

EFFECT OF MOTOR SKILLS AND COGNITION ON
ACTIVITIES OF DAILY LIVING IN CHILDREN
WITH DOWN SYNDROME

by

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INTRODUCTION: DOWN SYNDROME

The Center for Disease Control and Prevention estimates that each year one of every 691 children born in the United States has Down syndrome (Parker, Mai, & Canfield, 2010). There is a need for research focused on this population because Down syndrome is the most common genetic cause of intellectual disability in the United States (Center for Disease Control and Prevention (2006). On a personal note, 38% of Americans know a individual with Down Syndrome (Hurley, 2011).

Down syndrome (DS) is a genetic mutation most commonly due to an extra copy of the 21st chromosome resulting in gene overexpression (Roizen & Patterson, 2003). Trisomy 21 accounts for 95% of DS cases, but there are three types of Down syndrome. Trisomy 21 is also known as “nondisjunction”, a cell division error resulting three copies of chromosome 21 rather than the typical two in the embryo (Skallerup, 2008). Today, 329 genes mapped to chromosome 21 impact brain structure, behavior, physical functioning, cognition, and speech (Roizen & Patterson, 2003).

Trisomy 21 effects development patterns and results in phenotypically distinct characteristics in DS. Mosaicism, another type of DS, occurs in approximately 1% of the DS population. Mosaicism occurs when the nondisjunction of chromosome 21 takes place in only one of the initial cell divisions after fertilization (Skallerup, 2008). In translocation, another type of DS, part of chromosome 21 breaks off and attaches to another chromosome during cell division. The extra part of chromosome 21 causes Down syndrome characteristics to occur in those with translocation.

Common phenotypic characteristics present in individuals with DS include: low muscle tone, small stature, upward slant of the eyes, and a single deep crease across the

center of the palm. However, each person is unique and may possess the characteristics to a different degree or the characteristic may not be present (Skallerup, 2008). Individuals with Down syndrome have an increased risk for medical conditions including congenital heart defects, respiratory and hearing problems, Alzheimer's disease, childhood leukemia, thyroid conditions, and delayed physical development such as inferior muscle strength (Whitt Glover, et al., 2006; Skallerup, 2008). Fortunately, many of these conditions are treatable and many individuals with Down syndrome live healthy lives. Since 1983, the life expectancy for individuals with Down syndrome has increased from 25 years in 1983 to 60 years today.

Motor Development

Commonly, children with DS develop motor skills (fine and gross) in the same sequence at typically developing (TD) children, but "at a slower rate" (Mahoney, Robinson, & Fewell, 2001). Fine motor skills require the coordination of small muscle movement, especially movement of the hands like handwriting or buttoning a shirt. Gross motor skills require the coordination of large muscle groups resulting in large movements such as walking and running. Gross motor skills also incorporate postural control and stability. Motor development milestones such as crawling and walking provide an external manifestation of internal bodily changes such as bone growth, muscle development and coordination.

Motor Development Delays in Children with Down Syndrome

Due to delayed development, motor skills are less efficient (Mahoney et al., 2001). According to Esposito, MacDonald, Hornyak, & Ulrich (2012) muscle hypotonia, poor postural control and balance are phenotypic characteristics of DS. In addition

common factors include lower aerobic capacities, lower peak heart rates and decreased muscular strength (Esposito et al., 2012). These phenotypic characteristics affect neuromuscular and movement-related functions, making daily tasks more physiologically demanding for the individual (Barr & Shields, 2012). These features contribute to severely delayed developmental milestones such as standing, sitting, and creeping and crawling in the DS population (Cobo-Lewis et al., 1996). However, there is no standard or typical delay seen in children with DS, and the appearance of major motor milestones is highly variable. For example the onset of walking in children with DS can be delayed anywhere from six to eighteen months.

Expression of Motor Development: Physical Activity

Due to the physiological constraints manifested in individuals with Down syndrome, maintaining an active lifestyle can be more difficult. These constraints include less physical ability due to delayed motor development and less opportunity to be physically active (Menear, 2007). Other impediments include musculoskeletal, cardiovascular, biological, social, or environmental factors affecting individuals with Down syndrome (Barr & Shields, 2011). Therefore, individuals with Down syndrome often do not reap the benefits of physical activity due to these constraints.

Cognitive Development

Cognitive Development in Children with Down Syndrome

Poor physical activity output is also an expression of the degree of cognitive impairment and affects outcomes of activities of daily living (Esposito et al., 2012). Children with DS exhibit a wide variety of intellectual capacity (Bun, Roy, & Elliot, 2007). Intellectual capacity is often measured on the standardized Intelligence Quotient

Test (IQ Test), where an average IQ score is 100. An IQ below a 70 is classified as an intellectual disability. The majority of individuals with DS cognitively operate with a mild to moderate intellectual disability, with IQs ranging from 35 – 70 (Hurley, 2011). A “unique pattern of ability/disability” is also present in DS compared to peers of matched mental age according to Chua, Weeks, & Elliott (1996). This unique ability/disability pattern may reflect atypical cerebral organization in individuals with DS (Chua, Weeks, & Elliott, 1996). Jarrod (2007) relative strengths in some aspects of visuospatial processing

Adaptive Skills

Adaptive skills or activities of daily living (ADL) are the skills needed to maintain a normal quality of life. ADLS provide an indication of the impact a particular disability has on real-world functioning (Mervis, Klein-Tasman, & Mastin, 2000).

Disability Status

Adaptive skills are of crucial importance since adaptive skills are a key indicator of disability status. The Americans with Disabilities Act defines a disability as “any physical or mental impairment substantially limiting one or more major life activities” (A Guide to Disability Rights Laws, 2009). The “physical and mental impairments” discussed in the disability act are externally manifested through adaptive skills. Together, these unique physical and cognitive impairments result in substantial limitations. ADL assessments “provide an indication of the impact a particular disorder has on real-world functioning” (Mervis, Klein-Tasman, & Mastin, 2000). ADL testing is applicable for a variety of disabilities ranging from attention deficit hyperactivity disorder, to autism, to

cerebral palsy and how these specific disabilities impact daily living. If motor skills are never properly learned, compensatory strategies develop. (Mahoney et al., 2001).

Early Intervention Programs

Many children with DS participate in early intervention (EI) program, like physical, occupational, and speech therapy that attempt to bridge the gap between the child and TD peers and to minimize maladaptive skills (Shonkoff & Meisels, 2000). Maladaptive skills occur when children compensate for the deficits using inefficient and unsuccessful strategies. These maladaptive skills are concerning for developing children because future skills of increased complexity require strong foundations to build upon. EI programs usually begin ‘early in life’ or ‘early in the expression of the condition’ and continue through age five according to Hadders & Blauw (2005). The first years of life are a critical time for a child’s development, because children undergo rapid and volatile changes developmentally, acquiring essential life skills that will form the basis for more complicated skills later in life. Basic physical, cognitive, language, social, and self-help skills are achieved during these early years of life. These foundational skills precede future progress, and because children with Down syndrome usually face developmental delays, early intervention is highly recommended (Skallerup, 2008). An advantage of beginning intervention early in life is children receive more exposure to the motor patterns and behaviors they may struggle with in the future. Increased exposure and major goals of EI programs aim to increase the “rate of acquiring motor skills” and counteract “secondary problems resulting from the child's use of compensatory strategies” (Mahoney et al., 2001). Quality educational programs, stimulating home

environments, worthy health-care, and positive friend and family support all contribute to individuals with Down syndrome ability to reach full potentials and live fulfilling lives.

Despite physical and cognitive delays, individuals with Down syndrome regularly attend school, hold jobs, participate in lifestyle decisions, and contribute to society in many ways (McGuire & Chicoine, 2010).

Purpose

While it is known that children with DS lag in motor skills, no quantitative study has analyzed degrees of disability corresponding to biological and mental ages. The purpose of this study was to evaluate adaptive skills in preschool children with Down syndrome by examining the contributions of cognition and motor skills. Results of this purpose aimed to provide an initial step in understanding if intervention, therapy, and instruction should target improving motor skills and dexterity or cognition and understanding. An additional purpose of this study was to examine the relationship between cognition, motor skills, and activities of daily living in pre-school aged children with Down syndrome. Pinpointing the degree of impairment in specific motor, cognitive and adaptive tasks are crucial for EI programs. By identifying developmental deficits in specific subdomains of motor and cognitive development, early intervention services can devise evidence-based intervention and most effectively bridge the developmental gap between children with DS and their TD peers. Research on early cognitive and motor developmental trajectories in DS is important because practitioners can utilize results to formulate time-sensitive interventions that may prevent or offset potential future negative adaptive skills.

METHODS

Participants

Approval from the institutional review board was attained prior to the study. All parents of participants signed a university approved consent form before participation. Afterward all children were verbally asked if they want to participate in the presence of a witness. Ten participants with Down syndrome (4 male, 6 female) ranging in age from 36 to 65 months, with a mean age of 55 months, were recruited from Kinderfrogs program at the Starpoint School. The Kinderfrogs program is an early intervention program designed to provide developmentally appropriate educational experiences for preschool children with Down syndrome. Diagnosis is made off of parent report from a physician.

Recruitment

All participants with DS were recruited from the students at Kinderfrogs; a flyer was sent home with the parents of every class and information sessions were held at parent-teacher meetings to recruit participants. No effort was made to differentiate DS participants by trisomy 21, mosaicism, or translocation.

Apparatus

Three tests were used to evaluate participants' present level of performance. The Vineland Adaptive Behavior Scales 2nd edition measures adaptive skills, the Leiter International Performance Scale – Revised measures cognitive development, and the Peabody Developmental Motor Scales, 2nd edition measures motor development. Each test is a discriminative measure that is norm-referenced with the US population. The scores of each test are comparable to the average performance of the normative sample,

providing information to assess a quantitative delay and necessary intervention services (Tieman, Palisano, & Sutlive, 2005).

Vineland Adaptive Behavior Scales 2nd edition

Vineland Adaptive Behavior Scales 2nd edition (Sparrow, Balla, & Cicchetti, 2005) measures adapted behavior from childhood through adulthood. The system has four domains – communication skills, daily living skills, socialization skills, and motor skills. A composite maladaptive behavior index was calculated from disability in each of these domains. Each domain has additional sub-constructs that pertain to the domain. For example, the communication domain is composed of receptive language, expressive language and written language. The child's teacher filled out teacher rating forms evaluating the child (scale of 0 to 2, with 2 representing skill mastery) on a variety of executive motor functions. Teachers were provided a more accurate depiction of adaptive levels and avoided potential bias associated with parent or caregiver evaluations. A sum adaptive score was calculated as well as domain and subdomain age equivalents. To assess quantitative delays, scores were compared to peers, though the normative data included in the Vineland II manual,. The reliability of the Vineland II is 0.88 to 0.92 across domains and age. Validity of this test is 0.96 for ages 3-6.

Leiter International Performance Scale – Revised

Leiter International Performance Scale – Revised is designed for ages two through eighty-five. The Leiter assesses cognitive ability by measuring two factors, fluid reasoning and fundamental visual reasoning, through completion of eight tests. Each of the eight subtests are measured non-verbally. The final score is a measure of intelligence (IQ). Children were asked to perform a variety of non-verbal tasks, directly from the

Leiter manual, specific to both subdomains. Testing occurred in a therapy room at Kinderfrogs. After administering the test, composite and subdomain scores were calculated for each participant and compared to norm-referenced data included in the Leiter manual. The Leiter has demonstrated good concurrent validity with other measures of intelligence (the Wechsler scales) (Tsatsanis, Dartnall, Cicchetti, Sparrow, Klin, Volkmar, 2003). The Leiter is a non-verbal test of intelligence that has been found to be valid and reliable in youth with Down syndrome (Glenn & Cunningham, 2005).

Peabody Developmental Motor Scales, Second Edition

Peabody Developmental Motor Scales, Second Edition (Folio & Fewell, 2000) identifies fine and gross motor skills and delays in children from birth to age five. The Peabody (PMDS-2) contains subtests such as reflexes or visual-motor integration that examine motor skills in depth. Following the instructions in the Peabody manual, the administrator instructed the child to perform certain tasks; the test consisted of 127 gross motor tasks and 122 fine motor tasks. The execution of task is graded on a 0–2 scale, zero indicating the skill is not present and two indicating the skill is mastered. Testing was performed in the Starpoint gymnasium. After administering the test, a composite quotient and individual gross and fine motor quotients were calculated as well as age equivalents in the five subdomains. Normative data based off the population is included in the Peabody. Tieman et al. (2005) found that “the PDMS-2 has excellent reliability and validity as a discriminative measure.” The reliability of the total test varies by age, yet all are above 0.89.

Design And Analysis

All statistics were performed using Statistics Package for the Social Sciences (SPSS) version 20; statistical significance was set at $p < 0.05$ (SPSS Inc., Chicago, IL). Descriptive statistics were calculated to describe the sample as well as performance on each assessment. Means and standard deviations for chronological ages are found in Table 1. The process was repeated for each measure. Level of performance for each measure was expressed as a function of months. Performance on each test is reported in Table 1. Pearson product correlations were used to systematically examine the relationships between adaptive skills and motor abilities and between adaptive skills and intellectual abilities. Preliminary analysis included an independent sample t-test to determine if there were any gender differences.

To identify adaptive strengths and weaknesses frequency distributions were used to identify and quantify participants meeting specific criteria for adaptive skill proficiency. Bland-Altman plots were constructed to visually demonstrate participant deficits relative to their chronological ages. Age equivalents for each participant were calculated from performance on each of the three tests. Bland-Altman plots were also used to plot delays from mean group age. This was done to visually indicate which domains and subdomains possessed the greatest developmental delays.

RESULTS

Motor Development

The PMDS-2 measured five subdomains of motor development: stationary skills, locomotion, object manipulation, grasping and visual motor skills. Mean age equivalents for the group were calculated and compared to the group's mean age of 55 months. In

each of the five domains, the mean participant performance on the PMDS-2 scored below the group's mean age, indicating motor delays in each of these categories. Performance at 55 months would be considered to have no motor delays. Age delays range from 20 months in object manipulation to a 29 month age delay in grasping. The other three subdomains fell between this range: locomotor (23 month delay), stationary skills (25 month delay), and visual-motor integration (24 month delay). Please refer to figure 1 and figure two.

Table 1: Descriptive statistics of select tests and subtests

Variable	Performance (months)
	M \pm SD
Age (months)	55.0 \pm 10.4
Vineland	
Communication skills	44.0 \pm 7.5
Daily living skills	46.2 \pm 8.1
Social skills	44.5 \pm 7.7
Motor skills	40.3 \pm 4.2
Receptive language	41.3 \pm 6.4
Expressive language	39.9 \pm 8.0
Written language	52.6 \pm 12.3
Coping skills	47.8 \pm 12.7
Gross motor skills	36.3 \pm 0.9
Fine motor skills	44.3 \pm 8.0
Leiter-R	
Brief IQ	43.4 \pm 11.0
Fluid reasoning	44.4 \pm 10.6
Fundamental visual	42.3 \pm 15.2
Peabody	
Stationary skills	31.2 \pm 11.9
Locomotion skills	33.1 \pm 11.9
Object manipulation skills	35.1 \pm 10.7
Grasping skills	26.4 \pm 13.9
Visual motor skills	32.1 \pm 11.3

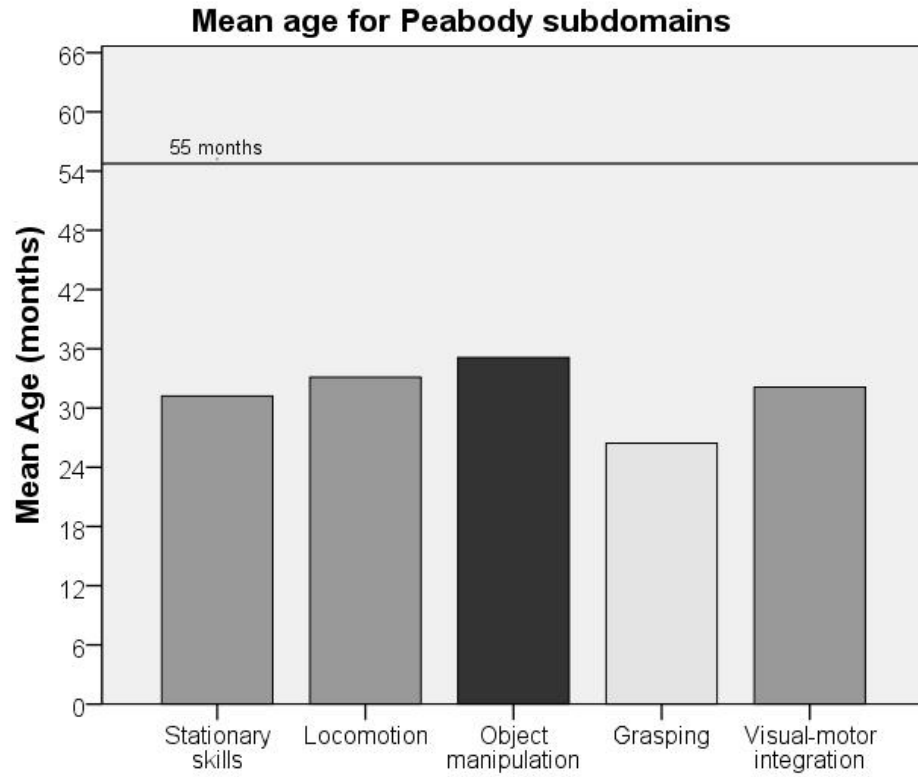
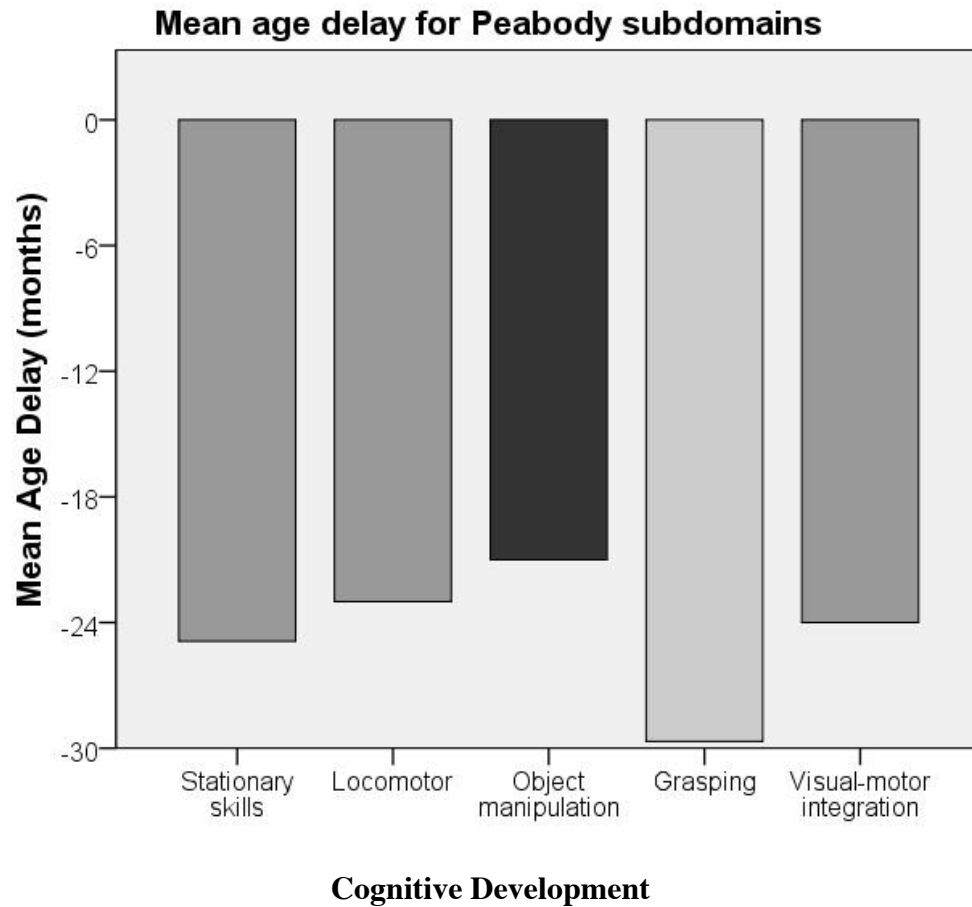
Figure 1: Mean age (months) for Peabody subdomains

Figure 2: Mean age (months) delay for Peabody subdomains



The Leiter-R test measured cognition total IQ, brief IQ and two subdomains of total IQ, fluid reasoning and fundamental visual reasoning. Figure 3 displays the total IQ delay, translated into months, for each individual participant. Information is presented as individual data for cognition because the Leiter tests only measures two subdomains of cognition while PMDS-2 and the Vineland both measured several domains. Performance of each participant on the two subdomains is displayed on figure 4. On average the sample possessed a mental age 13 months delayed from typically developing peers. A further break down the two subdomains of the Leiter test and the age equivalents were calculated as well. For the most part fundamental-visual intelligence are more delayed that the fluid reasoning component.

Figure 3: Mental age delay (months) for each participant

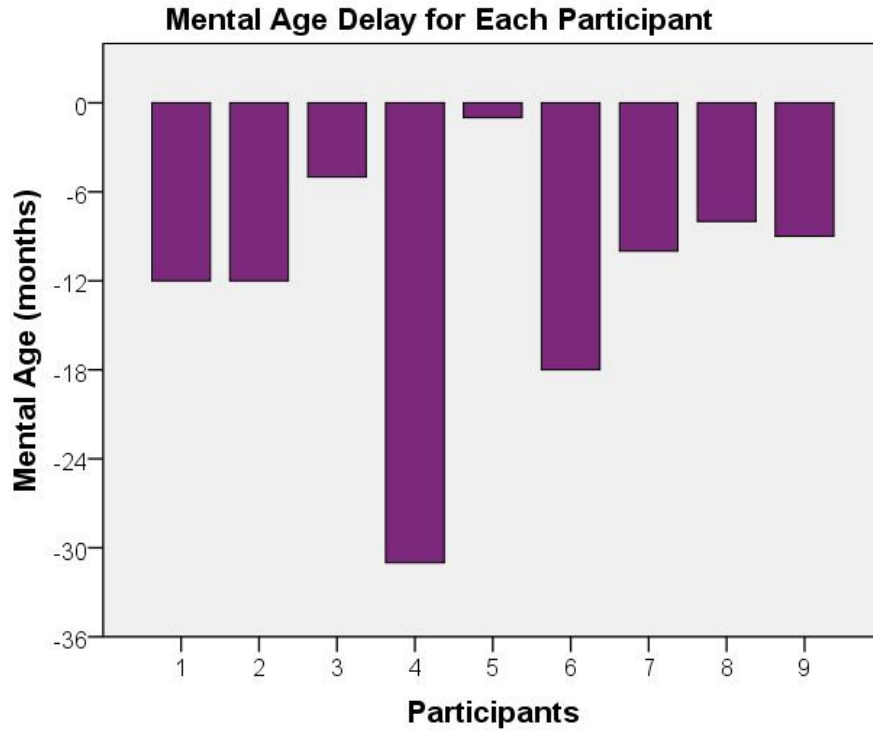
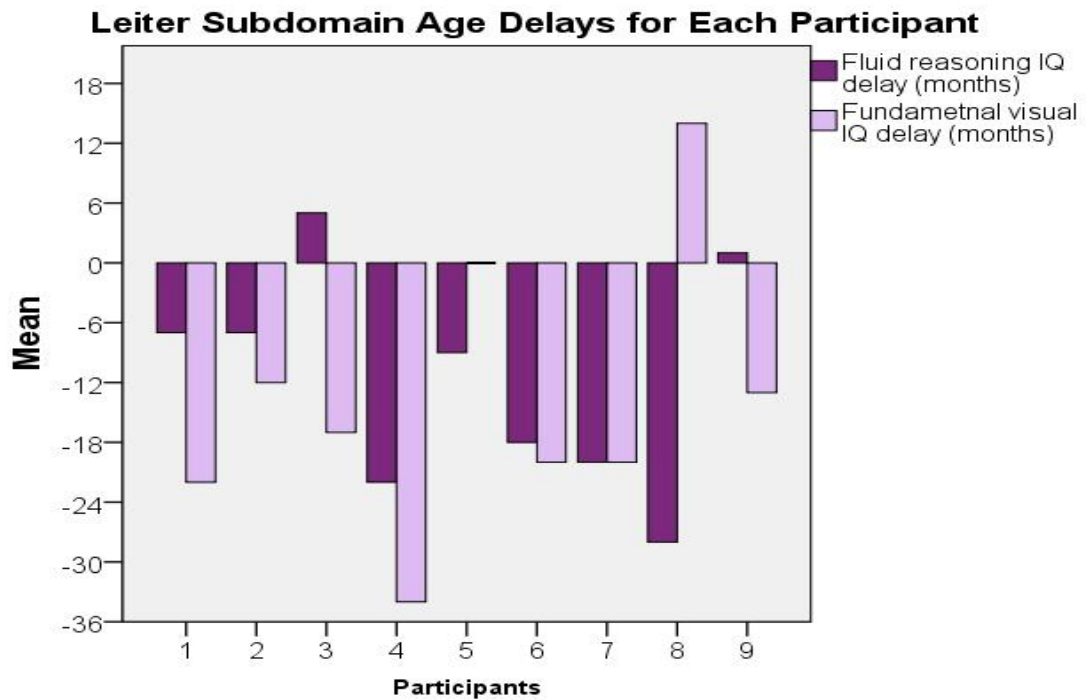


Figure 4: Leiter subdomain age delays (months) for each participant



Adaptive Skills

The Vineland measured adaptive skills in four major domains: communication, socialization, adaptive skills and motor skills. Each domain was composed of several subdomains. For example, the communication domain consisted of the subdomains: expressive language, receptive language, and written language. The other three domains had similar subdomain breakdowns. No general patterns emerged from Vineland testing demonstrated in figure 5. Participants were variable in the subdomains, demonstrating no obvious patterns. The relative strength of the written language sub-domain compared the other subdomains (receptive and expressive language) of communication is interesting to note. Figure 6 more clearly displays the large differences in adaptive capacities. The participants exhibited strength in written language, academic environment, community and coping skills. The participants exhibited adaptive weaknesses in the areas of expressive language, personal care, play and leisure and gross motor adaptations.

Figure 5: Mean age (months) for Vineland subdomains

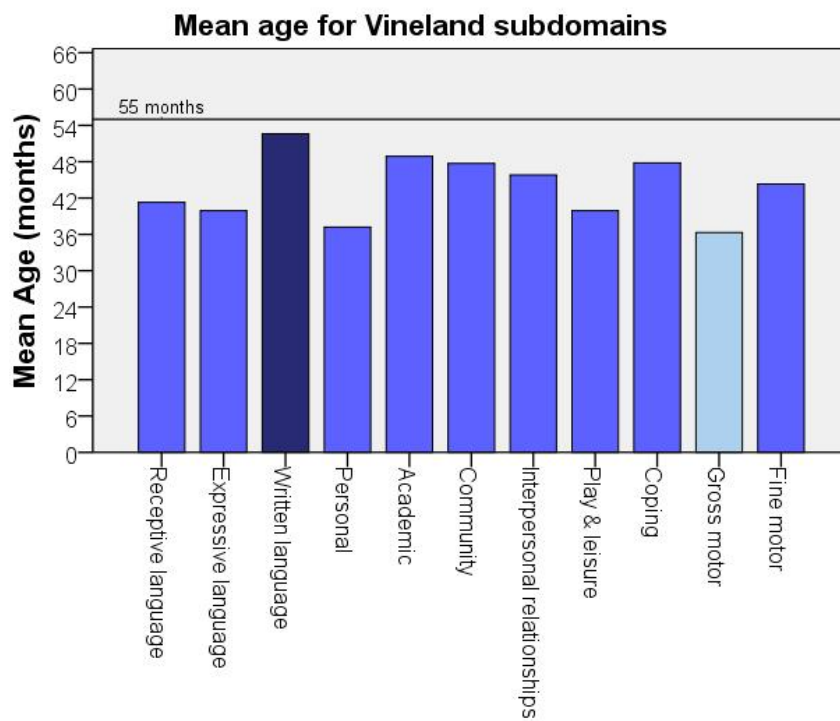


Figure 6: Mean age delay (months) for Vineland subdomains

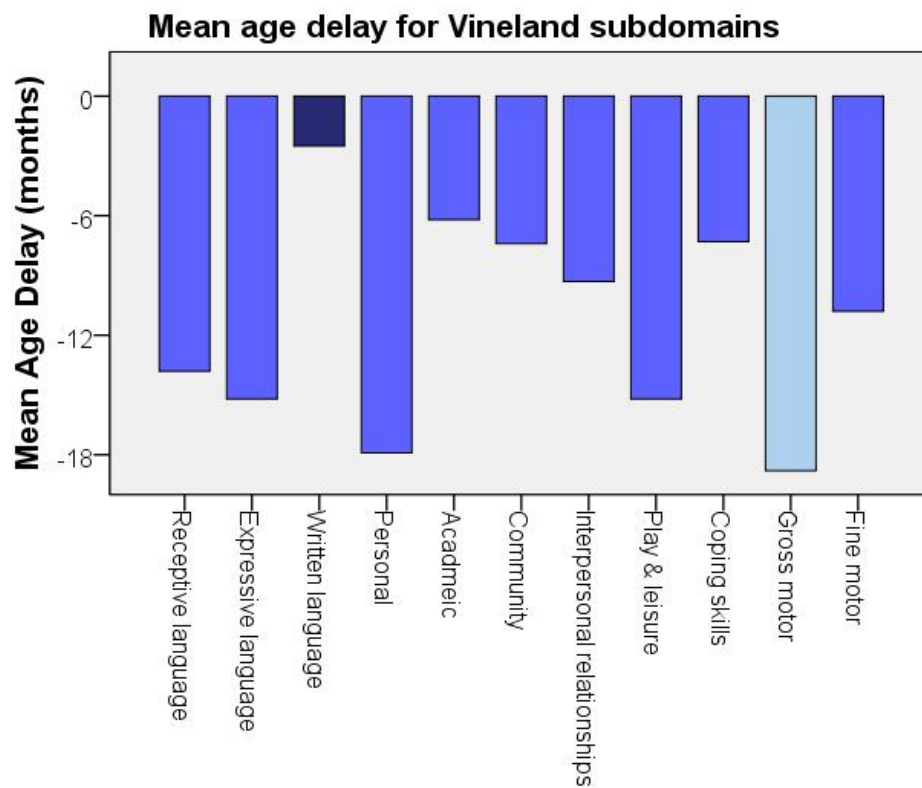


Figure 7: Mean age (months) for Vineland, Leiter, and Peabody subdomains

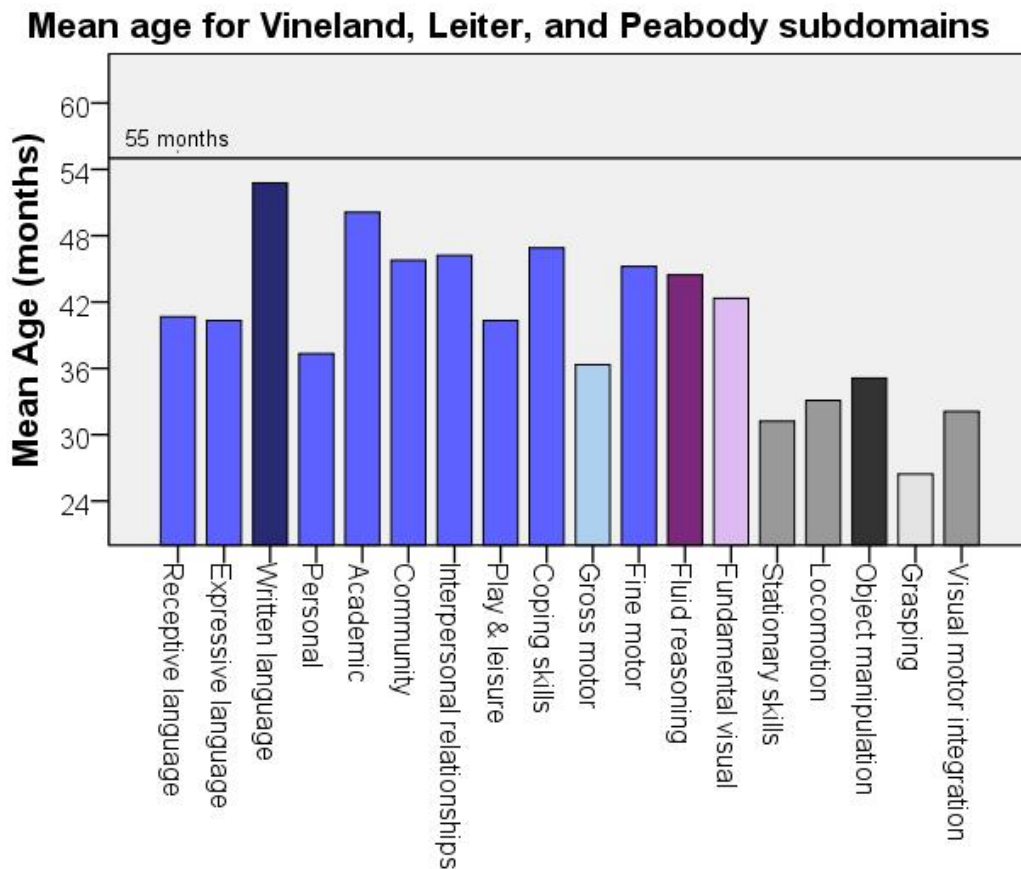


Figure 8: Adaptive communication skills

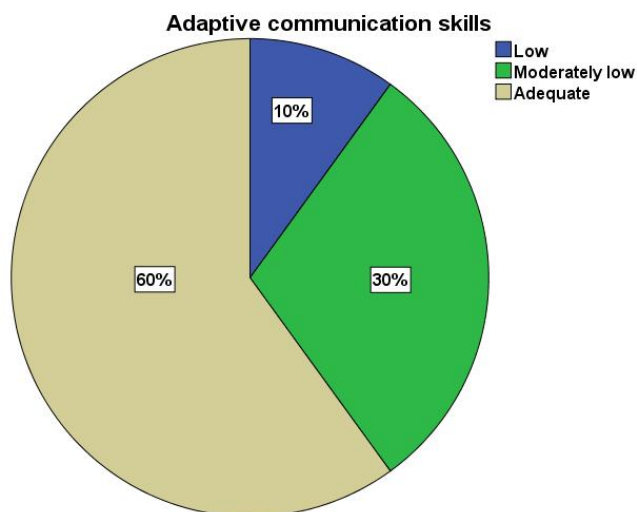


Figure 9: Adaptive socialization skills

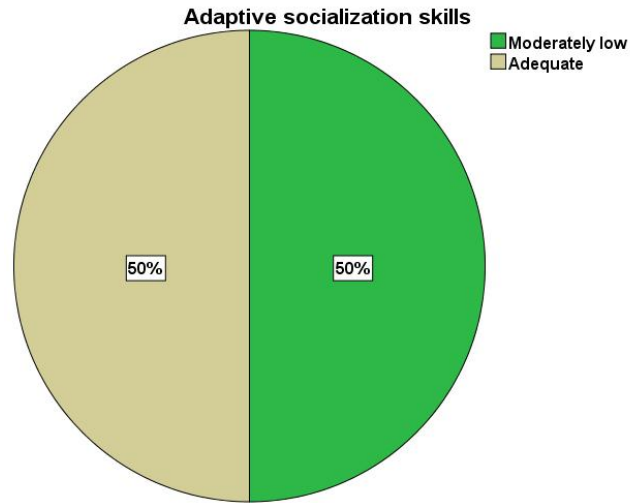


Figure 10: Adaptive motor skills

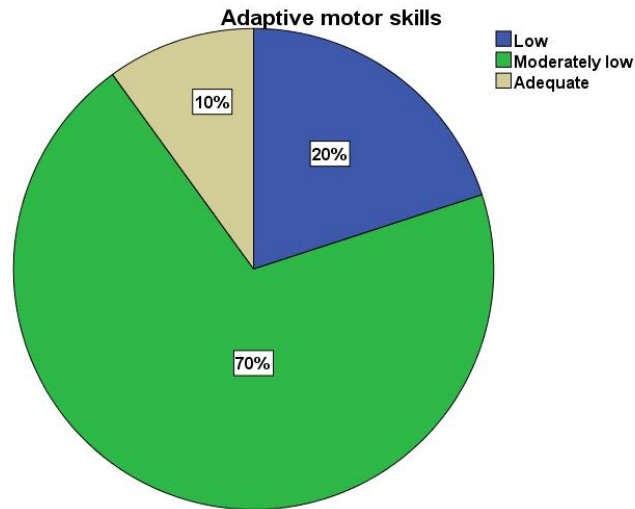
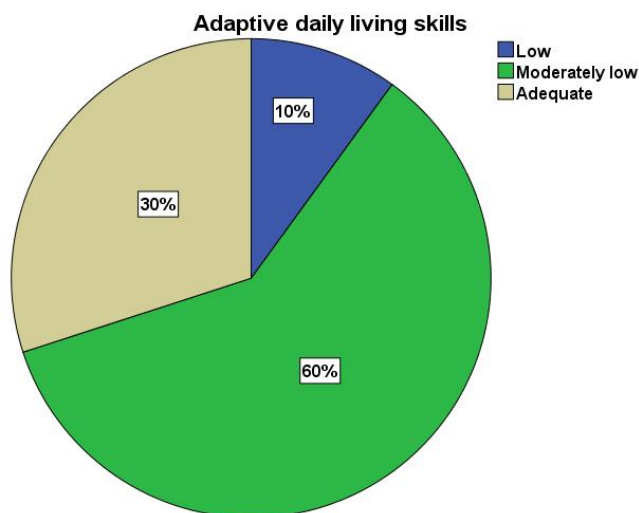


Figure 11: Adaptive daily living skills



DISCUSSION

The purpose of this study was to evaluate adaptive skills in preschool children with Down syndrome by examining the contributions of cognition and motor skills. Results of this purpose aimed to provide an initial step in understanding if intervention, therapy, and instruction should target improving motor skills and dexterity or cognition and understanding. An additional purpose of this study was to examine the relationship between cognition, motor skills, and activities of daily living in pre-school aged children with Down syndrome.

Particular patterns of strengths and weaknesses emerged from the data summarized in figure 7. Figure 7 displays the domains and subdomains of the PDMS-2, Leiter, and Vineland on the same graph. Overall, motor skills, displayed in grey, consistently display greater performance deficits from typically developing peers, compared to cognitive and adaptive performance deficits.

The Leiter results in purple on figure 7, display the relative performance of cognitive subdomains in comparison to the other tests run. The results of the Leiter indicated that fluid reasoning was less delayed than fundamental visual reasoning by 3 to 4 months. The age equivalents for the two cognitive subdomains only differ slightly demonstrating consistency in cognitive performance in this particular sample. Compared to motor development, cognitive development delay displays a smaller gap between typically developing peers with mean delays only being 9 to 13 months. Therefore, this sample of children with DS performed at a higher age than motor performance.

The PGMD-2 results in blue on figure 7, display the relative performance of motor subdomains in comparison to the other tests run. Consistently, motor delays were greater than both cognitive delays and all adaptive subdomains. Mean age delays were roughly an additional year (or 12 months) behind cognitive delays. The age equivalents for the five motor subdomains differ from a twenty-month deficit in object manipulation to a twenty-nine month deficit in grasping. This range demonstrates a lack of consistency in motor performance in this particular sample.

Composite adaptive scores were calculated for each domain of the Vineland, assigning each participant a numeric value from 1 to 5. Five was the highest-level successful adaptation, however participants in this study fell into levels 1 to 3 of the adaptive scale. The distribution of these three adaptive levels in the four adaptive domains can be seen in figures 8, 9, 10, and 11. The subjects exhibited strengths in adaptive communication domain and the adaptive socialization skills demonstrated by figures 8 and 9. The subjects exhibited strengths in the adaptive motor skills and adaptive daily living skills as seen by figure 10 and 11.

After running correlational tests, some significant positive correlations were found between motor development and adaptive skills in the following subdomains: stationary skills and communication skills (.731), visual motor skills and communication skills (0.738), locomotion and communication skills (.763), and object manipulation and communication skills (.792). Additionally, a significant relationship between cognitive development and adaptive skills in the subdomains of fundamental visual and adaptive motor skills (.733) was found. These positive correlations indicate that as performance in these adaptive skills increases, so does performance in the motor or cognitive skills. This strong relationship may be due to verbal instruction necessary to administer the PGMD-2 test. When instructing a child to perform a motor skill, visual demonstrations as well as verbal communication were provided. As communication skills improved, so did performance in the PGMD-2 subdomains. This may be due to receptive language deficits in the DS population. Literature on receptive language and children with DS is lacking. A further study evaluating the PGMD-2 test in specialized populations would be helpful in exploring these strong correlational values between PGMD-2 subdomains and adaptive communication skills.

Due to the novelty of this study, much of the results are not comparative to that completed by other in the field. The cognitive results were relatively on par with the average IQ scores of the DS population (Hurley, 2011). Much more exploration needs to be completed regarding developmental domains in children with DS, so early intervention services can most accurately target deficient skills and create appropriate interventions. These developmental deficits in specific subdomains of motor and cognitive development, provide a great tool for early intervention specialists. For

example, occupational therapists, therapists handling mostly fine motor rehabilitation, would be able to evaluate these results and recognize the relative strength of object manipulation skills (deficit of 20 months) and the relative weakness of grasping skills (deficit of 29 months). By understanding specific strengths and weaknesses of each intervention service, intervention therapists can devise evidence-based intervention and most effectively bridge the developmental gap between children with DS and their TD peers. Research on early cognitive and motor developmental trajectories in DS is important because practitioners can utilize results to formulate time-sensitive interventions that may prevent or offset potential future negative adaptive skills.

Practical Implications

Eligibility criteria for the Individuals with Disabilities Act (IDEA) require particular differences between chronological age and performance level. For example, a child who was 12 months old would be eligible for physical therapy services if his or her gross motor equivalent age was nine months or less. Therefore, this quantitative data clearly indicates that all of the participants qualify for disability services. Therapists often work with individuals with disabilities to mend this quantifiable gap between the child and their typically developing peers. Early intervention programs can use this data to devise evidence-based interventions according to capacities and deficiencies of children with DS. This particular sample displayed the greatest deviation from typically developing peers in realms of motor development. Therefore, specific intervention therapists that address motor development, specifically physical and occupational therapists, would be able to create evidence-based intervention programs to treat the most significant delays.

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ABSTRACT

Current literature lacks information about the relationship between specific domains of cognition and motor skills and adaptive skills in the Down syndrome population. Adaptive skills are of crucial importance since they are a key indicator of disability status. Adaptive skills, or activities of daily living, are the skills needed to maintain a normal quality of life. Therefore, it is necessary to identify which domains heavily impact adaptive skills so health services can promote these areas in early intervention programs. The purpose of this study was to evaluate adaptive skills (self-care activities) in children with Down syndrome by examining the contributions of cognition, fine motor skills, and gross motor skills. A cross-sectional approach and age-appropriate standardized tests were used to measure cognitive awareness, motor capacities, and daily living tasks in children with Down syndrome (N=9). Results of the sample indicated that children with Down syndrome developmentally lag from typically developing peers across all motor skills by 20 to 20 months. Cognitive testing indicated that children with Down syndrome developmentally lag from typically developing peers in cognitive domains only by 9 to 13 months. Adaptive strengths were evident in the communication and socialization adaptive domains and relative adaptive weaknesses were evident in the daily living skills and motor skills domains.