CHAPTER 31

# Individuals with Multiple Disabilities

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## INTRODUCTION

Individuals with hearing loss and additional disabilities represent a widely diverse and complex group. They differ in the type and degree of their hearing loss, the type and degree of their accompanying disability, and their overall level of functioning. Approximately 25% to 50% of newborns who are deaf or hard of hearing have additional neurodevelopmental conditions, most often cognitive, behavioral-emotional, and motor problems (Chilosi et al., 2010; Fortnum et al., 2006). Similarly, the Gallaudet Research Institute (GRI, 2011) indicated that approximately 41% of deaf or hard-of-hearing school-age children have additional disabilities. As seen in Table 31.1, the most prevalent of these conditions were intellectual disabilities, followed by learning disabilities and vision deficits. It is also possible that some disabilities may not become apparent until well into childhood or adolescence, further increasing these numbers.

There is also some evidence to suggest that the number of people with hearing loss who have additional disabilities is on the rise (Synnes et al., 2012). Several reasons have been suggested to account for this increase including improved survival rates among very low (<1,500 g) and extremely low (<1,000 g) birth weight infants who have a high risk of disability (Cristobal and Oghalai, 2008). Once-extraordinary measures are now routinely used to save preterm infants who, even a decade ago, may not have survived. Most agree that those who do survive the traumas of birth are at higher risk of lifelong disorders than full-term infants (Robertson et al., 2007; Stoinska and Gadzinowski, 2011; Wilson-Costello et al., 2005). However, some studies suggest that the technology and intervention that have improved survival rates have also resulted in improved overall outcomes for premature babies (Jonsdottir et al., 2012; Washburn et al., 2007).

Genetic causes also contribute to the number of individuals with hearing loss and additional disabilities. Approximately one-third of those with multiple handicapping conditions have a syndromic cause of hereditary deafness (Picard, 2004). The most common of these include Down, Usher, Pierre Robin, Treacher Collins, and CHARGE syndromes. In underdeveloped countries where consanguinity is high and genetic forms of hearing loss are more prevalent than in the developed world, education and counseling about inherited forms of hearing loss might lead to a decrease in inheritable hearing loss (Smith et al., 2005). Maternal infection remains a contributing causative factor of hearing loss. Although the prevalence of maternal rubella infection is down worldwide, cases of cytomegalovirus (CMV) are on the rise. CMV is associated with hearing loss and motor and cognitive deficits. Additional risk factors for developmental delays include environmental teratogens (i.e., factors that have adverse effects on embryos or fetuses), maternal substance abuse, and environmental deprivation.

Clearly, the high prevalence of infants and children with hearing loss and additional disabilities serves to emphasize the need for audiologists to acquire knowledge and competence to meet the challenges posed by their complex needs into adulthood. This chapter reviews some of the general characteristics of children and adults with hearing loss and

**TABLE 31.1**

**Percentage of Disabilities that Occur in Children with Hearing Loss**

| Additional Disability | % Children with Hearing Loss |
|---|---|
| No additional disabilities | 61.1 |
| Vision impairment (including deaf-blindness) | 5.5 |
| Intellectual disability | 8.3 |
| Autism | 1.7 |
| Orthopedic disability (including cerebral palsy) | 4.4 |
| Specific learning disability | 8 |
| Attention-deficit disorder/ attention-deficit hyperactivity disorder | 5.4 |
| Emotional disability | 1.8 |
| Other | 14.3 |

Note: Values were taken from Gallaudet Research Institute. (2011) *Regional and National Summary Report of Data from the 2009–2010 Annual Survey of Deaf and Hard of Hearing Children and Youth.* Washington, DC: Gallaudet Research Institute, Gallaudet University.

additional disabilities. Basic principles for assessment and suggestions for management of these special populations are offered. In considering these suggestions, a few points should be kept in mind. First, it is likely that young patients with hearing loss and other disabilities will have some conditions that have not been identified at the time of the audiologic assessment. Therefore, audiologists should be mindful of the possibility that unknown conditions might influence the testing and management of some patients. This is especially true of more subtle conditions such as attention deficits and emotional problems. Second, the combined effects of some conditions may confuse or delay a diagnosis of hearing loss. For example, a child with autism and hearing loss might be nonresponsive to sound, in part, because of "tuning out" behavior and, in part, because he or she truly cannot hear some sounds. Third, a lack of training or experience might lead audiologists to think that some individuals with multiple disabilities are untestable by behavioral measures, which could result in a reliance on physiological measures alone. Certainly, physiological measures contribute valuable information about the integrity of the auditory system. However, we should keep in mind that behavioral tests provide an indication of how an individual *uses* his or her hearing, a very important factor when considering management needs. Collectively, age-appropriate behavioral and physiological test methods can result in an accurate assessment of hearing in most individuals with multiple disabilities and will result in an improved ability for audiologists to develop management strategies.

## CUSTOMIZING THE HEARING ASSESSMENT

When evaluating individuals who have multiple disabilities, consideration must be given to any physical or cognitive limitations that could affect the assessment procedures. A thorough case history, review of prior evaluations, and keen observation can often identify the potential obstacles to assessment and may highlight individual strengths or interests that can be used to enhance the evaluation process. Obtaining as much information about the patient before the evaluation can help an audiologist prepare appropriately for the test session. For example, prior developmental testing or the use of developmental checklists will help audiologists determine an individual's ability to participate in behavioral tasks. Checklists are widely available and can be completed by parents or caregivers prior to their arrival at the clinic or while seated in the waiting room prior to the appointment. Likewise, when physical limitations exist (e.g., cerebral palsy (CP) or other gross motor deficits), modifications to any behavioral task requiring a motor response must be considered.

The widespread implementation of electronic medical records affords timely access to current medical histories and test results, thus avoiding repetitive tests and saving audiologists time in formalizing a profile of their patients. This is especially important when working with those who have multiple disabilities as they are likely to be receiving services from a number of professionals, thus providing a source of multidisciplinary information. More health systems today are moving toward an interdisciplinary model of care whereby several disciplines work together during a single consultation, assessment, or management session to provide an integrated plan of care. Interdisciplinary approaches to care can have an advantage over multidisciplinary care in that a patient's time is streamlined and communication among professionals should be enhanced. Another model of care is a transdisciplinary approach whereby representatives of several disciplines work together during the assessment and development of a care plan, but only a few members of the team provide the services. Regardless of the approach taken, communication among providers is of utmost importance when working with those who have multiple disabilities.

An initial observation without the patient's awareness can be helpful in determining typical behavior of the individual. Discretely observing the interactions between the patient and the caregiver in a waiting area can provide insight into the type, amount, and quality of communication or accommodation that may be effective (Dean and Harris, 2003). These initial observations aid in predicting how much cooperation can be expected and thus determining how to proceed with the assessment. For example, pretest observations of physical and cognitive engagement might reveal that an individual will not be able to participate in behavioral testing, and therefore, reliance on physiological measures will be necessary. Whether testing adults or children, individuals with multiple disabilities are more likely than typically developing individuals to require a heavy reliance on physiological measures over behavioral procedures. Observing the patient's behavior when his or her name is called in the waiting room can also provide some useful insight into the individual's level of functioning. Importance of the pretest interview cannot be overemphasized. Parents, care providers, therapists, and anyone who spends significant periods of time with the patient can provide valuable input about home and other environments, cognitive or physical limitations that might affect assessment or management, and potential compliance concerns.

Based on the review of case history information, previous evaluations, and observations of the patient, audiologists can prioritize the tests in the battery so that those likely to yield the most useful information and that are most easily obtained for the patient are conducted first. The order of the tests in the protocol might be quite different than that used with typically developing individuals. Audiologists should be mindful of the distinction between hearing sensitivity and responses of young children or those with developmental disabilities when interpreting the results of a behavioral test. Matkin (1977) coined the term *minimal response level* to describe the level at which a behavioral response to sound occurs, but also while recognizing that it might be elevated

as a result of nonsensory factors such as attention, motivation, or behavior.

## CUSTOMIZING TECHNOLOGY MANAGEMENT

There is ample evidence to suggest that children with hearing loss and additional disabilities are likely to be fit with hearing technology (e.g., hearing aids, cochlear implants) later than otherwise typically developing children (Kaga et al., 2007; Oghalai et al., 2012). It is also reasonable to assume that adults with multiple disabilities receive hearing technology at a lower rate than adults with hearing loss who have no additional disabilities. There can be several explanations for this delay or lack of intervention including delayed confirmation of precise hearing levels, family/caretaker priorities on other health concerns, or concerns regarding one's ability to secure, care for, and safely wear technology. One way to assist in individualizing the hearing technology candidacy and selection process is the use of functional auditory assessments.

It is not uncommon when assessing the hearing of some individuals with multiple disabilities to obtain little in the way of formal behavioral test results during an initial visit because of difficulty gaining a necessary level of cooperation. However, even without the patient's cooperation, useful information can be acquired through the use of functional auditory assessment tools. These assessments evaluate listening behaviors in real-world settings—outside the confines of sound-treated booths where most formal audiologic testing takes place. The goal of functional assessments is to tell us not only *what* an individual hears, but more importantly, how the individual *uses* what is heard in everyday situations. In addition, information can be obtained about how listening behavior might change in different settings, under different conditions, or with different speakers. This information can then be used to guide more formal evaluation and management plans for these patients. Typically, this information can be obtained from self-assessment, parent, teacher, or caregiver questionnaires. Although these tools have primarily been designed for use with children, it is reasonable to adapt such questionnaires for information gathering purposes when assessing the needs of individuals of any age who have cognitive or behavioral disorders.

The following sections provide some limited guidance when considering hearing technology options for those with a variety of disabilities. Although expectations for benefit will naturally need to be adjusted relative to expectations of typically developing individuals, there is reason to believe that these patients can obtain significant benefit from various forms of hearing technology for daily living activities and in educational settings (Kaga et al., 2007; Oghalai et al., 2012). Counseling families regarding appropriate expectations for their child or family member receiving hearing technology, especially if receiving a cochlear implant, requires relaying a clear message that improvements in hearing might have little if any impact on nonhearing-related developmental concerns.

## AUTISM SPECTRUM DISORDER

Autism spectrum disorder (ASD) is a developmental disorder characterized by symptoms appearing in early childhood and impairing day-to-day life function. These symptoms include qualitative impairments in social/communication interaction and repetitive and restricted behaviors, according to the *Diagnostic and Statistics Manual of Mental Disorders* (5th ed.) (DSM-5) (American Psychiatric Association, 2013). Under the umbrella of ASD, a patient's symptoms will fall on a continuum, with some showing mild symptoms and others, more severe. A diagnosis under the general diagnostic category of ASD is relatively new. Prior to the publication of DSM-5, there were five ASDs, each of which had a unique diagnosis: classic autism, pervasive developmental disorder (PDD), Asperger's disorder, Rett's syndrome, and childhood disintegrative disorder. With the exception of Rett's syndrome, these disorders are now subsumed into the diagnosis of ASD. Rett's syndrome is now its own entity and is no longer a part of the autism spectrum.

ASD is thought to have an early onset, with symptoms appearing before 24 months of age in most cases (Baghdadli et al., 2003; Ozonoff et al., 2010). Although a definitive diagnosis of autism is not generally made until the age of 3 years or later (Mandell et al., 2005), there are a growing number of reports of stable diagnoses following identification as young as 2 years (Chawarska et al., 2009). Prevalence estimates of ASD have increased steadily over time from reports of 1 to 5 children per 10,000 in the 1970s (Brask, 1972) to reports of 5 to 60 per 10,000 in the 1990s and early 2000s (Bertrand et al., 2001; Yeargin-Allsopp et al., 2003). Current numbers from the Centers for Disease Control and Prevention suggest a prevalence of 114 per 10,000 children (Baio, 2012; Rice, 2009). It remains to be seen whether there has been a true increase in prevalence of ASD over time or the reported changes in prevalence can be explained by changes in diagnostic criteria and increased awareness of the disorder by parents and professionals (Fombonne, 2003; Rutter, 2005). Boys are more likely to be affected with autism than girls, at a ratio of more than 3:1 (Van Bourgondien et al., 1987). About 50% to 70% of children with ASD also have an intellectual disability (LaMalfa et al., 2004).

There is no strong evidence to suggest that individuals with ASD have a greater risk of hearing loss than the general population. However, the presence of unusual sensory responses, including abnormal responses to sound, is considered an associated feature of ASD. For example, individuals with ASD might completely ignore sounds that would result in a reaction from typically developing individuals. Other times, they often appear to be overly sensitive to sound by covering their ears with their hands when loud or unexpected

sounds occur. In addition to these abnormal responses to sound, young children with ASD are known to lag behind on language milestones. Therefore, those with ASD will likely be referred to audiologists for hearing assessments as part of the developmental evaluation to rule out hearing loss as the cause of language delay. On average, behavioral responses to sound of children with ASD who have normal hearing are elevated and less reliable relative to those of typically developing children (Tharpe et al., 2006). Relatively little is known about higher order auditory abilities of individuals with ASD. However, altered temporal processing has been recorded in both adults (Samson et al., 2011) and children with ASD (Groen et al., 2009; Kwakye et al., 2011).

## Special Testing Considerations

Children with ASD who have hearing loss are diagnosed, on average, almost 1 year later than those without hearing loss (Mandell et al., 2005). Therefore, it is reasonable for audiologists to be alert to the general behavioral characteristics of childhood ASD to facilitate referral for evaluation when indicated. Several screening tools are available that can be used by audiologists. These include, among others, the Modified Checklist for Autism in Toddlers (M-CHAT) (Robins et al., 2001) and the Pervasive Developmental Disorder Screening Test II (PDDST-II) (Siegel, 1996).

Understanding the general behavioral characteristics of those with ASD can also be helpful to audiologists as they consider modifications to the traditional test battery. Because the majority of those with ASD exhibit cognitive deficits, behavioral abnormalities, and hypersensitivity to sensory stimulation, audiologists should be prepared to address those issues during the test session. For instance, transitions are often difficult for individuals with ASD. When possible, audiologists should avoid travel from room to room with the patient, taking care to escort the patient to the testing area immediately rather than keeping him or her in the waiting area. Audiologists will want to minimize physical contact with those who have tactile sensitivities. This may require initial testing in sound field, because of the possibility of aversion to the tactile stimulation created by earphone placement. A quick inquiry with the parent or caregiver might alert audiologists to any sensitivity that could affect testing.

Regardless of the chronologic age of the individuals, audiologists will need to use behavioral test procedures that are appropriate for their patient's cognitive level. This may mean that procedures typically used with infants and young children (described in Chapter 24) such as visual reinforcement audiometry (VRA) or play audiometric techniques will be used with older children or even adults. If VRA is used, one should consider minimizing the impact of the reinforcement by turning off the animation (if a lighted, animated toy is used) or using a video reinforcement. Other testing options for patients functioning at a developmental level of 2.5 years or greater are conditioned play audiometry (CPA) and tangible-reinforcement operant conditioning audiometry (TROCA) (Lloyd et al., 1968). Although not commonly seen in audiology clinics, TROCA is often used in pediatric practices that specialize in serving those with multiple disabilities. TROCA requires the patient to press a bar or a button whenever a sound is heard, which is paired with the dispensing of a tangible reinforcement (e.g., small piece of food). TROCA is noted to be particularly effective with children having cognitive or behavioral (e.g., ASD) disorders. A significant number of children with ASD receive other clinical services (e.g., speech therapy). A thorough review of reports from other providers as well as a brief discussion with a caregiver can alert audiologists to reward techniques that work with an individual patient.

Patients with ASD are often resistant to earphones or probes used for individual ear testing. Audiologists can ask the parent or caregiver to practice listening activities with headphones with the patient prior to the appointment. If a patient with ASD will not allow the placement of earphones or probes, audiologists might have to resort to sedated procedures. This is certainly true if one plans to fit hearing aids. Individuals with ASD are known to be difficult to sedate with currently available pediatric sedating agents and are at risk for seizures while under sedation (Mehta et al., 2004). Therefore, consultation with the physician in charge of administering and monitoring the sedation process will need to include notification of the patient's diagnosis of ASD.

## Special Management Considerations

For individuals with ASD, tactile sensitivities, and hearing loss, one can expect some resistance to wearing hearing technology. Therefore, maintaining consistent hearing aid or cochlear implant use might take longer to achieve with these individuals than with typically developing individuals. One technique for introducing amplification is to start by having the parent or caregiver gently massage the patient's ears several times a day until little or no resistance is offered. This may take anywhere from a few days to weeks. From there, one can introduce, to one ear only, a soft earmold without the device connected and build up wear time starting with a few minutes until the patient is willing to wear it for longer periods of time. Once the earmold is tolerated with little resistance, the device can be coupled to the earmold, and eventually, both devices can be introduced. Of course, this process will be slower or faster depending on the degree of tactile sensitivity and resistance offered. Hearing technology will need to be secured to the patient's clothing by use of retention devices designed specifically for that purpose. Such devices will leave the technology secured to the patient's clothing even if they are pulled from the ears. Once the individual becomes accustomed to wearing hearing technology, he or she may no longer need to use retention devices.

Loudness discomfort or hypersensitivity to sound has frequently been documented in children with ASD

(Tharpe et al., 2006). As such, it is essential that audiologists carefully adhere to prescriptive formulae for the selection and verification of hearing aid gain and output characteristics. Because it may be difficult or impossible to measure the patient's comfortable loudness levels, audiologists will often need to use age-appropriate normative targets provided by the prescriptive formulae. It is reasonable for audiologists to consider initially lowering the gain and output levels below those prescribed and gradually raising them as the patient becomes accustomed to the amplified sound. However, gain levels should always make speech audible for the patient.

## PHYSICAL DISABILITIES

Persons who are deaf or hard of hearing should have similar motor development and skills as those with normal hearing unless vestibular function is affected. That is, deafness alone does not affect motor abilities or balance function. In fact, 93% of children with deafness have average to above average motor skills (Lieberman et al., 2004). Environmental factors such as emphasis on physical skills in the school curriculum, opportunities for practice and play, and parenting styles are believed to influence physical development of children with hearing loss. Audiologists should be aware of expected gross motor milestones in typically developing children. If a child with hearing loss is not walking by 15 months of age, a referral for further evaluation by a developmental psychologist or pediatrician is warranted.

Vestibular abnormalities that can result in gross motor problems include cochlear malformations such as Mondini's deformity and cochlear hypoplasia. Other congenital causes of gross motor deficits in children with hearing loss include syndromes such as CHARGE syndrome and Usher syndrome type I (described in a later section) and CP. CP is a disorder of neuromotor function. Approximately 3% of children with hearing loss also have been diagnosed with CP, which is characterized by an inability to control motor function as a result of damage to or an anomaly of the developing brain (GRI, 2011; Roush et al., 2004). This damage interferes with messages from the brain to the body and from the body to the brain. The effects of CP vary widely from individual to individual. There are three primary types of CP:

- Spastic—characterized by high muscle tone (hypertonia) producing stiff and difficult movement
- Athetoid—producing involuntary and uncontrolled movement
- Ataxic—characterized by low muscle tone (hypotonia) producing a disturbed sense of balance, disturbed position in space, and general uncoordinated movement

These three types of CP can coexist in the same individual. CP can also be characterized by the number of limbs affected:

- Quadriplegia—all four limbs are involved
- Diplegia—all four limbs are involved and both legs are more severely affected than the arms
- Hemiplegia—one side of the body is affected and the arm is usually more involved than the leg
- Triplegia—three limbs are involved, usually both arms and a leg
- Monoplegia—only one limb is affected, usually an arm

CP is not a progressive condition. The damage to the brain is a one-time event. However, the effects may change over time. For example, with physical therapy a child's gross and fine motor skills may improve with time. However, the aging process can be harder on bodies with abnormal posture or that have had little exercise, so the effects may result in a gradual decline in motoric ability. It is important to remember that the degree of physical disability experienced by a person with CP is not an indication of his or her level of intelligence.

The brain damage that caused CP may also lead to other conditions such as learning disabilities or developmental delays. Approximately 20% of children with CP will also experience hearing or language problems (Robinson, 1973). The hearing loss is typically sensory/neural in nature. In addition, between 40% and 75% of individuals with CP will also have some degree of vision deficit.

### Special Testing Considerations

Individuals with motor delays may not respond behaviorally to auditory stimuli because their physical disabilities limit their ability to orient to sound (Moore, 1995). However, when testing children, VRA can still provide reliable information even for those with poor head and neck control. Modifications that might need to be made in the test arrangements for VRA include the use of an infant seat to provide additional head support. However, audiologists should ensure that head supports do not block the ears and impede sound field stimuli. If children with motor difficulties cannot make a head-turn response to sound, response modifications can be made. Modifications include alternative responses such as localizing to the sound stimuli with their eyes as opposed to head turns. CPA (see Chapter 24) might also require modifications. Response modifications might need to include options that do not require the use of fine motor skills. Examples of such modifications could include asking a child to drop a ball into a large bucket rather than having the child insert a peg in a pegboard, partial hand raising, or even just a head nod. Additionally, a variety of gross motor responses (e.g., hand motion) can be used to trigger an electronic switch that will, in turn, activate a computer screen programmed for appropriate visual reinforcement.

If the physical disability has a neuromotor component, such as with CP, physiological measures might be affected (Yilmaz et al., 2001). That is, abnormality in measures such as the auditory brainstem response (ABR) may be misinterpreted as indicative of hearing loss when, in fact, the

abnormality is in neurotransmission. Therefore, interpretation of the ABR must be made cautiously and in concert with the entire battery of auditory tests, behavioral and physiological. Sedation may be required when conducting ABR with individuals who have CP in an attempt to relax their head and neck or to reduce extraneous muscle movements, thus reducing myogenic artifact.

## Special Management Considerations

When selecting and fitting hearing technology on someone with physical impairments, there are a number of factors that must be considered, including the types of activities in which the individual participates (e.g., physical therapy) and his or her fine and gross motor ability (e.g., use of a wheelchair with head supports). When fitting children, it is important that the audiologist consider input from the parents and other professionals working with the child when determining amplification options. Children who require amplification for their hearing loss are typically fit with behind-the-ear (BTE) hearing aids. However, use of this type of aid may be inappropriate for children or adults with physical handicaps if they have poor head control (Tharpe et al., 2001). The close proximity of head supports or the person's own shoulders, if the head is leaning to one side, may result in excessive feedback or discomfort from BTEs. Problems with feedback might be reduced by selecting a hearing aid with a feedback cancellation feature, although care must be taken to ensure audibility across the speech spectrum is maintained. Another feature that might be beneficial for those with poor head control is a remote control. This can provide easier manipulation of the controls (e.g., volume control) of the hearing aid by caretakers (Roush et al., 2004).

Body-worn hearing aids and cochlear implant speech processors, although rarely used today, provide another option and would eliminate many of the problems that BTEs pose for patients with poor head control. However, body-worn hearing aids also require special consideration when being used with patients who have physical disabilities. For example, for very young children and for those of any age with oral-motor difficulties, the microphone of the aid may be vulnerable to food and drink. Moreover, clothes may rub on the microphone port, resulting in extraneous noise, and wheelchair harnesses can rub or press against the aid, resulting in discomfort or damage. Although children are not typically fit with in-the-ear (ITE) hearing aids, they may be an appropriate solution for adults or children who spend part of their day in atypical positions or who use a wheelchair with headrests.

## INTELLECTUAL DISABILITY

The term intellectual disability includes impairments of general mental abilities that impact adaptive functioning. Symptoms of intellectual disability first appear during the developmental period and diagnosis requires a comprehensive assessment of intelligence across conceptual, social, and practical domains (American Psychiatric Association, 2013). Adaptive skill areas include:

- Conceptual
  - Language
  - Reading
  - Writing
  - Math
  - Reasoning
  - Knowledge
  - Memory
- Social
  - Empathy
  - Social judgment
  - Interpersonal communication skills
  - Ability to make and retain friendships
- Practical/self-management
  - Personal care
  - Job responsibilities
  - Money management
  - Recreation
  - Organizing school and work tasks

As seen in Table 31.1, almost 10% of children with hearing loss also have intellectual disabilities (GRI, 2011). Those with an intellectual disability are at an increased risk for visual or hearing impairment or both (MacFarland, 2003). Detection and treatment of hearing loss in adults and children with intellectual disabilities is of utmost importance because hearing loss can exaggerate intellectual deficits by impeding the learning process (Roush et al., 2004).

Down syndrome, also referred to as trisomy 21, is the leading cause of hearing loss and intellectual disabilities and occurs in approximately 1 in 700 births in the United States (Parker et al., 2010). Audiologists are very likely to see a large number of children and adults with Down syndrome, a genetic disorder always associated with some degree of cognitive impairment. As individuals with Down syndrome age, there is a decline in intellectual ability. In fact, almost 100% of individuals with Down syndrome over 40 years of age demonstrate degenerative neuropathologic changes consistent with Alzheimer-type dementia (Zigman et al., 1995). Furthermore, some have speculated that the precocious aging of individuals with Down syndrome results in early presbycusis in this population (Dille, 2003). Hearing loss progresses more rapidly in adults with Down syndrome than those with other forms of intellectual disability or adults in the general population. Down syndrome is also frequently associated with conductive hearing loss and, less often, sensory/neural hearing loss. Although the majority of the conductive hearing losses in those with Down syndrome are secondary to middle ear effusion, some are the result of middle ear anomalies, such as ossicular malformations and damage to middle ear structures as a result of chronic

infection. In contrast to the typically developing population, the prevalence of middle ear effusion tends to remain high in individuals with Down syndrome regardless of age. Marcell and Cohen (1992) found that adolescents with Down syndrome have poorer hearing and greater incidence of conductive hearing loss than their peers with intellectual disability, but without Down syndrome. For a comprehensive review of hearing loss associated with Down syndrome, see Porter and Tharpe (2010).

## Special Testing Considerations

Little has been published on hearing assessment of adults with intellectual disability. However, it is well documented that audiologists must use test techniques that will bridge the difference between the chronologic and developmental age of individuals with cognitive disabilities to obtain valid test results (Diefendorf, 2003; Roush et al., 2004). The patient's mental or developmental age, not their chronologic age, should be considered when selecting appropriate test procedures and materials. Several investigators have evaluated the effectiveness of VRA with children having intellectual disabilities, including those with Down syndrome (Greenberg et al., 1978; Thompson et al., 1979). With typically developing children and those with intellectual disabilities, VRA is effective with infants as young as 6 months cognitive developmental age. However, children with Down syndrome require a cognitive developmental age of 10 to 12 months to successfully participate in a VRA procedure. Furthermore, behavioral thresholds of infants with Down syndrome have been found to be 10 to 25 dB poorer than those of typically developing infants when all had normal hearing verified via ABR (Werner et al., 1996). This elevation of behavioral thresholds is presumed to be the result of more inattentive behavior on the part of the children with Down syndrome relative to their typically developing peers. Moreover, this inattentive behavior provides additional reason to utilize a test battery that includes physiological measures when testing children with Down syndrome.

Although it is recommended that audiologists attempt to elicit a spontaneous head-turn response during the VRA conditioning process (Tharpe and Ashmead, 1993), some children with intellectual disability may not have developed auditory localization ability. Recall that auditory localization is a higher order skill than detection, the required skill for VRA. In such cases, several administrations of paired conditioning trials (pairing the stimulus and the reinforcer) may be required. If the patient does not respond to the auditory stimuli, the audiologist may be left with the question, "Does the patient not hear the stimuli, or can she or he not perform the task?" One method that can answer this question is for the audiologist to place the bone vibrator either in the patient's hand or on the head and, using a low-frequency stimulus at approximately 50 to 60 dB hearing level (HL), determine if the patient can perform the task using this vibrotactile cue. In this way, the patient is able to feel the stimulus and, thus, is not required to hear to participate. If the patient is able to cooperate for the task under these vibrotactile conditions, then the audiologist should return to the auditory stimuli and continue testing with the knowledge that the patient understands the task.

If using a play audiometric technique, it is often appropriate for the audiologist to demonstrate the play task to the patient with intellectual disability rather than attempting to explain the instructions verbally. Because learning the desired response behaviors may take longer for children and adults with intellectual disability, it may be useful to have them practice the listening task at home before coming to the clinic. It is important to keep the task as similar as possible to what actually will be expected in the clinical setting. Another approach is for the audiologist to demonstrate the task engaging the patient's parent or caregiver as the one being tested. The patient can then observe the procedure being conducted and see what is required. If the patient has use of some language, the audiologist should keep verbal instructions short, simple, and accompanied by gestures. Nonverbal expressions of reinforcement can be used generously (e.g., smiles, clapping, thumbs up) to indicate to the patient that he or she is complying with the task. Audiologists should keep in mind that the reinforcement is provided to support the response behavior of the patient, not to indicate if the patient is correct or incorrect (i.e., can hear or not hear the stimulus). Additional time will likely be needed to complete the play task, and the audiologist should expect response delays as a result of additional time needed for the patient to process the instructions and formulate a response. It is not unusual for patients with intellectual disability to have to return for more than one visit to complete testing. However, the visits should not be so far apart in time as to result in a significant delay in diagnosis. It is important in these cases to keep the examiner and the test procedures the same so that a routine can be established with the patient. This differs from testing with typically developing children where the examiner often has to change the task to keep the child's attention.

Whether using VRA, CPA, or conventional test procedures, it is recommended that control trials (no sound trials) be included throughout the testing session. This is especially true if working with individuals who have Down syndrome, because they typically are eager to please others and this often results in a high number of false-positive responses. Control trials are inserted randomly into the testing procedure at times when the audiologist would otherwise present the auditory signal. If a response is noted during a control trial, it is evidence of a false-positive result and should not be reinforced. This lack of a reward for false responses should reduce their frequency.

Although important for complete evaluation of all patients, it is particularly important to monitor the middle ear status of those with intellectual disabilities, because they

are known to have a higher degree of abnormal tympanometry and conductive hearing loss than the general population (May and Kennedy, 2010). Those with Down syndrome have an even higher incidence of otitis media than others with intellectual disability, because of the anatomic anomalies of the head and neck including the cochlea, ossicles, Eustachian tube, and nasopharynx. Chronic ear infections afflict approximately 70% of children with Down syndrome (Mitchell et al., 2003). In addition, those with Down syndrome are highly susceptible to impacted cerumen, because of narrow or stenotic external ear canals. Therefore, all hearing test procedures (e.g., ABR, VRA, play or conventional audiometry) should include the use of bone-conduction testing when possible. A conductive component can mask the presence of sensory hearing loss, thus delaying the fitting of amplification.

There will likely be a heavy reliance on physiological measures during the hearing assessment of patients with intellectual disabilities. One should be mindful of the impact of abnormal middle ear function on otoacoustic emissions (OAE) and ABR. That is, OAEs will be absent in the presence of impacted cerumen or middle ear effusion. Therefore, immittance audiometry will be an important component of the test battery. In a review of ABR studies in persons with Down syndrome, Dille (2003) concluded that ABR testing should be interpreted with caution, because it is likely that those with Down syndrome demonstrate a neural developmental time course that is uniquely different than that of typically developing individuals. Thus, comparing latency-intensity functions to normative values might result in erroneous conclusions. Widen et al. (1987) suggested that the ABR interpretation be based on both threshold of the response and latency-intensity series.

## Special Management Considerations

Because of the high incidence of middle ear disease in those with intellectual disability, especially those who are institutionalized or have Down syndrome, it is most efficient to have otologic examinations immediately prior to audiologic assessments. The otologic examinations can serve to ensure that the external canals are free of cerumen and that no active middle ear infection is present. Individuals with Down syndrome, regardless of age, should receive otologic and audiologic monitoring about every 3 months to manage cerumen and middle ear disease. By school age, between 45% and 93% of children with Down syndrome have had pressure-equalizing (PE) tubes (Mitchell et al., 2003; Shott et al., 2001). However, diligent audiologic and otologic monitoring is required because of the high failure and complication rates of PE tubes in those with Down syndrome (Iino et al., 1999).

For those requiring amplification, several issues must be considered. First, the implementation of prescriptive amplification fitting is recommended for all children and adults. Individual or age-appropriate ear acoustics should be taken into account in the hearing aid selection and fitting process. This is accomplished by measurement and application of the real-ear-to-coupler difference (RECD) (see Chapter 40). It is not uncommon for audiologists to use age-average RECD values as opposed to measuring them directly. However, one must consider the potential impact that any craniofacial anomaly (including Down syndrome) might have on this practice. Because of the typically smaller ear canals in individuals with Down syndrome, it is quite likely that an age-average RECD will result in an underestimation of ear canal sound pressure level, thus leading to overamplification.

Second, individuals with craniofacial anomalies or who have intellectual disabilities may have difficulty keeping hearing aids in place for a number of reasons. The use of wig tape or other hearing aid retention devices can help them stay in place behind the patient's ears.

Third, bone-conduction hearing aids may need to be considered for patients with chronic or recurrent middle ear disease or stenotic canals. Bone-anchored hearing aids have been used successfully in some children with Down syndrome (e.g., McDermott et al., 2008). In addition, for those with draining ears who use traditional air-conduction hearing aids, aids may need to be removed temporarily during times of active drainage.

Finally, the fitting of amplification may be delayed in individuals with intellectual disabilities because of other healthcare needs and concerns of the family. However, the earlier the amplification is introduced, the easier it may be to incorporate it into the patient's daily routine and the better the prognosis is for long-term acceptance. The parents or caretakers of patients with intellectual disabilities should receive careful and frequent instruction on the use and care of the amplification devices. Of course, to the extent possible, patients should be included in this educational process and encouraged to participate in the care of their devices.

## VISUAL IMPAIRMENT

The combination of vision and hearing deficits may be congenital or acquired later in life. Although often referred to as "deaf-blindness," one should keep in mind that the term "deaf-blind" typically refers to persons with dual sensory impairments who have some residual hearing and usable vision (Miles, 2003). Possible etiologies include syndromes such as:

- CHARGE syndrome—A specific pattern of birth defects represented by the acronym CHARGE: "C" for coloboma, "H" for heart defects, "A" for atresia choanae, "R" for retardation of growth and development, "G" for genitourinary problems, and "E" for ear abnormalities.
- Usher syndrome—The most common condition that involves both hearing and vision problems; an autosomal recessive disorder with primary symptoms that include

hearing loss and progressive retinitis pigmentosa. The vision difficulties include the onset of night blindness, which might become apparent during a child's school years, followed by loss of peripheral vision typically leading to severe low vision or blindness.

- Bardet–Biedl syndrome—A complex disorder that affects many parts of the body including the retina. Individuals with this syndrome have a retinal degeneration similar to retinitis pigmentosa.
- Goldenhar syndrome—A congenital birth defect that involves deformities of the face. Characteristics include a partially formed or totally absent ear (acrotia or anotia) and one missing eye.

Other causative factors for vision and hearing deficits occurring together include congenital prenatal infections (e.g., rubella, toxoplasmosis, herpes, CMV). The rubella epidemic of 1963 to 1965 contributed to the birth of more than 2,500 children with deaf-blindness in the United States. By 2011, there were almost 10,000 children in the United States alone who were considered to be deaf-blind (Teaching Research Institute, 2012). There are also postnatal causes of vision and hearing deficits (e.g., meningitis, asphyxia, stroke). The majority of individuals who are deaf-blind have additional disabilities such as physical impairments, cognitive impairments, and behavior disorders. In fact, more than 60% of individuals who are deaf-blind have intellectual disabilities (National Consortium on Deaf-Blindness, 2007).

Children with hearing loss are two to three times more likely to develop ophthalmic abnormalities than their normal-hearing peers (Guy et al., 2003). The irony is that people with hearing loss have a greater reliance on their vision for communication and environmental monitoring than those with normal hearing. Therefore, audiologists should encourage families of patients with hearing loss to have their vision monitored on a regular basis.

## Special Testing Considerations

One of the first things that an audiologist should determine is the patient's preferred sense (typically, it is tactile), and then the audiologist should let the patient explore the test environment for a short period of time or until the patient appears to be comfortable. In addition to the environment, the patient must be given time to "find the audiologist," rather than the audiologist imposing on the patient's space. It is important to remember that individuals who are deaf-blind may explore their environments tactilely, but many are also tactile-defensive, so they must be approached slowly. As the patient becomes more comfortable in the environment and with the test situation, the rules about space and touching may change (Mascia and Mascia, 2003).

During activities that require the audiologist to touch the patient (e.g., otoscopic examination, insertion of earphones), it is recommended that the patient be given as much involvement as possible. That is, the patient should be allowed to examine the equipment (e.g., otoscope, earphones) tactilely. Then, with the patient's hand still in contact, the otoscope, probe, or earphone can be slowly guided to the patient's ear. This process will require patience by the audiologist and may require more than one visit (Mascia and Mascia, 2003).

Auditory responsiveness of individuals who are deaf-blind may be compromised by their lack of curiosity. Thus, they may not turn toward the source of sound for a VRA procedure. As discussed in the section on individuals with intellectual disabilities, pairing the auditory stimuli with a vibrotactile stimulus may be necessary to condition the patient to the task (Mascia and Mascia, 2003). Once the patient has learned to respond consistently to the paired auditory and tactile stimulation, it can be assumed that the task is understood, and the tactile stimulation can be eliminated.

The selection of an appropriate reinforcement for behavioral tasks is critical. As previously mentioned, most individuals classified as deaf-blind have some residual vision. Therefore, even light perception can allow for successful implementation of visual reinforcement. This may require a slight dimming of the test suite lights to enhance the visual reinforcement for the patient. In some cases, a penlight positioned close to the patient and activated in response to a head turn or searching behavior can be implemented. If visual reinforcement is not possible, some patients may enjoy feeling specific textures, vibration, social praise, juice, food bits, or interesting toys. In any case, it will be important to consult with the patient's caregivers or teachers to assist in determining a desirable reinforcement.

It is also important when behaviorally assessing the hearing of a patient who is deaf-blind to determine an appropriate response to the stimulus. Parents, caregivers, and teachers may all be valuable resources in evaluating what kind of motor response can be expected from the patient in response to sound. Some possible responses include a head turn, reaching, arm raise, finger raise, or leg swing. Additionally, it may often be necessary to physically "show" the patient when and how to perform the response by manipulating the patient's hand, leg, or foot into place when the auditory stimulus is presented. This assistance can gradually be decreased using successive approximations until the child is able to respond with no cueing or assistance from the clinician.

## Special Management Considerations

It is likely that individuals with dual hearing and vision impairments will welcome the use of amplification when indicated. After all, the majority of this population has some degree of residual hearing ability, and enhancement of hearing could serve as an important supplement to less-than-optimal visual input. A survey of clinical audiologists confirmed the belief that those with vision and hearing difficulties could potentially benefit more from amplification

than those with hearing loss alone (Tharpe et al., 2001). In addition, amplification for those with dual impairments has a role beyond that of only enhancing speech perception ability (Wiener and Lawson, 1997). That is, audiologists need to consider more than just enhancing speech perception and must also focus on the role hearing has in orientation and mobility, which is essential to the development of successful independent living skills (Tharpe et al., 2002).

Experts in the rehabilitation of visual impairment use the term "orientation and mobility" to refer to one's location relative to environmental features and moving safely through one's environment. Much research has been conducted on hearing aid specifications designed to enhance speech perception ability, but considerably less research exists on enhancing the detection of environmental auditory cues. It is unknown whether there is a combination of hearing aid characteristics that can be used to enhance speech perception and also improve detection of environmental cues or that can possibly affect one or the other adversely. The need for an integrated approach is apparent for individuals with dual sensory impairments who need to coordinate the aspects of guiding, route instruction, and verbal communication. Even the limited research that has been done on sound localization with hearing aids has not considered the specific spatial hearing needs of persons with visual impairments. Because speech recognition is based mostly on frequencies above 500 Hz, it is common for hearing aids to attenuate frequencies below a cutoff level in the range of 500 to 1,000 Hz. This low-frequency cutoff is designed to reduce background sounds that can interfere with speech perception. However, that frequency range contains critical information for orientation and mobility with respect to traffic sounds (Wiener and Lawson, 1997) and environmental surfaces, such as walls (Ashmead et al., 1998). A third important property of hearing aids is the flexibility to switch between different programs. That is, hearing aids that are programmable can be set to optimize listening in different environments. Assuming that different listening needs require different hearing aid settings for optimal perception, this flexibility will be important to consider in rehabilitation strategies for those with vision and hearing impairments.

Numerous investigators have found that directional microphones provide an advantage when listening to speech in noise under laboratory conditions. However, omnidirectional microphones appear to enhance localization ability under certain laboratory conditions and, perhaps, in real-world settings (Tharpe et al., 2002). A considerable amount of research is still needed to enhance our knowledge in this area. In the meantime, one should be cautious when selecting microphone options for use by individuals with significant vision and hearing deficits. It appears reasonable to offer a switchable directional/omnidirectional microphone option to those with significant visual impairments who must rely on their hearing for getting around their environments safely. Instruction regarding careful head positioning during communication, especially when using a directional microphone, appears warranted.

## SUMMARY

The assessment and management of individuals with multiple disabilities is a great challenge for audiologists. However, with some knowledge of the characteristics of a number of disabilities, early planning for and adjustments to diagnostic procedures, and careful consideration of individual and family needs, one can obtain valid and reliable test results that lead to meaningful audiologic management.

Part of facing this challenge requires recognizing and admitting that no one can be an expert on all disabilities. With these patients, probably more than most, we must acknowledge that our expertise may be limited and that we must work with a multidisciplinary or, optimally, with an interdisciplinary team of professionals, the patient, and the patient's family in developing effective diagnostic and management strategies.

Finally, as with all patients, audiologists must consider the patient's and family's priorities as they relate to the hearing loss. For example, those with multiple disabilities may have other significant medical needs requiring substantial time and emotional energy. As such, the family may choose to defer the management of hearing loss until a time when they can more readily accept the challenge. Audiologists must be respectful of a family's decisions and be prepared to support and encourage families in their choices.

## FOOD FOR THOUGHT

1. You are suspicious that a child you are evaluating in clinic might have ASD. What additional tests/screenings might you conduct in addition to your traditional audiologic testing and what referrals might you make to other professionals?
2. You see that there is a 28-year-old patient with Down syndrome on your schedule for next week. This patient has used hearing aids for a few years. What pre-visit recommendations would you have for this patient's caregiver to prepare for this appointment?
3. You are attending an interdisciplinary team meeting to discuss a 9-year-old child who has hearing loss and significant vision problems that are not correctable with glasses. What are the most critical pieces of information you need from other team members and what is the most critical information for you to share with the others?

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