10 Programming for Children With Multiple Disabilities

A significant proportion of the population of deaf or hard-of-hearing children have one or more disabilities that are not caused by their hearing loss, even if they sometimes co-occur with it (Arnos & Pandya, in press). The Gallaudet Annual Survey of Deaf and Hard-of-Hearing Children and Youth (Holden-Pitt & Diaz, 1998; Mitchell & Karchmer, 2006) has indicated that at least 35% and perhaps over 50% of deaf and hard-of-hearing students in the United States have an additional, educationally significant condition or disability. Shallop (2008) reported equally high estimates of the proportion of deaf students using cochlear implants who have developmental conditions that complicate their progress. Despite the high prevalence of multiple disabilities combined with hearing loss, however, it remains difficult to locate either data-based studies of these children or carefully documented evidence regarding educational progress. On the basis of her review of the literature, Guardino (2008) went so far as to suggest that despite the increasing incidence of multiple disabilities among deaf and hard-of-hearing students, there appears to be a decrease over time in research on these students.
The lack of published research concerning children with multiple disabilities, especially at the group-comparison level, is in large part because their unique needs necessitate highly individualized programming that does not lend itself to generalization. Jones and Jones (2003) pointed out that the heterogeneity in both type and severity of developmental difficulties among deaf and hard-of-hearing children requires that decisions about appropriate educational placement and programming must be made on an individual basis. They stressed that interventions need to be family focused and to involve a team of specialists based on both child and family needs. Like Meadow-Orlans et al. (2003), Jones and Jones argued that it is crucial for a case manager to be available to coordinate services in such cases because the needs of these children are so complex. Programming for children with hearing loss and multiple disabilities can and should incorporate approaches and interventions that have shown evidence of success with other children with various types of disabilities. At the same time, it is critical that interventions be sensitive to and provide appropriate accommodations for hearing loss. Such support is required even in the earliest months of life, since early intervention shows positive effects for children with a wide range of disabilities.

VARIABILITY AMONG DEAF AND HARD-OF-HEARING CHILDREN WITH MULTIPLE DISABILITIES

Meadow-Orlans and her colleagues (Meadow-Orlans, Smith-Gray, & Dyssegaard, 1995; Meadow-Orlans et al., 2004) studied a small group of five infants with hearing loss plus a physical, cognitive, or emotional condition that was diagnosed during infancy. Etiology of the identified disabilities included cytomegalovirus (CMV) and birth trauma. There were three comparison groups: one consisting of infants with hearing loss who were identified as being “at risk” for but were not yet identified as having multiple disabilities, a second group of infants with hearing loss who were not considered to be at risk for any additional disability, and a group of hearing infants. Both the infants already identified as having multiple disabilities and those considered at risk had significantly lower birth weights than the hearing children and the children with hearing loss who were not deemed to be at risk.

Meadow-Orlans et al. (1995, 2004) were particularly interested in the levels of stress reported by the hearing mothers of the children identified as having multiple disabilities. Assessments of family stress and child behaviors were conducted using standardized parent-response instruments, the Parenting Stress Index (PSI, Abidin, 1986) and the Stress of Life Events (Holmes & Rahe, 1967) when children were 9 months old, and the Family Support Scale (Dunst,
Parents of children with multiple disabilities who responded to the survey and telephone interviews reported early experiences with their children as being highly stressful (see also Powers, Elliott, Patterson, Shaw, & Taylor, 1995), but they also expressed pride in their children's ability to overcome challenges. A number of parents reported significant problems identifying appropriate services for their children, a difficulty echoed by reports from other researchers (e.g., Ewing & Jones, Guardino, 2008). Finally, it is noteworthy that the hearing losses of those children in the study who had multiple disabilities were identified later than those of children without other disabilities. In some cases, it appeared that the other condition initially had "masked" evidence that the child had a significant hearing loss.

CHILDREN WITH COGNITIVE OR INTELLECTUAL DISABILITIES

Although estimates vary widely, it appears that a significant proportion of deaf and hard-of-hearing children with multiple disabilities have conditions that result in cognitive delays or learning disabilities. The 2005 Gallaudet Annual Survey (see also Holden-Pitt & Diaz, 1998) found that just over 8% of children who were included had "mental retardation" or cognitive delays. Looked at from the opposite direction, Guardino (2008) estimated that over 9% of children who have cognitive delays also have a hearing loss. In her review of the research, Guardino found that many of the data-based reports available about children with hearing loss plus cognitive challenges had been conducted in the 1980s. She suggested that the subsequent decrease in research involving this population resulted from the aging of the children who were affected by maternal rubella and, to some degree, by the non-categorical approach to education that has occurred since then. Whatever the cause, there remains a significant need for research into educational and family interventions oriented toward this combination of developmental challenges.

Knoors and Vervloed (in press) noted that of the children who have diagnosed cognitive delays in addition to hearing loss, approximately 30% have unknown etiologies. Of those with known etiologies, the majority has histories of pre- or perinatal CMV, rubella (German measles), kernicterus (severe jaundice/bilirubin encaphalopathy) or, particularly in the case of later onset of learning difficulty, infections such as meningitis. These etiologies typically have multiple developmental sequelae, and Knoors and Vervloed argued that while assessment and educational programming need to vary according to the profile of cognitive abilities, they also have to take into account the needs of children with hearing loss. In some cases, these children may be able to use spoken language, while for others a natural sign language or a total communication approach may be
Jenkins, & Trivette, 1984) and the Parenting Events Inventory (Crnic & Greenberg, 1990) when the children were 15 months of age. The results showed a bimodal distribution of stress ratings, with individual ratings falling either at the highest stress level or at a level lower than that typical for hearing parents of hearing infants. The low ratings fell in the area defined by the test as "suspect denial of stress," and the researchers suggested that such denial was taking place. They noted that a similar bimodal pattern of reported stress was obtained from the group of parents whose children were currently "at risk" but not identified as having multiple disabilities.

Although parents' ratings of stress varied, all five children in the Meadow-Orlans et al. (1995) study with confirmed multiple disabilities showed clear developmental delays by 12 months of age. Each child's profile of functioning was unique, but three of the five were described as uninterested in interacting with others and showed aberrant patterns of visual attention to people and objects. Two were reported to have extremely short attention spans. In contrast, over 70% of the children in the group identified as being at risk for but not identified with multiple disabilities showed no evidence of developmental delays or difficulties by 12 months of age. Meadow-Orlans et al., suggested that the early identification of hearing loss together with early intervention services had helped to prevent more instances of delays in this group. Other investigators also have reported varying developmental patterns in at-risk groups, including variability in the vocal behaviors achieved by children with moderate-to-severe hearing loss who have multiple disabilities (Nathani, Oller, & Neal, 2007). Although early intervention can provide positive developmental and family support, it cannot eliminate most organically based difficulties and delays.

In a study focused on older deaf and hard-of-hearing children, Meadow-Orlans et al. (2003) surveyed parents of 6- and 7-year-old children with hearing loss, some of whom had significant additional conditions affecting their development. The initial survey (n = 404) was followed by phone interviews with randomly selected parents (n = 62) and several face-to-face interviews. All of the children were enrolled in programs for deaf and hard-of-hearing students, but 32% of the respondents indicated that their children had educationally significant conditions in addition to hearing loss. Of the children reported to have additional complicating conditions, the largest specific group involved 12% of the sample, who were identified as having intellectual or cognitive delays. Significant proportions of those children also were reported to have vision loss, learning disability, attention deficit disorder, emotional or behavioral problems, cerebral palsy, or motor disabilities. Another 29% of the children with additional complicating conditions were reported to be in an "other" category, including children with brain damage, epilepsy, and health conditions. Clearly, this was a very heterogeneous group, and in that respect it is representative of children with hearing loss and additional developmentally relevant conditions.
more appropriate (van Dijk, van Helvoort, Aan den Toorn, & Bos, 1998, cited in Knors & Vervoorn, in press; van Dijk, Nelson, Postma, & van Dijk, in press). Van Dijk et al. (1998) found that a group of five deaf adults with moderate cognitive/intellectual disabilities living in a residential group home were able to learn and use signs that were taught during school time. Although sign-supported speech (using Signed Dutch) was the officially preferred mode of communication at school, van Dijk et al. noted that the participants spontaneously developed some sign structures that were like those in Sign Language of the Netherlands (NGT). Van Dijk et al. posited that more interaction with signing caregivers and other professionals who were fluent signers would have accelerated the participants’ signed communication abilities. Depending upon their level of cognitive functioning, some students may require instruction in using a selected and simplified set of signs or even picture- or symbol-based augmentative and alternative communication systems. These systems can involve communication boards on which symbols are manipulated, or they may be electronic, employing sometimes sophisticated software (see www.asha.org for information about augmentative and alternative systems).

Cochlear Implants, Cognitive Delays, and Language Development

Cochlear implants are sometimes provided to children who are deaf and have cognitive or related disabilities, but the effectiveness of the implants typically decreases compared with that of children who are only deaf. Parents therefore need to be informed that results cannot be expected to match those of children without cognitive disabilities (Pyman, Blamey, Lacy, Clark, & Dowell, 2000; Spencer, 2004). Pyman et al. found that basic auditory awareness and discrimination of vowels and consonants increased for children with motor and/or cognitive disabilities after 4 years of cochlear implant use. Nevertheless, only about 60% of the children could identify spoken words in sentences, while 80% of children in their study without cognitive disabilities could do so. Waltzman, Scalchunes, and Cohen (2000) similarly found increases in awareness of sound and increased evidence of being “connected” or “in touch” with their environment in a group of children with diverse multiple disabilities who received cochlear implants. Increases in language abilities were highly variable within the group, however, and children with greater cognitive disability were unable to complete the series of tests that were administered. A similarly wide range of functioning after cochlear implantation was shown in a German study by Hamzavi et al. (2000), in which 5 of the 10 participating children did not acquire spoken word reception or production skills after 3 years of using the implant, although 4 of the 5 lower functioning children gave evidence of some awareness of sound using the implants. Fukuda et al. (2003) presented single-case data on a
child with moderate developmental delay who had a sizable sign language vocabulary prior to cochlear implantation and who developed spoken language skills after implantation.

The type and severity of additional disabilities may be the determining factor for spoken language progress using cochlear implants. Holt and Kirk (2005) assessed the speech and spoken language development of 19 children with mild cognitive delays compared to 50 children without cognitive or any other identified disabilities; all had cochlear implants. Using a standardized parent report instrument completed at 6-month intervals, auditory skills at the awareness and word identification levels were found to advance for both groups, although children in the group with cognitive delays showed slower average progress and greater variability. Consistent with results of Pyman et al. (2000), children with cognitive disabilities required longer experience with their implants to achieve multiword/sentence understanding. Differences with the Waltzman et al. (2000) study were presumed to result from differences in type and severity of additional disabilities in the groups.

A final note: None of the investigators whose work is summarized above were able to identify specific predictors of outcomes of cochlear implantation for children with multiple disabilities, including mild cognitive delay and hearing loss. All called for further investigation of both predictors and methods for providing supportive therapy.

LEARNING DISABILITIES AND ATTENTION DEFICIT DISORDER

The term "learning disabilities" comprises a group of learning problems such as dyslexia, auditory processing disorder, visual perception difficulties, memory or executive function disorder, specific language impairment that is not due to hearing loss, and general cognitive or experiential deficits (Edwards, 2010). Learning disabilities so defined, regardless of whether children have a hearing loss, are considered to be of an organic origin, and medical testing typically indicates some central nervous system dysfunction. In an early electroencephalogram (EEG) study of 286 children in a special school for deaf children, Zwiercki, Stansberry, Porter, and Hayes (1976) found that 35 had obvious signs of neurological dysfunction and 21 had signs of minimal brain dysfunction. This finding suggested that a high proportion of children have learning disabilities not directly resulting from their hearing loss. Pisoni, Conway, Kronenberger, Hennings, and Anaya (2010) reached a similar conclusion from studies of children with cochlear implants, suggesting that many have dysfunctions or delays in basic neurocognitive functioning underlying information processing. Hawker et al. (2008) suggested that the language delays of some children using cochlear implants have
the same basis as specific language impairments in hearing children and do not require an explanation based on auditory experience. At this time, however, clear diagnostic guidelines for identification of specific language and learning disabilities in children with hearing loss continue to evade understanding, perhaps in part due to lack of sufficient descriptions of children’s performance and learning in varied contexts.

Laughton (1989, p. 74) proposed that children who are deaf or hard of hearing and also have learning disabilities will have “significant difficulty with the acquisition, integration, and use of language and/or nonlinguistic abilities” relative to peers with hearing loss only. Given the complexities associated with these processes in deaf children at large (see chapter 7), diagnosing learning disabilities remains a process of clinical judgment and problem solving on the part of clinicians who are conducting the assessment (see van Dijk et al., 2010). Perhaps as a result, both learning disabilities and attention dysfunctions appear to be overdiagnosed among those children (see Parasnis, Samar, & Berent, 2001). To some extent, this situation undoubtedly results from the overlap of behaviors symptomatic of learning disabilities in hearing children and behaviors due to late and inconsistent experience with language and resultant communication disabilities in children with hearing loss (Morgan & Vernon, 1994; Samar, Parasnis, & Berent, 1998).

On the other hand, Calderon (1998) suggested that learning disabilities tend to co-occur with hearing loss at a high rate due to shared etiologies, and this phenomenon may be one source of the cognitive differences between students with and without hearing loss summarized previously in this book (see, especially, chapter 7). Mauk and Mauk (1998) noted that estimates of the prevalence of learning disabilities in the population of deaf and hard-of-hearing children are highly variable, ranging from 3% to 60%. Samar et al. (1998) posited that relative lack of auditory input cannot explain the high rate of phonological and reading difficulties in the population of children with hearing loss, implying that this rate reflects learning disabilities. Given that learning disabilities are said to occur in 3% to 10% of hearing children, at least that rate could be expected for those with hearing loss (Edwards, 2010; Edwards & Crocker, 2008).

Deaf and hard-of-hearing children suspected of having learning disabilities are most often placed in classes for children with hearing loss. However, their special difficulties with integration of information in addition to delays in language development (regardless of modality of input) are likely to require a more highly structured educational environment for optimal academic development (Stewart & Kluwin, 2001). Their greater problems with memory, sequencing, and attention, relative to hearing children, as well as inconsistent performance over times and contexts also may require special educational supports beyond those effective for other deaf children. Importantly, those difficulties generally are found to characterize the learning behaviors of many deaf children.
(e.g., Marschark & Hauser, 2008), and it is important to determine the extent to which such findings are affected by the inclusion in research studies of students who actually have concomitant learning disabilities.

Reliable and valid assessment of learning disability in a deaf or hard-of-hearing child presents special difficulties and must employ varied methods and measures. Morgan and Vernon (1994) recommended a specific battery of tests including a case history (noting especially medical conditions and family history of reading or learning disabilities); two standardized measures of nonverbal cognitive functioning (to rule out overall cognitive delay); a measure of academic achievement; neuropsychological screening (to look for signs of dysfunction typically found in hearing, learning disabled children); and an evaluation of adaptive behaviors or daily function skills; plus testing of hearing, language, and communication skills using formal assessment tools (see also Hauser et al., 2008).

One of the signs of learning disability is a gap between potential—as indicated by a nonverbal cognitive or intelligence test—and achievement. However, virtually all of the relevant tests have norms and instructions appropriate only for the hearing student population, and this can lead to invalid and misleading test interpretation. Edwards (2010) therefore suggested using more than one test when assessing a specific psychological function in a child with hearing loss. The use of multiple tests and testing procedures, in fact, generally is recommended for all educational and developmental assessments.

Effective programming for children with hearing loss plus learning disabilities is complicated by the above-described lack of specific diagnostic approaches. The situation is made more complex by the necessity of cooperation among professionals in several different fields and the need for specialists who understand the particular effects of hearing loss (Laughton, 1989; Mauk & Mauk, 1998). Intervention-focused research in this area could be of much benefit, but additional work first is needed on identifying children with a combination of hearing loss and learning disabilities. Mauk and Mauk noted that simply using interventions designed for hearing children is neither sufficient nor appropriate, but there is a general lack of research in the area that might help to change the situation (Guardino, 2008).

Attention Disorders

Although they often co-occur, attention disorders (characterized by inattentiveness, hyperactivity, and/or impulsivity) can be present even when other learning disabilities are not. Like learning disabilities, the diagnosis of attention disorders in children with hearing loss is complex and remains more art than science. The communication histories of many children with hearing loss make it difficult to distinguish between those with organic attention and activity difficulties as opposed to patterns typical of deaf and hard-of-hearing children in general.
For example, we noted in the discussion of cognitive performance and cognitive styles (chapter 7) that selective and sustained attention is often attenuated for children with hearing loss compared to hearing children (e.g., Kritzer, 2009; Quittner et al., 1994), and many deaf or hard-of-hearing children appear to fit a category designated as “hyperactive.”

Kelly, Forney, Parker-Fisher, and Jones (1993) and Samar et al. (1998) found a greatly increased prevalence of attention and activity-level disorders in deaf and hard-of-hearing children with acquired hearing loss compared to those with an identified hereditary etiology. This suggests that some of the non-genetic causes of hearing loss during pre-, peri-, or post-natal periods (such as viral infections, prematurity, or meningitis) can have effects on the nervous system beyond auditory functioning. Kelly et al. (1993) suggested that interventions for children with hearing loss and attention disorders should be similar to those already used in practice with hearing children who have attention disorders: Classrooms should be designed so that visual distractions are minimized, basic study routines and techniques for organization should be explicitly taught, and visual organizers such as charts should be used whenever possible in the curriculum. Unfortunately, we were not able to find outcome data based on experimental manipulation of these or other intervention techniques for children with both hearing loss and attention disorders beyond occasional case studies. Because of the apparent prevalence of their co-occurrence, there is an urgent need for research and the development of assessment tools in this area.

AUTISM SPECTRUM DISORDERS

Diagnoses of childhood Autism-Spectrum Disorders (ASD) have increased over the past few decades. This disability can and does co-occur at a fairly high rate with hearing loss (Baily, de Choluy de Lenclaye, & Lauwerier, 2003; Rosenhall, Nordin, Sandstrom, Ahlsen, & Gillberg, 1999). The behaviors characteristic of deaf or hearing children with ASD differ significantly from those of children who are only deaf or hard of hearing, however, so they cannot be entirely explained by hearing loss itself (Gravel, Dunn, Lee, & Ellis, 2006). ASD generally is characterized by severe impairment of social interaction abilities, disruptions in eye contact with others, production of repetitive stereotyped movements, language delay and disorders, and cognitive impairment or uneven profiles of cognitive skills (Edwards & Crocker, 2008; Kanner, 1943, summarized in Vernon & Rhodes, 2009). Hyperactivity, attention span disorders, and aggression toward self or others also may be present although, as with other disorders, the presence and the severity of characteristics vary across individuals. (For more details, see Diagnostic and Statistical Manual of Mental Disorders-IV-RT of the American
Psychiatric Association, 2000.) For example, one of the subtypes currently considered to be within the spectrum of autism is Asperger Syndrome. Children with "Asperger's" typically pass language milestones at the expected ages and show average or even much higher levels of most aspects of cognition along with varied degrees of impairment of social behaviors (Volkmar, Klin, Schultz, Rubin, & Bronen, 2000). A diagnosis of ASD thus can represent a wide range of functioning skills and potential.

Autism Spectrum Disorders were once thought (incorrectly) to result from poor early interactive experiences, but they are now known to have organic, neurological, or physiological origins (Vernon & Rhodes, 2009), even if the exact mechanisms remain unknown (Rutter, 2005). Using generally accepted criteria for the identification of ASD, Jure, Rapin, and Tuchman (1991) concluded that about 4% to 5% of a population of 1,500 children with hearing loss also had autism. The etiology of the children so identified varied widely, however, and hearing loss and ASD can be associated with the same or similar etiologies (e.g., meningitis, epilepsy, congenital rubella syndrome, CHARGE;1 van Dijk et al., 2010). Deafblindness (see below) can lead to disrupted communicative behaviors and repetitive, stereotyped actions like those produced by many children with ASD, but Hoevenaars-van den Boom, Antonissen, Knoors, and Vervloet (2009) reported that deaf children with autism can be differentiated from deafblind children based on the quality of their social interactions.

Because children who are deaf or hard of hearing and have ASD vary greatly in their individual behaviors and abilities, treatment and educational interventions cannot be generalized for all children with the diagnosis. Nevertheless, Vernon and Rhodes (2009) indicated that there is consensus about the importance of early and intensive interventions, including treatments for both behavioral and communication aspects. Some interventions for autism alone employ signed language or other forms of visual communication (Bonvillian, Nelson, & Rhyne, 1981) and thus may be especially appropriate for children with autism plus hearing loss. Augmentative/alternative communication devices such as the Picture Exchange Communication System (PECS) also are used with some children who have severe ASD and might be appropriate for some children who also have hearing loss. In a recent review, however, Ostryn (2008) noted that there are few empirical studies of its effects, even with the general ASD population.

Lovaas (1987) developed a treatment approach referred to as Applied Behavior Analysis (ABA) in which negative and positive reinforcements are used to

1 CHARGE takes its name from the related symptomology: Coloboma (a keyhole type opening in iris and retina), Heart defect, Atresia of the choanae (blockage of the passages between the nasal cavity and the naso-pharynx), Retarded growth and/or development, Genital hypoplasia, and Ear anomalies/deafness.
modify the behaviors of children with ASD. This approach is labor intensive, highly structured, and greatly extended over time. Treatment involves parents, therapists, aides, and teachers working to improve a child's functioning in carefully defined, small steps. It is expensive to implement, but Lowaas reported data that showed some success with hearing children. Zacher, Ben-Itzchak, Rabinovich, and Lahat (2007) reported that 20 young children with ASD made more progress over a year's time in an ABA intervention program than 19 children of the same age and diagnosis who participated in a more developmentally oriented, eclectic program. This finding is consistent with other reports indicating that a highly structured program is necessary for children with ASD, especially if learning is to transfer beyond the teaching situation. Unfortunately, there appear to be no scientific studies available on the outcomes of interventions specifically with children with ASD and hearing loss beyond an occasional case study or personal report (Edwards & Crocker, 2008). An evidence base for practice is lacking.

DEAFBLINDNESS

There is a long history of programming and research involving children who have a combination of hearing and visual impairment, now referred to as deafblindness (van Dijk et al., 2010). Although total loss of either sense is rare, van Dijk et al. noted that the condition is characterized by enough loss in each area to preclude using it to compensate for loss in the other. Deafblindness can occur congenitally or at an early stage of life and, if so, has much more severe effects than if acquired later. Nevertheless the well-known stories of Helen Keller (who became deafblind at 19 months) and Laura Bridgeman (who became deafblind at 24 months) demonstrate the difficulty of communication development even when a child initially has sight and hearing.

Jan van Dijk and his colleagues developed a curriculum that is used in many countries to facilitate development of deafblind students. The curriculum stresses building relationships between the child and caregivers, gradually building awareness in the child of others, and supporting transition of communication behaviors from the concrete to the symbolic level. Chen, Klein, and Haney (2007) and van den Tillaart and Janssen (2006) developed curricula based on van Dijk's ideas, and a single subject, multiple baseline study conducted with four deafblind children indicated effectiveness of the approach. At least one comprehensive instrument for assessing behaviors of deafblind children with multiple difficulties, the Callier-Azusa Scales (Stillman, 1978; Stillman & Battle, 1986), also has been developed based on van Dijk's work.
Congenital Rubella Syndrome

Deafblindness can result from many of the same etiologies listed above for other disabilities, including a variety of pre-, peri-, and post-natal illnesses. Deafblindness can be, but is not always, associated with cognitive delays or deficits or with autism. Individuals who are deafblind due to rubella contracted during the early gestational period are particularly likely to also have a number of developmental difficulties, including intellectual deficits, behavioral difficulties, and repetitive stereotypical or obsessive movements like those found in children with autism (Munroe, 1999).

The incidence of congenital rubella syndrome (CRS) has decreased with the rise of vaccine use worldwide, but it is still an etiology that occurs in some parts of the world, and persons born during previous epidemics, although now adults, are still in need of special programming. There is some evidence that vision and hearing losses of deafblind persons with CRS worsen with age (Kingma, Schoenmaker, Damen, & Nune, 1997; Munroe, 1999; van Dijk, 1999), so continuing individualization and modification of interventions are necessary.

Genetic/Chromosomal Syndromes

A number of genetic/chromosomal syndromes are associated with deafblindness (see Arnes & Pandya, in press). These include but are not limited to CHARGE and Usher syndromes. CHARGE syndrome is the most prevalent etiology for deafblind people in the United States (Killoran, 2007). It can include "keyhole" openings in the irises and retinas of the eyes causing vision loss, blockage of passages between nasal cavity and nasopharynx, structural ear anomalies and hearing loss, balance problems, genital anomalies, hypotonia (low muscle strength), feeding and swallowing problems, and asymmetric facial palsy. Children with CHARGE can be medically fragile and require multiple surgeries early in life. Related behavior problems are common, characterized by a lack of impulse control. As with other conditions described above, severity of these impairments and the number of symptoms differ across children. Blake (2005) reported that the majority of a group of 30 individuals he studied required medications for behavior control and that two-thirds required substantial supervision and support. Van Dijk et al. (2010) noted that education and management of CHARGE children is particularly difficult and can be further complicated if supportive early interaction experiences are disrupted due to parental stress. Clearly, children with this syndrome, and their families, require consistent and specialized support.

Usher syndrome is another prevalent genetic cause of deafblindness, occurring in around 4% of children with hearing loss. There are several different
subtypes of Usher syndrome, and different characteristics suggest different emphases in educational interventions (Knoors & Vervloed, in press; van Dijk et al., 2010). Persons with Usher type 1 typically have significant hearing loss at or soon after birth, with visual loss occurring later. They usually are supported educationally through programs serving deaf children and with essentially the same methods—and arguments about language methods—as other children with hearing loss. Individuals with Usher type 2 tend to have lesser hearing losses (in the hard-of-hearing range), with vision loss typically occurring in adolescence. Persons with Usher type 3 have both hearing and vision functioning for a number of years before experiencing deterioration in both senses.

There is no intellectual disability associated with Usher syndrome, and van Dijk et al. (2010) indicated that clinical practice has suggested considerable emotional strength in students with this diagnosis. Vermeulen and van Dijk (1994) administered a personality assessment instrument to 16 adolescents with Usher syndrome and reported that these individuals showed strong ego functioning, social competence, and self-esteem. They noted that the group's scores indicated a relative lack of assertiveness, however, which they attributed to probable overprotection from parents and educators. Damon, Krabbe, Kilsby, and Mylanus (2005) surveyed 67 persons from six European Union countries who had a diagnosis of Usher syndrome and found that the respondents had generally positive attitudes and strived to maintain their independence. Respondents were particularly interested in methods and technologies that would support their socialization and independence.

Cochlear implants are considered to be a viable option for children with Usher syndrome or other children who have hearing loss in association with visual impairment. Yoshinaga-Itano (2003) reported on one child with profound hearing loss and progressive vision loss who began receiving intervention services at 6 weeks of age. Her hearing family used ASL along with some pidgin signed English for communication with her, and when she was 20 months old, she scored at the 99th percentile on the MacArthur Communicative Development Inventory Words and Sentences form (Fenson et al., 1993, 1994), using signs but compared to hearing norms. At that time she produced no spoken language. After cochlear implantation at 21 months of age, however, she began to use more vocal behaviors and to build auditory awareness. By 51 months of age, she had become primarily a spoken language user, a particularly fortunate transition due to deteriorating vision which seriously interfered with her reception of sign language. Yoshinaga-Itano presented this case as an example of the way that sign language can support emerging spoken language development when auditory reception is improved via use of a cochlear implant. This case also indicates that children who are eventually identified as deafblind do not necessarily experience significantly delayed early development.
A BROADER VIEW

The information in this chapter is limited in that it focuses on only some of the disabilities that can co-occur with hearing loss. One area that was not reviewed, for example, was the co-occurrence of motor/physical disabilities such as cerebral palsy with hearing loss. Meadow-Orlans et al. (2003) included comments from several parents of children with this combination of challenges, and it was clearly difficult for them to find appropriate programming and support. When motor disabilities complicate both expressive signing and speech, communication options are more limited and augmentative alternative methods must be considered.

Another area that was not discussed in this chapter was that of general emotional and behavioral disorders (Edwards & Crocker, 2008). These long have been evident as a potential problem area for deaf and hard-of-hearing children (e.g., Glenn, 1988; Meadow & Trybus, 1985), but such a wide variety of descriptors have been given across research reports (including, for example, inattentiveness, aggressiveness, anxiety, and even academic disorders) that it is not clear to what extent emotional/behavioral disorders form a discrete category. In addition, some emotional/behavioral problems of deaf and hard-of-hearing children have been posited to arise from basic communication and language delays, with concomitant disruption in the ability to communicate with parents and caregivers rather than indicating an organically based “disability.”

Despite the organization of this chapter by type of disability, current educational philosophies emphasize individual differences instead of such categorization. Evidence presented in each of the categories above illustrated a range of functional skills and needs, so that placement decisions cannot validly be based on etiology or labeling of the disability associated with the hearing loss. Accordingly, Ewing and Jones (2003) argued for a transdisciplinary approach to assessment and programming for multiply disabled deaf and hard-of-hearing children, which they described as characterized by indirect instead of direct service. This approach is highly collaborative, with around 10 specialists (including at least one knowledgeable with regard to deaf children) potentially needed to program sufficiently for a single child. Only one or two professionals are primary service deliverers or facilitators, however, so that communication with parents, therapists, and other educators can be more coherent and consistent. Such an approach would be responsive to parents’ complaints that they often have had to deal with too many professionals, some of whom give divergent recommendations and are seemingly unaware of recommendations from other specialists (Giangreco, Edelman, MacFarland, & Luiselli, 1997; Meadow-Orlans et al., 2003).

Ewing and Jones (2003) also recommended the use of person-centered instead of category-centered programming and offered the McGill Action Planning
System (Forest & Pearnpoint, 1992) as one example. A person-centered approach is based on identifying the strengths and learning abilities of a student, motivating factors, environments and contexts in which learning is facilitated, and specific instructional procedures that best promote learning. The process of identifying these components should include family, child, and professionals and would follow best from actual teaching-learning trials instead of use of standardized tests or procedures. Although this would be an ideal approach with all students, it may be a necessity for students with hearing loss plus additional disabilities. Furthermore, because there are few curriculum materials designed for specific combinations of disabilities, teachers need to be knowledgeable about a wide range of disabilities, even when they have the advantage of working with a supportive team.

SUMMARY: SERVING CHILDREN WITH MULTIPLE CHALLENGES

The presence of additional disabilities in the population of students with hearing loss continues and appears to be growing, as children who are born prematurely, have severe birth complications, or survive serious illness are increasingly likely to survive. As with other deaf and hard-of-hearing children, generalizations cannot be made about these children's academic and functional capabilities based on their etiologies, but it is clear that the effect of disabilities multiplies as they increase in severity and number.

- With one-third to as much as one-half of students with hearing loss being diagnosed as having some additional disability, educational planning must provide for handling a diversity of needs. Service provision for those children requires multiple specialists and, typically, more intensive service delivery than that for children with hearing loss alone. Collaboration across disciplines and among teachers and other service providers is critical.
- As with other students with hearing loss, those with multiple disabilities will vary in their abilities to acquire language skills, and options ranging from oral approaches to sign-only and augmentative or picture/computer-based approaches may be appropriate for specific individuals and must be available.
- In many cases, the additional difficulties shown by children identified as having multiple disabilities may be only mild cognitive delays or learning problems similar to those recognized as learning disabilities in the hearing population. The options for such children will differ
significantly from those for children with more severe learning challenges and educational placement decisions should differ accordingly.

- Ongoing assessment of developmental progress is critical, so that placement and service decisions can be modified as needed if those initially chosen prove ineffective.

Approaches that focus on individual children and carefully track their progress over time and with different interventions are necessary to effectively support the development of most children who have multiple disabilities. Even interventions specific to an individual child can produce useful research-based information, however, if they are carefully designed, conducted using rigorous single-subject methods, and well documented. Such studies, as well as comparison studies when they are possible, will be increasingly necessary if programming efforts are to meet the needs of the majority of deaf and hard-of-hearing students.