CHAPTER 2

Causes of Being Deaf and the Auditory Field

In order to understand different types of ways Deaf people are deaf, we need to understand what being "hearing" is. Deaf people call people who can hear and speak, "Hearing" or "Hearing people." Another phrase that is used for hearing people is, "Hearing world," referring to spaces populated by hearing people with speaking and hearing privileges, which is pretty much the entire world that we currently inhabit. That means that many of the services (e.g., fast-food drive-through, schools, workplaces, prisons, concert halls) and devices (e.g., doorbell intercoms in apartment buildings, loudspeakers at sporting events, automated registers) are designed *for* people who hear and speak, and *by* people who hear and speak.

People who may not hear or speak the same way hearing people do mostly identify themselves as Deaf, deaf, DeafBlind, DeafDisabled, hard of hearing, or late-deafened. If we meet someone from the deaf communities, how do we know which term to use to describe them? The best approach is to ask how they identify themselves. Most of the time, they'll tell you which term they identify with. But what if you want to refer to a group of people who do not hear or speak like hearing people do? You can use all those terms. Some people use this acronym, DDBDDHHLD (which represents the letters of those terms), to be inclusive of all the different identities within the Deaf communities (Council de Manos, 2019).

So, now do we know if they are DDBDDHHLD? Often caregivers who are puzzled when an infant or a young toddler, someone recovering from an illness, or a senior citizen does not respond to sound. Those caregivers might try making a sound behind them and see if they respond and turn their heads. If they don't respond, we still don't know how much they can hear, what kind of sounds they can hear, or if it's something else. There are ways to find the answers to those questions. Keep in mind, many Deaf individuals do not tend to want answers to those questions, as they are content with being Deaf. However, during their early formative years, and even throughout their childhood and adulthood, families of Deaf children may have been exposed to conflicting information (Humphries, Kushalnagar, Mathur, Napoli, Padden, Pollard, et al., 2014; Humphries, Kushalnagar, Mathur, Napoli, Padden, & Rathmann, 2014; Humphries et al., 2019) and misinformation (Hall, 2017) from medical and audiology professionals. For example, some professionals and organizations advocate for preventing deaf children from learning sign language before implantation. It is often standard practice for them to tell parents that they need to be using spoken language only and that the use of sign language should be avoided. This is counter to the fact that there is no evidence that shows sign languages harm the child's acquisition of spoken languages. In fact, evidence points to benefits of learning speech through sign language. There is also increasing evidence that depriving a deaf child of access to sign language access has a potential negative impact on the child (Hall, 2017; Humphries, Kushalnagar, Mathur, Napoli, Padden, Pollard, et al., 2014; Humphries, Kushalnagar, Mathur, Napoli, Padden, & Rathmann, 2014;). This is not to say all audiologists are uninformed and spread misinformation (Andrews & Dionne, 2008). For a more medically focused approach to audiology testing and rehabilitative issues, please see Martin and Clark (2019).

DETERMINING ONE'S HEARING LEVEL

Audiologists and Audiograms

According to the Center for Hearing and Communication (CHC, 2020, approximately 48 million Americans have some sort of hearing challenges. This number includes those who are born deaf or are part of Deaf culture. When confronted with the possibility of a difference in hearing, those people tend to be referred to an audiologist. An audiologist is a professional who specializes in detecting hearing levels and proposing different types of accommodations. The field an audiologist works in is called audiology. Pictured in Figure 2–1 is an audiologist working with a young client.

The purpose of an audiological evaluation is to measure the degree, type, and configuration of hearing levels by utilizing a physical examination of the ear, tests of hearing and listening, and tests of the middle ear function (Martin & Clark, 2019). An audiologist will first conduct a physical exam by looking at the outer ear for evidence of malformations. Then the audiologist uses an otoscope, which is an instrument that contains light and a magnifying glass, and inserts it into the ear to examine the ear canal and eardrum to see if there is excessive earwax or objects that could obstruct hearing. The audiologist also examines the condition of the eardrum and notes any excess fluid. A medical referral for further evaluation or treatment may be an outcome of this physical exam.

Next, the audiologist conducts tests of hearing tones or pure-tone audiometry. The individual enters a soundproof room and is fitted with earphones. Then the audiologist will leave the room and enter an adjacent room with a window through which both the client and the audiologist can see each other, as shown in Figure 2–2.

The audiologist will then proceed with turning on a machine that emits pure tones at selected pitches or frequencies to find the lowest tone that the individual responds to. The audiologist will also turn on low sounds, such as leaves rustling and water dripping, and then increase the loudness of the sounds to whispering, spoken language, a baby crying, a phone ringing, a dog barking, a running vacuum, a lawn mower, to a jet plane roaring by (Martin & Clark, 2019; Sheetz, 2012). The goal is to increase the sound level until the individual indicates they hear the sound by raising their hand. The audiologist varies the sounds, including the pitch level from low to high, to determine the hearing level in each ear. Figure 2–3 shows the frequencies and loudness levels of different types of sounds. Loudness is measured by decibels, which are units that measure how loud a sound is.

Figure 2–1. An audiologist at work. Photo courtesy of Brian Sattler. Used with permission.

Figure 2–2. An audiologist testing the hearing of a client in a soundproof testing booth. Photo courtesy of Brian Sattler. Used with permission.

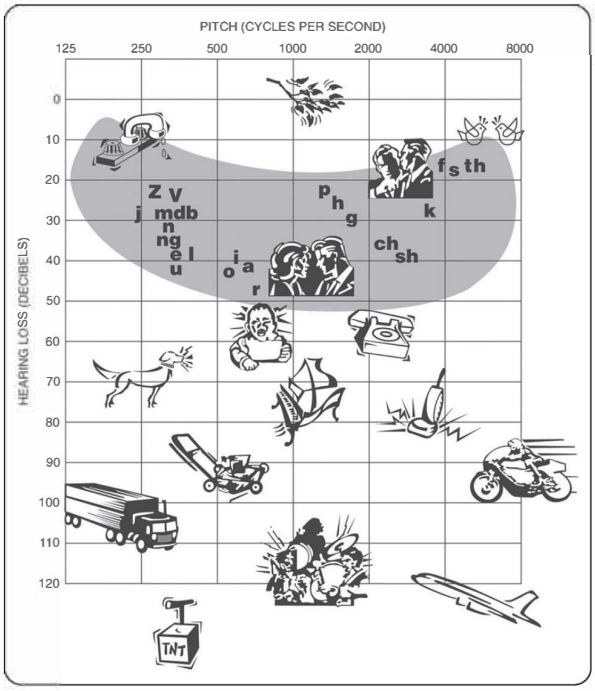

Figure 2–3. Diagram of the frequency and loudness levels of different sounds. Scheetz, Nanci A., *Deaf Education in the 21st Century: Topics and Trends.* © 2012. Printed and electronically reproduced by permission of Pearson Education, Inc., New York, NY.

The audiologist will also conduct tests to examine the "speech reception threshold," which means testing the quietest speech that can be heard. In another test, a standardized list of words is presented one at a time to the individual to assess the ability to recognize words across different loudness levels (Martin & Clark, 2019). Other tests may be administered to examine the functioning of the outer and middle ear. These are called "acoustic immittance measures" (Martin & Clark, 2019). These tests can detect blockage in the ear canal, fluid in the middle ear, or a puncture in the eardrum (Martin & Clark, 2019).

After these audiologic tests are completed, this information is then documented on an audiogram and recommendations are made for follow-up testing or medical referrals if necessary. Referrals also are made for assistive listening devices, speech and language counseling, or further audiologic rehabilitation (Martin & Clark, 2019).

The audiogram is a chart that measures sound from 0 to 120 decibels (dB) and pitch from 125 to 8,000 cycles per second (Sheetz, 2012). The hearing level of the right ear is indicated by a circle, while X is used to show the hearing level of the left ear. Here, the authors provide examples of their audiograms, as shown in Figures 2–4A–D.

Based on the information displayed in the audiograms and comparing these with Figure 2–3, we can surmise that Topher González Ávila, Raychelle Harris, and Irene Leigh (without her hearing aid) will not be able to hear a vacuum, a dog barking, a phone ringing, or a baby crying. We can surmise Jean Andrews can hear all of those, like most hearing people. However, Andrews and other hearing individuals in the late 60s age range may start to experience presbycusis, defined as changes in hearing levels, particularly in the higher frequencies, as they age.

Topher, Raychelle, and Irene will also not be able to hear spoken conversations. But they may or may not hear a lawn mower, an 18-wheeler truck, a bomb and possibly a

live band, a speeding motorcycle, and a jet plane, depending on the pitch, but this is not always necessarily accurate. Intriguingly, Harris is often alerted to someone knocking on the door when her Rottweiler, Samson, barks. According to Raychelle's audiogram and Figure 2–3, Raychelle is not supposed to be able to hear a dog barking. In other words, "hearing" is not an exact science, and everyone varies in their processing of sound.

The audiologist prepares the audiogram and gives the individual a specific label that corresponds with the hearing level (dB) as indicated in the audiogram. A person with a 10- to 15-dB hearing level would be labeled as having normal or typical hearing. At the next level, 16 to 25 dB would be identified as having a *slight* hearing loss. Someone receiving a *moderate* hearing label is able to hear sounds that are 41 to 55 dB or higher. Those testing at 56 to 70 dB would be told they have a *moderately severe* hearing level. A person with a *severe* hearing label would hear a range of 71 to 90 dB or higher. People with *profound* hearing levels would only be able to hear sounds that are 91 dB or above (Martin & Clark, 2019; Sheetz, 2012). Looking at Topher, Raychelle, and Irene's audiograms, as shown above, we see that audiologists would place them in the *profound* hearing level category, or from the view of some members of Deaf communities, they would receive the designation of an ASL sign, DEAF (with puffed cheeks), which is loosely translated as "truly Deaf" or "so Deaf." Table 2–1 shows each hearing level and the labels for each.

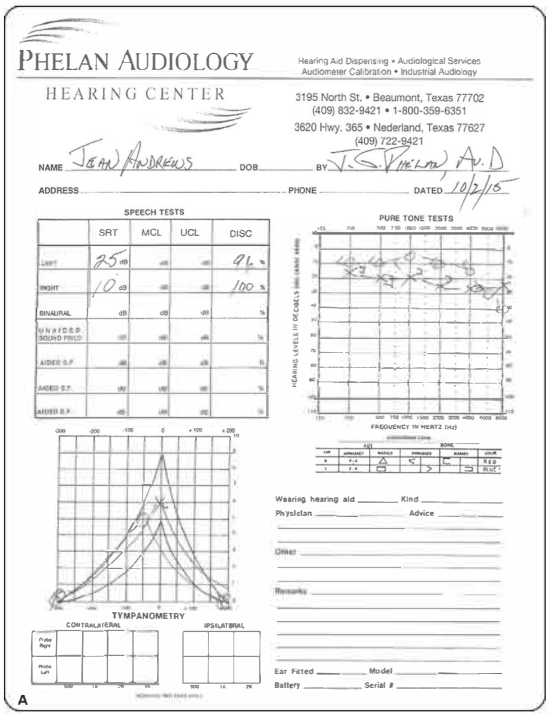

Figure 2–4. A. Jean Andrews' audiogram.

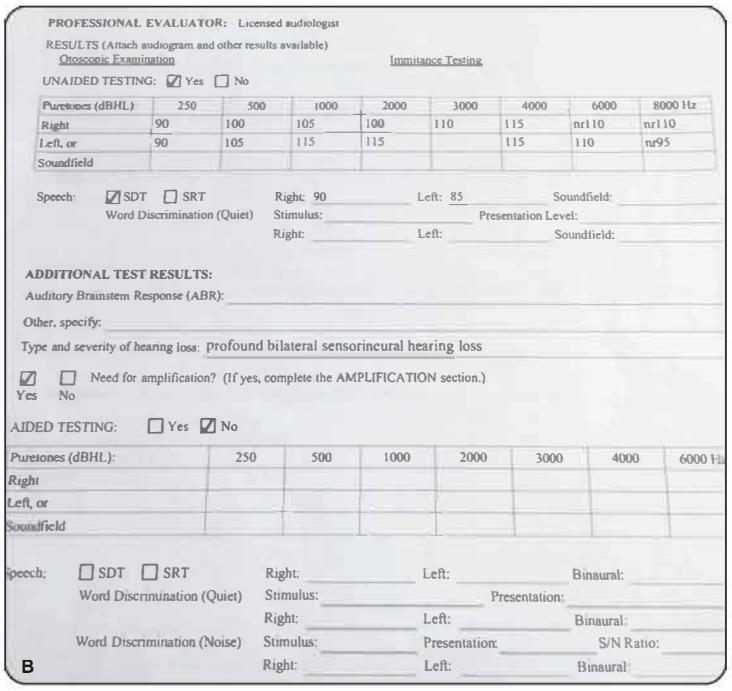

Figure 2–4. B. Topher González Ávila's audiogram.

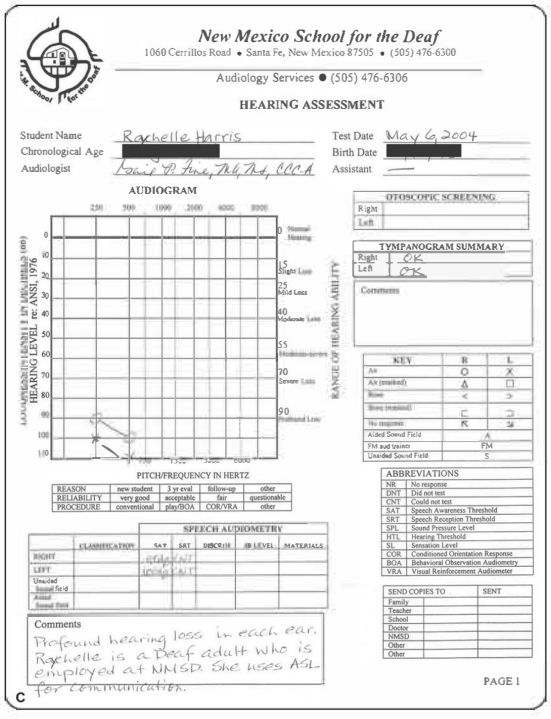

Figure 2–4. C. Raychelle Harris' audiogram.

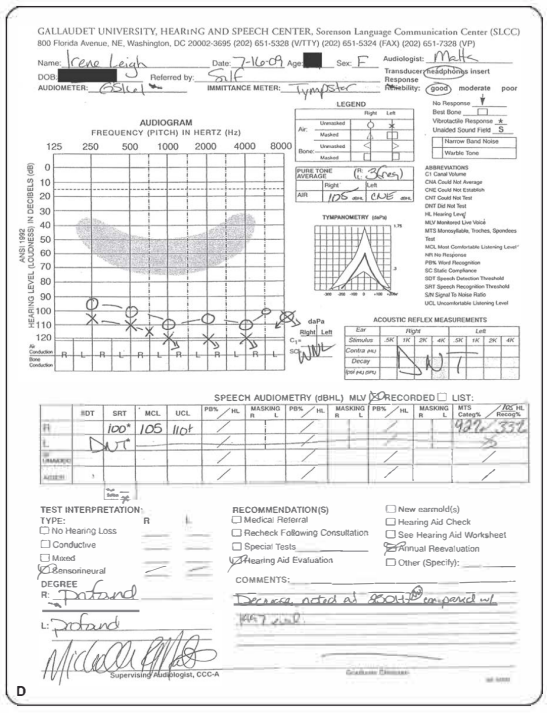

Figure 2–4. D. Irene Leigh's audiogram.

Table 2–1. Hearing Levels, Labels, and Examples

Hearing Level	Label	Implications in the Hearing World
−10 to 15 dB	Normal	Can participate seamlessly in spoken conversations
16 to 25 dB	Slight	Can converse in quiet environments; noisy environments can be difficult
26 to 40 dB	Mild	May be able to follow conversation if in quiet environment and topic is familiar
41 to 55 dB	Moderate	Quiet environment and conversations will need to be within 3–5 feet, may benefit from using an hearing aid
56 to 70 dB	Moderately severe	Will not be able to participate in conversations unless loud; will benefit from the above accommodations
71 to 90 dB	Severe	May identify environmental noises and loud sounds; may have difficulty producing intelligible speech
91+ dB	Profound	Does not usually rely on hearing or speech

Source: Adapted with permission of Scheetz (2012, p. 65).

Hearing people may have never seen an audiologist and may have never received an audiogram until their senior years when their family complains that they are not listening or responding only to very loud talking. On the other hand, Deaf people often grow up seeing countless audiologists and have stacks of audiograms from when they were younger. The audiogram is often used for different reasons, such as

qualifying for the Deaflympics, which is similar to the Olympics, but for Deaf athletes (International Committee of Sports for the Deaf, 2018, also briefly described in Chapter 1); receiving Vocational Rehabilitation and Social Security benefits; or being eligible for admission at an educational institution or program serving deaf students.

Hearing Labels

Audiologists and speech professionals typically use the term *hearing impaired* to describe all people with different types of hearing loss. As mentioned in Chapter 1, the term *hearing impaired* is not widely embraced by Deaf people. In any case, audiologists will further categorize people with hearing loss as deaf from birth or at the age when they lost their hearing. They also categorize those who have not spoken or heard language before they became deaf as *prelingually deaf*. This applies to people who were born deaf and did not receive language input, but this does not apply to deaf infants born into language-accessible households (e.g., signing households). In contrast, those who already sign, speak, and/or hear language before they became deaf are *postlingually deaf*. People who are *postlingually deaf* usually remember what it is like to speak and hear before their hearing levels changed (Marschark & Spencer, 2016).

Another term used by audiologists, *hard of hearing*, refers to people who have a slight to moderate hearing loss. *Hard-of-hearing* people often benefit from the use of hearing aids, assistive listening devices, and other forms of amplification (Martin & Clark, 2019). Some *hard-of-hearing* people do not benefit from those devices at all. For example, a person may be able to hear only high-frequency sounds such as a whistle, a bird chirping, or a doorbell but is unable to hear speech. Sometimes a person cannot hear low-frequency sounds and is challenged in understanding mostly adult men, whose speech registers in the low-frequency range (Martin & Clark, 2019; Sheetz, 2012). Some sounds such as *b* and *d* are low frequency—if a person is unable to hear low-frequency sounds such as these, imagine how much of the conversation would be predominately guesswork? On the other hand, people unable to hear high-frequency sounds such as *th* or *s* may also struggle with understanding people whose voice registers in the high-frequency range, which is the case for most adult women. Deaf and hard-of-hearing people often receive questions and comments from naïve hearing people asking why they are able to speak but not hear (both are different skills), or why they are able to hear a dog bark but not someone who is speaking (both have different decibel levels), or why they are able to hear a man speak but not a woman speak (both speak with different frequencies) and so on (Martin & Clark, 2019; Sheetz, 2012). You may also find hard-of-hearing people who identify as Deaf, even if they do hear.

In ASL signers within Deaf communities, there is an ASL phrase that is often transcribed as VERY-HARD-OF-HEARING (puffed cheeks). This means the opposite of what hearing people may think. That phrase is translated as the person being "almost hearing"! Similarly, LITTLE-HARD-OF-HEARING is translated as the person being able to hear just a little, but is overall, mostly Deaf. One of the classic pivotal early books on Deaf culture, titled *Deaf in America: Voices From a Culture*, goes into more detail about those terms (Padden & Humphries, 1988).

Often people are not able to separate the ability to hear from the ability to understand—for example, many Deaf people understand the spoken words for typical encounters such as, "Hello, how are you?" or "What's your name?" because those words are predictable and typically used in the beginning of most conversations between strangers. When the context of the conversation changes, Deaf people tend to try different ways to communicate, such as writing back and forth, typing on their cell phones, gesturing, and/or trying to read lips, which is usually the least effective way to communicate, as many sounds in the English language look the same on the lips such

as "ball" and "mall."

Try turning off the sound on your television or your computer device as you watch people speak. Are you able to follow what they are saying?

Additionally, there are also many other external factors influencing the ability to hear, such as background noise and reverberation, which many assistive hearing devices do not succeed in blocking. This can make it very difficult to hear conversation. If you enter noisy restaurants and have difficulty hearing conversation, you can understand how much harder this would be for people with different hearing abilities.

What Causes Changes in Hearing Levels?

What causes people to have varying hearing levels? People are put in either of those two categories: deaf before/at birth (congenital) and after birth (acquired). And within those categories, there are two areas in the ear where the hearing loss might occur. Issues in the outer and middle ear are called *conductive*. Issues happening inside the ear or within the auditory nerve are called *sensorineural* (Martin & Clark, 2019).

Genetic Causes

Genes that are inherited and gene mutations are the cause of deafness in approximately more than 50% of babies born deaf (Knoors & Marschark, 2014). So far, over 400 different genes have been found to cause people to become deaf, with scientists still trying to identify more genes (Smith, Shearer, Hildebrand, & Camp, 2014). Some of those genes make the baby deaf before birth, some during the toddler or teenager years, and some later in life. As for the hundreds of different deaf genes, approximately two-thirds of those "deaf" genes are nonsyndromic, meaning that these genes only cause the person to become deaf without any other physical changes. Connexin 26 is one example of a common nonsyndromic gene that many Deaf families may carry from generation to generation (Clark, 2003). The remaining genes are syndromic, which means that the affected person will not only be deaf but will also have additional conditions, including, for example, blindness, heart conditions, or intellectual development challenges, among other additional disabilities (Plante & Beeson, 2008). Examples of deaf (and additional disabilities) include people having Hunter syndrome (growth failure), Usher syndrome (progressive blindness), and Waardenburg syndrome (pigment abnormalities) (Scheetz, 2012; Vernon & Andrews, 1990).

Acquired

Those who are diagnosed as acquired became deaf due to external factors—not related to genetics. Those external factors that cause deafness develop during birth or after a baby is born and can happen any time during their lives. Examples include diseases such as meningitis, Ménière disease, premature births, fetal alcohol syndrome, or simply becoming elderly (Knoors & Marschark, 2014). For example, in the 1960s, there was a sudden, large increase of deaf children due to rubella, widely known as German measles. In 1969, a rubella vaccine was developed, and after that, the number of children contracting rubella was significantly reduced. The most common cause for hearing loss in adulthood is usually due to damage to the hearing mechanism. Such damage to the hearing mechanism can be the result of prolonged exposure to acute loud noise, the taking of drugs, the aging process, accidents that cause trauma to the hearing mechanism, and diseases that attack and damage the hearing mechanism (Martin & Clark, 2019; Sheetz, 2012).

Conductive

For those diagnosed as conductive, that term specifies challenges within the outer and middle ear. Examples include ears that are not fully open, earwax in the ear, ear infections, and physical injuries to the ear (such as a Q-tip puncturing an eardrum). Often external and middle ear issues can be fixed with medicine or surgery. Surgeries include removing excessive buildup of fluid, adding a tube, removing a blockage, repairing by adding a skin graft, or reconstruction of the damaged parts inside the ear. Conductive losses tend to be temporary (Martin & Clark, 2019; Sheetz, 2012). Figure 2–5 shows the external, middle, and internal sections of the ear.

Sensorineural

Sensorineural issues are limited to the cochlea inside the inner ear and the connecting auditory nerve. The cochlea looks like a very small snail and is the size of a pea. The cochlea transmits sound from the middle of the ear to the auditory nerve. The transmission process includes over 20,000 hairs inside the cochlea, where sounds move through waves of hair to the auditory nerve. Damage to the cochlea can include missing hair or a disorder where sound is not carried from the cochlea to the auditory nerve (Martin & Clark, 2019). People with sensorineural challenges sometimes experience drastic changes in the sensation of loudness; for example, someone might ask you to speak louder and then, in the next minute, ask you why you are shouting. Sensorineural issues cannot be repaired by medicine or minor medical intervention (e.g., adding a tube) and is usually permanent (Sheetz, 2012).

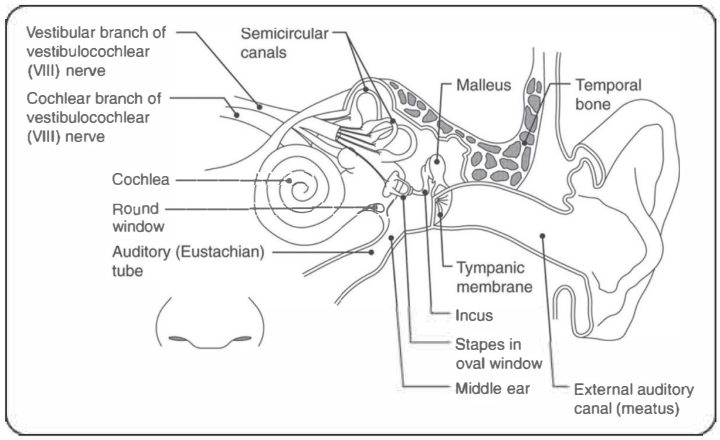

Figure 2–5. A diagram of the ear including external, middle, and internal sections. Courtesy of Marie A. Scheetz.

HISTORY OF AUDITORY TECHNOLOGY

The history of the relationship between the Deaf community and auditory technology is a complicated one, fraught with heartbreaking stories of coercion, suffering, and even death in the process of trying to create the ability to "hear" (Paludneviciene & Harris, 2011). For many centuries, there was a prevailing belief that people who were disabled at birth were being punished or were manifesting demonic origins, this being predetermined by the gods. Babies with disabilities, including deaf ones, would be abandoned, killed, or imprisoned. Simultaneously, attempts to cure deaf people have existed for centuries (Davis, 2006). Many of those "cures" only aggravated the damage for the deaf person. For instance, the use of hot oil with boiled worms in the ear or an operation on the ligament of the tongue to get them to speak were excruciatingly painful treatments. Other aggressive and assault-like actions such as

the repeated shaking of the head or forcing deaf people to shout so loudly that blood came out of their ears and mouths were often tried, in theory, that it would awaken their hearing (Winzer, 1993).

Can you imagine enduring those treatments for young children in order to "cure" their being deaf?

Other miracle cures sold by get-rich-quick medicine folks included magnetic head caps, vibrating machines, artificial eardrums, blowers, inhalers, massagers, magic oils, and creams, all with promises for permanent cures (Davis, 2006). Some charlatan-healers would strike the deaf person's head hard enough to fracture it, in hopes that the blow would shake something loose. Ear infections were treated with a white-hot iron applied and poked into the area behind the ear. Those "cures" persisted well into the 20th century (Winzer, 1993). Figure 2–6 shows a pamphlet proclaiming a cure for deafness.

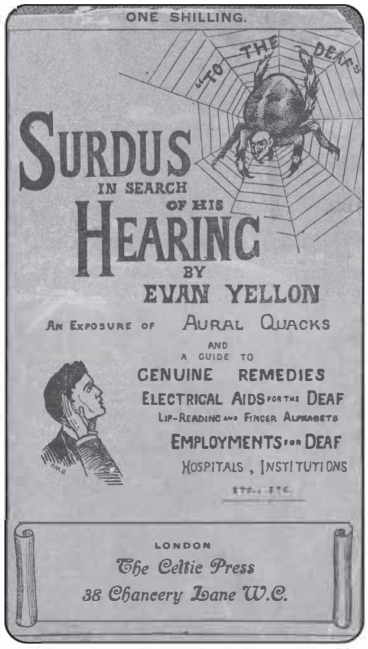

Figure 2–6. A 1906 advertisement proclaiming a cure for deafness. Courtesy of Gallaudet University Archives.

Although such cruel techniques have been abandoned in the United States, many deaf adults today remember being forced to speak English (and read lips) throughout their schooling, and if they tried to sign or gesture, they would be harshly disciplined by having their hands whipped with a ruler. Another cruel practice aimed at humiliation was to make the offending signer stand in the corner for hours (Baynton, 1996). The prohibition of deaf children from learning or using sign language still happens today, mostly without the explicit physical aspect of the punishment. However, implicitly, the punishment pervades as parents and educators may be instructed to avoid using sign language with deaf children. The thinking behind this is that if deaf children sign (or learn to sign), they will be less likely to want to learn how to speak and socialize with other deaf people. In turn, they will be more likely to successfully integrate into the hearing world (Knoors, Tang, & Marschark, 2014). However, this has not consistently proven to be the case (see Chapter 5 on Deaf education in this book).

For people with a medical perspective, utilizing auditory technology is usually the default mechanism for trying to make deaf people into hearing people. Auditory technology has evolved over time, starting with the development of ear trumpets, which were used to amplify sounds for hard-of-hearing individuals by collecting sounds and funneling them into the ear canal. The first wearable hearing aid was developed in 1936, and by the early 1950s, hearing aids could be worn on the body. See Hochheiser (2013) for more historical details.

In the 1960s through 1980s, at school, deaf children were required to wear body hearing aids upon arrival at school and to return these to the recharging station at the end of the day when going home (Conley, 2009). Body hearing aids involved a plastic case that was strapped to the chest or to the belt, with a cord attaching the case to a miniature speaker system connected to a plastic ear mold that fit in the ear canal (Welling & Ukstins, 2015). Figure 2–7 shows an old body hearing aid used in the late 1970s.

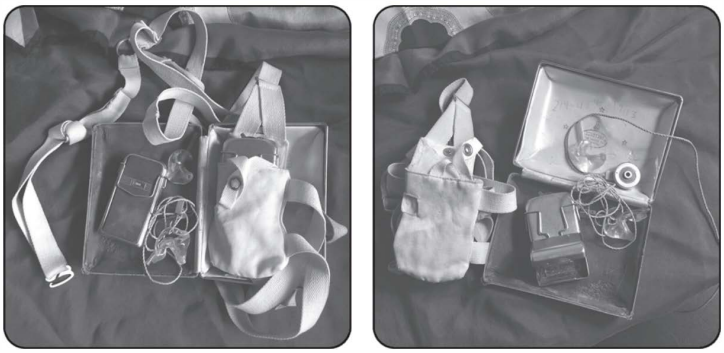

Figure 2–7. A body hearing aid used in the late 1970s. Photos courtesy of Steve Baldwin.

Deaf children were often forced to wear those types of hearing aids, which transmitted sound at very high and often painful levels that did not appropriately match their audiogram needs. Some people became used to it, but there were also many who did not (Sheetz, 2012).

Harris, one of the authors of this book, when in elementary school in the 1980s, was required by her Deaf school to wear a body hearing aid upon arrival for the full school day. Harris explained that she was receiving little or no benefit from the painful amplification of sounds—they just sounded very loud, and she had no idea what the noises were and where they were coming from. She could not concentrate in class while wearing the hearing aid. It was uncomfortable. She would often secretly disconnect or turn off the body aid, and the teachers would discipline her for turning it off once they realized what she was doing. Finally, her mother threatened the school with a lawsuit for forcing Harris to continue wearing the body hearing aid when there was no clear benefit for her. The school complied and allowed Harris to bypass wearing the body hearing aid when arriving at school. This sparked a movement for some other students at the school who felt the same way and removed their body hearing aids as well.

The U.S. Food and Drug Administration (FDA) approved cochlear implant surgery (see below for discussion about cochlear implants) in the United States for adults in 1984, then for young children ages 2 and up in 1990, and, in 2002, for children as young as 12 months old (Knoors & Marshark, 2014). Although the early cochlear implants worked for a number of deaf adults, others who underwent cochlear implant surgery in the 1980s and 1990s continue to share their traumatic stories online and post their videos online in various sites such as DeafVideo.tv or private Facebook groups. In those videos, also called vlogs (more on vlogs in Chapter 9), cochlear implant recipients would often discuss different side effects of the early cochlear

implant technology, such as frequent severe and debilitating headaches and vertigo, in addition to large, visible scars from the surgery, performed when they were much younger, as seen in Figure 2–8.

In their stories, many explain that they did not fully understand why they were having surgery. Some were told that they would become hearing (in some cases, children were given a coloring book that showed them becoming a flying superhero with a cape after cochlear implant surgery) and/or forced to undergo surgery against their will (DeafVideo.tv, 2019). Those traumatic experiences by members of the Deaf community generated an atmosphere of distrust and resistance against new auditory innovations that involve surgery and extensive speech training, taking time away from educational pursuits. Some Deaf people argue that the time spent on speech training and the risk for potential side effects such as an irregular location of the ears on both sides of the face (particularly for those with one implanted ear), vertigo, headaches, and facial paralysis, while very low, are not worth the efforts to hear and speak (Paludneviciene & Harris, 2011).

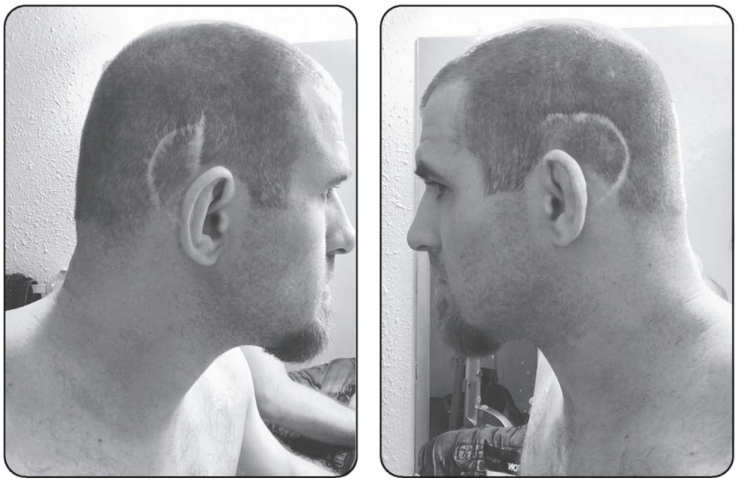

Figure 2–8. Images of a Deaf person with a large C-shaped scar on both sides of his head. Photo courtesy of Garrett Scott.

CURRENT AUDITORY INNOVATIONS AND REHABILITATION

Current auditory innovations are experiencing rapid transformation and major improvements. The medical field is expanding quickly, with fewer side effects, new experiments, updates, and releases. Audiologists, speech therapists, and teachers also work with new technology in identifying hearing differences and in developing hearing and speaking skills. This is called aural rehabilitation. There are new hearing level screening laws and organizations pushing to have hearing differences detected early in life for infants to ensure they have full access to language earlier, rather than later (Early Intervention for Infants and Toddlers, 2020), which is discussed more in detail in the next couple of chapters.

Hearing Level Screening

Some of you might ask, how do caregivers not notice an infant might be deaf until much later? Caretakers and parents may suspect something at first, for instance, when a loud noise during nap-time does not wake up the baby. Then a box is dropped behind the baby and the baby is spooked. The baby looks around, not because they hear the box but because they feel the box being dropped through vibrations that travel through the floor, or maybe feel the sudden movement of the air being pushed toward the baby as the box is being dropped, or maybe the baby sees a shadow of the

box being dropped, or a combination of all those signals. Likewise, when a parent arrives home, the baby looks at the door not because they hear the door opening and closing but because they see the sunlight that comes through the door as it is being opened. The baby looks up when a parent enters the bedroom not because the baby hears footsteps but because the baby smells the parent or even a slight wind blows into the room and alerts the baby. So all those signals, movements, and reactions can easily send confusing signals to caregivers.

Before early infant hearing screening laws in 1990, a child often was not identified as deaf until later in life, approximately age 2-1/2 or 3, when caregivers realized the child wasn't responding to spoken commands or loud noises regularly (Northern & Downs, 2014). Even today, with Universal Newborn Hearing Screening (UNHS) programs, some deaf children pass the screening as they are born hearing but develop progressive losses genetically caused or from diseases, or they may not be identified until they are older because parents do not provide the necessary follow-up. These delays happen more often when children have progressive or conductive hearing loss that might respond to surgery and/or medication (Northern & Downs, 2014).

Today, Universal Newborn Hearing Screenings (UNHS) and related public health programs are part of the Early Hearing Detection and Intervention (EHDI) system that is found in all 50 states and the District of Columbia. The National Center for Hearing Assessment and Management (NCHAM) manages these data.

Parents and caregivers are not the only ones who may miss signals that an infant who passes early hearing screening may be deaf. Most pediatricians (doctors specializing in working with infants and youth), primary care physicians (PCPs), nurses, and hospital technicians miss those signals too. They often have limited exposure to deaf babies. For one, they may have never seen a deaf child because it happens in about two to three babies per 1,000 born (NIDCD, 2016). Approximately 96% of deaf children have parents who hear (Mitchell, 2004) and those parents probably have never had exposure to deaf people or to a sign language.

Not only that, medical and audiology professionals typically do not receive training in issues related to culturally Deaf persons, the impact of early language deprivation, the use of sign language, and hearing loss in general in medical or professional schools (Andrews & Dionne, 2008; Meadow-Orlans, Mertens, & Sass-Lehrer, 2003). However, this is changing. Several major journal publications—*Journal of Clinical Ethics* (Kushalnagar et al., 2010), *Pediatrics* (Mellon et al., 2014), *Harm Reduction Journal* (Humphries et al., 2012), and *Maternal and Child Health Journal* (Hall, 2017)—printed articles discussing the importance of early identification. Not only that, a recent book publication titled *Language Deprivation and Deaf Mental Health*, edited by Glickman and Hall (2019), along with those journal publications are slowly transforming the medical field's perspective. Those publications are written by medical doctors, linguists, and educators. These Deaf scholars and their hearing colleagues explain the importance of early exposure to sign language, especially to ensure that the Deaf infant is not deprived of access to language early in life. The authors caution that if full language access is not provided early, deaf infants are at risk for cognitive, social, and academic delays as they grow older. This is discussed in detail in Chapters 4 and 5.

During early newborn hearing screening, a nurse or technician gives the infant a test using an AABR (automated auditory brainstem response), which works by recording brain activity with the baby's response to sound. If the baby does not register a response during the initial screening in either ear, the baby will be retested. If the same result is given for the second time, an audiologist will see the baby ideally within 3 weeks for a full diagnostic battery of hearing tests. The baby is then referred to an otolaryngologist (ear-nose-throat or ENT doctor) for a medical follow-up. At this

point, the baby receives an otolaryngologist's clearance to see an audiologist (Northern & Downs, 2014). After the parents see the pediatrician, newborn health screener technician, and otolaryngologist, the audiologist is the parents' next professional contact (Andrews & Dionne, 2008).

Deaf involvement in the EDHI system is supported by the Best Practice Guidelines published in the journal *Pediatrics*. Goal 10 states, "Individuals who are D/HH (Deaf and Hard of Hearing) will be active participants in the development and implementation of EHDI systems at the national, state/territory, and local levels; their participation will be an expected and integral component of the EDHI system" (Muse et al., 2013, p. 1337). Is this the case in your state? Check your local and state EHDI organizations for representation of professionals who are Deaf.

Many audiologists graduated from older audiology programs, which often follow the philosophy and recommendations of the Alexander Graham Bell Association for the Deaf and Hard of Hearing, known as AG Bell. They often do not recommend sign language and Deaf culture as an option equal to auditory devices, surgery, and rehabilitation for parents of deaf infants but may do so as a last resort after all other auditory resources have been exhausted. The profit margin for the medical cochlear implant surgery as well as the device itself is high, with the burden of payment falling on the insurance companies. There are ethics involved as to the risks and to the supposed benefits for this expensive operation and device implantation. Given these considerations, some in the Deaf community, and rightly so, claim that audiologists collaborate with medical doctors and the cochlear implant industry in supplying them with patients, generating millions of dollars in profits (Durr, 2011; Ringo, 2013).

Those audiologists often recommend that parents consider Listening and Spoken Language (LSL) programs for their deaf infant (Northern & Downs, 2014). LSL programs often tell parents not to use sign language and may encourage families with deaf children to avoid contact with the Deaf community and Deaf culture. Some audiologists, speech therapists, and medical professionals recommend against the addition of sign language to the deaf child's communication opportunities (Ringo, 2013). Some require parents to sign a contract agreeing to prevent their child from being exposed to sign language (Knoors & Marschark, 2014). Santini (2015) points out that LSL is simply a rebranding of oralism and oral education (discussed further in Chapter 5), which are approaches used to exclude sign language and Deaf culture from a deaf child's life. In deconstructing the mission of LSL, Santini (2015) claims that their program design is actually a mono-modal, limited language education approach focusing solely on training the deaf child to speak and hear.

Emerging new generations of audiologists are more likely to introduce the parents to different types of early childhood programs without excluding or putting the sign language/Deaf culture option last (Andrews & Dionne, 2008). The early childhood programs include ASL/English bilingual and bimodal programs (Nussbaum, Scott, & Simms, 2012) and Total Communication programs (Bodner-Johnson & Sass-Leher, 2003). These programs promote the use of sign language(s) for all children and include the teaching of spoken English skills for deaf children who may have some residual hearing or the use of hearing aids or cochlear implants (see below) and may benefit from spoken language exposure. For children who do not benefit from access to sound, signing is the option that provides full access to language. It is important to note that children who use cochlear implants or hearing aids still do not fully hear spoken languages but may use these devices to support their spoken English development, depending on visual cues such as speechreading and signing (Byrd, Shuman, Kileny, & Kileny, 2011; Marschark, Lang, & Albertini, 2002). Those approaches embrace the multimodal (speaking, signing, and writing), multilingual (ASL, English and other sign, spoken and written languages) forms of education, as opposed to LSL, which often excludes the multimodal, multilingual approach, specifically focusing on listening, speaking, writing, and the learning of one language, English (Ringo, 2013; Santini, 2015), again discussed in detail in Chapter 5.

An increasing number of professionals support the concept of providing deaf children with opportunities to learn sign language as early as possible as the safest route to follow so the child will not suffer from language deprivation (Humphries, Kushalnagar, Mathur, Napoli, Padden, & Rathmann, 2014; Kushalnagar et al., 2010; Mellon et al., 2014). In fact, sign language has been found to support the child's learning of spoken language. In one study of 87 children with severe to profound hearing loss from 48 to 87 months of age, children who were educated in the oral-aural method combined with cochlear implants and who also learned sign language were able to learn language on the same timetable as hearing children (Yoshinaga-Itano, Baca, & Sedey, 2010). Furthermore, there is wide variability and unpredictability in outcomes for auditory devices and spoken language-only approaches (Hall, Hall, & Caselli, 2019). Moreover, being able to speak is not the same as being able to listen to a teacher and understand everything that is being said in a noisy classroom. Neither does it mean the child is progressing in the learning of language. Thus, hearing aids, cochlear implants, and listening and spoken language approaches have limitations that can be remedied by providing full access to sign language. Chapter 5 elaborates on different educational pathways for deaf children.

Hearing Aids

Hearing aids are external devices that come in many forms. The most popular ones come with a mold that is inserted in the ear and connected to a device that fits behind the ear or inside the ear. The microphone, amplifier, and speaker all are all fitted in one small plastic case worn behind the ear, as shown in Figure 2–9 (Marschark & Knoors, 2014).

Some are inserted in the frames of eyeglasses. Some simply fit in the ear canal, are barely visible, and are called in-the-canal or completely-in-the-canal, and some aids are installed inside the middle of the ear (Sheetz, 2012).

Hearing aids are used to simply amplify and channel sound into the inner ear, but lately technological advances have allowed for more sophistication in how the device processes sound for amplification. For example, hearing aid devices now can reduce environmental sounds and focus on amplifying specific types of sounds such as human voices so the listener is not distracted or confused by background sounds. Those are called *digital* hearing aids. Some features include syncing the digital hearing aid with one's smartphone wirelessly using the Bluetooth feature (Sheetz, 2012). The effectiveness of the hearing aid depends on the deaf person's residual hearing—in other words, how much hearing there is, as indicated on the audiogram. If there isn't much hearing left, the hearing aid may not be as useful. Often a profoundly deaf person will turn the amplification much higher, and this at times may cause squealing, whistling, and severe distorting of sound, rendering the sound unintelligible if the earmold is not tightly fitted into the ear (Lane, Hoffmeister, & Bahan, 1996; Welling & Ukstins, 2015). However, there are techniques to minimize this problem.

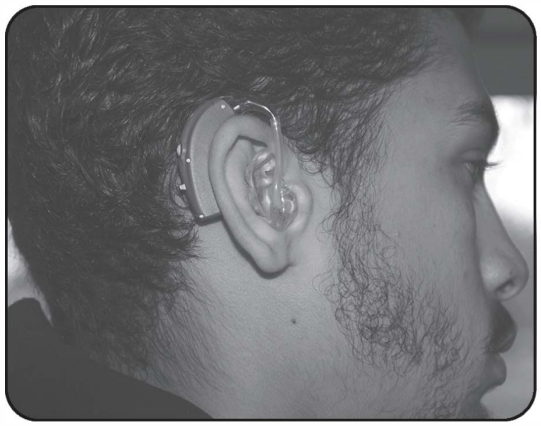

Figure 2–9. Image of a behind-the-ear hearing aid. Photo courtesy of Dezmond Moore.

Many members of the Deaf community wear hearing aids, which provide different types of benefits. Some individuals wearing hearing aids gain access only to environmental sounds such as sirens and someone knocking on the door. Some deaf people gain partial or full access to spoken language in specific scenarios such as a quiet room free of other noise and speaking with only one person. Some are able to manage a noisier environment with multiple people speaking (Knoors & Marschark, 2014). Some individuals simply wear hearing aids to listen to music and its rhythm and beats. Some deaf people wear their hearing aids with their hearing family members only and remove them for their daily routines. Some wear their hearing aids at work only, when communicating with hearing people. Bottom line, decisions to purchase and when to wear hearing aids greatly vary among individuals in the Deaf community. Customized digital hearing aids can range from $1,000 to $6,000 each and are often not included under most health plans (Sheetz, 2012). Sometimes people contact their local vocational rehabilitation services to help defray some of the hearing aid costs (Knoors & Marschark, 2014).

Cochlear Implants

For people who hear, sound travels through the ear and then finally arrives at the auditory nerve, which is connected to the inner ear. The auditory nerve then transmits the sound, now converted into electrical impulses, to the brain. The job of the brain is to interpret what you've heard. For people who have sensorineural hearing loss, the cochlea in the inner ear responsible for converting sound to electrical impulses is *not* working, so that when sound travels through the ear, the sound never arrives at the auditory nerve to be transmitted to the brain (Sheetz, 2012).

The way cochlear implants work is that there is an internal part (coil) that is surgically implanted in the cochlea (inside the inner ear) and directly attached to the auditory nerve. This implant has electrodes that allow external sounds to skip the cochlea that is not working and be converted into electrical impulses that can travel through the auditory nerve, which then sends signals to the brain—much like how people hear. In other words, the cochlear implant connects external sounds with the auditory nerve through the device that lies behind the ear. Cochlear implants do not amplify sound—instead, the sounds are transmitted directly to the auditory nerve. This device is attached to a magnet that is inserted behind the skin on the skull. The skull is slightly drilled in order to make a depression the size and depth of a quarter to fit a magnet on the side of the head. Then the external hearing aid, along with a magnetic field, is attracted to the magnet embedded under the skin behind the ear. This allows the recipient to take off or put on the device easily. Some people receive an

implant for one ear, and some receive implants for both ears (Knoors & Marschark, 2014). A diagram of an implanted ear with the cochlear implant device can be seen in Figure 2–10.

Unlike hearing aids, cochlear implants also do not depend on the amount of hearing the individual has left. Profoundly deaf people are usually better candidates for cochlear implants as long as their auditory nerve works because sometimes the surgery can wipe out the remaining hearing the person had prior to the surgery. This happens when the coil that goes through the cochlea (which is the size of a pea) is a little too rigid and damages the little hairs in the cochlea. This is why doctors usually recommend that hard-of-hearing people not receive a cochlear implant in both ears but rather in the ear that has the most hearing loss, so the other hard-of-hearing ear can work with the implanted ear (Paludneviciene & Harris, 2011).

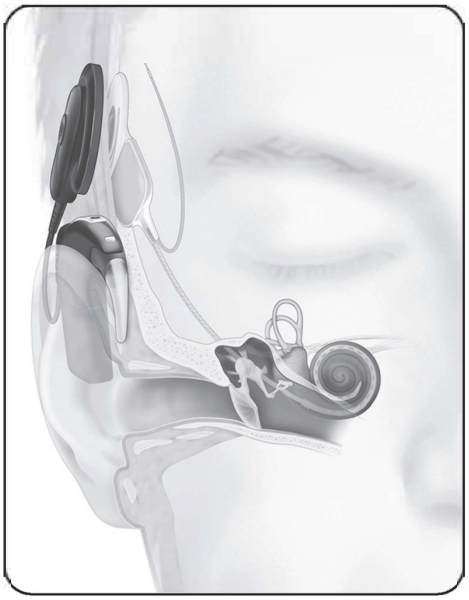

Figure 2–10. A drawing of a cochlear implant device on a human ear. Image courtesy of Cochlear Americas, ©2016.

Among medical professionals, there are a number of eligibility criteria for a successful experience with a cochlear implant. First, the infant needs to be deaf to qualify. The candidate also needs to be able to be scanned using magnetic resonance imaging (MRI). They need to have a functioning auditory nerve for the cochlear implant procedure. They need to be vaccinated against different possible infections, especially spinal meningitis. They need to be physically able to receive an implant and cleared for surgery. The caretakers need to have financial means to cover extra, unexpected costs that their insurance may not cover. The caretakers also need to have schedules that allow for, and have regular access to transportation, for frequent follow-up appointments and care. Not only that, they also need to ensure their infant will be enrolled in an educational program that includes listening and speaking practice opportunities. Of course, having realistic expectations about results and having support of family and friends also are helpful in having a successful experience (Hearing Link, 2012). For deaf adults considering implantation, in addition to the above criteria, it is better if they already have some ability to speak a language and understand a spoken language (e.g., English). If they have some benefit from using digital hearing aids, it is also important to determine whether the candidate will

receive more benefit from having a cochlear implant than their digital hearing aid. The candidate needs to also socialize with hearing, nonsigning people on a regular basis to make the surgery beneficial (Cochlear Implants, 2003; Hearing Link, 2012).

For people who have heard sounds all their lives, their brain has learned to identify and interpret these sounds through repeated auditory exposure. In contrast, those individuals who have been recently implanted need to train their brains to relearn or, in other words, *map* the impulses and the brain's interpretation of the impulses again. These impulses will be different from the sounds they are used to hearing. Adults who have had recent cochlear implantation and had some hearing before the surgery say that the sounds seem mechanical or computerized after surgery. They further commented that it takes time to make connections between the sound and the brain's interpretation of the sound so that they can recognize what the sounds mean (Chorost, 2005). Some children who receive implants pick up sounds in their brains quickly, some don't, and some are in between. Those who have just received the cochlear implant will need to attend regularly scheduled appointments with an audiologist to program the electrical impulses in the speech processor part of the cochlear implant in order to make sure the sound the person hears is at an appropriate loud level and can be interpreted by the brain. This is called "mapping" (Paludneviciene & Harris, 2011).

The American Academy of Otolaryngology-Head and Neck Surgery (2015) reports that the total cost of the cochlear implant, including evaluation, surgery, the device, and rehabilitation, can cost as much as $100,000. Some of the costs may be covered by insurance companies and Medicare, but not in every case. Those numbers do not include all of the costs associated with transportation and time off from work for multiple preoperative, postoperative surgery, and mapping appointments (Boudreault & Gertz, 2016). The devices also have an estimated shelf life of approximately 5 to 10 years, so an infant living well into his or her 70s may have to go in for multiple surgeries to replace, update, or upgrade the device. Likewise, lost or broken devices add to the overall total cost.

Aleki, a Deaf woman, had cochlear implant surgery at age 4. She used her cochlear implant until middle school when her processor broke. Her biological parents could not afford to replace her processor. When she was 17 and in foster care, the Department of Children and Families (DCF) covered the costs for a new processor. Now the current processor is 5 years old and about to die. Aleki set up a GoFundMe page to raise money for a new processor. She says her health insurance is only willing to pay up to $8,000. The total cost is $12,500. She is asking for $4,500 and so far has raised $120 in donations (Aleki, 2015).

The Cochlear Implant Controversy

With the Deaf community's past experience with medical doctors and audiologists, there understandably has been strong doubts and resistance on the part of the Deaf community toward cochlear implant technology that involves invasive surgery. In the 1980s through the early 2000s, there were reports of partial facial paralysis, painful tics caused by electrical stimulation, dizziness and vertigo, and even death as a result of obtaining a cochlear implant. The deaths were mainly caused by anesthesia before going into surgery or due to postoperative infection, particularly meningitis. Currently, patients are required to get vaccinated for meningitis before undergoing cochlear implant surgery, reducing postoperative infection leading to death (Boudreault & Gertz, 2016).

Those individuals receiving cochlear implants in the past have large scars on their head going around their ear, shaped as a big "C." Today, these scars are minor, with improved surgical techniques, and often happen behind the ear. A number of Deaf community members do not fathom putting people through an elective procedure that could potentially have serious or fatal consequences, even though the risk factors

are now lower than before. This sentiment runs even stronger when involving young children due to their inability to fully understand the potential consequences (Boudreault & Gertz, 2016; Paludneviciene & Harris, 2011).

In 1993, the National Association of the Deaf (NAD) published a statement discouraging cochlear implantation in children. But since the number of children undergoing cochlear implantation continued to increase, the NAD revised its position in the year 2000 to encourage access to sign language, especially for children with cochlear implants (NAD, 2000). The Food and Drug Administration (FDA) has since then strengthened its requirements and protocol for cochlear implants, mainly to protect patients from potential harm and death. The FDA has also lowered its recommended age of surgery for children to 12 months of age, considering the procedure to be sufficiently safe (FDA, 2014).

There are many inaccurate concerns about cochlear implants that often impede the ability to open a constructive dialogue about cochlear implants among members of the Deaf community. Many common misconceptions involving cochlear implants are that they prevent children from going swimming, going on rollercoasters, or playing sports. Some say cochlear implant users cannot drive hybrid cars, go scuba diving, or walk through Travel Security Agency (TSA) metal detectors at the airport. All of those are not true. Although cochlear implant devices are water resistant, not all of them are waterproof. The device may need to be removed for showering or swimming (Cochlear, 2015). Roller-coasters, due to their speed and unpredictability, can easily dislodge cochlear implants. Extra precautions will need to be taken with sports, possibly requiring the use of helmets. Cochlear implant users can drive hybrid cars without adverse effects. There is a maximum depth limit for cochlear implant users while scuba diving (FDA, 2014). Although walking through metal detectors is not a big problem, sometimes the magnet may activate the detector alarm, and it is best to for cochlear implant recipients to carry their "Patient Emergency Identification Card" with them at all times (Cochlear, 2015).

Cochlear implant users, like people with pacemakers for their heart, may experience some lifestyle changes after receiving the implant, particularly when it comes to physical contact, water, electronics, and magnets. Boxing and other aggressive sports are discouraged for cochlear implant users.

Although water resistant, the external device cannot be submerged in water (Cochlear, 2015). Cochlear implants sometimes set off or interact awkwardly with theft detection systems, metal detectors, radio transmitters, static electricity, and more. Cochlear implant users will need to communicate with health care workers if MRIs are needed, and possibly in some situations, the magnet may need to be surgically removed (then reinserted afterward) before being scanned by an MRI (Cochlear, 2015). Users have reported some frustration after receiving an implant ranging from inability to upgrade the implant, having implant damage (from impact), unavailability of replacement parts, infection requiring removal, long-term effects, implant failure, skin irritation, dependency on batteries, and dependency on audiologists to assist with programming the settings in the device. Demagnetized implants sometimes need to be surgically replaced (Weiss, 2012). On the lighter side, some cochlear implant users rub their hand through their hair only to find discarded staples or paper clips attached to their scalp, because of the magnet underneath the skin on their head. In any case, even with all those issues, cochlear implant use continues to rise, attesting to the satisfaction some feel with their cochlear implants.

Mario (age 5) and his older brother, Antonio (age 8), both bilateral cochlear implant users, were playing as sword fighters, using sticks in place of actual swords, in their backyard. Antonio's stick accidentally struck Mario behind his ear, around the area where the magnet was located. In the next few weeks, Mario complained to his parents that he was unable to understand most of his classmates and teacher (who speak English) at his school. It was discovered later that the magnet behind his ear broke during the impact. Mario needed surgery to have his magnet

replaced. Unfortunately, due to scheduling issues, Mario was not able to have surgery for another month. Fortunately, Mario and his Deaf family are fluent ASL signers and were able to communicate in ASL in the meantime.

Inspiration Porn

You may have seen (or will see) videos where deaf people or children are seen in an audiologist's office reacting to turning on of their cochlear implant device for the first time. Often those are personal video recordings of actual mapping appointments uploaded by caregivers. Those children who have been recently implanted with the devices are often crying, laughing, or smiling in slight shock. Or you've seen videos or articles of deaf children successfully speaking or hearing a word, a phrase, or a sentence.

Those types of videos tend to be uploaded to YouTube, become viral, and are often published via news outlets. Those are called "inspiration porn," a term coined by Stella Young, a disabled rights activist and writer. She argues that when disabled people are doing ordinary activities, for example, walking, driving, eating, or, in the case of Deaf people, speaking and comprehending spoken words and phrases, those uploaded pictures and videos are objectified by people who can do those things anytime, anywhere. Those people are called "abled people"; in other terms, they're not disabled.

With those uploaded materials, quotes such as, "The only disability in life is a bad attitude," "Your excuse is invalid," and "Before you quit. Try." are usually plastered on those photos or videos or written in articles about them, intended to make readers think, "Well, if that disabled person/child can do it, that means I shouldn't, ever, complain or feel bad about my life" (Pulrang, 2019; Young, 2012). Those pieces essentially make people feel good or better about their lives and feel happy about disabled people "overcoming" obstacles, essentially making those images, words, and videos "inspiration porn" for abled people (Heideman, 2015; Marcus, 2014). Marcus (2014) explains that few people actually look past those emotional moment videos, and the reality is often much more complicated and not "so shiny and perfect" as social media or media play it out to be (p. 1).

And those types of inspiration porn are actually "victim blaming" and "victim shaming" toward disabled and deaf people for not being able to walk, eat, speak, or hear like those "successful" disabled people can. If we can't walk, eat, speak, or hear like the media play them out to be, then "we didn't work hard enough like they did" (Pulrang, 2019; Young, 2012). We didn't go to speech therapy often enough, or we didn't practice hard enough, or we didn't have the right attitude. Inspiration porn is often very exploitative and only provides superficial pleasure and gratification for the reader. More often than not, the disabled person was never fully informed or asked for permission on how their image or video will be used (Pulrang, 2019).

Now that you understand how damaging and exploitative inspiration porn is, look back on your social media and news media activity. Do you remember liking or sharing those types of posts? Do you remember reading them and feeling inspired? Now when you look at those types of posts, can you identify words and phrases that are essentially victim blaming or shaming toward Deaf people who do not hear or speak? What are the typical words and phrases? What will you do now that you know what inspiration porn is about?

Genetic Engineering

Only a few dozen of the estimated 400 genes for deafness (see earlier section on genes) have been characterized, meaning that scientists understand the characteristics of these genes. The size and complexity of these genes make testing difficult. Tests are widely available for a few common forms of genes for deafness. The most widely used test is for connexin 26, which is the name of the protein that the gene *GJB2* produces.

Whether a person is deaf because of connexin 26 depends on the genetic status of the parents. The tendency of Deaf people to marry other Deaf people who communicate using sign language (linguistic homogamy) has resulted in a significant increase in the frequency of children who are deaf due to connexin 26. However, based on the ways in which most genes for deafness (not connexin 26, but rather recessive genes) are transmitted, there is no guarantee that the children will be deaf (Nance, 2004).

There are several purposes for genetic testing. Testing can be used to determine the genetic status of a deaf child or adult (diagnostic testing), carrier testing to find out which relatives may carry genes for deafness, and prenatal testing to determine the genetic status of a fetus. Genetic testing can be used to test embryos within days of egg fertilization with in vitro fertilization in a Petri dish to allow parents to select the desired genetic outcome (Johnston, 2005; Nance, 2003; Rolland & Williams, 2006).

More and more people are participating in DNA testing through different companies such as 23andMe, AncestryDNA, FamilyTreeDNA, and many more. They include connexin 26 testing, as well as Usher syndrome and some other syndromes involving deafness. This type of voluntary testing is controversial because when you participate, you are giving up your and your entire family tree's privacy in order to learn more about yourself and where you came from. Some of the information you learn may make you or others very uncomfortable, such as a surprise sibling, or zero biological connections with people you thought were your relatives, unexpected identification of a sperm or egg donor, and even aid in police investigations of your family members (Baig, 2019). Some people use this type of testing with their partners to decide whether to have a baby (or not), inadvertently altering the futures of those not yet fertilized or unborn babies.

Diagnostic testing in deaf infants or children can be beneficial in terms of knowing genetic influences related to preventing or preparing to deal with complex medical conditions associated with syndromic deafness (see above section). Diagnostic testing for common genes for deafness in infants and children is now considered a standard of care (Pandya & Arnos, 2006). It is natural for adults to be curious about causes, and some seek diagnostic testing to understand this along with their chances of having deaf or hearing children. Others will just let nature follow its course and wait to see their babies (Arnos, 2002). Most Deaf people are resistant to genetic testing, believing it may do more harm than good (see below) (Middleton, Hewison, & Mueller, 1998; Taneja, Pandya, Foley, Nicely, & Arnos, 2004). Hearing people are more likely to consider prenatal diagnosis for genetic deafness compared to deaf people (e.g., Martinez, Linden, Schimmenti, & Palmer, 2003; Middleton, 2004). Hearing and deaf people tend to think differently, with hearing people seeing deafness as a medical issue to be prevented or cured, while culturally Deaf people feel that "deaf" is not a medical problem but a proud identity and culture (Lane, 2005; Middleton, Emery, & Turner, 2010; Scully & Burke, 2019).

What happens when "deafness is cured"? *Zoom Focus: The End* is an award-winning movie produced by British Sign Language Broadcasting Trust (BSLBT), which commissions television programs made in British Sign Language by Deaf people for Deaf people. This movie has won over nine awards since being released in 2011. Watch and discuss among yourselves the movie's exploration of the potential impact of forced eugenics on Deaf people.

Genetic Controversy

There are social and psychological implications related to knowing more about genetic inheritance and choices about human characteristics. More and more people are thinking about this and about potential partners due to advances in genetic

technology that make it possible to, for example, choose partners based on genetic makeup or to choose the sex of the child.

People will make genetic decisions depending on their cultural and/or religious perceptions and life experiences. However, more and more people will become aware of what is possible related to manipulating the genes of their future children. Much will depend on their level of comfort in choosing to go with nature as opposed to making specific reproduction choices. This knowledge, however, means that people may be passing judgment on the value of certain kinds of human lives.

The process of prenatal testing creates the opportunity to decide how acceptable it is to have babies with disabilities, babies who will develop into individuals with their own unique identities. If one accepts prenatal testing to assess chances for having a child with a disability, this challenges the typical perspective that people with disabilities, including deaf persons who see themselves as culturally Deaf, are entitled to being born just like anyone else as well as being treated by society as equal to those who are hearing (Asch, 2001; Burke, 2006; Sandel, 2007).

But when people decide on reproductive choices that are not common, others may become upset. How so? To increase the chances of having a deaf baby, a Deaf couple visited a sperm bank and were informed that potential donors were eliminated if there was a possibility the child could be deaf (Mundy, 2002). How do you think they felt? What does this say about having a deaf child? In any case, they went ahead and asked a Deaf friend (with Deaf genes) to be the donor to increase their chances of having a Deaf baby. Deaf friends asking Deaf friends with Deaf genes to be donors does happen, but we have no way of actually knowing how many babies were born deaf this way. In 2006, a survey of clinics found that 3% reported intentionally using a screening tool to select an egg with a marker for disability based on parental decision (Wordsworth, 2015).

Have you ever looked at your loved ones and appreciated the color of their eyes, height, lip shape, intelligence, or athletic ability? Research shows that people who share the same ethnic cultural background (ethnic homogamy) and/or same language background (linguistic homogamy) tend to marry each other (Stevens & Schoen, 1988). By screening our potential partners based on their cultural, linguistic, and genetic traits, are we practicing a form of genetic engineering as we select our partners? What about choosing the sex of your child through genetic selection? Would you want to be able to choose to have a hearing or deaf child?

What was society's reaction? Public opinion ranged from supportive to fiercely oppositional. Clearly, there are a lot of people who think it is an unfair burden to purposefully have a deaf child. In yet another case in Australia, during in vitro fertilization, a couple was allowed to discard embryos carrying the connexin 26 gene mutation because these were viewed as defective (Noble, 2003). Not only that, in the United Kingdom, fertility legislation enacted in 2008 required that embryo selection must be based on the grounds of avoiding disease (Emery, Middleton, & Turner, 2010). From the perspective of British legislators supporting this effort, genes such as the connexin 26 gene mutation can easily fall into the disease category, and those who would prefer deaf children are not allowed to select embryos carrying the connexin 26 mutation. Deaf people have protested this restriction. In Russia, biologist Denis Rebrikov has started editing eggs donated by hearing women to inseminate five deaf couples so that they will give birth to hearing children. He is also working on creating gene-edited babies who are resistant to HIV (Cyranoski, 2019).

CRISPR, a gene editing kit, is available for people to purchase and use. You can order it on Amazon. It's revolutionary, cheap, effective, and easy to use. Anyone can become a genetic scientist by using that kit. Actually, professional scientists are divided on this issue—some feel this will encourage meaningful discoveries and engagement with science. Some feel this is a gateway for biohackers to accidentally create dangerous pathogens (Sneed, 2017).

Just think about the moral and ethical issues. Is it moral or ethical to discard embryos just because of the possibility of having a deaf child? What does this say about society's view of disability and of deaf people? Culturally Deaf people see themselves as normal and resent this perspective of society (e.g., Bova, 2008). Is society's attempt to control the number of deaf babies a form of eugenics? Eugenics is a philosophy that aims to improve the human race through different strategies, including selective breeding, forced segregation, forced sterilization, laws preventing marriage (and procreation) between less desirable people, and mass murder. Eugenics was popular in the late 1800s and throughout the early 1900s until Nazi Germany used this philosophy during the World War II years to murder people who were considered undesirable (Friedlander, 2002). Today's society can see the advances in genetic technology as either a medical triumph or as an example of cultural genocide (Nance, 2003).

Harris, one of the authors of this book, is Deaf because of a connexin 26 mutation, just like her father and sister. If legislation requires discarding of embryos carrying the connexin 26 gene, Harris, as well as her sister and father, would not exist. Scientists are fascinated with the connexin 26 mutation because they come with faster wound healing among other skin-related advantages. If we eradicate the connexin 26 mutation, what will happen to our ability to heal wounds? Because of the super skin-healing powers of connexin 26, she feels connexin 26 recipients should be honorary members of the X-men mutants. But let's ask the hard questions: Does removing the "bad" also come with removing the "good"? Who determines the "good" and the "bad" when it comes to genes? Why is being deaf bad if we have robust, proud Deaf communities all over the world? How do we measure goodness and badness when it comes to genes?

Do Deaf people and Deaf communities now face cultural and linguistic genocide? Is this moral or ethical? The reality is that even if genetic testing reduces the number of deaf babies, it is still expensive and not available in most parts of the world. Also, many families are not aware of their genetic heritages, which means that the possibilities of having deaf children continue, but for how long? Let's think about why some Deaf people do not want to be Deaf. Or why some Deaf people do not want to have Deaf children. Or why people in general do not want Deaf people to exist. Now imagine if the societies of the world were all accessible, welcoming, and equal—do you think those people would think differently? Imagine if you felt welcomed and embraced for you as you are. Would you want to change yourself? Would you want to change the futures of your children? Would you want to change other people? Probably not. Why not aim for a more accessible, welcoming, and equitable society?

CONCLUSIONS

Remember the three topics you aren't supposed to discuss with your friends unless you want to get into an argument and possibly lose friends in the process? Those topics are politics, religion, and sex. Well, you can add auditory devices, surgery, rehabilitation, and genetic engineering to the list! Those topics are also difficult to discuss and can result in emotionally charged discussions. It is important for people who are not deaf to approach this topic with an open mind and listen to Deaf people, their experiences, their opinions, and their preferences. Some people have had successful experiences with auditory devices, rehabilitation, and innovations. Some love their cochlear implants, or they have to wait for additional surgery or equipment to be able to use their implants again. Some people have had traumatic experiences with audiologists and speech therapists. Some people are content and are not interested in modifying or changing their hearing levels. Some members of the Deaf community believe that Deaf babies, children, and people do not need to be fixed or cured. Some feel that if our societies were accessible, equitable, and welcoming, none of us would feel the need to fix or cure. Like the Deaf gain perspective briefly

discussed in Chapter 1, Deaf people provide a unique perspective on the world and contribute to a diverse worldview. It is argued that by eliminating disability (rather than creating accessibility for all, as discussed in Chapter 9), we are interfering with the natural variations of life, biodiversity, and ecosystem that could later prove detrimental in ways we have never dreamed of. Cochlear implants, other auditory technology, and genetic advances are also seen as a significant threat to the well-being of the Deaf community. In response to that, some members of the Deaf community are trying to reach out to all parents of deaf children to educate them about the value of sign language and its benefits for all children.

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