RELATIONSHIP BETWEEN EXECUTIVE FUNCTIONING AND ADHERENCE IN YOUTH WITH SICKLE CELL DISEASE

A dissertation submitted
to Kent State University in partial fulfillment of the requirements for the degree of Doctor of Philosophy

By
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CHAPTER 1

INTRODUCTION

Executive Functioning (EF) is an umbrella term that refers to the high-level processes of a group of domains responsible for purposeful behavior (Anderson, 2002). Despite efforts to clarify EF and its domains, the concept remains vague and the domains have been under-specified (Lehto, Juujarvi, Kooistra, & Pulkkinen, 2003). Most researchers include inhibition, planning, organization, decision making, judgment, reasoning, cognitive flexibility (set shifting), and working memory as abilities involved in executive functioning (Alexander & Stuss, 2000; Anderson, 2002; Kelly, 2000; Lehto et al., 2003). Deficits in executive functioning not only influence cognitive processes but also affective responses and behaviors (Anderson, 2002). For example, individuals with executive dysfunction may exhibit argumentativeness, impulsivity, unwillingness to change behaviors, and poor interpersonal skills. Deficits in EF have been identified in several pediatric clinical populations, including patients with sickle cell disease (SCD) (Anderson, 2002).

SCD is a genetic blood disorder in which red blood cells form “sickle” shapes and lead to occlusion of blood vessels (vaso-occlusion), reduced blood flow to vital organs (ischemia), and a compromised immune system (Roseff, 2009). For children and adolescents with SCD, adhering to a prescribed medical regimen has been associated with a reduction in the risk of poor treatment outcome. For example, research studies show that adherence to medical regimens produce fewer number of pain crises in youth with SCD (Fisak, Belkin, von Lehe, & Bansal, 2011). Despite the relationship between positive medical outcomes and adherence, adherence is
lacking in youth with SCD. Previous research examining individual components of the regimen have found that two-thirds of youth have “less than optimal” or “poor” adherence to their medication regimen (Witherspoon & Drotar, 2006) and as few as 46% regularly attend clinic appointments (Modi, Crosby, Hines, Drotar, & Mitchell, 2012). Still, compared to other chronic illnesses (e.g., asthma, diabetes), little has been done to assess predictors of adherence in youth with SCD (Fisak et al., 2011; Quittner, Modi, Lemanek, Ievers-Landis, & Rapoff, 2008). The current study examined executive functioning and parent versus patient responsibility as predictors of adherence in these youth.

Research with various chronic illness populations shows that impairments in EF can directly affect a youth’s adherence to his or her medical treatment. For example, research with youth with chronic kidney disease indicates that deficits in attention can negatively influence a youth’s ability to learn new treatments and recall instructions given by the medical provider (Gerson et al., 2006). Memory deficits can inhibit a patient from remembering an added medication or treatment goal. Additionally, research with pediatric SCD populations as well as a review of chronic illness outcomes in general (for all ages) has shown that patients with executive impairments have poorer quality of life, which can lead to worse adherence and a decrease in the probability of completion of the treatment regimen (Fisak et al., 2011; Hopkins & Jackson, 2006).

Adolescence is a critical period in which to monitor adherence in any chronic illness population as this is the developmental period when adherence rates are lowest (Partridge, Avorn, Wang, & Winer, 2002). A likely cause for this poor adherence is the transition of responsibility for disease management from the parent to the adolescent during adolescence (Sawyer & Aroni, 2005). In addition to the difficulty of assuming new responsibilities, adolescents must wrestle with
normative developmental issues, including developing executive functioning. A review of studies looking at the relationship between age and adherence to chronic illness regimens demonstrates that adherence difficulties increase dramatically around age 11 and peak in mid-adolescence (Shaw, 2001).

Several barriers may be impeding adherence in adolescents. Adolescents are dealing with social pressures and have difficulty with regimens that require major lifestyle changes (e.g., dietary restrictions) or have perceived cosmetic side effects (e.g., hair loss as a side effect of hydroxyurea usage in SCD) (La Greca & Bearman, 2003). Adolescents with SCD identified forgetting to take medication or loss of medication as a primary barrier to following recommendations for pain management and pharmacological treatments (Modi et al., 2009). Further, they identified the desire to appear normal and questions about the efficacy of medications as additional barriers to following recommendations (Modi et al., 2009). A promising suggestion for improving pediatric adherence has been to tailor interventions based on the cognitive skills of youth, particularly executive functioning (McNally, Rohan, Pendley, Delamater, & Drotar, 2010). Executive functioning has often been suggested as a necessary prerequisite for adherence. However, while many authors have voiced the need for integrating EF training into adherence protocols, more empirical data is needed regarding the relationship between EF and adherence.

**Development of EF in Youth**

Historically, EF was thought of as a single construct, but has since morphed into a conceptualization as multiple inter-dependent components (Anderson, 2002). This conceptualization is supported by exploratory factor analyses (EFA) that have produced distinct, separable factors from a battery of EF tasks (Lehto et al., 2003; Miyake et al., 2000). Further, neuroanatomical findings of localized executive dysfunction in specific frontal systems, rather
than global executive dysfunction, suggest a nonunitary structure of executive functioning (Anderson, 2002). The differentiation between executive functioning and cognitive abilities (e.g., IQ score) has also not been made consistently clear, though EF typically is said to fit under the larger construct of cognitive functioning (Salthouse, 2005). Some skills, such as working memory, are part of many standardized intelligence tests and are also considered a construct of executive functioning, while others, such as shifting and inhibiting, have been found to be uncorrelated with general intelligence (Friedman et al., 2006).

Executive functioning deficits in youth include poor impulse control, disinhibition, difficulty planning and organizing, problems with goal setting and decision-making, working memory problems, difficulties with sequential processing, and mental inflexibility (Anderson, 2002). Factor analytic studies have distributed these variables into three to four factor solutions (Anderson, 2002; Kelly, 2000; Lehto et al., 2003; Levin et al., 1991; Miyake et al., 2000; Welsh, Pennington, & Groisser, 1991). There were similarities in the factor structures that emerged from these studies despite variability in test batteries and age ranges. Specifically, many of the solutions included planning, impulse control, concept reasoning, response speed, inhibition, and shifting.

The maturation of the various domains of executive functioning occurs at different developmental periods in youth, although the exact developmental profile is still unclear (Anderson, 2002). Attentional control (e.g., ability to inhibit behavior or exert impulse control, the capacity to attend to certain stimuli while ignoring others) is identified as early as 12 months, at which time infants have been seen to inhibit some behaviors in favor of a new response set (Anderson, 2002; Diamond & Goldman-Rakic, 1989). Improvements on tasks involving self-regulation and impulse control continue to be seen during early childhood (Anderson, 2002;
Espy, Kaufmann, McDiarmid, & Glisky, 1999). By nine years of age, children are monitoring and regulating their actions fairly well; however, an increase in impulsivity is seen during the onset of adolescence, around age 11 (Anderson, 2002).

Cognitive flexibility (e.g., the ability to shift between tasks, divide attention) has been identified in children as young as three, as they show some ability to switch between response sets. However, this ability weakens when rules become complex. The ability to conquer multidimensional switching tasks sees a marked increase between ages seven and nine, and continues to improve into adolescence (Anderson, 2002; Luciana & Nelson, 1998).

Goal setting (e.g., taking initiative, planning/organization) has been demonstrated in children as young as four years. Between the ages of six and seven, children’s goal setting abilities develop more rapidly and continue this rapid development until age nine or ten (Anderson, 2002; Welsh et al., 1991). Their simple planning and organizational strategies become more organized and complex and they perform easier tasks more effortlessly (Anderson, 2002). Other studies have also found gains in planning and organization during this developmental period (Anderson, Anderson, & Garth, 2001; Anderson, Anderson, & Lajoie, 1996; Levin et al., 1991). More improvements in several areas, including decision making, continue into adolescence (Anderson et al., 2001a; Anderson, Anderson, Northam, Jacobs, & Catroppa, 2001; Levin et al., 1991).

Information processing (e.g., efficiency and fluency of processing information, processing speed) shows improvement between the ages of three and five (Espy, 1997; Gerstadt, Hong, & Diamond, 1994). Significant gains in processing speed continue to occur between the ages of 9 and 15 (Anderson et al., 2001b, Kail, 1986). For example, 15-year-olds had significantly faster processing speeds than 11-to 13-year-olds on a task requiring the naming of
shapes and colors (Anderson et al., 2001b). Overall, the development and maturation of different executive functioning processes progresses from infancy to adolescence (Lehto et al., 2003).

**EF Deficits in Youth with Chronic Illness**

A portion of the research on EF in youth describes executive functioning deficits in youth with various chronic illnesses (e.g., Armstrong et al., 1996; Berkelhammer et al., 2007; Gerson et al., 2006; Gipson et al., 2006; Northam et al., 2001). Even though executive functioning deficits may be a predictor of nonadherence, executive functioning impairments are not recognized or assessed by physicians in up to 90% of patients (Hopkins & Jackson, 2006). This lack of recognition in clinical settings may stem from the fact that many neurocognitive deficits are not physically evident, and the patient may appear alert and oriented (Hopkins & Jackson, 2006). Further, heterogeneity in the presentation of executive functioning deficits exists between and within illness groups, making these impairments more difficult to identify (Jeste et al., 2003).

It is possible that components of the medical disorder itself have affected the youth’s executive functioning. First, structural lesions resulting from such disruptions as stroke and infectious abscesses can create EF deficits (Schillerstrom, Horton, & Royall, 2005). In addition, functional disruptions associated with the illness can affect executive functioning. These disruptions include metabolic dysfunctions, nutritional deficiencies, cytokine-mediated behavioral changes, and effects of medication (Schillerstrom et al., 2005). More research is needed to determine to what extent these disruptions are responsible for the EF deficits that have been found in youth with chronic illnesses and what effect they have on adherence (Fennell et al., 1990; Gerson et al., 2006; Gipson et al., 2006).

**EF Deficits in Youth with SCD**
SCD is an illness in which the medical disorder itself can affect executive functioning (Roseff, 2009). For example, chronic hemolytic anemia and vaso-occlusion seen in youth with SCD can cause hypoxia and strokes (Noll et al., 2001). A study examining magnetic resonance imaging (MRI) data in a sample of youth with SCD found that 59% of those that had experienced a stroke or infarct suffered structural damage to the frontal lobe, a key center for executive functioning (Gold, Johnson, Treadwell, Hans, & Vichinsky, 2008). A similar study utilizing neuroimaging found that 93% of youth with SCD that had suffered frontal lobe injury as a result of a silent cerebral infarct had accompanying neuropsychological deficits (Schatz, Brown, Pascual, Hsu, & DeBaun, 2001).

Previous research has consistently shown EF deficits in youth with SCD, especially in those that have experienced a cerebral infarction or other stroke. Youth with SCD with frontal-lobe infarction made significantly more perseverative errors on a card sorting task than youth with SCD without infarcts and siblings without SCD (Watkins et al., 1998). Additionally, attentional deficits have been found in children with SCD who have experienced stroke, evidenced by slow disengagement of attention on a visual orienting task (Craft, Schatz, Glauser, Lee, & DeBaun, 1994). In examining cognitive flexibility, children and adolescents with SCD who showed frontal infarcts on an MRI had a harder time manipulating information in working memory than the SCD group with no history of stroke or visible infarct (Brandling-Bennett, White, Armstrong, Christ, & DeBaun, 2003). An analysis of how the location of the infarct can affect cognitive flexibility in youth with SCD found that children and adolescents with infarcts in posterior brain regions performed more similarly to youth without a history of stroke or infarct on working memory tasks than youth with anterior or diffuse infarcts (White, Salorio, Schatz, &
DeBaun, 2000). Specifically, deficits were observed in the phonological loop, part of the working memory system, in children with anterior and diffuse infarcts (White et al., 2000).

Children with SCD also performed poorly on tasks requiring organization (e.g., object assembly) and the processing of information (Armstrong et al., 1996; Nabors & Freymuth, 2002). These youth demonstrated slow processing speed on a coding task, and this processing speed deficit was amplified in those with a history of stroke (Armstrong et al., 1996; Nabors & Freymuth, 2002). As executive functioning deficits likely affect a patient’s adherence to his or her medical regimen, and adherence is strongly related to outcome for chronic illnesses such as SCD, the EF-adherence relationship should be addressed (DiMatteo, Giordani, Lepper, & Croghan, 2002).

**The EF-Adherence Linkage: Differences in Youth versus Adults**

Though there is a body of literature on the EF-adherence linkage in adults, results from this research should not be generalized to the pediatric population. Adults are assumed to have normal levels of EF, so any deficits that exist are presumed to be due to the illness or injury. In youth, executive functioning is likely still developing, making it harder to differentiate between deficits due to illness/injury and those that have not developed normally in the first place.

The research in this area for both youth and adults is becoming more reliable as studies are using more objective measures of executive functioning and adherence as opposed to patient self-report, which is often influenced by social desirability and overestimates adherence (Rapoff, 1999). Objective measures of adherence in chronic illness populations include drug assays, pill counts, and electronic monitoring devices (e.g., memory chips in glucose reflectance meters for patients with diabetes) (La Greca & Bearman, 2003).

Research on this topic in the adult literature has focused largely on HIV populations because of the serious consequences of nonadherence to its treatment regimen, highly active
antiretroviral therapy (HAART). Any lack of adherence to this regimen could lead to HIV that is resistant to the prescribed medications and could result in increased mortality rates (Contardo, Black, Beauvais, Dieckhaus, & Rosen, 2009; Hinkin et al., 2004). Not surprisingly, this research has consistently found that executive functioning is positively associated with adherence in HIV patients (Contardo et al., 2009; Hinkin et al., 2004; Hinkin et al., 2002; Wagner, 2002).

The research on executive functioning and adherence is critically important for pediatric illness populations such as adolescents with SCD, as many of the medical regimens for these illnesses include various components that require the integration of several pieces of information (Bagner, Williams, Geffken, Silverstein, & Storch, 2007). For example, these youth must navigate complex medical regimens which may include remembering schedules and dosages, understanding and implementing both provider and parent instructions, and effectively communicating their own health care needs (Malee et al., 2009). Research in this area, particularly with younger children, has often relied on parent report of adherence and executive functioning, as parents are central to the health care of their children and often play a critical role in assisting with their care (Bagner et al., 2007). One study with youth with SCD found that those with the worst scores on a measure of planning and organization were the most adherent to their medication regimen (Treadwell et al., 2005). Caregivers of the most adherent youth were more involved in completion of the treatment regimen than caregivers in the less adherent groups, suggesting that parental responsibility plays a role in the adherence relationship.

**Responsibility’s Role in the EF-Adherence Relationship in Youth with SCD**

The division of responsibility between the parent and child for completion of the treatment regimen may influence adherence in youth with SCD. Particularly during the transition to adolescence, there generally comes a gradual shift in some responsibilities from the parent to the child. However, the adolescent’s drive for self-sufficiency is sometimes met with resistance
by a parent, who is reluctant to hand over the responsibility. It is imperative that both the parent and the child know who is responsible for ensuring the successful completion of the daily treatment regimen (Horton, Berg, Butner, & Wiebe, 2009).

Parental responsibility can take many forms, including reminding the child or adolescent to complete his or her treatment regimen, monitoring completion of the health care tasks, or performing the health care recommendation for the child (e.g., giving the medication directly to the child). Previous research with youth with diabetes found that more parental involvement, parental support, and parental monitoring are related to better adherence (Ellis et al., 2007; Horton et al., 2009; Palmer et al., 2011). As executive function contributes greatly to a patient’s decision-making ability, children and adolescents with EF deficits may rely more on their parents for completion of treatment recommendations (Schillerstrom et al., 2005). For youth with EF deficits, parents’ willingness to take responsibility for some of the treatment regimen can likely serve as a buffer against nonadherence. However, children and adolescents without parents or other support systems willing to take on some responsibility for treatment completion are likely at an increased risk for nonadherence.

The idea that responsibility may influence the EF-adherence relationship has been supported in a couple of studies examining youth with chronic illness (Malee et al., 2009; Treadwell et al., 2005). Children and adolescents with HIV infection had below average scores in distractibility, organization, processing speed, and working memory. Despite these impairments compared to normative data, their adherence to HIV medication was good overall (Malee et al., 2009). Further analyses determined that having an adult responsible for medication management was a significant predictor of adherence. Therefore, caregiver participation in ensuring completion of the treatment regimen can compensate for EF deficits in these youth.
(Malee et al., 2009). This pattern of caregivers’ participation in the regimen making up for EF impairments in these pediatric populations was similar to that found in the group of youth with SCD (Treadwell et al., 2005), in which the most adherent youth (whose caregivers were more involved) actually had the most EF deficits, suggesting it was caregiver responsibility that moderated this relationship. Despite these findings, many studies in the EF-adherence arena fail to examine parent’s role in their youth’s adherence. More work is needed to elucidate the role of responsibility in the EF-adherence relationship.

Although the current study focuses on responsibility as a key contributor to nonadherence, past research has identified specific treatment regimen factors, demographic factors, mental health concerns, and other family stressors as other potential influences. However, these influences have not been found significant in all cases, and results from any analysis should not be overgeneralized to all illness groups. Additionally, each illness likely has its own set of characteristics that are influenced by EF or that make adherence more difficult. For instance, family stressors may be more common in illnesses that are generally associated with low SES and minority race, such as SCD (Malee et al., 2009). We should be mindful of how an adolescent’s executive functioning could affect their adherence regardless of the aforementioned factors (Bagner et al., 2007). The purpose of this study is to explore the relationship between neurocognitive factors, particularly executive functioning, and adherence in adolescents with SCD, and to further examine responsibility’s role as a potential contributor to nonadherence. Further, as this study seeks to examine whether any deficits in the executive functioning-adherence relationship are a function of the disease versus a function of the normal developmental trajectory, this study will compare the EF-adherence relationship in the adolescents with SCD to a group of matched non-affected controls. Previous research has used
matched control groups to examine differences in youth with SCD (e.g., Berg, Edwards, & King, 2012).

**Specific Aims and Hypotheses**

**Aim 1:** Examine the nature of the relationship between neurocognitive factors and adherence in adolescents with sickle cell disease.

*Hypothesis 1:* Estimated IQ score (WRAT-4 score) will independently predict adherence rates in adolescents, with higher estimated IQ scores predicting more adherence.

Previous research has found that youth with SCD score lower than youth without SCD on measures of intelligence (Schatz, Finke, Kellett, & Kramer, 2002). These deficits may lead to difficulty implementing a complex medical regimen, such as that required for management of SCD.

*Hypothesis 2:* Executive functioning will be a stronger predictor of adherence than estimated IQ score.

A meta-analysis that examined different domains of cognitive functioning in youth with SCD found executive functioning to be the domain in which youth scored the lowest (Schatz et al., 2002). Components involved in executive functioning are directly involved in regimen completion (e.g., organizing a medication schedule) in youth with SCD, and these skills (i.e., organization/planning) likely facilitate adherence. Additionally, a positive relationship between EF and adherence has been demonstrated in the adult chronic illness literature (e.g., Contardo et al., 2009). Thus, there is a possibility that EF has an effect on treatment adherence in adolescents.

**Aim 2:** Investigate the nature of the relationship between parental role of responsibility and adherence in adolescents with sickle cell disease.
**Hypothesis 3**: Parental role of responsibility will independently predict adherence rates in adolescents with sickle cell disease, with higher rates of parent responsibility predicting more adherence.

Research addressing responsibility for regimen completion found that greater parent responsibility for ensuring regimen completion is associated with greater adherence across chronic illnesses (e.g., Ellis et al., 2007; Malee et al., 2009).

**Aim 3**: Compare the EF-adherence relationship in adolescents with SCD to a group of matched controls.

**Hypothesis 4**: The control group will demonstrate overall higher levels of executive functioning than the group with SCD.

Past research has noted EF deficits in youth with SCD (e.g., Armstrong et al., 1996; Nabors & Freymuth, 2002; Schatz & McClellan, 2006). Thus, it is likely they will score lower on measures of EF than a group of matched peers with no known medical condition.

**Hypothesis 5**: The EF-adherence relationship in the control group will be similar to that in the group with SCD, with higher rates of executive functioning predicting more adherence to daily activities.

One reason supporting the linkage between executive functioning and adherence in literature is that components of EF are critical to the initiation and follow-through of tasks (McNally et al., 2010). Therefore, youth with lower EF will likely be less adherent to daily tasks.

**Hypothesis 6**: Parental role of responsibility will independently predict adherence rates in adolescents in the control group, with higher rates of parent responsibility predicting more adherence to daily activities.
Similar to the group with SCD, when parents are more involved in ensuring completion of tasks, there is likely a greater chance for the execution of or adherence to these tasks (e.g., Treadwell et al., 2005).
CHAPTER 2

METHOD

Participants

Participants were 25 adolescents between 12-18 years old with SCD. Two participants refused to participate in the study for a refusal rate of 7%. The control group consisted of 25 adolescents with no current medical condition. The groups were not significantly different on age ($t(48) = -.43, p > .05$), gender ($\chi(1) = .00, p > .05$), or race ($\chi(2) = 4.93, p > .05$) (see Tables 1 and 2 for complete demographic information for both groups). Using G-power software, a large effect size requires 25 participants to yield a power of .8 for the proposed hypotheses. Further, a study with 15-30 participants is representative of what is found in the pediatric SCD literature (e.g., Treadwell et al., 2005).

Measures

When possible, psychometrically sound instruments that have been used with African-American youth were utilized in this study. Methods of collecting adherence data often differ based on age of participants. For instance, in studies using younger children (e.g., Witherspoon & Drotar, 2006), parent-report of adherence is often utilized. During the shift to adolescence, self-report is often utilized to account for the shift in responsibility (e.g., Modi et al., 2012; Modi et al., 2009). As the current study was with adolescents, self-report data was primarily utilized.

Assessment of Individual/Family Factors in Matched Controls

Parents completed a form with basic demographic information (e.g., age, medical history). Adolescents completed a packet that included the following measure:
Table 1

*Demographics of SCD Group*

<table>
<thead>
<tr>
<th></th>
<th>Mean (SD)</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child Variables</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td></td>
<td>44%</td>
</tr>
<tr>
<td>Age</td>
<td>14.10 (2.06)</td>
<td></td>
</tr>
<tr>
<td>Grade</td>
<td>8.26 (1.98)</td>
<td></td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
</tr>
<tr>
<td>African American</td>
<td></td>
<td>80%</td>
</tr>
<tr>
<td>Biracial</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Did Not Report</td>
<td></td>
<td>8%</td>
</tr>
<tr>
<td>Insurance</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Medicaid</td>
<td></td>
<td>76%</td>
</tr>
<tr>
<td>Private</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Did Not Report</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Number of ED visits</td>
<td></td>
<td>2.36 (2.31)</td>
</tr>
<tr>
<td>(previous year)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Parent Variables</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parent Relationship</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mother</td>
<td></td>
<td>80%</td>
</tr>
<tr>
<td>Father</td>
<td></td>
<td>4%</td>
</tr>
<tr>
<td>Grandmother</td>
<td></td>
<td>4%</td>
</tr>
<tr>
<td>Other</td>
<td></td>
<td>4%</td>
</tr>
<tr>
<td>Did Not Report</td>
<td></td>
<td>8%</td>
</tr>
<tr>
<td>Single Parent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td></td>
<td>83%</td>
</tr>
<tr>
<td>No</td>
<td></td>
<td>17%</td>
</tr>
<tr>
<td>Education Level</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Did not graduate HS</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>HS or GED</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Some College</td>
<td></td>
<td>40%</td>
</tr>
<tr>
<td>Associates or Bachelor’s Degree</td>
<td></td>
<td>24%</td>
</tr>
<tr>
<td>Did Not Report</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Insurance</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Medicaid</td>
<td></td>
<td>80%</td>
</tr>
<tr>
<td>Private</td>
<td></td>
<td>12%</td>
</tr>
<tr>
<td>Did Not Report</td>
<td></td>
<td>8%</td>
</tr>
</tbody>
</table>
Table 2

*Demographics of Control Group*

<table>
<thead>
<tr>
<th></th>
<th>Mean (SD)</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child Variables</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td></td>
<td>44%</td>
</tr>
<tr>
<td>Age</td>
<td>14.32 (1.55)</td>
<td></td>
</tr>
<tr>
<td>Grade</td>
<td>8.56 (1.69)</td>
<td></td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
</tr>
<tr>
<td>African American</td>
<td>80%</td>
<td></td>
</tr>
<tr>
<td>Biracial</td>
<td>4%</td>
<td></td>
</tr>
<tr>
<td>Other/Did Not Report</td>
<td>16%</td>
<td></td>
</tr>
<tr>
<td><strong>Insurance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Medicaid</td>
<td>72%</td>
<td></td>
</tr>
<tr>
<td>Private</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>Did Not Report</td>
<td>8%</td>
<td></td>
</tr>
<tr>
<td><strong>Parent Variables</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Education Level</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Did not graduate HS</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>HS or GED</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>Some College</td>
<td>32%</td>
<td></td>
</tr>
<tr>
<td>Associates or Bachelor’s Degree</td>
<td>20%</td>
<td></td>
</tr>
<tr>
<td>Graduate or Advanced Degree</td>
<td>8%</td>
<td></td>
</tr>
<tr>
<td><strong>Insurance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Medicaid</td>
<td>24%</td>
<td></td>
</tr>
<tr>
<td>Private</td>
<td>24%</td>
<td></td>
</tr>
<tr>
<td>Self-Pay</td>
<td>4%</td>
<td></td>
</tr>
<tr>
<td>Did Not Report</td>
<td>48%</td>
<td></td>
</tr>
</tbody>
</table>

*Note.* Only limited demographic data were collected on families of control group participants. Therefore, data such as who completed the packet and marital status was not gathered for the control group.

*Adherence and Responsibilities Questionnaire (Appendix A).* The Adherence and Responsibilities Questionnaire is a 27-item questionnaire that was modified for the current study from the Child Routines Inventory (CRI) of Sytsma, Kelley, and Wymer (2001) to adapt the questionnaire to an urban, low-income population. Teachers from the participants’ school were consulted in the adaptation of the measure, and they gave suggestions on which questions should be modified. For example, “I eat meals with family at the table each day” was changed to “I eat
dinner at dinner table” to reflect the school’s report that many of their students are in single-parent homes where a parent may work at night. This modified questionnaire measures adherence to activities of daily living (e.g., “I leave for school on time every day”; “I complete homework on time”), as well as who is taking responsibility for these tasks. Each activity asks for both an adherence and responsibility rating. Adherence responses are presented on a Likert scale ranging from 0 (never) to 4 (nearly always), with higher composite scores reflective of higher levels of adherence to daily activities (potential range = 0 to 108). Participants can respond “not applicable” if a particular activity is not part of their regular regimen. If a question is not applicable to a participant (e.g., they did not go to church or participate in a sport), this question was not included in the average, thus not affecting their total score.

For the responsibility rating, participants respond to each item using a Likert-type scale, from 1 (parent(s) take responsibility all of the time) to 5 (child takes responsibility all of the time), with higher composite scores reflective of more child responsibility (potential range = 22 to 110). To preserve clarity, five items only ask for an adherence rating and do not ask for a responsibility rating (e.g., “I get told by my parents or guardian what time to be home”). The original CRI exhibits good content validity, excellent internal consistency (α = .90), and good test-retest reliability (r = .86) (Sytsma et al., 2001).

**Assessment of Individual/Family Factors in Youth with SCD**

Adolescents and their caregivers completed a packet that contained the following measures:

*Adherence and Responsibilities Questionnaire.* This questionnaire (see description above) was also given to patients with SCD in order to allow direct comparisons with the control group on adherence and responsibilities ratings. However, the Self-Care Inventory (see
description below) was utilized to measure adherence toward the specific treatment regimen for SCD in these youth.

*Demographic Form (Appendix B).* Parents of youth with SCD completed a demographics form that included questions about gender, age, ethnicity, insurance coverage and education level of both themselves and their adolescent.

*Responsibilities Questionnaire (patient and parent version; Appendices C & D).* The Responsibilities Questionnaire is a five-item measure created for the current study to assess how much the parent and the youth are taking responsibility for different tasks related to a sickle cell regimen. The questionnaire was modified from the Asthma Responsibilities Questionnaire (McQuaid et al., 2001). Consultation with hematology physicians and nurse practitioners identified the five most common tasks in a sickle cell treatment regimen (e.g., restriction of physical activities, taking medications); these tasks were used to construct this questionnaire. Participants respond to each item using a Likert-type scale, from 1 (*parent(s) take responsibility all of the time*) to 5 (*child takes responsibility all of the time*), with higher composite scores reflective of more child responsibility (potential range = 5 to 25). An example of an item on the questionnaire is “Who takes responsibility for making sure I don’t do activities I’m not supposed to do?” The original ARQ has good convergent validity and internal consistency of alpha = .75-.78 for child report and alpha = .84-.87 for parent report (McQuaid et al., 2001). This scale has been used in samples that included African-American youth (e.g., Bruzzese, Unikel, Gallagher, Evans, & Colland, 2008; McQuaid, Kopel, Klein, & Fritz, 2003).

*Self-Care Inventory (SCI; patient and parent version; Appendices E & F).* The Self-Care Inventory is a four-item adherence measure adapted from the Self-Care Inventory for Diabetes (La Greca, Swales, Klemp, & Madigan, 1988). The SCI was adapted for this study to fit a sickle
cell population. The current SCI examines adherence to various aspects of the SCD treatment regimen including: following diet, activity restriction, medication, and attendance at appointments. Responses are presented on a Likert scale ranging from 1 (never) to 5 (always), with higher composite scores reflective of higher levels of adherence to treatment (potential range = 4 to 20). Participants can respond “not applicable” if a particular treatment is not part of their regimen. Internal consistency of this measure was reported to be greater than .80 in studies of children and adolescents with diabetes (e.g., Davis et al., 2001; La Greca, Swales, Klemp, Madigan, & Skyler, 1995). Validity data in a diabetes population demonstrated higher levels of self-care reportedly associated with better metabolic control (Davis et al, 2001). This scale has been used in samples that included African-American youth (e.g., Greening, Stoppelbein, Konishi, Jordan, & Moll, 2007; Lewin et al., 2009).

Neuropsychological Functioning

The neuropsychological functioning test battery assesses estimated IQ score, as well as multiple executive functioning domains including planning/organization, inhibition, and mental flexibility. New research suggests using this type of composite for executive functioning as opposed to a single measure (Schatz, Stancil, Katz, & Sanchez, 2014). The measures that were used in the current study include:

*Delis-Kaplan Executive Function System (DKEFS) Color-Word Interference Test* (Delis, Kaplan, & Kramer, 2001). The DKEFS Color-Word Interference Test is a measure of impulsivity, cognitive flexibility, and verbal inhibition. For the current study, the third component of this test (Inhibition) was used as a part of the executive functioning composite. This test is given to participants under time constraints (180 second time limit). This subtest, or the test from which it is derived (Stroop Color Word Test; Golden, 1978), has been used to
measure EF in chronic illness populations in research (e.g., Rosen et al., 2003; VanZutphen et al., 2007). Participants are required to inhibit their natural response by naming the color ink of the word which is printed in a different color ink from the color it actually names (e.g., *green* is printed in red ink). Completion time in seconds is the raw score, which is then converted to a scaled score. Scaled scores have a mean of 10 and a standard deviation of 3, with scores of 7 or lower indicating impairment. The DKEFS Color-Word Interference Test demonstrates validity and reliability, with alpha reliability estimates between .62 and .86 (Delis, Kramer, Kaplan, & Holdnack, 2004; Shunk, Davis, & Dean, 2006).

*DKEFS Trail Making Test* (Delis et al., 2001). The DKEFS Trail Making Test is a measure of set shifting (flexibility of thinking on visual-motor task) and processing speed. For the current study, the Number-Letter Switching task was used as a part of the executive functioning composite. This task requires participants to draw lines connecting a series of ascending numbers and letters while timed, alternating between numbers and letters (A-1-B-2). Completion time in seconds is the raw score, which is then converted to a scaled score. Scaled scores have a mean of 10 and a standard deviation of 3, with scores of 7 or lower indicating impairment. The DKEFS Trail Making Test demonstrates validity and reliability, with alpha reliability estimates between .57 and .81 (Delis et al., 2004; Shunk et al., 2006). This test has been used with African American youth (e.g., Staiano, Abraham, & Calvert, 2012).

*Rey Complex Figure Test* (RCFT; Meyers & Meyers, 1995; Rey, 1941). The Rey Complex Figure Test Copy Phase, a measure of planning, organization, and visuospatial constructional ability, was used as a part of the executive functioning composite in the current study. This task requires participants to copy a complex figure. For scoring, the figure is broken up into units and a score of 0, 0.5, 1, or 2 is assigned to each unit based on whether the unit is
correctly placed and/or accurately drawn (Meyers & Meyers, 1995). Raw scores are then transformed to $T$ scores ($M=50$, $SD=10$), with scores under 40 indicating possible impairment. The RCFT is reliable and valid and has been normed on a sample of children and adolescents (Meyers & Meyers, 1995).

Wide Range Achievement Test (WRAT)-4 Word Reading (Wilkinson & Robertson, 2006). The WRAT-4 Word Reading subtest is a reading test to estimate verbal IQ score and includes letter identification and word recognition. Raw scores are translated to standard scores, which have a mean of 100 and a standard deviation of 15 (potential range=55 to 145). Standard scores falling below 90 are considered below average. African-American youth were part of the norming process. Internal consistency is excellent (alpha=.92), and this test demonstrates internal and external validity (Wilkinson & Robertson, 2006).

Procedure

To collect the data from the sickle cell population, research assistants from Kent State University or hematology/oncology staff members recruited potential adolescent participants and their caregivers in the waiting room of the SCD clinic and/or during SCD support group meetings at the hospital. After consent from the parent/legal guardian and assent/consent from the adolescents were obtained, families were given a packet of measures, which included the measures described above. Research assistants were available to all participants if any questions arose during the completion of the questionnaires; specific questions could be clarified and items could be read to participants. To assess for cerebral compromise (e.g., stroke, cerebral infarct), imaging information was collected from the electronic medical record for each participant with SCD. The participants were then scheduled for neuropsychological testing at a subsequent visit. The neuropsychological evaluations were supervised by a licensed clinical neuropsychologist. During
the neuropsychological testing appointment, the adherence and responsibility questionnaires were readministered to get a current picture of these variables. The time between when the initial packet was completed and neuropsychological testing varied, as participants had different lengths of time between appointments (Mean time between initial packet and neuropsych measures = 8 months). This project was approved by the Institutional Review Boards of Kent State University and Akron Children’s Hospital.

The control group was recruited from I CAN schools in Cleveland, Ohio, a system of public charter schools. This school system is demographically similar (e.g. race, age, SES) to the sickle cell cohort that was used in this study. Adolescents were recruited through letters sent home to families. After assent and consent were obtained, graduate students and research assistants from Kent State University administered the packet of measures and the neuropsychological testing in the school setting. Adolescents in both the sickle cell and control group were given a gift card for their participation in the study.

**Analysis Plan**

**Preliminary Analyses**

To achieve consistent interpretation of data across measures, raw and/or standard scores on neuropsychological measures were transformed to T-scores ($M=50$, $SD=10$). To achieve a global executive functioning score, all T-scores on executive functioning measures were averaged, with lower T-scores indicative of worse executive functioning. Consistent with past research using overall adherence versus adherence to specific parts of the sickle cell regimen (e.g., Fisak et al., 2011), global adherence was calculated by averaging the adherence ratings for all treatment recommendations (e.g., taking medications, activity restrictions). Any recommendation not part of the participant’s regimen was not included in the average adherence score.
Histograms and z-scores identified outliers in the data (one participant in the control group and three participants in the SCD group were considered outliers on measures of EF). However, as these participants have scores that are in line with the range of EF that exists in the chronic illness literature, and as these participants did not significantly differ from the group on any other study variables, their data was retained. Age and severity (defined as objectively measured pain entrances to the hospital in the year prior to initial recruitment) were analyzed as potential covariates (see correlation matrices in Tables 3 and 4). As these correlations were not significant, these variables were not included in further analyses. Finally, all analyses were run with a bootstrapped sample with the same results.

**Primary Analyses**

_Hypothesis 1: Estimated IQ score will independently predict adherence rates in adolescents, with higher rates of estimated IQ score predicting more adherence._

A linear regression was conducted to determine whether estimated IQ score is associated with adolescent-reported adherence in youth with SCD. Estimated IQ score (WRAT-4 reading score) was entered as the independent variable and adherence was entered as the dependent variable. The model summary table of the output provided an R\(^2\) value, which was used to determine how much of the total variation in the dependent variable can be explained by the independent variable.

_Hypothesis 2: Executive functioning will be a stronger predictor of adherence than estimated IQ score._

Multiple regression was conducted to determine whether executive functioning is a stronger predictor of self-reported adherence than estimated IQ score. Estimated IQ score and
Table 3

*Correlations among Potential Covariates and Main Study Variables – Group with SCD*

<table>
<thead>
<tr>
<th>Variables</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Child Age</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>2. Severity</td>
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<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3. SCI</td>
<td>.02</td>
<td>-.02</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>4. Responsibility</td>
<td>.10</td>
<td>-.10</td>
<td>.05</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>5. EF (composite)</td>
<td>-.24</td>
<td>.29</td>
<td>-.04</td>
<td>.02</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>6. Adh - Daily</td>
<td>-.22</td>
<td>-.25</td>
<td>.24</td>
<td>-.20</td>
<td>.13</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>7. Resp - Daily</td>
<td>.36</td>
<td>-.29</td>
<td>.23</td>
<td>.05</td>
<td>-.08</td>
<td>.71**</td>
<td>-</td>
</tr>
</tbody>
</table>

Severity = ED visits in year prior to study enrollment; SCI = Self Care Inventory
Adh - Daily = Adherence to Daily Activities; Resp – Daily = Responsibility for Daily Activities

** p < .01

Table 4

*Correlations among Potential Covariates and Main Study Variables – Control Group*

<table>
<thead>
<tr>
<th>Variables</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Child Age</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>2. EF (composite)</td>
<td>-.18</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3. Adh - Daily</td>
<td>-.03</td>
<td>-.04</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>4. Resp - Daily</td>
<td>.23</td>
<td>-.01</td>
<td>.03</td>
<td>-</td>
</tr>
</tbody>
</table>

Adh - Daily = Adherence to Daily Activities; Resp – Daily = Responsibility for Daily Activities

effective functioning were entered as the independent variables and adherence was entered as the dependent variable.

An additional linear regression was conducted to examine the relationship between executive functioning and adolescent-reported adherence to daily activities, to allow for direct comparisons to the control group on identical measures. Executive functioning was entered as the independent variable and adherence to daily activities was entered as the dependent variable.
Hypothesis 3: Parental role of responsibility will independently predict adherence rates in adolescents with sickle cell disease, with higher rates of parent responsibility predicting more adherence.

A linear regression was conducted to determine whether adolescent-reported role of responsibility is associated with adolescent-reported adherence in adolescents with SCD. Responsibility was entered as the independent variable and adherence was entered as the dependent variable.

An additional linear regression was conducted to examine the relationship between adolescent-reported responsibility and adolescent-reported adherence to daily activities, to allow for direct comparisons to the control group on identical measures. Adolescent-reported responsibility to daily activities was entered as the independent variable and adolescent-reported adherence to daily activities was entered as the dependent variable.

Hypothesis 4: The control group will demonstrate overall higher levels of executive functioning than the group with SCD.

This hypothesis was analyzed by conducting an independent samples t-test with EF as the test variable and classification (SCD, control) as the grouping variable.

Hypothesis 5: The EF-adherence relationship in the control group will be similar to that in the group with SCD, with higher rates of executive functioning predicting more adherence to daily activities.

A linear regression was conducted to determine whether executive functioning is associated with adolescent-reported adherence in the control group. Executive functioning was entered as the independent variable and adherence was entered as the dependent variable.
Hypothesis 6: Parental role of responsibility will independently predict adherence rates in adolescents in the control group, with higher rates of parent responsibility predicting more adherence to daily activities.

As with the SCD group, a linear regression was conducted to determine whether adolescent-reported role of responsibility is associated with adolescent-reported adherence in adolescents in the control group. Responsibility was entered as the independent variable and adherence was entered as the dependent variable.
CHAPTER 3
RESULTS

Preliminary Analyses

There were no missing data on any variables used for any of the participants in the control group. For the group with SCD, one participant (4% of sample) did not complete the Self-Care Inventory, two participants (8%) did not complete neuropsychological testing, and four participants (16%) did not complete the Adherence and Responsibilities Questionnaire. Since these participants were missing entire questionnaires, multiple imputation is not appropriate. The participants with missing questionnaires did not significantly differ on any of the variables from the participants with complete packets. Thus, they were kept in analyses and their existing data was used.

Means of adherence, responsibility, estimated IQ score, and EF in both the SCD and control group are shown in Table 5. Youth with SCD had significantly lower estimated IQ scores than the control group, \( t(46) = -2.07, p < .05 \). For the group with SCD, adolescent-reported adherence and responsibility are significantly different between the SCD recommendations and the activities of daily living. Specifically, these adolescents report more adherence to \( t(19) = 4.22, p < .01 \) and having parents more involved with \( t(19) = -5.23, p < .01 \) SCD recommendations versus activities of daily living. When comparing the adherence and responsibility ratings on activities of daily living between the SCD group and the control group, no significant differences emerged.

Preliminary analyses were run to explore and investigate patterns of executive functioning that may inform the primary hypotheses. Defining clinical impairment as T-scores
Table 5

Descriptives of the Main Study Variables

<table>
<thead>
<tr>
<th>Variable</th>
<th>N</th>
<th>Mean</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Estimated IQ Score - SCD</td>
<td>23</td>
<td>91.96</td>
<td>12.02</td>
</tr>
<tr>
<td>Estimated IQ Score - Control</td>
<td>25</td>
<td>99.68</td>
<td>13.73</td>
</tr>
<tr>
<td>Executive Functioning – SCD</td>
<td>23</td>
<td>38.25</td>
<td>12.51</td>
</tr>
<tr>
<td>Executive Functioning – Control</td>
<td>25</td>
<td>44.41</td>
<td>7.48</td>
</tr>
<tr>
<td>Adherence – SCD Regimen (patient)</td>
<td>24</td>
<td>4.28</td>
<td>.57</td>
</tr>
<tr>
<td>Adherence – SCD Regimen (parent)</td>
<td>20</td>
<td>4.04</td>
<td>.71</td>
</tr>
<tr>
<td>Responsibility – SCD Regimen (patient)</td>
<td>25</td>
<td>2.78</td>
<td>1.21</td>
</tr>
<tr>
<td>Responsibility – SCD Regimen (parent)</td>
<td>20</td>
<td>2.11</td>
<td>.80</td>
</tr>
<tr>
<td>Adherence – Daily Living – SCD</td>
<td>21</td>
<td>3.66</td>
<td>.44</td>
</tr>
<tr>
<td>Adherence – Daily Living – Control</td>
<td>25</td>
<td>3.88</td>
<td>.37</td>
</tr>
<tr>
<td>Responsibility – Daily Living - SCD</td>
<td>21</td>
<td>4.15</td>
<td>.39</td>
</tr>
<tr>
<td>Responsibility – Daily Living - Control</td>
<td>25</td>
<td>4.02</td>
<td>.47</td>
</tr>
</tbody>
</table>

Note. Higher responsibility scores = more child responsibility of 1.5 standard deviations below the mean (T-score ≤ 35; Putzke, Williams, Daniel, Bourge, & Boll, 2000), seven adolescents with SCD and three adolescents in the control group were categorized as impaired. Examining these adolescents compared to adolescents with composite EF scores that were not in the impaired range, impaired adolescents had significantly lower adherence to daily activities ($t(17)=-2.39, p<.05$). There were no significant differences between the groups on responsibility to daily activities, responsibility to SCD tasks, or adherence to SCD tasks. Parent-report was examined to compare results. A trend ($t(16)=-2.04, p=.058$) was found, such that parents of adolescents with SCD in the impaired group reported more parent involvement in completion of the SCD regimen than the group that was not impaired. There were no differences in parent-report of adherence.

Imaging information collected from the electronic medical record for each participant with SCD found that eight participants (32%) had abnormal brain imaging (e.g., visible infarct, hypoxia). However, these participants did not significantly differ on estimated IQ score or
composite EF from participants with normal imaging data, $t(21) = -.51, p > .05$. Additionally, there were no differences in adherence and responsibility data based on results of brain imaging.

**Relationship between Neurocognitive Factors and Adherence in Youth with SCD**

*Hypothesis 1:* Estimated IQ score will independently predict adherence rates in adolescents, with higher estimated IQ scores predicting more adherence.

In a linear regression examining the relationship between estimated IQ score (WRAT-4 reading score) and adolescent-reported adherence to the SCD treatment regimen, higher levels of estimated IQ score did not predict higher levels of adherence, $F(21, 1) = .01, p > .05$.

*Hypothesis 2:* Executive functioning will be a stronger predictor of adherence than estimated IQ score.

In a multiple regression examining whether executive functioning is a stronger predictor of self-reported adherence than estimated IQ score, the two predictors explained only 0.2% of the variance, $F(2, 20) = 0.02, p > .05$. As both predictors accounted for a negligible amount of variance in the regression, neither can be thought of as the stronger predictor of adolescent-reported adherence.

An additional linear regression was conducted to examine the relationship between executive functioning and adolescent-reported adherence to daily activities, to allow for direct comparisons to the control group on identical measures. Higher levels of EF did not predict higher levels of adherence to daily activities, $F(17, 1) = 0.30, p > .05$.

**Relationship between Role of Responsibility and Adherence in Youth with SCD**

*Hypothesis 3:* Parental role of responsibility will independently predict adherence rates in adolescents with sickle cell disease, with higher rates of parent responsibility predicting more adherence.
In a linear regression examining the relationship between adolescent-reported responsibility and adolescent-reported adherence to the SCD treatment regimen, higher levels of parental responsibility did not predict higher levels of adherence, $F(22, 1) = 0.05, p > .05$.

An additional linear regression was conducted to examine the relationship between adolescent-reported responsibility and adolescent-reported adherence to daily activities, to allow for direct comparisons to the control group on identical measures. Higher levels of adolescent-reported responsibility predicted higher levels of adherence to daily activities, $F(18, 1) = 18.33, p < .01$.

**Exploratory Analyses**

Responsibility should be examined as a potential moderator of the EF-adherence relationship in youth with chronic illnesses, such as SCD. However, as tests of moderation are generally underpowered, and due to our small sample size, we are unable to fully investigate this research question. In an exploratory analysis using the PROCESS macro (Hayes, 2012), the overall moderation model for youth with SCD was not significant, $F(1, 19) = 2.82, p > .05$, although the model accounted for 12.83% of the variance in adolescent-reported adherence. In the control group, the overall moderation model was also not significant, $F(1, 21) = 0.43, p > .05$, accounting for only 2.02% of the variance in adolescent-reported adherence.

**Comparisons between Youth with SCD and Healthy Controls on Measures of EF**

*Hypothesis 4: The control group will demonstrate overall higher levels of executive functioning than the group with SCD.*

An independent samples t-test was conducted to compare EF in the SCD and Control conditions. Adolescents in the control group had statistically significantly higher EF scores ($M=44.41, SD=7.48$) than adolescents with SCD ($M=38.25, SD=12.51$), $t(46)=-2.09, p<.05$. 
When examining the components of EF individually, planning/organization (measured by the RCFT) was the most impaired in both groups, with over half of the sample falling below average in this domain (see Table 6).

**Relationship between Executive Functioning and Adherence in Healthy Controls**

*Hypothesis 5: The EF-adherence relationship in the control group will be similar to that in the group with SCD, with higher rates of executive functioning predicting more adherence to daily activities.*

A linear regression examining the relationship between EF and adolescent-reported adherence to daily activities determined that higher levels of EF did not predict higher levels of adherence in healthy controls, $F(23, 1) = 0.03, p > .05$. This finding was similar to that in the group of youth with SCD.

**Relationship between Role of Responsibility and Adherence in Healthy Controls**

*Hypothesis 6: Parental role of responsibility will independently predict adherence rates in adolescents in the control group, with higher rates of parent responsibility predicting more adherence to daily activities.*

In a linear regression examining the relationship between adolescent-reported responsibility and adolescent-reported adherence to daily activities, higher levels of parental responsibility did not predict higher levels of adherence in healthy controls, $F(23, 1) = 0.01, p > .05$. This finding was unlike that in the group with SCD, in which greater adolescent-reported responsibility predicted higher levels of adherence.
Table 6  

*Descriptive of Different Components of EF*

<table>
<thead>
<tr>
<th>EF Test</th>
<th>N</th>
<th>Mean</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>RCFT – SCD</td>
<td>23</td>
<td>31.18</td>
<td>21.52</td>
</tr>
<tr>
<td>RCFT - Control</td>
<td>25</td>
<td>37.80</td>
<td>13.56</td>
</tr>
<tr>
<td>TMT - SCD</td>
<td>23</td>
<td>41.04</td>
<td>11.48</td>
</tr>
<tr>
<td>TMT - Control</td>
<td>25</td>
<td>46.16</td>
<td>9.07</td>
</tr>
<tr>
<td>CWI - SCD</td>
<td>23</td>
<td>42.52</td>
<td>12.47</td>
</tr>
<tr>
<td>CWI - Control</td>
<td>25</td>
<td>49.28</td>
<td>8.44</td>
</tr>
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RCFT = Rey Complex Figure Test-Copy; TMT = DKEFS Trail Making Test #4 (Letter/Number Switching); CWI = DKEFS Color-Word Interference #3 (Inhibition)
CHAPTER 4
DISCUSSION

The results of this study indicate that adolescents with sickle cell disease have poorer executive functioning than their healthy peers, and these findings remain even after accounting for impairments evident on brain imaging. The fact that EF is lower in the adolescents with SCD compared to the group of healthy controls irrespective of comorbid neurologic complications provides support for regular neuropsychological monitoring in this population with a chronic illness. Specifically, this monitoring could detect a change or decline in EF, such as that from cerebral compromise as a result of SCD, which could be helpful in providing simplified treatment plans or increasing parental involvement in execution of the treatment regimen.

Further, the present study suggests a need for increased awareness to additional factors that may be influenced by these deficits in EF, such as adherence and responsibility for treatment regimen completion, which are also affected by the transitional period of adolescence. This study extends previous literature that has described EF deficits in youth with SCD without investigating its relationship to adherence (e.g., Schatz & Buzan, 2006; Schatz et al., 2014; Schatz et al., 2001; Watkins et al., 1998).

The current study differs from studies of adults with chronic illnesses that investigate the EF-adherence relationship because, in taking a developmental perspective, this study elucidates the parent’s role and how it may influence the adolescent’s adherence. Thus, while the
relationship between executive functioning and adherence was not substantiated in the present study, factors such as responsibility may be influencing this relationship. The current study is an initial step in analyzing these relationships and making recommendations for clinical implications and future research.

As executive functioning abilities are critical for successful completion of purposeful tasks, such as treatment regimen completion, and several EF abilities are still underdeveloped during adolescence, the present study sought to further explore EF difficulties faced by adolescents with SCD. This study expands on research that has examined only one specific ability or domain of EF (e.g., working memory in White et al., 2000) and aimed to assess three of the most widely cited domains of executive functioning (planning/organization, cognitive flexibility, inhibition). Planning/organization was most impaired in both groups in the present study. During adolescence, an increase in impulsivity may lead to difficulties with planning, organization, and decision making. These difficulties may be further complicated by cognitive and emotional sequelae of a SCD diagnosis (e.g., anxious or depressed mood). This planning/organization deficit supports the need for parent involvement since planning and organization are critical to adherence. For instance, youth with SCD must organize complex medication regimens and plan regular clinic appointments, and a failure to do so may considerably compromise their health.

Further, as objective measures do not necessarily align with subjective ratings, this study sought to improve our confidence in the EF findings by obtaining the data objectively. The study demonstrated that adolescents in the control group had statistically significantly higher EF scores than adolescents with SCD. Specifically, 30% of our sample of adolescents with SCD show global EF impairment (< 1.5 SD below the mean). This finding is similar to previous literature
that has noted EF deficits in youth with SCD (Berkelhammer et al., 2007; Gold et al., 2008; Schatz et al., 2001; Treadwell et al., 2005). In the control group, only 12% show global impairment. As adolescents in both groups were demographically similar to account for changes due to the developmental trajectory, the differences in EF are likely due to a combination of structural and functional disruptions of the disorder. Side effects of medication may affect some components of EF and these should be discussed with a doctor. Additionally, as depression and anxiety are more common in children with chronic illnesses, these mood concerns may play a role in the disruption of EF. For example, one study found a link between anxiety and EF deficits in youth with chronic pain (Hocking et al., 2011). Future research should examine potential contributors to the EF deficits.

As a large percentage of the youth with SCD had clinically significant impairment in EF, and to examine how these EF deficits could impact adherence to the treatment regimen, the adolescent’s EF was compared to adolescent-reported adherence. These results were examined with consideration of parental involvement in regimen completion during the transitional period of adolescence. Results failed to show a significant relationship between either estimated IQ score or executive functioning and adolescent-reported adherence. However, parents of youth with lower EF reported more involvement in completion of the SCD regimen than parents of youth with higher EF. As the literature suggests, having an adult take responsibility for completion of the regimen can compensate for EF deficits in youth with chronic illnesses, thus possibly eliminating the hypothesized relationship between EF and adherence (Malee et al., 2009; Treadwell et al., 2005). Additionally, parents that are aware of EF deficits may take a more involved role with regimen completion.
The Health Belief Model (HBM) is one theoretical approach that has been used to understand variables related to the likelihood of adhering to a medical regimen. The Health Belief Model’s main premise relates a perception of an illness and the efficacy of its treatment with adherence to prescribed recommendations (Strecher, Champion, & Rosenstock, 1997). From a developmental perspective, parents may identify barriers (e.g., low EF) that would affect the degree to which they believe their adolescent is capable of effectively implementing treatment recommendations (Charach, Volpe, Boydell, & Gearing, 2008). This identification of barriers along with other health beliefs (e.g., the illness is severe, there are benefits of following the recommendations) may increase the parent’s willingness to take responsibility for ensuring regimen completion.

Adolescence is a common time period during which parents transfer responsibility for the completion of daily tasks to their child. The importance of parental responsibility is in line with research that has utilized the Health Belief Model with youth with chronic illnesses. For instance, one study found that the factor of the HBM most closely related to adherence in youth with diabetes was willingness and ability to follow cues to action, including “supportive participation of significant others in self-care” (Bond, Aiken, & Somerville, 1992; Gillibrand & Stevenson, 2006). Bush and Iannotti (1990) developed the Children’s Health Belief Model, in which they emphasized the caretakers’ role in influencing the child’s health. Following the creation of this model, adherence research citing the HBM more often gives attention to systemic factors such as responsibility for regimen (e.g., Logan, Zelikovsky, Labay, & Spergel, 2003). Since parents are more likely than their adolescents to have better understanding of the risks of non-adherence, when they are involved, follow-through of treatment recommendations is expected to be more consistent. Models of adherence in youth should continue to include parent/family factors.
Overall perception of responsibility differed between the youth with SCD and their parents. A lack of consistent findings between parent- and adolescent- report can serve as a reminder that if the adolescent’s EF is low, or just as a result of the developmental trajectory, he/she may not be an accurate reporter, which could lessen the confidence in the self-report data. Obtaining objective measurement along with taking into account parent report may provide more complete information into these relationships. It is likely that parents can more easily intervene with some components of the SCD treatment regimen than others. The current study analyzed each treatment component individually. While parent-report suggested that they took more responsibility for the completion of the SCD regimen than adolescent-report suggested, adolescents’ and parents’ agreed that parents took significantly more responsibility for medication administration and ensuring attendance at clinic appointments versus monitoring the adolescent’s diet and activity restrictions. Knowing which parts of the regimen are monitored less by parents can be beneficial to health care professionals in providing education around how to more easily follow these treatment recommendations.

The present study’s utilization of a measure of adherence and responsibility to daily activities allowed for comparisons on adherence and responsibility to disease-specific recommendations in the group with SCD. That is, this study examined differences between youth with SCD’s perception of their adherence and responsibility to their treatment regimen activities and more general daily activities that are likely completed by most adolescents in their peer group. Adolescent-reported adherence and responsibility were significantly different between the SCD recommendations and the activities of daily living, such that adolescents report more adherence to and having parents more involved with SCD recommendations versus activities of daily living. We can hypothesize that these adolescents view their SCD tasks as more critical to
complete than activities of daily living, and thus they adhere more to these tasks regardless of who is responsible. These results are compatible with the HBM framework relating a person’s belief about the seriousness of a disease and his or her health behavior, as well as with literature showing a significant association between patient’s perceptions of disease severity and adherence. For example, a meta-analysis concluded that nonadherence is more than one-and-a-half times more likely when an individual does not perceive a disease or illness to be severe. Further, the odds of adhering were nearly 2.5 times greater if patients perceived the disease to be severe (Dimatteo, Haskard, & Williams, 2007).

An alternate but equally as probable explanation for the greater adherence rating to the SCD tasks versus the tasks of daily living can be attributed to demand characteristics. Specifically, as we collected data in the Sickle Cell Clinic, the adolescents may have been more likely to report adherence to these tasks. Additionally, adolescents may view many of the activities of daily living as unnecessary (e.g., clean bedroom daily). In the present study, higher levels of adolescent-reported responsibility predicted higher levels of reported adherence to daily activities. Consistent with literature suggesting that adherence difficulties peak in mid-adolescence amidst a struggle with social pressures and egocentric beliefs, these adolescents may be pulling away from parents that are nagging them to perform these chores which they see as futile, resulting in more nonadherence when there is more parental involvement (Shaw, 2001).

A key difference in the present study from the adult EF literature is in the identification of the processes responsible for the deficits in EF. As opposed to adults, youth’s executive functioning may not be fully developed; therefore, we must consider that some deficits may not be due to illness/injury but instead to underdeveloped baseline EF. In order to assess which findings may be due to their medical condition as opposed to the developmental period, the
group of adolescents with SCD was compared to a group of matched controls. Examining the control group on identical measures of EF and adherence, higher levels of EF did not predict higher levels of adherence. This finding was similar to that in the group of youth with SCD. However, only three of the 25 healthy controls were considered impaired (<1.5 SD below the mean), so these findings are likely affected by range restriction. As mentioned for the group of adolescents with SCD, some of the activities described in the questionnaire (e.g., cleaning room) may be seen as unimportant to these adolescents. The HBM describes perceived benefits as one construct that is central to predicting a person’s behavior. If an adolescent identifies the value or usefulness of an activity or behavior, they are more likely to engage in that activity or behavior. Thus, a rating of importance or perceived benefits would clarify these findings by shedding light on a potential reason for poor adherence.

Finally, the present study provided information on adherence and the division of responsibility during the transitional period of adolescence in a group of healthy controls. This information is important as all youth, irrespective of disease status, strive for independence during this time period. In the control group, adolescents reported that they are mostly responsible for taking care of their own daily tasks and activities, and higher levels of parental responsibility did not predict higher levels of adherence. Since there is not an overall need for parental involvement for many of these daily self-care activities for adolescents (e.g., bathing, brushing teeth, dressing), future research should look at what other variables may be affecting adherence. Also, an important line of research could assess how these adherence ratings are related to objective performance in school (i.e., grades). Finally, the questionnaires used in this study were modified to fit the current sample. Future research that expands on this preliminary study could benefit from a better adherence measure for healthy children. This questionnaire is
likely influenced by family environment. For instance, questions about exercise, sport participation, and church attendance will likely not apply for those students whose families do not emphasize or encourage participation in those activities.

**Limitations and Future Directions**

The current study has some additional limitations which must be considered. First, the small sample size, although on par with what is often published in the sickle cell literature (e.g., Schatz et al., 2014; Treadwell et al., 2005; Valrie, Gil, Redding-Lallinger, & Daeschner, 2008), limits the robustness of the findings and makes it difficult to run advanced statistics (i.e., moderation). For example, in contrast to what has been found in previous research, our findings did not differ based on imaging data. However, these data were limited by sample size. Specifically, the majority of the participants with abnormal imaging data in our study had not experienced overt stroke, perhaps restricting the range of observed neurocognitive dysfunction (Schatz & McClellan, 2006). Conversely, other youth may have experienced cerebral events that have not shown up on imaging. These youth may have impaired executive functioning even though their brain imaging has not been coded as abnormal, potentially attenuating the differences between groups. Further, the sample size limits the amount of confounding variables that can be analyzed. While our study examined two potential covariates suggested by previous literature (age, severity), several others (e.g., perceived regimen complexity, mental health factors, family stress and other life stressors) may be involved.

Other methodological limitations include lack of parent report in the control group, which prohibits comparisons between parent and adolescent report on all variables. As adolescents may not always be accurate reporters, taking into account parent report can be helpful in understanding the complete picture. Additionally, the current study used questionnaires that were
created for other chronic illnesses and have not been validated in the sickle cell population. However, they demonstrate good reliability in the present study. We repeated these measures to examine test-retest reliability. The responsibility questionnaire demonstrated test-retest reliability over time. However, we did see change in responses to the adherence questionnaire over time. Due to the nature of these questionnaires, the developmental period of adolescence, and the time between administrations, this finding is not surprising. In fact, we do not expect stability in adherence and responsibility during the time period of adolescence, and the reliability of the responsibility questionnaire could be attributed to transfer of responsibility already having been completed prior to the initial administration of the questionnaire. The usage of EF and adherence ratings from the same time point for analyses was based on these concerns of potential instability in adherence ratings. In future studies, objective measures of adherence should be used when possible to limit overestimation of self-reported adherence. Overall, our sample of youth with SCD and their parents reported greater adherence to the medical regimen than is typically found in studies examining adolescents with SCD. Possible reasons for these high adherence ratings are a clinic culture of accountability (almost all of our families have been coming to the same clinic for many years), greater parent responsibility at younger ages (our mean age = 14, which is considered early adolescence), communication issues between physician and family (the family believes they are adhering, but they do not fully understand some or all parts of the treatment regimen), or the aforementioned overestimation due to demand characteristics or other factors.

Our findings suggest the importance of examining a different, but just as significant, transition period: the period between adolescence and young adulthood. A possible hypothesis for our lack of significant findings between EF and adherence is the over-involvement of parents.
in our sample, an involvement that is likely less during the transition from adolescence to young adulthood (e.g., age 18+). As adolescents transition to adult care, there is less expectation that parents remain involved in treatment management. A systematic review by Jordan, Swerdlow, and Coates (2013) determined that youth with SCD experience an increased mortality rate as they transition out of pediatric clinics into adult care. The review cites the decrease in parent involvement and poor communication/coordination between providers as possible reasons for the health decline during this transition period (Jordan et al., 2013). A second review of youth with various chronic illnesses during the transition from pediatric to adult medical care, found that adherence declines across domains during this transition period, including attendance at medical appointments, medication adherence, and adherence to other illness specific tasks (Pai & Ostendorf, 2011).

Future research should employ larger sample sizes in order to examine if responsibility moderates the EF-adherence relationship. A moderating effect would bolster support for parents remaining involved in implementation of treatment recommendations, especially when an adolescent’s EF is impaired. Additionally, research should aim to explore the connection between parental EF and child adherence. We have already identified research that indicates that youth with EF deficits can still be adherent when parents play a primary role in the execution of the regimen (Treadwell et al., 2005). However, patients with EF deficits whose parents’ EF is also impaired likely would be at a disadvantage when it comes to adherence, as neither parent nor child has the executive capacity to carry out the regimen. Parental executive functioning is critical for understanding the benefits and risks for a given recommendation, understanding how to carry out the recommendation, and making sound decisions regarding the youth’s health care (Schillerstrom et al., 2005).
As our study did not find a link between EF and adherence, future research should consider gathering a better real-world assessment of executive functioning (Malee et al., 2009; Treadwell et al., 2005). Some youth appear more impaired on subjective self-report measures than in performance-based measures of EF (Vriezen & Pigott, 2002). Conversely, there is evidence that some youth perform well on standardized EF instruments but have trouble with EF in daily activities (Vriezen, Pigott, & Pelletier, 2001). Structured assessment likely lessens the demands on executive functioning, and deficits are not as readily visible. In testing situations, the structure and plan provided by the examiner may serve as the youth’s executive control, covering up the true picture of the child’s executive functioning (Gioia, Isquith, Kenworthy, & Barton, 2002). Therefore, neuropsychological testing results from research may not be enough to say that a child or adolescent will be adherent to his or her treatment regimen (Vriezen & Pigott, 2002). Gathering several types of ratings from multiple informants, along with behavioral observations in a real-world setting, might clarify the true picture of the youth’s executive functioning.

**Clinical Implications**

Based on what we know about the EF-adherence relationship in youth with SCD, along with some of the potential influences on this relationship, there are several implications for improving adherence. When implementing any treatment regimen or clinical intervention, health care professionals, parents, teachers, and any other caregivers that may be involved with the pediatric patient should pay specific attention to the youth’s developmental level and related considerations and challenges (Ou, Feldman, & Balkrishnan, 2010). Youth with SCD face challenges beyond those of other illness groups. For instance, research consistently shows that adherence is greater when the child or adolescent perceives his/her illness to be severe. For example, one study showed that patients with asthma were less adherent to their medication.
regimen than oncology patients, who noted more benefit to taking their medication than the patients with asthma (Horne & Weinman, 1999). This research reinforces the importance of continued education on the benefits of adherence to all youth with SCD, especially when they are feeling well and not experiencing pain crises. Further, youth with illnesses such as cystic fibrosis and hemophilia typically have a smoother transition from adolescent to adult care, as they have access to more comprehensive adult care centers. When youth with SCD transition to adult care, they are typically managed by their primary care providers. Therefore, it is imperative that there is good communication between providers during any transition period.

As many youth with SCD exhibit EF deficits, clinicians should begin considering executive functioning when giving treatment recommendations, and some measure of EF may be necessary as a part of the treatment protocol. The present research supports the assertion that parents often play a larger role in regimen completion if their child exhibits EF deficits. However, as adolescents aim for more independence during adolescence, parents may inevitably take less responsibility, and the relationship between EF and adherence may be more apparent. Youth with SCD should receive neuropsychological assessment fairly regularly, especially as cerebral compromise can change the course of EF mid-illness.

Our results demonstrate that youth with SCD may be at greater risk for EF deficits than their healthy peers. As caregivers may not always be present to ensure proper treatment regimen implementation, children and adolescents with deficits in executive functioning may need simplified treatment regimens (Bagner et al., 2007). Further, EF should be considered when discussing the treatment regimen with young patients. Concrete interventions, such as sticky notes and phone alarms, could be helpful for patients with impairments in executive functioning, as these interventions put little demand on the EF system and serve as cues for implementation of
the treatment recommendations (Rosen et al., 2003). Further, health care professionals should educate families on the importance of parental involvement for all parts of the sickle cell regimen, even through adolescence.

Finally, an interdisciplinary treatment team is important in working with these pediatric patients. The present study found that youth report less adherence to diet and activity restrictions than medication recommendations and attendance at appointments. Behavioral health specialists, hematology staff, developmental-behavioral pediatricians, and nutritionists could work together to modify or improve difficult areas of the treatment regimen to increase the chance of successful implementation (Bagner et al., 2007; Vanzutphen et al., 2007). Psychologists could provide additional information on EF, help to identify roles of family members in treatment implementation, and discuss barriers to treatment success.

**Conclusion**

Research should continue to disentangle the complex relationship between executive functioning and adherence in youth with chronic illness such as SCD, paying particular attention to the role of parent- versus adolescent- responsibility for treatment regimen implementation. The majority of neuropsychological deficits are not being assessed in patients with chronic illnesses (Hopkins & Jackson, 2006). The current findings show that youth with SCD have lower EF than their healthy peers, which provides support for regular neuropsychological screening in youth with SCD, to monitor any changes in EF which may require further simplification of the treatment regimen. The findings also suggest that doctors should talk to parents about their health beliefs and their role in ensuring regimen completion. Physicians and other health care professionals can advise how we think of developmental trends in chronic illness. Paying more
attention to factors that influence adherence to illness management during critical developmental periods can be instrumental in improving treatment outcomes.
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APPENDIX A
ADHERENCE AND RESPONSIBILITIES QUESTIONNAIRE

Routines are events that occur regularly: at about the same time, in the same order, or in the same way every time. Please rate how often you engaged in each routine in the last month. Circle a rating ranging from 0 (never) to 4 (nearly always). If an item does not apply to you, please mark “N/A”.

Then, if applicable, check who is responsible for making sure a task is completed: (If you weren’t told to do something, just check N/A.)

<table>
<thead>
<tr>
<th>How often does this occur?</th>
<th>Who is responsible for making sure the task is finished?</th>
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</thead>
<tbody>
<tr>
<td>0 = Never</td>
<td>My parent(s) do it all of the time</td>
</tr>
<tr>
<td>1 = Rarely</td>
<td>My parent(s) do it most of the time</td>
</tr>
<tr>
<td>2 = Sometimes</td>
<td>My parent(s) and I do it equally</td>
</tr>
<tr>
<td>3 = Often</td>
<td>I do it most of the time</td>
</tr>
<tr>
<td>4 = Nearly Always</td>
<td>I do it all of the time</td>
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<tr>
<td>N/A = Not applicable</td>
<td>N/A</td>
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</thead>
<tbody>
<tr>
<td>1. I wake up on time on weekdays.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
<td>O</td>
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<tr>
<td>2. I get dressed on time.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
<td>O</td>
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<tr>
<td>3. I have a set routine for getting ready in the morning (e.g. wash my face, brush my teeth, brush my hair, use deodorant).</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
<td>O</td>
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<tr>
<td>4. I straighten my bedroom daily.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
<td>O</td>
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<tr>
<td>5. I shower, bathe, and/or wash my hands and face daily.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
<td>O</td>
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<tr>
<td>6. I leave for school on time every day</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>N/A</td>
<td>O</td>
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<td>Task</td>
<td>How often does this occur?</td>
<td>Who is responsible for making sure the task is finished?</td>
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<td>7. I eat a snack after school</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>8. I complete homework <strong>on time.</strong></td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<td>9. I study/review for tests</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<td>10. I organize my things for the next day</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>11. I spend time doing fun activities with my family</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>12. I eat dinner at dinner table</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>13. I complete chores regularly (e.g. wash dishes, clean room, mow</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
<td></td>
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<td>the lawn, prepare meals, take care of children)</td>
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<td>14. I talk with my family about my day</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<td>15. I go to bed <strong>on time</strong></td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<td>16. I participate in sports</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>17. I attend sports practices and games</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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<tr>
<td>18. I participate in extracurricular activities (excluding sports)</td>
<td>0 1 2 3 4 N/A</td>
<td>O O O O O O O O</td>
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</table>

**Notes:**
- **on time** indicates the task is completed on time.
- N/A = Not applicable.
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<tr>
<th></th>
<th>19. I attend after school activities (e.g., sporting events, dances, tutoring, Saturday school etc.)</th>
<th>20. I exercise regularly</th>
<th>21. I attend church</th>
<th>22. I ask for permission from a parent or guardian before going somewhere</th>
<th>23. I get told by my parents or guardian what time to be home</th>
<th>24. I remind my parents or guardian before leaving home for school or other activities</th>
<th>25. I talk to my parents or guardian about how/when I will get home from school.</th>
<th>26. I use good manners</th>
<th>27. I have specific and consistent consequences for misbehavior (e.g., remove computer, lose privileges or grounded)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
<td>0 1 2 3 4 N/A</td>
</tr>
</tbody>
</table>
APPENDIX B
DEMOGRAPHIC FORM

Are you the primary caregiver for the patient being seen at Akron Children’s Hospital?
☐ Yes       ☐ No

What is your relationship to the patient?
☐ Mother       ☐ Father       ☐ Stepmother       ☐ Stepfather
☐ Grandmother   ☐ Grandfather   ☐ Legal guardian   ☐ Other: ____________________________

Teen Date of Birth: ___________________  Parent Date of Birth _______________

Who currently resides in the teen’s residence?

<table>
<thead>
<tr>
<th>Relationship to Teen</th>
<th>Age</th>
<th>Relationship to Teen</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td></td>
<td>6.</td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td></td>
<td>7.</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td></td>
<td>8.</td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td></td>
<td>9.</td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td></td>
<td>10.</td>
<td></td>
</tr>
</tbody>
</table>

How would you describe your racial or ethnic background? (please check all that apply)
☐ African American ☐ Asian ☐ Caucasian ☐ Hispanic ☐ Native American ☐ Pacific Islander ☐ Bi-Racial ☐ Other ☐ Prefer not to Respond

How would you describe your teen’s racial or ethnic background? (please check all that apply)
☐ African American ☐ Asian ☐ Caucasian ☐ Hispanic ☐ Native American ☐ Pacific Islander ☐ Bi-Racial ☐ Other ☐ Prefer not to Respond

What is the mother’s (or female guardian’s) highest level of education? (if applicable)
☐ Some High School ☐ High School or GED ☐ Some College or Professional School
☐ Associates or Bachelors Degree ☐ Some Graduate School ☐ Graduate or Advanced Degree

What is the father’s (or male guardian’s) highest level of education? (if applicable)
☐ Some High School ☐ High School or GED ☐ Some College or Professional School
☐ Associates or Bachelors Degree ☐ Some Graduate School ☐ Graduate or Advanced Degree

Are you a single parent? ☐ Yes ☐ No

What type of insurance provider do you have?
What type of insurance provider does your teen have?

☐ Medicaid/Buckeye/Care Source/HMO  ☐ Private Insurance  ☐ Self Pay/No Insurance

Teen’s Grade Level: _____________

Teen’s estimated grade point average for the current grading period: _____________

Teen’s estimated grade point average for the previous year: _____________
APPENDIX C
RESPONSIBILITIES QUESTIONNAIRE

Below are different tasks that your doctor may have asked you to do.
Circle who does each of the tasks: (If you weren’t told to do something, just check N/A.)

<table>
<thead>
<tr>
<th>Task</th>
<th>My parent(s) do it all of the time</th>
<th>My parent(s) do it most of the time</th>
<th>My parent(s) and I do it equally</th>
<th>I do it most of the time</th>
<th>I do it all of the time</th>
<th>N/A</th>
</tr>
</thead>
<tbody>
<tr>
<td>Making sure I follow my diet</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Making sure I don’t do activities that I’m not supposed to do</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Making sure I have enough medicine</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Making sure I take my medication</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Making sure I go to my appointments</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
</tbody>
</table>
APPENDIX D
RESPONSIBILITIES QUESTIONNAIRE – PARENT FORM

Below are different tasks that your doctor may have asked your child to do.
Circle who does each of the tasks: (If your child wasn’t told to do something, just check N/A.)

<table>
<thead>
<tr>
<th>Task</th>
<th>I do it all of the time</th>
<th>I do it most of the time</th>
<th>My child and I do it equally</th>
<th>My child does it most of the time</th>
<th>My child does it all of the time</th>
<th>N/A</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Making sure my child follows his/her diet</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>□</td>
</tr>
<tr>
<td>2. Making sure my child doesn’t do activities he/she is not supposed to do</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>□</td>
</tr>
<tr>
<td>3. Making sure my child has enough medicine</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>□</td>
</tr>
<tr>
<td>4. Making sure my child takes his/her medication</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>□</td>
</tr>
<tr>
<td>5. Making sure my child goes to his/her appointments</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>□</td>
</tr>
</tbody>
</table>

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APPENDIX E

SELF-CARE INVENTORY

Please rate each of the items according to HOW OFTEN YOU FOLLOWED YOUR TREATMENT INSTRUCTIONS in the past week. Use the following scale:

1 = Never
2 = Mostly not, but once in a while
3 = About half of the time
4 = Most of the time
5 = Always
NA = Not told to do it

In the past week, how often have you followed the instructions for:

1. Following your diet
2. Taking medicine (write in each of your meds):
   Medication 1: _____________
   Medication 2: _____________
   Medication 3: _____________
   Medication 4: _____________
3. Going to appointments
4. Not doing activities you’re not supposed to

1 2 3 4 5 NA
APPENDIX F
SELF-CARE INVENTORY – PARENT FORM

HOW OFTEN HAS YOUR CHILD FOLLOWED HIS/HER TREATMENT INSTRUCTIONS in the past week? Use the following scale:

1 = Never
2 = Mostly not, but once in a while
3 = About half of the time
4 = Most of the time
5 = Always
NA = Not told to do it

In the past week, how often has your child followed the instructions for:

1. Following his/her diet 1 2 3 4 5 NA

2. Taking medicine (write in each of his/her meds):
   Medication 1: ___________ 1 2 3 4 5 NA
   Medication 2: ___________ 1 2 3 4 5 NA
   Medication 3: ___________ 1 2 3 4 5 NA
   Medication 4: ___________ 1 2 3 4 5 NA

3. Going to appointments 1 2 3 4 5 NA

4. Not doing activities he/she is not supposed to 1 2 3 4 5 NA

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