factors appear to be related. Early and/or frequent otitis media appears to have long-term impact on auditory processing skills. Mild fluctuating hearing loss, associated with otitis media, seems to affect the development of the central auditory system. Subsequently, this may affect higher-order language processing skills.

Some auditory processing disorders and their resulting problems are thought to be genetic. Parents often relate similar learning and/or listening difficulties as a child. Processing disorders may be related to difficulties or illnesses occurring during prime gestation periods of pregnancy, or trauma during birth. There may be a link between the presence of these disorders and the treatment of infant jaundice, although this

relationship is not well understood. Bile deposits in the CNS seem to be the culprit. There appears to be a higher incidence of APDs in children with other sensory processing disorders and other language/learning disorders, although significant research is needed to understand the causation and related factors. APDs are not the result of these other disorders, and frequently, the cause remains unknown.

In adults with later onset APDs, etiologies range from vascular problems, significant head trauma (particularly closed-head injuries), sensorineural hearing loss, and neurologically based disorders such as Parkinson's disease or multiple sclerosis.

The etiologies of central auditory processing disorders are not clearly defined. Lewis (1986) estimates that three to seven percent of all school-aged children exhibit some type of learning disability. The National Center for Learning Disabilities indicates almost 2.9 million U.S. school children are classified as having specific learning disabilities. In addition, they also receive some special education services through the public school system. This number does not reflect any children in private schools, religious schools, or who are home-schooled. It also does not include any children under elementary school ages. Additionally, the number of children with significant reading disabilities is estimated to be 17 to 20 percent, according to the National Assessment of Education Progress in 1994 and 1998. This information does not subcategorize APD-related disabilities; however, research in this arena would estimate that a substantial number of these children would be identified with an APD if complete evaluations were performed.

The effect of auditory processing difficulties can be devastating, but many parents, clinicians, and physicians are committed to improving diagnostic and treatment options.

## Diagnosis of Auditory Processing Disorders

APDs are characterized as an auditory deficit that can be diagnosed by a professional audiologist. However, due to the multiplicity of symptoms and learning manifestations associated with APDs, a team approach to diagnosis is advised. It is critical to evaluate and understand the child's educational, social, speech/language, cognitive, visual, and medical status and needs.

Medical providers are instrumental in determining whether pathological conditions exist that may cause learning difficulties in children. APDs symptomatology may mimic speech/language deficits, thus a complete evaluation of receptive (understanding) and expressive (production) speech is a critical part of any APD evaluation. Cognitive skills and deficits can reflect significant information regarding the individual's capacity for learning; assessing other factors which may impact learning abilities can also shed light on potential challenges. Parents are critical sources of information because they can relay developmental history, milestones, difficulties encountered in school and home environments, and past and current medical information. Parents often are the driving force in obtaining further diagnostic procedures for APDs.

## Remediation/Treatment of Auditory Processing Disorders

Two major compounding factors in auditory processing, as with any brain-based skill, are maturation levels of the central nervous system (CNS) and the amount of brain plasticity. On this basis, many parents are advised to "wait and see" rather than pursuing learning supports that can help their child achieve better academic, social, and personal growth during the maturational process. These delays can often lead to increased frustration, deterioration of self-esteem, and poor school attitudes and behaviors. For this reason, prompt screening is advised. However, once an APD is diagnosed in a child, regular monitoring, approximately every one to two years, should be ensured so that maturational changes can be appropriately reflected in support planning.

The maturation of the CNS must be considered in the diagnosis and treatment of APDs. The current research in brain plasticity and its impact on learning is also an important consideration. The ability of the auditory cortex to reorganize continues throughout life reflecting the ability to acquire new skills and behaviors. In summary, Bellis states that neuromaturation of some portions of the auditory system may not be complete until age 12 or later; however, management of APDs in children should be undertaken as early and aggressively as possible.

A variety of available resources address treatment techniques and classroom accommodations that may be used to meet the needs of individuals with APDs. The first step is to complete an audiological evaluation. Hearing may fluctuate, particularly in children who are prone to otitis media, and this will affect their listening abilities. Many schools and audiologists have checklists or parent surveys which help screen children for APDs and other learning difficulties. Parents may wish to request a special education services assessment through their school system.

In general, APD test batteries are only completed on children aged seven and older. Some pediatric audiologists, with specific experience in testing younger children, may administer some of the tests in the APD battery from approximately age five and up.

### Treatment and Therapeutic Supports

For many years, educators, speech pathologists, audiologists, and other providers have struggled to find appropriate treatment approaches that lead to improved learning abilities and quality of life for individuals with auditory processing disorders. While this continues to be an area with substantial ongoing changes and developments, there are options available for adults and children. Management protocol depends on the auditory profile of the individual. Treatment options may include, but are not limited to, modifying the listening environment, utilizing personal FM systems or other assistive listening devices, designing treatment programs to enhance processing skills, and academic accommodations.

## Health Care Provider Tips

- Service providers in the medical community have the opportunity and professional responsibility to help families connect with the appropriate personnel in the educational setting, and to collaborate with the school personnel who provide services to students with hearing loss.

- Develop and maintain partnerships with families to empower them to advocate for their children's best interests. Some families may not have the ability to advocate for their children because of financial or employment hardships. Home-schooled students and students at private schools may not be receiving the recommended hearing screenings required by the IDEA.

- Research professional guidelines that provide frameworks for hearing screening programs and the legislation and mandates in your state (state mandates do vary). Recognize the legislative regulations that affect the deaf and individuals with hearing loss:

    - The Individuals with Disabilities Education Act (IDEA) of 2004 requires access to a free and appropriate education (FAPE) for all children with disabilities.

    - Section 504 of the Rehabilitation Act of 1973 prohibits recipients of federal funds from discriminating against "otherwise qualified individuals."

    - The Americans with Disabilities Act (ADA), modeled after the Rehabilitation Act of 1973, replaces the word "handicap" with "disability" and applies to all employers, not just those receiving federal funds. It ensures the provision of information access in all public settings.

- The process of the assessment and placement component for IDEA or 504 begins with a concern about a child identified by the parent, teacher, school nurse, physician, or other individual or agency. The first step is a hearing screening or audiological assessment to confirm or rule out hearing loss as the problem. The assessment can help to determine the appropriateness of a special education referral.

- Auditory neuropathy is a unique hearing disorder in which the inner ear, or cochlea, is normal but the transmission from the cochlea to the auditory brainstem is impaired. Auditory neuropathy can affect people of all ages, from infancy through adulthood.

- The characteristic symptom of auditory neuropathy is poor speech perception abilities. Speech discrimination is typically worse than predicted by the degree of hearing loss depicted on an audiogram.

- Auditory neuropathy is diagnosed using auditory brainstem response (ABR) and otoacoustic emissions (OAE) tests.

- Be sensitive to the fact that the diagnosis of auditory neuropathy is challenging for families. Each child is unique and may respond to the various treatments and communications options differently. A multidisciplinary team approach with the family and other caregivers is recommended.

- Recurrent otitis media is one of the most common medical disorders in children and is the most common cause of hearing loss in children. Obtain a careful history from the parents, perform a physical examination (don't forget to insufflate), and complete diagnostic evaluations. These are all critical in determining the effect of otitis media on the child's hearing.

- All children with recurrent otitis media, or chronic otitis media with effusion, should have their hearing assessed. The purpose of this is to determine the presence and type of hearing loss (e.g., conductive, sensorineural, or mixed hearing loss) and the degree of hearing loss through further audiometric testing and an otolaryngology referral.

- Methods of hearing testing are behavioral testing, tympanometry, otoacoustic emissions, and brain stem auditory evoked response. Recognize that the reliability of some of these tests is determined by the cooperativeness of the child. Assess infants below six months of age with a complete battery of tests because behavioral testing alone is not appropriate for this age.

- There are both non-surgical management and surgical interventions for treating recurrent otitis media. Research has demonstrated that infants who are screened, diagnosed, and receive early intervention by six months of age have near normal language outcomes by age three. Those infants whose hearing loss is detected later than six months often demonstrate delays the rest of their lives.

- Most states have implemented Early Hearing Detection and Intervention (EHDI) programs through their state health departments. [See the Bibliography for print and online resources.]

- Even if an infant passes a newborn hearing screening, it could develop hearing loss later. If an infant has a high risk factor, such as a family history of childhood hearing loss, the infant should be monitored with ongoing audiologic testing.

- Refer a child immediately to Child Find or an audiologist for screening and diagnosis whenever a parent expresses concern about the child's hearing or speech.

- Each child is unique and develops speech and language at his or her own rate. If a child does not meet the majority of the milestones in one age range, he or she should be referred to an audiologist and to a speech pathologist. Remember that hearing can be tested at any age.

- Audiologic services are mandated in the public schools (PL94-142). Public schools provide diagnostic capabilities for older children and rehabilitative supports.

- Non-otitis related pediatric hearing loss may be divided into sensorineural (SNHL) and conductive (CHL) categories. Congenital hearing loss/deafness (unilateral or bilateral) is more common than CHL.

- Syndromic SHNL is associated with recognized genetic syndromes. While the list of syndromes with associated SNHL is fairly long, the three most common are: Treacher-Collins, Goldenhar's, and Branchio-Oto-Renal.

- The atresia of the external auditory canal is the most common cause of congenital, non-otitis related CHL. There is a strong gender predisposition with a male to female ratio of 3 to 2. The right ear (61 percent) is more commonly affected than the left ear (39 percent).

- Individuals who have auditory processing disorders frequently have hearing abilities within normal ranges but are not as efficient or effective as one would predict in acting upon the auditory information they receive. Some of the symptoms of APDs may mimic hearing loss, so it is critical to assure that normal hearing is present prior to proceeding with an evaluation of auditory processing skills.

- APDs are a multi-faceted set of problems that occur in different listening situations. People must be able to receive, analyze, organize, store, and retrieve auditory information for optimal hearing.

- Individuals who have an APD are diverse and may present a wide variety of symptoms. Many of these symptoms include learning difficulties, poor understanding of language or speech, lowered academic performance, and/or attention difficulties which appear similar to hyperactivity.

- Children with APDs often fail to develop sound-symbol relationships in reading and language decoding. Children with APDs are often viewed by teachers and parents as having substantial inconsistencies in their learning abilities and behavioral problems.

- APDs may be related to difficulties or illnesses occurring during critical gestation periods in pregnancy, or trauma during birth.

- In adults with a later onset APD, etiologies range from vascular problems, significant head trauma (particularly closed-head injuries), sensorineural hearing loss, and neurologically based disorders such as Parkinson's disease or multiple sclerosis.

- Use a multidisciplinary team approach to diagnose an APD.

- Utilize the knowledge of parents. They are critical sources of information because they can relay developmental history, milestones, difficulties encountered in school and home environments, and past and current medical information. Parents are often the driving force in obtaining further diagnostic procedures.

- Recommend prompt screening for APDs. Once an APD is diagnosed in a child, regular monitoring every one to two years should be ensured to reflect maturational changes in support planning.

# TREATMENT OPTIONS

## Cochlear Implants

Cochlear implants are electronic devices that transform sound waves into digital electronic signals and transmit these signals directly to the nerve tissue in the inner ear. Unlike conventional hearing aids that amplify sound waves, cochlear implants stimulate the nerves of the inner ear directly. The device has a surgically implanted component and an external processor. The external processor contains a microphone, the electronics that encode the signal, and a power supply. The information is transmitted by induction to the internal device implanted beneath the scalp. Patients return for the fitting of the external sound processor three to four weeks after surgery. Measurements are made at that time to determine the necessary levels to stimulate the nerve. These adjustments are made several times throughout the first year to establish the patient's best hearing. Annual check-ups are recommended.

Cochlear implants were initially approved for adults who had hearing at some point in their lives and then lost their hearing to the degree where conventional hearing aids were of no benefit. The majority of these patients cannot use telephone-based technology to supplement hearing. They have however, an auditory memory of sounds and words.

The implants were later approved for children who were born with a severe to profound hearing loss. Children with severe to profound congenital hearing loss do best when implanted at the earliest possible time because the auditory cortex of the brain needs sound stimulation to develop. Currently, cochlear implantation is approved for children 12 months of age and older. If children receive the implant at age five or older, they may get sound awareness from the device but they will neither be able to understand words nor develop speech. Language recognition is one of the goals of cochlear implantation. Children and adults who have been deafened as a result of meningitis should be implanted as soon as possible since calcification of the cochlea can result from the infection, thus making insertion of the electrode into the cochlea difficult or impossible.

Risks involving surgery are quite low. The surgery is a 2 to 2 1/2 hour outpatient procedure done by an ear surgeon. The serious immediate risks are infection, with potential loss of the implant, and injury to the facial nerve. There is risk of facial nerve injury because, during surgery, the electrode array passes directly over the facial nerve in the mastoid, on its way to the cochlea. Patients with cochlear implants are at greater risk for meningitis compared to people without cochlear implants because the inner ear fluid comes into contact with the spinal fluid. All patients with cochlear implants should receive the pneumococcal vaccine.

Eligible patients can be referred to a center that performs cochlear implantation. Audiologists at these centers are able to perform the necessary testing and preoperative counseling to determine candidacy. With the advent of universal newborn hearing screening, most children will see an audiologist who can make the referral to an appropriate center.

## Hearing Aids

When Alexander Graham Bell was inventing the telephone, he was not trying to invent the wonderful and versatile instrument that is used for communication today. He was trying to invent the first hearing aid to be used by his wife. This section explains hearing aids, their size, and technology.

### Size

Years ago, there were body hearing aids (usually worn over the chest) which connected to a receiver, coupled with an earmold in the ear. Now, the size of hearing aids varies from completely in the canal

(CIC), which is barely visible to the observer, to behind the ear (BTE), which is worn behind the ear. The latter is coupled to plastic tubing which leads to the earmold that is housed in the concha of the outer ear and the ear canal. There are many types of earmolds, which vary according to acoustic and comfort levels. Some of these earmolds include in-the-ear (ITE) which sits fully in the concha, in-the-canal (ITC), and half-shell, which is a size between the ITE and ITC. As a general rule, the smaller the hearing aid, the more expensive it is for the patient. Usually, the size depends on the patient's preferences of the hearing aid aesthetics and cost.

## Technology

The technologies available for aids include analog, programmable, and digital. Analog hearing aids have been available for many years. To custom-fit physically, an earmold impression is taken and then sent to a lab for the hearing aid or earmold to be made. To be custom-fit for the configuration of the hearing loss, an audiogram is sent to the lab. If the person has good hearing in the low frequencies but poor hearing in the high frequencies, the hearing aid response will have more amplification in the high frequencies and less in the low frequencies. These analog hearing aids are generally effective in amplifying speech in quiet atmospheres. These aids, however, have a limited capacity in helping people hear speech when background noise is present.

In an effort to address the limitations of analog hearing aids, the hearing aid industry produced programmable hearing aids. These hearing aids have different settings for different environments. For example, the settings can be adapted for use in the following environments:

- In a relatively quiet environment (e.g., one-on-one or small group setting)
- With background noise (e.g., in a restaurant or grocery store)
- With loud background noise (e.g., where speakers would have to speak up to hear themselves)
- With a telephone

Depending on the manufacturer and the model of the hearing aid, the number of settings varies widely. It has been found that too many settings per hearing aid are confusing for the user.

Part of the confusion arises from the difficulty in moving from one hearing aid program setting to another setting. Moving from setting to setting is accomplished in one of two ways: using a remote control or a button on the hearing aid itself. Each of these methods has advantages and disadvantages. Though using the remote control to change settings is easy, the possibility of losing or misplacing the remote control is a disadvantage. Changing settings using buttons on the hearing aid can frustrate some hearing aid users, who have lost some of the sensation in their fingertips. For these users, distinguishing and navigating the closely set, small buttons on the hearing aid is a challenge.

There was initial confusion in the industry as these hearing aids were termed "digitally programmable" hearing aids, despite the fact that they were analog hearing aids. In the mid to late 1990s, technology was gleaned from other industries and true digital hearing aids arrived on the scene.

The microphone of a digital hearing aid monitors incoming signals many times per second. It can distinguish speech from background noise and amplify the signals identified as speech. For example, a patient complained that her new, expensive, digital hearing aids only worked intermittently. For instance, when she initially entered the kitchen she could hear the overhead fan; after a few seconds, she could no longer hear it. In this case, her digital hearing aids were working well; the fan was not identified as speech, thus amplification of the fan noise stopped.

In the advent of Digital Signal Processing (DSP), digital hearing aids offer many capabilities beneficial to the patient. For instance, some patients may hear some frequencies well but have significant hearing loss in the next range of frequencies. The need for amplification of varying frequencies differs among patients. Digital hearing aids address this need.

Gain is the term used to describe how much the hearing aid amplifies. Although analog technology can do this to some extent, DSP gives the hearing aid more flexibility by regulating how much to amplify, and at which specific frequencies.

Acoustic feedback is a problem for hearing aid users. Acoustic feedback can happen with any amplified sound. It occurs when sound broadcasts from the receiver and comes back into the microphone repeatedly. With hearing aids, it sounds like a high pitched whistle, and often causes much embarrassment or frustration to the user. With analog hearing aids, the solution is to either tighten the physical fit or reduce the gain. Digital Feedback Reduction (DFR) can now sense when feedback is present and reduce or eliminate it automatically.

Directional microphones are often used with digital hearing aids. This is not a new concept, but one revived from the late 1960s. The concept employs two microphones; one that is positioned to better pick up the speaker in front of the listener, and one designed to pick up signals from all directions. DSP reduces the sound coming from all directions, thereby, enhancing the signal coming from the front.

According to the hearing aid industry, 80 percent of hearing aids sold today are digital. When digital technology was first incorporated into hearing aids, they were quite expensive, and remain so. However, hearing aid manufacturers have developed an array of product lines with fewer "bells and whistles" to make digital hearing aids more affordable to a greater number of people.

## Determining the Use of Hearing Aids

If a patient has difficulty hearing conversations, a complete audiologic evaluation must be performed. This enables the audiologist to determine if there is significant enough of a hearing loss to warrant the use of hearing aids, or if there are other causes for the difficulties in communication. The audiologist may also determine if there are situations that require medical or surgical interventions.

If it is determined that a hearing aid may be beneficial, the individual's expectations regarding the hearing aid need to be discussed. When hearing aids are ordered and fitted, the user must be educated regarding their use.

## Issues in Hearing Aid Use

Hearing aids cannot replace "normal" hearing. However, hearing aids can help address a number of accompanying problems associated with hearing loss. With tinnitus, hearing aids may help to cover up or mask this problem. To address recruitment, hearing aids have components that do not allow the amplification to become too loud. Speech discrimination may be impaired when the cochlea or VIII nerve pathways do not work as efficiently as they once did. Unfortunately, hearing aids do not help this problem as well as they do for other problems.

When people live with hearing loss for a while and are first fitted with hearing aids, they start hearing forgotten sounds, but with a different quality. There may even be sounds in the environment that they never heard before. When the brain hears a new sound, the sound is processed to determine if it is important enough to continue to process. New hearing aid users often comment that it is noisy with hearing aids. For example, a new user wore her hearing aids to a family picnic and was bothered by a foreign sound. Finally, she asked her granddaughter what the sound was. The granddaughter replied, "Those are birds chirping, Grandma."

Successful hearing aid users are those who wear their hearing aids during most waking hours. This gives the wearer the time to become accustomed to the new hearing aid and sounds. As patients become more accustomed to amplification, proper adjustments can be made about sound quality based on the patient's preferences.

Unsuccessful hearing aid users are persons who purchase hearing aids due to family or peer insistence, or only for use on special occasions, such as family reunions or holiday get-togethers. These events usually present complicated conversations, significant background noise, and many people speaking at once. If the hearing aid user has not had a chance to adapt to and distinguish the sounds in the environment, these situations may be frustrating to the hearing aid user. These persons may conclude that hearing aids are not helpful and may eventually abandon wearing them.

## The Baha® System of Direct Bone Conduction

Bone Anchoring Hearing Aid or Baha® system is an implantable, osseointegrated, temporal bone prosthetic device specifically designed for patients with mixed or conductive hearing loss, or unilateral sensorineural hearing loss, also known as Single Sided Deafness (SSD).

In the 1950s, Swedish Professor Per Ingvar Branemark discovered that when titanium, in its purest form, is exposed to air, it develops an oxide layer that is permanent and will not separate from the titanium. Professor Branemark found that in this oxide layer, an active biological field forms that allows living tissue to grow and bond permanently with the titanium. He also found that the retention properties of this process strengthen over time. First applied to oral implants, this process—osseointegration—proved successful. Its use was gradually expanded to include research into how osseointegration could be used to help those with hearing loss. The result was the Baha® system of direct bone conduction.

The Baha® system is comprised of three parts—a titanium fixture, a connecting abutment, and a detachable sound processor. The fixture is implanted into the temporal bone during a 30 to 60 minute outpatient surgical procedure. Three months later (six months for pediatric cases), allowing for thorough osseointegration, the sound processor is fitted via the snap abutment. Via the abutment, the sound processor receives sound and sends it to the functioning cochlea using the skull bone as a pathway to bypass the outer and middle ear. Direct bone conduction delivers clear, efficient sound without invading the middle or inner ear structures. Presently, there are two sound processors available. The ear-level Divino sound processor uses digital signal processing. It has an integrated directional microphone, an adjustable AGCo, and Low Tone Trimmers. The body-worn Cordelle, with K-amp technology, provides an additional 13 dB of gain over the Divino. Additionally, two ear-level sound processors, still in use by many Baha® wearers, are the Compact with AGCo (with an external directional microphone), and the Classic with linear output.

### Patient Indications

The Baha® system was first FDA cleared for use in the United States in 1996 to treat mixed and conductive hearing loss in patients age 18 and older. In 1999, FDA indications were expanded to include children age five and older. In 2001, the system was cleared for bilateral implantation, and in 2002, it was cleared for use in the treatment of unilateral, profound sensorineural hearing loss, or Single Sided

Deafness (SSD). Also in 2002, the Baha® Softband was introduced, designed to be worn by children under age five.

Patients with mixed or conductive hearing loss who cannot benefit from conventional air conduction amplification may possibly benefit from the Baha® system. Chronic otitis media, congenital aural atresia and microtia, cholesteatoma, middle ear dysfunction or disease, and external otitis are examples of conductive hearing losses where conventional amplification treatments may not prove beneficial. Patients with mixed or conductive hearing loss, who could benefit from the Baha® system must:

- Be age five years or older
- Have a bone conduction pure tone average (BC PTA) in the indicated ear, better than or equal to 45 dB
- Have monosyllabic word discrimination score of 60 percent or better
- In the case of bilateral fitting, there must be a symmetrical bone conduction PTA defined as less than 10 dB difference in the bone conduction PTA, or less than 15 dB at 500, 1,000, 2,000, and 4,000 Hz

Common causes of Single Sided Deafness (SSD) include acoustic neuroma tumors and/or tumor removal surgery, sudden deafness, neurological degenerative disease, Meniere's disease, viral infection, and trauma. Patients with previous SSD had only one possible treatment option in the past, called the contra lateral routing of signal (CROS) aid, a system with limited performance and satisfaction due to its placement in the functioning ear only.

Unlike the criteria for mixed and conductive hearing loss, SSD patients must have normal hearing in one ear, defined as an air conduction PTA score of 20 dB, or better. The Baha® system uses transcranial routing to send sound received by the sound processor on the deaf side to the cochlea in the normal hearing ear. Clinical studies have proven that the Baha® system eliminates the head shadow effect, improves speech intelligibility in noise, and has greater perceived benefit than CROS aids.

### Conclusions

The Baha® system, first used in Europe more than 25 years ago, has become standard treatment for certain types of conductive and mixed hearing loss and Single Sided Deafness. It is estimated that there are currently 25,000 people benefiting from the Baha® system's unique direct bone conduction application, enabling clear and natural sound to be transmitted to a functioning cochlea. Studies have demonstrated Baha® system benefits, for both patient indications and advantages, over conventional amplification.

## Assistive Devices

Assistive devices are a category of products, other than hearing aids, intended to fill gaps in communication. Not all assistive devices and strategies can solve all problems. It helps to understand the problem and the potential solutions. It is helpful for most people with hearing loss to build a toolbox or array of technologies to manage the challenges that different listening situations bring.

Patients can take advantage of residual hearing, vision, and touch to improve understanding and the intent of the stimulus. Multiple strategies may be necessary to ensure effective communication. Some of these devices can be used in conjunction with hearing aids to add more information or clarity to the communication.

### Auditory Strategies

One strategy is to alter the auditory stimulus (besides amplification) so that it fits within the range of the individual's residual hearing. For most people, hearing loss varies across the frequency range; therefore, frequency-shaping often adds to clarity. Another strategy is to transpose the signal into a frequency range where there is more residual hearing. Transmitting the signal through radio waves, light waves, wires, or electro-magnetic fields can be helpful in managing the effects of distance and competing background noise.

### Visual Strategies

This strategy tries to convert auditory information into visual information. Closed captioning, real-time captioning, and voice-recognition technologies convert speech to text. Auditory alarms can be converted to flashing light signals.

### Tactile Strategies

Tactile signals, such as vibrations, can be helpful to relay information to patients. For example, converting speech to tactile vibration can add to an individual's ability to speechread. Auditory alarms can be converted to tactile signals as well.

## Health Care Provider Tips

- Cochlear implants have been approved for adults and children 12 months or older. Cochlear implants are electronic devices that transform sound waves into digital electronic signals and transmit these signals directly to the nerve tissue in the inner ear. The implant surgery is a 2 to 2 1/2 hour outpatient procedure done by an ear surgeon.

- Be proactive with patients who get cochlear implants. Remind them that serious, immediate risks are infection, with potential loss of the implant, and injury to the facial nerve. Recall that patients with cochlear implants are at greater risk for meningitis compared to people without cochlear implants. All patients with cochlear implants should receive the pneumococcal vaccine.

- Usually the size of the hearing aid depends on the patient's preferences of the hearing aid aesthetics and cost. Inform patients that there are three types of technologies available: analog, programmable, and digital. Each technology has its complications and benefits that the patient should explore. Remind patients that hearing aids cannot replace "normal" hearing.

- Successful hearing aid users are those who wear their hearing aids during most waking hours to adjust to the new hearing aid and sounds. Unsuccessful hearing aids users are persons who purchase hearing aids due to family or peer insistence, or only for use on special occasions. Over time, proper adjustments can be made for sound quality based on the patient's preferences.

- The Baha® system is an implantable, osseointegrated, temporal bone prosthetic device specifically designed for patients with mixed or conductive hearing loss, or unilateral sensorineural hearing loss, also known as Single Sided Deafness (SSD).

- This technology is available for patients with a mixed or conductive hearing loss, and who cannot benefit from conventional air conduction amplification. Chronic otitis media, congenital aural atresia and microtia, cholesteatoma, middle ear dysfunction or disease, and external otitis are examples of conductive hearing losses where conventional amplification treatments may not prove beneficial.

- Clinical studies prove that the Baha® system eliminates the head shadow effect, improves speech intelligibility in noise, and offers greater perceived benefit than CROS aids.

- Assistive devices are a category of products, other than hearing aids, intended to fill gaps in communication. Although not all assistive devices and strategies can solve all problems, patients can build a toolbox or array of technologies to help with hearing in different listening situations.

# CONCLUSION

People with hearing loss are diverse, possessing varying ranges of hearing ability and different cultural identities. Those who identify themselves as part of the Deaf community see themselves as a distinct cultural group. The majority of people with hearing loss, however, may not identify with their hearing loss in this manner. More likely, these individuals still identify with the hearing mainstream. This cultural chasm challenged the collection of information for the writing of this handbook.

Although identifiers like hard of hearing are intended to be all-embracing, the term falls victim to the cultural bias against hearing loss. This bias is loaded with deaf prejudice, ageism, stigma, and more. Of those with hearing loss, 97.8% will not be clinically deaf, so there is a legitimate reason to create a handbook addressing this group's needs. The term "person with hearing loss" is used to highlight the person, not the hearing loss. It makes the distinction that a large number of these individuals may not identify as hard of hearing.

Another challenge is the lack of research and information on the demographics of hearing loss and its impact on people of color, people who are limited English proficient, and immigrants. In comparison, a large amount of research exists regarding deaf culture and people. For example, Gallaudet University (founded in 1864) is the world's only university in which all programs and services are specifically designed to accommodate deaf and hard of hearing students. The University is known for quality research on the history, language, culture, and other topics related to deaf people. We know from this work the impact of deafness on children and individuals both from a biological and cultural point of view. There are still questions regarding the cultural impact of hearing loss on diverse individuals.

It is this lack of information and resources that serves as a testament to the need for research and initiatives devoted to understanding exactly how cultural and ethnic identity can affect the medical and life experiences of people with hearing loss. The multidimensional nature of identity necessitates the broadening of the clinical and social views regarding people with hearing loss and to design programs, laws, and policies that acknowledge this diversity within each individual.

For example, a growing number of younger people are losing their hearing due to noise. How will this affect the person's identity, self-esteem, and health? In the clinical encounter, do the person's race/ethnicity and cultural beliefs about hearing loss need to be considered? People with hearing loss, deaf or not, may experience bias and prejudice from staff, negatively impacting quality health care. How can this bias and prejudice be addressed? If the person does not communicate well in English, are there institutional resources that support staff to get an interpreter and utilize other aids for communication? This handbook discusses these issues and raises questions about appropriate care for individuals with hearing loss.

Hearing loss is often an invisible disability. Organizations must ensure that communication tools and resources, such as interpreters, are readily accessible for patients. It is the responsibility of health care providers to understand the intricacy of perspectives surrounding hearing loss and the diverse backgrounds of their patients. Providers should implement changes needed in the health care system to provide the highest quality of care.

# RESOURCES

## SCHOOL-BASED HEARING SCREENING INFORMATION

(California, Colorado, Maryland, Ohio, Oregon, Virginia, Washington)

### California

(Excerpted from Health and Safety Code Section 1685, Appendix B "Testing Standards for Hearing Impaired," Section 2951 of the *California Administrative Code, Title 17, Public Health*)

**Authoritative Agency**: State Department of Health

#### Target Populations:

1. Each pupil shall be given a screening test in kindergarten or first grade, and subsequently in second, fifth, eighth, and tenth or eleventh grades.
2. Each pupil enrolled in classes for the physically handicapped, educationally handicapped, special education programs, or ungraded classes, shall be given hearing tests when enrolled in the program and every third year thereafter.

#### Test Frequencies and Screening Levels:

Pure tone audiometric screening tests, either group or individual, shall be conducted at a level not to exceed 25 decibels and shall include the frequencies 1,000, 2,000, and 4,000 Hz. Failure to respond to any of the required frequencies at the screening level constitutes a failure of the screening test.

Pure tone air conduction threshold tests shall include the frequencies 250, 500, 1,000, 2,000, and 4,000 Hz and shall be given to:

- All students who fail the screening tests
- All pupils who are to be considered for further audiological or otological evaluation

#### Referral Criteria:

The schools shall provide the parents or guardians of children who fail the hearing tests with a written notification of the test results and recommend that a medical evaluation be obtained whenever the test demonstrates:

- A hearing level of 30 decibels or greater for two or more frequencies in an ear at 250, 500, 1,000, 2,000 or 4,000 Hz, or a hearing level of 40 decibels or greater of any one of the frequencies tested, 250 through 4,000 Hz, on two threshold tests completed at an interval of at least two weeks, or there is evidence of pathology (e.g., an infection of the outer ear, chronic drainage, or a chronic earache).

### Colorado

(Excerpted from Standards of Practice for Audiology Services in the Schools, Colorado Department of Education, Exceptional Student Services, revised September, 2004)

**Authoritative Agency**: State Department of Education

### Target Populations:

1. All students in grades kindergarten, one, two, three, five, seven, and nine, or comparable secondary levels.
2. All transfer students entering without current screening records, within two months of school enrollment.
3. All students who have failed the previous year's screening and who were not cleared by an audiologist.
4. All students receiving special education and/or related services (students who are being assessed for special education services should have been screened within the preceding twelve months).
5. All children enrolled in public-funded early childhood programs.
6. All infant and preschool children upon referral through existing community Child Find processes.

### Test Frequencies and Screening Levels:

- 500 Hz - 20 dB for preschool through fifth grades; (tympanometry may replace 500 Hz for PS through third grade); 25 dB is acceptable if ambient noise levels are high; 500 Hz is optional for sixth through twelfth grades if negative history of HL;

- 1,000 Hz-20 dB;

- 2,000 Hz-20 dB;

- 4,000 Hz-20 dB;

- 6,000 Hz-25 dB for six through twelfth grades (optional for all other grades).

### Referral Criteria:

A hearing screening referral results from any one or more of the following:

1. Pure tone referral criteria—any designated frequency in either ear.
2. Acoustic-emitting screening includes tympanometry, static admittance, and equivalent ear canal volume measurements. One of the following constitutes a referral:
    - Flat tympanogram and equivalent ear canal volume outside the normal range on two successive occurrences in a four to six week interval;
    - Low peak admittance on two successive occurrences in a four to six week interval; or
    - Abnormally wide tympanometric width (gradient) (TW>200 daPa) on two successive occurrences in a four to six week interval.
3. Visual inspection includes otoscopy and/or a cursory observation of the ear. Either of the following conditions constitutes a referral:
    - Structural abnormalities of the outer ear, ear canal, or eardrum; or
    - Ear canal drainage.
4. Rescreening for all pure tone referrals within the same session or within two weeks of the initial screening using the same frequencies, levels, and referral criteria.

## Maryland

(Excerpted from Annotated Code of Maryland, Education Article, 7-404, Hearing and Vision Screening Tests)

**Authoritative Agency**: State Department of Health/Education

### Target Populations:

1. Unless evidence is presented that a student has been tested within the past year, the screenings required shall be given in the year that a student enters a school system, and enters the fourth, fifth, sixth, and ninth grades. Students may be referred for screenings at other times if problems are suspected.
2. Further screening shall be done in accordance with the bylaws adopted by the State Board.

## Ohio

(Excerpted from Ohio Department of Health, Policies for Hearing Conservation Programs for Children, Requirements and Recommendations, Revised 2001)

**Authoritative Agency**: State Department of Health

### Target Populations:

1. Preschool children
   a. Preschoolers enrolled in school programs shall be screened upon entrance to the preschool program and annually thereafter. Children who cannot be screened shall be referred to an audiologist and/or physician for evaluation.

2. School-aged children in regular classes
   a. School-aged children shall be screened at five grade levels: kindergarten, first grade, third grade, fifth grade, and ninth grade. Students may be tested in additional grades.

3. In addition, the following children shall be screened:
   a. those students new to a school (and not tested within the last 12 months)
   b. those students referred by a teacher
   c. those students who were referred within the past year with no documented follow-up, regardless of grade level
   d. those students absent during the previous hearing screening
   e. those students at risk for noise exposure (e.g., band, vocational education, industrial education, automotive mechanics, etc.)

4. To establish a baseline audiogram, testing of high school seniors who have taken courses involving noise exposure is strongly recommended.

Guidelines for the management of children who wear hearing aids or have a documented hearing loss:

- Children wearing hearing aids shall not be screened in the schools.

- Children identified with a hearing loss, who do not wear hearing aids, shall remain in the school hearing screening program.

- For children who fall into the a and b categories above, follow-up with parents to make sure that the child is under a doctor's care.

- School-aged children in special classes shall be screened at the ages that correspond to the grade levels listed in the previous paragraphs above. Children who cannot be screened shall be referred for audiologic and medical evaluation.

The consensus of the Ad Hoc Hearing Advisory Committee is that children in special classes should still be included in the screening program even though the children may need to be referred on a periodic basis.

### Test Frequencies and Screening Levels:

1. Required Screening Procedures
    a. External observation of the ear
        i. The screener shall note on the hearing screening form any reported symptoms.
        ii. Reported symptoms include:
            - discharge from a child's ear canal
            - any malformation of the ear
            - soreness or pain in or about the ears
2. Pure-tone air conduction audiometry
    a. Each child shall be screened at 1,000 Hz, 2,000 Hz, 4,000 Hz, and at 20 dB HL in the right ear and in the left ear. No other frequencies or intensities are to be used.
3. Optional Screening Test
    a. Tympanometry
        i. The Ohio Department of Health recommends tympanometry as a useful tool for screening middle ear problems. Tympanometry does not, however, measure hearing and shall not be used without pure tone screening.
        ii. Anyone who screens using tympanometry shall follow the guidelines outlined in the Criteria for Referral (Section VI).

### Referral Criteria:

- Observation.

- Immediate referral for discharge from ear canal, malformation of the ear, and soreness or pain in or about the ears.

- Pure tones - students failing two screenings within a six week period will result in parents receiving written notification that their child has failed a hearing screening test and that an examination by a physician and/or audiologist is recommended. (No diagnostic statement about the child's suspected hearing problem shall be mentioned in the parents' notification.)

## Oregon

(Excerpted from Revised School Health Services Manual, Regulations OAR 581-022-0705 Health Services, updated September, 2003)

**Authoritative Agency**: State Department of Education

### Target Populations:

Hearing screening shall be offered annually for all students in as many of the following categories as resources allow, starting with:

- First entry into school;

- Every year from pre-kindergarten through third grade;
- Seventh and eleventh grades;
- Upon entrance into special education;
- Upon grade repetition;
- Parent or school staff referrals; and
- School-age children who already receive regular audiologic management need not participate in a screening program.

The frequency of screenings and the participating population should be determined in consultation with appropriate local health care providers.

### Test Frequencies and Screening Levels:

20 dB HL at 1,000 Hz and 4,000 Hz (including 500 Hz if environment is within acceptable noise level).

### Referral Criteria:

Students failing to respond to one or more frequencies in either ear should be re-instructed, have the earphones repositioned, and re-screened. Re-screening may occur in the same session, on the same day, or within two weeks of screening. When the student fails after re-instruction and re-screening, she or he should be referred to an audiologist for evaluation.

- Students failing the follow-up audiological testing must be referred to appropriate medical and/or further audiological evaluation to occur within four to six weeks.
- Follow-up should occur within one month and no longer than three months to ensure students' medical, educational, and rehabilitative needs are met.

## Virginia

[Excerpted from: Code of Virginia, Section 22.1-273 and Section 22.1-214; Virginia Department of Education (Effective January, 1994). *Regulations Governing Special Education Programs for Children with Disabilities in Virginia*; SUPTS. MEMO. No. 159 (August 1987) and SUPTS. MEMO. No. 168 (September, 1987).]

**Authoritative Agency**: State Department of Education

### Target Populations:

In Virginia, hearing screening is required as follows:

- Component of the School Entrance Health Form: Part II - Comprehensive Physical Examination Report. (See Code of Virginia, 22.1-270.)
- Grades three, seven, and ten - unless tested as part of the School Entrance Health Form: Part II - Comprehensive Physical Examination Report. (See Code of Virginia, 22.1-273.)
- All children within 60 administrative working days of initial enrollment in a public school (see Regulations Governing Special Education Programs for Children with Disabilities in Virginia, effective January, 1994).

- The hearing of each child with a disability shall be tested during the eligibility process prior to placement in a special education program (see Regulations Governing Special Education Programs for Children with Disabilities in Virginia, effective January, 1994).

### Test Frequencies and Screening Levels:

- Five-hundred (500) Hz, 1,000 Hz, 2,000 Hz, and 4,000 Hz at 20 dB. "Note: If the location is too noisy to use 20 dB, a new location must be secured. Screening should never be conducted at intensities greater than 25 dB."

- Tympanometry screening is recommended for all students, kindergarten through third grade and all preschool-aged students, in the early childhood special education programs, or four-year-old programs.

### Referral Criteria:

- If the student fails to respond in either ear to two or more frequencies, a re-test should be scheduled within a two-week period. Referrals should be made if the second screening results do not improve. If the screening is part of the special education eligibility process, the school should be responsible for obtaining an audiological evaluation.

- Students with flat tympanograms, low static compliance (Peak Y), or abnormally wide tympanograms should be rescreened in four to six weeks.

- Parents, of all students who do not perform satisfactorily on a hearing screening and subsequent re-test (within two weeks), are notified by school health personnel. Parents should be advised to have the child evaluated by an audiologist or by their health care provider.

## Washington

(Excerpted from WAC Chapter 246-760 Auditory and Visual Standard-School Districts, Updated 9/30/02)

**Administrative Authority**: State Department of Public Instruction

### Target Populations: (WAC 246-760-020)

Schools shall conduct auditory and visual screening of children:

1. In kindergarten and grades: one, two, three, five, and seven;
2. For any child showing symptoms of possible loss in auditory or visual acuity referred to the district by parents, guardians, or school staff; and
3. If resources permit, schools shall annually screen children at other grade levels.

### Test Frequencies and Screening Levels: (WAC 246-760-030)

- 1,000, 2,000, and 4,000 Hz at 20 dB HL

### Referral Criteria: (WAC 246-760-050)

If a child does not respond to one or more frequencies in either ear:

- The school must rescreen the child within six weeks;

- Notify the child's teachers of the need for preferential positioning in class because of the possibility of decreased hearing; and

- Notify the parents or legal guardian of the need for audiological evaluation if the student fails the second screening.

Schools shall notify parents or legal guardian of the need for medical evaluation if:

- Indicated by audiological evaluation; or
- Audiological evaluation is not available.

| NATIONAL RESOURCES | PHONE | WEB and LOCATION ADDRESS |
|---|---|---|
| **Public Awareness** | | |
| **American Academy of Audiology** provides the highest quality of hearing health care service to children and adults. | 800-AAA-2336 | http://www.audiology.org<br><br>11730 Plaza America Drive, Suite 300<br>Reston, VA 20190 |
| **American Academy of Otolaryngology – Head and Neck Surgery** represents specialists who treat the ear, nose, throat, and related structures of the head and neck. | 703-836-4444 | http://www.entnet.org<br><br>One Prince Street<br>Alexandria, VA 22314 |
| **American Speech-Language and Hearing Association** is the professional, scientific, and credentialing association for audiologists, speech-language pathologists, and speech, language, and hearing scientists. | 800-638-8255<br>301-897-5700 TTY | http://www.asha.org<br><br>10801 Rockville Pike<br>Rockville, MD 20852 |
| **American Tinnitus Association** promotes relief, prevention, and the eventual cure of tinnitus. | 800-634-8978 | http://www.ata.org<br><br>Portland, OR |
| **The Better Hearing Institute** educates the public about the neglected problem of hearing loss and what can be done about it. | 800-EAR-WELL | http://www.betterhearing.org<br><br>515 King Street, Suite 420<br>Alexandria, VA 22314 |
| **Healthy Hearing** provides primary information on hearing loss news and consumer education. | 800-567-1692 | http://www.healthyhearing.com<br><br>5282 Medical Drive, Suite 150<br>San Antonio, TX 78229 |
| **Hearing Loss Association of America (formerly Self Help for Hard of Hearing People)** provides support for persons with hearing loss through information, education, advocacy, and support. | 301-657-2248<br>301-657-2249 TTY | http://www.hearingloss.org/<br><br>7910 Woodmont Ave,<br>Suite 1200<br>Bethesda, MD 20814 |

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| **National Cancer Institute-Head and Neck Cancer** provides comprehensive information about tumors of the head and neck. | 800-4-CANCER<br>800-332-8615 TTY | http://www.cancer.gov/cancerinformation/cancertype/headandneck |
| **National Institute on Deafness and Other Communication Disorders** conducts and supports research and research training in disease prevention, health promotion, and the special biomedical and behavioral problems associated with people having communication impairments and disorders. | 800-241-1044<br>800-241-1055 TTY | http://www.nidcd.nih.gov<br><br>31 Center Drive, MSC 2320<br>Bethesda, MD 20892 |
| **Family Resources** | | |
| **BEGINNINGS for Parents of Children Who Are Deaf or Hard of Hearing** provides emotional support and access for families with deaf children or children with hearing loss; age birth through 21. | 800-541-HEAR Voice/TTY | http://www.ncbegin.org<br><br>Raleigh, NC |
| **National Family Association for Deaf-Blind** identifies, coordinates, and disseminates information related to children and youth who are deaf-blind; age birth through 21. | 800-255-0411 | http://www.nfadb.org<br><br>141 Middle Neck Road<br>Sands Point, NY 11050 |
| **Adult/Youth Resources (General)** | | |
| **American Society for Deaf Children** provides support, encouragement, and information to families raising children who are deaf or with hearing losses. | 800-942-ASDC<br>717- 334-7922 TTY | http://www.deafchildren.org<br><br>Gettysburg, PA |
| **Helen Keller National Center for Deaf-Blind Youths and Adults** Center provides services to youth and adults who are deaf-blind. | 516-944-8900 Ext. 326 | http://www.hknc.org<br><br>141 Middle Neck Road<br>Sands Point, NY 11050 |

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| **League for the Hard of Hearing** provides rehabilitation and human services for infants, children, adults, and seniors with hearing loss, who are deaf, and deaf-blind. | 954-731-7208 Voice/TTY | http://www.lhh.org<br>50 Broadway, 6th Floor<br>New York, NY 10004 |
| **National Dissemination Center for Children with Disabilities** provides information on children/youth disabilities, programs, and services. | 800-695-0285 Voice/TTY | http://www.nichcy.org<br>Washington, DC |
| **National Information Clearinghouse on Children Who Are Deaf-Blind** provides extensive resources and personalized services for anyone requesting information about, or for, deaf-blind children. | 800-438-9376<br>800-854-7013 TTY | http://www.dblink.org<br>141 Middle Neck Road<br>Sands Point, NY 11050 |
| **Education Resources** | | |
| **Council of Academic Programs in Communication Sciences and Disorders** provides educational and professional support for programs that educate undergraduate and graduate students in the communication sciences and disorders. | 952-920-0966 | http://www.capcsd.org<br><br>Minneapolis, MN |
| **Educational Audiology Association** specializes in the management of hearing and hearing impairment within the educational environment. | 800-460-7EAA | http://www.edaud.org |
| **The Post secondary Education Programs Network** assists institutions to effectively serve students with hearing loss or the deaf. | 865-974-0607 Voice/TTY | http://www.pepnet.org<br>Claxton Complex A508<br>Knoxville, TN 37996 |
| **Language Services** | | |
| **American Sign Language Access** promotes sign language accessibility. | | http://www.aslaccess.org<br><br>4217 Adrienne Drive<br>Alexandria, VA 22309 |

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| **The National Cued Speech Association** champions effective communication, language development, and literacy through the use of Cued Speech. | 919-828-1218 Voice/TTY | http://www.cuedspeech.org<br><br>23970 Hermitage Road<br>Cleveland, OH 44122 |
| **The Registry of Interpreters for the Deaf** provides sign language interpreting and transliterating services for the deaf and persons with hearing loss. | 703-838-0030<br>703-838-0459 TTY | http://www.rid.org<br><br>333 Commerce Street<br>Alexandria, VA 22314 |
| **Technology Assistance/Hearing Aid** | | |
| **ABLEDATA** provides information about assistive technology products and rehabilitation equipment available from domestic and international sources. | 800-227-0216 | http://www.abledata.com<br><br>8630 Fenton Street, Suite 930<br>Silver Spring, MD 20910 |
| **MCI Global Relay Services** provides Telecommunications Relay Service (TRS) to enable those with hearing or speech disabilities to communicate. | 866-735-0373<br>866-735-0193 TTY | http://globalrelay.mci.com |
| **The National Captioning Institute** provides access to television's entertainment and news through the technology of closed captioning. | 800-950-0958 TTY | http://www.ncicap.org<br><br>1900 Gallows Road, Suite 3000<br>Vienna, VA 22182 |
| **National Center on Accessible Media** provides accessible media for the deaf and people with hearing or sight losses. | 617-300-2000 | http://main.wgbh.org<br><br>125 Western Avenue<br>Boston, MA 02134 |
| **Telecommunications Equipment Distribution Program Association—State Programs** provides information on state programs that offer free communication equipment to persons with hearing loss. | | http://www.tedpa.org/statprog/map.htm |
| **Sprint Relay Services** provides relay services for the deaf and individuals with hearing loss | 800-676-3777<br>800-877-8973<br>TTY/TDD/ASCII | http://www.sprintrelayonline.com |

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| **Legal and Policy** | | |
| **American Deafness and Rehabilitation Association** supporting positive public policies for individuals who are deaf or have a hearing loss. | | http://www.adara.org<br><br>Myersville, MD |
| **Hard of Hearing Advocates** creates and implements programs and solutions where persons with hearing loss have undue problems. | 508-875-8662 | http://hohadvocates.org<br><br>245 Prospect Street<br>Framingham, MA 01760 |
| **National Association of the Deaf** promotes, protects, and preserves the rights and quality of life of the deaf and individuals with hearing loss U.S. | 301-587-1788<br>301-587-1789 TTY | http://www.nad.org<br><br>814 Thayer Avenue<br>Silver Spring, MD 20910 |
| **The United States Access Board** provides information about ADA Accessibility Guidelines. | 202-272-0080<br>202-272-0082 TTY | http://www.access-board.gov<br><br>1331 F Street, NW, Suite 1000<br>Washington, DC 20004 |
| **Research** | | |
| **The American Hearing Research Foundation** funds research in hearing and balance disorders, and also educates the public about the disorders. | 312-726-9670 | http://www.american-hearing.org<br><br>8 South Michigan Avenue, Suite #814<br>Chicago, IL 60603 |
| **The National Organization for Hearing Research Foundation** funds and supports research into the causes, preventions, treatments, and cures of hearing loss and deafness. | 610-664-3135 Voice/TTY | http://www.nohrfoundation.org<br><br>225 Haverford Avenue,<br>Suite #1<br>Narberth, PA 19072 |

| REGIONAL RESOURCES | PHONE | WEB and LOCATION ADDRESS |
|---|---|---|
| **Colorado** | | |
| **Hands & Voices** | 866-422-0422 | http://www.handsandvoices.org<br><br>Denver, CO |
| **The LISTEN Foundation** | 303-781-9440 | http://www.listenfoundation.org<br><br>6950, E. Belleview Ave,<br>Suite 203<br>Englewood, CO 80111 |
| **National Center for Voice and Speech** | 303-446-4834 | http://www.ncvs.org<br><br>1101 13th Street,<br>Denver, CO 80204 |
| **Georgia** | | |
| **American Hearing** | 404-929-0067 | http://www.americanhearing.com |
| **Hawaii** | | |
| **Kapiolani Deaf Center** | 808.734.9210 (TEL/TTY)<br>808.734.9238 (FAX) | http://kapiolani.hawaii.edu/object/kdc.html<br><br>Manono Building, Room 102<br>Kapiolani Community College<br>4303 Diamond Head Road<br>Honolulu, HI 96816-4421 |
| **Mid-Atlantic** | | |
| **The Alexander Graham Bell Association for the Deaf and Hard of Hearing** | 202-337-5220<br>202-337-5221 TTY | http://www.agbell.org<br><br>3417 Volta Place, NW<br>Washington, DC 20007 |
| **Kennedy Krieger Institute** | 800-873-3377<br>443-923-2645 TTY | http://www.kennedykrieger.org<br><br>707 North Broadway<br>Baltimore, MD 21205 |
| **Maryland Technology Assistance Program** | 800-TECH TAP<br>866-881-7488 TTY | http://www.mdtap.org |

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| **Laurent Clerc National Deaf Education Center** | 202-651-5031 \|<br>202-651-5636 TTY | http://clerccenter.gallaudet.edu<br><br>800 Florida Avenue NE<br>Washington, DC 20002 |
| **Northern California** | | |
| **Bay Area Communication Access** | 415-356-0405<br>415-356-0376 TTY | http://bacainterp.com<br><br>443 Tehama Street<br>San Francisco, CA 94103 |
| **California Ear Institute** | 650-494-1000 | http://calear.com<br><br>1900 University Avenue,<br>Suite 101<br>East Palo Alto, CA 94303<br><br>5801 Norris Canyon Road<br>Suite 200<br>San Ramon, CA 94583 |
| **California Relay Service** | 877-546-7414<br>800-867-4323 TTY | http://www.ddtp.org/california_relay_service<br><br>505 Van Ness Avenue<br>Telecommunications Division<br>San Francisco , CA 94103 |
| **California Telephone Access Program** | **English**:<br>800-806-1191<br>800-806-4474 TTY<br><br>**Spanish**:<br>800-949-5650<br>800-896-7670 TTY<br><br>**Hmong**:<br>866-880-3394<br><br>**Cantonese**:<br>866-324-8754<br><br>**Mandarin**:<br>866-324-8747 | http://www.ddtp.org/CTAP<br><br>Stockton, CA |
| **Deaf and Disabled Telecommunications Program** | 877-546-7414<br>800-867-4323 TTY | http://www.ddtp.org |
| **Golden Gate Hearing Services** | 415-332-0932 | 1400 Franklin Street<br>San Francisco, CA 94109 |

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| **H.E.A.R. Hearing Education and Awareness for Rockers** | 415-409-EARS | http://www.hearnet.com<br>San Francisco, CA |
| **Hearing and Deafness Resources Center** | 415-923-4485 | 890 Beach Street<br>San Francisco, CA 94109 |
| **Hearing Society for the Bay Area** | 415-693-5870<br>415-921-8990 TTY | http://www.hearingsociety.org<br><br>49 Powell Street, Suite 400<br>San Francisco, CA 94102 |
| **Northwest** | | |
| **Direction Service** | 541-686-5060<br>541-284-4740 TTY | http://www.directionservice.org<br><br>3411 A Willamette Street<br>Eugene, OR 97405 |
| **Hearing, Speech and Deafness Center** | 206-323-5770 Voice/TTY | http://www.hsdc.org<br><br>1625 19th Avenue<br>Seattle, WA 98122 |
| **Oregon Lions Sight and Hearing Foundation** | 800-635-4667 | http://www.orlions.org<br><br>1410 SW Morrison Street,<br>Suite 760<br>Portland, OR 97205 |
| **Vestibular Disorders Association** | 800-837-8428 | http://www.vestibular.org<br><br>Portland, OR |
| **Ohio** | | |
| **Council on Education of the Deaf** | 330-672-0735<br>330-672-2396 TTY | http://www.deafed.net<br><br>405 White Hall,<br>College of Education<br>Kent State University<br>Kent, OH 44242 |
| **Southern California** | | |
| **House Ear Institute** | 800-388-8612<br>213-484-2642 TDD | http://www.hei.org<br><br>2100 W. 3rd. Street<br>Los Angeles, CA 90057 |
| **John Tracy Clinic** | 213-748-5481 | http://www.jtc.org |

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| **Roles, Rights, & Responsibilities for Deaf and Hard of Hearing** | 818.677.2611 Voice/TTY | http://ncod.csun.edu<br><br>California State University, Northridge<br>18111 Nordhoff Street<br>Northridge, CA 91330 |
| **Signing Exact English Center for the Advancement of Deaf Children** | 310-430-1467 Voice/TTY | http://www.seecenter.org<br><br>S.E.E. Center For the Advancement of Deaf Children<br>P.O. Box 1181<br>Los Alamitos, CA 90720<br><br>Long Beach Center<br>740 East Wardlow Road<br>Long Beach, CA 90807 |

| KAISER PERMANENTE RESOURCES | PHONE | WEB and LOCATION ADDRESS |
|---|---|---|
| **National** | | |
| **Kaiser Permanente and Disability Rights Advocates Health Access Program** | 510-25-3634 Voice/TTY | http://kpnet.kp.org/ada/index.html<br><br>1800 Harrison Street, 19th Floor<br>Oakland, CA |
| **Northern California General** | | |
| **Northern California Newborn Hearing Screening Program** | | http://insidekp.kp.org/california/perineo/clinical/reg_programs/nhsp.htm<br><br>1950 Franklin Street<br>Oakland, CA 94612 |
| **Northern California San Mateo County - Redwood City Medical Center** | | |
| **Ambulatory Care Services Audiology Policies** | | http://insidekp.kp.org/california/redwoodcity/policies/ambulatorycareservices/headandnecksurgery_audiology_toc.htm |

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| **Hearing Center/Audiology General Information** | 650-299-2977 | http://insidekp.kp.org/california/redwoodcity/departments/head_necksurgery/hearingcenter_audiology/index.htm<br><br>1800 Broadway, #5<br>Redwood City, CA 94063 |
| **Head & Neck Surgery General Information** | 650-299-2570 | http://insidekp.kp.org/california/redwoodcity/departments/head_necksurgery/index.htm<br><br>Hospital Tower 4FL<br>1150 Veterans Blvd.<br>Redwood City, CA 94063 |
| **ASL Language Services** | 800-874-9426<br>RWC KFH ID: #295714<br>Clinic ID: #20117 | http://insidekp.kp.org/california/redwoodcity/services/communication/languageservices.htm |
| **Northern California Santa Clara County – Santa Clara Medical Center** | | |
| **Hearing Aid Center General Information** | 408-553-6900 | http://insidekp.kp.org/california/santaclara/departments/ent_headandnecksurgery/hearingaidcenter/index.htm<br><br>2894 Homestead Road<br>Santa Clara, CA 95051 |
| **ENT/Head and Neck Surgery Services Homestead MOB** | 408- 851-2950 | http://insidekp.kp.org/california/santaclara/departments/ent_headandnecksurgery/index.htm<br><br>2nd Floor Dept., #290<br>710 Lawrence Expressway<br>Santa Clara, CA 95051 |
| **Northern California Santa Clara County – Santa Teresa Medical Center** | | |
| **Head and Neck Surgery** | 408-972-6580 | http://www.permanente.net/kaiser/pages/d406-top.html<br><br>280 Hospital Parkway,<br>Building B<br>San Jose, CA 95119 |
| **Northern California Sonoma County – Santa Rosa** | | |
| **Hearing Aid Center** | 707-566-5201<br>707-566-5259 TDD | 3333 Mendocino Avenue, #240<br>Santa Rosa, CA 95403 |

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| **Northern California Placer County – Roseville** | | |
| Hearing Aid Center | 916-771-6680 | 2120 Professional Drive, #220<br>Roseville, CA 95661 |
| **Northern California Contra Costa County – Walnut Creek** | | |
| Hearing Aid Center | 925-295-4327 | 710 South Broadway, #209<br>Walnut Creek, CA 94596 |
| **Northern California Solano County – Vallejo** | | |
| Hearing Aid Center | 707-645-2500 | 761 Broadway, #200<br>Park Place Medical Office Building<br>Vallejo, CA 94589 |
| **Southern California Service Area – San Diego** | | |
| Head and Neck Surgery | 619-280-5998 | http://hns.zion.ca.kp.org |
| **Southern California Service Area – Valley Insider** | | |
| Sign Language Request | | http://valleys.ca.kp.org/Departments/dept.html |

# BIBLIOGRAPHY

## Definitions and Demographics


Alexander, F. M., Gannon, J. R. (1984). Deaf Heritage. Silver Spring, MD: National Association of the Deaf.

Bauman, Neil. 2003. "Ototoxic Drugs Exposed, Second Edition." Center for Hearing Loss Help. 49 Piston Court, Stewartstown, PA 17363. http://www.hearinglosshelp.com

Bogardus ST, Yueh B, Shekelle PG. "Screening and Management of Adult Hearing Loss in Primary Care." The Journal of the American Medical Association, Vol 289(15) 2003, pp. 1986-1990.

Center of Disease Control (2002). Summary of Health Statistics for US Adults: National Health Interview Survey, 1997. Retrieved from http://www.cdc.gov/nchs/data/series/sr_10/sr10_205.pdf

Centers for Disease Control and Prevention, National Center for Health Statistics, Table 59. Vision and hearing limitations among adults 18 years of age and over, by selected characteristics: United States, selected years 1997-2004.

Deaf Life, 10, 112. 1997, October "For hearing people only."

Greinwahl Jr., J. MD, & Hartnick, C. 2002). Evaluation of Children with Sensorineural Hearing Loss. Archives of otolaryngology/ Head and Neck Surgery, 128, 84-87.

Gallaudett University, Sign Languages of the World, by Country. Accessed May 29, 2007 http://library.gallaudet.edu/dr/faq-world-sl-country.html

Gallaudet Research Institute, Annual Survey of Deaf and Hard of Hearing Children and Youth, 2006-2007 Regional and National Summary. Accessed August 5, 2008 http://gri.gallaudet.edu/Demographics/

Hearing Loss Association of America. Facts on Hearing Loss. (2005) Bethesda, MD. http://www.hearingloss.org/learn/factsheets.asp. 08 Sept. 2006

Hearing Review (2005) Hearing loss, hearing and hearing aids: Hear-it. 09 Sept. 2006 www.hear-it.org

Kochkin, S. "MarkeTrak VII: Hearing Loss Population Tops 31 Million People." The Hearing Review, Vol. 12(7) July 2005, pp. 16-29.

Kochkin, S. "MarkeTrak VII: Obstacles to adult non-user adoption of hearing aids." The Hearing Journal, Vol. 60 (4), April 2007, pp. 27-43.

Lane, H. (1984). When the mind hears: a history of the deaf. New York: Random House, Inc.

Lane, H., Hoffmeister, R., Bahan, B. (1996). A journey into the deaf-world. DawnSignPress.

Li, Y., Ban, L. (2003). Parental Decision Making and the Choice of Communication Modality for the Child who is Deaf. Archives of Pediatric Adolescent Medicine, vol. 157, 162-168.

Lotke, M. (1995). She Won't Look at Me. Annals of Internal Medicine, 123, 54-57.

McCreary Stebnicki, J., RN, MSN, & Coering, H., RN, PhD (1999). The Culture of the Deaf. Journal of Transcultural Nursing, vol. 10, no. 4, 350-357.

National Association of the Deaf on the World Wide Web: http://www.nad.org

Noonan, D. (2005). A Little Bit Louder, Please. Newsweek. Retrieved from June 6th issue, from http://www.msnbc.com.html

Oghala, J., Chen, L., Breunen, M., Tonini, R., Mandolidis, S., (2002). Neonatal Hearing Loss in the Indigent. Laryngoscope, 112, 281-286.

Padden, C., Humphries, T. (1990). Deaf in America: voices from a culture. Boston: Harvard University Press.

Scott, D. M. (2005, Nov. 29). Hearing research: Children from culturally and linguistically diverse populations. The ASHA Leader, p. 4, 22-23.

Smith, R. (2005). Hereditary Hearing Loss in Humans: Where are we in 2005? The Registry, vol. 13, no.1, 1-2.

Tamaska, P., Malia, T., Stein, C., Graenflo, D., Meador, H., Zizove, P. (2000). Preventive Attitudes and Beliefs of Deaf and Hard of Hearing Individuals. Archives of Family Medicine, vol. 9 (6), 518-525.

WebMD.com: http://www.webmd.com

Wiley, T., Torre III, P., Cruckshonles, K., Nondahl, D., Tweed, T. (2001). Hearing Sensitivity in Adults Screened for Selected Risk Factors. Journal of American Academy of Audiology, 12, 337-347.

## Cultural Perspectives, Beliefs, and Behaviors

Berlin EA. & Fowkes WC, Jr.: "A teaching framework for cross-cultural health care—Application in family practice, In Cross-cultural Medicine." West J. Med. 1983, 12: 139, 93-98

Centers for Disease Control and Prevention. "Economic costs associated with mental retardation, cerebral palsy, hearing loss, and vision impairment," United States, 2003. MMWR 2004;53:57-9.

Hearing Loss Web: http://www.hearinglossweb.com/index.htm

Jackson L. Understanding, eliciting, and negotiating client's multicultural health beliefs. Nurse Practitioner. 1993;18(4) 30-34.

Kleinman, A. "Patients and Healers in the Context of Culture." The Regents of the University of California. 1981.

Prochaska, J. O., DiClemente, C. C., & Norcross, J. C. (1992). In search of how people change: Applications to addictive behaviors. American Psychologist, 47(9), 1102-1114.

Prochaska, J. O., Norcross, J. C., & DiClemente, C. C. (1994). "Changing for Good: A Revolutionary Six-Stage Program for Overcoming Bad Habits and Moving your Life Positively Forward." New York: Avon Books.

Prochaska, J. O., Redding, C. A., & Evers, K. E. (1997). The transtheoretical model and stages of change. In K. Glanz, F. M. Lewis, & B. K. Rimer (Eds.), "Health Behavior and Health Education: Theory, Research, and Practice." 2nd ed. San Francisco: Jossey-Bass Publishers.

Velicer, W. F, Prochaska, J. O., Fava, J. L., Norman, G. J., & Redding, C. A. (1998) Smoking cessation and stress management: Applications of the Transtheoretical Model of behavior change. Homeostasis, 38, 216-233.

Zimmerman, GL., Olsen, CG., and Bosworth, MF. "A 'Stages of Change' Approach to Helping Patients Change Behavior." Am Fam Physician 2000;61:1409-16. Wright State University School of Medicine, Dayton, Ohio, http://www.aafp.org/afp/20000301/1409.html

## Communicating with the People with Hearing Loss

Barnett, S. (2002) Cross-Cultural Communication with Patients Who Use American Sign Language. Family Medicine, vol. 34, no. 5, 376-382.

Harlan Lane, Robert Hoffmeister, and Ben Bahan. "A journey into the deaf-world." San Diego, California DawnSignPress, 1996.

Hines, J. (2000). Communication Problems of Hearing Impaired Patients. Nursing Standard, vol. 14, no. 19, 33-37.

Lotke, M. (1995). She Won't Look at Me. Annals of Intern Medicine, 123, 54-57.

McCreary Stebnicki, J., RN, MSN, & Coering, H., RN, PhD (1999). The Culture of the Deaf. Journal of Transcultural Nursing, vol. 10, no. 4, 350-357.

McCreary Stebnicki, J., RN, MSN, & Coering, H., RN, PhD (1999). The Culture of the Deaf. Journal of Transcultural Nursing, vol. 10, no. 4, 350-357.

Tamaska, P., Malia, T., Stein, C., Graenflo, D., Meador, H., Zizove, P. (2000). Preventive Attitudes and Beliefs of Deaf and Hard of Hearing Individuals. Archives of Family Medicine, vol. 9 (6), 518-525.

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## Risks Factors

Bluestone, Charles, D., MD, Stool, 2nd ed., W.B. Saunders company, 1990, 3-59.

Crandell, C., Mills, T., Gauther, R. (2004). Knowledge, Behaviors, Attitudes about Hearing Loss and Hearing Protection among Racial/Ethnically Diverse Young Adults. Journal of the National Medical Association, vol. 69, no. 2,176-186.

Duff, B. (2001). Hearing Loss in Children with Craniofacial Anomalies. Medicine and Health /Rhode Island, vol. 84, no. 12, 399-400.

Greinwahl Jr., J. MD, & Hartnick, C., (2002). Evaluation of Children with Sensorineural Hearing Loss. Archives of otolaryngology/ Head and Neck Surgery, 128, 84-87.

Honeycutt, A., Dunlap, L., Chen, H., Homsi, G. (2004). Economic Costs Associated with Mental Retardation, Cerebral Palsy, Hearing Loss, and Vision Impairment-U.S., 2003. Morbidity and Mortality Weekly Review, vol. 53, No. 3, 57-59.

Kountakes, S., Skoulas, I., Phillips, D., Chang, C. (2002). Risk Factors for Hearing Loss in Neonates: A Prospective Study. American Journal of Otolaryngology, vol. 23, no. 3, 133-137.

Mann, T., Cuttler, K., Campbell, C. (2001). Newborn Hearing Screens May Give A False Sense of Security. Journal American Academy of Audiology, 12, 215-219.

Murphy, M., Gates, G., (1999). Hearing Loss: Does Gender Play a Role? Medscape general Medicine, vol.1, no. 2.
http://www.medscape.com

National Institute of Health Consensus Statement. (1993). Vol. 11, no. 1.

Noonan, D. (2005). A Little Bit Louder, Please. Newsweek. Retrieved from June 6th issue, from http://www.msnbc.com.html

Oghala, J., Chen, L., Breunen, M., Tonini, R., Mandolidis, S., (2002). Neonatal Hearing Loss in the Indigent. Laryngoscope, 112, 281-286.

Rabinowitz, P., Duren, R., Is Acculturation Related to the Use of Hearing Protection? (2001). AIHAJ, 62, 611-614.

Smith, R. (2005). Hereditary Hearing Loss in Humans: Where are we in 2005? The Registry, vol. 13, No.1, 1-2.

Wang, S., (2005). Listen: iPods Can Damage Your Ears. Philadelphia Inquirer.
http://www.timesleader.com.html

Wiley, T., Torre III, P., Cruckshonles, K., Nondahl, D., Tweed, T. (2001). Hearing Sensitivity in Adults Screened for Selected Risk Factors. Journal of American Academy of Audiology, 12, 337-347.

Woolley, A., Kirk, K., Neumann, A. (1999). Risk Factors for Hearing Loss from Meningitis in Children: The Children's Hospital Experience. Archives of Otolaryngology-Head and Neck Surgery, vol. 125, no. 5, 509-514.

## Occupational Exposures, Disability, and Legal Issues

Adapted from http://www.nad.org/healthcare and http://www.nad.org/adaq&a (Oct. 7, 2005); National Association of the Deaf (NAD) Law and Advocacy Center, Silver Spring, MD. For more information, contact the NAD Law and Advocacy Center at (301) 587-7730 or nadlaw@nad.org.

For more information about Kaiser Permanente California ADA Compliance programs and resources see our website: http://kpnet.kp.org/ada/

## Idiopathic Sudden Sensorineural Hearing Loss

Byl, F., (1984). Sudden Hearing Loss: Eight Years Experience and Suggested Prognostic Table. Laryngoscope, 94, 647-651.

Chandrasekhar, S., (2001). Intratympanic Dexamethasone for Sudden Sensorineural Hearing Loss: Clinical and Laboratory Evaluation. Otol Neurotol, 22, 18-23.

Cinamon, U., Bendet, E., Kronenberg, J., (2001). Steroids, Carbogen or Placebo for Sudden hearing Loss: A Prospective Double-Blind Study. Eur Arch Otorhinolaryngol, 258, 477-480.

Ciuffetti, G., Scardazza, A., Serafini G., Lombardini, R., Mannarino, E., Simoncelli, C. (1991). Whole-blood filterability in sudden deafness. Laryngoscope, 101, 65-67.

Fetterman, B., Sanders, J., Luxford, W., (1996). Prognosis and Treatment of Sudden Sensorineural Hearing Loss. Am J Otol, 17, 529-536.

Fisch, U., Murata, K., Hossli, G., (1976). Measurement of oxygen tension in human perilymph. Acta Otolaryngol, 81, 278-282.

Ginaoli, G., Li, J., (2001). Transtympanic Steroids for Treatment of Sudden Hearing Loss. Otolaryngol-Head Neck Surg, 125, 142-146.

Jackler, R., Brackmann, D., (2005). Neurotology (2nd ed.). Philadelphia: Elsevier Mosby.

Kanzaki, J., Taiji, H., Ogawa, K.,(1988). Evaluation of Hearing Recovery and Efficacy of Steroid Treatment in Sudden Deafness. Acta Otolaryngol Suppl, 456, 31-36.

Katz, J., Stecker, N.A., & Henderson, D. (1992). Introduction to central auditory processing. In J. Katz, N.A. Stecker & D. Henderson (Eds.), Central auditory processing: A Transdisciplinary view St. Louis: Mosby Year Book, Inc.

Kopke, R., Hoffer, M., Wester, D., O'leary, M., Jackson, R., (2001). Targeted Topical Steroid Therapy in Sudden Sensorineural Hearing Loss. Otol Neurotol, 22, 475-479.

Moskowitz, D., Lee, K., Smith, H., (1984). Steroid use in Idiopathic Sudden Sensorineural Hearing Loss. Laryngoscope, 94,664-666.

Schuknecht, H., Kimura, R., Naufal, P., (1973). The pathology of sudden deafness. Acta Otolaryngol, 76, 75-97.

Stokroos, R., Albers, F., Tenvergert, E., (1998). Antiviral Treatment of Idiopathic Sudden Sensorineural Hearing Loss: A Prospective, Randomized, Double-Blind Clinical Trial. Acta Otolaryngol, 118, 448-495.

Stokroos, R., Albers, F., Schirm, J., (1999). Therapy of Idiopathic Sudden Sensorineural Hearing Loss: Antiviral Treatment of Experimental Herpes Simplex Virus Infection of the Inner Ear. Ann Otol Rhinol Laryngol, 108, 423-428.

Tran Ba Huy, P., Sauvaget, E., (2005). Idiopathic Sudden Sensorineural Hearing Loss is not an Otologic Emergency. Otol Neurotol, 26, 896-902.

Tucci, D., Farmer, J., Kitch, R., Witsell, D., (2002). Treatment of Sudden Sensorineural Hearing Loss with Systemic Steroids and Valacyclovir. Otol Neurotol, 23, 301-308.

Wilson, W., (1984). Why Treat Sudden Hearing Loss. Am J Otol, 5, 481-483.

Wilson, W., Byl,F., Laird, N., (1980). The Efficacy of Steroids in the treatment of Idiopathic Sudden Hearing Loss. Arch Otolarygol, 106, 772-776.

Wilson, W., Veltri, R., Laird, N., Sprinkle, P. (1983). Viral and epidemiologic studies of idiopathic sudden hearing loss. Otolarygol Head and Neck Surg, 91, 653-658.

## Autoimmune Hearing Loss

Etanercept therapy for immune-mediated cochleovestibular disorders: a multi-center, open-label, pilot study. Arthritis Rheum 2005;53:337-342.

Broughton SS, Meyerhoff WE, Cohen SB. Immune-mediated inner ear disease: 10-year experience. Semin Arthritis Rheum 2004;34:544-548.

Harris JP, Sharp PA. Inner ear autoantibodies in patients with rapidly progressive sensorineural hearing loss. Laryngoscope 1990;100:516-524.

Matteson EL, Choi HK, Poe DS, Wise C, Lowe VJ, McDonald TJ, Rahman MU.

McCabe BF. Autoimmune sensorineural hearing loss. Ann Otol Rhinol Laryngol 1979;88:585-589.

Ruckenstein MJ. Autoimmune inner ear disease. Curr Opin Otolaryngol Head Neck Surg 2004;12:426-430.

## Pediatric Population Sections

Americans with Disabilities Act of 1990, Public Law 101-336, 42, U.S.C. 12101 et seq.: U.S. Statutes at Large, 104, 327-328 (1991).

American Speech-Language-Hearing Association. (1995). ASHA desk reference for audiology and speech-language pathology, II, 71-82.

American Speech-Language-Hearing Association. (1993). Guidelines for audiology services in the schools. Asha, 35(Suppl. 10), 24-32.

American Speech, Language and Hearing Association. How Does Your Child Hear and Talk. Developmental milestones for speech and language. http://www.asha.org/public/speech/development/child_hear_talk.htm 08 Sept. 2006

Beltman JA, Jonkers Y, Nuijten I, et.al. Definition of a critical region on chromosome 18 for congenital aural atresia by arrayCGH. Am J Hum Genet 2003;72(6):1578-84.

De La Cruz A, Teufert KB. Congenital aural atresia surgery: Long-term results. Otolaryngol Head Neck Surg 2003;129:121-127.

Educational Audiology Association, 1997. EAA Position Statement: Recommended Professional Practices for Educational Audiology. www.edaud.org

Hood LJ. Auditory neuropathy: What is it and what can we do about it? The Hearing Journal. Aug 1998. 51(8): 10-18 cited from www.nidcd.nih.gov/health/hearing/neuropathy

Johnson, C.D., Benson, P., Seaton, J. (1997). Educational Audiology Handbook. San Diego: Singular Publishing Group, Inc.

Joint Committee on Infant Hearing, 2000. Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. Pediatrics, Vol. 106 No. 4.

Lambert PR. Congenital aural atresia. In: Bailey BJ, Calhoun KH, editors. Head and Neck Surgery - Otolaryngology, 3rd Ed. Philidelphia: Lippincott Williams & Wilkins; 2002:1745-1757.

Matkin, N. (1994). Key considerations in the provision of family centered services. Paper presented at the Colorado State Symposium on Deafness, Colorado Springs, CO.

Morton NE. Genetic epidemiology of hearing impairment. Ann New York Audiological Soc 1991;630:16-31.

National Center for Hearing Assessment and Management (NCHAM)
www.infanthearing.org

Penn, T. (1999). School-based hearing screening in the United States. Audiology Today, 11, 20-21.

Rehabilitation Act of 1973, Section 504, 29, U.S.C. 794 U.S. Statutes at Large, 87, 335-394 (1973).

Snyder, N., Diggs, C., Whitmire, K., Karr, S., Clarke, C., Franklin, R. (2005). IDEA reauthorization signed into law. The ASHA Leader, 10, 1 and 22.

Steel KP. A new era in the genetics of deafness. N EnglJ Med 1998;339(21):1545-7.

Gurtler N, Lalwani AK. Etiology of syndromic and nonsyndromic sensorineural hearing loss. Otolaryngol Clin North Am 2002;35(4):891-908.

U.S. Department of Education. (September 29, 1992). Assistance to states for the education of children with disabilities program and preschool grants for children with disabilities; Final rule (34 CFR Parts 300 and 301). Federal Register, 57, 189.

Yeakley JW, Jahrsdoerfer RA. CT evaluation of congenital aural atresia: What the radiologist and surgeon need to know. Journal of Computer Assisted Tomography 1996;20(5):724-731.

Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early and later-identified children with hearing loss. Pediatrics. 1998; 102:

## Treatment Options

American Speech-Language-Hearing Association, "Assistive Technology: What are Assistive Listening Devices?" May 29, 2007 http://www.asha.org/public/hearing/treatment/assist_tech.htm

American Speech-Language-Hearing Association, "Cochlear Implants." May 29, 2007
http://www.asha.org/public/hearing/treatment/cochlear_implant.htm

Assistech Inc. "Assistive Devices Network: Deaf and Hard of Hearing." (list of hearing assistive devices) May 29, 2007 http://www.assistivedevices.net/

Entific Medical Systems, "About Baha® - A difference patients can hear." May 29, 2007
http://www.entific.com/aboutbaha.asp

Federal Communications Commission, Consumer & Governmental Affairs Bureau, Disability Rights Office, May 29, 2007 http://www.fcc.gov/cgb/dro/

Federal Trade Commission, FRC Consumer Alert, "Sound Advice on Hearing Aids." May 29, 2007
http://www.ftc.gov/bcp/conline/pubs/health/hearing.shtm

Healthy Hearing, "Helpful Information on Hearing Loss, Hearing Aids, Cochlear Implants, Tinnitus, and more" May 29, 2007 http://www.healthyhearing.com/

Hearing Aids 101, "Hearing Aids Information and Advice." May 29, 2007, http://www.hearingaids101.com/

National Institute on Deafness and Other Communication Disorders, National Institutes of Health, "Cochlear Implants", May 29, 2007 http://www.nidcd.nih.gov/health/hearing/coch.asp.

National Institute on Deafness and Other Communication Disorders, National Institutes of Health, "Hearing Aids," May 29, 2007 http://www.nidcd.nih.gov/health/hearing/hearingaid.asp

University of Maryland, Medical Center, Maryland Hearing and Balance Center, "Baha Implant." http://www.umm.edu/otolaryngology/baha.html

U.S. Food and Drug Administration, Center for Devices and Radiological Health, "Cochlear Implants." May 29, 2007. http://www.fda.gov/cdrh/cochlear/index.html

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1. In what context did you receive the Handbook(s)?

____Training/Workshop _____Individual Request ____Other: specify (____)

2. Please rate the effectiveness of the Handbook(s) as learning tools:

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|---|---|---|---|---|---|
| Latino | 1 | 2 | 3 | 4 | 5 |
| Disability | 1 | 2 | 3 | 4 | 5 |
| African American | 1 | 2 | 3 | 4 | 5 |
| Asian and Pacific Islander (API) | 1 | 2 | 3 | 4 | 5 |
| Lesbian, Gay, Bisexual and Transgender (LGBT) | 1 | 2 | 3 | 4 | 5 |
| People with Hearing Loss | 1 | 2 | 3 | 4 | 5 |

3. Please rate the effectiveness of the Handbook(s) in improving cross-cultural clinical skills:

| | Not at All | | Somewhat | | Extremely |
|---|---|---|---|---|---|
| Latino | 1 | 2 | 3 | 4 | 5 |
| Disability | 1 | 2 | 3 | 4 | 5 |
| African American | 1 | 2 | 3 | 4 | 5 |
| Asian and Pacific Islander | 1 | 2 | 3 | 4 | 5 |
| Lesbian, Gay, Bisexual and Transgender | 1 | 2 | 3 | 4 | 5 |
| People with Hearing Loss | 1 | 2 | 3 | 4 | 5 |

4. Describe what you like about the Handbook(s):

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