

THESIS

PREDICTORS OF FUNCTIONAL PERFORMANCE IN
SCHOOL-AGED CHILDREN WITH DOWN SYNDROME

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ABSTRACT

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This project examined whether executive functioning, language ability, and/or intelligence quotient predicts functional performance in children with Down syndrome, the most common neurogenetic syndrome associated with intellectual disability. Functional performance is the performance of tasks universal to all children-- such as self-care, mobility, and social interaction. Identifying patterns of functional performance in Down syndrome is critical as it is a foundation for optimal outcomes for the child, their family, and community. Executive functioning is an umbrella term used to describe thinking skills that are involved in goal-directed behavior. Children with Down syndrome are predisposed to specific areas of relative strengths and challenges in executive functioning, but it is unclear whether this phenotypic profile affects functional performance. Children with Down syndrome and students with mixed developmental disabilities were matched for mental and chronological age using the Leiter International Performance Scale-Revised. Functional performance and executive function were measured by parent-report, using the Pediatric Evaluation of Disability Inventory and the Behavior Rating Inventory of Executive Function- Preschool Version, respectively. Language and mental ability were measured using two standardized assessments, the Oral and Expressive Language Scales of the Oral and Written Language Scales and the Leiter International Performance Scale-Revised. Results indicated that children with

Down syndrome and children with mixed developmental disabilities had similar functional profiles with strength in mobility and relative challenges in social function and self-care. Executive function was the only significant predictor of functional performance for relative children with Down syndrome, while intelligence quotient was the only significant predictor of functional performance for children with mixed developmental disabilities. Findings suggest differential targets for interventions to improve functional performance outcomes in children with Down syndrome and mixed developmental disabilities.

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CHAPTER 1: INTRODUCTION

Down syndrome is the most common neurogenetic disorder associated with intellectual disability, with an estimated 6,000 live births annually (Parker et al., 2010). The prevalence of Down syndrome has increased by greater than 31% since 1979 (Shin et al., 2009) and is estimated to be present in 8 individuals per 10,000 people in the United States (Presson et al., 2013). Improved services and medical care, including deinstitutionalization and enhanced surgical interventions for congenital heart defect, have led to an increase in life expectancy for individuals with Down syndrome (Bittles & Glasson, 2004; Presson et al., 2013).

Individuals with Down syndrome receive a variety of intervention treatments to improve their life quality. These interventions are based on their individual strengths and challenges. Individuals may receive care from occupational therapists, speech therapists, physical therapists, physicians, special educators, and social workers. Most children with Down syndrome receive access to free, public special education programs as endorsed by the Individuals with Developmental Disabilities Act (National Institute of Health, 2014). In a prior study using data from the Special Education Elementary Longitudinal Study (SEELS) dataset (2003-2004), Fidler and colleagues (in press) examined the range of educational services that children with Down syndrome utilized. Parents of 104 children with Down syndrome, many of whom were placed in regular classrooms yet spent a majority of their day in the special education resource room, reported receipt of one or more of the following services: speech language (81.7%), occupational therapy (63.5%), physical therapy (29.8%), social work (25.0%), respite care (21.2%), audiology (19.2%), assistive technology/devices (17.3%), tutoring

(16.4%), psychological services (14.4%), reader/interpreter/signer (7.7%) and orientation/mobility services (2.9%) (Fidler, Most, & Daunhauer, in press). Due to the increased prevalence and improved life expectancy of this population, there is need for further research to provide foundational evidence for intervention development that will promote optimal outcomes and increased quality of life.

Individuals with Down syndrome have a high probability of demonstrating a distinct behavioral phenotype associated with their genotype, or genetic makeup, with areas of relative strength and challenge emerging in early childhood. A phenotype is an outcome of a particular genotype that includes aspects of behavior (Hodapp & Dykens, 2004). Behavioral phenotypes are probabilistic for individuals with neurogenetic disorders. Not all individuals within a particular neurogenetic disorder will display the characteristics associated with the disorder's behavioral profile, but they will have a higher probability of doing so. In addition, each neurogenetic disorder does not differ from other neurogenetic disorders on every behavior (Hodapp & Dykens, 2004; Hodapp, 2001; 2004). It is also important to note that behavioral phenotypes may become more pronounced over time, so that individuals who are older chronologically may display more pronounced patterns of strengths and challenges (Hodapp, 2001). Moreover, as children grow older, their experiences in their various environments may also influence the development of their strengths and challenges (Hodapp, 2001).

The behavioral profile of individuals with Down syndrome includes relative strengths in visuospatial processing, receptive language, and social relationships (Daunhauer & Fidler, 2011; Daunhauer & Fidler, 2013; Fidler, 2005; Fidler, Philosfky, & Most, 2009). Individuals with Down syndrome are also likely to show relative challenges

in motor skills and communication, in particular expressive language (Fidler, 2006; Fidler, Hepburn, & Rogers, 2006). The behavioral phenotype varies across children with Down syndrome. While children with Down syndrome are more likely to demonstrate this pattern of behavior, not all children with Down syndrome will show these behaviors. In addition, these behaviors may not be unique to children with Down syndrome. Other etiological groups may also display these strengths and challenges (Daunhauer & Fidler, 2013; Fidler, 2005; Fidler, 2006; Fidler, Most, Booth-Laforce, & Kelly, 2008; Fidler & Daunhauer, 2011; Hodapp & Dykens, 2004; 2012; Hodapp, 1997; 2001; 2004).

Although the past few decades of research have focused on the characterization of the behavioral phenotype of individuals with Down syndrome, there is a paucity of research examining how the behavioral phenotype relates to functional performance among children with Down syndrome (see Daunhauer, 2011 for a review). According to the Individuals with Disabilities Act (IDEA, 2004), functional performance refers to tasks an individual performs outside of an academic context, or routine tasks of daily living. Types of functional performance tasks include self-care such as dressing, aspects of social function, including playing games with peers, and aspects of mobility, such as going up and down stairs (National Dissemination Center for Children with Disabilities, 2010). Children with Down syndrome and other developmental disabilities may have difficulty performing one or more of these types of tasks, which may decrease health and well-being and increase family burden (Schieve et al., 2011). The proposed study aims to: (1) examine between-group and within-group differences in functional performance profiles for children with Down syndrome and children with mixed developmental disabilities matched for chronological (CA) and mental age (MA), (2)

identify to what extent intelligence quotient (IQ), language ability, and executive function predict functional performance in children with Down syndrome and children with mixed developmental disabilities, and (3) determine whether the predictors of functional performance differ for children with Down syndrome and children with mixed developmental disabilities matched for CA and MA. Describing the functional performance profiles of children with Down syndrome will inform targeted intervention approaches for practitioners, parents, and teachers by identifying areas of strength and challenge.

CHAPTER 2: BEHAVIORAL PHENOTYPE AND THE FUNCTIONAL PROFILE IN CHILDREN WITH DOWN SYNDROME

Down syndrome is caused by an extra chromosome on chromosome 21 in 95% of individuals with Down syndrome (CDC, 2014). Translocation causes about 4% of occurrences of Down syndrome and involves part of chromosome 21 being translocated on other chromosomes (CDC, 2014). Mosaicism accounts for approximately 1% of occurrences of Down syndrome and involves only some cells having three copies of chromosome 21 (Shin, Siffel, & Correa, 2010). Individuals with Down syndrome typically experience both intellectual and developmental delays (NIH Down Syndrome Working Group, 2007). In 2007, the NIH Down Syndrome Working Group created research objectives regarding research on Down syndrome including, but not limited to four areas: 1) identifying more information about the cognitive phenotype, 2) investigating the effectiveness of interventions, 3) performing longitudinal studies, and 4) exploring new intervention techniques. In the revised plan, there is greater emphasis on expanding research on epigenetic and environmental determinants that facilitate or constrain the development of the Down syndrome behavioral phenotype (NIH Down Syndrome Working Group, 2014). Examining the current body of inquiry on the Down syndrome phenotypic profile is closely aligned with this NIH research plan and critical to identifying potential targets for interventions involving children with Down syndrome.

Over the past two decades, there has been growing research characterizing the behavioral phenotype of Down syndrome across the lifespan. Variations in the behavioral phenotypes of individuals is dependent on the overexpression of specific genes, as well as interactions between genes and environmental factors such as home

residential settings versus institutionalization or access to early intervention services (Chapman and Hesketh, 2000). Hodapp (2004) hypothesized that these environmental factors have direct and indirect effects on individual differences of the behavioral phenotypes among individuals with neurogenetic disorders. Children with Down syndrome may be more likely to choose environments that reflect their strengths and challenges in specific areas of activities. Rosner and colleagues (2004) examined the activities in which children with Down syndrome, Prader-Willi syndrome, and Williams syndrome participated. Children with Down syndrome were more likely to participate in arts and crafts and other visual-motor activities (Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). The following section reviews the body of evidence regarding the behavioral phenotype of individuals with Down syndrome in the major domains of development.

Cognitive development. Individuals with Down syndrome typically have IQs in the mild to moderately low range (CDC, 2013). Throughout childhood, IQ in Down syndrome steadily declines (Hodapp, Evans, & Gray, 1999). Children with Down syndrome under 3 years of age typically have an IQ in the 60s and 70s. IQ in children with Down syndrome decreases to the 40s and lower 50s at 5-7 years and by 9-11 years decreases to the upper 30s and lower 40s (Hodapp et al., 1999). The reasons for this decline remain poorly understood, however patterns of strength and challenge have been identified in areas of cognitive development.

The Down syndrome cognitive phenotypic profile likely emerges in early infancy and there is evidence for emerging strengths in visual processing from this period of development (Fidler, 2005). Overall, infants and children with Down syndrome have

relative strengths in visual processing and imitation and have relative challenges in visual exploration and attention (Brown et al., 2003; Carney, Henry, Messer, Danielsson, Brown, & Ronnberg, 2013b; Fidler et al., 2006). In a study examining early learning in toddlers with Down syndrome, toddlers demonstrated strengths in visual processing (Fidler, Hepburn, & Rogers, 2006). Delays in visual processing and receptive language tasks were not observed at this age as they have been in older children with Down syndrome (Klein & Mervis, 1999; Wang & Bellugi, 1994). These findings support the concept that the behavioral phenotype is not static, but a dynamic process that is important to consider across the lifespan (Fidler et al., 2006).

In school-aged children with Down syndrome, relative strengths in visuospatial processing emerge (Fidler, Most, & Philofsky, 2009). Researchers examining CA-matched middle to high school aged students with Down syndrome and Williams syndrome and MA-matched typically developing peers, found that the group with Down syndrome demonstrated within-group relative strength in visuospatial short-term memory and relative challenge in verbal short-term memory. However, Carney and colleagues found no statistically significant differences between individuals with Down syndrome and individuals with Williams syndrome for verbal short-term memory and the rate of visuospatial short-term memory. Children with Down syndrome had significantly lower performance on the verbal short-term memory task as compared to their performance on the visuospatial memory task. This lower performance remained consistent over time (Carney, et al., 2013b). This finding suggests domain specific challenges in verbal short-term memory in individuals with Down syndrome (Carney et al., 2013b).

Additionally, a handful of researchers also have examined the Down syndrome phenotype from an information processing perspective. In this work researchers have compared event-related potentials (ERPS) in infants with Down syndrome to typically developing infants with mixed results. For example, Karrer and colleagues (1995) found that the ERP morphology was similar in infants with and without Down syndrome, indicating that similar underlying mechanisms for processing speed may be present in infants with and without Down syndrome. In addition, infants with Down syndrome performed slightly faster than their typically developing peers on the visual memory task and at a similar pace on focused attention and visual fixation (Karrer, Wojtascek, & Davis, 1995). However, in another study on ERPs in infants with Down syndrome, when compared to their typically developing peers, the infants with Down syndrome demonstrated challenges in habituation to visual stimuli, a skill that requires visual memory (Karrer, Karrer, Bloom, Chaney, & Davis, 1998). While this small amount of evidence in the literature using neurological measures has mixed findings, research using behavioral assessments of information processing indicates that infants and toddlers with Down syndrome demonstrate delays in visual exploration and sustained attention (Brown et al., 2003; Gunn, Berry, & Andrews, 1982; Miranda & Fantz, 1973).

Executive function. Executive function is an umbrella term describing complex cognitive processes involved in goal-directed behavior, such as working memory, shifting, inhibition and planning (Carlson, 2005; Kerr & Zelazo, 2004; Zelazo & Muller, 2011). Executive functions may predict academic achievement in typically developing children (Brock, Rimm-Kaufman, Nathanson, & Grimm, 2009). For example, more competent performance on aspects of executive functioning have been found to be

related to higher levels of math and literacy skills in typically developing children and children at-risk for developmental delays (for a review see Daunhauer & Fidler, 2011, 2013). Duckworth and Carlson (2013) further proposed that executive functioning may play a larger role than intelligence in certain academic and life outcomes. Harms and colleagues (2014) conducted a longitudinal study and found that the executive function of inhibitory control at age 8 predicted academic competence at age 12 independent of verbal ability. The authors speculated that executive function might be more predictive than IQ due to its role in the foundational skills required for later development of cognitive abilities (Harms, Zayas, Meltzoff, & Carlson, 2014). For example, having the ability to inhibit automatic responses and the ability to be cognitively flexible can help a student learn and apply new materials (Duckworth & Carlson, 2014; Harms et al., 2014). Several studies have also shown associations between executive functioning difficulties and functional performance outcomes for individuals with developmental and intellectual disabilities (Gilotty et al., 2002; Tazari et al., 2007; Zingerevich & LaVesser, 2009).

Relative to research involving children with mixed developmental disabilities, there is a paucity of research on executive function for children with Down syndrome (Daunhauer, Fidler, & Will, 2014b). Emerging research indicates that school-age children and adolescents with Down syndrome demonstrate a profile of relative strengths and challenges in executive functioning including: relative strengths in shifting (Daunhauer et al., 2014a), and challenges in planning (Fidler et al., 2005bc; Fidler et al., 2006; Kasari & Freeman, 2001) and in working memory (Carney, Brown, & Henry, 2013; Costanza et al., 2013). Planning difficulties have been observed in young children with Down syndrome (Fidler, Hepburn, Mankin, & Rogers, 2005) and are also evident in

older children with Down syndrome (Daunhauer et al., 2014a; Fidler et al. 2015; Lee et al., 2011). Fidler, Hepburn, Mankin and Rogers (2005) examined nonverbal requesting and problem solving in toddlers with Down syndrome and found that children with Down syndrome had difficulties with problem solving, revealing early deficits in goal-directed behaviors. This study was one of the few that examined goal-directed behaviors in individuals with Down syndrome.

However, the existing data use a variety of methods including behavioral tasks and informant-report on executive function in everyday contexts. In addition, there is little replication in examining skills over time. Studies on planning and working memory indicate more pronounced deficits specific to Down syndrome (Fidler, Most, & Philofsky, 2009; Costanzo et al., 2013; Carney, Brown, & Henry, 2013a). However, existing studies on inhibition in Down syndrome have mixed evidence (Lee et al., 2011; Daunhauer & Fidler, 2013; Daunhauer et al., 2014a). In one study, individuals with Down syndrome had lower performance on a verbal task of inhibition, but not a visual task measuring inhibition (Constanzo et al., 2013). In another study, children and adolescents with Down syndrome did not differ from MA-matched typically developing children on inhibition tasks (Carney et al., 2013a). In addition, there is mixed evidence in the literature for problems in cognitive flexibility (shifting) in individuals with Down syndrome. One study found that five-year-olds with Down syndrome performed a delayed non-matching to object task significantly better than were their developmentally matched peers with autism (Dawson et al., 1998). Using a caregiver report of executive function, Daunhauer et al. (2014a) found that children with Down syndrome did not perform significantly differently from mental age-matched peers on items measuring

children's ability to flexibly solve problems or switch attention in everyday contexts. Conversely, in another study, almost 41% of a group of school-aged children with Down syndrome could not shift on the dimensional card sorting task (Edgin, 2003) and another study with children, adolescents, and adults with Down syndrome and Williams syndrome revealed that individuals with Down syndrome had significantly lower performance on the shifting tasks (Costanzo et al., 2013). Clearly more research is needed to understand these divergent findings.

Some of these divergent findings may be due to measurement difficulties. Research regarding test-retest reliability has been limited to tasks measuring executive function (Beck et al., 2005). Wiebe et al. (2011) conducted a confirmatory factor analysis that concluded that a single latent EF variable was the most parsimonious model for 3-year-old children. Yet, McAuley and White also used confirmatory factory analysis and found different abilities in inhibition, working memory, and processing speed from 6 to 24 years (McAuley & White, 2011). In many studies, the executive function batteries are composed of entirely different tasks, making it difficult to compare and replicate across studies. However, there have been great efforts to establish laboratory tasks to measure executive function developmentally young children (Espy, Kaufmann, Glisky, & McDiarmid, 2001; see Carlson, 2005 and Garon et al., 2008 for a review). In addition, there are many studies that are using similar tasks for preschoolers which is helping to establish validity and reliability for some tasks, however more work is needed to establish measures that underlie common definitions of executive function to be replicated across age groups, and in typical and atypical populations (Carlson, 2005). In addition, the use of informant-report in contrast to laboratory tasks is an

important methodological issue with implications for intervention development. This study utilized a parent report on executive function that is a reliable measure of executive function in everyday life.

Given there is no universally accepted executive function battery for laboratory-based tasks, and that previous research has established that parent-reported executive function skills may be associated with functional skills (Gilotty et al., 2002; Mangeot et al., 2002; McClean et al., 2014; Pugliese et al., 2014), this study used a parent report measure allowing for the examination of everyday executive function skills and how they related to functional performance skills. Furthermore, more research is needed to explore executive functioning in Down syndrome and its relationship with other factors such as academic achievement and functional performance (Daunhauer et al., 2014b) to provide further understanding and implications for intervention. The proposed study aims to explore the relationship between executive functioning and functional performance.

Language development. An additional area to consider in the Down syndrome phenotypic profile is language development. Infants and children with Down syndrome show areas of relative strength in receptive language (Miller & Leddy, 1998). Infants with Down syndrome demonstrate strengths in prelinguistic vocalizations and receptive language, but delays in the development of expressive language beginning with relative challenges in vocal imitation (Fidler, 2005; Mahoney et al., 1981; Miller & Leddy, 1998). Although infants with Down syndrome experience language delays in infancy, an increased learning of language occurs during toddlerhood with a growth in vocabulary size occurring during this time (Fidler, 2006). Toddlers with Down syndrome also show

within-syndrome pattern of strengths in receptive language and weaknesses in expressive language (Fidler et al., 2006). In a study examining the relationship between expressive language and cognitive development, toddlers with Down syndrome showed less instrumental, or object, requesting than the children with a mixed etiology of mental age- matched developmental disabilities and mental-aged matched typically developing children. However, the toddlers with Down syndrome did not show a deficit in social requesting (Fidler et al., 2005c). Relative strengths in receptive language and relative weaknesses in expressive language become more pronounced during childhood (Fidler et al., 2009). Nonverbal communication skills facilitate social interactions for children with Down syndrome and are considered a relative strength (Fidler et al., 2005c).

Social development. The use of social behaviors in everyday life is referred to as social function, a component of functional performance (Haley, Coster, Ludlow, Haltiwanger, & Andrellos, 1992). Social development has been considered an area of relative strength for individuals with Down syndrome. Children with Down syndrome demonstrate aspects of relative strengths in joint attention and nonverbal socioemotional functioning, including developing relationships with others and emotionally signally as observed increased smiling behavior in infants (Carvajal & Iglesias, 2000). However, recent research indicates that individuals may also demonstrate specific challenges in areas of social development including recognizing intentions (Hahn, Fidler, Hepburn, and Rogers, 2013) and recognizing emotions (e.g. Adamson, Bakeman, Deckner, & Ronski, 2009), as well as the over-use of social skills to avoid challenging tasks (e.g. Cebula 2010; Fidler, 2005b; Fidler et al., 2009).

Relative strengths in social development. An aspect of positive emotional signaling includes smiling frequency. Fidler, Barrett, and Most (2005) examined age-related smiling frequency in children, adolescents, and adults with Down syndrome. They found that while smiling is frequent in childhood its frequency decreases with age. This finding is important when considering that the behavioral phenotype may not be static across the lifespan, so it is important to examine the phenotype across different age ranges and longitudinally (Fidler et al., 2005b).

Levels of joint attention have been found to be appropriate for their developmental level in children with Down syndrome (Wishart, 2007). Relatedly, Fidler and colleagues (2008) examined the function of social behaviors including orientation and social engagement in children with Down syndrome. They examined the emergence of these social skills in infants and toddlers with Down syndrome and a mental age-matched comparison group of children with developmental delays. The authors found that young children with Down syndrome had relative strengths in orienting and engagement behaviors compared to other areas of development. They also found that developmental competence of these behaviors grew significantly more rapidly from 12 months to 30 months than in the comparison group.

Challenges in recognizing emotion. When considering the social behaviors displayed by infants with Down syndrome, heightened attention to people could indicate poorer ability to switch attention efficiently between people, objects, and the environment. While infants with Down syndrome do spend time sharing attention with objects and adults, they spend less time coordinating attention (for a review see Cebula et al., 2010). In early childhood, a study revealed that during interactions with parents,

children with Down syndrome were more likely to be unengaged during contexts encouraging instrumental requesting than during contexts encouraging simple interactions (Adamson et al., 2009). This pattern was not observed in typically developing children (Adamson et al., 2009). Children with Down syndrome also made fewer social referencing looks than typically developing children (Adamson et al., 2009). Difficulties in emotion recognition and/or the ability to use the information to guide behavior are also important to consider. Emotion recognition difficulties have not been well researched in individuals with Down syndrome.

Hahn and colleagues (2013) examined the relationship among aspects of social cognition, intentionality, and intersubjective skills, joint attention, and affect sharing in children with Down syndrome and developmental disabilities. Intentionality is an individual's ability to interpret someone else's prescribed meaning e.g. understanding the intended goal of a task such as putting keys in a cup when the individual watches someone attempt it unsuccessfully. Intentionality assesses the social cognition processes of social problem solving. Results indicated that higher rates of affect sharing in children with Down syndrome were associated with poorer intention reading abilities for children with Down syndrome. This finding was unique to children with Down syndrome, as the observed pattern was not observed in children with other developmental disabilities matched for MA and CA (Hahn et al., 2013). These findings highlighted how children with Down syndrome may not use their social skills to effectively problem solve and function in social situations.

Co-morbid autism spectrum disorder. It should also be noted that some individuals with Down syndrome have specific challenges in social behavior that result

in a co-morbid diagnosis of autism spectrum disorder (ASD; Fidler, 2006). In a population-based study of ASD in children with Down syndrome ages 2-11 years, the prevalence was estimated to be approximately 7% (DiGiuseppi et al., 2010). This prevalence rate in children with Down syndrome is higher than in the general population (Frieden et al., 2014). Symptoms of individuals with Down syndrome that meet the criteria for ASD include: poor use of eye gaze, restricted interests, and social isolation as observed during activities involving communication and play (Hepburn et al., 2008).

Using social skills to avoid challenges. Although the development of social skills may help children adjust to their environment, children with Down syndrome may use these very skills to opt out of other opportunities for learning (Fidler, Most, Booth-LaForce, & Kelly, 2008). Fidler and colleagues (2005b) found that young children with Down syndrome were more likely to ask for help when presented with a challenging task (object retrieval) than two comparison groups: (1) children with mixed developmental disabilities matched for both MA and CA; and (2) a group of children with typical development matched for MA. Another study found that children with Down syndrome used social-based strategies to complete challenging object and sorting tasks (Wishart, 2007). However, the social-based strategies used were inappropriate and counterproductive to the task at hand. For example, children with Down syndrome would increase eye contact with their partner rather than the task. This study revealed the possible difficulties in core aspects of interpersonal functioning in Down syndrome, and although individuals are highly sociable, there may be qualitative and quantitative differences in how social cognition develops and is applied in learning contexts (Wishart, 2007). This overuse of social strategies observed within children with Down

syndrome may have a large downstream effect on development as missed opportunities for learning may lead to greater developmental delays (Cebula et al., 2010; Pitcairn & Wishart, 1994). Finally, the over-use in social skills to avoid challenging activities and tasks may be related to the lower levels of mastery motivation observed from infancy to the early school years in this population (Glenn, Dayus, Cunningham, & Horgan, 2001; Gilmore, Cuskelly, & Hayes 2003; Ruskin et al., 1994).

Overall, individuals with Down syndrome display both patterns of strength and challenge in social skills. However, it is not entirely clear how these social skills relate to the performance of social tasks. There is a need to examine how these social skills relate to functional performance in everyday life. Social functioning is a component of functional performance that helps children perform everyday tasks. In this study, we examine predictors of social functioning of children with Down syndrome and mixed developmental disabilities.

Motor development. An additional area of challenge for individuals with Down syndrome is motor development. Individuals with Down syndrome typically have low muscle tone (CDC, 2013). Motor functions are required for many of the tasks involved in functioning of everyday life, such as the ability to hold a toothbrush and move throughout a room (Frank & Ebsensen, 2014; Haley et al., 1992).

Motor development is delayed from infancy in children with Down syndrome, although it follows a similar developmental sequence to that of typically developing children (Frank & Ebsensen, 2014; Palisano et al., 2001). In children and adolescents with Down syndrome, motor impairments are apparent in fine and gross motor tasks and praxis skills (Fidler et al., 2009). Fidler, Hepburn, Mankin, and Rogers (2005)

examined praxis skills in young children with Down syndrome, mixed developmental disabilities, and typically developing children using parent self-report. They found that children with Down syndrome demonstrated lower (less competent) scores on the motor functioning scale than their MA- matched peers without disabilities and children with other disabilities. They also found that children with Down Syndrome performed significantly lower on both the praxis battery and the object retrieval task, and that there was a positive association between praxis skills and daily living skills in the children with Down syndrome. These findings suggest that a specific profile of praxis difficulties in young children Down syndrome, and evidence of an association between praxis and performance of mobility tasks in children with Down syndrome (Fidler et al., 2005b).

Over the past two decades, relative strengths and challenges have been researched in aspects of cognitive, language, social, and motor development for children with Down syndrome. There has been less research on associations between the behavioral phenotype and aspects of everyday living, including functional performance and adaptive behavior (Daunhauer, 2011).

Functional Performance and Adaptive Behavior

Functional performance and adaptive behavior are two related constructs that capture the extent to which individuals carry out important tasks of everyday life. Adaptive behavior refers to those skills that individuals need to meet the demands of everyday life including self-care, safety, and money management (Sparrow, Cicchetti & Balla, 1984). For example, adaptive behavior has been found to account for 42% to 46% of the variance in both residence and work independence for adults with intellectual disabilities (Woolf, Woolf, & Oakland, 2010). Functional performance, a term

from rehabilitation science, is related to the concept of adaptive behavior. Functional performance refers to activity performance, but in a specified range of everyday activities ubiquitous in childhood (e.g., getting dressed, brushing teeth, and playing with peers; Haley et al., 1992). Functional performance stems from a rehabilitation perspective that takes into account varying contexts and levels of assistance and/or accommodation (Daunhauer, Gerlach-McDonald, & Khetani, 2014c). Assistance includes the involvement of an individual aiding the child to complete the task; whereas accommodations involve modifying the task so that the child is able to complete the task (Haley et al., 1992).

Understanding and examining predictors of functional performance and adaptive behavior is critical for building knowledge to inform the design of targeted interventions that promote positive life outcomes in children with Down syndrome (Haley, Raczek, Coster, Dumas, & Fragala-Pinkham, 2005) for their families. In a study involving mothers of children and adults with Down syndrome, Bourke and colleagues (2008) found that mothers had better mental health scores when their children required less assistance with functional activities such as self-care and problem solving. In another study, higher levels of functional performance increased the likelihood of obtaining employment after high school in older individuals with Down syndrome (Foley et al., 2012). While not specific to Down syndrome, a recent study by Khetani and colleagues (2013) found that difficulties in functional abilities are associated with greater restrictions in participation in community activities for preschool-aged children who received early intervention services due to having IDD or being at risk for a developmental delay. Difficulties in mobility, toileting, speech, feeding, safety awareness, and friendships were significantly

associated with participation difficulties in 7-9 activities including neighborhood outings, community-sponsored activities, and recreation and leisure. Indicators of functional abilities, rather than the individuals' service eligibility, were associated with community participation difficulty. These findings highlight the critical importance of examining predictors of functional performance in young children with Down syndrome to facilitate the onset of positive developmental trajectories leading to life optimal outcomes (Rhitman et al., 2010).

The Down syndrome functional performance profile appears to be measurable in early childhood (e.g. Daunhauer, 2011; Dykens, Hodapp, & Evans, 1994; Fidler, Hepburn, & Rogers 2006) with more pronounced differences in relative strengths and challenges detectable with older age groups (Coe et al., 1999; Dolva et al., 2004; Dolva et al., 2007; Loveland & Kelley, 1988; 1991; Rihman et al., 2010; Volman et al., 2007). These findings regarding functional performance profiles becoming more detectable with age corresponds with current thinking regarding neurogenetic phenotypic profiles changing dynamically over time (Fidler, Osaki, & Hepburn, 2011). In both adaptive behavior and functional performance for individuals with Down syndrome, a distinctive profile of relative strength and challenges emerges beginning in early childhood that suggests socialization and mobility as relative strengths and self-care and communication as relative challenges (for a review see Daunhauer, 2011).

Functional performance and adaptive behavior in Down syndrome. Volman, Visser, and Lensvelt- Mulders (2006) examined the relationships among motor ability, mental ability, and the performance of functional skills in Dutch children with Down syndrome. Results revealed that limitations were found in the self-care and social

function domain, whereas children performed relatively well on measures of mobility. Within the Down syndrome participants, inter-individual variability was large across all domains. The study found that measures of motor ability were more predictive of functional performance than mental ability. This finding may provide the implication that focusing on underlying motor abilities will improve functional skills. However, as noted by Daunhauer (2011), the measure they used for mental ability potentially incorporated some aspects of motor ability, and involved only one task to assess intelligence (Volman, Visser, & Lensvelt-Mulders, 2006). This study highlights the importance of how understanding underlying skills may provide the foundation for intervention development. This study also demonstrated the importance of using reliable and valid measures of the constructs in order to validly examine the associations between mental ability and functional performance. The current study aimed to examine the relationship between mental ability and functional performance with a measure that examines multiple aspects of mental ability due to the paucity of current literature on this relationship.

Additional studies indicate a specific profile of functional performance for children with Down syndrome. Dolva, Coster, and Lilja (2004) examined parent-reported functional performance in 5-year old children with Down syndrome in Norway. For the self-care domain, parents reported that their children had more difficulties in activities requiring fine motor skills. Parents' main concerns in regards to their child starting school were language functioning and self-care performance (Dolva, Coster, & Lilja, 2004). In a follow-up study, Dolva, Lilja, and Hemminsson (2007) used the PEDI to examine the relationship of age of entry into mainstream education and the functional

performance skills of Norwegian children with Down syndrome. Results revealed that functional performance skills were lower on the self-care and social functioning domains for students who postponed entry into mainstream education classrooms than students with Down syndrome who entered the classroom at six years. The researchers concluded that specific skills seemed to be desirable for students to enter mainstream schools and that this may be culturally dependent (Dolva, Lilja, & Hemminsson, 2007). This study highlights how functional skills have important implications for intervention and education, and how focusing on foundational aspects of functional skills early in childhood may be beneficial and contingent on the culture. It should be noted that neither of these studies had comparison groups.

Only a few studies have used comparison groups when examining functional performance in children with Down syndrome. In a study examining within group and between group comparisons, Mancini et al. (2003) examined functional performance in children with Down syndrome at 2 years of age and 5 years of age to chronologically age-matched typically developing children. In children with Down syndrome, social function was a relative challenge in both age groups as compared to typically developing children. However, each group only had 10 children in the sample size (Mancini et al., 2003). Further research is warranted with larger sample sizes and comparison groups in order to examine between group differences to further characterize the behavioral phenotype of Down syndrome.

In addition, the Vineland Adaptive Behavior Scales (VABS) has been used to assess adaptive behavior (Fidler, Hepburn, & Rogers, 2006). In one study that used the VABS, toddlers with Down syndrome demonstrated slightly elevated scores on the

socialization domain, and no differences on the communication, daily living skills, or motor skills from typically developing children or children with other developmental disabilities. However, within group differences were observed among toddlers with Down syndrome in the relative strengths in socialization and relative weaknesses in communication (Fidler et al., 2006). Dykens, Hodapp and Evans (1994) examined children from 1-11 years of age using the VABS and found relative strengths in daily living skills and socialization skills and relative weaknesses in expressive communication (Dykens, Hodapp, & Evans, 1994). However, other studies have led to more evidence supporting the adaptive profile that includes relative strength in socialization and relative challenges in communication and daily living skills (Loveland & Kelly, 1988, Loveland & Kelley, 1991; Rihtman et al., 2010). Through examining within-group effect sizes across the above-described research, Daunhauer (2011) found that a distinctive profile emerges after early childhood that suggests daily living skills, also known as self-care, may be a relative challenge to everyday functioning in addition to communication for this population (Daunhauer, 2011). However, more research is needed to examine this profile and determine whether this profile is specific to individuals Down syndrome.

By further characterizing the specificity of the behavioral phenotype in Down syndrome, including aspects of functional performance, potential areas for targeted interventions could be identified to promote optimal development (Daunhauer, 2011; Daunhauer & Fidler, 2013).

Examining the behavioral phenotype from the perspective of the International Classification of Functioning (ICF) is critical to conceptualizing function for children with

Down syndrome. Additionally, this framework can provide a common language for both research and intervention.

International Classification of Functioning (ICF)

The International Classification of Functioning (ICF) was developed by the World Health Organization (WHO) to classify types of disability and functioning (World Health Organization, 2001 2002, 2013). Functioning is defined by the ICF as being the dynamic interaction between an individual's environment, health, and personal factors. The ICF provides a way to measure disability by conceptualizing disability within both medical and social models. Regardless of the presence or absence of a diagnosis, the ICF can be utilized to describe functioning in an individual, including positive and negative aspects. The relationships between health condition, environmental, and personal factors are dynamic and complex, and it is important to collect information about these factors and explore the associations (WHO, 2013). The umbrella term of functioning includes the domains of: body structure and functions, activities, and participation. The ICF aims to provide a universal common language for individuals across different fields and disciplines (WHO, 2002, 2013). An example is using the definitions and classifications for individuals with developmental disabilities in order to help coordinate care and services. However, it is important to recognize that the ICF is applicable to all people in all contexts in describing their functioning and health (WHO, 2013).

A major drawback to the ICF and ICF-CY is the ambiguity in use of the terms activity and participation that has resulted in multiple interpretations of these two concepts. Examples of activities according to the ICF include dressing, grooming, and

walking, etc., while participation entails work, recreation, education, and community involvement, etc. There has been considerable debate about how to use ICF language to describe participation, and it has been criticized for being inadequate for guiding development of measurements to assess levels of functioning (Dijkers, 2010). Another difficulty highlighted in the literature regarding the ICF was that it was not applicable to assessing functioning in children (Ostenjo et al., 2006). In order to address some of these issues, the World Health Organization developed the ICF-CY specifically for children and youth in 2007.

Functional performance focuses on the skills a child uses to partake in everyday activities whereas participation focuses on the involvement in life situations (Bedell et al., 2013). In Adolfsson and colleagues' (2011) review on measures of functional performance and participation, measures of functional performance often children from infancy to early childhood and measures of participation assess children from preschool-adolescence. The current study focused on the relationship between foundational developmental skills, executive functioning, and functional performance. Examining functional performance for individuals with Down syndrome in this manner may be critical for developing targeted interventions that facilitate optimal outcomes.

Predictors of Adaptive Behavior in Developmental Disabilities

There is a paucity of research specifically examining the relationship of IQ and functional performance for individuals with Down syndrome. One study has found significant correlations between IQ and adaptive behavior in middle-late childhood, in that higher IQ scores are associated with higher adaptive behavior performance (Rhitman et al., 2010). There is emerging evidence for a relationship between executive

function, whether using parent–reported measures or laboratory tasks, and adaptive behavior in individuals with other developmental disabilities. Studies in children with autism spectrum disorder (ASD) in late childhood and adolescence have found significant relationships between the overall general executive dysfunction as reported by parents and problems with adaptive behavior (Gilotty et al., 2002; McClean et al., 2014; Pugliese et al., 2014; Liss et al., 2001). In a study that utilized both a parent-report measure and laboratory-based measure of executive function, only the parent-report measure of executive function was significantly associated with adaptive behavior (Pugliese et al., 2014).

Studies that only utilized laboratory tasks found mixed results for children with autism. In a study comparing a group of elementary school-aged children with autism to a group of peers with developmental language disorder, researchers found that one task examining executive function was associated with the daily living skills domain of adaptive behavior for the group with autism. However, multiple executive function tasks were significantly correlated with adaptive behavior for the group with developmental language disorder. The results indicated that executive function may not play a role in adaptive behavior for elementary school-aged children with autism but may for children with developmental language disorder (Liss et al., 2001), however, given the modest sample sizes these results should be considered preliminary and subject to further replication. In another study in children with ASD only a planning-based task was associated with adaptive behavior (Ozonoff et al., 2004).

Moreover, further studies have examined predictors of adaptive behavior in individuals with traumatic brain injury (TBI), attention deficit hyperactivity disorder

(ADHD) heavy prenatal exposure to alcohol, and oppositional defiant disorder. Overall, executive dysfunction as measured by parent-report was associated with adaptive behavior for children with TBI (Mangeot et al., 2002). In addition, higher performance on laboratory-based tasks was associated with higher adaptive behavior scores 3 years after injury, but not for children 3 months after injury (Brookshire et al., 2011), suggesting that executive dysfunctions may become more pronounced over time. Similarly, higher performance on laboratory-based executive function tasks was also associated with higher adaptive behavior scores for children with ADHD (Clark et al., 2002; Ware et al., 2012), histories of prenatal alcohol exposure (Ware et al., 2012), and children with Oppositional Defiant Disorder (Clark et al., 2002).

Overall, these studies provide evidence towards the relationship between executive function and adaptive skills in a variety of developmental disorders. For studies that used both laboratory-based tasks and parent-report measures of executive function, parent-report measures were better predictors of adaptive outcomes (Mangeot et al., 2002; McClean et al., 2014; Ware et al., 2002). In a study that used a typically developing control group, the relationship between executive function and adaptive behavior was not significant, suggesting that a relationship between executive function and adaptive behavior may be unique to children experiencing executive dysfunction (Ware et al., 2012). However, more research is warranted to examine the relationship between executive function and adaptive skills. The current study aimed to examine the relationship between executive function and functional performance, as functional performance also incorporates the level of assistance required to perform tasks in everyday life. Given the findings in other developmental disorders, examining predictors

of functional performance in children with Down syndrome could help provide needed targets for phenotypic-specific interventions for children with Down syndrome.

Predictors of Functional Performance in Children with Down syndrome

Based on the current state of inquiry in this area, possible predictors of functional performance include cognitive development, language development, and executive functioning as these areas are specific areas of relative challenge in children with Down syndrome (Daunhauer, Fidler, & Will, 2014; Daunhauer & Fidler, 2013). The relationship between mental age and functioning in everyday life is well established in children with intellectual disabilities in general (for a review, see Harrison & Boan, 2004), and the demands required in functional performance (i.e., getting dressed) require executive functioning. It is critical to explore the relationship between executive function and functional performance in children with Down syndrome given the specific challenges in executive functioning reported by Lee et al. (2011) and Daunhauer et al. (2014a). In addition, Daunhauer et al. 2014b's study examining school function and executive function found executive function to be the only significant predictor of functional performance in the school context. However, it should be noted that Daunhauer et al. (2014b) did not include a comparison group and did not include function in the context of everyday home life. Further characterizing executive function and functional performance of a child interacting at home with a parent could provide innovative targets for intervention.

The purpose of this study was to examine the relationship between the Down syndrome phenotypic profile and functional performance by answering the following questions:

1. Do children with Down syndrome have: (a) between group differences in functional profiles when compared to children with mixed developmental disabilities (matched for MA- and CA); and (b) does either group demonstrate within group differences in functional performance profiles?
 - a. Currently there is a paucity of evidence to support a directional hypothesis for this research question. However, evidence suggests a Down syndrome-specific profile of relative strengths and challenges in functional performance for children with Down syndrome (e.g., Fidler et al., 2006; also see Daunhauer 2011 for a review).
2. What factors best predict functional performance for children with Down syndrome (IQ, EF, or overall language development)?
 - a. Hypothesis 2a. Based on Daunhauer, Fidler, & Will (2014) it is expected that EF will be a significant predictor of functional performance as reported by the parents or caregivers of children with Down syndrome.
3. Do predictors of functional performance (IQ, EF, or overall language development) for children with Down syndrome differ from those for children with mixed developmental disabilities matched for MA- and CA?
 - a. Hypothesis 3a. This study is among the first to conduct between-group comparisons of models predicting functional performance outcomes. Prior literature suggests there may be differences due to the specific behavioral phenotype in individuals with Down syndrome (see Daunhauer, 2011 for a review; Daunhauer, Fidler, & Will, 2014).

CHAPTER 3: METHODS

Participants

The study sample included 39 children between 5-10 years of age and their parents in two groups: (1) a group of children with a confirmed diagnosis of Down syndrome ($n = 22$); and (2) a group of children with mixed developmental disabilities ($n = 17$) who were participating in a larger study at Colorado State University. A diagnosis of Down syndrome was confirmed using parent report of genetic testing. Additional inclusion criteria for both groups included: MA fell in the equivalent 2:0 to 5:11 CA range used for the Behavior Rating Inventory of Executive Function-Preschool Version (BRIEF-P; Gioia, Espy, & Isquith, 2003), no history of traumatic brain injury, no medical or other genetic conditions beyond those associated with developmental disabilities, and an absence of the diagnosis of comorbid autism spectrum disorder as reported by the parents. The mixed developmental disabilities group was mental age-matched to the Down syndrome group on both chronological age and nonverbal mental age using the Leiter International Performance Scale-Revised (Leiter-R, Roid & Miller, 1997). See Table 1 for further participant characteristics. The mixed developmental disabilities group included children with a range of developmental disabilities including idiopathic intellectual disability, fragile X, and Williams syndrome.

Participants were recruited from the Rocky Mountain Down Syndrome Association, the Poudre School District in Fort Collins, CO; JFK Partners, a University Center of Excellence in Developmental Disabilities at the University of Colorado-Denver; and the Department of Human Genetics of the Emory School University School of Medicine in Decatur, GA. Participants were recruited through flyers and referrals at

these organizations as well as through physician and teacher referral, mailings to eligible participants, and information booths at organization events.

Measures

Demographics. Parents completed the Medical History and Intervention Information Questionnaire (MHIIQ) questionnaire regarding the child and family. Items about the child included date of birth, gender, race/ethnicity, diagnosis, health history, history of social and language development, as well as items about the child's service and educational history. Parents were also asked to report on their age, race/ethnicity, employment status, level of education, current marital status, and gross family income.

Mental Age and IQ. The Leiter International Performance Scale-Revised (Leiter-R, 1997) was used to determine mental age. The Leiter-R is a cognitive assessment that was administered to the child participants in a developmentally appropriate laboratory setting. The Brief-Intelligence Quotient (Brief-IQ) composite score was calculated to obtain an overall nonverbal mental age of the participants. Four subtests are composed of the Brief-IQ composite score and include: Figure Ground (FG), Form Completion (FC), Sequential Order (SO), and Repeated Patterns (RP). Figure ground involves finding and identifying specific items that are presented in a picture. Form completion involves interpreting incomplete visual information to answer questions about a picture. Sequential order involves correctly interpreting the sequence of objects in picture. The repeated patterns domain involves correctly placing items in a sequential pattern according to a visual display. The administration of the Leiter-R requires minimal verbal instruction from the examiner and requires no verbal response from the examinee, making the instrument suitable for populations of individuals who may have

language delays, such as those with Down syndrome. The Leiter-R was standardized using a national sample of typical developing children ($n = 1,719$). The authors also report high test-retest reliability across age groups ($r_s = 0.80-0.90$). Concurrent validity has been reported using the WISC-III Full Scale and Performance IQ as a standardized criterion measure ($r = 0.85$) (Roid & Miller, 1997).

Language. The Oral and Written Language Scales (OWLS, Carrow-Woolfolk, 2008) was used to assess language development in this study. The OWLS is designed for use with individuals between 3-21 years of age and has three scales: Listening Comprehension, Oral Expression, and Written Expression. For this study, participants were assessed using the Listening Comprehension and Oral Expression domains. The Listening Comprehension Scale is a measure of receptive language. The examiner reads a prompt out loud and the participant can answer nonverbally by pointing to a picture on a page. The Oral Expression Scale is a measure of spoken language. The examiner reads a prompt and the participant must answer a question, complete a question, or generate a sentence. The OWLS was standardized in a population of typically developing children ($n = 1,985$). High test-retest reliability was reported ranging from 0.76-0.81. High interrater reliability was also reported (0.95). The OWLS has been tested on content and construct validity ($r = 0.70$), as well as with a variety of other measures of language and cognitive ability to demonstrate high criterion-related validity ($r_s=0.70-0.90$; Carrow-Woolfolk, 2008). The raw scores are calculated by totaling how many items a child gets correct on each domain and are converted to a standardized score compared to the population of typically developing children. This study used the OWLS Oral Composite which is a standardized composite score combining the two

domains of listening comprehension and oral expression.

Executive function. The Behavior Rating Inventory of Executive Functioning-Preschool Version (BRIEF-P; Gioia, Espy, & Isquith, 2003) is a standardized parent and teacher-report assessment of executive function behaviors in a child's everyday life. The BRIEF-P is composed of two domains including behavioral regulation and one composite score, the Global Executive composite. The Global Executive Composite was administered to the parents in this study. This composite scores is based on sum of the following clinical scales: Inhibit, Emotional Control, Shift, Working Memory, and Plan/Organize. Higher scores on the Global Executive Composite indicate that a child is experiencing more problems with executive function.

Mental age scores for each participant from the Leiter-R were used to determine if participants from both groups would be included in the current study (i.e., in order for participants to be included their MA had to fall in the equivalent 2:0 to 5:11 CA range used to standardize the BRIEF-P). These procedures were critical in that overall developmental level was already accounted for when considering executive functioning with this assessment. Given that the child's chronological age may have been greater than their MA; raw scores of the Global Executive Composite were used in this analysis. The BRIEF-P has demonstrated adequate test-retest reliability with parents (0.78-0.90) and adequate convergent and discriminate validity with other behavior measure, such as the Child Behavior Checklist. The authors conducted a confirmatory factor analyses to validate the construct of executive functioning (BRIEF-P, Gioia, Espy, & Isquith, 2003).

Functional performance. The Pediatric Evaluation of Disability Inventory (PEDI) (Haley, et al., 1992) is a parent-report assessment designed to measure the functional performance and skill development of children with disabilities. The PEDI is distinguished from a developmental model in that it does not focus on children reaching motor, cognitive, and social developmental milestones. Instead, the PEDI measures a child's ability to engage in tasks of everyday life that place demands on these skills (Haley et al. 2010).

The PEDI authors developed this assessment based on a disablement framework that incorporated impairments, functional limitations, and social role performance. The framework incorporated a developmental and contextual framework leading to measurement constructs of discrete functional skills and performance of functional tasks in a child's natural environment, thus creating the functional skills and caregiver assistance scales (Haley et al., 1992). The conceptual basis of the PEDI framework was explored using the ICF framework (Ostenjo, Bjorkbaekmo, Carlberg, & Vollestad, 2006). Analysis revealed that the ICF could be a framework to assist with clarification of measurement constructs of the PEDI scales, but the ICF has limitations for examining functioning in early childhood (Ostenjo et al., 2006).

The PEDI is comprised of two sections including the Functional Skills Assessment and Caregiver Assistance. Each section includes three domains: Self-Care, Mobility, and Social Function. The functional skills assessment asks whether or not the child is capable or incapable of performing that skill. For example, in the self-care domain in regards to tooth brushing (e.g., holds toothbrush), in the mobility domain

in regards to car transfers (e.g. gets in and out of car with little assistance or instruction), and in the social function domain in regards to functional use of communication (e.g. names things) and play (e.g. interacts with peers). The caregiver assistance scale measures the level of assistance needed in these domains (i.e. 5=independent, 4=supervision, 3=minimal assistance, 2= moderate assistance, 1=maximal assistance, and 0= total assistance) and the level of modifications needed (N= no modifications, C= child-oriented modifications, R= specialized rehabilitation equipment, and E= extensive modifications). The PEDI is primarily used for children from the ages of 6 months- 7.5 years, but can be used in older children whose functional abilities fall below that of a 7.5-year-old child with no disabilities. Internal consistency, inter-interviewer reliability, and inter-interviewer reliability were reported in a normative sample and a clinical sample ($r_s=0.95-0.99$). In addition, studies have reported construct, concurrent, discriminant, and evaluative validity ($r_s=0.70s-0.90s$; Haley et al., 1992; Haley et al., 2010; Erkin et al., 2007; Stahult et al., 2011). In this study, a composite of the functional skills and caregiver assistance skills was obtained by adding the functional skills domain and caregiver assistance domain raw scores. The population was standardized using a sample of participants under 7.5 years of age. Given that this sample had participants greater than this age chronologically, the authors of the PEDI recommend use of the raw scores for analysis (Haley et al., 1992). A higher score on this composite indicates that the children performs at a higher skill level on the task and performs these tasks more independently.

The PEDI has been used in many studies with different childhood conditions, such as cerebral palsy, musculoskeletal disorders, spina bifida, mitochondrial diseases, Down syndrome and others to investigate the relationship between diagnosis and functional performance (Graveline, Young, & Hwang, 2000; Haley et al., 2010; Rogac, Meznaric, Zeviani, Sperl, & Neubauer, 2011; Tsai, Yang, Chan, Huang, & Wong, 2002). In addition to exploration of the relationship between diagnosis and functional performance, the PEDI has been utilized to compare outcomes among different diagnostic groups (Tsai et al., 2002). The PEDI is a valuable tool to examine functional performance in children with disabilities. By examining the PEDI in children with Down syndrome, it will be beneficial to compare outcomes of the PEDI to children with other disabilities as well as within-group differences to reveal outcomes that may be specific to the behavioral phenotype of Down syndrome, as well as provide information for targeted interventions. Overall, the PEDI is an appropriate tool to examine functional performance in children with Down syndrome and children with developmental disabilities.

Analysis plan. We examined the functional profile of children with Down syndrome and children with mixed developmental disabilities. We also performed repeated measures ANOVAs to examine the within-group functional skills and caregiver assistance domain differences. All variables were examined for normality within each group. Groups were compared with one-way ANOVAs to determine group differences on all demographic variables, mental age, and chronological age. Missing data was addressed by comparing the groups for significant differences to determine the effect the missing data had on group differences.

We performed separate multiple linear regression analysis to examine the nature of the relationships between IQ, executive function, and language and the outcome variable of functional performance on the PEDI in children with Down syndrome and children with mixed developmental disabilities. Multi-collinearity was addressed between all of the predictors in the regression models, and adjusted models were run to address any issues with multi-collinearity.

CHAPTER 4: RESULTS

Functional Profile of Functional Performance

Down syndrome group. A repeated measures ANOVA was performed to examine the within-subject differences between scores on the self-care, mobility, and social function functional skills domains of the PEDI for children with Down syndrome. There were statistically significant differences between scores on the domains, $F(2, 20) = 49.1, p < .001$. A post-hoc paired-samples t -test was conducted to determine the magnitude of the differences between domains. Individuals with Down syndrome had significantly higher scores on mobility functional skills than self-care functional skills, $t(21) = 6.57, p < .001$. Individuals with Down syndrome also had significantly higher scores on mobility functional skills than social function functional skills, $t(21) = 9.02, p < .001$. The difference between self-care and social function functional skills was not significant. These results indicate that as reported by the parents, children with Down syndrome had relative strength in mobility skills and relative challenges in self-care and social function.

A repeated measures ANOVA was performed to examine the within-subject differences between these same areas (self-care, mobility, and social function) for caregiver assistance domains of the PEDI for children with Down syndrome. There were statistically significant within-group differences between scores on the domains, $F(2, 20) = 21.42, p < .001$. A post-hoc paired-samples t test was performed to determine the magnitude of the differences between domains. Caregivers reported on average that their children required significantly higher levels of assistance on the self-care domain than the mobility domain, $t(21) = 4.55, p < .001$. Caregivers also reported significantly

higher levels of caregiver assistance on the social function domain, $t(21) = 6.72, p < .001$. There was no significant difference between the social function and self care caregiver assistance domains. These results indicate that as reported by the parents, children with Down syndrome receive more assistance with self-care and social function tasks than mobility tasks.

Mixed developmental disabilities group. A repeated measures ANOVA was performed to examine the within-subject differences between scores on the self-care, mobility, and social function functional skills domains of the PEDI for children with mixed developmental disabilities. There were statistically significant differences between scores on the domains, $F(2, 15) = 15.03, p < .001$. A post-hoc paired-samples t test was conducted to determine the magnitude of the differences between domains. As reported by the parents, children with mixed developmental disabilities had significantly higher scores on mobility functional skills than self care functional skills, $t(16) = 5.66, p < .001$. Children with mixed developmental disabilities also had significantly higher scores on mobility functional skills than social function functional skills, $t(16) = 4.12, p = .001$. The difference between self-care and social function functional skills was not significant. These results indicate that as reported by the parents, children with mixed developmental disabilities also had a relative strength in mobility skills and relative challenges in self-care and social function skills.

A repeated measures ANOVA was performed to examine the within-subject differences between these same areas (self-care, mobility, and social function) for caregiver assistance domains of the PEDI for children with mixed developmental disabilities. There were statistically significant within-group differences between scores

on the domains, $F(2, 15) = 5.51, p = .016$. A post-hoc paired-samples t test was performed to determine the magnitude of the differences between domains. Caregivers reported on average that their children required less assistance on the mobility domain than the self-care domain, $t(16) = 2.72, p = .02$. Caregivers also reported that individuals required less caregiver assistance on the mobility domain than the social function domain, $t(16) = 3.41, p < .001$. The social function caregiver assistance and self-care caregiver assistances scores were not significantly different. The results indicate that the individuals with mixed developmental disabilities, as reported by their caregivers, required less assistance with mobility tasks than self-care and social function tasks.

Overall, parents reported that children with Down syndrome and children with mixed developmental disabilities both had significantly higher scores on the mobility functional skills domain and required less caregiver assistance on this domain than the self-care and social function domains. A one-way ANOVA was performed to determine whether children with Down syndrome and children with mixed developmental disabilities had significantly different scores on the PEDI. All results were not significant. See Table 2 for complete results. These findings indicate that children with Down syndrome and children with mixed developmental disabilities were reported to have a similar functional profile.

Predictors of Functional Performance

Down syndrome group. A multiple linear regression analysis was performed to examine the extent to which IQ, language ability, and executive function predicting functional performance in school-aged children with Down syndrome. See Table 3 for

results. Findings from this analysis showed that the overall regression was statistically significant, $F(3, 18) = 4.86, p = .012$. This means the combination variables included in the overall prediction equation were significant predictors of parent-reported functional performance. The regression equation can be written as follows:

$$\text{Functional Performance} = -1.96 \text{ IQ} + .592 \text{ Language Ability} + -.836 \text{ Executive Function}$$

The regression indicates that a 1-point increase in IQ is associated with a 1.96 decrease in functional performance controlling for language ability and executive function. Similarly, a 1-point increase in executive function difficulties controlling for IQ and language ability is associated with a .836 decrease in functional performance, indicating that greater executive function difficulties were associated with lower functional performance. A 1-point increase in language ability controlling for executive function and IQ was associated with a .592 increase in functional performance. Together, the predictors in the equation accounted for 44.8% of the variance in functional performance, Adjusted $R^2 = .356$. According to Cohen (1988), this is a medium to large effect size. Findings also indicated that only executive function, $t = -3.19, p = .005$ was a significant predictor of functional performance. The standardized regression weight was, $\beta = -.56$ ($sr = -.60$).

Inspection of the zero-order correlation between IQ and language ability and inspection of the collinearity statistics suggested that the intercorrelation between these two predictor variables created a confound with multicollinearity. To address this problem with multicollinearity, these two variables were combined into a linear composite variable. A multiple regression analysis was performed with the linear composite variable. See Table 4 for the results. The results of the analysis were

statistically significant, $F(2, 19) = 5.89, p = .010$. Together the variables accounted for 31.8% of the variance in functional performance, Adjusted $R^2 = .318$. According to Cohen (1988) this is a medium effect size. Executive function was the only significant predictor of functional performance, $t = -3.23, p = .004$. The standardized regression weight was $-.583$ ($sr = -.60$).

Mixed developmental disabilities group. A multiple linear regression analysis was also performed to examine the extent to which IQ, language ability, and executive function predicted functional performance as reported by parents in the mixed developmental disabilities group. See Table 5 for the results. Findings from this analysis showed that the overall regression was statistically significant, $F(3, 8) = 8.59, p = .007$. This means that the variables included in the overall prediction equation were significant predictors of functional performance. The linear regression equation can be written as follows:

Functional Performance = 1.546 IQ + .372 Language Ability + .328 Executive Function

The regression equation indicates that a 1-point increase in IQ controlling for language ability and executive function was associated with a 1.55 increase in functional performance. A 1-point increase in language ability controlling for IQ and executive function was associated with a .372 increase in functional increase, and a 1-point increase in executive function difficulties controlling for IQ and language ability was associated with a .328 increase in functional performance, indicating that greater executive function difficulties were associated with higher functional performance. Together the predictors in the equation accounted for 76.3% of the variance in

functional performance, Adjusted $R^2 = .674$. According to Cohen (1988) this is a large effect. Findings also indicated that only IQ, $t=3.20$, $p=.013$, was a significant predictor of functional performance. The standardized regression weight was 1.043.

Inspection of the zero-order correlation between IQ and language ability and inspection of the collinearity statistics suggested that the intercorrelation between these two predictor variables created a confound with multicollinearity. To solve this problem with multicollinearity, these two variables were combined into a linear composite variable. A multiple regression analysis was performed with the linear composite variable. See Table 6 for the results. The results of the analysis were statistically significant, $F(2, 9) = 7.16$, $p = .014$. Together the variables accounted for 61.4% of the variance in functional performance, Adjusted $R^2 = .528$. According to Cohen (1988) this is a large effect size. The language ability and IQ composite was the only significant predictor of functional performance, $t = -3.51$, $p = .007$. The standardized regression weight was $-.728$ ($sr = -.76$).

Comparison of regression models. Comparison of fit of the models from the Down syndrome and mixed developmental disabilities group revealed no significant differences between the respected R^2 values, $Z = 1.41$, $p > .05$. A comparison of the structure of the model from the two groups was also conducted by applying the model derived from the group from the mixed developmental disability group to the data from the Down syndrome group. The predicted outcomes based on the Down syndrome model and the predicted outcomes based on the mixed developmental disabilities group were not statistically significant according to the Steiger's Z test ($Z = .72$, $p > .05$). In addition, a comparison of the regression weights for executive function and the

OWLSIQ linear composite for the two groups indicated that they were not significantly different from each other ($Z = .369$, $p > .05$, $z = .80$, $p > .05$).

Post-hoc analysis. Given that the regression models for the two groups did not significantly differ on fit, structure, outcomes, or predictors; an additional post-hoc regression analysis was conducted. See Table 7 for the results. This analysis included all participants combined from both the Down syndrome and mixed developmental disabilities groups to examine the extent to which the IQ and language ability composite and executive function predicted functional performance as reported by parents for all participants. The results of the analysis were statistically significant, $F(2, 31) = 6.54$, $p = .004$. Together the variables accounted for 30% of the variance in functional performance, Adjusted $R^2 = .251$. According to Cohen (1988) this is a small to medium effect size. Executive function was the only significant predictor of functional performance, $t = -2.721$, $p = .011$. The standardized regression weight was $-.45$ ($sr = -.51$).

Table 1. Participants Characteristics by Group

Participant Characteristics	Down Syndrome n=22	Other DD n=17	X ² or F; (p)
	<i>M or %(SD)</i>	<i>M or % (SD)</i>	
CA, Mo	82.5 (10.53)	81.0 (12.98)	.159 (.693)
MA, mo*	49.5(6.70)	50.4(11.68)	.094 (.760)
Leiter-R Brief IQ	61.8(8.16)	68.3(18.00)	2.10(.157)
BRIEF-P General Executive Composite	118.0(21.0)	136.7(15.22)	9.50 (.004)
OWLS Oral Composite	102.5 (14.94)	121.4(23.12)	8.46(.007)
Child Percent Male	54.5%	58.8%	.068(.796)
Child Percent White, Non- Hispanic	86.4%	76.5%	.315(.578)
Mother's age, yr	40.2(5.97)	41.0(7.30)	.144(.706)
Mother, bachelor's degree or more	77.2%	76.4%	.313(.579)
Mother Percent White, Non-Hispanic	90.9%	88.2%	.852 (.362)
Father's age, yr	42.0 (5.640)	42.38(6.51)	.484 (.491)
Father, bachelor's degree or more	86.3%	76.5%	.512(.479)
Father Percent White, Non-Hispanic	86.4%	88.2%	.238 (.629)
Percent Family Income >80,000	68.0%	70.6%	.033(.856)

* Derived from Leiter-R Brief IQ

Table 2. Pediatric Evaluation of Disability Inventory Raw* Score Item Averages by Domain

PEDI Scale	DS M (SD)	DD M (SD)
Functional Skills		
Self-Care	.782 (.141)	.745 (.112)
Mobility	.949 (.054)	.896 (.145)
Social Function	.760 (.104)	.677 (.163)
Caregiver Assistance		
Self-Care	.694 (.216)	.734 (.216)
Mobility	.908 (.152)	.862 (.172)
Social Function	.629 (.213)	.664 (.206)

*Raw scores were used due to the ages of the sample size being greater than the normed population and per the manual recommendations (Haley et al, 1992)

Table 3. Results of multiple regression analyses to assess the relationship between functional performance and language, IQ, and executive function in children with Down syndrome

Outcome Variable	Predictor Variable	<i>b</i>	<i>SE</i>	β	<i>p</i>	<i>R</i>
Functional performance	IQ	-2.283	1.47	-.597	.138	-.344
	Language	.799	.799	.382	.331	.229
	Executive Function	-.794	.266	-.535	.008	-.575

F (3, 18) = 4.89, *p* = .012, Adjusted R² = .357

Table 4. Results of adjusted multiple regression analyses to assess the relationship between functional performance and language and IQ composite, and executive function in children with Down syndrome

Outcome Variable	Predictor Variable	<i>b</i>	<i>SE</i>	<i>B</i>	<i>p</i>	<i>R</i>
Functional performance	IQ / Language Composite	-.226	.241	-.169	.360	-.210
	Executive Function	-.867	.268	-.583	.004	-.595

F(2,19)= 5.89, *p* = .010, Adjusted R² = .318

Table 5. Results of multiple regression analyses to assess the relationship between functional performance and language, IQ, and executive function in children with mixed developmental disabilities

Outcome Variable	Predictor Variable	<i>b</i>	<i>SE</i>	B	<i>p</i>	<i>R</i>
Functional performance	IQ	1.55	.484	1.043	.013	.749
	Language	-.301	.372	-.261	.443	-.275
	Executive Function	-.250	.328	-.136	.468	-.260

F(3,8)= 8.59, *p* = .007, Adjusted R² = .674

Table 6. Results of adjusted multiple regression analyses to assess the relationship between functional performance and language and IQ composite, and executive function in children with mixed developmental disabilities

Outcome Variable	Predictor Variable	<i>b</i>	<i>SE</i>	B	<i>p</i>	<i>R</i>
Functional performance	IQ / Language Composite	.492	.140	.728	.007	.726
	Executive Function	-.438	.381	-.239	.468	-.238

F(2,9)= 7.16, *p* = .014, Adjusted R² = .528

Table 7. Results of post-hoc multiple regression analyses to assess the relationship between functional performance and language and IQ composite, and executive function in combined participants including children with Down syndrome mixed developmental disabilities

Outcome Variable	Predictor Variable	<i>b</i>	<i>SE</i>	B	<i>p</i>	<i>R</i>
Functional performance	IQ / Language Composite	.240	.141	.258	.100	.291
	Executive Function	-.747	.220	-.517	.002	-.238

F(2,31)= 6.54, *p* = .004, Adjusted R² = .251

CHAPTER 5: DISCUSSION

Individuals with Down syndrome display a pattern of strengths and challenges in various domains of development. There is a paucity of research examining how the Down syndrome behavioral phenotype may relate to individuals' functional performance profile in everyday life. This study examined the functional performance profile of a group of children with Down syndrome and a comparison group of individuals with mixed developmental disabilities to better understand whether or not individuals with Down syndrome present with a unique functional performance profile. In addition, this study examined whether IQ, language ability, or executive function predicted functional performance in individuals with Down syndrome and individuals with mixed developmental disabilities.

Functional performance implications

Functional performance profiles. Consistent with previous findings in the areas of functional performance and adaptive behavior (Daunhauer, 2011; Fidler et al., 2006), individuals with Down syndrome displayed a within-individual profile of relative strengths and challenge in areas of functional performance. Individuals with Down syndrome showed relative strength in mobility and relative challenges in both social function and self-care. Although research on children with Down syndrome has shown gross motor delays in early childhood (de Campos et al., 2009), it appears that by school age they present with a relative strength in the performance of mobility tasks compared to self-care and social function tasks. An example of mobility functional skills includes “walks down full flight [of stairs] with no difficulty” (Haley et al., 1992, p.108), and an example of caregiver assistance for mobility tasks is the amount of assistance needed from

independent, with no need for assistance, to maximal the child needs assistance needed for the child to “climb and descend a full flight of stairs (12-15 steps)” (Haley et al., 1992, p.154). Parents reported that children with Down syndrome had more competent functional skills and required less caregiver assistance on these mobility tasks than self-care and social function tasks. Examples of performing self-care functional skills include “prepares toothbrush with toothpaste” and “ties shoelaces” (Haley et al., 1992, p.111). An example of caregiver assistance for self-care tasks involves how much assistance the child requires with grooming “brushing teeth, brushing or combing hair and caring for nose” (Haley et al., 1992, p.128). Examples of the performance of social function functional skills include “play activities or games that have rules” and “makes up elaborate pretend sequences from imagination” (Haley et al., 1992, p.116). An example of caregiver assistance for social function is how much assistance the child requires in the “ability to plan and carry out joint activities with a familiar peer” (Haley et al., 1992, p.162). Parents reported that their children had similar levels of functional skills in self-care and social function, and required similar levels of caregiver assistance.

Compared to the sample of individuals with developmental disabilities, there was no significant difference between groups in the overall functional performance levels. The sample of individuals with mixed developmental disabilities also displayed a similar functional profile of relative strength in mobility and relative challenge in social function and self-care. Therefore, the results in this study do not support that children with Down syndrome present with a unique functional profile.

As emphasized by Hodapp & Dykens (2004), each neurogenetic disorder does not differ from other neurogenetic disorders on every behavior. Furthermore, by choosing a comparison group of children with intellectual disabilities this study followed the current best recommendations for examining whether a profile is unique to a particularly etiology group such as children with Down syndrome (Dykens & Hodapp, 2007; Hodapp & Dykens, 2001; Seltzer et al., 2004). However, given the modest sample size and high variability across all variables in the comparison group, these results should be considered preliminary and subject to replication. Furthermore, the current findings on the functional performance profiles have implications for future intervention. Given that there is a relative strength in mobility for both groups; it may be beneficial to use this strength in interventions that target their areas of challenge. For example, perhaps interventionists can incorporate motor strengths such as locomotion within games to help children be successful while scaffolding social function skills such as taking turns or following rules with their peers. Targeting both strengths and challenges in early childhood may help to promote optimal development (Fidler & Nadel, 2007).

Predictors of functional performance. In the Down syndrome group, executive function was the only significant predictor of the functional performance composite score. Furthermore, intelligence was the only significant predictor of functional performance for the mixed developmental disabilities group. For individuals in the group with mixed developmental disabilities, IQ scores ($M = 68.3$, $SD = 18.00$) were more variable than in children with Down syndrome ($M = 61.8$, $SD = 8.16$). This may explain in part, why IQ might have been a larger predictor than executive function for this extremely small sample size of children with mixed developmental disabilities. In the

adjusted regression models, the language measure was combined with IQ due to their colinearity, suggesting that this particular language measure assessed an overall level of comprehension. In addition, when examined, the two group's regression models did not significantly differ from each other in fit, structure, outcomes, or predictors. Overall further replication of these models in larger samples is warranted. In a post-hoc linear multiple linear regression analysis examining the same variables with the combined groups, executive function was the only significant predictor of functional performance.

Therefore, when considering the regression results from Down syndrome group and the post-hoc analysis regression of the combined groups, the larger pattern of results from this study indicated that executive function is a significant predictor of functional performance. Specific deficits in executive function have been documented from early childhood in individuals with Down syndrome (Daunhauer et al., 2014a). This finding of executive function as a predictor of functional performance in children with Down syndrome is congruent with the broader, extant literature in this area. Executive function is recognized as an important indicator of overall school success in typical development (Duckworth & Carlson, 2014; Harms, Zayas, Meltzoff, & Carlson, 2014) , as well as predictive of academic achievement, such as math (Brock et al., 2009; Fuhs et al., 2014) and reading ability (Monette et al., 2014). For individuals with developmental disabilities executive function has been recognized as a significant predictor of academic outcomes in children with autism (Akbar, Loomis, & Paul, 2013). Executive function difficulties have also been associated with adaptive skill deficits for children with autism (Gilotty et al., 2002; Zingerevich & LaVesser, 2009). Moreover, in a project related to the current study, executive function was the only significant predictor

of school-based functional performance in elementary-aged students with Down syndrome (Daunhauer et al., 2014b). The current study results provides specific evidence for a relationship between executive function and functional performance in school-aged children with Down syndrome in their everyday tasks outside of an academic context. However, given the results comparing regression models with the comparison groups and the modest-sized samples, these findings are subject to replication.

Tarazini, Mahone, and Zabel (2007) proposed an interactional model of adaptive demands and executive dysfunction for children with neurological disorders. They proposed that interventions targeting executive function should promote independence in self-care. Therefore, executive function skills may be an important area of targeted intervention for parents, educators, and practitioners to consider for children with Down syndrome. Future research should examine whether or not this interactional model is dependent on the behavioral phenotype of neurogenetic disorders. In addition, it will be important to examine executive function and functional performance skills over time to investigate the emerging profiles in early childhood and their potential implications for later childhood and adolescence. Examining this functional profile in relationship to the development of executive function in toddlerhood may help to identify specific areas of target, such as working memory or self-care skills, before they become significant areas of specific challenge (Fidler & Nadel, 2007; Fidler et al., 2011).

Given the results on the functional performance profile, targeted interventions should focus on both self-care skills and social function in order to facilitate optimal functional performance outcomes in both individuals with Down syndrome and

individuals with mixed developmental disabilities. Additionally, findings from the predictors of functional performance indicate that executive function may be an important target for interventionists aiming to improve functional performance outcomes for children with Down syndrome. The results from this study indicate that the relationship between executive function and functional performance in children with mixed disabilities is less clear. The current study results also raise questions regarding whether the functional performance profile and use of executive function as a predictor of functional performance is unique to the Down syndrome phenotypic profile.

Limitations and Future Directions

There are many limitations to this study that must be taken under consideration when interpreting the generalizability of the results. First, this study had modest sample sizes, and in particular, the mixed developmental disabilities group was extremely small due to missing data and challenges with the mental-age matching of participants. In addition, the mixed developmental disabilities group had a heterogeneous population of individuals with a variety of diagnoses including other neurogenetic syndromes, such as Williams syndrome or Fragile X syndrome, as well as learning disabilities. The heterogeneous population of this group may have led to the findings that intelligence was the most important predictor of functional outcomes in this particular sample. Future researchers should examine the relationship between executive function, intelligence, and functional performance in larger samples of mixed developmental disabilities (Seltzer et al., 2004). It may be also helpful for future studies to use comparison groups of other neurogenetic syndromes, such as Fragile X syndrome or Williams syndrome, in order to compare and contrast the functional profiles associated

with phenotypes. A larger sample size will also allow for the examination of functional skills and caregiver assistance separately which can lead to the development of targeted interventions for the individuals as well as caregivers. In addition, larger sample sizes will allow for the inclusion of more variables, including environmental factors, aspects of motor development, and medical conditions.

Clearly larger sample sizes are critical to gaining the data needed to develop effective interventions.

Furthermore, the language ability measure used in this study was colinear with IQ, indicating that the two measures did not substantially contribute to functional performance outcomes in different ways. Given that the particular language measure used for this study assessed overall language ability, it may be helpful to focus on specific aspects of language development and the relationship to functional performance outcomes in future studies. For example, studies show that lexical and grammatical development is specifically delayed in children with Down syndrome (Polisenka & Kapalkova, 2014). Examining how this development influences functional performance, and in particular social function and communication, may help to reveal how targeted language may influence functional performance outcomes.

In addition, the current study's analyses did not take environmental factors into account, such as type of school classroom the children attend, such as special education classrooms in contrast to inclusion classrooms, family structure, types of intervention services received, or aspects of the home environment as predictors of functional performance. Environmental factors may play a role on an individual's capabilities within their everyday life. The present study examined aspects of language

and cognitive development, which does not necessarily take into account all possible factors that influence functional performance outcomes. Future studies should incorporate larger sample sizes to explore whether or not the relationship between executive function and functional performance exists in larger, geographically representative populations, and whether this relationship is consistent in other developmental disabilities or typically developing children.

Additionally, a composite score from the BRIEF-P (Gioia, Espy, & Isquith, 2003) ,a parent report measure of executive function in everyday life, was used as a predictor of functional performance in the current study. Future studies should also examine the domains of executive function separately to see whether or not different aspects of executive function, such as working memory or planning, are more strongly associated with functional performance outcomes. The use of laboratory-based tasks may also help to examine how strengths and challenges in specific aspects executive function may be related to functional performance outcomes. The use of both methodologies would provide the examination of task performance and executive function skills in everyday life. The combination of these measures may provide important implications for the development of interventions using specific tasks aimed at improving skills in everyday life or targeting everyday life skills.

Summary

In conclusion, parents of children with Down syndrome reported a functional performance profile of strength in mobility and challenges in self-care and social function. This functional performance profile was not unique to the children with Down syndrome in this study, as parents reported children with mixed developmental

disabilities had a similar functional profile. However, executive function was a significant predictor of functional performance in children with Down syndrome, whereas intelligence was the only significant predictor of functional performance in children with mixed developmental disabilities. Executive function may be an important area for targeted interventions aimed at improving functional performance outcomes for children with Down syndrome. Future research should examine these outcomes over time at various ages to examine the developmental trajectories of these outcomes to help form targeted early interventions for parents, teachers, and practitioners.

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