Transition Readiness in Adolescents with Blood Disorders: The importance of family functioning and parent-adolescent communication style

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Dedications

This is dedicated all who have provided mentorship along the way, especially Mary T. Lutz.

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Abstract

Transition Readiness in Adolescents with
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Bleeding disorders, including hemophilia, an X-linked recessive disorder of blood coagulation, and von Willebrand disease (VWD), a more common inherited blood disorder, both result in prolonged bleeding and can involve similar treatment components (Kazazian, 1993; Kelley, 1996; Lemanek, Buckloh, Woods, & Butler, 1995; Paper & Kelley, 2002). While a majority of children with blood disorders (90% of children with hemophilia) are expected to live into adulthood, very little research has evaluated the variables thought to predict readiness for transition or successful transition from pediatric to adult health care in young adults with blood disorders (Blum et al., 1993; Gortmaker & Sappenfield, 1984). The purpose of the current study was to evaluate factors proposed by Holmbeck and Shapera (1999), including demographics, parenting-adolescent interaction, and family functioning, that are thought to predict successful adjustment in adolescence. The study also examined differences between adolescents with hemophilia and those with VWD, explored how factors predict adjustment during adolescent transition to adult health care and assessed factors thought to predict readiness for transition. Results from the study suggest few differences between adolescents with VWD and those with hemophilia and partial support for the Holmbeck and Shapera (1999) model. Demographic variables were found to play a significant role in predicting transition outcome. Results should be interpreted

cautiously given the small sample size of the study and low power. Findings suggest that male adolescents and those from a low socioeconomic background may be more likely to experience poorer family functioning and increased maladjustment. Results provided support for a multi-tiered intervention program that provides minimal services to well-functioning families surrounding transition to adult care, but provides more intensive services to families that have multiple risk factors.

CHAPTER 1: INTRODUCTION

Current research has attempted to determine the importance of programs in facilitating the transition process and increasing adolescent and young adult adjustment after transition from pediatric to adult health care. The proposed study evaluated variables thought to be important in adolescent adjustment (Holmbeck & Shapera, 1999), and applied this framework to understand adjustment in adolescents with blood disorders, hemophilia and VWD, and to predict readiness for transition to adult clinics, in order to provide additional information to treatment providers regarding how to assess readiness for transition and how to structure transition programs in their pediatric care settings. To gain a better understanding of hemophilia and VWD, a review of the literature detailing the etiology, clinical manifestations, and treatments of blood disorders will be discussed. Given that few studies assessing the psychosocial implications of VWD on children, adolescents, and their families have been published, the literature reviewed for this project will focus mainly on hemophilia. Information regarding the psychosocial impact of chronic illness and hemophilia on child and adolescent adjustment will be included. A detailed explanation of the theoretical models of adolescent development and the transition process will be highlighted. A discussion of the effects that family functioning and parenting practices have on psychological adjustment will complete the literature review. Given the limited amount of information available on family functioning and parenting in hemophilia or VWD, research that evaluated how these variables impact adolescent adjustment to other types of chronic illnesses will be included.

CHAPTER 2: LITERATURE REVIEW

Incidence and Etiology of Hemophilia

Hemophilia, an X-linked recessive disorder of blood coagulation, causes a deficiency in factor clotting and affects between 1 in 5,000 newborn males within the general population (Soucie, Evatt, & Jackson, 1998). There are on average 18,000 individuals diagnosed with hemophilia within the United States and 2 million worldwide (Soucie et al., 1998). Children with hemophilia are classified into different subtypes based on the type of factor deficiency they experience. Factor VIII deficiency, known as hemophilia A or classic hemophilia, is the most common deficiency found in 80% of all cases, followed by factor IX deficiency, called hemophilia B or Christmas disease (Lemanek, Buckloh, Woods, & Butler, 1995; Merck, 2003). Kasper and Dietrich (1985) indicated that hemophilia A is four times more common than hemophilia B. The disorder is not prevalent in any given ethnic, religious, or racial groups, although it has been classified in the past as the "royal disorder" due to the number of royal families in Europe affected by hemophilia during the late 1800's and early 1900's.

Both factor VIII and factor IX are X-linked disorders (Thompson, Gustafson, & Ware, 1998). In the case of hemophilia males pass down hemophilia to their daughters, who then carry the disease. It is expected that a father with hemophilia will bare sons unaffected by the disorder and daughters who carry the gene for hemophilia. Those daughters, who are carriers of the gene, will pass the gene onto their daughters, who

will also be carriers, and half of their sons will have hemophilia (Bussing & Johnson, 1992). For families with a genetic link to hemophilia, the disease may affect more than one child. Hemophilia can also be caused by genetic mutation, and is present at birth (Handford, Mayes, Bixler, & Mattison, 1986). Around 25% of all children diagnosed with hemophilia have no family history of the disorder, thus have hemophilia as a result of genetic mutation (Lemanek et al., 1995).

Individuals with hemophilia experience low levels of factor VIII or IX in the blood, which results in delayed clotting following injury or spontaneous bleed. Typically bleeding in the body is regulated by constriction of the blood vessels, activity within the platelets, and blood clotting factors. Blood clotting factors involve proteins within the plasma of the blood that aid the body in decreasing bleeding and minimizing the time it takes for bleeding to cease (Merck, 2003). Decreased levels of clotting factors in the plasma, as is seen in hemophilia, cause delayed clotting time and spontaneous bleeds if repeated bleeding occurs in a given joint or organ (Lemanek et al., 1995).

After birth hemophilia can be detected by measuring the level of factor VIII in the contained umbilical cord blood. Typically, factor IX deficiency is more difficult to diagnose, as newborns can have low levels of factor IX at birth, but not meet criteria for a diagnosis of hemophilia. Individuals diagnosed with factor IX deficiency can have more severe medical complications than those with factor VIII. In addition factor VIII and IX deficiency can be measured by a prolonged partial thromboplastin time, which indicates the time that it takes to convert substances with procoagulatant properties (Lemanek et al., 1995). Some have suggested that prolonged bleeding during

circumcision or bleeding in the mouth during infancy are early symptoms of the disorder (Montogomery & Scott, 1992). Individuals with mild hemophilia may go undiagnosed, although they may experience more excessive bleeding after surgery or major injury than expected (Merck, 2003). Typically hemophilia is noted by 18 months of age, as many infants with hemophilia have experienced their first bleeding episode by age one year (Merck, 2003). Individuals suffering from rare hemophilia disease mutations often are difficult to diagnose (Kazazian, 1993). Currently, there is no method to clearly detect females who carry the gene (Kazazian, 1993).

While a majority (90%) of children with hemophilia are now expected to live into adulthood, very little research has evaluated the impact that hemophilia/HIV- (Human Immunodeficiency Virus Negative) has on adolescent and young adult development. Concerns in the past have surrounded exposure to viruses transmitted by a tainted blood supply, given that most individuals with hemophilia relied on human blood in the treatment of the disorder. Prior to 1985, blood was not screened for disease including human immunodeficiency virus (HIV) and hepatitis B and C. A majority (70% in severe cases and 20% in mild hemophilia cases) of those individuals infused with blood before 1985, contracted HIV (Centers for Disease Control, 1987; National Hemophilia Foundation, 1991). The HIV crisis that drastically altered the field of hemophilia research during the 1980's and 90's is now beginning to change, as many adolescents with hemophilia have not contracted HIV (Blum et al., 1993; Gortmaker & Sappenfield, 1984). Therefore, focusing on how adolescents with hemophilia/HIV- adjust to the disease is a relatively new research area. Given the medical nature of this blood disorder and changes in the area of hemophilia treatment, it is important to

discuss clinical manifestations associated with the disorder, as well as treatment options for hemophilia.

Clinical Manifestations and Treatment of Hemophilia

There are a number of clinical manifestations of hemophilia, with a majority of individuals diagnosed with the disorder experiencing bleeding in their joints and organs. The frequency and intensity of bleeding episodes varies depending on severity of the disease and injury. The bleeding that individuals with hemophilia experience is frequent, specific to organs and joints, and is precipitated spontaneously or by trauma. Decreased clotting factor levels within the blood can prolong the time it takes for the bleeding to stop in a child with hemophilia (Montogomery & Scott, 1992). The clotting of superficial cuts or scrapes is often not a concern, as the systems to stop this type of bleeding remain intact (Bussing & Johnson, 1992). Although individuals may appear to be physically "normal", many experience intense pain episodes and joint and muscle degeneration because of the abnormal bleeding, which can lead to chronic orthopedic disability (Brown & DeMaio, 1992; Handford et al., 1986). Not only do chronic disabilities associated with hemophilia affect the quality of life in individuals with the disease, but restrictions on level and type of activity, pain experienced with the disease, and medical treatment can impact quality of life.

Differing levels of severity impact the frequency of bleeds, type of medical treatment, and pain associated with the disease. Hemophilia has been conceptualized into three classifications including mild (5-30% activity), moderate (2-5% activity), and

severe (< 2% activity), based on the factor activity in the plasma. Many patients represent the severe disease group and level of disease severity is determined by the amount of bleeding that an individual experiences (Kazazian, 1993; Kessler, 1991). Individuals with severe hemophilia most likely will experience significantly more spontaneous bleeding episodes in the joints without any trauma or injury, while those with moderate disease severity will show abnormal bleeding after trauma with little to no spontaneous bleeding episodes. Lemanek and colleagues (1995) note that if there is re-injury to the same area in children with hemophilia, a spontaneous bleed in the affected area may occur. Those with mild hemophilia experience bleeding only in the presence of severe trauma or after surgery (Montgomery & Scott, 1992).

Young infants and children can experience abnormal bleeding in the mouth that accompanies teething or trauma regardless of severity level. Bleeding from hematomas that result from minor injuries can also occur. As children become more active, bleeding episodes within individual's organs and joints increase. Trauma to vital organs including the head, neck, and stomach can be fatal and require emergency care (Lemanek et al., 1995). In addition spontaneous bleeds in children and adolescents with hemophilia have been linked to psychological factors including stress and anxiety (Baird & Wassen, 1985; Lemanek et al., 1995). Other symptoms including acute pain, swelling, and restricted movement, often caused by bleeding, can be present in children with hemophilia.

Hemophilia and Pain

Children with hemophilia experience different kinds of pain, including reoccurring acute pain, caused by bleeding episodes, and chronic pain, resulting from repeated internal bleeding in the joints (musculoskeletal). Often times utilization of factor replacement products can stop acute bleeds, thus decreasing pain, however chronic pain can become debilitating and significantly affect quality of life (Lemanek, et al., 1995). Pain that accompanies severe bleeding episodes and restricts child activity level is thought to contribute to school absenteeism in this population. Results of a recent study indicated that on average children missed 27 days of school a year, which constituted 15% of the total school year (Colegrove & Huntzinger, 1994). Given that a majority of adolescents and adults with hemophilia experience hemophilic arthropathy, chronic pain management is a concern for treatment providers (Dietrich, 1986; Varni, Walco, & Katz, 1989). Recent movements within the field have focused on decreasing school absenteeism, pain, and bleeding episodes by increasing families' abilities to treat medical complications associated with hemophilia at home.

Medical Treatment

The home care movement has become the standard practice for treating hemophilia today, as health care providers and families coordinate treatment and collaborate in the care of children with the disorder. Hospitals typically utilize a comprehensive care model to provide services to these families, so that a multidisciplinary team can help families treat bleeding episodes, provide support and education, and manage disease-related complications (Lemanek et al., 1995). Although

the hospital and clinic staff is involved in the medical care of these children, families are the primary healthcare providers as they administer routine medical care and manage disease complications (Lemanek et al., 1995).

Beginning in the early 1970's coagulant factor concentrates revolutionalized the treatment of hemophilia, as the treatment allowed children to use coagulant factor through intravenous infusion of human plasma (Brown & DeMaio, 1992). This procedure replaces the deficient clotting factor within the child's blood with lyophilized coagulation factor concentrates. The medical advance allowed families to begin treating their children in the home, rather than relying on hospital-based services, thus reducing dependency on medical professionals and increasing reintegration into family and community life (Brown & DeMaio, 1992). The application of behavioral techniques to teach families about home care practices, have been found to increase parent proficiency in administering treatment and improve long-term adherence (Gilbert & Varni, 1988; Sergis-Deavenport & Varni, 1983).

Although the positive advantages of receiving care in the home environment are numerous for children with hemophilia, the strain that home care places on families is important to discuss. Given that few children with hemophilia rely on medical centers to provide routine care, parents and children must intravenously infuse routine factor replacement products at home (Brown & DeMaio, 1992). Although professionals advise patients on the amount of factor replacement to use, based on their disease severity, there is variation in the dosage of factor replacement used in children with comparable levels of disease severity (Handford, Charney, Ackerman, Eyster, & Bixler, 1980). Typically children and adolescents with milder disease may use replacement

products to treat a bleed that occurs or engage in short-term prophylaxis, while others with more severe disease may use factor products based on a long-term prophylaxis model. Prophylaxis treatment can occur on a daily or weekly basis (e.g., long-term prophylaxis) or may be used prior to engaging in a higher-risk activity (e.g., sport games, surgery) that may result in a bleed (e.g., short-term prophylaxis) (Berntorp et al., 2003). It has been thought that prophylaxis treatment has resulted in better health related quality of life, as well as decreased arthropathy and bleeding episodes (Berntorp et al., 2003; Bullinger and von Mackensen, 2003; Fischer, Van Der Bom, & Van Den Berg, 2003), although these procedures can be distressing to children and families as they rely on intravenous infusion of product.

In addition to the administration of factor replacement products and medications, families and children with hemophilia have other responsibilities associated with home care. Children and adolescents with hemophilia must avoid surgery and drugs that may aggravate bleeding (Merck, 2003). Families must also utilize strategies that minimize risk for injury and bleeds, by restricting the activity level of the affected child and helping the child avoid situations that may cause injury. Thus children and adolescents with hemophilia are unable to play contact sports and engage in other activities that are potentially risky. For parents of adolescents, negotiating developmentally appropriate activities while adhering to medical restrictions can often be challenging.

Management of pain is also a critical goal in treatment, thus families may need to provide psychological support, as well as medical interventions, to decrease both chronic and acute pain episodes. Understanding the difference between chronic and

acute pain, the treatments that accompany both types of pain, and child communication about intensity of pain is essential. Some research has indicated that psychological interventions, including hypnosis, were effective in decreasing pain experienced, but not in reducing bleeding episodes (LeBaron & Zeltzer, 1985). In addition to hypnosis, cognitive-behavioral interventions including muscle relaxation, breathing exercises and guided imagery have been helpful in reducing chronic pain experienced by children with hemophilia (Varni, 1981; Varni, Gilbert, & Dietrich, 1981).

Not only must families be aware of and monitor child activities and pain responses, but they also must be educated about early signs of disease complications. Since routine care is not provided in a hospital or clinic setting, early professional treatment for bleeds and other disease complications must be sought. Severe disease complications that demand medical intervention, above that received in the home environment can be disruptive to the family and child's daily life.

Finally, a critical piece of home care is constant monitoring of treatment provided. Many families providing home care to a child or adolescent with hemophilia are required by medical staff to keep journals detailing activity level, treatment administered, pain episodes, and disease complications. Journals are often a way for medical professionals to assure that a family is adhering to medical protocol and to remain knowledgeable about the child's disease progression. Of note is that compliance with medical regimens has been documented by some as being poor in children and adolescents with hemophilia. In one study, almost 75% of children and adults sampled were found to have problems with adherence to medical regimen (Weiss et al., 1991). Home care of children and adolescents is complicated, and requires dedication and

organization within the family. Family members must adhere to medical regimens that often interfere with daily activities and be on alert for signs of medical complications, while continuing to juggle multiple family demands. The stress of being a parent or caregiver to a child or adolescent with this potentially fatal disease can be overwhelming for some families and may lead to increased anxiety or distress (Lemanek et al., 1995).

Individuals with hemophilia have been reported to experience high levels of anxiety and stress, as they live with the constant concern of injury or spontaneous bleeding activity. While there is limited research detailing nonmedical interventions utilized with this population, LaBaw (1992) explored the use of hypnosis as a way to decrease levels of anxiety in children with hemophilia. Children with hemophilia included in the study were required to meet weekly over the course of three months. During these sessions children talked about the implications of having hemophilia, were taught relaxation strategies, and were inducted into two trances. Later, education regarding hypnosis taught the children ways to self-induce trances at home. LaBaw asserted, through case studies, that hypnosis was an effective strategy for children suffering from hemophilia in reducing anxiety and spontaneous bleeding episodes. Another report indicated that use of hypnosis with children and adolescents with hemophilia decreased their need for blood transfusions by 25 to 50 percent (Swirsky-Sacchetti & Margolis, 1986). Of note is that there was not a control group for this study.

Children with hemophilia often require extensive medical procedures, including venipuncture, and are restricted from participating in high-risk activities like contact

sports (Nichols et al., 2000). In addition, concerns regarding their psychological adjustment to the illness (e.g., anxiety, stress) have been noted (Lemanek et al., 1995). Many times medical providers are acutely aware of the challenges that families and children with hemophilia face in daily management of the disease. Drotar and colleagues (1997) surveyed medical providers of psychosocial services at pediatric hemophilia treatment centers to determine the types of problems encountered by children with hemophilia and their families. A total of 71 providers across 53 treatment centers responded, including social workers, nurses, and psychologists. The surveys were designed to measure structure of psychological services provided, presenting psychosocial problems, allocation of time, and obstacles to access services. Child and parent adjustment to hemophilia was the problem with the highest reported frequency, followed by financial problems related to hemophilia, maternal adjustment to HIV, child noncompliance with treatment for hemophilia, child adjustment to HIV, and parental noncompliance with treatment for hemophilia, respectively. In general providers rated problems with behavioral adjustment to be the most frequent (M = 3.1, SD = 2.4, range 0-5) and academic problems to be the second most common. Overall family relationship problems, parent adjustment problems, and peer relationship issues were seen relatively frequently by providers. Providers also indicated that around one fifth of the time they spent with families was consumed with issues surrounding problematic parent and child adjustment to hemophilia. Many times these psychosocial interventions involved both the child and mother, while some involved the mother alone, the family, or the parent dyad. Interestingly professionals sampled for this study reported that a large proportion of their pediatric clinic were in need of psychological

services (45%), but did not receive them. A majority of interventions provided by these medical professionals included giving informational or educational support to families (Drotar, Agle, Eckl, & Thompson, 1997). In addition to discussing the etiology, clinical presentation and treatment of hemophilia, it is essential to review VWD, a disease that has not been studied extensively in the literature.

Etiology, Clinical Presentation and Treatment of VWD

Similar to hemophilia, VWD is an inherited blood disorder resulting in prolonged bleeding, however VWD affects a much larger proportion of the population (1-2%) and both females and males can be diagnosed with the disease (Paper & Kelley, 2002). Although VWD is the most common bleeding disorder, many individuals with the disease go undiagnosed, especially those with milder forms. Individuals with VWD are classified into three different types. Type 1 VWD, the most prevalent type, affects between 70-80% of individuals with VWD, is typically associated with patients who have low levels of VWF and is a milder form of the disease. In comparison Type 2 VWD is found in 15-30% of individuals with VWD and is classified as a lack of functioning VWF in the blood. Individuals with Type 2 VWD have enough VWF in their blood, but the protein does not function properly. There are four subtypes of Type 2 VWD, with Type 2N being the most similar to hemophilia A as joint and muscle bleeding may be apparent in these patients (White & Montgomery, 2000). Finally Type 3 VWD is the rarest and most severe form of the disease. Individuals with Type 3 VWD produce no VWF and often show decreased levels of factor VIII in their blood. Clinical

symptoms between hemophilia A and Type 3 VWD are similar; where as other types of VWD are less severe than hemophilia (Paper & Kelley, 2002; White & Montgomery, 2000).

Discovered by a Finnish physician in 1926, VWD is a disease that results from a lack of VWF protein or dysfunctional VWF in the blood. A lack of functioning VWF in the blood causes platelets to not adhere to an injured blood vessel wall or other platelets, resulting in prolonged bleeding. VWF also serves to stabilize and transport factor VIII, a factor associated with hemophilia A. Thus some individuals with low levels of factor VIII may appear, on blood test, to have similar results, as those with hemophilia A. Unlike hemophilia, which is an X-linked disorder, VWD is inherited by a deficit on autosome chromosome 12. Compared to X-linked disorders that affect primarily males, autosomal disorders affect males and females equally. In families where one or both parents pass down a defective VWF gene, the child may be affected (Paper & Kelley, 2002). Infants are not typically tested at birth for VWD unless doctors are aware of family history. Typically VWD is not diagnosed until later in life, and in many cases individuals live with VWD without knowing their diagnosis or receiving medical treatment (Paper & Kelley, 2002).

VWD can be diagnosed through a complex series of medical tests, and many advocate that specialized care through a pediatric hematologist be obtained as very few general practitioners are educated about symptoms associated with VWD or have the proper laboratory equipment to diagnose the disorder (Paper & Kelley, 2002). Given the numerous subtypes of the disorder, and various presentations, including decreased VWF and factor VIII, diagnosis and clinical presentation can be complicated.

Although some subtypes of VWD have similar clinical presentations to hemophilia, symptoms associated with VWD typically include bruising, frequent and intense nose bleeds, heavy and prolonged menstruation, and prolonged bleeding following surgery. As noted above those with Type 2 N or 3 VWD may also experience bleeding into joints and muscles. In individuals with VWD bruising can occur without injury, and nosebleeds may be caused by injury or changes in climate (i.e., moving to a drier climate). It is not that VWD causes the bleeds to occur, but instead that the disease results in prolonged bleeding from injury or agitation. While these are the most common symptoms of VWD, some patients experience more serious problems including gastrointestinal bleeding, rectal bleeding, and bleeds into joints/muscles. Patients with VWD often do not experience spontaneous bleeding. Similar to hemophilia, surgery and trauma can be problematic for individuals with VWD, as they will experience difficulty controlling bleeding following these events.

While not as prevalent, those with more severe forms of VWD may show similar problems with pain to those with hemophilia, and women with heavy menstrual bleeding may also indicate more discomfort. For those experiencing abdominal or joint bleeding associated with VWD, pain management can also be very important (Paper & Kelley, 2002). Since disease severity is often less, compared to individuals with hemophilia, those with VWD may experience infrequent contact with medical providers and daily health management may be minimal (Paper & Kelley, 2002).

Medical treatments for VWD are typically not as complex as they are with hemophilia, however this can vary depending on the severity of symptoms associated with VWD. Individuals with VWD are usually educated about how to prevent bleeds

from occurring, how to stop bleeding, and how to cope with pain that accompanies bleeding episodes. Those with VWD are encouraged to keep their nostrils moist to prevent nosebleeds and to engage in preventative dental hygiene to lessen problems with bleeding during dental work. Once a bleed begins, VWD patients are sometimes instructed to use local measures (i.e., direct pressure, ice, topical agents, nasal packing, topical hemostatic agents) to control bleeds (Paper & Kelley, 2002). When these less invasive measures are ineffective, medication to rapidly increase the rate at which VWF and factor VIII are being released into the blood can be prescribed (desmopressin acetate). Desmopressin can be administered through nasal spray or by injection into the vein or under the skin. This treatment option is typically used for individuals with Type 1 VWD, as those with Type 2 and Type 3 VWD do not benefit to the same degree from the medication. For women who experience menorrhagia, heavy menstrual bleeding, a combination of estrogen and progesterone in the form of oral contraceptives can be helpful in decreasing symptoms. Other medications like antifibrinolytics can help to prevent reoccurring bleeds. Patients, who continue to exhibit symptoms that cannot be controlled by medication like desmopressin, may require more aggressive measures including infusion of factor concentrate (Paper & Kelley, 2002). Similar to individuals with hemophilia who self-infuse, adolescents and adults with VWD who require factor concentrate are encouraged by medical staff to learn how to self-administer the factor products. Keeping a record of treatment is also standard practice for VWD patients who self-infuse factor concentrate products. Additional treatment options include exercise like physical training and aerobic exercise, which have been found to increase VWF supply in the blood (Paper & Kelley, 2002). Although risky sports that involve

the potential for serious injury are not advisable, guidelines for sport activities are typically less restrictive for VWD patients than for hemophilia patients. While there have not been any studies conducted to evaluate if nonpharmacologic interventions are helpful in decreasing pain in patients with VWD, health care practitioners advocate the use of relaxation, deep breathing, and guided imagery with this population (Paper & Kelley, 2002).

Given the clinical nature of hemophilia and VWD, and the fact that both diseases have a chronic course, concerns over how families and children with blood disorders adjust to psychological demands of the illness have been raised. While there are very few published studies evaluating the psychosocial affects of VWD in children, some recent literature focusing on health-related quality of life in an adult sample suggested that women with VWD endorsed similar levels of quality of life as males with severe hemophilia and who were HIV+ (Barr et al., 2003). This study suggests the importance of evaluating similar constructs and adjustment in a younger sample. Due to the lack of published material regarding the psychosocial implications of VWD, the remainder of the literature review will focus on research that has been conducted with children and adolescents with hemophilia.

Psychosocial Adjustment to Hemophilia

Adjustment is a measure of how an individual adapts psychologically, socially and physiologically to a chronic illness (Grey, Cameron, & Thurber, 1991). Often variables like disease outcome, self-esteem, social competency, and quality of life are

used to reflect child and family adjustment to a chronic condition. Pless and Roghmann (1971) reported that while children with a chronic illness are at a higher risk to develop psychiatric disorders compared to the general population, only a minority show psychiatric disturbances. In addition, studies have indicated that high levels of stress and anxiety, along with lifestyle change, that often accompany chronic illness, and hemophilia specifically, can place families at increased risk for problems with adjustment (Wallander, Varni, Babani, Banis, & Wilcox, 1989). Within the hemophilia literature, children and adolescents' personality traits, behavior problems, and intellectual abilities have been assessed to determine the impact that hemophilia has on psychological adjustment.

Personality Traits

Throughout the early literature evaluating the impact of hemophilia on children and families, many studies focused on personality development in the child with hemophilia and personality differences between children with the disorder and non-ill peers. A review of the literature concerning personality changes, found few significant differences between personality constructs in children with hemophilia and matched samples, however children with more severe disease and greater physical handicap experienced more depression, less extroversion, less sociability, and higher scores on a scale of neuroticism (Bussing & Johnson, 1992; Olch, 1971).

In an early study, Olch (1971) administered the Rorschach and Draw-a-Person tasks to a group of children with hemophilia and controls. Results suggested that there was not a "hemophiliac" personality pattern that distinguished the children with

hemophilia from those without the disease. Handford and colleagues (1986) also looked at personality traits in boys with hemophilia using a personality questionnaire with Early School (ESPQ), Childhood (CPQ) and High School (HSPQ) forms. The children (age 5 to 19 years) sampled were receiving comprehensive medical and psychological services free of charge from a funded hemophilia project at the time of the study. In order to compare differences on measures of personality to a control group, and among children with different levels of disease severity, children were categorized into mild-moderate and severe levels of hemophilia. Compared to control participants, the children with hemophilia were found to have scores indicating that they were significantly brighter, more stable, and more secure, based on trait patterns produced by the personality measure. In comparing across the disease severity levels, individuals classified as having severe hemophilia, were found to be more submissive, serious, and high on a scale of self-control than the mild-moderate group who rated high on scales assessing assertiveness, enthusiasm, and low-self control. The authors concluded that the children with more severe disease evidenced these personality traits because they were more likely to sustain serious injury and suffer from greater medical complications than those with a less severe disease pattern, thus making them more cautious and reserved (Handford et al., 1986). Handford and colleagues (1986) also found that personality traits changed with time, as older children with hemophilia reported higher levels of enthusiasm, assertiveness, and sense of adventure than younger children with the disorder. Overall results indicated that disease severity may have an impact on personality traits endorsed by children with hemophilia, although no consistent “personality” pattern was determined (Handford et al., 1986; Olch, 1971).

Studies focusing on personality patterns of children with hemophilia have become scarce. Early studies that evaluated differences in personality in children with the disease, found that severity of disease and age of the child impacted their scores on scales assessing assertiveness, submission, self-control, and enthusiasm. Recent literature focuses more on objective measurements of adjustment and typically includes appropriate normative samples.

Behavior Problems

Various studies evaluating behavior problems in children with hemophilia have used objective measures and a variety of control groups including sibling, age-matched peers, and other illness groups. In an early study examining adjustment with objective rating scales, Klein and Nimorwicz (1982) completed a pilot study assessing the relationship between knowledge of disease and psychological distress in a sample of mothers, fathers, children, and siblings (N = 12 families) with hemophilia. Child participants and siblings ranged in age from 8 to 23 years and a majority of the children sampled came from intact, two-parent families. Mother, father, and child (age 15 years and above) distress was rated using the Symptom Checklist-90 Revised, which assesses psychiatric problems in a variety of populations (SCL-90; Derogatis, 1977), the Beck Depression Inventory (BDI; Beck, 1972), and the Self-Evaluation Questionnaire (STAI; Spielberger, Gorsuch, & Lushene, 1970). Children, younger than age 15 years, were administered the Children's Depression Inventory (CDI; Kovacs, 1979) and the Revised What I Think and Feel questionnaire (RCMAS; Reynolds & Richmond, 1978). A 58-item measure assessing hemophilia knowledge was devised to measure general

understanding of disease processes and treatment of the disorder. Klein and Nimorwicz (1982) found that mothers sampled reported a higher level of psychological distress on the SCL-90 compared to other family members, and in comparison to the general population, as 7 out of the 12 mothers had scores in the clinically significant range. Fathers who completed the measures showed scores in the average range, although their scores on a measure of somatozation were in the lower end of the significant range. Children with hemophilia and their siblings, who were older than age 15 years ($N = 11$), were found to have scores on the SCL-90 that were in the middle of parent-reported levels of distress. Thus they appeared to be experiencing less distress than mothers reported, but more distress than fathers reported. BDI and STAI anxiety scores were within the normal range for a majority of the older children sampled, although 3 out of 11 families did have mild elevations on the depression measure. The younger subset of children and siblings ($N = 7$) did not indicate any significant levels of distress on the CDI or the RAMAS compared to a normative sample. Siblings also did not show signs of distress on these measures.

A more recent study evaluated the social adjustment of children with chronic blood disorders and their siblings (Clemente et al., 2003). This multicenter study evaluated siblings and children with either hemophilia or β–thalassaemia in 115 families using the Social Adjustment Scale, an unpublished questionnaire developed by the authors to assess adjustment across various areas including school, social leisure activities, family relationships and overall adjustment. Children with hemophilia were found to evidence higher levels of social adjustment than children with β-thalassaemia and similar levels of social adjustment compared to unaffected siblings in the areas of

school and family, although all three groups scored lower on the social adjustment measure than a normative sample. The authors noted that all children in the study reported a number of difficulties in the area of leisure activities, with 43% of children with hemophilia and 43% of siblings reporting social dysfunction on this subscale. Results also suggested that older children (>11 years) with chronic illness reported significantly more social adjustment difficulties than younger children in the area of school functioning. The authors indicated that while children with hemophilia appear to evidence similar levels of social adjustment compared to controls, more attention to children as they reach adolescence is important. In addition they noted the high rates of social dysfunction in the sample, both of children with chronic illness and their siblings, suggesting that a child with a chronic illness may impact both their own, as well as their siblings, level of adjustment in socially based leisure activities (Clemente et al., 2003).

In a study assessing symptoms of anxiety and interfamilial stress, Bussing and Burket (1993) evaluated a group of children with hemophilia and age-matched controls. Both parents and children in the study were administered the interview version of the Schedule for Affective Disorders and Schizophrenia for School-Age Children (KSADS; Puig-Antich & Ryan, 1986) and the Index of Family Relations scales. A total of 23 children with hemophilia participated in the study [6 children were HIV+, along with samples of children with asthma (N=37) and healthy controls (N = 31)]. There were no significant differences found between the hemophilia/HIV- group and healthy controls on measures of psychological adjustment. Compared to the asthma group, the hemophilia/HIV- group had significantly less psychopathology, specifically anxiety

diagnoses, and reported experiencing lower rates of family stress (Bussing & Burket, 1993).

Further studies comparing children with hemophilia (HIV-) and a matched peer sample indicated that children with hemophilia do experience some difficulties with internalizing problems Trzepacz and colleagues evaluated a sample 40 boys with hemophilia and 40 matched classmates (Trzepacz, Vannatta, Davies, Stehbens, & Noll, 2003). In phase one of the study researchers visited the school class of each of the children with hemophilia and administered questionnaires evaluating social reputation and acceptance. In addition the researchers had teachers and students nominate children in the class for 30 roles in the Revised Class Play (RCP) and they completed Best Friend ratings (RCP; Masten, Morison, & Pellegrini, 1985; Best Friends; Bukowski & Hoza, 1989). Following phase one, a classmate for each participant with hemophilia was identified as a comparison peer. Children were matched on race, gender, and date of birth. In phase two of the study all participants and their families were administered the Children's Depression Inventory (CDI; Kovacs, 1992), Roberts Apperception Test for Children (RATC; McArthur & Roberts, 1982), the Loneliness and Social Dissatisfaction Questionnaire (Asher, Hymel, & Renshaw, 1984), the Self-Perception Profile for Children (Harter, 1985), the Child Behavior Checklist (CBCL; Achenbach, 1991), and the Vocabulary and Block Design subtests from the Wechsler Intelligence Scale for Children-Revised (WISC-R; Silverstein, 1975) (Trzepacz et al., 2003).

Results indicated no significant differences between children with hemophilia and their peers on demographic, socio-economic, or intellectual variables. In addition no differences were seen on the measures of social reputation (RCP), social acceptance

(Best Friends), or social competence (CBCL) using parent, teacher, peer, and self ratings. Children with hemophilia were found to have significantly higher scores on a measure of childhood depression (CDI) and reported lower scores on measures of scholastic competence, athletic competence and global self-worth compared to their peers. Mothers indicated higher levels of somatic complaints and internalizing problems, while fathers reported higher scores on measures of anxiety/depression in children with hemophilia. Both parent and child reported scores on depression, competence, and anxiety/internalizing behavior measures remained within the average range. The authors noted significant differences between children with varying levels of disease severity, as children with more severe disease had significantly less aggressive-disruptive behaviors at school, more symptoms of depression, and more themes of aggression and rejection on the Robert's Apperception measure. Overall results suggest that children with hemophilia demonstrate more difficulties with emotional functioning than peers, although they do not show clinically significant levels of symptoms. In other areas of functioning, including social abilities and behavioral adjustment, there were no differences (Trzepacz et al., 2003).

Gartstein, Short, Vannatta, and Noll (1999) evaluated psychological adjustment using the CBCL (Achenbach, 1991), the CDI, and a demographic questionnaire in various samples of children with chronic illness, including hemophilia, sickle cell disease (SCD), cancer, and juvenile rheumatoid arthritis (JRA). Within the hemophilia subgroup of the study, 21 families participated, and all children sampled were Caucasian males. In order to compare adjustment of children with hemophilia, to their peers, a control group was devised with children matched on age, race, gender, and

school classroom (i.e., children in the control group were from the same classrooms as the children with hemophilia). Results indicated that mothers of children with hemophilia rated their children as experiencing significantly more Internalizing Behavior Problems on the CBCL than mothers of children in the comparison group. Specifically elevations were seen on the subscales of somatic concerns, which is often elevated for individuals with chronic illness, and anxiety/depression. Father report of child adjustment did not yield any differences between children with hemophilia and without hemophilia. Children with hemophilia reported having more significant symptoms on the CDI than their matched peers, although scores were not in the clinically significant range (M = .25; Gartstein et al., 1999).

As a part of the larger Hemophilia Growth and Development Study, Nichols and colleagues (2000) reported on changes in adaptive functioning and emotional adjustment over the course of 4 years between children with hemophilia/HIV- and those who were hemophilia/HIV+. Participants included 333 boys, ages 6-19 years, of which 207 children were hemophilia/HIV + and 126 were hemophilia/HIV-. A majority of the individuals were classified as having a severe form of hemophilia. The parents of the children sampled completed the Pediatric Behavior Scale (PBS; Lingdren & Koeppl, 1987), along with the Vineland Adaptive Behavior Scale (Sparrow, Balla, & Cicchetti, 1984). Results indicated that children with hemophilia who were HIV- had better adaptive functioning, and demonstrated less change in emotional adjustment, than children who were HIV+. All of the children sampled showed decreased levels of externalizing behavior problems on the PBS over time and measures assessing anxiety and depression were within the normal range.

In comparison to studies showing that children with hemophilia have not endorsed clinically significant levels of psychological distress (Gartstein et al., 1999; Nichols et al., 2000), others have indicated that boys with hemophilia demonstrate difficulty in social functioning. It is possible that constraints placed on children and adolescents' participation in sport activities impacts social relationships. This may be especially relevant during adolescence, when inclusion in a sports team helps to form social relationships. In addition other studies found that boys were often reluctant to share medical information with peers and teachers, which could pose as an obstacle to healthy adjustment (Oremland, 1986).

In general, studies evaluating behavior and psychological adjustment to hemophilia have been mixed. Some of the earlier studies have indicated significant levels of psychological distress in mothers and older adolescents with hemophilia, while additional studies have shown elevated scores on measures of child anxiety, depression, and somatic complaints compared to a normative group, although results were not clinically significant (Gartstein et al., 1999; Klein & Nimorwicz, 1982; Trzepacz et al., 2003). While, medical professionals report that parent and child maladjustment to hemophilia is the most prevalent psychosocial problem in this population (Drotar et al., 1997). However, when compared to other children with chronic illness, those with hemophilia demonstrated significantly less anxiety, had better adjustment, and scored in the normal range on measures of behavioral functioning (Bussing & Burket, 1993; Colegrove & Huntzinger, 1994). Of note is that some of these studies had low sample sizes (Gartstein et al., 1999; Klein & Nimorwicz, 1982). While families and their children with hemophilia do not show poor adjustment compared to other disease

groups, they are at risk to experience more problems with psychological adjustment than non-ill peers even though scores are in the non-clinical range (Gartstein et al., 1999). When discussing behavioral and emotional concerns of children with hemophilia it is important to also explore neuropsychological implications of hemophilia and how cognitive abilities and academic success may affect behavioral/social functioning.

Intellectual Abilities, Neuropsychological Functioning and Academic Concerns

Recent research has discounted early accounts, that children with hemophilia demonstrate above average to superior intellectual capacities (Olch, 1971), and has instead shown that these children often have average abilities on measures assessing intelligence (Colegrove & Huntzinger, 1994). Despite average intelligence, children with hemophilia have consistently been found to have decreased academic achievement compared to what is expected given their intellectual abilities (Smith et al., 1997). Many indicate that problems with academic achievement may stem from chronic absenteeism, which also leads to problems in social functioning (Fowler, Johnson, & Atkinson, 1985; Weitzman, 1986). However, some studies have shown no relationship between absenteeism and decreased academic achievement. It may be that other factors including decreased social competence and lower adaptive functioning impact academic achievement in children with hemophilia.

Olch (1971) explored the intellectual and academic capacities of boys with hemophilia. Results on intelligence measures (45 children ages 2 to 21 years) indicated slightly above average levels of intelligence for children with hemophilia, however a

number of the children sampled demonstrated below average levels of academic achievement. Lack of school attendance was significant, as over one-third of the children missed at least one-fourth of the school year. Given the number of medical complications that children with hemophilia experience, and the nature of the disease, absence from school is a concern for many families (Woolf et al., 1989). Other studies assessing school functioning in children with hemophilia, have found that boys with the disease often experience difficulties with school adjustment and lag behind other children in academic achievement (Markova & McDonald, 1980; Woolf et al., 1989).

A more recent study conducted by Colegrove and Huntzinger (1994) examined social competency, school absenteeism, and academic achievement in 37 children with hemophilia (ages 8 to 19 years). A majority of the children came from middle class (61%) Caucasian families, and had a more severe form of the disease (86% boys with severe hemophilia). The children were administered the Wechsler Intelligence Scale for Children-Revised (WISC-R; Wechsler, 1974), along with the Wide Range Achievement Test-Revised (WRAT-R; Jastak & Wilkinson, 1984) during the baseline assessment. Caregivers also completed the CBCL. At the time of the second assessment, children were given the Woodcock Reading Mastery Tests-Revised (WRMT-R; Woodcock, 1987) and the Social Skills Rating System (SSRS; Gresham & Elliot, 1990). Parents completed the caregiver version of the SSRS, while teachers were asked to report on the Harter Self-Perception Profile for Children (Harter, 1985), the CBCL, and the SSRS for teachers. Teachers were aware of the children's hemophilia status. Results indicated that on average children missed 27 days of school a year, which constituted 15% of the total school year. Verbal, performance, and full scale IQ scores were within the average

range, and measures of academic achievement yielded scores that were lower than expected based on results of intelligence measures. Interestingly absenteeism was found to be associated with decreased teacher ratings on measures of competence (academic, social, and athletic), physical attractiveness, and adaptive functioning. In addition, absenteeism was correlated with higher teacher ratings on a scale of problem behavior. There were no differences seen in scores between children with hemophilia /HIV+ and children with hemophilia/HIV- (Colegrove & Huntzinger, 1994).

Some have asserted that with the introduction of home care, absenteeism in children with hemophilia would decrease, since hospital services would not be utilized as frequently. One study demonstrated differences between Finnish children with hemophilia and their age-matched peers on measures of school absenteeism and achievement at baseline (1971-72), were no longer significant at a later time period with a similar group of participants (1988-89). Given that the introduction of home based care was in 1980's, it is possible that less obtrusive medical care played a role in decreased in absenteeism and increased school achievement (Kvist, Kvist, & Rajantie, 1990). Woolf and colleagues (1989) found different results indicating that school absenteeism continued to be an issue for children with hemophilia after the introduction of home care, as children sampled missed, on average, 18 days of school per school year.

A predecessor of the Hemophilia Growth and Development Study evaluated the academic and neuropsychological functioning of children with hemophilia (Nichols et al., 2000). Participants were the same as those reported in prior descriptions of this study, and included children with hemophilia/HIV+ and those with hemophilia/HIV-.

Measures included, the Beery Visual-Motor Integration Test (Beery VMI; Beery, 1989), the WRAT-R, the Vineland, the Grooved Pegboard Test (Trites, 1977), the Trail Making Test (Reitan, 1971), Word Fluency (FAS; Spreen & Benton, 1977), the Judgment of Line Orientation (JOLO; Benton, Hamsher, Vaney, & Spreen, 1983), the Boston Naming Test (Kaplan, Goodglass, & Weintraub, 1983), the PBS, and the Rey Auditory-Verbal Learning Test (Rey AVLT; Lezak, 1995). Overall there were no significant differences found on measures between children with hemophilia/HIV+ and those with hemophilia/HIV- (Smith et al., 1997). Scores on neuropsychological tests were in the average range for both groups of children. All of the children showed below average adaptive behavior and lowered scores on academic achievement measures, compared to what was expected given IQ results. Loveland and colleagues (1994) suggested that lower academic achievement may be related to absenteeism and psychosocial factors, however statistical analyses were not conducted.

A subsequent study (Sirois et al., 1998) explored relationships between neuropsychological and neurological findings in children with hemophilia. While children with hemophilia/HIV- showed normal MRI scans, they were more likely to present with coordination or gait abnormalities compared to children with hemophilia/HIV+. Overall, participants with lowered performance on the neuropsychological measures assessing cognitive functioning were more likely to have academic problems, a history of head trauma, and lower parental level of formal education compared to children who did well on the cognitive measures, regardless of HIV status. In general neuropsychological functioning was not related to neurological findings, although interactions with cognition and gait abnormalities were described.

Sirois and colleagues (1998) indicated that problems with gait and coordination were related to hemophilia, rather than to HIV status, and thus concluded that hemophilia complications have a negative impact on neurological functioning (Loveland et al., 1994).

Overall, results have consistently indicated average intellectual abilities for children with hemophilia, with studies demonstrating that many children with hemophilia are performing worse on academic achievement measures than what is expected given their level of intelligence. Deficits in academic achievement have been attributed to classroom absenteeism and discrepancies between IQ level and achievement appears to increase as children get older. Although it was expected that home care would decrease children's absence from school, recent studies indicate that school absenteeism is a chronic problem for this population. School absenteeism has been linked to decreased teacher ratings of academic and behavioral competence, and adaptive functioning in children with hemophilia. It is possible that psychosocial factors including peer relationships, family functioning, and psychological adjustment to hemophilia, may play a role in decreased academic achievement.

Variables that Affect Psychological Adjustment in Hemophilia

Although the degree to which hemophilia impacts children and adolescents' psychological adjustment is debated within the literature, numerous studies suggest that specific factors can influence the role that illness plays in adjustment. Variables such as child age, gender, stress, and family functioning have been shown to be important contributors to child, adolescent, and family adjustment to hemophilia (Drotar et al.,

1996; Madden, Terrizzi, & Friedman, 1982). Medical factors including illness variables, defined as frequency of hospitalization, frequency of ER visits, child reported pain frequency, child reported pain intensity, and age at diagnosis, have not been supported as playing a significant role in child and family adjustment to chronic illness (Drotar et al., 1996; Hurtig, Koepke, & Park, 1989; Hurtig & White, 1986; Lutz, Barakat, Smith-Whitley, & Ohene-Frempong, 2004). While there are varying opinions as to those factors that are the most important predictors of successful adjustment and the connections between the predictor variables, the focus of the current study is to further evaluate how psychosocial factors, specifically family functioning, impact adolescent adjustment to hemophilia, particularly surrounding adolescent adaptation to increased responsibility that accompanies the transition from pediatric to adult health care.

Adolescent Development and Health Care

As an older child enters into adolescence and then moves into adulthood a number of challenges are noted, both developmentally and with regard to health care for individuals with chronic illness (Schidlow & Fiel, 1990). First it is important to realize that adolescents do not function like children in the context of a family, nor are they independent adults (Court, 1993). Much of the research that has evaluated the impact of chronic illness in adolescence has not been sensitive to the differences in development and health care needs that are apparent across different stages of adolescence (Hamberg, 1974). Thus, a young adolescent at age 12 years has different needs than an 18-year-old adolescent who is preparing to transition out of his or her home environment and away

from pediatric care settings. Second, it is essential to discuss how chronic illness impacts adolescent development, how adolescent health care needs are different from younger children, and why transition is necessary for young adult patients. During this review of the literature, a discussion of the changes that accompany adolescence will be completed, followed by information about how changes typically seen during adolescent development are impacted by chronic illness.

Adolescence is viewed as an evolving period when changes in biological, psychological, and social roles are apparent (Holmbeck, 1994). As with other stages of development, there is a substantial amount of individuality and variability in when and how adolescents progress through this period of development. Some have conceptualized adolescence in two segments, one being the change from childhood to adolescence and the second the shift from adolescence to young adulthood (Steinberg, 1996). Holmbeck and Shapera (1999) proposed a model indicating how individuals progress through the stages of adolescence (see Figure 1). According to this model, the relationship between developmental changes that occur during adolescence and various psychosocial factors impacts adjustment across settings. How adolescents resolve conflicts depends on developmental changes and adolescent relationships with parents, adults, and peers. Holmbeck and Shapera (1999) go on to propose that the relationship between developmental changes, including physical maturity or cognitive advancement, and outcome is mediated and moderated by other factors (i.e., self-perception, family functioning, parental relationships). In an attempt to understand the model proposed by Holmbeck and Shapera (1999) a description of physical, cognitive, and social changes expected during adolescence will follow.

Developmental Changes in Adolescence

Included in the primary changes seen in adolescence are physical maturity, psychological/cognitive changes, and shifts in social roles. Physical maturity during adolescence includes changes in body proportion, the growth of body hair, changes in voice quality, increased strength and coordination in males, and the onset of menarche in females (Tanner, 1962). While variation in physical development is noted, research has indicated that pubertal development occurs about 2 years earlier in females than in males. Interestingly pubertal timing and pubertal status has been found to have an impact on adolescent adjustment and family interactions, in that females who physically mature at a faster rate have an increased likelihood to experience adjustment problems, while early physical development in males is met with increased socially acceptable behaviors (i.e., increases in athletic ability, more social relationships). The manner in which the family or support network reacts to pubertal development of an adolescent has been found to significantly affect how the adolescent adjusts to these changes (Holmbeck, 1996; Paikoff & Brooks-Gunn, 1991).

In addition to physical changes, adolescents also experience psychological and cognitive changes. According to Piaget (1972), adolescence is the period during which individual's transition from pre-operational thought to formal operational thought. It is during this time that higher-level cognitive reasoning is developed. Thus, individuals in the formal operational stage are able to think more abstractly about complex problems. Adolescents should be able to think about future events and be more realistic in their thought patterns (Holmbeck & Shapera, 1999). Other developmental theories, specifically the information processing theory, assert that adolescents analyze

information more efficiently, retain a greater capacity of information in working memory, have an increased knowledge base, and are better at regulating cognitive processes (Keating, 1990).

Finally changes in social roles are observed within adolescence. These changes in social roles can be seen in various contexts, as adolescents experience alterations in family relationships and status, are able to vote or enroll in the armed services during late adolescence (political), are able to begin working (economic shift), and are given increased responsibility within society (i.e., are able to drive, can be tried in the adult legal system).

Little has been reported within the literature about how physical maturity impacts disease severity or illness related factors in hemophilia or how chronic illness may influence physical development. This said, cognitive and social changes accompanying adolescence certainly have an effect on individuals with hemophilia. Adolescents exhibiting advancements in cognitive development and who appear to have a better understanding of complex medical regimes may be given more responsibility for medical self-care. Given limitations on their physical activity level, boys with hemophilia may have difficultly fitting in with peer groups who support increased involvement in athletic activities during this stage of development. Since many adolescents define social relationships through their peer group, adolescents with hemophilia may find themselves struggling to relate with certain peer networks. In addition increased social relationships, along with decreased time spent at home, may impact adherence to medical regimens as adherence to medical regimen decreases in adolescence. The model proposed by Holmbeck and Shapera (1999) details that various

interpersonal contexts might mediate or moderate how these primary changes impact developmental outcome and can be applied to understanding how a chronic illness may affect adolescent development.

Contexts that mediate or moderate the relationship between developmental changes and outcome. Holmbeck and Shapera (1999) outline four basic contexts that mediate and moderate the relationship between developmental changes and outcome, including the family, peers, school, and work. Specifically the family will be the focus of this review since it is of interest to the current study. Despite historical writings about adolescence as a period of "storm and stress", more recent research has not suggested that adolescence is a time of conflicted parent-child interactions (Holmbeck & Hill, 1991). Although conflicts between parents and adolescents are not supported to the degree that many previously believed, adolescence is conceptualized as a time when emotional distance between the parent and child occurs. Research suggests that the goal of parents during this time of development is to respect adolescent need for increased responsibility and decision-making, while continuing to maintain a high level of interaction and cohesiveness with the adolescent (Holmbeck, 1996). Studies have found that parents who lack flexibility have children with increased adjustment problems (Fuligni & Eccles, 1993). Holmbeck and Shapera (1999) further explain those variables that are of interest when studying the family context. Specifically they note that assessment of parent-child conflict, attachment between parent and child, parent-child collaboration (problem-solving), parenting styles, and parenting practices are important when evaluating family and adolescent adjustment.

As peer networks become more mature and provide adolescents with a support system outside of their family structure, peer relationships grow and develop. Although many adolescents focus on peer relationships during this time of development, the family remains as a base for the adolescent when they are attempting to pursue peer friendships. Hartup (1983) indicated that secure family relationships lead to successful peer relationships during adolescence and that adolescent relationships that occur across settings (e.g., friendships at school, work and home) provide adolescents with opportunities to hone value systems, increase self-reliance, and develop relationships (Holmbeck, 1996). Holmbeck (2003) also asserted that other intrapersonal and demographic factors including, gender, ethnicity, socio-economic status, individual perception, family structure, and the community environment can mediate and moderate the relationship between developmental changes and outcome. In addition to understanding how numerous variables impact typical adolescent development, it is also imperative to appreciate how chronic illness during adolescence may interfere with typical development and how interactions between family members may be altered by the presence of a chronic illness.

Adolescence and Chronic Illness

Developmentally, adolescence is conceptualized as a time for individuals to gain independence from their family network, solidify peer relationships, and develop a coherent sense of self (Berk, 2001). In addition, it requires individuals to make decisions regarding sexual practices, vocation, substance use, and future life plans. Thinking about these issues, and making decisions regarding personal values is

challenging for any individual, but can be intensified for adolescents who are also attempting to manage a chronic illness. Adolescents with chronic illness are gaining independence in their medical care, making choices about treatment, monitoring their own care, transporting themselves to medical appointments, and communicating with health care professionals about their medical needs (Chesler & Anderson, 1987).

Gaining medical independence can be troublesome for many adolescents, as some may find that parents assume they are able to handle more responsibility for their medical care than is developmentally appropriate, while others are unable to exert independence because of the level of parental involvement (Pless, Heller, Belmonte, & Zvagulis, 1988). Parents of children with chronic illness and health care professionals can become overprotective, which hinders adolescents need for increased autonomy (Cameron, 1985). In other situations, adolescents may take on too much responsibility for medical care early in development without having the ability to manage all of their needs resulting in poor health outcome. For example some younger adolescents may administer their own medications without parental supervision, which could be problematic when complex medication regimens are prescribed. Research has indicated that physical, social, and psychological development for adolescents with chronic illness may be stressed or delayed because of illness, as they may not achieve appropriate levels of independence and autonomy because of family over involvement (Delengowski & Dugan-Jordan, 1986; Rosen, 1992). Furthermore, Bullinger and von Mackensen (2003) reported that in a sample of 320 children with hemophilia and their parents, both adolescents and parents indicated significant concerns about how to plan for the adolescent's future.

Blum and colleagues (1993) reported that providing adolescents with programs that enhance medical independence may increase adolescent ability to take responsibility for self-care. Although some have asserted that a majority of individuals with chronic illness (84%) do not present with significant functional limitations and make the transition into adulthood without experiencing maladjustment, other studies have indicated that there are a significant number of adolescents with functional impairments who have difficultly managing the challenges that accompany transitioning to adult health care systems (16%) (Gortmaker, Perrin, Weitzman, Homer, & Sobol, 1993; Newacheck, McManus, & Fox, 1991). In addition many adolescents and families have reservations about leaving pediatric health care settings (Rosen, 1992), although a majority of adolescents with a chronic illness transition into adult care systems successfully (Gortmaker et al., 1993). In this study it was unclear if the adolescents and young adults sampled had access to, or participated in, transition programs offered through their pediatric hospital prior to transition. Programs that address parent and patient anxiety regarding the transition from the pediatric to adult health care settings and that acknowledge the change from intense parental involvement in child medical care to decreased involvement in adult care can be helpful in assuring successful transitioning for the adolescent in question (Blum et al., 1993).

Transition from Pediatric to Adult Health Care

With improvements in medical care, the life span of children with chronic illness including hemophilia, cancer, and sickle cell disease have increased (Gortmaker & Sapenfield, 1984; Lewis-Gary, 2001; Schidlow & Fiel, 1998). Gortmaker and

Sapenfield (1984) indicated that survival rates for children with chronic illness have greatly increased and mortality rates have decreased, thus the incidence of chronic illness in children has remained stable. Studies have reported that up to 80% of children with chronic illness survive into adulthood, and that approximately 90% of children with hemophilia live into adulthood (Blum et al., 1993; Gortmaker & Sappenfield, 1984). Together, these data suggest that the number of adolescents with a chronic illness who will transition into adult care settings has grown and will remain stable, resulting in new medical challenges for providers, adolescents effected by disease complications, and their families. As children with chronic illness progress in age, new treatment challenges arise including transitioning adolescents to adult care, addressing differences between pediatric and adult care treatment models, understanding the capabilities of pediatric professionals in treating young adults, and responding to the lack of information about what transition services are effective in producing successful outcomes.

A number of pediatric professionals have continued to treat individuals beyond age 21 because of a lack of comprehensive transition programs and identified adult care physicians (Council on Child & Adolescent Health, 1988; Fulginiti, 1992). Over the past 10 years, many children's hospitals and comprehensive care units have voiced concerns about treating individuals over the age of 21 years (Bachrach & Greenspun, 1990; Blum et al., 1993). The debate about when to transition an adolescent into adult care is not only impacted by the age of the adolescent, but also by their developmental abilities.

One of the advantages of the pediatric subspecialty is the availability of care that is based on a developmental model for treating children and their families. While there are many reasons supporting transition from pediatric to adult health care, it is also important to note that pediatric health care professionals are more aware of family contexts that individuals continue to live within throughout young adulthood and are sensitive to issues/treatment of adolescents and young adults who experience developmental delays. While the benefits of receiving care in this type of setting are numerous, one potential downfall is that the field of pediatrics is separate from adult medical care, which can affect collaborative care for young adult patients transitioned into adult health care. Pediatric health care professionals also have not received specialized training in adult disease processes, raising concerns about their ability to treat previous pediatric patients who have entered into young adulthood. Professionals, and hospitals, often are wary about the liability of providing services to young adults that are outside the scope of their training. Others have voiced concerns about treating adolescents and young adults, citing that pediatric physicians often communicate treatment options through parents, without providing important information to adolescent patients. Pediatricians may shy away from addressing important adolescent and young adult issues, including sexual activity, substance abuse, independent living, and risk-taking behaviors when parents are highly involved in the care (Blum et al., 1993). With decreased parental involvement that accompanies care in adult health care settings these issues may be better addressed.

There are many conflicting views about the pros and cons of receiving care in a pediatric versus an adult care center, and the influx of medical liability lawsuits in this

country over physicians practicing outside of the scope of their training has prompted many hospital administrators to issue policies that prohibit medical professionals from treating individuals over the age of 21 years (Bachrach & Greenspun, 1990). The implementation of these policies has caused concern within the medical community, as it has lead to a flux of young adult patients with chronic illness seeking adult medical care and resulted in increased concerns about what services are needed to help create a successful transition from pediatric to adult medical care. In addition health care professionals surveyed reported that they are inconsistent in how they transition adolescents and many indicated that they do not adequately prepare adolescents or families for the transition process (Por et al., 2004).

Research conducted in the 1980's noted a critical need for transition programs that provide older adolescents with support and information to prepare for transition into adult medical care (Barbero, 1982). Research has also indicated that a lack of transition programs has caused young adults to receive treatment for chronic illnesses from internists and family practitioners after discharge from pediatric care settings (Bachrach & Greenspun, 1990). Although clinic disbandment is often very different from transition to adult health care similarities can be drawn, as both transition and disbandment result in a cessation of the relationship between a patient and a physician, and that referral to another physician is needed. Some studies assessing the effects of clinic disbandment have shown that when a coordinator of care is not identified for youth and their families, a large proportion of patients do not receive adequate medical services or follow-up care. In addition higher rates of morbidity are found when these medical services were not arranged for clinic patients (Kaufman et al., 2000). This

provides striking evidence into the importance of transition programs that provide support and identification of competent adult health care providers for young adult patients (Bachrach & Greenspun, 1990).

In response to concerns about the effects of not providing transition services to adolescents, young adults, and families, groups including the Council on Scientific Affairs from the American Medical Association, have recommended, as early as 1993, transition programs that provide "uninterrupted, coordinated, developmentally appropriate, psychosocially sound, and comprehensive" health care to individuals with chronic illness (Blum et al., 1993; p. 570) and facilitate transition into adult health care (Council on Scientific Affairs, 1993). Despite numerous policy statements advocating the use of formal transition programs, empirical studies have not evaluated the success of these programs, nor has research adequately identified those factors thought to influence successful transition or transition readiness (Blum et al., 1993). Since data regarding the effectiveness of transition programs are not prevalent, a review of different models of transition programs cited in the literature will be reviewed.

Definitions of transition. Prior to discussing types of transition programs it is important to understand how the term "transition" has been defined and used within pediatric literature. Some have conceptualized transitioning as a "progressive, developmental process toward the assumption of adult role responsibilities" and indicated that programs should focus on the changing and anticipated needs of the adolescents (Modrcin, 1989; p. 221), while others asserted that transition is a

"multifaceted, active process" that encompasses medical, psychological, and educational needs of the adolescent and their family (Blum et al., 1993; p. 573).

Although many of these definitions of transition are similar, transition from adolescent to adult health care can happen in multiple ways, across different health care settings. For some change from a pediatric to adult health care provider does not occur in adulthood, as some chronic illness conditions are managed by primary care physicians who continue to provide care regardless of patient age (i.e., allergies and asthma). Even when providers remain static, a shift in how the patient is treated (i.e., increased autonomy, decreased family involvement) is appropriate. Thus, transition is not always equivalent to a change in health care personnel or a shift in location of services.

In addition, transition has been conceptualized as a process that occurs at varying rates depending on the individual and family being treated, therefore there is no set age when an adolescent should transition into adult health care. The decision of when to transition from pediatric to adult health care should be based on developmental readiness, severity of illness, complexity of treatment, personal characteristics of the individual, and availability of adult health care physicians. Those with more complex disease processes and severe functional limitations may require lengthier preparation for transition or a more structured transition intervention to aid in the transition process.

The issues surrounding transition from pediatric care to adult care systems are diverse and include assessing adolescent readiness to transition, development of interventions that aid transition, and enhancing professionals understanding of how transitioning affects adolescents and young adults with chronic illness. Adolescents

surveyed have expressed their concern about transition, providing evidence that programs that address adolescent concerns and research that provides practical information to practitioners on how to transition patients successfully are needed (Capelli, MacDonald, & McGrath, 1989). In order to adequately study the process of transition, it is important to understand how to assess adolescent readiness for transitioning.

Assessment of readiness. Assessment of adolescent readiness to transition has not been well reported within the literature, as a number of transition studies define readiness as "developmental readiness" and "a degree of social maturity". These general definitions of readiness make it difficult for physicians or other medical staff to determine how to identify those adolescents who are ready to move into adult health care. Some health care settings have used age limit requirements (i.e., patients between ages 18 and 21 years) as cut-offs for transitioning, and in fact some hospitals have issued policies that they will not treat individuals over a certain age (American Academy of Pediatrics, 1996). This said, many practitioners feel that there are other variables, both medical and developmental, that are important to consider when assessing readiness for transition (Capelli et al., 1989). Some of these variables are disease specific, while others apply to various illness conditions.

Researchers often cite psychological or developmental maturity as an essential factor in readiness for transition (Por et al., 2004). In studying a group of 40 health care professionals practicing in a London based hospital who treated adolescents with cystic fibrosis (CF) and sickle cell disease (SCD), medical staff reported that a variety of

criteria including mental maturity ($N = 21$), age ($N = 6$), willingness to transfer ($N = 5$), ability to care for self ($N = 5$), level of support ($N = 4$), and individual qualities ($N = 3$) influenced their decision about when an adolescent was ready to transition.

In a study evaluating a transition program in adolescents with Insulin-Dependent Diabetes Mellitus (IDDM), investigators at a hospital in Finland conceptualized readiness for transition by disease specific medical variables including cessation of growth and full pubertal maturity, as measured by medical outcomes (Salmi et al., 1986). These researchers indicated that adolescents, who had not reached full growth or pubertal maturity, still required services of pediatric physicians (i.e., including how to manage disease during hormone changes accompanying puberty). Of note is that sexual maturity should not necessarily be used as a way to determine cognitive maturity, as the two are not directly related. Both studies noted above did not provide a clear idea of how developmental maturity was assessed, and thus it was assumed that psychological readiness was determined by physician observation and treatment compliance (Salmi et al., 1986).

To provide a more objective measure, Cappelli and colleagues developed a questionnaire to determine psychological readiness for transition in a Canadian sample of individuals with cystic fibrosis (CF) (Capelli, MacDonald, & McGrath, 1989). Using a sample of pediatric ($N = 22$) and adult ($N = 10$) health care professionals, along with some adults with CF ($N = 3$), information regarding behaviors and knowledge needed for adult CF care was gathered (Capelli et al., 1989). Pediatric and adult health care professionals included pediatricians, nurses, social workers, and psychiatrists. Participants were interviewed regarding factors they felt were the most essential,

desirable or necessary in successful transition and a transition questionnaire was created. From the responses of the individuals interviewed, a 24-item questionnaire was devised to assess readiness. Included in the readiness measure were questions addressing knowledge of the disease and items assessing self-care responsibilities. Knowledge questions were as follows: (a) knowledge of effects of medications used to treat the disease (b) reasons for use of particular treatments (c) knowledge of the proper way to administer treatment (d) knowledge of one's own medications and doses (e) ability to make appropriate medical decisions based on hypothetical situations. Self-care questions included assessment of: (a) responsibility of contacting the provider when adolescent was ill (b) responsibility of arranging, transporting to, and keeping clinic or doctor appointments (c) independence in maintaining treatment regimen (Cappelli et al., 1989).

Following completion of the measure, a sample of 36 adolescents with CF were administered the questionnaire and parents were asked to subsequently rate how likely they felt the adolescent would transition successfully. Follow-up after transition was not reported. Results indicated that parent ratings of successful adolescent transition were associated with the self-care scale of the readiness measure described above (Cappelli et al., 1989). Overall the authors indicated that the questionnaire was better able to predict readiness for successful transition, as determined by parent rating, than was adolescent age. Given the number of participants ($N = 32$) and the limited number of measures used, results should be interpreted cautiously. In addition it is important to note that none of these individuals had actually been transitioned during the course of the study.

Despite limitations, the study provided important data indicating that self-care responsibility was important for successful transition. Thus research has suggested that age and physical/sexual maturity are not the best predictors of readiness to transition, because psychological maturity, physical maturity, and age do not always correspond. Research, across illness groups, should continue to explore how to determine readiness to transition by evaluating if health care professionals, parents, and adolescents' ratings of adolescent medical self care predict successful transition outcomes and readiness for transition. Understanding when an adolescent is ready for transition is perhaps the first essential piece to ensuring successful transition outcome. As physicians or other health care providers feel that an adolescent is ready to transition to adult health care it is important to facilitate the transition process for the identified adolescent and their family. While there is a lack of empirically based research on how to improve the transition process, a number of studies have assessed adolescent/family needs during transition and have made recommendations about the important components in a transition program.

Adolescent needs during transition and the components of a successful transition program. There is a lack of research that empirically evaluates elements of successful transition and most research has focused on clinical impressions from professionals working within transition programs and adolescents with a chronic illness. Blum and colleagues (1993) highlighted seven factors that were consistently identified by health care professionals as being necessary for successful transition through review of current programs that provide transition interventions: (a) improving professional

and environmental support (i.e., building support networks that facilitate healthy maturation in the adolescent) (b) increasing environmental support systems (i.e., family, school, health care systems) (c) providing cooperative care that is organized toward patients' needs (d) encouraging adolescent participation in medical treatment and health care (e) maintaining family support (i.e., establishment of autonomy and independence with their families) (f) parent support and (g) heightened professional sensitivity to the transition period (Blum et al., 1993).

Similar ideas were also discussed by a group of adolescents with a chronic illness who were interviewed about transition to adult health care (Soanes & Timmons, 2004). Adolescents reported having the following needs during the transition time: (a) for staff to address the lack of familiarity with adult services and distress over ending familiar relationships with pediatric staff (b) for time to build relationships with adult medical staff (c) for adequate preparation and discussion of transition before it occurred (d) for flexibility and individualized transition plans and (e) for support from others during the process (Soanes & Timmons, 2004).

Other critical issues for successful transition into adult health care have been identified though adolescent and parent interview. After interviewing a group of adolescents with IDDM and cancer for a pilot study, Hagen and Barclay (1987) reported that providing adolescents with honest information about their health, their course of treatment, and options regarding their health care allows adolescents to assert control over their treatment. Adolescents ($N = 6$, age range 15-24 years) were interviewed in a semi-structured group setting for a 3-hour period. The provision of information about financial and insurance issues was important for adolescents. Similar to adolescents in

the Soanes and Timmons study (2004), adolescents interviewed also identified a need to gain support from medical staff and other adolescents with the same chronic illness.

While factors identified were derived from observation of transition interventions (Blum et al., 1993) and gained through unstructured interviews (Hagen & Barclay, 1987; Soanes & Timmons, 2004), it is important to realize that more objective measures of the essential components of successful interventions and the effects that structured interventions have on adolescent adjustment to transition are essential. Equally important is research that explores how different illness groups may react to transition and how to construct interventions that effectively meet the needs of individual illness populations.

In an effort to determine parent and adolescent perspective on transition programs using more structured methods, Telfair, Myers, and Drezner (1994) assessed the transition needs of 36 adolescents (13 to 19 years old) and 60 young adults (21 to 30 years old) with SCD. Using a non-standardized Sickle-Cell Transfer Questionnaire (SCTQ) young adults were asked to provide information about their experience and feelings about transitioning into adult health care and adolescents and their parents provided their perspective on how they would feel and what they would need during transition. For adolescents with SCD the main concern surrounded financial responsibilities and insurance within the adult health care settings, especially for older adolescents and young adults who may be released from their parent's insurance company at 18 years of age (Newacheck, 1990). In addition adolescents surveyed were unsure of how physicians would relate to the impact that SCD has on their life and how empathetic adult physicians would be about symptoms they were experiencing.

Parents, voiced some of the same concerns as adolescents, however they also felt anxiety about their children taking more responsibility for their medical care and managing pain episodes independently. Overall participants acknowledged feeling mixed emotions about the transition, and indicated that a transition program would help them gain control over their medical treatment. Specifically adolescents were interested in receiving more information about adult health care, meeting adult physicians, obtaining problem-solving tools in disease management, gaining independence, and learning how to navigate through the medical system. Older adolescents (over the age of 16 years) liked the idea of transitioning more than younger adolescents, and those with severe disease symptoms were found to have more concerns about transition than those with less severe disease symptoms. Caregivers, who were married, were significantly more likely to feel relief when their children transitioned, than unmarried caregivers (Telfair et al., 1994).

Court (1993) also evaluated adolescents' feelings about transition programs and transition into adult care settings. Adolescents with diabetes receiving treatment in an Australian hospital were mailed questionnaires. A total of 105 adolescents participated, including 48 males and 57 females between the ages of 15 to 18 years. Adolescents indicated their most important needs as being: information about their disease, new developments in treatment, emergency care, and accessibility to phone advice. When asked about the key factors that were essential to treatment in adult health care settings, adolescents identified short waiting times, confidentiality, and informality. Participants reported that it was extremely important for them to have their own identified specialist who remained consistent and who they were able to access regularly.

When considering transition it appears essential to individualize the transition plan, as adolescents within similar transition stages may have different needs (Lewis-Gary, 2001). Addressing parent and adolescent concerns and feelings about transition, as well as allowing the adolescent opportunities to make developmentally appropriate decisions regarding healthcare and teaching skills to help them gain medical independence are crucial. Programs that provide comprehensive, and collaborative, care that increases adolescent health care responsibility and works with parents to decrease family involvement can facilitate increased independence, readiness for transition, and successful transition (Konsler & Jones, 1993). In addition to addressing parent and adolescent needs through structured transition programs, increasing medical staff knowledge of essential components of transition would be important in providing consistent support during transition. In general there has been limited training for physicians and other health care professionals regarding these issues, however collaborative training between pediatric and adult physicians is important (Blum et al., 1993; Lewis-Gary, 2001). Improving medical staff training on a systematic level (e.g., through medical school coursework) has been suggested as a way to increase provider knowledge and to better address families' needs during the transition process (Hagood, Lenker, & Thrasher, 2005). When medical staff do not attend to transition needs and transition programs are not in place, adolescent adjustment may be affected.

The impact of transition. Studies that assessed the impact of transition, and utilized standard outcome measures, find significant differences on outcome measures after transition, with poorer adjustment reported. As none of the health care settings

where the studies took place provided access to a transition program, results indicate how adolescents respond to transition without formalized intervention services in place. For example, in an early study, Salmi et al. (1986) evaluated the metabolic control of 61 Finnish adolescents with IDDM over the course of a 2-year period covering transition. Results indicated that measurements of HbA1 were stable for the first year of the study, while participants remained in pediatric care settings. A slight deterioration was noted upon transition to the adult health care settings, specifically for female participants 3 to 6 months after transition. Overall metabolic control significantly improved from the time of the first visit in the pediatric health care setting to the last visit in the adult health care setting. This improvement was more significant for females than for males, and the authors attribute these improvements to changes within the adolescents themselves, as the practices of the physicians were reportedly the same across both settings. Individuals with a shorter disease history were found to have an easier transition, as recorded by more stable metabolic rates, than individuals with a long disease history (Salmi et al., 1986).

In a study conducted by Wysocki and colleagues, health and adjustment was studied in late adolescents with insulin-dependent diabetes mellitus (IDDM) to assess adjustment during transition (Wysocki, Hough, Ward, & Green, 1992). A structured transition intervention was not provided. The sample of 44 women and 37 men, between the ages of 18 and 22 years, were administered standard measures that included demographic, social support, knowledge, and adjustment questionnaires. Compared to a normative group without chronic illness, participants in this study did not show evidence of elevated scores on measures of psychopathology. This said, the results

suggested that many of these individuals had poor treatment compliance, limited health care use, and poor glycemic control. On a measure of quality of life, older adolescents with IDDM rated themselves as having lower quality of life compared to younger adolescents with IDDM, who were participating in another study by the authors. Of particular interest was that a majority (65%) of the individuals sampled for the study continued to live at home, while 15% lived in a college dormitory. In addition these individuals rated themselves as having fewer social supports and greater social insularity on measures assessing behavior. Overall individuals' retrospective rating of adjustment in early adolescence was significantly correlated with adherence, health care utilization, and quality of life in later adolescence, indicating that poor adjustment might be a factor that identifies those adolescents in need of additional services during the transition period.

Since a large portion of the transition literature does not include control groups, it is difficult to determine if positive (or negative) changes seen after transition are related to the transition or are due to maturation (Blum et al., 1993). This said, results from these studies suggest that without a transition program in place, difficulties with treatment compliance can occur. In addition, they indicated that the developmental transition from adolescence to young adulthood may be particularly challenging for adolescents with a chronic illness and that increased support during this time is appropriate. Overall results provided support for more formalized transition programs during this time of transition.

Development of transition programs. Rosen (1992) has presented very specific guidelines to follow when devising a transition program based on other accounts of transition interventions, which include: holding the transition program at the adult health care setting, introducing the families to the idea of transition early in pediatric health care settings, providing adequate information regarding health education and self-care, giving information that is directed toward and relevant to the adolescent, encouraging decision making by the adolescent and including the adolescent when explaining treatment procedures (Court, 1991; Nasr, Campbell, & Howatt, 1992; Schidlow & Fiel, 1990). Further steps, such as obtaining adolescent assent for medical treatment is also important. Of importance is that Rosen indicated that empirical evidence supporting the use of transition programs is needed. When developing a transition program, Rosen (1992) stated that, care provided in the adult health care setting should be equivalent to that in the pediatric health care setting in terms of quality and intensity and that collaboration between the pediatric and adult physicians during the transition process is necessary. In an attempt to better clarify how these components of transition can be practiced, transition program models have been proposed.

Court (1991) conceptualized transition programs by three general models depending on the number of steps taken to transition patients: pediatric health care to adult health care, pediatric health care to transition clinic to adult health care, and pediatric health care to adolescent health care to young adult health care to adult health care based on previous work conducted with adolescents who have diabetes (Baum & Kinmonth, 1985). The article written by Court also interviewed 70 (male = 32, female = 38; *M* age = 29.5 years) individuals with IDDM who had been transitioned into adult

health care within the last 5 years. According to Court (1991) direct transition from pediatric health care to adult health care was the most common transition model and he highlighted pros and cons to each type of program in his review. Some studies have suggested that there are lower dropout rates if older adolescents are transitioned to young adult health care settings before being placed in adult health care (Baum & Kinmonth, 1985). Studies that have evaluated transition outcome have not used control groups nor have they specifically evaluated a transition intervention program (Betz, 2004; Rettig & Athreya, 1991). Unfortunately there is a crucial lack of information about how these transition program models are implemented and the effects that transition programs have on adjustment following transition.

An early study (Nasr, Campbell, & Howatt, 1992) assessed transition in 40 adolescents with cystic fibrosis. A portion of the adolescents included in the study (33%) had attended a transition program, while the other participants were recruited from an adult CF site and had not attended a transition program. Few details were provided about the components of the transition program, with the exception that the program entailed meeting with the adult pulmonary fellow in the pediatric pulmonary clinic during the year prior to transition. Qualitative data was gathered from these adolescents regarding transition. No formalized measures were used. Most of the adolescents who attended the transition program felt that these visits with the fellow were helpful (9 out of 13) and 42% of the complete sample felt that attending a transition program would be "helpful". Other adolescents (17%) encouraged transitioning directly to adult care without attending a transition program.

Only one study, evaluating the impact of a residential weekend focusing on transition from pediatric to adult healthcare services with 13 adolescents with diabetes, has been published (Cuttell, Hilton, & Drew, 2005). Developed in England, this program focused on independent living skills (e.g. cooking meals, budgeting, shopping for food), career skills (e.g. interviewing for job), and health care skills (e.g. alcohol awareness, smoking, driving, sexual health). In addition, education regarding transition and opportunities to speak with adults with diabetes who had recently transitioned were provided. While standardized measures were not used to evaluate the effectiveness of the transition program, all adolescents surveyed reported that the weekend was "useful", "fun", and "educational". They also indicated that they felt more confident to care for their illness independently and more comfortable with the transition to adult health care. As a portion of the transition program was educationally based, the authors asked a group of adolescents with diabetes from the same clinic who did not attend the weekend to fill out a questionnaire evaluating disease knowledge (*M* percentage = 58.6). Participants in the intervention also completed a pre (*M* percentage = 50.4) and post (*M* percentage = 76.5) assessment of the same questionnaire, suggesting that the intervention did significantly impact knowledge of disease (Cuttell et al., 2005).

While these are important data, evaluation of other transition programs using more participants and standardized questionnaires is necessary. In addition, assessment of those variables thought to facilitate successful transition and transition readiness, including family functioning and parent involvement, is crucial, as family factors may play an important role in predicting adolescent's readiness for transition to adult health

care, their ability to handle increased medical independence and their success following transition.

Family Functioning

In addition to the normal daily stressors that a family with a healthy child manages, families with children who have a chronic illness are required to deal with stressors involving symptoms associated with the disease and treatment protocol. Families managing a chronic illness may be confronted with financial difficulties, changes in family roles, and constant monitoring of the chronic condition (Shannon, 1996). In both research and clinical settings, the importance of evaluating child adjustment to chronic illness with respect to family and environmental surroundings is essential. A number of theories have been developed using an approach that stresses the necessity of including all of the systems that interact and impact a child's adjustment (Brofenbrenner, 1979; Kazak, 1989). The impact that chronic illness can have on family structure is multifaceted as family functioning, parenting styles, and parent-child interaction may be impacted by disease variables, and in turn may influence the relationship between disease factors and adjustment. Thus childhood chronic illness is seen as not only having an impact on the child with the illness, but also creates change for every family member. In addition how family members and caregivers deal with the illness will have an impact on how the child manages and adjusts to their condition.

Specifically, measures of family functioning give information on how a family attempts to maintain balance in the face of stressors associated with chronic illness. Family functioning is thought to be a combination of factors including family cohesion

and family adaptability. Richmond (1917) introduces the terminology of family cohesion and defines its as the "degree of emotional bonding between family members". Family cohesion is thought to be critical to a family's ability to survive and flourish as a unit. Others have explained cohesion or family functioning as the compatible needs of individuals that allow them to function together as a group (Shaw, 1981). Family adaptability, the ability of a family to maintain flexibility in light of external stressors, is also crucial in determining how a family adapts to childhood chronic illness. Discussion of how family functioning can impact adjustment to hemophilia and other chronic illnesses will follow.

Family Functioning and Psychological Adjustment to Hemophilia

Manco-Johnson (1996) states that although the literature on family functioning in hemophilia is limited, that practitioners working with children with hemophilia have extensive experience regarding family functioning issues. She goes on to cite that family issues that can impact treatment include, "acceptance of the diagnosis, perception of the problem, parenting style, hemophilia-related communication, social support, disclosure issues, financial concerns, and perceived benefits and toxicity of the therapy plan" (Manco-Johnson, 1996, p. S22). The presence of a child with hemophilia can affect a family's ability to function in a number of ways. The genetic nature of hemophilia has been related to feelings of guilt and depression in a number of parents who have children with hemophilia (Varekamp et al., 1990). Families also struggle with balancing the need for children to engage in age-appropriate activities, like sports, while restricting risk-taking behaviors or play.

Varekamp and colleagues (1990) evaluated 126 mothers of children with hemophilia between ages 0 to 12 years in a Dutch hemophiliac clinic. Family burden was measured by assessing how care for a child with hemophilia impacted everyday activities in the life of the family (including housekeeping, leisure activities, external relations, and holidays). The impact of chronic illness on marital status was also assessed by asking parents if they had grown closer or farther apart as a result of their child's illness. Mothers interviewed indicated that care for their child "almost never" interfered with their ability to engage in everyday activities. Those things that were most often disrupted by medical care for the child were housekeeping, keeping a regular daily schedule, relaxing after work, time for leisure activities, going out with spouse or partner, and holidays. Overall 52% of the mothers surveyed indicated that hemophilia had not impacted their marital relationships, with 45% of those mothers reporting that they had grown closer with their spouse since the diagnosis of hemophilia. Although half report no significant changes in marital relationships, 48% did endorse negative changes in the relationship with their spouse emphasizing the impact that a diagnosis of hemophilia can have on parents of the affected child. Child age and disease severity did not significantly impact level of family burden.

Confirmation of childhood chronic illness impact on family burden and family relationships was reported in a study assessing the effects of treatment attitudes and family stress on treatment adherence. Seventy-five male participants ranging in age from 8 to 20 years and their mothers were evaluated (Van Sciver, D'Angelo, Rappaport, & Woolf, 1995). The sample was comprised of 31 boys with hemophilia (*M* age = 15.1 years), 22 male patients with sickle cell disease (*M* age = 11.9 years), and 22 children

with asthma (*M* age = 11.4 years). Measures including healthcare ratings of medical adherence to treatment protocol, the Medical Compliance Incomplete Stories Test (M-CIST; Koocher, Czajkowski, & Fitzpatrick, 1987), and the Impact of Illness Scale (Stein & Reissman, 1980) were administered. Overall significant differences between groups on measures of treatment adherence were seen, as males with hemophilia were rated as being "somewhat compliant" with treatment protocol, but "resistant" toward medical intervention. Although there were no significant differences seen in family stress among these three groups, compared to normative groups, all three groups of children had elevated levels of family strain. It is important to realize that the complex treatment associated with many chronic illnesses, especially hemophilia, can put added strain on a family system that is attempting to cope with demands placed on them by daily life in addition to the stress of complicated and invasive medical regimens.

Two studies have evaluated the impact of hemophilia on family functioning and family relationships. In children and adolescents with hemophilia elevated levels of family strain are noted, along with decreased positive relationships between parents of children with hemophilia. Since there is limited information regarding family functioning in adolescents with hemophilia and their families, a broader review of the pediatric literature on family functioning will be provided.

Family Functioning in Relation to Adjustment in Chronic Illness

Family functioning has been evaluated with regard to the effects that family cohesion has on adjustment to a variety of chronic illnesses. Higher ratings of family functioning are linked to decreased conflict, better organization, and increased

adjustment in chronic illness. Perrin, Ayoub, and Willet (1993) examined mother, child and teacher report of adjustment and the factors related to adjustment. The sample included 187 children (range 7 to 18 years) who were either healthy or who had a chronic illness (cerebral palsy, spina bifida, rheumatoid arthritis, scoliosis, muscular dystrophy, seizure activity). The CBCL was administered as a measure of adjustment, along with the Social Competence Scale (SCS) of the CBCL, the Personal Adjustment and Role Skills Scale (PARS; Walker, Stein, & Perrin, 1990) and the HRI. Family functioning was assessed by the Family Environment Scale (FES; Moos & Moos, 1981). Children's self-concept was obtained through report on the Piers-Harris Scale (CSCD). Families who reported high cohesion, better organization, high involvement in social activities, minimal conflict, and flexibility with family rules (when necessary) had children with better psychological adjustment (Perrin et al., 1993).

Davis, Tucker, and Fennell (1996) conducted a study of 22 pediatric patients with renal failure (*M* age = 9.92 years) and 12 patients who had recently received a kidney transplant (*M* age = 10.45 years). Results indicated that family cohesion had a significant positive relationship with daily living skills in the kidney transplant group and had an impact on communication skills, socialization skills, and overall functioning in the renal failure patients, such that families who reported higher levels of family cohesion had better functioning in the area of daily living skills, communication, socialization, and adjustment.

In a sample of 42 children (*M* age = 8.4 years, *SD* = 2.3 years) with congenital or acquired limb deficiencies, Varni and others evaluated family functioning and child temperament in relation to adjustment (Varni, Rubenfeld, Talbot, & Setoguchi, 1989).

The CBCL was administered as a measure of psychological and social adjustment, while family functioning was assessed by the FES. Additional measures included the Colorado Childhood Temperament Inventory (Rowe & Plomin, 1977) and the Degree of Limb Loss Scale (Varni et al., 1989). Results supported that family functioning and child temperament accounted for a significant amount of the variance in predicting psychological and social adjustment to limb loss over and above that accounted for by demographic factors. Greater family cohesion and organization, and less family conflict, resulted in better adjustment within this sample (Varni et al., 1989).

Lavigne, Nolan, and McLone (1988) used parent report to examine the effects of child coping and family cohesiveness on adjustment to myelomeningocele. The CBCL was administered as a measure of adjustment, while coping was measured through a 49-item Coping Inventory and scores representing family cohesiveness were obtained from the FES. The combination of family cohesiveness, overall coping ability, and temperament correctly predicted 94% of the children as being maladjusted as compared to adjusted on the total behavior scale. Thus Lavigne et al. (1988), through an indiscriminant functional analysis, conclude that the combination of factors, including family functioning and coping, has the ability to predict a child's adjustment to a chronic illness.

Another study assessing family functioning and coping in children with SCD also indicated significant associations between coping and family functioning (Lutz, Barakat, Smith-Whitley, & Ohene-Frempong, 2004). In a sample of 73 caregivers and 23 children (*M* age = 7.54 years, *SD* = 5.49 years) measures including a demographic form, the KIDCOPE (Spirito, Stark, & Williams, 1988), the COPE (Carver, 1997), the

McMaster Family Assessment Device (FAD; Epstein, Baldwin, & Bishop, 1983), the CBCL, and the Miami Quality of Life questionnaire (QOL; Armstrong et al., 1999) were administered. Additional measures of disease severity and stress of disease were also computed. Relationships between disease severity and adjustment and disability stress and adjustment were not found. Child active coping strategies were found to be associated in the expected direction with increased family functioning. Child gender and age were also found to play significant roles in family functioning and adjustment, with parents of male children reporting lower family functioning. The child sample for this study was small, thus increased sample size is needed to support the results.

Overall family functioning has been shown to be an important variable when considering how, and the degree to which, a child adjusts to stressors associated with a chronic illness. Additionally the interaction of other variables, like coping and demographic factors, with family functioning has been found to better account for psychosocial adjustment to chronic illness. In addition to discussing family functioning, parent and child interactions are important to assess as they may be altered by the presence of a chronic illness. Understanding parent and children interactions and parenting styles may be particularly important to facilitate adjustment during transition.

Parenting Styles and Parent-Adolescent Interaction

Most research on parenting styles has evaluated how parenting affects adjustment in a population of children and adolescents without chronic illness. Given the impact that parenting styles have on adolescent adjustment in individuals without

chronic illness, it is important to explore the effects of parenting style on those with a chronic illness condition. In addition, as adolescents' transition from pediatric to adult health care, the role of parental figures drastically changes. It is unknown if parenting styles impact adjustment during this crucial time when parents must balance the need to support their adolescent, with the need to provide them with increased independence, as the literature is scarce. First discussion of the theory surrounding parenting styles will be provided, followed by a summary of the literature that investigates the impact of parenting on adolescents and young adults without chronic illness. Finally literature detailing how parenting style affects adolescents with chronic illness, specifically hemophilia, will be provided.

Parenting Styles

Child-rearing practices and parenting styles have been found to impact child, adolescent, and adult development in various ways. Some studies have explored parenting styles, parental attitudes about childrearing, and patterns of parent-child interactions, while other research has focused on parental beliefs, which explores parental goals and attributions for behavior across a variety of contexts. Thus parental beliefs are viewed as situation specific, while parenting styles are more general (Coplan, Hastings, Lagace-Seguin, & Moulton, 2002). Based on various parental attributes, including parental warmth, responsiveness, demandingness, and control, different types of parenting styles have been proposed.

By observing Caucasian middle-class parents interactions with their preschool children during the mid 1960's, Baumrind (1971) gathered information about two

general dimensions of parenting styles: demandingness and responsiveness. Parents high in demandingness were found to set high expectations for their children and required their children to meet those standards, as opposed to parents with low demandingness who did not place demands on their children or try to impact or change their children's behavior. In addition parents who were high in responsiveness were very open and responsive to their children, while those with low responsiveness were rejecting and unengaged with their children (Berk, 2001). Based on these two dimensions, three parenting styles were devised.

An authoritarian parenting style has been defined as a style in which parents exert control or power over their children without warmth or flexibility. Often authoritarian parents do not value two-way communication with their children and they have a strict set of standards that they expect their children to follow. These parents are typically rated as low in respect to warmth, but high in control. Authoritarian parents tend to be coercive, harsh, and demanding toward their children, as they value obedience, respect, and order (Baumrind, 1971). In comparison, authoritative parents also set high standards for their children, however, these parents are willing to be flexible and receptive to their child's needs. Thus these parents exert control, while remaining warm and nurturing. Authoritative parents often discuss feelings with their children and provide explanations for punishment or consequences that result from their children's behavior. Parents who subscribe to a permissive parenting style tend to be nurturing and accepting of their children, however they do not make any demands or impose control. Children raised by permissive parents tend to make developmentally inappropriate decisions, and they often do not have household rules that they must abide

by. Many permissive parents lack confidence in their child rearing abilities and utilize ineffective parenting techniques (Berk, 2001).

Maccoby and Martin (1983) conceptualized a typology of parenting styles that included four patterns of parenting that vary with respect to levels of parental demandingness and responsiveness based on Baumrind's work (1971). In addition to the authoritative and authoritarian parenting styles, Maccoby and Martin (1983) added indulgent (low in demandingness and high in responsiveness) and uninvolved (low in both demandingness and responsiveness) parenting styles.

A number of variables, including ethnicity, SES, child temperament, parent temperament, and cultural expectations, influence parenting style (Coplan et al., 2002; Richter, Eisemann, & Richter, 2000). Individuals with lower SES tend to engage in authoritarian parenting styles and use more punitive child rearing practices (Kaufmann et al., 2000), although benefits of authoritative parenting are seen across all levels of SES (Kaufmann et al., 2000; Shumow, Vandell, & Posner, 1998).

Parenting style has been found to influence parent behavior, specifically parental discipline, and parenting style is thought to affect child adjustment. Most research has indicated that children raised by authoritative parents have better adjustment, as evidenced by increased independence, self-respect, peer relationships, and quality relationships with parents (Baumrind, 1971; Feldman & Wentzel, 1990; Kaufmann et al., 2000). Kaufmann et al. (2000) assessed the relationship between parenting style, specifically authoritative and authoritarian styles, child competence and behavior problems in a sample of elementary school children and their mothers. They reported that authoritative parenting was associated negatively with emotional distress and

behavior problems, and was positively related to healthy adjustment. Regression analyses indicated that authoritative parenting was highly predictive of healthy adjustment in this sample of children, even when age, grade, ethnicity, and SES were controlled.

Support for the positive impact of authoritative parenting on child adjustment was gathered from Baumrind's (1971) study in which researchers observed parent-child interactions in a preschool setting. Children and adolescents were observed at two additional time points (10 years and 15 years) and objective scales were not used in this study. Children with authoritative parents tended to be successful in academic and social situations and were highly motivated to accomplish their own goals. Children of authoritarian parents demonstrated decreased self-esteem, increased substance abuse, decreased satisfaction in peer relationships, and increased delinquent behaviors (Baumrind, 1991; Baumrind, 1971; Parish & McClusky, 1992). Children of permissive parents were found to be more dependent, have decreased achievement, and be more immature. These children also tended to be disobedient and overly demanding toward adults (Baumrind, 1971). The impact that parenting styles have on adjustment through the lifespan varies, as many indicate that conflicts and changes, which accompany different developmental stages, present new challenges for parents and adolescents. It is during these challenges that parent style and interaction with the adolescent may impact adjustment, as adolescents with more responsive parents may find it easier to navigate challenges (both developmental and medical), while adolescents with parents who are permissive or show a lack of flexibility may have more difficulty. Overall,

parenting styles may play a crucial role in adolescent adjustment to transition from pediatric to adult health care.

Parenting Style and Adolescence

While theories highlighted in previous sections, including the parenting theory of Maccoby and Martin (1983) are discussed within adolescent literature, other theories including the individuation theory attempt to integrate parenting style with developmental tasks seen specifically during adolescence (Grotevant & Cooper, 1986). As children progress into adolescence, developmental goals and tasks change. For adolescents, optimal parenting strategies that promote the attainment of autonomous functioning, individualization, and independence from family are different than the optimal parenting strategies needed for children to meet childhood developmental milestones (i.e., increased peer relationships, independent decision making) (Aquilino & Supple, 2001). Parenting behavior can greatly hinder or improve adolescents' abilities to achieve success in this developmental stage. Parents who encourage appropriate freedom and autonomy can provide adolescents with opportunities to practice decision-making skills and explore social roles in a secure environment (Aquilino & Supple, 2001). Providing adolescents with a safe environment to assume responsibility for medical self-care activities, while continuing to have the support of family members, is especially important to the development of independence and self-confidence in adolescents with chronic illnesses. In comparison, adolescents with disengaged or uninvolved parents may not have adequate psychosocial support and may feel overwhelmed by medical demands accompanying adult health care. These

adolescents may have a more difficult time solving medical problems that arise if there is not a parent available for consultation.

Parenting styles have also been evaluated with respect to adolescent autonomy. Eccles, Early, Frasier, Belansky, and McCarthy (1997) sampled 7th and 8th grade early adolescents and their families. The sample represented diverse ethnic groups, as 49% of the participants were African-American and 33% were Caucasian. A majority of the primary parents interviewed were mothers. Measures gathered information about demographic background, family dynamics, family and peer relationships, resources, family stressors, and adolescent development. Results indicated that positive and consistent regulation by parents was related to low levels of behavior problems and a higher Grade Point Average (GPA), whereas excessive psychological control was related to poor adolescent functioning. Adolescent girls who perceived high levels of parental connectedness and emotional support were more likely to be successful in the domains of psychological and behavioral development. In addition parents who did not employ strategies to help regulate their adolescent's behavior and provided limited support for autonomy in their adolescents, had adolescents who were more likely to be a part of a peer group involved in problem behaviors and who did not endorse conventionally positive adolescent behaviors (Eccles et al., 1997).

A study of 386 adolescents (8th-9th grade) assessed the relationships between parenting styles, academic achievement, and substance use (Cohen & Rice, 1997). Parents and adolescents were found to disagree on the type of parenting styles utilized in their respective families, as adolescents felt that their parents were less authoritative and more authoritarian than the parents reported. The sample was comprised of White

(55%), Hispanic (24%) and Asian (18%) families; parents represented diverse educational backgrounds. Adolescents who perceived their parents to be authoritative had higher academic achievement and lower substance use, compared to adolescents who rated their parents as permissive or authoritarian. Adolescents who perceived their parents to be less authoritative and more permissive were more likely to use tobacco and alcohol. The authors point out that parent perception of their parenting styles was not significantly related to child substance use. Baumrind (1991) also found that authoritative parents, who were highly demanding and responsive, had adolescents with less problematic drug use and better competence.

Radziszewska, Richardson, Dent, and Flay (1996) evaluated smoking, academic grades, and depressive symptoms across a sample of adolescents who varied in terms of ethnicity, gender, and SES status. The sample included 340 Asian, 519 African-American, 1827 Hispanic, and 1305 White adolescents, with 2063 participants being female and 1930 participants being male. Authoritative parenting style was found to be associated with the best outcomes, while the unengaged parenting style was associated with the worst outcomes (outcome included academic achievement and depression scores). Permissive and autocratic (authoritarian) styles were associated with intermediate results. Ethnicity and adolescent gender were found to moderate the association between parenting styles and outcome, with African American boys with unengaged parents and Asian females with both unengaged and autocratic parents having higher depression scores than any other ethnic groups. Across all of the ethnic groups, there was no support that autocratic (authoritarian) parenting led to better outcomes compared to other parenting styles. Smoking was not significantly different

across parenting styles. Of note is that all data was collected via child report (Radziszewska et al., 1996).

As results have indicated, parents who employ styles conducive with developmental progression have adolescents with better adjustment. Aquilino and Supple (2001) indicate that successful obtainment of developmental goals in adolescence, specifically individuation, leads to positive adjustment and increased competence in young adulthood. Although research has evaluated how parenting style impacts adolescent adjustment, fewer studies have shown how parenting style impacts adjustment in adolescents with a chronic illness.

Parenting Styles and Hemophilia

As children become adolescents, parent-child relationships change and more focus is placed on peer friendships. Adolescents with chronic illness have been thought to experience delays in development, in that they may not have the opportunities to or be able to meet the developmental tasks posed in adolescence (Hauser et al., 1979). For some adolescents with chronic illness deficits in physical, social or intellectual abilities greatly impact individuation from the family unit. In addition some have indicated that adolescents who are required to abide by a strict medical regimen may experience difficulties separating from their parents for numerous reasons. It may be that they rely heavily on their parents for help with medical care or that parents' attempts to control disease management interfere with adolescents' independence (Chassin, Presson, Rose, Sherman, & Prost, 2002). Parents may feel that adolescents are less compliant with

medical regimens, if they are not monitored, which is supported by research suggesting that adherence to medical regimens is the worst during adolescence (Delamater, 1993).

Very few studies have evaluated child-rearing practices in parents of children with hemophilia. Mayes, Handford, Kowalski, and Schaefer (1988) assessed parenting attitudes and practices and child personality traits in the families of 22 males with hemophilia (ages 5 to 19 years) who were treated in a comprehensive care unit over a 6-year period. Children were administered a personality questionnaire at the beginning of their treatment and again six years later. Both mothers and fathers completed the Roth Mother-Child Relationship Evaluation as a pre-test and post-test (MCRE; Roth, 1961). Results indicated that over time, personality traits reported by the children changed significantly as boys rated themselves as being less serious and sensitive, happier, more enthusiastic, and more self-reliant at post-test. There was not a comparison group to assess if these changes in personality styles were specific to boys with hemophilia or if they were seen in healthy matched controls as well. Parents did not differ on measures assessing parental attitude change over the treatment period. In comparison to the normative group, mothers were reportedly higher on a measure of acceptance and lower on a measure of overprotection and overindulgence at both pre and posttest. Fathers were also lower than the normative group on the measures of overprotection and overindulgence, but did not show significant differences from controls on a measure of acceptance or rejection. Parenting styles did not change over the course of the study. Maternal acceptance was correlated with child obedience and submissiveness, while high paternal acceptance and low paternal overprotection and overindulgence were significantly associated with child self-control and less child shyness. There were no

significant differences between parenting styles or attitudes in children with different levels of disease severity.

An earlier study conducted by Madden, Terrizzi, and Friedman (1982) interviewed 24 mothers of children with hemophilia (age range 3-18 years). Using a questionnaire devised for the study, Maternal Psychological Response to Hemophilia and Child Adjustment, mothers were asked to rate their level of distress about issues related to hemophilia including distress over diagnosis, feelings of guilt, and fear over medical complications. In addition they were asked to rate the level of support they received from their family and their child's adjustment to the disease (fewer problems than an average child, similar to an average child, more problems than an average child). Results indicated an association between mother adjustment to the disease and child adjustment. The authors asserted that mother parenting practices may mediate the interaction between mother and child adjustment, as mothers who experience extreme levels of distress about their child's illness may be more likely to become withdrawn or inconsistent with regard to parenting practices leading to decreased child adjustment. Statistically analyses were not run to explore this hypothesis. Mothers with severe levels of distress in this study were more likely to report having little control over their child, having difficultly establishing appropriate boundaries with the child, and having trouble forming high expectations for the child. Given that parenting styles were not directly measured, along with the nonstandardized assessment device used in this study, results should be interpreted cautiously. It may be that parents with high levels of distress may also evidence problems with parenting practices.

Consistent with the broader literature, results of the research provided note better adjustment in families where parents are not overly indulgent or overprotective and where parents set appropriate limits for their children with hemophilia. While one study found that parents in their sample showed less overprotection and overindulgence than a normative sample, another study reported that parenting style may be related to parent and child adjustment (Madden et al., 1982; Mayes et al., 1988). Research within the general pediatric literature provides more information about how parenting style may be influenced by chronic illness and the impact that parenting practices have on child adjustment.

Parenting Styles and Adolescents with a Chronic Illness

Using a sample of 64 families with children who had juvenile rheumatoid arthritis (JRA) ages 8 to 14 years and 64 families without children with a chronic illness (matched control group), Gerhardt and colleagues (2003) assessed child-rearing practices. In addition to surveying parents, the authors also interviewed 16 professionals working within the JRA clinic to assess their perception of parenting style. Demographic and disease related information was obtained and a questionnaire, the Child-Rearing Practices Report (CRPR; Block, 1965; Block, 1980), was administered to both mothers and fathers participating. Health care providers showed general agreement in their assessment of parenting practices, as many conceptualized families into three domains including protectiveness, discipline problems, and worry. Overall mothers of children with JRA reported more worry about their child's health and rough play behaviors, while fathers indicated worry about their child's health and difficulty disciplining their child. The authors reported that having a child with chronic

illness may affect parenting practices that are specifically related to the child's medical condition, but not affect general parenting practices. Most families showed parenting practices that were similar to the control group. When evaluating the impact of disease complications on parenting style, results suggested that difficulties in parenting were associated with increased disease complications. The higher the medical stress the more likely parents were to worry and become protective of their children. A relationship between parenting practices and length of disease was not found. Although this provides information about how parenting practices are affected by disease severity and are different in families with chronic illness, more information about child adjustment and parenting practices is needed.

Other studies have attempted to understand parenting practices by observing parent and child interactions. In a sample of 42 female children with JRA and a group of 42 matched control participants (*M* age = 9.96, *SD* = 2.40 years) parent and child interactions were recorded. Parents and their children completed demographic measures, and then were asked to engage in a memory game. Parents were instructed to have their child study a poster that displayed pictures of animals for 3 minutes. Following this study time, parents were to take the poster away and have the child generate as many of the animals as possible in a 10-minute period. Researchers allowed parents to talk with their children, but they were asked to not give the children the answers. Parents were not told that the purpose of this study was to assess parent and child interaction. Interestingly results showed that mothers of children with severe JRA were more likely to be directive with their child (i.e., gave more clues, prompted the child, provided more structure, and set more rules) than mothers of children with mild

JRA or no chronic illness. In general mothers did not differ in the amount of positive feedback given to children or in the frequency of questions asked. Of note is that children in the three groups did not differ in terms of their success on the task, how frequently they asked for parental help, or on off-task behavior. In an attempt to explain the results, Power, Dahlquist, Thompson, and Warren (2003) suggested that mothers with children who had more severe JRA may have been more anxious about their child's performance causing them to be more directive. These mothers may also have felt that their child needed more help, in that those children with severe disease may need additional help in daily activities and self-care, which parents may generalize as needing more help in other areas including cognitive tasks. Also of note is that mothers of children with more severe disease were more likely to follow up their children's incorrect and correct answers with negative and positive feedback respectively, as compared to the other mothers who used more specific cuing to provide feedback to their child. Power and colleagues (2003) felt that this type of interaction may have made the game less enjoyable for the children with severe disease in comparison to the other groups.

Within the pediatric literature more studies appear to be emerging that are looking at parent and child interaction and parenting style through the use of objective rating scales or coded observational methods. Results have shown differences in parenting practices among parents of children with chronic illness compared to those without chronic illness when they are in medical situations. Studies have not shown that global parenting practices are affected (Gerhardt et al., 2003). Other research has indicated significant differences in parent directiveness when the child has severe

disease compared to other children with milder disease complications. Further exploration of parent-child interaction within chronic illness is warranted, as few of these studies have focused on how parent and child interaction changes in adolescence and how parenting practices impacts adjustment.

Purpose of Present Study

As medical interventions become more advanced, children with chronic illness are living longer. In the case of blood disorders, the AIDS epidemic significantly impacted the type of long-term care adolescents and children with hemophilia and other blood diseases received. Currently medical professionals working with this population of adolescents are facing new challenges as they are attempting to find ways to assess transition readiness and to successfully transition adolescent patients from pediatric health care into adult health care. The purpose of the current study was to evaluate how factors proposed by Holmbeck and Shapera (1999), including demographic, family functioning, and parent-adolescent interaction, interact to predict adjustment during adolescence and readiness for transition to adult health care. The study also examined differences in psychosocial adjustment between adolescents with hemophilia and those with VWD. Finally the study explored families' feelings and concerns about transition and the need for a transition program for adolescents with blood disorders.

The findings obtained from the study will add to the small, but growing research that provides information about adolescents' transition from pediatric to adult health care and provided more information about family functioning, parent-adolescent

interaction, and adjustment in children with blood disorders. While numerous position papers have identified a need for research regarding the effect of transition on adjustment, few studies have effectively evaluated transition in pediatric populations (Bachrach & Greenspun, 1990; Blum et al., 1993; Fulginiti, 1992; Schidlow & Fiel, 1993). Many studies that evaluated transition have used subjective measures to assess adolescent feelings about transition, but have not included objective measures or parent informants (Capelli et al., 1989; Salmi et al., 1986). In addition studies assessing adjustment to hemophilia have not evaluated the effect of multiple factors, like family functioning and parent-adolescent interaction, on outcome in adolescence, and there are no studies published assessing the psychosocial implications of VWD on adjustment in pediatric populations.

The current study aimed to expand the literature regarding blood disorders and transition from pediatric to adult health care in a number of ways: (a) The study included adolescent and parent perception of transition, and used more objective and standardized measures. (b) This research was particularly important for hemophilia and VWD populations, as adolescents with blood disorders have not been studied extensively in the pediatric literature. In addition the lifespan of individuals with hemophilia has increased, indicating a need to address issues of transition with this population. (c) Information about variables thought to impact adolescent readiness for transition and adjustment to chronic illness were gained. Previous studies evaluating transition have not clearly defined how to assess readiness for transition (Salmi et al., 1986) and this study evaluated if medical independence and self efficacy predict adolescent readiness. This will provide physicians and other health care providers with

additional information to help them determine readiness for transition in their adolescent patients. (d) Finally the study examined what factors influence adolescent adjustment during transition. As adolescence can be a challenging developmental period, additional research that evaluates how adolescent development, family functioning, family interactions, and adjustment are impacted by chronic illness is important. The current study had three hypotheses.

Hypothesis I

That there would be significant differences in measures of disease, family functioning, parent-adolescent interaction, and adjustment between adolescents with hemophilia and those with VWD.

Hypothesis IA. That adolescents with hemophilia would show higher physician-, parent-, and adolescent-rated disease severity, higher physician-, parent-, and adolescent-rated disease interference, and higher illness stress than adolescents with VWD.

Hypothesis IB. That adolescents with hemophilia would show lower family functioning, more negative parent-adolescent interactions, fewer positive parent-adolescent interactions, and lower adjustment than adolescents with VWD.

Review of medical literature indicated that adolescents with hemophilia typically have more severe disease and more complex medical regimens than

adolescents with VWD (Paper & Kelley, 2002). In addition, adolescents with hemophilia usually have a number of restrictions placed on the types of activities they can engage in, while adolescents with VWD rarely are restricted from activities. Given the differences in medical severity and intensity of medical treatment, it was thought that physician-, parent-, and adolescent-rated disease severity and interference would be different between the groups, with adolescents with hemophilia demonstrating higher rated disease severity and interference reflective of increased medical severity and the intensity of medical regimens.

Literature has also suggested that adolescents with hemophilia may experience elevated levels of stress, anxiety, and family conflict related to the presence of the chronic illness (Van Sciver et al., 1995; Varekamp et al., 1990; Wallander et al., 1989). Literature has emphasized that chronic illnesses that interfere with physical activity level and participation in sports can be especially challenging for adolescent males (Oremland, 1986). In a study assessing adjustment in children and adolescents with SCD, Lutz and colleagues (2004) found that parents of male adolescents reported lower family functioning compared to female adolescents (Radziszewska et al., 1996).

In addition to differences expected between adolescents with hemophilia and those with VWD on measures of family functioning, it was expected that differences in the severity of the medical illness and complexity of the medical regimen between adolescents with hemophilia and those with less severe VWD would lead to differences on measures of adjustment. In comparison to other children with chronic illness, children with hemophilia have been found to display increased internalizing symptoms on measures of adjustment. Given that adolescents with hemophilia require increased

medical interventions and administer daily factor replacement products, it was thought that they would evidence problems with maladjustment compared to those with VWD, as more severe disease can have a greater impact on functioning.

Hypothesis II

That adolescent- and parent-rated family functioning (FAD) and parent-adolescent interaction (SCIFF) would moderate the relationship between disease variables and adolescent- and parent-rated adjustment. That the association between disease severity, disease interference, and illness stress and adolescent adjustment would be moderated by family variables, such that at high levels of family functioning and more positive parent-adolescent interaction, disease variables would have a weaker association with adolescent maladjustment, while at low levels of family functioning and more negative parent-adolescent interaction disease variables would have a stronger association with adolescent maladjustment.

The adolescent development model (Hombeck & Shapera, 1999) proposed that the relationship between primary developmental changes in normal adolescence and developmental outcomes (i.e., adjustment) are moderated by interpersonal factors (i.e., family, peer, school, and work) and demographic variables (i.e., ethnicity, gender, SES). While factors thought to impact adolescent development have been studied within the broader child-clinical literature, few studies have evaluated how changes associated with chronic illness, including disease severity, disease interference, and illness related stress, are impacted by family variables in predicting outcome. Adolescents with

chronic illness and their families are confronted with a number of daily stresses associated with disease variables, which are above and beyond daily hassles experienced by families with healthy adolescents (Shannon, 1996). This is especially significant in the case of hemophilia and VWD, where individuals must monitor bleeding episodes, manage acute and chronic pain, and administer daily factor replacement products (Lemanek et al., 1995; Merck, 2003). As home care is often the preferred method of treatment, adolescents with blood disorders and their families take on more responsibility for medical care and are encouraged to rely less on medical staff to handle illness management (Brown & Demaio, 1992; Lemanek et al., 1995).

An important component of adjustment to chronic illness and disease related stress is family involvement and positive family interaction. Few studies have evaluated the effects of family functioning, parenting styles and family interaction in hemophilia, however, overall results suggest that adolescents with chronic illness, including those with blood disorders, and their families are at increased risk to experience stress associated with medical variables (Davis et al., 1996; Lavigne et al., 1998; Van Sciver et al., 1995; Varni et al., 1989). Studies evaluating parenting in the general population have found that parents who have positive interactions with their adolescents and regulate adolescent activities have adolescents with decreased behavior problems and increased academic achievement (Cohen & Rice, 1997; Eccles et al., 1997; Radziszewska et al., 1996). Other literature focusing on chronic illness populations indicated that difficulties in parenting were associated with increased disease complications (Gerhardt et al., 2003; Power et al., 2003).

Hypothesis III

That disease, family and adjustment variables would predict a significant portion of the variance in physician-, parent-, and adolescent-rated adolescent and parent transition readiness.

Hypothesis IIIA. That disease severity and disease interference would predict a significant amount of the variance in physician-, parent-, and adolescent-rated transition, such that adolescents with low disease severity and disease interference would have higher rated self and parent transition readiness.

Hypothesis IIIB. That family functioning and parent-adolescent interaction would predict a significant amount of the variance in physician-, parent-, and adolescent-rated transition readiness, such that families with higher family functioning and more positive/fewer negative parent-adolescent interaction would have higher rated self and parent transition readiness.

Hypothesis IIIC. That parent and adolescent transition variables and current adolescent adjustment would predict a significant amount of the variance in physician-, parent-, and adolescent-rated transition readiness , such that parents and adolescents reporting fewer transition concerns, fewer negative transition feelings, higher adolescent medical independence, higher adolescent self-efficacy, and higher adolescent adjustment would have higher rated self and parent transition readiness.

Previous literature has suggested that disease and family variables are associated with adjustment to chronic illness and adjustment during transition (Cohen & Rice, 1997; Davis et al., 1996; Eccles et al., 1997; Perrin et al., 1993; Salmi et al., 1986). Individuals experiencing increased disease severity may be more likely to experience problems associated with transition. As these individuals most likely have more complex medical regimens and increased reliance on pediatricians, transition to a new adult physician may prove difficult (Salmi et al., 1986). Interestingly disease severity has not been linked to adjustment within the literature (Lutz et al., 2004), indicating that other variables such as negative parent and child perception of disease severity/interference and increased illness-related complications may be more indicative of reliance on health care professionals and poor adjustment both prior to and after transition.

Given that transition readiness is a measurement of how well young adults with chronic illness adjust to changes associated with developmental progression and the shift from pediatric to adult health care, it is expected that family variables will continue to be important. The research suggests that parents who are flexible in their parenting, but who remain involved, and families with greater family functioning (better problem-solving abilities, more family involvement, a higher level of organization) have children with better adjustment (Blum et al., 1993).

Transition readiness is linked to higher medical independence, since many adult health care settings rely more on the individual patient, and not the family, to adhere with medical treatments and regimens. Thus, adolescent medical independence and self-efficacy may play an important role in determining readiness for transition and

transition success (Hagen & Barclay, 1987; Konsler & Jones, 1993). In addition adolescents who feel that they have control in making medical decisions, who feel capable of handling disease complications, and who have some responsibility for medical care would more likely find the change to adult health care and increased medical independence less troubling. In addition literature suggests better adjustment prior to transition predicts better adjustment after transition (Wysocki et al., 1992).

CHAPTER 3: METHOD

Participant Recruitment

Data for this study were obtained from the Hemophilia Outpatient Clinic at St. Christopher's Hospital for Children and the National Hemophilia Foundation, Delaware Valley Chapter. A total of 27 adolescents and families were introduced to the study and 85% of families consented to participate in study ($N = 23$). Families who did not sign consent indicated being too busy to complete the study ($N = 3$) or having extenuating family circumstances that interfered with enrollment in the study ($N = 1$). All of the families who signed consent, with the exception of one, completed data collection. Attempts to gather data on the one family who did not finish the study stopped when the family failed to attend the second scheduled homevisit and were unable to be reached via the phone (i.e., number was disconnected). Overall 10 adolescents diagnosed with hemophilia and 12 adolescents diagnosed with VWD of varying severity levels were included in the sample ($N = 22$). File review was not collected on two families due refusing to consent to have medical records released or lack of response from medical staff to complete file review when participants were collected from outside of St. Christopher's Hospital. The one family who refused to have consent of medical records released had an adolescent who was not interested in completing the video taped interaction and at that point decided that they did not want to participate in the study, thus consent was not provided. The family did consent to have collected data be used in the current study.

The primary source of recruitment was identification of eligible families at St. Christopher's outpatient clinic. For 59% of the families recruitment occurred during their regularly scheduled medical appointment in the outpatient clinic. Medical staff discussed the study with families and if families were interested in participating in the study, they were introduced to the research assistant. Families either completed the study immediately following their medical appointment or arranged a time with the research assistant for a follow up appointment. If a research assistant was unavailable during the clinic visit, the medical staff obtained permission from the family to be contacted about the study.

Since this recruitment effort did not yield a sufficient number of subjects, additional methods of recruitment were employed to increase sample size. These methods included: 1) Sending a mailing to 50 eligible study participants at St. Christopher's Hospital (12 hemophilia and 38 VWD) that described the study. A self-addressed post card provided families with information on how to participate in the study. 2) Including a description of the study in the National Hemophilia Foundation newsletter. Families were provided with information on how to participate in the study. 3) Attending the National Hemophilia Foundation Family Weekend and recruiting participants from this activity. 4) Broadening inclusion criteria by increasing the age range of participants. 5) Introducing home visits to increase participant retention in the study. These various recruitment methods allowed for an additional 9 families to be recruited.

Overall, 23% of families were introduced to the study in clinic and completed the study during a regularly scheduled clinic appointment, 36% of the families were

introduced to the study in clinic and completed the study during a homevisit, 23% of the families were introduced to the study during the family weekend and completed the study during the weekend event, 14% of the families were introduced to the study during the family weekend and completed the study during a homevisit, and 5% of the families were recruited from the National Hemophilia Foundation newsletter and completed the study during a homevisit. There were no additional participants recruited from the clinic mailing.

Inclusion criteria for the study were as follows: 1) Adolescent participants must have been between ages 12-21 years. 2) Adolescents must have met the diagnosis for hemophilia or VWD and HIV-negative status was documented. Exclusion criteria for the study were as follows: 1) Adolescents were diagnosed with a developmental disability or mental retardation as reported by the parent or as indicated by chart review. 2) If a family had two children with hemophilia or VWD the oldest child (if younger than age 21 years) was asked to participate first in the study and the younger child was asked to participate second. One family in the study had two children who met criteria for the study. Since the older child was not interested in participating, the younger child was included.

Instruments

Instruments used in the study included measures of demographic, disease, family functioning, parent-adolescent interaction, transition, and adolescent adjustment variables.

Measures of Demographic and Disease-Related Variables

Demographic and disease-related variables were assessed through completion of file review (see Appendix A) and a *General Information Form* (GIF) by the primary parent and the adolescent (see Appendices B-C). The GIF took adolescents and parents about 10-15 minutes to complete. Information regarding the child's factor level or VWD type, number of spontaneous bleeds, number of prolonged bleeds, number of hospital admissions, number of phone calls to nursing services or the hospital, number of blood transfusions, self-infusion practices, and number of medications administered were obtained through medical file review. For those patients who received medical care at a hospital other than St. Christopher's, medical review and consent forms for release of medical information were sent to their respective hospitals. A total of seven adolescents who participated in the study received care from a hospital other than St. Christopher's. Consent was obtained from six of those families to request medical information from their respective hospitals and file reviews were returned for five families. Information pertaining to treatment and contact between the family and the medical staff was limited to the past year. Chart review for each patient took place a minimum of three weeks after measures were completed or were sent out immediately to the other hospitals.

Disease Severity and Illness Stress

Based on methods specified in the literature, level of medical severity was calculated differently depending on disease population. For adolescents with hemophilia, those with mild hemophilia were defined as individuals who did not

experience spontaneous hemorrhages, had significant hemorrhage only after major trauma and had circulation level of Factor VIII or IX ranging from 5-30%. Children were defined as having moderate hemophilia if they experienced rare spontaneous hemorrhages, had significant hemorrhage only after minor trauma, and had circulation level of Factor VIII or IX ranging from 2-5%. Severe hemophilia was defined by children who experienced spontaneous hemorrhages into soft tissues, joints, and muscles, had bleeding after any type of trauma or minor surgery, and had circulating level of Factor VIII or IX between 0-1%.

For those with VWD, medical severity was calculated by disorder subtype, such that those individuals with Type 2N and 3 VWD were considered as having more severe VWD than those with Type 1 VWD. Those adolescents with VWD who experienced low VWF and factor VIII levels, similar to that of children with hemophilia, were considered to have more severe VWD.

In addition physicians, parents, and adolescents were asked to rate how severe they felt each adolescent's disease was on a 5-point Likert type scale. Physician-, parent-, and adolescent-rated disease severity were used separately as measures of disease severity for both groups. A second measure was used to assess perception of disease interference on daily life. Again physician-, parent-, and adolescent-rated disease interference were used as separate measures. Finally, a measure of illness stress was the average additive score comprised of number of absences from school during the previous school year, number of calls to hospital or nursing services, number of blood transfusions, and number of hospital admissions (Drotar et al., 1991). This measure of

illness stress was calculated in the same way across both the hemophilia and VWD groups in order to create a common measure of illness stress.

Family Functioning

Adolescents and their parents were asked to complete the McMaster Family Assessment Device (FAD) (Epstein, Baldwin, & Bishop, 1983) to evaluate family functioning. The FAD is a 60-item scale that assesses 7 domains of family functioning: general functioning, problem solving, communication, roles, affective responsiveness (expression of positive emotion), affective involvement, and behavioral control. Designed as a screening measure to indicate family problems, the FAD is rated by respondents using a 4-point Likert-type scale. Internal-consistency of the seven scales on the FAD has been reported as being moderate (ranging from .72 to .92), while test-retest estimates were .66 to .73. The FAD has been found to delineate between clinic and non-clinic families (Epstein et al., 1983). Lower scores on the FAD imply better family functioning. In the study the parent- and adolescent-rated general family functioning scale was used to assess family functioning. The FAD generally takes parents and adolescents 15-20 minutes to complete.

Parent-Adolescent Interaction

In addition to measuring family functioning using the FAD, the System for Coding Interactions and Family Functioning (SCIFF; Lindahl & Malik, 2000) was utilized to provide a behavioral assessment of family conflict, parenting style, and problem solving. Previous research completed with the SCIFF has been conducted with

a variety of family structures (1-and 2-parent families) and with diverse ethnic groups, including European-American, Hispanic-American, and African-American families (Lindahl, 1998; Lindahl & Malik, 1999). Originally used in a study that assessed family functioning in a multiethnic sample of boys ages 7 to 12 years with externalizing behavioral disorders, the SCIFF is based on theories including systemic family theory, structural family theory, and social learning theory (Minuchin, 1974). Within this study Lindahl (1998) was able to classify boys into the correct diagnostic category [no psychological/behavior problems, Attention-Deficit Hyperactivity Disorder (ADHD), Oppositional Defiant Disorder (ODD), and both ADHD and ODD] with 90% accuracy using the SCIFF coding system in evaluating family interaction. Of note is that the scales of coercive parenting and conflicted relationships distinguished clinic from non-clinic families. Thus codes in the system assess the structural or organizational nature of the family, capture patterns of dysfunction among family members, and evaluate affective relationships within the family (Lindahl & Malik, 2000).

In order to code family interactions, families in the study were asked to engage in a videotaped conversation about a recent argument that involved the adolescent and other family members present. Families were instructed to review the topic of the conflict, describe the role that each family member played in the conflict, and discuss how they would resolve similar disagreements in the future (SCIFF; developmental task). In addition the families were asked to have a second discussion about a problem or conflict the family had recently experienced related to the adolescent's medical care or hemophilia/VWD diagnosis (SCIFF; medical task).

Four family level codes included in the measure (Negativity/Conflict, Positive Affect, Cohesiveness, Focus of Problem) are rated on a 5-point Likert type scale ranging from 1 (*very low*) to 5 (*high*). Codes on the SCIFF provide ratings of affective family interaction, tension within the family, family cohesiveness, and respect among family members. The additional two scales, Parenting Style and Alliance Formation, are categorical ratings. Four types of parenting styles rated are: Democratic, Hierarchical/Autocratic, Lax and Inconsistent. Parenting Style assesses communication, decision-making, parental authority, and family members contributions to the family unit. Alliance formation indicates alliance between family members and labels families as exhibiting Balanced, Marital, Parent-Child, or Disengaged relationships. In addition to family codes, parents and children also receive individual ratings. Parental marital communication and responsiveness is rated, and each parent receives a rating for their interaction with the child (Rejection/Invalidation, Coerciveness, Triangulation, Withdrawal, and Emotional Support). Children receive three ratings that assess their affective state (Anger/Frustration, Sadness, and Positive Affect) and two behavioral ratings (Withdrawal and Opposition/Defiance). Since many of the families included in the study did not have two parents present during data collection, scales assessing triangulation and marital relationships were not coded.

Previous research using the SCIFF with other populations has indicated that trained research assistants have been reliable and effective raters. Training of research assistants (2 assistants) involved a review of the SCIFF manual to discuss and familiarize raters with the coding system. Then several tapes rated by the primary investigator were reviewed with the raters and each item was discussed. As a group the

raters then rated criterion tapes and ratings were compared and discussed in a group supervision format with the primary investigator. Each tape was reviewed by two different raters. Weekly training meetings were conducted to discuss discrepancies between raters (Lindahl & Malik, 2000). If there were significant discrepancies between raters, based on low inter-rater agreement that was calculated for each tape, the interaction was reviewed in training meetings with the primary investigator. If raters were unable to agree on a rating during the training meeting the primary investigator provided separate ratings for the interaction. Then the scores from the two raters and the primary investigator were averaged. Training continued for 3 weeks until the raters had achieved an adequate reliability (.66 percentage agreement at week 2, .80 percentage agreement at week 3). Raters were closely supervised after weekly training ended and reliability between raters was calculated after every 6 coded interactions. Reliability continued to remain at or above .70. Tapes were labeled using a participant number for confidentiality and tapes were destroyed after they had been reviewed for relevant data. One participating family completed the video interaction in Spanish. The tape was then translated by one of the assistants who was fluent in Spanish in order for it to be coded.

Validation studies have demonstrated adequate interrater reliability for a majority of the individual codes. Overall Pearson correlations ranged from .70 to .80 for all scales except Parental Triangulation (r = .60-.65) and Mother Withdrawal (r = .50), indicating marginal reliability for these two scales. Child codes were found to have satisfactory reliability (r = .65-.80). In studies assessing validity of the SCIFF, results have suggested that SCIFF code of negativity was positively correlated with

family conflict and negatively correlated with positive family affect and cohesiveness. In addition families with higher rated family conflict, as measured by other family functioning scales, had higher scores on measures of rejection, coercion, triangulation, and lower emotional support scores. While family cohesion was negatively associated with rejection, coercion, and triangulation, it was positively associated with support (Malik & Lindahl, 1999). Parent-adolescent interaction was measured using the family scales of negativity/conflict and positive affect, on both the developmental and the medical task (e.g., negative SCIFF parent-adolescent interaction, positive SCIFF parent-adolescent interaction, medical negative SCIFF parent-adolescent interaction, and medical positive SCIFF parent-adolescent interaction). Combined, the developmental and the medical task on the parent-adolescent interaction took about 30 minutes for each family.

Transition Measure

In order to assess adolescents and parents' feelings about and concerns with transition from pediatric to adult health care systems, a comprehensive measure assessing transition was used (see Appendices D-E). In research with a sample of adolescents with SCD, Telfair and colleagues (1994) have utilized a transition questionnaire, which was modified for the current study to address concerns specific to hemophilia and VWD. In addition questions were reformatted so that adolescents and parents responded using a 5-point Likert type scale. The questionnaire included scales assessing, Transition Concerns and Feelings, Transition Programs, and Adolescent Medical Self-Care. Questions on the measure were appropriate for adolescents with

hemophilia and VWD as questions are specific to disease complications, medical treatments, and transition issues of adolescents with blood disorders. The transition measure took 20 to 30 minutes to complete.

Transitioning concerns and feelings. Included in the questionnaire was a list that asked participants to indicate concerns they had about transitioning into adult health care. Some potential concerns included: "I will not have enough information to know what to expect", "I wonder if I will have to take responsibility for myself", "I have concerns about being treated like an adult", and "I am scared about getting to know new health care providers". Adolescents and parents were asked whether concerns were 0 (*not true*) to 4 (*very true*) for them. In addition adolescents and parents were asked to describe how they might have felt if they learned that they/or their child was going to be transitioned. Again participants were asked to rate if the feelings listed would be 0 (*not true*) to 4 (*very true*) for them. For both the concerns and the feelings section four additive scores were computed, with higher scores indicating more parent- and adolescent-rated concerns and more parent-and adolescent-rated negative feelings regarding transition.

Finally, physicians, parents, and adolescents were asked to predicted adolescent and parent transition readiness using a 5-point Likert type scale.

Transition program. In addition to assessing individuals' thoughts and feelings about transition, Telfair et al. (1994) asked adolescents if a transition program would make the transition to adult health care easier, if they would like additional support

during the transition time, if a program should give them more control in the process, and if a program should allow them to meet other adults with hemophilia who have transitioned successfully. Information about what type of programming should be offered (i.e., information about adult care centers, problem solving strategies, meeting adult health care providers) was included. Adolescents and parents were asked to rate the importance of a transition program and indicate those things they would like to see addressed in a transition program using a 5-point Likert type scale. Additive scores was computed, with a higher score indicating greater parent- and adolescent-rated desire/need for a transition program.

Adolescent medical independence and self-efficacy. In the third section of the transition questionnaire, an assessment of functional skills explored how adolescents and parents handled disease symptoms, how they responded to medical problems ranging from mild to severe, how often they utilized health care professionals, and how adherent they were with their medial regimen. Another measure in this section investigated adolescent medical independence, by having parents and adolescents rate how responsible and independent the adolescent was in their medical care using a 5-point Likert type scale. In this portion of the questionnaire, adolescents indicated if they made their own health care appointments, if they knew who to contact if they needed medical help, if they were responsible for their medications, and if they took medications without parental reminders. Parent- and adolescent-rated medical independence scores were calculated, with higher scores indicating more medical independence and responsibility. Finally the self-efficacy section of the measure

required adolescents and parents to indicate how confident adolescents were in being able to handle daily management of their disease. Parent- and adolescent-rated self-efficacy scores were calculated, with higher scores indicating greater self-efficacy.

Adolescent Adjustment Measures

A number of measures including adolescent behavior problems on the Behavioral Assessment System for Children (BASC, parent and adolescent report) and the Brief Symptom Inventory (BSI; Derogatis & Spencer, 1982) were used to assess adolescent adjustment to hemophilia and VWD.

The Behavioral Assessment System for Children (BASC; Reynolds & Kamphaus, 1992) is a measure that assessed personality, self-perception, and behavior of children and adolescents across multiple settings. For the study, parent (PRS) and adolescent (SRP-A) ratings of adolescent behavior were gathered from adolescents age 12 to 18 years and their respective parents. The parent version of the BASC included a rating form for parents of adolescents, age 12 to 18 years, and included 138 questions. Parents were asked to rate their adolescent on a 4-point sale indicating if their adolescent 0 (n*ever*) to 3 (*almost always*) engaged in a behavior. From parental ratings, eleven Clinical Scales (Aggression, Anxiety, Attention Problems, Atypicality, Conduct Problems, Depression, Hyperactivity, Learning Problems, Somatization, and Withdrawal) and four Adaptive scales were computed (Adaptability, Leadership, and Social Skills). In addition composite scores that summarized child behavior, like Externalizing Problems (Hyperactivity, Aggression, and Conduct Problems), Internalizing Problems (Anxiety, Depression, and Somatization), School Problems

(Attention Problems and Learning Problems), and Adaptive Skills (Adaptability, Social Skills, and Leadership), were recorded. A behavioral symptoms index (BSI) assessing overall behavior is included. Reliability and validity scales, which evaluate parental inconsistency and negative responding, provide information regarding the accuracy of parent report. Internal consistency reliabilities for the PRS ranged from .85 to .90, while test-retest reliability for the adolescent version of the PRS was around .70. Scales on the PRS correlate with similar scales on other measures of child adjustment (CBCL). For analyses, the Behavioral Symptom Index (BSI), a composite score of the overall level of problem behavior in the adolescent was used as a measure of parent-rated adolescent adjustment. Higher scores on the BSI are associated with increased problem behavior.

Similar to the parent version, the self-report BASC included 118 statements that assessed clinical problems (Anxiety, Attitude to School, Attitude to Teachers, Atypicality, Depression, Locus of Control, Sensation Seeking, Sense of Inadequacy, Social Stress and Somatization) and adaptive behavior (Interpersonal Relations, Relations with Parents, Self-Esteem, and Self-Reliance). Adolescents rated their behavior by answering true or false to a series of statements about their feelings, beliefs, and behavior. Composite scores included Clinical Maladjustment (Anxiety, Atypicality, Locus of Control, Social Stress, and Somatization), School Maladjustment (Attitude to School, Attitude to Teachers, and Sensation Seeking), and Personality Adjustment (Relations with Parents, Interpersonal Relations, Self-Reliance, and Self-Esteem). Finally the Emotional Symptoms Index (ESI), a global rating of serious emotional disorders, was reported. Similar to parent measures, the self-report also

included reliability and validity measures. The SRP has an internal consistency of .80 for the adolescent version, and test-retest reliability was .76. For analyses the Emotional Symptoms Index (ESI), a global measure of serious emotional disturbance was used to measure adolescent-rated adjustment. Higher scores on the ESI are indicative of more serious emotional problems. Both parents and adolescents typically finished the BASC in a 20 to 30 minute time frame.

In addition to the BASC, the BSI (Derogatis & Spencer, 1982) was administered to all adolescents included in the sample, to obtain ratings of adjustment for adolescents ages 19-21 who could not complete the BASC because of their age (see Appendix N). The BSI is a 53-item self-report scale designed to assess psychological symptoms in medical and non-medical populations. Individuals completing the measure were asked to respond to questions using a 5-point Likert-type scale that ranged from 0 (*not at all*) to 4 (*extremely*) based on how they had been feeling in the past week. Nine subscales were measured including somatization, obsessive compulsive, interpersonal sensitivity, depression, anxiety, hostility, phobic anxiety, paranoid ideation, and psychoticism. In addition three global scales, the global severity index (GSI), the positive symptom distress index (PSDI), and the positive symptom total (PST), were calculated from items included in the measure. Scores on all 53 items are involved in computing the GSI total score. This GSI scale was used for the study as a global measurement of adjustment. Internal consistency for the nine scales in the measure range from .71 to .85, with the GSI test-retest reliability at .90 (Derogatis & Spencer, 1982). The BSI took adolescents 10 to 15 minutes to complete.

Procedure

All data were collected through standard procedures at the St. Christopher's Hospital for Children, the National Hemophilia Foundation, and Drexel University. Protocols for the data collection, as well as subsequent changes in participant recruitment, were approved by the Institutional Review Board of Drexel University. Parents of children with hemophilia or VWD and who met criteria for the study were identified and approached by the medical staff in clinic, during the NHF family weekend, or through mailings. If families were interested in participating in the study, they were introduced to the research assistant or provided with information on how to contact the research assistant. Data collection was either completed following a medical visit, during the NHF family weekend, or through a homevisit that was arranged by the research assistant. During these meetings informed consent of the parents and adolescent assent were obtained prior to the beginning of the study. Following consent/assent, the research assistant or primary investigator administered the questionnaires to the parent and the adolescent through interview format. Approximately one and one-half hour was needed by each family to complete the questionnaires. After completion of the forms parents and adolescents were asked to engage in a videotaped conversation about a recent family argument surrounding a developmental and medical task that involved the family members present for the study (SCIFF developmental and medical task). Families were instructed to review the topic of the conflict, describe the role that each family member played in the conflict, and discuss how they would resolve similar disagreements in the future. It took families, on

average, two hours to complete all measures for the study. All measures used in the study were labeled using a participant number for reasons of confidentiality and videotapes were destroyed after they were coded for relevant data.

Approach to Data Analysis

A total of 22 families were recruited for the proposed study, with 10 families in the hemophilia group and 12 families in the VWD group. A SPSS database was constructed including all relevant clinical and demographic information. The sample was described with respect to demographic and clinical variables. All data were analyzed using a probability level of .05 as the standard measure of significance. With a medium effect size (.50) and alpha set at .05, power was calculated at .1994 for the first hypothesis, .210 for the second hypothesis and, .316 for the third hypothesis. Given concerns regarding low power, findings of marginal significance ($p < .10$) were examined cautiously.

Approach to Preliminary Analyses

Given concerns about the small sample size, the group of adolescents with hemophilia and group of adolescents with VWD were combined to evaluate preliminary analyses. Preliminary analyses included exploration of the relationships among demographic, disease, family functioning (FAD), parent-adolescent interaction (SCIFF), and child adjustment (BASC; BSI) measures. Those demographic and disease

variables found to have significant associations were controlled in regression analyses as appropriate. Zero-order correlations were computed among family variables.

Approach to Analysis of the First Hypothesis

Hypothesis IA. Independent sample t-tests were run to evaluate the hypothesis that adolescents with hemophilia and adolescents with VWD would show differences on family functioning, parent-adolescent interaction (SCIFF), and adolescent adjustment variables (BASC, BSI).

Hypothesis IB. Independent samples t-tests were run to evaluate the hypothesis that adolescents with hemophilia and adolescents with VWD would show differences on measures of physician-, parent-, and adolescent-rated disease severity, physician-, parent-, and adolescent-rated disease interference, and illness stress. Initially it was planned that disease severity and disease interference variables would be collapsed among raters to create a composite score. Since initial correlations suggested that physician-, parent-, and adolescent-rated disease severity and disease interference were not significantly correlated, ratings were evaluated separately.

Approach to Analysis of the Second Hypothesis

The second hypothesis stated that the association between disease severity (i.e., physician-, parent-, and adolescent-rated disease severity, physician-, parent-, and adolescent-rated disease interference, and illness stress) and adolescent adjustment

(BASC; BSI) would be moderated by family functioning and parent-adolescent interaction variables (SCIFF). The criteria for moderation were not met for the variables of physician-, parent-, and adolescent-rated disease severity, physician- and adolescent-rated disease interference, illness stress, or parent-and adolescent-rated BASC. The criteria for moderation were met for the variables of parent-rated disease interference and adolescent-rated BSI total problems, since parent-rated disease interference was significantly correlated with adolescent-rated BSI total problems and parent-rated family functioning. As a result of these preliminary correlations only one regression analysis was run to explore the association between parent-rated disease interference and adolescent-rated BSI total problems. This regression analysis was run based on the procedure outlined in Holmbeck (1997; 2002) and Baron and Kenny (1986). Prior to running the analysis, all continuous variables included in the regression were centered. According to procedures outlined in Holmbeck (2002) the variables of parent-rated family functioning and adolescent-rated BSI total problems were centered by subtracting the grand mean from each individual score so that simple regression slopes could be calculated when a significant moderator relationship was found. Within the hierarchical regressions adolescent-rated BSI total problems was entered as the dependent variable. Control variables were entered on the first step of the regression equation to control for variance associated with this variable based on the correlations computed during the preliminary analyses. Parent disease-rated interference was entered on the second step, parent-rated family functioning was included on the third step, and the product of parent-rated disease interference and parent-rated family

functioning was entered on the fourth step. The interaction was examined as a test of the moderator hypothesis.

Approach to Analysis of the Third Hypothesis

All transition readiness scores were in the moderate range (3), resulting in reduced variance and an inability to create high and low transition readiness groups. Most variables were normally distributed, however given the small sample size transition readiness scores were evaluated as continuous variables (physician-rated adolescent transition readiness, $M = 3.50$, $SD = 1.00$, range = 1-5; physician-rated parent transition readiness, $M = 3.60$, $SD = .75$, range = 2-5; parent-rated adolescent transition readiness, $M = 2.59$, $SD = 1.10$, range = 0-4; parent-rated self transition readiness, $M = 2.55$, $SD = 1.06$, range = 0-4; adolescent-rated self transition readiness, $M = 2.24$, $SD = 1.04$, range = 0-4; adolescent-rated parent transition readiness, $M = 2.62$, $SD = .60$, range = 2-4). In addition, transition readiness scores were planned to be collapsed among raters to create a composite score. Initial correlations suggested that physician-, parent-, and adolescent-rated perception of transition readiness were not significantly correlated; transition readiness scores were evaluated separately. Four regression analyses were run in a stepwise fashion with physician- and parent-rated transition readiness (parent and adolescent) entered as the dependent variables and predictor variables entered based on significant correlations from preliminary analyses. There were no significant correlations among demographic, disease, family functioning, parent-adolescent interaction, or adjustment variables and adolescent-rated self and

parent transition readiness. Therefore regression analyses were not run with adolescent-rated self and parent transition readiness.

CHAPTER 4: RESULTS

Sample Description

Hemophilia Sample

The demographic and disease composition of the sample was evaluated (see Tables 1 and 2). There were a total of 10 adolescents included in the hemophilia group with a mean adolescent age of 15.59 years (*SD* = 3.14, range = 13-21). One hundred percent of the adolescents in the hemophilia sample were male. Ethnic backgrounds included 50% Caucasian, 40% African-American, and 10% who did not identify with any of the ethnic groups listed.

The 10 parents within the hemophilia group were comprised of biological mothers (80%) and biological fathers (20%). Given the small sample size the multiple informants were collapsed to form one parent group. Parents reported a mean age of 40.90 years (*SD* = 5.67, range = 35-51). Similar to adolescent ethnic background, a majority of the parents were Caucasian (60%), while others reported ethnicities including African-American (40%). Five parents in the sample reported that they were currently married (50%), while others were separated (30%), never married (10%), or remarried (10%). Overall 50% of parents were married, while 50% were not. Median family income was between $50,000-$74,999 (50%), with 20% reporting incomes below $19,999 and 30% reporting incomes above $75,000.

In the hemophilia sample, one adolescent did not consent to have medical records released. All other file reviews were completed. Out of those with complete medical information 89% had a diagnosis of hemophilia 8A and 11% had a diagnosis of

hemophilia 9B. With regard to disease severity, 22% of the adolescents with hemophilia had moderate disease (factor levels 2-5%) and 78% had severe disease (factor levels < 2%). Medical file review suggested that 56% of the adolescents experienced spontaneous bleeds (*M* = 1.89, *SD* = 2.52, range = 0-6) and 33% experienced prolonged bleeds (*M* = 1.00, *SD* = 1.73, range = 0-3).

Few adolescents with hemophilia experienced a hospitalization during the past year (22%; *M* = .22, *SD* = .44, range = 0-1). Adolescents attended at least two outpatient clinic visits (*M* = 2.44, *SD* = 1.94, range = 1-7), while 25% reported that they used homecare services. Parents reported a mean of 14.8 school absences (*SD* = 37.09; range = 0-120), while file review suggested a mean of .67 phone calls to clinic (*SD* = .33, range = 0-1) and .33 transfusions (*SD* = .58, range = 0-1).

One hundred percent of the sample reported taking daily prescriptions, with a mean number of 1.89 medications (*SD* = 1.16, range = 1-4). File review suggested that adolescents with hemophilia used factor replacement products approximately 52.33 times (*SD* = 89.78, range = 0-156). Parents reported that adolescents using factor replacement products administered the medication either as needed (PRN = 30%) or were on prophylaxis treatment (more than 3 times per week = 20%, 1-3 times per week = 30%, once every two weeks = 10%, once every month = 10%). Parents reported that adolescents with hemophilia learned to self-infuse factor replacement products at age 11.9 years (*SD* = 2.96 years, range = 7-17 years), while adolescents reported that the mean age that they learned to self-infuse was 12.11 years (*SD* = 3.1 years, range = 7-16 years). Thirty-six percent (36%) of parents reported that their adolescents self-infused some of the time, while 27% of parents indicated that their adolescent never or rarely

infused and 36% of parents indicated that their adolescent infused most or all of the time. Reasons why adolescents did not infuse, according to parents, was because 14% were learning to infuse, 26% were in pain/could not find a vein, 14% had port access or 43% found it easier for a parent to infuse. Thirty-six percent (36%) of adolescents indicated that they infuse all of the time, with 36% of adolescents reporting that they never or rarely infuse and 27% of adolescents replying that they infuse some to most of the time. Reasons why adolescents did not infuse, according to adolescents, was because 40% were in pain/could not find a vein, 40% did not want to infuse, or 20% found it easier for parent to infuse. Twenty-percent of the adolescents sampled had another medical diagnosis, including 10% with hepatitis C and 10% with asthma.

Parents reported that 10% of adolescents in the hemophilia group had psychological difficulties. Parents also reported that 30% of the adolescents experienced learning difficulties and received special education accommodations at school.

VWD Sample

The demographic and disease composition of the sample was evaluated (see Tables 1 and 2). There were a total of 12 adolescents included in the sample with a mean adolescent age of 16.08 years (*SD* = 2.27, range = 13-21). There were more females included in the VWD group (83%) than males (17%). Ethnic backgrounds included 42% Caucasian, 17% African-American, 33% Latino, and 8% who did not identify with any of the ethnic groups listed.

The 12 parents within the sample were comprised of biological mothers (92%) and grandparents (8%). Given the small sample size multiple informants were collapsed to form one parent group. Parents reported a mean age of 42.33 years (*SD* = 7.11, range = 31-54). Similar to adolescent ethnic background, a majority of the parents were Caucasian (42%), while others reported ethnicities including African-American (17%), Latino (25%), and other ethnicities (17%). Over half of parents in the VWD sample reported that they were currently married (58%), while 25% were divorced, 8% were separated, and 8% were never married. Overall 58% of parents were married, while 41% were not. Median family income was between $10-19,999 (25%), with other incomes below $10,000 (17%), between $20-34,999 (17%), between $50-74,999 (17%), and above $75,000 (25%).

VWD diagnosis types varied with physicians reporting that 82% of adolescents with VWD had Type 1, 9% had Type 2, and 9% had Low Levels VIII. One adolescent did not consent to have medical records released in the VWD sample, thus medical file review was not completed on this participant. Medical file review suggested that 18% of the sample experienced spontaneous bleeds (*M* = .18, *SD* = .40, range = 0-1) and 55% experienced prolonged bleeds (*M* = 1.45, *SD* = 1.75, range = 0-5).

Approximately half of adolescents sampled had experienced a hospitalization during the past year (56%; *M* = .64, *SD* = .92, range = 0-3). Adolescents had attended at least two outpatient clinic visits (*M* = 1.73, *SD* = 1.10, range = 0-4), while none reported that they used homecare services. Parents reported a mean of 22.33 school absences (*SD* = 28.17; range = 0-88), while file review suggested a mean of 1.45 phone calls to clinic (*SD* = 2.70, range = 0-9) and .18 transfusions (*SD* = .60, range = 0-2).

Eighty-eight percent (88%) of the sample reported taking daily prescriptions, with a mean number of 3.18 medications (*SD* = 3.06, range = 0-10). File review suggested that factor replacement was prescribed PRN for one adolescent in the VWD group however file review and parent report did not indicate that the factor replacement was used during the past year with this adolescent. Forty-one percent (41%) of the adolescents sampled had another medical diagnosis, including 8% with hepatitis C and 33% with asthma.

Parents reported that 25% of adolescents in the VWD group had psychological difficulties. Parents also reported that 17% of the adolescents experienced learning difficulties and received special education accommodations at school.

Preliminary Analyses

Descriptive Information on Measures for Hemophilia Sample

Descriptive information on each measure for adolescents with hemophilia is presented in Table 3. Physicians indicated that the mean for disease severity was in the moderate range (*M* = 3.11, *SD* = .93, range = 2-4); parents reported that the mean for disease severity was also in the moderate range (*M* = 3.70, *SD* = 1.42, range = 1-5); adolescent-rated disease severity also had a mean within the moderate range (*M* = 3.50, *SD* = 1.35, range = 1-5). Physicians indicated that the mean impact of disease interference on daily functioning was in the low range (*M* = 2.44, *SD* = 1.13, range = 1-4); parents reported that the mean impact of disease interference on daily functioning was also in the low range (*M* = 2.00, *SD* = 1.05, range = 1-4). Adolescents with

hemophilia rated that the mean impact of disease interference on daily function was very low to low (*M* = 1.90, *SD* = 1.10, range = 1-4). The mean for illness stress was 3.97 (*SD* = 9.87, range = 0-30.25).

Parents reported a mean for general family functioning of 1.62 (*SD* = .34, range = 1.08-2.00), while adolescents with hemophilia indicated a mean for general family functioning of 1.70 (*SD* = .48, range = 1.25-2.58). Both parents and adolescents with hemophilia reported that, on average, they agreed with statements associated with better family functioning. On negative SCIFF parent-adolescent interaction the mean was 2.50 (*SD* = 1.15, range = 1.5-5), suggesting that a majority of the families of adolescents with hemophilia displayed low (2) levels of tense or conflicted interactions. The mean for medical negative SCIFF parent-adolescent interactions was 1.61 (*SD* = .60, range = 1-2.5), indicating that families of adolescents with hemophilia displayed very low (1) to low (2) levels of tense or conflicted interactions. On positive SCIFF parent-adolescent interactions the mean was 2.39 (*SD* = 1.19, range = 1-4), suggesting that a majority of the families of adolescents with hemophilia displayed low (2) levels of positive affect. The mean for medical positive SCIFF parent-adolescent interaction was 2.61 (*SD* = 1.34, range = 1-5), indicating that families of adolescents with hemophilia displayed low (2) to moderate (3) levels of positive affect.

Results on the parent-rated BASC total behavior problems indicated *T*-scores within the average range (*M* = 53.14, *SD* = 9.26, range = 42-66), with 20% (*N* = 2) of parents reporting that their adolescent with hemophilia had clinically significant behavior problems (*T* scores $\geq$ 60) compared to a normative sample. Results on the adolescent-rated BASC total emotional problems indicated *T*-scores within the average

range (*M* = 43.43, *SD* = 5.26, range = 38-53), with none of adolescents with hemophilia reporting that they experienced clinically significant emotional problems (*T* scores ≥ 60) compared to a normative sample. The adolescent-rated BSI total behavioral symptom index mean was .42 (*SD* = .46, range = .06-1.66). Compared to the normative sample a BSI index mean of .42 is within the normal range for male adolescents (*T*-score = 44). One adolescent with hemophilia had a score on the BSI in the clinical range (*T* ≥63) (5%).

Descriptive Information on Measures for VWD Sample

Descriptive information on each measure for adolescents with VWD is presented in Table 4. Physicians indicated that the mean for disease severity was in the moderate range (*M* = 3.00, *SD* = .45, range = 2-4); parents reported that the mean for disease severity was in the low range (*M* = 2.50, *SD* = 1.00, range = 1-5); adolescent disease severity also had a mean in the low range (*M* = 2.33, *SD* = 1.15, range = 1-4). Physicians indicated that the mean impact of disease interference on daily functioning was in the moderate range (*M* = 3.09, *SD* = .54, range = 2-4); parents reported that the mean impact of disease interference on daily functioning was in the low range (*M* = 2.58, *SD* = 1.56, range = 1-5). Adolescents with VWD rated the mean impact of disease interference on daily function as being low (*M* = 2.17, *SD* = .94, range = 1-4). The mean for illness stress was 6.56 (*SD* = 8.48, range = .75-25.50).

Parents reported a mean for general family functioning of 1.74 (*SD* = .38, range = 1.08-2.50); adolescents with VWD indicated a mean for general family functioning of 1.82 (*SD* = .37, range = 1.25-2.33). Both parents and adolescents with VWD reported

that, on average, they agreed with statements associated with better family functioning. On negative SCIFF parent-adolescent interaction the mean was 2.04 (*SD* = 1.59, range = 1-5), suggesting that a majority of the families of adolescents with VWD displayed low (2) levels of tense or conflicted interactions. The mean for medical negative SCIFF parent-adolescent interaction was 1.64 (*SD* = .92, range = 1-4), indicating that families of adolescents with VWD displayed very low (1) to low (2) levels of tense or conflicted interactions. On positive SCIFF parent-adolescent interaction the mean was 3.55 (*SD* = 1.27, range = 1-5), suggesting that a majority of the families of adolescents with VWD displayed a moderate (3) level of positive affect. The mean for medical positive SCIFF parent-adolescent interaction was 3.18 (*SD* = 1.15, range = 1-5), indicating that families of adolescents with VWD displayed moderate (3) levels of positive affect.

Results on the parent-rated BASC total behavior problems indicated *T*-scores within the average range (*M* = 52.70, *SD* = 13.58, range = 36-80), with 25% (*N* = 3) of parents reporting that their adolescent with VWD had clinically significant behavior problems (*T* scores $\geq$ 60) compared to a normative sample. Results on the adolescent-rated BASC total emotional problems indicated *T*-scores within the average range (*M* = 49.10, *SD* = 9.39, range = 38-68), with one adolescent with VWD reporting that he experienced clinically significant emotional problems (*T* scores = 69) compared to a normative sample. The mean on the adolescent-rated BSI total behavioral symptom index was .60 (*SD* = .52, range = .04-1.66). Compared to the normative sample a BSI mean index score of .60 were within the normal range for male adolescents (*T*-score = 48) and for female adolescents (*T*-score = 49). Two adolescents with VWD had scores on the BSI in the clinical range (*T* = 62) (17%).

Association of Demographic Variables with Family Functioning, Parent-adolescent Interaction, and Adjustment Measures

To determine the appropriate control variables, Pearson and Spearman correlations were computed using the combined hemophilia and VWD samples to examine the association between demographic, disease, family functioning, parent-adolescent interaction, and adjustment variables (see Table 5).

Preliminary correlations and analyses with demographic variables. Spearman correlations suggested that males, adolescent minority status, and low family income were associated with higher disease severity/interference, lower family functioning, poorer adjustment, and higher transition readiness scores. Given significant correlations with adolescent gender, independent samples t-tests were run to assess for significant differences between males and females. Significant differences were found between the groups with males showing higher parent-rated disease severity [t (16) = 2.93, p = .010 (male M = 3.67, SD = 1.44; female M = 2.30, SD = .67)], lower positive SCIFF parent-adolescent interaction [t (18) = 1.60, p = .005 (male M = 2.32, SD = 1.17; female M = 3.89, SD = 1.02)], higher parent-rated adolescent transition readiness [t (20) = 3.25, p = .004 (male M = 3.17, SD = .72; female M = 1.90, SD = 1.1)], and higher parent-rated self transition readiness [t (20) = 3.11, p = .006 (male M = 3.08, SD = .79; female M = 1.90, SD = .99)]. No significant differences were seen between the groups on the measure of negative SCIFF parent-adolescent interaction.

Given significant correlations with adolescent ethnicity, independent samples t-tests were run to assess for significant differences between adolescents of Caucasian

ethnicity and adolescents of other ethnicities. Significant differences were found between the groups with adolescents of minority status reporting lower family functioning [t (19) = -2.63, p = .016 (Caucasian M = 1.53, SD = .32; Minority M = 1.95, SD = .40)]. No significant differences were seen between the groups on measures of adolescent-rated disease interference and physician-rated adolescent transition readiness.

Based on Pearson correlations, adolescent age showed no significant associations with disease, family functioning, parent-adolescent interaction, or adjustment measures. Decreased family income was found to be significantly associated with increased adolescent-rated BSI total problems (r = -.53, p = .010), increased adolescent-rated BASC emotional total problems (r = -.71, p = .001), decreased physician-rated adolescent transition readiness (r = .53, p = .016), decreased physician-rated parent transition readiness (r = .44, p = .050), decreased parent-rated adolescent transition readiness (r = .50, p = .019) and decreased parent-rated self transition readiness (r = .64, p = .001).

Correlations with disease variables. Higher parent-rated disease severity was associated with lower adolescent-rated family functioning (r = .45, p = .042), lower positive SCIFF parent-adolescent interaction (r = -.60, p = .005), and lower medical positive SCIFF parent-adolescent interaction (r = -.49, p = .028), while higher adolescent-rated disease severity was associated with lower positive SCIFF parent-adolescent interaction (r = -.49, p = .027). Higher physician-rated disease interference was associated with lower adolescent-rated family functioning (r = .68, p = .001) and

lower medical positive SCIFF parent-adolescent interaction (r = -.46, p = .050). Higher parent-rated disease interference was associated with higher adolescent-rated BSI total problems (r = .47, p = .026) and higher adolescent-rated disease interference was associated with lower adolescent-rated family functioning (r = .61, p = .003). No significant correlations were seen between disease severity, disease interference, and illness stress variables and parent-rated family functioning, negative SCIFF parent-adolescent interaction, medical negative SCIFF parent-adolescent interaction, parent-rated BASC total behavior problems or adolescent-rated BASC total emotional problems. Correlations evaluating associations between disease variables and family functioning, parent-adolescent interaction and adjustment indicated that higher disease severity and disease interference were related to lower family functioning, lower positive SCIFF parent-adolescent interactions, and poorer adjustment.

Correlations with family functioning, parent-adolescent interaction, and adjustment variables. Additional Pearson correlations were conducted to explore associations of family functioning and parent-adolescent interaction with adjustment variables. Decreased parent-rated family functioning was significantly associated with increased adolescent-rated BSI total problems (r = .52, p = .013), increased parent-rated BASC total behavior problems (r = .51, p = .035), and increased adolescent-rated BASC total emotional problems (r = .61, p = .009). Poorer family functioning was associated with poorer adjustment.

Examination of Hypotheses

Hypothesis One: That there would be significant differences in measures of disease, family functioning, parent-adolescent interaction, and adjustment between adolescents with hemophilia and those with VWD

Independent samples t-tests were run to determine differences between adolescents with hemophilia and those with VWD on disease, family functioning, parent-adolescent interaction, and adjustment measures (see Table 6). No significant differences between the groups were found on physician-rated disease severity [t (11) = .33, p = .748], physician-rated disease interference [t (11) = -1.58, p = .144], parent-rated disease interference [t (20) = -1.00, p = .328], adolescent-rated disease interference [t (20) = -.61, p = .546, or illness stress [t (15) = -.58, p = .573]. A significant difference between the groups was found on parent-rated disease severity [t (20) = 2.32, p = .031] and adolescent-rated disease severity [t (20) = 2.18, p = .041]. Parents of adolescents with hemophilia rated their adolescents as experiencing greater disease severity (M = 3.7) than parents of adolescents with VWD (M = 2.5) on a 5 point scale with 5 indicating severe disease. Adolescents with hemophilia also rated themselves as experiencing greater disease severity (M = 3.71) than adolescents with VWD (M = 2.45) on a 5 point scale with 5 indicating severe disease.

No significant differences were found between the groups on parent-rated family functioning [t (20) = -.76, p = .454] or adolescent-rated family functioning [t (19) = -.62, p = .542]. No significant differences were found between the groups on negative [t (18) = .72, p = .482], positive [t (18) = -2.08, p = .052], medical negative [t (18) = -.07,

p = .945] or medical positive [t (18) -1.03, p = .328] SCIFF parent-adolescent interaction variables.

No significant differences were found between the groups on adolescent-rated BASC total emotional problems [t (15) = -1.44, p = .171], adolescent-rated BSI total problems [t (20) = -.87, p = .396], or parent-rated BASC total behavior problems [t (15) = .08, p = .941]. Results suggested no significant differences between the groups on measures of disease, family functioning, parent-adolescent interaction, and adjustment, with the exception of a significant difference found on measures of parent- and adolescent-rated disease severity. Overall the hypothesis that adolescents with hemophilia would show higher disease severity, higher disease interference, lower family functioning, less positive parent-adolescent interaction, and poorer adjustment than adolescents with VWD was not supported. Given the lack of significant differences between adolescents with hemophilia and adolescents with VWD, the two groups were combined for the rest of the analyses.

Hypothesis Two: That the association between disease severity, disease interference, and illness stress and adolescent adjustment would be moderated by family variables

Initial correlations indicated that the criteria for moderation were not met for physician-rated disease severity, physician-rated disease interference, parent-rated disease severity, adolescent-rated disease severity, adolescent-rated disease interference, and illness stress, as these variables were not found to be correlated with any of the adjustment measures. Based on initial correlations, one moderator analysis was performed to examine whether parent-rated family functioning moderated the

association between parent-rated disease interference and adolescent adjustment (adolescent-rated BSI total problems), after controlling for the effects of family income. On step 1 of the regression analysis, total family income did account for a significant portion of the variance in predicting adolescent-rated BSI total problems [R^2 = .35, F change = 10.68, p = .004]. On step 2, parent-rated disease interference [R^2 change = .06, F change = 1.96, p = .178] did not independently account for a significant portion of the variance in predicting adolescent-rated BSI total problems. On step 3, parent-rated family functioning [R^2 change = .05, F change = 1.64, p = .217] did not independently account for a significant portion of the variance in predicting adolescent-rated BSI total problems. On step 4, interaction was tested and parent-rated family functioning was found to moderate the association between parent-rated disease interference and adolescent-rated BSI total problems [R^2 change = .12, F change = 4.81, p = .042].

Post-hoc analyses were completed to further evaluate parent-rated family functioning as a moderator of the association between parent-rated disease interference and adolescent-rated BSI total problems as suggested by Holmbeck (2002). Parent-rated family functioning, the variable hypothesized to moderate the relationship between parent-rated disease interference and adolescent-rated BSI total problems was recoded into two new conditional moderator variables for post-hoc analyses. To generate the "high" and "low" parent-rated family functioning variables, the standard deviation of parent-rated family functioning was subtracted, for the high score, and added, for the low score, from the centered score of parent-rated family functioning (Aiken & West, 1991). Once high and low parent-rated family functioning scores were obtained new interaction terms were created by multiplying high/low parent-rated

family functioning scores by parent-rated disease interference. Centered parent-rated disease interference, one of the new conditional moderators (high or low parent-rated family functioning) and the new interaction term were entered simultaneously into a regression equation. Results from these moderation analyses indicated that the interaction term of parent-rated disease interference and low parent-rated family functioning significantly predicted adolescent-rated BSI total problems [R^2 change = .46, F change = 5.12, p = .010.]. The interaction term of parent-rated disease interference and high parent-rated family functioning was also found to significantly predict adolescent-rated BSI total problems [R^2 change = .46, F change = .51, p = .010]. Post-hoc probing supported parent-rated family functioning as a moderator of the association between parent-rated disease interference and adolescent-rated BSI total problems. In addition, significance (t) tests for each simple slope indicated that the high and low parent family functioning variables did not independently account for a significant amount of the variance in predicting adolescent adjustment. Two equations were generated from these analyses so that simple regression lines could be plotted to interpret the significant interaction (See Figure 2):

For low parent family functioning (1 SD above the mean):

Adolescent-rated BSI total problems = .53(Low parent-rated family functioning) -.08(Parent-rated disease interference) + .34(Interaction of low parent-rated family functioning and parent-rated disease interference) + .23

t (18) = -.75, p = .462

For high parent family functioning (1 SD below the mean):

Adolescent-rated BSI total problems = .53(High parent-rated family functioning) + .17(Parent-rated disease interference) + .34(Interaction of high parent family functioning and parent disease interference) + .62

t (18) = 2.00, p = .060

Results suggest only partial support for this hypothesis as parent-rated family functioning was found to moderate the association between parent-rated disease interference and adolescent-rated BSI total problems when controlling for family income. Other disease severity, disease interference, and illness stress variables were not associated with adjustment.

Hypothesis Three: Prediction of physician-, parent-, and adolescent-rated transition readiness using family functioning, parent-adolescent interaction, and adolescent adjustment variables

Based on initial significant correlations, regression analyses were performed to examine the effects of demographic, disease, transition, and adjustment variables on transition readiness. Family income, adolescent ethnicity, physician-rated disease interference, parent-rated disease interference, adolescent-rated disease interference, parent-rated family functioning, adolescent-rated family functioning, positive SCIFF parent-adolescent interaction, parent-rated transition concern, parent-rated transition feeling, and adolescent-rated BSI total problems, were entered in a stepwise fashion into the first regression with physician-rated adolescent transition readiness entered as the dependent variable. Family income [*Beta* = .54, t = 2.61, p = .018, R^2 = .29, F (1, 17) =

6.82, p = .018] and parent-rated disease interference [*Beta* = -.46, t = -2.27, p = .038, R^2 = .46, F (1, 16) = 5.14, p = .038] were found to significantly predict physician-rated adolescent transition readiness, predicting 39% of the variance. Results suggested that adolescents with higher family income and lower parent-rated disease interference had higher physician-rated adolescent transition readiness scores. Other demographic, disease, family functioning, parent-adolescent interaction, transition, and adjustment variables did not significantly predict physician-rated adolescent transition readiness.

Family income, adolescent ethnicity, physician-rated disease severity, parent-rated disease interference, parent-rated family functioning, adolescent-rated family functioning, positive SCIFF parent-adolescent interaction, parent-rated transition concern, parent-rated transition feeling, and adolescent-rated BASC total emotional problems were entered in a stepwise fashion into the second regression with physician-rated parent transition readiness entered as the dependent variable. Adolescent ethnicity [*Beta* = -.53, t = -2.23, p = .044, R^2 = .28, F (1, 13) = 4.96, p = .044] was found to significantly predict physician-rated parent transition readiness, predicting 22% of the variance. Results suggested that adolescents of Caucasian ethnicity had higher physician-rated parent transition readiness scores. Family functioning, parent-adolescent interaction, transition, and adjustment variables did not significantly predict physician-rated parent transition readiness.

Adolescent gender, family income, parent-rated family functioning, adolescent-rated family functioning, positive SCIFF parent-adolescent interaction, parent-rated transition concern, parent-rated transition feeling, parent-rated adolescent self-efficacy, adolescent-rated transition concern, and adolescent-rated BSI total problems were

entered in a stepwise fashion into the regression with parent-rated adolescent transition readiness entered as the dependent variable. Adolescent gender [*Beta* = -.63, *t* = -3.40, *p* = .003, R^2 = .39, *F* (1, 18) = 11.56, *p* = .003], family income [*Beta* = .38, *t* = 2.13, *p* = .048, R^2 = .52, *F* (1, 17) = 4.54, *p* = .048], and parent-rated transition feeling [*Beta* = -.48, *t* = -2.44, *p* = .027, R^2 = .81, *F* (1, 16) = 5.96, *p* = .027], were found to significantly predict parent adolescent transition readiness, predicting 65% of the variance. Results suggested that adolescent males, adolescents with higher family income and adolescents with fewer parent-rated negative transition feelings had higher parent-rated adolescent transition readiness scores. Family functioning, parent-adolescent interaction, and adjustment variables did not significantly predict parent-rated adolescent transition readiness.

Adolescent gender, family income, parent-rated family functioning, adolescent-rated family functioning, positive SCIFF parent-adolescent interaction, parent-rated transition concern, parent-rated transition feeling, parent-rated adolescent medical independence, parent-rated adolescent self-efficacy, adolescent-rated transition feeling, and adolescent-rated BASC total emotional problems were entered in a stepwise fashion into a regression with parent-rated self transition readiness entered as the dependent variable. Family income [*Beta* = .69, *t* = 3.52, *p* = .003, R^2 = .47, *F* (1, 14) = 12.42, *p* = .003], adolescent gender [*Beta* = -.50, *t* = -3.32, *p* = .006, R^2 = .71, *F* (1, 13) = 11.01, *p* = .006], parent-rated transition concern [*Beta* = -.42, *t* = -2.69, *p* = .020, R^2 = .82, *F* (1, 12) = 7.24, *p* = .020], and parent-rated adolescent medical independence [*Beta* = .36, *t* = 3.93, *p* = .002, R^2 = .93, *F* (1, 11) = 15.47, *p* = .002], were found to significantly predict parent self transition readiness, predicting 93% of the variance. Results suggested that

adolescent males with higher family income, fewer parent-rated transition concerns, and higher parent-rated adolescent medical independence had higher parent-rated self transition readiness scores. Family functioning, parent-adolescent interaction and adjustment variables did not significantly predict parent-rated self transition readiness.

Exploratory Analyses

SCIFF Parent-Adolescent Interaction Measure

Exploratory analyses were also conducted to evaluate potential differences between adolescents with hemophilia and VWD on other variables included in the parent-adolescent interaction measure, as well as to assess differences between parent-adolescent interactions when families discussed a developmental versus a medical task. In the initial hypotheses the variables of negative, positive, medical negative, and medical positive SCIFF parent-adolescent interaction were used to evaluate differences between adolescents with hemophilia and adolescents with VWD. In an attempt to better understand the parent-adolescent interaction measure additional analyses were run to explore other variables included on the SCIFF.

Differences between groups on additional parent-adolescent interaction variables. Independent samples t-tests were run to explore differences between individuals with hemophilia and VWD on other variables of the SCIFF parent-adolescent interaction measure that were not included in the initial hypotheses (see Figure 3). Results suggested significant differences between the groups, with

adolescents with hemophilia having lower levels of cohesiveness [t (18) = -2.13, p = .047 (hemophilia M = 3.28, SD = 1.09; VWD M = 4.23, SD = .90)], higher adolescent withdrawal [t (9) = 2.55, p = .030 (hemophilia M = 2.33, SD = 1.25; VWD M = 1.23, SD = .41)], lower adolescent positive affect [t (18) = -2.53, p = .021 (hemophilia M = 2.33, SD = 1.37; VWD M = 3.82, SD = 1.25)], and lower medical adolescent positive affect SCIFF parent-adolescent interaction [t (18) = -2.16, p = .045 (hemophilia M = 2.28, SD = 1.28; VWD M = 3.50, SD = 1.24)]. There were no significant differences seen between the groups on other SCIFF parent-adolescent interaction variables (See Figure 3).

Differences between developmental and medical task on SCIFF parent-adolescent interaction variables. Paired sample t-tests were run to explore potential differences between how families (combined sample) interacted when asked to discuss a developmental task in comparison to how families interacted when asked to discuss a medical task (see Figure 4). Results showed significant differences between the developmental and medical tasks, with the developmental task having higher negative SCIFF parent-adolescent interaction [t (19) = 2.37, p = .029 (developmental M = 2.25, SD = 1.39; medical M = 1.63, SD = .78)], lower cohesiveness SCIFF parent-adolescent interaction [t (19) = -2.36, p = .029 (developmental M = 3.80, SD = 1.08; medical M = 4.05, SD = .96)], higher parental rejection SCIFF parent-adolescent interaction [t (19) = 2.35, p = .030 (developmental M = 1.95, SD = 1.07; medical M = 1.50, SD = .65)], and lower parent emotional support SCIFF parent-adolescent interaction [t (19) = -2.43, p = .025 (developmental M = 3.00, SD = 1.39; medical M = 3.53, SD = 1.29)]. Differences

between the medical and developmental tasks on adolescent oppositional behavior SCIFF parent-adolescent interaction approached significance [t (19) = 1.36, p = .055], with adolescents showing higher oppositional behavior on the developmental task (M = 1.88, SD = .96) than on the medical task (M = 1.58, SD = .63). Overall results suggest that families demonstrate better interactions when discussing a medical task as opposed to a developmental task.

Differences on SCIFF variables between families of different ethnicities. Independent samples t-tests were run to explore differences on parent-adolescent variables among ethnic groups. Results suggested that there were no significant differences on SCIFF variables between Caucasian and minority adolescents.

Differences between one- and two-parent families on SCIFF. Independent samples t-tests were run to explore differences between adolescents from one- and two-parent families on variables of the SCIFF parent-adolescent interaction measure, as there were concerns that one-parent families may interact differently than two-parent families on the interaction task based on previous literature (Uhlendorff, Arteit, & Krappmann, 2002). Results suggested no significant differences between the groups, with one- and two-parent families showing similar levels of negative interactions, positive interactions, cohesiveness, parental rejection, parental coerciveness, emotional support, parental withdrawal, adolescent anger, adolescent sadness, adolescent withdrawal, adolescent oppositional behavior, and adolescent positive interactions across both developmental and medical tasks of the SCIFF.

Transition Questionnaire

The transition measure for the current study included a number of scales that evaluated concerns about transition, feelings about transition, the importance of a transition program, essential components of a transition program, as well as two scales that evaluated the adolescents' medical independence and self-efficacy.

Differences between adolescents with hemophilia and VWD on transition measures. Independent samples t-tests were run to explore potential significant differences between adolescents with hemophilia and adolescents with VWD on transition measures. No significant differences were found on the measures. Trends were seen on measures of parent feelings about transition [t (20) = -2.08, p = .051], with parents of adolescents with VWD reporting more negative feelings about transition (hemophilia M = 1.35, SD = .66; VWD M = 2.03, SD = .83). Both adolescents with hemophilia and VWD, as well as their parents, reported few concerns and few negative feelings. Both groups indicated that they would be interested in a transition program, with parents of adolescents with VWD reporting a slight, but not significant, increase in interest. Parents of both adolescents with hemophilia and VWD indicated that adolescents were "sometimes" independent in their medical care and parents "sometimes" felt comfortable with their adolescents' ability to care for their medical needs.

Descriptive information for transition scales in hemophilia sample. The mean score for parent transition concern was 1.59 (*SD* = .52, range = .80-2.47), for parent transition feeling was 1.35 (*SD* = .66, range = .54-2.23), and for parent transition reasons was 2.84 (*SD* = .63, range = 1.88-4.00). Overall parents reported few concerns about their adolescent with hemophilia transitioning to adult health care, as parents reported that they were "unlikely" (1) to have the concerns listed on the questionnaire when transitioning occurred. Parents of adolescents with hemophilia also reported having few negative feelings about transition, with most parents reporting that it was "unlikely" (1) for them to feel negatively about the transition when it occurs. Parents of adolescents with hemophilia reported that on average they "may" (2) benefit from a transition program. The mean score for parent-rated adolescent medical independence was 2.23 (*SD* = .63, range = 1.48-4.0) and the mean score for parent-rated adolescent self-efficacy was 2.92 (*SD* = .66, range = 1.86-3.71). In general, parents reported that adolescents with hemophilia were "sometimes" (2) medically independent in taking care of their own health care needs and parents felt that adolescents with hemophilia were efficacious in their ability to care for their own health needs "sometimes" (2) to "most of the time" (3).

The mean score for adolescent transition concern was 1.78 (*SD* = .60, range = .67-2.67), for adolescent transition feeling was 1.51 (*SD* = .40, range = 1.0-2.15), and for adolescent transition reasons was 2.40 (*SD* = .68, range = 1.13-3.75). Overall adolescents with hemophilia reported few concerns about transitioning to adult health care, as they reported that they were "unlikely" (1) to have the concerns listed on the questionnaire when transitioning occurred. Adolescents with hemophilia also reported

having few negative feelings about transition, with most adolescents reporting that it was "unlikely" (1) for them to feel negatively about the transition when it occurs. Adolescents with hemophilia reported that on average they "may" (2) benefit from a transition program. The mean score for adolescent-rated medical independence was 2.35 (*SD* = .45, range = 1.83-2.93) and the mean score for adolescent-rated self-efficacy was 2.94 (*SD* = .54, range = 2.29-3.93). In general adolescents with hemophilia reported that they were "sometimes" (2) independent in taking care of their own health care needs and that they were efficacious in their ability to care for their own needs "sometimes" (2) to "most of the time" (3).

Descriptive information for transition scales in VWD sample. The mean score for parent transition concern was 1.94 (*SD* = .54, range = 1.27-2.87), for parent transition feeling was 2.03 (*SD* = .83, range = .62-3.54), and for parent transition reasons was 2.93 (*SD* = .87, range = 1.25-4.00). Overall parents reported few concerns about their adolescent with VWD transitioning to adult health care, as parents reported that they were "unlikely" (1) or "may" (2) have the concerns listed on the questionnaire when transitioning occurred. Parents of adolescents with VWD reported having some negative feelings about transitioning, with most parents reporting that they "may" (2) feel negatively about the transition when it occurs. Parents of adolescents with VWD reported that on average they "may" (2) or "would" (3) benefit from a transition program. The mean score for parent-rated adolescent medical independence was 2.17 (*SD* = .50, range = 1.59-3.28) and the mean score for parent-rated adolescent self-efficacy was 2.47 (*SD* = .46, range = 1.93-3.21). In general parents reported that

adolescents with VWD were "sometimes" (2) independent in taking care of their own health care needs and parents felt that adolescents were efficacious in their ability to care for their own needs "sometimes" (2).

The mean score for adolescent transition concern was 1.73 (*SD* = .53, range = .60-2.40), for adolescent transition feeling was 1.97 (*SD* = .65, range = 1.08-3.38), and for adolescent transition reasons was 2.71 (*SD* = .65, range = 1.38-3.75). Overall adolescents with VWD reported few concerns about transitioning to adult health care, as adolescents reported that they were "unlikely" (1) to have the concerns listed on the questionnaire when transitioning occurred. Adolescents with VWD also reported having some negative feelings about transition, with most adolescents reporting that they were "unlikely" (1) or "may" (2) feel negatively about the transition when it occurs. Adolescents with VWD reported that on average they "may" (2) benefit from a transition program. The mean score for adolescent-rated medical independence was 2.33 (*SD* = .67, range = 1.48-3.52) and the mean score for adolescent-rated self-efficacy was 2.71 (*SD* = .58, range = 1.86-3.50). In general adolescents with VWD reported that they were "sometimes" (2) independent in taking care of their own health care needs and that they were efficacious in their ability to care for their own needs "sometimes" (2).

Assessment of the discussion of transition, timing of transition, need for a transition program, and important components of a transition program. Given concerns about small sample size and the practical need to provide hematology physicians with information on transition for their patients, both with hemophilia and

VWD, the two groups were combined. In the combined sample a majority of parents (*N* = 17, 77%) and adolescents (*N* = 14, 64%) reported that they had not discussed transition with their physician. Those parents who did report discussing transition indicated that they talked about the timing of transition (*N* = 3), changing to a new adult physician (*N* = 1), and increasing adolescent responsibility (*N* = 1). Adolescents reported that changing to a new adult physician (*N* = 4) and timing of transition (*N* = 2) were discussed, while two adolescents reported that they did not remember what had been discussed.

On average parents reported that the best age for adolescents to transition is 21-25 years (*N* = 9, 41%), while adolescents reported that the best age for transition was 18-21 years (*N* = 12, 55%). This said, on average parents reported that they would like their adolescent to transition at age 19.59 years (*SD* = 2.77, range = 14-25 years), while adolescents reported that they would like to transition at age 19.55 years (*SD* = 2.44, range = 14-25 years).

In general, parents reported that they felt that a transition program would be helpful for their adolescent (*N* = 20, 91%) and for themselves (*N* = 19, 86%), while most adolescents reported that a transition program would be helpful for themselves (*N* = 17, 81%) and for their parents (*N* = 17, 81%). When asked about important components of a transition intervention program, few parents strongly indicated that they wanted to learn about how to help the adolescent care for his/her own health care needs (23%) or learn more about hemophilia/VWD (27%). On the other hand, parents strongly ("very true") reported wanting a transition program to help their adolescents learn to independently care for their own health care needs (46%), help their adolescents

talk to/educate others about their disease (41%), help parents work with other health care providers/insurance companies (41%), help adolescents discuss feelings about the hereditary nature of their disorders (50%), and help adolescents discuss feelings about having children (50%). Adolescents in general did not report feeling as strongly, as their parents, about the components needed in a transition program. They strongly indicated that they did not want to learn about how to independently care for their own health care needs (48%) or learn more about hemophilia/VWD (57%). Many adolescents were interested in discussing these topics (62%). Overall many parents and adolescents reported that they would like to learn more about adult health care programs, meet physicians in adult health care settings, learn how to solve medical problems that arise, help promote transfer of responsibility for health care from parent to adolescent, and discuss feelings regarding transition.

Correlations of transition variables and transition readiness. Pearson and Spearman correlations were conducted to explore associations between transition variables and physician-, parent-, and adolescent-rated transition readiness. Results suggested significant associations between parent-rated transition concerns and transition readiness, as more parental transition concerns were associated with lower physician-rated adolescent transition readiness ($r = -.52$, $p = .020$), lower parent-rated adolescent transition readiness ($r = -.62$, $p = .002$), and lower parent-rated self transition readiness ($r = -.72$, $p \leq .001$). Similar associations were also seen as more negative parent feelings about transition and lower parent-rated adolescent self-efficacy were associated with lower physician-rated adolescent transition readiness (feelings, $r = -.47$,

p =. 036), parent-rated adolescent transition readiness (feelings, r = -.74, $p \leq .001$; efficacy, r = .58, p = .005) and parent-rated self transition readiness (feelings, r = -.85, $p \leq .001$; self-efficacy, r = .55, p = .008). Results suggest that parents who reported more concerns about transition, had more negative feelings about transition, and felt less secure about their adolescents' ability to care for their own health had lower rated self and adolescent transition readiness.

Negative adolescent feelings about transition was associated with lower parent-rated adolescent transition readiness (r = -.55, p = .010) and lower parent-rated self transition readiness (r = -.50, p = .022). Fewer adolescent reported reasons for a transition program, lower adolescent-rated medical independence, and lower adolescent-rated self-efficacy were associated with lower adolescent-rated self transition readiness (reasons, r = .53, p = .014; health, r = .56, p = .008) and lower adolescent-rated parent transition readiness (self-efficacy, r = .46, p = .035). Overall correlations suggested that adolescents with more negative feelings about transition, adolescents who felt that there were fewer reasons to have a transition program, adolescents with had lower rated medical independence, and adolescents who felt insecure in their own ability to care for their health care needs had lower rated self and parent transition readiness.

Finally, higher adolescent-rated BASC total emotional problems was significantly associated with lower physician-rated parent transition readiness (r = -.54, p = .039), lower parent-rated adolescent transition readiness (r = -.50, p = .040) and lower parent-rated self transition readiness (r = -.61, p = .009). Poor adolescent

adjustment was correlated with lower physician, adolescent and parent transition readiness scores.

Differences on transition measures between female and male adolescents. Given significant associations between adolescent gender and transition variables, independent t-tests were run to explore the potential significant differences between males and females on these variables. Significant differences were found between male and female adolescents on parent feelings about transition [t (20) = -2.45, p = .024] and parent-rated adolescent self-efficacy, [t (20) = 2.30, p = .032]. On average, parents of female adolescents reported more negative feelings about transition (female M = 2.14, SD = .78; male M = 1.37, SD = .70) and parents of female adolescents reported lower adolescent self-efficacy (female M = 2.39, SD = .43; male M = 2.92, SD = .61). No differences were seen between females and males on other transition variables.

CHAPTER 5: DISCUSSION

Overview of Findings

The current study assessed differences on psychosocial and adjustment variables between adolescents with hemophilia and VWD and evaluated how demographic, family functioning, and parent-adolescent interaction variables impacted adolescent adjustment to chronic illness (Holmbeck & Shapera, 1999). In addition, the study explored family and adolescent feelings about transition and the need for a transition program.

Findings should be interpreted cautiously given the small sample size and low power. Overall findings did not support the hypothesis that adolescents with hemophilia would evidence increased disease severity, poorer family functioning, more negative parent-adolescent interaction, and poorer adjustment than adolescents with VWD. In general, results indicated minimal differences in disease, family functioning, interaction, and adjustment variables between adolescents with hemophilia and VWD, although adolescents with hemophilia and their parents rated their own or their adolescent's disease severity as significantly higher than adolescents with VWD or their parents. Both groups reported relatively mild disease severity, mild disease interference, and good adjustment.

Findings provided partial support for the adolescent development model proposed by Holmbeck and Shapera (1999) and the hypothesis that family variables would moderate the association between disease variables and adjustment. Evaluation

of the moderation hypothesis suggested that parent-rated family functioning moderated the association between parent-rated disease interference and adolescent-rated BSI total problems when controlling for family income. Other moderator relationships were not supported.

Results also provided partial support for the hypothesis that demographic and transition variables would significantly predict transition readiness. Overall findings indicated that family income accounted for a significant portion of the variance in predicting physician-rated adolescent transition readiness, while family income and physician-rated disease severity accounted for a significant portion of the variance in predicting physician-rated parent transition readiness. Additional regressions suggested that adolescent gender, family income, and transition variables significantly predicted parent-rated adolescent transition readiness and parent-rated self transition readiness. No variables significantly predicted adolescent-rated self or parent transition readiness and none of the family or adjustment variables accounted for a significant amount of the variance in predicting transition readiness.

Exploratory analyses on the SCIFF parent-adolescent interaction measure suggested that adolescents with hemophilia had lower levels of cohesiveness, higher adolescent withdrawal, and lower adolescent positive affect. Overall it appeared that there were more significant differences between adolescents with hemophilia and those with VWD on parent-interaction variables on the developmental task than on the medical task. Findings suggested more negative interaction, lower cohesiveness, higher parental rejection, and lower emotional support on the developmental task compared to the medical task. No significant differences between one- and two-parent families and

adolescents of different ethnicities were seen on the SCIFF measure. Additional exploratory analyses evaluating transition measures, suggested similar levels of concerns and need for a transition program in both groups. Adolescents and their families with VWD reported slightly more negative feelings about transition. Significant differences were seen between male and female adolescents on the transition measure, as parents of female adolescents reported more negative feelings about transition and rated their adolescent as having less self-efficacy than male adolescents.

Differences between Adolescents with Hemophilia and VWD on Disease, Family Functioning, Parent-adolescent Interaction, and Adjustment Variables

Findings suggested that there were no significant differences between the groups on measures of disease severity or disease interference, with the exception of significant differences on measures of parent- and adolescent-rated disease severity. Adolescents with hemophilia and their parents reported a higher level of disease severity than those with VWD. Additional analyses indicated that there were no significant differences between the groups on measures of family functioning, negative or positive medical and developmental SCIFF parent-adolescent interaction, transition and adjustment variables. Exploratory analyses indicated that the groups did differ on additional parent-adolescent interaction variables. Results suggested that adolescents with hemophilia and their parents showed significantly decreased levels of cohesiveness, increased adolescent withdrawal, and decreased adolescent positive affect during the developmental task, while adolescents with hemophilia also demonstrated decreased positive affect during the medical task. Given the small sample size and reduced power, the ability to detect

significant differences between the groups that exist is reduced. Therefore, with a larger sample size and increased power true differences are more likely to be seen. This said, even with increased sample size significant differences may not be seen between the groups. More recent data has suggested minimal differences between disease groups, indicating that diagnosis alone does not significantly predict outcome (Gartstein et al., 1999). Separating the groups by severity levels (e.g., high versus low severity) might have led to more significant findings.

The hypothesis that adolescents with hemophilia and VWD would differ on measures of family functioning, parent-adolescent interaction, and adjustment was based on literature that hemophilia is a more severe disease, involving increased invasive medical interventions, in comparison to VWD (Brown & DeMaio, 1992; Paper & Kelley, 2002; White & Montgomery, 2000). Models of risk and resistance factors have suggested that risk variables, including higher disease severity and more invasive treatment, negatively impact adolescent adjustment, while resistance factors, including family functioning and parent-adolescent interaction, serve to buffer the effects of risk factors on outcome (Wallander et al., 1989). Thus, it was conceptualized that higher levels of disease severity would be related to poorer functioning. Despite theoretical models detailing the impact of severity on outcome, the current results do not suggest that severity or other outcome variables differed between the groups based on physician-rated disease severity, disease interference or illness stress, which was gathered from file review. It may be that more subtle aspects of family functioning may be affected, but that global measures did not accurately assess or cannot address these changes. Thus, it may be that the theoretical model proposed by Wallander and

colleagues (1989) is correct and that sampling bias and measurement error account for the lack of significant findings.

While it is widely accepted that hemophilia tends to be more a more severe disease, by medical standards, it is important to realize that the range of disease severity and the degree of disease interference on daily life can vary greatly across individuals with hemophilia and VWD. The adolescents with VWD who were included in the sample may have had similar levels of disease severity or interference as adolescents with hemophilia. It may be that many of the adolescents with VWD recruited for the current study represented individuals with more severe disease symptoms. Since recruitment occurred in clinic, many of the adolescents with VWD who were approached to participate in the study attended clinic on a frequent basis and were known to medical staff. Higher medical severity and the associated need for medical consultation may have increased their rate of clinic attendance. Other adolescents with VWD, with less severe disease or disease interference, may have not attended clinic visits and therefore were not recruited for the study. In addition, symptoms that adolescents with VWD experienced were mostly related to heavy menstrual bleeding and may be more problematic, as well as more difficult to control, compared to bleeds in the hemophilia sample. Differences in symptom presentation may also have impacted the level of severity and illness interference between the groups. Finally, it may be that adolescents in the hemophilia sample who participated had less severe disease. It is unclear if clinic attendance is related to disease severity in adolescents with hemophilia or if those adolescents with higher disease severity may have been less likely to participate in a study that requires a time commitment. It is also possible that

adolescents with hemophilia who had higher disease severity also had more intensive home based interventions or services that reduced their clinic attendance. Therefore, differences on disease severity, disease interference, and illness stress variables may not be found because of a lack of representativeness within the groups.

In reviewing these findings it is important to discuss how severity was measured, as it appears that diagnosis did not significantly represent disease severity or predict family functioning, adolescent-parent interaction or adjustment. Results from the study suggested that perception of disease interference was not significantly different between groups, suggesting that diagnosis may not accurately separate less severe from more severe disease conditions. Literature has not found a link between medically based ratings of disease severity (e.g., factor levels, diagnosis) and outcome, suggesting that separating adolescents by diagnosis or using objective measures based on file review may not be an accurate way to categorize those with less or more severe disease (Barakat et al., 1997; Lutz et al., 2004; Stuber et al., 1997). Alternative measurement approaches to disease severity are discussed later.

It is also unclear if measures of family functioning and parent-adolescent interaction accurately assessed family variables in this sample. It appeared that evaluation of additional parent-interaction variables, particularly cohesiveness and adolescent behavior (i.e., affect and withdrawal), was important in highlighting potential differences between the groups. Results suggest the need to evaluate family functioning and interaction using multiple variables, as assessing global measures of positive and negative interaction was not as informative.

Additional concerns regarding the validity of family functioning measures in minority samples have been noted within the literature (Barakat, Lash, Lutz, & Nicolaou, 2005). Current results suggested that adolescents of minority status reported lower levels of family functioning than Caucasian adolescents. It is unclear if measures of family functioning accurately assess differences in family functioning and interaction that may accompany different ethnic backgrounds. Although the parent-adolescent interaction measure was originally used with a Hispanic population, other family functioning measures have not been extensively used with minority samples. Compounding concerns regarding the validity of the family functioning measures with ethnic minority families, lack of variance in family functioning scores was observed, with a majority of families reporting moderately high levels of family functioning.

In a related point, literature has suggested that other variables including socio-economic status, adolescent gender, and adolescent age impact outcome more than severity (Casey, Brown, & Bakeman, 2000; Lutz et al., 2004). It may be that differences on parent-adolescent interaction variables, specifically cohesiveness, adolescent affect and adolescent withdrawal between the hemophilia and VWD group were more due to differences in the gender between participants in the group (i.e., males in the hemophilia group and females in the VWD group) than to differences between disease variables. Previous studies have indicated that demographic variables can serve as potential risk factors for poorer adjustment, as males, older adolescents, and adolescents with lower SES have been shown to be at greater risk for poorer family functioning and adjustment to illness, especially in children with other blood disorders like sickle cell disease (SCD). Hurtig and White (1986) found a significant drop in

social competence and an increase in externalizing behavior problems in older boys with SCD. Hurtig and Park (1989) supported these findings in a later study that indicated that adolescent boys had the poorest rates of social competence compared to younger males and females with SCD. Higher rates of internalizing problems have also been noted in adolescence, with studies suggesting that older children have more internalizing symptoms than younger children and that male adolescents have higher rates of internalizing symptoms than female adolescents in a sample of adolescents with SCD (Brown et al., 1993; Kell, Kliewer, Erickson, & Ohene-Frempong, 1998). Lemanek and colleagues (1986) have also suggested that lower income may be related to increased behavior problems seen in a sample of children with SCD.

Overall findings suggest that adolescents in the current sample do not report severe levels of disease nor do groups differ on measures of functioning. Questions regarding the accuracy of using diagnosis as a way to measure disease severity are raised, especially given pediatric literature that suggests that diagnosis does not impact functioning and conflicting findings regarding disease severity. As mentioned, sampling bias may play a role in the lack of variance on disease severity measures. In addition how disease severity was measured may have impacted findings, as there is a lack of evidence that supports the use of diagnosis as a measure for severity. Additionally concerns about parent and adolescent ratings of disease severity and illness interference are raised, as family and adolescent knowledge of disease may influence their perception of disease severity, such that families with more disease knowledge may rate disease differently than families with less disease knowledge. The use of adolescent ratings for severity may also be problematic as their understanding of disease

severity may be related to their stage of development. Concerns about the reduced variance in the family functioning and parent-interaction measures and the use of the measures with minority participants are also apparent. While more general measures of family functioning and parent-adolescent interaction did not suggest differences between the groups, exploratory findings indicated that variables of cohesiveness, adolescent positive affect, and adolescent withdrawal were different. In addition, it appears that demographic variables may play a larger role in understanding family functioning and adjustment to illness than severity or diagnosis in this sample.

Moderation of the Association between Disease Variables and Adolescent Adjustment by Family Functioning and Parent-adolescent Interaction Variables

Initial correlations suggested few significant associations between disease variables and adolescent adjustment, further emphasizing the decreased role that medical severity and illness interference play in predicting adjustment. While it is sometimes difficult to accurately measure medical severity, the current study attempted to obtain objective and subjective variables to assess severity of disease, including multiple informant ratings, measures of health care utilization, and diagnosis. The lack of a relationship between disease severity and adjustment has been noted numerous times across other studies (Barakat et al., 2005).

Results should be interpreted cautiously, given that only one significant moderator effect was found and concerns about measurement error. Although there was one significant moderation finding, it may be that family functioning does not act as a moderator between disease severity and outcome, but that other variables (e.g., gender,

ethnicity, SES) moderate the relationship between disease severity and outcome. It is also plausible to hypothesize that there is not a significant relationship between disease severity and outcome, thus moderation is not possible, as a number of other studies have been unable to find significant associations between severity and adjustment (Casey et al., 2000). This said, the significant moderation finding will be discussed.

The significant moderator effect indicated that the impact of disease interference on adolescent adjustment may vary depending on the level of family functioning, such that at low levels of family functioning (poor family functioning) disease severity was significantly associated with adjustment and at high levels of family functioning (better family functioning) disease severity was not associated with adjustment. It appears that high family functioning may act as a buffer or a resistance factor against the impact of disease severity on adjustment. This result provides partial support for the developmental model proposed by Holmbeck and Shapera (1999) which indicates that interpersonal factors including family functioning, parenting, school/work, and peer variables moderate the association between developmental/disease variables (e.g., physical/disease, cognitive, psychological) and outcome (e.g., adjustment, autonomy).

Certainly families who perceive themselves as having intact problem solving skills, communication, emotional support, and appropriate division of family roles are more likely to be able to effectively handle the stress that comes with having an adolescent with a chronic illness (Davis et al., 1996; Perrin et al., 1993). In turn, families with high levels of conflict and low levels of cohesion, problem-solving and emotional support, may be more likely to have adolescents who demonstrate difficulties with adjustment. Previous literature has suggested that children and adolescents raised

by authoritative parents have better adjustment and increased quality relationships with peers and family (Baumrind, 1971; Feldman & Wentzel, 1990; Kaufmann et al., 2000). Additionally, families with a high degree of parental guidance and positive family interactions have been found to have adolescents with low levels of behavioral problems and greater success at school. Greater levels of emotional support and family cohesion have also been related to better psychological and behavioral functioning in adolescent girls (Eccles et al., 1997). Poor family functioning may result in adolescents who are non-adherent with their disease regimen and demonstrate difficulties managing illness complications because of a lack of parental support and guidance. This may be magnified during adolescence, when adolescents are attempting to separate from families and form their own identity, while continuing to need parental supervision surrounding disease management (Barakat, Smith-Whitley, & Ohene-Frempong, 2002; Baskin et al., 1998). These results also add support to the importance of assessing and promoting family functioning in adolescents and their families with chronic illness, especially during adolescence when families are preparing for transition.

In addition to focusing on interpersonal factors, the Holmbeck and Shapera (1999) model also recognizes the impact of demographic and intrapersonal factors like ethnicity, gender, and socioeconomic status on adjustment. Results from the moderation analyses suggested that family income played an important role in adolescent adjustment. Results are supported by other studies that have also implicated the impact of demographic variables, including low SES, on adjustment in a chronic illness sample (Lutz et al., 2004).

In many of these studies it is unclear if low SES or minority status, variables that are highly correlated, impact adjustment. Research has suggested that children of lower SES are exposed to more negative life events and higher levels of stress, which can impact health outcomes (Brady & Matthews, 2002; Evans & English, 2002). In addition to having increased exposure to violence, these children can also experience discrimination associated with lower income and ethnicity (Selner-O'Hagan, Kindlon, Buka, Raudenbush, & Earls, 1998). It is thought that children who are exposed to more negative life events will interpret other life events as more threatening, than children with fewer negative life events. Chen and colleagues (2004) evaluated a sample of high school students and found that those of low SES were more likely to interpret ambiguous situations/stimuli as threatening compared to students of higher SES. These students also were found to have higher levels of diastolic blood pressure and heart rate reactivity. Results suggest that increased life stress or stressful life events may have a negative impact on adolescent perception of future situations and effect overall physical well-being (Chen, Langer, Raphaelson, & Matthews, 2004).

These results have been supported in the chronic illness literature, as Chen and colleagues (2003) found that adolescents with asthma from low income families had significantly more exposure to stressful experiences during their lives, had lower beliefs about their ability to control their health, and showed more physiological markers associated with severe disease than adolescents with higher SES (Chen, Bloomberg, Fisher & Strunk, 2003; Chen, Fisher, Bacharier, & Strunk, 2003). In discussing the results, the authors suggest that increased negative stress appraisals (e.g., appraisal that a situation is threatening) and a diminished sense of control was more associated with

income than with having a chronic illness (Chen et al., 2003). These results add support to the idea that adolescents of low SES in the sample may view stressors associated with their chronic illness, including transition, as more threatening. Adolescents and families of higher SES who have more access to health care and other resources, and who are faced with fewer stressful life events, may be more likely to show better adjustment and better family functioning. In addition, their perception of disease and stressors associated with disease may be more neutral. Greater stress appraisals have also been found to be linked to worse physical outcome, as a result of higher levels of negative physiological markers associated with ongoing stress.

Results suggest the need for research that evaluates the interaction of demographic variables and family functioning on adjustment. Studies that continue to explore the relationship between stress appraisals and adjustment may help guide treatment planning, as interventions have been effective in changing how individuals think about or perceive situations. It may be that addressing adolescent's appraisals of stressors associated with a chronic illness could result in lower stress and better adjustment. It appears from these results that the focus of research should be on evaluating how family factors may moderate the relationship between demographic variables (i.e., gender, ethnicity, income) and adjustment, as well as evaluating other combination of associations between variables.

Prediction of Parent and Adolescent Transition Readiness

Since longitudinal data could not be gathered in the scope of this study, transition readiness scores were obtained from physicians, parents and adolescents in order to assess variables that play an important role in predicting outcome when transition occurs. It should be noted that there was limited variance in the transition readiness scores, with a majority of physicians, parents, and adolescents rating transition readiness to be in the average range. A larger sample size and a sample that is better representative of the range of disease severity in adolescents with hemophilia and VWD are necessary.

Current results did not support family functioning or adjustment as significant predictors in transition readiness. It was expected that those adolescents with intact family support, good family functioning, and a high level of current adjustment would be more likely to have better transition readiness scores than other adolescents. Research has indicated a link between family functioning and overall adjustment in children and adolescents with chronic illness (Perrin et al., 1993). In addition Davis and colleagues (1996) found that higher levels of family cohesion positively and significantly impacted daily living skills and overall adjustment in children with renal failure. Results suggest that family functioning can serve as a protective factor in buffering adverse events associated with a chronic illness, as it may help adolescents and children cope with or handle stressors that accompany development or their chronic illness and promote successful adjustment (Davis et al., 1996). Overall adaptation to chronic illness has, in turn, been linked to future adjustment, such that adolescents who have protective factors (e.g., high family functioning), which promote successful

adjustment, would be predicted to continue having protective factors and experience positive adjustment as they become older (Wysocki et al., 1992). Results suggest that family functioning and adjustment would be of importance in successful transition to adult medical care and would facilitate increased medical, as well as developmental, independence that accompanies adulthood.

In addition, when measuring transition readiness physicians, parents, and adolescents were simply asked how they felt that the adolescent or parent would transition into adult health care and was not based on actual behavior. Therefore the reliability and validity of the measure for transition readiness is questionable. Very little guidance was provided as to what defined readiness for transition. It may be that informants had different ideas of what a successful transition or transition readiness means, as some may think that successful transition is simply to move to adult health care with few difficulties, while others may think that successful transition is for the adolescent to effectively take on adult responsibilities associated with their illness. Differing perceptions of transition readiness may impact results, as family functioning may not significantly predict an adolescents' ability to move to a new physician, but may impact their ability to successfully take on additional responsibilities. More precise definitions of transition readiness and measurement of actual behavior during and after transition may have yielded different results.

Problems with family functioning and adjustment measures may also explain the nonsignificant findings. As mentioned before, concerns with measuring family functioning in an ethnically diverse sample have been noted within the literature and it is unclear if measures of family functioning adequately represent day-to-day family

interactions. While the current study attempted to use multiple measures of family functioning to obtain valid and reliable measurements of family relationships, more research on the use of family measures with different ethnic and SES samples needs to be completed. In addition many of the families in the sample reported high levels of family functioning, intact adolescent adjustment, and high levels of transition readiness. It may be that increased variability on adolescent adjustment and transition readiness scorers is needed in order to see the effect that family functioning has on these measures. In addition it is possible that at higher levels of family functioning there are few associations between disease variables and adjustment.

While family functioning and adjustment measures did not predict transition readiness, demographic factors including income level, ethnicity, and gender, as well as disease interference and transition variables, had a significant impact on transition readiness. Family income showed the most robust findings, as it predicted three out of the four transition readiness scores. Families with increased income may be able to access resources that facilitate adjustment and they may feel increased control over the transition process (Chen et al., 2003). Families with greater resources are also more likely to have freedom to choose who they will transition to, as opposed to lower income families who may have to transition to a specific hospital for adult care based on location. Low income families may also be more likely to utilize emergency services because medical assistance may not be accepted at some clinics, which can seriously hinder success following transition. Research has also suggested that minority families and those families with lower SES may be more wary of health care

professionals and the medical system, which could result in unsuccessful transition outcome (Lutz et al., 2004).

Interestingly physicians also rated lower SES families to have more difficulty transitioning. It was unclear if this was because of significant differences in adjustment between high and low SES families or if it was based on physician perception of adjustment in low SES families. Physicians may feel better connected and more comfortable with higher income families, who may have a greater level of education, may be more like the physician, and who physicians may perceive as more capable of handling the chronic illness. While there is a lack of literature assessing patient income status on physician perception of patient adherence and adjustment, Taira and colleagues have indicated that physicians sampled were more likely to discuss interventions (e.g., diet and exercise) for high-risk behaviors with patients of high income than with patients of lower income (Taira, Safran, Seto, Rogers, & Tarlov, 1997). Patients with lower SES may also ask fewer questions during medical visits and therefore discussions surrounding medical concerns may occur less frequently (Roter et al., 1997). Results suggest that physician or medical staff perception of income level may impact treatment decisions. More studies assessing physician relationships with families of low SES and the impact of medical team perception of families on medical decision making are needed to better understand the role of family income on adjustment and health outcome.

Adolescent gender also played a significant role in regressions evaluating parent-rated transition readiness. Male adolescents were found to be rated as experiencing greater transition readiness than females by their parents and parents also

rated their own transition readiness as being greater when they had a male adolescent. This may be in part due to parental perception that males are more capable of handling their medical needs or increased independence that accompanies adulthood. Research has suggested that parents of female adolescents are more restrictive and less likely to allow females to engage in activities that promote independence. In addition, female adolescents may ask for parental help to deal with medical complications more frequently than males, giving parents the perception that the adolescent is unable to handle medical concerns independently. Literature has also suggested that female patients are more likely to visit doctors and discuss medical concerns with physicians, thus female adolescents and their parents may have more frequent contact with and rely more heavily on medical staff, which may impact their perception of the transition process (Brink, van Dulmen, Messerli-Rohrbach, & Bensing, 2002; Hall, Irish, Roter, Ehrlich, & Miller, 1994; Wu, Howard, McGowan, Frau, & Dai, 2004).

Adolescent ethnicity and disease interference were each significant in one analysis, however these results were inconsistent and should be interpreted cautiously given the lack of other findings. It may be that adolescent ethnicity impacts comfortability with the medical team and lack of trust with the medical system, which impacts transition readiness. In addition, children of minority status may be confronted with increased social stressors that could influence outcome to a greater degree than Caucasian adolescents (Barbarin & Christian, 1999; Baskin, et al., 1998; Evans & English, 2002). Disease interference may also impact transition readiness, as those with greater levels of disease interference may evidence more difficulty transitioning to a new physician, may be less comfortable with the transition process, or may have disease

variables that significantly impact the adolescents' ability to care for their illness independently (Salmi et al., 1986).

While ethnicity and disease interference were found to have a role in predicting transition readiness, parent-rated transition variables were found to more consistently play a role in parent-rated adolescent and self transition readiness. It may be that associations between the variables reflect an informant bias across parent-rated measures. Parents who have more concerns and negative feelings about transition may be more likely to have lower rated self and adolescent transition readiness. Certainly those parents who are nervous or scared about transitioning their adolescent to adult health care may feel that they are less capable of handling the transition successfully and may feel that their adolescent is not ready for transition. In addition, they may feel that their own apprehensions about transition will impact their adolescent's ability to transition successfully or their own apprehensions may be based on the appraisal that their adolescent will not be able to handle the increased responsibility that accompanies transition. Parents appraisal of adolescents' ability to independently handle their own medical needs also predicted parent own transition readiness, indicating that parents who feel more comfortable with their adolescent assuming medical independence were more likely to feel that transition to adult health care would be successful and easier for them as parents. Results suggest that adolescents' feelings or concerns about transition did not impact their perception of their own or their parent's transition readiness.

Again results strongly suggest the role of demographic variables, especially SES, in predicting future transition readiness. In addition, assessment of parents' feelings about transition appeared to be important in understanding how an adolescent

or family may react to transition. Results also provide some support for a transition intervention program, as programs that decrease parental concerns and negative feelings and increase their perception of adolescent medical independence may result in increased transition readiness. In addition results may be generalized to general developmental progression and increased independence that accompanies adulthood, as demographic variables and parental feeling about adolescent increased responsibilities may play a role in adolescent readiness to move into adulthood. Certainly variables thought to be important in facilitating successful transition in health care will also be variables that facilitate successful developmental progression.

Differences between groups on additional parent-adolescent interaction variables. While overall positive and negative interactions were not different, exploratory analyses suggest that adolescents with hemophilia and their parents may interact differently on the interaction measure compared to adolescents with VWD and their parents. More differences between the groups were seen on the developmental task versus the medical task and seemed to surround the adolescents' behavior during the interaction, as opposed to overall differences in family communication patterns, with the exception of family cohesiveness. For example, one interaction between a 15-year-old female with VWD and her mother focused on the daughter's distress over an argument with her sibling. Both the mother and the daughter sat in close physical proximity, made eye contact throughout the interaction, and were able to discuss the concern without intervention or prompting from the research assistant. The mother appeared sensitive to the daughter's concerns, noting that she was also upset at the

sibling's behavior and reflected some of the daughter's feelings. In comparison, a number of the development task discussions with the hemophilia group surrounded behavioral problems. One mother and her son discussed the son's inability to abide by curfew. The son was disengaged in the interaction, only speaking when the mother asked him a direct question. They sat at a distance from each other and the son looked at the table during the interaction, making little eye contact with his mother. At times the mother expressed her frustration at the lack of participation from her son in the activity; however the son did not comment or react to his mother's demands to discuss the concern. The interactions of these two families, categorized as the adolescent's responsiveness/engagement in the activity and the mother's reaction to their adolescent's behaviors, provided examples of differences noted.

It is possible that disease severity or diagnosis, as well as gender, played a role in differences in interaction or how parents interacted with their adolescents. While disease severity was not found to be significantly different on most measures used, and was not significantly associated with family functioning and adjustment, it should be noted that adolescents with hemophilia and their parents reported higher levels of disease severity, which may explain differences in how these families interacted. In addition, a trend in the expected direction was seen on parent-rated disease interference, suggesting that adolescents with hemophilia may be rated by their parents as having a higher level of disease interference than adolescents with VWD in a larger sample. With a larger sample size true differences between the groups are more likely to be seen. This may lend credence to the idea that increased disease severity and interference, in addition to increased invasive medical procedures, may significantly

impact how the adolescent with a chronic illness interacts with family members. Also increased severity and interference may impact how parents treat adolescents when discussing a developmentally based task. Research has suggested that the presence of a chronic illness can stunt family growth and adolescent ability to meet developmental milestones, as parents may be more protective and not allow adolescents to engage in developmentally appropriate activities or tasks (Delengowski & Dugan-Jordan, 1986; Rosen, 1992). When discussing developmental tasks increased negative interactions may be associated with overall family conflict surrounding parental protectiveness or adolescent attempts to become independent.

Alternatively, research has indicated an inconsistent relationship between diagnosis and outcome, suggesting that even with a larger sample size there are not significant differences between the chronic illness groups based on diagnosis (Gartstein et al., 1999). This implies that other factors, potentially gender, may explain differences in parent-adolescent interaction highlighted in the case examples provided above. As reported, interaction between the mother and the daughter was different from that of the mother and the son. Results suggested that males showed higher disease severity and less positive parent-adolescent interactions. It may be that males appeared or were more withdrawn from the task and less positive in comparison to females who engaged in the interaction. Research has suggested that parents of female children are more likely to discuss emotionally charged topics with their child, in comparison to parents with male children (Kuebli, Butler, & Fivush, 1995). In addition, female adolescents often receive more opportunities in society and with peer groups to discuss concerns and to express emotions surrounding difficult situations. Research has indicated that

females will discuss issues involving emotional closeness and trust, as opposed to males who have been found to discuss recognition and mastery issues (Buhrmester, 1996; Buhrmester & Prager, 1995; Oxley, Dzindolet, & Miller, 2002). Also it should be noted that many of the parent participants in the study were mothers and that adolescent females may be more at ease discussing development tasks with their mothers than adolescent males. A study assessing mother and child communication around sexual behaviors in adolescence indicated that mother's discussions about sexuality were more comprehensive and involved more discussion of factual and moral issues with daughters than with sons.

Differences between parent-adolescent interaction styles on the developmental compared to the medical task. Finally, when evaluating the parent-adolescent interaction scale families showed differences in the way they interacted on the developmental task compared to the medical task. Overall families in the study demonstrated higher levels of conflict/tension, decreased cohesiveness, increased parental rejection, and decreased parental emotional support when discussing a developmental task (e.g., driving, curfew, behavior) than when discussing a medical task (e.g., concerns with medical management, administration/use of medications, limitations given the presence of chronic illness). Results suggest that family communication patterns, styles of interacting, and behavior during interaction is dynamic and depends on the topic being addressed. In addition it appeared that discussion of a medical concern decreased the amount of negative interactions (e.g.,

rejection, conflict) and increased positive interactions (e.g., cohesiveness and emotional support).

Differences between the groups (hemophilia and VWD) seemed to dissipate from the developmental to the medical task, suggesting that families interacted in a more similar way on the medical task than on the developmental task. While additional analyses were not done to evaluate the impact of disease diagnosis on these changes in interactions, review of mean scores from the groups on both the developmental and the medical task suggested that the hemophilia group had more marked changes in interaction style from the developmental to the medical task.

To date there has not been a study that has evaluated differences in how families interact around every day developmental issues in comparison to medical problems. Results highlight the need to better understand the dynamic nature of family interaction, as it appears that families may change their approach or style of interaction when faced with different topics of discussion.

Parental empathy and support of an adolescent with a chronic illness may impact how they interact with their adolescent surrounding medically oriented topics. This may be increased in chronic illnesses like hemophilia and VWD where there is a genetic component to the disease. Literature has suggested that parents of children who have a genetic disease may feel more guilt or responsibility for their child's illness (Varekamp et al., 1990). In addition, parents may have a better understanding of the disease and disease components because they may have lived with a family member who had the illness (e.g., a father in the case of hemophilia) or may have the illness themselves (e.g., in the case of VWD). In addition, parents are usually more involved with adolescent's

medical care and may perceive the medical topic discussed during the interaction as more of a joint concern. Therefore, less negative interactions and more cohesion may be observed because of the parents' role in medical care. In comparison, increased conflict may surround developmental tasks and disagreements about developmental issues may not evoke the same degree of parental empathy or understanding. Many of the topics chosen by families in the sample (e.g., disobedience, violation of rules, poor academic achievement) are more likely to be perceived as the individual adolescent's problem, as opposed to a general family issue. Many parents also voiced their frustration and anger over their adolescent's behavior, which was not observed during the medical tasks.

In addition, the nature of the task may have impacted the results. During many of the parent-adolescent interactions, parents and adolescents had difficulty deciding on a medical topic to discuss. They consistently reported having few problems with their disease (e.g., disease did not interfere with day-to-day life) and reported that they rarely argued or even discussed medical concerns. Given the mild disease severity and disease interference ratings provided by physicians, parents, and adolescents, it is not surprising that they reported few concerns about their medical illness. In comparison parents and adolescent were generally able to quickly come up with developmental topics that they had recently had an argued about.

Decreased negative interactions may in part be due to decreased disagreements about medical topics compared to increased negative emotions surrounding developmental topics that are more typical during adolescence and occur with greater frequency. In addition, parents and adolescents may rarely talk about medical concerns

in daily functioning and thus the novelty of discussing a medical topic may have impacted how they interacted with each other. Other studies have indicated that having a child with chronic illness may impact parenting practices related to the child's medical condition, but not change general parenting practices (Gerhardt et al., 2003).

Needs Assessment for a Transition Program

Given the increased survival rates of children with blood disorders, as well as other chronic illnesses, a great deal of focus within medical and pediatric psychology research has been the exploration of family needs during transition to adult health care providers and the necessity of structured transition programs to facilitate this process. In order to better understand the perceptions of families with an adolescent with a blood disorder an assessment of the need for transition services was conducted as a part of the larger study.

Necessity of a transition program. Overall parents and adolescents in both groups reported few concerns and negative feelings about the transition process. It is important to note that none of the adolescents sampled were in the process of transitioning to adult care, therefore it is unclear if parental and adolescent concerns or feelings would change as families go through transition. Correlations of transition variables and adolescent age were not significant, indicating that concerns and negative feelings about transition were not associated with adolescent age and approach of transition. Previous literature has indicated that age has not been significantly associated with adolescence readiness to transition (Cappelli et al., 1989), while

additional research has suggested that increased adolescent age was associated with more positive feelings about upcoming transition (Telfair et al., 1994). Other studies, which have evaluated adolescents prior to transition, have suggested that many adolescents feel alright about transition when interviewed (Boyle, Farukhi, & Nosky, 2001; Telfair et al., 1994).

In addition to assessing parent and adolescent feelings about the transition process, the needs assessment focused on adolescent medical independence and adolescent self-efficacy. These variables are important as previous research has suggested that adolescent medical independence is predictive of future success following transition (Cappelli et al., 1989). Results from the study suggest that both parents and adolescents feel that adolescents are "sometimes" independent in their own health care with increased independence significantly associated with increased adolescent age. Parents and adolescents also reported that adolescents "sometimes" to "most of the time" demonstrated self-efficacy. While greater independence has been associated with better adjustment and transition, it is unclear how much independence is necessary for optimal functioning.

Literature suggests that degree of independence will vary depending on the individual needs of the adolescent (Lewis-Gary, 2001; Modrcin, 1989). It appears that families sampled have adopted a developmental model for increased adolescent independence, as medical independence was associated with increased age. In addition, results indicate that adolescents sampled were able to handle medical independence with moderate levels of self-efficacy. Interestingly, measures of self-efficacy were not

associated with age, suggesting that adolescents did not feel more capable of handling their medical concerns as they became older.

An additional analysis showed that female adolescents were rated by their parents as demonstrating lower levels of self-efficacy than male adolescents, potentially indicating that parents are less confident in female adolescents' ability to handle medical responsibilities. It may be that female adolescents are not provided with as many opportunities to practice independent medical skills resulting in parental perception of reduced self-efficacy or that females are more likely to ask for help when they experience medical problems, which may impact parental perception of their ability to competently handle their own medical needs. It is also possible that differences between female and male rated self-efficacy may be a result of diagnosis. More specifically, treatment of adolescent males with hemophilia may be more proactive in nature (e.g., prophylaxis) allowing adolescents and their parents to feel more control over the disease. In comparison, adolescent females with VWD may have symptoms that are more variable and are not amendable to proactive treatment regimens potentially resulting in lower self-efficacy.

Based on parental and adolescent report, overall results suggest that a transition program may be effective in increasing transition readiness in families who are displaying a number of concerns about transition and who report negative feelings about the transition process. Findings do not indicate that a transition program is a necessity for all families, although many families reported interest in attending a program. Given these findings additional support is gathered that programs that work to reduce parental concerns, parent/adolescent negative feelings, increase independence, and increase self-

efficacy could influence transition readiness. It may be that a more general information program could be helpful for families who show few negative feelings/concerns and who generally function well, while more intensive programs that facilitate the transition process and that address other areas thought to influence adolescent outcome (e.g., poor family functioning, parent-adolescent interactions) would be more appropriate for families with high level concerns, who show higher risk factors (e.g., gender, ethnicity, income), or who evidence difficulties with adjustment. It also could be that families are unaware of the impact of transition and do not realize the potential benefits of a transition intervention.

Results from the transition need assessment were similar to those found in other transition studies that have evaluated different illness samples. Telfair and colleagues (1994), using the same transition scale, found similar results with most parents and adolescents reporting mixed emotions and levels of concern about transitioning, with overall results suggesting that they felt "okay" with the process. In addition many adolescents and parents in his sample endorsed being interested in a transition program and indicated interest in various components of a transition program. While Telfair and others found that older adolescents and adolescents with less disease severity were more likely to feel positively about transition, the current study found less consistent results. Adolescent age and many of the disease severity/interference variables were not found to impact feelings or concerns about transition. Differences between the current study and Telfair's results may be due to ethnic and income differences, as Telfair's sample of children with SCD was primarily African-American. Boyle and colleagues (2001) also found no associations between feelings about transition and age or disease severity in a

group of adolescents with cystic fibrosis. In addition many of the studies within the literature suggest that adolescents feel that transition around age 18-19 years of age is appropriate, similar to findings in the current study (Boyle et al., 2001).

Limitations of Current Study and Implications for Future Research

Sample Size, Demographic Limitations and Implications for Future Research

A major limitation for the current study is the small sample size. The genetic nature of hemophilia and low incidence of the disease made it especially difficult to recruit adolescents with a diagnosis of hemophilia. While initial attempts had been made to complete the study using multiple sites, it became apparent that other hospitals had low numbers of patients who met the inclusion criteria. In an attempt to increase sample size, participants were gained from the National Hemophilia Foundation. Even with additional participant pools and loosening inclusion criteria limited numbers of participants were recruited. Given the high incidence rate of VWD it was expected that more participants from this disease population would be recruited, but this was also a struggle. In reviewing data collection it appeared to be especially difficult to recruit patients out of clinic given that many of these adolescents attend clinic irregularly and sometimes did not present for annual visits. Concerns regarding the representativeness of sample are noted, as it is unclear if adolescents in the current sample accurately represent adolescents within the general hemophilia/VWD population because of lower disease severity.

As a result of a small sample size, power was significantly reduced. Reductions in power resulted in significant changes to the planned analyses and a limited ability to achieve significant results. It would be expected that with a larger sample size additional significant results would be obtained, especially on measures where trends were observed and regression analyses. Thus, the variables that may potential predict transition readiness and outcome variables may have been overlooked. Concerns about small sample sizes have been noted within other studies assessing adolescents with hemophilia, as it is a rare disorder and many treatment centers do not have a large number of patients with the diagnosis. Other research has reported sample sizes of 20 to 40 subjects, highlighting the difficulty in collecting participants from these disease groups (Colegrove & Huntzinger, 1994; Garstein et al., 1999; Logan, Gibson, Hann, & Parry-Jones, 1993; Trzepacz et al., 2003;). Those studies that have been able to gather a large number of participants have used multi-site recruitment strategies in an attempt to increase sample size. Additional studies will need to focus on collaboration with other hemophilia treatment centers in order to have sufficient sample sizes. During the current study the use of home visits significantly increased recruitment of participants and would likely help increase numbers in future studies.

In addition to reduced sample sizes and the need for studies with greater participant recruitment, another limitation of the current study was the differences in the ethnic backgrounds between the groups, as well as the large number of females included in the VWD sample. Additional concerns regarding the demographic representativeness of the study are also noted. Participants for the current study were gathered from a clinic sample that was primarily Latino. While the ethnic background

of the study participants matched that of the clinic where data was gathered, it is not necessarily representative the ethnic background of the general population of individuals with blood disorders. In the current study, results suggested that a majority of adolescents with hemophilia were Caucasian (50%), while a smaller subset were African-American (40%) or of another ethnicity (10%). In a large multi-site study, using 14 comprehensive hemophilia care centers, 333 boys ages 6-19 were identified (Nichols et al., 2000). A majority of these participants were Caucasian (72.4%), with 15% Latino, 10.8% African-American, and 1.8% of other ethnicities. Comparison of results from both studies suggest some differences in the ethnic backgrounds of adolescents in the current study and those in other research with hemophilia. Within the VWD sample a larger portion of the adolescents in the current study were Latino (33%). Given that there has not been any published research evaluating VWD in pediatric populations it is unclear how the ethnic background for VWD patients represents the overall sample. Given concerns regarding the representativeness of the current sample, generalization of results should be done cautiously. Since there were limited data in the literature regarding the psychological adjustment and level of family functioning in individuals with VWD it is unclear if any differences between the hemophilia and VWD group are due to disease variables (disease severity) or are impacted gender and ethnicity. Obtaining a sample of adolescents in both groups that more widely match national demographics for ethnicity and recruiting a more equal sample of male/female adolescents with VWD could help answer questions posed.

Measure Limitations and Implications for Future Research

In many earlier studies it has been questioned if disease severity, as calculated by diagnosis, is an accurate reflection of the severity of the chronic illness. In the current study disease severity ratings were based on physician, parent, and adolescent perception. Results suggested that disease severity varies greatly, with disease severity based on diagnosis or illness type not being highly correlated with perception of severity/interference. In addition it appeared that each informant had different perceptions of an adolescent's level of disease severity and disease interference. Again developmental understanding of disease severity and family knowledge of disease may have played a role in different perception scores. Additional research studies should also attempt to collect perception of disease severity and disease interference, as it appears to provide information that goes beyond severity ratings that are based on diagnosis. When collecting ratings from adolescents and parents it will be important to understand if disease knowledge influences their scores. In addition future studies should use multiple informants when collecting data. Results suggested that physician, parent, and adolescent report were different and added their own unique view of the variables assessed.

As a part of the current study family functioning and parent-adolescent interaction measures were used. While both measures have been developed and normed, there has been little published about their validity with families of minority status and some have criticized their use with minority families (Barakat et al., 2005). As research in the area of family functioning and parent-adolescent interaction evolves, the development of more sensitive measures will be essential to better understand

relationships between demographic, family functioning, and adjustment variables. Additionally, using African-American individuals to help code parent-adolescent interactions may increase validity of the measure. Many studies within the pediatric psychology literature have also begun to encourage father participants in studies. In the current study two fathers and one grandparent were included. In order to expand research within the field, the use of multiple parents in a family when studying family functioning and parent-adolescent interaction would be important.

Although the ethnic sensitivity of the parent-adolescent measure needs to be addressed, overall results suggest that the SCIFF interaction variables were positively correlated with other similar scales and negatively correlated with dissimilar scales indicating good internal consistency within the scale. The lack of associations between the parent-adolescent interaction variables and family functioning measure brings into question the construct validity of the measures. It may be that differences in how the measures are obtained (e.g., parent/adolescent report versus observation) may be responsible for the lack of correlation. Given the small sample size of the current study investigators were unable to use all of the scales from the measure in the analyses, thus those scales thought to best represent the overall tone of the interaction (e.g., negative/positive interaction) and that were highly correlated with a number of the other scales were used. This said, it appears from the exploratory analyses that additional scales on the SCIFF add their own unique variance and should be included in the analyses when sample size permits. In addition the study highlights the importance of realizing differences in family interaction styles dependent upon the topic of discussion. More research that focuses on the dynamic nature of family interactions and differences

between developmental and medical interaction tasks is important in better understanding how to promote more effective communication within a family.

Transition Needs Assessment Limitations and Future Research Implications

In the current study the transition questionnaire was given in an attempt to gain additional information about the need for a transition program within this disease group. While the transition questionnaire gathered a great deal of qualitative information, it is not normed, nor have psychometric information been published on the measure. The use of a more structured measure and one that has good psychometric properties would be needed in future studies that are evaluating the transition process.

Although information gained from the transition measure will be helpful in describing the needs and interest of adolescents with hemophilia and VWD in a transition program, the current study was unable to longitudinally assess the process of transition nor implement and evaluate a transition program. Within the literature some studies have conducted similar needs assessments, although they have typically been done with less standardized measures, have not taken into account other variables (e.g., adjustment, family functioning), and have not relied on physician-, parent-, and adolescent-rated transition readiness. Future studies need to focus on evaluating adolescents prior to and following transition to see if outcomes warrant the need for a transition program. In addition, more studies evaluating the essential components of transition and the effectiveness of transition programs are necessary.

Clinical Implications

Results gathered from this study have a number of clinical implications for practice. Most striking is the impact of demographic variables on family functioning, parent-adolescent interaction, and adjustment variables. Overall findings most strongly suggest that adolescents of lower SES are those most likely to have impaired family functioning, fewer positive parent-adolescent interactions, and poorer adjustment. Results also suggest that males and adolescents of minority status may also show poorer adjustment and family functioning, although these relationships inconsistent. Males were also found to have parents who rated fewer concerns and negative feelings associated with transition, while females were reported to have lower levels of transition readiness and less parent-rated self-efficacy. Results highlight the need for clinicians to pay particular attention to the impact that disease may have on adolescents with certain demographic characteristics, as demographic variables may be more important than disease variables.

Although level of SES or gender can not be changed, physicians who are more aware of the associations between demographic variables and adjustment/transition may be in a better position to intervene. Providing interventions that are tailored to the unique needs of adolescents of lower SES and helping parents and adolescents gain access to services that promote successful adjustment are essential. Medical staff may find that increased interaction with social workers, who can help families of lower SES navigate the medical system, will result in better continuity of care as families may be more likely to follow-up with adult physicians after transition. In addition providing

more support for these families during the transition process and potentially using a cognitive behavioral type intervention to address negative appraisals associated with transition may be helpful (Chen et al., 2004). In addition, interventions should focus on different needs of female and male adolescents. For example, interventions that encourage better family functioning in males and increased communication between males and their parents would be important in assuring better medical and developmental outcomes. In turn, interventions that help parents of female adolescents feel more comfortable with their adolescent's increased medical independence and that allow parents and female adolescents to problem solve through medical situations would increase parental and adolescent comfort level. Not only do these interventions support successful transition from pediatric to adult health care, but they are also important for successful developmental progression into young adulthood. It appears that males may be more likely to evidence difficulties with general developmental issues and family functioning, while females show less concerns surrounding development. It may be that chronic illness and the implications of the disease more significantly impacts males ability to gain independence and engage with peers in a developmentally appropriate manner, compared to females who may find that illness has a smaller impact on general development and independence. Demographic variables were also found to impact transition, as parents of females with a chronic illness were more likely to have increased difficulty with the transition.

In addition to demographic variables, results from the parent-adolescent interaction task indicated that families may naturally increase positive communication and support when topics surround medical care, which is important in how clinical care

is provided. More specifically families may appear to be functioning well as a unit when in a medical setting, given the focus on medical topics, however problems surrounding developmental issues may not be as apparent. This may interfere with medical staff's abilities to accurately assess family functioning and adolescent adjustment. In addition, these results may suggest that families "rise to the occasion" when faced with medical concerns or problems and therefore need less intervention surrounding medically based tasks. In reality some of the families may actually need more intervention in learning how to cope with normal adolescent development issues. While these services may be needed, it is important to consider that providing these types of interventions may be well outside of the scope of pediatric clinics. Some larger clinics may employ psychologists or other mental health professionals to provide individual treatment or group interventions to their patients. This is certainly not feasible in all settings, therefore medical teams may need to be discuss how they can quickly and effectively screen for difficulties with family functioning or adjustment and then have resources for the families to access if intervention is warranted. Within the pediatric psychology literature, empirically supported intervention programs that have focused on cognitive-behavioral techniques and family functioning have been found to be effective in reducing medical symptoms associated with diabetes and asthma, as well as improving parental distress surrounding cancer diagnosis (Boardway, Delamater, Tomakowsky, & Gustafsson, Kjellman, & Cederblad, 1986; Kazak, 2005). More recently interventions that attempt to combine family therapy with cognitive behavioral interventions have been shown to be effective with young adult cancer survivors (Kazak, 2005).

Results do not necessarily suggest universal, immediate need for a transition program. Within pediatric health care settings there has been a great deal of focus on the need for comprehensive transition programs. While these programs would not negatively impact adolescents or their families, and may help some families who are at higher risk, they do not appear to be necessary for all families. It may be that healthcare providers' concerns and negative feelings about transitioning their patients is more problematic than family and patients' concerns about the transition process. This highlights the need for additional medical provider training around adolescent development and transition issues. Although parents and adolescents reported that a transition program would be helpful, findings are more suggestive that interventions that promote better family functioning, improved parent-adolescent interaction and communication, and address adolescent adjustment concerns would be more appropriate and effective in facilitating transition. Even with additional information about the transition process and guidance around transition, if families are unable to communicate and demonstrate poor family functioning adolescent and parent ability to successfully transition will be compromised. Others have also encouraged the use of a multi-tiered approach to intervention (Barakat, Kunin-Batson, & Kazak, 2003). Suggesting that no transition intervention, or at most a general information transition program, should be offered to families who evidence few concerns regarding transition and who show few risk factors. The intensity of this program should then increase for families who display more risk factors (e.g., demographic factors, poor family functioning, increased concerns with transition, decreased adolescent medical independence). Adolescents and their families who show minimal concerns may be involved in a general education

program, but may also have opportunities to engage in interventions to address concerns with family functioning, transition, or medical independence. If families continue to struggle with the idea of transition following intervention, intensity of the program may increase to having social workers follow individual families to increase transition success. In addition, medical providers may create transition plans for each of their families and make sure that individuals of the medical teams are working together to help the family follow this plan for transition.

Conclusions

Overall results did not provide support for differences between adolescents with hemophilia and VWD on disease, family functioning, or adjustment variables. Both groups of adolescents demonstrated, on average, intact family functioning and adjustment. Limited support was found for family variables role in moderating the relationship between disease interference and adjustment, with family functioning appearing to serve as a buffer against the potential impact of disease factors on adjustment. Finally, family and adjustment variables did not appear to predict parent or adolescent transition readiness. Demographic variables were found to play an important role in family interactions and indicated an increased need to promote family functioning in male, minority, and low SES samples. Future research should focus on increasing sample size within hemophilia research, using multiple informants, evaluating the process of transition and effectiveness of transition programs, and further exploring how family dynamics and interactions change based on different discussion

topics. Finally, results suggest that interventions with this population and surrounding transition should be multi-tiered, in that families with low level concerns or negative feelings may find a general information program about transition beneficial, while families who display increased difficulties with family functioning, poor adjustment, or a high degree of concern about transition may benefit from a more intensive intervention that focuses on promoting family functioning, adolescent adjustment, and medical independence. Interventions should be tailored to meet the unique needs of male and females with blood disorders, as results suggested that males may have more difficulty with family interactions, while females may be more insecure about caring for their medical needs.

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Table 1

Demographic Variables for the Hemophilia and VWD Sample and their Parents

	Hemophilia				VWD			
Demographic Variables	*N*	Adolescents	*N*	Parents	*N*	Adolescents	*N*	Parents
Gender	10				12			
Males		100%				17%		
Females						83%		
Relationship	10				12			
Mother				80%				92%
Father				20%				
Grandparent								8%
Age	10		10		12		12	
M		15.59		40.90		16.08		42.33
SD		3.14		5.67		2.27		7.11
Range		13-21		35-51		13-21		31-54
Ethnicity								
Caucasian		50%		60%		42%		42%
African-American		40%		40%		17%		17%
Latino						33%		25%
Other		10%				8%		17%

Table 1 (*continued*).

	Hemophilia			VWD		
Demographic Variables	*N*	Adolescents	*N* Parents	*N*	Adolescents	*N* Parents
Marital Status						
Married			50%			58%
Divorced						25%
Separated			30%			8%
Never Married			10%			8%
Remarried			10%			
Education						
1-8						8%
9-12			50%			25%
Some college/vocational			20%			42%
College graduate			20%			17%
Professional School			10%			8%
Income						
Below $19,999			20%			42%
$20,000-34,999						17%
$35,000-49,999						
$50,000-74,999			50%			17%
$75,000-99,999			20%			17%
Over $100,000			10%			8%

Table 2

Health Status of Adolescents in Sample

Health Variables	*N*	Hemophilia Adolescents	*N*	VWD Adolescents
Diagnosis				
Hemophilia	10	45%		
VWD			12	55%
Diagnosis Type				
Hemophilia A		89%		
Hemophilia B		11%		
VWD Type 1				82%
VWD Type 2				9%
VWD Low Levels VIII				9%
Severity Level for Hemophilia				
Mild (> 5 %)				
Moderate (2-5 %)		22%		
Severe (< 2 %)		78%		
Medications				
M		1.89		3.18
SD		1.16		3.06
Range		1-4		0-10
Spontaneous Bleeds				
M		1.89		.18
SD		2.52		.40
Range		0-6		0-1
Prolonged Bleeds				
M		1.00		1.45
SD		1.73		1.75
Range		0-3		0-5

Table 2 (*continued*).

Health Variables	*N*	Hemophilia Adolescents	*N*	VWD Adolescents
Hospitalization				
M		.22		.64
SD		.44		.92
Range		0-1		0-3
Outpatient Clinic Visits				
M		2.44		1.73
SD		1.94		1.10
Range		1-7		0-4
Factor Replacement				
M		52.33		
SD		89.78		
Range		0-156		
Usage				
PRN		30%		
Prophylactic				
> 3 times per week		20%		
1-3 times per week		30%		
once every 2 weeks		10%		
once a month		10%		
Other Health Condition				
Hepatitis C		10%		8%
Asthma		10%		33%
Learning Problems		30%		17%
Psychological Disorders		10%		25%

Table 3

Descriptive Information of Clinical Variables for Adolescents with Hemophilia and their Parents

Clinical Variables	Adolescents		Parents		Physician	
	M	*SD*	*M*	*SD*	*M*	*SD*
Disease						
Disease Severity	3.50	(1.35)[a]	3.70	(1.42)[a]	3.11	(.93)[b]
Disease Interference	1.90	(1.10)[a]	2.00	(1.05)[a]	2.44	(1.13)[b]
Illness Stress	3.97	(9.87)[a]				
Family Functioning	1.70	(.48)[b]	1.62	(.34)[a]		
SCIFF Interaction						
Negative	2.50	(1.15)[b]				
Positive	2.39	(1.19)[b]				
Medical Negative	1.61	(.60)[b]				
Medical Positive	2.61	(1.34)[b]				
Adjustment						
BSI	.42	(.46)[a]				
Adolescent BASC	43.43	(5.26)[c]				
Parent BASC	53.14	(9.26)[c]				
Transition Readiness						
Adolescent	2.11	(1.05)[b]	3.10	(.74)[a]	3.89	(.78)[b]
Parent	2.56	(1.05)[b]	3.0	(.82)[a]	4.00	(.71)[b]

Note. [a] $N = 10$, [b] $N = 9$, [c] $N = 7$

Table 4

Descriptive Information of Clinical Variables for Adolescents with VWD and their Parents

Clinical Variables	Adolescents		Parents		Physician	
	M	*SD*	*M*	*SD*	*M*	*SD*
Disease						
Disease Severity	2.33	(1.15)[a]	2.50	(1.0)[a]	3.00	(.45)[b]
Disease Interference	2.17	(.94)[a]	2.58	(1.56)[a]	3.09	(.54)[b]
Illness Stress	6.56	(8.48)[a]				
Family Functioning	1.82	(.37)[a]	1.74	(.38)[a]		
SCIFF Interaction						
Negative	2.04	(1.59)[b]				
Positive	3.55	(1.27)[b]				
Medical Negative	1.64	(.92)[b]				
Medical Positive	3.18	(1.15)[b]				
Adjustment						
BSI	.60	(.52)[a]				
Adolescent BASC	49.10	(9.39)[c]				
Parent BASC	52.70	(13.58)[c]				
Transition Readiness						
Adolescent	2.33	(1.07)[a]	2.17	(1.19)[a]	3.18	(1.07)[b]
Parent	2.67	(.65)[a]	2.17	(1.11)[a]	3.27	(.65)[b]

Note. [a] $N = 12$, [b] $N = 11$, [c] $N = 10$

Table 5.

Correlations between Demographic, Family Functioning, Parent-Adolescent Interaction, and Adjustment Variables

	Gender	Adolescent Ethnicity	Family Income	Parent FAD	Adolescent FAD	Adolescent BSI
	r, *p*	*r*, *p*	*r*, *p*	*r*, *p*	*r*, *p*	*r*, *p*
Adolescent Ethnicity	.17, ns					
Family Income	-.28, ns	-.63, .002 *				
Parent FAD	-.11, ns	.17, ns	-.32, ns			
Adolescent FAD	-.08, ns	.56, .008 *	-.33, ns	.26, ns		
Adolescent BSI	.13, ns	.18, ns	-.53, .010 *	.52, .013 *	.12, ns	
Parent BASC	-.16, ns	.08, ns	-.20, ns	.51, .035 *	.11, ns	.44, .040 *
Adolescent BASC	.13, ns	.42, ns	-.71, .001 **	.61, .009 **	.25, ns	.80, .000 **
SCIFF Negative	-.55, .001 *	.05, ns	-.11, ns	.25, ns	.12, ns	.20, ns
SCIFF Positive	.57, .008 *	-.17, ns	.15, ns	-.13, ns	-.25, ns	-.20, ns
SCIFF Medical Negative	.02, ns	-.13, ns	.02, ns	.04, ns	-.16, ns	-.05, ns
SCIFF Medical Positive	.38, ns	-.14, ns	.13, ns	-.16, ns	-.26, ns	-.11, ns

Table 5 (*continued*).

	Parent BASC *r*, *p*	Adolescent BASC *r*, *p*	SCIFF Negative *r*, *p*	SCIFF Positive *r*, *p*	SCIFF Medical Negative *r*, *p*
Adolescent Ethnicity					
Family Income					
Parent FAD					
Adolescent FAD					
Adolescent BSI					
Parent BASC					
Adolescent BASC	.57, .006 **				
SCIFF Negative	.42, ns	.23, ns			
SCIFF Positive	-.23, ns	-.12, ns	-.69, .001 **		
SCIFF Medical Negative	.44, ns	.15, ns	.53, .016 *	-.28, ns	
SCIFF Medical Positive	-.27, ns	-.12, ns	-.63, ns	.80, .001 **	-.40, ns

Note. ns = not signficant. *$p < .05$, **$p < .01$. Sample size was $N = 22$ for ethnicity, income, parent FAD, adolescent BSI, parent BASC, and adolescent BASC. Sample size was $N = 21$ for adolescent FAD. Sample size was $N = 20$ for SCIFF negative, SCIFF positive, SCIFF medical negative, and SCIFF medical positive.

Table 6

Differences between Adolescents with Hemophilia and VWD on Disease Severity, Family Functioning, Parent-Adolescent Interaction and Adjustment Variables

Variable	Hemophilia *M*	VWD *M*	*t*	(*df*)	*p*
Disease Severity					
Physician	3.11	3.00	.33	(11)	.748
Parent	3.70	2.50	2.32	(20)	.031*
Adolescent	3.50	2.33	2.18	(20)	.041*
Disease Interference					
Physician	2.44	3.09	-1.58	(11)	.144
Parent	2.00	2.58	-1.00	(20)	.328
Adolescent	1.90	2.17	-.61	(20)	.546
Illness Stress	3.97	6.56	-.58	(15)	.573
Family Functioning					
Parent	1.62	1.74	-.76	(20)	.454
Adolescent	1.70	1.82	-.62	(19)	.542
Parent-Adolescent Interaction					
Negative	2.50	2.05	.72	(18)	.482
Positive	2.39	3.55	-2.08	(18)	.052

Table 6 (continued).

Variable	Hemophilia *M*	VWD *M*	*t*	(*df*)	*p*
Medical Negative	1.61	1.64	-.07	(18)	.945
Medical Positive	2.61	3.18	-1.03	(18)	.328
Adjustment					
Parent BASC	53.14	52.70	.08	(15)	.941
Adolescent BASC	43.43	49.10	-1.44	(15)	.171
BSI	.41	.60	-.87	(20)	.396

Note. *$p \leq .05$, ** $p \leq .01$

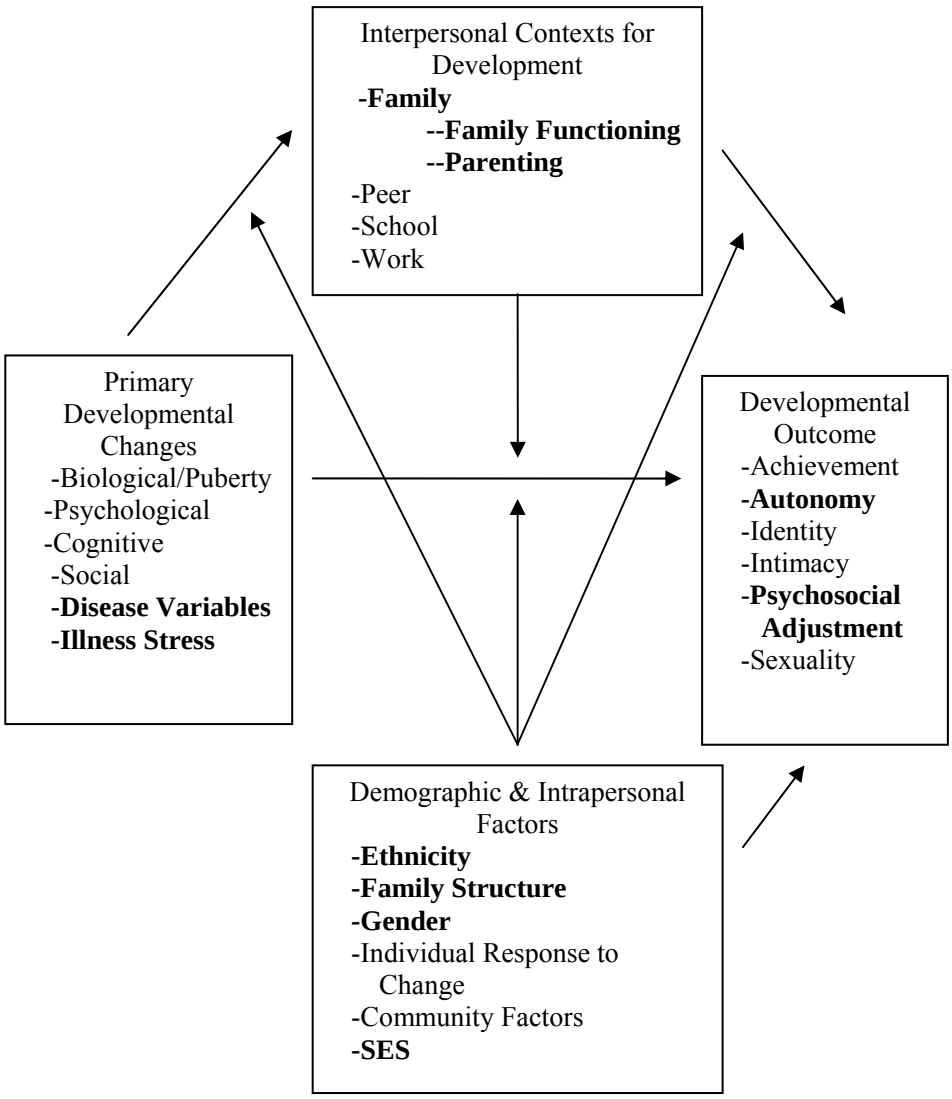

Figure 1.

Risk and Resistance Factors Impact on Adjustment in Adolescence

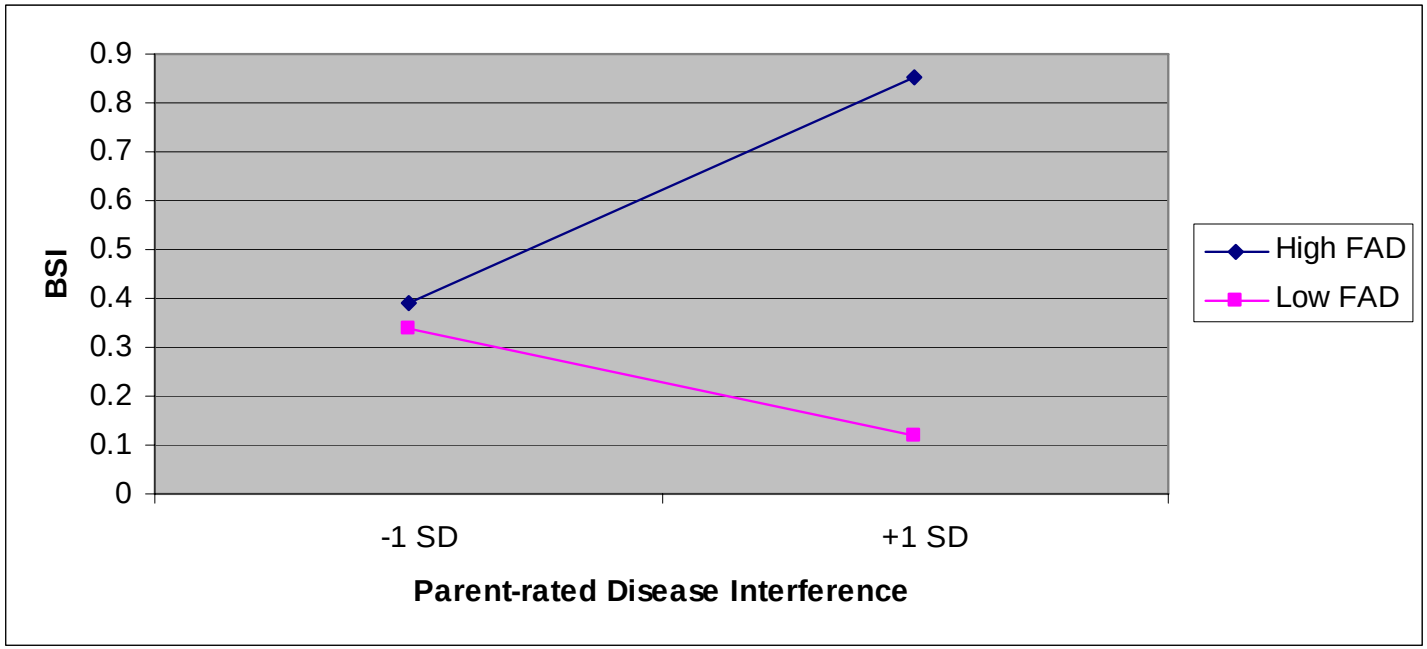

Figure 2.

Post-hoc Analyses of Moderation

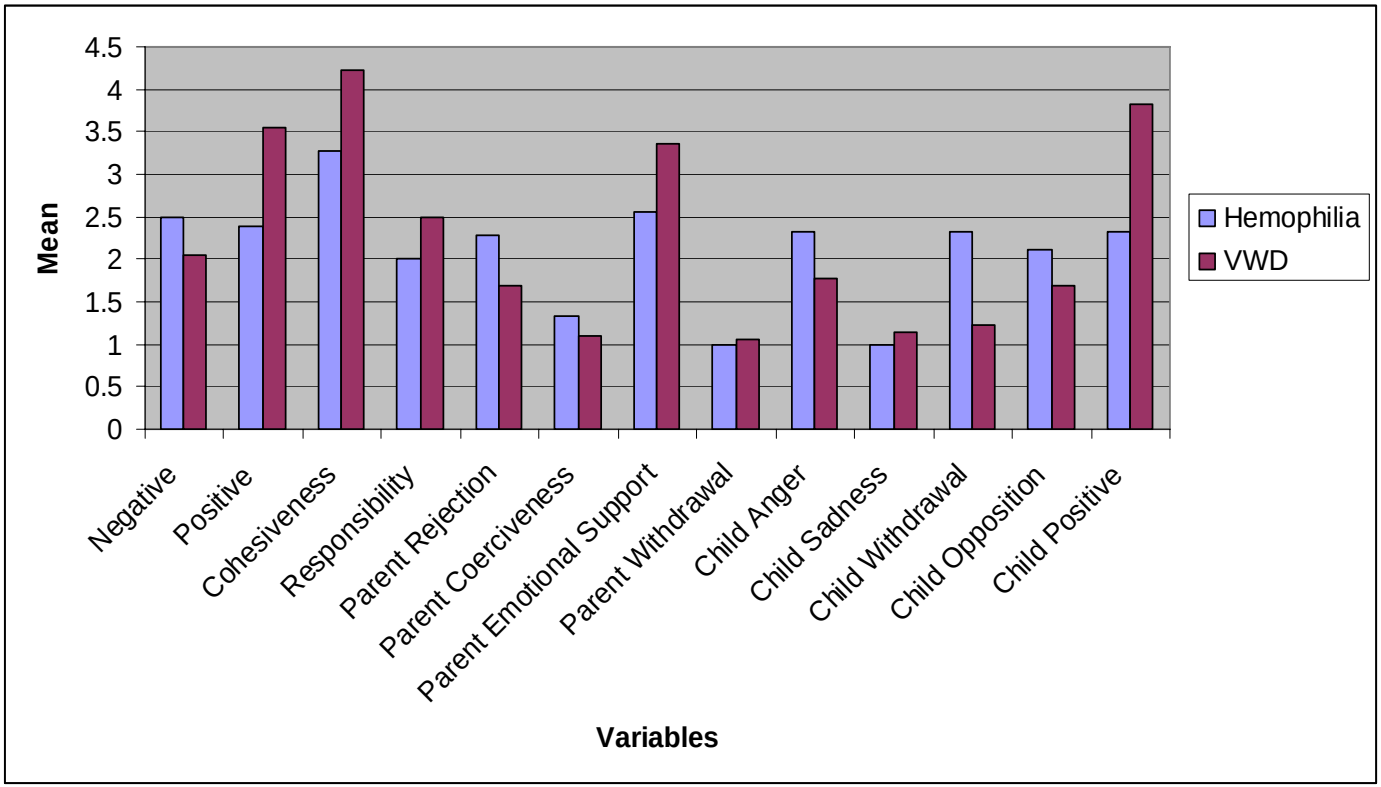

Figure 3.

Differences in SCIFF Mean Scores Between Adolescents with Hemophilia and Adolescents with VWD on the Developmental Task

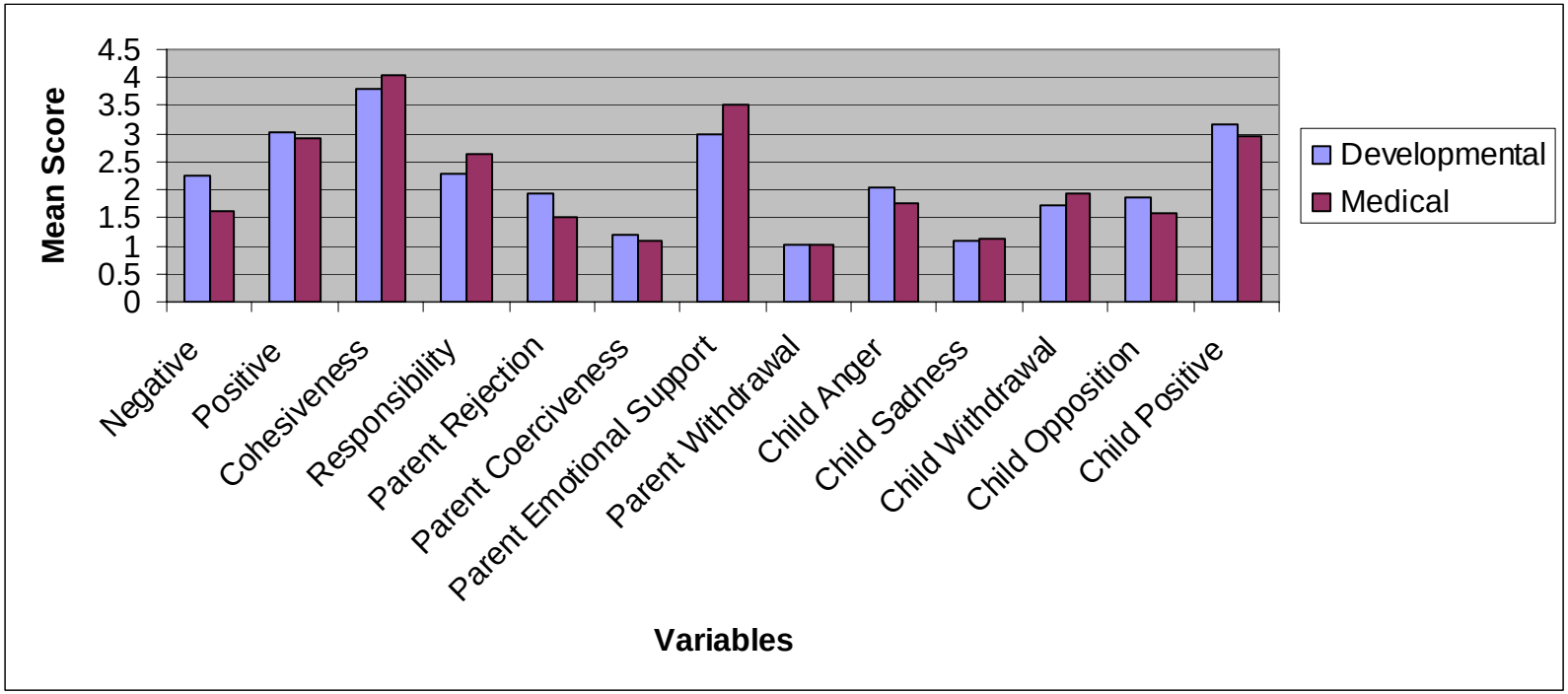

Figure 4.

Differences in SCIFF Mean Scores between Developmental and Medical Task

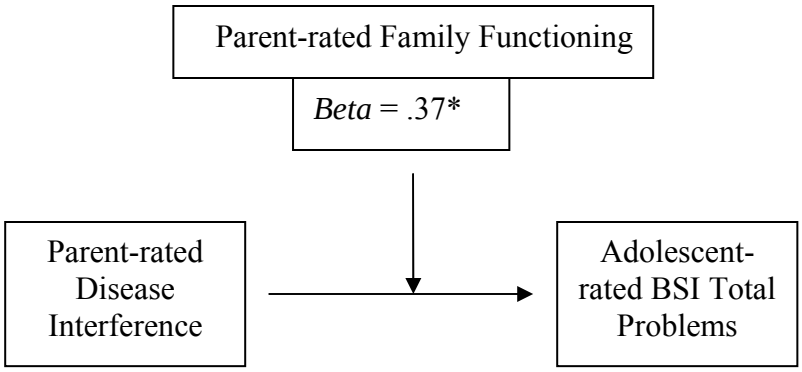

Figure 5.

Moderation of the Association of Parent-rated Disease Interference and Adolescent-rated BSI Total Problems by Parent-rated Family Functioning

Figure Captions

Appendix A: File Review Data Sheet

Date of Birth _____ Date of Consent _____ Date of Review_____

VWD **Hemophilia**

Type:_____

_____

_____

Hemophilia	Factor Level	[ ]	5-30%
		[ ]	2-5%
		[ ]	< 2%
	Hemorrhage	[ ]	No spontaneous
		[ ]	After major trauma
		[ ]	Rare spontaneous
		[ ]	After minor trauma
		[ ]	Spontaneous
		[ ]	Any trauma
VWD		[ ]	Type 1 VWD
		[ ]	2N VWD
		[ ]	3 VWD
		[ ]	Low VIII levels

Date <u>One Year</u> prior to Consent _____

# Hospitalizations _____	# Phone calls _____
# Home visits _____	# Transfusions_____
#Outpatient visits _____	# Spontaneous Bleeds_____
# of medications_____	# Prolonged Bleeds_____
# School absences_____	# Factor replacement_____

Types of Medications

Prescription Type Dosage

_____
_____
_____

Notable memos in chart regarding psychological problems or delays_____
_____

Self-infusion (Age, other family members, who typically infuses the child):

Age_____

Others_____
_____
_____

Notes about discussions regarding transition_____
_____
_____

Medical insurance company/type_____

Medical Staff Rating

1. Compared to other adolescents with hemophilia/VWD how **severe** is this child's illness?

1 2 3 4 5
Not at All Extremely

2. How **much** does this child's illness interfere with his/her daily functioning?

1 2 3 4 5
Not at All Extremely

Appendix B: General Information Form (GIF) Parent

1. Today's date: _____

2. Child with Hemophilia/VWD
   a. Gender: Male Female (Circle one)
   b. Date of Birth _____
   c. Child's Age _____
   d. Does your child live at home? Yes No (Circle one)
      If not where do they live?_____

3. Which of the following best describes your child with hemophilia/VWD's ethnic background?

   [ ] White
   [ ] African-American
   [ ] Latino
   [ ] Asian-American
   [ ] Other _____

4. Number of members in child's household including parents, siblings, grandparents, aunts, uncles, and cousins if they reside in the household. _____

5. Number of other children in the household with hemophilia/VWD?

Age	Gender (Male or Female)	Relationship
_____	_____	_____
_____	_____	_____
_____	_____	_____

Primary Caregiver #1 (You)
Age: _____

6. Which of the following best describes your relationship to the child?

   [ ] Mother
   [ ] Father
   [ ] Grandparent
   [ ] Legal Guardian
   [ ] Other _____

7. Which of the following best describes Primary Caregiver #1's (your) ethnic background?

[ ] White
[ ] African-American
[ ] Latino
[ ] Asian-American
[ ] Other _____

8. What is the highest level of education completed by Primary Caregiver #1 (you)?

[ ] 1 – 8th grade
[ ] 9 – 12th grade
[ ] Some college/ vocational school
[ ] College graduate
[ ] Professional school/graduate school

9. Check and answer those applicable to Primary Caregiver #1 (you).

[ ] Married: What year? _____
[ ] Separated: What year? _____
[ ] Divorced: What year? _____
[ ] Widowed: What year? _____
[ ] Never Married
[ ] Remarried: What year? _____

10. What is your family's total annual income? (Check one)

[ ] Less than $10,000
[ ] $10,000 - $19,999
[ ] $20,000 - $34,999
[ ] $35,000 - $49,999
[ ] $50,000 - $74,999
[ ] $75,000 - $99,999
[ ] $100,000 - 124,999
[ ] Over $125,000

Primary Caregiver # 2

Age: _____

11. Which of the following best describes Primary Caregiver # 2's relationship to the child?

[ ] Mother
[ ] Father
[ ] Grandparent
[ ] Legal Guardian
[ ] Other _____

12. Which of the following best describes Primary Caregiver # 2's ethnic background?

[ ] White
[ ] African-American
[ ] Latino
[ ] Asian-American
[ ] Other _____

13. What is the highest level of education completed by Primary Caregiver # 2 ?

[ ] 1 – 8th grade
[ ] 9 – 12th grade
[ ] Some college/ vocational school
[ ] College graduate
[ ] Professional school/graduate school

14. Check and answer those applicable to Primary Caregiver # 2.

[ ] Married: What year? _____
[ ] Separated: What year? _____
[ ] Divorced: What year? _____
[ ] Widowed: What year? _____
[ ] Never Married
[ ] Remarried: What year? _____

These questions are about your child with hemophilia/VWD:

15. Does he/she have any learning problems? [ ] Yes [ ] No

16. What is the highest school grade, he/she has completed? _____
How many days of school has your child missed in the past 12 months? _____

17. Does he/she or did he/she ever receive special education services?
[ ] Yes [ ] No

Please describe briefly including type of placement e.g., learning disabled resource room:_____
_____
_____

18. Does your child have any psychological difficulties? [ ] Yes [ ] No
Please describe briefly_____
_____

19. Please list below your child's current extracurricular activities:

_____
_____
_____
_____
_____

20. What is the name of your child's illness? _____

21. How old was your child when the illness was diagnosed? _____

22. How long has your child been receiving care at the Hemophilia Clinical Program of St. Christopher's Hospital?

_____ years _____months

23. Has your hematology doctor spoken with you about transition from pediatric clinic to adult care?

[ ] Yes [ ] No

If so what briefly describe what you have discussed:_____
_____

24. When do you feel is the best age for adolescents with hemophilia/VWD to transition into an adult care program?

[ ] 12-16 years of age
[ ] 16-18 years of age
[ ] 18-21 years of age
[ ] 21-25 years of age
[ ] Older than 25 years of age

25. What age do you feel would be best for your child to transition into an adult care program? _____ years

26. Please list the type of insurance coverage your child currently has:

_____

27. Please list below all activities that you took primary responsibility for related to the care and treatment of your child's illness in the past 12 months. Include information on how frequently you engaged in these activities.

_____
_____
_____

28. Please list below all activities that your child took primary responsibility for related to the care and treatment of his/her illness in the past 12 months. Include information on how frequently he/she engaged in these activities.

_____
_____
_____

29. Does your child regularly take any prescription medications? [ ] Yes [ ] No
Please list medications:

_____
_____

If your child does not need infusions please skip to question number 34.

30. How old was your child when they learned to self-infuse? _____ years

31. How often does your child use factor replacement products?_____

32. How often does your child self-infuse when you or another caregiver are with him/her?

[ ] Never
[ ] Rarely
[ ] Some of the time
[ ] Most of the time
[ ] All of the time

33. If your child does not always self-infuse, please describe briefly why others infuse?

_____

34. Compared to other adolescents with hemophilia/VWD how **severe** is your child's illness?

1 2 3 4 5

Not at All Extremely

35. How **much** does your child's illness interfere with your child's daily functioning?

1 2 3 4 5

Not at All Extremely

Appendix C: General Information Form (GIF) Adolescent

1. Today's date: _____

Please answer the following questions about your illness and care.

2. Please list below your current extracurricular activities:

_____
_____
_____

3. What is the name of your illness?_____

4. How was old were you when the illness was diagnosed? _____

5. How old were you when you first learned about your illness? _____

6. How long have you been receiving care at the Hemophilia Clinical Program at St. Christopher's Hospital?

_____ years _____months

7. Has your hematology doctor spoken with you about transition from pediatric clinic to adult care?

[ ] Yes [ ] No

If so what briefly describe what you have discussed:_____

8. When do you feel is the best age for adolescents with hemophilia to transition into an adult care program?

[ ] 12-16 years of age
[ ] 16-18 years of age
[ ] 18-21 years of age
[ ] 21-25 years of age
[ ] Older than 25 years of age

9. What age do you feel would be best for you to transition into an adult care program?

___years

10. Please list the type of insurance coverage you currently have:_____

11. Please list below all activities that you took primary responsibility for related to the care and treatment of your illness in the past 12 months. Include information on how frequently you engaged in these activities._____
_____
_____

12. Please list below all activities that your parents took primary responsibility for related to the care and treatment of your illness in the past 12 months. Include information on how frequently they engaged in these activities._____
_____
_____

13. Do you regularly take any prescription medications? [ ] Yes [ ] No
Please list medications:
_____
_____

If you do not need infusions please skip to question number 34.

14. How old were you when you learned to self-infuse? _____ years

15. How often do you use factor replacement products?_____

16. How often do you self-infuse when your parents or another caregiver are with you?

[ ] Never
[ ] Rarely
[ ] Some of the time
[ ] Most of the time
[ ] All of the time

17. If your do not always self-infuse, please describe briefly why others infuse for you?
_____
_____

18. Compared to other adolescents with hemophilia/VWD how **severe** is your illness?
1 2 3 4 5

Not at All Extremely

19. How **much** does your illness interfere with your daily functioning?
1 2 3 4 5

Not at All Extremely

Appendix D: Transition Questionnaire Parent

Transition Concerns and Feelings

A. Concerns

Sometimes adolescents and parents have concerns and questions about what it would be like to have a child move from a child and adolescent medical care program to an adult program. Please read through the following questions and indicate what concerns you have about your child being transitioned into an adult care program.

1. I think that my child will not have enough information to know what to expect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I wonder if my child will be able to take responsibility for him/herself.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. I wonder if my child will be able to make decisions on his/her own.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I wonder if my child will have a tough time getting the health care people to talk to him/her instead of me.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. I wonder if my child will know how to get or ask for the information he/she needs.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. I have concerns about my child being treated as an adult.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. I am not concerned about my child talking to other people about his/her health alone.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. I am concerned about how my child would get back and forth from his/her care program.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. I am <u>not</u> concerned about how my child will pay for his/her medical care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. I am concerned about whether health care professionals will allow me to talk with them about my child's medical care and include me in my child's health care appointments.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. I am concerned that the adult health care people might not know my child and not understand how hemophilia or VWD works.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. I am concerned about my child being seen as drug seeking when he/she is in pain or needs medications.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. I do no want my child to leave the health care people we have known for a very long time and who understand my child/family.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. I am scared about getting to know new health care providers.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

15. I am afraid new staff would not believe me or my child.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

B. Feelings

Imagine for a minute that you are finding out that your child is going to be transitioned into an adult care program. Tell us how you <u>might feel</u> when you hear about this transition.

1. I will be excited/happy.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I will feel afraid.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. I will be relieved.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I will feel unsure.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. I will have <u>no</u> feelings one-way or the other.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. I will be worried

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. I will be nervous.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. I feel it will be the right time for my child to move on.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. I will be anxious.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. I will be okay with the idea.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. I will be angry.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. I will feel deserted/abandoned.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. I will feel I had no control over the decision.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. Other feelings:_____

C. Readiness

1. I feel that my child will adjust to transition into an adult health care system very well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I feel that I will be able to adjust to this transition very well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

Transition Programs

A. Reasons for Transition Program

1. Do you think a program that will help your child and other adolescents with hemophilia or VWD go from a child and adolescent treatment center to an adult treatment center is needed? (Please check one):

☐YES ☐ NO

2. Do you think a program that helps you and other parents of children and adolescents with hemophilia or VWD go from a child and adolescent treatment center to an adult center is needed?

☐YES ☐ NO

Please read the following questions and indicate why you think such a transition program is <u>or</u> is not needed.

3. It would make it easier to transition care and help me to prepare my adolescent for the transition.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I would know more about what to expect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. It would provide more support in helping my child meet his/her health care needs.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. It would help my child to be treated as an adult.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. It would give my child more control over his/her health care decisions and life.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. It would give me the chance to meet other parents of adolescents with hemophilia or VWD who are doing well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. It would not help me that much because enough help is already provided.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. It would help me prepare for my child's transition.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

Other reasons to have/not have this program. (Please state them below.)

_____

_____

B. Components of Transition Program

If a transition program was provided to you and your child, what are some of the ways and/or kinds of help it should offer.

1. It should provide information about adult health care programs.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. It should provide opportunities to allow my child and myself to meet adult health care providers.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. It should include information about ways that I can help my child solve problems he/she is having with his/her medical health care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. It should not provide information to help my child to care for his/her own health care needs independently.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. It should help my child to make health care decisions on his/her own.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. It should not help me to learn more about hemophilia or VWD.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. It should provide information to me about ways that I can help my child talk to and educate others about his/her medical condition.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. It should provide me with someone who I can talk to about transitioning or other problems.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. It should work with me to help me facilitate my child's independence in their medical health care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. It should provide time for me to discuss my child's health care needs and my feelings about my child being a new patient with an adult medical health care provider.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. It should provide me with information about how to handle and deal with other health care professionals like the emergency room, insurance companies, hospital staff ect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. Transition programs should focus on my child's concerns about how his/her disease may affect his/her decision to have children in the future and his/her feelings about the genetic nature of their disorder.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. Transition programs should focus on learning more about hemophilia or VWD (i.e., learning more about the medical nature of the disorder).

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. It would help if transition programs included information about vocational and career choices for my child.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

15. Other ways and or help you feel the program should offer:_____

Adolescent Medical and Self-Care

A. Functional Skills

1. Please describe what your child does to treat his/her **non-hemophilia/VWD** problems (i.e. a cold).

_____Tries to take care of it independently without involving parents or calling the doctor/nurse.
_____Tries to take care of it independently without involving parents, but calls the doctor/nurse.
_____Tries to take care of it at home with parents help, without calling the doctor/nurse.
_____Tries to take care of it at home with parents help, but also calls the doctor/nurse.
_____Does nothing and takes his/herself to the clinic, doctor's office, or calls home health nurse.

2. Please describe what your child does if he/she has a "**not so bad**" or **mild** problem related to his/her hemophilia or VWD (i.e., bruising).

_____Tries to take care of it independently without involving parents or calling the doctor/nurse.
_____Tries to take care of it independently without involving parents, but calls the doctor/nurse.
_____Tries to take care of it at home with parents help, without calling the doctor/nurse.
_____Tries to take care of it at home with parents help, but also calls the doctor/nurse.
_____Does nothing and takes his/herself to the clinic, doctor's office, or calls home health nurse.

3. Please describe what your child does when he/she has hemophilia or VWD related **pain**.

_____Tries to take care of it independently without involving parents or calling the doctor/nurse.
_____Tries to take care of it independently without involving parents, but calls the doctor/nurse.
_____Tries to take care of it at home with parents help, without calling the doctor/nurse.
_____Tries to take care of it at home with parents help, but also calls the doctor/nurse.
_____Does nothing and takes his/herself to the clinic, doctor's office, or calls home health nurse.

4. Please describe what your child does when he/she has a "**moderate**" problem related to his/her hemophilia or VWD (i.e., more severe nose bleed, minor swelling resulting from a bleed).

_____Tries to take care of it independently without involving parents or calling the doctor/nurse.
_____Tries to take care of it independently without involving parents, but calls the doctor/nurse.
_____Tries to take care of it at home with parents help, without calling the doctor/nurse.

_____Tries to take care of it at home with parents help, but also call the doctor/nurse.
_____Does nothing and takes his/herself to the clinic, doctor's office, or calls home health nurse.

5. Please describe what your child does when he/she has a "**severe**" problem related to his/her hemophilia/VWD (i.e., swollen joint that is spontaneous or following injury, injury to head).
_____Tries to take care of it independently without involving parents or calling the doctor/nurse.
_____Tries to take care of it independently without involving parents, but calls the doctor/nurse.
_____Tries to take care of it at home with parents help, without calling the doctor/nurse.
_____Tries to take care of it at home with parents help, but also call the doctor/nurse.
_____Does nothing and takes his/herself to the clinic, doctor's office, or calls home health nurse.

6. If you said your child takes care of his/her "**mild**", "**moderate**", or "**severe**" problems with hemophilia/VWD at home, please describe what he/she or you do (If he/she does not take care of it at home, skip to '7' below).
a. To take care of mild problems at home _____
b. To take care of moderate problems at home_____
c. To take care of severe problems at home_____

7. If you said your child takes care of his/her pain related to hemophilia/VWD at home, please describe what he/she or you do (If he/she does not take care of it at home, skip to '8' below):

a. To take care of pain problems at home_____

8. Do you think there are things your child can do to reduce the number of problems he/she has related to his/her hemophilia/VWD? (Please check only one). ☐ YES ☐ NO

What types of things can your child do to reduce the number of problems he/she has with his/her medical condition:_____

9. How often do you think your child should have a general health check-up (a visit not following an emergency room visit or hospital visit). (Please check only one)
_____Once a year
_____Twice a year (Every six months)
_____Three times a year (Every four months)
_____Four times a year (Every three months)
_____Only when he/she needs to

10. Think for a moment about when your child had a clinic visit, after these visits how well would say your child follows the doctor's instructions from that time to the next visit. (Please check only one).
_____Very well (He/she follows instructions **all** of the time).
_____Well (He/she follows instructions **most** of the time).
_____Somewhat (He/she follows instructions **some** of the time).
_____Not too well (He/she **rarely** follows instructions).
_____Not at all well (He/she **does not** follow instructions).

11. I know my child's hemoglobin factor level or VWD type. ☐ YES ☐ NO

Please indicate your child's factor level or VWD type_____

If your child has hemophilia:

Does your child have **mild** **moderate** **severe** hemophilia.

Does your child have **hemophilia A** or **hemophilia B**

B. Adolescent Health Care List

Below are statements about thoughts and behaviors your child may or may not have/do regarding his/her health. Please circle the answer that best describes how true the following statements are.

1. My child always takes care of all of his/her own health care needs.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

2. My child wants to know more about how his/her hemophilia/VWD affects him/her.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

3. My child is responsible for administering his/her own medications or self-infuses factor products.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

4. It is easy for my child to talk with his/her doctor.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

5. I feel that my child has little or no control over how hemophilia/VWD affects him/her.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

6. I allow my child to speak with his/her doctor alone during visits.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

7. I allow my child to go to the doctors or clinic by themselves. (If child does not drive: I will allow my child to go to the doctors or clinic by themselves when they are old enough to drive).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

8. My child gets embarrassed if he/she has to take medications in front of his/her friends.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

9. My child administers medications (i.e. nasal spray) or self-infuses factor as prescribed, independently.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

10. I feel my child having hemophilia/VWD will affect what happens to him/her in the future.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

11. When my child has an emergency he/she knows how to get in contact with the right medical people.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

12. My child is responsible for getting him/herself to and from the doctor or clinic.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

13. My child is responsible for getting his/her own medicine and medical supplies.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

14. My child makes his/her own health care appointments.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

15. My child knows how street drugs and/or alcohol affects his/her hemophilia/VWD.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

16. My child has access to sex education and birth control information when he/she needs it.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

17. My child tells his/her friends about his/her condition.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

18. My child takes proper precautions when playing sports or doing other activities without parental supervision.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

19. My child tells teacher/employers or potential employers about his/her condition.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

20. I believe having hemophilia/VWD will make it harder for my child to live away from home.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

21. I feel my child can take care of his/her condition by him/herself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

22. I feel my child can handle the times when he/she has pain.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

23. My child can go to the clinic by him/herself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

24. If my child has a problem with people in the clinic or from home health care, he/she does not know what to do or who to contact.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

25. My child knows how to talk to a medical person or anyone else about how hemophilia/VWD affects him/her.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

26. I am allowing my child to choose which doctor he/she will use for an adult doctor.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

27. My child knows about his/her insurance and carries medical coverage and other identification with him/her.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

28. My child makes sure he/she understands the doctor or nurses instructions.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

29. My child makes sure all the medical questions he/she have are answered.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

C. Efficacy Questions

The following questions ask about how sure you are about how your child can deal day-to-day with hemophilia or VWD. There are no right or wrong answers, we just want to know what you think. So for each question tell us how true the following statements are.

1. I am sure that my child can do something to stop or reduce a bleeding episode.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

2. I am sure that my child can keep doing most of the things he/she does day-to-day.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

3. I am sure that my child can reduce hemophilia/VWD pain by using methods other than taking extra medication.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

4. I know my child can do something to help him/herself feel better if he/she is feeling sad or blue.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

5. As compared to other people with hemophilia/VWD my child can manage his/her life from day-to-day.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

6. My child can manage his/her hemophilia/VWD symptoms so that he/she can do enjoyable things.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

7. My child cannot deal with the frustration of having hemophilia/VWD.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

8. I am sure that my child feels that he/she can talk to me about his/her treatment concerns.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

9. My child can solve medical problems alone, and ask for help if the complications progress.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

10. My child can decide on how best to treat his/her medical symptoms (i.e., deciding to use factor replacement).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

11. I am sure that my child can speak with medical professionals about his/her health care concerns.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

12. I feel my child is able to deal with doctors and nurses by him/herself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

13. I feel my child knows enough about how hemophilia/VWD affects him/her.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

14. I think that my child can handle how hemophilia/VWD will affect important areas of his/her life in the future (i.e., marriage, having a family).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

Appendix E: Transition Questionnaire Adolescent

Transition Concerns and Feelings

A. Concerns

Sometimes adolescents and parents have concerns and questions about what it would be like to go from a child and adolescent medical care program to an adult program. Please read through the following questions and indicate what concerns you have about being transitioned into an adult care program.

1. I think that I will not have enough information to know what to expect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I wonder if I will have to take responsibility for myself.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. I wonder if I will be able to make decisions on my own.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I wonder if I will have a tough time getting the health care people to talk to me instead of my parents.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. I wonder if I will know how to get or ask for the information I need.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. I have concerns about being treated as an adult.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. I am not concerned about talking to other people about my health.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. I am concerned about how I would get back and forth from my care program.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. I am not concerned about how I will pay for my medical care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. I am concerned about whether or not my parents will let me answer questions or talk to the health care people myself.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. I am concerned that the adult health care people might not know me and not understand how hemophilia or VWD works.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. I am concerned about being seen as drug seeking when I am in pain or need medications.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. I do no want to leave the health care people I have know for a very long time and who understand me.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. I am scared about getting to know new health care providers.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

15. I am afraid new staff would not believe me.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

B. Feelings

Imagine for a minute that you are finding out that you are going to be transitioned into an adult care program. Tell us how you might feel when you hear about this transition.

1. I will be excited/happy.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I will feel afraid.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. I will be relieved.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I will feel unsure.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. I will have no feelings one way or the other.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. I will be worried

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. I will be nervous.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. I feel it will be the right time to move on.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. I will be anxious.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. I will be okay with the idea.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. I will be angry.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. I will feel deserted/abandoned.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. I will feel I had no control over the decision.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. Other feelings:_____

C. Readiness

1. I feel that I will adjust to transition into an adult health care system very well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. I feel that my parents will be able to adjust to this transition very well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

Transition Programs

A. Reasons for Transition Program

1. Do you think a program that will help you and other adolescents with hemophilia or VWD go from a child and adolescent treatment center to an adult treatment center is needed? (Please check one):

☐YES ☐ NO

2. Do you think a program that helps your parents and other parents of children and adolescents with hemophilia or VWD go from a child and adolescent treatment center to an adult center is needed?

☐YES ☐ NO

Please read the following questions and indicate why you think such a transition program is <u>or</u> is not needed.

3. It would make it easier to transition care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. I would know more about what to expect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. It would provide more support in helping me meet my health care needs.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. It would help me to be treated as an adult.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. It would give me more control over my health care decisions and life.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. It would give me the chance to meet young adults with hemophilia or VWD who are doing well.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. It would <u>no</u>t help me that much because enough help is already provided.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. It would help my parents prepare for transition.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

Other reasons to have/not have this program. (Please state them below.)

_____

_____

B. Components of Transition Program

If a transition program was provided to you and your parents, what are some of the ways and/of kinds of help it should offer. (Please check <u>all</u> that apply):

1. It should provide information about adult health care programs.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

2. It should provide opportunities to allow my parents and myself to meet adult health care providers.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

3. It should include information about ways that I can solve problems I am having with my medical health care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

4. It should <u>not</u> provide information to help me care for my own health care needs independently.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

5. It should help me to make health care decisions on my own.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

6. It should <u>not</u> help me to learn more about hemophilia or VWD.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

7. It should provide information to me about ways that I can talk to and educate others about my medical condition.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

8. It should provide me with someone who I can talk to about transitioning or other problems when I need it.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

9. It should work with my parents to help them to allow me to take a more independent role in my health care.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

10. It should provide time for me to discuss my health care needs and feelings about being a new patient with an adult medical health care provider.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

11. It should provide me with information about how to handle and deal with other health care professionals like the emergency room, insurance companies, hospital staff ect.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

12. Transition programs should focus on my concerns about how my disease may affect my decision to have children in the future and my feelings about the genetic nature of my disorder.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

13. Transition programs should focus on learning more about hemophilia or VWD (i.e., learning more about the medical nature of the disorder).

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

14. It would help if transition programs included information about vocational and career choices for me.

0	1	2	3	4
Not true	Unlikely	Maybe	True	Very True

15. Other ways and or help you feel the program should offer:_____

A. Functional Skills

1. Please describe what you do to treat your **non-hemophilia/VWD** problems (i.e. a cold).

_____Try and take care of it independently without involving my parents or calling the doctor/nurse.
_____Try and take care of it independently without involving my parents, but call the doctor/nurse.
_____Try and take care of it at home with my parents help, without calling the doctor/nurse.
_____Try and take care of it at home with my parents help, but also call the doctor/nurse.
_____Do nothing and take myself to the clinic, doctor's office, or call my home health nurse.

2. Please describe what you do when you have a "**not so bad**" or **mild** problem related to your hemophilia or VWD (i.e., bruising).

_____Try and take care of it independently without involving my parents or calling the doctor/nurse.
_____Try and take care of it independently without involving my parents, but call the doctor/nurse.
_____Try and take care of it at home with my parents help, without calling the doctor/nurse.
_____Try and take care of it at home with my parents help, but also call the doctor/nurse.
_____Do nothing and take myself to the clinic, doctor's office, or call my home health nurse.

3. Please describe what you do when you have hemophilia or VWD related **pain**.

_____Try and take care of it independently without involving my parents or calling the doctor/nurse.
_____Try and take care of it independently without involving my parents, but call the doctor/nurse.
_____Try and take care of it at home with my parents help, without calling the doctor/nurse.
_____Try and take care of it at home with my parents help, but also call the doctor/nurse.
_____Do nothing and take self to the clinic, doctor's office, or call my home health nurse.

4. Please describe what you do when you have a "**moderate**" problem related to your hemophilia or VWD (i.e., more severe nose bleed, minor swelling resulting from a bleed).

_____Try and take care of it independently without involving my parents or calling the doctor/nurse.
_____Try and take care of it independently without involving my parents, but call the doctor/nurse.
_____Try and take care of it at home with my parents help, without calling the

doctor/nurse.
_____Try and take care of it at home with my parents help, but also call the doctor/nurse.
_____Do nothing and take myself to the clinic, doctor's office, or call my home health nurse.

5. Please describe what you do when you have a "**severe**" problem related to your hemophilia/VWD (i.e., swollen joint that is spontaneous or following injury, injury to the head).
_____Try and take care of it independently without involving my parents or calling the doctor/nurse.
_____Try and take care of it independently without involving my parents, but call the doctor/nurse.
_____Try and take care of it at home with my parents help, without calling the doctor/nurse.
_____Try and take care of it at home with my parents help, but also call the doctor/nurse.
_____Do nothing and take myself to the clinic, doctor's office, or call my home health nurse.

6. If you said you take care of your "**mild**", "**moderate**", or "**severe**" problems with hemophilia/VWD at home, please describe what you or your parents do (If you do not take care of it at home, skip to '7' below).

a.To take care of mild problems at home_____
b.To take care of moderate problems at home _____
c.To take care of severe problems at home_____

7. If you said you take care of your pain related to hemophilia/VWD at home, please describe what you or your parents do (If you do not take care of it at home, skip to '8' below):

a. To take care of pain problems at home _____

8. Do you think there are things you can do to reduce the number of problems you have related to your hemophilia/VWD? (Please check only one). ☐ YES ☐ NO

What types of things can you do to reduce the number of problems you have with your medical condition:_____

9. How often do you think you should have a general health check-up (a visit not following an emergency room visit or hospital visit). (Please check only one)
_____Once a year
_____Twice a year (Every six months)
_____Three times a year (Every four months)
_____Four times a year (Every three months)
_____Only when I need to

10. Think for a moment about when you have a clinic visit, after these visits how well would say you follow the doctor's instructions from that time to the next visit. (Please check only one).
_____Very well (I follow instructions **all** of the time).
_____Well (I follow instructions **most** of the time).
_____Somewhat (I follow instructions **some** of the time).
_____Not too well (I **rarely** follow instructions).
_____Not at all well (I **do not** follow instructions).

11. I know my hemoglobin factor level or my VWD type. ☐ YES ☐ NO

Please indicate your type _____

If you have hemophilia:

Do you have **mild** **moderate** **severe** hemophilia.

Do you have **hemophilia A** or **hemophilia B**

B. Adolescent Health Care List

Below are statements about thoughts and behaviors you may or may not have/do regarding your health. Please circle the answer that best describes how true the following statements are.

1. I always take care of all of my health care needs.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

2. I want to know more about how my hemophilia/VWD affects me.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

3. I am responsible for administering my own medications or self-infusing factor products.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

4. It is easy for me to talk with my doctor.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

5. I feel I have little or no control over how hemophilia/VWD affects me.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

6. I speak with my doctor alone during visits.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

7. I am allowed to go to the doctors or clinic by myself. (If you do not drive: I will be allowed to go to the doctors or clinic by myself when I am old enough to drive).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

8. I get embarrassed if I have to take my medications in front of my friends.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

9. I administer my medications (i.e., nasal spray) as prescribed or self-infuse factor, independently.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

10. I feel that having hemophilia/VWD will affect what happens to me in the future.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

11. When I have an emergency I know how to get in contact with the right medical people.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

12. I am responsible for getting myself to and from the doctor or clinic.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

13. I am responsible for getting my own medicine and medical supplies.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

14. I make my own health care appointments.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

15. I think about how street drugs and/or alcohol affects my hemophilia/VWD.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

16. I get sex education and birth control information when I need it.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

17. I tell my friends about my condition.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

18. I do what I need to do to play sports or do other activities safely.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

19. I tell teacher/employers or potential employers about my condition.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

20. I believe having hemophilia/VWD will make it harder for me to live away from my parents.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

21.I feel I can take care of my condition by myself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

22. I feel I can handle the times when I have pain.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

23. I can go to the clinic by myself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

24. If I have a problem with people in the clinic or from home health care, I do not know what to do or who to contact.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

25. I know how to talk to a medical person or anyone else about how hemophilia/VWD affects me.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

26. I am considering which doctor I will use for my adult doctor.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

27. I carry my medical coverage and other identification with me.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

28. I make sure I understand the doctor or nurses instructions.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

29. I make sure all the medical questions I have are answered.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

C. Efficacy Questions

The following questions ask about how sure you are in dealing day-to-day with hemophilia or VWD. There are no right or wrong answers, we just want to know what you think. So for each question tell us how true the following statements are.

1. I am sure that I can do something to stop or reduce a bleeding episode.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

2. I am sure that I can keep doing most of the things I do day-to-day.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

3. I am sure that I can reduce my hemophilia/VWD pain by using methods other than taking extra medication.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

4. I can do something to help myself feel better if I am feeling sad or blue.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

5. As compared to other people with hemophilia/VWD I can manage my life from day-to-day.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

6. I can manage my hemophilia/VWD symptoms so that I can do the things I enjoy doing.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

7. I cannot deal with the frustration of having hemophilia/VWD.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

8. I am sure that I can talk to my parents about my treatment concerns.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

9. I can solve medical problems alone, and ask for help if the complications progress.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

10. I can decide on how best to treat my medical symptoms (i.e., deciding to use factor replacement).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

11. I am sure that I can speak with medical professionals about my health care concerns.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

12. I feel I am able to deal with doctors and nurses by myself.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

13. I feel I know enough about how hemophilia/VWD affects me.

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

14. I think that I can handle how hemophilia/VWD will affect important areas of my life in the future (i.e., marriage, having a family).

0	1	2	3	4
Not true	Unlikely	Sometimes	True	Very True

Vita

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Publications

Barakat, L.P., Lutz, M.J., Nicolaou, D.C., & Lash, L.A. (2005). Parental locus of control and family functioning in the quality of life of children with sickle cell disease. *Journal of Clinical Psychology in Medical Settings. In press*.

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