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Profiles of Neuropsychological Functioning in Children and Adolescents with Spina Bifida

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LOYOLA UNIVERSITY CHICAGO

PROFILES OF NEUROPSYCHOLOGICAL FUNCTIONING IN CHILDREN AND ADOLESCENTS WITH SPINA BIFIDA

A DISSERTATION SUBMITTED TO THE FACULTY OF THE GRADUATE SCHOOL IN CANDIDACY FOR THE DEGREE OF DOCTOR OF PHILOSOPHY

PROGRAM IN CLINICAL PSYCHOLOGY

BY

RACHEL WASSERMAN

CHICAGO, IL

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ABSTRACT

The current study examined neuropsychological performance among children with spina bifida (SB) to determine if there are distinct subgroups or “profiles” of cognitive functioning. 96 children with SB myelomeningocele (ages 8-15) completed a brief assessment battery. Hierarchical and non-hierarchical cluster analyses were used to identify and confirm a cluster solution. Hypothesized predictors of cluster membership included lesion level, number of shunt surgeries, history of seizures, age, ethnicity, socio-economic status, family stress, and family environment. Outcomes included independence, academic success, expectations for the future, and quality of life.

Ward’s cluster method indicated a 3-cluster solution, and was replicated with 2 other cluster methods. The following labels were applied to the clusters: "Average to Low Average Cognitive Ability, Impaired Motor" (n=39), "Average to Low Average Cognitive Ability" (n=32), and "Extremely Low to Borderline" (n=25). SES and shunt status significantly predicted group membership. Cluster membership significantly predicted independence, academic success, parent expectations for the future, and child reported physical quality of life.

Cluster analyses identified 3 distinct cognitive profiles with different patterns of cognitive strengths and weaknesses. These clusters proved to distinguish the groups on future outcomes as well. Findings from this study highlight the variability in cognitive profiles among children with SB. Clinical implications and future research are discussed.
CHAPTER ONE
INTRODUCTION

The purpose of this study was to examine neuropsychological performance among children with spina bifida to determine if there are distinct groups or “profiles” of cognitive functioning. Spina bifida myelomeningocele (SBM) is a congenital birth defect that produces orthopedic, neurological, urinary, and psychological difficulties. Neuropsychological functioning in children with spina bifida, has been shown to predict social development (Rose & Holmbeck, 2007), quality of life (Hetherington, Dennis, Barnes, Drake, & Gentili, 2006), and functional independence (Heffelfinger et al., 2008). However, the neuropsychological sequelae of SBM are complex and heterogeneous due to differences in the severity of neuropathology. For instance, SBM is associated with malformations of brain structures (e.g. Chiari II malformation; delayed maturation of gray and white matter; and hydrocephalus; Argento, Warschausky, Shank, & Hornyak, 2011). Children with SBM demonstrate considerable variability with respect to the nature of their neurological insults and cognitive deficits (Yeates, Loss, Colvin, & Enrille, 2003). Thus, it has been challenging to identify a neuropsychological phenotype for children with spina bifida.

A profile of neuropsychological functioning for children with spina bifida has been described by combining findings across many studies (Argento et al., 2011; Dennis & Barnes, 2010; Dennis, Landry, Barnes, and Fletcher, 2006; Fletcher & Dennis, 2009).
These reviews have demonstrated how children with spina bifida and/or hydrocephalus differ from their typically developing counterparts across various neuropsychological constructs such as reading (Barnes & Dennis, 1992), verbal discourse (Barnes & Dennis, 1998; Dennis & Barnes, 1993), narrative content (Dennis, Jacennik, & Barnes, 1994), math skills (Barnes, Pengelly, Dennis, Wilkinson, Rogers, & Faulkner, 2002), attention (Brewer, Fletcher, Hiscock, & Davidson, 2001), executive functions (Fletcher et al., 1996), memory (Scott et al., 1998; Yeates, Enrile, Loss, Blumenstein, & Delis, 1995), and intelligence (Fletcher et al., 1992; Soare & Raimondi, 1977). Most of these studies compare children with spina bifida to typically developing children or population norms.

While these studies have provided valuable information about group differences for children with and without spina bifida, they have not addressed the cognitive heterogeneity within this group. Indeed, researchers have found that performance on neuropsychological measures varies among children with spina bifida (Fletcher et al., 2005). Thus, children with spina bifida do not always demonstrate the same level or pattern of performance deficits. Significant within group differences could be indicative of variations of severity within the same profile (Figure 1, top) or different patterns of performance that are indicative of multiple profiles (Figure 1, bottom). Fletcher, Ostermaier, Cirino, and Dennis (2008) report evidence for the latter. Even though no statistical comparisons were conducted, data provided by Fletcher and colleagues (2008) suggest that “the modal profile is most apparent for the group of children who are not Hispanic and who have lower level (lumbar or sacral) spinal lesions” (pg. 9). Hence,
there is evidence for more than one neuropsychological profile of children with spina bifida (e.g., Hispanic children and children with upper level lesions may have qualitatively different profiles than other children). More generally, it is likely that children with spina bifida demonstrate varying patterns of neurobehavioral functioning.

Information about the cognitive ability of children with spina bifida is important for neuropsychological assessment and clinical intervention. When neuropsychologists evaluate an individual’s cognitive functioning, they assess many cognitive domains to obtain a profile of relative strengths and weaknesses. The individual’s profile allows neuropsychologists to recommend appropriate interventions. Just as neuropsychologists use an individual's profile to determine such interventions, so might clinicians use a literature-based profile to create much needed group interventions. In fact, Fletcher and Dennis (2009) suggest that researchers "focus on core deficits" when creating and evaluating interventions for children with spina bifida. However, an intervention based on the current literature would address the typical profile of deficits, but not necessarily the deficits of a particular individual. Because there is considerable heterogeneity among children with spina bifida, such an intervention may not be appropriate for every participant or even the majority of participants. For example, a comprehensive intervention that addresses all known core deficits might be excessive for children with only one or two areas of concern. On the other-hand, an intervention that focuses on one deficit (e.g. attention) may not be comprehensive enough for children with co-occurring deficits (e.g. a math skill deficit in combination with an attention deficit).
Cluster analytic techniques can be used to detect whether relatively homogeneous subgroups exist within a larger, more heterogeneous group (Steele & Aylward, 2007). Children with spina bifida are certainly a heterogeneous group, considering their cognitive performance. By identifying subgroups of children with spina bifida, it is possible that more tailored interventions could be designed to address the different types of cognitive weaknesses within the larger group. The current study aimed to determine whether there are subgroups of children with similar neuropsychological profiles, within a larger group of children with spina bifida. It was hypothesized that 4 subgroups of children with spina bifida exist with distinct profiles. A more detailed description of each subgroup is included in the following chapter. Briefly, the four subgroups were hypothesized to include children with: 1. Generally higher functioning (than group 2) with significant variability within neuropsychological domains; 2. Generally lower functioning (than group 1) with significant variability within neuropsychological domains. 3. Generally higher functioning (than group 4) with similar performance within neuropsychological domains. 4. Generally lower functioning (than group 3) with similar performance within neuropsychological domains (see Figure 2).

In addition to identifying subgroups with similar neurocognitive profiles, predictors and outcomes of group membership were also investigated. Several risk factors have been associated with differences in cognitive functioning, such lesion level, number of shunt revisions, a history of seizures, age, ethnicity, SES, family stress, and family environment (Argento et al., 2011 & Dennis et al., 2006; Fletcher et al., 2008). It was expected that these factors would predict group membership. Additionally, cognitive
ability can influence other areas of functioning (Fletcher & Dennis, 2009). Thus, it was expected that group membership would predict differences in the following outcome variables: independence and self-care, academic success, expectations for the future, and quality of life. Each of these outcomes is a potential area of intervention for children with spina bifida (Argento et al., 2011; Fletcher & Dennis, 2009). Determining what outcomes are associated with a particular cognitive profile might aid in developing more specific interventions that are tailored to a subgroup’s overall strengths and weaknesses.

The current study aimed to address several limitations of past work. This study examined individual differences within spina bifida, rather than comparing children with spina bifida to norms or a typically developing group. Additionally, instead of examining one cognitive construct (e.g., attention), the current study assessed many constructs (intelligence, attention, comprehension of complex language, affect recognition, executive functioning, and manual dexterity) to generate sub-group specific, multidimensional profiles of strengths and weaknesses. Cluster analysis was used to determine whether subgroups with similar neuropsychological profiles exist within the larger group. These subgroup profiles have the potential to be more informative than general statements about the neuropsychological functioning of children with spina bifida. Finally, predictors and outcomes of profile membership were examined.

The following sections include a review of the current literature pertaining to the hypotheses of this study. Specifically, the literature review explored present neuropsychological profiles for children with spina bifida, support for subgroups of children with similar cognitive profiles, and predictors and outcomes of
neuropsychological performance. As well, methods are discussed, including descriptions of the data collection process and measures used. Data analytic procedures that address the hypotheses of this study are explained. Finally, results are reported and conclusions, clinical implications, and future directions are discussed.
CHAPTER TWO

REVIEW OF THE RELEVANT LITERATURE

Brain Abnormalities Associated with SBM

Spina bifida myelomeningocele (SBM) is associated with several brain malformations that influence cognitive outcomes (Fletcher & Dennis, 2009; Juranek & Salman, 2010). The most commonly associated brain abnormality is the Chiari II malformation. Most children with SBM demonstrate this complex anomaly of the midbrain, hindbrain and cervical spinal cord (Fulton & Yeates, 2010; Juranek & Salman, 2010; Fletcher & Dennis, 2009). A Chiari II malformation is characterized by “a significantly smaller posterior fossa (cerebellum and brain stem) with its contents crowded and distorted in appearance” (Juranek & Salman, 2010, pg. 23). Children with SBM may also present with additional, less frequent brain malformations, such as tectal beaking (an abnormality of the midbrain, Fletcher & Dennis, 2009) and dysgenesis of the corpus callosum (Futon & Yeates, 2010). In addition to structural abnormalities, Chiari II malformation can cause an obstruction of the flow of cerebral spinal fluid in the third and/or fourth ventricles (Fletcher & Dennis, 2009). Thus, about 80-90% of children with SBM also present with hydrocephalus, an accumulation of cerebral spinal fluid in the ventricles of the brain (Fulton & Yeates, 2010). When the cerebral spinal fluid does not drain properly, the ventricles expand and create pressure on the surrounding brain structures. Secondary complications, due to hydrocephalus, can include the destruction of
white matter axons near the lateral ventricles and the stretching of neural fibers, particularly the corpus callosum (Del Bigio, 2010; Fulton & Yeates, 2010). If the hydrocephalus is so severe that it is expected to cause further complications, then a shunt is surgically placed in the brain shortly after birth to drain the excess fluid (Argento et al., 2011). Overall, the most common brain malformations in individuals with SBM occur within the cerebellum, corpus callosum, and cerebral cortex. However, a great amount of heterogeneity in the size, shape, and appearance of these brains structures has been documented for individuals with spina bifida (Juranek & Salman, 2010). Due to these differences in brain anomalies, individuals with SBM experience a variety of neurological insults and, thus, present with inconsistent neurocognitive profiles.

**Neuropsychological Profiles in Children with Spina Bifida**

**Domains of Cognitive Functioning**

A neuropsychological profile typically includes a description of performance across cognitive domains, emphasizing particular areas of strength or weakness (e.g., Argento et al., 2011; Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009). Profiles include cognitive domains such as intelligence (IQ), academic achievement, attention and executive functioning, language, social-emotional processing skills, and motor ability (e.g. Argento et al., 2011; Fulton & Yeates, 2010; Wills, 1993). These descriptions of cognitive functioning are created by reviewing results from many studies that examine different areas of cognitive functioning. A neuropsychological profile of children with spina bifida has been proposed in the literature (Argento et al., 2011; Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009; Fulton &
Yeates, 2010). The following is a description of the neuropsychological profile for children with spina bifida, as presented in the literature.

Authors suggest that children with SBM typically present with low to low average IQ (Argento et al., 2011; Fulton & Yeates, 2010). Thus, it is implied that, as a group, their intelligence is lower than population norms. This implication is supported by studies that include children with and without SBM. Children with SBM have demonstrated lower intelligence compared to their typically developing peers (Hampton, Fletcher, Cirino, Blase, Drake, Dennis, & Kramer, 2011). Still, the overall measure of IQ may not be a good description of their intellectual functioning, because children with spina bifida often demonstrate a discrepancy between verbal and non-verbal IQ. It is suggested that children with SBM perform more poorly on measures of non-verbal IQ because of fine-motor and spatial processing deficits associated with cerebellar dysfunction (Fletcher et al., 2008; Lee et al., 2005). This discrepancy is particularly noted with the Weschler or Stanford Binet intelligence tests (Fletcher et al., 2008). For example, one study found that a group of children with spina bifida performed within the average range on verbal IQ, and borderline range for non-verbal IQ (Vinck, Maassen, Mullaart, & Rotteveel, 2006). While this discrepancy between verbal and non-verbal intellectual functioning is often noted in the literature (i.e., Erikson, Baron, & Fantie, 2002; Fletcher et al., 2008; Fulton & Yeates, 2010), it is not always found to be significant (Dennis et al., 1981; Hommet et al., 1999). One reason for these differences in findings could be the amount of variability within the sample. In fact, researchers have concluded that children with SBM and hydrocephalus demonstrate the largest amount of variability in their IQ scores, when
compared to children with other types of spina bifida (Barf, Verhoef, Jennekens-Schinkel, Post, Gooskens, Prevo, 2003).

Children with spina bifida are noted to show strengths in certain areas of academics that are analogous to their pattern of intellectual functioning. Generally, children with spina bifida score in the average range for basic academic skills like word reading, spelling, and basic math operations (Fletcher et al., 2008). However, children with spina bifida score lower than would be expected on measures of complex skills like math application, calculation, and reading comprehension (Argento et al., 2011; Erickson et al., 2002; Fulton & Yeates, 2010). When compared with typically developing children, children with spina bifida use fewer mature strategies (i.e., adding or multiplying rather than counting) to solve complex math problems (Barnes, Wilkinson, Khemani, Boudesquie, Dennis, & Fletcher, 2006). Overall, the profile of academic functioning generally reflects a similar pattern of intellectual strength and weakness (i.e., higher verbal than non-verbal abilities). Still, some studies of academic functioning have found conflicting evidence. For instance, Hampton and colleagues (2011) concluded that children with spina bifida performed more poorly than controls on measures of word recognition as well as math calculation. Thus, as with intelligence, studies suggest much variability in academic performance among individuals with spina bifida.

Children with spina bifida are often described as having deficits in both attention and executive functioning (Argento et al., 2011; Fletcher et al., 2008). However, children with spina bifida do not show deficits in all areas of attention. Often, children with spina bifida perform more poorly on measures of selective and divided attention, than they do
on measures of sustained attention (Brewer et al., 2001; Erickson et al., 2002). More specifically, Argento and colleagues (2011) summarize recent research and suggest that children with spina bifida show difficulties in attention because they have impairments in shifting attention from one stimulus to another. Reportedly, children with spina bifida take longer to orient their attention to a relevant stimulus (i.e., focusing) and struggle to inhibit their return of attention to a previously-attended to stimulus (Dennis & Barnes, 2010). For children with spina bifida, this pattern of attention deficits has been associated with midbrain malformations, such as tectal beaking and smaller posterior brain volume (Dennis et al., 2005), rather than anterior systems (frontal lobes) that are generally related to ADHD and issues with sustained attention (Burmeister, Hannay, Copeland, Fletcher, Boudousquie, & Dennis, 2005). The ability to shift attention is related to executive functioning. As previously mentioned, children with spina bifida are described as having deficits in executive functioning when compared to able-bodied peers (Argento, et al., 2011; Fletcher et al., 2008; Hampton et al., 2011; Lindquist, Uvebrant, Rehn, & Carlsson, 2009; Roebroeck et al., 2006). Additionally, children with hydrocephalus perform worse on measures of executive functioning than those without hydrocephalus (Barf et al., 2003; Vinck et al., 2006). In sum, children with spina bifida show deficits in specific areas of attention and overall executive functioning.

Additionally, children with spina bifida often show issues with both gross and fine motor skills. Gross motor functioning is usually dependent upon the level of the spinal lesion, such that higher lesions lead to greater gross motor deficits. Depending on the level of the lesion, children with spina bifida may require the use of braces or a
wheelchair to ambulate. Often, fine motor functioning is also impaired bilaterally in children with spina bifida (Erickson et al., 2002). More specifically, children with spina bifida show difficulty with motor planning (Erickson et al., 2002; Fletcher et al., 2008), motor timing (Dennis et al., 2004), and motor speed (Barf et al., 2003; Hetherington & Dennis, 1999). Due to these deficits, children with spina bifida perform more poorly than typically developing children on measures of fine motor skills (Hampton et al., 2011). This pattern of fine motor difficulties, particularly deficits in motor timing, has been associated with decreased volume of the cerebellum (Dennis et al., 2004). Also, children with shunted hydrocephalus perform more poorly than those without shunts (Hampton et al., 2011). Overall, it is suggested that individuals with spina bifida present with various fine and gross motor functioning, depending on their level of lesion and shunt status.

According to the literature, children with spina bifida generally show difficulties with social skills and social-contextual language (Argento et al., 2011; Erikson et al., 2002; Fletcher et al., 2008; Fulton & Yeates, 2010). More basic social skills like eye contact and emotional IQ seem relatively intact in adults with spina bifida (Iddon, Morgan, Loveday, Sahakian, & Pickard, 2004). However, children with spina bifida show deficits in pragmatic communication skills (Fulton & Yeates, 2010). For example, children with spina bifida are noted to struggle with matching conversation topics to an evolving social context (Fletcher et al., 2008); making inferences and understanding non-literal language (Barnes & Dennis, 1998); and conveying meaning concisely (Dennis & Barnes, 1993). Erickson and colleagues (2002) also suggest that children with spina bifida have difficulty comprehending non-verbal social cues, such as gestures and body
positioning. The pattern of social/language deficits common to children with spina bifida has been described as cocktail party syndrome (Tew, 1979). This syndrome is defined as “hyperverbosity; fluent, well-articulated speech containing perseverations and stereotyped phrases; and an over-familiarity of manner” (Argento et al., 2011, pg 561).
As a whole, these descriptions of social-emotional functioning suggest that children with spina bifida have deficits in processing non-verbal communication and complex language, which leads to social skill deficits.

**Overarching Pattern of Functioning Within Cognitive Domains**

As is apparent from this literature review, children with spina bifida often show a characteristic pattern of strengths or weaknesses within each cognitive domain. For instance, children with spina bifida show greater difficulty with measures of non-verbal IQ than verbal IQ and greater difficulty with selective attention than sustained attention.
To identify a phenotype of neuropsychological functioning for children with spina bifida, Dennis and colleagues (2006) have taken a different approach to the traditional neuropsychological profile that describes functioning across domains (i.e., intelligence, academic skills, attention, etc.). These researchers have examined underlying similarities in the pattern of strengths and weaknesses within each cognitive domain (e.g., strength in sustained attention and weakness in shifting attention). Dennis and colleagues (2006) suggest that the neuropsychological profile is best described by these overarching strengths and weaknesses that reflect inconsistencies in the traditional neuropsychological profile.
For children with SBM, Dennis and colleagues (2006) describe overarching strength in associative processing and weakness in assembled processing. Associative processing is defined as, “data-driven and based on the formation of associations, enhancement, engagement, and categorization” (Dennis et al., 2006, pg. 289). According to Dennis and colleagues (2006), associative processing requires the engagement of one domain. Some examples of tasks requiring associative processing include recognizing faces or decoding familiar words. Intact associative processing is reportedly related to intact motor learning/adaption from movement repetition; intact recognition and categorization of faces and shapes; intact memory without intension to memorize (implicit memory); intact grammar and vocabulary; intact word recognition; intact math facts; and intact behavior activation (Dennis et al., 2006). Dennis and colleagues (2006) also suggest that strength in associative processing may depend on environmental influences, such as poverty, parenting, and education. They suggest that children with a less enriching environment may demonstrate less strength in associative processing skills than children from a more enriching environment. Thus, it is suggested that children with spina bifida maintain the ability to engage with one stimulus or idea at a time, but the level of achievement is dependent on one’s environment.

In contrast, “assembled processing” is used to describe the pattern of impairments that is demonstrated by children with SBM. Assembled processing is “based on dissociation, suppression, disengagement, and contingent relations” (Dennis et al., 2006, pg 289). As described by Dennis and colleagues (2006), assembled processing requires the disengagement from one stimulus and use of several cognitive domains at the same
time. For example, making inferences from oral language requires the application of real
world knowledge to the current discourse. It requires the individual to disengage from the
content of the conversation, to shift attention to his/her own real-world knowledge, and to
then apply the knowledge to the content of the conversation. For instance, suppose that
one person says to another, “I need to find shoes for the winter.” To further the
conversation, the listener must first recall relevant knowledge about winter (i.e., winter is
usually cold and snowy) and then apply it to the context of the conversation (i.e., the
speaker may need shoes that are warm and waterproof). Dennis and colleagues (2006)
propose that impairment in this type of processing is a result of primary and secondary
neurological insults (e.g., brain malformations and issues related to hydrocephalus).
Impaired assembled processing is reportedly associated with impaired motor control
(hand, eye coordination), impaired coordinate or relational perception (figure/ground
delineation), impaired explicit memory, impaired constructed meaning (applying world
knowledge and context to language), impaired reading comprehension, impaired math
estimation, and impaired behavioral regulation (Dennis et al., 2006). Overall, these
researchers suggest that children with spina bifida generally present with functional
deficits due to weaker assembled processing skills and functional assets due to stronger
associative processing skills.

**Neuropsychological Profiles for Subgroups of Children with Spina Bifida**

Dennis and colleagues (2006) suggest that the neuropsychological profile for
children with spina bifida is determined by specific neurological insults and
environmental factors. However, children with SBM do not always experience the same
neuropsychological insults or exhibit the same brain malformations. In fact, a large amount of heterogeneity in the size and appearance of brain structures has been reported for children with spina bifida (Juranek & Salman, 2010). Children with SBM experience different neuropsychological insults that most likely lead to differences in their neurocognitive functioning. Thus, a single neurocognitive profile may not be appropriate for most children with SBM.

In the current literature, children with spina bifida have been categorized into subgroups based on criteria for neuropsychological disorders (e.g. Burmeister et al., 2005; Yeates et al., 2003). Typically, these subgroups are identified by examining prevalence rates of neuropsychological diagnoses within a larger group of children with spina bifida. For example, about 50% of children with spina bifida display a cognitive pattern consistent with non-verbal learning disorder (NVLD; Yeates et al., 2003) and about one third of children with spina bifida meet criteria for attention-deficit hyperactivity disorder (ADHD; Burmeister et al., 2005). Thus, subgroups of children with spina bifida may include those who meet criteria for ADHD, NVLD, both, or neither. However, children with spina bifida often do not display typical symptoms or behavior associated with these diagnoses. While children with spina bifida do have issues with attention and non-verbal learning, they do not exhibit the same pattern of neuropsychological impairments typically seen in children with ADHD (Brewer et al., 2001) or NVLD (Hommet et al., 1999; Ris et al., 2007). Thus, these diagnostic categories, and method of sub-grouping, may not be appropriate for children with spina bifida.
Still, it is possible that subgroups of children with spina bifida exist with similar neuropsychological profiles. Not all children with spina bifida show the same pattern of neuropsychological functioning. While generic patterns and models of neuropsychological performance have been suggested in the literature, there is still a large amount of variability within this population (Barf et al., 2003; Snow, Prince, Souheaver, Ashcraft, Stefans, & Edmonds, 1994; Wills, 1993). Researchers conclude that variability is the norm for children with SBM, and that “the prototypal SB patient is an untenable abstraction” (Barf et al., 2003, pg. 817). Thus, one phenotypic profile may not be the best description of neuropsychological functioning for children with spina bifida. Moreover, Fletcher and colleagues (2008) suggest that this variability is due to specific predictors. These researchers suggest that the profile of cognitive functioning “varies in a principled way, with sociodemographic factors, biological variables, and environmental variables” (Fletcher et al., 2008, pg. 319). It follows that children with similar predicting factors should present with similar neuropsychological profiles. Therefore, subgroups of children with spina bifida may exist that have similar neuropsychological profiles as well as a similar array of predictive correlates.

Rather than determine how many children with spina bifida meet diagnostic criteria for a specific disorder, this researcher aimed to identify subgroups (clusters) and then determine what factors were common within each subgroup (i.e., inattention, non-verbal deficits, etc.). A model of neurocognitive functioning by Dennis and colleagues (2006) was used to predict neuropsychological characteristics of each potential cluster. Dennis and colleagues (2006) suggest that biological factors, such as Chiari II
malformation, hydrocephalus, shunt malfunction, and lesion level affect assembled processing skills and functional deficits. These researchers suggest that greater biological severity is associated with greater cognitive impairment. Thus, to predict specific clusters, this researcher assumed the level of general cognitive functioning would depend on biological severity, such that children with more severe biological risk factors would perform at a generally lower cognitive level. Dennis and colleagues (2006) also suggested that strength in associative processing skills and functional assets (i.e., vocabulary) are reduced by environmental factors such as poverty, low SES, and poor parenting. They state, “environmental moderators are important, not because of their influence on assembled processing, but because they reduce SBM assets in associative processing” (Dennis et al., 2006, pg. 293). Thus, it was expected that positive environmental predictors (i.e., higher SES) would be associated with higher performance on measures that require associative processing (i.e., vocabulary), relative to other scores in each individual's profile. These relatively higher performance scores would create more variability within the neuropsychological profile.

Based on these assumptions, it was expected that four clusters of individuals with similar neuropsychological profiles would emerge from a larger sample of youth with spina bifida. The distinctive features of each group were based on an interaction between biological and socio-environmental factors (see Figure 2). It was expected that the first cluster would be distinguished by higher scores than the second cluster, with variability across measures of neuropsychological functioning. It was expected that individuals in this cluster would have fewer biological risk factors, and therefore their deficits in
assembled processing would be less severe. The variability in scores were expected to be due to the presence of fewer environmental risk factors, whereby associative processing skills and neurocognitive strengths were expected to be intact (see Figure 3). Thus, due to an enriching environment, associative processing skills would be higher than assembled processing skills. The second cluster was expected to include individuals with variability in their scores (due to fewer environmental risk factors and intact assembled processing skills), but generally lower scores than the first cluster (due to greater biological severity, i.e., more shunt surgeries, see Figure 3). The third and fourth clusters were expected to include individuals with less variability in their profiles. It was expected that their profiles would be more consistent because of greater environmental risk factors (i.e., low SES) and thus fewer functional assets (areas of relatively higher performance). Moreover, it was expected that the third cluster would include individuals with fewer biological risk factors, and thus higher scores than individuals in the fourth cluster (see Figure 3). In sum, the first and third clusters would show greater overall functioning, due to fewer biological insults, while the second and fourth clusters would display generally lower functioning because of more severe biological insults (see Figure 3).

**Predictors of the Neuropsychological Profile**

**Biological Predictors**

The location of the spinal cord lesion is a biological factor that may affect the child's neuropsychological profile. Generally, more extensive motor and cognitive impairments are associated with higher level lesions (Argento et al., 2011; Fulton and Yeates, 2010). Specifically, less of the spinal cord is damaged with lower lesions and
thus fewer muscle groups are affected. Higher level lesions affect limb functioning and fine motor skills (Erickson et al., 2002). While the association between lesion level and motor functioning is well established, there are mixed findings regarding the relationship between lesion level and cognitive functioning. Some researchers note no significant association between lesion level and cognitive performance (Lomax-Bream, Barnes, Copeland, Taylor, & Landry, 2007; Roebroeck et al., 2006) or sustained attention (Erickson et al., 2002). Still, lesion level is reportedly related to academic achievement, such as functional reading outcome (Hetherington et al., 2006). Overall, findings from the literature suggest that lesion level is related to motor functioning and may be predictive of academic performance and general cognitive functioning. Lesion level is a variable that is present at birth and may be associated with general cognitive functioning. A higher lesion level would contribute to greater biological severity. Thus, it was hypothesized that lesion level would predict group membership, such that a greater proportion of children in cluster 2 and 4 would have high lesion levels (see Figure 2).

To treat hydrocephalus, children with SBM typically undergo a shunt placement surgery shortly after birth (Argento et al., 2011). In this surgery, a shunt is placed in the brain to drain excess cerebral spinal fluid. Still, it is possible for a shunt to fail or become infected, and thus require shunt revision or replacement. A surgical intervention is required each time a shunt is revised or replaced. Thus, further neurological damage is possible with every shunt surgery. Indeed, a greater number of shunt-related surgeries has been associated with decreased full-scale IQ (Barf et al., 2003) and performance IQ (Hetherington et al., 2006); poorer executive functioning (Brown et al., 2008); and lower
functional math skills (Hetherington et al., 2006). Thus, the literature suggests that a greater number of shunt revisions is associated with poorer neurocognitive outcomes. However, other studies suggest that shunt revisions have no effect on IQ (Dennis et al., 1981), sustained attention (Erickson et al., 2002), or neuropsychological outcomes (Hampton et al., 2011). Thus, the literature includes mixed findings as to whether a greater number of shunt surgeries is associated with differences in neuropsychological functioning. According to the model proposed by Dennis and colleagues (2006), a greater number of shunt malfunctions may be associated with increased functional deficits. Thus, it was expected that the number of shunt surgeries would be associated with greater biological severity. It was hypothesized that the number of shunt surgeries would successfully predict the individual's neuropsychological profile, such that a greater amount of shunt surgeries would be related to an increased chance that the individual was in cluster 2 or 4 (see Figure 2).

Whether a child has a history of seizures is another biological factor that could influence neuropsychological outcomes in children with spina bifida. Epilepsy is often associated with hydrocephalus (Erikson et al., 2002), and is more common in children with spina bifida and hydrocephalus than in children without hydrocephalus (Yoshida et al., 2006). One study found that children with spina bifida and a history of seizures displayed poorer meta-cognitive skills (executive functioning, Brown et al., 2008). There are few studies that investigate the impact of epilepsy on neuropsychological outcomes in children with spina bifida. Still, for adults with spina bifida, epilepsy has been associated with mental retardation (Barf et al., 2003). Also, in a population of otherwise healthy
children, children with epilepsy performed more poorly on measures of attention (Williams, Griebel, & Dykman, 1998). Thus, according to the literature, a history of seizures may predict lower cognitive and executive functioning scores in children with spina bifida. It was expected that a history of seizures would be associated with greater neurocognitive deficits. In other words, it was hypothesized that the chance of an individual being in cluster 2 or 4 would increase if he/she had a history of seizure disorder (see Figure 2).

**Sociodemographic Predictors**

The neuropsychological profile may differ depending on the child’s age. It is possible that a child’s performance relative to same-aged peers may improve or deteriorate as the child matures. For instance, children with spina bifida show difficulties with complex math skills. However, children are not expected to understand complex math skills until mid to late childhood. Researchers have found that when children with hydrocephalus are compared to typically developing peers, their relative math abilities decrease with age (Wills, 1993). There are mixed findings in the literature regarding age as a predictor of other areas of cognitive functioning. For instance, a review of the literature suggest that as children with spina bifida become older, they may show improved sustained attention and reduced behaviors associated with “cock-tail party syndrome” (Erickson et al., 2002). On the other hand, other researchers have found no cognitive differences in children of different ages (Dennis et al., 1981). Overall, the literature suggests that age may predict a child’s level of academic achievement, attention, and social language, but may not affect more stable cognitive domains such as
intelligence. It is possible that older children may demonstrate greater variability in their neurocognitive profile. Thus, it was hypothesized that the chance of an individual being in cluster 1 or 2 (those with greater variability) would increase as the child's age increases.

Ethnicity is another factor that may affect a person's neuropsychological profile, and is particularly relevant to children with spina bifida. Mexican-American (Hispanic) mothers are 2 times more likely to have a child with a neural tube defect (Berry, Bloom, Fley, & Palfrey, 2010). Thus, the prevalence rate of Hispanic children with spina bifida is higher than would be expected (Lary & Edmonds, 1996). There are several ways in which ethnicity might affect performance on neuropsychological measures. Differences in language, cultural norms, patterns of social/family interactions, and importance placed on certain types of intelligence/learning may affect how a child performs on neuropsychological tests (Sattler, 2008; Sternberg, 2004). Specifically, Hispanic-American children may perform more poorly on academic measures and measures of language. The average reading level of Hispanic-Americans in the 12th grade is about 4 years behind that of Euro-American and Asian American youth (Sattler, 2008). This pattern of lower verbal scores for Hispanic-American children has been demonstrated in children with spina bifida as well. Fletcher and colleagues (2008), report that on average Hispanic children with SBM show lower verbal intelligence than nonverbal intelligence. Thus, the literature suggests that ethnicity may predict a child's neuropsychological profile, particularly verbal and reading abilities. These constructs are typically described as relative strengths for children with SBM. Thus, it is possible that Hispanic children
with spina bifida do not show relative strengths in associative processing skills, and thus perform more consistently across neurocognitive measures. It was hypothesized that the individual's ethnicity would successfully predict the individual's neuropsychological profile, such that Hispanic youth would have a greater chance of being in cluster 3 or 4 (see Figure 2).

**Environmental Predictors**

The neuropsychological profile may also differ, depending on the child's environment. Low socioeconomic status (SES) is a risk factor for neural-tube defects (Wasserman, Shaw, Selvin, Gould, & Syme, 1998). Thus, many children with spina bifida are born into a family that is economically disadvantaged. In typically developing children, it is well established that low SES is a risk factor for poorer cognitive, academic, and socio-emotional outcomes (McLoyd, 1998). Therefore, it is possible that SES may affect neuropsychological outcomes in children with spina bifida as well. In fact, Swartwout, Garnaat, Myszka, Fletcher, and Dennis (2010) have found an interaction between SES and ethnicity in predicting IQ performance in children with spina bifida. They report that low SES, Hispanic children display higher non-verbal abilities than verbal abilities. This profile is opposite from the typical profile (higher verbal than non-verbal IQ) that is reported for non-Hispanic children and high SES, Hispanic children (Swartwout et al., 2010). While SES does appear to affect neuropsychological performance in children with spina bifida, it reportedly has no effect on behavioral outcomes of executive dysfunction (Brown et al., 2008). Overall, the literature suggests that children from low SES families, may show greater difficulty with verbal cognitive
measures, but no differences in executive functioning. Because measures of verbal IQ most likely map onto associative processing skills (Fletcher et al., 2008), it is possible that youth from lower SES display fewer functional assets. It is also possible that children from lower SES families show greater difficulty with verbal cognitive measures because of a less enriching family environment. Thus, individuals with low SES and less enriching environments might also display a more consistent profile, as there would not be specific areas of strength. In the model proposed by Dennis and colleagues (2006), it is suggested that low SES leads to reduced functional assets. Thus, in the current study, it was hypothesized that SES and family environment would successfully predict group membership. Specifically, individuals from families with low SES, low enrichment, and high stress would more likely be a member of cluster 3 or 4 (less variable profile), and individuals with higher SES, higher enrichment, and lower stress would more likely be a member of cluster 1 or 2 (more variable profile; see Figures 2 and 3).

**Outcomes of Neuropsychological Profiles**

A person's neuropsychological profile may predict how that individual functions in every-day life. Children with spina bifida are often delayed in their every-day functioning and adaptive behavior. Such areas of delay include independence, academic success, expectations for the future, and quality of life. Thus, interventions might be appropriate to help children with spina bifida achieve a similar level of every-day functioning as their peers. By examining what outcomes are associated with which neuropsychological profile, we can better determine which subgroups might be most at risk, and in need of intervention.
Individuals with spina bifida do not achieve similar levels of independence as their same-aged peers (Friedman, Holmbeck, DeLucia, Jandasek, & Zebracki, 2009), and show deficits in adaptive behavior (Holler, Fennel, Crosson, Boggs, & Mickle, 1995). Thus, for children with spina bifida, independence is an important area for intervention. It is possible that differences in the neuropsychological profile could be associated with differences in independence/self-care achievement. In fact, research supports some associations between neuropsychological performance and independence outcomes. For children with spina bifida, higher executive functioning is associated with functional independence (Heffelfinger et al., 2008) and autonomy development (Tuminello, Holmbeck, & Olsen, 2011). Also, writing fluency has significantly predicted personal living skills and community living skills (Barnes, Dennis, & Hetherington, 2004). Thus, a pattern of neuropsychological performance may predict one's level of independence/self-care. It was expected that individuals with higher scores on neurocognitive measures would achieve a greater level of independence. Specifically, it was hypothesized that subgroups would be associated with greater independence as follows (from the highest level of independence to the lowest): cluster 1, cluster 3, cluster 2, and cluster 4 (see Figures 2 and 3).

When compared with typically developing peers, many individuals with spina bifida are less successful in their academic careers. For example, young adults with spina bifida are less likely to attend college or to be employed by age 18/19 (Zukerman, Devine, & Holmbeck, 2011). Thus, predictors of academic success are important to understand so that appropriate interventions can be developed. Some studies suggest that
one's neuropsychological profile may be a salient predictor of academic success. Many children with spina bifida (about 25%) have a specific math disability and about 3% have a reading disability (Barnes et al., 2006). These specific learning disorders may have an impact on academic success. For children with spina bifida, academic achievement is associated with verbal IQ (Swartwout et al., 2010). Also, writing fluency is a significant predictor of whether an individual with spina bifida attends college, such that better writing fluency leads to an increased chance of attending college (Barnes et al., 2004). Overall, the literature suggests that one's neuropsychological profile many predict academic success. Thus, it was expected that group (cluster) membership would be associated with level of academic achievement. Specifically, it was expected that the first cluster of youth with spina bifida, those with higher overall scores and intact strengths, would be associated with the greatest amount of academic success. It was also hypothesized that the following clusters would be associated with lesser levels of academic achievement as follows (from highest level of academic achievement to the lowest): cluster 3, cluster 2, and cluster 4 (see Figures 2 and 3).

As previously mentioned, many individuals with spina bifida lag behind their typically developing peers in regards to meeting developmental milestones such as going to college. Most youth with spina bifida report that they would like to live independently, and many report that they would like to have a family, own a home, and get married (Betz & Redcay, 2005). However, young adults with spina bifida are generally delayed in achieving these goals. Youth with cognitive delays may have greater difficulty achieving these milestones. Thus, parents of children with cognitive delays might have fewer
expectations for their child’s independence, career, and social development. There is some evidence that parental expectations for the future might differ, depending on the neuropsychological profile. For children with spina bifida, research suggests that higher reading ability is associated with more ambitious parental expectations for their child’s career (Creed, Conlon, and Zimmer-Gembeck, 2007). Thus, it was expected that parental expectations for their child's future would differ depending on the child's neuropsychological profile, such that parents of youth with higher cognitive functioning would have greater expectations for the future. It was hypothesized that the following clusters would be associated with parental expectations for the future as follows (from greatest expectations to least): cluster 1, cluster 3, cluster 2, and cluster 4 (see Figures 2 and 3).

There are mixed findings regarding quality of life in youth with spina bifida. Some research suggests that children with spina bifida report lower quality of life than would be expected (Lemelle et al., 2006). However, other studies suggest that children with spina bifida report moderate to high average levels of quality of life (Sawin, Brei, Buran, & Fastenau, 2002). Thus, it appears that reported quality of life varies among individuals with spina bifida. It is possible that one’s neuropsychological profile may explain some of the variability in reported quality of life. Hetherington and colleagues (2006), report that for children with spina bifida, functional math skills were associated with quality of life. Another study of children with spina bifida concluded that executive functioning was associated with subjective quality of life (Barf, Post, Verhoef, Prevo, & Goosken, 2010). However, there was no reported association between quality of life and
reading skills, cognitive skills, intelligence, memory, or word production (Barf et al., 2010; Hetherington et al., 2006). Still, it is possible that a profile that indicates a higher cognitive performance, rather than individual measures, may predict greater quality of life. As with other outcomes, it was hypothesized that group (cluster) membership would be associated with quality of life, such that the following groups would display various levels of quality of life as follows (from highest level of quality of life to the lowest): cluster 1, cluster 3, cluster 2, and cluster 4 (see Figures 2 and 3).

The Current Study

While a neuropsychological phenotype is presented in the literature (Argento et al., 2011; Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009; Fulton & Yeates, 2010), it may not be the best indicator of neuropsychological functioning for all individuals with spina bifida. There is a large range of neuropsychological functioning in children with spina bifida. Thus, the group’s average performance may not apply to many or most children with spina bifida. Still, differences in the phenotypic profile may vary systematically as a function of biological and socio-environmental factors (Fletcher et al., 2008). Thus, it is possible that the larger, heterogeneous group could be divided into smaller, more homogeneous groups. In other words, more than one profile may be necessary to best describe this groups’ neuropsychological functioning. The current study aimed to identify subgroups of children with spina bifida, with similar neuropsychological profiles. These subgroup profiles have the potential to provide more clinically useful information than the overall group profile. From the subgroup profiles,
clinicians, teachers, and other care-takers would have a better understanding of the various ways that individuals with spina bifida may present.

Additionally, the profile described in the literature is a compilation of several individual studies of neuropsychological functioning. Few single studies include a comprehensive neuropsychological battery for all of the participants (e.g., Hampton et al., 2011). Thus, the neuropsychological phenotype for children with spina bifida is a compilation of findings from studies with different participants. One issue with this approach is that the hypothesized general profile may not describe strengths and weaknesses within individuals, but rather strengths and weaknesses across the larger groups. For example, if a participant group in one study was particularly strong in verbal skills and a participant group in another study was weak in non-verbal skills, than researchers may conclude that, as a group, children with spina bifida are generally stronger in verbal skills than non-verbal skills. However, this superiority of verbal functioning in one group may actually be due to group differences on demographics or other factors, rather than demonstrating a significant discrepancy across all individuals with spina bifida. The current study included measures of several cognitive domains to compile a comprehensive neuropsychological profile for each participant. Thus, the current study investigated a profile based on strengths and weaknesses within individuals with spina bifida.

Another issue that the current study aimed to address is the lack of participants’ diversity in previous research. Several of the previous studies have excluded children with lower intelligence (e.g., excluded IQ: <70 Dennis et al., 1981; <70, Hampton et al.,
It may be important to exclude participants based on IQ to rule out confounding effects of global cognitive impairment (Vinck et al., 2006). However, this practice is not conducive to understanding the range of cognitive functioning in children with spina bifida. Thus, the phenotype that is described in the literature may not be representative of children with spina bifida who have low IQ. Additionally, the cut-off point for IQ is not agreed upon or consistent across studies. Thus, the current study aimed to provide a better description of neuropsychological functioning in a more intellectually diverse sample of children with spina bifida.

In addition, previous studies have not included an ethnically diverse participant sample. Unfortunately, many researchers of neurocognitive functioning in children with spina bifida have not reported the ethnicity of their participants (e.g., Barf et al., 2003; Dennis et al., 1981; Hommet et al., 1999; Iddon et al., 2004; Lindquist et al., 2009; Jenkinson et al., 2011; Snow, 1999; Snow et al., 1994). Therefore, it is impossible to know whether conclusions from these studies and the subsequent neuropsychological phenotype, are generalizable across children of different ethnic backgrounds. In fact, Fletcher and colleagues (2005) suggest that there are cognitive differences between children of different ethnicities. Specifically, Fletcher and colleagues (2005) suggest that the current cognitive phenotype for children with spina bifida is most applicable to non-Hispanic children who have lower level spinal lesions. To address this concern, the current study included a group of children from ethnically diverse backgrounds.
Additionally, the current study examined ethnicity as a potential predictor of neuropsychological functioning.

Overall, the current study aimed to make a significant contribution to the literature by providing a more generalizable and clinically useful description of neuropsychological functioning in children with spina bifida. To address the variability in cognitive ability within children with spina bifida, the current study identified subgroups of children with similar neuropsychological profiles. Additionally, the study included a comprehensive neuropsychological battery, to determine individual strengths and weaknesses. Finally, the current study included more diverse participants, so that the findings of this study may be more generalizable to all children with spina bifida.

**Study Hypotheses**

**Hypothesis I.** Four cluster groups would emerge from the analysis. Varied performance across measures of associative and assembled processing would result in four distinct subgroups. The mean neuropsychological profile for each of these subgroups would be exemplified by variability and higher functioning than other clusters (cluster 1), variability and lower functioning than other clusters (cluster 2), consistency and higher functioning than other clusters (cluster 3), and consistency and lower functioning than other clusters (cluster 4; see Figure 2 and 3). Because the cluster analysis is an exploratory technique and the 4 cluster solution is not guaranteed, the following hypotheses may need to be adjusted to reflect the results of the cluster analysis.

**Hypothesis II.** Biological factors (lesion level, shunt status, number of shunt surgeries, and seizures), socio-demographic factors (age and ethnicity), and
environmental factors (socioeconomic status, family environment, family stress) would predict group membership. Biological factors would predict performance on tasks of assembled processing and the level of overall functioning (low vs. high), such that individuals with more severe biological factors would be associated with clusters 2 and 4, whereas individuals with fewer biological risk factors would be associated with clusters 1 and 3 (see Figure 2 and 3). Socio-demographic factors would predict performance on tasks of associative processing and group membership, such that older individuals and non-Hispanic individuals would more likely be in cluster 1 or 2 (more variable profiles), whereas younger individuals and Hispanic individuals would more likely be in cluster 3 or 4 (less variable profiles; see Figures 2 and 3). Finally, it was hypothesized that higher SES, greater personal growth in the family environment, and less family stress would be associated with better performance on tasks of associative processing and greater variability in the neurocognitive profile. Specifically, individuals with higher SES, greater personal growth in the family environment, and less family stress would more likely be a member of clusters 1 or 2 (more variable profile), whereas lower SES individuals would more likely be in cluster 3 or 4 (less variable profiles; see Figures 2 and 3).

**Hypothesis III.** It was hypothesized that group membership would predict levels of independence, academic success, expectations for the future, and quality of life. Specifically, subgroups would be associated with outcome variables in the following order from the highest level of outcome to the lowest: variability and higher functioning (cluster 1), variability and lower functioning (cluster 2), consistency and higher
functioning (cluster 3), and consistency and lower functioning (cluster 4; see Figure 2 and 3).
CHAPTER THREE

METHODS

Participants

The focus of the current study was on adolescents with spina bifida from a larger, longitudinal study on psychosocial adjustment in adolescents with spina bifida (Devine, Holbein, Psihogios, Amaro, & Holmbeck, 2012). Only youth with myelomeningocele were included in the current study, as brain malformations are usually associated with only myelomeningocele (Fletcher & Brei, 2010). Families and a close friend of a child with spina bifida, ages 8-15 years old, were recruited from four main sources: a children’s hospital, a children’s hospital that exclusively serves children with physical disabilities, a university-based medical center, and a statewide spina bifida association. Subjects were eligible for participation if they were able to speak and read English or Spanish, if at least one primary caregiver could participate, if they were cognitively able to complete questionnaires and neuropsychological measures, and if they lived within 300 miles of Chicago, IL. Families were recruited in many ways. The four organizations identified eligible families and mailed recruitment letters and initiated contact via phone. During the phone call, eligibility was determined and the first home visit was scheduled with the family. Additionally, some families were recruited in clinic. A research assistant met with patients during their outpatient clinic appointment to discuss the study and schedule the first home visit. Posters and flyers were also posted in the hospitals. From
these flyers, families contacted researchers to discuss eligibility and participation in the study. If the family was eligible, then researchers scheduled the initial home visit with the family. Two-hundred and forty-six families were approached during recruitment. Of the original 246 families, 163 agreed to participate; however, 21 of those families could not be contacted or later declined, and 2 families did not meet inclusion criteria, resulting in a sample size of 140 families (57% participation rate). Of these 140 children with spina bifida, 53.6% were female, the mean age was 11.40, 53.3% were Caucasian, 27.9% were Hispanic, 12.9% were African American, and 5.7% were of another ethnicity. There were no significant differences between those who participated and those who declined on the following characteristics: type of SB (i.e., myelomeningocele vs. other), $\chi^2(1)=0.0002$, shunt status, $\chi^2(1)=0.003$, and occurrence of shunt infections $\chi^2(1)=1.08$ (Devine et al., 2012). Although the larger study continued collecting data every 2 years, the current study used only data from the first time-point.

Additionally, the current study only included individuals with myelomeningocele (MM), and only those who completed every neuropsychological measure. Mother-report and medical chart information were used to determine whether the child had MM. In cases of discrepancy, medical chart information was preferred. Of the 44 participants who were excluded in the current study: 14 had some other form of spina bifida (e.g., lipomeningocele) and 30 did not complete the entire neuropsychological battery. Participants did not complete the neuropsychological battery for several reasons including fatigue, refusal to complete home visits, or low comprehension. Of those who did not complete the battery, 12 participants attempted every measure but were unable to
complete the battery due to low comprehension. There were no significant differences between those who did and did not complete the neuropsychological battery on the following characteristics: age, SES, race and IQ (WASI full, 2-scale IQ). There were significant differences in gender and shunt status, such that a greater percentage of males and children with shunts completed the battery and were included in this study (see Table 3).

The final participants in the current study included 96 families of children with spina bifida. Of the 96 children with spina bifida, 49% were female, the mean age was 11.13, 55.2% were Caucasian, 26% were Hispanic, and 18.8% were of another ethnicity. Parent report indicated 86.5% of the children almost always spoke English, 5.8% spoke it very often, 2.9% spoke it moderately often, and 4.8% were unknown.

Medical information was gathered from the medical chart. For the current 96 participants, medical chart review indicated almost half the children had spinal lesions in the lumbar level (41.7%), 32.3% were sacral, 13.5% were thoracic, and 12.5% were missing; 83.3% had a shunt; 53.1% had at least one shunt revision (20.8% missing); and 12.5% had a history of seizures (15.6% missing). Data was missing because the medical record data was not collected or because the lesion level was “unknown” in the medical record.

**Design and Procedure**

Trained graduate and undergraduate research assistants collected data from participants during 2 home visits that lasted about 3 hours each. For families that spoke Spanish, at least one Spanish-speaking research assistant participated in the data
collection. Families were compensated $50 for the first visit and $100 for the second. Additional participants were provided the following: $50 for peer participation, $10 for health professional questionnaire, and $25 for teacher questionnaire. After obtaining consent from the parents and assent from the child, families were asked to complete several questionnaires. To maintain confidentiality, family members were asked to fill out the questionnaires independently. As well, to ensure that the child understood the questionnaires, research assistants offered to read each question aloud and any Likert scale responses were displayed on a laminated card for the child to choose from. During the first home visit, the family identified one teacher and health professional to participate in filling out questionnaires. After the first home visit, researchers contacted the teacher and health-care professional, who completed questionnaire data through the mail. The adolescent with spina bifida also participated in about two hours of neuropsychological assessments that took place over both home visits (1 hour during each visit). Trained research assistants administered all neuropsychological assessments. All neuropsychological assessments were conducted in English, but instructions were clarified in Spanish if needed. After the home visit, the neuropsychological measures were scored and checked by another research assistant. For all participants, medical information about their physical status was gathered from the mother’s questionnaire and from the child’s medical chart (medical chart release was acquired during the home visit).
Measures

Neuropsychological Profile

Measures from the neuropsychological battery were used to provide a profile of neuropsychological performance. All neuropsychological measures provide age-based performance norms. Morris, Blashfield, and Satz (1981) suggest that researchers using cluster analysis should minimize the number of measures for each construct to reduce redundancy and to ease interpretability of the subgroups. Thus, only one or two measures were included for each neuropsychological construct. Table 4 summarizes specific abilities and measures associated with "associative and assembled" processing (see Table 4). Measures are listed by cognitive domain.

Intelligence. Two subtests from the Wechsler Abbreviated Scale of Intelligence (WASI) were used to assess verbal and non-verbal intellectual ability (Wechsler, 1999). To determine relative performance on verbal and nonverbal measures, these two subtests were kept separate, rather than combing them for a full-scale IQ score. Verbal intellectual ability was assessed with the vocabulary subtest from the WASI. The vocabulary subtest is a 42-item task similar to the Vocabulary subtests of the Wechsler Intelligence Scale for Children (WISC-III) and the Wechsler Adult Intelligence Scale (WAIS-III), except that the WASI subtest includes low-end picture items. Items 1-4 require the examinee to name pictures. Items 5-42 are orally and visually presented words which the examinee defines. Vocabulary is a measure of the individual’s expressive vocabulary, verbal knowledge, and fund of information. In addition, it is a good measure of crystallized intelligence and general intelligence (g). The average reliability coefficient for children 6-16 years old
was .89 (Wechsler, 1999). Standard scores are provided with a mean of 10 and standard deviation of 3.

Non-verbal intellectual ability was measured with the matrix reasoning subtest from the WASI. The matrix reasoning subtest is similar to the Matrix Reasoning subtest in the Wechsler Adult Intelligent Scale-III (WAIS-III). It is a series of 35 incomplete gridded patterns that the examinee completes by pointing to or stating the number of the correct response from five possible choices. Matrix Reasoning is a measure of nonverbal fluid reasoning and general intellectual ability. The average reliability coefficient for children 6-16 years old was .92 (Wechsler, 1999). This subtest provides a standard score with a mean of 10 and standard deviation of 3.

**Academic achievement.** The Wide Range Achievement Test 3 (WRAT3) was used to measure the development of basic skills of reading, spelling, and arithmetic (Wilkinson, 1993). The reading subtest measures the ability to read single words. For the spelling subtest, participants are required to spell individual words. The arithmetic measures the participant’s ability to complete math problems of increasing complexity. If the participant is not able to complete algebraic problems, this subtest also measures more basic math skills such as counting. The WRAT3 has demonstrated adequate internal consistency across subscales. The spelling subscale has a median coefficient alpha of .89, the reading subscale has a median coefficient alpha of .90, and the arithmetic subscale has a median coefficient alpha of .85 (Wilkinson, 1993). For each subtest, an individual achieves a standard score with a mean of 100 and a standard deviation of 15.
**Attention/ executive functioning.** Several measures were included to assess various aspects of attention and executive functioning. The planned connections subtest of the Cognitive Assessment System (CAS) was used to assess non-verbal planning skills that are a part of executive functioning. The CAS is an assessment battery designed to evaluate cognitive processing in children 5-17 years of age. The planned connections subtest has demonstrated adequate reliability with reliability coefficient ranging from .66-.86 (M=.77). Confirmatory factor analytic results indicate adequate construct validity, such that the planned connections subtest loaded onto the “planning” construct (maximum likelihood factor loadings range from .647 to .777). The CAS is designed to measure non-verbal cognitive processing so that it is unbiased toward minority children. Each subtest yields a scaled score with a mean of 10 and a standard deviation of 3 (Naglieri & Das, 1997).

Verbal executive functioning was assessed with the verbal fluency subtest of the Delis Kaplan Executive Function System (D-KEFS). The D-KEFS provides normative and qualitative data assessing higher level executive functions (Delis, Kaplan, & Kramer, 2001). The D-KEFS verbal fluency subtest includes three sections. To complete this subtest the participant is requested to generate as many words as possible in 60 seconds, under three separate conditions. In the first condition, the participant is given a letter and is asked to provide as many words as possible that begin with that letter (Letter Fluency). For the second condition, the participant is required to generate words within a specific category (Category Fluency). In the third condition, the participant is required to produce words within specific categories, by shifting between one category and another (Category
Switching). The switching subtest produces two scores for fluency (how many words were said correctly), and accuracy (how many times they correctly switched from one category to the next). The letter fluency subtest has demonstrated moderate to high internal consistency coefficients, but the category fluency and category switching have demonstrated lower scores of internal consistency (Delis et al., 2001). The verbal fluency task has been included in other measures of executive functioning and has previously shown evidence of validity (Delis et al., 2001). For each section of the verbal fluency subtest, scaled scores are provided with a mean of 10 and a standard deviation of 3.

Several subtests from the Test of Everyday Attention for Children (TEA-Ch) were administered to assess visual and verbal attention, as well as selective, sustained, and divided attention. The TEA-Ch yields age-scaled scores and percentiles based on a normed population of 293 children (Manly, Robertson, Anderson, & Nimmo-Smith, 1999). To assess selective/focused visual attention, the Sky Search subtest was administered. To complete this task, the participant must circle pairs of items where both items are the same, as quickly as possible. This task results in three scores: number of targets identified, efficiency of task (how quickly they were able to identify the correct targets), and attention score (efficiency, controlling for motor speed). The manual reports adequate test-retest reliability for time per target ($r = .80$) and attention score ($r = .75$; Manly et al., 1999). To assess sustained auditory attention, the Score subtest was administered. For this subtest, the participant is required to listen for and count the number of “scoring sounds” on an audiotape. The manual reports adequate test-retest reliability, with 76.2% of children scoring within 1 standard deviation of their time 1
score upon retesting (Manly et al., 1999). The Sky Search Dual Task subtest was administered to assess sustained-divided visual/auditory attention. For this task, the participant is required to circle pairs of identical items, while simultaneously counting the number of “scoring sounds” on an audiotape. The manual reports adequate test-retest reliability ($r = .81$; Manly et al., 1999). To assess auditory divided attention, the Score Dual Task was administered, in which the participant must listen for and count the number of “scoring sounds” on an audiotape, while simultaneously listening for the name of an animal in a news broadcast. The manual reports adequate test-retest reliability, with 71.4% of children scoring within 1 standard deviation of their time 1 score upon retesting (Manly et al., 1999). A scaled score with a mean of 10 and standard deviation of 3 is yielded for each subtest.

**Fine motor.** Fine motor ability was assessed using the Lafayette Instrument Grooved Pegboard Test (Model #32025). This test measures speed and accuracy of hand-eye coordination. This test requires more complex visual-motor coordination, as it consists of 25 holes with randomly positioned slots. The pegs, which look like a key with a round side and a square side, must be rotated to match the hole before they can be inserted (Lafayette Instrument, 2002). The test is scored by the length of time, in seconds, required to complete each trial. Norms are available for age and sex (Trites, 1977). Normative data for each hand (dominant and non-dominant) was used to calculate a z-score ($M=0$, $SD=1$) and standardized score for each hand ($M=100$, $SD=15$).

**Social-emotional processing.** The Diagnostic Analysis of Nonverbal Accuracy 2 (DANVA2) was used to assess social-emotional processing. Two subtests from the
DANVA2 were administered in which the participant was required to label the stimulus as “happy, sad, angry, or fearful”. The first subtest, the Child Facial Expression Test, consisted of 24 photographs of child facial expressions; 12 female and 12 male showing an equal number of high and low intensity emotions. The subtest has good internal consistency, with coefficient alphas ranging from .69 to .81 (Nowicki, 2003). The second subtest, Child Paralanguage Test, includes 32 voice trials, with an equal number of male and female voices for each of the four high and four low intensity trials of each emotion. Scores have shown to be internally consistent for eight-year-old (alpha = .74) and ten-year-old (alpha = .76) children (Nowicki, 2003). For each subtest, age-based normative data was used to calculate a z-score (M=0, SD=1) and standardized score (M=100, SD=15).

**Social-contextual language.** Two subtests from the Comprehensive Assessment of Spoken Language (CASL) were used to assess social-contextual language skills (Carrow-Woolfolk, 1999). The CASL is a norm-referenced oral language assessment battery of tests for children and young adults aged 3 through 21 years old. The Inference subtest was used to measure comprehension of complex language in which meaning is not directly available from lexical or grammatical information. For this subtest participants were asked to answer questions that rely on contextual cues. For example, one item from this subtest states, "Before Jim left for work, he put on a heavy woolen coat. What was the weather like?”. The Pragmatic Judgment subtest was also used to measure awareness of the appropriateness of language in relation to the situation in which it is used. For example, one item from this subtest asks, "Suppose the telephone rings.
You pick it up. What do you say?". Adequate internal reliability is reported for each subtest of the CASL (Chronbach’s alphas range from .64 to .94). Standardized scores are provided for each subtest with a mean of 100 and a standard deviation of 15.

**Predictors of the Neuropsychological Profile Cluster Membership**

**Demographics.** Mother questionnaire data were used to assess the child's age and ethnicity. This information was gathered with the Parent Demographic Questionnaire (PDQ), which was developed for this study. This questionnaire was designed to assess a variety of demographic information about the child, caregiver(s), and family.

**SES.** The PDQ was completed by the child's parents and also included questions about the caregiver's employment status, marital status, education, occupation, and income. The Hollingshead Four Factor Index of socioeconomic status was used to assess SES (Hollingshead, 1975). SES was derived by assigning a score to mothers’ and fathers’ occupations and education level. Education and occupation scores were combined and these scores were averaged across caregivers to calculate the family SES. In the case of single-parent families, or two-parent families in which only one parent was employed, that individual’s score was used to represent the family. Family SES scores range from 8-66 and higher scores reflect higher SES.

**Enrichment of the child’s environment.** The Family Environment Scale (FES, Form R) was completed by the child’s mother and father (Moos & Moos, 1994). It measures people’s perceptions of their actual family environments. It includes three dimensions: relationship, personal growth, and system maintenance. The current study will use the achievement orientation subscale from the personal growth dimension. This
The achievement orientation subscale from the personal growth dimension had poor reliability ($\alpha = .39-6.63$).

The Family Stress Scale (FSS) was also used to measure the enrichment of the child’s environment (Quittner, Glueckauf, & Jackson, 1990). It is a 19 item questionnaire that assesses common stressors in families with children with spina bifida, on a 5 point scale. Higher scores indicate higher amount of perceived stress. There are 13 non-disease specific items (e.g., outings in the community) and 6 disease-specific items (e.g., catheterization). It was completed by mothers and fathers. The FSS showed good internal consistency ($\alpha = .88$ to .92) in the current study.

Medical information. The Medical History and Adherence Questionnaire was adapted from the Parent-Report of Medical Adherence in Spina Bifida Scale (PROMASB, Holmbeck et al., 1998), which was developed for a previous study on youth with spina bifida by the same investigator. The measure is designed to obtain disease-specific medical information. Information about the adolescent's lesion level, shunt status, history of shunt surgeries, and seizure history was gathered from the Medical History and Adherence Questionnaire, which was completed by the youth's parents.

Outcomes of the Neuropsychological Profile

Independence. The Scales of Independent Behavior-Revised (SIB-R; Bruininks, Woodcock, Weatherman, & Hill, 1996) was used to assess an individual's level of independent functioning. The SIB-R is a norm-referenced measure that assesses fourteen
areas of adaptive behavior (e.g., gross-motor skills, language comprehension, money and value) and eight areas of maladaptive behavior (e.g., socially offensive behavior, destructive to property). In the current study, parents completed a checklist on four of the adaptive behavior subscales: Fine-Motor, Money and Value, Language Comprehension, and Time and Punctuality. Each item was ranked on a four-point Likert scale, indicating how well the child completes each task: (0) never or rarely, even if asked, (1) does, but not well, or about ¼ of the time, may need to be asked (2) does fairly well, or about ¾ of the time, may need to be asked, and (3) does very well, always or almost always, without being asked. The total raw score was used for each subscale, with a higher score indicating greater independence. Excellent internal consistency was found for the current study ($\alpha = .92-.95$).

**Academic success.** Teachers of participants in this study completed the Teacher Report Form (TRF; Achenbach & Rescorla, 2001). The TRF is comprised of 118 problem items and is provides normative data for children ages 6-18. The TRF yields adaptive functioning subscales. To assess academic success, the academic performance subscale was used in this study. For this scale, T-scores were used, where a higher t-score would indicate greater academic success.

**Parental expectations for the future.** Questions about the Future-P, is a parent-reported questionnaire that was used to assess parental expectations for his/her child's future. The Questions about the Future questionnaire was developed for a previous study on youth with spina bifida. The measure asks the respondent to rate statements about the child’s future on a four-point scale, from 1 (very unlikely) to 4 (very likely). Eight
statements reflect future employment and educational achievement, transportation, living independence, relationships, and ability to have and raise children. The mean score from this measure was used in subsequent analysis. Thus, a higher score indicated the parent expected his/her child to achieve more developmental and independence milestones in the future. Internal consistency was excellent in the current study (α = .94-.95).

Quality of life. Youth with spina bifida and their parents completed a questionnaire measure of the youth’s quality of life, the Pediatric Quality of Life Inventory Version 4.0 Generic Core Scales (PedsQL; Varni, Seid, & Kurtin, 2001). This questionnaire measures health-related quality of life in children and adolescents ages 2 to 18 years old. The measure consists of 23 items applicable for pediatric populations with acute and chronic health conditions that load onto 5 scales: physical health, emotional functioning, social functioning, school functioning, and psychosocial health (which is a combination of emotional, social, and school functioning). This measure utilizes a five-point Likert scale with response categories ranging from ‘0- never a problem’ to ‘4- almost always a problem’. In the current study, scores from the first 4 scales were used in the analyses. Adequate internal consistency has been demonstrated in the current study for each of the scales for parent report (α = .59-.82) and child report (α = .65-.72).

Statistical Treatment

Data Analyses

A cluster analysis was used to determine whether subgroups of children with similar neuropsychological profiles exist within the larger group of children with spina bifida. Cluster analysis is a "person centered" statistical tool that identifies groups of
individuals with similar scores on multiple dimensions. There is no significance test in cluster analysis and thus a power analysis is inapplicable. One issue with cluster analysis is that the analysis will provide a cluster solution whether or not subgroups are actually present in the sample (Steel & Aylward, 2007). Thus, the following precautions were taken to maximize external validity of findings in this study. Henry, Tolan, and Gorman-Smith (2005) suggest standardizing all measures used in a cluster analysis. Thus, only age-normed standard scores (mean= 100, standard deviation= 15) were used in the cluster analysis. Hierarchical and nonhierarchical analyses, as outlined by Steele & Aylward (2007), Henry and colleagues (2005), and Fisher and colleagues (2000), were used to identify and confirm the number of subgroups. Hierarchical clustering with Ward's linkage was used to identify the subgroups and average-linkage and $K$-means analysis were used to confirm the solution. Additionally, variables that were not used to develop the clusters were used to predict cluster membership. As well, the cluster solution was used to predict outcome variables (e.g., independence). These predictor and outcome analyses also provide information about the validity of the cluster solution (Henry et al., 2005). If group membership was found to be a significant predictor of related outcomes in the directions predicted, then the validity of the subgroups would be supported. To determine subgroup distinctions, the mean scores of each subgroup were examined. A cognitive profile of strengths and weaknesses was identified for each subgroup and subgroups were labeled to reflect the average to low average cognitive profile. If the first hypothesis was not supported and a different cluster solution was identified, hypotheses II and III would be altered appropriately.
A multinomial logistic regression was used to evaluate associations between the predictor variables and cluster categories. ANOVA and MANOVA analyses were used to evaluate associations between group membership and outcome variables. Power analyses were conducted using GPower (Faul, Erdfelder, Lang, & Buchner, 2007) to assess what sample size is appropriate for the proposed statistical analyses. Assuming a power of .90, an alpha of .05, a sample of 59 would be required to detect a medium effect size with the most complicated regression analyses. A sample of 84 participants would be required to detect a large effect size with the most complicated ANOVA analyses, assuming that there are indeed 4 subgroups. For the MANOVA analysis, if there are 4 subgroups, a sample size of 96 is required to detect a large effect size. Thus, the current study had the sample size necessary to detect medium to large effects. If a different cluster solution was identified with more than 4 groups, these power analyses would be revisited.

**Hypothesis I**

Four cluster groups were hypothesized to emerge from the analysis. The mean neuropsychological profile for each of these subgroups would be exemplified by variability and higher functioning (cluster 1), variability and lower functioning (cluster 2), consistency and higher functioning (cluster 3), and consistency and lower functioning (cluster 4; see Figure 2 and 3).

To test the first hypothesis, hierarchical and nonhierarchical analyses were conducted, as outlined by Steele & Aylward (2007) and Henry and colleagues (2005), to identify and confirm the number of subgroups. The following 22 variables were subjected to hierarchical cluster analysis: (1) Verbal IQ (WASI vocab), (2) Non-verbal IQ (WASI
matrix reasoning), (3) Math (WRAT arithmetic), (4) Word reading (WRAT reading), (5) Spelling (WRAT spelling), (6) Non-verbal executive functioning (CAS planned connections), (7) Letter fluency (D-KEFS letter fluency), (8) Category fluency (D-KEFS category fluency), (9) Category switching fluency (D-KEFS category switching), (10) Category switching accuracy (D-KEFS category switching), (11) Visual selective attention (TEA-CH sky search), (12) Visual attention efficiency (TEA-CH sky search), (13) Attention score (controlled for motor ability, TEA-CH sky search), (14) Verbal sustained attention (TEA-CH score), (15) Multi-modal (visual/verbal) divided attention (TEA-CH sky search dual task), (16) Verbal divided attention (TEA-CH score dual task), (17) Dominant fine-motor (Grooved pegboard), (18) Non-dominant fine motor (Grooved pegboard), (19) Non-verbal emotion recognition (DANVA faces), (20) Verbal emotion recognition (DANVA paralanguage), (21) Inferences (CASL), and (22) Pragmatic judgment (CASL). Hierarchical methods were used to identify the number of clusters that maximized differences between clusters on the neuropsychological variables, then a nonhierarchical cluster analysis ($k$-means) was used to confirm the number of clusters identified by Ward's method. This method provided a robust identification of clinically meaningful clusters of participants. Because this was an exploratory analysis, it was possible a different cluster solution would emerge. If a different cluster solution was identified, hypotheses II and III would be altered appropriately.

**Hypothesis II**

Biological factors (lesion level, number of shunt surgeries, and seizures), sociodemographic factors (age and ethnicity), and environmental factors (socioeconomic
status, family stress, and family environment) were expected to predict group membership. Biological factors were expected to predict the level of functioning (low vs. high), such that individuals with more severe biological factors are expected to be members of clusters 2 and 4, whereas individuals with fewer biological risk factors were hypothesized be members of clusters 1 and 3 (see Figure 2 and 3). Socio-demographic factors were hypothesized to predict group membership, such that older individuals and non-Hispanic individuals would more likely be in cluster 1 or 2 (more variable profiles), whereas younger individuals and Hispanic individuals will more likely be in cluster 3 or 4 (less variable profiles; see Figures 2 and 3). Finally, it was hypothesized that higher SES individuals would more likely be a member of clusters 1 or 2 (more variable profile), whereas lower SES individuals, individuals with less personal growth in their family environment, and higher family stress would more likely be in cluster 3 or 4 (less variable profiles; see Figures 2 and 3).

A multinomial logistic regression was conducted to evaluate associations between the predictor variables and cluster categories, to determine how accurately we could predict group membership based on the predictor variables. The dependent variable was group status (individual's cluster). The predictors were lesion level, number of shunt surgeries, history of seizures, age, ethnicity, SES, personal growth in the family environment, and family stress.

**Hypothesis III**

Group membership was expected to be associated with different levels of independence, academic success, expectations for the future, and quality of life (QOL).
Specifically, it was expected subgroups would be associated with outcome variables in the following order from the highest level of outcome to the lowest: variability and higher functioning (cluster 1), variability and lower functioning (cluster 2), consistency and higher functioning (cluster 3), and consistency and lower functioning (cluster 4; see Figures 2 and 3).

ANOVAs or MANOVAs were conducted to determine whether the identified clusters differed on each of the following outcomes: independence, academic success, expectations for the future, and quality of life. Because these outcome variables were unrelated separate constructs, they were not combined into a single MANOVA analysis. First, a MANOVA was conducted to determine whether the clusters differ on the level of independence. For this analysis, group (cluster) status was used as the independent variable (IV) and the 4 subscales from the SIB-R were used as the dependent variables (DVs). To assess differences in academic success, an ANOVA analysis was conducted, such that group status was the IV and the academic performance subscale from the TRF was the DV. An ANOVA was also used to identify differences in parental expectations for the future, such that group status was the IV and the mean parental expectations score was the DV. If the mother and father reports on this measure were significantly correlated, then they would be combined to include one composite score for parental expectations of the future. If the parents’ scores were not correlated, then two separate ANOVAs would be run to assess mother and father expectations for the future. Differences in QOL were assessed with 2 MANOVA analyses (parent and child report separately), such that group status was the IV and the each scale scores (4 total) for child-
reported QOL were the DVs. If parent and child report were found to be significantly correlated, they would be combined and only one MANOVA would be conducted. If any of these analyses were found to be significant, then post-hoc tests of group differences were used to determine which group means differ significantly from others.
CHAPTER FOUR

RESULTS

Preliminary Analyses

Demographics

As previously discussed, analyses were run with participants who completed the entire battery. As seen in Table 3, preliminary analyses revealed there were no significant differences between those who did and did not complete the neuropsychological battery on the following characteristics: age, SES, race, and IQ (WASI full, 2-scale IQ). There were significant differences in gender and shunt status, such that a greater percentage of males and children with shunts completed the battery and were included in this study (see Table 3).

Standardization of Cluster Variables

Prior to conducting the cluster analysis, all of the neuropsychological scores were converted to standard scores. While all of the scores were standardized, some of them were in the form of z-scores, t-scores, or scaled scores. To be able to compare scores across measures, they were converted to the same type of standard score (m=100, sd=15). Converting the scores to the same scale also reduced the chance that the cluster analysis would prioritize variables with a larger range in their scores. For example, a difference between standard scores 85 and 100 would appear to be a greater distance than between scaled scores 7 and 10.
Combining Mother and Father Report

Prior to examining the main hypotheses of the study, the relationship between mother and father report on questionnaire measures was examined. Mother and father report were significantly correlated for all questionnaire scales (SIB-R, future expectations, and quality of life; $r = .40$ to $.87, p < .01$). Thus, mother and father report were combined to form a composite measure of parent report. This composite score was used in all of the following analyses. Child report was not significantly correlated with parent report, and thus was run separately in subsequent analyses.

Outliers

Univariate outliers were examined for each neuropsychological, predictor, and outcome variable. An outlier was defined as a score greater than 3.29 standard deviations from the mean (Tabachnick & Fidell, 2013). One outlier was identified among the neuropsychological variables: one participant's score on DANVA language was -3.41 standard deviations from the mean. After reviewing the raw data, it was decided to keep the outlier in the dataset, as it seemed valid, and was not extreme. Several outliers were identified for grooved pegboard. Because the standard score was calculated with a population mean and standard deviation, some research participants performed very poorly and received negative standard scores. As most standard scores are positive, a negative standard score would be difficult to interpret. Tabachnick and Fidell (2013) suggest one way to adjust an outlier whereby the outlier score is adjusted to remain deviant, but not as deviant. Thus, any grooved pegboard standard score <20 was changed to a standard score of 20. A standard score of 20 was chosen, because this score is
possible, but still extremely low (-5.33 SD from the mean). Outliers were also identified
for number of shunt surgeries. Thus, three participants with more than 8 shunt surgeries
were recoded to 8 shunt surgeries. Finally, one outlier was identified for the Family
Stress Scale. This participants' score was changed to one more than the next extreme
score (Tabachnick & Fidell, 2013).

**Skewness**

Skewness was examined for all predictor and outcome variables. The predictor
variable "number of shunt surgeries" was skewed after outliers were adjusted. However,
this variable was not adjusted for skewness because it would be difficult to interpret the
"square root" or "logarithm" of number of surgeries (Tabachnick & Fidell, 2013).

**Multivariate Outliers**

Multivariate outliers were examined using methods described by Tabachnick &
Fidell (2013). Among the 87 participants with complete data for predictor variables, there
were no multivariate outliers.

**Hypothesis I**

It was hypothesized that four cluster groups would emerge from the analysis.
Varied performance across measures of associative and assembled processing would
result in four distinct subgroups. The mean neuropsychological profile for each of these
subgroups would be exemplified by variability and higher functioning (cluster 1),
variability and lower functioning (cluster 2), consistency and higher functioning (cluster
3), and consistency and lower functioning (cluster 4; see Figure 2 and 3).
To test the first hypothesis, hierarchical and nonhierarchical analyses were conducted, as outlined by Steele & Aylward (2007) and Henry and colleagues (2005), to identify and confirm the number of subgroups. The following 22 variables were subjected to hierarchical cluster analysis: (1) Verbal IQ (WASI vocab), (2) Non-verbal IQ (WASI matrix reasoning), (3) Math (WRAT arithmetic), (4) Word reading (WRAT reading), (5) Spelling (WRAT spelling), (6) Non-verbal executive functioning (CAS planned connections), (7) Letter fluency (D-KEFS letter fluency), (8) Category fluency (D-KEFS category fluency), (9) Category switching fluency (D-KEFS category switching), (10) Category switching accuracy (D-KEFS category switching), (11) Visual selective attention (TEA-CH sky search), (12) Visual attention efficiency (TEA-CH sky search), (13) Attention score (controlled for motor ability, TEA-CH sky search), (14) Verbal sustained attention (TEA-CH score), (15) Multi-modal (visual/verbal) divided attention (TEA-CH sky search dual task), (16) Verbal divided attention (TEA-CH score dual task), (17) Dominant fine-motor (Grooved pegboard), (18) Non-dominant fine motor (Grooved pegboard), (19) Non-verbal emotion recognition (DANVA faces), (20) Verbal emotion recognition (DANVA paralanguage), (21) Inferences (CASL), and (22) Pragmatic judgment (CASL).

SPSS (v21.0, released in 2012) was used for all analyses. Squared Euclidean distance was used as the similarity measure because it considers elevation of scores in addition to pattern of scores (e.g., it would group people with high standard scores separately from people with low standard scores). Ward’s clustering method was chosen for the first cluster analysis because it maximizes between group differences, while also
minimizing within group differences. It is also very commonly used in the behavioral literature (Clatworthy, Buick, Hankins, Weinman, & Horne, 2005). It achieves a cluster solution by minimizing the within-group sum of squared Euclidean distances between each individual and its cluster mean, at each stage.

Because cluster analysis is an exploratory method, the most conservative precautions were used to support the stability of the cluster solution, as exemplified by Fisher and colleagues (2000). The cluster analysis incorporated 3 separate analyses. First, a hierarchical, agglomerative clustering method (Ward’s method) was used to identify a cluster solution. Ward’s method is an agglomerative method, which starts with as many groups as there are participants and combines similar subjects or groups until there is only a single cluster. The most appropriate cluster solution is decided by examining the agglomeration coefficients for a significant "jump" in value. This "jump" indicates a combining of clusters that were not in fact similar (Aldenderfer & Blashfield, 1984). Second, as recommended by Borgen and Barnett (1987), another method of hierarchical clustering (average linkage, between groups) was used to validate the first cluster solution. Average linkage is similar to Ward's method because it is also an agglomerative method, however it uses different criteria to determine similarity between an individual and a cluster. The average linkage method considers the average distance between participants' data in one cluster vs. another. Whereas, Wards method considers the increase in sum of squares within clusters when they are combined (Aldenderfer & Blashfield, 1984). The stability of the cluster solution is supported if both hierarchical analyses indicate the same number of clusters with similar cluster membership (Borgen &
Barnett, 1987). Last, a nonhierarchical analysis was conducted (K-means), which groups participants based on a specified number of clusters. The cross-method stability of the cluster solution is further supported if the nonhierarchical analysis results in similar cluster profiles. To determine whether the cluster profiles are similar, the mean profile scores for each cluster were plotted and examined. Additionally, overlap between one clustering method and another was determined by recording each participant's cluster membership (as exemplified by Steele, Dreyer, & Phipps, 2004 and Fisher et al., 2000).

**Cluster Analysis**

Ward's method indicated that a three-cluster solution best fit the data. A large increase in the agglomeration coefficient suggests that two very distinct clusters have been combined. The agglomerative or grouping, schedule provided by Ward's method indicated a notable increase in agglomeration statistic after the three-cluster solution. When 3 clusters were reduced to 2 clusters the agglomeration coefficient increased by 85,300, which is compared to relatively trivial increases (i.e., 47,439; 42,135; etc; see Table 5). The mean scores for each cluster, based on Ward's method, are shown in Table 6. The standard score profiles are presented in Figure 4.

The average linkage within-in groups method also indicated a three-cluster solution, due to the notable increase in the agglomeration statistic after the three-cluster solution. When 3 clusters were reduced to 2 clusters the agglomeration coefficient increased by 2068, which is compared to relatively trivial increases (i.e., 323, 438, etc; see Table 7). This large increase indicated 2 very different clusters were combined. Mean profile scores for each of the three clusters were plotted. The mean profile for each of the
three clusters generated by the average linkage method paralleled the profiles generated by Ward's method (see Figures 4 and 5). In addition 82% of the participants classified by Ward's method were classified in a similar cluster generated by the average linkage method (see Table 8). This level of consistency is greater than that found to be adequate in previous studies, i.e., 69.2% and 73% in Fisher and colleagues, 2000 and Steel and colleagues, 2004, respectively. Thus, the cluster solution developed by Ward's method was replicated statistically, using a second agglomerative method.

K-means, set at a three-cluster solution, also created similar cluster profiles, as those created by Ward's method (see Figures 4 and 6). Additionally, 81% of the participants classified by the Ward's method were classified in a similar cluster generated by the K-means analysis (see Table 9). This level of consistency is greater than that found to be adequate in previous studies, i.e., 70% in Fisher et al., 2000. Thus, the three cluster solution was replicated using K-means, a non-hierarchical method.

The first hypothesis was not supported, as an alternative cluster solution was identified. Still, given the statistical reliability of the clusters, a label or description was developed for each cluster, based on the group's mean profile (see Table 6 and Figure 4). Weschler (1999) provides the following classifications for standard score ranges: below 69 "extremely low", 70-79 "borderline", 80-89 "low average", 90-109 "average", 110-119 "high average", 120-129 "high". These classifications were used to interpret the cluster profiles. Clusters from the original clustering method (Ward’s method) are described below.
Cluster 1 "average to low average cognitive, impaired motor" (n = 39, 41%).

These participants performed in the average range on measures of intelligence, academic achievement, and social-emotional processing (see Table 6). They performed in the low average to average range on measures of executive functioning and social-contextual language. Most notably, they performed in the extremely low range on measures of fine motor ability. For the most part, their attention performance was in the average range, except for measures of visual selective attention and efficiency. The efficiency subtest, which is a timed task that requires participants to circle items with a pen, may have been effected by the participants' fine motor impairments. It is also possible that their fine motor impairments hindered their performance on the executive functioning task: CAS Planned Connections, which is also a timed task that requires participants to draw lines between boxes. Based on this profile of strengths and weaknesses, this cluster was labeled "Average to Low Average Cognitive, Impaired Motor".

Cluster 2 "average to low average cognitive" (n = 32, 33%). Participants in cluster 2 performed in the average range on measures of intelligence, academic achievement, social-emotional processing, and social-contextual language (see Table 6). They also performed in the average range on measures of executive functioning, except for one measure that required fine motor skills (CAS Planned Connections), which may have been affected by their fine motor ability. While their fine motor performance was markedly higher than cluster 1 (see Figure 4), it was still in the borderline range. Interestingly, these participants performed in the low average range on several measures of attention (auditory and dual attention). They performed in the average range on
measures of visual attention. Due to the this profile of scores, this cluster was labeled "Average to Low Average Cognitive".

**Cluster 3 "extremely low to borderline" (n = 25, 26%).** Participants in cluster 3 performed in the extremely low to borderline range on all measures except for 2 subtests: WRAT spelling (low average) and TEACH number of identified targets (average). The most notable aspect of this cluster's profile is their consistent performance in the extremely low to borderline range. Thus, this cluster was labeled "Extremely Low to Borderline".

**Modification of Hypothesis II**

It was hypothesized that biological factors (lesion level, number of shunt surgeries, and seizures), socio-demographic factors (age and ethnicity), and environmental factors (socioeconomic status, family environment, family stress) would predict group membership. Because an alternate cluster solution was identified, the following hypothesis has been altered to reflect the new cluster solution.

**Biological Factors**

Because higher lesion level is associated with greater motor impairment (Erickson et al., 2002), the new hypothesis suggested lesion level would predict performance on motor tasks, such that individuals with high lesion levels would be members of clusters 1 and 3, whereas individuals with lower lesion levels would be more likely to be members of cluster 2 (see Figure 4). Number of shunt surgeries and seizures would predict performance on cognitive tasks, such that those with greater number of shunt surgeries and positive seizure history would be associated with cluster 3, whereas individuals with
fewer shunt surgeries and negative seizure history would be more likely to be members of clusters 1 and 2.

**Socio-demographic Factors**

It was hypothesized that socio-demographic factors would predict performance on associative processing tasks (e.g., verbal IQ, spelling, and reading). Specifically, it was thought that these factors would predict whether there was a split between verbal and non-verbal abilities. Cluster 2 was the only cluster without a notable difference between verbal and non-verbal measures of IQ and academic achievement (see Figure 4). Thus, it was hypothesized that younger individuals and Hispanic individuals would more likely be members of this cluster (cluster 2), whereas older individuals and non-Hispanic individuals would more likely be members of cluster 1 or 3 (more variable profile).

**Environmental Factors**

Finally, it was hypothesized that higher SES, greater personal growth in the family environment, and less family stress would be associated with better performance on tasks of associative processing (including verbal abilities). Specifically, individuals with higher SES, greater personal growth in the family environment, and less family stress would more likely be a member of clusters 1 or 3 (more variable profile), whereas other individuals would more likely be in cluster 2.

**Hypothesis II Results**

A logistic regression was conducted to evaluate associations between the predictor variables and cluster categories, to determine how accurately group membership is predicted by the predictor variables. The dependent variable was group
status (individual's cluster membership). The predictors were lesion level, number of shunt surgeries, history of seizures, age, ethnicity, SES, personal growth in the family environment, and family stress. The logistic regression produced an error indicating a quasi-complete separation in the data. In other words, one of the predictors or a combination of the predictors nearly perfectly predicted cluster membership. It was determined that seizure status was the predictor causing this issue, as there was no error when seizure status was removed. Thus, seizure status was isolated so that two regressions were completed, one with seizure status and one with all other predictors.

First, a logistic regression with all variables except for seizure status was conducted. The results indicated the model explained a significant amount of the original variability $\chi^2(18) = 33.93, p<.05$, and was a good fit of the data. Of the 7 predictors, SES had a significant main effect on cluster membership $\chi^2(2) = 11.77, p<.01$. More specifically, SES significantly predicted whether a participant was placed in either the "average to low average cognitive, impaired motor" group or the "extremely low to borderline" group, $b = 0.09$, Wald $\chi^2(1) = 8.79, p<.01$. As well, SES significantly predicted whether a participant was placed in the "average to low average cognitive, impaired motor" group or the "average to low average cognitive" group, $b = 0.06$, Wald $\chi^2(1) = 4.78, p<.05$. The models suggested that participants with higher SES were more likely to be placed in the "average to low average cognitive, impaired motor" than either of the other two groups.

A second logistic regression was complete with seizure status as the predictor and cluster membership as the outcome. The results showed the model explained a significant
amount of the original variability $\chi^2(2) = 6.42, p<.05$, and was a good fit of the data. Seizure status had a significant main effect on cluster membership $\chi^2(2) = 6.42, p<.05$. More specifically, seizure status significantly predicted whether a participant was placed in either the "average to low average cognitive, impaired motor" group or the "extremely low to borderline" group, $b = 0.09$, Wald $\chi^2(1) = 8.79, p<.01$. As well, seizure status significantly predicted whether a participant was placed in the "average to low average cognitive" group or the “extremely low to borderline” group, $b = 2.34$, Wald $\chi^2(1) = 4.35, p<.05$. The models suggested those with a history of seizures were more likely to be placed in the "extremely low to borderline" group.

The second hypothesis was partially supported. When all predictors were included in the model, the model significantly predicted group membership. Further analyses indicated SES and seizure status were the only predictors with a significant main effect. All other hypothesized effects were not supported.

**Modification of Hypothesis III**

It was hypothesized that group membership would predict levels of independence, academic success, expectations for the future, and quality of life. Because an alternate cluster solution was identified, the following hypothesis has been altered to reflect the new cluster solution. Specifically, subgroups would be associated with outcome variables in the following order from the highest level of outcome to the lowest: "average to low average cognitive" (cluster 2), "average to low average cognitive, impaired motor" (cluster 1), and "extremely low to borderline" (cluster 3; see Figure 4).
Hypothesis III Results

Independence

A MANOVA was conducted to examine the association between cluster membership and level of independence. The four scales from the SIB-R were used as dependent variables. Using Wilk's statistic, the results suggested that cluster membership significantly predicted level of independence $\lambda = 0.81, F(8,172) = 2.41, p < .05$. Separate univariate ANOVAs on the outcome variables revealed significant effects of cluster membership on each scale individually: fine motor, $F(2, 89) = 4.82, p < .05$; money $F(2, 89) = 8.44, p < .01$; language $F(2, 89) = 7.14, p < .01$; and time $F(2, 89) = 4.69, p < .05$. Post-hoc tests revealed participants in the "average to low average cognitive" group had significantly greater levels of independence than those in the "extremely low to borderline" group, for each subscale: fine motor ($p < .05$), money ($p < .01$), language ($p < .01$), and time ($p < .05$). Participants in the "average to low average cognitive, impaired motor" cluster also had significantly greater levels of independence than those in the "average to low average cognitive, impaired motor" cluster, for the fine motor ($p < .05$) and money ($p < .05$) subscales (see Table 10).

Academic Success

An ANOVA was run to test the association between cluster membership and academic success, with teacher reported academic success as the dependent variable. The Levene statistic indicated the variances of the clusters were not homogeneous. Thus, Welch's F and Games-Howell post hoc statistics were reported, as these measures are more robust when the assumption of equal variances is violated. Results indicated that
group status significantly predicted academic success, Welch's $F(49.6) = 17.22$, $p < .01$.

Post-hoc analyses revealed significantly less academic success for participants in the "extremely low to borderline" group than those in the "average to low average cognitive, impaired motor" group ($p < .01$) and the "average to low average cognitive" group ($p < .01$; see Table 10).

**Expectations for the Future**

An ANOVA was run to test the association between cluster membership and expectations for the future, with parent reported future expectations as the dependent variable. The Levene statistic indicated the clusters had unequal variances, and Welch's F and Games-Howell post hoc statistics were reported. Results suggested group status significantly predicted parental expectations for the future Welch's $F(51.86) = 14.53$, $p < .01$. Post-hoc analyses indicated significant differences between each group ($p = .00$ to .04), such that parents of participants in the "average to low average cognitive" group had the highest future expectations, followed by participants in the "average to low average cognitive, impaired motor" group, and finally the "extremely low to borderline" group (see Table 10).

**Quality of Life**

Two MANOVAs were conducted to examine the relationship between cluster membership and quality of life. Parent and child report were analyzed separately. The dependent variables included the 4 subscales for each measure of QOL. Using the Wilk's statistic, results indicated cluster membership did not have a significant effect on parent reported quality of life, $\lambda = 0.85$, $F(8,176) = 1.83$, $p = .08$. However, cluster membership
significantly predicted child reported quality of life, $\lambda = 0.83$, $F(8,172) = 2.12$, $p < .05$.

Follow-up univariate ANOVAs revealed cluster membership significantly predicted the physical scale, $F(2, 90) = 5.18$, $p < .01$, but none of the other scales. Post-hoc tests indicated participants in the “average to low average cognitive” group reported significantly greater physical quality of life than participants in the "extremely low to borderline" group ($p < .01$).

In sum, the third hypothesis was mostly supported. The effects of group membership on independence, academic success, expectations for the future, and child-reported QOL were significant. Parent reported QOL was the only outcome not predicted by cluster membership. Additionally, most results were in the hypothesized direction (cluster 2, 1, 3 in order from highest to lowest outcome; see Table 10). Academic success was the only significant scale for which the results were not in the hypothesized direction, as group 1 (“average to low average cognitive, impaired motor”) was rated as having higher academic success than group 2 (“average to low average cognitive”).

**Exploratory Analyses**

The following analyses were not proposed at the beginning of the study, but were included after the analyses were completed. During the preliminary analyses, it was discovered that several participants ($n=12$) were not able to complete the neuropsychological battery due to low comprehension. Thus, this group of participants was seen as qualitatively different from participants included in the cluster analyses. It was determined that including these participants as a 4th group would be clinically meaningful because they represent youth with spina bifida who are so low functioning
that they are not able to understand and implement the tasks for the neuropsychological battery. Additionally, including these participants in subsequent analyses would contribute to the literature as participants in this range of functioning are often excluded from research (e.g., excluded IQ: <70 Dennis et al., 1981; <70, Hampton et al., 2011; <90, Iddon et al., 2004; <70 Lindquist et al., 2009; <80, Snow, 1999; <75, Vinck et al., 2006).

**Four Cluster Solution (with "Non-completers")**

The previous cluster analysis revealed a 3 group solution as the best fit for the data. However, the cluster analysis only included participants who completed the entire neuropsychological battery. Thus, the following analyses were completed to examine predictors and outcomes of the cluster solution, including the 4th group, which was labeled “non-completers.”

**Predictors of the 4 cluster solution.** It was hypothesized that biological factors (lesion level, number of shunt surgeries and seizure history), socio-demographic factors (age and ethnicity), and environmental factors (socioeconomic status, family environment, family stress) would predict group membership. To build upon the previous analyses, it was predicted group four would be composed of participants with higher lesion levels, more shunt surgeries, positive seizure history, younger age, Hispanic ethnicity, lower SES, less personal growth in the family environment, and less family stress. The logistic regression again produced an error indicating a quasi-complete separation in the data due to the seizure status variable. Thus, two regressions were completed again, one with seizure status and one with all other predictors.
A logistic regression with all variables except for seizure status indicated the model explained a significant amount of the original variability $\chi^2(27) = 52.26, p<.01$, and was a good fit of the data. Of the 7 predictors, SES had a significant main effect on cluster membership $\chi^2(3) = 12.43, p<.01$. However, post-hoc analyses revealed SES did not significantly distinguish the “non-completers” group from any other group. A second logistic regression was complete with seizure status as the predictor and the 4-group cluster membership as the outcome. The results showed seizure status had a significant main effect on cluster membership $\chi^2(3) = 17.85, p<.01$. More specifically, seizure status significantly predicted whether a participant was placed in either the "average to low average cognitive, impaired motor" group or the "non-completers" group, $b = -2.58$, Wald $\chi^2(1) = 9.53, p<.01$. As well, seizure status significantly predicted whether a participant was placed in the "average to low average cognitive" group or the “non-completers” group, $b = -3.84$, Wald $\chi^2(1) = 10.17, p<.01$. The models suggested those without a history of seizures were more likely to be placed in the "average to low average cognitive, impaired motor" group and the “average to low average cognitive” group rather than the "non-completer" group. Thus, the hypothesis was partially supported, as only seizure history predicted group membership.

**Four cluster solution predicting outcomes.** It was expected group membership would predict levels of independence, academic success, expectations for the future, and QOL. Specifically, it was expected the additional cluster would be associated with the lowest scores across all outcome measures. As these analyses have previously been
completed with the 3 cluster solution, only post-hoc findings entailing the fourth cluster will be reported here. For a full summary of means for each outcome see Table 11.

A MANOVA was conducted to examine the association between cluster membership and level of independence. The four scales from the SIB-R were used as dependent variables, and the 4 cluster solution was used as the independent variable. Using Wilk's statistic, the results suggested that cluster membership significantly predicted level of independence $\lambda = 0.58$, $F(12, 249) = 4.73$, $p < .01$. The Levene statistic indicated the fine motor subscale had unequal variances. Thus, Games-Howell post hoc statistics were reported, as it is more robust when the assumption of equal variances is violated. Separate univariate ANOVAs revealed significant effects of cluster membership on each scale individually: fine motor, $F(3, 97) = 6.14$, $p < .01$; money $F(3, 97) = 8.28$, $p < .01$; language $F(3, 97) = 10.70$, $p < .01$; and time $F(3, 97) = 14.19$, $p < .01$. Post-hoc tests revealed for independence with money, participants in the "average to low average cognitive" group had significantly higher levels of financial independence than participants in the "non-completer" group ($p < .01$). Participants in the "non-completers" group also showed significantly lower amounts of independence with language, as compared to those in the "average to low average cognitive, impaired motor" group ($p < .01$) and the "average to low average cognitive" group ($p < .01$). Finally, participants in the "non-completers" group showed significantly less independence with time than participants in any of the other three groups: "average to low average cognitive, impaired motor" ($p < .01$), "average to low average cognitive" ($p < .01$), and "extremely low to borderline" ($p < .01$; see Table 11).
Four cluster solution predicting academic success. An ANOVA was run to test the association between cluster membership and academic success, with teacher reported academic success as the dependent variable. The Levene statistic indicated the variances of the clusters were not homogeneous. Thus, Welch's F and Games-Howell post hoc statistics were reported. Results indicated that group status significantly predicted academic success, Welch's $F(44.91) = 24.09, p<.01$. Post-hoc analyses revealed significantly less academic success for participants in the "non-completers" group than those in the "average to low average cognitive, impaired motor" group ($p<.01$), and the "average to low average cognitive" group ($p<.01$; see Table 1).

Four cluster solution predicting expectations for the future. An ANOVA was run to test the association between cluster membership and expectations for the future, with parent reported future expectations as the dependent variable. The Levene statistic indicated the clusters had unequal variances. Thus, Welch's F and Games-Howell post hoc statistics were reported. Results suggested group status significantly predicted parental expectations for the future Welch's $F(31.69) = 15.31, p<.01$. Post-hoc analyses indicated significantly less ambitious expectations for the future for participants in the "non-completers" group, compared to those in the "average to low average cognitive, impaired motor" group ($p<.05$), and the "average to low average cognitive" group ($p<.05$; see Table 1).

Four cluster solution predicting quality of life. Two MANOVAs were conducted to examine the relationship between cluster membership and quality of life. Parent and child report were analyzed separately. The dependent variables included the 4
subscales for each measure of QOL. Using the Wilk's statistic, results indicated cluster membership did not have a significant effect on parent reported quality of life, \( \lambda = 0.84, F(12,251.64) = 1.48, p = .13 \). However, cluster membership significantly predicted child reported quality of life, \( \lambda = .74, F(12,243.7) = 2.50, p < .01 \). Follow-up univariate ANOVAs revealed cluster membership significantly predicted the physical scale \( F(3, 96) = 4.54, p < .01 \) and the social scale \( F(3, 96) = 2.79, p < .05 \). Post-hoc analyses indicated no significant differences between "non-completers" and any other group for physical and social quality of life.

The exploratory hypotheses were mostly supported, as the 4 cluster solution significantly predicted all outcomes, except for QOL. Additionally, all significant effects were in the direction hypothesized (cluster 4 having the lowest outcome score, see Table 11). Interestingly, the "non-completers" group and the "extremely low to borderline" group had similar outcomes except for independence with time.

**Shunt status predicting 4 cluster solution.** Number of shunt surgeries was expected to predict cluster membership, but this hypothesis was not supported for the 3 cluster solution. It is possible that a significant effect was not found because of insufficient power. In fact, the variable “number of shunt surgeries” was skewed, which may have reduced the power to detect a significant effect. Thus, it was decided to use a similar measure, shunt status, as a predictor. Number of shunt surgeries was originally chosen as a predictor because it is continuous and provides more information than shunt status. However, shunt status as a dichotomous variable (does the child have a shunt: yes or no) was found to be less skewed than number of shunt surgeries. Therefore, it was
decided to conduct an exploratory analysis to determine whether shunt status would predict cluster membership.

Shunt status was obtained from each participant’s medical chart. A logistic regression was conducted with shunt status as the independent variable and cluster membership (4 cluster solution) as the dependent. The results showed shunt status had a significant main effect on cluster membership $\chi^2(3) = 15.24, p<.01$. More specifically, shunt status significantly predicted whether a participant was placed in either the "average to low average cognitive, impaired motor" group or the "average to low average cognitive" group, $b = -1.52$, Wald $\chi^2(1) = 5.56, p<.05$; the "average to low average cognitive" or the "extremely low to borderline" group, $b = -2.53$, Wald $\chi^2(1) = 5.43$, $p<.05$. The "non-completers" group only included participants with shunts, thus post-hoc analyses with this 4th group indicated extremely high and unreliable Wald $\chi^2$ statistics. The models suggested that those with shunts were more likely to be placed in the "extremely low to borderline" group (cluster 3) and the "average to low average cognitive, impaired motor" group (cluster 1) rather than the "average to low average cognitive" group (cluster 2). Cluster 4 ("non-completers"), was composed solely of participants with a shunt. In short, participants were more likely to be placed in the "average to low average cognitive" group (cluster 2) if they did not have a shunt (see Table 12). Thus, the hypothesis was supported.
CHAPTER FIVE

DISCUSSION

The purpose of this study was to examine neuropsychological performance among children with spina bifida to determine if there are distinct groups or “profiles” of cognitive functioning. It was predicted that four cluster groups would emerge from the analysis: variability and higher functioning (cluster 1), variability and lower functioning (cluster 2), consistency and higher functioning (cluster 3), and consistency and lower functioning (cluster 4; see Figure 2 and 3). Biological, sociodemographic, and environmental variables were examined as possible predictors of cluster membership. Additionally, measures of independence, academic success, expectations for the future, and quality of life were examined as possible outcomes of cluster membership.

The results of this study suggested a 3 cluster solution best fit the data. Participants in the "average to low average cognitive, impaired motor" cluster performed in the average range on measures of intelligence, academic achievement, and social-emotional processing; the low average to average range on measures of executive functioning and social-contextual language; and in the extremely low range on measures of fine motor ability (cluster 1). Participants in the "average to low average cognitive" cluster performed in the average range on measures of intelligence, academic achievement, social-emotional processing, social-contextual language, and executive functioning; low average to average on measures of attention; and borderline on measures
of fine motor skills (cluster 2). Finally, participants in the "extremely low to borderline" cluster (cluster 3) performed in the extremely low to borderline range on all measures except for WRAT spelling (low average) and TEACH number of identified targets (average). Because the proposed 4 cluster solution was not supported, the predictor and outcome hypotheses were adjusted to reflect the 3 cluster solution. It was hypothesized that younger individuals, Hispanic individuals, low SES individuals, individuals with less emphasis on personal growth in the family environment, and individuals with more family stress would more likely be placed in cluster 2; whereas, individuals with a greater number of shunt surgeries, and positive seizure history would more likely be placed in cluster 3. Additionally, cluster membership was predicted to be associated with outcome variables in the following order from the highest level of outcome to the lowest: cluster 2, cluster 1, and cluster 3. Exploratory analyses were also included to examine predictors and outcomes of a 4th group of participants who were not able to complete the neuropsychological battery due to low comprehension. As well, shunt status was included in exploratory analyses, to examine whether it predicted cluster membership.

The study built upon the current literature by examining individual differences within children with spina bifida, rather than comparing children with spina bifida to norms or a typically developing group. Additionally, instead of examining one cognitive construct (e.g., attention), the current study assessed many constructs (intelligence, attention, comprehension of complex language, affect recognition, executive functioning, and manual dexterity) to generate sub-group specific, multidimensional profiles of strengths and weaknesses. These subgroup profiles have the potential to be more
informative than general statements about the neuropsychological functioning of children with spina bifida. Moreover, few previous studies have included participants with low IQ scores (<70). Hispanic populations have also been under-represented in previous research. Because participants in the current study were more intellectually and ethnically diverse, findings may be more representative of children with spina bifida. The following section includes a review of the hypotheses, a description of the findings, and a discussion of possible explanations for the findings. Finally, suggestions for future directions and clinical applications based upon the study are discussed.

Hypothesis I

It was hypothesized that varied performance across measures of associative and assembled processing would result in four distinct subgroups: variability and higher functioning, variability and lower functioning, consistency and higher functioning, and consistency and lower functioning. Scores from 22 neuropsychological measures were used to create the clusters. Contrary to the hypothesis, results indicated that a 3 cluster solution best fit the data: average to low average cognitive, impaired motor (cluster 1); average to low average cognitive (cluster 2); and extremely low to borderline (cluster 3). In examining the hypothesized areas of strength and weakness (see Table 4), all clusters indicated hypothesized patterns in the general areas of academic functioning, visual executive functioning, and fine motor skills. Cluster 3, "extreme low to borderline," also performed as hypothesized in general areas of intelligence, attention, and social/emotional processing. Cluster 1 "average to low average cognitive, impaired motor" indicated a pattern that was consistent with hypothesized strengths and weakness
in the area of intelligence; and cluster 2 "average to low average cognitive" was consistent in the area of attention. None of the clusters demonstrated hypothesized strengths and weaknesses in the area of verbal executive functioning. Of note, the subgroup profiles indicated different patterns of strengths and weaknesses, rather than merely different levels of the same profile (see Figure 4). While the slopes of the profile for clusters 1 and 3 were similar, cluster 2 had different slopes, indicating a distinctly different neuropsychological profile. These distinct profiles suggest one cognitive phenotype for all children with spina bifida may be too limiting and may not adequately represent the population.

Snow and colleagues' (1994) also found a 3 cluster solution when they examined neuropsychological functioning in adolescents and young adults with spina bifida. Snow and colleagues (1994) used different measures, the Halstead-Reitan Neuropsychological Test Battery and Wechsler Intelligence Scale. Thus, comparisons to their findings were difficult. Still, the clusters identified by Snow and colleagues (1994) were distinguished by the following: mostly borderline functioning in IQ, visual scanning, and abstraction abilities (cluster 1); average IQ and low average visual scanning and abstraction abilities (cluster 2); and mostly extremely low functioning in IQ, visual scanning, and abstraction abilities. Overall, Snow's participants seemed to be lower functioning than the participants in the current study. Although, their sample was different from the current study in that it was smaller (n=37), the participants were older (M=17.65 years), and fewer participants had shunts (70% had shunts; Snow et al., 1994).
There may be several reasons that the cluster solution was not accurately predicted. First, it is possible that cognitive differences are not easily predicted, due to a multitude of factors that affect cognitive functioning in youth with spina bifida. Fletcher and Dennis (2009) proposed the cognitive profile of individuals with spina bifida would vary in a "principled manner as a function of" certain factors (e.g., hydrocephalus and poverty). However, they list several possible factors without mentioning which factors may be more influential or may interact with other factors to determine cognitive functioning. Thus, while Fletcher and Dennis (2009) provide evidence for each factor individually, there is little discussion of how interactions or additive effects might affect cognitive outcomes. Second, the sample in the current study was small (n=96), thus it is possible more clusters exist, but were not identified due to a limited sample size. Third, the neuropsychological battery given in the current study was not appropriate for individuals with intellectual disabilities. Several of the tasks were complicated and required adequate understanding of directions to complete the task. Thus, a number of participants were not able to complete the battery due to limited comprehension. The current study aimed to include cognitively diverse participants, and thus it was decided to include those who could not complete the battery as a fourth subgroup.

**Hypothesis II**

The second hypothesis proposed that biological factors (lesion level, number of shunt surgeries, and seizures), socio-demographic factors (age and ethnicity), and environmental factors (socioeconomic status, family environment, and family stress) would predict group membership. Predictors were examined to identify possible causes
of differences in cognitive functioning and to validate the cluster solution. Analyses were run with the original 3 cluster solution as well as the 4 cluster solution (with "non-completers subgroup).

Regarding the 3 cluster solution, it was hypothesized that younger individuals, Hispanic individuals, low SES individuals, individuals with less emphasis on personal growth in the family environment, and individuals with more family stress would more likely be placed in cluster 2; whereas, individuals with a greater number of shunt surgeries, and positive seizure history would more likely be placed in cluster 3. Due to an error, two analyses were run, one with and one without seizure status. Both models significantly predicted cluster membership. However, only SES and seizure status were found to have a significant main effect on cluster membership. Participants were more likely to be placed in the "average to low average cognitive, impaired motor" group (cluster 1) if their family's SES was higher. Those with a history of seizures were more likely to be placed in the "extremely low to borderline" group (cluster 3).

For the 4 cluster solution, it was predicted cluster 4 ("non-completers") would be composed of participants with higher lesion levels, positive seizure history, younger age, Hispanic ethnicity, lower SES, less personal growth in the family environment, and more family stress. Again, both models (with and without seizure status) were found to significantly predict cluster membership. Contrary to the first analysis, only seizure history had a main effect of cluster membership, such that participants with a history of seizures were more likely to be placed in cluster 4 (non-completers) than cluster 1 (average to low average cognitive, impaired motor) or cluster 2 (average to low average
cognitive). Additionally, because the variable "number of shunt surgeries" was skewed, exploratory analyses were also conducted with shunt status. It was determined that shunt status significantly predicted cluster membership. Specifically, those without a shunt were more likely to be placed in cluster 2 (average to low average cognitive). Groups 1 (average to low average cognitive, impaired motor), 3 (extremely low to borderline), and 4 (non-completers) were more likely to include participants with a shunt.

Because only 3 of the 9 predictors had a significant effect on the cluster solution, the external validity of the cluster solution was not well supported by the hypothesized predictors. There are several possible reasons why cluster membership was not predicted by most of these variables. First, issues with the data may have reduced the power to detect significant effects. For example, the variable "number of shunt surgeries" was skewed, which may have reduced the power to detect a significant effect. Second, it is possible the current study examined less salient predictors, and that other biological or environmental predictors (e.g., brain malformations or education) may have a greater effect on cluster membership. For example, Hampton and colleagues (2011) identified differences in cognitive profiles due to differences in hydrocephalus status, which was not measured in the current study. In fact, Fletcher and Dennis (2009) list several other biological predictors of cognitive functioning that were not included in this study (e.g., structural issues with cerebellum and brainstem, callosal dysgenesis, white matter loss, etc.).

Alternatively, the current study did identify SES, seizure status, and shunt status as significant predictors of cluster membership. These findings are congruent with past
research. In typically developing children, it is well established that low SES is a risk factor for poorer cognitive, academic, and socio-emotional outcomes (McLoyd, 1998). Still, it is also known that lower SES is associated with poorer school conditions (Aikens & Barbarin, 2008). Thus, it is possible the educational environment also has an effect on cognitive outcomes; however, educational environment was not examined as a part of this study. Positive seizure history and positive shunt status have also been associated with poorer cognitive outcomes (Brown et al., 2008; Hampton et al., 2011). In fact, seizures in adults with spina bifida have been associated with mental retardation (Barf et al., 2003). While seizure and shunt status are both risk factors on their own, they have also been associated with hydrocephalus (Erikson et al., 2002). Unfortunately, hydrocephalus status was not examined in this study. It is possible hydrocephalus status would also predict cluster membership. Indeed, Hampton and colleagues (2001) found different cognitive profiles for youth with spina bifida depending on whether they had no hydrocephalus, shunted hydrocephalus, or arrested hydrocephalus (hydrocephalus without a shunt). Overall, only a few predictor variables were found to have significant associations with cluster membership, which may indicate a need to further examine other possible predictors of cognitive differences.

**Hypothesis III**

The third hypothesis of this study explored outcomes of cluster membership. It was hypothesized that group membership would predict levels of independence, academic success, expectations for the future, and quality of life. Outcomes were examined to determine the utility of cluster profiles in predicting real-world functioning
and to validate the cluster solution. Analyses were run with the original 3 cluster solution as well as the 4 cluster solution (with "non-completers" cluster).

For the 3 cluster solution it was hypothesized that clusters would be associated with outcome variables in the following order from the highest level of outcome to the lowest: "average to low average cognitive" (cluster 2), "average to low average cognitive, impaired motor" (cluster 1), and "extremely low to borderline" (cluster 3; see Figure 4). Cluster membership was found to significantly predict independence (fine motor, money, language, and time), academic success, parental expectations for the future, and child-reported quality of life (physical). Cluster membership was not a significant predictor of parent-reported quality of life or child-reported emotional, social, or school related quality of life. Besides academic success, outcome means for each scale were in the hypothesized direction (in order of highest to lowest: cluster 2, cluster 1, and cluster 3; see Table 10). Cluster 1 ("average to low average cognitive, impaired motor") showed greater academic success than cluster 2 ("average to low average cognitive"), still clusters 1 and 2 had greater academic success than cluster 3 ("extremely low to borderline").

While the difference in academic success between clusters 1 and 2 was not statistically significant, it is still noteworthy. Indeed, participants in cluster 1 achieved higher scores on measures of academic achievement from the neuropsychological battery (see Figure 4). Thus, it is not surprising that they would also score higher on teacher reported academic success. Additionally, while clusters 1 and 2 achieved similar IQ scores, participants in cluster 1 achieved higher scores than participants in cluster 2 on measures of auditory attention (see Figure 4 and Table 6). Previous researchers have
found an association between measures of attention and academic achievement for children with spina bifida (Loss, Yeates, & Enrile, 1998). Auditory attention may be most important for learning in lecture-based academic settings. Thus, it is possible cluster 2 ("average to low average cognitive") represents participants who have difficulties with auditory attention and academic performance, compared to participants in cluster 1 ("average to low average cognitive, impaired motor").

For the 4 cluster solution, it was predicted cluster 4 ("non-completers") would be associated with the lowest level of each outcome, compared to the other three clusters. Using the 4 cluster solution, cluster membership was found to be associated with independence (fine motor, money, language, and time), academic success, parental expectations for the future, and child reported quality of life (physical and social). There were no significant associations between cluster membership and parent reported quality of life or child reported emotional or school related quality of life. For most outcome scales, means were in the hypothesized direction, such that participants in cluster 4 had the lowest scores (see Table 11). The only exception was social quality of life, for which participants in cluster 4 reported greater social related quality of life than participants in clusters 1, 2, or 3 (see Table 11). Because participants in cluster 4 most likely were the lowest functioning of the 4 groups, as they were the participants who were not able to complete the battery due to low comprehension, it is possible they did not fully understand the questionnaire, or were not aware of their social difficulties. These scenarios are more likely than this group truly experiencing a better social quality of life,
considering parents of participants in cluster 4 had the lowest average rating for their child's social quality of life (see Table 11).

Cluster membership was expected to be associated with parent and child reported quality of life, but in fact was only associated with child reported QOL. It is fairly common to obtain different results for child versus parent report of quality of life (Eiser & Morse, 2001). Still, both reports are considered valid (Theunissen, Vogels, Koopman, Verrips, Zwinderman, Verloove-Vanhorick, & Wit, 1998). Only the physical and social subscales of self-reported quality of life were predicted by cluster membership. It is possible that the physical subscale was predicted due to large differences in fine motor functioning, that correspond to differences in physical quality of life (see grooved pegboard scores, Figure 4 and Table 6). Participants in cluster 3 performed most poorly on measures of fine motor ability and also reported the lowest physical quality of life. Overall, the results indicated cluster membership was not a good predictor of parent or child reported quality of life, but was a good predictor of many other functional outcomes.

In sum, this study provided support for the existence of subgroups of children with spina bifida with similar neuropsychological profiles. More specifically, three subgroups were identified. The cluster solution was replicated using hierarchical and non-hierarchical clustering methods, and the validity of the cluster solution was supported by significant associations with several predictor variables (SES, seizure history, and shunt status) and outcome variables (independence, academic success, and expectations for the future). Several factors were identified that suggest the cognitive profile differs between
subgroups of youth with spina bifida and has significant implications for future functioning. The findings of this study were particularly noteworthy, because the significant influences of the cognitive profiles on functional outcomes were found across methods and reporters.

**Study Limitations and Future Directions**

There are several limitations in the current study that could be improved upon in future research identifying and predicting cognitive profiles of children with spina bifida. First, the measures in the neuropsychological battery were not specifically chosen for this study, as this study used archival data from a larger, longitudinal research program. Thus, while this study included a wide range of neuropsychological functioning, it did not include all areas of neuropsychological functioning that have been determined to be pertinent for children with spina bifida (see Table 4). To provide a more complete profile, future researchers would benefit from using a neuropsychological battery that includes measures for all areas of identified strengths and weaknesses for children with spina bifida.

In addition, while several precautions were taken to increase the validity of these findings, the cluster analysis method is inherently exploratory and thus this cluster solution needs to be replicated. Unfortunately, due to small sample size, it was not possible to attempt replication by splitting the sample in the current study. Additionally, several factors in the current study limited the statistical power to detect predictors and outcomes of cluster membership. Due to the study's small sample size, the analyses could detect only medium to large effects. Future researchers could benefit from a larger
sample size that would allow them to test for replication and detect smaller effects. While it is difficult to obtain a large sample size in a pediatric population, this could be achieved by collaborating across multiple sites.

As well, the current study used only cross-sectional data. Thus, it is unclear whether the cluster solution identified in the current study would hold up across time. It is possible that a participant may move from one cluster to another. For example, if a child's shunt becomes infected and requires replacement, or if a child continues to have seizures, he/she may have a reduction in cognitive functioning and thus may move to a lower functioning cluster. Therefore, much could be learned from documenting a child's neuropsychological functioning over time, as well as factors that may contribute to changes in cognitive functioning. Additionally, because the study was cross-sectional, it is uncertain whether the child's cognitive profile would predict future functioning. In the current study, the child's cognitive profile was assessed at the same time as his/her outcome variables. Thus, the profile is simply indicative of current functioning, not future functioning. Longitudinal research that examines future functioning could determine which groups of children with spina bifida are at risk for long term concerns.

Another limitation of the current study was the fact that neuro-imaging and neurobiological information were not included as predictors of neuropsychological profiles. Unfortunately, information about hydrocephalus and structural abnormalities were not available for this study. However, Fletcher and Dennis (2009) have suggested the neural phenotype and secondary insults to the brain are likely causes of differences in the cognitive profile. Thus, it is possible that other predictors, such as hydrocephalus
status or structural abnormalities, are stronger predictors of one's neuropsychological profile than the predictors examined in this study.

While the current study examined several predictors of cluster membership, it did not test interactions. The current study failed to find main effects for several predictor variables, such as number of shunt surgeries, age, ethnicity, family stress, and family environment. It is possible that 2 or more of these variables may interact, rather than have a main effect on one's neuropsychological profile. Interactions were not included in the current study because predictor variables were primarily used to validate the cluster solution, and because of our small sample size and lack of power to reliably test interactions. However, interactions may be important to examine as researchers move forward in determining potential causes of cognitive differences.

Finally, several precautions were taken to prepare the data for cluster analysis, but there were still outliers for the grooved pegboard subtests. Because cluster analysis is sensitive to outliers, it is possible that a greater emphasis was placed on fine motor ability when the clusters were determined. Thus, it is recommended that future researchers either choose neuropsychological measures that rarely produce outliers, or adjust all outliers before conducting cluster analyses.

**Clinical Implications**

There are several suggestions for working with this population that can be made based on the current findings. Results suggest there is no "one" neuropsychological profile for children with spina bifida. Rather, individuals in this population present with a wide range of functioning and may be better categorized by several "subgroup" profiles.
Because profiles vary, it is important that professionals (e.g., teachers, doctors, nurses, etc) do not use the established "phenotype" as a sole basis for their interactions with an individual with spina bifida. For example, it may be necessary for a doctor or teacher to obtain specific information about the individual's cognitive functioning before developing an appropriate lesson plan or discussing medical decisions. Additionally, psychologists developing group interventions for children with spina bifida may want to develop more specific interventions for subgroups of children with spina bifida. For example, an interventionist may want to create separate interventions for those of higher and lower cognitive ability. Additionally, among the higher cognitive group, the interventionist may want to create an intervention with smaller groups for those with attention difficulties.

Several suggestions can also be made for neuropsychologists assessing a child with spina bifida. First, findings suggest that several children with spina bifida could be classified as having mild mental retardation. Several participants in the current study were so cognitively impaired that they could not complete the neuropsychological battery, due to low comprehension. Thus, it may be important for neuropsychologists to use measures that have a lower floor, are sensitive to lower levels of functioning, and are easier to complete. Because these children may not be accurate reporters, it also may be necessary to obtain information from adult care-givers and teachers.

Additionally, participants in the current study showed deficits in fine motor functioning that seemed to be independent of cognitive functioning. Thus, it is crucial that neuropsychologists include measures of fine motor functioning in their assessments of children with spina bifida. Several practical recommendations can be made to address
fine motor issues. For example, it could be recommended that a child receive an extra copy of class notes, be provided a computer and typing lessons, have an assistant or classmate take notes for him/her, etc. In addition, it may be important to take fine motor deficits into consideration when interpreting scores on other measures. For example, several IQ and academic measures require a participant to draw or write within a certain amount of time. A participant with fine motor deficits may perform more poorly than he/she should on such tasks.

Finally, this study suggested the neuropsychological profile was a good indicator of functional outcomes. Thus, this study provided support for the utility of neuropsychological assessments in determining how a child with spina bifida should be performing in school and functioning independently. Thus, whenever there are concerns about academic performance or independence at home, a neuropsychological evaluation may be helpful in determining what can/should be expected of the individual.
APPENDIX A

TABLES AND FIGURES
Table 1: Hypothesized predictors and outcomes of neuropsychological profiles

<table>
<thead>
<tr>
<th>Predictors</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biological</td>
<td>Independence</td>
</tr>
<tr>
<td>Lesion level</td>
<td>Academic success</td>
</tr>
<tr>
<td>Number of shunt surgeries</td>
<td>Expectations for the future</td>
</tr>
<tr>
<td>History of seizures</td>
<td>Quality of life</td>
</tr>
<tr>
<td>Sociodemographic</td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td></td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
</tr>
<tr>
<td>Environmental</td>
<td></td>
</tr>
<tr>
<td>Socio-economic status</td>
<td></td>
</tr>
<tr>
<td>Family stress</td>
<td></td>
</tr>
<tr>
<td>Family environment</td>
<td></td>
</tr>
</tbody>
</table>
Table 2: Demographic variables for included vs. excluded participants

<table>
<thead>
<tr>
<th>Demographic characteristics</th>
<th>Included</th>
<th>Excluded</th>
<th>Statistical test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child age in years (n=138), M (SD)</td>
<td>11.13 (2.41)</td>
<td>11.76 (2.57)</td>
<td>t(136) = -1.40</td>
</tr>
<tr>
<td>Child gender (n=134)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male, % (n)</td>
<td>51% (49)</td>
<td>29% (11)</td>
<td>χ²(1) = 5.37*</td>
</tr>
<tr>
<td>Female, % (n)</td>
<td>49% (47)</td>
<td>71% (27)</td>
<td></td>
</tr>
<tr>
<td>Child ethnicity (n=133)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White, % (n)</td>
<td>55% (53)</td>
<td>57% (21)</td>
<td>χ²(1) = 0.03</td>
</tr>
<tr>
<td>Other, % (n)</td>
<td>45% (43)</td>
<td>43% (16)</td>
<td></td>
</tr>
<tr>
<td>Shunt status (n=131)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>With shunt, % (n)</td>
<td>81% (76)</td>
<td>62% (23)</td>
<td>χ²(1) = 5.02*</td>
</tr>
<tr>
<td>Without shunt, % (n)</td>
<td>19% (18)</td>
<td>38% (14)</td>
<td></td>
</tr>
<tr>
<td>Hollingshead SES (n=132), M (SD)</td>
<td>41.06 (16.15)</td>
<td>36.72 (15.47)</td>
<td>t(130) = 1.43</td>
</tr>
<tr>
<td>FSIQ (n=134), M (SD)</td>
<td>84.50 (18.67)</td>
<td>81.68 (21.79)</td>
<td>t(132) = 1.55</td>
</tr>
</tbody>
</table>

Note. * = p<.05. The Hollingshead (1975) Four Factor Index of socioeconomic status (SES) is based on a composite of maternal education, paternal education, maternal occupational status, and paternal occupational status.
Table 3: Demographic variables for participants who completed vs. did not complete the neuropsychology profile due to low comprehension and/or intellectual disability

<table>
<thead>
<tr>
<th>Demographic characteristics</th>
<th>Completed</th>
<th>Did not complete</th>
<th>Statistical test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child age in years (n=108), $M (SD)$</td>
<td>11.13 (2.40)</td>
<td>11.91 (2.39)</td>
<td>$t(105) = -1.02$</td>
</tr>
<tr>
<td>Child gender (n=108)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male, % (n)</td>
<td>51% (49)</td>
<td>42% (5)</td>
<td>$\chi^2(1) = 0.38$</td>
</tr>
<tr>
<td>Female, % (n)</td>
<td>49% (47)</td>
<td>58% (7)</td>
<td></td>
</tr>
<tr>
<td>Child ethnicity (n=107)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White, % (n)</td>
<td>55% (53)</td>
<td>55% (6)</td>
<td>$\chi^2(1) = 0.00$</td>
</tr>
<tr>
<td>Other, % (n)</td>
<td>45% (43)</td>
<td>45% (5)</td>
<td></td>
</tr>
<tr>
<td>Shunt status (n=104)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>With shunt, % (n)</td>
<td>81% (76)</td>
<td>100% (10)</td>
<td>$\chi^2(1) = 2.32$</td>
</tr>
<tr>
<td>Without shunt, % (n)</td>
<td>19% (18)</td>
<td>0% (0)</td>
<td></td>
</tr>
<tr>
<td>Hollingshead SES (n=103), $M (SD)$</td>
<td>41.06 (16.15)</td>
<td>34.00 (12.30)</td>
<td>$t(101) = 1.34$</td>
</tr>
</tbody>
</table>

Note. *= p<.05. The Hollingshead (1975) Four Factor Index of socioeconomic status (SES) is based on a composite of maternal education, paternal education, maternal occupational status, and paternal occupational status.
<table>
<thead>
<tr>
<th>Domain</th>
<th>Assets Associative Processing</th>
<th>Deficits Assembled Processing</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>From Fletcher &amp; Dennis, 2009: Measure</td>
<td>From Fletcher &amp; Dennis, 2009: Measure</td>
</tr>
<tr>
<td>MOTOR</td>
<td>Adaptation (mirror drawing)</td>
<td>Online control (tracking)</td>
</tr>
<tr>
<td></td>
<td>CAS planned connections; Grooved pegboard</td>
<td></td>
</tr>
<tr>
<td>PERCEPTION</td>
<td>Categories (face recognition)</td>
<td>Relations (mental rotations)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>WASI Matrix Reasoning</td>
</tr>
<tr>
<td>MEMORY</td>
<td>Implicit (priming)</td>
<td>Explicit (episodic recall)</td>
</tr>
<tr>
<td></td>
<td>Stipulation (word definitions)</td>
<td></td>
</tr>
<tr>
<td>LANGUAGE</td>
<td>WASI Vocab; DKEFS letter and category</td>
<td>DKEFS switching; CASL Inferences and Pragmatic Judgment</td>
</tr>
<tr>
<td>READING</td>
<td>Decoding (word recognition)</td>
<td>Comprehension (text meaning)</td>
</tr>
<tr>
<td>MATH</td>
<td>Numbers (exact calculation)</td>
<td>Algorithms (estimations)</td>
</tr>
<tr>
<td>ATTENTION</td>
<td>TEACH sky search</td>
<td>TEACH score; sky search DT; score DT</td>
</tr>
<tr>
<td>SOCIAL/EMOTIONAL</td>
<td>DANELVA Faces and Voices</td>
<td></td>
</tr>
</tbody>
</table>
Table 5: Agglomeration coefficients and change across steps in Ward's cluster analysis

<table>
<thead>
<tr>
<th>Number of clusters</th>
<th>Agglomeration coefficient</th>
<th>Change in coefficient</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>338,736</td>
<td>13,634</td>
</tr>
<tr>
<td>9</td>
<td>352,370</td>
<td>15,937</td>
</tr>
<tr>
<td>8</td>
<td>368,307</td>
<td>19,765</td>
</tr>
<tr>
<td>7</td>
<td>388,072</td>
<td>22,123</td>
</tr>
<tr>
<td>6</td>
<td>410,195</td>
<td>26,622</td>
</tr>
<tr>
<td>5</td>
<td>436,817</td>
<td>42,135</td>
</tr>
<tr>
<td>4</td>
<td>478,952</td>
<td>47,439</td>
</tr>
<tr>
<td>3</td>
<td>526,391</td>
<td>85,300</td>
</tr>
<tr>
<td>2</td>
<td>611,691</td>
<td>200,620</td>
</tr>
<tr>
<td>1</td>
<td>812,311</td>
<td>---</td>
</tr>
</tbody>
</table>

*Note:* A large increase in the agglomeration coefficient suggests that two very distinct clusters have been combined. When 3 clusters were reduced to 2 clusters the agglomeration coefficient increased by 85,300, which is compared to relatively trivial increases (i.e., 47,439; 42,135; etc).
Table 6: Mean standard score (and standard deviation) for each Ward’s cluster

<table>
<thead>
<tr>
<th>Measure</th>
<th>Cluster 1 (Average to Low Average Cognitive, Impaired Motor)</th>
<th>Cluster 2 (Average to Low Average Cognitive)</th>
<th>Cluster 3 (Extremely Low to Borderline)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WASI Vocabulary</td>
<td>96.96 (16.87)²</td>
<td>Avg</td>
<td>67.96 (11.22)³ Extremely Low</td>
</tr>
<tr>
<td>WASI Matrix Reasoning</td>
<td>90.27 (15.20)²</td>
<td>Avg</td>
<td>64.42 (11.22)³ Extremely Low</td>
</tr>
<tr>
<td>WRAT Reading</td>
<td>107.46 (13.62)²</td>
<td>Avg</td>
<td>78.16 (17.90)² Borderline</td>
</tr>
<tr>
<td>WRAT Spelling</td>
<td>107.56 (13.98)²</td>
<td>Avg</td>
<td>83.04 (15.40)² Low Avg</td>
</tr>
<tr>
<td>WRAT Math</td>
<td>89.79 (15.35)²</td>
<td>Avg</td>
<td>68.32 (16.74)² Extremely Low</td>
</tr>
<tr>
<td>CAS Planned Connections</td>
<td>80.64 (12.63)²</td>
<td>Low Avg</td>
<td>64.60 (8.03)² Extremely Low</td>
</tr>
<tr>
<td>DKEFS Letter Fluency</td>
<td>89.48 (18.17)²</td>
<td>Low Avg</td>
<td>71.20 (12.61)² Borderline</td>
</tr>
<tr>
<td>DKEFS Category Fluency</td>
<td>87.82 (16.61)²</td>
<td>Low Avg</td>
<td>68.60 (11.32)² Extremely Low</td>
</tr>
<tr>
<td>DKEFS Switching Fluency</td>
<td>91.28 (17.72)²</td>
<td>Avg</td>
<td>72.80 (14.15)² Borderline</td>
</tr>
<tr>
<td>DKEFS Switching Accuracy</td>
<td>91.15 (17.15)²</td>
<td>Avg</td>
<td>76.00 (13.84)² Borderline</td>
</tr>
<tr>
<td>TEACH Sky Search # of Targets</td>
<td>100.26 (19.60)²</td>
<td>Avg</td>
<td>92.80 (17.08)² Avg</td>
</tr>
<tr>
<td>TEACH Sky Search Efficiency</td>
<td>75.00 (14.51)²</td>
<td>Low Avg</td>
<td>66.80 (10.69)² Extremely Low</td>
</tr>
<tr>
<td>TEACH Sky Search Attention</td>
<td>78.21 (17.49)²</td>
<td>Low Avg</td>
<td>73.20 (15.60)² Borderline</td>
</tr>
<tr>
<td>TEACH Score</td>
<td>97.05 (15.08)²</td>
<td>Avg</td>
<td>73.80 (11.66)² Borderline</td>
</tr>
<tr>
<td>TEACH Sky Search Dual Task</td>
<td>91.54 (27.65)²</td>
<td>Low Avg</td>
<td>63.40 (14.77)² Extremely Low</td>
</tr>
<tr>
<td>TEACH Score Dual Task</td>
<td>93.88 (13.88)²</td>
<td>Low Avg</td>
<td>68.00 (10.51)² Extremely Low</td>
</tr>
<tr>
<td>Grooved Pegboard Dom</td>
<td>35.05 (21.76)²</td>
<td>Extremely Low</td>
<td>25.68 (12.94)² Extremely Low</td>
</tr>
<tr>
<td>Grooved Pegboard Non-Dom</td>
<td>34.00 (18.57)²</td>
<td>Extremely Low</td>
<td>25.08 (14.92)² Extremely Low</td>
</tr>
<tr>
<td>DANVA Faces</td>
<td>96.90 (16.02)²</td>
<td>Avg</td>
<td>73.69 (19.71)² Borderline</td>
</tr>
<tr>
<td>DANVA Voices</td>
<td>93.27 (9.53)²</td>
<td>Avg</td>
<td>77.58 (13.64)² Borderline</td>
</tr>
<tr>
<td>CASL Inferences</td>
<td>89.33 (12.79)²</td>
<td>Low Avg</td>
<td>66.76 (17.72)² Extremely Low</td>
</tr>
<tr>
<td>CASL Pragmatic Judgment</td>
<td>91.64 (12.64)²</td>
<td>Avg</td>
<td>69.84 (14.93)² Extremely Low</td>
</tr>
</tbody>
</table>

Note: Superscripts with same letters are not significantly different from each other. Those with different letters are significantly different.
Table 7: Agglomeration coefficients and change across steps in average link cluster analysis

<table>
<thead>
<tr>
<th>Number of clusters</th>
<th>Agglomeration coefficient</th>
<th>Change in coefficient</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>8,879</td>
<td>203</td>
</tr>
<tr>
<td>9</td>
<td>9,082</td>
<td>185</td>
</tr>
<tr>
<td>8</td>
<td>9,267</td>
<td>681</td>
</tr>
<tr>
<td>7</td>
<td>9,948</td>
<td>179</td>
</tr>
<tr>
<td>6</td>
<td>10,127</td>
<td>606</td>
</tr>
<tr>
<td>5</td>
<td>10,733</td>
<td>438</td>
</tr>
<tr>
<td>4</td>
<td>11,171</td>
<td>323</td>
</tr>
<tr>
<td>3</td>
<td>11,494</td>
<td>2068</td>
</tr>
<tr>
<td>2</td>
<td>13,562</td>
<td>3539</td>
</tr>
<tr>
<td>1</td>
<td>17,101</td>
<td>---</td>
</tr>
</tbody>
</table>

*Note:* A large increase in the agglomeration coefficient suggests that two very distinct clusters have been combined. When 3 clusters were reduced to 2 clusters the agglomeration coefficient increased by 2068, which is compared to relatively trivial increases (i.e., 323, 438, etc).
Table 8: Overlap in cluster membership between Ward's cluster solution and average linkage within groups

<table>
<thead>
<tr>
<th>Ward's method</th>
<th>Average linkage (within groups)</th>
<th>Agreement</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>33 5 1</td>
<td>85%</td>
</tr>
<tr>
<td>2</td>
<td>7 4 21</td>
<td>65%</td>
</tr>
<tr>
<td>3</td>
<td>0 25 0</td>
<td>100%</td>
</tr>
</tbody>
</table>

Overall agreement (79 out of 96 cases): 82%

Note: Highlighted numbers indicate cases that overlapped between cluster methods.
Table 9: Overlap in cluster membership between Ward's cluster solution and k-means

<table>
<thead>
<tr>
<th>Wards Method</th>
<th>K-means method</th>
<th>Agreement</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28 6 5</td>
<td>72%</td>
</tr>
<tr>
<td>2</td>
<td>3 3 26</td>
<td>81%</td>
</tr>
<tr>
<td>3</td>
<td>1 24 0</td>
<td>96%</td>
</tr>
</tbody>
</table>

Overall agreement (78 out of 96 cases): 81%

*Note:* Highlighted numbers indicate cases that overlapped between cluster methods.
Table 10: Means for each outcome variable by cluster (Ward's method)

<table>
<thead>
<tr>
<th>Scale</th>
<th>Cluster 1 (n=39): Average cognitive, impaired motor</th>
<th>Cluster 2 (n=32): Average cognitive, low avg attention</th>
<th>Cluster 3 (n=25): Extremely low to borderline</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Average</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Independence</td>
<td>Fine motor</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Cluster 1</td>
<td>Cluster 2</td>
<td>Cluster 3</td>
</tr>
<tr>
<td></td>
<td>43.09(^a)</td>
<td>48.36(^{ab})</td>
<td>41.56(^b)</td>
</tr>
<tr>
<td></td>
<td>Money</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>23.42(^c)</td>
<td>32.10(^{cd})</td>
<td>19.44(^d)</td>
</tr>
<tr>
<td></td>
<td>Language</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>39.58</td>
<td>44.40(^e)</td>
<td>35.29(^e)</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>42.55</td>
<td>47.33(^f)</td>
<td>39.46(^f)</td>
</tr>
<tr>
<td>Academic success</td>
<td>46.38(^g)</td>
<td>45.00(^h)</td>
<td>36.90(^{eh})</td>
</tr>
<tr>
<td>Future expectations</td>
<td>3.35(^{ij})</td>
<td>3.68(^{ik})</td>
<td>2.96(^{jk})</td>
</tr>
<tr>
<td>QOL (Parent report)</td>
<td>Physical</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1.85</td>
<td>2.38</td>
<td>2.15</td>
</tr>
<tr>
<td></td>
<td>Emotional</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.59</td>
<td>2.76</td>
<td>2.55</td>
</tr>
<tr>
<td></td>
<td>Social</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.23</td>
<td>2.54</td>
<td>2.28</td>
</tr>
<tr>
<td></td>
<td>School</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.38</td>
<td>2.47</td>
<td>2.05</td>
</tr>
<tr>
<td>QOL (Child report)</td>
<td>Physical</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.39</td>
<td>2.76(^l)</td>
<td>2.03(^l)</td>
</tr>
<tr>
<td></td>
<td>Emotional</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.68</td>
<td>2.50</td>
<td>2.63</td>
</tr>
<tr>
<td></td>
<td>Social</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.67</td>
<td>2.80</td>
<td>2.29</td>
</tr>
<tr>
<td></td>
<td>School</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.48</td>
<td>2.45</td>
<td>2.02</td>
</tr>
</tbody>
</table>

*Note.* Means with the same letters are significantly different from each other (*p*<.05).
Table 11: Means for each outcome variable by cluster, for 4 cluster solution

<table>
<thead>
<tr>
<th>Scale</th>
<th>Cluster 1 (n=39): Average cognitive, impaired motor</th>
<th>Cluster 2 (n=32): Average cognitive, low avg attention</th>
<th>Cluster 3 (n=25): Extremely low to borderline</th>
<th>Cluster 4 (n=12): Non-completers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Independence</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fine motor</td>
<td>43.09&lt;sup&gt;a&lt;/sup&gt;</td>
<td>48.36&lt;sup&gt;ab&lt;/sup&gt;</td>
<td>41.56&lt;sup&gt;b&lt;/sup&gt;</td>
<td>33.89</td>
</tr>
<tr>
<td>Money</td>
<td>23.42&lt;sup&gt;c&lt;/sup&gt;</td>
<td>32.10&lt;sup&gt;de&lt;/sup&gt;</td>
<td>19.44&lt;sup&gt;d&lt;/sup&gt;</td>
<td>13.72&lt;sup&gt;e&lt;/sup&gt;</td>
</tr>
<tr>
<td>Language</td>
<td>39.58&lt;sup&gt;f&lt;/sup&gt;</td>
<td>44.40&lt;sup&gt;gh&lt;/sup&gt;</td>
<td>35.29&lt;sup&gt;g&lt;/sup&gt;</td>
<td>26.72&lt;sup&gt;h&lt;/sup&gt;</td>
</tr>
<tr>
<td>Time</td>
<td>42.55&lt;sup&gt;i&lt;/sup&gt;</td>
<td>47.33&lt;sup&gt;jk&lt;/sup&gt;</td>
<td>39.46&lt;sup&gt;il&lt;/sup&gt;</td>
<td>23.00&lt;sup&gt;ijkl&lt;/sup&gt;</td>
</tr>
<tr>
<td>Academic success</td>
<td>46.38&lt;sup&gt;mn&lt;/sup&gt;</td>
<td>45.00&lt;sup&gt;op&lt;/sup&gt;</td>
<td>36.90&lt;sup&gt;mo&lt;/sup&gt;</td>
<td>35.63&lt;sup&gt;np&lt;/sup&gt;</td>
</tr>
<tr>
<td>Future expectations</td>
<td>3.35&lt;sup&gt;r&lt;/sup&gt;</td>
<td>3.68&lt;sup&gt;rqst&lt;/sup&gt;</td>
<td>3.00&lt;sup&gt;s&lt;/sup&gt;</td>
<td>2.23&lt;sup&gt;rt&lt;/sup&gt;</td>
</tr>
<tr>
<td>QOL (Parent report)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physical</td>
<td>1.85</td>
<td>2.38</td>
<td>2.15</td>
<td>1.72</td>
</tr>
<tr>
<td>Emotional</td>
<td>2.59</td>
<td>2.76</td>
<td>2.55</td>
<td>2.49</td>
</tr>
<tr>
<td>Social</td>
<td>2.23</td>
<td>2.54</td>
<td>2.28</td>
<td>2.06</td>
</tr>
<tr>
<td>School</td>
<td>2.38</td>
<td>2.47</td>
<td>2.05</td>
<td>2.01</td>
</tr>
<tr>
<td>QOL (Child report)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physical</td>
<td>2.39</td>
<td>2.76&lt;sup&gt;a&lt;/sup&gt;</td>
<td>2.03&lt;sup&gt;u&lt;/sup&gt;</td>
<td>1.80</td>
</tr>
<tr>
<td>Emotional</td>
<td>2.68</td>
<td>2.50</td>
<td>2.63</td>
<td>2.83</td>
</tr>
<tr>
<td>Social</td>
<td>2.67</td>
<td>2.80</td>
<td>2.29</td>
<td>3.26</td>
</tr>
<tr>
<td>School</td>
<td>2.48</td>
<td>2.45</td>
<td>2.02</td>
<td>2.00</td>
</tr>
</tbody>
</table>

*Note.* Means with the same letters are significantly different from each other (*p*<.05).
Table 12: Shunt status and cluster membership, for 4 cluster solution

<table>
<thead>
<tr>
<th>Shunt status</th>
<th>Cluster 1 (n=39): Average cognitive, impaired motor</th>
<th>Cluster 2 (n=32): Average cognitive, low avg attention</th>
<th>Cluster 3 (n=25): Extremely low to borderline</th>
<th>Cluster 4 (n=12): Non-completers</th>
</tr>
</thead>
<tbody>
<tr>
<td>With shunt % (n)</td>
<td>89.7% (35)</td>
<td>65.6% (21)</td>
<td>96.0% (24)</td>
<td>100% (12)</td>
</tr>
<tr>
<td>Without shunt % (n)</td>
<td>10.3% (4)</td>
<td>34.4% (11)</td>
<td>4.0% (1)</td>
<td>0% (0)</td>
</tr>
</tbody>
</table>
Figure 1: Hypothetical profiles indicative of neurocognitive heterogeneity
Figure 2: Hypothesized clusters with similar neuropsychological profiles

<table>
<thead>
<tr>
<th>Less Biological Severity</th>
<th>Greater Biological Severity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cluster 1: Generally higher functioning (than cluster 2) with significant variability within the neurocognitive profile.</td>
<td>Cluster 2: Generally lower functioning (than cluster 1) with significant variability within the neurocognitive profile.</td>
</tr>
<tr>
<td>Cluster 3: Generally higher functioning (than cluster 4) with similar performance within the neurocognitive profile.</td>
<td>Cluster 4: Generally lower functioning (than cluster 3) with similar performance within the neurocognitive profile.</td>
</tr>
</tbody>
</table>
Figure 3: Hypothesized level of functional assets and deficits for hypothesized clusters

<table>
<thead>
<tr>
<th>Cluster</th>
<th>Cluster 1</th>
<th>Cluster 2</th>
<th>Cluster 3</th>
<th>Cluster 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associative processing</td>
<td>High</td>
<td>Medium</td>
<td>High</td>
<td>Low</td>
</tr>
<tr>
<td>Assembled processing</td>
<td>Low</td>
<td>High</td>
<td>Medium</td>
<td>Low</td>
</tr>
</tbody>
</table>
Figure 4: Wards linkage cluster profiles
Figure 5: Average linkage cluster profiles
Figure 6: K-means cluster profiles
REFERENCE LIST


Hollingshead, A. A. (1975). Four-factor index of social status. Unpublished manuscript, Yale University, New Haven, CT.


Iddon, J. L., Morgan, D. J., Loveday, C., Sahakian, B. J., & Pickard, J. D. (2004). Neuropsychological profile of young adults with spina bifida with or without


VITA

Rachel Wasserman was born and raised near Orlando, FL. Before attending Loyola University Chicago, she attended the Illinois Institute of Technology, where she earned a Bachelor of Science in Psychology in 2007.

While at Loyola, Dr. Wasserman participated in several clinical practicums in areas of pediatric psychology and pediatric neuropsychology. She conducted in her predoctoral internship at Texas Children's Hospital, Baylor College of Medicine. Dr. Wasserman also served on several committees, including the Clinical Psychology Information Committee and the Practicum Committee. As well, Dr. Wasserman was awarded the Child and Family Research Assistantship, the Advanced Doctoral Fellowship, and the American Psychological Association Graduate Student Ethics Prize.