SELF-DETERMINATION IN ADOLESCENTS AND ADULTS WITH FRAGILE X SYNDROME: THE RELATIONSHIP BETWEEN SELF-REPORT, PARENT PERCEPTIONS, AND INDIVIDUAL CHARACTERISTICS

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ABSTRACT

Adrienne N. Villagomez: Self-determination in adolescents and adults with fragile X syndrome: The relationship between self-report, parent perceptions, and individual characteristics. (Under the direction of Barbara H. Wasik and Anne C. Wheeler)

Self-determination is a characteristic that encompasses an individual's abilities and attitudes to act as one's own agent (Wehmeyer, 1992). Individuals with increased self-determination may act according to their beliefs and preferences more than individuals who are less self-determined, which in turn may lead to increased decision-making opportunities. Encouraging self-determination is a key strategy to reduce disparities faced by individuals with intellectual and developmental disabilities (I/DD; Shogren, Wehmeyer, Reese, & O'Hara, 2006). Although fragile X syndrome (FXS) is the most common known genetic cause of intellectual disability and autism spectrum disorder, no studies have examined factors associated with self-determination in this population.

Eighty-six individuals with FXS (56 males and 30 females) between the ages of 12- and 40-years old and their parents completed questionnaires about self-determination. Individuals with FXS completed the Arc's Self-Determination Scale (Adolescent and Adult versions, Wehmeyer, 1995); their parents completed the Self-Determination Questionnaire (Carter et al., 2013), which covered a range of skills and experiences and parental perceptions of importance. Data on parental perceptions of barriers and strategies to encourage self-determination were examined qualitatively. Cognitive ability (i.e., FSIQ), functional skills, autism symptoms, and anxiety data were also collected on individuals with FXS.

Age, adaptive behavior, and social avoidance predicted scores on the autonomy domain of the self-report measure and gender predicted scores on the remaining three domains. Parent-reported self-determined behaviors were predicted by adaptive behavior. Most parents rated all self-determined behaviors as "very important" for their child. The strength of the relationship between self- and parent-report was significantly influenced by all child variables (e.g., ASD, FSIQ). Many parents identified emotional support, behavioral support, and providing opportunities as important areas of support for their child's development of self-determination. In comparison, parents reported anxiety, self-efficacy, communication, and cognitive functioning as the most significant barriers to their child's development of self-determination.

Findings from the present study highlight the importance of considering functional skills in individuals with FXS in predicting and ultimately promoting the development of self-determination. These findings also underscore the need for parent-report as well as information on the role of family in the development of self-determination.

This dissertation is dedicated to my grandfather, who made my education his priority.

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LIST OF ABBREVIATIONS

ASD	Autism spectrum disorder
DD	Developmental disability
FMRP	Fragile X mental retardation protein
FSIQ	Full scale intelligence quotient
FXS	Fragile X syndrome
ID	Intellectual disability
I/DD	Intellectual/developmental disability
IQ	Intelligence quotient
NVIQ	Nonverbal intelligence quotient
SD	Self-determination
SDPQ	Self-determination Parent Questionnaire
SDS	Arc's Self-determination Scale
VIQ	Verbal intelligence quotient

CHAPTER 1: INTRODUCTION

Individuals with intellectual disabilities (ID) and developmental disabilities (DD) experience significant disparities in health care access and quality when compared with individuals without these disabilities (Durvalsula & Beange, 2001; Fisher, 2004; Havercamp, Scandline, & Rother, 2004; Marks & Heller, 2003). Likewise, they also experience deficits in health autonomy (i.e., the ability to exercise autonomy in relation to healthcare in the areas of maintenance, understanding, and communication) (Wullink, Widdershoven, van Schrojenstein Lantman-de Valk, Metsemakers, & Dinant, 2009). As a function of their combined intellectual and developmental disabilities (I/DD), these individuals frequently experience significant difficulty maintaining their health and understanding the effects of their behavior on their health (Havercamp et al., 2004; Horwitz, Kerker, Owens, & Zigler, 2000; Ouellette-Kuntz, 2005). They also experience patient/provider communication challenges (Aaron & Chesley, 2003; Smedley, Stith, & Nelson, 2002). Healthcare disparities for individuals with I/DD may be improved by a variety of interventions focused on improving health autonomy. For instance, because individuals with I/DD are less frequently involved in the decision-making process about their healthcare and healthcare needs, interventions including education and supports to increase choice in healthcare decisions are necessary to address the health disparities of individuals with I/DD and increase health autonomy (Shogren, Wehmeyer, Reese, & O'Hara, 2006).

Engaging individuals with I/DD in their healthcare requires careful consideration of their skills and ability, as well as their attitudes and beliefs. Decisional capacity and self-determination are two factors which may not only inform health autonomy, but may also potentially improve

health autonomy and reduce healthcare disparities faced by individuals with I/DD. Decisional capacity encompasses the extent to which an individual is able to engage in a decision making process by understanding the consequences and making a decision free from coercion. Applebaum, Grisso, Frank, O'Donnell, & Kupfer, (1999) proposed a taxonomy for evaluating consent capacity for individuals with disabilities that includes four components: *understanding*, *appreciation, reasoning*, and *expressing a choice*. Understanding encompasses an individual's ability to comprehend factual information related to the nature of the health care research or procedure. Within a research context, the component includes understanding the purpose and nature of the study, as well as its logistics, benefits and risks, and the ability to withdrawal. This aspect of decisional capacity may be linked to previous experiences with medical procedures and decision-making experience. The second component, appreciation of the situation, involves recognizing the effects of research participation specific to the individual. Appreciation that the purpose of research recruitment is not for personal benefit or individualized care is an important concept related to understanding the personal implications the research study has on an individual's own circumstance. The third component, reasoning, encompasses the process of comparing alternatives with regard to consequences. Reasoning includes a preliminary choice, consequential reasoning for a given choice, comparative reasoning, consistency, and the ability to weigh risks and benefits. Finally, the ability to express a choice voluntarily and free from coercion is necessary, but not sufficient, to conclude decisional capacity in individuals with I/DD.

Including individuals with I/DD in the decision-making process in healthcare decisions involves consideration of several factors. First, each element of decisional capacity may be evidenced to varying degrees in a given individual. While some individuals may need increasing

levels of support to engage in the decision-making process, determining the degree to which they can engage in the decision-making process can increase the opportunity for health autonomy and improved healthcare. Individuals with ID in the mild to moderate range have demonstrated aspects of decisional capacity and are able to provide consent to standard health care (Cea & Fisher, 2003). Within the context of more complex healthcare decisions, support and educational approaches can be used to allow individuals with ID to be increasingly engaged and take an active role in health care decisions. While decisional capacity can increase an individual's health autonomy by determining an individual's ability to engage in the decision-making process, decisional capacity does not encompass other skill sets and abilities; it also excludes attitudes and beliefs that are important to health autonomy.

The concept of self-determination is one avenue to understand the attitudes, beliefs, and behaviors, which support decisional capacity and health autonomy. Self-determination encompasses abilities and attitudes that can contribute to decisional capacity and health autonomy for individuals with I/DD. For instance, self-determination includes the skills and attitudes necessary for an individual to advocate for oneself and voice his or her opinion. Among other strategies as a way to address health disparities, promoting self-determination in the I/DD population is key (Shogren, Wehmeyer, Reese, & O'Hara, 2006). Self-determination may facilitate an individual's opportunity to be included in health-related decision-making. Individuals with increased self-determination may act according to their beliefs and preferences more than individuals who are less self-determined which may lead to increased decision-making experiences and opportunities and, as a result, enhanced attitudes and beliefs about their ability to make decisions (i.e., self-determination).

The issue of health autonomy for individuals with I/DD is increasingly important. Health autonomy is particularly important for individuals with FXS as significant advancements in understanding the molecular basis of FXS have led to an increase in clinical trials to target symptoms of FXS. Given the concerns of health autonomy in individuals with I/DD, particularly for those with FXS, this study addressed the gap in the literature regarding self-determination of individuals with FXS by exploring parent- and self-report of self-determination using a sample of parents and their adolescent and adult children with FXS.

Outcomes from the current study were intended to inform future efforts to engage individuals with one major category of I/DD, namely those with fragile X syndrome (FXS), in healthcare decisions by illustrating the range of self-determination currently reported by individuals with FXS and parent perceptions of their children's self-determination. With increased understanding of the relationship between self-determination and individual characteristics (e.g., presence of autism spectrum disorder), gaps in healthcare access and quality can be improved. Further understanding of self-determination in individuals with FXS can inform efforts to increase health autonomy and decisional capacity as the disparities for individuals with I/DD in healthcare are addressed. Further, this study provided a basis for investigating how self-determination relates to decisional capacity in order to inform future efforts in shared decision making.

CHAPTER 2: REVIEW OF THE LITERATURE

Chapter two provides an overview of research on fragile X syndrome (FXS) and self-determination. First, literature on FXS including a general description of the genetic syndrome, the cognitive profile, and behavioral profile were reviewed. Second, this literature review provided an overview of topics within the research field of self-determination including historical information, theoretical perspectives, measures of self-determination, as well as factors that contribute to self-determination, and parent perceptions. The review concludes with an introduction to the present study.

Fragile X syndrome

Fragile X syndrome (FXS) is a genetic syndrome considered to be the most common hereditary cause of intellectual disability and is characterized by a range of physical, cognitive, and behavioral characteristics. FXS was originally referred to as Martin-Bell syndrome when it was first described as a disorder in 1943 by Martin and Bell. FXS was later confirmed by molecular studies of variations on the X chromosome (Lubs, 1969; O'Donnell & Warren, 2002).

FXS is an X-linked genetic syndrome and therefore affects males more than females. Prevalence rates of FXS at the full-mutation level are estimated at one in 3,600-4,000 males and one in 4,000-6,000 females (Beckett et al., 2005; Crawford et al., 2002). FXS results from a mutation of the X chromosome in the region of the fragile X mental retardation (*FMR1*) gene. FXS results from expansion of the cytosine-guanine-guanine (CGG) trinucleotide repeat sequence, which causes hypermethylation and silencing of the *FMR1* gene and interrupts encoding the fragile X mental retardation protein (FMRP; Verkerk et al., 1991). Whereas

neurotypical individuals have between 5 and 44 CGG repeats in the *FMR1* gene, individuals with said premutation have between 55 and 200 repeats and individuals with full mutation have 200 or more repeats. Full mutation of the *FMR1* gene results in a lack of FMRP caused by disruption of the methylation process. FMRP regulates production of proteins that are essential for neuronal migration, neurogenesis, and synaptic plasticity throughout development. FMRP plays several roles at the neural level (Willemsen, Oostra, Bassell, & Dictenberg, 2004). Several studies have documented a correlation between behavioral and cognitive abilities and FMRP expression levels in individuals with FXS (Loesch et al., 2003; Tassone et al., 1999). Therefore, it is widely accepted that low levels of FMRP play a causal role in the FXS phenotype (Farzin & Koldewyn, 2014).

As FXS is linked to a mutation on the X chromosome, males with a full mutation are typically more affected than females because, for females, only one of their two X chromosomes carries a repeat expansion on the *FMR1* gene (Farzin & Koldewyn, 2014). Due to the presence of a second X chromosome, females typically have higher cognitive functioning than males, however they are vulnerable to emotional and behavioral problems (Lachiewicz, 1995; Lachiewicz & Dawson, 1994). Females will produce a relatively higher level of FMRP, which will result in less pronounced and fewer symptoms associated with FXS.

Classical physical features of FXS include a narrow and elongated face, prominent and large ears, high arched palate, large head circumference, and macroorchidism (i.e., enlarged testicular volume in adult men). These physical features are associated with connective tissue problems, which often result in loose joints, flat feet, low muscle tone, heart murmurs, and other skeletal problems (Farzin & Koldewyn, 2014). The behavioral and cognitive phenotype is also well documented in the literature.

Cognitive Profile. Cognitive delays in development have a complex and dynamic role in neurodevelopmental disorders. Cognitive delay is often a nonlinear process and cannot simply be defined by performance on standardized measures in comparison to typically developing peers (Cornish et al., 2013). It is not known whether delay remains stable across time, illustrated by plateau in performance, or whether progress for those with FXS simply occurs at a slower rate than seen in typically-developing peers (Cornish et al., 2013). Although cognitive profiles in genetic syndromes are becoming increasingly well documented, intellectual delay does not necessarily imply global impairment across multiple cognitive domains (Cornish et al., 2013).

Intellectual impairment is the most frequent cognitive characteristic in FXS, with intellectual disabilities in the mild to severe range. Approximately 95% of adult males with FXS have an estimated IQ below 70 (Farzin & Koldewyn, 2014). Males have an average IQ in the 40s, whereas females often have an average IQ between 70 and 90. FXS is characterized by significant impairments in attention, impulsivity, and working memory (Cornish et al., 2013). Cognitively, individuals with FXS exhibit strengths and weaknesses both between and within domains, including relative strengths in vocabulary, long-term memory, and face perception and relative weaknesses in inhibitory control, short-term memory, numerical processing, visual motor integration and coordination, and selective and sustained attention (Aumgardner et al., 1995; Crowe & Hay, 1990; Simon & Finucane, 1996). Language delays are also prominent in individuals with FXS, particularly in the area of expressive language.

Behavioral Profile. The behavioral phenotype of FXS includes hyperactivity, impulsivity, aggression, and gaze aversion, as well as characteristics related to anxiety and attention-deficit hyperactivity disorder (Cohen et al., 1991; Sudhalter et al., 1990). Additionally, many individuals exhibit symptoms of autism spectrum disorder (ASD), including poor eye

contact, stereotyped behaviors, sensory aversions, and hand flapping and communication abnormalities (Bailey et al., 1998; Hagerman et al., 1986).

Autism in FXS. ASD is characterized by behavioral impairments in social communication and social interaction as well as restricted and repetitive behaviors, interests, and activities (APA, 2013). Individuals with FXS exhibit several shared behaviors with ASD, including self-injurious behavior, perseverative behavior, motor stereotypies, poor eye contact, and odd or delayed speech (Harris et al., 2008; Rogers, Wehner, & Hagerman, 2001).

Approximately one third of boys with FXS meet DSM-IV criteria for autism, while an additional one third meet criteria for pervasive developmental disorder (Harris et al., 2008; Rogers, Wehner, & Hagerman, 2001). Additionally, many individuals with FXS who do not meet criteria for an ASD diagnosis exhibit behaviors of the autism phenotype, and an estimated 90% of males with FXS exhibit at least one autistic behavior (Brock & Hatton, 2010). These data result in wide variability in reported prevalence rates of ASD in FXS. Reports of co-morbidity of ASD diagnoses in males with FXS range from 15-52% (Hernandez et al., 2008). Recent revisions and changes in the diagnostic criteria of ASD in the Diagnostic and Statistical Manual (DSM-5) further complicate the estimating prevalence and conceptualizing co-morbidity of ASD in individuals with FXS (Wheeler et al., 2014).

The broader autism behavioral phenotype is well documented in the fragile X population. Often, an ASD diagnosis precedes identification of FXS in children, due to the prominent behavioral symptoms of ASD (Bailey, Raspa, Bishop, & Holiday, 2009). Predictors of autism in FXS include increased problem behavior, lower adaptive behavior, lower IQ and developmental skills, increased age, and lower FMRP levels (Cohen, 1995; Hatton et al., 2006; Kau et al., 2004; Rogers et al., 2001). A series of studies suggest that children with FXS without ASD share

similar profiles with children diagnosed with developmental delay (DD), whereas children with FXS and comorbid ASD were more similar to children with idiopathic autism due to sensory characteristics and impairments in imitation skills (Rogers et al., 2001; Rogers, Hepburn, Stackhouse, & Wehner, 2003; Rogers, Hepburn, & Wehner, 2003). Literature suggests that children with FXS+ASD exhibit poorer developmental outcomes than children with FXS or idiopathic autism (Kaufman et al., 2004). Males with FXS males and comorbid ASD are more likely to experience poorer outcomes (e.g., weaker communication and social skills, lower adaptive behavior scores, more significant behavior problems, and greater cognitive impairment) than FXS males without ASD and individuals with idiopathic autism (Bailey et al., 2000). Although less research has been conducted on females with FXS, several overlapping ASD features have been described. One study investigated symptoms of ASD in females between the ages of five and 80 years (31 females with full-mutation and 43 females with premutation; Clifford et al., 2007). Using two gold standard diagnostic tools, the Autism Diagnostic Observation Scale-Generic (ADOS-G; Lord et al., 1991) and the Autism Diagnostic Interview-Revised (ADI-R; Lord, Rutter, & Le Couteur, 1994), Clifford et al. (2007) found a prevalence estimate of 13% for ASD in females with an *FMR1* mutation.

Social and communication impairments. Systematic review of behavioral markers suggests that social impairments are the most significant predictor that differentiates FXS+ASD from FXS (Brock & Hatton, 2010). Specifically, social withdrawal (e.g., avoidance and indifference) and adaptive socialization behaviors are often independent predictors of ASD in individuals with FXS (Budimirovic, Bukelis, Cox, Gray, Tierney, & Kaufmann, 2006). A high level of social withdrawal and social anxiety in individuals with FXS is often related to autistic behavior (Roberts et al., 2007). Significant social withdrawal is often only seen in individuals

with FXS with severe autistic symptoms (Budimirovic et al., 2006; Budimirovic & Kaufmann, 2011). Similarly, impairments in adaptive socialization behaviors, such as difficulty recognizing emotions and applying rules related to social interactions, are often strong predictors of ASD in FXS (Kaufmann et al., 2004). In cross-sectional (e.g., Kaufmann et al., 2004) and longitudinal studies (e.g., Hernandez et al., 2009), impaired adaptive socialization was the greatest contributor to ASD diagnosis and severity in the FXS population over communication parameters and overall cognition (Budimirovic & Kaufmann, 2011).

Although consistent predictors of autism in the communication domain have not been identified, some research suggests that individuals with FXS perform higher on measures of receptive language than expressive language, whereas individuals with FXS+ASD do not exhibit this strength (Lewis et al., 2006; Philofsky, et al., 2004). This finding is consistent for young children as well as adolescents and young adults. Retrospective parent-report of early language milestones in individuals with FXS who later received an ASD diagnosis suggests that children with FXS+ASD met language milestones significantly later (i.e., 10-12 months) than children with FXS (Hinton et al., 2013). Although the relationship between expressive language delays and ASD may weaken as children age, this association is evidenced in young children with FXS.

Social behavior profiles for individuals with FXS and FXS+ASD have been proposed in the literature. Based on a longitudinal study, Hernandez et al. (2009) proposed a profile of autistic behaviors in FXS, which is primarily characterized by impairment in social interactions, specifically, peer relationships, socially-relevant communication, severity of social withdrawal, imitative play, and delays in adaptive communication and receptive language. Kau et al. (2003) proposed a social behavior profile in boys with FXS+ASD that includes adaptive socialization abnormalities, in addition to distinct patterns of adaptive behavior and aberrant behaviors (e.g.,

attentional problems, stereotypic behavior, and irritability). Kaufmann et al. (2004) expanded the social behavioral profile by proposing that social interaction deficits (e.g., peer interaction and imaginative play) are distributed on a continuum for individuals with FXS, FXS and pervasive developmental disorder (FXS+PDD), and FXS+ASD. Diagnosis is complicated by recent changes to the DSM, which preclude the diagnosis of PDD (APA, 2013). As compared to individuals with idiopathic autism, individuals with FXS+ASD show significantly less impairment in discrete social behaviors, such as shared enjoyment, social smiling, and quality and amount of social interactions (Hall et al., 2010; Kau et al., 2004).

FXS is commonly characterized by social avoidance and anxiety that can cause an individual to exhibit symptoms similar to the autism phenotype. Therefore, social markers must be interpreted with caution when used to identify comorbid ASD in FXS (Roberts et al., 2007). For instance, despite interest in social interaction, high levels of social avoidance is often observed and is commonly attributed to difficulty warming up in novel situations (Roberts et al., 2007). Although individuals with FXS+ASD tend to exhibit milder social withdrawal and less impaired social responsiveness in comparison to individuals with idiopathic autism, individuals with FXS+ASD typically exhibit increased social avoidance (Rogers et al., 2001; Wolff et al., 2012). Additionally, while males with FXS and comorbid ASD exhibit similar levels of social approach as males with FXS, males with FXS and comorbid ASD consistently display poor eye contact (i.e., less modulation over time), whereas males with FXS exhibit improved eye contact over increasingly familiar social situations (Roberts et al., 2007). For this reason, there is significant debate surrounding ASD diagnoses in FXS, raising questions regarding the difference between idiopathic autism and co-morbid ASD in FXS.

Restricted and repetitive behaviors and interests. Although restricted and repetitive behaviors and interests (RRBIs) are well documented in the FXS population, there is a paucity of research comparing RRBIs in individuals with FXS and individuals with FXS+ASD. For example, self-injurious behavior is observed in approximately 58% of males with full-mutation FXS (Symons et al., 2003). Research suggests that RRBIs do not differentiate individuals with FXS+ASD from individuals with FXS; however, differences between individuals with FXS+ASD and individuals with idiopathic autism are documented in the literature. Specifically, males with FXS+ASD exhibited significantly less ritualistic and compulsive behavior, but increased repetitive motor behaviors than males with idiopathic autism (Wolf et al., 2012).

Symptom stability. Few studies have addressed the stability of autistic behaviors in individuals with FXS through longitudinal investigation, and little is known regarding changes in severity of autistic behaviors with age. Although clinical reports have proposed that young children with FXS may exhibit increased autistic behaviors, a paucity of research supports this notion (Hatton et al., 2006). Cross-sectional analyses of children with FXS between the ages of 1.5 and 14.7 years suggests that autistic behaviors are relatively stable and that social withdrawal increases slowly but significantly over time in males with FXS+ASD (Hatton et al., 2006). The stability of ASD diagnoses in males with FXS was also investigated in a longitudinal evaluation (Hernandez et al., 2009). Findings suggested that as the FXS+ASD population exhibited less severe impairments over time, the FXS group demonstrated increasing autistic behaviors (i.e., social impairments), which resulted in less differentiation between the groups over time and approximately 70% diagnostic agreement over time.

Parent report of ASD in FXS. Although there is considerable debate surrounding the nature of ASD in FXS, parent report of ASD symptoms in FXS provides additional insight into

the perceived behavioral phenotype. Per parent report, repetitive and stereotyped behaviors were the strongest predictors of ASD in FXS (Brock & Hatton, 2010). Increased repetitive behavior was also positively correlated with social impairments. Another study using retrospective parent report found that children with FXS and comorbid ASD acquired language and motor milestones significantly later than children with FXS not diagnosed with ASD (Hinton et al., 2013). Based on a national survey of parents of children with FXS, 46% of full-mutation males were diagnosed with ASD (Bailey, Raspa, Olmsted, & Holiday, 2008). In comparison, Wheeler et al (2015) found that fewer males (28%) and females (11%) with FXS met criteria for ASD based on the DSM-5 (APA, 2013) than on the DSM-IV-TR (APA, 2000).

Anxiety. Symptoms of anxiety and social withdrawal are core features of the FXS phenotype. Diagnosing anxiety disorders in individuals with ID, particularly those with FXS, is primarily conducted through questionnaires and behavioral checklists used to identify anxiety symptoms (Cordeiro, Ballinger, Hagerman, & Hessl, 2011). A national survey of parents of children with FXS found 70% of males and 56% of females were treated for symptoms of anxiety or had a formal diagnosis (Bailey et al., 2008). One study used the Child Behavior Checklist (CBCL) parent-report questionnaire and found that among 38 females between the ages of 4- and 11-years old, 47% scored above the 98th percentile on social withdrawal (Lachiewicz, 1992). Cordeiro et al. (2011) used a diagnostic clinical interview and a questionnaire normed with an ID sample to assess the prevalence of anxiety disorders in FXS, examine factors associated with anxiety disorders in FXS, and to compare rates of anxiety disorders in FXS to the population of individuals with ID and Williams Syndrome. Nearly 60 males and 39 females between the ages of five and 33 years old participated in the study. Parents were administered the Anxiety Disorders Interview Schedule for DSM-IV: Parent Report

Version (ADIS-IV; Silverman & Albano, 2004) to measure the severity of present symptoms of anxiety. The Anxiety Depression and Mood Scale (ADAMS; Esbensen et al., 2003) is a 28-item questionnaire designed and normed with individuals with ID. Of the sample, 82.5% of individuals met criteria for at least one anxiety disorder and 58.3% of individuals met criteria for multiple anxiety disorders (Cordeiro, Ballinger, Hagerman, & Hessl, 2011). Individuals most frequently met criteria for specific phobia, social phobia, and selective mutism. Individuals with below average IQ had a higher rate of specific and social phobia than individuals without intellectual disabilities. Significantly more individuals with an ASD diagnosis were diagnosed with selective mutism. In comparison to idiopathic ID, individuals with FXS have significantly higher rates of several anxiety disorders (Cordeiro et al, 2011).

Self-Determination

During the past 20 years, self-determination has been used within the broader field of disability research as a multi-faceted construct (Wehmeyer, 1999). Self-determination refers to an individual acting as the primary causal agent in his or her life, including making decisions and choices regarding one's quality of life (Wehmeyer, 1996a). Thus, self-determination should be defined as consisting of behaviors that are free from undue external interference or influence. Self-determination can refer to the skills and behaviors, opportunities, and supports that individuals have to act as causal agents (Shogren, Wehmeyer, Reese, & O'Hara, 2006). Self-determination is a dispositional characteristic in that it refers to the manner in which an individual's behavior is relatively consistent across different situations due to the organization of psychological, cognitive, and physiologic elements (Wehmeyer, 1999). Self-determination is a trait that emerges across the lifespan. As individuals learn various skills and gain attitudes that allow them to be causal agents in their lives, they act in a more self-determined manner.

The term self-determination was initially used exclusively in the fields of philosophy, and political science, and later in the field of psychology in the 1970s (Wehmeyer, 1999). In philosophy, the construct was used in topics related to determinants of behavior. In the field of political science, it was a term used to refer to self-governance, freedom, and independence. In psychology; self-determination has been included in theories of personality and motivation. Later, self-determination was a term used within other fields, including the disability field. Self-determination was first used during the normalization movement for people with disabilities that occurred during the 1990s, which changed the way service delivery and supports for individuals with ID were conceptualized, particularly within the field of special education and transition from high school (Shogren, Wehmeyer, Matthew Reese, & O'Hara, 2006; Wolfensberger, 1972).

Self-determination as a concept within the field of special education began in 1990 when the Office of Special Education Programs (OSEP) in the U.S. Department of Education funded several projects on the promotion of self-determination (Wehmeyer, 1999). The OSEP initiative marks the beginning of wide recognition of self-determination for individuals with I/DD, particularly within the field of special education. As an educational construct, self-determination is defined as, "the attitudes and abilities that lead individuals to define goals for themselves and to take the initiative in achieving these goals" (Federal Register, 1989, p. 38166). During the OSEP initiative, significant changes again occurred in service delivery, where individuals with ID received community-based supports rather than institutionalization (Wehmeyer, 1999). Relatedly, the way the term 'disability' was conceptualized also began to change, transitioning from a medical model to a model that focused on social and environmental factors (Shogren, et al., 2006).

When considering the self-determination of individuals with I/DD, two main factors are proposed in the literature as important considerations. First, the *capacity* to engage in self-determined behaviors or act in a self-determined manner (i.e., skills an individual has to make choices or decisions, problem solve, and advocate for oneself) needs to be assessed (Wehmeyer & Garner, 2003). Second, the *opportunity* an individual has to practice self-determination within various environments and individuals who may support or inhibit self-determined behaviors should also be considered. Acting in a self-determined manner requires not only certain abilities, but also attitudes (Ward, 1998; Wehmeyer, 1992a). Some individuals with I/DD may have ability, but lack the attitude or vice versa. For example, some individuals with I/DD may have the ability to make decisions, but lack the attitudes and beliefs that their decisions can have an effect on their lives, making it less likely that they will make decisions, despite their cognitive ability to do so. Conversely, some individuals with I/DD may have attitudes and beliefs that lend them to act in a self-determined manner, such as feeling self-efficacious, yet lack the cognitive skills or ability to effectively make decisions. These individuals may be more likely to make a decision, but not necessarily a decision that would result in a satisfying outcome. Although capacity and opportunity are two factors that play a role in the lives of all individuals, careful consideration of each of them for individuals with I/DD is increasingly important in order to successfully address barriers to the development of self-determination.

Theoretical Perspectives of Self-Determination

Depending on the theoretical perspective, self-determination is defined or described in varying ways, (i.e., what is considered self-determined behavior) and empirically validated using various measures and assessment. Within the Functional Model of Self-Determination (Wehmeyer, 1999), self-determination is defined by the purpose or function of the behavior,

which is discussed in greater detail below as it lends itself well to conceptualizing self-determination for the present study.

Alternative theories define self-determination by considering the interaction of an individual's capacities in various opportunities to improve their situations and get what they believe they want and need in life (Mithaug et al., 2003). In the Theory of Self-Determination, capacity refers to an individual's ability, knowledge and perceptions, which enable an individual to be self-determined in the context of school and learning. Opportunities refer to chances for an individualto apply ability and knowledge (Wolman, 1994). While the Functional Model of Self-Determination is focused on personal characteristics, the Self-Determined Learning Theory emphasizes the *process* in which individuals become self-determined (Shogren et al., 2008). Another model, the Ecological Model of Self-Determination (Wehmeyer et al., 2011), is derived from Bronfenbrenner's ecological systems theory (Bronfenbrenner, 1979). Within this theory, self-determination is characterized by a complex process that occurs as the result of both the individual and environment (Wehmeyer et al., 2011). Finally, the Self-Determination as Self-Regulation theory (Mithaung, 1998) posits that self-determination is a form of self-regulation uninterrupted by external influence and always occurs within a social context. Although these models vary, several common threads can be observed, such as the developmental perspective each take on self-determination, the role of personal capacity, and the recognition of environment and context. Each theory uses a person-environment fit model with self-determination as the outcome (Wehmeyer et al., 2011).

Functional Model of Self-Determination. The Functional Model of Self-Determination is a theory based on the functions of self-determined behavior, rather than the specific behaviors themselves (Wehmeyer, Kelchner, & Richards, 1996). This theory conceptualizes self-

determination within a person-environment interactional framework (Wehmeyer et al., 2011). Self-determination is a construct that cannot be defined as a response class (e.g., a set of behaviors) as almost any behavior can be argued as an attempt to exert control and the nonoccurrence of a given action or behavior may also be illustrative of self-determination (Wehmeyer, 1999). Additionally, defining self-determination as a response class neglects cultural differences, which may influence what is considered acceptable and unacceptable self-determined behavior in a social realm.

The Functional Model of Self-Determination definition of self-determination includes carefully chosen terms including 'causal agent' and 'undue external influence or interference' to describe self-determination and self-determined behavior (Wehmeyer, 1999). The concept of causal agency refers to how an individual can be the cause for things to happen in his or her life and that an action that can be purposeful (Wehmeyer, 1999). Within the Functional Model of Self-Determination, causal agents are individuals who act with purpose to shape or influence their future. Similarly, the term 'undue' as it relates to external influence is used to recognize that humans are not entirely autonomous or independent; rather they are interdependent to varying degrees in various realms of their lives. This definition is careful to explain that self-determination does not translate to complete autonomy with an absence of influence; rather it suggests that external influence and interference are subjective and contextual, varying for each individual.

Wehmeyer's (1999) Functional Model of Self-Determination suggests that an individual's relative self-determination is influenced primarily by four main factors; (a) *Capacity*, which is influenced by learning and development, (b) *Opportunity,* which takes into consideration the environment and individual experience, (c) *Perception and Beliefs*, and (d)

Supports. Capacity, opportunity, and perceptions and beliefs each interact with one another and influence the four essential characteristics of self-determination (i.e., autonomy, self-regulation, psychological empowerment, and self-realization). *Supports* encompasses any support mechanisms (e.g., resources, accommodations) and is considered an independent influence on the four essential characteristics as well (i.e., does not interact with the other three factors in the model.

Skills and Characteristics of Self-Determination. Attitudes and abilities are the component elements of self-determined behavior that are identified by four essential characteristics of self-determined behavior as outlined by the Functional Model of Self-Determination (Wehmeyer, 1999). 'Essential characteristics' is a term used to imply that an individual's behaviors must reflect each of these four characteristics to some degree in order to be considered self-determined. As will be discussed further, several factors may impact the degree to which these characteristics are present (e.g., age, capacity, opportunity, circumstance) and the degree to which self-determination is expressed. Autonomy, Self-Regulation, Psychological Empowerment, and Self-Realization are each necessary but not sufficient alone for self-determined behaviors. Whereas autonomy and self-regulation are two functional characteristics, which are primarily focused on actions and behaviors, psychological empowerment and self-realization emphasize the importance of cognitive contributions to self-determination.

Autonomy. Autonomy is considered to be synonymous with individuation; a term used in the field of developmental psychology to refer to the formation of individual identity (Damon, 1983). Individuation is a "progression from dependence on others for care and guidance to self-care and self-direction" (Sigafoos, Feinstein, Damond, & Reiss, 1988, p. 432). Subsequently,

behavioral autonomy is considered the outcome of individuation (Wehmeyer, 1999). Behavioral autonomy then encompasses actions that are the result of people acting according to their interests, preferences and/or abilities, as well as independently.

Behavioral autonomy is further separated into four different types of activities: self- and –family care activities, management activities, recreational/leisure activities, and social/vocational activities (Wehmeyer, 1999; Sigafoos, Feinstein, Damond, & Reiss, 1988). Self- and family care activities encompass family-oriented functions, personal care, household functions, and daily living activities. Management activities refer to interactions with the environment to a given degree. For example, such activities include engaging with community resources, and fulfilling responsibilities and obligations. Recreational activities refer to the degree with which an individual incorporates personal preferences and interests when choosing to engage in an activity. Similarly, social/vocational activities include social involvement and vocational activities.

Self-Regulation. Self-regulation refers to an internal response system that allows individuals to inspect their environments. It includes response repertoires in order to make future decisions on how to act, to engage in an action, evaluate the outcome, and revise plans as needed (Wehmeyer, 1999; Whitman, 1990). Self-regulated behaviors include self-management strategies such as self-instruction, self-monitoring, self-evaluation, and self-reinforcement. Also included are goal-setting and attainment, problem-solving, decision-making, and strategies of observational learning (Agran, 1997).

Psychological Empowerment. Psychological empowerment is a multi-dimensional construct that encompasses more than one dimension of perceived control. From a cognitive standpoint, psychological empowerment refers to personal efficacy. From a personality

standpoint; it refers to locus of control. Psychological empowerment also includes motivational domains (Wehmeyer, 1999; Zimmerman, 1990). This multidimensional account of perceived control, or psychological empowerment, was found to be a valid construct in previous research (Zimmerman & Rappaport, 1988).

Self-Realization. Self-realization is a construct that refers to an individual's knowledge and awareness of what he or she does well and acts accordingly (Wehmeyer, 1999). Self-realization involves use of comprehensive and relatively accurate knowledge of oneself, including strengths and weaknesses. Self-realization refers to self-knowledge and self-understanding, which is formed through experience. It is also influenced by interpretation of the environment, reinforcement, and attributions of an individual's own behavior.

Measures of Self-Determination

Though there is a paucity of research seeking the perceptions of self-determination of individuals with disabilities (Chambers et al., 2007), one study asked 778 individuals with I/DD to rate the importance of self-determination and found that individuals with I/DD rated the importance of self-determination higher than their professionals and families (Schalock et al., 2005). Three norm-referenced measures of self-determination that incorporate self-report are documented in the literature. Each has been used to varying degrees for research, educational planning, and to identify strengths and limitations in self-determination of individuals with disabilities. These measures can be used to promote self-determination by identifying current levels of self-determination, as well as interests and abilities related to self-determination (Wehmeyer, 2011). Scales also vary by theoretical perspective and focus. While some measures are global assessments of self-determination, others focus on specific aspects (e.g., academic).

The Arc's Self-Determination Scale. The Arc's Self-Determination Scale (SDS; Wehmeyer & Kelchner, 1995) is a tool designed to enable individuals to become more self-determined by providing an avenue to evaluate their own beliefs about themselves and identify relative strengths and weaknesses related to self-determination (Wehmeyer, 1996). The Arc's SDS is a measure that is linked to and operationalizes the functional model of self-determination (Wehmeyer et al., 2011). This tool provides a global measure of self-determination that encompasses the four functional characteristics as described in the functional model of self-determination. These four characteristics serve as scale domains and some are further divided into subdomains (Wehmeyer, 1996).

The autonomy domain was divided into two subdomains, independence and acting on the basis of preferences, beliefs, interests and abilities. Similarly, the self-regulation domain was divided into interpersonal cognitive problem solving, and goal-setting and task performance. The full scale includes 72 items, with 32 multiple-choice items in the Autonomy domain, 9 items in the Self-Regulation domain, 16 items in the psychological empowerment domain, and 15 items in the Self-Realization domain.

Items were developed by adapting questions from existing measures within each domain and by author generation (Wehmeyer, 1996). All items were modified to read at a fourth-grade level or below and items were formatted to be more accessible to individuals with I/DD (i.e., multiple choice items where applicable). For example, in addition to author-generated items, the Autonomy domain adapted items from the Autonomous Functioning Checklist (AFC; Sigafoos, Feinstein, Damond, & Reiss, 1988), a parent-report measure of behavioral autonomy in the areas of self and family care; management; recreational activity; and social and vocational activity. Part of the Self-Regulation domain was adapted from the Means End Problem-Solving technique

(MEPS; Platt & Spivack, 1989), which provides a scenario with a beginning and ending to a story and asks respondents to generate the means by which the given outcome was achieved. The Self-Realization domains also used author-generated items and adapted items from the Short Index of Self-Actualization (Jones & Crandall, 1986) based on a factor analysis that suggested 11 representative items. All items were included in a pilot study, exploratory factor analysis, and correlation analysis to demonstrate adequate construct validity and concurrent criterion-related validity (Wehmeyer, 1996). Construct validity was documented through discriminative validity and factorial validity. The Arc's SDS adequately differentiated between students with cognitive disabilities and those without disabilities (Wehmeyer, 1995). Factor analysis indicated that the four scales accurately reflect the constructs that they were used to measure. Internal consistency reliability for the complete scale, excluding the self-regulation domain, was .90. Cronbach's alpha for the autonomy domain was .90, .73 for the psychological empowerment domain, and .62 for the self-realization domain.

American Institutes for Research Self-Determination Scale. The American Institutes for Research Self-Determination Scale (AIR; Wolman, Campeau, Dubois, Mithaug, & Stolarski, 1994) is a measure that collects student-, parent- and teacher-report of student capacity and opportunity for self-determination. The self-report version consists of 24 questions that yield Capacity and Opportunity domain scores. Capacity is defined as behaviors that students engage in to meet their goals and needs. Opportunity is defined by a student's feelings about their opportunity to engage or perform a given behavior. This measure was normed with 450 students with and without disabilities (Wolman et al., 1994) and demonstrates reliability and validity in measures of capacity and opportunity. Although the AIR measure captures different aspects of

self-determination than the Arc's SDS (Wehmeyer & Kelchner, 1995), the two measures are related (r = .50; Wehmeyer, 2011).

Self-Determination Assessment Battery. The Self-Determination Assessment Battery (SDAB; Field, Hoffman, & Sawilowsky, 2004) was developed within the context of an intervention theory. The SDAB consists of multiple domains that measure behavioral, cognitive, and affective aspects of self-determination based on student-, parent-, and teacher-report. Additionally, the assessment includes an observation checklist and a self-determination knowledge pretest for students, which assesses knowledge and skills taught through the Steps to Self-Determination curriculum (Field & Hoffman, 1994). The Student Scale consists of 92 items that comprise four subscales; General Positive, General Negative, Specific Positive, and Specific Negative to provide information regarding student's perceived strengths and weaknesses. Although the SDAB measure is designed primarily to evaluate self-determination within the settings of home, education, and related environments, it emphasizes the educational setting. The Parent- Perception Scales and Teacher-Perception Scales are each 30 items about behaviors, abilities and skills associated with self-determination, which relate specifically to Field and Hoffman's (1994) intervention curriculum. The SDAB has a high level of internal consistency reliability that ranges from .83 to .97 (Field, Hoffman, & Sawilowsky, 2004). Construct validity was also demonstrated through the use of a known intervention, the Steps to Self-Determination (Field & Hoffman, 1992).

Factors Contributing to Self-Determination

Several variables contribute to self-determination (Wehmeyer et al., 2011). The following discussion highlights variables pertinent to the topic of self-determination in individuals with FXS.

Gender. Previous research suggests mixed and limited findings on the effect of gender on self-determination. Wehmeyer and Garner (2003) found no differences in self-determination by gender. However, Nota et al. (2007) found that women exhibited higher levels of self-determination than men when measured by the Evaluation of Self-Determination Instrument (ESI, Nota et al., 2007), which assessed self-determination in various daily activities, expressing ideas, opinions, and emotions, and choices and desires. However, this measure was administered to staff members who supervised individuals with intellectual disabilities and did not consist of self-report. In one study using a self-report measure, the Arc's Self-Determination Scale, females also tended to score higher than males (Shogren, 2007).

Age and life stages. A developmental trend in self-determination begins in adolescence and gradually increases through adulthood before leveling off (Wehmeyer, 1999). However, other studies suggest that age does not predict level of self-determination (Wehmeyer & Garner, 2003). Nota et al. (2007) found no differences in self-determination across age ranges in a study of 141 adults with intellectual disabilities between the ages of 16 and 65 years old (mean age 35.75 years). Although age alone elicits mixed findings, life experiences may strongly influence self-determination (Wehmeyer et al., 2011). For example, while age can predict high and low autonomy, individuals with I/DD experience a wide range of opportunities and experiences at different rates (Wehmeyer et al., 2011). Life experiences may ultimately influence their level of self-determination regardless of their age.

Cognitive ability. Although research demonstrates a consistent correlation between intellectual ability and self-determination, (Stancliffe et al., 2000; Wehmeyer, 1996; Wehmeyer & Garner, 2003) the relationship is relatively weak and complex. Though overall self-determination will be limited by the complexity of knowledge and skills acquired by an

individual as a function of intellectual disability, if opportunities to develop such knowledge and skills are restricted, the individual's opportunity to act in an increasingly self-determined manner is also inhibited (Wehmeyer & Garner, 2003; Wehmeyer 2006). In other words, if one assumes that an individual cannot gain the skills needed to act in a self-determined manner and thus limits opportunities and appropriate supports to act in a self-determined manner, many individuals with I/DD may not have the opportunity to fully develop self-determination regardless of their ability to do so.

Wehmeyer and Garner (2003) used discriminant function analysis to examine self-determination and autonomy in 301 adults with I/DD. One hundred and forty-nine individuals were identified as having a mild intellectual disability (i.e., IQ score between 56 and 75), 93 were identified as having an intellectual disability in the severe range (IQ score of 55 or lower), and 59 individuals were identified as having a developmental disability. Individuals ranged between 19 and 73 years old with a mean age of 38.5 years. The Arc's Self-Determination Scale (Wehmeyer & Kelchner, 1995) and the Autonomous Functioning Checklist (AFC; Sigafoos et al., 1988) were used as self-report measures of self-determination and autonomy, respectively. Additionally, the Adult Version of the Nowicki Strickland Internal-External Scale (ANS-IE; Nowicki & Duke, 1974) and the Life Choices Survey (LCS; Kishi et al., 1988) were used. Wehmeyer and Garner (2003) found that intellectual ability was not a significant contributor to autonomy or self-determination. Rather, opportunities to make choices contributed significantly to both autonomy and self-determination. Wehmeyer and Garner (2003) found main-effect differences among disability groups on self-determination scores. Specifically, individuals in the severe intellectual disability group had lower self-determination scores than individuals with mild intellectual disabilities or developmental disabilities. Although there were also significant

correlations between IQ score and self-determination scores among individuals with the same intellectual disability, Wehmeyer and Garner (2003) reported that IQ was not a significant predictor for self-determination or autonomous functioning and there were no significant differences in autonomous functioning scores among the three groups. IQ scores were not predictive of high versus low self-determination or autonomous functioning groups, suggesting that individuals across intellectual ability can exhibit comparable levels of self-determination with appropriate supports in place.

Adaptive behavior. Though there is only a modest correlation with intellectual ability and self-determination, some research suggests that adaptive behavior may have a strong influence on self-determination. For example, social ability, as measured by the Social Ability Evaluation Scale for Adults with Mental Retardation (VAS-ARM; Marchesini, & Nota, 2001) was significantly correlated with self-determination (Nota et al., 2007). Social ability was measured based on basic social abilities (e.g., saying hello, accepting compliments) and interaction management (e.g., expressing wants and needs, following advice). Social skills were also identified as a moderating factor in self-determination of students with emotional disturbances and learning disabilities (Carter, Lane, Pierson, & Glaeser, 2006; Pierson, Carter, Lane, & Glaeser, 2008).

Autism. There is a dearth of research addressing differences in self-determination among adolescent students with various disabilities (Carter, Lane, Pierson, & Glaeser, 2006). Moreover, there are only two studies investigating self-determination of individuals with ASD (Chou, Palmer, Wehmeyer, & Lee, in press; Fullerton & Coyne, 1999). As social skills are a potential contributor to global self-determination, the question of whether self-determination differs in individuals with autism is an important one. Moreover, impairments in social understanding

could potentially limit opportunities to act in a self-determined manner (Wehmeyer, Shogren, Zager, Smith, & Simpson, 2010).

Fullerton and Coyne (1999) conducted interviews with 23 students with ASD to determine how individuals with ASD understand concepts related to self-determination and found that self-knowledge played an important factor in self-determination. Chou et al. (in press) compared individuals with ID, learning disability (LD), and autism spectrum disorder (ASD) on scores from the Arc's Self-Determination Scale (Wehmeyer & Kelchner, 1995; Chou, Palmer, Wehmeyer, & Lee, in press). Ninety-five of 222 middle- and high-school students in the sample had ASD. A one-way between-subjects multivariate analysis of variance was run on six dependent variables related to self-determination. Chou et al. (in press) found that students with autism expressed lower levels of autonomy in comparison to students with LD. In comparison to students with LD and ID, scores across each factor of self-determination were lower for students with ASD.

Environmental factors. Although cognitive ability influences the degree of support an individual with I/DD needs to become increasingly self-determined, the degree of self-determination is influenced more by the environment and supports than by cognitive capacity (Wehymer et al., 2011). Wehmeyer and Bolding (1999) investigated the role of the environment on self-determination while controlling for intellectual ability in a matched-samples design. Two-hundred-seventy-three individuals with ID who worked or lived in one of three different environments that promoted or limited self-determination (e.g., community-based; community-based congregate such as a group-home or sheltered employment; or a non-community-based congregate, such as an institution) were matched by IQ as well as gender and age where feasible. Wehmeyer and Bolding (1999) found significant differences in self-determination and autonomy

based on environment. Individuals who lived or worked in non-congregate community-based settings expressed significantly less self-determination. Opportunities within different environments were considered a strong influence on reported levels of self-determination in individuals with intellectual disability, regardless of an individual's degree of disability.

Wehmeyer and Bolding (1999), however, did not account for the fact that the individuals grouped by environment may already express higher levels of self-determination, which contributed to the environments in which they were currently working or living. Therefore, Wehmeyer and Bolding (2001) repeated the study with individuals with ID before and after they moved from more restrictive to less restrictive working or living environments and found significant positive changes in self-determination when individuals moved to less restrictive environments. These two studies suggest that environment can play a strong role in the expression of self-determination of individuals with I/DD.

Though the school setting has been the primary focus in efforts to promote self-determination, self-determination prospects are influenced by opportunities individuals have not only at school, but also at home (Mithaug, Mithaug, Agran, Martin, & Wehmeyer, 2003). The home environment provides numerous opportunities to practice skills and develop attitudes related to acting in a self-determined manner. For individuals with disabilities to act in an increasing self-determined manner, these skills not only need to be developed, but individuals need repeated exposure to opportunities to apply newly learned skills and generalize these skills (Sands, Bassett, Lehmann, & Spencer, 1998). In fact, one study used discriminant function analysis to illustrate that level of intelligence was not a strong of a predictor of self-determination, whereas opportunities for an individual to make choices in one's environment was a significant predictor (Wehmeyer & Garner, 2003).

Zhang (2005) found that children and youth without disabilities from Caucasian families exhibited increased personal independence compared with African American and Asian counterparts. Additionally, Zhang (2005) found that children without disabilities of parents who graduated from college experienced increased opportunities to act in a self-determined manner (e.g., express their interests, make decisions, and set goals) than children of parents who did not graduate from college. Children of higher-income families had more opportunities to be engaged in skills related to self-determination. These findings suggest a relationship between socioeconomic status and parenting-practices related to self-determination for children. Parents of children with disabilities were less likely to provide opportunities to make choices and decisions, set goals, and recognize their needs than parents of children without disabilities (Zhang, 2005). This finding is consistent with the literature suggesting that children with disabilities are provided fewer opportunities to engage in trial-and-error activities, make choices and decisions, and goal setting (Bannerman, Sheldon, Sherman, & Harchik, 1990). Within the home, families affect the development of self-determination (Turnbull & Turnbull, 1996). In addition to family characteristics, including cultural beliefs, values, expectations, and coping, family interactions such as role expectations and cohesion, family functions, and family lifespan issues all may impact self-determination.

Adult Outcomes. In the past, individuals with I/DD have experienced limited opportunities to act in a self-determined manner, such as making choices and expressing preferences (Wehmeyer & Metzler, 1995). Five studies have investigated the impact of self-determination on outcomes for individuals with I/DD. Wehmeyer et al. (1996) found that individuals who were self-determined in comparison to those less self-determined were significantly different across multiple behavioral indicators of autonomy and control. Similarly,

students with I/DD with higher levels of self-determination exhibited more positive post-school outcomes, such as employment and financial independence, than students with lower levels of self-determination at one and three years post-graduation (Wehmeyer & Palmer, 2003; Wehmeyer & Schwartz, 1997). Quality of life for individuals with I/DD is also influenced by self-determination. Wehmeyer and Schwartz (1998) and Lachapelle et al. (2005) found that self-determination scores predicted higher quality of life.

Parent Perceptions of Self-Determination

The topic of self-determination has been primarily addressed in the school setting, particularly with regard to the field of special education and transition services for youth with disabilities. Less is known about parent perspectives of self-determination, despite the significant role they play to facilitate and promote self-determination in their children with I/DD. Parent support and involvement in the development of self-determination is recognized as a critical factor for youth with disabilities (Abery, 1994, Field & Hoffman, 1994; Martin & Marshall, 1998; Wehmeyer, 1996). Parents and caregivers have an enduring impact on their children and support their children, particularly those with disabilities. Parents, in comparison to schools and educators, have a different vantage point to view self-determination and promote the attitudes and behaviors related to self-determination (Carter et al., 2013). Moreover, parents spend a significant amount of time with their children and have innumerable opportunities to promote self-determination in their children. There is a paucity of research investigating how parents value self-determination as a goal for their child and how they view the opportunities for self-determination outside of school (Zhang, 2006; Zhang et al., 2005).

Similar to the effect some variables (e.g., age) have on an individual's self-determination, these factors also contribute to how parents interpret current self-determination of their children,

the potential for their children's self-determination, and the importance of fostering self-determination. Culture, for instance, affects one's perceptions and behaviors. It is a learned and shared knowledge or interpretation of reality that likely affects how parent's parent and emphasizes behaviors related to self-determination for their children. Importantly, culture also determines and defines how an individual identifies, understands, or accepts disability (Wehmeyer et al., 2011). Parent assessment of their child's self-determination may be influenced by a myriad of variables including child, parent, and school specific factors (Shogren, 2011; Wehmeyer et al., 2011). Parents play an important role in helping their children develop the skills, knowledge, and beliefs related to self-determination and in providing opportunities and reinforcement for their children to act in a self-determined manner (Field & Hoffman, 1999). Parents can serve as models for their children to develop skills, attitudes, and behaviors related to self-determination.

Carter et al. (2013) surveyed 627 parents of children with ID or ASD to assess parent perspectives of self-determination. They assessed how parents rated the importance of seven component skills of self-determination, how parents rated their child's performance on each of these skills, and how they perceived their child's overall capacity for self-determination. Carter et al. (2013) also investigated factors that were associated with the level of importance and performance parents rated for their child's self-determination. Parents of students who participated in the study were identified across 34 randomly selected public school districts (Carter et al., 2013). Each student received special education services under intellectual disability or autism, was enrolled in kindergarten through 12th grade, and was between the ages of five and 18 years old. Of all parents in the sample, 85.8% of the parents or caregivers who responded to the survey were mothers, 11.5% were fathers, nearly 1% were grandmothers, and the remaining

2% indicated other (e.g., adoptive parent, foster parent, step-parent). Approximately 40% of respondents reported that their child was eligible for free or reduced-price lunch (FRL). The average age of the students reported was 11.72 years old and almost three-quarters of the students were male. Students who were older than 18 years were excluded from the study as their disabilities were considered to be more severe and less likely to experience the same variety of educational environments. Three hundred and five students included in the study sample were eligible for special education under autism, 190 were identified as having an intellectual disability, 28 were reported to have both autism and intellectual disability, and 94 were reported to have another disability (e.g., cerebral palsy). The majority (80%) of parents described their child's disability in the mild/moderate range and 20% described their child's disability in the severe/profound range. Nearly 40% of parents reported that their child had a one-on-one assistant in school at the time of the survey.

Carter et al. (2013) developed a questionnaire for parents that paralleled a survey used in previous studies of teachers (Stang et al., 2009), special educators (Carter et al., 2008), and paraprofessionals (Carter, Lane, & Sisco, 2011). The survey asked parents to rate the importance and ability of their children in several component skills: decision-making, choice-making, goal-setting, problem-solving, self-advocacy and leadership, self-awareness and self-knowledge, and self-management and self-regulation (Wehmeyer et al., 2007). Additionally, items from the AIR Self-Determination Scale (Wolman, Campeau, DuBois, Mithaug, & Stolarski, 1994) parent version were also incorporated to assess students' self-determination capacities (i.e., the extent beliefs are connected to expectations, actions, choices, and results). Over 70% of parents rated decision-making, choice-making, problem-solving, self-regulation, self-management, self-awareness and self-knowledge as being very important skills for their children and over 60% of

parents rated self-advocacy, leadership, and goal setting as somewhat important. Parents consistently rated their child's skill level as low, particularly in the areas of self-awareness and self-knowledge. Carter et al. (2013) found low correlations between importance and skill performance as rated by parents. Disability level, intellectual disability, and FRL status were significant predictors of how important parents perceived self-determination. Parents who described their child's disability in the severe/profound range rated the importance of learning skills related to self-determination lower than parents of children in the mild/moderate disability range.

Carter et al. (2013) is the only identified study to assess parent perception of specific skills associated with self-determination. One previous study assessed parent beliefs of global self-determination for high school students between the ages of 16 and 21 years old with low- and high-incidence disabilities (Grigal, Neubert, Moon, & Graham, 2003). Beliefs regarding teaching self-determination in school curricula, including students in Individualized Education Plan (IEP) meetings, and their child's opportunities to express and make choices in school were assessed. The parent survey included nine statements about self-determination including their child's ability to express their interests and abilities, participation in IEP meetings, decision-making, self-advocacy, and goal setting. Two hundred and thirty-four parents/caregivers responded to the survey, with 83.5% of the parents being mothers, 10% fathers, 6% grandparents and 2% legal guardian. The majority of parents (69%) reported their child had a high-incidence disability and 31% reported their child had a low-incidence disability. Nearly three-quarters (70%) of students that were reported on by parents were males. Almost all (96%) parents/caregivers agreed that their children should be active participants in the IEP meetings and 98% of parents believed that their children should be taught self-determination skills in

school. Parents' beliefs regarding the importance of teaching self-determination was not influenced by their child's disability status (i.e., high- vs. low-incidence).

Present Study

In the present study, self-determination in adolescents and adults with FXS was investigated through self-report and parent-report to contribute to the movement toward increased health autonomy to address healthcare disparities. Due to increased understanding of the molecular basis of FXS, clinical drug trials continue to become increasingly prevalent for individuals with FXS. Understanding self-determination in the FXS population will contribute to future efforts to determine the ability of individuals with FXS to engage in informed consent to research (i.e., decisional capacity). The primary aims of this study include: (1) examining the psychometric proprieties of self- and parent-report measures of self-determination in FXS; (2) identifying strengths and weaknesses in self-determination of adolescents and adults with FXS; (3) understanding how parent's perceive self-determination in their children with FXS;(4) investigating the relationship between self-reported and parent-reported self-determination; and (5) examining the impact of personal characteristics relevant in the FXS population, including autism and anxiety. Each of these five aims is described below.

The first aim of this study is to examine the psychometric properties of two self-determination measures. This initial aim is required due to the nature of measures currently available to assess self-determination. First, because there are no standardized parent-report measures of self-determination, the psychometric properties of the parent-report measure used in this study were evaluated. Second, as the self-report measure has been typically used with individuals less severely affected by intellectual disability and other developmental disabilities, the validity of using the Arc's Self-determination Scale for individuals with FXS was examined.

Therefore, aim 1 is to support subsequent analyses using the self- and parent-report measures of self-determination.

Aim 2 of the present study allowed the characterization of self-determination in adolescents and adults with FXS through self- and parent-report. Despite previous efforts to document self-determination in individuals with I/DD, there is a paucity of research elucidating the characteristics of self-determination in individuals with comorbid developmental disabilities and moderate to severe intellectual disability), such as those with FXS. Despite the significant role parents serve in the lives of individuals with ID in terms of opportunities to practice and act in a self-determined manner, there is also a paucity of research examining parent perspectives of self-determination. However, understanding parent perceptions of self-determination is an important and necessary step to furthering the development of self-determination in individuals with developmental disabilities. Due to increased understanding of the molecular basis of FXS, clinical drug trials continue to become increasingly prevalent for individuals with FXS. Understanding self-determination in the FXS population will contribute to future efforts to determine the ability of individuals with FXS to engage in informed consent to research (i.e., decisional capacity).

Aim 3 serves to further examine the role of individual characteristics on self-reported and parent-reported self-determination. Although research suggests that intellectual disability is moderately correlated with self-determination, intellectual functioning does not necessarily predict self-determination (Wehmeyer & Garner, 2003). There is a dearth of research investigating how individual characteristics (e.g., autism spectrum disorder and anxiety) affect self-determination. It is important to understand how different factors influence overall self-determination in order to understand if self-determination plays a role in decisional capacity.

Aim 4 of this study addresses a gap in the self-determination literature by investigating the relationship between self- and parent-reported behaviors of self-determination in adolescents and adults with I/DD. Currently no known studies have compared parent assessments of their children's self-determination with their adolescent and adult children's self-reported self-determination. An investigation of the relationship between self- and parent- report will contribute to understanding whether self-report is consistent with parent-report.

Finally, aim 5 of this study serves to support and further illustrate the findings of the previous aims by describing what factors parents perceive to be significant barriers to the development of self-determination in their children as well as factors that support the development of self-determination. By identifying themes in parent responses to open-ended questions, aim 5 will inform the interpretation of quantitative findings from aims 2-4.

Research Aims and Questions

Aim 1: Examine the psychometric properties of the parent-report and self-report measures of self-determination.

Questions:

1. Is the original four factor model of the *Arc's SDS* valid for use with individuals with FXS?
2. What factors underlie the items of the parent-report *Self-Determination Parent Questionnaire*?

Aim 2: Describe the characteristics of self-determination in adolescent and adult males and females ages 12-40 with FXS on the self-report *Arc's Self-determination Scale* and the parent-report *Self-Determination Questionnaire*.

Questions:

1. What are the shape, range, mean, and variance of scores on each of the four subdomains (i.e., autonomy, self-regulation, psychological empowerment, and self-realization) and total self-determination score of the *Arc's Self-determination scale* for male and female adolescents and adults with FXS?
2. What are the shape, range, mean, and variance of parent-reported self-determined behaviors and ratings of importance of self-determination in male and female adolescent and adult children with FXS on the SDPQ?
3. What are the correlations between individual characteristics (i.e., ASD, anxiety, intellectual functioning, adaptive behavior, and age) and self-reported self-determination on the Arc's SDS in males and females with FXS separately?
4. What is the correlation between each of the following individual characteristics (i.e., ASD, anxiety, intellectual functioning, adaptive behavior, and age) and parent-reported self-determination and ratings of importance of self-determination in males and females with FXS on the SDPQ?

Aim 3: Examine predictors of self-determination in individuals with FXS, including autism spectrum disorder (ASD), anxiety, intellectual functioning, adaptive behavior, and age to determine how these characteristics influence self-reported and parent-reported self-determination in male and female adolescents and adults with FXS.

Questions:

1. To what extent does individual ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender predict self-reported self-determination subdomain scores (autonomy, self-regulation, psychological empowerment, and self-realization) in individuals with FXS?

2. To what extent does individual ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender predict parent-reported self-determined behaviors in their children with FXS?
3. To what extent does individual ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender predict parent-reported ratings of importance of self-determination?

Aim 4: Determine the relationship between self-reported self-determination and parent-reported behaviors and importance of self-determination in adolescents and adults with FXS.

Questions:

1. To what extent do individual symptoms of ASD, symptoms of anxiety, intellectual functioning, adaptive behavior, age, and gender moderate the relationship between self-reported and parent-reported self-determination?
2. To what extent do individual ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender moderate the relationship between self-reported self-determination and parent-reported ratings of importance of self-determination?

Aim 5: Identify themes among parent-reported supports and barriers they believe contribute most to the development of self-determination in their adolescent and adult children with FXS to better inform the interpretation of the quantitative findings from aims 1-4.

Questions:

1. What themes arise in parent responses to open-ended questions about supports and barriers of the development of self-determination in their adolescent and adult children with FXS?

CHAPTER 3: METHODOLOGY

Participants

The present study included 86 individuals recruited for a larger study, the *Decisional Capacity and Informed Consent in FXS*, a research study that has been approved by the University of North Carolina at Chapel Hill's Institutional Review Board (IRB)[1]. 56 males (22 adolescents 12-18 years old and 34 adults between 19-40 years old) and 30 females (12 adolescents and 18 adults) were included in analyses (see Table 5). A purposeful sampling method was implemented because individuals were required to meet specific criteria to be included in the study (Merriam, 2009). The project recruited participants based on documentation of full-mutation FXS, age, gender, and location. All individuals have documented full-mutation FXS and have completed the full assessment battery. All participants also had a parent or caregiver complete rating scales and questionnaires regarding their child's functioning (e.g., adaptive behavior).

A power analysis was conducted to ensure that 85 participants was a large enough sample size to detect any effects that result from inclusion of six independent variables in a multiple regression analysis. G*Power 3.1 (Faul, Erdfelder, Lang, & Buchner, 2007) was used to assess power with an alpha level set at .05, the inclusion of 86 cases, and six independent variables. The power was calculated at .80 to detect an overall R^2 of .20 or more on a multiple regression analysis with a sample size of 70.

[1] Additional information can be found under the Procedures section of this chapter.

Measures

Cognitive functioning. The *Stanford Binet Intelligence Scales-5th edition* (SB5; Roid, 2003) was used to measure cognitive functioning. The SB5 provides scores for verbal and nonverbal ability across five domains: Fluid Reasoning, Knowledge, Quantitative Reasoning, Visual-Spatial Reasoning, and Working Memory. A full scale IQ is a composite score from the verbal and nonverbal subtests. The SB5 (Roid, 2003) has high reliability in the nonverbal reasoning domain, the verbal reasoning domain, and the full scale IQ score. Internal consistency ranges from .95 to .98 across all age groups. Reliability for the factor indexes (e.g., Fluid Reasoning) range from .90 to .92. Subtest reliability ranges from .84 to .89. Concurrent and criterion validity are also well documented using the SB-4 (Thorndike, Hagen & Sattler, 1986), the SB-form L-M (Terman & Merrill, 1960), the Woodcock-Johnson III Tests of Cognitive Ability (Woodcock, McGrew, & Mather, 2001), the Universal Nonverbal Intelligence Test (Bracken & McCallum, 1998), the WAIS-III (Wechsler, 1997), the WISC-III (Wechsler, 1991), and the WPPSI-R (Wechsler, 1989).

The floor effect is a statistical phenomenon that occurs when data points fall in the lowest range of possible values on a given measure and is a known limitation to many standardized IQ tests, particularly for individuals with severe intellectual disability. A floor effect often results in skewed distribution with limited variability because a given target population obtains scores that are in the lowest possible values or potentially lower than the normative sample (Jackson, 2011). Standardized IQ tests, including the SB5, have documented limited range and precision for individuals with such ID, including individuals with FXS (Sansone et al., 2014). As a result, Sansone et al. (2014) developed and validated a method to conduct a raw z-score transformation based on the norm sample.

Based on the findings from Sansone et al. (2014), each of the indices subtest standard scores that comprise the NVIQ were transformed to z-scores and then rescaled with a mean of 100 and standard deviation of 15 in order to be comparable to original standard scores.

To obtain a transformed index score each subtest raw score was first, rescaled to a z-score, resulting in a deviation score. To convert each participant's raw score to a normalized z-score for each subtest, the mean raw score from the standardization sample by age band raw score and subtest was subtracted from the participant's raw score and divided by the standard deviation of the norm sample by age band and subtest ($z_{ij} = (r_{ij} - \mu_{ij})/ \sigma_j$). Next, the deviation score for each subtest was converted from a mean of 0 and a standard deviation of 1 to a scale consistent with the original subtest standardized scores (i.e., with a mean of 10 and standard deviation of 3). Finally, the mean of the deviation scores of the nonverbal reasoning index (i.e., all five subtests) was calculated and rescaled with a mean of 100 and a standard deviation of 15, which is analogous to the subtest standardized score combination that is used to generate the index score. The same procedure was used for the verbal reasoning index and the full-scale IQ.

Adaptive Behavior. The *Scales of Independent Behavior, Revised* (SIB-R; Bruininks, Woodcock, Weatherman, & Hill, 1996) is a norm-referenced parent-report measure used to assess adaptive behavior and maladaptive behavior. The SIB-R includes 14 areas of adaptive behavior and also assesses internalized, externalized, and asocial maladaptive behavior. The adaptive behavior items include a 4-point Likert scale to assess gross motor, fine motor, social interaction, language, comprehension, language expression, eating and meal preparation, toileting, dressing, personal self-care, domestic skills, time and punctuality, money and value, home/community, and orientation. Each item is written in precise behavioral statements.

The SIB-R Adaptive Behavior composite is comprised of several subdomains, including motor, social communication, personal living, and community living. The motor subdomain consisted of items related to gross and fine motor skills. The social communication subdomain includes receptive and expressive language and reading and writing. Personal living skills include skills related to personal hygiene and safety awareness.

The SIB-R is a norm-referenced measure and has extensive measures of reliability and validity. Reliability measures include documented internal consistency, test-retest reliability, and inter-rater reliability. Validity is also documented for the adaptive and maladaptive behavior scales.

Autism spectrum disorder. The *Autism Diagnostic Observation System, Second Edition* (ADOS-2; Lord et al., 2012) is a semi-structured assessment used to assess symptoms of autism spectrum disorder. The ADOS-2 provides tasks that elicit opportunities for individuals to engage in social communication and reciprocity. Symptoms of repetitive behavior and restricted interests are also observed during ADOS-2 assessment. Behavioral observation is then coded and entered into the ADOS-2 algorithm, which provides a total score that illustrates symptom severity. Research reliability across examiners was obtained.

The ADOS-2 (Lord et al., 2012) has well documented reliability and validity. Interrater reliability for domain scores (e.g., Social Affect) were calculated using correlations. The social affect domain had a correlation of .98, the restricted and repetitive behaviors had a correlation of .80, and the overall total had a correlation of .96 on Module 2. The social affect domain had a correlation of .92, the restricted and repetitive behaviors had a correlation of .91, and the overall total had a correlation of .94 on Module 3. The social interaction domain had a correlation of .93, the communication domain had a correlation of .84, the social interaction and

communication domain had a correlation of .92, and the restricted and repetitive behaviors had a correlation of .82 on Module 2. Test-retest reliability was also assessed using intraclass correlations and demonstrated good stability. The correlations for Module 2 were .84 for the social affect domain, .73 for the restricted and repetitive behaviors, and .83 for the overall total domain. The correlations for Module 3 were .81 for the social affect domain, .82 for the restricted and repetitive behaviors, and .87 for the overall total domain. Test-retest validity was not available for Module 4. Predictive validity, which is determined by the extent to which the ADOS-2 algorithm for each module accurately identifies individuals on the spectrum, is strong. Sensitivity for module 2 is 98% and specificity is 90% when distinguishing between autism and non-spectrum individuals. On the module 3, sensitivity is 91% and specificity is 84% for autism and non-spectrum individuals. Module 4's sensitivity and specificity are each 93% when distinguishing between ASD and non-spectrum individuals.

The *Social Communication Questionnaire Liftetime Form (SCQ;* Rutter, Bailey, & Lord, 2003) is a parent-report questionnaire used to assess developmental history of social communication impairments. The SCQ consists of 40 yes or no items and has a cut off score of 15. Scores greater than 15 suggest symptoms consistent with ASD. The SCQ is based on the DSM-IV (APA, 2000) and the content of the *Autism Diagnostic Interview-Revised* (Lord, Rutter, & LeCouteur, 1994). The measure is commonly used for screening, progress of symptoms, and research. The SCQ has internal consistency that ranges from .81 to .93 (Rutter, Bailey, & Lord, 2003), has been shown to differentiate children with ASD from children with other disabilities, and has a high correlation with the ADI-R (i.e., average agreement between items is 70.8%).

Best practice for a diagnosis of autism spectrum disorder includes consideration of developmental history using parent report as well as current functioning through behavior

observation. In the present study, the SCQ is used as a measure of developmental history based on parent report and the ADOS-2, the gold standard assessment of ASD, is used to assess symptoms related to ASD. Diagnostic determination of participants in the present study consisted of those who met criteria for ASD on both the SCQ and the ADOS-2.
Autism spectrum disorder for each participant was determined based on whether they met criteria for ASD on both the ADOS-2 (Lord et al., 2012) and the SCQ (Rutter, Bailey, & Lord, 2003).

Anxiety. Anxiety is measured through a parent-reported rating scale. The *Anxiety, Depression, and Mood Scale (ADAMS;* Esbensen, Rojahn, Aman, & Ruedrick, 2003) consists of 28 items and serves as a screener for psychiatric disorders in individuals with ID. The ADAMS General Anxiety scale and Social Avoidance scale was used to assess overall levels of anxiety in each participant. Each item is scored on a 4-point Likert scale (0 = "not a problem" to 3 = "severe problem"). The scale's psychometric properties were evaluated and normed with 265 individuals with ID and also validated with a total of 129 psychiatric patients with ID (Esbensen, Rojahn, Aman, & Ruedrick, 2003).

The General Anxiety scale consisted of seven items, including ratings on each participant as nervous, tense, worried, and anxious. The remaining three items asked parents to report if their child does not relax or settle down, experiences panic attacks, and trembles when frightening situations are not present. The Social Avoidance scale also consisted of seven items related to social avoidance, including communication, withdrawn and shy behavior, avoidance of others, a lack of facial expressions, avoidance of eye contact, and avoidance of peers.

The development of the ADAMS (Esbensen, Rojahn, Aman, & Ruedrick, 2003) scale included a measure of reliability and validity, as well as normative information by age. Retest

reliability was calculated using intraclass correlation coefficients. The full scale's reliability was .81 and fell in the descriptive category of excellent (Cicchetti, 1994). The General Anxiety subscale's retest reliability was .78 and the social avoidance retest reliability was .83. Item retest reliability ranged between .45 and .78. Interrater reliability was also calculated using intraclass correlation coefficients with a one-way random effects model. The full scale had a .48 interrater reliability, which falls in the fair range (Cicchetti, 1994). The General Anxiety subscale and the Social Avoidance subscale had interrater reliabilities of .39 and .61, respectively. Internal consistency was measured using Cronbach's alpha to assess each item's correlation with the remaining items of the particular subscale. Internal consistency on the General Anxiety subscale was .83 and .80 on the Social Avoidance subscale, which are within the acceptable range set for research (Nunnally, 1967). The General Anxiety subscale has an interfactor correlation with the Social Avoidance subscale (r=.514). Normative information was also made available by age and intellectual disability. Individuals between the ages of 10 and 29 years had a General Anxiety subscale mean score of 5.73 (SD= 4.83) and mean score of 6.52 (SD = 4.76) on the Social Avoidance subscale. Individuals between the ages of 30 and 39 years had a General Anxiety subscale mean score of 5.17 (SD = 4.93) and a mean score of 5.90 (SD = 4.41) on the Social Avoidance subscale.

Self-Determination. Self-determination was measured by self-report and parent-report. The Arc's Self-Determination Scale, Adolescent and Adult Versions (Wehmeyer & Kelchner, 1995) were used to assess self-reported self-determination. Participants were administered the Adolescent Version if they were still in school at the time of assessment. The Arc's SDS Adolescent Version was normed with 500 adolescents with cognitive disabilities (Wehmeyer, 1996). The Adult Version is currently being normed (Wehmeyer, in press). The Arc's SDS was

chosen as a measure of self-determination because the tool can be used to assess global self-determination by capturing attitudes and beliefs as well as skills and behaviors related to self-determination. The Arc's SDS provides a total self-determination score and subscale scores for the four domains; Autonomy, Self-Regulation, Psychological Empowerment, and Self-Realization. Across 72 items, a total of 148 points are possible. Higher scores are indicative of higher levels of self-determination.

Concurrent criterion-related validity was documented by using three conceptually-related measures, the Nowicki-Strickland Internal-External Scale (ANS-IE; Nowicki & Duke, 1974), the Intellectual Achievement Responsibility Questionnaire (IARQ; Crandall, Katkovsky, and Crandall, 1965), and the Self-Efficacy Scale (SES; Sherer, Maddux, Mercandante, Prentice-Dunn, Jacobs, & Rogers, 1982). Correlations among the SDS domains and the three measures ranged between .25 and .5, indicating moderate to strong relationships. Construct validity was documented through discriminative validity and factorial validity. The Arc's SDS adequately differentiated between students with cognitive disabilities and those without disabilities (Wehmeyer, 1995). Factor analysis indicated that the four scales accurately reflect the constructs that they were used to measure. Internal consistency reliability for the complete scale, excluding the self-regulation domain, was .90. Cronbach's alpha for the autonomy domain was .90, .73 for the psychological empowerment domain, and .62 for the self-realization domain.

The *Self-Determination Questionnaire* (SDPQ) is used to assess parent-reported self-determination. This is a nonstandardized measure developed by Carter et al. (2013) as a questionnaire for parents that paralleled a survey used previously in previous studies of teachers (Stang et al., 2009), special educators (Carter et al., 2008), and paraprofessionals (Carter, Lane, & Sisco, 2011). Currently, standardized parent-report measures of self-determination are not

available. The SDPQ covers seven core observable skills related to self-determination, four broad areas of self-determination (autonomy, self-regulation, psychological empowerment, and self-realization), and seven statements regarding how their children have their wants and needs met. The four broad areas of self-determination were added to the otherwise original questionnaire format for the purpose of investigating overall parent attitudes about different components of self-determination. For each of these three sections, parents were asked to rate how well their child performs a given skills, acts according to the four areas of self-determination, and has their wants and needs met. The three-point Likert scale scored "0" for not well, "1" for somewhat well, and "2" for very well. Parents were also asked to rate all of the items within these three sections by level of importance. The three-point Likert scale scored "0" for not important, "1" for somewhat important, and "2" for very important. Finally, the SDPQ included two open-ended questions, which asked parents to list two ways she or he encourages self-determined behavior in their child and two factors that she or he considers being the most significant barriers to the development of self-determination in their child. Adequate validity and reliability for the Self-Determination Parent Questionnaire is documented in the literature (Carter, Lane, Peirson, & Glaeser, 2006; Carter et al., 2009). In the most recent study using the measure, Cronbach's alpha was .87 (Carter et al., 2013).

Demographics. Demographic data were obtained through a demographic survey form completed by the participant's primary caregiver. Demographics data that were collected includes race/ethnicity, caregiver education level, household income, and forms of public assistance and resources received (e.g., Medicaid, food stamps, supplemental security income, welfare/public assistance, and respite care).

Procedures

Data Source. The *Decisional Capacity and Informed Consent in Fragile X Syndrome* is a research study that began in April, 2013 and will continue through June, 2016. The project is supported through funding by the Research Triangle Institute (RTI International) and the National Institutes of Health. The project is implemented by the Carolina Institute for Developmental Disabilities at the University of North Carolina at Chapel Hill and RTI International. The *Decisional Capacity and Informed Consent in Fragile X Syndrome* research study has been approved by the University of North Carolina at Chapel Hill's Institutional Review Board (IRB) and acceptance of the application is located under Appendix 1).

The Decisional Capacity and Informed Consent in Fragile X Syndrome project has two primary aims: (a) to develop a valid scale to measure decisional capacity and assess the range of decisional capacity in adolescents and adults with FXS and (b) to develop a tablet-based application to aid and enhance decisional capacity and informed consent.

The *Decisional Capacity and Informed Consent in Fragile X Syndrome* study includes a large neurocognitive assessment battery for individuals with FXS and a variety of rating scales and questionnaires for parent-report of their child's functioning. A total of nine standardized measures were administered to individuals with FXS including measures of intellectual functioning, autism, executive function, memory, self-determination, reading comprehension, and oral comprehension. A total of 8 parent-report measures were administered, five of which are standardized. These scales included measures of autism, anxiety, attention-deficit/hyperactivity disorder, adaptive behavior, sensory processing, decision-making history, decisional capacity, and self-determination.

Recruitment. Participants were recruited through three main efforts: (1) identification through the FX research registry maintained at the University of North Carolina at Chapel Hill; (2) follow-up on families who participated in prior studies conducted by the principal investigator, Don Bailey; (3) announcement of the research study through organizations, including the National Fragile X Foundation. After families were recruited, they completed a screening to determine eligibility for the study. Individuals who were very low-functioning (e.g., nonverbal) are ineligible for the study due to the perceived difficulty fully engaging in the assessment battery and potentially engaging in the consent process in a meaningful way in the future. Families who were successfully screened and enrolled in the study were scheduled for a data collection visit at the participant's home or school, based on family preference. Prior to the visit, the participant's primary caregiver received a packet in the mail containing the consent and assent forms and all parent-rating scales, with exception to the adaptive behavior scale, which is completed as an interview with the parent during the evaluation. Assent and consent forms were reviewed and signed at the start of the initial visit. Data collection often took place over two scheduled visits lasting a total of approximately five hours. Participants and primary caregivers were each reimbursed $60 for their time and participation in the study.

Statistical Analysis

The present study employed a mixed methods approach to examine self-determination in adolescents and adults with FXS with both quantitative and qualitative procedures employed. The first four aims were designed to explore the factors underlying two measures of self-determination (i.e., self- and parent-report), examine the relationships between individual characteristics (e.g., anxiety symptoms) and self- and parent-reported self-determination, and identify the relationship between self- and parent-report. Using a qualitative approach, this study

also explored factors that parents perceived as the greatest barriers and supports to the development of self-determination in their aging children with FXS. This descriptive analysis of qualitative data provided enhanced understanding and breadth of knowledge of the quantitative results (Creswell & Plano Clark, 2011).

Data were analyzed using IBM SPSS software (version 22.0). Independent variables included the following: (1) symptoms of autism spectrum disorder was calculated from the ADOS-2 algorithm; (2) intellectual functioning was measured using the FSIQ score on the SB5; (3) symptoms of anxiety was measured using the total score from the ADAMS general anxiety scale; (4) adaptive behavior was measured by the SIB-R total score. Self-determination self-report and parent-report will primarily serve as dependent variables. All variables were analyzed as continuous variables, with the exception of gender. A table of constructs, measures used, and variables is presented in Table 2. Statistical analysis and reasoning for each of the questions listed under the four aims of this proposal are detailed below. Any subsequent analyses were directed by initial findings.

Descriptive analysis of two open-ended questions on the parent-report SDPQ was conducted to identify the frequency of various themes among parent perceptions of supports and barriers to self-determination. First, two raters independently reviewed parent responses on the SDPQ. The second stage of the analysis consisted of defining themes. Third, the data were coded using the identified themes and analyzed for inter-rater reliability. Consensus scoring was used to resolve discrepancies. Some themes were further divided into subthemes. Detailed information on consensus scoring and themes of supports and barriers are provided in their respective subsections below.

CHAPTER 4: RESULTS

Descriptive data

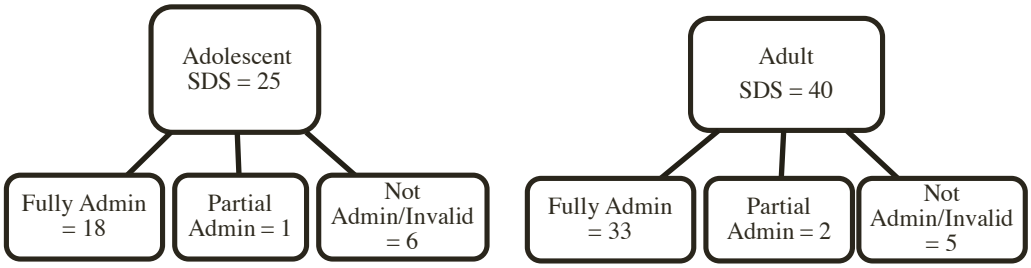
Both parent-report and self-report measures of self-determination were completed for the majority of participants (76%; $n = 65$). Twenty participants have only parent-report data and one participant received only the self-report form. The self-determination self- and parent-report measures were added to the Decisional Capacity project protocol approximately six months after the larger study's initiation of active data collection. Because parent-report measures could be collected retroactively (i.e., after collecting participant data at the home visit), whereas the self-report measure required direct administration, there are more parent-report measures in the present study.

Missing items. Subscale scores were calculated in instances where at least 80% of the items were completed. Based on general guidelines for principal component analyses, 80% of the data for a given subscale was deemed sufficient for a valid subscale score. A subscale score was not calculated for participants with less than 80% of the items completed. For instance, for the Self-Determined Behavior subscale on the *Self-Determination Parent Questionnaire* (SDPQ; Carter et al., 2013) participants needed 11 of 13 items for a subscale score to be calculated. For the Importance of Self-Determination subscale of the SDPQ, six out of seven items were needed for the subscale to be calculated. One participant's SD behaviors subscale scores on the SDPQ were not calculated and three participants' Importance of SD subscale scores were not calculated due to missing data.

Of the 65 participants in the present study who completed the *Arc's Self-Determination*

Scale (SDS; Wehmeyer & Kelchner, 1995) self-report form, approximately two-thirds completed the adult form (n = 40) and one-third completed the Adolescent form (n= 25). Several participants were either not administered the self-report form based on the examiner's judgment of the participant's ability to reliably provide self-report data, or the examiner identified their self-report as invalid post- administration. Of the 65 self-report participants, six adolescents (24%) were excluded, and five adults (13%) were excluded. Individuals who were not administered the form or whose form was identified as invalid were combined to form a separate subgroup (n = 11). Additionally, there were a few participants (i.e., 1 participant on the Adolescent form and 2 participants on the Adult forms) with partial completion of the SRS due to inability to complete more demanding subsections (e.g., Self-Regulation domain). Examiners reported various factors such as perceived difficulty with expressive language or working memory deficits as reasons for not administering the full SDS. Individuals who have partial data were included in all analyses, but did not have total scores due to missing section scores. Lastly, four participants who were administered the Adolescent form had a missing total score because they earned a raw score of 0 on the Self-Regulation domain (See Figure 1 for flow chart).

Figure 1. SDS administration flow chart



The 11 individuals who were identified as not able to reliably complete the Arc's SDS differed significantly in several ways from the sample who completed the self-report form.

Participants who did not reliably complete the SDS were younger, had lower nonverbal IQ, and fewer daily living skills than participants who did complete the self-report measure (Table 1). Participants who did not reliability complete the SDS form were also more likely to meet criteria for ASD, and more likely to be male (Table 2). Participants did not differ in verbal IQ, full scale IQ, or parent-ratings of generalized anxiety, and social avoidance (Table 1).

Table 1. T-tests and descriptive statistics for SDS Administered and Not Administered Groups

	SDS Administered		SDS Not Administered			
	n	*M(SD)*	*n*	*M(SD)*	*t*	*df*
Age	54	21.74(7.37)	11	17.45(2.34)	3.50**	52
NVIQ	54	62.35(19.59)	11	37.27(8.93)	6.61**	33
VIQ	54	58.73(21.92)	11	67.55(28.78)	-1.15	61
FSIQ	52	58.15(19.35)	11	66.91(28.49)	-.97	12
Adaptive Behavior	53	59.83(19.35)	8	25.25(14.70)	3.24**	59
Generalized Anxiety	54	5.69(4.06)	11	6.09(4.68)	-.30	63
Social Avoidance	54	5.59(4.85)	11	7.18(3.76)	-1.02	63

*$p < .05$, two-tailed
**$p < .01$, two-tailed

Table 2. Chi-Square and descriptive statistics for SDS Administered and SDS Not Administered Groups

	SDS Administration			
	Administered	Not Administered	χ^2	*p*
Gender			6.75	.00
Males	28	10		
Females	26	1		
ASD Status			5.75	.02
ASD	12	6		
No ASD	41	4		

Demographic data. Families were asked to provide demographic information, including race/ethnicity, household income, caregiver education level, and number of resources and public assistance the family received (e.g., Medicaid, food stamps, supplemental security income, welfare/public assistance, and respite care; Table 3). Roughly one-quarter (24%; $n = 21$) of families had more than one child in the present study. The majority of individuals were Caucasian (84%). Only 11% identified as non-white, which included four individuals who

identified as African American, 2 who identified as Hispanic, 2 who identified as Asian / Pacific Islander, and 2 who identified as Other (e.g., biracial). Of families who reported a household income (63%; n = 54), most (55%) reported an income of $65,000 or higher. A total of 8% reported an income under $64,999 and 37% did not report a household income. However, there was more reported variability in the number of public assistance and resources families received. A total of 36% reported receiving zero resources, 40% reported between one and three resources, and 19% reported between four and six resources. A total of 6% did not report whether they received public assistance or resources. The majority of parents were well educated, with 35% who reported a college degree, 21% who reported a graduate degree, 26% who reported some college or an associate's degree, and just 6% who reported a high school degree. A total of 13% of caregivers did not provide education information.

Child characteristics. Descriptive statistics for child characteristics (e.g., age, FSIQ) are listed in Tables 4 and 5 for the parent-report sample and the self-report sample, respectively. The mean age for the parent-report sample was 20.76 years for the entire sample, 20.73 for males, and 20.83 for females. The average converted FSIQ for the entire sample was 56.11. Males in the current sample did not have significantly lower converted FSIQ scores than females (Table 6). However, males had significantly lower NVIQ scores, and the Shapiro-Wilk test of normality suggested that normality could be assumed for FSIQ.

Table 3. Demographics for Total Sample

	Total (n = 86)	%	Male (n=56)	%	Female (n=30)	%
Age						
12-17 years	34	40%	22	39%	12	14%
18-40 years	52	60%	34	61%	18	22%
Race/Ethnicity						
American Indian/Alaska Native	0	0%	0	0%	0	0%
Asian	1	1%	1	2%	0	0%
Native Hawaiian Other Pacific Islander	1	1%	1	2%	0	0%
Black or African American	4	5%	3	5%	1	3%
Caucasian	72	84%	45	80%	27	90%
Hispanic or Latino	2	2%	2	4%	0	0%
Other	2	2%	1	2%	1	3%
Missing	4	5%	3	5%	1	3%
Household Income						
Less than $35,000	3	4%	3	5%	0	3%
$35,000 to $44,999	2	2%	1	2%	1	0%
$45,000 to $54,999	1	1%	1	2%	0	0%
$55,000 to $64,999	1	1%	1	2%	0	0%
$65,000 or more	47	55%	32	57%	15	50%
Missing	32	37%	18	32%	14	47%
Caregiver Education Level						
Less than High School	0	0%	0	0%	0	0%
High School Graduate	5	6%	4	7%	1	3%
Additional Training / Some College	18	21%	11	20%	7	23%
Associates Degree	4	5%	4	7%	0	0%
College Degree/Some Post College	30	35%	18	32%	12	40%
Graduate Degree	18	21%	13	23%	5	17%
Missing	11	13%	6	11%	5	17%
Public Assistance/Resources						
0 reported assistance/resources	31%	36%	14	25%	17	57%
1-3 reported assistance/resources	34	40%	24	43%	10	33%
4-6 reported assistance/resources	16	19%	14	25%	2	7%
≥ 7 reported assistance/resources	0	0%	0	0%	0	0%
Missing	5	6%	4	7%	1	3%

Table 4. Parent-report sample demographics

	Parent-report Sample											
	Total Sample (N = 85)				Males (N = 55)				Females (N=30)			
	N	*M*	*SD*	Range	N	*M*	*SD*	Range	N	*M*	*SD*	Range
Age	85	20.76	6.77	12-39	55	20.73	6.74	12-38	30	20.83	6.94	12-39
FSIQ	82	56.11	20.89	16-105	55	53.80	23.00	16-105	27	60.81	15.06	27-88
NVIQ	83	55.95	20.67	16-102	54	46.22	15.56	16-93	29	74.07	16.46	31-102
VIQ	83	56.95	23.04	11-111	55	54.27	24.77	11-111	28	62.21	18.51	15-89
Adaptive Behavior	79	52.47	30.77	1-141	50	37.68	21.88	1-113	29	77.97	27.14	8-141
Generalized Anxiety	85	6.24	4.18	0-16	55	6.51	4.22	0-16	30	5.73	4.13	0-15
Social Avoidance	85	5.85	4.40	0-18	55	6.16	4.22	0-18	30	5.27	4.74	0-16
SD Behaviors	84	14.80	7.37	1-38	54	12.65	5.47	2-30	30	18.67	8.76	1-38
Importance of SD	82	11.38	2.95	2-14	54	11.07	3.12	2-14	28	11.96	2.53	6-14

Table 5. Self-report sample demographics

Self-report Sample												
	Total Sample (N = 65)				Males (N = 38)				Females (N=27)			
	N	*M*	*SD*	Range	N	*M*	*SD*	Range	N	*M*	*SD*	Range
Age	65	21.02	6.96	12-39	38	20.92	6.86	12-38	27	21.15	7.24	12-39
FSIQ	63	59.68	21.22	16-105	38	59.39	24.62	16-105	25	60.12	15.10	27-88
NVIQ	65	58.11	20.50	24-102	38	46.37	13.84	24-76	27	74.63	16.64	31-102
VIQ	63	60.27	23.24	11-111	38	60.24	26.27	11-111	25	60.32	18.22	15-89
Adaptive Behavior	61	55.30	30.31	1-141	35	37.31	20.80	1-85	26	79.50	23.54	31-141
Generalized Anxiety	65	5.75	4.14	0-16	38	6.16	4.35	0-16	27	5.19	3.82	0-15
Social Avoidance	65	5.86	4.70	0-16	38	6.21	4.53	0-18	27	5.37	4.96	0-16
Autonomy	54	54.30	21.15	17-96	28	49.21	19.84	17-84	26	59.77	21.51	22-96
Self-Regulation	51	5.35	4.92	0-21	25	2.32	2.19	0-7	26	8.27	5.07	1-21
Psychological Empowerment	54	11.56	2.92	5-16	28	9.64	2.38	5-14	26	13.62	1.86	10-16
Self-Realization	54	10.52	2.87	4-15	28	8.79	2.71	4-12	26	12.39	1.60	9-15
SD Total Score	54	81.43	26.29	28-142	28	69.68	22.20	28-107	26	94.08	24.73	46-142

Table 6. Descriptive statistics for child characteristics

	Gender t-test		
	t	*df*	*p*
FSIQ	-1.65	73	.10
Adaptive Behavior	-7.21	77	.00
Generalized Anxiety	.817	83	.41
Social Avoidance	.897	83	.39

The average adaptive behavior composite score was 52.47. Males had an overall lower adaptive behavior composite than females (Table 6). Table 7 lists scores for adaptive behavior subdomains (e.g., personal living) on the SIB-R for males, females, and the sample as a whole. The distributional shape of adaptive behavior scores was examined to determine the extent to which assumptions of normality were met. Based on skewness, kurtosis, and the Shapiro-Wilk test of normality, normality was assumed.

Table 7. Descriptive statistics for Adaptive Behavior SIB-R subdomains

	Males (n = 56)		Females (n = 30)		Sample (N = 86)	
	M(SD)	Range	*M(SD)*	Range	*M(SD)*	Range
Motor Skills	54.51(25.37)	19-144	81.79(26.49)	27-145	64.40(28.81)	19-145
Social Communication	50.07(20.06)	12-104	84.20(24.42)	12-128	62.26(27.13)	12-128
Personal Living	55.40(22.38)	7-130	89.40(21.78)	26-130	67.40(27.44)	7-130
Community Living	35.75(25.97)	1-132	76.17(25.73)	28-131	52.22(31.81)	1-132

Scores on the ADAMS parent-report measure of Generalized Anxiety and Social Avoidance subscales (21 total points possible in each subscale) were similar between males and females. The average score on the Generalized Anxiety subscale was 6.24 for the entire sample, 6.51 for males, and 5.73 for females (Table 6). Scores on the Social Avoidance subscale were slightly lower for the sample. The distributional shape of Generalized Anxiety subscale scores was examined to determine the extent to which assumptions of normality were met. As a whole, skewness, kurtosis, and the Shapiro-Wilk test of normality suggested that normality could not be assumed. Similarly, normality could not be assumed for the Social Avoidance subscale.

A total of 22 males (39%) and 2 (7%) of females met criteria for ASD (based on scores on the ADOS-2 and SCQ). As a whole, a little over one quarter (28%) of the entire sample met criteria for ASD, which is consistent with prevalence estimates of ASD in FXS (Hernandez et al., 2008). Significantly more males met criteria for ASD than females (Table 8).

Table 8. Autism spectrum disorder status

	Males		Females		Sample		Chi-square		
	f	%	*f*	%	*f*	%	χ^2	*df*	*p*
Does not meet criteria for ASD	31	55.4%	27	90.0%	58	67.4%			
Meets criteria for ASD	22	39.3%	2	6.7%	24	27.9%			
							10.84	1	.00

Note: ASD determined by meeting cutoff scores on ADOS-2 algorithm and SCQ

Rationale for raw score data analyses

Using two different versions (Adolescent and Adult formats) of the SDS self-report measure presented with challenges for combined data analyses. To examine the present study

sample as a whole while using two versions of the Arc's SDS (i.e., Adult and Adolescent forms) with different norm samples and different types of standard scores, percentile scores or raw score data could be used. The norm samples from the SDS Adolescent and Adult formats were compared to determine if raw scores or percentile scores were more appropriate for subsequent analyses.

The norm samples of the *Arc's Self-Determination Scale* Adolescent and Adult Forms were compared to the present study samples of adolescents and adults with FXS to inform subsequent analyses. The two samples were comparable based on gender and race/ethnicity (Table 9; Figure 2). The norm sample on the SDS for adolescents was 44.6% male, 42% female, and 13.4% gender unknown. The present study sample of adolescents was 56% male and 44% female. The norm sample on the SDS for adults was 54.6% male and 45.5% female. The present study sample for adults was 57.5% male and 42.5% female. The norm sample and the present study sample primarily identified as Caucasian, with 56.8% of adolescents (22.5% identified as African-American) and 91.8% of adults in the norm sample. In the present study sample, 80% of adolescents and 87.5% of adults identified as Caucasian.

There were differences between the norm sample and the present study sample based on age for adolescents and adults (Table 10). Figure 3 illustrates that in comparison to the norm sample, the present study was younger in age, with most individuals being 15 years old, in comparison to 17 years old. Figure 4 shows a similar pattern for adults, the present study sample is younger than the norm sample. Most adults in the present study were between the ages of 18 years and 29 years, whereas most adults in the norm sample were between the ages of 41 and 60 years old.

Table 9. Comparison of norm and present study sample demographics

	SDS Norm Sample				Present Study Sample			
	Adolescents		Adults		Adolescents		Adults	
Gender								
Male	223	44.6%	100	54.6%	14	56.0%	23	57.5%
Female	210	42.0%	83	45.5%	11	44.0%	17	42.5%
Gender Unknown	67	13.4%	-	-	-	-	-	-
Race/Ethnicity								
African American	78	22.5%	12	7.1%	2	8.0%	1	2.5%
American Indian	2	0.6%	-	-	0	0.0%	0	0.0%
Asian	6	1.7%	1	0.6%	1	4.0%	0	0.0%
Caucasian	197	56.8%	156	91.8%	20	80.0%	35	87.5%
Hispanic	61	17.6%	1	0.6%	1	4.0%	0	0.0%
Pacific Islander	-	-	-	-	0	0.0%	0	0.0%
Other	3	0.8%	-	-	0	0.0%	2	5.0%
Missing	-	-	-	-	1	4.0%	2	5.0%
Disability Status								
Autism spectrum disorder	2	0.5%	17	10.1%	3	12.0%	4	10.0%
Intellectual disability	128	35.2%	104	61.5%	9	36.0%	20	50.0%
Intellectual disability + ASD	-	-	-	-	5	20.0%	6	15.0%
Emotional disorder	15	4.1%	17	10.1%	-	-	-	-
Learning disability	160	44.0%	15	8.9%	-	-	-	-
Orthopedic impairment	1	0.3%	-	-	-	-	-	-
Other health impairment	6	1.6%	9	5.3%	-	-	-	-
Speech / Hearing impairment	2	0.5%	3	1.8%	-	-	-	-
Traumatic brain injury	-	-	4	2.4%	-	-	-	-
No disability (excluding FXS)	50	13.7%	-	-	6	24.0%	9	22.5%
Missing	-	-	-	-	2	8.0%	1	2.5%

Figure 2. Norm sample and present study sample gender comparison

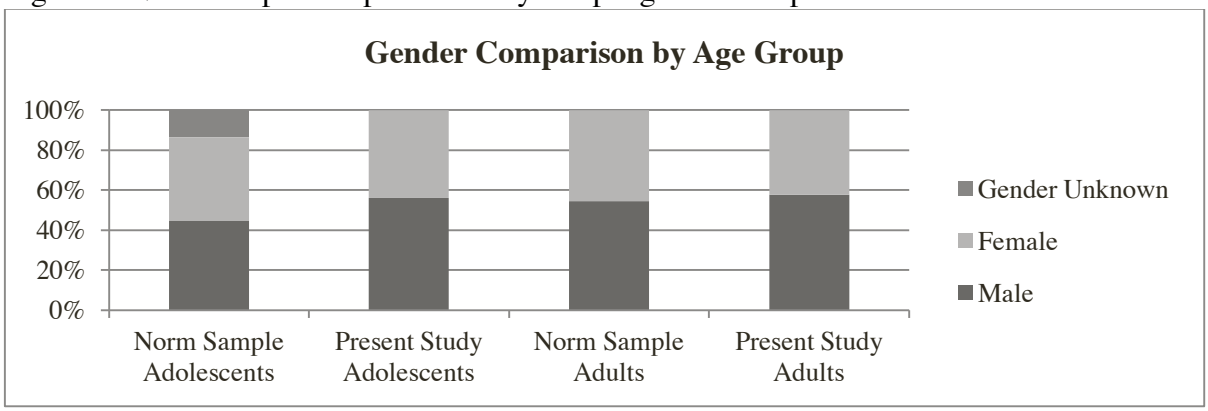


Table 10. Comparison of norm and present study sample age

	SDS Norm Sample				Present Study Sample			
	Adolescents		Adults		Adolescents		Adults	
Age								
12	-	-	-	-	4	16.0%	-	-
13	-	-	-	-	2	8.0%	-	-
14	2	1.1%	-	-	4	16.0%	-	-
15	23	13.0%	-	-	9	36.0%	-	-
16	40	23.0%	-	-	3	12.0%	-	-
17	53	29.7%	2	1.2%	3	12.0%	-	-
18	35	20.0%	5	3%	-	-	5	20.0%
19	8	4.6%	10	6%	-	-	3	12.0%
20	4	2.3%	6	3.6%	-	-	3	12.0%
21	7	4.0%	6	3.6%	-	-	1	4.0%
22	3	1.7%	7	4.2%	-	-	3	12.0%
23	-	-	2	1.2%	-	-	4	16.0%
24	-	-	7	4.2%	-	-	3	12.0%
25	-	-	8	4.8%	-	-	3	12.0%
26	-	-	5	3%	-	-	2	8.0%
27	-	-	5	3%	-	-	0	0.0%
28	-	-	8	4.8%	-	-	2	8.0%
29	-	-	5	3%	-	-	3	12.0%
30	-	-	7	4.2%	-	-	1	4.0%
31	-	-	4	2.4%	-	-	1	4.0%
32	-	-	8	4.8%	-	-	0	0.0%
33	-	-	5	3%	-	-	1	4.0%
34	-	-	2	1.2%	-	-	2	8.0%
35	-	-	4	2.4%	-	-	0	0.0%
36	-	-	1	0.6%	-	-	1	4.0%
37	-	-	5	3%	-	-	0	0.0%
38	-	-	4	2.4%	-	-	1	4.0%
39	-	-	1	0.6%	-	-	1	4.0%
40	-	-	-	-	-	-	0	0.0%
41-50	-	-	29	17.4%	-	-	-	-
51-60	-	-	17	10.2%	-	-	-	-
> 61	-	-	5	3%	-	-	-	-

Figure 3. Norm sample and present study sample age comparison for adolescent sample

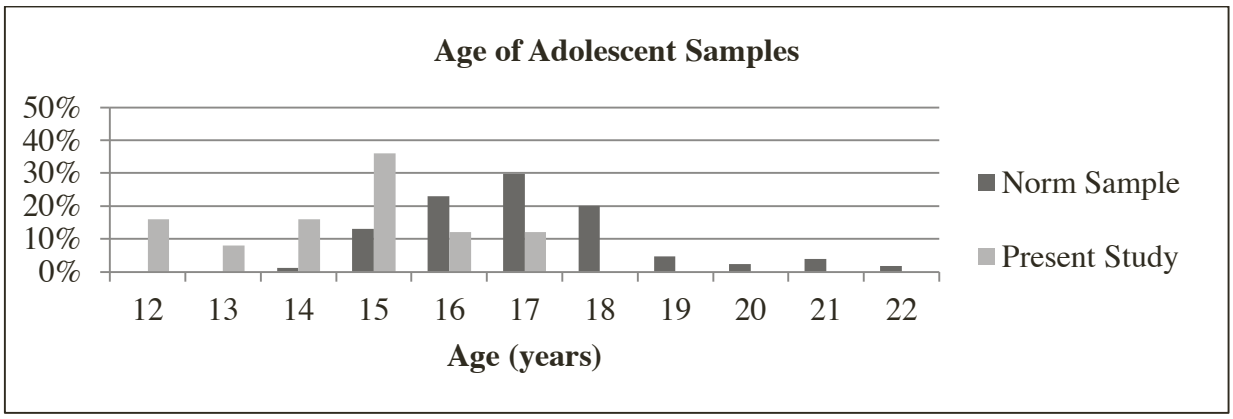
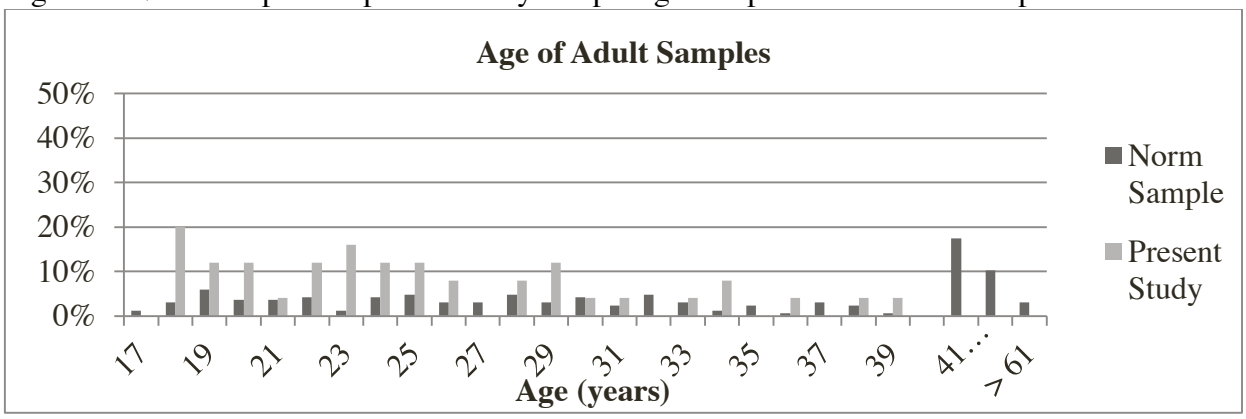


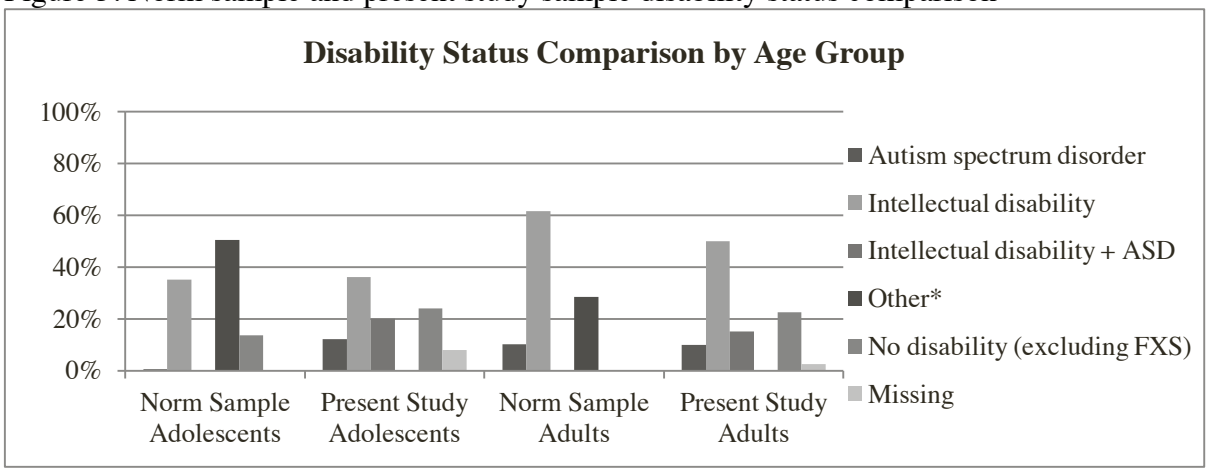
Figure 4. Norm sample and present study sample age comparison for adult sample



Additionally, there were differences in disability status, above and beyond the present study sample that consisted exclusively of individuals with full mutation FXS (Table 9; Figure 5). The norm samples did not include details on severity of disability and as a result comparisons could not be made regarding the degree of intellectual disability. However, there were still several differences based on disability categories. Individuals in the present study sample were organized into five disability categories: (1) intellectual disability; (2) ASD; (3) intellectual disability and comorbid ASD (ID+ASD); (4) no disability (i.e., FXS, but ID and/or ASD were not identified); and (5) missing data (2 adolescents and 1 adult). In comparison, the norm sample covered a wider range of disability status, which included intellectual disability, ASD, learning

disability, emotional disorder, other health impairment, orthopedic impairment, speech/hearing impairment, and traumatic brain injury. There were also some adolescents identified as having no disability. A similar percentage of adolescents and adults were identified as having an intellectual disability across the norm sample (35.2% adolescents; 61.5% adults) and the present study sample (36% adolescents; 50% adults). Similarly, percentages of individuals with ASD were consistent, with exception to fewer adolescents being identified as having ASD in the norm sample. While less than 1% of adolescents in the norm sample were identified as having ASD, 12% of adolescents in the present study were identified as ASD. 10% of the adult norm sample and adult present study sample were identified as having ASD. While some similarities between the norm sample and present study sample exist, based on percentages of individuals identified with ID and ASD, the remaining norm sample includes more individuals with impairments which are likely less severe, whereas 20% of the adolescent and 15% of the adult population in the present study are likely to be considered more severely impaired (i.e., identified as having ID and comorbid ASD).

Figure 5. Norm sample and present study sample disability status comparison



*Other includes emotional disorder, learning disability, orthopedic impairment, other health impairment, speech/hearing impairment, and traumatic brain injury

Finally, while adolescent and adult participants in the present study were enrolled based only on full mutation FXS and being between the ages of 12 and 40 years, they were more likely to have a wider range of employment status (e.g., unemployed). In comparison, data from participants in the norm sample for the SDS Adult Version were more specific. The norm sample was collected during a research study to examine the relationship between self-determination and employment. As a result, all participants in the norm sample were also already in contact with an employment support agency. The norm sample collected for the Adolescent Version consisted of 500 students from urban, suburban, and rural schools across five states and who were identified as receiving special education services by their school district.

In conclusion, for two primary reasons the raw scores were chosen for the subsequent analyses in the present study. First, based on what is known about each sample, there are several differences, specifically in the areas of age, disability status, and employment status for adults. Second, although using the raw scores for the subsequent analyses limits the opportunity to make comparisons to a broader I/DD population that is well documented in the literature, significant findings related to within group differences for individuals with FXS would have been overlooked.

Aim 1: Examine the psychometric properties of the parent-report and self-report measures of self-determination, respectively.

Question 1. *Is the original four factor model of the Arc's SDS valid for use with individuals with FXS?*

A confirmatory factor analysis on the Arc's SDS (Wehmeyer & Kelchner, 1995) was originally proposed to test whether the previously derived subdomains (i.e., autonomy, psychological empowerment, and self-realization) and total score were valid for use with

individuals with FXS, specifically those with intellectual disability in the moderate to severe range. The results were intended for comparison with the previous factor structure using a sample of individuals with intellectual disability differing from the normed sample (Wehmeyer, 1996). However, the present study included a sample that required use of both the adolescent and adult versions of the Arc's SDS and, as a result, two confirmatory factor analyses were necessary. Due to the small sample size of adolescents and adults in the present study, the results of each confirmatory factor analysis would have been significantly limited due to small sample sizes and thus were dropped from the present study.

Question 2. *What factors underlie the items of the Self-Determination Parent Questionnaire?*

The exploratory factor analysis on the SDPQ included a total of 77 cases (missing data from 8 participants in the sample). Preliminary analyses were used to determine the factor analysis with the best fit. First, 8 items were excluded due to concerns for multicollinearity, as indicated by the determinant of the correlation matrix, which was calculated at 3.46E-9 and was lower than the necessary minimum value of .00001. The 8 items removed were not original to the SDPQ (Carter, 2013), but were added to overlap with the four areas of self-determination suggested by Wehmeyer (1999) and used in the Arc's Self-Determination Scale (Wehmeyer & Kelchner, 1995) self-report form. After these items were removed, a second factor analysis also had too low of a determinant of 5.70E-6. The correlation matrix was assessed for items with high correlations. Two items (i.e., Begins Work and Tries Another Plan) from the "Wants and Needs" section of the SDPQ (Carter et al., 2013) were highly correlated (r =.80). After removing the item "My child begins work on plans to meet his or her goals as soon as possible", the determinant was 3.04E-5.

A principal axis factor analysis was conducted on 20 items of the *Self-Determination Parent Questionnaire (SDPQ;* Carter et al., 2013*)* with oblique (oblimin) rotation. An oblique rotation was used for two primary reasons. First, from a theoretical standpoint, the items in the SDPQ are all related and likely correlated, as they all describe behaviors related to self-determination. The main difference was that approximately half of the items focus on parent perspectives about the importance of skills related to self-determination for their children while the other half of the items relate to their child's skill level across different skill domains. Results showed that factor 1 and 2 are positively correlated (r=.40). Second, preliminary results from exploratory factor analyses were run using both oblique and orthogonal rotations and showed similar factors. Less than 7% of the variance was explained by additional factors after the first two factors were identified. As a result, a final factor analysis forced items into a two-factor model.

The Kaiser-Meyer-Olkin measure of sampling adequacy (KMO; Kaiser, 1970) confirmed that the sample was adequate for the analysis, KMO = .81 and falls in the meritorious range (Hutcheson & Sofroniou, 1999). KMO values for individual items were also reviewed. For Measures of Sampling Adequacy items were assessed in the anti-image correlation matrix (Table 11). All items ranged between .58 and .88, which was higher than the acceptable minimum value of .5 (Field, 2013). An initial analysis was run to obtain eigenvalues for each factor. There were 2 factors that met Kaiser's criterion of 1 and in total explained 43% of the variance. A total of 2 factors were retained. Factor 1 consists of 13 items and factor 2 consists of seven items.

Table 11. Anti image correlation matrix table

Choice - I	.58*																			
Choice -B	-0.18	.70*																		
Decisions -I	-0.14	-0.01	.86*																	
Decisions-B	0.16	-0.37	0.09	.82*																
Problem Solve – I	0.17	-0.26	-0.38	0.27	.77*															
Problem Solve - B	-0.14	0.18	0.04	-0.26	-0.18	.82*														
Goal Set - I	0.21	-0.02	-0.01	-0.17	-0.33	-0.03	.82*													
Goal Set - B	0.10	0.03	-0.10	-0.14	0.16	-0.26	0.14	.88*												
Advocacy - I	-0.18	0.29	-0.18	-0.23	-0.14	0.23	-0.18	-0.10	.82*											
Advocacy -B	0.03	-0.15	0.10	0.10	0.05	-0.17	-0.05	-0.15	-0.13	.85*										
Manage - I	-0.17	0.01	0.00	-0.04	-0.01	-0.13	-0.07	0.04	-0.16	0.07	.87*									
Manage - B	-0.11	-0.01	-0.19	-0.12	0.02	-0.08	-0.08	-0.11	0.03	-0.31	-0.08	.83*								
Aware - I	-0.24	0.03	-0.08	0.07	0.01	-0.13	-0.51	-0.11	-0.14	0.01	-0.28	0.15	.81*							
Aware - B	0.22	-0.11	0.10	0.04	-0.13	0.09	0.10	-0.13	-0.11	-0.08	-0.09	-0.30	-0.06	.86*						
Knows Likes	0.00	-0.39	-0.19	-0.04	0.17	-0.12	0.26	0.13	-0.14	0.00	0.09	-0.02	-0.14	-0.22	.68*					
Sets Own Goal	-0.10	0.15	-0.11	-0.30	-0.17	0.20	0.01	0.02	0.23	-0.18	0.01	0.17	-0.03	-0.29	-0.07	.81*				
Meet Own Goal	-0.03	-0.07	0.02	0.03	0.10	-0.20	-0.07	-0.01	-0.14	-0.03	0.26	0.05	-0.03	0.09	-0.05	-0.39	.86*			
Check Progress	-0.11	-0.20	-0.01	0.05	-0.01	-0.16	-0.07	-0.21	-0.02	0.14	-0.16	0.08	0.24	-0.11	0.21	-0.29	-0.16	.83*		
Ppl Listen To Child	-0.15	0.21	0.00	-0.12	-0.05	0.19	-0.17	-0.02	0.16	-0.21	-0.07	0.15	0.07	-0.16	-0.23	0.14	-0.11	-0.18	.77*	
Tries Another Plan	0.16	-0.08	0.07	0.06	0.06	-0.19	-0.05	-0.05	-0.11	0.20	-0.07	-0.26	0.09	0.05	0.09	-0.20	-0.30	0.11	-0.25	.83*

*Note: Items marked with "-I" asked about how important parents perceived that behavior and items marked with "-B" indicate ratings of behavior frequency

The pattern matrix (Table 12) lists the factor loadings after rotation, which provided information about the unique contribution of each variable to the factor. Table 13 provides factor loadings after rotation to illustrate the relationship between factors. Table 14 illustrates the correlation matrix for items included in the final factor analysis. There were 91 (47%) nonredundant residuals with absolute values greater than .05. Guidelines suggested that under 50% of residuals greater than .05 is acceptable (Field, 2013). Items in factor 1 that cluster together represented observed skills related to self-determination and factor 2 represented parent perceptions of importance of self-determination.

The two factors of the SDPQ each had high reliability (Factor 1 Cronbach's $\alpha = .90$; Factor 2 Cronbach's $\alpha = .85$). All items in factors 1 and 2 had a correlation above .3 with the respective total subscale. All items in factors 1 and 2, with the exception of choice-making (Cronbach's $\alpha = .87$), had Cronbach's α scores below the subscale reliability, which suggested that removing this item will not increase the reliability of the subscale (Table 15). The increase in reliability for the second factor was insubstantial when the choice-making item was removed (i.e., a change in alpha from .85 to .87). As a result, this item was not excluded from the factor due to conceptual validity.

The exploratory factor analysis was used to explore the best way to measure the constructs of interest (i.e., self-determined behaviors, and importance of self-determination) through parent-report. As a result, two subscales (self-determined behaviors and importance of self-determination) were identified from the factor analysis and used in the subsequent data analyses on the SDPQ.

Table 12. Pattern matrix - Exploratory factor analysis results for the Self-Determination Parent Questionnaire

ITEM	FACTOR 1	FACTOR 2
My child sets his or her own goals to satisfy wants or needs. (S)he thinks about his or her own abilities when setting goals.	**.774**	-.032
My child figures out how to meet goals alone. (S)he makes plans and decides what to do independently.	**.756**	-.070
Decision-Making Skills - SKILL LEVEL Examples: making decisions about one's future, independently or with help	**.705**	.002
Self-Awareness and Self-Knowledge - SKILL LEVEL Examples: being able to identify one's strengths, limitations, preferences, and interests	**.673**	.063
If a plan doesn't work, my child tries another one to meet his or her goals.	**.626**	-.027
My child checks his or her own progress when completing his or her plan. (S)he asks others what they think of his or her progress.	**.622**	.010
Goal-Setting Skills - SKILL LEVEL Examples: being able to set and track goals, as well as develop plans to achieve goals	**.621**	.023
Self-Advocacy and Leadership Skills - SKILL LEVEL Examples: knowing one's rights, communicating effectively, being an effective leader	**.566**	.065
Choice-Making Skills - SKILL LEVEL Examples: being able to make choices that reflect one's own preferences	**.555**	.065
Self-Management and Self-Regulation Skills - SKILL LEVEL Examples: monitoring own behavior, providing own reinforcement, self-directing own learning	**.515**	.153
Problem-Solving Skills - SKILL LEVEL Examples: being able to identify a problem, think of possible solutions, evaluate each solution	**.501**	.241
People listen when my child talks about what (s)he wants and is good at.	**.474**	.064
My child knows what (s)he needs, likes, and is good at.	**.435**	-.067
Self-Awareness and Self-Knowledge - IMPORTANT Examples: being able to identify one's strengths, limitations, preferences, and interests	-.095	**.882**
Goal-Setting Skills - IMPORTANT Examples: being able to set and track goals, as well as develop plans to achieve goals	.074	**.782**
Self-Advocacy and Leadership Skills - IMPORTANT Examples: knowing one's rights, communicating effectively, being an effective leader	.040	**.726**
Self-Management and Self-Regulation Skills - IMPORTANT Examples: monitoring own behavior, providing own reinforcement, self-directing own learning	-.011	**.714**
Problem-Solving Skills IMPORTANT Examples: being able to identify a problem, think of possible solutions, evaluate each solution	.046	**.645**
Decision-Making Skills - IMPORTANT Examples: making decisions about one's future, independently or with help	.075	**.619**
Choice-Making Skills - IMPORTANT Examples: being able to make choices that reflect one's own preferences	-.027	.351
Eigenvalues	6.26	2.25
% of variance	31.27%	11.26%
α	.90	.85

Note: Factor loadings over .40 are highlighted in bold

Table 13. Structure matrix

	Factor 1	Factor 2
SetsOwnGoal	0.761	0.279
MeetOwnGoal	0.728	0.234
DecisionMakingDO	0.706	0.285
SelfAwareDO	0.698	0.333
GoalSetDO	0.631	0.273
CheckProgress	0.626	0.26
TriesAnotherPlan	0.615	0.224
ProblemSolveDO	0.598	0.443
SelfAdvocLDO	0.592	0.292
SelfMgmtDO	0.576	0.36
ChoiceMakingDO	0.521	0.141
PplListenToChild	0.499	0.254
KnowsLikes	0.409	0.108
SelfAwareLEARN	0.259	0.844
GoalSetLEARN	0.388	0.811
SelfAdvocLEARN	0.332	0.742
SelfMgmtLEARN	0.276	0.71
ProblemSolveLEARN	0.305	0.663
DecisionMakingLEARN	0.324	0.649
ChoiceMakingLEARN	0.114	0.34

Extraction Method: Principal Axis Factoring
Rotation Method: Oblimin with Kaiser Normalization

Table 14. Correlation matrix with 1-tailed significance

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
Choice Making _IMP	1.00	0.16	0.00	0.38	0.23	0.04	0.13	0.30	0.01	0.25	0.00	0.18	0.00	0.48	0.10	0.26	0.23	0.10	0.16	0.37
CM_SD	0.12	1.00	0.04	0.00	0.03	0.03	0.16	0.02	0.38	0.00	0.26	0.00	0.28	0.00	0.00	0.00	0.00	0.00	0.06	0.03
DM_IMP	0.30	0.20	1.00	0.09	0.00	0.01	0.00	0.05	0.00	0.05	0.00	0.00	0.00	0.01	0.02	0.01	0.05	0.04	0.07	0.14
Decision Making _SD	0.04	0.47	0.16	1.00	0.16	0.00	0.00	0.00	0.01	0.00	0.02	0.00	0.03	0.00	0.00	0.00	0.00	0.00	0.00	0.00
Problem Solve LEARN	0.09	0.21	0.60	0.11	1.00	0.01	0.00	0.23	0.00	0.05	0.00	0.02	0.00	0.00	0.41	0.00	0.06	0.02	0.06	0.08
Problem Solve DO	0.20	0.21	0.25	0.49	0.27	1.00	0.00	0.00	0.01	0.00	0.00	0.00	0.00	0.00	0.09	0.00	0.00	0.00	0.03	0.00
Goal SetL EARN	0.13	0.11	0.47	0.32	0.65	0.40	1.00	0.04	0.00	0.01	0.00	0.01	0.00	0.01	0.38	0.00	0.01	0.01	0.01	0.01
Goal Set DO	0.06	0.23	0.19	0.49	0.09	0.54	0.21	1.00	0.01	0.00	0.03	0.00	0.04	0.00	0.09	0.00	0.00	0.00	0.01	0.00
Self Advoc LEARN	0.28	0.04	0.51	0.29	0.47	0.28	0.60	0.26	1.00	0.01	0.00	0.00	0.00	0.01	0.12	0.05	0.03	0.07	0.06	0.04
Self Advoc DO	0.08	0.33	0.19	0.39	0.19	0.40	0.26	0.45	0.27	1.00	0.07	0.00	0.03	0.00	0.01	0.00	0.00	0.01	0.00	0.01
Self Mgmt LEARN	0.35	0.07	0.40	0.23	0.41	0.37	0.55	0.22	0.53	0.17	1.00	0.01	0.00	0.01	0.43	0.09	0.29	0.02	0.04	0.08
Self Mgmt DO	0.11	0.32	0.32	0.42	0.23	0.43	0.28	0.45	0.30	0.52	0.28	1.00	0.04	0.00	0.01	0.00	0.01	0.02	0.04	0.00
Self Aware LEARN	0.36	0.07	0.50	0.21	0.50	0.37	0.72	0.20	0.62	0.21	0.63	0.21	1.00	0.03	0.16	0.06	0.08	0.20	0.07	0.16
Self Aware DO	-0.01	0.43	0.28	0.45	0.32	0.30	0.27	0.44	0.29	0.49	0.26	0.52	0.22	1.00	0.00	0.00	0.00	0.00	0.00	0.00
Knows Likes	0.15	0.50	0.24	0.33	0.03	0.16	-0.04	0.16	0.14	0.30	0.02	0.27	0.11	0.43	1.00	0.01	0.02	0.24	0.01	0.15
Sets Own Goal	0.07	0.37	0.27	0.53	0.31	0.36	0.30	0.40	0.19	0.40	0.16	0.30	0.18	0.57	0.28	1.00	0.00	0.00	0.00	0.00
Meet Own Goal	0.09	0.32	0.19	0.48	0.18	0.47	0.29	0.42	0.23	0.36	0.07	0.28	0.16	0.39	0.25	0.69	1.00	0.00	0.00	0.00
Check Progress	0.15	0.33	0.20	0.41	0.25	0.41	0.27	0.45	0.17	0.26	0.24	0.23	0.10	0.42	0.08	0.60	0.54	1.00	0.00	0.00
Ppl Listen To Child	0.12	0.18	0.17	0.32	0.18	0.22	0.29	0.27	0.18	0.33	0.20	0.21	0.17	0.39	0.27	0.37	0.40	0.37	1.00	0.00
Tries Another Plan	-0.04	0.22	0.13	0.41	0.17	0.44	0.29	0.39	0.21	0.26	0.16	0.40	0.12	0.35	0.12	0.50	0.60	0.38	0.39	1.00

*Note: Items marked as "_IMP" asked about how important parents perceived that behavior and items marked with "_LEARN" indicate behavior rating

Table 15. Reliability item-total statistics

ITEM	Corrected Item-Total Correlation	Cronbach's alpha if Item Deleted
FACTOR 1		
ChoiceMaking DO	0.507	0.892
DecisionMaking DO	0.671	0.886
ProblemSolveDO	0.594	0.890
GoalSetDO	0.672	0.887
SelfAdvocDO	0.614	0.888
SelfmgmtDO	0.546	0.890
SelfAwareDO	0.694	0.885
KnowsLikes	0.398	0.896
SetsOwnGoal	0.758	0.879
MeetOwnGoal	0.739	0.880
CheckProgress	0.666	0.885
TriesAnotherPlan	0.650	0.886
PplListenToChild	0.508	0.897
FACTOR 2		
ChoiceMakingLEARN	0.316	0.868
DecisionMaking LEARN	0.620	0.831
ProblemSolveLEARN	0.601	0.833
GoalSet LEARN	0.704	0.817
SelfAdvoc LEARN	0.692	0.819
SelfMgmt LEARN	0.620	0.832
SelfAware LEARN	0.772	0.809

Aim 2: Describe the characteristics of self-determination in adolescent and adult males and females ages 12-40 with FXS on the self-report *Arc's Self-determination Scale* and the parent-report *Self-Determination Questionnaire* in order to examine predictors of self-determination.

Question 3. *What are the shape, range, mean, and variance of scores on each of the four subdomains (i.e., autonomy, self-regulation, psychological empowerment, and self-realization) and total self-determination score of the Arc's Self-determination scale for male and female adolescents and adults with FXS?*

Autonomy domain

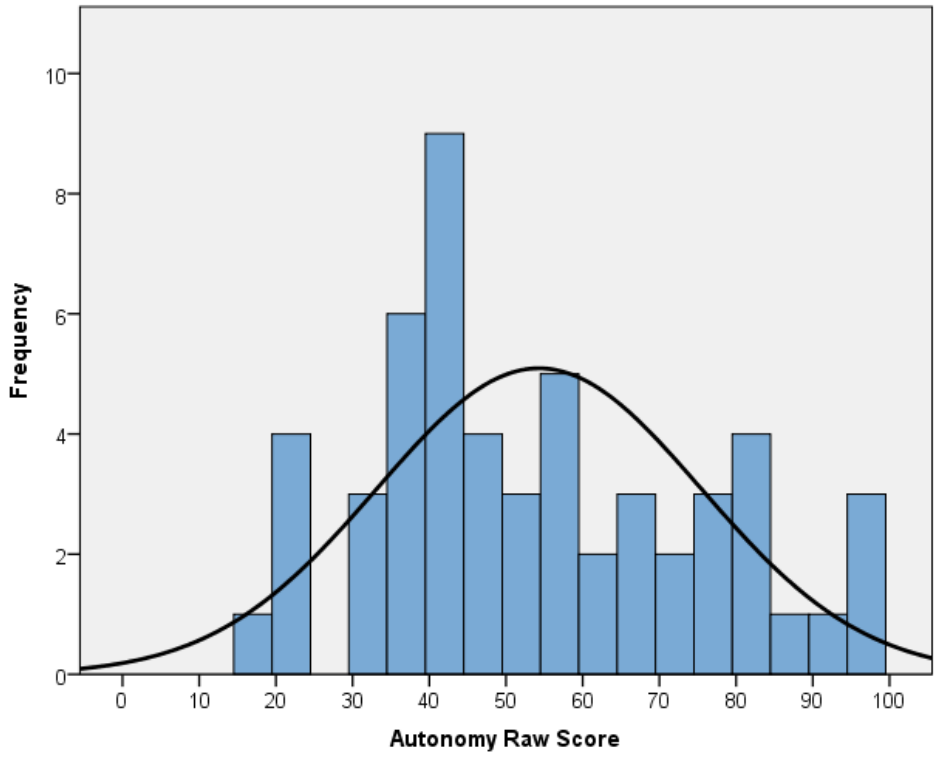
There were 96 total points possible on the Autonomy domain. Scores were obtained for a total 54 participants (Table 16). The Shapiro-Wilk normality test confirmed that the data were not significantly different from a normal distribution. The data were slightly platykurtic with a score of -.76. Figure 6 shows the raw score distribution for the Autonomy domain.

Table 16. Arc's SDS section raw score descriptive statistics

	n	Points Possible	*M(SD)*	Range	Skewness *(SE)*	Kurtosis *(SE)*
Autonomy	54	96	54.30(21.15)	17 – 96	.37(.33)	-.76 (.64)
Self-Regulation	47	21	5.81(4.86)	0 – 21	1.04(.35)	.96(.68)
ICPS*	47	(12)	2.77(3.27)	0 – 11	.80(.33)	-.71(.66)
Goal Setting	47	(9)	2.57(2.36)	0 – 9	.96(.33)	.69(.66)
Empowerment**	54	16	11.56(2.92)	5 – 16	-.23(.33)	-1.09(.64)
Self-Realization	54	15	10.52(2.87)	4 – 15	-.79(.33)	-.10(.64)
SD Total Score	47	148	85.53(24.86)	28 – 142	.23(.35)	-.23(.68)

*Interpersonal Cognitive Problem Solving
**Psychological Empowerment

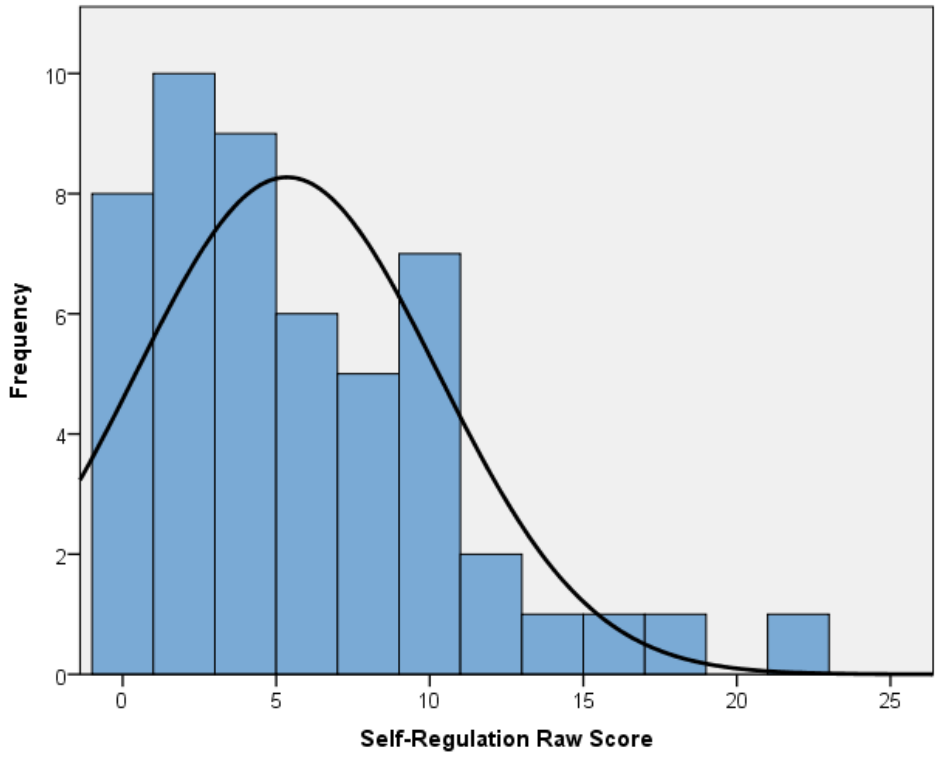
Figure 6. Autonomy Raw Score Histogram



Self-Regulation domain

The Self-Regulation domain included 21 total points possible. Scores on the Self-Regulation domain were obtained for a total 42 participants. The mean for the overall sample was 5.81 (Table 14). The Shapiro-Wilk normality test confirmed that the data were significantly different from normal distribution. The data appeared positively skewed and leptokurtic with a score of .96 (standard error = .68; Figure 7).

Figure 7. Self-Regulation Raw Score Histogram



The Self-Regulation domain consisted of two subdomains, (a) Interpersonal Cognitive Problem-Solving and (b) Goal Setting, which accounted for 12 and 9 points, respectively. The Interpersonal Cognitive Problem-Solving subdomain had a mean score of 2.77 (SD = 3.27; range 0 – 11; Table 16; Figure 8) and the Goal Setting subdomain had a mean score of 2.57 (SD = 2.36; range 0 – 9; Figure 9). The Shapiro-Wilk normality test confirmed that the data were significantly different from normal distribution for both subdomains (Interpersonal Cognitive

Problem Solving $S\text{-}W = .81, df = 47, p < .01$; Goal Setting $S\text{-}W = .89, df = 47, p < .01$). The scores on the Interpersonal Cognitive Problem Solving were leptokurtic with a score of -.71 (standard error = .66; Figure 8). The scores on the Goal Setting were also leptokurtic with a score of .69 (standard error = .66; Figure 9).

Figure 8. Self-Regulation – Interpersonal Cognitive Problem Solving Raw Score Histogram

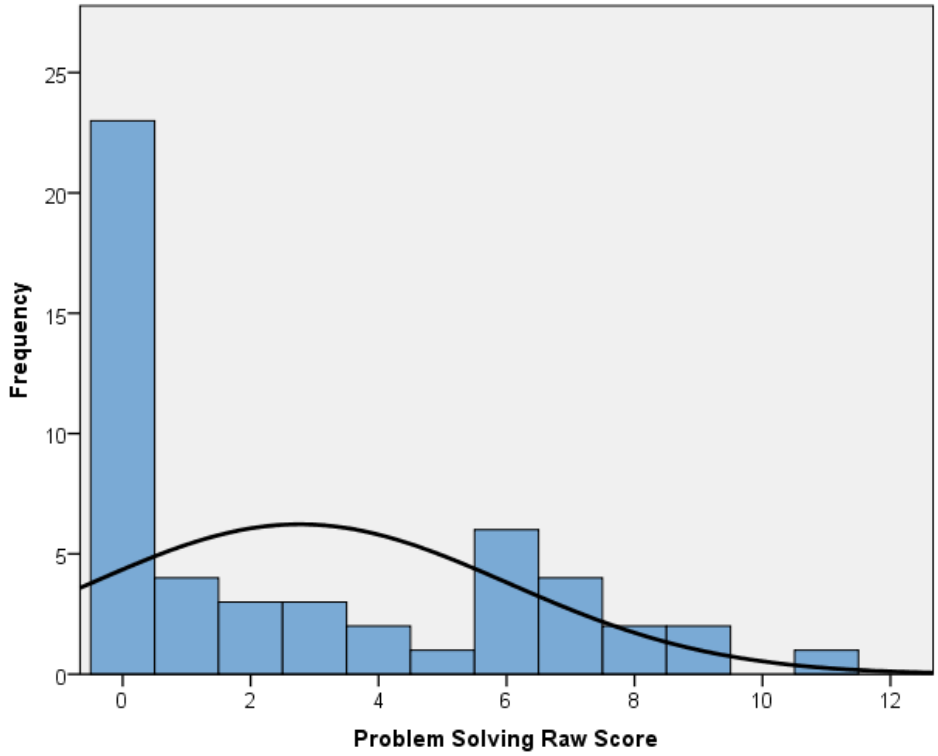
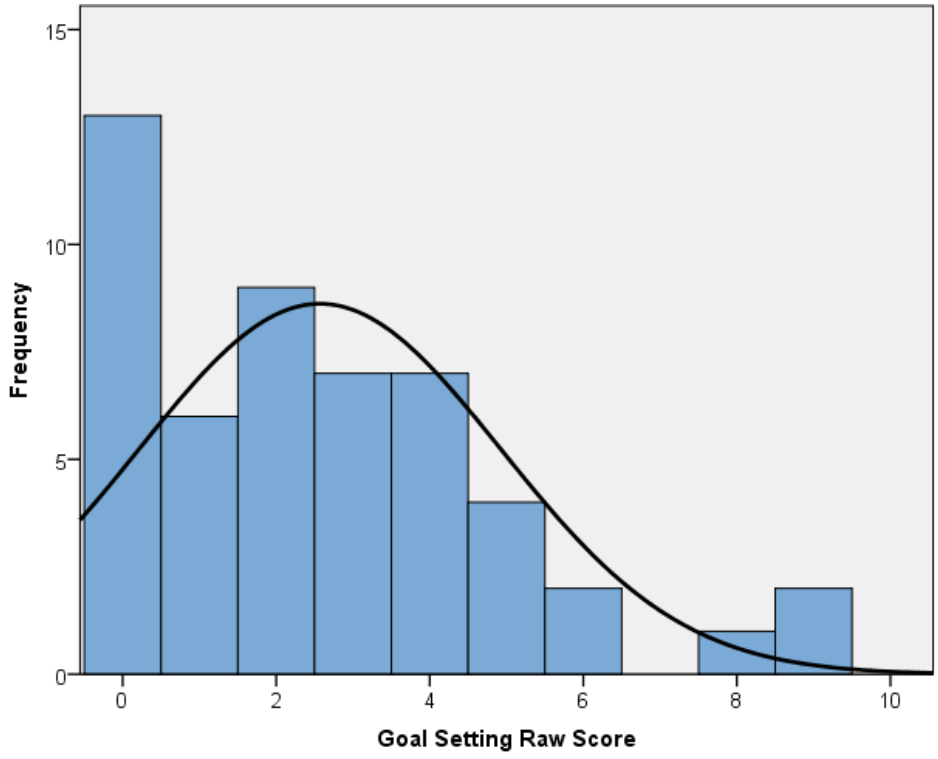


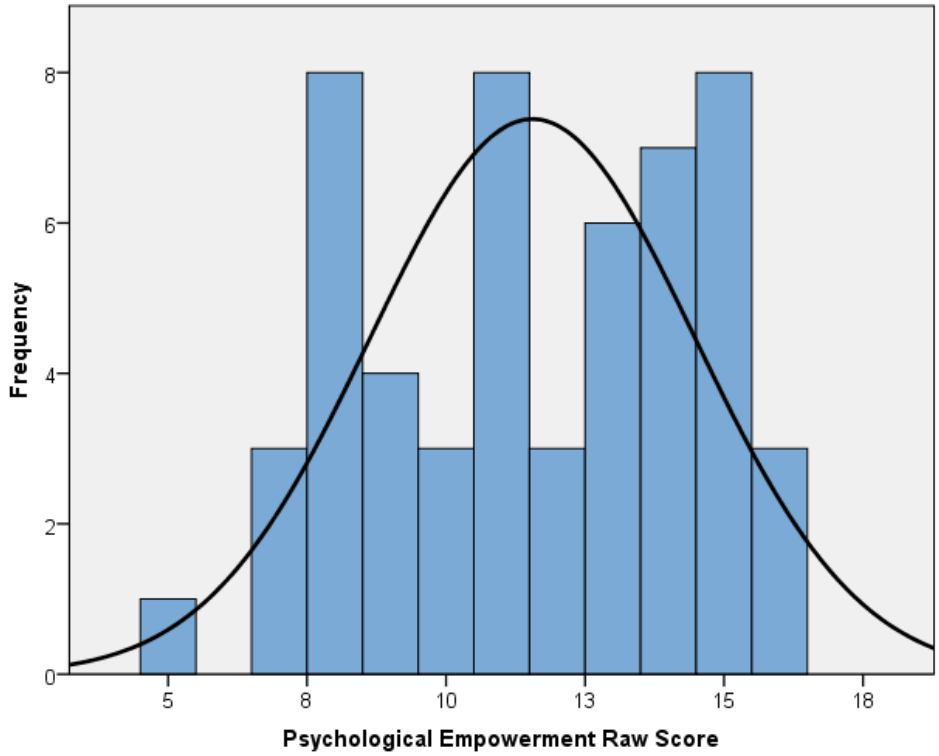
Figure 9. Self-Regulation – Goal Setting Raw Score Histogram



Psychological Empowerment domain

There was 16 total points possible on the Psychological Empowerment domain. Scores on the Psychological Empowerment domain were obtained for a total 47 participants. The mean for the overall sample was 11.56 (SD =2.92; range 5 – 16; Table 16). The Shapiro-Wilk normality test suggested that the data were significantly different from a normal distribution, ($S\text{-}W = .94, df = 54, p = .01$). The data were platykurtic with a score of -1.09 (standard error = .64). Figure 10 shows the raw score distribution for the Psychological Empowerment domain.

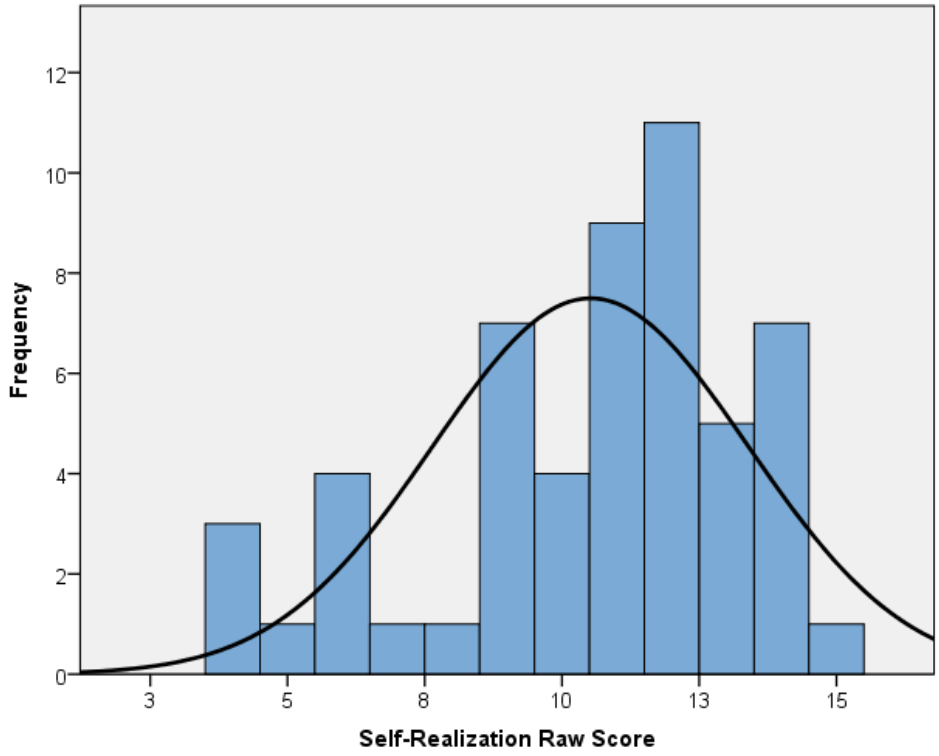
Figure 10. Psychological Empowerment Raw Score Histogram



Self-Realization domain

There were 15 total points possible on the Self-Realization domain. Scores on the Self-Realization domain were obtained for a total 54 participants. The mean for the overall sample was 10.52 (Table 16). The minimum score was 4 and maximum score was 15. The Shapiro-Wilk normality test suggested that the data were significantly different from a normal distribution. The data were negatively skewed. Figure 11 shows the raw score distribution for the Self-Realization domain.

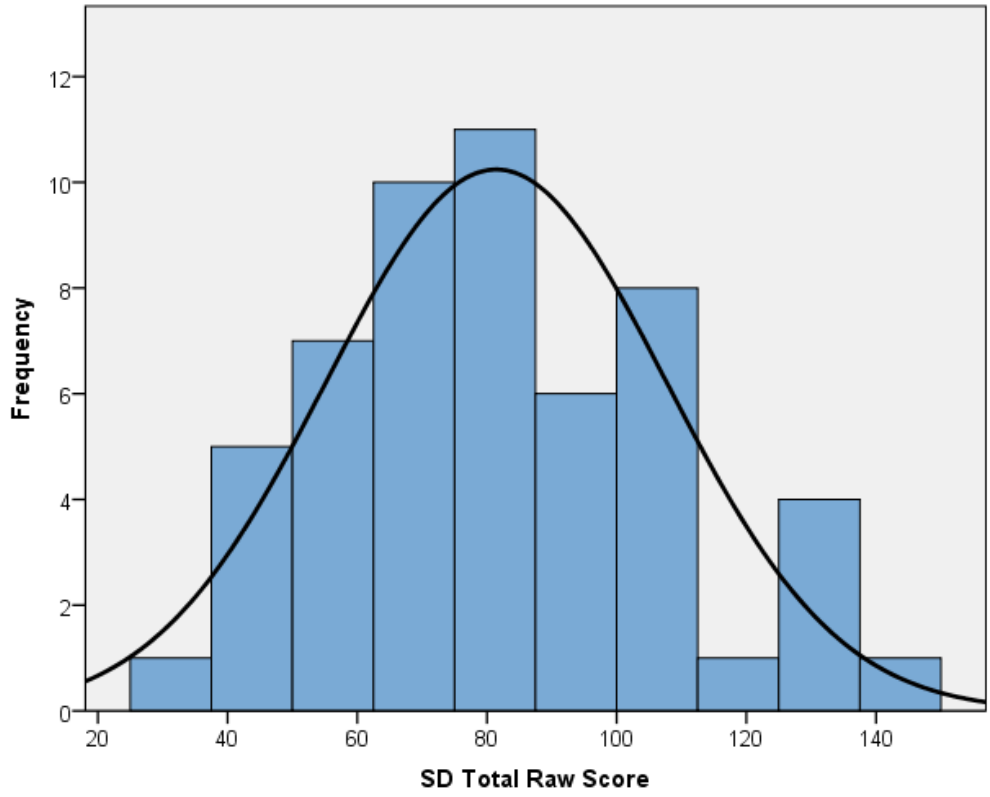
Figure 11. Self-Realization Raw Score Histogram



Self-Determination Total Score

The total score on the Self-Determination Scale (comprised of the four domain scores) had a total of 148 points possible. Self-Determination total scores were calculated for a total 47 participants. The mean for the overall sample was 85.53 (Table 16). The Shapiro-Wilk normality test suggested that the data were not significantly different from a normal distribution. Figure 12 shows the raw score distribution for the Self-Determination Total Score.

Figure 12. Self-Determination Total Raw Score Histogram



Question 4. *What are the shapes, means, and standard deviations of parent-reported self-determined behaviors and importance of self-determination reported by parents on the Self-determination Questionnaire?*

Two subdomains emerged for the SDPQ (Carter et al., 2013) based on the exploratory factor analysis (Aim 1, Question 2). The first subdomain was comprised of items that asked parents to rate their child's behaviors related to self-determination in Section I (e.g., How well do you feel your child does this [behavior] now?), as well as six of seven items of the Wants and Needs questions in Section V. The second subdomain that emerged in the factor analysis was comprised of parent ratings of importance of skills related to self-determination in section I (e.g., How important do you feel it is for your child to learn this skill now?) for each of the 7 skill areas (e.g., decision-making). Due to multicollinearity among the four broad domains of self-

determination proposed by Wehmeyer (1999) in Section II of the SDPQ and the behavior skills in Section I, the broader items were dropped from the factor analysis. However, descriptive statistics and correlations on these items with Section I items are included below. The rationale for maintaining the discrete skills in Section I over the broader self-determination domains in Section II was that Section I asked parents to rate more observable and discrete skills than Section II, which seemed more valid and more amenable to intervention.

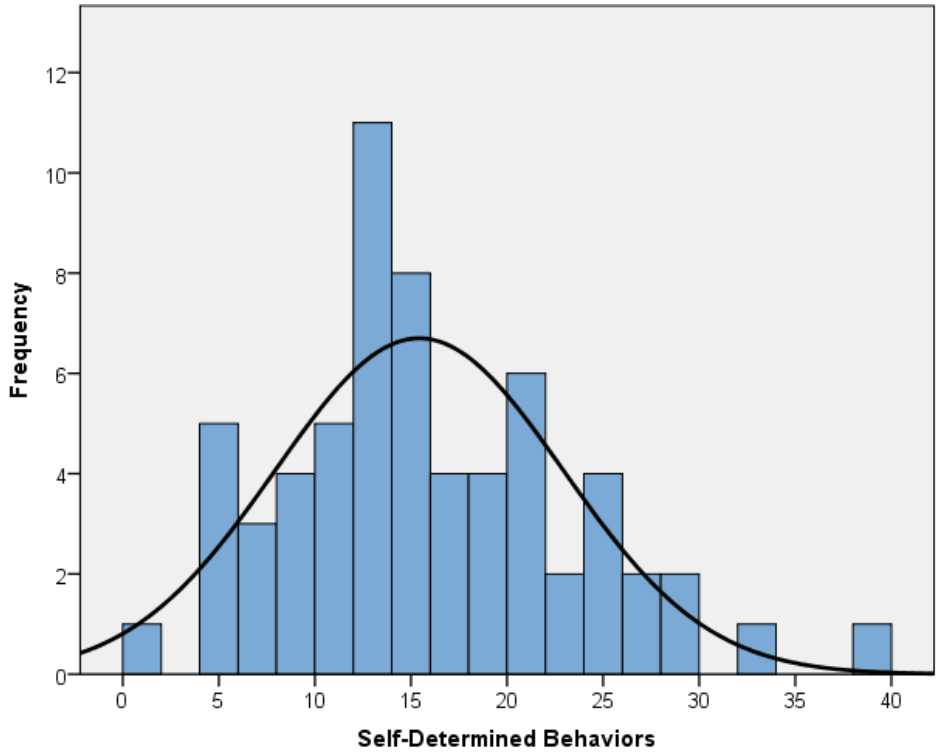
Self-Determination Skills Subscale

The average total score on the Self-Determination Behaviors subscale for the entire sample was 14.80(Table 17). The total scores for males (SD = 5.47) was significantly lower than the total scores for females. The distributional shape of SD Skills subscale scores was examined to determine the extent to which assumptions of normality were met. Skewness, kurtosis, and the Shapiro-Wilk test of normality ($S\text{-}W$ = .96, df = 84, p = .01) suggested that normality was not assumed (Figure 13).

Table 17. Descriptive statistics for the self-determined behaviors subscale (N = 84)

	n	*M(SD)*	Skewness *(SE)*	Kurtosis *(SE)*
Males	54	12.65(5.47)	.49(.33)	1.20(.64)
Females	30	18.67(8.73)	.15(.43)	-.39(.83)
Entire Sample	84	14.80(7.37)	.73(.26)	.67(.52)

Figure 13. Sample distribution of Self-Determination Behaviors subscale scores (N=83)



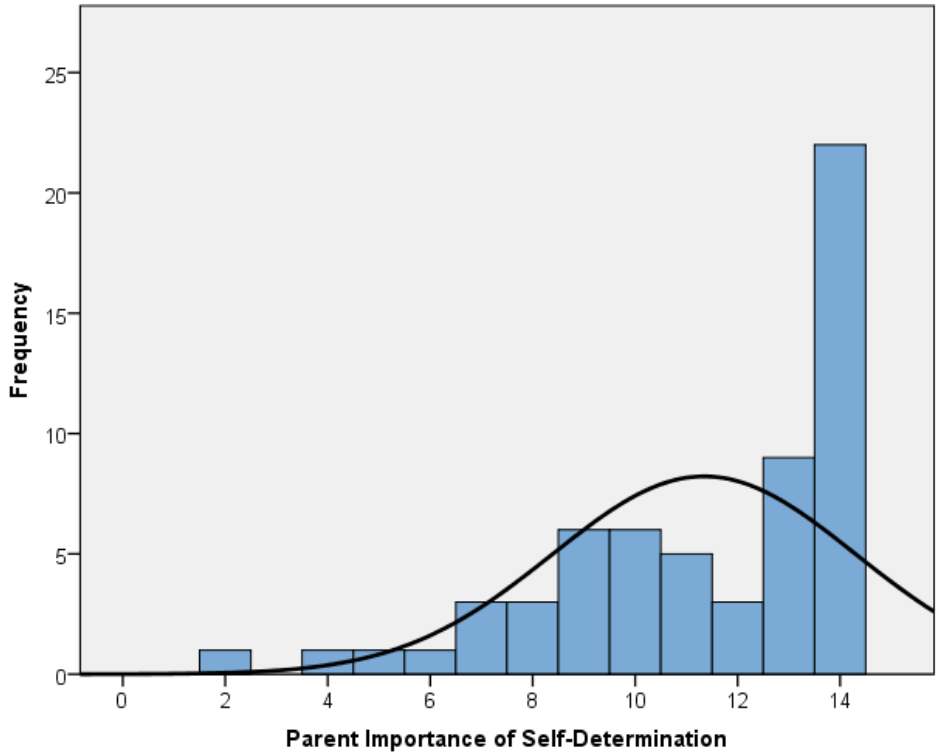
Importance of Self-Determination Subscale

Scores on the Importance of Self-Determination subscale were obtained for a total 82 participants. The mean for the overall sample was 11.38 (Table 18). The data were negatively skewed. The Shapiro-Wilk test confirmed that the data were significantly different from a normal distribution, ($S\text{-}W = .85, df = 54, p < .01$). The data are also leptokurtic, which suggested a heavy-tailed distribution as illustrated in Figure 14.

Table 18. Descriptive statistics for the importance of self-determination subscale (N = 82)

	n	*M(SD)*	Skewness *(SE)*	Kurtosis *(SE)*
Males	54	11.07(3.12)	-.98(.33)	.34(.64)
Females	28	11.96(2.53)	-.98(.44)	-.30(.86)
Entire Sample	82	11.38(2.95)	-1.03(.27)	.42(.53)

Figure 14. Sample distribution of Importance of SD Subscale scores (N = 82)



Question 5. *What is the correlation between individual characteristics and self-reported self-determination in males and females with FXS separately?*

There were several correlations between domain scores and individual characteristics (Table 19). The Autonomy domain was significantly positively correlated with age and adaptive behavior, and negatively correlated with social avoidance. The Self-Regulation domain was significantly positively correlated with adaptive behavior, and negatively correlated with ASD. The Psychological Empowerment domain was significantly positively correlated adaptive behavior, and negatively correlated with ASD. The Self-Realization domain was significantly positively correlated with adaptive behavior, and negatively correlated with ASD. The Self-Determination Total Score was significantly positively correlated with age and adaptive behavior, and negatively correlated with social avoidance.

Table 19. Correlation matrix – Predictor variables and Arc SDS subdomains

	Age	FSIQ	ASD	Adaptive Behavior	Anxiety	Social Avoidance	Autonomy Raw Score	Self-Regulation Raw Score	Psychological Empowerment Raw Score	Self-Realization Raw Score	Self-Determination Total Score
Age	--										
FSIQ	-0.03	--									
ASD	**-.263***	-0.12	--								
Adaptive Behavior	0.10	0.15	**-.362****	--							
Generalized Anxiety	0.13	-0.06	0.09	-0.07	--						
Social Avoidance	-0.08	-0.01	0.20	-0.15	**.411****	--					
Autonomy Raw Score	**.351***	-0.15	-0.09	**.449****	-0.06	**-.411****	--				
Self-Regulation Raw Score	0.01	0.17	**-.331***	**.573****	-0.12	-0.22	**.357***	--			
Psychological Empowerment Raw Score	-0.04	-0.15	**-.361****	**.525****	-0.04	-0.21	**.341***	**.527****	--		
Self-Realization Raw Score	0.05	0.04	**-.339***	**.551****	-0.23	-0.12	**.310***	**.508****	**.534****	--	
Self-Determination Total Score	**.283***	-0.12	-0.22	**.590****	-0.09	**-.411****	**.951****	**.593****	**.551****	**.512****	--

**Correlation is significant at the 0.01 level (2-tailed).
*Correlation is significant at the 0.05 level (2-tailed).

Several correlations among child characteristics were identified. ASD was negatively correlated with adaptive behavior, indicating that individuals with fewer daily living skills were more likely to meet criteria for ASD. Younger participants were also more likely to meet criteria

for ASD. Lastly, there was a significant correlation between parent-reported symptoms of social avoidance and generalized anxiety, which is consistent with previous research in I/DD (Esbensen et al., 2003). Full scale IQ was not significantly correlated with any other individual characteristics.

Question 6. *How does performance on the 'Interpersonal Cognitive Problem Solving' and the 'Goal Setting and Task Performance' subsections of the Self-Regulation domain differ by points earned and qualitatively by age and gender?*

The two subsections of the Self-Regulation domain of the *Arc's Self-Determination Scale* were analyzed separately to examine differences in performance by age and gender (Table 20). There were significant gender differences on the Interpersonal Cognitive Problem Solving subdomain, as well as the Goal Setting subdomain. Females earned more points than males on both subdomains. There were not significant differences between adolescents and adults on the Interpersonal Cognitive Problem Solving or the Goal Setting subdomain.

Table 20. Descriptive statistics for Self-Regulation subdomains by gender and age groups

	Gender				**Age Group**			
	Male	Female			Adolescent	Adult		
	M(SD)	*M(SD)*	*t*	*df*	*M(SD)*	*M(SD)*	*t*	*df*
Problem-Solving	.44(1.04)	5.00(3.11)	3.49**	52	2.61(3.36)	2.85(3.26)	.99	63
Goal Setting	1.88(1.62)	3.23(2.78)	-2.13*	40	2.00(1.82)	2.88(2.58)	-1.42	49

*$p < .05$, two-tailed
**$p < .01$, two-tailed

Data from these subsections were also reviewed from a content standpoint to explore qualitative differences by age and gender. Of 23 adolescents, 7 (6 males) were unable to complete and/or were not administered this section of the SDS. Of 40 adults, 7 (6 males) were unable to complete and/or were not administered this section of the SDS. Data on 8 adolescent

females and 8 adolescent males, as well as 16 adult females and 17 adult males were reviewed and reported on below.

Interpersonal Cognitive Problem Solving

Participants were presented with six questions that provided the beginning and end of an interpersonal situation in a story format. Participants were asked to provide an action that happened in the middle of the story in order to connect the beginning of the story (the dilemma) with the end of the story (the solution). For example, one item begins with, "Your friends are acting like they are mad at you. You are upset about this" and ends with, "The story ends with you and your friends getting along just fine." Each item received a maximum of 2 points possible with the option of earning 1 point for partial credit. The Interpersonal Cognitive Problem Solving (ICPS) subsection had a maximum of 12 points.

Two of 8 females (25%) and all 8 adolescent males (100%) earned 0 points on the Interpersonal Cognitive Problem Solving subsection. Many of them gave responses that were inconsistent with the story prompt and several said they "did not know" in response to each of the six prompts. Of the remaining adolescent females, 1 adolescent female earned a total of 2 points (1 point on each of 2 questions). Five adolescent females earned total subsection scores that ranged between 6 and 9 points (i.e., earned 1 or 2 points on each item). One-point responses typically provided one action or step, in comparison to 2-point responses, which gave an action from the participant as well as a response from other actors in the story (e.g., your friends). For example, in the prompt described above, one adolescent female responded, "Try to talk to them about it and see what's wrong" and earned 1 point for stating an action she could take in the story, but not providing an action or response from the other actors in the story (i.e., her friends). One example of a 2-point response provided by an adolescent female was, "You wonder why

they are mad so you text them. They say that they were just having family problems and tired. You text back, 'I understand.'"

Most adult males (n = 12; 70.6%) and one of 16 adult females (6.3%) earned 0 points on the Interpersonal Cognitive Problem Solving subsection. Eight adults (4 males; 4 females) earned credit (i.e., 1 or 2 points) on half or fewer of the questions for a total score of 3 or less. One adult male earned 4 points total by providing a 1-point response (i.e., one action to complete the story) on four of six items. 11 adult females earned a total score that was between 4 and 11 points. An example of a common one-point response to a story prompt about not knowing anyone at a new job and the story ending with having many friends at the new job, one individual stated, "introduce yourself" and earned 1 point. A more detailed 2-point response was, "I start by asking about things I see on their desks that show their interests like a snow globe or a postcard, I start being friendly to the people and they all were happy to be my friend."

More adult males than adolescent males completed the Interpersonal Cognitive Problem Solving subsection. However, adult and adolescent females were better able to complete this subsection in comparison to adolescent and adult males. There was also more variability in performance across items for adults than adolescents. Adults were better able to respond and earn credit on items 35, 36, and 38, which were about responding to friends who appear mad, forgetting materials needed for work or school, and being at a new job, but not knowing anyone and wanting friends. In comparison, items 33 and 37, which were about negotiating in a planning meeting at school or work and wanting to be elected for a committee at work or school, were the lowest scoring items. Adults appeared to have the most difficulty with two items that were less common interpersonal situations than the others. For example, while item 37, which was the lowest scoring item, is about being on a committee at work, while the highest scoring item (item

38) is about making friends at a new job. The situation of trying to make friends in a new environment is more relatable and ordinary, in comparison to being on a committee at work (i.e., a situation that is likely less often encountered and less understood).

Goal Setting

Participants were asked three questions regarding plans for the future (i.e., after graduation for adolescents and in five years for adults). All participants were asked if they had a specific goal or plan and then were asked to list up to four things they should do to meet that goal related to 1) living situation, 2) employment, and 3) transportation.

Living situation. Of 16 adolescents who completed the Self-Regulation section, the majority of them (n = 11; 6 males and 5 females) reported that they did not plan for where they would live after they graduate high school. Of the remaining five adolescents, one adolescent male shared which state he wanted to live in and another reported that he wants to live at college. Two of the remaining three adolescent females shared where they wanted to live and gave multiple steps they would need to take (e.g., work hard in school, talk to parents, save money).

Of 33 adults (17 male; 16 female) who completed the Self-Regulation section, over half (69%; n = 18; 12 female; 6 male) of the adults reported that they did not plan for where they would live in five years. The remaining males shared where they wanted to live (n =11) and three reported one step they would need to take in order to achieve that goal. The majority of males shared either a location (e.g., city, state, or type of housing) or with whom they wanted to live (e.g., grandparents, brother, friend). The remaining females (n = 4) reported where they wanted to live (e.g., region of the country) in the future and provided multiple steps (e.g., apply for a job in that area, be hired, save money).

Employment. Of the 16 adolescents who completed the Self-Regulation section, 12 adolescents (6 males; 6 females) reported that they did not have plans or goals related to employment. The remaining four adolescents (2 male; 2 female) shared where they want to work after they graduate (e.g., at Disney, at a dance studio, at a store, at a factory), however, none provided steps to take to get the job they want.

Nearly half (*n*=14; 6 male; 8 female) of the 33 adults that completed the Self-Regulation section did not report future plans for employment. Several adult males (*n* = 7) stated where they wanted to work (e.g., office, Bojangles, Dominos, Pizza Hut, video game store), but did not state any steps they could take to work toward their goal. Four males and three females reported where they want to work and provided one step. For example, one adult male shared that he wanted to do woodworking and stated that he would need to get a degree. One female shared that she wanted to work in an office and would have to apply for a job. Five females shared plans for employment and provided multiple steps to reach their goal. For example, one adult female reported that she wanted to work in an elementary school and listed several steps including going to school, getting a master's degree, and getting her teaching license.

Transportation. Nine (6 males; 3 females) of 16 adolescents reported they did not have plans or goals related to transportation after they graduate. One male and one female gave plans (e.g., a car, a bus) without giving steps to take toward their plan. The remaining five adolescents gave steps that included saving money, and getting a permit and license. 13 (8 males; 5 females) of 33 adults did not report a goal related to transportation. Seven adult males and four adult females stated a goal related to transportation (e.g., bus, bike, car, family car, community transportation), but did not provide any additional details. Two adult males and five adult females provided a goal and one or more steps for transportation.

Question 7. *What is the correlation between child variables and parent-reported self-determination and importance of self-determination in males and females with FXS?*

In the total sample there were several correlations between subscale scores and individual characteristics (Table 21). The parent-reported SD Behaviors subscale was correlated with adaptive behavior, and negatively correlated with ASD, and social avoidance. On the parent-rated Importance of SD subscale, there were few correlations with child characteristics, which was attributed to the lack of variability and negative skew of the subscale data. There was a small positive correlation between the Importance of SD subscale and adaptive behavior. The Importance of SD subscale and the SD Behaviors subscale were positively correlated.

Table 21. Correlation matrix – Predictor variables and SDPQ subscales

	1	2	3	4	5	6	7	8
Age	--							
FSIQ	-.30	--						
ASD	**-0.27***	-.11	--					
Adaptive Behavior	0.10	0.15	**-.39****	--				
Anxiety	0.14	-0.06	0.12	-0.05	--			
Social Avoidance	-0.07	-0.01	0.22	-0.14	**0.40****	--		
SD Behaviors	0.17	0.02	**-0.36****	**0.53****	-0.03	**-0.30****	--	
Importance of SD	0.04	-0.15	-0.04	**0.23***	0.12	-0.02	**0.37****	--

** Correlation is significant at the 0.01 level (2-tailed).

* Correlation is significant at the 0.05 level (2-tailed).

There were several correlations among child characteristics. ASD was negatively correlated with adaptive behavior, indicating that individuals with fewer daily living skills were more likely to meet criteria for ASD. Younger participants were also more likely to meet criteria for ASD. Lastly, there was a significant correlation between parent-reported symptoms of social avoidance and generalized anxiety, which is consistent with previous research in I/DD (Esbensen et al., 2003).

In the second aim, data on the self- and parent-report from the present study sample were described and differences by gender were explored. Participants, particularly males and adolescents, exhibited marked difficulty on the self-regulation section of the SDS. While the majority of parents rated nearly all behaviors related to self-determination as "very important," parents of females reported that their child exhibited higher levels of self-determined behaviors than did parents of males. Adaptive behavior was significantly positively correlated with all domains on the SDS and three of the four domains were significantly negatively correlated with ASD. Additionally, the Autonomy domain was significantly positively correlated with age and negatively correlated with social avoidance. Parent-reported SD behaviors were also positively correlated with adaptive behavior and negatively correlated with ASD and social avoidance.

Aim 3: Examine predictors of self-determination in individuals with FXS, including autism spectrum disorder (ASD), anxiety, intellectual functioning, adaptive behavior, and age to determine how these characteristics influence self-reported and parent-reported self-determination in male and female adolescents and adults with FXS.

Question 8. *To what extent do ASD, anxiety, intellectual functioning, adaptive behavior, and age predict self-reported self-determination subdomain scores in individuals with FXS?*

First, multiple regression analyses were used to explore whether gender moderated the effects of age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior as predictors of self-reported behaviors of self-determination across each subdomain and the total score. After centering the variables and creating interaction variables, the six predictors and the interactions were entered into a simultaneous regression model.

Results suggested that while gender did not moderate self-reported skills of self-determination for any of the subdomain scores or total score, gender was a main effect for some

subdomains. Specifically, gender was not a moderator (i.e., the interaction variable did not account for a significant proportion of the variance) for Autonomy, (Table 22), nor was there a main effect. Gender was not a moderator of Self-Regulation (Table 23), however, there was a main effect. Similarly, gender was not a moderator of Psychological Empowerment (Table 24), but there was a main effect. Although gender did not moderate Self-Realization (Table 25), there was a main effect. Lastly, gender was not a moderator of the SD Total Score (Table 26), and it did not have a main effect. For the multiple regressions that indicated gender was a main effect, it was retrained in the subsequent regression models. Gender was dropped from the models predicting Autonomy and the Total Score.

Multiple regression analyses were conducted to examine predictors of self-determination in individuals with FXS. Independent variables included ASD, anxiety, FSIQ, adaptive behavior, and age. In some cases, gender was also included. Five multiple regressions were run to identify predictors of each of the four subdomains of the Arc SDS (i.e., autonomy, self-regulation, psychological empowerment, and self-realization) and the total score.

Table 22. Multiple regression with gender as moderator of Autonomy subdomain Score (n = 50)

		b	Std. Error	β	t	p
1	(Constant)	31.466	13.568		2.319	.025
	Age	.872	.382	.306	2.283	.027
	FSIQ	-.196	.130	-.178	-1.511	.138
	ASD	9.537	6.738	.188	1.415	.164
	Adaptive Behavior	.342	.090	.475	3.784	.000
	Generalized Anxiety	.080	.746	.015	.107	.915
	Social Avoidance	-1.378	.665	-.277	-2.072	.044
2	(Constant)	27.216	15.484		1.758	.086
	Age	.912	.391	.320	2.333	.025
	FSIQ	-.200	.131	-.181	-1.524	.135
	ASD	10.632	7.044	.210	1.509	.139
	Adaptive Behavior	.298	.118	.414	2.529	.015
	Generalized Anxiety	.027	.757	.005	.036	.972
	Social Avoidance	-1.339	.674	-.269	-1.988	.053
	Gender	4.126	7.055	.098	.585	.562
3	(Constant)	35.235	18.610		1.893	.066
	Age	.759	.439	.266	1.728	.093
	FSIQ	-.246	.155	-.222	-1.591	.120
	ASD	11.103	10.339	.219	1.074	.290
	Adaptive Behavior	.228	.132	.316	1.731	.092
	Generalized Anxiety	.247	.962	.045	.257	.799
	Social Avoidance	-1.648	.854	-.331	-1.929	.062
	Gender	4.247	7.856	.101	.541	.592
	Age x Gender	-.063	.903	-.011	-.069	.945
	FSIQ x Gender	.048	.335	.020	.142	.888
	ASD x Gender	-3.879	23.218	-.033	-.167	.868
	Adaptive Behavior x Gender	.184	.268	.101	.685	.497
	Generalized Anxiety x Gender	2.713	2.043	.252	1.328	.193
	Social Avoidance x Gender	-2.481	1.870	-.251	-1.327	.193

Note. R^2 = .42 for Step 1 (p = .00); ΔR^2= .01 for Step 2 (p = .56); ΔR^2 = .07 for Step 3 (p = .60)

Table 23. Multiple regression with gender as moderator of Self-Regulation subdomain Score (n = 47)

		b	Std. Error	β	*t*	*p*
1	(Constant)	1.719	3.518		.489	.628
	Age	-.065	.102	-.096	-.638	.527
	FSIQ	.013	.034	.049	.380	.706
	ASD	-2.431	1.793	-.193	-1.356	.183
	Adaptive Behavior	.085	.023	.501	3.722	.001
	Generalized Anxiety	.099	.200	.077	.497	.622
	Social Avoidance	-.166	.172	-.138	-.966	.340
2	(Constant)	-2.299	3.658		-.629	.533
	Age	-.039	.096	-.058	-.409	.685
	FSIQ	.007	.032	.025	.203	.840
	ASD	-1.340	1.735	-.106	-.772	.445
	Adaptive Behavior	.040	.028	.236	1.446	.156
	Generalized Anxiety	.072	.188	.055	.382	.705
	Social Avoidance	-.136	.162	-.113	-.841	.406
	Gender	4.248	1.671	.428	2.543	.015
3	(Constant)	-4.796	4.126		-1.162	.253
	Age	.013	.104	.020	.130	.898
	FSIQ	.041	.035	.157	1.188	.243
	ASD	-1.574	2.319	-.125	-.679	.502
	Adaptive Behavior	.037	.030	.216	1.232	.227
	Generalized Anxiety	-.125	.224	-.097	-.560	.579
	Social Avoidance	.045	.192	.038	.236	.815
	Gender	3.132	1.740	.315	1.800	.081
	Age x Gender	-.099	.209	-.074	-.471	.641
	FSIQ x Gender	.157	.075	.279	2.105	.043
	ASD x Gender	-3.171	5.165	-.112	-.614	.544
	Adaptive Behavior x Gender	.079	.060	.187	1.314	.198
	Generalized Anxiety x Gender	-.474	.467	-.186	-1.016	.317
	Social Avoidance x Gender	.127	.417	.055	.305	.763

Note. R^2 = .36 for Step 1 (p = .00); ΔR^2= .09 for Step 2 (p = .02); ΔR^2 = .12 for Step 3 (p = .19)

Table 24. Multiple regression with gender as moderator of Psychological Empowerment subdomain Score (n = 50)

		b	Std. Error	β	t	p
1	(Constant)	13.187	1.920		6.867	.000
	Age	-.093	.054	-.233	-1.725	.092
	FSIQ	-.035	.018	-.222	-1.878	.067
	ASD	-2.096	.954	-.295	-2.198	.033
	Adaptive Behavior	.047	.013	.460	3.646	.001
	Generalized Anxiety	.157	.106	.206	1.491	.143
	Social Avoidance	-.146	.094	-.208	-1.546	.129
2	(Constant)	9.522	1.841		5.171	.000
	Age	-.059	.046	-.148	-1.274	.210
	FSIQ	-.037	.016	-.241	-2.403	.021
	ASD	-1.151	.838	-.162	-1.374	.177
	Adaptive Behavior	.009	.014	.087	.632	.531
	Generalized Anxiety	.111	.090	.146	1.238	.223
	Social Avoidance	-.112	.080	-.160	-1.399	.169
	Gender	3.558	.839	.602	4.242	.000
3	(Constant)	8.666	2.286		3.791	.001
	Age	-.072	.054	-.180	-1.335	.190
	FSIQ	-.035	.019	-.228	-1.867	.070
	ASD	-.230	1.270	-.032	-.181	.857
	Adaptive Behavior	.014	.016	.137	.855	.398
	Generalized Anxiety	.154	.118	.201	1.301	.202
	Social Avoidance	-.113	.105	-.161	-1.074	.290
	Gender	3.766	.965	.637	3.902	.000
	Age x Gender	-.007	.111	-.008	-.061	.952
	FSIQ x Gender	-.006	.041	-.018	-.147	.884
	ASD x Gender	3.413	2.852	.206	1.197	.239
	Adaptive Behavior x Gender	.026	.033	.101	.783	.439
	Generalized Anxiety x Gender	-.049	.251	-.032	-.194	.847
	Social Avoidance x Gender	-.052	.230	-.038	-.228	.821

Note. R^2 = .42 for Step 1 (p = .00); ΔR^2= .18 for Step 2 (p = .00); ΔR^2 = .02 for Step 3 (p = .91)

Table 25. Multiple regression with gender as moderator of Self-Realization subdomain Score (n = 50)

		b	Std. Error	β	t	p
1	(Constant)	8.032	1.997		4.023	.000
	Age	.022	.056	.055	.389	.699
	FSIQ	-.007	.019	-.046	-.371	.712
	ASD	-.809	.992	-.115	-.816	.419
	Adaptive Behavior	.051	.013	.511	3.859	.000
	Generalized Anxiety	-.098	.110	-.129	-.889	.379
	Social Avoidance	.016	.098	.024	.168	.867
2	(Constant)	4.978	2.056		2.421	.020
	Age	.050	.052	.127	.969	.338
	FSIQ	-.010	.017	-.062	-.548	.587
	ASD	-.022	.935	-.003	-.023	.982
	Adaptive Behavior	.020	.016	.198	1.267	.212
	Generalized Anxiety	-.136	.101	-.180	-1.351	.184
	Social Avoidance	.044	.089	.064	.496	.622
	Gender	2.965	.937	.506	3.165	.003
3	(Constant)	5.046	2.513		2.008	.052
	Age	.039	.059	.098	.657	.515
	FSIQ	-.022	.021	-.141	-1.043	.304
	ASD	.983	1.396	.139	.704	.486
	Adaptive Behavior	.022	.018	.220	1.243	.222
	Generalized Anxiety	-.077	.130	-.102	-.593	.557
	Social Avoidance	-.046	.115	-.067	-.401	.691
	Gender	3.559	1.061	.607	3.355	.002
	Age x Gender	-.174	.122	-.220	-1.425	.163
	FSIQ x Gender	-.032	.045	-.098	-.716	.479
	ASD x Gender	1.265	3.135	.077	.404	.689
	Adaptive Behavior x Gender	.001	.036	.005	.038	.970
	Generalized Anxiety x Gender	.376	.276	.251	1.364	.181
	Social Avoidance x Gender	-.147	.253	-.106	-.581	.565

Note. R^2 = .36 for Step 1 (p = .00); ΔR^2= .12 for Step 2 (p = .00); ΔR^2 = .04 for Step 3 (p = .77)

Table 26. Multiple regression with gender as moderator of SD Total Score (n = 50)

		b	Std. Error	β	t	p
1	(Constant)	55.433	15.778		3.513	.001
	Age	.701	.444	.197	1.577	.122
	FSIQ	-.238	.151	-.172	-1.571	.124
	ASD	3.897	7.836	.062	.497	.621
	Adaptive Behavior	.526	.105	.585	5.005	.000
	Generalized Anxiety	.311	.868	.046	.359	.722
	Social Avoidance	-1.693	.774	-.273	-2.189	.034
2	(Constant)	39.951	17.349		2.303	.026
	Age	.844	.438	.237	1.928	.061
	FSIQ	-.250	.147	-.181	-1.700	.096
	ASD	7.888	7.892	.125	1.000	.323
	Adaptive Behavior	.366	.132	.407	2.773	.008
	Generalized Anxiety	.117	.848	.017	.138	.891
	Social Avoidance	-1.552	.755	-.250	-2.056	.046
	Gender	15.030	7.905	.286	1.901	.064
3	(Constant)	45.704	19.376		2.359	.024
	Age	.752	.494	.211	1.522	.137
	FSIQ	-.282	.165	-.204	-1.706	.097
	ASD	10.282	10.593	.163	.971	.338
	Adaptive Behavior	.282	.149	.314	1.889	.067
	Generalized Anxiety	.048	1.049	.007	.046	.964
	Social Avoidance	-1.596	.903	-.257	-1.767	.086
	Gender	14.795	9.620	.282	1.538	.133
	Age x Gender	-.333	1.019	-.048	-.327	.746
	FSIQ x Gender	.163	.378	.054	.431	.669
	ASD x Gender	-1.690	26.192	-.011	-.065	.949
	Adaptive Behavior x Gender	.295	.303	.146	.973	.337
	Generalized Anxiety x Gender	2.561	2.304	.200	1.111	.274
	Social Avoidance x Gender	-2.520	2.110	-.207	-1.194	.240

Note. R^2 = .50 for Step 1 (p = .00); ΔR^2= .04 for Step 2 (p = .06); ΔR^2 = .05 for Step 3 (p = .67)

Autonomy

Multiple regression analysis was used to test if individual characteristics (i.e., age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted scores on the

Autonomy subdomain (Table 27). The results of the regression indicated that the model explained 42% of the variance. Social avoidance, adaptive behavior, and age significantly predicted Autonomy scores. FSIQ, autism status, and generalized anxiety were not significant predictors. As age and adaptive behavior increased, autonomy increased. As social avoidance increased, autonomy decreased.

Table 27. Multiple regression predicting Autonomy subdomain score (n = 50)

	b	*SE b*	*β*	*t*	*p*
(Constant)	31.466	13.568		2.319	.025
Age	.872	.382	.306	2.283	**.027**
FSIQ	-.196	.130	-.178	-1.511	.138
ASD	9.537	6.738	.188	1.415	.164
Adaptive Behavior	.342	.090	.475	3.784	**.000**
Generalized Anxiety	.080	.746	.015	.107	.915
Social Avoidance	-1.378	.665	-.277	-2.072	**.044**

Self-Regulation

Multiple regression analysis was used to test if individual characteristics (i.e., gender, age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted scores on the Self-Regulation subdomain (Table 28). The results of the regression indicated that the model explained 36% of the variance. Gender significantly predicted self-regulation scores. Females had higher scores on the Self-Regulation domain. Age, autism status, FSIQ, adaptive behavior, generalized anxiety, and social avoidance were not significant predictors of self-regulation. The lack of significant variables may have been due to the fact that many of the individuals with more impairments (e.g., very low FSIQ, and/or ASD) may not have been administered the SDS.

Table 28. Multiple regression predicting Self-Regulation subdomain Score (n = 47)

	b	*SE b*	*β*	*t*	*p*
(Constant)	1.949	3.301		0.59	0.558
Gender	4.248	1.671	0.428	2.543	**0.015**
Age	-0.039	0.096	-0.058	-0.409	0.685
FSIQ	0.007	0.032	0.025	0.203	0.84
ASD	-1.34	1.735	-0.106	-0.772	0.445
Adaptive Behavior	0.04	0.028	0.236	1.446	0.156
Generalized Anxiety	0.072	0.188	0.055	0.382	0.705
Social Avoidance	-0.136	0.162	-0.113	-0.841	0.406

Psychological Empowerment

Multiple regression analysis was used to test if individual characteristics (i.e., gender, age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted scores on the Psychological Empowerment subdomain (Table 29). The results of the regression indicated that the model explained 59% of the variance. Gender and FSIQ significantly predicted scores on the Psychological Empowerment subdomain. Age, adaptive behavior, ASD, generalized anxiety, and social avoidance were not significant predictors. Females and individuals with higher FSIQ reported higher levels of psychological empowerment.

Table 29. Multiple regression predicting Psychological Empowerment subdomain Score (n = 50)

	b	*SE b*	*β*	*t*	*p*
(Constant)	9.522	1.841		5.171	0.000
Gender	3.558	0.839	0.602	4.242	**0.000**
Age	-0.059	0.046	-0.148	-1.274	0.21
FSIQ	-0.037	0.016	-0.241	-2.403	**0.021**
ASD	-1.151	0.838	-0.162	-1.374	0.177
Adaptive Behavior	0.009	0.014	0.087	0.632	0.531
Generalized Anxiety	0.111	0.09	0.146	1.238	0.223
Social Avoidance	-0.112	0.08	-0.16	-1.399	0.169

Self-Realization

Multiple regression analysis was used to test if individual characteristics (i.e., gender, age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted scores on the Self-Realization subdomain (Table 30). The results of the regression indicated that the model explained 48% of the variance. Gender significantly predicted scores on the Self-

Realization subdomain. Age, FSIQ, adaptive behavior, ASD, anxiety, and social avoidance were not significant. As daily living skills increased, self-realization increased. Females had higher levels of self-realization (i.e., self-awareness of strengths and weaknesses).

Table 30. Multiple regression predicting Self-Realization subdomain Score (n = 50)

	b	*SE b*	*β*	*t*	*p*
(Constant)	4.978	2.056		2.421	0.020
Gender	2.965	0.937	0.506	3.165	**0.003**
Age	0.05	0.052	0.127	0.969	0.338
FSIQ	-0.01	0.017	-0.062	-0.548	0.587
ASD	-0.022	0.935	-0.003	-0.023	0.982
Adaptive Behavior	0.02	0.016	0.198	1.267	0.212
Generalized Anxiety	-0.136	0.101	-0.18	-1.351	0.184
Social Avoidance	0.044	0.089	0.064	0.496	0.622

SD Total Score

Multiple regression analysis was used to test if individual characteristics (i.e., age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted scores on the Self-Determination Total Score (Table 31). The results of the regression indicated that the model explained 50% of the variance. Social avoidance significantly predicted total SD scores, as did adaptive behavior. FSIQ, age, autism status, and generalized anxiety, were not significant. As social avoidance increased, total SD scores decreased. Individuals with more daily living skills had higher total SD scores.

Table 31. Multiple regression predicting SD Total Score (n = 50)

	b	*SE b*	*β*	*t*	*p*
(Constant)	55.433	15.778		3.513	.001
Age	.701	.444	.197	1.577	.122
FSIQ	-.238	.151	-.172	-1.571	.124
ASD	3.897	7.836	.062	.497	.621
Adaptive Behavior	.526	.105	.585	5.005	**.000**
Generalized Anxiety	.311	.868	.046	.359	.722
Social Avoidance	-1.693	.774	-.273	-2.189	**.034**

Question 9. *To what extent do individual ASD, anxiety, intellectual functioning, adaptive behavior, and age predict parent-reported self-determination?*

First, a multiple regression analysis was used to explore whether gender moderates the effects of age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior as predictors of parent-reported behaviors of self-determination. After centering the variables and creating interaction variables, the six predictors and the interaction were entered into a simultaneous regression model. Results suggested that gender did not moderate parent-reported skills of self-determination nor was there a gender main effect (Table 32). As a result, gender was dropped from the model used in subsequent analyses.

Multiple regression analysis was used to test if child characteristics (i.e., age, FSIQ, ASD, anxiety, social avoidance, and adaptive behavior) significantly predicted parent-reported behaviors of SD (Table 33). The results of the regression indicated that the model explained 39% of the variance. Adaptive behavior significantly predicted parent-reported SD behaviors. FSIQ, age, autism status, generalized anxiety, and social avoidance were not significant. As adaptive behavior increased, parent-reported self-determination increased.

Table 32. Multiple regression with gender as moderator of SD Behavior Subscale Score (n = 73)

		b	Std. Error	β	t	p
1	(Constant)	11.157	3.364		3.317	0.001
	Age	0.042	0.104	0.042	0.407	0.685
	ASD	-0.285	1.789	-0.018	-0.159	0.874
	Adaptive Behavior	0.133	0.024	0.578	5.443	0.000
	Generalized Anxiety	0.082	0.179	0.05	0.457	0.649
	Social Avoidance	-0.318	0.187	-0.182	-1.696	0.095
	FSIQ	-0.034	0.033	-0.1	-1.018	0.312
2	(Constant)	9.126	3.75		2.434	0.018
	Age	0.054	0.104	0.054	0.519	0.605
	ASD	0.025	1.801	0.002	0.014	0.989
	Adaptive Behavior	0.112	0.03	0.488	3.765	0.000
	Generalized Anxiety	0.076	0.178	0.046	0.429	0.670
	Social Avoidance	-0.296	0.187	-0.17	-1.58	0.119
	FSIQ	-0.04	0.034	-0.117	-1.184	0.241
	Gender	2.245	1.859	0.155	1.207	0.232
3	(Constant)	-0.397	11.423		-0.035	0.972
	Age	0.524	0.331	0.526	1.583	0.119
	ASD	0.236	7.776	0.015	0.03	0.976
	Adaptive Behavior	0.121	0.096	0.527	1.26	0.213
	Generalized Anxiety	-1.264	0.617	-0.766	-2.049	0.045
	Social Avoidance	0.882	0.621	0.506	1.419	0.161
	FSIQ	-0.024	0.116	-0.071	-0.207	0.836
	Gender	2.218	2.197	0.153	1.01	0.317
	Age x Gender	-0.395	0.24	-0.574	-1.649	0.105
	ASD x Gender	0.756	6.977	0.057	0.108	0.914
	Adaptive Behavior x Gender	0.003	0.067	0.022	0.049	0.961
	Generalized Anxiety x Gender	1.09	0.477	0.97	2.285	0.026
	Social Avoidance x Gender	-1.03	0.487	-0.851	-2.113	0.039
	FSIQ x Gender	-0.723	1.982	-0.129	-0.365	0.717

Note. R^2 = .39 for Step 1 (p = .00); ΔR^2= .01 for Step 2 (p = .23); ΔR^2 = .06 for Step 3 (p = .34)

Table 33. Multiple regression predicting SD Behavior Subscale Score (n = 73)

	b	*SE b*	β	*t*	*p*
(Constant)	11.157	3.364		3.317	0.001
Age	0.042	0.104	0.042	0.407	0.685
FSIQ	-0.034	0.033	-0.100	-1.018	0.312
ASD	-0.285	1.789	-0.018	-0.159	0.874
Adaptive Behavior	0.133	0.024	0.578	5.443	**0.000**
Generalized Anxiety	0.082	0.179	0.05	0.457	0.649
Social Avoidance	-0.318	0.187	-0.182	-1.696	0.095

Question 10. *To what extent do gender, age, intellectual functioning, adaptive behavior, ASD, and anxiety predict parent-reported importance of self-determination?*

Due to the significantly negatively skewed data on parent perceptions of the importance of SD and lack of variability, multiple regression analysis were not conducted to examine predictors (Figure 14). Rather, several independent *t*-tests were conducted to examine if parents who rated all SD skills as "very important" differed in any way from parents who did not rate all skills as "very important." Several variables were explored, including child gender, age, FSIQ, adaptive behavior, autism spectrum disorder, general anxiety, social avoidance, and parent ratings of their child's skill level. Additionally, parent variables, including caregiver education, income, and level of public assistance (e.g., Medicare) were examined. Of 82 participants (3 were excluded because their subscale could not be calculated), 31 parents (37%) rated all SD skills as "very important." Each of the t-tests and chi-square tests compared the parents who rated all skill items in Section I as "very important," equivalent to a score of 14, to parents who did not rate all items as "very important" (i.e., scores of 13 and lower). Table 34 provides descriptive statistics and *t*-test results and table 35 lists chi-square results.

Table 34. Descriptive Statistics and t-tests for Importance of SD subscale (*n* =85)

Variables	*n*	*M*	*SD*	*t*	*df*	*p*
Age				1.62	83	.11
Total score = 14	34	22.21	7.05			
Total score < 14	51	19.80	6.47			
FSIQ				-.60	77	.55
Total score = 14	29	54.17	20.03			
Total score < 14	50	57.12	21.59			
Adaptive Behavior				2.96	77	.00*
Total score = 14	31	64.65	33.90			
Total score < 14	48	44.60	26.02			
Generalized Anxiety				.44	55	.66
Total score = 14	34	6.50	5.02			
Total score < 14	51	6.06	3.55			
Social Avoidance				-.24	83	.81
Total score = 14	34	5.71	4.33			
Total score < 14	51	5.94	4.49			
SD Behavior Subscale				2.20	78	.03*
Total score = 14	31	16.32	7.63			
Total score < 14	49	13.06	5.90			

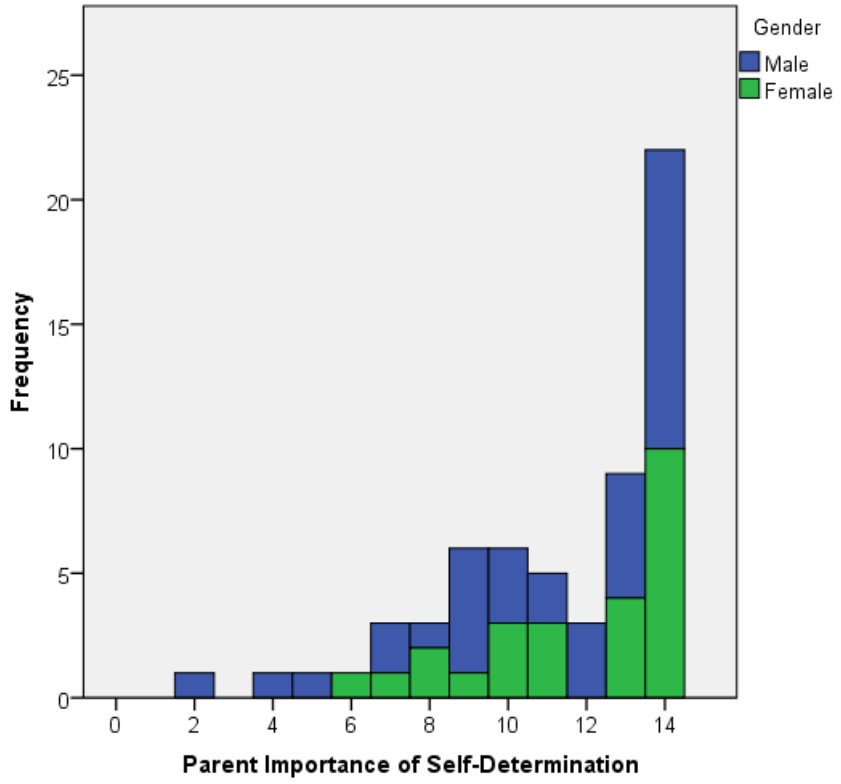
Table 35. Chi-square Child Characteristics and Importance of SD subscale (*n* =78)

	Importance of SD			
	Score = 14	Scores < 14	χ^2	*p*
Age			2.85	.09
Adolescents	8	23		
Adults	21	26		
Gender			1.35	.25
Males	18	36		
Females	13	15		
ASD Status			.64	.43
ASD	7	16		
No ASD	22	33		

Child characteristics. A chi-square test of independence was performed to examine the relation between gender and ratings of "very important" on the Importance of SD subscale. There was no significant association between gender and Importance of SD, (Figure 15), nor was there a significant difference in the age of child for parents who rated all skills as "very important". A chi-square test of independence was also performed to examine the relation between adolescents and adults and ratings of "very important" on the Importance of SD

subscale. There were not differences in Importance of SD subscale scores for adolescents and adults. FSIQ did not affect parent ratings of importance of SD. A chi-square test of independence was conducted to examine the relationship between autism status and ratings of "very important" on the Importance of SD subscale. There were not differences in Importance of SD subscale scores for individuals with ASD and those without an ASD diagnosis. No significant difference was found in the child's parent-reported generalized anxiety for parents who rated all items as very important. There was also not a significant difference in the child's parent-reported social avoidance for parents who rated all items as very important. The only significant difference between parents who rated all skills as "very important" and those who did not was their child's level of adaptive functioning. Lastly, parents who rated their children's SD higher were more likely to rate all skills as "very important".

Figure 15. Sample distribution of Importance of SD Subscale by Gender



Parent characteristics. Demographic information, including caregiver education, income, and public assistance and resources received were also examined as possible factors related to differences in parent perceptions of importance of self-determination. Over half (55%) of families who rated all skills as "very important" reported having between one and three public assistance resources, whereas nearly half (43%) of families who did not report all skills as "very important" reported zero resources. Parents who rated all skills as "very important" in comparison to families who did not were similar in range of caregiver education level and income. Frequencies of family income, caregiver education, and public assistance resources are reported in Table 36.

Table 36. Importance of SD frequencies by parent demographics

	Importance of SD	
	Score = 14 N (%)	Scores < 14 N (%)
Public Assistance		
0 resources	8 (26%)	22 (43%)
1 – 3 resources	17 (55%)	15 (29%)
4-6 resources	6 (19%)	10 (20%)
≥ 7 reported assistance/resources	0 (0%)	0 (0%)
Missing	0 (0%)	4 (8%)
Caregiver Education		
Less than High School	0 (0%)	0 (0%)
High School Graduate	0 (0%)	5 (10%)
Additional Training / Some College	10 (32%)	8 (16%)
Associates Degree	2 (7%)	2 (4%)
College Degree/Some Post College	11 (36%)	18 (35%)
Graduate Degree	4 (13%)	12 (24%)
Missing	4 (13%)	6 (12%)
Income		
Less than $35,000	3 (10%)	0 (0%)
$35,000 to $44,999	1 (3%)	1 (2%)
$45,000 to $54,999	1 (3%)	0 (0%)
$55,000 to $64,999	1 (3%)	0 (0%)
$65,000 or more	13 (42%)	32 (63%)
Missing	12 (39%)	18 (35%)

Subscale item descriptive data. Descriptive statistics and frequencies of the seven items that asked parents about the importance of various skills related to self-determination were examined for differences by gender (Tables 37 and 38). Although 37% of families rated all seven

skills as "very important," more parents of females (43%) than males (33%) rated all skills as "very important". Item frequencies were explored to determine if specific skills were more likely to be considered less important for males and females than other skills (i.e., rated as "not important" or "somewhat important"). The two items with the lowest frequency of "very important" parent-ratings for males were Goal-Setting (*n*=25) and Self-Advocacy and Leadership (*n*=27). Similarly, parents of females were also more likely to rate Goal-Setting as less important (*n*=19) than other skills. There were no significant differences between males and females on choice making, decision-making, problem-solving, goal-setting, self-management and self-regulation, or self-awareness and self-knowledge skills. Parents of females were significantly more likely to rate self-advocacy and leadership skills as "very important" than parents of males.

Table 37. Descriptive Statistics for Importance of SD subscale items by gender

	Gender						
	Males (*n*=50)		Females (*n*=29)				
Importance of SD skills	*M*	*SD*	*M*	*SD*	*t*	*df*	*p*
Choice-Making	1.76	.47	1.72	.59	.295	81	.77
Decision-Making	1.65	.65	1.59	.73	.40	81	.69
Problem-Solving	1.70	.54	1.79	.50	-.67	80	.50
Goal-Setting	1.39	.63	1.64	.56	-1.80	80	.08
Self-Advocacy and Leadership	1.35	.73	1.71	.46	-2.74	76.86	.01*
Self-Management and Self-Regulation	1.67	.51	1.82	.39	-1.52	69.01	.13
Self-Awareness and Self-Knowledge	1.56	.57	1.71	.46	-1.36	66.01	.18

Table 38. Frequencies for Importance of SD subscale items by gender

	Gender			
	Males (*n*=54)		Females (*n*=29)	
Importance of SD skills	*f*	%	*f*	%
Choice-Making				
"Not important"	1	1.8%	2	6.7%
"Somewhat important"	11	20.0%	4	13.3%
"Very important"	42	76.4%	23	76.7%
Decision-Making				
"Not important"	5	9.1%	4	13.3%
"Somewhat important"	9	16.4%	4	13.3%
"Very important"	40	72.7%	21	70.0%
Problem-Solving				
"Not important"	2	3.6%	1	3.3%
"Somewhat important"	12	21.8%	4	13.3%
"Very important"	40	77%	23	76.7%
Goal-Setting				
"Not important"	4	7.3%	1	3.3%
"Somewhat important"	25	45.5%	8	26.7%
"Very important"	25	45.5%	19	63.3%
Self-Advocacy and Leadership				
"Not important"	8	14.5%	0	0%
"Somewhat important"	19	34.5%	8	26.7%
"Very important"	27	49.1%	20	66.7%
Self-Management and Self-Regulation				
"Not important"	1	1.8%	0	0%
"Somewhat important"	16	29.1%	5	16.7%
"Very important"	37	67.3%	23	76.7%
Self-Awareness and Self-Knowledge				
"Not important"	2	3.6%	0	0%
"Somewhat important"	20	36.4%	8	26.7%
"Very important"	32	58.2%	20	66.7%

In summary, analyses in aim 3 suggested that while gender did not moderate the relationship between predictor variables and self-reported self-determination, gender was a significant predictor of scores on the Self-Regulation, Psychological Empowerment, and Self-Realization domains on the SDS. In comparison, age, adaptive behavior, and social avoidance predicted scores on the Autonomy domain. Adaptive behavior was the only significant predictor of parent-reported self-determined behaviors. Relatedly, parents of children with more daily living skills were more likely to rate all SD behaviors as "very important."

Aim 4: Determine the relationship between self-reported self-determination and parent-reported behaviors and importance of self-determination in adolescents and adults with FXS.

Question 11. *To what extent do ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender moderate the relationship between self-reported and parent-reported self-determination?*

Child characteristics were examined as moderators of the nature and strength of the relationship between parent-reported and self-reported self-determination. The nature of the relationship was assessed using individual regressions. The strength of the relationship between self- and parent-report was compared for groups (e.g., gender) through correlation coefficients. To assess the effect of continuous variables (e.g., FISQ) on the relationship between parent- and self-reported SD, the continuous variables were split into equal groups of high, medium, and low.

Gender

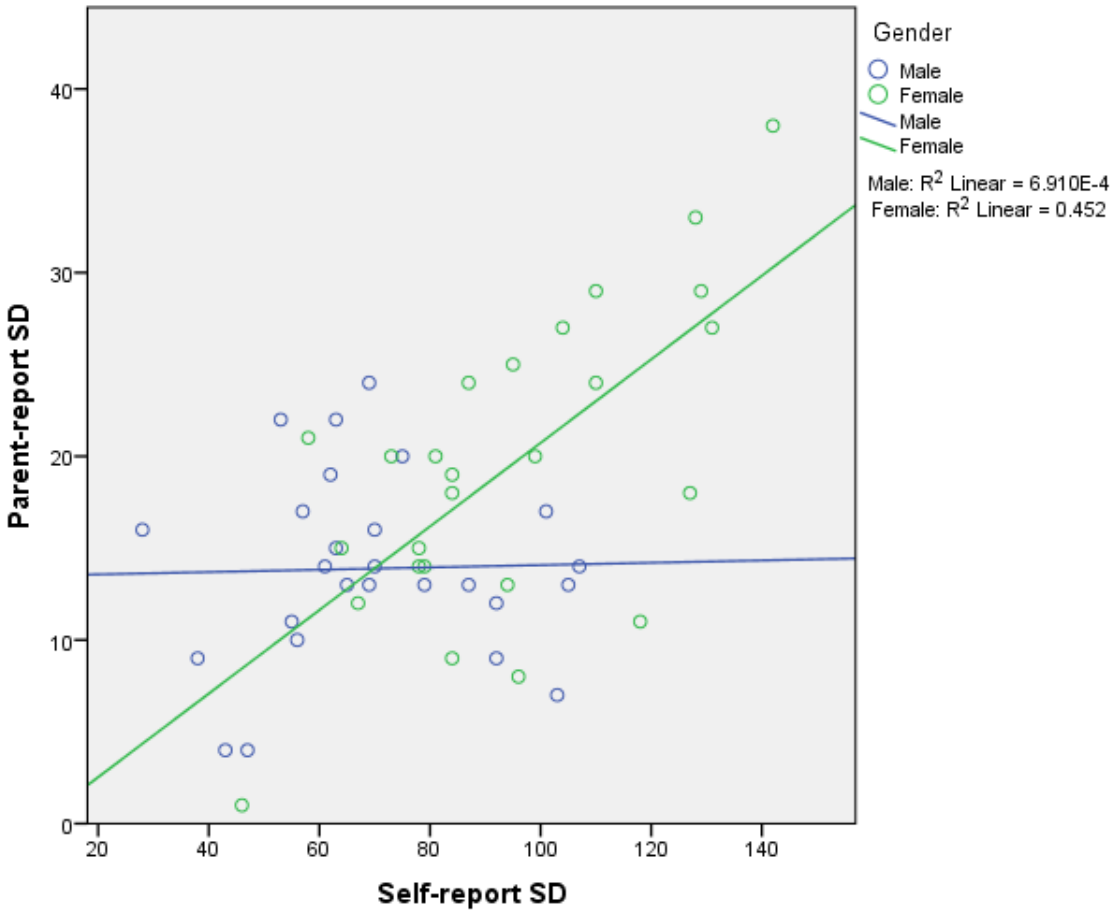
To test if gender moderated the nature of the relationship between parent- and self-reported self-determination, an individual regression analysis was conducted (Table 39; Figure 16). In the first step, parent-reported self-determination and gender were included. These variables accounted for a significant amount of the variance in self-reported self-determination. To avoid multicollinearity, the variables were centered before creating the interaction term, which was added to the model. The interaction variable accounted for a significant proportion of the variance in self-reported SD, suggesting that gender does moderate the relationship between parent- and self-reported self-determination.

Table 39. Linear model of gender as moderator of parent- and self-reported self-determination (*n*=53)

	b	*SE B*	*t*	*p*
Model 1				
Constant	65.42	9.19	7.12	.00
SD Behaviors (Parent-report)	1.48	.43	3.48	.00
Gender	-16.33	6.23	-2.62	.01
Model 2				
Constant	55.60	10.16	5.48	.00
SD Behaviors (Parent-report)	1.99	.48	4.12	.00
Gender	12.50	15.49	.81	.42
SD Behaviors x Gender	-1.88	.93	-2.02	.05

Note. R^2 = .38 for Step 1 (p = .00); ΔR^2= .05 for Step 2 (p = .05)

Figure 16. Scatterplot of parent- and self-reported SD by gender



A two-sample test on correlations was conducted to assess whether gender moderated the strength of the relationship between parent- and self-reported self-determination. For males, self-reported self-determination is negatively correlated with parent-report (n =55; r = -.281), whereas there is a significant positive correlation between parent- and self-report for females (n

= 30; r = .672, p < .01). The two-sample test on correlations z-score of -4.65 indicated that the moderating effect of gender on parent- and self-report is statistically significant. While parents of females rate their self-determination similarly to one another, parents of males rate their children at a range of levels that do not correlate highly with self-reported self-determination.

Age

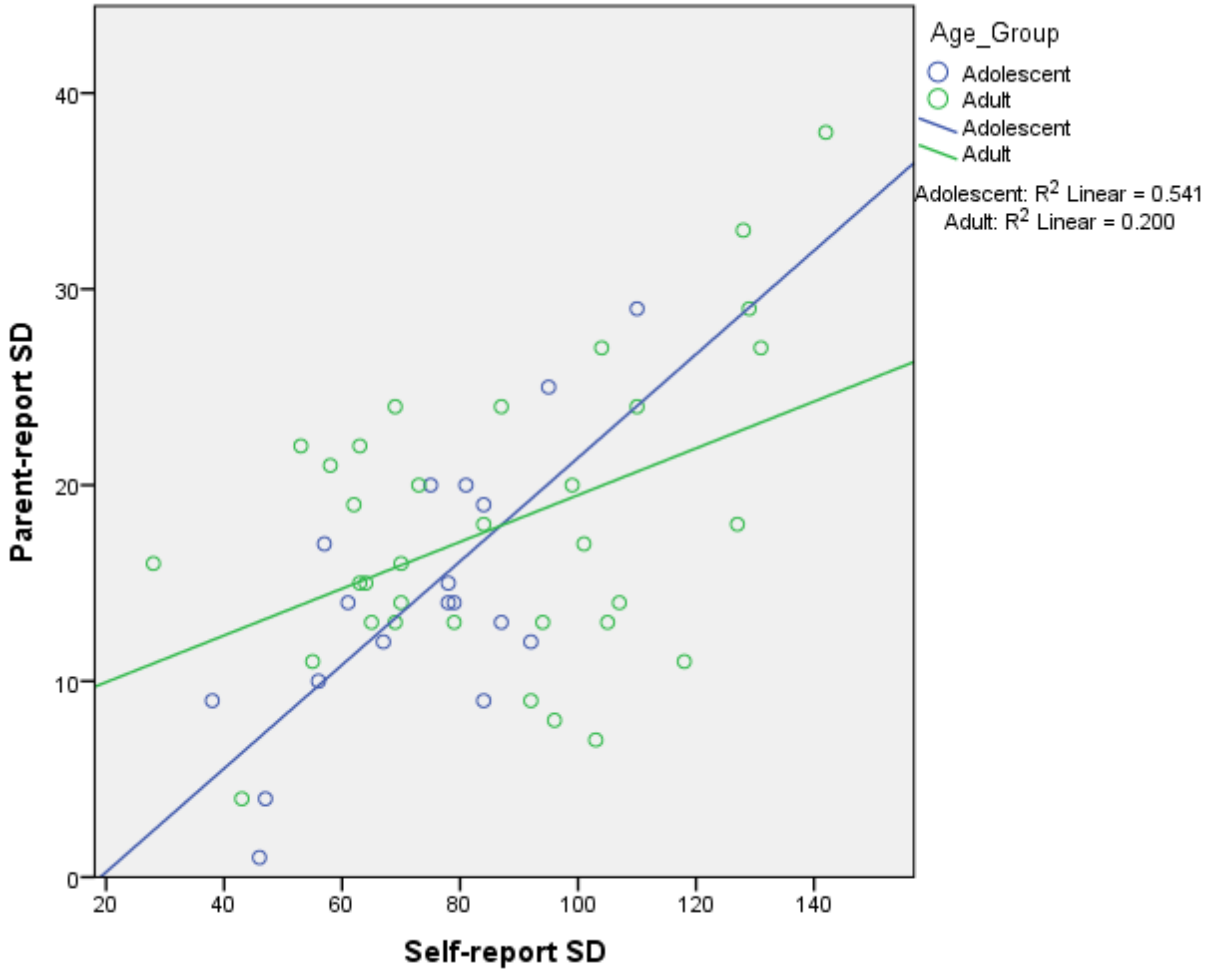
Age was examined as a moderator of the nature and strength of the relationship between parent- and self-reported self-determination by comparing adolescents and adults. An individual regression analysis was conducted. The first step, which included parent-reported self-determination and age group, showed that these variables accounted for a significant amount of the variance in self-reported self-determination (Table 40; Figure 17).The variables were centered before creating the interaction term to avoid multicollinearity which was added to the model in step 2. The interaction variable did not account for a significant proportion of the variance in self-reported SD. As a result, age was not identified as a moderator of the nature of the relationship between self- and parent-reported self-determination.

Table 40. Linear model of age as moderator of parent- and self-reported self-determination (n=53)

	b	*SE B*	*t*	*p*
Model 1				
Constant	38.13	10.82	3.53	.00
SD Behaviors (Parent-report)	1.78	.42	4.30	.00
Age	.64	.41	1.56	.13
Model 2				
Constant	37.53	10.84	3.47	.00
SD Behaviors (Parent-report)	1.89	.43	4.40	.00
Age	.61	.41	1.49	.14
SD Behaviors x Age	-2.89	2.99	-.96	.34

Note. R^2 = .33 for Step 1 (p = .00); ΔR^2= .01 for Step 2 (p = .34)

Figure 17. Scatterplot of parent- and self-reported SD by age group



A two-sample test on correlations was conducted to assess whether age moderated the strength of the relationship between parent- and self-reported self-determination. For adolescents, self-reported self-determination is negatively correlated with parent-report (n =34; r = -.379). However, a significant positive correlation occurred between parent- and self-report for adults (n = 51; r = .448, $p < .01$). The two-sample test on correlations z-score of -3.83 indicated that age significantly moderated the strength of the relationship between parent- and self-report.

FSIQ

To examine whether children's full-scale IQ moderated the nature of the relationship between parent- and self-reported self-determination, an individual regression analysis was

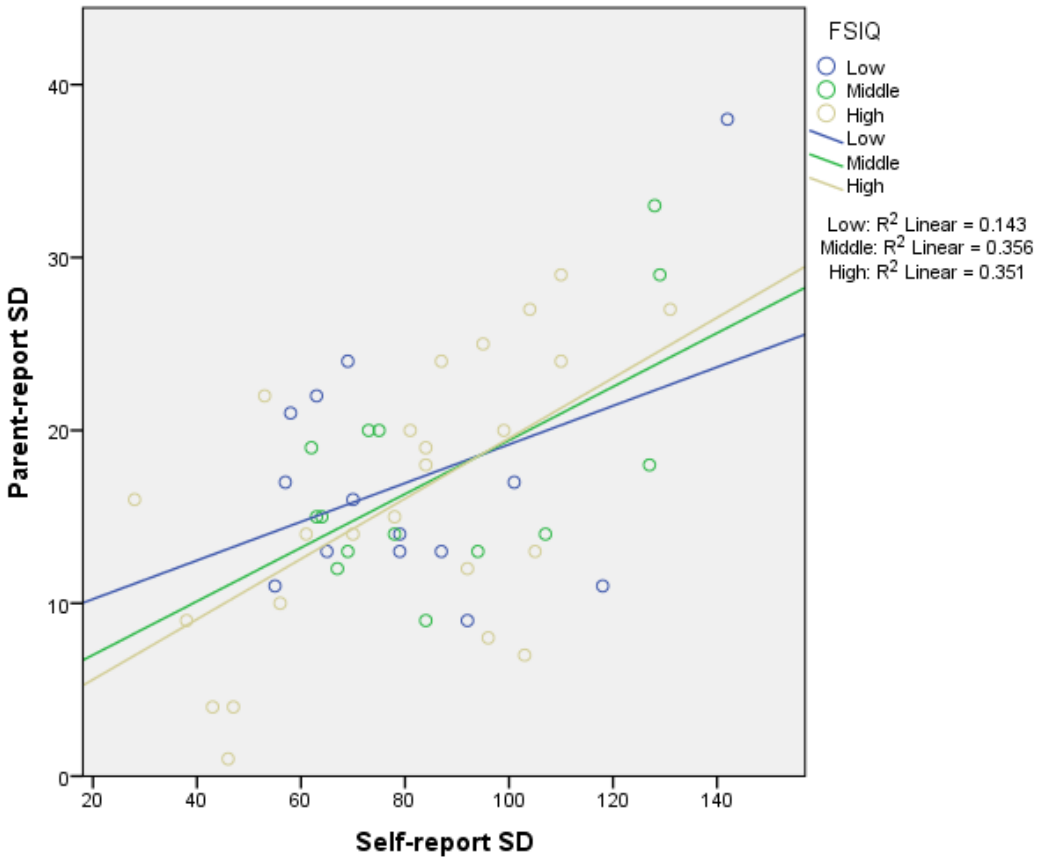
conducted (Table 41; Figure 18). In the first step, parent-reported self-determination and FSIQ were included. These variables accounted for a significant amount of the variance in self-reported self-determination. To avoid multicollinearity, the interaction term was created using centered variables. When added to the model, the interaction variable did not account for a significant proportion of the variance in self-reported SD.

Table 41. Linear model of FSIQ as moderator of parent- and self-reported self-determination

	b	*SE B*	*t*	*p*
Model 1				
Constant	57.75	13.16	4.39	.00
SD Behaviors (Parent-report)	1.81	.46	3.91	.00
FSIQ	-.10	.16	-.62	.54
Model 2				
Constant	63.56	25.70	2.47	.02
SD Behaviors (Parent-report)	1.81	.46	3.88	.00
FSIQ	-.20	.40	-.50	.62
SD Behaviors x FSIQ	.14	.53	.26	.79

Note. R^2 = .26 for Step 1 (p = .00); ΔR^2= .00 for Step 2 (p = .79)

Figure 18. Scatterplot of parent- and self-reported SD by FSIQ



To assess if FSIQ moderated the strength of the relationship between parent- and self-report, a test for the equality of three correlation coefficients was conducted by splitting the sample into low, middle, and high IQ groups. The low IQ group (FSIQ score of 46 or below; n = 27) self- and parent-report was positively correlated, r = .438. In comparison, the middle IQ group (FSIQ scores between 47 and 64; n = 26) had a negative correlation between self- and parent-report (r = -.419), while the high IQ group (FSIQ score ≥ 65; n = 32) had a significant positive correlation between self- and parent-reported self-determination (r = .592, p < .01). The chi-square test indicated that the moderating effect of FSIQ on parent- and self-report was statistically significant, $\chi^2(2, N = 85) = 17.66, p < .01$.

Adaptive behavior

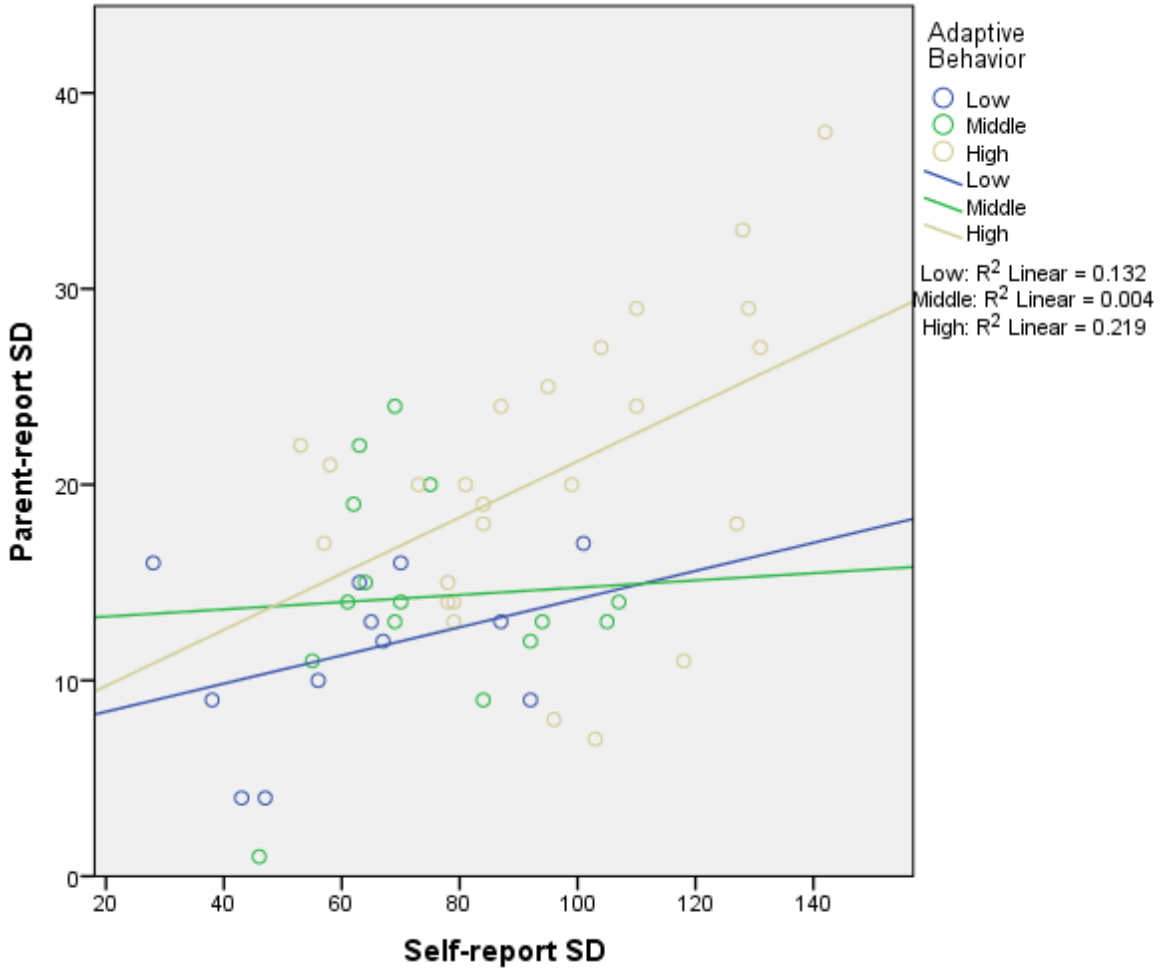
Adaptive behavior was examined as a moderator of the relationship between parent- and self-reported self-determination. To assess the nature of the relationship between parent- and self-report, an individual regression was conducted (Table 42, Figure 19). In the first step, parent-reported self-determination and adaptive behavior were included. These variables accounted for a significant amount of the variance in self-reported self-determination. To avoid multicollinearity, the interaction term was created using centered variables. The interaction variable did not account for a significant proportion of the variance in self-reported SD.

Table 42. Linear model of adaptive behavior as moderator of parent- and self-reported self-determination

	b	*SE B*	*t*	*p*
Model 1				
Constant	44.49	7.55	5.89	.00
SD Behaviors (Parent-report)	.98	.53	1.87	.07
Adaptive behavior	.35	.13	2.68	.01
Model 2				
Constant	55.90	14.90	3.75	.00
SD Behaviors (Parent-report)	.79	.57	1.39	.17
Adaptive behavior	.18	.24	.76	.45
SD Behaviors x Adaptive Beh	.30	.39	.89	.38

Note. R^2 = .39 for Step 1 (p = .00); ΔR^2= .01 for Step 2 (p = .38)

Figure 19. Scatterplot of parent- and self-reported SD by adaptive behavior



To assess if adaptive behavior moderated the strength of the relationship between parent- and self-report, a test for the equality of three correlation coefficients was conducted by splitting the sample into low, middle, and high adaptive behavior groups. The low adaptive behavior group (adaptive behavior scores ≤ 26; n = 26) self- and parent-report was positively correlated, r = .363. In comparison, the middle adaptive behavior group (scores between 27 and 62; n = 30) had a negative correlation (r = -.430) between self- and parent-report, while the high adaptive behavior group (score ≥ 63; n = 29) had a significant positive correlation between self- and parent-reported self-determination (r= .561, p < .01). The chi-square test indicated that the moderating effect of adaptive behavior on parent- and self-report was statistically significant, χ^2

$(2, N = 85) = 17.33, p < .01$.

ASD

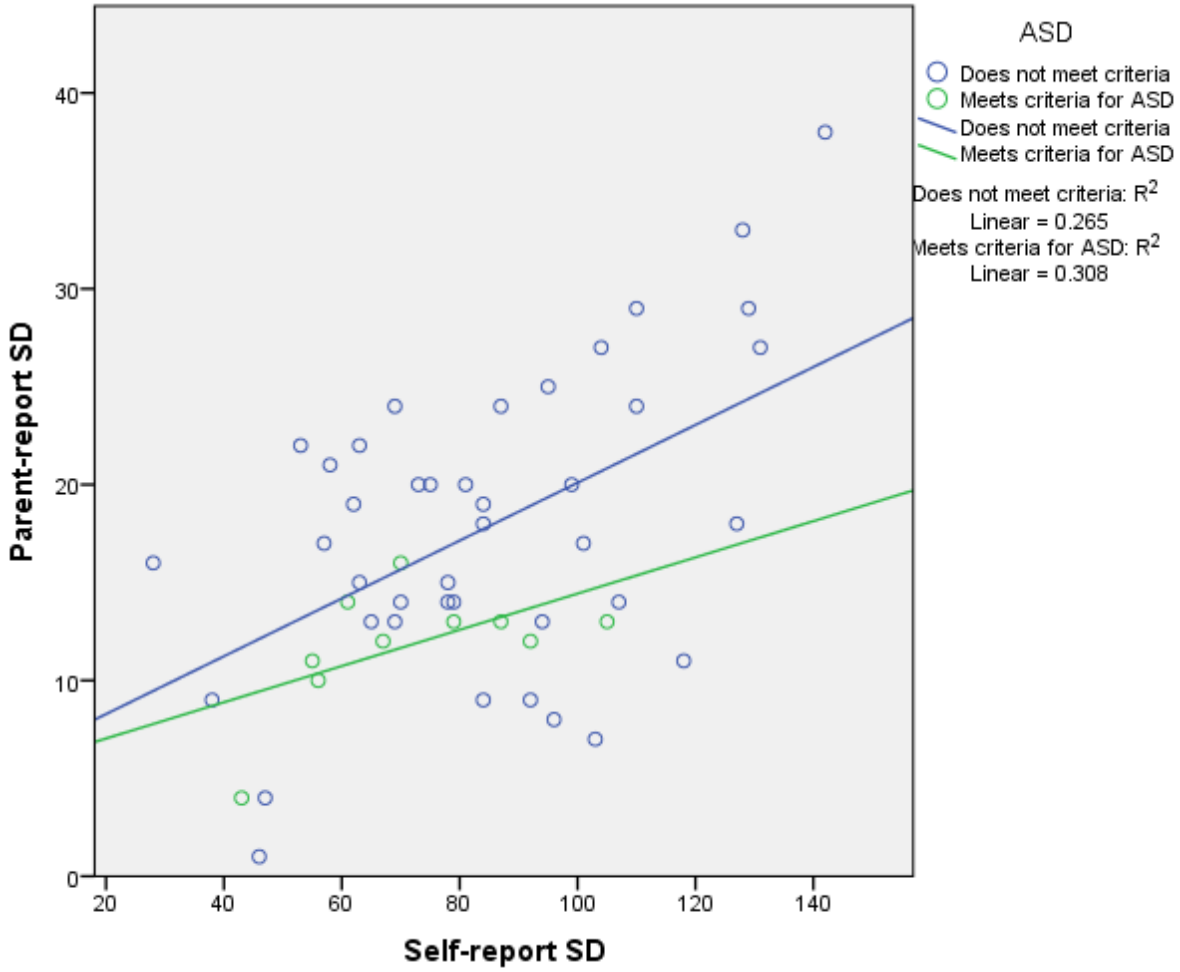
Autism spectrum disorder (ASD) was examined as a moderator of the nature and strength of the relationship between parent- and self-reported self-determination by comparing individuals who met criteria for ASD and those who did not meet criteria. The first step in the individual regression analysis included parent-reported self-determination and ASD status. These variables accounted for a significant amount of the variance in self-reported self-determination, (Table 43; Figure 20). The variables were centered and an interaction term was created, which was added to the model in step 2. The interaction variable did not account for a significant proportion of the variance in self-reported SD.

Table 43. Linear model of ASD as moderator of parent- and self-reported self-determination

	b	*SE B*	*t*	*p*
Model 1				
Constant	51.09	8.61	5.93	.00
SD Behaviors (Parent-report)	1.87	.44	4.20	.00
ASD	-1.63	8.26	-.20	.86
Model 2				
Constant	52.07	8.80	5.91	.00
SD Behaviors (Parent-report)	1.81	.46	3.98	.00
ASD	-19.86	29.79	-.67	.51
SD Behaviors x ASD	1.52	2.38	.64	.53

Note. $R^2 = .29$ for Step 1 ($p = .00$); $\Delta R^2 = .01$ for Step 2 ($p = .53$)

Figure 20. Scatterplot of parent- and self-reported SD by ASD



A two-sample test on correlations was conducted to assess whether ASD status moderates the strength of the relationship between parent- and self-reported self-determination. For individuals who met criteria for ASD, self-reported self-determination was negatively correlated with parent-report (n =23; r = -.481). However, there was a significant positive correlation between parent- and self-report for individuals who did not meet criteria for ASD (n = 58; r = .515, $p < .01$). The two-sample test on correlations z-score of -4.18 indicated that ASD status significantly moderated the strength of the relationship between parent- and self-report.

Anxiety

To examine whether child anxiety moderated the nature of the relationship between

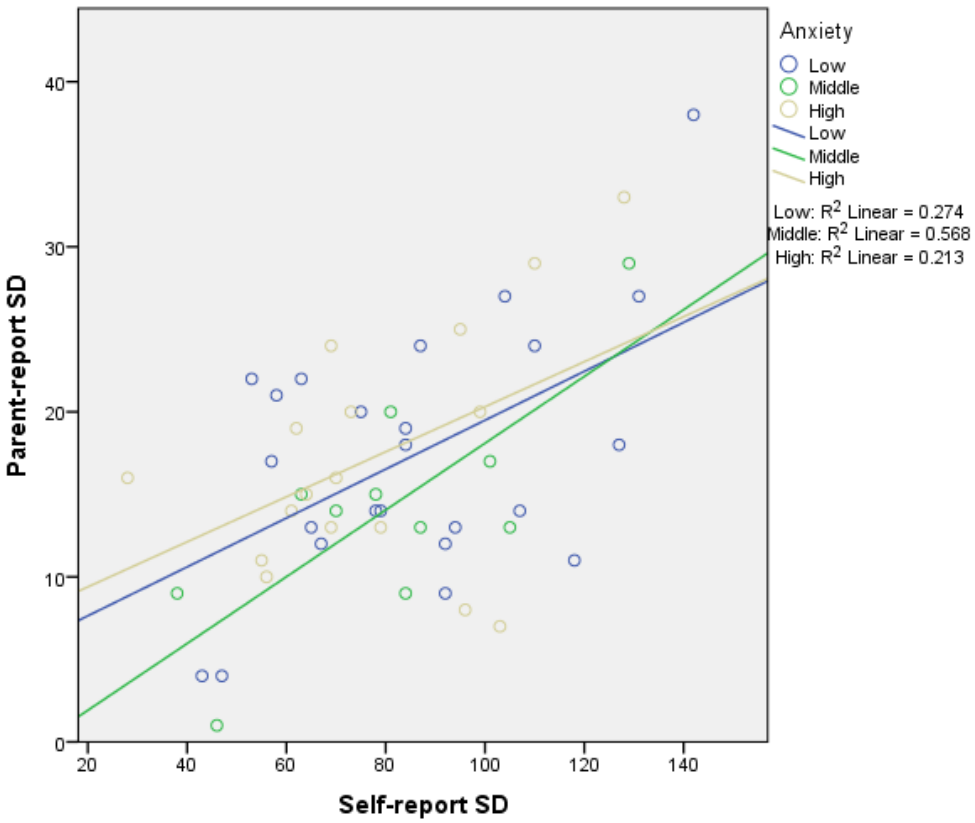
parent- and self-reported self-determination, an individual regression analysis was conducted (Table 44; Figure 21). In the first step, parent-reported self-determination and parent-rated child anxiety were included. These variables accounted for a significant amount of the variance in self-reported self-determination. When the interaction term was added to the model, it did not account for a significant proportion of the variance in self-reported SD.

Table 44. Linear model of anxiety as moderator of parent- and self-reported self-determination (*n* = 53)

	b	*SE B*	*t*	*p*
Model 1				
Constant	49.86	9.31	5.36	.00
SD Behaviors (Parent-report)	1.90	.42	4.48	.00
Anxiety	.07	.77	.08	.93
Model 2				
Constant	41.72	12.46	3.35	.00
SD Behaviors (Parent-report)	1.83	.43	4.26	.00
Anxiety	1.61	1.75	.92	.36
SD Behaviors x Anxiety	-.42	.42	-.98	.33

Note. R^2 = .29 for Step 1 (p = .00); ΔR^2= .01 for Step 2 (p = .33)

Figure 21. Scatterplot of parent- and self-reported SD by anxiety



To assess how child anxiety moderated the strength of the relationship between parent- and self-report, a test for the equality of three correlation coefficients was conducted by splitting the sample into low, middle, and high anxiety groups. The low anxiety group (score of 4 or below; $n = 26$) self- and parent-report was significantly positively correlated ($r = .584$, $p < .05$). The mid anxiety group (scores between 5 and 8; $n = 28$) also had a significant positive correlation between self- and parent-report ($r = .596$, $p < .05$). In comparison, the high anxiety group (score ≥ 8; $n = 31$), had a non-significant negative correlation between self- and parent-reported self-determination ($r = -.350$). The moderating effect of child anxiety on parent- and self-report was statistically significant, $\chi^2(2, N = 85) = 19.26, p < .01$.

Social avoidance

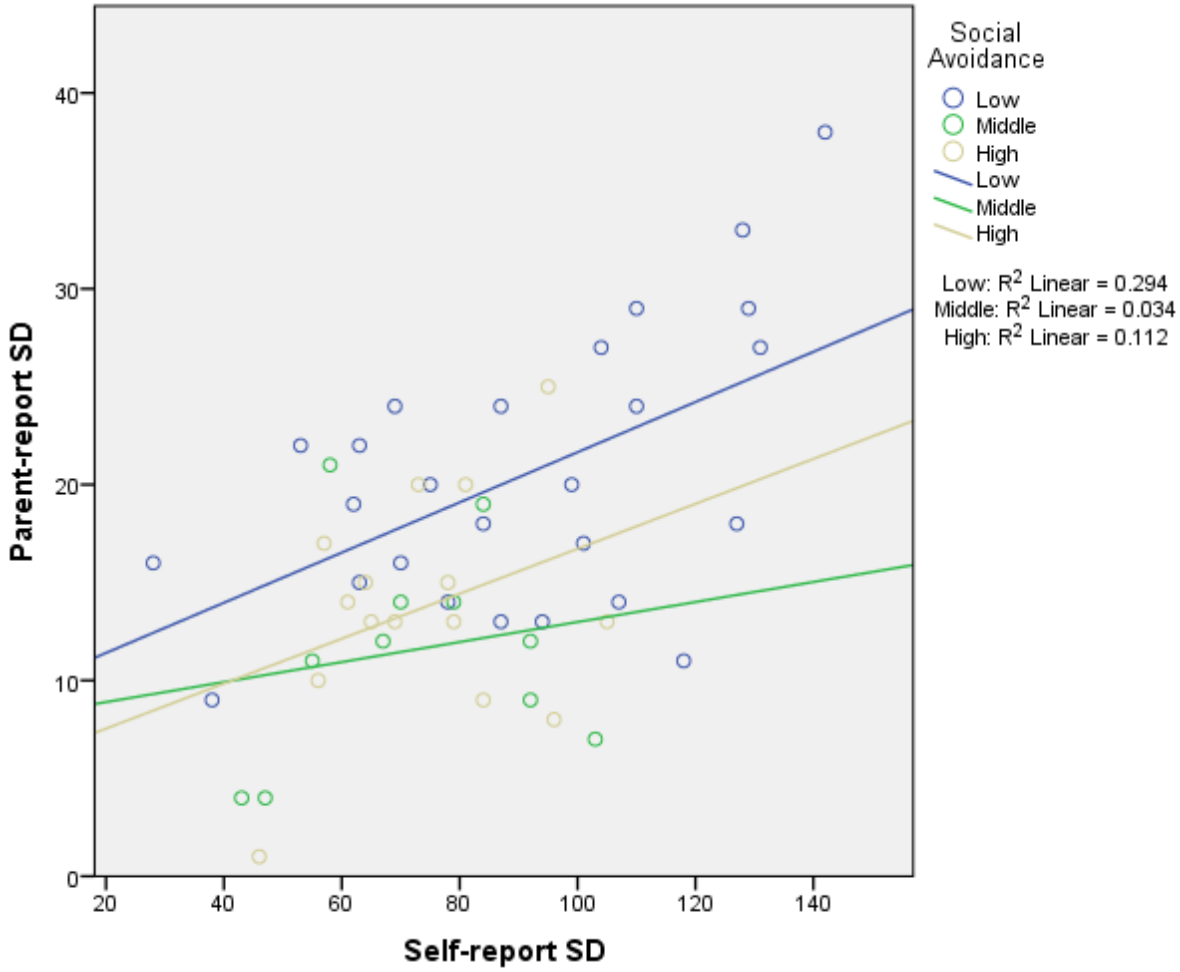
Social avoidance was examined as a moderating variable on the relationship between parent- and self-reported self-determination. To assess the nature of the relationship between parent- and self-report, an individual regression was conducted (Table 45, Figure 22). Parent-reported self-determination and social avoidance accounted for a significant amount of the variance in self-reported self-determination. The interaction between parent-reported self-determination and social avoidance did not account for a significant proportion of the variance in self-reported SD.

Table 45. Linear model of social avoidance as moderator of parent- and self-reported self-determination

	b	*SE B*	*t*	*p*
Model 1				
Constant	59.92	10.48	5.71	.00
SD Behaviors (Parent-report)	1.63	.46	3.53	.00
Social Avoidance	-.93	.71	-1.32	.19
Model 2				
Constant	62.84	10.54	5.96	.00
SD Behaviors (Parent-report)	1.37	.49	2.80	.01
Social Avoidance	-1.08	.71	-1.53	.13
SD Behaviors x Social Avoidance	-4.88	3.29	-1.49	.14

Note. $R^2 = .32$ for Step 1 ($p = .00$); $\Delta R^2 = .03$ for Step 2 ($p = .14$)

Figure 22. Scatterplot of parent- and self-reported SD by social avoidance



To assess if social avoidance moderated the strength of the relationship between parent- and self-report, a test for the equality of three correlation coefficients was conducted by splitting the sample into low, middle, and high social avoidance groups. The low (scores ≤ 3; $n = 23$) and middle (scores 4 through 6, $n = 28$) social avoidance groups' self- and parent-report were significantly positively correlated ($r = .363$, $p < .05$; $r = .612$, $p < .01$, respectively). In comparison, the high social avoidance group (scores ≥ 7; $n = 34$) had a significant negative correlation ($r = -.463$, $p < .05$) between self- and parent-report. The chi-square test suggests that the moderating effect of social avoidance on parent- and self-report is statistically significant, $\chi^2 (2, N = 85) = 19.26, p < .01$.

Question 12. *To what extent does individual ASD, anxiety, intellectual functioning, adaptive behavior, age, and gender moderate the relationship between self-reported self-determination and parent-reported importance of self-determination?*

When the question to explore how child characteristics moderate the relationship between self-reported self-determination and parent-reported importance of self-determination was developed, more variability in parent-reported importance of self-determination was anticipated. Due to a lack of variability in parent-reported ratings of importance of self-determination, the proposed analyses (i.e., six individual regressions) was not be completed.

Aim 4 findings revealed that the nature of the relationship between parent- and self-reported self-determination was not moderated by any child variables (e.g., age, gender, IQ, ASD, adaptive behavior, anxiety, or social avoidance). However, the strength of the relationship between parent- and self-reported self-determination was significantly moderated by each of these variables. There were negative correlations between self- and parent-report for males, adolescents, and individuals with ASD and positive correlations for females, adults and individuals who did not meet criteria for ASD. Individuals in the low or high FSIQ and adaptive behavior groups had positively correlated self- and parent-reported SD, whereas individuals in the mid IQ and adaptive behavior groups had negatively correlated self- and parent-reported SD. Individuals with high levels of anxiety and social avoidance had self- and parent-reported SD that were negatively correlated, whereas individuals with low levels of anxiety and avoidance were positively correlated.

Aim 5: Examine themes among parent-reported supports and barriers that they believe contribute most to the development of self-determination in their adolescent and adult children with FXS. This aim serves to inform the interpretation of quantitative findings from aims 1 through 4.

Question 13. *What themes arise in parent responses to open-ended questions about supports and barriers of the development of self-determination in their adolescent and adult children with FXS?*

The purpose of aim 5 was to explore themes among parent-reported supports and barriers that they believed contribute to self-determination in their adolescent and adult children with FXS. Findings informed interpretation and supplemented findings from the quantitative results in aims 1 through 4.

Coding

Parents were asked to respond to the following two items, "Think about ways in which you encourage any of the skills [specific behaviors related to self-determination] listed above. Briefly share two ideas for how other parents might help their children with disabilities develop any of these skills," and "What do you consider to be the biggest barrier(s) to your child developing any of these skills or becoming more self-determining?" These open-ended questions were the same as those used in Carter et al (2013). Because parents were given the opportunity to provide two separate ideas for each item (i.e., two responses to each prompt), parent responses were often coded and counted in two or more different themes. Additionally, one response from a parent may have been coded for more than one theme. For example, the response, "Communication deficit. Mental deficit," was coded for two explicit themes and counted once in each of the following themes: (a) communication and (b) IQ. In comparison, if a parent provided two responses for supports that were each coded for same theme (e.g., emotional support), their response was counted only once in the emotional support category. For example, one parent wrote "never give up" as her first response, and "praise whatever success you have" as her second response. Each response was coded as emotional support and counted only once in the

emotional support category. In other words, one parent's response was never coded or counted twice in the same theme.

No response. Many parents did not respond to the open-ended questions on the *Self-Determination Parent Questionnaire*. Nearly one-third (*n*=25; 29.41%) of parents did not respond or only partially responded to the prompt about strategies to support self-determination. Of these 25 parents, 17 (68%) parents did not respond to the prompt in its entirety and eight (32%) parents provided one and two responses. Similarly, nearly one-third (*n*=26; 30.5%) of parents did not respond or partially responded to the prompt about barriers to self-determination. Of these 26 parents, 11 (42%) parents did not respond to the prompt in its entirety and 15 (58%) parents provided one and two responses. A total of 10 parents did not respond to either prompt, whereas seven only skipped the supports prompt and one parent skipped the barriers prompt.

Responses not coded. A small number of responses to both prompts were not coded for any theme. These responses were often examples of behavior, an expression or belief, or were not applicable responses to the prompt (e.g., providing a way to support self-determination in response to the prompt about barriers). There were eight parent responses were not coded in the supports prompt and four responses that were not coded in the barrier prompt. Each of these responses were coded as such and can be found in Appendices 3 and 4.

Supports

A total of six themes, three of which included a total of nine subthemes, were identified in parent responses to encouraging self-determination (Table 46). Raters agreed on 89.4% of all responses (total = 170 responses, i.e., two responses per parent). Raters came to consensus on 18 responses. The number of themes identified and coded in each response ranged from zero (i.e., no response) to four (i.e., 1 response with multiple themes identified).

Table 46. Themes in parent responses to encouraging skills related to self-determination

Theme:	Frequency:	Response examples:
Behavioral support		
Behavioral support was defined by parent response that detailed a conventional behavioral strategy (e.g., visual support, positive reinforcement), modeling appropriate behaviors, and expressing demands and expectations for a specific behavior. Any reference of a strategy that would increase a child's skill set was coded here.		
Behavioral strategies	**14**	
		• *"Using social stories."* • *"Visual learning techniques such as an action plan to help understand the outcome/goal."* • *"Prepare for changes – act out or talk out what is to happen."*
Expectations	**2**	
		• *"Encourage and expect involvement in household duties, chores, and responsibilities."* • *"Self-regulation – He can learn to know when it is time to take a shower after the end of the day."*
Modeling	**7**	
		• *"As they got older, I would demonstrate problem solving and decision making by talking through a problem and listing my choices, then the effects of each choice. That way, they could see how it works."* • *"Learn and show how you live."* • *"Lead by example: Think advocacy is important? Go to advocacy day and discuss with your child. Take them to advocacy day."*
Parent does not know	**4**	
Defined by parents who expressed that they did not know how to encourage or support the development of self-determination.		
		• *"I need suggestions!"* • *"Don't know."*
Emotional support	**23**	
Emotional support was coded when parents described love, encouragement, and warm parenting practices as a way to support their child's development of self-determination.		
		• *"Love them a lot."* • *"Encourage activities that are appropriate for their developmental age, to build self-worth."* • *"Tell them their strengths. Emphasize them. Have not really been great at this!"*
External resources	**7**	
External resources was defined by services and supports outside of the home, including schools and community agencies that are needed to support the development of self-determination.		
		• *"Additional schooling to give her needed skills."* • *"The biggest help has been enrolling him in a small school for special needs kids. The atmosphere is one of encouragement and support along with focus on the discipline it takes to succeed."* • *"Advocate for services to support community involvement."*
Opportunities		
Opportunities were coded under two subthemes (i.e., choices, and experiences) when parents responses explicitly stated that they offered their child choices or when they alluded to the importance of having various life experiences as a means of developing self-determination.		

Choice	**17**	
		• *"Talk with your child about things they like to do. Try to offer choices in what they can choose to do so they learn about making decisions/choices."* • *"We encourage him to make his own choices. We give limited number of choices and let him choose."*
Experience	**32**	
		• *"Exposing him to all situations and typical peer settings."* • *"If there is something he really wants, like pizza – ask him to call Dominos and order it, pay for it, ask for it at the counter."* • *"Let him make mistakes, so he can learn from mistakes."*
Skill development		
Skill development was coded when parents referred to supporting specific skills related to self-determination. A total of four subthemes were identified within this theme, including goal-setting, problem-solving, self-advocacy, and self-awareness. Key words, including goal, problem-solve, self-advocacy, and self-awareness were required.		
Goal-setting	**8**	
		• *"Breaking goals down into small attainable steps is helpful and seems to provide feelings of progress and success."* • *"Encourage and teach your child about setting goals for learning skills and use a chart for tracking progress and give rewards to encourage them to continue on."* • *"We help her design a flow chart that lists her goals and ways to achieve them."*
Problem-solving	**8**	
		• *"I encourage him to help problem solve, instead of taking the easy route and solving problems for him."* • *"We often have to revisit decisions he must make. His first response is 'no.' When I don't react, but allow him to calm himself, he can think and problem solve better. It can take time."*
Self-advocacy	**4**	
		• *"Self-advocacy – it is okay to ask for help and be able to communicate to others what makes you feel successful."* • *"Teaching her how to advocate for herself. Doing it for her the first two years of high school, explaining what I was doing. Helping her in her third year if she was not being successful and letting her do it alone her fourth year – helped a lot."*
Self-awareness	**7**	
		• *"We help him understand what he is feeling and then explain (and show) how he might overcome his feelings (i.e., being scared of an event but going and having a good time)."* • *"An ongoing goal is to teach him how to be more self-aware of his behavior and to recognize when he may be getting over-stimulated, how to communicate that effectively in a more productive manner."*
No Strategy	**5**	
No Response	**24**	

Raters also came to consensus on five parents' responses that did not provide a strategy in at least one of the two opportunities to do so. For example, some responses simply described a behavior such as, "he can make his own bed and put clean sheets on his bed." Other responses that were not coded as a strategy to encourage self-determination were more philosophical in nature or described an attitude or disposition such as, "Strive to make yourself the best you can possibly be. You don't need to compete with anyone except yourself!" These responses are labeled "No strategy" in Appendix 3.

Themes. The Behavioral Support theme was comprised of three subthemes; Behavioral Strategies, Expectations, and Modeling. A total of 14 responses were coded as Behavioral Strategies and identified when parents made specific reference to a common technique or strategy that is often used in positive behavioral support. Parents, particularly those of males, made reference to techniques including social stories, visual schedules, and prompting. The Expectations subtheme was used specifically for two responses that suggested parent expectations would help support their child's development of self-determination. Responses within Behavioral Support were also coded as Modeling when parents described how their day-to-day parenting behaviors or explicit demonstrating would help their children develop self-determination. 5% (n= 3) of parents of males and 8% (n=4) of parents of females made reference to modeling as a behavioral support.

The Opportunities theme is comprised of two subthemes, Choice and Experience. Many parents referred to giving their child opportunities to make choices and/or giving them the opportunity to learn from real world experience as a strategy to develop self-determination. 18% (n =10) of parents of males and 23% (n = 7) of parents of females were coded for offering their children choices as a strategy to support self-determination. Experience was the most frequently

coded theme among parents of males (42%; *n* = 23) and parents of females (30%; *n* = 9). Many parents made statements related to exposure, practice, and learning from mistakes that were coded under the subtheme, Experience.

The Skill Development theme consisted of four subthemes, Goal-setting; Problem-solving; Self-advocacy; and Self-awareness. Parent responses were coded under one of the four subthemes in Skill Development when parents specifically referred to a skill related to self-determination. This code required a more elaborate or thoughtful response than simply restating a skill that was listed in previous sections of the questionnaire. For example, the response, "self-regulation – He can learn to know when it is time to take a shower after the end of the day" did not detail how that parent supported self-regulation. In comparison, "Breaking goals down into small attainable steps is helpful and seems to provide feelings of progress and success" suggested that the parent has acted in a certain way to support the development of goal-setting in their child. 2% of parent of males (*n* = 1) and 23% of parents of females (*n* = 7) reported strategies related to Goal-Setting and 7 % of parents of males (*n* = 4) and 13% of parents of females (*n* = 4) reported Problem-Solving skills to support self-determination. 5% of parents of males (*n* = 3) and 13% of parents of females (*n* = 4) provided a response that was specific to helping their child develop Self-awareness. Lastly, 2% of parents of males (*n* = 1) and10% of parents of females (*n* = 3) made reference to helping support Self-advocacy skills in their children.

The remaining three themes coded in the Supports section were Emotional Support, External Resources, and Parent Does Not Know. The Emotional Support theme was coded when parents shared an approach to support self-determination that focused on providing love, encouragement, and warm parenting practices as a method to support self-determination. Emotional support was the second most common theme and was coded for 25% of parents of

males (n = 14) and 30% females (n = 9). 9% of parents of males (n = 5) and 7% of parents of adult females (n = 2) responses referred to External Resources (i.e., services and supports outside of the home, including schools and community agencies) as important for supporting self-determination. Lastly, 7% (n = 4) of parents of males shared that they did not know or needed suggestions on how to support self-determination, which were coded as, "Parent does not know."

Barriers

A total of 12 themes, one of which was comprised of four subthemes, were identified in parent responses as barriers to their child's development of self-determination (Table 47). Raters agreed on 96.47% of all responses (total = 170 responses, i.e., 2 responses per parent). Raters came to consensus on six responses. The number of themes identified and coded in each response ranged from zero (i.e., no response) to four (i.e., one response with multiple themes identified). Of the responses that the raters came to consensus on, four were of parents that did not respond to the prompt appropriately. For example, one response was, "teach by example," which would have been appropriate for the previous prompt on supports, but was not coded in this section. These responses are labeled "No code" in Appendix 4. A total of three parents responded that they did not know what was serving as a barrier to their child's self-determination.

Table 47. Themes in parent responses to barriers related to the development of self-determination

Theme:	**Frequency:**	**Response examples:**
Asking for help	**3**	
Asking for help was coded exclusively when parents reported indicated that asking for help was a specific significant barrier to their child's self-determination.		
		• *"Asking for help when needed. He is extremely shy."* • *"Knowing what she can and can't do and asking for help with things she is weak in."*
Academic skills	**4**	
Academic issues were defined as parent responses that referred to difficulties in reading, writing, and math.		
		• *"Can't read."* • *"Reading and math comprehension."* • *"Can't write down ideas."*
Anxiety	**21**	
Anxiety consisted of parent responses that referred to thoughts, feelings, and behaviors related to anxiety and fear.		
		• *"His anxiety has been the biggest barrier to being confident and independent."* • *"Her anxiety limits her experience."* • *"Anxiety and reactivity can keep him from reaching goals. He reacts strongly to corrective feedback and be offensive in his response, i.e., yelling, making fist."*
Behavior and mood	**12**	
Behavior and mood included parent responses that described difficulties with mood (e.g., emotionality, attitudes) and behaviors (e.g., impulsivity, hyperactivity, perseveration, and disruptive behavior) that impaired their self-determination.		
		• *"Attitude."* • *"There's such a challenge to getting him to see beyond what he wants."* • *"Impulse control."*
Cognitive functioning		
Cognitive functioning included parent responses that referred to various cognitive functions such as executive function (e.g., attention), and understanding. Many responses also referred to overall IQ. The theme of cognitive functioning is comprised of five subthemes listed below.		
Developmental Level	**3**	
		• *"Mental age, maturity, understanding 'life'."* • *"I think most of these skills an be taught over time at varying ages. However, there is a higher level of 'thinking' and 'analysis' that naturally happens from trial and error, therefore, there may be limitations to some skills during maturation."*
Executive function	**6**	
		• *"Does not have the executive functioning skills."* • *"Higher level problem solving skills are always being addressed and worked on by everyone involved in her life."*
IQ	**14**	
		• *"Below normal IQ."* • *"Mental deficits."*
Understanding	**11**	
		• *"Lack of understanding."* • *"He doesn't necessarily understand what he's deciding or why."* • *"Being able to fully understand what is going on and reacting on his own."*
Communication	**11**	
Communication was defined as difficulties with effectively communicating thoughts and feelings through language		

that limited self-determination.	
	• *"Having the right words to use to describe his feelings or decisions."* • *"Lack of language and communication skills."* • *"Lack of expressive language. He can be difficult to understand and he has difficulty expressing his thoughts."*
Confidence and Self-efficacy	**16**
The Self-efficacy and Confidence theme was defined as any parent response that referred to a lack of confidence, self-esteem, or self-efficacy as a barrier, and/or referred to a reliance on others or a need to please others over their own personal preferences that served as a barrier to self-determination.	
	• *"Her reliance on other people to make decisions for her."* • *"No self confidence. Have to keep reassuring them that they can do it… may take time but they can do it."* • *"Self-confidence. Wants to not have FXS and be 'normal'."*
Don't know	**3**
Several parents' responses indicated that they did not know what factors were barriers to their child's development of self-determination.	
	• *"Don't know"* • *"Not sure"*
Lack of resources	**7**
Lack of resources included limited resources or a lack of public resources such as housing, transportation, and employment opportunities, as well as limited supports (e.g., staffing, community agencies).	
	• *"Lack of public resources – transportation, adequate housing, waiting lists!"* • *"Economically, providing services to support growth and experiences."* • *"Community may have a 'transition plan' that is easiest or cheapest and try to mold your child to fit their services. Ignoring personal interest and strengths of child."*
Motivation	**6**
This theme was defined by parents who referred to their child's lack of motivation to acquire new skills related to self-determination and parent's difficulty motivating them. Motivation was often identified in conjunction with anxiety.	
	• *"Lack of initiative."* • *"Don't know how to motivate him."* • *"Fear of failure. Motivation."*
Lack of opportunity	**4**
A lack of opportunity was identified when parents described missed opportunities due to parents limiting opportunities for their child to have an experience that may help with self-determination.	
	• *"The parent enabling them and sheltering them and not allowing the child to grow up or letting go."* • *"His stepfather and I allowing him more freedom."* • *"The lack of employment opportunities that suit his abilities."*
Self-awareness	**6**
Self-awareness was coded when parents specifically identified a lack of insight or awareness as a barrier to the development of self-determination.	
	• *"He is unaware of the extent to which his intellectual disability impairs his ability to function as a normal independent adult."* • *"Goal does not equal skill set. Not completely aware of her own strengths and weaknesses."* • *"Lack of self-awareness and self-monitor skills."*

Themes. Two themes related to emotional and behavior regulation were identified in parent responses; Anxiety, and Behavior and Mood. 16% of parents of males (*n* = 9) and 40% of parents of females (*n* = 12) suggested that thoughts, feelings, and behaviors related to anxiety and fear were among the most significant barriers to their child's self-determination. The Behavior and Mood theme was coded for parent responses that referred to their child's emotionality and challenging behaviors (e.g., perseveration, disruptive behaviors, hyperactivity). 15% of parents of males (*n* = 8) and 13% of parents of females (*n* = 4) identified Behavior and Mood issues as a barrier.

Parents identified a lack of three main characteristics (i.e., Self-awareness, Self-efficacy, and Motivation), which served as significant barriers to self-determination. A lack of self-awareness, or insight to one's strengths and weaknesses, was identified as a barrier to self-determination by 5% of parents of males (*n* = 3) and 10% of parents of females (*n* = 3). A lack of Self-efficacy, Confidence, or Self-esteem was also identified as a significant barrier to self-determination and was shared by 15% of parents of males (*n* = 8) and 27% of parents of females (*n* = 8). 5% of parents of males (*n* = 3) and 10% of parents of females (*n* = 3) identified a lack of motivation as a significant barrier.

Many parents identified poor communication and difficulty asking for help as barriers to self-determination for their children. 16% of parents of males (*n* = 9) and 7% of parents of females (*n* = 2) suggested that their child's difficulty expressing their thoughts and feelings effectively was a significant barrier. 2% of parents of males (*n* = 1) and 7% of parents of females (*n* = 2) specifically recognized difficulty asking for help as a barrier to their child's self-determination.

The Cognitive Functioning theme consists of four subthemes; Executive Function, Developmental Level, IQ, and Understanding. Parent responses were coded under one of the four subthemes in Cognitive Functioning based on how they described the specific difficulties related to cognitive functioning. 2% of parents of males(n = 1) and 7% of parents of females (n = 2) specifically referred to their child's level of maturity, or Developmental Level. 16% of parents of males (n = 9) and 7% of parents of females (n = 2) made reference to their child's level of Understanding as a significant barrier. Many parents suggested that their child's overall IQ was a significant barrier to their ability to develop self-determination (22% of parents of males (n =12); and 7% of parents of females (n = 2)). 5% of parents of males (n = 3) and 10% of parents of females (n = 3) said that deficits in executive function (e.g., attention, problem solving) were barriers to their child's self-determination. 5% of parents of males (n = 3) and 3% of parents of females (n = 1) identified specific academic skills (e.g., reading) as barriers to their child's self-determination.

Lastly, some parents identified barriers that were not intrinsic to their child, but rather referred to an environmental barrier, such as a lack of opportunity or a lack of resources as significant barriers to self-determination. 9% of parents of males (n = 5) and 7% of parents of females (n = 2) identified a Lack of Resources, which included references to public and community resources such as housing, transportation, employment opportunities, and other social supports (e.g., staff and community agencies). 5% of parents of males(n = 3) and 3% of parents of females (n = 1) identified a Lack of Opportunity, which included references to parents sheltering their child or not allowing certain experiences for their child as a significant barrier.

In summary, several themes emerged among parent-reported supports and barriers to self-determination. Some of the most frequent strategies to support self-determination reported by

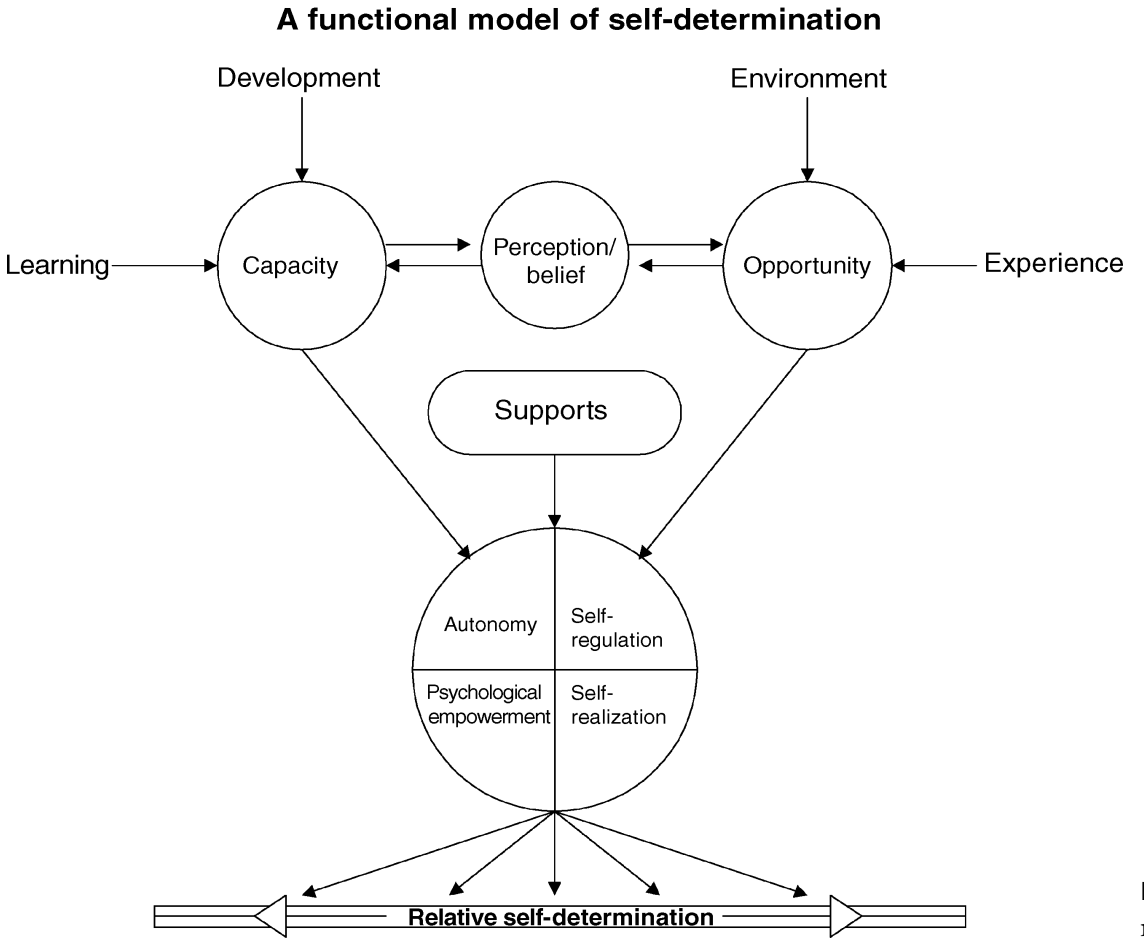
parents included emotional support, behavioral support, and providing opportunities. Fewer parents suggested specific skill development (e.g., self-advocacy), and external resources. Anxiety, self-efficacy, communication, and cognitive functioning were among the most commonly reported barriers to self-determination. Parents also indicated difficulties with asking for help, academic skills, behavior and mood, self-awareness, and motivation as barriers. Some parents referred to a lack of resources or opportunity as significant barriers as well. Approximately one-third of parents only partially responded or did not respond to the open-ended prompts on supports and barriers and a small number of parents explicitly stated that they did not know what served as supports or barriers to self-determination.

CHAPTER 5: DISCUSSION

Defining self-determination

The present study explored self-determination, as defined by the Functional Model of Self-Determination (Figure 23; Wehmeyer, 1996), in adolescents and adults with FXS. Within this model, self-determination is conceptualized as choices and decisions made through causal agency (i.e., the ability to make or cause things to happen) that contribute to one's quality of life (Wehmeyer, Kelchner, & Richards, 1996). Volitional action (i.e., making a conscious choice), which enables one to be the causal agent to maintain or improve quality of life, was later added to be more inclusive toward individuals with more severe impairments, who at one point were considered by some as unable to be self-determined (Wehmeyer, 2005). Recently, Shogren et al. (in press) reconceptualized the functional model's theoretical framework and proposed causal agency theory as an avenue to reframe self-determination. This revised framework is within the context of positive psychology, a strengths-based perspective in disability research, and with a new focus on the fit between capacity and environment (Shogren, 2013). Within the new model, the basic framework of the functional model was maintained and causal agency theory was incorporated to include an emphasis on how a person becomes self-determined and focuses more on human agency (Shogren, Wehmeyer, Palmer, & Forber-Pratt, in press). Aspects of self-determination theory (Deci & Ryan, 2002), which originated in motivational psychology, were also incorporated to emphasize how basic needs motivate causal agency and result in improved wellbeing within a social environment.

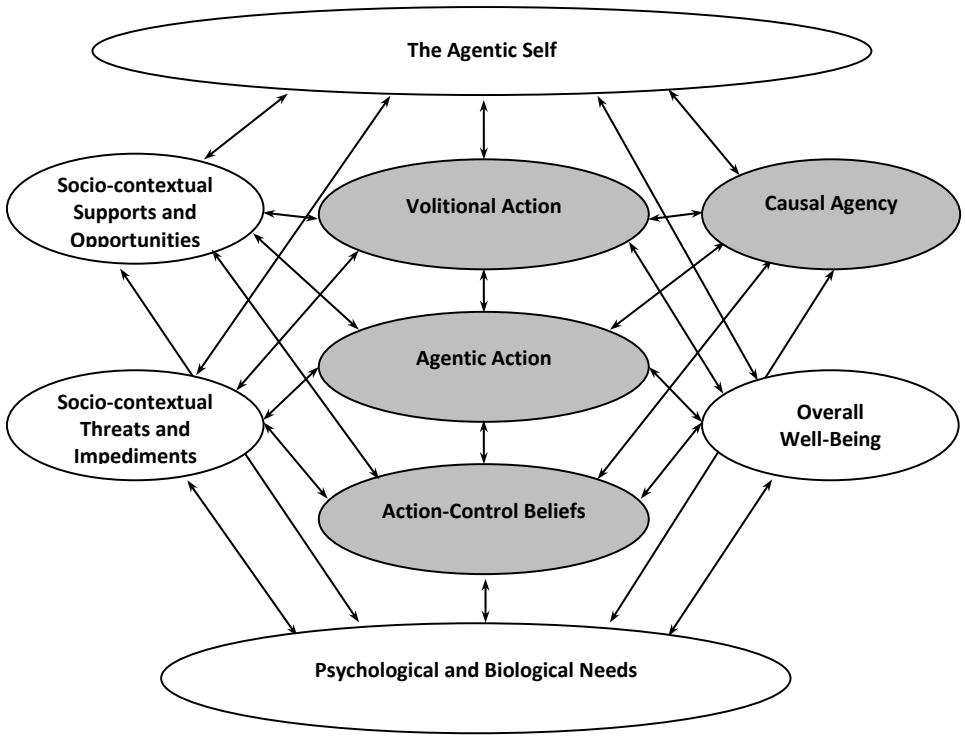
Figure 23. Functional Model of Self-Determination (Wehmeyer et al., 2003).



Under causal agency theory, the essential components from the original functional model (e.g., autonomy and self-realization) have also been revised. What was previously characterized as autonomy is now *volitional action*, defined by self-initiation and actions that function to allow an individual to act autonomously (Shogren, Wehmeyer, Palmer, & Forber-Pratt, in press). Self-regulation is *agentic action*, which is action that is self-directed, self-regulated, and enable individuals to work toward goals and respond to opportunities and challenges. Psychological empowerment and self-realization are encompassed by *action-control beliefs*, which include control expectancy (i.e., a belief related to the self and the goal), capacity beliefs (i.e., belief about self and means to achieve the goal), and causality beliefs (i.e., a belief about the usefulness

of the means to achieve goals). Positive action beliefs enable an individual to act in a manner that is empowered and self-aware. Through these subtle changes, self-determination is characterized less by the specific action or the belief that motivates that action and more by the *function* the action serves, and whether it enables an individual to act as a causal agent (Shogren, Wehmeyer, Palmer, & Forber-Pratt, in press). These multiple layers of human agency as described by the reconceptualized functional model are illustrated in Figure 24.

Figure 24. Layers of human agency



The definition of self-determination was also revised as a dispositional characteristic that allows one to serve as a causal agent (Shogren, Wehmeyer, Palmer, & Forber-Pratt, in press). As a dispositional characteristic, self-determination can fluctuate over time and across contexts and may depend on opportunities and supports. With opportunities to engage in self-determined action, individuals serve as causal agents, have their basic needs met, and as a result, improve

their wellbeing. Overall, Shogren et al. (in press) argue that causal agency theory provides a framework for enhancing supports and focusing on wellbeing. The revision of the functional model informs future interventions and directions for research and compliments findings from the present study, particularly parent perceptions of supports and barriers, which are well aligned with the new emphasis on socio-contextual supports.

Importance of self-determination

Though limited in number, outcome studies on self-determination provide compelling evidence of how significant self-determination is for individuals with intellectual and developmental disabilities (I/DD). In addition to academic outcomes, research studies show that self-determination is linked to more positive independent living and employment outcomes (Martorell, Gutierrez-Rechacha, Pereda, & Ayuoso-Mateos, 2008; Shogren, Wehmeyer, Palmer, Rifenbark, & Little, in press; Wehmeyer & Palmer, 2003; Wehmeyer & Schwartz, 1997), leisure and recreation habits (McGuide & McDonnell, 2008), and more positive quality of life and life satisfaction (Nota, Ferrari, Soresi, & Wehmeyer, 2007; Shogren, Lopez, Wehmeyer, Little, & Pressgrove, 2006; Wehmeyer & Schwartz, 1998) for individuals with I/DD. Specifically for individuals with FXS, self-determination may also be increasingly important because of the increase in clinical trials. Clinical trials are an important consideration because they require individuals to engage in and assent or consent to research studies that could potentially have lasting effects on their cognitive and behavioral presentation and functioning and quality of life. Without sufficient self-determination, individuals with FXS may be less likely to advocate for themselves, particularly regarding research participation.

However, few studies to date have explored self-determination within specific I/DD populations. Significant heterogeneity among individuals diagnosed with intellectual disability,

autism spectrum disorder, and genetic syndromes (e.g., FXS) that are captured under developmental disability is well documented in the literature. Moreover, while areas of strength and weakness can be identified between groups, there is also significant variability within groups (e.g., autism spectrum disorder recognized as a spectrum of functioning). As a result, research that examines various characteristics, such as self-determination, would benefit from empirical studies that document functioning within specific I/DD populations. As FXS is the leading hereditary cause of intellectual disability and recognized as the leading genetic cause of ASD, FXS is one such population that would be important to explore specific intra-individual differences in self-determination.

It is important to explore how the wide range of relative strengths and impairments that are characteristic of individuals with FXS might influence self-determination. Finally, exploring self-determination within this population is also very important to determine whether individuals with a known etiologic cause of I/DD have different levels of self-determination resulting purely from understanding the origins of their difficulties and impairments (i.e., attitudes and beliefs about their disability status). Relatedly, parents of individuals with FXS may have different perceptions of self-determination for their child than do parents of children with a developmental disability that does not have a clear etiology.

Predicting self-determination

Adaptive behavior was the most consistent predictor of self-determination in parent- and self-reported self-determination and parents of children with more developed daily living skills were more likely to rate all SD behaviors as "very important." Gender did not moderate the relationship between predictor variables and self-reported self-determination, however it was a significant predictor of scores on the on the Self-Regulation, Psychological Empowerment, and

Self-Realization domains on the SDS. In comparison, age, adaptive behavior, and social avoidance predicted scores on the Autonomy domain.

The significant positive correlation between adaptive behavior and self-reported attitudes and beliefs (i.e., scores on the psychological empowerment and self-realization domains) suggests that all aspects of self-determination can be supported by the development of adaptive behavior, not just autonomy. It is possible that as individuals develop increasing daily living skills, they feel more self-efficacious and as a result more empowered and also aware of skills and tasks for which they need help (i.e., self-realization). These findings suggest that functional skills are essential in order to be able to act autonomously and advocate to have needs met. That adaptive behavior is the strongest predictor and these daily living skills can be taught indicates promising future targets for intervention to increase self-determination in individuals with FXS. However, because causality cannot be determined, it is also possible that individuals with lower self-determination have reduced motivation to improve daily living skills and increase autonomy.

While there is limited research on the trajectory of adaptive skills in individuals with FXS, research shows that daily living skills of individuals with ASD increasingly lag behind same-age peers over time (Carter et al., 1998). Moreover, research suggests that the gap between adaptive behavior and intellectual functioning for individuals with ASD continues to increase into adolescence (Kanne et al., 2011). Children with ASD and with IQ's below 70 developed daily living skills at a slower rate than children with ASD with higher IQ's (Freeman, Del'Homme, Guthrie, & Zhang, 1999). Smith et al. (2012) examined the development of daily living skills and the influence of ID in individuals through adolescence and adulthood with ASD and found that skills continued to improve through early 20's, plateaued and slowly declined in the early 30's. These rates were exacerbated by intellectual disability (i.e., individuals with ASD

and ID had fewer daily living and demonstrated a slower rate of change). In comparison to individuals with Down syndrome, who continue to gain daily living skills throughout adulthood, Smith et al. (2012) proposed that the slowing improvement of skills for individuals with ASD may contribute to poorer adult outcomes for individuals with ASD. The relationship between age and functional skills in adolescents and adults with ASD and ID and ASD may be intimately related to self-determination. Additional research that also includes measures of self-determination is warranted.

Klaiman et al. (2014) found that the most meaningful gains in adaptive behavior for individuals with FXS occurred after 14 years of age, further stressing the importance of interventions in late childhood and early adolescence. Moreover, Hustyi and colleagues (2015) found that for individuals with FXS who were matched to a control group by sex, age, IQ, and autism symptomology had similar levels of independent living skills, the daily living skills of individuals with FXS may be significantly impacted by symptoms of autism. Hustyi et al. (2015) hypothesized that severe social anxiety may be correlated with ASD symptoms and partly account for the poorer daily living skills in individuals with FXS. Similar hypotheses can be made about why other factors, such as autism spectrum disorder and anxiety were not predictors of self-determination in the present study. For instance, although anxiety was not a direct predictor of self-determination in the present study, it is possible that individuals with increased anxiety also experience more challenges with adaptive behavior, which subsequently decreases their self-determination. Similarly, research shows that adolescents and adults with FXS and comorbid ASD have more behavioral and emotional problems, lower levels of adaptive behavior (Smith et al., 2012) and less independent daily life outcomes (Hartley et al., 2011), which further suggests that adaptive behavior may be a mediating factor between other individual differences

and self-determination outcomes. Future research is needed to examine adaptive behavior as a mediating variable.

Relationship between parent- and self-report

The present study also explored the relationship between parent- and self-reported self-determination. Findings indicated that the nature of the relationship between parent- and self-reported self-determination was not moderated by any child variables. However, when differences in the magnitude of the association between parent- and self-report were explored, the data indicated significant differences by all factors. This finding suggests that for part of the sample in the present study, parents and children did not report similar levels of self-determination. The data suggested that males, adolescents, and individuals who met criteria for ASD reported levels of self-determination that were less consistent with their parents. Similarly, individuals with lower IQ, fewer daily living skills, high anxiety, and high social avoidance also had more discrepant reports of self-determination than their parents. Although these findings are limited by the fact that there were roughly twice as many parents who reported on their child's self-determination, they suggest two potential explanations. First, it is possible that individuals who are more impaired may be less able to reliably report on their self-determination. Second, it is possible that parents of children who they perceive as more impaired or less able to be self-determined may underestimate their child's self-determination. Of course, these two possibilities are not mutually exclusive and signify the need for additional research to further explore the relationship between self- and parent-report. At present the only research that might corroborate the second explanation, is Carter's (2013) study that suggested parents of children with more severe impairments (e.g., mild versus moderate to severe intellectual disability) were more likely to report lower levels of self-determination. Because a paucity of research has used both parent-

and self-report measures of self-determination, little is known about the consistency of reporting among parents and children. A review of the findings from the present study suggests that parents and children report comparable self-determined behaviors when children are older, have higher cognitive and adaptive skills, and lower levels of behavioral and emotional difficulties (e.g., anxiety and ASD).

Parent perspectives

Despite the significance of the parent role in the development of self-determination in children with intellectual and developmental disabilities, it is only recently that research has explored the parent role. As a result, a paucity of empirical research that includes parent perspectives of self-determination is available. Wehmeyer (2014) recently recognized the lack of focus on the role that parents play in supporting their child's development of self-determination and decision making and suggested extrapolation from family systems and child development research. The present study adds to the current research on the family role with findings on parent perspectives of the importance of self-determination, as well as parent perspectives on factors that support and challenge the development of self-determination. In comparison, the teacher role in self-determination is further documented, likely because of the initial focus on self-determination in the special education setting (Wehmeyer, 2014).

Importance of self-determination. In the present study, child skill level per parent-report did not influence the degree of importance parents placed on self-determination. Parents reported generally high levels of importance across all self-determined behaviors (e.g., choice making, self-regulation). Parents of males and females nearly equally reported choice making, decision making and problem solving as very important. However, parents of females were more likely to report goal-setting, self-advocacy, self-regulation, and self-awareness as "very important" than

were parents of males." This finding suggests that parents of males may not prioritize higher-order, more advanced skills (e.g., self-awareness) as much as parents of females. This difference may be due to perceptions of their child's intellectual disability or current emotional or behavioral challenges. Because self-determination is a dispositional characteristic that changes over time and experience, longitudinal studies may provide further insight into parent perceptions of importance over time.

In the original study using the SDPQ, more than 60% of parents of younger children (i.e., grades kindergarten through grade 12) who qualified for special education under ID or ASD, rated skills related to self-determination as "somewhat" or "very" important (Carter et al., 2013). This finding suggests that age may not influence parent perceptions of how important self-determination is for their child. In comparison to the present study where the variability in parent ratings of importance was too limited of variability to examine predictors, Carter et al. (2013) found that disability level, intellectual disability, and free and reduced lunch status were significant predictors of importance. Specifically, parents of children who were identified as having a disability in the severe or profound range rated importance of self-determination lower than parents of children who were less severely impaired. Two possible reasons might account for these differences in findings. First, the population in the Carter et al. (2013) study was younger and included more families who had children that qualified for free or reduced lunch, whereas the present study sample consisted primarily of high income and well-educated families. Given the limited research that suggests parents with higher levels of education and higher SES are more likely to give their children with special needs more opportunities to practice skills related to self-determination (Zhang, 2005), families of lower SES (i.e., children that qualify for free and reduced lunch) may have less awareness of self-determination and as a result may not

prioritize these skills for their children to the same degree. Second, the present study sample may be unique in that the parents all share an understanding of the genetic underpinnings of their child's disability, which might positively influence the way parents perceive their child's disability.

Supporting self-determination. The open-ended questions on the SDPQ allowed for parents to directly report what aspects of their child's lives supports and challenges the development of self-determination. Findings from the open-ended questions on the SDPQ suggest that parents have great insight on how to support self-determination. Just as adaptive behavior was the most significant predictor of self-determination based on parent- and self-report, parents frequently reported providing opportunities and experiences for their children as key strategies for improving self-determination. Considering daily living skills are learned behaviors, it makes sense that opportunities and experiences are crucial to the development of adaptive behavior. Whether parents were referring to general skills versus what they perceive to be specific behaviors related to self-determination remains unclear.

Also among the most common themes of support were emotional support and behavioral techniques. Although emotional support and behavioral techniques were not quantifiable measures that could be used as predictor variables in the present study, one can conceive how emotional support, which leads to greater self-esteem and self-efficacy, would support self-determination. Future research is needed to explore the role of emotional support in the development of self-determination for individuals with I/DD.

While the present study appears to be the first to obtain parent perspectives on strategies to support self-determination for their adolescent and adult children, Summers et al. (2014) reported on qualitative data on strategies used to develop self-determined skills (e.g., choice-

making) in young children with disabilities. Families emphasized strategies that supported developing opportunities for choice making, self-regulation, and engagement. They emphasized deliberate multiple opportunities for practice and creating an accessible environment (e.g., structural supports for children with physical disabilities). As the child's first teacher, parents teach self-determined behaviors throughout childhood and as the child develops more independence, the role of the parents does not diminish, rather it changes in nature (Palmer, 2010).

Barriers to self-determination. In comparison to parent-report of supports, which were relatively consistent with quantitative findings (i.e., adaptive behavior as most significant predictor), few parents referred to low adaptive behavior, or a lack of opportunities to practice such basic skills in a real life setting as a barrier. Instead, parents were more likely to reference anxiety, behavior and mood, self-efficacy, communication, and cognitive functioning. Wehmeyer (2014) suggested that the need for supports to enable children with disabilities might also inadvertently foster dependency. A minority of parents in the present study acknowledged this obstacle and reported that by not allowing their children various opportunities, they may limit their child's development of self-determination. In other words, these parents reported barriers that were most consistent with the main findings that adaptive behavior was the strongest predictor of self-determination. Because Carter et al. (2013) have not published on the qualitative data that were collected on parent perceptions of supports and barriers as proposed in the original article, this is the first known study to highlight parent perceptions of obstacles to self-determination.

Despite parents' emphasis on the importance of self-determined behaviors, there was a high level of no response in the open-ended questions on the SDPQ, which suggests that

although parents believe self-determination is important, many may not feel equipped to shape self-determined behaviors in their children and are unsure of the barriers. The relatively high nonresponse rate in the present study suggests that many parents have difficulty identifying supportive and challenging factors related to their child's development of self-determination. Relatedly, a small number of parents explicitly stated that they did not know how to answer these open-ended questions. In fact, one parent even confessed that she felt that she needed suggestions on how to support her child's development. Although speculative, the high rate of parents who do not feel confident self-identifying supportive and challenging factors for their children suggests that future efforts to educate and empower parents around self-determination for their children would be beneficial.

Measuring self-determination

The present study used two different measures for self- and parent-reported self-determination. Particular challenges with the self-report measure indicated many advantages to parent-report. However, a parent-report measure of self-determination also has its own inherent limitations. Findings from the present study related to administration and structural validity of the self- and parent-report measures suggest several next steps for research on self-determination.

Challenges of self-report. The present study illustrated significant challenges with accessibility of the Arc's SDS for a more severely impacted population. Wehmeyer (2014) argues that while individuals with disabilities may have difficulty executing several skills related to self-determination with complete independence, they can still serve as causal agents in their own lives, particularly with adequate supports and opportunities. In line with this argument, it may be that it is not that individuals with FXS in the present study have low levels of self-

determination, but that there are significant limitations with measurement of self-determination in a more severely affected population of I/DD and the findings from the present study may be an underestimate of self-determination in adolescents and adults with FXS.

The Arc's SDS presented difficulty with several demands of the measure, including the language demands, reading level, and general accessibility. For example, the multiple response format in the autonomy domain used five multiple response options, while three may have been sufficient and more accessible for a lower functioning population. The self-regulation domain also presented unique challenges for this population due to the working memory demands necessary to understand and hold the beginning and end of a story in one's working memory, while developing a response that fits as the middle sequence in the story. In comparison, the response formats of the psychological empowerment and self-realization domains were more accessible as they each had only two multiple-choice options. The psychological empowerment domain, which states two opposing statements about various beliefs, might be increasingly accessible by decreasing the language demands by using one statement and adopting the agree/disagree format of self-realization domain. These minor changes to the Arc's SDS may increase accessibility and validity in measuring self-determination in lower functioning individuals.

The present study indicated that individuals who were adolescent, male, or identified as meeting criteria ASD were less likely to be administered the Arc's SDS based on the examiner's judgment of the participant's ability to validly complete the self-report measure. Individuals with lower NVIQ and fewer daily living skills were also less likely to be administered the self-report measure. Of the participants who were administered the self-report measure, individuals had the most difficulty on the Self-Regulation domain. It was suspected that working memory

demands and expressive and receptive language demands were significant barriers to completing this section of the self-report measure. This domain is likely an underestimate of participants' self-regulation, as conceptualized in self-determination (i.e., interpersonal cognitive problem solving and goal setting). However, while there are reasons to believe that the present study samples' performance on this domain is an underestimate of their skills due to the difficulty of the task, qualitative analysis suggests that skill deficits related to interpersonal cognitive problem solving and goat setting also likely exist. For example, while adolescents had ideas and goals for future jobs, they were unable to provide basic steps to reach their goals (e.g., specific job training, job coach, apply for the job). Additionally, because social anxiety and social impairments are hallmark features in FXS, it is not surprising that they would have more difficulty on items related to interpersonal cognitive problem solving. The challenges with self-report with individuals with more significant I/DD suggest that parent-report might be an alternative approach to measurement when necessary.

Confirmatory factor analysis could not be completed on the self-report measure due to small sample sizes of adolescents and adults who completed the respective adolescent and adult forms of the Arc's Self-determination Scale in the present study. The different standard scores used on the two measures (i.e., percentile scores and standard scores) is a significant limitation in exploring differences across adolescents and adults. Future efforts are needed to transform the adolescent form norm sample scores to standard scores, which would allow for future studies to use both measures simultaneously.

Parent-report as a proxy measure of self-determination. Because self-determination is not limited to individuals who are identified as higher functioning, measures that are accessible for all individuals should be available. However, it is common for psychological measures to use

parents and teachers as proxy reporters when individuals cannot self-report, and are even used when individuals can self-report to provide additional information for comparison. Many advantages to parent-report exist, including gaining a second impression of an individual's current functioning, and assessing how equipped parents feel to support their child's self-determination. Additionally, parent-report is also advantageous when the reliability of self-report is at question.

Despite a relatively small sample size as a limitation, the exploratory factor analysis indicated that the parent-report measure had a solid two-factor structure, which made it easy to identify two subscales (i.e., self-determined behaviors and importance of self-determination). While the measure demonstrated the utility of parent-report as a reliable measure of behaviors and skills related to self-determination, one of the main limitations was that parents were not asked to report on their child's attitudes and beliefs related to self-determination. It remains unknown whether parents are capable of accurately identifying their child's attitudes and beliefs related to self-determination. However, currently numerous standardized and well-documented parent-rating scales that cover a range of behaviors (e.g., daily living, social communication, internalizing and externalizing) and mood issues (e.g., depression, anxiety) are used. Future measures of self-determination through parent-report could assess attitudes and beliefs (two of the four essential components identified by the functional model) by asking parents to report on how frequently a child expresses their thoughts, feelings, and attitudes related to things such as success, failure, and self-awareness.

Informing intervention

In additional to health care disparities, studies have documented plateaued functional living skills (Smith, Maenner, & Mailick Seltzer; 2012), poorer quality of life, fewer housing

options, and increased unemployment and social isolation for individuals with disabilities (Wehman, 2013). While these disparities have been addressed by the education system through a focus on transition from high school to adulthood, less has been done to individualize intervention for individuals with moderate to severe intellectual disability. Similarly, there is limited research exploring intervention in the home or family environment. Although many positive outcomes ranging from employment to quality of life have been documented in the self-determination literature, these findings are limited to a less impaired population of individuals with disabilities and as a result leave many questions regarding how these findings apply to a more severely affected population.

The present study findings support the notion that, regardless of skill level, the development of self-determination is very important to parents. Moreover, while parents readily stress the importance of self-determination, they not only have difficulty identifying factors that support and challenge the development of self-determination, but their ideas are not consistent with empirically identified predictors of self-determination. This discrepancy suggests a great need for intervention both in terms of services for children as well as education and support for parents. Furthermore, the significant discrepancy between skill level and the emphasis placed on the importance of self-determination suggests that intervention is an important next step.

Direct interventions are a significant next step because findings from the present study indicate a strong relationship between adaptive behavior and self-determination and previous research shows a strong relationship between self-determination and quality of life. If self-determination is strongly predicted by daily living skills, then further developing daily living skills is an excellent avenue to support and increase independence. Because adaptive behavior, in comparison to IQ, is a shapeable skillset, significant opportunities for intervention and growth

exist, suggesting promising outcomes for self-determination in individuals with FXS. Self-determination encompasses a set of skills that can be practiced in all areas of functioning, especially in terms of school and advocating for services. Within the school setting, self-determination is important for all individuals with disabilities. However, as with most interventions, it is important that approaches be individualized and determined by a child's skill level. For example, males with FXS will need significantly different interventions that focus on basic lower-level skills, while females with FXS may be more prepared to practice higher-order skills, such as self-advocacy. Wehmeyer et al. (2013) found that a 3-year high school intervention focused on developing self-determination in students ID and LD led to significant positive outcomes including increased employment, career goals, and community access than peers with ID and LD that did not complete the intervention. Many instructional programs provided in the school setting that support the development of self-determination encourage family involvement (Wehmeyer, 2014).

One specific finding from the present study that might directly inform intervention is the self-report on the self-regulation section of the Arc's SDS. While most adolescents reported on employment goals, very few were able to provide one or more steps to take to work toward that goal. This finding suggests a significant need for an explicit area of intervention.

Research is needed to identify family- and parent-related variables that may influence the development of self-determination for individuals with I/DD. Research on how parents and siblings can help family members with I/DD develop functional daily living skills is an important first step that might begin by determining how different family systems approach teaching moments for self-determined behaviors and daily living skills. Parent and family research is particularly important because, while many interventions have been school-based, there are

endless opportunities to help children practice daily living skills and self-determined behaviors in the home environment. Davis and Wehmeyer (1991) suggested several strategies for parents to promote self-determination in education, including allowing their children various experiences, modeling self-confidence, exploring disability-related differences with them, encouraging them to take responsibility, and using every opportunity to make choices and providing feedback. They also shared that it is important to emphasize the process of goal setting and attainment, and set ambitious and attainable goals and expectations for children. Moreover, parents can encourage the siblings of children with I/DD in helping create a positive family environment for fostering self-determination (Wehmeyer, 2014). For example, siblings can support, advocate, and serve as a role model, particularly in the school setting.

Another important avenue for research focused on parents relates to parenting styles and identifying how parenting styles influences whether a parent provides more opportunities for choices and experiences for their children. For example, parents who experience high levels of anxiety themselves may not only have children who also experience more pronounced anxiety, but they may offer fewer opportunities for their children to take their own risks and practice newly learned skills. Similar to individual interventions, parent interventions should also be individualized and applied based on family need, culture, and goals for their children. Home visiting might be one avenue to support families and provide parent education to support self-determination from a young age.

One of the most important factors when considering how to intervene at the family level is culture. Self-determination as a normalization movement originated in Europe and specifically promotes values associated with self-determination that are central to western culture and society (Frankland, Turnbull, Wehmeyer, & Blackmountain, 2004). As one might expect, Eastern and

other non-Western cultures that have differing cultural norms (e.g., collectivism) will likely operationalize self-determination very differently (Zhang, 2005). These cultural differences then lead to a wide range of parenting styles, with Western cultures exhibiting parenting practices related to self-determination that may be discouraged by non-Western cultures (Zhang et al., 2005). While the research is limited, there is evidence that suggests parenting styles do impact a child's development and acquisition of skills related to self-determination (Zhang, Katsiyannis, & Zhang, 2002; Zhang, Wehmeyer, & Chen, 2005). In addition to cultural differences that must be reflected in the form of parent and family interventions, other factors, including SES and education must also be carefully considered. Zhang (2005) explored how culture, SES, and special education status affected parent engagement in fostering self-determination and found that parents with college degrees involved their children more frequently in daily decision making, discussing the future, and goal setting than parents with less education. Zhang (2005) also found that parents of children with disabilities involved their children less in these same activities, which was consistent with previous research that found parents of individuals with disabilities provide fewer choice making and trial-and-error activities (Bannerman, Sheldon, Sherman, & Harchik, 1990). However, because Zhang's (2005) study included a limited number of families (20%) with children who received special education under limited categories (e.g., learning disability, emotional/behavioral disorders), additional research is needed to corroborate these findings in families with children who experience similar or more significant impairments (e.g., intellectual disability, autism spectrum disorder).

In addition to direct interventions for parents, research on environmental variables is important for determining factors that contribute to self-determination in conjunction with family-systems research. Brotherson et al. (2008) and Summers et al. (2014) found that family

characteristics (e.g., housing) influenced how the family supported self-determination and created opportunities for their young children with disabilities. Lastly, professionals also play an important role in supporting the family. An emphasis on self-determination should be encouraged at an early age and include opportunities for individuals to express preferences, practice decision making, and increase autonomy (Palmer, 2010). As children age, the role of parents should be increasingly collaborative in nature to support collective decision making. It will be important for professionals working with families to support parents as their roles change.

Limitations

As mentioned above, measurement challenges are the primary limitation to this study. However, several other limitations should be considered when interpreting the results. The relatively homogenous sample in the present study in terms of race, SES, and caregiver education is one such limitation. The lack of variability in race, income, and education status of caregivers significantly limits the generalizability of the findings from the present study. Cultural differences, including race and ethnicity, income, and parent education are likely important characteristics that affect how important parents perceive self-determination for their child. Currently there are no known studies exploring these factors as variables that affect perceptions of importance. However, there is qualitative research that documents that importance of cultural variables. Specifically, one study indicated that parents with college degrees involve their children more in daily living skills (i.e., chores) decision making, and goal setting (Zhang, 2005). Similarly, these factors might also influence parent perceptions of supports and barriers, as well as general awareness of self-determination. Additionally, future research may benefit from

having comparison groups in order to better understand whether these findings are unique to FXS.

There were two primary limitations related to measurement. First, because the norms for the Adult SDS form were not published prior to the start of the present study, the age of the norm sample (i.e., over 40 years old) was not known, which resulted in norm sample that was not comparable to the present study sample. Second, another measurement-related factor that may have affected the results was measuring ASD dichotomously (i.e., met criteria or did not meet criteria) rather than as a continuous variable (i.e., symptom count). Use of ASD as a dichotomous variable may have resulted in IQ as a confounding variable because individuals with FXS and comorbid ASD are more likely to have lower IQ (Hustyi et al., 2015).

The present study used raw scores and did not make comparisons to the norm sample, due primarily to significant differences between the norm sample and the present study sample. While this lack of comparison may be considered a limitation through one lens, exploring individual differences within group was also an important first step to understand self-determination in individuals with FXS. By using raw scores, the present study found many significant differences within the group of individuals with FXS.

Priming parents through completion of the rating scale section on the SDPQ prior to answering open-ended questions on supports and barriers may also be another limitation in the present study. Although it was a minority, many parents' responses were similarly worded to the questions they completed prior to the open-ended section. Conversely, it may be that parents identified with some of the behaviors and skills related to self-determination and might suggest that parents may be receptive to interventions that provide parent education on approaches to supporting the development of self-determination in their children. Lastly, the high no response

rate on the open-ended section is something that could be avoided with a simple change. By offering parents an opportunity to report that they "do not know" what serves as supports or barriers for their child's self-determination, future research could better differentiate between parents who accidently or intentionally skipped the item from those who did not respond because they did not know what to write.

Future Directions

Results from the present study suggest that parent-report may be an appropriate alternative to assessing self-determination in individuals who are lower functioning, particularly until there is a more accessible self-report measure of self-determination. However, a parent's ability to provide insight into their child's feelings and beliefs related to self-determination is limited when compared to personal insight and self-report. The SDPQ is a good first step toward developing a standardized parent-report measure. Future efforts are needed to incorporate other variables related to self-determination, rather than being solely focused on behaviors or skills (e.g., choice making, problem solving). Relatedly, the present study is the first known study to provide initial psychometrics (i.e., exploratory factor analysis) on this parent-report measure. With additional minor changes as described earlier, this measure would benefit from testing with a larger sample that includes parents with children with a wider range of I/DD.

While the present the study provided insight into parent knowledge of ways to support self-determination and barriers to their child's self-determination, research is needed to determine how confident parents feel in their ability to help their children develop skills related to self-determination. Relatedly, parent focus groups could inform how receptive parents are to various interventions. While the family system may be the most important context for the

development of self-determination for individuals with I/DD, there is a paucity of research on the role of families (Wehmeyer, 2014).

Further exploration of the relationship between adaptive behavior and self-determination is an important next step. While it was expected that autism spectrum disorder and anxiety would be important predictors of self-determination in the FXS population, these hypotheses were not supported. Exploring these factors with adaptive behavior as a mediating variable may provide insight into the role of adaptive behavior in self-determination. For example, individuals with high anxiety or severe social impairments may not practice as many adaptive daily living skills as those with less social anxiety or social impairments. In Hustyi et al. (2015), even when controlling for IQ, individuals with FXS and high levels of autism symptomology were more dependent than those who did not meet criteria for ASD. This finding serves as an example of how adaptive behavior may be a mediating factor between ASD and self-determination.

As changes to the functional model have been proposed, future efforts are needed to develop appropriate measures (Shogren, Wehmeyer, Palmer, & Forber-Pratt, in press). Based on the findings from the present study, I would argue for future measures to be made accessible for all individuals with disabilities. Changes including reading level, formatting, and fewer multiple-choice response options would likely dramatically enhance accessibility for individuals with I/DD. Additionally, like many standardized psychological measures, measures of self-determination would benefit from a standardize measure that includes compatible parent-teacher- and self-report scales.

While individuals with FXS exhibit a unique behavioral phenotype, they also experience a group cohesion that may be a strong protective factor to self-determination that other individuals with intellectual disability and their families may not experience. Exploring whether

this understanding of their disability helps support self-determination and self-advocacy is also an important next step.

Conclusion

Research on contributing factors to self-determination for individuals with disabilities suggests a complex relationship with IQ, mixed findings on age and gender, and adaptive behavior commonly identified as a significant predictor. However, there is a paucity of research on self-determination for individuals with more significant impairments, as well as for individuals of specific I/DD groups (e.g., genetic syndromes). The purpose of the present study was to extend this research by exploring self-determination within fragile X syndrome (FXS) through self-report and parent-report. Results suggested that adaptive behavior is the most consistent predictor of self-determination in adolescents and adults with FXS based on both self- and parent-report measures. Despite the strong predictive value of adaptive behavior, parents did not explicitly identify the development of daily living skills as an important factor in supporting self-determination, nor did they identify a lack of daily living skills as a barrier to self-determination. Findings from the present study highlight the importance of future efforts emphasizing parent participation from both a measurement and intervention standpoint for individuals with significant impairments.

APPENDIX 1: IRB DOCUMENTATION

To: Adrienne Villagomez
School of Education Deans Office

From: Non-Biomedical IRB

Approval Date: 2/10/2015
Expiration Date of Approval: 2/09/2016
RE: Notice of IRB Approval by Expedited Review (under 45 CFR 46.110)
Submission Type: Initial
Expedited Category: 5.Existing or non-research data
Study #: 14-1349

Study Title: Self-determination in adolescents and adults with fragile x syndrome: The relationship between self-report, parent perceptions, and individual characteristics

This submission has been approved by the IRB for the period indicated. It has been determined that the risk involved in this research is no more than minimal.

Study Description:

Purpose: This study will explore self-determination in adolescents and adults with fragile x syndrome (FXS) using a self-report and parent questionnaire. This study will examine characteristics related to self-determination (e.g., cognitive functioning, autism spectrum disorder, anxiety). This study is a dissertation that will be using data collected from an ongoing research study approved by UNC (IRB Number 13-1128).

Participants: Approximately 100 males and females with FXS and their parents or primary caregiver recruited for the Decisional Capacity and Informed Consent in Fragile X Syndrome (IRB Number 13-1128).

Procedures (methods): Participants will be scheduled for a visit at home or in school where they will complete standardized measures of underlying cognitive and social/behavioral factors thought to be related to self-determination. Prior to the visit, the participant and/or their primary caregiver will be sent the consent/assent forms and self-report/parent-report measures.

Regulatory and other findings:

The IRB has determined that the study-specific rationale provided by the investigator is sufficient to justify the waiver of informed consent according to 45 CFR 46.116(d).

Investigator's Responsibilities:

Federal regulations require that all research be reviewed at least annually. It is the Principal Investigator's responsibility to submit for renewal and obtain approval before the expiration date.

You may not continue any research activity beyond the expiration date without IRB approval. Failure to receive approval for continuation before the expiration date will result in automatic termination of the approval for this study on the expiration date.

Your approved consent forms and other documents are available online at http://apps.research.unc.edu/irb/index.cfm?event=home.dashboard.irbStudyManagement&irb_id=14-1349.

You are required to obtain IRB approval for any changes to any aspect of this study before they can be implemented. Any unanticipated problem involving risks to subjects or others (including adverse events reportable under UNC-Chapel Hill policy) should be reported to the IRB using the web portal at http://irbis.unc.edu.

Please be aware that additional approvals may still be required from other relevant authorities or "gatekeepers" (e.g., school principals, facility directors, custodians of records).

The current data security level determination is Level I. Any changes in the data security level need to be discussed with the relevant IT official. If data security level II and III, consult with your IT official to develop a data security plan. Data security is ultimately the responsibility of the Principal Investigator.

This study was reviewed in accordance with federal regulations governing human subjects research, including those found at 45 CFR 46 (Common Rule), 45 CFR 164 (HIPAA), 21 CFR 50 & 56 (FDA), and 40 CFR 26 (EPA), where applicable.

APPENDIX 2: PRESENT STUDY MEASURES

Construct	Measure	Variable(s)	Level	Number
Self-determination (self-report)	*Arc's Self-determination Scale (Arc's SDS)*	Total score	Continuous	Raw score
		Autonomy subdomain score	Continuous	Raw score
		Self-regulation subdomain score	Continuous	Raw score
		Psychological Empowerment subdomain score	Continuous	Raw score
		Self-realization subdomain score	Continuous	Raw score
Parent-reported self-determination	*Self-determination Parent Questionnaire (SDPQ; nonstandardized)*	Self-determined Behaviors subscale score	Continuous	Raw score
		Importance of SD subscale score	Continuous	Raw score
Intellectual ability	*Stanford Binet Intelligence Scales, 5th Edition (SB5)*	Nonverbal IQ	Continuous	Standard score converted
Adaptive behavior	*Scales of Independent Behavior-Revised (SIB-R)*	Adaptive behavior composite score	Continuous	Standard Score
ASD Determination		Meets criteria ADOS-2 and SCQ	Nominal	Yes/No
	Autism Diagnostic Observation Schedule, 2nd Edition (ADOS-2)	Overall Total (SCI+RRB)	Continuous	Algorithm score
	Social Communication Questionnaire (SCQ)	Cutoff score = 15	Nominal	Yes/No
Anxiety	*Anxiety Depression and Mood Scale (ADAMS)*	Generalized Anxiety Total Score	Continuous	Raw score
Social avoidance	*Anxiety Depression and Mood Scale (ADAMS)*	Social Avoidance Total Score	Continuous	Raw score

APPENDIX 3. PARENT RESPONSES ON THE SDPQ OPEN-ENDED QUESTION 1 BY PARTICIPANT ID

ID	Gender and age group	Code	Encourage Skills Response 1	Code	Encourage Skills Response 2	Total # of codes	# of codes used
1001	Adult male	Experience	Keeping active in community - church activities; Special Olympics	Experience	Maintaining work (home, public) ethic - gives them feeling of accomplishment, can do, pride, and give back to society - also feels like everyone else - good citizen!	2	1
1011	Adult male	Choice	We always try to provide choices so NAME can have a sense of some control in his life. We provide choices from something as simple as what to eat for dinner, to what type of car to buy for the family!	Self-awareness	An ongoing goal is to teach NAME how to be more self-aware of his behavior and to recognize when he may be getting over-stimulated, how to communicate that effectively and in a more productive manner.	2	2
1021	Adult male	Don't know	Don't know they have it different	Resources	Getting him into school as fast as I could.	2	2
1031	Adult male	Experience	practice	Emotional support	LOVE	2	2
1032	Adolescent female	Emotional support	Never give up.	Emotional support	Praise whatever success you have.	2	1
1033	Adolescent male	Emotional support	Patience.	Emotional support	Love.	2	1
1041	Adolescent male	Behavioral strategy	Use tokens to help with self-regulation; i.e.. Limit his by having them cost a token & limit # of tokens	Behavioral strategy	Use social stories to help with psychological empowerment, self-regulation, & choice making skills	2	1
1061	Adolescent female	Emotional support	Don't let your child tell you they cannot do something, you need to push them to their fullest potential.	Choice; Emotional support	Let your child make choices on their own that are within reason. Praise them every day. Tell them how great they are, beautiful, smart, etc…and how much you love them.	3	2
1071	Adult male	Experience	Get them involved with outside activities- special Olympics/job.	Resources	Talk to other parents/teachers and find out what benefits their child is	2	2

					receiving and how they went about getting that resource.		
1081	Adult male	No strategy	Preston likes to help out. He loves to vacuum, he has his own vacuum. So he vacuums his bedroom and upstairs.	No strategy	He can make his own bed and put clean sheets on his bed.	2	1
1091	Adult female	No response		No response		0	0
1101	Adult male	Behavioral strategy	Set routines, although these too can get to be "obsessive"	Behavioral strategy	Prepare for changes - act out or talk out what is to happen	2	1
1102	Adult male	Emotional support; Behavioral strategy; Experience	Patience, repeating, repeating a act as a haircut. Breaking down steps to independence I go, go inside, sign him in, wait, pay, tip, leave after years it has gotten that he can walk in, give sign-in info, pay (with a $20) and give 1 dollar tip…	Behavioral strategy	A weekly plan - we do laundry M W F - it was a daily "obsession" - so now will get clothes sorted with dark night before - ready to wash. It's the assurance that it will be done - again much trial and error - still can't manage the actual job…	4	3
1111	Adolescent male	Behavioral strategy	Use visual reminders throughout the house to reinforce (e.g. "All food stays in the kitchen").	No response		1	1
1141	Adolescent male	Self-awareness	We help him understand what he is feeling and then explain (and show) how he might overcome his feelings (i.e. being scared of an event but going and having a good time).	Experience	We encourage Josh to make decisions and express them, giving him the time to sort through his thoughts (i.e. deciding what to order on a menu at a restaurant).	2	2
1151	Adult male	Choice	We encourage him to make his own choices. We give limited number of choices and let him choose.	Experience	Let them try, as long as they are safe. Empower them to do and try.	2	2

1161	Adolescent male	Experience	Allow their child to make the mistake- see where they need help.	Emotional support	Don't push too hard.	2	2
1172	Adult male	Experience ; Emotional support	Encourage to make minor decisions and provide praise for selections.	Modeling	Explain and re-explain in various words and examples acceptable behaviors and why. Very important in maintaining a job where things are not always fair and other personalities are involved.	3	3
1181	Adult male	Choice; Experience	Establish a "chore" list of tasks to be done daily but allow some choice within tasks. Ex: Pick out and set out clothes for the next day but can choose which shirt and which pants and shoes after checking weather on TV.	Experience	If there is something he really want, like pizza- ask him to call Dominos and order it, pay for it, ask for it at counter.	3	2
1191	Adult male	Behavioral strategy	Try to explain everyday choice in ways NAME can understand.	Problem solving	I encourage NAME to help problem solve, instead of taking the easy route and solving problems for him.	2	2
1192	Adult male	Emotional support	Encourage activities that are appropriate for their developmental age, to build self-worth.	Problem solving	Help them, step by step with problem-solving skills, this being a weakness even as adults.	2	2
1211	Adult male	No response		No response		0	0
1212	Adult male	No response		No response		0	0
1241	Adult male	Modeling	I need to communicate with my son about what he needs to learn and know.	Experience	Let him make mistakes, so he can learn from mistakes.	2	2
1251	Adolescent male	No response		No response		0	0
1271	Adult female	No response		No response		0	0

1281	Adult male	Modeling	Learn and show as you live life.	No response		1	1
1301	Adult male	Behavioral Strategy; Self-awareness	Self awareness/self knowledge- To help identify his strengths and interests he could have a schedule of a daily chore. NAME loves to help around the house.	Expectations	Self regulation- NAME can learn to know when it is time to take a shower after the end of the day.	3	3
1302	Adult male	No response		No response		0	0
1321	Adolescent male	Don't know	Not sure.	Don't know	Not sure.	2	1
1322	Adolescent male	Don't know	Don't know	Don't know	Don't know	2	1
1331	Adult male	Choice; Experience	Making choices- without having to pay money- is supported by regular trips to the public library. Waiting in line, counting out the correct number of videos, expressing needs clearly, etc.	Experience	Taking inventory of foods consumed during the day to decide if snack should be fruit/vegetable or bagel/toast.	3	2
1332	Adult female	No response		No response		0	0
1333	Adolescent female	No response		No response		0	0
1351	Adult female	Choice	Allow and encourage child to make their own decisions, perhaps offering different options.	Self-awareness	Openly and honestly communication about the strengths and weaknesses and work to compensate or work around weaknesses and stress assets.	2	2
1361	Adolescent male	Choice	Allowing child to make a few food choices at grocery store.	Resources	Knowing all resources available to your child.	2	2
1371	Adolescent male	Don't know	I need suggestions!	No response		1	1
1372	Adolescent male	Resources	The biggest help has been enrolling him in a small school for special needs kids. The atmosphere is one	No response		1	1

			of encouragement and support along with focus on the discipline it takes to succeed.				
1381	Adult male	Emotional support	We constantly demand excellent and smart behavior.	Experience	We involve him in many activities and events.	2	2
1382	Adolescent male	Experience	Insist on children being independent in every way possible that doesn't jeopardize their safety.	Behavioral strategy; Experience	Expose them to as many experiences as possible and encourage good behavior.	3	2
1391	Adult female	No response		No response		0	0
1392	Adult male	Self-advocacy; Emotional support	Talk openly about ways to work around negative impacts of their disability. Create positive options rather than emphasizing the problems.	Expectations; Experience	Encourage and expect involvement in household cuties, chores, and responsibilities.	4	4
1401	Adult female	Experience; Self-advocacy	Teaching her how to advocate for herself. Doing it for her the first two years of high school, explaining what I was doing. Helping her in her third year if she was not being successful and letting her do it alone her fourth year - helped a lot.	No response		2	2
1402	Adolescent male	Choice	Providing choices to help him show preferences and interests.	Experience	Exposing him to all situations and typical peer settings.	2	2
1411	Adolescent female	Experience	I encourage her with her computer skills. She prefers the computer and Ipad so I found apps that she can learn from. So I have her work on those as well as letting her surf the web.	Behavioral strategy	When the behavior gets a little rough, I taught her how to calm herself down (and count backwards etc.) so I just have to prompt her a little to use these tactics to calm herself	3	2

1461	Adolescent male	Emotional support	Talk to them.	Emotional support	Love them a lot.	2	1
1481	Adult male	No response		No response		0	0
1491	Adult male	No response		No response		0	0
1501	Adolescent male	Behavioral strategy	Using social stories.	Problem solving	Using pros and cons of a decision.	2	2
1521	Adolescent male	No strategy	Autonomy - NAME knows how to get snacks independently.	No strategy	Self-Regulation - He is able to use breathing strategies to calm self.	2	1
1531	Adolescent male	Behavioral strategy	Visual learning techniques such as an action plan to help understand the outcome/goal.	Experience	Give individual the opportunity to make decisions starting at a young age.	2	2
1581	Adolescent female	Goal setting	When she has a school project due, I ask her to define mini-goals along the way and to plan to finish at least a day or two early.	Modeling	I talk to her about decisions and plans of my own.	2	2
1591	Adolescent female	Experience ; Self-awareness	Provide as broad of a base of experiences as possible. She then has more to draw from in regards to likes/dislikes and strengths.	Emotional support	Regularly point out her strengths in small things to help provide tangible feedback for self-monitoring.	3	3
1611	Adult male	Experience	My one son wants to be "independent" and we encourage this whenever possible. We are not obtaining guardianship of him at 18 but will allow him to experience his adult rights with guidance. Second FX son this is not possible.	Emotional support	A person will be who they are going to be with little regard to therapies. Wished I had spent less time trying to "do" everything I could for my sons and less time "worrying" - They develop when their brains are good and ready	2	2
1621	Adult male	Choice; Experience	I try to give NAME choice when appropriate as much as possible and experience	Emotional support; Problem solving	We often have to revisit decisions he must make. His first response is "No." When I don't' react,	4	4

			reasonable consequences of his decisions.		but allow him to calm himself, he can think and problem solve better. It can take time.		
1631	Adolescent female	No response		No response		0	0
1641	Adolescent female	Goal setting; Experience	Start young setting manageable goals with clearly defined positive outcomes: Example: Before we can (do something the child likes) we need to clean the house. What do you want to do to help? Ok, so you are going to ____ in the next 30 minutes and then we	Modeling	Lead by example: Think advocacy is important? Go to advocacy day and discuss with your child. Take them to advocacy day.	3	3
1651	Adult male	Experience	Find an interest and figure out how your child can spend more time, or productive time doing an activity that incorporates that interest.	Emotional support; Behavioral strategy	We have capitalized on NAME's love of repetition to say something positive about him that he can repeat.	3	3
1661	Adult female	Choice	Training to give options and then encouraging her to make the choice	Resources	Additional schooling to give her needed skills	2	2
1671	Adult male	Choice	Give choices- not a lot, two or three.	Emotional support	Tell them their strengths. Emphasize them. Have not really been great at this!	2	2
1681	Adult female	No response		No response		0	0
1691	Adult male	Choice	Talk with your child about things they like to do. Try to offer choices in what they can choose to do so they learn about making decisions/choices .	Goal setting; Behavioral strategy	Encourage and teach your child about setting goals for learning skills and use a chart for tracking progress and give rewards to encourage them to continue on.	3	3
1711	Adolescent female	Experience ; Choice	Providing lots of opportunities for children to make their own choices and see the	Experience ; Problem solving	We try to build these skills at home on a regular basis by encouraging our daughter to do things	4	3

			outcomes of those choices including a variety of activities in different settings.		on her own and replying to many questions with "What do you think?"		
1731	Adult male	No response		No response		0	0
1732	Adult female	Goal setting	We help her design a flow chart that lists her goals and ways to achieve them.	Self-awareness	We point out her strengths and define her weaknesses and ask her how she can use that knowledge to improve in her weak areas.	2	2
1741	Adult female	Goal setting; Resources	Discuss her goals with her and help find ways to accomplish- look for outside help- BUR/OOD	Self-awareness	Be realistic- discuss strengths and weaknesses and how to use them for future planning.	3	3
1742	Adolescent male	Choice	Give options - help decide.	No response		1	1
1751	Adult female	Experience	Developing as many experiences as possible.	No response		1	1
1752	Adolescent male	Experience	Parents can offer as many experiences to develop interest, provide vocabulary for child to express likes - dislikes.	Resources	Advocate for services to support community involvement.	2	2
1761	Adult female	Modeling; Goal setting	Behavior modeling of others who set goals and developing a step by step process (breaking goals into small parts)	No response		2	2
1762	Adult female	Emotional support; Goal setting	Being a good listener, providing encouragement through the goal setting process.	Problem solving	Higher level problem solving skills strategies.	3	3
1763	Adult female	Goal setting	Breaking goals down into small attainable steps is helpful and seems to provide feelings of progress and success.	Emotional support	Providing emotional support and a sounding board for concerns and perceived road blocks.	2	2
1771	Adult female	Choice	Since they were little, I have tried to offer choices, offering more choices as they got	Modeling; Problem solving	As they got older, I would demonstrate problem solving and decision making by talking through a	3	3

			older.		problem and listing my choices, then the effects of each choice. That way, they could see how it works.		
1772	Adult male	No response		No response		0	0
1782	Adult female	No response		No response		0	0
1801	Adolescent male	Experience	Give them opportunities	Behavioral strategy	Be very reinforcing	2	2
1801	Adolescent female	No strategy	Strive to make yourself the best you can possibly be. You don't need to compete with anyone except yourself!	No strategy	Find your passion and make that your life's journey! Advocate for yourself; be a best friend to yourself!	2	1
1802	Adolescent female	Emotional support	Self-realization - I praise her for things she does well and point out to her what she's done such a good job at	Emotional support	Self-regulation - praise for job well done	2	1
1811	Adult female	Choice; Experience	Provide the child with every opportunity possible to make choices, decisions, and to take care of themselves.	Emotional support	No matter how long it takes - even years - continue with you belief in and effort for your child to become as independent as possible.	3	3
1812	Adult male	No response		No response		0	0
1831	Adult male	Experience	Give them jobs that have to be done on a regular basis.	Emotional support	Be patient with their learning.	2	2
1841	Adult female	Experience	Involvement in church small groups doing activities the person with disabilities likes to do such as baking/cooking. She enjoys church.	Emotional support	Praising them when they are in a happy state.	2	2
1842	Adolescent male	No strategy	With a sports interest tell me more about each player/team.	Experience	Find a way to help out a team - loves football so he helped manage the team this past fall.	2	2

1851	Adult female	Experience ; Emotional support; Self-advocacy	We have always encouraged NAME to self advocate for herself and to focus on her strengths. We have encouraged her to step out of her comfort zone and to try new things and to not be afraid of failure. It is ok to make mistakes.	No strategy	NAME has always been self-motivated. She wants to succeed and do well.	4	4
1862	Adolescent female	Self-advocacy	Self-advocacy - It is okay to ask for help and be able to communicate to others what makes you successful.	Choice; Problem solving; Behavioral strategy	Choice-making - encourage to make "good" choices and what those consequences will be because of the choice. We prompt them to verbalize what they perceive their consequences to be. Sometimes we do this exercise in written form.	4	4

APPENDIX 4. PARENT RESPONSES ON THE SDPQ OPEN-ENDED QUESTION 2 BY PARTICIPANT ID

ID	Gender and age group	Code	Barriers Response 1	Code	Barriers Response 2	Total # of codes	# of codes used
1001	Adult male	Lack of resources	Lack of public resources - transportation, adequate housing, waiting lists!	Lack of resources	Lack of general public knowledge, understanding - even researchers - of this population and its broad range of variability with each and every FX affected person!! It's quite overwhelming and questionnaires like this just bring it all back!!	2	1
1011	Adult male	IQ	His cognitive ability, his inability to make important decisions based on meaningful input and information.	Communication	Lack of expressive language. He can be difficult to understand and he has difficulty expressing his thoughts.	2	2
1021	Adult male	Don't know	Don't know	Don't know	Don't know	2	1
1031	Adult male	IQ	Intellectual ability/aptitude	No code	Love, teach by example	1	1
1032	Adolescent female	IQ	Mental capacity.	Executive function	Attention.	2	2
1033	Adolescent male	IQ	Mental capacity.	No response		1	1
1041	Adolescent male	IQ	His IQ	Lack of resources	Amount of time needed to work on them consistently	2	2
1061	Adolescent female	Lack of opportunity	The parent enabling them and sheltering them and not allowing the child to grow up or letting go.	No code	If the parent does not show any of these behaviors (patience, empathy, kindness, happy) then the child will never learn these good behaviors. Children learn what they live, show by example!	1	1
1071	Adult male	Lack of opportunity	His stepfather and I allowing him more freedom.	Lack of resources	Isolation- we do not live in a neighborhood that has other his age or transportation to get him there.	2	2

1081	Adult male	Understanding	Being able to fully understand what's going on and reacting on his own.	Understanding	Understand what it means to take a bath/brush teeth and do a good job.	2	1
1091	Adult female	No response		No response		0	0
1101	Adult male	Anxiety	Anxiety, change	Communication	Not able to express self - especially on demand - people don't realize this, have so much inside that none gets out.	2	2
1102	Adult male	Anxiety; Academic skills; IQ	Anxiety - afraid of failure - response on demand - not able to keep records will always need help with math, writing/reading IQ too low to have these skills	Motivation	Has to be motivated - and really don't know what will or will not trigger that - works one day and next day not!	4	4
1111	Adolescent male	Behavior and mood	Impulse control	Understanding	Not truly understanding the consequences for something you do.	2	2
1141	Adolescent male	Communication	Having the right words to use to describe his feelings or decisions.	No response		1	1
1151	Adult male	Understanding; IQ	Lack of understanding. His limited cognitive ability.	No response		2	2
1161	Adolescent male	No code	Nothing he can't do- but a matter of time.	Self-efficacy	Him saying he can't- but helping him.	1	1
1172	Adult male	Understanding	His ability to reason and recognize and weigh options.	Lack of opportunity	Opportunities for options not directed/implemented by parent but by him.	2	2
1181	Adult male	No response		No response		0	0
1191	Adult male	Communication; IQ	Communication deficit. Mental deficit.	No code	He is	2	2
1192	Adult male	IQ	Mental deficits.	No response		1	1
1211	Adult male	Self-efficacy	No self confidence. Have to keep reassuring them that they can do it…may take time	Self-efficacy	Low self-esteem. Have to keep reassuring them that they can do it…may take time but they	2	1

			but they can do it.		can do it.		
1212	Adult male	Self-efficacy	No self-confidence. Have to keep reassuring them they can…may take time but they can do it.	Self-efficacy	Low self-esteem. Have to keep reassuring them they can…may take time but they can do it.	2	1
1241	Adult male	Executive function	Learning to process information.	Anxiety; Asking for help	Asking for help when needed. He is extremely shy.	3	3
1251	Adolescent male	No response		No response		0	0
1271	Adult female	No response		No response		0	0
1281	Adult male	Executive function	Ability to focus.	Behavior and mood	Behavior issues.	2	1
1301	Adult male	Understanding	NAME wants to help in the daily chores of the house but he does not know when or what can be done without one of us instructing him.	Understanding	NAME has a hard time remembering to bathe himself. He has to be verbally instructed by an adult.	2	1
1302	Adult male	Communication	Lack of communication.	IQ	Lack of cognitive ability.	2	2
1321	Adolescent male	Self-efficacy	Self confidence.	Don't know	Not sure.	2	2
1322	Adolescent male	Don't know	Don't know	Don't know	Don't know	2	1
1331	Adult male	Anxiety	Hard to say- anxiety plays such a strong role in making healthy choices difficult.	Behavior and mood	There's also such a challenge to getting Z to see beyond what he wants.	2	2
1332	Adult female	Self-awareness	There is a constant balance between what makes her safe or happy and what might hurt someone's feelings.	Anxiety	She gets so anxious, she curls into herself.	2	2
1333	Adolescent female	No response		No response		0	0
1351	Adult female	Academic skills	Reading and math comprehension.	Motivation	Lack of initiative.	2	2
1361	Adolescent male	Lack of resources	Lack of resources within community.	No response		1	1
1371	Adolescent male	Motivation	Don't know how to motivate him.	No response		1	1
1372	Adolescent male	Anxiety	His anxiety has been the biggest	No response		1	1

			barrier to being confident and independent.				
1381	Adult male	Self-awareness	He still has a very hard time with self-awareness, i.e., nose running, dirty face, etc.	Behavior and mood; self-efficacy	He lies a lot. Answers questions with what he thinks we want to hear rather than the truth.	2	2
1382	Adolescent male	IQ	Below normal IQ.	Behavior and mood	Impulsivity.	2	2
1391	Adult female	No response		No response		0	0
1392	Adult male	Self-awareness	NAME is unaware of the extent to which his intellectual disability impairs his ability to function as a normal independent adult.	Lack of opportunity	The lack of employment opportunities that suit his abilities.	2	2
1401	Adult female	Anxiety	Anxiety and feeling overwhelmed.	No response		1	1
1402	Adolescent male	Communication	Lack of language and communication skills.	Anxiety	Anxiety in new settings.	2	2
1411	Adolescent female	Behavior and mood	Behaviors - anytime routine is derailed - tantrums	Self-awareness; Asking for help	Knowing what she can and can't do and asking for help with things she is weak in.	3	3
1461	Adolescent male	Motivation	Attitude	Behavior and mood	Listening	2	2
1481	Adult male	No response		No response		0	0
1491	Adult male	Self-efficacy	Self-confidence	Self-efficacy	Concern about disappointing others	2	1
1501	Adolescent male	Academic skills	Reading.	Behavior and mood	Impulsive.	2	2
1521	Adolescent male	Understanding	Recognizing/ understanding what these concepts mean.	Understanding	Creating strategies that he could follow/ understand to achieve self-determinations goals.	2	1
1531	Adolescent male	IQ	Concrete thinking vs. abstract thinking. Visual learner.	Self-efficacy	Self confidence. Still lacks the confidence to make a decision. Seeks approval.	2	2
1581	Adolescent female	Anxiety; Self-efficacy	She is a people pleaser and would struggle with making a decision that might upset or	Behavior and mood	She is often distracted.	3	3

			disappoint others.				
1591	Adolescent female	Anxiety	Her anxiety limits her experience	Lack of support	Lack of adult support to help her learn patterns of decision making.	2	2
1611	Adult male	Developm ental level	Mental age, maturity, understanding "life"	No response		1	1
1621	Adult male	Behavior and mood	NAME can be rigid in his thinking and very resistant to new things.	Anxiety; Behavior and mood	NAME's anxiety and reactivity can keep him from reaching goals. He reacts strongly to corrective feedback and be offensive in his response, i.e., yelling, making fist	3	2
1631	Adolescent female	No response		No response		0	0
1641	Adolescent female	Anxiety	Anxiety.	Self-awareness	Goal does not equal skill set. Not completely away of her own strengths and weaknesses.	2	2
1651	Adult male	Academic skills	Literacy- NAME cannot read.	Anxiety	Anxiety- NAME mixes up time and days and he is often anxious about his schedule.	2	2
1661	Adult female	Executive function	Does not have the executive functioning skills	No response		1	1
1671	Adult male	Self-efficacy	Self-confidence. Wants to not have FXS and be "normal."	Anxiety	Panic/anxiety attacks in job situations. Loses confidence. Fears to try again. Fears humiliation. Aaron is very high functioning. He sees his lack- his difference- his inabilities. Compares self to cousins (who high achievers!).	2	2
1681	Adult female	Lack of resources	NAME does not have anyone - a professional - except her family and friends to help her plan and set goals.	Lack of resources	She may not have the funding she needs to follow through with her plans.	2	1
1691	Adult male	Self-awareness	Lack of self awareness and self monitor skills.	Executive function	Attention span.	2	2

1711	Adolescent female	Anxiety; Self-efficacy	She has trouble making decisions and forming her own opinions. She is very strongly influenced by what others want her to do.	Self-efficacy	Lack of confidence in her choices and abilities is probably the biggest barrier.	2	2
1731	Adult male	No response		No response		0	0
1732	Adult female	Understanding	She can't always recognize which is the salient information that she should act on.	No response		1	1
1741	Adult female	Developmental level	Immaturity	Self-efficacy; Communication	Not able to be assertive and communicate effectively	3	3
1742	Adolescent male	Understanding	He doesn't necessarily understand what he's deciding or why.	No response		1	1
1751	Adult female	Self-efficacy	Her reliance on other people to make decisions for her.	Anxiety	Her fear of making a mistake.	2	2
1752	Adolescent male	Lack of resources	Economically, providing services to support growth and experiences.	Lack of resources	Community may have a "transition plan" that is easiest or cheapest and try to mold your child to fit their services. Ignoring personal interest and strengths of child.	2	1
1761	Adult female	Behavior and mood	Emotionality	Executive function	Higher level problem solving skills are always being addressed and worked on by everyone involved in NAME's life.	2	2
1762	Adult female	Self-efficacy	Level of confidence.	Anxiety	Fear, insecurities	2	2
1763	Adult female	Self-efficacy	Lack of confidence.	Anxiety; Motivation	Fear of failure. Motivation.	3	3
1771	Adult female	Communication; Asking for help	When she has a problem, she does not know how to ask for help.	Self-efficacy	She has trouble coming up with possible choices, and difficulty finding out the effects of those choices.	3	3

1772	Adult male	Communication	Lack of verbal skills.	No response		1	1
1782	Adult female	No response		No response		0	0
1801	Adolescent male	IQ	Not smart	Communication	Not verbal enough	2	2
1801	Adolescent female	Lack of resources	A lack of peer acceptance and a lack of community	Lack of resources	A lack of quality infrastructure and support!	2	1
1802	Adolescent female	Anxiety	Anxiety	Understanding	Confusion/inability to understand, frustration	2	2
1811	Adult female	IQ; Behavior and mood	Only her own cognitive and emotional limitations and who knows for sure what they are.	No response		2	2
1812	Adult male	No response		No response		0	0
1831	Adult male	Academic skills	Can't read.	Academic skills	Can't write down ideas.	2	1
1841	Adult female	Understanding	Lack of understanding.	Anxiety; Motivation	Lack of desire and motivation.	3	3
1842	Adolescent male	Communication	Being able to convey his preferences in a way others will understand.	Understanding	Not sure if he understand the importance of many of the things listed above.	2	2
1851	Adult female	Self-efficacy	Lack of self confidence. Not always believing in herself and constantly questioning her decisions.	Anxiety	Fear of failure and making mistakes.	2	2
1862	Adolescent female	Developmental level	I think most of these skills can be taught over time at varying ages. However, there is a higher level of "thinking" and "analysis" that naturally happens from trial and error, therefore there may be limitations to skills during maturation	No response		1	1

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