

Chapter 1

**A LIFESPAN DEVELOPMENTAL APPROACH
TO ASSESSING AND ADDRESSING
NEURODEVELOPMENTAL DISORDERS:
A COMPREHENSIVE GUIDE TO MEETING
INDIVIDUAL AND CAREGIVER NEEDS**

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ABSTRACT

Individuals living with neurodevelopmental disorders (NDs) require substantial specialized supports and resources across the lifespan. Parents and siblings are typically at the forefront of caring for these individuals and ensuring that individualized medical and psychosocial needs are met. Within their roles as primary caregivers, numerous benefits and challenges are often encountered while working to promote the best quality of life possible for the entire family unit. Using a lifespan developmental approach, this chapter highlights the experiences of those caring for individuals diagnosed with NDs, with an emphasis on the unique needs of those acting as caregivers for individuals diagnosed with Smith-Magenis, Williams, or Down syndrome. Challenges associated with each predominant developmental period are explored, including methods of

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assessing specific challenges via psychological or neuropsychological evaluation. Example recommendations made to address these challenges are provided, and how primary caregivers can seek out resources aimed at addressing challenges is discussed. Advocating within educational systems, independent living and vocational planning, and overall family-oriented quality of life are emphasized. Given that siblings are frequently called upon to assume caregiver roles and financial responsibility as they age, distinct challenges within this relationship dynamic are considered. Legal concerns regarding guardianships and alternatives to guardianships are also highlighted.

Keywords: Neurodevelopmental disorders, Smith-Magenis syndrome, Williams syndrome, Down syndrome, caregivers, siblings

INTRODUCTION

“[Having a sibling with a neurodevelopmental disorder] has taught me to respect life the way it is made.”

12 year-old sibling of 5.5 year-old diagnosed with Down syndrome

Promoting the best quality of life possible for individuals with neurodevelopmental disorders (NDs) and their families requires significant time and attention to both individual- and family-based needs. For many of these families, substantial specialized care and resources are needed across the lifespan of the diagnosed individual. Parents and siblings are typically at the forefront of providing supportive care and ensuring that individualized medical and psychosocial needs are met. Within their roles as primary caregivers¹, numerous benefits and challenges are often encountered. Primary caregivers must learn to cope with these challenges in such a way that meets the child’s specific needs while also preserving the well-being of the entire family unit. Therefore, this chapter investigates the experiences of those caring for individuals diagnosed with NDs, with an emphasis on the unique lifelong needs of those acting as caregivers for individuals diagnosed with Smith-Magenis syndrome (SMS), Williams syndrome (WS), or Down syndrome (DS). Challenges associated with each predominant developmental period are explored, including those encountered during infancy, early childhood, middle childhood, adolescence, and adulthood. Methods of assessing specific individual and family needs via developmental, psychological, neuropsychological, and vocational evaluation will be discussed within the context of each developmental period. Example recommendations made to address identified challenges are presented, along with examples of how caregivers can seek out resources aimed at addressing those identified needs. Approaches to coping with developmental and behavioral concerns are highlighted. Common challenges related to educational systems, independent living, vocational planning, and overall family-oriented quality of life are emphasized. Given that siblings are frequently called upon to assume supportive roles and financial responsibility as they age, distinct challenges within this relationship dynamic are considered. Legal concerns regarding guardianship and alternatives to guardianships are also discussed.

¹ For the purposes of this chapter, the term “primary caregivers” or “caregivers” generally refers to parents and/or other legal guardians.

WHAT ARE NEURODEVELOPMENTAL DISORDERS?

"My daughter was just diagnosed today. It has been 32 years of trying to find out if there was some unifying reason for all of her differentnesses."

Parent of 32 year-old diagnosed with SMS

By definition, neurodevelopmental disorders (NDs) are a classification of disabilities that predominately affect the development and functioning of the nervous system (Reynolds and Goldstein, 1999). Multiple disorders have been classified as NDs, including intellectual disability, learning disorders, autism spectrum disorders (ASD), and attention-deficit/hyperactivity disorder (ADHD). Those diagnosed with NDs experience a variety of challenges, such as problems with speech and language, motor function, adaptive behaviors, emotional expression, memory, and age-appropriate learning. Although specific problem areas and needs often change across the lifespan, the majority of children who are diagnosed with a ND will continue to experience challenges as they age. Addressing these ever-changing needs can be quite difficult, and day-to-day management typically requires intensive time and commitment in the form of home- and school-based interventions, community-based therapies, and medication management. As expected, the totality of these stressors can place tremendous demands on primary caregivers and siblings.

HOW ARE NEURODEVELOPMENTAL AND GENETIC DISORDERS RELATED?

"Children with SMS require lots and lots of patience and understanding. They have mental disabilities that cause major behavior meltdowns and delayed learning."

Parent of a child diagnosed with SMS

"Now that I know that there is a legitimate medical reason for her delays, I am not so anxious for my daughter to "catch up" to an appropriate stage of development for her age. I am more relaxed knowing that she has her own timetable, and that she will grow and progress eventually."

Parent of a child diagnosed with SMS

In the United States (U.S.), approximately 12-13% of children between the ages of 3 and 17 are diagnosed with one or more NDs (Pastor and Reuben, 2009). Both medical communities and educators have observed what appears to be a genuine increase in the prevalence of children with NDs (Grupp-Phelan, Harman, and Kelleher, 2007; Kelleher, McNerny, Gardner, Childs, and Wasserman, 2000; U.S. Department of Education, 2007). Recent research suggests that the percentage of children diagnosed with such a disability will continue to rise, in part due to increased public awareness and modifications in diagnostic criteria (Froehlich, Lanphear, Epstein, Barbaresi, Katusic, and Kahn, 2007; Grandjean and

Landrigan, 2006; Newschaffer, Falb, and Gurney, 2005; Prior, 2003; Rutter, 2005; Visser, Bitsko, Danielson, Perou, and Blumberg, 2010).

Genetics likely plays a salient role in the etiology of NDs, and children diagnosed with genetic disorders are often found to have some type of ND as an expression of the genetic condition. Throughout the world, more than 7.6 million children are born with some type of severe genetic condition annually (World Health Organization, 2005), resulting in a large number of families who must learn to cope with related stressors, including medical problems, developmental delays, and/or intellectual impairments.

Approximately half of intellectual disabilities have a known genetic basis (Emery and Rimoin, 1990), with the expression of the disability typically being attributed to both genetic and environmental factors (Dykens, Hodapp, and Finucane, 2000). Based on the current DSM-IV-TR (American Psychiatric Association (APA), 2000) diagnostic criteria, an individual can be diagnosed with an intellectual disability if there is evidence of impaired intellectual functioning prior to age 18 and if they also demonstrate deficits in adaptive functioning. Impaired intellectual functioning is typically defined as having an intelligence quotient (IQ) of approximately 70 or less, based on a standardized, well-validated intelligence assessment. Adaptive functioning refers to daily living skills, such as communication, self-care behaviors, independent living, or social/interpersonal skills (American Association of Intellectual and Developmental Disabilities, 2012; Schroeder, 2000). The DSM-IV-TR further categorizes intellectual disabilities into four sub-groups, including 1) mild (i.e., intellectual quotient of 50-55 to approximately 70), 2) moderate (i.e., IQ of 35-40 to 50-55), 3) severe (i.e., IQ of 20-25 to 35-40), or 4) profound impairment (i.e., IQ below 20-25; Greenberg et al., 1996; Martens, Wilson, and Reutens, 2008; Meyer-Lindenberg, Mervis, and Berman, 2006). The majority of individuals with Smith-Magenis, Williams, or Down syndromes, the primary foci of this chapter, will have IQs falling in the range of mild to moderate intellectual impairment.

Of note, the term “intellectual disability” is the preferred term utilized among disability sectors when describing intellectual impairments. However, the term “mental retardation” continues to be utilized, most often in law and public policy sectors. It is anticipated that the term “mental retardation” will be largely replaced with “intellectual disability” or “intellectual developmental disorder” when the DSM-5 is released in 2013 (Comer, 2013). In addition, “mental retardation,” which is now categorized in the grouping of “Disorders Usually First Diagnosed in Infancy, Childhood, or Adolescence” will be listed in a new grouping called “Neurodevelopmental Disorders” along with ADHD, learning disorders, communication disorders, coordination disorders, and autism spectrum disorders. Other commonalities among both neurodevelopmental and genetic disorders, such as problems with learning and attention, will be discussed elsewhere in this chapter.

DIAGNOSING SMITH-MAGENIS, WILLIAMS, AND DOWN SYNDROMES

This chapter provides a broad overview of individual, primary caregiver, and family needs among those coping with NDs. Discussion topics will include challenges that often emerge across specific developmental periods, perceived family benefits that result from the

disorder, and available resources and services aimed at assisting individuals and families coping with NDs. In addition to this broad overview, special attention will be paid regarding the needs of those living with and supporting individuals diagnosed with Smith-Magenis syndrome (SMS), Williams syndrome (WS), and Down syndrome (DS).

Smith-Magenis Syndrome

“I feel so very blessed to have my brother in my life. His bright spirit and his sweetness - even in the face of overwhelming obstacles he faces daily - are a constant inspiration to me. My relationship with him has made me a better person - a more loving, patient, kind and happy person. My relationship with my developmentally-typical siblings is closer because of the bond that we share in having a brother with special needs. My brother is an essential part of my family and of my life. And while there certainly have been challenges (I don't think I slept a full night once during my high school years), I can honestly say that I wouldn't trade my experience of being his sister for anything in the world.”

26 year-old sibling of 13 year-old with SMS

Smith-Magenis syndrome (SMS) is a chromosomal disorder caused by either a deletion of a portion of chromosome 17 or a mutation of the *RAI1* gene, which lies within the SMS chromosome 17p11.2 region (Elsea and Girirajan, 2008; Smith et al., 2006). This syndrome affects approximately 1 out of 15,000 to 1 out of 25,000 live births worldwide and is typically a sporadic syndrome that is not inherited from a parent. Individuals are affected equally across genders and racial groups. The syndrome is characterized by physical, developmental, and behavioral features (Smith et al., 2006). Common features among those diagnosed with SMS include craniofacial anomalies, a hoarse voice, feeding problems in infancy, low muscle tone, developmental delays, including early speech and language problems, intellectual disability, decreased pain sensitivity, chronic sleep disturbances, hyperactivity, arm hugging, hand squeezing, attention problems, emotional lability, and self-injurious behaviors that include skin picking and nail yanking (Elsea and Girirajan, 2008; Slosky, Foster, and Elsea, 2012; Smith et al., 2006). Intellectual functioning typically falls within the mild to moderate range of intellectual disability, with a mean full scale IQ score of 47 (range: 20 to 78, Greenberg et al., 1996). Specific intellectual weaknesses are often seen on sequential processing tasks; however, relative strengths are observed in areas of acquired knowledge and reading (Dykens, Finucane, and Gayley, 1997). Medical problems also commonly exist, including congenital heart defects, seizure disorders, peripheral neuropathy, scoliosis, vision problems, and urinary tract anomalies (Edelman et al., 2007; Smith et al., 2006).

SMS was first documented in the literature in the early 1980s (Smith, McGavran, Waldstein, and Robinson, 1982). Diagnostically, SMS may first be suspected based on phenotypic appearance and behaviors. The small chromosomal deletion indicative of diagnosis was often overlooked until fluorescent in situ hybridization (FISH) became more readily accessible (Dykens et al., 2000). Comparative genomic hybridization (CGH) microarray is now commonly utilized to verify a suspected SMS diagnosis.

Williams Syndrome

“It is a privilege to live with someone that has Williams syndrome because you grow a new-found respect for all the different people we live with day to day.”

14 year-old sibling of 7 year-old with WS

Williams syndrome (WS) is a rare ND caused by a hemizygous contiguous gene deletion of the 7q11.23 region of chromosome 7 (Peoples et al., 2000). This region of chromosome 7 spans approximately 28 genes, including the elastin gene (*ELN*), which is deleted in over 95% of individuals with WS (Schubert, 2009). This syndrome affects 1 in 7,500 to 1 in 20,000 people worldwide, with equal rates across genders and racial groups. As with SMS, it is a sporadic syndrome typically not inherited from a parent (Schubert, 2009; Stromme, Bjomstad, and Ramstad, 2002). WS can be conceptualized as a multi-system disorder with distinct physical, developmental, and social features and medical complications. Common features include intellectual disability and learning difficulties, developmental delays, craniofacial anomalies, low birth weight, and feeding problems (Burn, 1986). Physically, people with WS have a distinct appearance with a wide mouth, full lips, full cheeks, short upturned noses, periorbital fullness, broad brows and foreheads, and curly hair (Martens, Wilson, and Reutens, 2008). Common medical concerns include supra-aortic stenosis, hypertension, hypercalcemia, vision, hearing, and dental problems, glucose intolerance, subclinical hypothyroidism, early onset puberty, gastrointestinal problems, and genitourinary tract problems (Burns, 1986; Cambiaso et al., 2007; Pober, 2010; Stagi et al., 2005; Wessel et al., 2004). Socially, WS is characterized by a unique interpersonal demeanor, and individuals are often described as having a “cocktail party” personality, defined as being overly friendly, highly sociable, and displaying high degrees of empathy (Schubert, 2009). Despite being exceedingly friendly and empathetic, those with WS also demonstrate a poor understanding of social relationships and boundaries, making it difficult to develop and maintain friendships. Cognitively, individuals with WS show a broad range of intellectual ability, with an average IQ of 50-60 (range: 40 to 100), in the range of mild to moderate intellectual disability (Martens et al., 2008; Meyer-Lindenberg et al., 2006).

Verbal abilities tend to be strong, but weaknesses exist with respect to visuospatial processing and reasoning (Martens et al., 2008). People with WS often show a strong affinity towards music, display strong emotional reactions towards music, and are noted to display creativity in their musical abilities (Martens et al., 2008).

Children with WS are usually first identified by their characteristic facial appearance (Burns, 1986). FISH testing can be utilized to verify the diagnosis, as the chromosome deletion is usually too small to visualize on karyotype (Borg, Delhanty, and Baraitser, 1995; Elcioglu, Mackie-Ogilvie, Daker, and Berry, 1998). However, testing may also be done by CGH microarray, which is the current recommended methodology.

Down Syndrome

“If only the world was as simple in their perspective and as pure as a kid with Down syndrome, the world would be a better place. We could learn a lot from them all.”

15 year-old sibling of 10 year-old with Down syndrome

Down syndrome (DS), or trisomy 21, is a chromosomal condition caused by an extra copy of chromosome 21. In 94% of cases of DS, there is full trisomy; 2.4% of cases are mosaic trisomy, and in 3.3% of cases there is a translocation involving chromosome 21 (Jones, 2006). The syndrome affects approximately 1 in 800 births worldwide, with a slightly increased incidence of DS in males as compared to females, with the ratio being 1.15 to 1 (Canfield et al., 2006). Individuals with DS show distinct physical, developmental, and social features. Cognitively, people with DS have an IQ in the range of 35 to 70, or the mild to moderate range of intellectual disability (Jones, 2006). Overall, people with DS have a good rhythm and enjoy music. Although delayed, social functioning tends to be approximately three years more advanced than mental age (e.g., if intellectual functioning is at a 6 year-old level, social functioning is at an approximately 9 year-old age level). Physical features of individuals with DS may include short stature, epicanthal folds, hypotonia (i.e., poor muscle tone), single transverse palmar crease, protruding tongue, small ears, and a short appearing neck. Around 50% of children with DS are born with a congenital heart defect. People with DS may experience gastrointestinal problems such as Hirschsprung disease, which can cause chronic constipation, and urinary tract abnormalities (Jones, 2006). Persons with DS experience accelerated aging with initial signs of aging typically beginning in the fourth decade of life; 60 to 75% of individuals with DS will eventually develop Alzheimer disease. Leukemia is also more common in individuals with DS (Wiseman et al., 2009).

DS is typically identified at birth based upon physical appearance. A standard peripheral blood karyotype will identify the trisomy 21, including translocations and other variants involving chromosome 21 that may be associated with DS. Many individuals may be identified prenatally. Parental screening tests, including ultrasound and maternal serum screening, may lead to diagnostic testing, such as chorionic villus sampling (CVS) or amniocentesis, which identify DS through karyotypic analysis of cells from the placenta or fetus.

Comparison and Summary of Syndromes

Individuals with SMS, WS, and DS all demonstrate unique physical, developmental, and social traits. However, the extent to which individuals will be affected by the specific features of their syndromes varies both across and within the diagnoses. For example, in comparison to SMS, individuals with WS or DS tend to have a good-natured disposition and demonstrate fewer behavioral concerns. However, individuals with WS and DS tend to have more medical complications than individuals with SMS. In addition to medical concerns, individuals with WS display high incidences of psychiatric conditions, including anxiety and depression. These psychiatric problems often progress with age (Schubert, 2009) and are not seen as frequently among those with SMS or DS. One unique psychiatric concern seen in individuals

with WS is auditory allodynia, which is a substantial aversion to, or fear of, certain sounds that are not typically found aversive (Levitin, Cole, Lincoln, and Bellugi, 2005). With respect to cognitive functioning, individuals with SMS, WS, and DS all function within the mild to moderate range of intellectual disability. The specific pattern of cognitive strengths and weaknesses may vary by diagnosis.

Regardless of the specific syndrome, the complexity of these medical, developmental, intellectual, and social concerns, as well as the varied presentation, requires significant medical and psychosocial management by an integrated multidisciplinary team of highly trained professionals. While these teams include physicians and other medical professionals, families coping with NDs such as SMS, WS, and DS, benefit from psychosocial support teams, including case managers, psychologists, social workers, and educational advocates, as well as other ancillary providers such as physical, occupational, and speech/language therapists. The services required vary depending upon specific individual and family needs, which often change across the developmental lifespan.

CHALLENGES, ASSESSMENTS, AND INTERVENTIONS: INFANCY AND EARLY CHILDHOOD

“It has given us a greater appreciation for the value of the lives of those who society may not see as perfect. We have all grown in patience, love, understanding, and compassion, and we wouldn't trade it, as challenging as it is.”

Parent of a 7 year-old without WS and a 4 year-old with WS

“I feel very ill equipped to handle the aggressive and impulsive behaviors of our son with WS. And these behaviors cause me consistent and extreme stress. We have yet to find professionals (doctors, therapists, teachers, psychologists, etc.) that are able to truly provide us with ways to help our son in these critical areas. It is quite discouraging as a caregiver to realize that the "professionals" are at a loss as to how actually help my child and they are supposed to be the ones with the education, experience, expertise, etc. I can explain the behaviors to my spouse, my friends, medical professionals, etc. but when it is you dealing with the same stuff every day from this little person who does not seem to be improving, it is a heavy weight to carry daily.”

Parent of a child diagnosed with WS

Developmental Challenges

Children with NDs often face numerous challenges that emerge as early as infancy and/or early childhood. Common concerns in this developmental period include problems related to sleep and feeding, maladaptive behavior, poor motor development, and delays in speech and language development.

Sleep, Feeding, and Behavioral Concerns

Sleep-related concerns can be tremendous barriers to wellness among families coping with NDs. This is especially true among individuals and families coping with SMS. Those with SMS usually experience nocturnal wakefulness and persistent daytime hypersomnia, shortened sleep cycles, problems falling asleep, and reduced periods of rapid eye movement (REM) sleep (Smith, Dykens, and Greenberg, 1998b). Research has demonstrated that these sleep problems are highly detrimental to caregiver and family well-being (Hodapp, Fidler, and Smith, 1998). Moreover, families report a greater number and intensity of maladaptive behaviors when sleep disturbances increase (Dykens and Smith, 1998). Sleep problems are also present among individuals with WS. Specific problems often include significant bedtime resistance and periods of wakefulness throughout the night (Annaz et al., 2010).

Feeding problems are also not uncommon among individuals diagnosed with NDs. For example, children with WS often experience both reflux and constipation in addition to atypical weight gain and abdominal pain of unknown etiology (Poher, 2010). They are also at risk for hypercalcemia, which can lead to decreased feeding, irritability, and colic (Cooper-Brown et al., 2008). Similar feeding and digestive problems are observed in children with SMS and DS (Smith et al., 2012). For example, infants with DS often experience oral motor and dental problems (Cooper-Brown et al., 2008). These problems collectively contribute to feeding difficulties, which may include aversions to certain food textures, chewing and swallowing difficulties, choking, hypotonia, and poor motor coordination, which can limit self-feeding behaviors. The severity of feeding problems may lead children with these disorders to be labeled as failure to thrive (FTT), and the chronicity of these problems may not resolve without intensive feeding interventions (e.g., feeding clinics, modified feeding schedules, feeding tubes).

Significant maladaptive behaviors, which are observed somewhat frequently among individuals with NDs, can create high levels of stress among caregivers and families, which, in turn, can negatively impact both individual and family functioning. Individuals with SMS typically display more maladaptive behaviors than those with WS or DS. Among individuals with SMS, these maladaptive behaviors often include problems with aggression and self-injury, including self-biting and nail-pulling (Colley, Leversha, Voullaire, and Rogers, 1990; Stratton et al., 1986). Disobedience, tantrums, stubbornness, destruction of property, enuresis, and encopresis are also common problems (Dykens and Smith, 1998). A study of 19 children with SMS found that, on average, the children displayed 13 different stereotypic behaviors, including self-hugging, teeth grinding, hand flapping, and repetitive rocking (Martin, Wolters, and Smith, 2006). Based on parent report, the children also displayed an average of five self-injurious behaviors such as self-hitting and/or self-biting, headbanging, skin picking, and insertion of fingers or objects into body orifices other than the mouth.

Motor Development and Speech and Language Concerns

Problems with motor and/or speech and language development are also common among children with NDs, including those with SMS, WS, or DS; however, the etiology and expression of these difficulties vary by diagnosis. For example, a study of 11 children with SMS, ages 5 to 34 months, found moderate to severe delays in both motor functioning and expressive language acquisition (Wolters et al., 2009); however, receptive language was less severely affected (Smith et al., 2012). Results showed that toddlers (ages 24 to 34 months) demonstrated more significant deficits than infants in these domains, suggesting that these

delays become more apparent over time. Problems with motor and speech and language development have been attributed to generalized cognitive deficits, as well as to hypotonia, oral-motor abnormalities, and middle ear dysfunction. Neuropathies in the upper and lower extremities may also play a role in the motor delays experienced by children with SMS, as well as contributing to self-injury.

Children with WS have delays in both fine and gross motor development, due to problems with musculoskeletal development (Burns, 1986; Udwin and Yule, 1991). One study assessing psychomotor development in young children with WS (less than 42 months of age) found severe delays in motor functioning in the majority of children as compared to typically developing children, prompting researchers to recommend occupational and physical therapies (Tsai, Wu, Liou, and Shu, 2008). Visuospatial deficits have also been reported among children with WS, which further contribute to problems with tasks requiring fine motor coordination (Nakamura et al., 2009). Language acquisition can be significantly delayed as well, with expressive language skills developing at approximately three years of age (Mervis et al., 2000).

Children with DS are also at risk for motor and/or speech and language delays as a function of impaired cognitive functioning, hypotonia, and craniofacial abnormalities that often include a protruding tongue (Canfield et al., 2006; Jones, 2006). Language deficits tend to be more significant than general cognitive, motor, or social deficits, although deficits often exist in all of these domains (Clibbens, 2001; Fowler, 1990; Miller, 1992; Rondal and Comblain, 1996; Tager-Flusberg, 1999). In terms of language development, specific problems with morphology and syntax have been identified, with related deficits present well into adulthood. However, research results have varied widely when exploring language development among children with DS. Specifically, some research findings suggest that nonverbal cognitive functioning and receptive language development tend to be more advanced than expressive language development (Miller, 1999). Others have found no differences between nonverbal and verbal cognitive ability, while further research has shown delays in both receptive and expressive language.

Early Child Interventions Programs and Developmental Assessments

Fortunately, a number of interventions are available to help children and families meet the wide range of developmental needs that are present in SMS, WS, and DS. Identification of specific areas of strength and weakness is important when designing individually tailored interventions. This process typically begins with a developmental assessment, which can be utilized to evaluate children from birth through ages 5 to 6. Most children with complex genetic disorders, who are known to be at risk of significant neurodevelopmental delays, will qualify for such an assessment. Developmental assessments are usually conducted by a psychologist, psychological examiner, or neuropsychologist, although additional aspects of developmental assessments may be completed by developmental pediatricians, speech and language pathologists, occupational therapists, or physical therapists. It is becoming more common for developmental assessments to be completed in interdisciplinary care clinics that include some or all of these specialty services, and they are often completed through state-run early intervention programs, in hospitals, and in other community-based settings. Primary caregivers who bring their children to a developmental assessment can expect to participate in

an intake interview, during which they will be asked to provide background information about their child's medical and developmental history, and to discuss their ongoing concerns. Clinicians will typically make note of behavioral observations while engaging the child in a formal, standardized assessment of their development in the areas of early cognitive ability, receptive and expressive language, fine and gross motor functioning, and social skills.. Depending on the primary presenting concerns and the age of the child, the evaluation may also include assessments of pre-academic skills, behavioral and emotional functioning, and/or attention and impulsivity. Each child's performance on these measures will be compared to what would be expected given the child's age, based on normative data. Once the assessment is complete, a report will be written and provided to the caregiver(s). A typical report will consist of an overview of the child's medical, developmental, family, and social history, a detailed description of the assessment results, a summary of results with any associated diagnoses, and a list of recommendations and resources aimed at helping the primary caregiver(s) meet both the child and family's needs.

Examples of Recommendations Found in a Developmental Assessment

Clinicians are able to provide a variety of recommendations following the completion of a developmental assessment, and these are typically listed within the report that caregivers receive. These recommendations should be specific to each child's age, developmental needs, cultural and language background, and make note of available services and resources in the community. Although not an exhaustive list, below are examples of recommendations that may be provided for an infant who is found to have cognitive and/or neurodevelopmental delays:

Referral for Early Intervention Services

- 1) The child's parent(s) are encouraged to share the results of this assessment with Early Intervention Services. Early Intervention Services can assist in setting up appropriate local services and therapies for children under the age of 3 years.

Recommendation to Participate in Occupational Therapy, Physical Therapy and/or Speech and Language Therapy

- 2) It is strongly recommended that the child (and family) participate in occupational therapy, physical therapy, and speech and language therapy. Referrals for each of these services can be provided if desired by the family. For children in bilingual families, it is further recommended that these services be completed with a bilingual therapist.

Recommendations to Address Fine and Gross Motor Delays

- 3) The following recommendations are provided to the child's parent(s) to address his/her delays in motor development. These activities can be incorporated into the child's play activities within the home environment.
 - a. Engage the child in activities that require him/her to sit independently.

- b. Encourage the child to play on the floor in a prone position (i.e., on his/her stomach) so that he/she builds strength in his/her arms and legs. This position will also encourage neck strength, fine motor control as he/she reaches for toys, and gross motor control, which will help him/her learn to crawl.
- c. Encourage the child to stand and bounce while holding onto furniture or adults for support. This will help him/her develop leg strength and balance.
- d. Play hide-and-seek games that encourage the child to find toys hidden under washcloths or cups to develop fine motor control.
- e. While reading stories together, encourage the child to turn the pages of the books.
- f. Fine motor skills can be practiced in conjunction with self-care behaviors, such as feeding himself/herself. In addition to allowing the child to hold his/her own bottle, practice using sippy cups. He/she can also practice using a pincer grasp to pick up cereal or grasping a spoon while eating rice cereal or other table foods.

Recommendations to Address Speech and Language Delays

- 4) The following recommendations are provided to the child's parent(s) to promote language development. The parents are encouraged to consistently integrate these activities into the child's daily routine.
 - a. Encourage the child to mimic words and word sounds in his/her environment.
 - b. Begin to teach the child the words for common objects in his/her environment, as well as the names for colors and body parts. He/she will learn how to understand these words first and then learn how to say the words as he/she ages.
 - c. Use songs to practice words, numbers, and counting.
 - d. Pair nonverbal gestures with words in the child's environment. For example, point to objects when talking about them so that he/she learns to associate words with the objects.
 - e. Spend time reading to the child. Infants enjoy brightly colored books that include simple language and nursery rhymes. Cloth books and "touch and feel" books are also recommended.
 - f. Although television time should be limited, allowing the child to utilize television programs for learning activities can help promote his/her language development. His/her parent is encouraged to work with him/her to understand and express words when watching television.
 - g. Once the child has mastered the naming of common objects, encourage his/her use of words when making requests. Require the child to attempt to use words before providing him/her with the desired object, and praise him/her for effort.
 - h. For children with severe problems mastering spoken language, sign language, picture exchange programs (e.g., Picture Exchange Communication Systems), and/or augmentative assistive language devices should be utilized. There are a number of such devices available, including many low cost iPad and tablet applications that can be downloaded. Utilization of these resources can facilitate adaptive verbal and non-verbal communication and decrease the frequency of behavioral outbursts and self-harming behaviors.

Recommendations to Address Feeding Problems

- 5) For picky eaters and infants who are reluctant to try new foods, take things slowly and be patient, choose feeding times when the child is feeling well, happy, and not too hungry, expect messes and distractibility, keep mealtimes fun, and focus on the positive while feeding the child. When an infant or young child refuses a new food, wait a few weeks before trying it again and consider mixing it with a preferred food.

Recommendations for a pre-school aged child who is found to have cognitive and/or neurodevelopmental delays *may* include some or all of the following:

Implementation of 504 Plan or Individualized Education Plan (IEP)²

- 1) The child's parents are encouraged to share the results of the current evaluation with appropriate school personnel so that they may be considered in the development of a 504 Plan or an Individualized Education Plan (IEP). Results of this evaluation indicate that the child is exhibiting deficits that will require classroom accommodations and curriculum modifications to be successful in school.

Recommendations for Therapy Services to Address Behavioral Concerns

- 2) The child and family would benefit from participating in outpatient and/or home-based individual/family-based therapy to help address communication difficulties, oppositional behaviors, aggression, and self-injurious behaviors. Applied behavior analysis, parent-child interaction therapy, and other evidence-based parenting strategies may be especially helpful in addressing these concerns. An in-home therapist may be especially beneficial in setting up consistent expectations for the child within the home and helping to create cohesive behavioral expectations between home and school.

Recommendations for Address Behavioral Concerns in the Home

- 3) Specific behavioral interventions can be applied in the home to promote the child's social and emotional well-being. The following strategies and techniques are recommended:
 - a. Maintain predictable classroom and home routines; keep the daily schedule as consistent as possible. A schedule builds a sense of safety, reduces anxiety, sets reasonable expectations, and breaks things down into manageable components. Keeping the child on a routine will help him/her manage his/her behaviors and give him/her the best chance to succeed. Create visual schedules for the child so that he/she may be reminded of "what comes next" throughout the day.
 - b. Activities that require the child to sit still should not be more than 15 to 20 minutes in length. After 15 to 20 minutes, allow the child to get up and move around before returning to the task he/she needs to complete.
 - c. Before addressing the child, speak his/her name and make eye contact; keep oral directions short and uncomplicated. Ask him/her to repeat the directions or demonstrate that he/she understands what is required.

² See the section entitled *Advocacy, Psychoeducational Assessments, and Academic Accommodations* for more information on IEPs and 504 Plans.

- d. Actively ignore minor, undesired behaviors that are not harmful to oneself or others. Parents often unintentionally provide children with attention for inappropriate behavior, which can inadvertently increase the likelihood that the child will engage in that same inappropriate behavior in the future. Therefore, it is important to ignore, minor negative behaviors, while at the same time, providing specific praise and attention when the child engages in appropriate, positive behavior.
- e. When agitated, have the child move to a separate area away from peers and siblings (in a different room close by if possible). Many children are able to calm themselves if given the opportunity and materials. Allowing the child to engage in activities that require organization and repetition may help the child calm down. Doing puzzles, counting, singing, listening to music or a story, or writing letters and shapes repetitively may help facilitate this process. These strategies should only be used when the child has become overwhelmed or upset, and not when the child has misbehaved.
- f. Try to identify the child's strengths that can be publicly announced or praised. When praising the child, label specific aspects of what is being done well, and use brief statements. For example, rather than saying, "Great job!" say "I really like it when you sit nicely and eat your lunch!" or "Good job saying 'thank you'!"
- g. Try to offer immediate reinforcement for appropriate behavior (i.e. stickers for chart, free-time, etc.). Waiting too long for rewards may lose the desired effect.
- h. When the child is having a tantrum, ensure that he/she is safe (e.g., move him/her away from hard floors, walls, or any dangerous objects), but do not engage with him/her in any other way (e.g., do not talk to him/her or provide other forms of attention). He/she is likely using self-injurious behaviors as a form of communication and control. Engaging with him/her while he/she is participating in such behaviors will only prolong the behaviors. As the child begins to understand that tantrums and self-injury no longer produce their desired outcomes, the behaviors will reduce in frequency and severity.
- i. When the child is aggressive with others or property belonging to others, remove him/her from the situation whenever possible. Follow through with consequences such as time-outs or removal of fun activities. Do not interact with him/her while he/she is in time-out. If he/she gets up from his/her time out location, simply bring him/her back without speaking to him/her. When the time out is over, give him/her a one sentence explanation of why he/she was punished and have him/her repeat the explanation to ensure that he/she understands. If a time-out is assigned due to the child's oppositional or defiant behavior, have the child follow the original instruction and assign a second time-out if he/she again fails to comply.

Recommendations to Address Fine Motor Delays

- 4) The following recommendations are provided to the child's parent(s) to address his/her fine motor difficulties. These activities should be incorporated into the child's play activities within the home environment.
 - a. Provide the child with a variety of writing materials (e.g. pencils, colored pencils, crayons, paint brushes) to strengthen his/her basic writing skills.

- b. Model the appropriate technique for gripping a pencil and how to position the paper when writing.
- c. Provide the child with supplies that will aid him/her in appropriately gripping his/her pencil (e.g., oversized pencils, pencil grips).
- d. Engage the child in fine motor play that includes tracing, coloring within the lines, stringing beads, lacing cards, building objects with blocks or Legos, putting puzzles together, and forming objects with play dough.
- e. Encourage the child to correct or rewrite poorly formed letters; monitor his/her practice to ensure letters are formed correctly.
- f. For some children, fine motor deficits may be severe. In such cases, expecting the child to engage in traditional written communication may not be realistic. Continuing to insist that the child participate in challenging fine motor tasks may result in significant frustration, tantrums, and other aggressive behaviors. In such cases, it is recommended that the child be encouraged to utilize tablets or other assistive devices. These skills can promote learning, decrease disruptive behaviors, and improve overall quality of life. Typing can also be introduced as the child ages.
- g. Reinforce his/her successful efforts when engaging in traditional writing activities and/or utilization of technology-based assistive devices to participate in written communication. Provide corrective feedback as needed.

Recommendations to Address Gross Motor Delays

- 5) The following recommendations are provided to the child's parent(s) to address his/her gross motor difficulties. These activities can be incorporated into the child's play activities within the home environment.
 - a. Ride tricycles or scooters. Be sure to have the child wear a helmet and other protective pads to protect the child and teach good self-care behaviors.
 - b. Practice kicking and throwing balls, pushing carts, and pulling wagons.
 - c. Participate in obstacle courses and relay races.
 - d. Walk across balance beams, jump on trampolines, and climb and swing on playground equipment.
 - e. Dig in the dirt and plant gardens.

Recommendations to Address Speech and Language Delays

- 6) It is recommended that the child's family and school personnel actively work on his/her conversational skills to increase the likelihood of communication success and competence. Given the child's sporadic use of phrases and sentences to obtain items and activities, and his/her limited ability to echo the speech of others, it is recommended that his/her family members learn skills to improve his/her independent speech. The most effective method for teaching independent speech is by teaching a child to request desired items. There are several steps in the progression toward independent requesting.
 - a. First, a parent must know the items and activities that are desired by the child. These may include edible items such as chips, juice, cookies, peanuts, or crackers. These may also include items or access to activities such as watching a cartoon, playing with a musical toy, mom singing, or throwing a ball back and forth.

Anything that your child enjoys can be used to teach requesting. The more items and activities that are available, the more opportunities to teach requesting there are.

- b. The next step is to restrict access to the most desirable items. This does not mean putting all of the child's toys away. It simply means putting his/her favorite toys out of reach so that it creates a motivation for the child to ask for the item. If the child is able to go and get desired items by himself/herself, he/she has no reason to communicate with you. However, if he/she needs the parent to get things for him/her, he/she is more likely to communicate verbally. Favorite toys, videos, snacks, and other items should be put out of reach but in sight.
- c. Once the child is unable to get things on his/her own, the parent should follow the child until he/she indicates that he wants something. The child may indicate by pointing, pulling you to the item, crying, or glancing at the item. If the parent knows what the child wants, he/she should say the name of the item and pause for the child to repeat it. If the child does not repeat the name, say the name once more and then deliver the item anyway. Do not withhold items for too long or the child may become frustrated and learn that talking is too difficult. The goal is for the child to learn that talking is a good way to get his/her needs met. With snack items, the parent should give the child only a small portion. By giving only a small amount, the child will need to come back to ask for more and so there is another opportunity to teach the child to ask. If the parent gives the child a full glass of juice instead of only a few sips, then he/she may become full and not want anymore. Therefore, only give small portions to increase the number of opportunities to teach.
- d. With activities, only allow a small amount of time with the items. Try not to take things away, but when the child puts a toy down, the parent should pick it up. Then, when he/she wants it again, the parent has it, and there is another opportunity to teach him/her to ask. Always say the name of the item and pause to see if he/she will repeat what the parent said before giving the item or snack. Requesting items and activities is best done in the context of playtime.
- e. Games can also be played that promote language acquisition.
 - i. Sing songs using preschool tapes or videos.
 - ii. Play "what is it?" games by putting objects in a bucket and labeling the objects as they are pulled from the bucket.
 - iii. Play games such as I Spy, Pictionary, or acting games that require verbalization.

Recommendations to Address Sleep Concerns

- 7) The child's parents are encouraged to speak to a pediatrician concerning his/her sleep difficulties, as there may be behavioral interventions that can help him/her to fall asleep. It will be important to implement a consistent sleep routine (e.g., bath, then story, then lights out at 8:30 every night). Suggestions for establishment of this routine include slowly weaning his/her daily naptime, along with waking him/her up earlier in the morning. As much as possible, allow the child to help determine a plan that will make him/her more comfortable at night and set up a consistent, calming

sleep environment (e.g. getting a soft blanket, putting pictures of the family beside his/her bed, selecting soothing music to listened to before bed, etc.).

- 8) Encourage the child to fall asleep independently by putting him/her to bed when drowsy, but still awake. If the child avoids bedtime, set consistent limits, and if the child gets out of bed during the night, matter of factly walk him/her back to his/her bed.
- 9) Specific sleep concerns related to SMS should be addressed with a pediatrician and sleep specialist. Sleep phase shift programs may be successful in creating and sustaining a more normative sleep schedule. Medications such as melatonin and other modifications such as bright light therapy may be required to sufficiently address sleep concerns in this population.

Recommendations to Address Feeding Problems

- 10) For young children who exhibit behavioral problems during mealtimes, try to conceptualize the mealtime as an opportunity to teach the child appropriate behaviors and manners. Establish a routine that includes sit-down, family style meals, and keep the television turned off. Set reasonable time limits for meals and provide only small portions of preferred foods to encourage the child to expand his/her diet. Avoid “clean your plate” rules that may lead children to develop unhealthy eating habits, and include the child in conversation and social interaction during the meal to make it enjoyable.
- 11) Establish a set of mealtime rules, starting with only 2 or 3. Remind the child of the rules before each meal and provide specific praise for appropriate mealtime behavior. Have the child practice the correct behavior the first two times he/she breaks a rule and reserve time-out for the third time the rule is broken. Avoid providing the child with snacks in between meals if he/she did not eat appropriately at mealtime.

Recommendations to Address Development of Self-Care Behaviors

- 12) The following recommendations are provided to the child’s parent(s) to promote development of self-care behaviors. They are encouraged to consistently integrate these activities into the child’s daily routine.
 - a. Encourage the child to practice self-care behaviors such as getting dressed and putting on his/her own shoes. Be patient with him/her as these activities will likely take a significant amount of time at first.
 - b. Practice allowing the child to button his/her own buttons and zip his/her own zippers. This will help improve fine motor skills as well.
 - c. Assist the child in learning appropriate bathroom hygiene, such as washing his/her own hair, brushing his/her own teeth, and taking care of his/her own toileting behaviors. To ensure cleanliness, it may be helpful to allow the child to attempt these behaviors independently at first and then assist him/her as necessary.
 - d. Set expectations for the child to complete simple household chores such as putting away his/her own toys, putting his/her jacket in the closet, helping set the dinner table, or putting his/her laundry in the hamper. Setting these expectations now will help the child develop independent living skills as he/she ages.

- e. Use positive reinforcement (e.g., praise, small rewards) to encourage the child to engage in self-care behaviors.

Promoting Social Skill Development

- 13) The following recommendations are being made to promote the child's social skill attainment.
 - a. Model the appropriate use of manners (e.g., saying please and thank you, refraining from interrupting others during conversations, sharing toys and games, raising his/her hand to ask a question in class).
 - b. The child may also benefit from learning simple scripts for initiating play, such as "That looks like fun. Can I play too?" or "Would you like to play with these blocks with me?" Such scripts could be practiced at home and school, and the child should be prompted to use these phrases when he/she is around other children.
 - c. Play activities should include activities that require turn-taking and one-on-one interactions. Examples include throwing a ball back and forth or playing games such as Candyland or Chutes and Ladders that have simple rules and require at least two people to play. Other activities may include racing cars side by side, playing with puppets, or building or making something with manipulatives.
 - d. Model and practice pretend play with the child. This could include any number of activities such as playing dress-up or house, pretend cooking, playing pirates or dinosaurs, or pretending to be superheroes.

Recommendations to Promote Toilet Training

- 14) The following recommendations are provided to the child's parent(s) to promote toilet training.
 - a. Determine whether the child is ready to be toilet trained. The child must achieve bladder control prior to initiating toilet training. Signs of bladder control include remaining dry during short (i.e., 30 minute) naps, urinating all at once rather than "dribbling" throughout the day, and engaging in facial expressions suggesting that the child needs to urinate or is urinating.
 - b. Recognize that toilet training is a multi-step process that is more manageable if broken down into smaller steps. Focusing on mastering one step at a time will increase the child's chances of success.
 - c. The child is more likely to eliminate on the toilet once he/she feels comfortable sitting on the toilet to potty chair. The child will feel more comfortable if this activity is perceived as being enjoyable. Reading stories or singing toileting songs that are only accessible when sitting on the toilet or potty chair can reinforce the child's desire to use the toilet. Use praise or small rewards such as stickers, hugs, or M&M's to encourage these behaviors.
 - d. After the child is comfortable sitting on the toilet or potty chair, setting up a "sitting schedule" can increase the chances of successful elimination. Set up the schedule around the times that the child is most likely to naturally want to eliminate such as immediately after waking, 30 minutes after a meal, shortly after active play, and/or after bathing. On days when the primary caregiver is home with the child, the caregiver is encouraged to provide the child with a favorite

beverage to help ensure the need to eliminate during scheduled times. When the child successfully eliminates on the toilet or potty chair, be sure to use lots of praise. Other small rewards may also help reinforce this behavior.

- e. Teach the child to ask to utilize the toilet or potty chair by requiring that the child ask permission prior to going into the bathroom. The parent can prompt the child to do one of the following: 1.) make the sign for potty 2.) Say “potty,” or 3.) Say, “I need to go to the potty.” Praise the child for asking, and then immediately take him/her to the toilet or potty chair.
- f. If the child continues to demonstrate difficulties transitioning out of diapers, it may be helpful to have the child wear underpants under the diaper. This will help the child sense when he/she is wet, thus increasing discomfort and the chance that the child will want to eliminate on the toilet or potty chair.

Recommendations to Promote Pre-Academic Skill Development

- 15) The following recommendations are provided to the child’s parent(s) to strengthen pre-academic skills development. They are encouraged to use these techniques at home, perhaps incorporating them into play.
 - a. Practice identifying numbers and shapes using age appropriate games and books.
 - b. Integrate learning opportunities into daily activities. For example, have the child count objects in the grocery store, identify common road signs while driving, identify left and right turns while driving, or identify letters and numbers in the home environment.
 - c. Use songs to practice learning letters or counting.
 - d. Use matching games to identify objects, associative pairs, quantities, and upper and lowercase letters.
 - e. Engage the child in sorting activities to arrange toys/objects by color, size, functions, features, or categories.
 - f. Use blocks or alphabet and number magnets to help the child learn sequencing skills.
 - g. Spend time reading to the child. While reading, help him/her identify letters and matching initial words sounds to pictures within the stories (e.g., “c” goes with “cat”). Consider choosing books that provide examples of how to behave appropriately in social situations, that are reassuring and foster development of healthy self-esteem, and that are imaginative, humorous, and silly.
 - h. Utilize repetition to learn how to write letters, especially those in the child’s name. Repetition or songs can be used to help the child learn his/her address and phone number as well.
 - i. Although videogame/computer and television time should be limited, allowing the child to utilize these tools for learning activities can help promote his/her pre-academic skills. His/her parents are encouraged to work with him/her to learn skills when playing on the computer or watching television.

Addressing Safety Concerns

- 16) The following recommendations are provided to help the family address safety concerns.

- a. Teach appropriate social boundaries such as remaining in close proximity to caregivers, how to refrain from speaking to unfamiliar others, and asking permission to go places with peers or other adults.
- b. For particularly active children, who enjoy exploring their environment and have difficulty remaining within arm's reach of their caregivers when outside or in public places, use of a child tracking device may be helpful. Several companies produce GPS systems that include a tracking bracelet worn by the child and a parent monitor that both sounds an alarm when children wander off too far, and that also can be used to determine the location of the child.
- c. Practice teaching the child how to cross the street safely.
- d. If the child has a tendency to leave the house without permission, it may be beneficial to place locks on doors that are out of the child's reach and/or install alarms on doors and windows. Door alarms may be particularly helpful for caregivers of children who have a tendency to get out of bed at night and roam the house.

Many developmental assessment reports will also include some type of recommendation for when the child should return to complete a follow-up assessment.

Early Developmental Resources for Families

As stated in the preceding section, developmental assessment results may include recommendations for occupational, physical, and/or speech and language therapies, feeding clinics, and/or behavioral interventions, depending on the individualized needs of the child and family. The following sections offer brief descriptions of what these resources are and the benefits of each for families coping with SMS, WS, or DS.

Occupational and Physical Therapy

Evaluations for occupational and/or physical therapy may be conducted as part of a larger developmental assessment, as described in the preceding section, or may be conducted independently. Occupational therapy evaluations can be helpful in assessing a variety of potential concerns including visual-motor and visual-perceptual skills (e.g., handwriting, cutting with scissors), upper extremity utilization (e.g., range of motion, bilateral hand coordination, strength), gross motor control, and activities of daily living (e.g., hygiene, dressing, feeding; Children's Healthcare of Atlanta, 2012). The content of a physical therapy evaluation can vary as well. However, broadly speaking, caregivers can expect that the initial evaluation will include an objective assessment of posture, joint functioning, skeletal alignment, movement analysis, strength and flexibility, reflexes, pain, and sensory processing (Physician Therapyworks, 2012). Most hospital systems offer occupational and physical therapy services. Larger cities may also have community-based rehabilitation clinics offering these services. Public educational systems should provide these services for qualifying children; however, caregivers may want to inquire as to how frequently the services can be offered, as services may be limited depending upon the school system's resources.

Speech and Language Therapy

Speech and language evaluations are typically conducted by speech and language pathologists but may be conducted as part of larger development evaluations as well. The age-based evaluation will likely include an assessment of receptive and expressive language, verbal fluency, voice and resonance, oral motor skills, articulation, and hearing ability. If deficits exist, ongoing speech and language therapy will be recommended. In addition to the services offered through hospitals and pediatric rehabilitation centers, speech and language therapy should be available through the child's public school system.

Sign language, in combination with other augmentative and alternative communication systems, has been shown to be useful in promoting language acquisition and generalized communication skills among individuals with NDs (Clibbens, 2001). These interventions can also reduce frustrations and maladaptive behaviors among children with communication difficulties. New technologies, such as iPads or the DynaVox®, have been shown to be especially helpful. For many children with NDs, the utilization of sign language or other alternatives to spoken language can be phased out as speech improves; however, for some, these speech and language alternatives may continue to be beneficial throughout the individual's life.

Feeding Clinics

Feeding clinics are available to help families with children and adolescents who experience challenges with feeding behaviors. Such clinics may comprise a number of pediatric specialty services; however, most of these clinics are run by speech and language pathologists, psychologists, nutritionists, or a combination of these services. Feeding clinics can be found at most children's hospitals and/or pediatric rehabilitation centers that offer speech and language therapy. When caregivers bring their child to a feeding clinic, they can expect an evaluation that will consist of an intake assessment of feeding behaviors, medical concerns, and developmental delays. Problems with chewing and swallowing, food and texture aversions, and mealtime anxiety will be explored. The child's anatomy and behaviors related to drinking and eating will be explored to the extent that the child may be observed engaging in these activities. It may also be necessary to conduct a swallowing study, in which a videofluoroscopy is utilized to assess whether food and liquids can move through the mouth and throat safely and effectively.

As stated previously, children with SMS, WS, and DS are likely to experience significant problems with feeding behaviors, requiring ongoing supports beyond the initial feeding clinic evaluation. Thus, these families are encouraged to request a referral from their pediatrician to participate in intensive feeding therapy or seek out such services on their own. In addition to feeding therapy, the pervasive feeding and gastrointestinal problems seen among these children may also require ongoing care from a pediatric gastroenterologist, especially if feeding problems are severe enough to require feeding tubes. Moreover, kidney problems, enuresis, and frequent urinary tract infections are often reported, which may require additional assistance from pediatric nephrologists and/or urologists (Poerber, 2010). The family's pediatrician can assist with making these referrals as well.

Behavioral Therapy

Maladaptive behaviors can be among the most challenging issues faced by caregivers. While behavioral concerns can exist among most children (and adults) with NDs, extreme and persistent maladaptive behaviors are more often reported among those with SMS as compared to those with WS or DS (Elsea and Girirajan, 2008; Slosky, Foster, and Elsea, 2012; Smith et al., 2006). It is important to remember that for individuals with SMS, WS, or DS, the etiology of these behaviors is typically both genetic and environmental. This suggests that although standard behavioral interventions can be very helpful in reducing and effectively managing maladaptive behaviors, knowledge of the specific syndrome can aid in developing a behavioral support plan that takes into consideration a specific syndrome's medical, cognitive, and behavioral profile. Therefore, in addition to ongoing behavioral therapy, psychoeducation can be a highly important first step in helping caregivers better understand the behaviors they are most likely to encounter. This educational process needs to be initiated immediately following diagnosis so that caregivers can preemptively prepare for potential concerns before they develop and/or become severe. For example, if caregivers are aware that self-injurious behaviors are likely to develop, they can actively spend time prompting or helping the child participate in more adaptive methods of coping with frustration (i.e., playing outside, squeezing silly putty or play dough, taking a bath) from the time the child is very young. As stated in a previous section, language delays or deficits can lead to significant frustration and behavioral problems. If caregivers are aware that speech and language problems are likely, they can teach sign language or the use of assistive devices before delays are apparent. Finding professionals who are specially trained to understand the complexity of these severe maladaptive behaviors can be difficult. However, more children's hospitals or larger pediatric medical clinics will have pediatric psychologists or developmental specialists who are familiar with addressing behavioral concerns among children with NDs. Early intervention programs also employ psychologists and certified behavior analysts who are trained in assessing the function of specific behaviors. Knowledge of syndromic profiles can greatly aid these professionals in conducting functional behavioral assessments that take etiology into account.

CHALLENGES, ASSESSMENTS, AND INTERVENTIONS: THE MIDDLE CHILDHOOD YEARS

"I am very happy to have my brother. I cannot imagine him being anything but himself, my little brother with Down syndrome. He can get on my nerves, but mostly I like having him around and wish that I could take him to school and show him off - plus teach some of the bullies how he is not different from them. He CAN read and do all things, and I would love to show them my brother and have him with me at my school. They don't let him go to my school because he has Down syndrome, and this school system separates them in other classes and my school does not have those classes. He has to go to a different elementary than this one. I think my brother should be able to go to the same school that I go to."

14 year-old sibling of 8-year-old diagnosed with DS

“The hardest part in raising him and for our other two children was the school years. Everything was a battle from getting services to finding teachers who wanted him in their class to IEP wording to appropriate assignments to friendships in school. I look back on those years and wonder how I retained any sanity at all. I wish we could have done those years better for everyone's sake.”

Parent of 24 year-old with DS and 22 year-old sibling without DS

Developmental Challenges

New and ongoing developmental challenges exist during the middle childhood years (i.e., ages 6 to 11, approximately), particularly for children with NDs. Previously established home-, school-, and community-based interventions often need to continue throughout the elementary and middle school years. In addition to already implemented interventions, new developmental needs and milestones specific to this period need to be considered. Common concerns include problems with social relationships, learning, inattention, hyperactivity, and impulsivity.

Peer Relationships and Bullying

Peer relationships become increasingly important as children enter the middle childhood years. Regardless of whether NDs exist, the significance of friendships in promoting quality of life and relationship development has been well-established (e.g., Bagwell, Schmidt, Newcomb, and Bukowski, 2001; Bierman, 2004). Irrespective of disability status, children desire similar characteristics in friendships, such as shared social interactions and experiences, empathy, and trust (Heiman, 2000; Turnbull, Blue-Banning, and Pereira, 2000). Inadequate social skills, poor self-esteem, and previous peer rejection tend to predict a lack of friendships (Bierman, 2004; Odom et al., 2006). Given that children with severe NDs are likely to experience problems with social skills, making and maintaining friendships can be especially challenging (Shokoohi-Yekta and Hendrickson, 2010). Children with NDs have commonly expressed feelings of loneliness due to their struggles to make friends (Solish, Perry, and Minnes, 2010). Additionally, when a child is perceived as being different or less socially competent, he/she can become a target of bullying.

In general, children without NDs participate in significantly more social activities than children with NDs (Solish et al., 2010). Additionally, children with NDs typically encounter most of their friend and peer interactions at school as compared to home environments (Shokoohi-Yekta and Hendrickson, 2010). Geisthardt, Brotherson, and Cook (2002) found that children with physical disabilities are more likely to experience friendships within the home environment than children with behavioral problems or intellectual disability. While this study did not explicitly address social concerns among children with SMS, WS, or DS, the results suggest that children with these disorders may experience significant problems making friends based on the cognitive and behavioral deficits displayed. Living in a secluded area further prevents children with NDs from having friendships outside of school, although living near other children does not guarantee interactions with peers (Geisthardt et al., 2002).

Based on this literature, social concerns and potential problems with peer relationships are important considerations for children with SMS, WS, and DS. However, for children with

WS, these problems are particularly salient. Individuals with WS display a significant lack of stranger anxiety, which is often described as having a “cocktail party personality” (Pober, 2010, p. 245). Being highly sociable and empathetic may be perceived as a good character trait; however, this hyper-sociability can also cause problems and lead to significant concerns among primary caregivers. Studies have found that children with WS often fail to recognize threatening facial cues and do not understand social boundaries, which may lead them to associate with people who may cause them harm (Schumann, Bauman, and Amarai, 2010). While this leads to a need for increased monitoring around unfamiliar adults, social skills deficits and difficulty understanding social cues can also cause problems with peer relationships, as these children may be easily manipulated into engaging in undesired activities. Despite possessing hyper-sociability, children with WS also report feeling socially isolated and experiencing anticipatory anxiety in social situations (Dykens, 2003). Children with WS have been described as demonstrating problems with obsessive thinking and being overly attentive (Davies, Udwin, and Howlin, 1998). With respect to socialization, such behaviors may translate into poor boundaries, which may lead potential friends to become overwhelmed and shy away from ongoing social interactions.

Learning Disorders

It is not uncommon for children with genetic disorders such as SMS, WS, and DS to experience problems with learning. These difficulties may be the direct result of intellectual disability, a specific learning disorder, attention problems, or a combination of the all three. Although problems with learning may be identified prior to school entry, specific learning disorders, which are a type of NDs in and of themselves, are most commonly identified during the primary and/or secondary school years. The DSM-IV-TR (APA, 2000) provides diagnostic criteria for three types of learning disorders. These include mathematics disorder, reading disorder, and disorder of written expression. All three learning disorder diagnoses require a level of achievement in reading, mathematics, or written expression that is significantly lower than would be expected given a child’s age, IQ, and education level. The soon-to-be published DSM-5 (Comer, 2013) will place learning disorders within a new grouping called “neurodevelopmental disorders.” It is anticipated that mathematics disorder will be renamed as dyscalculia, and reading disorder will be renamed dyslexia.

Given these diagnostic criteria, while children with SMS, WS, and DS are likely to experience problems with learning, they may be unlikely to meet full criteria for diagnosis of a learning disorder, given their intellectual disability. At the same time, they may benefit from interventions that are designed for children with specific learning disabilities.

Inattention and Hyperactivity

Like intellectual disability and learning disorders, children with genetic disorders such as SMS, WS, and DS often display problems with inattention, impulsivity, and hyperactivity. Depending upon the intensity and number of symptoms, a child demonstrating problems in these domains may meet DSM-IV-TR diagnostic criteria for attention-deficit/hyperactivity disorder (APA, 2000). An ADHD diagnosis requires that a child display ongoing symptoms of inattention and/or hyperactivity and impulsivity in multiple settings (e.g., school and home) for a period of no less than six consecutive months with symptoms beginning prior to age 7. For example, children with ADHD may demonstrate significant difficulties completing chores or homework, paying attention in class, or following directions. They may seem

forgetful, be easily distracted, or avoid tasks that require sustained attention. Children with ADHD may also have significant problems sitting still to the extent that they are constantly fidgeting, climbing on furniture or other objects at inappropriate times, and/or talking excessively. They may have difficulties with peer interactions because it is hard for them to take turns or engage in play activities for age-appropriate lengths of time. Three different categories of ADHD diagnoses currently exist. If symptoms are primarily inattentive in nature, the diagnosis is ADHD, Predominantly Inattentive Type. If symptoms are primarily hyperactive-impulsive in nature, the diagnosis is ADHD, Predominantly Hyperactive-Impulsive Type. Most children display a combination of inattentive and hyperactive-impulsive symptoms, which is diagnosed as ADHD, Combined Type. Although currently classified within the DSM-IV-TR as a “disruptive behavior disorder,” the DSM-5 (Comer, 2013) will re-categorize ADHD as a “neurodevelopmental disorder.”

Problems with inattention, hyperactivity, and impulsivity have been documented among children diagnosed with DS, SMS and WS (Colley, Leversha, Voullaire, and Rogers, 1990; Stratton et al., 1986). Among those with WS, one study found that more than 80% of children meet diagnostic criteria for ADHD, an anxiety disorder, or both disorders (Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, and Mervis, 2006), while a recent study on ADHD among those with DS revealed a prevalence of approximately 44% (Ekstein, Glick, Weill, Kay, and Berger, 2011). Pharmacological intervention to treat ADHD and other psychiatric disorders in individuals with genetic syndromes often does not show adequate success (Laje, Bernert, Morse, Pao, and Smith, 2010). Of note, pharmacologic intervention should be considered on an individual basis with careful monitoring and the recognition that some medications may exacerbate existing sleep or behavioral problems and may cause undesired weight loss (Smith et al., 2012).

Advocacy, Psychoeducational Assessments, and Academic Accommodations

Regardless of the child’s diagnosis or the severity of delays or deficits, most families want their child to engage in age-appropriate activities, such as attending school, reaching his/her learning potential, building relationships with teachers and peers, and participating in extracurricular activities. For children with NDs, meeting academic needs requires a continuous partnership among the school system, family, and child’s medical and psychosocial teams. An important initial step for caregivers is to work with the child’s school to help personnel better understand the child’s genetic syndrome diagnosis. Such psychoeducation on the child’s diagnosis needs to begin as soon as the child enters school and should continue each time the child encounters new educators and support staff. Given the wide variety of complex diagnoses and needs that children have in educational settings, primary caregivers cannot expect that schools will be knowledgeable about specific diagnoses prior to their child entering the school system. Proper education and training often needs to be provided, and for rare diagnoses, such as SMS or WS, the caregivers and/or child’s hospital-based team may be called upon the help with such training. Although this can be frustrating and time consuming, it also offers them an opportunity to advocate for the child. In-services between families and school personnel can not only provide disorder-specific education, but they can also help form partnerships that are designed to best serve the child as he/she ages.

Such interactions can also help the child learn to feel more comfortable advocating for himself/herself.

Schools in the United States are expected to educate children within the least restrictive environment (LRE), which means that many children with NDs will spend at least a portion of the school day with typically developing children (National Center for Learning Disabilities, 2012). As such, it can be helpful for schools to address children's perceptions of individuals with NDs and offer in-services or interventions to educate the child's classmates on their diagnosis as well (Staub, 1998; Vignes et al., 2009). Offering classmates an opportunity to ask questions about the syndrome or disorder and its associated features can be very important in helping to promote a supportive environment for the child (Rillotta and Nettelbeck, 2007). These interactions can also help to alleviate caregivers' concerns about peer relationships and potential bullying. The child's peers may benefit from being provided with the opportunity to ask questions about the physical manifestations of the disorder or syndrome, such as facial features, as well as medical concerns (Rillotta and Nettelbeck, 2007). Allowing classmates to ask questions about the child's learning needs, classroom assistance, maladaptive behaviors, and social concerns can foster compassion, minimize stereotyping, promote normalization, and improve the likelihood that the child with a ND will be able to develop adaptive and meaningful friendships as he/she moves through the educational system (Hemmeter, 2000; Rillotta and Nettelbeck, 2007).

In addition to the benefit of in-services aimed at educating teachers and classmates, children with NDs are eligible to receive specialized school-based accommodations through the Individuals with Disabilities Education Act (IDEA) or Section 504 of the Rehabilitation Act (National Center for Learning Disabilities, 2012). IDEA is a federal statute that was developed to provide free and appropriate education to children between the ages of 3 and 21, who have specific disabilities that adversely impact their educational attainment. Children qualifying under IDEA receive federally mandated and funded supplemental services and/or supports in excess of those provided within the general curricula. These accommodations are provided free of charge to the family and may include assistance with learning disorders, developmental delays, intellectual deficits, social and emotional difficulties, behavioral concerns, and managing complex medical issues. In addition to the IDEA, Section 504 is a broad civil rights law that protects the rights of those with a physical or mental disability in a way that enables the individual to participate in education with his/her peers. The law requires that schools eliminate barriers that may prevent the individual from fully participating in his/her education.

In order to obtain and ensure that educational accommodations are provided through IDEA and Section 504, a formal educational plan needs to be written, either in the form of an Individualized Education Plan (IEP) or 504 Plan. Children with diagnosed intellectual deficits, learning disorders, and/or other NDs, including those with SMS, WS, or DS should qualify for an IEP, and it is recommended that a plan be put in place as soon as the child enters the public school system (i.e., as young as age 3). Of note, private school systems are not obligated to implement an IEP or 504 Plan, but they may be open to establishing such a plan or implementing similar accommodations upon request. Once a referral is made by the parent, school, or other entity, a meeting will be held to determine what types of accommodations are needed. These meetings, which are held at least annually once a plan has been established, are important opportunities for family and others providing support to build partnerships with the child's educational team. In most cases, school systems require that a

child complete a formal psychoeducational assessment prior to implementation of an education plan. Some schools prefer to conduct the psychoeducational assessments themselves, with the assistance of a licensed or certified school psychologist. Families also have the option of having the assessment completed by a licensed psychologist or neuropsychologist in the community. For children with rare disorders and/or intensive care needs, having the assessment completed by a provider specializing in pediatric psychology can be beneficial, as this type of provider may have more specialized training in addressing the specific needs often found among children with unique genetic or medical disorders. These assessments are often completed in follow-up to earlier developmental assessments, and much like developmental evaluations, they often begin with interviews regarding the child's developmental history, current level of functioning, and specific concerns from family members and others. Most psychoeducational evaluations include formal assessments of cognitive functioning, adaptive functioning, academic achievement, and emotional and behavioral functioning. Depending on the primary presenting concerns, the assessment may also include evaluation of attention and impulsivity, memory, executive functioning, speech and language, and/or motor functioning. Once the assessment is complete, a report will be written and provided to the primary caregiver(s). As with developmental assessments, a typical report will consist of an overview of the child's medical, developmental, family, and social history, a detailed description of the assessment results, a summary of results with any associated diagnoses, and a list of recommendations and resources aimed at helping meet both the child and the family's needs.

Examples of Recommendations from Psychoeducational Assessments

A variety of recommendations can be found within the contents of a psychoeducational assessment report. These recommendations will be specific to each child's age, cognitive and developmental needs, cultural and language background, and available services and resources. As with developmental assessments, if significant deficits are present, the first recommendation made is likely to suggest that primary caregivers provide the assessment report to the school and request that a meeting be held to *discuss developing an IEP or 504 Plan*. If such a plan already exists, the report may advise that the school consider modifying or updating the current plan to best meet the child's current needs. Although not an exhaustive list, below are examples of recommendations that may be included in a psychoeducational assessment report.

Recommendation for Special Education Services or Resource Room Assistance

- 1) The results of this assessment indicate that the child displays significant cognitive and/or learning deficits. It is recommended that the child participate in special educational services and/or have access to resource room assistance to help support his/her significant educational needs.

Recommendations to Address Problems with Inattention and Hyperactivity

- 2) The following recommendations are provided to address the child's identified problems with inattention, distractibility, and hyperactivity.

- a. The child may benefit from taking tests in the resource room or library, where he/she can work without distractions.
- b. Avoid numerous directions or assignments. Allow the child to finish one assignment or direction at a time before going on to the next.
- c. Maintain a predictable classroom routine; keep the daily schedule as consistent as possible. Prepare the child ahead of time when you know that the routine must be changed or when transitions are going to take place.
- d. Allow the child to sit near the front of the classroom; stand in close proximity to the child while teaching. This will help keep him/her focused on the classroom activities.
- e. Before addressing the child, speak his/her name and make eye contact; keep oral directions short and uncomplicated. Ask him/her to repeat the directions until it is certain that he/she understands what is being asked.
- f. Be aware of the child's frustration level. Knowing when the child is about to lose focus or become frustrated may prevent inappropriate behavior and feelings of failure. Do not be afraid to discuss this with the child so that both of you can identify the factors that lead to frustration.
- g. Allow the child to take breaks from schoolwork throughout the day. This will help decrease frustration and increase learning and productivity. Children with significant attention problems may need to take multiple breaks each hour.
- h. As the child ages, encourage the utilization of assignment notebooks. This will promote organizational skills and increase the likelihood that assignments will be completed successfully. Children with fine motor problems may benefit from using an audio recording device to record assignments.

Recommendations to Address Problems with Mathematics Achievement

- 3) The following recommendations are provided to address the child's deficits in mathematics achievement:
 - a. The child should receive remedial, one-on-one instruction in mathematics.
 - b. It will be beneficial to ensure that the child has fully automatized his/her math facts before attempting to have him/her learn more abstract, problem-solving concepts. The better his/her understanding of basic math concepts (i.e., adding, subtracting, multiplying, dividing), the easier it will be for him/her to apply basic knowledge to higher level math problems and to complete math problems efficiently. Utilizing flashcards, charts, and computer programs may assist the child in learning his/her basic math facts and increasing the speed with which he/she is able to recall and apply these facts.
 - c. Based on measured visual-spatial difficulties, the child may benefit from math worksheets that include structured workspace. Consistency in presentation is important so that examples should be written using either horizontal or vertical alignment, not both. Graph paper may also help with organizational efforts.
 - d. Any difficulties with reading may also impact the child's ability to successfully complete mathematical word problems. Therefore, his/her parents and teachers are encouraged to read word problems to the child, to help him/her read through problems aloud, and/or to provide pictures that accompany word problems. The child should be taught specific strategies for identifying important pieces of

information in the problem and determining the operation needed to solve the problem.

- e. Providing manipulative objects and utilizing them in instruction (e.g., rulers, beads, blocks) will likely improve the child's opportunities for success in math.
- f. At home, enjoy time as a family, engaging in tasks that encourage learning. Spend time playing structured age-appropriate games/activities that require visuo-spatial reasoning (e.g., Legos/blocks, puzzles, mazes, etc.).
- g. At the child ages, it will be especially important to focus on practical mathematical skill development such as telling time, reading thermometers and car gauges, using money, maintaining a checkbook, and understanding basic banking and credit principles.

Recommendations to Address Problems with Reading and Writing Achievement

- 4) The following recommendations are provided to address the child's deficiencies in reading and written expression:
 - a. The child should receive remedial, one-on-one instruction in reading and written language.
 - b. Prepare a copy of the homework assignments and hand it to the child at the end of the day. The goal is to create a comfortable and successful environment. In this case, having the child accomplish the homework is more important than the difficulty encountered in copying his/her assignments.
 - c. Provide audiotaped copies of text books so that comprehension of class materials is not negatively influenced by reading deficits. This provision will also help the child's parent(s) assist him/her in completing course assignments.
 - d. The child may benefit from oral testing and from having test questions and items read to him/her so that his/her knowledge of course material is being assessed rather than her reading ability. Worksheet instructions should also be read aloud to the child.
 - e. For children with significant motor problems that deter hand written activities, consider allowing the use assistive devices such as computers and tablets to complete assignments.
 - f. Encourage the child's appreciation for reading by allowing him/her to choose age appropriate reading materials (books, comic books, magazines) and by reading with him/her, discussing the characters, exciting parts of the story, etc. The child may enjoy picking out books-on-tape from his/her local library and following along in the books as the stories are read to him/her. This strategy may be particularly helpful for improving reading fluency.
 - g. At home, enjoy time as a family, engaging in activities that encourage reading and writing skills (e.g., Boggle, Scrabble, Pictionary, word finds, etc.). Allow the child to choose family activities among a pre-determined list of options. Allowing him/her to determine the activity will give him/her a sense of control and encourage him/her to take an active role in his/her own learning.

Improving Confidence and Effort

- 5) The following recommendations are provided as methods for encouraging the child's continued engagement and effort in the classroom and when completing homework:

- a. Focus on building academic self-confidence. The child may show a strong desire to learn but need more time and more intensive instruction than other students to accomplish and master tasks. It will be important to promote confidence in his/her ability to learn as he/she ages so that he/she does not become discouraged by his/her cognitive and academic weaknesses.
- b. Try to identify the child's strengths that can be publicly announced or praised. In this way, the child's peers will perceive him/her positively, and he/she will continue to seek out positive attention.
- c. Address the child in a calm, clear tone when encouraging academic behaviors. If he/she feels overly pressured to accomplish academic tasks, he/she is more likely to feel anxious and lose confidence. At the same time, do not allow the child to avoid participation in academic work, as this may inadvertently increase anxiety related to school performance.

Addressing Behavioral Concerns within the Home

- 6) Specific behavioral interventions can be applied in the home to promote the child's social and emotional well-being. The following strategies and techniques are recommended:
 - a. Develop a consistent schedule within the home. A schedule builds a sense of safety, reduces anxiety, sets reasonable expectations, and breaks things down into manageable components. Keeping the child on a routine will help him/her manage his/her behaviors and give him/her the best chance to succeed. He/she will be more invested in the schedule if he/she is able to help create it. The schedule needs to be posted prominently in the home. Although weekends tend to be more variable than weekdays, a schedule needs to be developed for the weekends with consistent bedtimes and hygiene routines. While being consistent, it is also important to foster flexibility and to ensure that the child does not become overly reliant on routine and resistant to change.
 - b. Develop a list of household rules. Try to keep this list to no more than 5 of the most important rules, and allow the child (and his/her siblings) to help create the list. Specific age-appropriate consequences for breaking each rule should be written down and discussed so that the child will be aware of what the consequence will be if he/she disobeys.
 - c. Always address the child in a calm, clear tone when redirecting him/her or encouraging academic behaviors. Avoid yelling and using loud tones of voice or physical discipline. While it is important to use firm tones so that the child knows what is expected of him/her, yelling and/or harsh tones will likely result in defiant behaviors. If he/she feels overly pressured to accomplish tasks, he/she is more likely to feel anxious and lose confidence.
 - d. Create a Focus Zone within the home and school. This is a place where the child can go to calm down or to have quiet time. A Focus Zone could be a small area with a desk or table that is within sight and sound supervision. He/she should be encouraged to use the space, and it should not be considered a punishment. Things to have within the Focus Zone include: 1) Art Materials: Paper, crayons, and beads; 2) Puzzles and age-appropriate games; 3) Toys that require the child to

- put things together such as Legos; 4) Relaxing music; and 5) Comfortable seat, pillows, etc. TV and video games are not recommended. The Focus Zone should not be used as playroom. Encourage the child to use this area to calm down, and then return to his/her normal activities.
- e. Daily exercise and outdoor activities will allow the child to expel excess energy and calm down.
 - f. If he/she wets the bed, have him/her complete the appropriate hygiene and assist him/her in changing his/her bedding. Avoid punishing him/her for this behavior. Simply have him/her help clean up any mess that has been made and then move on.
 - g. Consequences for refusing to listen can include such things as loss of free time, early bed time, loss of fun outings, and loss of access to toys or video games. He/she should have the opportunity to earn these privileges back. The child should always be held accountable when he/she breaks rules. Avoid using corporal punishment, as this may result in increased acting out behaviors.
 - h. Children respond well to praise and compliments. When the child does something positive, be sure to acknowledge him/her and possibly allow him/her to earn rewards. This is especially important with school work and following household rules and expectations.
 - i. When the child refuses to do what is asked of him/her, inform him/her of the potential consequences of his/her actions and give him/her two to three choices for appropriate ways to behave if possible. This will assist the child in feeling as though he/she is maintaining some sense of power and control. Enforce the consequences immediately if he/she does not do what is asked.
 - j. Behavior management books such as *1-2-3 Magic* by Thomas Phelan and *S.O.S.: Help for Parents* by Lynn Clark are recommended to assist in reducing oppositional, defiant, and aggressive behaviors.

Promoting Healthy Weight and Physical Activity

- 7) Children with NDs are at high risk for problems related to obesity, beginning in the middle childhood years. While this may be related to metabolic problems, it will be important to promote healthy diet and exercise behaviors in the child. The family may benefit from working with a dietician in learning about portion control and how to develop healthy meal plans. The child's weight should be closely monitored and his/her eating habits should be closely supervised as much as possible. He/she should be praised for making healthy food choices.
- 8) The child should be encouraged to participate in physical activity on a regular basis. It will be important to identify exercises and activities that are enjoyable for the child so that he/she is more motivated and interested in maintaining them over time. He/she may enjoy karate, yoga, swimming, playing tag, taking walks with a friend or family member, gardening, participating in community service activities that require some physical exertion, dancing, or bowling, for example, He/she should be praised for his/her efforts, and physical activity logs and pedometers may be particularly helpful for monitoring. If significant physical limitations are present, the child and family may benefit from consulting with a physical therapist.

Promoting Social Skill Development

- 9) The following recommendations are being made to promote the child's social skill attainment.
 - a. Use role-playing or social stories to teach new skills or desired interactions with others.
 - b. Practice initiating and maintaining conversations by teaching the child how to shake hands, introduce himself/herself, ask reciprocal questions, and maintain eye contact.
 - c. Look for times throughout the day to help the child better understand perspective taking. For example, ask the child to identify emotions of people in television shows or storybooks.
 - d. Have the child participate in clubs, volunteer activities, library reading groups, school groups, or other activities that the child enjoys that will encourage him/her to practice socializing with other children his/her age.
 - e. The child would benefit from participating in a social skills group. Such groups may be offered through the child's school or through a local mental health clinic.

Similar to developmental assessments, the final recommendation in a psychoeducational or neuropsychological assessment report is likely a statement regarding when the child and family should return for a follow-up assessment. It may also include recommendations regarding referrals to other disciplines, including speech/language, occupational, and physical therapists, psychologists or behavioral specialists, nutritionists, or sleep medicine clinics, for example.

CHALLENGES, ASSESSMENTS, AND INTERVENTIONS: THE TEENAGE YEARS AND BEYOND

“In my opinion the parents with young Williams children all think they are going to be better as adults than for the most part they really are going to be. I think [families are given] too optimistic of a rosy future for those with children. It's good to be encouraging and positive, but you need to sprinkle it with a dose or realism.”

Parent of an individual diagnosed with WS

Developmental Challenges

As those with NDs become teenagers, additional challenges emerge for individuals and caregivers. As is common among those with NDs, challenges encountered during infancy, early childhood, and middle childhood may continue to exist, improve, or worsen over time. With this in mind, therapies and resources already in place prior to entering adolescence may continue to be required (e.g., school-based academic accommodations), while others may no longer be necessary (e.g., feeding clinics, speech and language therapy). During adolescence (i.e., approximately ages 11 to 18) and beyond, common concerns include issues related to sexual development, romantic relationships, and vocational planning. Many caregivers must

also make considerations for semi-independent or community-based living and determine how to best address legal concerns such as guardianships and advanced care directives.

Sexual Development and Romantic Relationships

Addressing sexual development and romantic relationships can be a difficult topic for caregivers, regardless of whether or not their adolescent is diagnosed with a ND (Lesseliers and Van Hove, 2002). When the adolescent has a ND, additional challenges and safety concerns exist. The literature has discussed two common and contradictory perceptions when it comes to addressing sexuality among those with NDs (McCarthy, 1999). The first suggests that those with NDs need to be protected from society due to the inherent vulnerabilities that exist in a sexually provocative world. This could be interpreted to mean that the topic of sexuality and romantic relationships should be avoided, with the somewhat misguided presumption that this will protect individuals with NDs from desiring sexual interactions and/or being victimized. The second view acknowledges that most individuals with NDs will want to engage in sexual relationships, just like most individuals without NDs (McCarthy, 1999; Melberg-Schwier and Hingsburger, 2000). Unfortunately, normative sexual desire, combined with poor social boundaries and reduced intellectual ability, may lead those with NDs to pursue inappropriate and perhaps dangerous sexual encounters, making ongoing sexual education and discussion imperative.

In further considering the second (and more commonly accepted) viewpoint, primary caregivers are then charged with determining how and when to begin discussing sexual development and romantic desires with their child with a ND (Lesseliers and Van Hove, 2002; Thorin and Irving, 1992). This has been identified as a significant source of stress for primary caregivers, who are often unsure of what information to provide, how to present the information in a way that will be understandable, and when to begin discussions. Depending upon the level of intellectual, behavioral, and/or emotional impairment, there are also numerous questions regarding whether to “allow” romantic and potentially sexual relationships. Appropriate methods of pregnancy prevention must also be explored. In some cases, adolescents and adults with NDs will be reasonably well-supervised within their school and living environments. However, completely negating opportunities for sexual contact is nearly (if not completely) impossible, making it even more important that sexual education and social skills training are provided in a timely and developmentally sensitive manner.

Many primary caregivers struggle to acknowledge that their child with a ND may want to engage in sexual interactions (Melberg-Schwier and Hingsburger, 2000; Lesseliers and Van Hove, 2002). Picking up on any cues related to potential sexual interest (e.g., flirting behaviors, masturbation, wanting to watch television or movies with sexual content) displayed by the adolescent may be an important first step in promoting awareness for parents. These cues can also act as conversation starting points. As compared to parents, professionals working with individuals with NDs have been found to be more likely to notice sexual interest and behaviors. Additionally, while parents may be less inclined to acknowledge sexual interests, many professionals believe that those with NDs, like all other individuals, have a fundamental right to sexual expression. With this in mind, professionals are likely to work with individuals with NDs to normalize their sexual desires, discuss independent decision-making, and learn self-advocacy and appropriate social boundaries. A primary goal is to help primary caregivers feel capable of openly discussing issues surrounding intimacy and sexuality. In this way, medical and/or psychological professionals may act as a valuable

resource in helping primary caregivers address sexual development and romantic relationships in their adolescent and adult children with NDs.

Life Skills and Vocations

Many individuals with significant NDs, such as SMS, WS, or DS, want to participate in work-related activities. Engaging in work activities allows individuals with NDs to find purpose and self-fulfillment, interact with and contribute to the community, and build self-esteem and life skills. What constitutes successful vocational attainment depends on the goals of the individual and what opportunities are available (Smith, Wilson, Webber, and Graffam, 2004). Vocational success may be dictated by the number of hours and environments in which it is feasible for the individual to work; employer expectations; and the cognitive, developmental, behavioral/emotional, physical, and medical problems of the individual. The American Association of Intellectual and Developmental Disabilities (AAIDD) stipulates that those with such challenges must be afforded opportunities that promote well-being in a variety of environments, including employment settings (AAIDD, 2010, 2012).

Historically, those with significant NDs have been perceived as unable to work (Marks, 1999; Oliver, 1990; Smith et al., 2004), and until recently, have often been excluded from work environments and society at large unless needed to fill a void in the workforce. For example, throughout much of the 20th century, those with NDs were called upon to work during times of war, when labor shortages existed, and were returned to institutions when the wars ended. Although much has changed with respect to how those with NDs are supported, stigmas still exist that can make employment an ongoing challenge. Statistics suggest that individuals with NDs are much less likely to be employed than those without such a disability (Smith et al., 2004). This is an important statistic to consider, especially given that community-based employment is associated with increased quality of life (Eggleton et al., 1999). In the U.S., all individuals with disabilities are protected by anti-discrimination legislation that requires that employers provide reasonable accommodations that allow the individual to complete his/her work-related tasks (Smith et al., 2004). These laws indicate that those with NDs should be able to work in an environment that supports satisfactory work experience such that no discrimination should exist in hiring practices; however, employment rates continue to be low. Employers who are more likely to employ those with NDs have been found to demonstrate greater awareness of anti-discriminatory processes and are more likely to favor social justice. Person-environment fit has also been found to be particularly important in that it is not only salient that the person be able to perform his/her job but that he/she feels accepted and valued as a member of the work team (Hagner, 1992). Social skills have been described as a predictor of whether or not an individual will be accepted in the work place, and intervention studies have shown that many individuals with NDs can be taught specific task-related social skills, such as asking for assistance and using verbal problem-solving strategies (Holmes and Fillary, 2000; Martella et al., 1993).

“Natural supports” are defined as “assistance provided by people, procedures, or equipment in a given workplace that (a) leads to desired personal and work outcomes, (b) is typically available or culturally appropriate in the work place, (c) is supported by resources from within the work place, facilitated to the degree necessary by human service consultation” (Butterworth et al., 1996, p. 106). These “natural supports” are imperative in helping those with NDs succeed in the workplace and may require co-worker training, on-site job coaches, and community-based partnerships. Job coaches or job instructors can help

individuals break down work tasks, establish methods of performing tasks successfully, provide prompts, and build independence and self-confidence (Callahan and Garner, 1997). While the supports can require extensive time and training to implement, they should not deter employers from hiring individuals with specialized needs. Through partnerships with community-based agencies, such as rehabilitation services, employers may be able to offer specially created or “carved jobs,” that enable the individual to be successful in the workplace (Gilbride and Hagner, 2005, p. 295).

Vocational Assessments and Planning

As discussed in the previous section, as individuals with NDs age, it is important to consider how to best plan for vocational goals. A formal vocational assessment can provide a helpful starting point for taking steps towards vocational planning. Such an assessment can be completed through a state-run vocational rehabilitation services program or through most mental health clinics that specialize in meeting the needs of adolescents and young adults. School systems will often be equipped to help families facilitate this process as well. Most vocational assessments will be conducted by a licensed psychologist, school psychologist, or psychological examiner. Again, as with developmental and psychoeducational assessments, family and other support persons are typically asked to participate in an interview regarding the child’s current level of functioning and related concerns. This interview will be followed by a formal assessment of cognitive functioning, vocational interests, personality, adaptive functioning, and behavioral and emotional functioning. Depending on specific presenting concerns and the overall purpose of the evaluation, the assessment may also include an evaluation of academic achievement, memory, attention, and/or executive functioning, much like a psychoeducational evaluation. In medical settings, the assessment may also include additional measures of functional competence that address medication adherence and schedules, as well as awareness of medical factors associated with the ND. Once the assessment is complete, a report will be written and provided to the individual and/or primary caregivers. As with other assessment reports, a typical vocational assessment report will consist of an overview of the individual’s medical, developmental, family, and social history, a detailed description of the assessment results, a summary of results with any associated diagnoses, and a list of recommendations and resources aimed at helping address the individual’s vocational and life skill needs.

Examples of Recommendations Provided in Vocational Assessments

The following are examples of recommendations that may be made as part of a vocational assessment for an individual with a ND. The specific recommendations made will vary substantially depending upon the severity of intellectual disability, learning disorders, and problems with adaptive functioning, medical limitations, and/or mood and behavioral problems that exist. However, in general, there will be a recommendation to provide the assessment report to either the individual’s school or local vocational rehabilitation services, who can then provide additional help in facilitating vocational placements and supervision (if

needed). Additionally, the recommendations may include further suggestions for online vocational exploration, vocational counseling, and/or job shadowing.

Referral to Vocational Rehabilitation Services

- 1) The individual is encouraged to share the results of this assessment with Vocational Rehabilitation Services and/or to provide a copy of the results to administrative officials at his/her current educational facility. Vocational Rehabilitation Services may be especially helpful in facilitating the individual's ongoing vocational development.

Options for Additional Vocational Exploration

- 2) The individual can take advantage of the following career resources to continue with his/her vocational exploration:
 - a. O*NET Online (<http://online.onetcenter.org/>): O*NET Online provides comprehensive information on key attributes and characteristics of workers and occupations. On this site, the individual can take a free skills search assessment that will provide additional information on the types of careers for which he may be well-suited.
 - b. Occupational Outlook Handbook (<http://www.bls.gov/oco/>): The Occupational Outlook Handbook provides the most up-to-date information available on hundreds of jobs and includes information on the amount of training and education needed, earnings, expected job prospects, and work conditions.

Vocational Counseling

- 3) Vocational counseling, which may be available through his/her school, is recommended to assist the individual as he or she continues to explore vocations of interest. In addition to facilitating the development of a specific vocational path and the steps that need to be taken to further his/her vocational development, engaging in such counseling will allow the adolescent to consider how his/her personal values, cognitive/academic strengths and limitations, and openness to new ideas influence his/her ability to obtain a satisfying vocation.

Vocational Shadowing

- 4) Once vocational interests have been solidified, it may be beneficial to shadow professionals in vocations that the individual wants to pursue. This will allow the individual to network with others in fields of interest and get an in-depth view of what the vocation entails on a day-to-day basis.

Vocational assessments may also result in other recommendations offered to promote the quality of life of the individual and/or his/her family. For example, recommendations may be given to promote adaptive functioning. Again, the examples below represent areas that may need to be addressed in adolescence or adulthood and do not represent an exhaustive list that will meet all individual needs.

Adaptive Functioning Skills

- 5) The following recommendations are provided to the individual and his/her primary caregivers to promote development of adaptive functioning skills:
 - a. Focus on developing practical mathematics skills, such as counting money, making correct change, and understanding basic banking and credit principles.
 - b. Practice setting longer-term goals (e.g., work-related goals, social goals) and how to break these larger goals into smaller goals that can be achieved in a reasonable amount of time.
 - c. Continue to encourage writing skills by having the individual write short notes or letters to friends. This will help him/her maintain the writing skills and fine motor dexterity he/she has developed thus far.
 - d. Assist the individual in taking more control over his/her medication adherence. Although the individual may need to be supervised in doing this, using pills boxes or cell phone/watch alarms may help the adolescent monitor his/her own medications more independently.

Community Living

Individuals with SMS, WS, and DS typically experience pervasive problems with daily living skills, or adaptive functioning, to the extent that they will require some level of monitoring throughout their lives and are unable to live independently as adults (Schubert, 2009). Deinstitutionalization has created an impetus for policymakers and researchers to focus on more appropriate long-term living arrangements for individuals with significant disabilities as they reach adulthood (Emerson, 2004). With this in mind, community integration has now become the primary goal, with its emphasis on independent or semi-independent living (Racino, 1995). Successful community integration implies that individuals with NDs have the opportunity to not only reside in the community, but to work and recreate in ways that are similar to those without NDs. Many individuals with NDs who are unable to live independently will reside in publicly-funded, supportive housing programs that provide supervised living and supportive services (Wong and Stanhope, 2009). Community-based care following deinstitutionalization has been greatly influenced by Wolfensberger's normalization principle (Flynn and Aubry, 1999; Wolfensberger, 1983). This principle stems from Nirje's work on social role valorization and indicates that individuals with NDs need to be integrated into culturally-rich normative community settings (Flynn and Aubry, 1999). Within these settings, quality of life will be improved if individuals can actively participate in socially valued roles and interactions. The principle argues that institutions are harmful, because they perpetuate atypical behavioral interactions and ways of functioning, which in turn increases public stigmas regarding those with disabilities. Competence and self-worth are believed to be enhanced via participation within the larger society. Residential settings can vary widely (Wong and Stanhope, 2009). They may include intensive therapeutic care with extensive treatment and rehabilitation, small group living scenarios emphasizing peer relationships and community involvement, living with a surrogate family, or residing with family members with in-home supportive services. Some residential settings serve to transition those with mild to moderate NDs to independent living. Others are structured to offer long-term supervised care in a supportive atmosphere. Programs such as the 1981 Home

and Community Based Services Waiver have effectively reduced the number of people residing in larger institutional settings (i.e., settings with more than 15 individuals) by providing Medicaid funding for non-institutional services. As of 2004, 83.5% of individuals with NDs were residing in settings with 15 people or less; nearly half (46.2%) were residing in settings with 3 people or less (Prouty, Smith, and Lakin, 2005). Home-based supports tend to be a newer avenue of support (Wong and Stanhope, 2009). These allow individuals to remain in their own home environments while offering daily or weekly therapies and respite care for family members.

Guardianships, Alternatives to Guardianships, and Advance Care Directives

As children with neurodevelopmental disorders (NDs) move through adolescence, society also begins to presume adulthood. The following information has been gathered from various U.S. state specific sources. Although specific details differ by state, a person with or without a ND is generally considered an adult if any one of the following criteria are met: 1.) the individual has been emancipated by a court of law, 2.) the individual is 18 years of age or older, or 3.) the individual has been married. In some states, an individual is also considered emancipated if she has been pregnant. It is important to remember that once people meet the definition of adulthood, whether or not they have a ND, it is assumed that they should be able to make informed decisions by the legal system, health care providers, governmental agencies and society in general. Parents who have had de facto natural guardianship of their child do not automatically retain guardianship or legal decision making authority for the adult. This has special implications for adults with NDs.

The only legal alternative available to caregivers to retain guardianship of an adult is to initiate a court proceeding, which can be costly and may be unnecessary depending upon the severity of the ND and the needs of the individual. Such a proceeding seeks to determine whether the adult's ND severely impacts his/her decision making ability to such an extent that he/she is in need of protection. This, however, does not need to be the first course of action for an adult with a ND (O'Sullivan, 1999), even for people with severe limitations in cognitive function and decision-making abilities. There may be multiple ways for family members to continue supporting the individual in decision-making endeavors without seeking formal adult guardianship. For example, unless the person with ND has elected otherwise, family members are still able to attend and participate in Individualized Education and Support Planning meetings and may be able to give needed medical consents as next of kin.

With changes in privacy laws such as the *Health Insurance Portability and Accountability Act of 1996*, it has become more difficult for parents to gain access to the health records of their adult children (HIPAA, 1996). However, there are ways for caregivers to initiate conversations with the adult child and the healthcare provider team to discuss how medical decisions will be made. A discussion including the individual with a ND, the caregiver, and the individual's provider often results in a plan that is agreed to by all parties. This may include the completion of releases of information that allow the caregiver to continue communicating directly with the healthcare provider or those that allow the caregiver to continue obtaining medical information. The primary stipulation in allowing the individual with the ND to complete release forms is ensuring that consent has been provided through an informed process. In non-emergent situations, providers have an obligation to

obtain informed consent from service recipients and to make every effort to present the information in language and terms that is understandable (Beauchamp and Childress, 1994). Formal cognitive assessments may be needed to determine whether the individual with the ND has the cognitive capacity to provide informed consent for medical care and/or sign release forms allowing caregivers permission to continue actively participating in care.

Guardianship Alternatives

Individual state processes vary, but in general, procedures are available to ensure that caregivers and other family members can exercise certain authorities without undergoing a full guardianship procedure (Dinerstein, 2006). Some of these procedures are detailed below, but caregivers and family members should discuss the best course of action with the service providers who provide routine support to the individual with ND, such as physicians and social service agencies, before proceeding. Individual state offices typically publish regulations governing decision making that impact people with NDs (Pennsylvania Code Title 55, ch.6000 subchapter R, 2011). Similarly, state specific advocacy groups (Disability Rights Texas, 2011) also have publications and staff available that provide education regarding state-specific options. These referenced sources outline the following potential alternatives to guardianship.

Representative Payee. People with NDs who qualify for benefits under the Social Security Administration (SSA) but who are unable to manage their fiscal affairs can participate in the representative payee program. This program provides for the management of Social Security and Supplemental Security Income payments to people with NDs, who are not capable of independently managing their payments. In most cases, a family member or friend serves as the representative payee, but an agency, such as a supports provider, can also serve in this capacity. The representative payee helps a person with NDs manage his/her benefits and has certain responsibilities, such as ensuring that the benefits are used as intended, saving any remaining benefits amounts, and maintaining adequate financial records. More information is available from the SSA (www.ssa.gov).

Power of Attorney (POA). The execution of a POA enables an individual to act on behalf of another person in certain situations. Consent for the POA must be given by an individual who understands the process and the ramifications of giving another person the authority to make decisions on their behalf. An attorney should be consulted about the possible uses of a POA. There are jurisdictional differences, but a POA typically remains active if an individual is no longer able to understand the POA process, and it carries broad authority beginning the day the document is executed. This type of POA is called a durable POA. The POA process is much less costly and cumbersome than a full guardianship determination, and most families of people with ND who get a POA execute a durable POA and not a limited POA. The latter can be used for brief periods of time where an individual is not available to act on their own behalf. Many healthcare and service providers, as well as governmental agencies, will accept a POA in situations requiring decisions related to accessing records. If a POA is executed, caregivers should provide copies to service providers and governmental agencies such as the SSA.

Health Care Proxy (HCP). A HCP appoints another individual, or agent, to make healthcare decisions for a person in the event that the person becomes unable to make such decisions. Again, states differ widely in the extent of authority of the HCP and the process by which a HCP can be appointed. Generally, an HCP is executed by a person who is appointing

a trusted friend or family member to make medical decisions in the event that the individual executing the document becomes incapacitated, or unable to do so. Most jurisdictions leave the decision of incapacitation to the primary physician.

Some jurisdictions also recognize health care representatives and health care agents, the former only requiring an agreement drafted between the two parties, while the latter requires the services of a notary. Some states have also enacted statutes that give default decision-making authority to certain individuals, such as family members, in a prescribed, hierarchical arrangement. Typically, people who work for agencies that support people with NDs are not able to serve in this capacity for a particular individual, except as a last resort, unless the agency employee also happens to be a family member. Likewise, certain rules may apply for individuals residing in state administered developmental centers. State or county offices that are responsible for administering programs for people with developmental disabilities are often the best resource for specific information.

Advanced Directives (AD). In instances where a HCP has been appointed, an individual also usually executes an AD. This is a separate legal document that contains instructions to healthcare providers and that specifies what procedures are desired or not desired in the event that the person becomes unable to make such decisions on their own behalf. The use of dialysis, respirators, cardio pulmonary resuscitation (CPR) techniques, tube feedings, and organ donation all can be expressly addressed in an AD. Discussing these issues with a family member with a ND can be difficult, but it can prevent scenarios during which caregivers are asked to make these decisions quickly during a time of crisis. In addition, certain decisions that an individual can make for himself/herself as part of an AD cannot be made once the person is unable to make the decision for himself/herself, even if a legal guardian has been appointed. The contents of an AD for people with ND can become controversial, and an attorney should be consulted when formulating such a document. For example, in most jurisdictions, a person acting on behalf of another cannot decline life sustaining treatment for another individual, unless the other individual meets specific criteria for a “terminal condition.” This is true even for a person who is a court appointed legal guardian.

Adult Guardianship

In most cases and situations, use of the above procedures is sufficient to support people with NDs in managing their own lives. On occasion, legal guardianship is the only alternative. A guardian is a person or an agency that has been appointed by a court to act on behalf of another person. This ultimately removes that person’s right of self-determination. A court appointed guardian can be a single person, or in some instances, more than one person can act as co-guardians. In guardianship proceedings that appoint co-guardians, special attention should be given to whether consent from both parties is needed, or if one or the other party can consent independently. It is also possible to include guardian succession in a decision. A guardianship decision, or decree, once rendered, can only be modified by the court.

For the guardianship determination to proceed, an attorney must be engaged by the person petitioning the court for guardianship. In addition, an attorney may be appointed by the court to represent the interests of the individual with a ND. Before the court proceedings, the individual with a ND will need to be evaluated by a licensed clinician to determine his/her current competence, or ability to make decisions. Part of this evaluation will include a determination of potential risk to the person should a guardian not be appointed. This

evaluation, depending on the jurisdiction, can be completed by a licensed psychologist, a psychiatrist, or a primary care physician. A court date will be determined, and all parties involved may receive a summons to appear in court, including close family members. In certain circumstances, a physician or psychologist can attest that the presence of the person with the ND would not be in the person's best interest, and an exception to attendance can be made. During the hearing, the hearing master or judge will listen to the evidence, witnesses may be called to testify, and both attorneys will have the opportunity to ask the witnesses questions. A determination is then made, granting either general or limited guardianship. General Guardianship is sometimes referred to as 'plenary' guardianship and may be appropriate for individuals with NDs who have been found generally incapable of decision making in all aspects of their lives. Limited Guardianship can cover specific decision making circumstances. Specific circumstances include issues arising in residential, educational, financial, health care, legal, and/or job related settings.

A legal guardianship determination transfers the rights of the individual to the guardian, and, therefore, is the most restrictive alternative. The concept of "support," which enables an individual to participate in decision making, has supplanted "surrogacy," or making decisions on behalf of an individual, as the method of choice for meeting the needs of individuals with NDs (AAIDD, 2010, 2012). As such, professional and family organizations, such as the American Association on Intellectual and Developmental Disabilities (AAIDD) and The ARC, have issued a joint position statement on guardianship that recommends such an action as a last resort measure (AAIDD and The ARC, 2012).

PRIMARY CAREGIVERS AND SIBLINGS: CHALLENGES, BENEFITS, AND FAMILY WELL-BEING

Primary Caregivers

"I can only speak to what it has been like for our family. Our child with Down syndrome...has taught our family so many lessons; the most important for me has been acceptance and unconditional love. When I am at my worst, [he] opens his arms to me in an embrace. He loves me, holds me, and shows me the power of unconditional love. Because of his example, I have been able to offer that to my other children. [He] has shown us the importance of inclusion and the beauty of every single person, regardless of ability. He has shown us the joy woven throughout each day and the importance of learning things in small steps instead of giant gulps. He has shown me what it is like to be genuinely joy-filled for someone else's achievement or special moment. His level of compassion and empathy are incredible and often humbling. We also have witnessed the pain of being different and the difficulties that our society has with people who are different."

Parent of 11 year-old child with DS and 11 year-old child without DS

"We still do not have a good handle on how to care for him; we just want his quality of life to be as good as we can make it, but that remains a mystery."

Parent of a child diagnosed with WS

Parenting a typically developing child is a monumental task that requires tremendous time, emotional, and financial commitments. In addition to meeting these typical child care needs, caregivers of individuals who are diagnosed with one or more NDs also face a number of unique and often unexpected challenges, which may include added financial burdens due to medical care costs and specialized therapies, advocating for educational needs, and determining how to provide for their child as they age (Davies, Howlin, and Udwin, 1997). Caregivers of children diagnosed with SMS, WS, or DS must learn to cope with and readily adapt to the physical and behavioral features of the syndrome as they emerge across the lifespan. Many of these challenges have been discussed throughout the course of this chapter and may include any number of problems, such as feeding, sleep, and other developmental concerns; motor and/or language deficits; intellectual disability; significant maladaptive behaviors; social deficits; learning and attention problems; and learning to cope with complex medical problems (Poher, 2010).

Primary caregivers must not only manage individual child needs, but simultaneously address these challenges while working to provide the best quality of life possible for the entire family system (Silver, Westbrook, and Stein, 1998). The significant demands of the direct support role leads to a high degree of stress and burden (Fidler, Bailey, and Smalley, 2000). For example, questionnaires completed by parents of 36 children with SMS showed that the degree of maladaptive behavior displayed by the child with SMS was the best predictor of parental stress and pessimism (Hodapp et al., 1998). The burden is then compounded by the knowledge that the direct support role will continue throughout the course of the child's life, as most individuals with SMS, WS, or DS will be unable to live independently as adults (Davies, Howlin, and Udwin, 1998; Udwin, Webber, and Horn, 2001). Over time, the associated burdens can lead to problems with primary caregiver well-being (Graham, Ballard, and Sham, 1997; Hasselkus, 1988), especially given that the direct support role is unrelenting and oftentimes unexpected (Eicher and Batshaw, 1993).

Research exploring primary caregiver well-being and NDs continues to emerge. Established literature cites the importance of social support in promoting the psychological and physical health of these primary caregivers (Barakat and Linney, 1992; Erickson and Upshur, 1989). With the intensity of caregiver demands associated with supporting a child with SMS, WS, or DS, it may be expected that primary caregivers would display poor self-care behaviors or demonstrate coping difficulties. In fact, one study of SMS caregiver well-being supported this statement with findings suggesting that maternal caregiver well-being was directly impacted by perceived child health vulnerability, caregiver satisfaction, and benefit finding (Foster et al., 2010). Paternal caregiver well-being was most influenced by depressive symptoms and benefit finding. Overall, this study found significantly increased rates of depressive and anxiety symptoms, disrupted sleep, and difficulties maintaining annual physical exams among both mothers and fathers providing direct support to individuals with SMS. These findings and other similar studies have identified an increased need for counseling services for these caregivers (Foster et al., 2010; Scallan et al., 2010).

Resources for Primary Caregivers

The following resources are provided to help primary caregivers address the personal needs and challenges of providing direct support to individuals with NDs.

Books and other written resources: There are a number of books and other written resources available to caregivers raising children with NDs. Although certainly not exhaustive, this list provides a sampling of available written works that have been recommended by others raising children with specialized needs.

- 1) *Swan Mothers: Discovering our True Selves by Parenting Uniquely Magnificent Children* by Natalia Erehnah (2012)
- 2) *You Will Dream New Dreams: Inspiring Personal Stories by Parents of Children with Disabilities* by Kim Schive and Stanley D. Klein (Editors) (2001)
- 3) *A Different Kind of Perfect: Writings by Parents Raising a Child with Special Needs* by Cindy Dowling and Neil Nicoll (Editors) (2006)
- 4) *The Elephant in the Playroom: Ordinary Parents Write Intimately and Honestly about the Extraordinary Highs and Heartbreaking Lows of Raising Kids with Special Needs* by Denise Brodey (2007)
- 5) *Where We Going Daddy?* by Jean-Louis Fournier (2010)
- 6) *When your Child has a Disability* by Mark L. Batshaw MD (2000)
- 7) *Special Children, Challenging Parents: The Struggles and Rewards of Raising a Child with a Disability* by Robert A. Naseef (2001)

Online resources and networks: There are also a number of online resources available to caregivers. The following includes a list of general resources as well as more specialized online supports for those caring for individuals with SMS, WS, and DS.

- 1) General Online Supports and Networks
 - a. Parents Helping Parents (www.php.com)
 - b. Parent to Parent USA (www.p2pusa.org/p2pusa/SitePages/p2p-links.aspx)
 - c. Alliance of Genetic Support Groups (www.geneticalliance.org/)
 - d. Exceptional Parent Magazine (www.eparent.com/)
 - e. Siblings of Kids with Special Needs, University of Michigan Health System (www.med.umich.edu/yourchild/topics/specneed.htm)
- 2) Smith-Magenis syndrome
 - a. Parents and Researchers Interested in Smith-Magenis syndrome (PRISMS; www.prisms.org)
 - b. Smith-Magenis syndrome: National Institutes of Health, Genetics Home Reference (ghr.nlm.nih.gov/condition/smith-magenis-syndrome)
- 3) Williams syndrome
 - a. Williams syndrome Association (www.williams-syndrome.org)
 - b. Williams syndrome: PubMed Health (www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002105/)
 - c. Williams syndrome: National Institutes of Health, Genetics Home Reference (ghr.nlm.nih.gov/condition/williams-syndrome)
- 4) Down syndrome
 - a. National Down syndrome Society (www.ndss.org)

- b. National Association for Down syndrome (www.nads.org)
- c. Down syndrome: PubMed Health (www.ncbi.nlm.nih.gov/pubmedhealth/PMH0001992/)

Finding individual and family therapists. There are many approaches to seeking out individual and/or family counseling services. Depending on the primary caregiver's comfort level, it may be beneficial to ask his/her primary care physician or the family's pediatrician about referral sources for therapy. Physicians are often well-networked within their respective communities and may be able to identify appropriate mental health professionals. As discussed in previous sections of this chapter, many children and adolescents with NDs participate in a variety of specialized therapy services. The professionals conducting these therapies may also be able to provide referrals for primary caregivers interested in engaging in psychotherapy. Alternately, individuals with medical insurance can go to the insurance company's website to find out more about the mental health benefits that are available to them and providers in their area. Depending upon the specific goals of therapy, it may be beneficial for the caregiver to seek out a therapist with a background in health psychology, as these care providers are more likely to have training in addressing caregiver needs. It is recommended that primary caregivers *consider asking potential therapists about their area of expertise* to help determine whether the therapist will be a good fit in meeting the person's needs (Martin, 2006; Stoppler, 2005). The following additional questions may also be helpful to ask potential therapists when determining who to see:

- 1) What is your approach to therapy? How are therapy sessions conducted (individually, group, family, etc.)?
- 2) What are my treatment options? How long can I expect treatment to take?
- 3) What are your views on the utilization of medication to treat mental health concerns?
- 4) Are there any alternative therapies or options to consider (e.g., changes in diet or exercise, biofeedback, acupuncture) that may be beneficial?
- 5) What is your level of education? What type of licensure do you have?
- 6) What are the costs for your services? What insurance do you accept? Are there alternate payment options?
- 7) What policies are in place for cancellations and/or emergencies?
- 8) What are the hours you are available? How often do I need to be seen for therapy?

Siblings

"My sister is very strong willed and knows what she wants to do. She doesn't let others influence what she does. She has a set plan in her mind and that's what she sticks to, and I love and hate that about her. Growing up with [my sister] was hard because I never understood why [she] was getting so much more attention from my parents. As well as why she got away with so much stuff. She did something and would get sent to her room for an hour, and I did the same thing and got grounded for two weeks, which seemed very unfair at the time. Now I understand it more...I still don't like it, but I understand why."

16 year-old sibling of 22 year-old diagnosed with DS

“I was 13 years old when [my sister] was born. She did change our lives considerably. It was stressful going thru junior high school and high school with her, as I was her main caregiver, because both my parents worked. When I came home from school, my responsibility was to care for both of my sisters. My younger sister was 8 years old when [our sister] was born, and she admits to being ashamed of her. She would not bring her girlfriends to the house because of [our sister]. I, on the other hand, being the oldest sibling, was involved in her life, to the extent that I used to babysit other developmentally disabled children who went to the same training center as [our sister]. She was certainly a challenge. Our parents are deceased now, and my younger sister and I are [our sister]'s conservators. It has been very hard in the later years, as we are both involved in her life and her care. She has now been diagnosed with Alzheimer's making this much more challenging to deal with. I would say that [our sister] has definitely changed our lives. Everyone in our direct family knows, loves, and understands [our sister], even the youngest of my grandchildren. [Our sister] has the mental age of 4 years old is 51 years old chronologically and has Alzheimer's...we are learning more every day, and each day has its own challenges.”

64 year-old sibling of 51 year-old with DS

“My responsibility for him is scary. I get stressed trying to keep him out of trouble and safe. I enjoy his hugs and his nickname for me because I know he loves me. He misses me when I am gone and expresses his joy to see me; that always makes me feel good. He teaches me patience, and he is very loving so he is easy to care about. He shows love and acceptance toward me and so I feel the same toward him. He has directed me to a goal for myself to be an advocate for the disabled. I also have gained a desire to learn sign language and other skills to work with the disabled.”

17 year-old sibling of 15 year-old with SMS

“It is TOUGH growing up with a SMS sibling. 'Fair' is not part of the equation. Do not get to do many things because of sibling. Do not get to do things with both parents...one is always with the SMS individual. Embarrassing situations. Hard to have friends over. Don't get to do casual trips. Everything must be planned out.”

Parent of a 17 year-old without SMS and a 15 year-old with SMS

It is important to consider the dynamic of the family and how family members both positively and negatively influence one another when determining how to best promote quality of life among the entire family, while effectively caring for a child with a ND. Research in this area has focused primarily on children with disabilities and their parents, while the way other children in the family are influenced has been often overlooked. Siblings of individuals with disabilities have unique experiences, concerns, benefits, and intra-familial relationship factors that directly result from living with a sibling with a disability. Conway and Meyer (2008) found that siblings experience many of the same emotional concerns as parents and other caregivers who provide direct support, including feelings of isolation and guilt, support demands, a need for information, and concerns about the future of the individual with a disability. In addition to these concerns, siblings may face unique issues not faced by their parents, such as resentment and embarrassment, social problems, and a pressure to achieve. While potential negative emotions and experiences have been reported, a number of positive experiences have also been acknowledged. One study on families with children

with Down syndrome and Rett syndrome found that siblings demonstrated increased tolerance, better awareness of differences, and greater maturity when compared to peers (Dyke, Mulroy, and Leonard, 2009). Siblings of children with NDs have also been described as having an especially caring and compassionate nature (Dyke et al., 2009). It should be noted that siblings' experiences vary depending on the type of illness or condition their sibling has. For example, siblings of individuals with SMS need to learn to adjust to and to cope with their sibling's severe maladaptive behaviors (Moshier et al., 2012). By comparison, siblings of individuals with WS or DS will likely experience the unique challenges and benefits associated with living with an individual who has significant medical and/or psychiatric problems. The typically developing sibling may feel that he or she has less in common with the sibling with SMS, WS, or DS, resulting in greater emotional separation between the siblings.

In addition to influencing the sibling relationship, the presence of a child with a disability in the family affects the relationship between the well-sibling and his/her parents (Moshier et al., 2012; Neece, Blacher, and Baker, 2010; Schuntermann, 2009). The greater the number and severity of stereotypic and self-injurious behaviors displayed by a child with SMS, the more time parents must spend addressing them and providing direct attention to the child with the disability (Moshier et al., 2012). In families of children with WS, parents may need to spend large quantities of time attending to the psychiatric concerns of the child with WS (Leyfer, Woodruff-Borden, and Mervis, 2009). Similarly, the presence of a child with DS in a family means that parents will likely spend more time addressing medical concerns for that child (Graff et al., 2012). This results in less time available for the parent to spend with the sibling (Schuntermann, 2009). Furthermore, the more difficult a child's temperament, the more pronounced the degree of differential parenting, defined as the extent to which a parent's expectations, treatment of, and quality of time spent with a child with a disability differs with regards to other children in the family (Rivers and Stoneman, 2008). Research has found that families of children with disabilities tend to show greater rates of differential parenting than families of children without disabilities. Siblings' responses to differential parenting range from resentment and anger to acceptance. Studies have found differential parenting to be associated with negative sibling outcomes, such as greater competition and increased conflict (Rivers and Stoneman, 2008; Taylor, Fuggle, and Charman, 2001). However, children who are able to understand why they are being treated differently from their sibling, and perceive this as being fair, tend to be more accepting of the parenting differences.

Resources for Siblings

The following resources are provided to help support siblings in addressing personal needs and challenges.

Books and other written resources: There are a number of books and other written resources available to siblings of individuals with NDs. This list provides a sampling of available written works that have been recommended by others living with and/or caring for those with NDs.

- 1) *A Boy Alone* by Karl Taro Greenfeld (2009)

- 2) *At Home in the Land of Oz: Autism, My Sister, and Me* by Anne Clinard Barnhill (2007)
- 3) *Brothers and Sisters* by Laura Dwight (2007)
- 4) *Brothers and Sisters of Disabled Children* By Peter Burke (2003)
- 5) *Thicker than Water: Essays by Adult Siblings of People with Disabilities* from Woodbine House (Editors) (2009)
- 6) *The Sibling Slam Book: What It's Really Like to have a Brother or Sister with Special Needs* by Don Meyer and David Gallagher (2005)

Online resources and networks: There are also a limited number of online resources available to siblings of individuals with NDs.

- 1) Sibling Support Project (www.siblingsupport.org)
- 2) Kids Health (www.kidshealth.org)

Finding individual and family therapists. Siblings of individuals with NDs may benefit from seeking professional assistance to help support appropriate coping and adjustment. It can be helpful for siblings to have a safe and confidential environment in which they can voice frustrations, ask questions regarding their sibling's diagnosis and prognosis, learn to strengthen relationships with siblings and parents, assess benefits, and improve their quality of life. It can be especially beneficial for siblings to have such supports during naturally developmentally stressful times such as the teenage years or when transitioning into adult caregiver roles. It is recommended that children and adolescent siblings seek out therapy with professionals specializing in NDs, such as pediatric psychologists. Many hospitals and communities also offer Sibshops, or group-based support systems, for siblings. A list of Sibshops can be found at www.siblingsupport.org/sibshops/index_html.

CONCLUSION

The ever-changing needs of individuals with NDs necessitate substantial specialized support, intervention, and resources across the lifespan. As these individuals age, each developmental period presents unique challenges for primary caregivers, who must learn to cope in such a way that promotes individual well-being and the family's overall quality of life. Understanding the potential concerns that may arise can help caregivers prepare to meet developmental needs in an active and adaptive manner. For infants and young children, developmental assessments can be utilized to identify strengths and weaknesses in such a way that leads to individualized therapies (e.g., speech and language, occupational, physical, and behavioral therapies) and initial academic planning. In middle childhood, psychoeducational evaluations and the resulting recommendations provided can further assist families in developing academic plans (e.g., IEPs) and continuing necessary therapies. Additionally, challenges with social skills and peer relationships may need to be continuously addressed. Throughout adolescence and beyond, challenges with puberty and romantic relationships, vocations, and adult living arrangements all need to be addressed. Vocational assessments can offer assistance in determining appropriate job-related skills and settings.

Considerations also need to be made with respect to individual decision-making capacities and whether guardianships or alternative to guardianships need to be pursued. This can often be a cumbersome and involved process for caregivers, including adult siblings, who often are charged with transitioning into caregiver responsibilities as parents age. Due to the intense demands associated with caring for an individual with a ND, caregiver and sibling needs must be considered and supportive resources implemented.

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