HEARING LOSS AND CO-OCCURRING DEVELOPMENTAL DISABILITY: RECOMMENDATIONS FOR IMPROVED CLINICAL PRACTICE

Capstone Project

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By
Mandi Grumm, B.A.

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CAPSTONE COMMITTEE:

Gail Whitelaw, Ph.D., Advisor
Christy Goodman, Au.D.
Julie Hazelbaker, Ph.D.
Peter Paul, Ph.D.
ABSTRACT

It is well-known that children with developmental disabilities are at greater risk for hearing loss. Although proper audiologic care is imperative for all children with hearing loss, those diagnosed with multiple disabilities often face unique challenges due to varying degrees of cognitive and physical impairment. For some children with critical health concerns, diagnosing and treating hearing loss may not always be a primary goal for families. Pediatric audiologists must be equipped to support these families by identifying, diagnosing, and providing intervention options for children with developmental disabilities. However, traditional audiologic protocols are not always appropriate for patients with unique needs. The field of special education has a wealth of knowledge and proficiency in working with children with developmental delays. Pediatric audiologists should be flexible in incorporating innovative techniques for diagnosing and treating children with co-occurring hearing loss and developmental disability.

Communication barriers are common in children with developmental delays and common in children with hearing loss, further complicating oral communication development when hearing loss and developmental disability occur together. Interdisciplinary teams consisting of a range of pediatric specialists can support families in their endeavors towards a better quality of life for their children with multiple disabilities. Providing high-quality early intervention services can address individual needs across disciplines through offering appropriate comprehensive evaluations, as well as evidence-based interventions and therapies specific to each child’s needs. Although more research on this topic is desperately needed, pediatric audiologists working collaboratively with family-centered interdisciplinary teams can certainly facilitate successful hearing and communication outcomes for children with multiple disabilities.
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VITA

2011……………………………………………………………… B.A. Speech & Hearing Science
B.A. in Psychology
Minor in Disability Studies
The Ohio State University

2013-2014……………………………………………………………… LEND Audiology Trainee
Nisonger Center
The Ohio State University
Wexner Medical Center

2014-2015……………………………………………………………… Audiology Extern
Little Ears Hearing Center
Louisville, KY

FIELDS OF STUDY

Undergraduate Major Fields: Speech and Hearing Science, Psychology
Undergraduate Minor: Disability Studies

Graduate Major Field: Doctor of Audiology (Au.D.)
Focus on Early Intervention/Pediatric Audiology
Graduate Interdisciplinary Specialization: Disability Studies
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CHAPTER I

INTRODUCTION

Developmental disabilities are chronic conditions occurring during the first twenty-one years of life that limit an individual’s development in physical, learning, or behavioral domains. These conditions include attention-deficit/hyperactivity disorders, autism spectrum disorders, cerebral palsy, fetal alcohol spectrum disorders, hearing loss, intellectual disabilities, language and learning disabilities, muscular dystrophy, and vision impairment (Centers for Disease Control and Prevention [CDC], 2014). Developmental disabilities affect a significant proportion of children worldwide. In the United States, approximately 15% of children aged 3-17 years have at least one developmental disability (Boyle, et al., 2011). This staggering statistic of nearly 10 million children currently living with developmental disabilities in the US requires substantial medical, educational, and social supports. Rates for intellectual disability diagnoses continue to increase considerably each year (Cooper, Melville, & Morrison, 2004). Autism spectrum disorder diagnoses have skyrocketed by 78% in the past decade alone (CDC, 2014). It is difficult to determine whether continuing rises in the prevalence of pediatric developmental disabilities are due to greater awareness and differential diagnostic methods, increases in rates of birth and survival for children with disabilities, or a combination of factors.

People with developmental disabilities have significantly greater healthcare needs than the general population. In addition to needs related to developmental concerns, children with disabilities are more likely to experience medical or neurological co-morbidities (Schieve, et al., 2012). Therefore, children with special needs require significantly greater pediatric and specialist services than their typically developing peers. However, children across the range of developmental disabilities experience statistically disproportionate barriers to receiving
necessary health-related services (Cooper, Melville, & Morrison, 2004; Lennox & Kerr, 1997). Reasons for lack of appropriate care in this population vary, but children with special health care needs in low-income and racial minority groups continue to experience the lowest rates for access to medical and specialty health care (van Dyck, et al., 2004). Poor service acquisition and lack of compliance exacerbate delays, particularly when developmental disabilities co-occur with other conditions.

Childhood hearing loss is one of many categories included in the umbrella term of developmental disability. Approximately 2 to 3 out of every 1000 children in the United States are born with hearing loss (Vohr, 2003). Congenital hearing loss can result from a variety of factors. While genetics contribute to roughly half of congenital hearing loss cases, environmental factors are thought to contribute to approximately a quarter of infant hearing impairments (CDC, 2014). Environmental, acquired hearing loss can result from maternal infection during pregnancy. The most common maternal infections involved in congenital hearing loss include toxoplasmosis, rubella, cytomegalovirus (CMV), and herpes (Smith, et al., 2014). Hearing loss may occur as part of a syndrome with additional identifiable symptoms. However, the majority of mutations causing hearing loss are non-syndromic (CDC, 2014). Congenital hearing loss may also be caused by physical abnormalities of the outer, middle, or inner ear structures.

Some children acquire hearing loss after birth, resultant from illness, ototoxic medications, head injury, or trauma. Children may develop permanent hearing loss as a symptom of viral infection, such as meningitis, or as a side-effect of ototoxic drugs typically used in chemotherapy treatments (Smith, et al., 2014). Temporary hearing losses often occur with cases of otitis media, especially in young children. Childhood hearing loss can fluctuate, remain stable,
or decline progressively over time. For many individuals, the precise cause of their hearing loss is unknown.

All states and US territories have established Early Hearing Detection and Intervention (EHDI) programs to manage surveillance and policies for addressing infant hearing loss. Data from the most recently published national Hearing Screening and Follow-Up Survey indicates that approximately 1.4% of infants who received universal newborn hearing screening were diagnosed with permanent hearing loss (CDC, 2010). However, nearly 15% of children ages 6-19 years have some degree of measurable hearing loss as assessed by clinical audiologic measures (Niskar, et al., 1998). Further research is required in order to more fully define true incidence and prevalence data relating to degrees of hearing impairment, causes, and permanence of hearing loss in the pediatric population.

With continued advancements in medical care, more children with cognitive or intellectual, physical, neurological, and developmental delays are surviving infancy and childhood. Risk factors for childhood hearing loss, outlined by the Joint Committee on Infant Hearing (2007), include pre-natal and post-natal conditions that also pose risks for additional developmental disabilities. For example, infections such as rubella, cytomegalovirus, herpes, toxoplasmosis, meningitis, and measles have been linked not only to hearing loss, but also to higher rates of autism (Szymanski, et al., 2012). Furthermore, neonatal concerns including prematurity and low birth weight increase risk of childhood hearing loss as well as risks for numerous other developmental delays.

Children with hearing loss are considerably more likely to receive an additional diagnosis of disability or delay than their typically developing peers. The Gallaudet Research Institute publishes demographic data for a large national sample of children with hearing impairment.
Recent data suggests that as many as 40% of children with hearing loss experience co-occurring diagnoses (Gallaudet Research Institute [GRI], 2013). This statistic has significant clinical implications for the pediatric audiology patient population. The most common occurrences of coexisting disabilities with hearing loss include intellectual disability, learning disability, developmental delay, ADD/ADHD, orthopedic impairment, blindness/low-vision, speech/language impairment, and autism spectrum disorder (GRI, 2013). Therefore, it is imperative that pediatric audiologists are equipped to provide clinical care for these patients with special needs.

Lack of standardized developmental screenings that have been validated for children with hearing loss often delays diagnosis of additional disabilities (Wiley & Meinzen-Derr, 2013). For example, in children who are deaf or hard of hearing, the identification of autism spectrum disorders occur approximately one year later than diagnoses of autism in children with normal hearing (Mandell, Novak, & Zubritsky, 2005). Currently, clinicians must rely on the existing standardized assessments that were normed for children with normal hearing acuity, in addition to parent reports and clinical observations, to aid in the diagnosis of children with concomitant developmental delays. Precise dual-diagnoses of speech and language disorders can be challenging when hearing loss is present. Hearing loss of any degree can delay the progression of communication skills (Yoshinaga-Itano, Johnson, Carpenter, & Stredler-Brown, 2008). Oftentimes, speech-language pathologists are tasked with determining which delays are likely due to hearing loss and which delays may be due to additional disorders. Preliminary data for the Language Environment Analysis (LENA) device yields promising possibilities for language assessment in children with hearing loss. The automatic vocalization analysis programming within LENA has been useful in identifying apraxia, dysarthria, “deaf” speech, and “auditory
neuropathy” speech in addition to screening speech patterns typical in children with autism spectrum disorders (Yoshinaga-Itano, 2014). Although there is no replacement for comprehensive standardized speech and language evaluations, employing all available assessment tools may assist in narrowing down specific delays for each patient.
IDENTIFICATION/DIAGNOSIS

The unique challenges associated with assessing children with hearing impairment and additional disability begin with detection and diagnosis. Appropriate and timely diagnostic information is necessary to guide parents and professionals in intervention processes. The Joint Committee on Infant Hearing (JCIH) makes specific recommendations for screening, diagnosis, and treatment timelines. Regardless of developmental ability, newborns should receive electrophysiologic hearing screening by one month of age, follow-up comprehensive audiologic and medical evaluations by three months, and appropriate intervention by six months of age (Joint Committee on Infant Hearing [JCIH], 2007). However, several additional factors come into play when other medical or developmental issues occur with hearing loss. For this reason, follow-up is often quite poor in this special patient population (Herer, 2012; Park et al., 2012). Paired with an increase in prevalence for children with disabilities to experience progressive hearing loss, hearing impairment in this population is often overlooked. Although medical and health factors may complicate the priority of hearing loss diagnosis and management, early detection and intervention is crucial for children with developmental disabilities.

Infants born with any of the risk factors outlined by the Joint Committee on Infant Hearing (2007) and those who do not pass their newborn hearing screening should receive a comprehensive audiologic evaluation to diagnose or rule out hearing loss as soon as possible. Audiologic assessment for infants younger than six months of age can be completed with electrophysiologic testing, regardless of developmental ability (Diefendorf, 2003). Information obtained from the case history, immittance testing, otoacoustic emissions (OAEs), and threshold
auditory brainstem response (ABR) testing can provide the audiologist with definitive data regarding the infant’s hearing status.

The case history is an essential audiologic tool in uncovering medical risk factors for hearing loss, risks for inherited hearing loss, and parental concern relating to development of auditory, speech, and language milestones. In addition to milestones related to hearing, audiologists may uncover additional areas of developmental concern that warrant referral for further assessment. Oftentimes, audiologists are one of the first specialists to evaluate children on the autism spectrum, before they are diagnosed with autism (Egelhoff, Whitelaw, & Rabidoux, 2005). Therefore, audiologists working with children should be knowledgeable and familiar not only with indicators of hearing loss and communication delays, but also typical ranges for achievement of developmental milestones in order to provide appropriate referrals when concerns arise. A careful examination of the outer ear and tympanic membrane can alert the audiologist to potential indicators for medical referral such as pinna malformations associated with particular syndromes, impacted cerumen which may impede audiologic testing, or suspected otitis media which may contribute to fluctuating hearing loss and negatively affect the health status of the middle ears.

Immittance testing is another valuable diagnostic tool that does not require active participation of the patient. Tympanograms plot admittance as a function of pressure applied to the ear canal (Shanks & Shelton, 1991). This assessment of middle ear mobility can yield important diagnostic data regarding health status of the middle ears. Additionally, acoustic reflex testing can reveal objective information about a patient’s afferent auditory system, function of the auditory brainstem, integrity of the facial nerve, and functional status of the middle ear (Borg, 1973). Advances in wideband measures of energy reflectance and admittance will likely
provide supplementary data in the infant population (Feeney & Sanford, 2005). However, norms for infants and neonates are currently being developed for wideband reflectance measures (Hunter & Shshnaz, 2014).

Otoacoustic emissions, produced by outer hair cells within the cochlea and reflected back through the tympanic membrane, can be evoked by acoustic stimuli and measured clinically (Kemp, 1978). Information obtained from OAE testing can be used to determine site of lesion for hearing loss and to verify and support other audiologic test findings. This information is particularly useful in the assessment of difficult to test patients, such as young children and those unable to provide reliable behavioral responses.

The auditory brainstem response evaluation is the most important tool in measuring auditory thresholds and integrity of the auditory nervous system in infants and in patients who are unable to reliably participate in behavioral test measures. Valid frequency-specific threshold information can be obtained through comprehensive ABR test measures to estimate behavioral hearing thresholds in infants and children (Siningger, 1993). Auditory steady-state response (ASSR) testing employs a similar test procedure and is useful in determining estimated hearing thresholds for patients who are unable to provide reliable behavioral results. Objective electrophysiologic information can be used for the initial hearing aid fitting and in the development of aural habilitation plans. At least one ABR test is recommended as part of the comprehensive audiologic evaluation for all children under three years of age to confirm degree and configuration of hearing impairment (JCIH, 2007). However, behavioral audiologic testing should be completed as soon as developmentally appropriate for a determination of each patient’s functional hearing abilities.
The combination of electrophysiologic and behavioral measures allows for a more comprehensive picture of each child’s auditory proficiencies. Diefendorf (2003) stated, “behavioral tests provide the critical link between electrophysiologic measures and the child’s daily use of audition.” This cross-check principle should be employed for every newly diagnosed pediatric patient, as soon as they are able to provide reliable responses in the sound booth (Jerger & Hayes, 1976). A comparison of the results for an audiologic test battery should yield data in agreement for each patient. When discrepancies exist, further information may need to be obtained before making diagnostic decisions. The Bayley Scales of Infant Development can be used as an indicator for developmental age when determining aptness of behavioral testing for each individual. These standardized scales estimate mental ability and can determine approximate developmental age for children with cognitive impairment (Boyle, et al., 1994; Diefendorf, 2003).

Visual reinforcement audiometry (VRA) is typically performed for otherwise typically developing children between the ages of six months to two years. Operant conditioning paradigms paired with visual reinforcement can yield valid behavioral detection thresholds in children who are unable to complete standard behavioral audiometry (Diefendorf & Gravel, 1996). The basic premise for this operant conditioning task in the assessment of hearing acuity is the training of the patient to turn his or her head in response to auditory stimuli. Responses are then reinforced with a visual incentive, such as a video, toy, or flashing light. Behavioral audiologic assessment for children with disabilities employs similar tasks as those used for children without additional impairments; however test procedure modifications may be necessary based on individual abilities. For example, infants with motor deficiencies may require more stability during testing. Research conducted by Roush and colleagues (2004) suggests that VRA
results can be improved in this patient population by providing appropriate physical support through more stable infant seats in the testing booth. Specifically, a high chair with fastened tray and straps, or small chair in which the child can place his or her feet on the ground may be helpful in facilitating postural stability. Judging head-turn responses is often complicated in children with disabilities due to involuntary motor movements, hyperactivity, and vision impairment (Diefendorf, et al., 2011). Increasing the number of control trials for each stimulus presentation should be considered if validity and reliability of the patient’s responses are questionable. Limiting distractions in the sound booth is another important modification, especially for children who may become over-stimulated. Children may become uninterested with the task and habituate, or cease responding, after a period of time. When habituation to test stimuli occurs, attention may be improved by switching ears or changing the stimulus frequency (Diefendorf, et al., 2011). Additionally, the speed of testing may need to be slowed to accommodate the central processing abilities of each patient. Although there are many modifications that can be made for children with developmental delays, flexibility of the audiologist is key in determining specific testing protocols for each child.

Conditioned play audiometry (CPA) is suitable for children who are able to respond to stimuli with physical motor movements. Typically developing children three years of age and older can usually be conditioned to complete this task. In CPA, the child is trained to perform a task, such as dropping objects in a box, placing pegs in a hole, or putting rings on a cone each time frequency-specific or speech stimuli is detected. Supplementary instruction and modeling of expected behavior may be necessary for testing children with disabilities using this procedure and added time may be required in order to condition the desired behavioral response for children with developmental disabilities. For a child with visual impairment, tactile cues such as
using the bone oscillator paired with the auditory stimulus may assist in teaching CPA test procedures (Diefendorf, et al., 2011). Modifications may also need to be addressed if the child does not have the fine motor precision or visual coordination to grasp a block and drop it in a bucket or place a peg in a board (Roush, et al., 2004). For instance, a child with fine motor difficulties may consistently touch or point to a block when he or she detects the auditory stimuli and the audiology assistant may throw it in the bucket for them as a motivator to keep their attention. Audiologists should carefully observe the motor abilities of each child in order to construct a feasible behavioral task that can be repeated with each presentation of auditory stimuli. Oftentimes, children with special needs require greater response time due to processing delays. A slightly longer duration stimulus may be used and time between stimulus presentations may be extended in order to provide the patient with ample time to detect and respond to the sound. Children with disabilities may also benefit from greater reinforcement for responses. Fatigue and exertion can quickly lead to challenging behavioral outbursts. Praise and encouragement can go a long way in terms of sustaining attention for the completion of testing. For these reasons, behavioral audiologic assessment strategies may differ in this special population.

Behavioral observation audiometry (BOA) can be used to elicit minimal response levels for children who are developmental unable to participate in traditional VRA or CPA responses. BOA can assess hearing acuity through monitoring unconditioned reflexive and orienting behaviors in response to stimulus presentation. For patients with multiple modality involvement, the audiologist can carefully observe and record sucking, eye-blinking, eye widening, and startle responses following presentation of auditory stimulus (Diefendorf & Gravel, 1996). Enlisting a parent and additional audiologist or assistant may be helpful in determining these responses.
For patients able to complete conventional audiometry, a variety of response options are appropriate, as long as they are consistent and time-locked with the presentation of auditory stimuli. Responses may include hand raising, clapping, head nodding, high-fiving, saying “beep” or “I hear it,” blinking eyes, or pushing a patient-response button (Diefendorf, et al., 2011). This response selection can be patient-driven for children with special needs. For example, if a child waves at the audiologist or makes an exaggerated smile in response to stimuli presentation, this behavior should be reinforced and used in testing.

Comprehensive audiologic evaluations include responses to frequency-specific stimuli, such as pure tones, narrow-band noise, and warble tones in addition to responses to speech stimuli. Therefore, it is imperative that speech testing be included in the test battery to help assess functional auditory profiles for each patient. A speech awareness threshold (SAT) or speech detection threshold (SDT) is defined as the level at which the patient can just detect the presence of a speech signal. Diefendorf and colleagues (2011) suggest allowing a familiar voice, such as a parent or caregiver, to present the speech stimuli while the audiologist monitors the VU meter in cases where attention to recorded speech materials and to the audiologist’s voice is limited. Versatility in determining the speech stimulus presentation, such as calling the child’s name or speaking short phrases that are of interest to the patient should be considered.

Speech reception thresholds (SRTs) are the intensity at which the patient is able to identify familiar spondee words with 50% accuracy. For children with expressive and/or receptive language impairments, modifications in test protocols may need to be addressed. Children may repeat words, point to pictures on a picture board, or point to body parts in response to speech stimuli (Diefendorf, et al., 2011). Children who exhibit echolalia may easily be able to mimic or repeat words back, although they may not necessarily have the language
skills necessary for identifying or comprehending the particular word. Speech reception tasks can be presented as an interactive “game” for children when colorful picture boards or toys are incorporated in the test procedures.

Speech perception testing is a higher-level task that requires receptive vocabulary competency. For children with multiple modality involvement, extra caution should be taken when choosing a specific speech measure (Diefendorf, et al., 2011). Word lists are typically normed for particular age ranges in typically developing participants. Therefore, the developmental level of each patient should be matched carefully to the selected speech perception test. Speech perception testing yields important information for each patient’s audiologic profile. However, due to language, vocabulary, and attention requirements, speech recognition and discrimination tasks are not always feasible for children with more significant speech and language delays.

Although visual coordination and fine motor skills may not be appropriate for certain behavioral tasks, audiologists can build on each child’s individual strengths to engage in conditioned response behaviors to auditory stimuli. Valid audiologic diagnostic appointments for children with disabilities often require extra time, patience, and creativity (Widen & Keener, 2003). Many audiologic procedures, including otoscopy, tympanometry, otoacoustic emissions, and ear-specific behavioral testing may feel overwhelming and stressful for children with sensory issues because probe insertion and transducer placement requires ear manipulation and somewhat intrusive equipment placed into and on the child’s ears. Audiologists must be prepared to handle behaviors that arise during this testing while providing comfort and reassurance during test procedures. Children with developmental disabilities may necessitate frequent breaks during testing. Likewise, a comprehensive audiologic evaluation may require more than one office visit
to complete (Davis & Stiegler, 2005; Cloppert & Williams, 2005). However, obtaining this information is extremely important for determining the child’s hearing status and is essential for the programming and fitting of hearing instruments.

Obtaining test results consistent with normal detection abilities for auditory stimuli does not necessarily indicate normal hearing in all pediatric patients. Parents may report auditory symptoms of inattentiveness or hypersensitivity to sounds, difficulty listening in settings with competing background noise, or inability to localize sounds (Davis & Stiegler, 2005). These indicators should alert the audiologist to a potential need for assessment beyond the peripheral auditory system. The American Academy of Audiology (2010) publishes clinical practice guidelines for the assessment and management of central auditory processing disorders (APD or CAPD). Some children with developmental disabilities and normal peripheral hearing may experience difficulties with competing stimuli, temporal processing, loudness perception, and listening in background noise (Alcantara, Weisblatt, Moore, & Bolton, 2003; Davis & Stiegler, 2005). However, these central processing measures are rarely assessed as part of the traditional audiologic test battery. Depending on specific concerns and related diagnoses, an evaluation for central auditory processing disorder may be indicated.

In addition to identification and initial diagnosis of pediatric hearing loss, the Joint Committee on Infant Hearing provides guidelines for the monitoring of hearing status and communication development. For infants and children with risk factors for progressive hearing loss, such as congenital CMV, syndromes associated with late-onset or progressive hearing loss, and children who have received chemotherapy, hearing should be monitored at least every six months until three years of age, and at regular intervals subsequently, dependent upon the particular risk factor (JCIH, 2007). Following diagnosis of infant hearing loss, audiologists
should see pediatric patients at a minimum of every three months for the first two years of amplification use and every four to six months thereafter (American Speech-Language-Hearing Association [ASHA], 2004). Initially, infants diagnosed with hearing loss will require more frequent audiologic appointments for earmold impressions and fittings due to the rapid rate with which their external ears grow. This also provides the audiologist with opportunities to engage in additional audiologic counseling with parents, check hearing aids, and monitor communication progress. Parent concern is perhaps the most important determining factor in the frequency of audiologic evaluation. Therefore, children should receive audiologic monitoring at any time if caregivers develop concern regarding a change in hearing status or delays in speech-language development.
CHAPTER III

HABILITATION

The benefits of early intervention for children with developmental disabilities have been well researched and documented. Therefore, all infants and children who have received a diagnosis of hearing loss should be considered candidates for early intervention services. Families of every newly-diagnosed child with hearing impairment should receive un-biased information on communication options, including hearing technologies and manual communication routes (JCIH, 2007). Early identification of hearing loss, accompanied by timely interventions, has demonstrated successful communication, language, social, and cognitive skills, consistent with each child’s chronological age and cognitive abilities (Moeller, 2000). In order for early intervention to be successful, recommendations for follow-up and referrals for related assessments should be made immediately following diagnosis of hearing loss.

Given the benefits of early intervention, auditory habilitation should begin as soon as possible if the child meets candidacy criteria. Although communication outcomes may differ significantly depending on each patient’s intellectual capacity, the provision of access to sound can facilitate the best oral communication development. Speech and language communication outcomes may be predicted by developmental age, expressive language ability, degree of hearing loss, mode of communication, and presence of additional disabilities (Yoshinaga-Itano & Sedey, 1999). Speech consists of articulation, voice, and fluency of spoken communication. Speech milestones for typically developing children vary significantly by age, with cooing and babbling expected at 4-6 months and two word utterances that are mostly intelligible for familiar listeners by 24 months of age (Apel & Masterson, 2012). Language consists of the rules delineating word meanings, guidelines for word combinations in the formation of meaningful sentences, and
pragmatic social policies. Developmental language milestones may include gestures such as waving, pointing, and head shaking by 12 months and asking simple who, what, and where questions by 24 months of age (Apel & Masterson, 2012). Progress should be monitored regularly in the early intervention course to ensure that developmental appropriate communication outcomes are achieved for each child.

For families that desire auditory and spoken language communication outcomes for their child with hearing loss, hearing aids are often the starting point in the intervention process. Acoustic amplification strategies are recommended for those with mild to moderately-severe hearing impairment and must be tried before pursuing cochlear implants in patients with severe to profound losses. There is a paucity of research studies conducted specifically on children with co-occurring developmental disabilities who use hearing aids. However, hearing aids have proven beneficial, not only for typically developing children with hearing loss, but also for children with varying degrees of disability (Roush, et al., 2004). Therefore, developmental disabilities should not necessarily be a contraindication for hearing aid interventions, regardless of the cognitive or functional abilities of the child.

The American Academy of Audiology (2013) has developed and updated practice guidelines for pediatric amplification. Audiologists working with special populations should be familiar with these guidelines and should be comfortable in using and modifying protocols to meet the needs of their patients. Pediatric patients with any type and degree of hearing loss, including minimal/mild loss and unilateral hearing loss, should be considered candidates for amplification due to increased risk for delays in communication and academic performance (Yoshinago-Itano, DeConde Johnson, Carpenter, & Stredler Brown, 2008; American Academy of Audiology [AAA], 2013). In children with other developmental concerns, the provision of
early audiologic intervention is especially important in order to limit additional communication and academic difficulties, potentially compounded by the presence of hearing loss. Generally, binaural behind-the-ear (BTE) hearing aids are recommended for infants and children with bilateral sensorineural hearing impairment (AAA, 2013). Following an unsuccessful trial of hearing aids, bone-anchored hearing systems or cochlear implantation should be considered valid options for children meeting audiologic candidacy criteria for these devices.

For children with permanent conductive or mixed hearing loss, for whom traditional hearing aids are not possible due to anatomical malformations or medical middle ear concerns, bone-anchored hearing aids should be considered (AAA, 2013). Typically, bone-anchored devices are worn on a soft headband for infants and young children. Following a trial period, surgical osseointegration can occur for children five years of age and older (Roman, Nicollas, & Triglia, 2011). Bone-anchored hearing systems have also proven successful for children with co-occurring developmental disabilities. McDermott and colleagues (2008) found significant benefit across communication, educational, and health domains for children with Down syndrome using bone-conduction hearing aids. In addition to quantitative communication improvements post-implantation, 100% of parents and caregivers reported subjective enhanced quality of life for these children (McDermott, et al., 2008). Similarly, bone-anchored hearing aid (BAHA) use in children with severe behavioral problems has shown promising benefit in improved health and quality of life for children with more significant developmental delays (Doshi, McDermott, Reid, & Proops, 2010). Participants in the latter study were unable to complete behavioral audiometry measures or a BAHA trial prior to surgical implantation. Therefore audiologists relied exclusively on objective electrophysiologic measures for the programming and fitting of the bone-anchored systems (Doshi, McDermott, Reid, & Proops, 2010). However, outcomes of the
study revealed that all children accepted and consistently wore their devices for eight hours or more per day without incident after a period of habituation.

Bone-anchored hearing aids can also be used in pediatric cases of single-sided deafness. Although current literature presents some mixed results on the effectiveness of BAHA devices for children with profound unilateral hearing loss, these devices have been shown to improve speech-in-noise perception and ease of listening in some pediatric patients with single-sided deafness (McKay, 2010). There are a number of implantable hearing devices currently being researched and used worldwide. Other countries are presently offering these implantable devices for children, although research is severely limited in the pediatric population at this time (Roman, Nicollas, & Triglia, 2011). Currently, research on implantable devices in children with co-occurring developmental disability is non-existent in the literature. In the United State, these options may be on the horizon for special populations, contingent on outcomes in future research studies.

Historically, cognitive disabilities were a contra-indication for cochlear implantation. However, children with various disabilities in addition to profound hearing impairment indicate strong potential for significant improvements in expressive and receptive language. For children with autism spectrum disorder and cochlear implants, increased vocalizations, improvements in social communication, environmental awareness, and improved behavior is consistently reported (Beers, et al., 2014; Donaldson, Heavner, & Zwolan, 2004). These improvements typically occur at a slower rate for children with cochlear implants and additional developmental delays (Waltzman, Scaleunes, & Cohen, 2000; Cruz, et al., 2012). However, children across the wide range of developmental disabilities with severe to profound hearing impairment have demonstrated success as cochlear implant users (Wiley, Jahnke, Meinzen-Derr, & Choo, 2005;
Cruz, et al., 2012; Waltzman, Scalchunes, & Cohen, 2000; Fukuda, et al., 2003). Improved language abilities are a fundamental goal for cochlear implantation in the pediatric population. Language outcomes are dependent on a number of complex factors in children with co-occurring developmental disabilities, most importantly measures of nonverbal cognition for each individual (Meinzen-Derr, Wiley, Grether, & Choo, 2009). Positive subjective benefit, relating to improved environmental awareness of sounds and enriched quality of life, has been consistently reported by parents of implanted children with co-occurring developmental disability, even in the absence of significant quantitative communication improvements post-implantation. In a study that evaluated cochlear implant outcomes for children with CHARGE syndrome, a disease of multiple organ involvement occurring with ear anomalies and associated hearing impairment, 100% of parents reported improved response and reaction capability for their implanted children, even for those who were unable to develop any functional spoken communication skills (Arndt, et al., 2014). The majority of children with additional disabilities consistently wear and use their cochlear implant processors during most or all waking hours (Wiley, Jahnke, Meinzen-Derr, & Choo, 2005). This data implies device tolerance and use for children who willingly wear their processors.

Based on younger ages of implantation and inability to diagnose certain disabilities in infancy, cochlear implants are often implanted in children prior to the manifestation of additional handicaps (Wiley, Jahnke, Meinzen-Derr, & Choo, 2005). Autism spectrum disorders in particular, are typically diagnosed after cochlear implantation has been performed (Meinzen-Derr, et al., 2014). It is difficult to separate language delays caused by hearing loss and subsequent language delays caused by co-occurring disabilities for children with co-morbid diagnoses because both factors likely contribute synergistically to delays. However, the course of
development for auditory communication skills in children with multiple handicaps differs drastically from those of children with hearing impairment as their only disability. For children with multiple handicaps and cochlear implants, the rate of oral communication development is typically slower and often does not reach levels obtained by typically developing implanted children (Donaldson, Heavner, & Zwolan, 2004; Waltzman, Scalchunes, & Cohen, 2000). Nevertheless, this data reveals significant improvements in oral expressive and receptive skills, social interactions, and general connectedness to the environment for this unique population. In addition to quantitative data supporting the use of cochlear implants for children with additional disabilities, families perceive substantial benefits related to the quality of their children’s lives post-implantation (Wiley, Jahnke, Meinzen-Derr, & Choo, 2005). Many of the current objective tools used to measure cochlear implantation progress do not apply to children with additional developmental disabilities. Therefore, it is important to document objective and subjective data for these children in order to appropriately address outcomes. Observations from parents, teachers, and therapists can be helpful tools in evaluating and monitoring progress.

A comparison of the language abilities of children with cochlear implants and intellectual disabilities to their hearing peers with matched nonverbal cognitive abilities demonstrates a significant gap in language achievement (Meinzen-Derr, Wiley, Grether, & Choo, 2011). Research conducted by Waltzman and colleagues (2000) found that while 100% of the implanted children in their study with normal development had exclusively oral communication outcomes, only 59% of the implanted children with additional disabilities used oral only communication. Children with multiple handicaps including hearing loss, exhibit a wide range of communication outcomes on a continuum including behavioral communication, behavior and sign, sign only, sign and some spoken communication, and oral communication outcomes (Wiley, Jahnke,
Meinzen-Derr, & Choo, 20015). For children with co-morbid hearing impairment and autism spectrum disorder, communication outcomes vary significantly, with approximately even distribution between auditory/oral-only communication, augmentative communication modalities, and a combination of communication approaches (Meinzen-Derr, et al., 2014). Similarly, a mixture of varied oral and manual communication outcomes were found for cochlear implanted children with CHARGE syndrome (Arndt, et al., 2014). Therefore it is impossible to predict the precise language outcomes for multiply involved children utilizing audiologic intervention. This evidence supports the necessity for practitioners to be knowledgeable in other communication modalities in order to appropriately counsel families on realistic audiologic expectations and additional options for communication.

Due to difficulties in predicting outcomes for children with heterogeneous handicaps, Trimble and colleagues (2008) suggested incorporating a modified version of the Battelle Developmental Inventory Screening Test to evaluate children with multiple disabilities prior to cochlear implantation. The purpose of this instrument is to screen and evaluate early childhood developmental milestones (Newborg, 2005). The information obtained from the Battelle may be helpful in counseling families and setting realistic expectations for language development after implantation. For instance, children with known intellectual disabilities may experience functional limitations in their ability to develop speech and language skills, commensurate with their degree of intellectual disability. Whether individuals receive acoustic amplification in the form of hearing aids, electrical stimulation through cochlear implants, or a combination electro-acoustic system, auditory access is the foundation for oral expressive and receptive language skills, as supported by the auditory feedback loop. Children with co-occurring hearing impairment and developmental disabilities are certainly able to learn these skills; however, their
communication improvements with auditory access typically occur at lower rates (Cruz, et al., 2012). Successful hearing aid and cochlear implant outcomes depend on numerous factors, one of the most important of which is the degree of disability for each individual.

Frequency modulation (FM) systems are an effective hearing assistive technology that can be used in conjunction with hearing aids, bone-anchored systems, and cochlear implants, or independently for children without hearing impairment. Personal FM systems work via the transmission of a signal from a remote microphone to receivers worn by a listener. Essentially, the child receives the speech signal directly to his or her ear(s), which overcomes some of the challenges associated with distance and listening in environments with background noise. For some children with developmental disabilities that affect their processing skills, personal FM systems can be advantageous, even for children without hearing loss. For instance, in children with autism spectrum disorders (ASD) or attention deficit hyperactivity disorders (ADHD), FM systems have been shown to significantly increase on-task behaviors, improve listening behaviors in noise, and yield better ratings regarding classroom performance from their teachers (Schafer, et. al, 2013). Similarly, children with auditory processing disorders (APD) have demonstrated great success with the use of personal FM systems. Specifically, children with APD exhibit better academic performance, improved speech recognition in noise, and enhanced psychosocial ratings when consistently using an FM system (Johnston, et al., 2009). Overall, personal FM system studies in children with processing or learning disabilities offer positive findings. Audiologists should keep an open mind when considering habilitative options for children with and without hearing loss and provide individualized recommendations for children with developmental disabilities.
A 2013 review of the national Children with Special Health Care Needs (CSHCN) survey indicates troubling data for children with co-occurring hearing loss and developmental disabilities. Approximately 5% of the 40,465 parents of children with special needs reported that their children experienced hearing difficulties (Russ, Kenney, & Kogan, 2013). However, only 1% of these children were found to consistently use their hearing aids. Reasons for these inconsistencies in hearing aid use for this population were unclear. However, increased sensory and developmental co-morbidities reported in the study may have played a role in children’s rejection of their hearing technologies. In order for children to receive necessary benefit from their assistive hearing technologies, the devices must be used consistently to support auditory brain development. Hearing aids, cochlear implants, and personal FM systems provide access, stimulation, and development of auditory neural pathways necessary for spoken communication (Flexer, 2015). Additionally, children learn largely through incidental listening, or over-hearing. When hearing instruments are worn during all waking hours, access to critical language models can occur regularly.

For children with hearing loss occurring with developmental disabilities, parents and caregivers are generally responsible for placing these instruments on the child’s ears and encouraging daily use. For this reason, audiologic counseling for all participants involved in each child’s daily care is essential for compliance (Gabbard & Schryer, 2003). Familiarity with daily listening checks, regular maintenance, cleaning, and trouble-shooting of devices is necessary in the special needs population because it is likely that the child will not have the language to tell a caregiver when their device malfunctions. Fortunately, there are a number of safety strategies and retention devices available for hearing aids and cochlear implants. Although these are often used for small children, children with developmental disabilities of any age can certainly benefit
from these strategies which include tamper-proof battery doors and locks, caps, headbands, clips, cords, and double-sided hypoallergenic tape.

The ultimate goal of every auditory (re)habilitation plan is to improve functional communication abilities for each patient. However, fitting appropriate technology is just a small piece of the intervention process. Pediatric audiologists should provide consistent follow-up services that include objective and subjective outcome measures for every pediatric patient. This is particularly important for patients with co-occurring developmental disabilities because they likely will be unable to provide reliable responses relating to their hearing aid use, performance, and listening challenges.

Evidence-based pediatric hearing aid fitting protocols should be followed to positively support the development of auditory skills for infants and children who wear hearing instruments. Probe-microphone real ear measures are a valid, repeatable, and reliable method of assessing performance of hearing aids (Bagatto, et al., 2005; AAA, 2013). Hearing aid and assistive listening devices should be verified by the audiologist at the initial fitting and at subsequent follow-up visits to provide evidence that speech is audible and over-amplification is not occurring. These procedures include the measurement of real-ear to coupler difference (RECD) as well as on-ear real ear measures (REM). For some children who are unable to sit still or tolerate probe placement, simulated real ear measures (SREM) using age-matched norms can be substituted. In addition to hearing aid verification procedures, FM and remote microphones systems can and should be verified for use on children with normal peripheral hearing via real ear measurements (Schafer, et al., 2014; AAA, 2008).

For all children with hearing loss, and especially for those with additional disabilities, a method for systematically tracking each child’s auditory development and performance over
time is paramount. Clinical validation measures can provide data to support the need for additional hearing technologies or accessories, document the need for environmental/classroom modifications, and demonstrate individual benefit and progress when aided with hearing instruments. A battery of outcome evaluation tools for the evaluation of hearing aid fittings in infants and children is available through the University of Western Ontario Pediatric Audiological Monitoring Protocol (UWO PedAMP). Currently, these assessments are available to audiologists online, free of charge, and offer standardized procedures for recording hearing aid or cochlear implant use, monitoring progress, and tracking of communication outcomes.

Additionally, clinical data is available for the use of the UWO PedAMP protocol in children with comorbidities (Bagatto, et al., 2011). Current data suggests that although children with complex factors score lower on tests of auditory development than age-matched typically developing peers, they do experience improvements in performance with hearing aid use.

Caregiver reports can add important data in measuring and tracking outcomes. The UWO PedAMP protocol consists of four clinical tools that should be administered at specific stages for infant fittings. First, the Infant Hearing Program Amplification Benefit Questionnaire is a quick tool to assess parental and caregiver acceptance of hearing technology, familiarity with device maintenance, and satisfaction with outcomes (Bagatto, et al., 2011). Another central tool in the process is data collected through specific probe-microphone verification measures. The LittlEARS Auditory Questionnaire is administered to gain information regarding receptive and expressive auditory behaviors when aided with hearing instruments (Tsiakpini, et al., 2004). The final tool included in this protocol is the Parent’s Evaluation of Aural/Oral Performance of Children (PEACH). This short questionnaire provides data relating to communication outcomes in different listening environments (Ching & Hill, 2005). Parent and caregiver validation
questionnaires can be especially helpful in assessing progress in the child’s regular listening situations because the audiologist generally interacts with the child in a very controlled environment.

A combination of objective and subjective monitoring techniques provide a multidimensional approach to documenting auditory performance over time. Erber (1982) described a hierarchy of auditory skills comprised of detection, discrimination, identification, and comprehension. Although patients with comorbid developmental disabilities may not be able to participate in more advanced levels of auditory testing, evaluating children across the hierarchy of functional auditory ability can provide useful insight into each patient’s performance ability. The Ling 6 sound test, comprised of /m/, /ah/, /oo/, /ee/, /sh/, and /s/ can be used as a functional detection, discrimination, and identification task for children with and without additional disabilities, depending on individual cognitive and language aptitude. These six speech sounds can provide functional auditory information across the range of human speech (Smiley, Martin, & Lance, 2004). Meaningful insight into auditory performance can be measured relatively easily and quickly across varying listening environments. In addition to assessing unaided and aided performance, Ling stimuli can also be utilized in the assessment of signal-to-noise and figure-ground relationships (Smiley, Martin, & Lance, 2004).

When developmentally able to participate in higher level aided testing measures, audiologists may select from a number of speech perception tests are for the assessment of functional speech and language development. This information is frequently used to monitor progress and modify hearing instrument settings. However, studies for children with co-occurring intellectual disabilities reveal that traditional audiologic measures are not always possible due to developmental level of these children (Donaldson, Heavner, & Zwolan; 2004).

There are several reasons why acoustic and electric auditory inputs may not be appropriate for every child with hearing loss and additional developmental concerns. If manual communication outcomes are preferred by the child’s family, hearing aids or cochlear implants may not be desired. In other instances, children with access to sound may prefer to communicate through manual or signed approaches. Sometimes, the presentation of disabilities hinders successful auditory-oral communication. American Sign Language is a valid option for manual communication and language development. For children with more severe forms of cognitive, intellectual, developmental, or motor-planning disabilities, augmentative alternative communication (AAC) may be the most appropriate communication method (Davis, Barnard-Brak, Dacus, & Pond, 2010; Horn & Kang, 2012). In fact, Davis and colleagues (2010) considers deaf individuals with multiple disabilities “ideal candidates” for AAC systems, given their unique communication barriers. Aided AAC strategies, ranging from low-tech photographs and symbols to high-tech communication boards and electronic speech generating devices, can facilitate effective communication in this unique population. This alternative communication can be used to compensate for limited verbal communication skills by integrating symbols, devices, techniques, and strategies to enhance communication (Schlosser & Sigafoos, 2006). In addition to aided AAC systems, unaided modes of augmentative and alternative communication may include gestures, facial expressions, non-speech vocalizations, and signs. Children with hearing impairment and concomitant disabilities are not limited to choosing only one of these options.
Often, a combination of these communication routes can be used to support effective expressive and receptive communication.
CHAPTER IV

INTERDISCIPLINARY TEAM APPROACH

The primary goal for children with hearing loss is the provision of appropriate interventions early in each child’s development. However, children with hearing loss co-occurring with varying degrees of cognitive and/or physical impairment typically require a number of specialized services. Effective teams require coordination of varying professionals, organizations, and agencies (Nelson, Houston, Hoffman, & Bradham, 2011). Interdisciplinary, multidisciplinary, and transdisciplinary teams are uniquely qualified to support families by providing collaborative assessments and habilitation plans for individuals with multiple disabilities. This wide assortment of professional team involvement should include family members and caregivers, healthcare specialists, and special educators. The provision of high-quality assessments can address individual needs across disciplines by offering appropriate evaluations, as well as evidence-based and therapies specific to meet each child’s needs.

Effective teams include the patient and family members as well as autonomous healthcare providers including a developmental pediatrician, orthopedic surgeon, genetic counselor, physical therapist, occupational therapist, psychiatrist, dentist, dietician, social worker, psychologist, pharmacist, nurse, speech-language pathologist, audiologist, and team coordinator who come together to make team decisions in the best interest of the patient (Patel, Pratt, & Patel, 2008). Child-life specialists, early intervention teachers, and special educators should also be included to assess and provide input related to educational needs and development. However, the specific make-up of each team should be dependent on the particular needs of each patient.

Pediatricians should serve as the medical home, or anchor, in each child’s team. Current position statements for both the American Academy of Pediatrics (2007) and the Joint
Committee on Infant Hearing (2007) emphasize the value of the medical home for regular surveillance and developmental screenings, especially for children with special health care needs. The medical home approach emphasizes service provision that is accessible, family-centered, continuous, coordinated, and comprehensive (Strickland, et al., 2009). Pediatricians serve a number of roles for children with developmental disabilities and delays, including consultant, coordinator of care, and referral source for specialty services. Diefendorf (2003) emphasizes the priority of medical management over audiologic care in children who may require vital surgeries or medical therapies prior to the provision of audiologic services. Although pediatric audiologists interact with children with hearing loss on a daily basis, hearing loss is a low-incidence disability and many physicians are not familiar with specific audiologic protocols. Audiologists should communicate regularly to share information with pediatricians, especially when they share a patient.

With the implementation of universal newborn hearing screenings, audiologists are often one of the first specialists to provide diagnostic evaluation for children with hearing loss. Audiologists are uniquely qualified to assess, identify, diagnose, and provide a range of intervention options for individuals with hearing loss (AAA, 2004; ASHA, 2004). Therefore, pediatric audiologists will remain permanent members of the interdisciplinary team for children with hearing loss and co-occurring developmental delays, providing consistent monitoring of hearing status and aural habilitation planning, as well as services related to hearing instrument maintenance and repair.

Oftentimes, audiologists and speech-language pathologists are the first to recognize symptoms of autism spectrum disorder in young children (Rabidoux, 2005). Parents may initially present to appointments with concerns regarding their child’s hearing status or language
development when more complex diagnoses underlie their child’s auditory attention or communication delays. Therefore, in addition to expertise in assessing each discipline’s specialty populations, clinicians across a wide array of pediatric healthcare specialties should be familiar with symptomatology and knowledgeable of risk factors for developmental disabilities. Likewise, they should be comfortable in making appropriate referrals when indicators outside of their diagnostic expertise arise.

Following confirmed diagnosis of hearing impairment, the child’s parents and pediatrician should be informed of the diagnosis immediately. One of the most critical goals for all identified deaf and hard of hearing infants is timely and coordinated entry into early intervention programs (JCIH, 2007). Pediatric audiologists must be familiar with their state’s birth-to-three programs and provide referrals promptly to assist families in receiving necessary services. Additionally, children who have been identified with hearing loss should receive medical evaluation by an otolaryngologist (JCIH, 2007). The purpose of the otolaryngology assessment is to establish or rule out etiologies for the hearing loss, identify related conditions, and to provide recommendations for surgical intervention, if indicated. Otolaryngologists may order radiologic imaging studies to assess anatomy of auditory structures and recommend genetic counseling to verify the presence or absence of an inherited pathology (Doyle & Ray, 2003). Pediatric otolaryngologists provide a unique medical perspective for children with hearing impairment and their expertise makes them a fundamental member of the interdisciplinary team.

A number of pediatric hearing loss cases occur with vision impairment. Therefore, children identified with hearing impairment should receive a visual acuity assessment by an optometrist experienced in assessing infants (JCIH, 2007). If vision impairment is identified, teachers of the visually impaired may be added to the interdisciplinary team, in order to
incorporate appropriate interventions for blind and visually impaired services and to educate other professionals on adaptive aids utilized for the child with visual impairment.

When working together on a team, flexibility is a core component in obtaining valid and reliable information regarding a child’s hearing and communication status (Rabidoux, 2005). Speech-language pathologists represent an integral component of the interdisciplinary team for children with communication delays. Within the field of speech and language pathology, specialization in auditory verbal therapy (AVT) emphasizes provision of communication therapies for individuals with hearing loss for whom listening and spoken language outcomes are desired. In addition to specific knowledge relating to communication strategies for children with hearing impairment, auditory verbal therapists receive extensive education and training in hearing technologies and effective educational accommodations (AG Bell Academy, 2015). Research on auditory verbal interventions demonstrates significant benefit for children with hearing loss. In fact, speech, language, and reading performance for children with hearing loss in AVT programs have been found to be equivalent to outcomes for age-matched control groups with normal hearing (Dornan, et al., 2010). However, current research studies on AVT do not include children with co-occurring developmental disabilities. Therefore, speech language pathologists and auditory verbal specialists should be prepared to design individualized therapy plans promoting developmentally appropriate communication outcomes for children with more complex needs. Itinerant teachers of deaf/hard of hearing children will regularly join the interdisciplinary team through providing home-based early intervention services. Many children with hearing loss will continue to receive itinerant special education services specific to their hearing loss as they transition into school based services.
Although many professionals on the interdisciplinary team do not have specific training relating to pediatric hearing loss, they assess and monitor other functional areas of the patient’s life. Oftentimes, with supplementary training and communication from audiologists, these professionals can incorporate hearing objectives into their therapies and intervention. For example, an occupational therapist may design therapy plans to reduce behaviors related to sensory defensiveness by extending hearing aid use time in patients that have difficulty wearing their hearing aids. Additionally, OTs may add hearing instrument maintenance, insertion, and removal into their functional activities plans. Social workers may assist families in finding funding resources for hearing instruments and therapists may work with families to ease concerns relating to social stigmas of hearing loss.

For school-aged children, audiologists may provide itinerant services for students and resources for classroom teachers with goals of supervising assistive listening devices and FM use, improving classroom acoustics, and monitoring educational outcomes related to hearing loss. Likewise, teachers of deaf and hard of hearing children are trained in facilitating appropriate educational supports for students and their classroom teachers. A number of evidence-based standards for teachers of deaf and hard of hearing students have been established by the Council for Exceptional Children and Council on the Education of the Deaf (Easterbrooks, 2008). Unfortunately, classroom acoustics often provide poor listening environments for children (Knecht, Nelson, Whitelaw, & Feth, 2002). Therefore a combination of teacher education regarding ways to improve classroom listening conditions and evaluation and monitoring of student performance can provide invaluable data regarding each child’s academic functional abilities. Teacher questionnaires, including the Screening Instrument for Targeting Educational Risk (SIFTER) are available online, free of charge for preschool,
elementary, and secondary students to assess classroom performance for children who use hearing technologies (Anderson, 2015). Establishing educational programs for children with hearing impairment in mainstream educational placements is a complex task. When combined with additional needs relating to co-occurring developmental disabilities in students with hearing loss, educational audiologists, teachers of the deaf and hard of hearing, mainstream classroom teachers, and special educators must work together to provide systematic observation and assessment to ensure that appropriate educational supports are in place.

Although professional team collaboration of trained specialists is crucial in the interdisciplinary process, parents and family members should also be considered fundamental representatives on the team. Parents are usually the primary caretakers for children with developmental disabilities and are tasked with not only bringing their child to medical and therapy appointments, but are also responsible implementing habilitation strategies at home and in functional real-world settings. Parents know and understand their child on a deeper level than their child’s healthcare and educational providers and will be involved through all transitions from early intervention, to educational placements and services, and post-secondary decisions. Parents are typically the primary advocates for access and resources for high-quality care across settings for their child with disabilities. Incorporating a family-centered approach into pediatric practices can allow the patient’s family to be involved in decision processes (Bailey, Raspa, & Fox, 2012). Parents and caregivers must be on board with decisions made by the team because they will be tasked with implementing them in the home environment.

Research on parents of children with developmental disabilities indicates significant levels of chronic negative stress on parents, especially for mothers of children with disabilities (Miodrag & Hodapp, 2010). Despite increased stress, parents of children with developmental
disabilities can also experience heightened resilience when faced with the increased demands of their children with special needs. Peer and Hillman (2014) found several factors that contribute to parental resilience in this population, including coping style, optimism, and social support. In addition to evaluating needs of the child, trained psychologists, social workers, and family support specialists on the interdisciplinary team can assess parental stress and provide resources to support the cultivation of resilience for each family (Peer & Hillman, 2014).

All practitioners can encourage families through the provision of supportive counseling. However, most providers cannot truly empathize with parents of children with multiple disabilities. Fortunately there are many family-to-family support groups that range in size, location, and emphasis of specific disabilities. Social media has certainly contributed to access of these valuable supports for parents. For instance, Hands and Voices, a parent-driven group for families of children with hearing impairment, has chapters across the United States and internationally. Membership includes access to resources regarding parent advocacy, special education laws and resources, and parent mentorship program for parents of newly diagnosed children with hearing impairment referred to as “Guide By Your Side” (Hands and Voices, 2014). Additionally, resources and support groups are available for “Deaf Plus,” a positive family support network for individuals with hearing loss and co-occurring disabilities, and “Deaf-Autism,” a group focusing the specific needs of children with autism and hearing loss (Hands and Voices, 2014). Deaf-blind support groups are also available online through Hands and Voices.

Sibling involvement is often overlooked in the interdisciplinary team. However, siblings are an integral component of the nuclear family and should be included in the team dynamic as appropriate. Research on sibling involvement in the pediatric audiologic clinic is limited.
However, current information suggests that the majority of siblings have positive feelings toward their sibling with cochlear implants and many take on a protective, authoritarian role over their younger siblings with hearing loss (Wiseman, Davis, Kelly, Warner-Czyz, & Loy, 2014). Therefore, audiologists should strive to include siblings in the counseling process as siblings can provide unique perspectives regarding their family dynamics and may prove to be beneficial in aural rehabilitation outcomes.

Incorporating family-centered approaches into pediatric care is best practice for children with disabilities (Bailey, Raspa, & Fox, 2012). Parents and caregivers must be on board with decisions and recommendations made by the team because they will be tasked with implementing them in the home environment. In order to best support these children, interdisciplinary teams should support parents by encouraging and valuing their perspectives and opinions. There are numerous benefits to this method of interdisciplinary coordination for the patient, family, and specialists. These include improved quality of care, comprehensive service provision, and better assessment, diagnostic, and management outcomes. Interdisciplinary teams are uniquely qualified to serve children with complex needs. A comprehensive picture of the whole child is necessary in order to facilitate successful communication outcomes for children with hearing loss and co-occurring developmental delays.
CHAPTER V

SPECIAL EDUCATION

It is essential that all pediatric specialists, including audiologists, provide appropriate services to children with disabilities. Not only is this an ethical principle, but suitable identification, diagnosis, and intervention services for children with hearing loss are mandated by federal law (Kreisman, & John, 2010). In 1973, Congress passed the Rehabilitation Act. This law prohibits discrimination on the basis of disability for any program or agency receiving federal funding. Section 504 of this federal law forbids the exclusion of individuals with disabilities and promotes equal opportunity and access through the provision of reasonable accommodations (Rehabilitation Act, 1973). Shortly afterward, the Education for All Handicapped Children Act was enacted by the United States Congress in 1975. This mandate required all public schools to provide equal educational access to all children, regardless of disability. Many revisions to this law have been made since its inception. Currently, it is referred to as the Individuals with Disabilities Education Improvement Act and is known colloquially as IDEA (Individuals with Disabilities Education Act [IDEA], 2004). Finally, the No Child Left Behind Act seeks to improve educational outcomes for students in the public school system with the goal of closing achievement gaps for disadvantaged students, including those with disabilities, by imposing new requirements for standards, assessments, accountability and parent involvement (No Child Left Behind [NCLB], 2001).

The Individuals with Disabilities Education Act includes service delivery to individuals with hearing impairment and deafness, learning disabilities, autism, visual impairment, speech and language problems, emotional disturbance, cognitive disability, orthopedic impairment,
traumatic brain injury, and other health impairments in the public school system (IDEA, 2004). IDEA is separated into two sections. Part B defines provision of services for children with disabilities ages 3-21 years through Individualized Education Plans (IEPs). Part C defines early intervention services for infants and children from birth through two year of age. Individualized Family Service Plans (IFSPs) outline early intervention care for children receiving services through part C (IDEA, 2004). The number of infants and young children served under IDEA part C in 2011 was 336,895 (US Department of Education, 2014). This number represents 2.8 percent of the birth through age two population in the United States. In 2011, 745,954 preschool children three to five years of age were served under IDEA part B, representing 3.9 percent of their age-matched peers. Elementary and secondary students ranging in age from 6-21 years old included 5,789,884 individuals served through part B (US Department of Education, 2014). This number represents 8.4 percent of students in the United States.

Six key principles for educating students with disabilities are described in the Individuals with Disabilities Education Act. First, zero reject forbids schools from excluding students from a free and appropriate public education (IDEA, 2004). The zero reject philosophy contends that no child can be rejected or denied an education, regardless of the severity of their disability. Second, fair and nondiscriminatory evaluations must be conducted in order to determine the extent of the disability and appropriate services (IDEA, 2004). Third, specially designed education is mandated through the development of an Individualized Education Plan (IEP) for each student (IDEA, 2004). Fourth, the least restrictive environment principle requires schools to provide instruction in the best setting for that child while emphasizing inclusion with non-disabled peers as appropriate (IDEA, 2004). Fifth, procedural due process allows students and their families to request an unbiased hearing when disagreements occur (IDEA, 2004). Finally, the principle of
parent and student participation encourages involvement in the decision making processes for special education services (IDEA, 2004). The most recent revision of IDEA aligns closely with NCLB. The No Child Left Behind Act (2001) addresses inclusion of these students in mandated accountability testing.

Any child with hearing loss or a combination of hearing loss and co-occurring hearing loss and developmental disability warrants appropriate hearing healthcare and educational audiologic services. Children with hearing impairment routinely require accommodations and supports in their educational programs. In fact, recent findings suggest that greater than 95% of children with hearing loss receive specialized supports through their schools (GRI, 2013). These support personnel include interpreters, paraprofessionals, speech-language pathologists, physical therapists, occupational therapists, social workers, psychologists, tutors, assistive technology specialists, itinerant teachers of the Deaf/hard-of-hearing, auditory-verbal therapists, and educational audiologists. The amount and frequency of these services vary widely for each child, dependent on individual need and available resources. However, research conducted by Soukup and Feinstein (2007) indicates that the IDEA mandates have been difficult to implement for children with hearing loss and intellectual disabilities. Unfortunately, many deaf and hard of hearing educators do not have specific training for educating dually diagnosed children. Similarly, special education professionals are typically not specialized in instructional approaches for children with hearing loss. Kreisman and John (2010) have summarized due process trials under the Individuals with Disabilities Education Act involving children with hearing loss and children with auditory processing disorders. Oftentimes, parents must advocate for the unique needs of their multiply-involved children to ensure that all appropriate
accommodations and modifications for their child’s hearing loss and additional disabilities are accounted for in their child’s educational planning.

Specific accommodations are often recommended for children with hearing loss. A different set of accommodations are typically endorsed for children with developmental disability, dependent on their specific diagnosis and functional abilities. Therefore, children with co-occurring hearing loss and disability require very specialized accommodations and modifications to promote equal access to classroom instruction, particularly in mainstreamed educational settings. In 2011, ninety-five percent of children aged 6-21 years of age receiving public education services through IDEA part B were educated in regular, mainstream classrooms for at least some portion of the school day (US Department of Education, 2013). Therefore, classroom teachers, deaf and hard of hearing educators, and special education professionals must be prepared to design and implement educational programming for children with multiple disabilities.

Leppo and colleagues (2013) provide literature review and discussion of reasonable accommodations for deaf and hard of hearing students with co-occurring disabilities. Accommodations uniquely relevant to children with hearing loss may include sign language interpreters, note takers or speech-to-text technology for lectures, assistive listening devices, closed captioning of videos used in the classroom, and extra time for tests and standardized assessments. Given the vast range of diversity in additional disabilities, these typical accommodations may not be relevant for all children with comorbid diagnoses. Therefore, the heterogeneity of this student population requires individualized accommodation considerations (Leppo, Cawthon, & Bond, 2013; IDEA, 2004). Additionally, accommodations and individualized plans are not static for children with hearing loss and co-occurring disabilities.
Frequent monitoring of progress within different educational contexts is essential for verifying accommodation provisions.

Special educators have a wealth of knowledge and practice in teaching and modifying behaviors of children with developmental disabilities. A number of successful approaches have been published in special education literature. These methods and strategies for specific behavioral modifications and social expectations can and should be adapted for use across familiar and non-familiar environments. Although no known studies have assessed the efficacy of these strategies in the pediatric audiology clinic, it can be reasonably inferred that these methods have a place in the pediatric audiolologic clinic for certain patients.

The field of applied behavioral analysis (ABA) has a strong history of addressing the unique needs of people with disabilities, especially in behavioral intervention strategies for individuals with autism spectrum disorders. ABA research has demonstrated significant success in reducing aberrant behaviors and increasing socially appropriate behaviors (Axelrod, McElrath, & Wine, 2012). The systematic processes underlying behavior modification in applied behavior analysis can certainly be used in the audiology clinic. In fact, operant conditioning tasks elicited with visual reinforcement audiometry and conditioned play audiometry already utilize the most basic principles of ABA. Through pairing auditory stimuli with expected behavioral responses and reinforcing correct responses, the audiologist is fundamentally conducting and recording audiologic data for discreet trials. Audiologists teach expected behavioral responses to these tasks through a range of prompts, including vocal, visual, gestural, and physical prompting. Correct behaviors are also modeled to teach behavioral audiometry in the sound booth.

Reinforcement for correct responses should vary according to each patient’s interests and motivators. For example, reinforcers may include a short break to watch the video screen or play
with a preferred toy, a small edible treat, or verbal praise and a high-five. Children utilizing token boards or star charts may continue to use them in the audiology clinic in order to earn their reinforcers after a number of correct responses. In addition to incorporating ABA principles into diagnostic audiometry, applied behavior analysis methods can also be integrated into teaching self-sufficiency in caring for and maintaining hearing instruments for children with disabilities. For instance, chaining strategies may be used to teach the children the process of opening their hearing aid battery door, removing the dead battery, inserting a new battery, turning the hearing aid back on, and re-inserting it in their ear. These principles may also be useful in teaching appropriate ways to clean and store hearing instruments.

Many individuals with hearing loss benefit from visual strategies to support communication. Research also substantiates the use of visual supports for children with developmental disabilities, particularly those with autism spectrum disorders. Visual activity schedules typically depict a sequence of expected events and transitions through a series of pictures or symbols. Significant improvement in classroom transitions for students with autism have been demonstrated with the use of visual schedules (Pierce, Spriggs, Gast, & Luscre, 2013). Likewise, visual schedules or agendas have been recommended for generalized use across educational, home, and community settings. Chebuhar and colleagues (2013) studied the use of picture schedules for individuals with autism spectrum in healthcare settings. Results of this study indicated reduced parental stress and a decrease in maladaptive behaviors exhibited by children in pediatric dental, psychiatric, surgery, and other specialty clinics (Chebuhar, McCarthy, Bosch, & Baker, 2013). Although data in this pilot project represent a small sample of children with developmental disabilities, outcome results certainly encourage trial use of
visual schedules within the pediatric audiology clinic. (See Appendix A for sample visual activity schedules for audiologic assessment and intervention.)

Video modeling, consisting of recorded video examples and narration for expected behaviors in particular settings, is another visual tool for children on the autism spectrum. Video models have proven successful in teaching appropriate behaviors and facilitating transitions for children with developmental disabilities. In fact, behaviors can be acquired more quickly through video modeling than through live modeling of the expected behavior (Charlop-Christy, Le, & Freeman, 2000). These visual tools have direct applications in the pediatric audiology clinic. Prior to the appointment, the patient could watch a video of another child experiencing a comprehensive audiologic evaluation, with the audiologist narrating the steps. This video could provide verbal expectations for behaviors and participation in the audiology clinic, with the addition of visual supports to familiarize the child to the clinical environment and to the audiologist performing the procedures.

Social stories consist of text describing a specific social situation and expected behaviors associated with the event. Although the degree of lasting benefit with social story intervention varies somewhat across research findings, many studies demonstrate success for some individuals (Karkhaneh, et al., 2010). Wide heterogeneity across students with autism spectrum disorders may factor into this range of variability in outcome data. Effective social stories for children with autism spectrum disorders address who is involved in the scenario, what is expected to occur, where the event transpires, when the event takes place, and why it occurs (Gray, 2000). Stories should be written at the child’s functioning level with a goal to share information in a reassuring manner. Although further research on this topic is warranted, current single case studies consistently demonstrate positive effects for higher functioning children with
autism spectrum disorders in the short-term (Karkhaneh, et al., 2010). In the pediatric audiology clinic, social stories could be used to effectively prepare the patient for audiologic procedures and outline expectations and reasoning for behavioral participation in the evaluation. Stories could begin with the arrival at the clinic, procedures for signing in and waiting in the lobby, waiting for their name to be called, and continue to narrate the audiologic evaluation protocol sequence. As frequent recitation is encouraged for best outcomes, parents could go over the social story with their child several times prior to the appointment, and re-read material while waiting in the waiting area to encourage familiarity with expected outcomes. (See Appendix B for sample social story for audiologic assessment expectations.)

Power Cards are another strategy used in special education, particularly for those with autism spectrum disorders. In this approach, a preferred movie, television, or video game character is fixed to text that outlines steps for expected behaviors or routines (Gagnon, 2001; Keeling, Myles, Gagnon, & Simpson, 2003). The basic premise for this educational approach is the capitalization on a particular interest as a motivator for improving behaviors in particular settings or situations. In the audiology clinic, this could include picture cards of Spiderman with the following steps: “Spiderman says to wear the headphones,” “Spiderman says to listen for the beeps,” and so forth. Although this is a relatively quick and easy modification to add to the pediatric audiologic evaluation, the individual nature of this strategy makes widespread implementation impossible for numerous patients with autism spectrum disorders. Power cards may be a useful tactic for certain patients, depending on their particular “hero” motivators. However, patients’ motivators may change over time, rendering ineffective use in follow-up appointments, especially when these children may only interact with their audiologist on a bi-annual or annual basis. Due to these challenges in a clinical environment, power cards may be
better suited to assisting parents, caregivers, and teachers in encouraging the child to develop proper behaviors and routines related to use, care, and maintenance for hearing instruments in their daily lives. (See Appendix C for sample power cards incorporating hearing instrument retention strategies and hearing assessment.)

Pediatric audiologists should be aware of the challenges experienced by children with varying disabilities and comorbid hearing loss. However, due to the individual nature of children with these impairments, each patient should be evaluated based on his or her own distinctive qualities. Collaboration with other professionals in a family-centered approach is essential in creating comprehensive assessment and intervention strategies. Certain testing paradigms may require modifications based on each patient’s developmental abilities. The result of these best practices are certainly worthwhile, as children with varying disabilities co-occurring with hearing loss can absolutely receive benefit from these approaches, contributing to a better quality of life.
CHAPTER VI

FUTURE DIRECTIONS

Further research across several areas of intervention, habilitation, healthcare collaboration, and education planning is desperately needed for children with hearing loss and concomitant developmental disabilities. However, young children with multiple disabilities are widely heterogeneous in terms of their characteristics, capabilities, and learning needs (Horn & Kang, 2012). Therefore, single-case designs are often the most appropriate model for studying outcomes in this special population. Currently, the evidence base does not contain enough replications of such interventions for children with co-occurring developmental disabilities. Interdisciplinary coordination and collaboration can lead to development of better tools and strategies for children with special needs across settings. For pediatric healthcare specialists, special education resources should be considered in order to limit communication barriers and to encourage participation in diagnostic assessments and interventions. Currently, specific tools for assessing and monitoring auditory skill development of children with additional developmental disabilities do not exist. Audiologists and healthcare practitioners should continue to adapt and modify strategies for children with co-occurring hearing loss and developmental disabilities, closely monitor individual outcome data, and publish their results in order to widen the clinical evidence base for these pediatric patients.
CONCLUSIONS

Increases in survival rates for medically fragile children are a relatively new phenomenon. A range of co-occurring developmental disabilities is very common in children with hearing loss. Heterogeneity in this complex population makes development of standardized assessments for each disability subgroup nearly impossible. Therefore, audiologists and healthcare providers across a range of disciplines must modify their approaches in order to best assess and provide habilitation for children with multiple special needs. Children with hearing loss and accompanying disabilities often require substantial services, modified assessment techniques, and unique treatment planning to accommodate their distinctive needs, especially if spoken language outcomes are desired.

It is important that clinicians realize that hearing aids and assistive listening technologies are not the only, or even primary, concern for many families of children with co-occurring developmental disabilities. In addition to hearing devices, families are often tasked with learning and incorporating a number of assistive technologies, including mobility devices, positioning and orthotic apparatuses, self-help equipment, adapted toys and switches, and specialized communication devices (Horn & Kang, 2012). Each of these assistive strategies requires extensive financial resources, time, training, and commitment. Teams of qualified pediatric specialists are uniquely trained to support families by designing interdisciplinary intervention plans and monitoring progress across developmental domains. Legal statutes exist to minimize educational barriers and provide appropriate educational and (re)habilitation access for children with disabilities in the United States. These policies, together with family-centered
interdisciplinary resources, promote the attainment of efficacious communication and learning outcomes for children with concomitant disabilities.

Further research on this diverse population is necessary in order to better understand the functional abilities and needs of children with co-occurring hearing loss and developmental disabilities. This information is crucial for improving assessment techniques and intervention strategies across disciplines. Within the field of pediatric audiology, the development of appropriate test materials for quantitative monitoring of progress for children with multiple disabilities is essential for counseling families on their child’s auditory development and in providing realistic expectation data for children with hearing loss and additional disabilities.
REFERENCES


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<tr>
<td>Ear Tickles</td>
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<tr>
<td>Wear Headphones</td>
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<tr>
<td>Play Listening Games</td>
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**You did it!**

- **You did it!**
- **You did it!**
- **You did it!**
- **You did it!**
### Earmold Impressions Schedule

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<td>Putty in Ears</td>
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<tr>
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<td><img src="image4" alt="Wait" /></td>
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</table>

**You did it!**

*Stars for each step completed.*

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60
Sometimes, I need to have my hearing checked.
My audiologist checks my ears and checks my hearing.

When I get to the audiology office, I need to sign in at the desk.
The receptionist may ask my parents to fill out paperwork.

I can play quietly in the lobby while I wait.
When the audiologist calls my name, I will follow her.

I will sit still while the audiologist touches and looks in my ears.
If I feel nervous, I can hold my Mom’s hand.

Next, I get to go in the sound booth.
The audiologist will open the door and I will step in and sit on the chair.

The audiologist will put headphones on my ears.
I will listen and follow directions for the listening game.

When the hearing test is finished, I will earn a prize for my hard work.
My parents will be proud of me for following directions at the audiology office!
Below is a Power Card example for an elementary school student who struggles to keep his hearing aids on while at school. He has frequent behavioral outbursts in which he throws his hearing aids in the classroom. This Power Card example encourages the child to keep his hearing aids on and utilizes his favorite hearing-aid-wearing comic book hero, Blue Ear. Limited teacher and student reactions to disruptive outbursts involving hearing aid throwing is also important to ensure that these behaviors are not inadvertently reinforced.

Blue Ear loves listening with his hearing aids. He wears them at home and at school. Sometimes it is difficult for him to leave his ears alone. Sometimes he gets frustrated and angry. However, he has learned that listening is important for learning. He needs to wear his hearing aids all the time to hear and talk to his teachers and friends. When he is angry, he never throws his hearing aids. Instead, he sits in his special bean bag chair to cool off. He keeps his hearing aids on no matter how he is feeling.

Just like Blue Ear, it is important to keep your hearing aids on at school and never throw them. This would make Blue Ear proud. Blue Ear would like all boys who love him to remember these three things:

1. Keep your hearing aids on at school.
2. When you are upset, you can take a cool-down in your special bean-bag chair. Throwing your hearing aids is never ok.
3. Tell your teacher if you think there is a problem with your hearing aids.
Below is a Power Card example for a kindergarten-aged girl with hearing loss and global developmental delays who is fearful in the audiologic clinic and cries during testing. These behaviors often make behavioral audiologic results difficult/impossible to obtain. Her favorite character is Sofia the First.

Sofía the First has fun at the audiology clinic! She sits quietly on her mommy’s lap and lets Mandi [audiologist’s name] look in her ears. She lets Mandi put a squishy earphone in her ears. This doesn’t hurt. Sometimes it even tickles and makes funny sounds! After this, Sofía the First goes into the listening booth for her hearing test. She gets to wear a different pair of headphones. She follows Mandi’s directions and waits for the beeps. Sofía the First drops a block in the bucket every time she hears a sound.

After playing the fun listening games, Sofía the First gets to choose a special sticker. Her mommy is so proud of her hard work!

Just like Sofía the First, it is important to follow directions when you visit your audiologist. These listening games help make sure your pink princess hearing aids are working right just for you! There is no need to cry or be sad. Your audiologist is very nice and you will have a great time.

Sofía the First would like all girls who love her to remember these three things:

1. The audiology clinic is a fun place with awesome toys.

2. Your audiologist will touch your ears to look in them and help you wear headphones for listening games.

3. You will earn a special prize when you do your best listening and your parents will be so proud of you!