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Measurement of Disease Specific Social Support in Adolescents with Cystic Fibrosis

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MEASUREMENT OF DISEASE SPECIFIC SOCIAL SUPPORT IN ADOLESCENTS WITH CYSTIC FIBROSIS

By

David H. Barker

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MEASUREMENT OF DISEASE SPECIFIC SOCIAL SUPPORT IN ADOLESCENTS CYSTIC FIBROSIS

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This study documented the creation and initial validation of the Perceived Adolescent Social Support: Cystic Fibrosis (PASS-CF) inventory. The inventory was developed from semi-structured interviews of adolescents with cystic fibrosis (CF) and measured both supportive and non-supportive behaviors provided to adolescents by their family and friends. This study reports the findings from these interviews, results of the pilot testing of the measure, exploratory analyses of the utility of individual items, and the relationships between supportive and non-supportive behaviors and important clinical outcomes, such as treatment adherence, health-related quality of life (HRQoL), and other health outcomes. In particular, the study compared two measurement models suggested by popular definitions of social support. The "perceived support" model emphasized adolescents' cognitive appraisals of the support provided to them by family and friends, and the functional support model emphasized the utility of specific behaviors in managing CF. Results provided support for both models and provided insights into important next steps in the study of social support in adolescents with CF.

Dedication

I dedicate this work to my wife Linsi and three boys Bryce, Kevin and Zack. Thank you for your love and support.

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Chapter 1

Review of the Literature

Managing a chronic illness is stressful for the individuals affected, as well as their families and friends (Kazak, Rourke, & Navsaria, 2009; Reiter-Purtill, Waller, & Noll, 2009). Chronic illness adds to the daily responsibilities of individuals and families, changes daily routines and social roles, strains resources, and can limit work-related and recreational activities. Successfully navigating these challenges requires flexibility and support from family and friends. This support may be needed for many different tasks (e.g., providing medication, reminding to do treatments, empathic listening, facilitating recreational activities) and can be provided by different individuals including family, friends, teachers, and medical teams (Duncan, Duncan, & Strycker, 2005). Recent theory suggests that to be helpful, support must match the needs of the individual (Decker, 2007). These needs are likely to vary depending on the demands of the particular illness, but to date, few studies have examined how illness-specific support relates to disease management and health outcomes (Gallant, 2003).

Understanding the relationships between illness-specific support and disease management is particularly timely given the ongoing challenge of improving youths' adherence to their daily treatments (DiMatteo, 2004a; Kripalani, Yao, & Haynes, 2007; McDonald, Garg, & Haynes, 2002; Modi & Quittner, 2006a; Osterberg & Blaschke, 2005). Poor treatment adherence is a major contributor to both treatment failure and increased morbidity in adolescents with chronic illness conditions (DiMatteo, Giordani, Lepper, & Croghan, 2002; Simpson et al., 2006). The importance of family and peer influences has been highlighted across illness groups (De Civita & Dobkin, 2004; Kazak et al., 2009), and is particularly evident in the substantial decrease in adherence during adolescence (La Greca & Mackey, 2009).

Adolescence is a time of transition, marked by the increasing influence of peers and friends and more time spent outside of the home, resulting in increased independence and less parental supervision (Modi et al., 2008; Shroff Pendley et al., 2002). Consequently, routine patterns of disease management can be disrupted, and roles and responsibilities redefined. Amidst the turmoil of this transition, supportive behaviors that facilitated disease management may decrease or disappear, and obstructive behaviors may appear or increase. Family and friends may also reallocate support provision, with friends taking on a more important role (Gallant, 2003). Identifying which types of supportive behaviors and sources of support are most effective during this transition may help teens, families and clinicians navigate the challenges of adolescence while maintaining appropriate disease management.

Similar to other chronic illnesses, adolescents with cystic fibrosis (CF) show a considerable decrease in adherence (Zindani, Streetman, Streetman, & Nasr, 2006). The consequences of non-adherence are significant for these youth, given that there is also an increase in hospitalizations, a marked decrease in lung function, and increased morbidity related to the illness during adolescence (Quittner, Barker, Marciel, & Grimley, 2009). Surprisingly, few studies have directly assessed social support in this population, and no study has examined the relationships between social support and disease management (Gallant, 2003; Graetz, Shute, & Sawyer, 2000). The purpose of this dissertation was to develop and validate an illness-specific inventory of supportive and non-supportive behaviors for adolescents with CF.

Brief Overview of Cystic Fibrosis

CF is the most common life-shortening, autosomal recessive condition in Caucasian populations, affecting approximately 30,000 individuals in the United States (Suaud & Rubenstein, 2010). The mutations in the cystic-fibrosis-transmembrane-conductance-regulator gene interfere with the transport of water and salt across the cell membranes of epithelial cells in the pancreas, intestine, liver, reproductive tract, lungs, and sweat glands (Suaud & Rubenstein, 2010). Disruption of this transport process leads to the accumulation of thick secretions which contribute to organ damage resulting in morbidity and shortened life span. The major cause of death is due to recurrent infections in the lungs, which eventually lead to respiratory failure. Advances in the medical management over the last 20 years have resulted in longer life expectancy for patients who adhere to their treatments (37.8 years; Cystic Fibrosis Foundation, 2009). These gains in life span underscore the importance of helping teens adhere to their daily treatments.

CF is among the most complex, time-consuming pediatric chronic conditions to manage, requiring substantial daily effort to complete daily medications and treatments, taking between 2 to 4 hours per day (i.e., airway clearance, nebulized medications, oral medications; Modi & Quittner, 2006a). Proper disease management also requires dietary changes to increase caloric intake to 110-200% of the recommended daily allowance (Stallings, Stark, Robinson, Feranchak, & Quinton, 2008). In addition to the daily treatment regimen, patients attend quarterly clinic visits. CF is also marked by frequent pulmonary infections which require intravenous (IV) antibiotic treatment and extended hospital stays (Quittner et al., 2009).

Managing hospitalizations, periods of sickness, and time-consuming daily treatments not only require significant support from family, but limit the time youth have for socializing with friends and peers (Carson & Hieber, 2001). These limitations likely interfere with the provision of support from friends and peers during a time when this support is most needed. Moreover, due to the risk of spreading life-threatening, antibiotic resistant bacteria, infection control policies recommend complete segregation among patients with CF, effectively eliminating contact between patients (Saiman & Siegel, 2004). These policies, while medically necessary, limit the possibility of support from other adolescents with CF and increase teens' sense of social isolation (Russo, Donnelly, & Reid, 2006). In sum, the complexity and burdensome nature of the treatment regimen, paired with the well-documented decline in treatment adherence and reduced time with peers, suggest that understanding the impact of social support may be a key to improving treatment adherence in youth with CF.

Overview and Measurement of Social Support

Social support has been shown to play an important role in how individuals adapt to stress (Cohen & Wills, 1985; Kessler, Price, & Wortman, 1985) and there is a vast literature addressing its influence on psychological well-being (Cohen & Wills, 1985; Coyne & Downey, 1991). Fewer studies have focused on how social support influences disease management, and findings from these studies have been inconsistent (Gallant, 2003; Nausheen, Gidron, Peveler, & Moss-Morris, 2009). Additionally, this literature has been criticized for not adequately explaining the mechanisms through which support influences adherence and health outcomes (Decker, 2007; DiMatteo, 2004b; Glasgow, Strycker, Toobert, & Eakin, 2000; Simoni, Frick, & Huang, 2006). This lack of theory has resulted in different definitions and measures of support. The definitions that have received the most attention include those that emphasize individuals' perceptions of support, followed by those that focus on the functional role of specific supportive behaviors (e.g., providing reminders about medications; Penninx, Kriegsman, van Eijk, Boeke, & Deeg, 1996). These two popular definitions imply different measurement models and mechanisms through which support influences disease management. Understanding which mechanisms influence disease management will facilitate the creation of targeted interventions to improve management, and by extension, health outcomes (Quittner, 2000).

How support is measured depends on which definition is used. Definitions that focus on perceived support suggest a measurement model that focuses on individuals' evaluations of the support they receive. These evaluations can be defined as a latent construct that influences respondents' perceptions of specific supportive behaviors (Figure 1.1). Consequently, specific behaviors are deemphasized because they are thought to be the vehicle used to access individuals' perceptions of support. Measures based on this model typically combine ratings of specific behaviors into composite scores that are expected to demonstrate strong internal consistency.

This model also implies that social support influences health outcomes by way of individuals' perceptions about the support that is being provided. Studies using this model have reported strong and consistent links among measures of perceived social support and measures of other social-cognitive processes (i.e., perceived distress, self-efficacy, control-beliefs, trait anxiety, positive state of mind; Gonzalez et al., 2004; Lakey & Cassady, 1990), as well as self-reported outcomes that have significant emotional and cognitive contributions (depression, HRQoL; Varni, Rubenfeld, Talbot, & Setoguchi, 1989; Zeller & Modi, 2006), but reported inconsistent links with disease management (DiMatteo, 2004b; Gallant, 2003).

In contrast to models emphasizing individuals' perceptions, the functional support model places more emphasis on the specific behaviors that are provided by supportive others. Using this model, the utility of each behavior is important in its own right and is thought to directly influence the desired outcome. For example, reminding teens to take medications may directly influence how frequently they take them, regardless of how "supportive" they perceive the reminders to be. In essence, the frequency of each behavior is viewed as an unobserved construct measured by participants' reports of frequency—the behavior, rather than the *perception* of the behavior, is the focus of the model (Figure 1.2). This model does not require individual behaviors to be correlated, suggesting that composite scales are likely to have low internal-consistency. Because the functional-support model focuses on the behavioral mechanisms that influence disease management, behaviors must be matched to the specific outcome they are thought to influence. Composite scores would dilute this "matching" between behaviors and outcomes and may not be appropriate for this model of support.

Many measures of social support are a mixture of both definitions. Participants are often asked both about their perceptions of supportive behaviors (Figure 1.1; e.g., Sherbourne & Stewart, 1991) and the frequency of these behaviors (Figure 1.2; e.g., Duncan et al., 2005). Most measures also yield composites of several supportive behaviors; an approach consistent with a perceptual definition of support (Gallant, 2003). This mixture of mechanisms has likely contributed to the inconsistent results reported by studies investigating the relationship between social support and disease management (Gallant, 2003).

Clearly specifying which mechanisms influence disease management has important methodological and treatment implications. Methodological considerations include decisions about which behaviors should be measured, whether participants should be asked about behavioral frequency or perceived supportiveness, whether a composite scale or a count of individual behaviors should be created, and whether behaviors should be matched to the specific outcomes they influence. It is also possible that these mechanisms differentially influence important emotional, social, and health outcomes. For example, perceptions of support may influence emotional outcomes, while functional supportive behaviors may influence specific management activities. Different definitions of support also lead to different intervention strategies. Perception-based models indicate that interventions targeting individuals' perceptions will facilitate disease management. In contract, functional models suggest that interventions focused on increasing specific supportive behaviors provided by families and friends should improve disease management (e.g., treatment reminders, scheduled treatment times).

Social Support and Disease Management. Although the mechanisms through which social support influences disease management have not been adequately described, there is evidence from adults with chronic medical conditions that social support from family and friends, regardless of how it is defined, improves adherence and health outcomes (DiMatteo, 2004b; Koenig et al., 2008; Lett et al., 2007). Far less, however, is known about social support in pediatric populations (Decker, 2007). Research in a number of pediatric conditions has examined the influence of family and peer processes (e.g., parent-child conflict, peer-group identification) on health outcomes (De Civita & Dobkin, 2004; Fiese & Everhart, 2006; White, Miller, Smith, & McMahon, 2009; Wysocki et al., 2008), and examined how support provided to caregivers relates to caregivers' adjustment (Lewandowski & Drotar, 2007; Quittner, Glueckauf, & Jackson, 1990), but only a few studies have explicitly examined the links among supportive behaviors from family and friends and disease management.

Two of these studies exemplify the utility of focusing on disease-specific support in the context of pediatric type-1 diabetes (Bearman & La Greca, 2002; La Greca & Bearman, 2002). These studies reported the validation of two measures, the Diabetes Social Support Questionnaire-Family (DSSQ-Family) and the DSSQ-Friends. Generally, the investigators found that these disease-specific measures contributed to the prediction of self-reported treatment adherence, beyond that provided by a measure of general support. There were, however, important differences in these predictive relationships depending on how the questionnaires were scored.

These studies compared two commonly used strategies for scoring measures of support. The questionnaires asked participants to rate both the frequency of specific, illness-related behaviors and how supportive they perceived these behaviors to be. The authors then compared two scoring systems: the *normative* system which used the frequency of supportive behaviors and the *individualized* system which multiplied the frequency by supportiveness ratings (La Greca & Bearman, 2002).

Although not explicitly stated by the authors, the *normative* scoring system emphasized a functional model of support, while the *individualized* system was a mixture of a functional and perceptual models. Moreover, both studies evaluated composite scores for these two scoring systems—a strategy consistent with a perceived support model (Figure 1.1). It is not surprising, therefore that the composite of individualized scores, which included teens' perceptions of support, produced higher estimates of internal consistency and was more predictive of self-reported adherence than the composite of the frequency ratings. The frequency ratings, tied to a functional support model, were inconsistent with forming a composite score (Figure 1.2), likely contributing to the lower observed reliability, and by extension, lower predictive utility of the composites. Although the combination of frequency and teens' perceptions in the individualized scores complicated identification of the mechanisms of support, the higher predictive power of this composite relative to the frequency composite suggested that teens' perceptions of support play a role in their disease management.

There was also evidence from the DSSQ-Friends that the functional roles of specific behaviors also influenced disease management. Bearman and La Greca (2002) reported that the global composite scores from the DSSQ-Friends were not predictive of a global rating of adherence. However, a significant relationship was found when specific behaviors addressing glucose monitoring were matched to questions that directly assessed adherence to monitoring. This finding supports the functional definition of support and highlights the importance of matching specific behaviors to desired outcomes. In sum, La Greca and Bearman's work suggests that *both* perceptual and functional mechanisms play a role in influencing teens' disease management.

Social Support in the Context of CF. As mentioned previously, successful management of CF requires completing multiple daily treatments and making substantial dietary modifications. Managing CF also influences how much time youth can spend with healthy peers and completely restricts access to others with CF. An increased need for support from others, coupled with reduced access to that support, suggests that facilitating support from family and friends is a promising pathway to improve disease management. Surprisingly, there has only been one study that focused on support provided by family and friends of adolescents with CF. In this study, Graetz and colleagues (2000) used structured interviews to identify supportive and non-supportive behaviors provided by family and friends, and related these behaviors to emotional and social functioning. They found that families provided considerably more support related to CF management (e.g., reminders about treatment, helping with treatment related tasks) than friends, while friends provided more companionship (e.g., acceptance of the teen, praise and encouragement). Moreover, compared to younger teens, older teens felt they received less support from family members. In contrast, there were no age differences in support from friends. There were also no gender differences in the support provided by family or friends.

The work of Graetz and colleagues is unique not only because it is currently the only study of social support in CF, but because it inquired about non-supportive behaviors such as, nagging, teasing, and talking about the illness in public. The inclusion of non-supportive behaviors addressed recent criticisms of the broader literature on social support for its exclusive focus on the *benefits* of supportive behaviors, while ignoring the potentially negative effects of non-supportive behaviors (Gallant, 2003; Helgeson, Lopez, & Kamarck, 2009; Kyngäs, Hentinen, & Barlow, 1998; Lawhon, Humfleet, Hall, Reus, & Muñoz, 2009). These non-supportive behaviors were found to be uniquely predictive of symptoms of depression, anxiety and oppositional behavior. Unfortunately, this study did not include measures of treatment adherence and was, therefore, unable to identify which supportive or non-supportive behaviors best facilitated disease management.

Currently, there is no disease-specific measure of social support for patients with CF. Because of the significant decline in treatment adherence during adolescence, there is a critical need to better understand how families and friends support teens' management of their illness. A disease-specific measure of support would facilitate social support research in CF. The Perceived Adolescent Social Support-CF (PASS-CF) was designed to capture the most common supportive and non-supportive behaviors identified by teens and to include ratings of both the frequency and perceived helpfulness of these support behaviors. The remainder of this document details the development and initial validation of the measure and is organized to present results on the three studies that elucidated this process. The first study involved the formative research that facilitated the creation of the measure, including collection and analysis of semi-structured interviews with adolescents with CF. The second study included the process of item generation and efforts to pilot the measure using cognitive interviews that accessed how teens understood and interpreted the items in the measure. Finally, the third study examined the properties of individual items, and the initial validation of the PASS-CF. Throughout the initial validation, there was an emphasis on comparing the perceived support and functional support models.

Chapter 2

Formative Research

Before constructing an illness-specific inventory of social support, it was important to understand how support was experienced by adolescents with CF. Therefore, adolescents were interviewed individually about what their family and friends have done to facilitate or impede their management of CF. This section details the process of collecting and analyzing this important qualitative information.

Methods Used to Collect and Code Adolescent Interviews

Twenty-four adolescents with CF were recruited from two specialty care clinics in South Florida and Cincinnati. Participants were individually interviewed about the supportive and non-supportive behaviors provided by family and friends. Parent consent and adolescent assent were obtained prior to participation in the study. The interviews used a semi-structured protocol that was divided into four sections addressing both supportive and non-supportive behaviors. Each section began with a general question "*What do your [family/friends] do to help you with your CF?*" or "*What do your [family/friends] do that gets in the way of your treatments?*" These questions were followed by more specific queries about different components of disease management (oral medications, nebulized medications, airway clearance, pancreatic enzyme supplements, diet, exercise, illnesses, clinic visits, hospitalizations), as well as behaviors related to emotional and social well-being.

Following each supportive or non-supportive behavior identified by participants, they were asked how often the behavior occurred and rated the behavior on a 7-point scale from "*very unsupportive*" to "*very supportive*." Participants were also asked to list the first name and relationship of the individuals in their family and friendship groups, indicate how close they felt to that person, and specify whether that person was aware of their illness. Interviews were audio recorded and then transcribed in preparation for coding.

The transcripts were coded in two phases. The first phase identified each supportive or unsupportive behavior in the transcripts, and the second phase employed an iterative process to generate a set of categories which described these behaviors. Once behaviors were identified, coders recorded the location in the transcript, a basic description of the behavior, the source of the behaviors (e.g., parent, sibling, friend), the reported frequency of the behavior, and participants' ratings of supportiveness.

The second phase used an iterative process to classify supportive behaviors. This process began with a set of theoretical categories that were derived by examining previous literature and consulting with experts in the field of CF. Behaviors were then assigned to the most appropriate category and each category was qualitatively evaluated in terms of how well the behaviors matched others in the category. This information informed modifications to the original categories and the creation of sub-categories to provide more complete descriptions of the behaviors. This process was repeated until each category contained a homogeneous set of supportive or non-supportive behaviors. During each iteration, assignments of behaviors to categories were independently conducted by two coders. The reformulations of categories were performed collaboratively with a group of researchers familiar with CF. Importantly, this group included individuals who were not involved in other aspects of the coding which provided an outside perspective to avoid decisions that were idiosyncratic to the original coders.

Results of Formative Research

Demographics. Because the aim of the instrument development phase was to capture the breadth of potential supportive and non-supportive behaviors, it was important that participants differed in their basic demographic information and disease severity. The average age of participants was 15.73 (Standard Deviation [SD] = 1.83), 50% were female, and 17% were Hispanic. Participants' lung function, measured using the "gold standard" spirometry measure for CF, percent of patients' predicted forced expiratory volume in 1 second (FEV_1 % Predicted), was 73.66% (SD = 24.10), which was similar to the national average for CF patients (76.1%; Foundation, 2009).

The size of family networks ranged from 4 to 10 (average = 6.71), with adolescents describing the relationships with about half (52%) of these individuals as "very close." The size of friendship networks ranged from 1 to 13 (average = 7.75), with 78% of those individuals having some knowledge of the adolescent's diagnosis. Importantly, it appeared that adolescents differed in how much they disclosed to their friends. The majority of adolescents (58%; *n* = 14) told everyone they listed in their network about their diagnosis, while a smaller number reported that only a few of their friends (17%; *n* = 4) knew about the diagnosis. Qualitatively, these participants noted that many of their friends were told about their diagnosis by a parent or family member without their permission. The remaining participants shared their diagnosis with some, but not all, of their friends (25%; *n* = 6). These findings suggested that disclosing their diagnosis to their friends was challenging for many teens. Further, decisions about disclosure may influence how much treatment-related support they received from their friends.

Categorizing Supportive and Non-supportive Behaviors. The interviews generated 489 behaviors from family members and 413 behaviors from friends. To test how well coders were able to identify specific behaviors in the transcripts, 20% of the transcripts were re-coded by independent research assistants. Comparisons of the two sets of codes indicated that raters were able to identify a similar number of behaviors from the transcripts (intraclass correlation coefficients across the six transcripts = .95). The behaviors were then assigned to the first set of theoretically-derived categories, which included *informational, emotional, tangible,* and *relational* (i.e. providing a sense of belonging to a social group) support. During this iterative process, additional categories were added and subcategories were formed, resulting in a hierarchical classification depicted in Tables 2.1 and 2.2.

To better understand the similarities and differences in the behaviors displayed by family members and friends, the most frequently endorsed behaviors were compared in terms of how many youth endorsed the behavior, their ratings of supportiveness, and how frequently the behavior occurred (Tables 2.3 and 2.4). There were 19 family and 18 friend behaviors endorsed by at least five teens. Of the 28 unique behaviors, 9 were provided both by family members and friends, indicated by darkened bullets in tables 2.3 and 2.4.

Although the limited number of adolescents who endorsed any given category precluded statistical comparisons between behaviors from families and friends, it appeared that family behaviors were viewed as more supportive and more frequent than friend behaviors. It is also important to note that participants were reluctant to rate either family members or friends as being unsupportive for behaviors related to doing treatments, such as nagging. Qualitatively, they often mentioned that the person “cared for them” and wanted them to do their treatments, but the constant reminding was annoying. For example, when discussing parental reminders to do treatments, one adolescent stated, “*Their intentions are good but the way they pursue it isn’t that wonderful. I’d rather them tell me to do it instead of them yelling at me to do it. I mean I’m a person, too, I forget things.*”

These findings extend work completed by La Greca and colleges (1995) who studied adolescents with Type-1 diabetes. They also used structured interviews to address support from family and friends. The findings suggested that some youth rated non-supportive behaviors, such as nagging, as supportive. The authors emphasized the individual differences among adolescents’ perspectives when determining which behaviors were supportive or non-supportive, but did not articulate why some youth may rate nagging or other non-supportive behaviors as supportive.

This formative research provided a map of the supportive and non-supportive behaviors provided by family and friends of teens with CF. It also identified some of the challenges of measuring support in this population, which included a reluctance to rate behaviors as unsupportive. Findings from this qualitative study were used to generate standard items for the PASS-CF, enabling a more systematic examination of support received by adolescents with CF.

Chapter 3

Item Generation and Cognitive Interviews

Generating Items for the PASS-CF

Interview transcripts were used to generate items for each category using the adolescents' own language. Items were then reduced by focusing on the most frequently identified behaviors. The resulting measure included 24 items for family members and 27 items for friends (Appendix A). The transcripts were also used to identify an appropriate time-frame for participant recall of these behaviors. The recall windows varied significantly depending on the context of the behavior. For example, supportive behaviors that addressed hospital stays, periods of illness, and periods of emotional distress were reported in terms of the most recent event. In contrast, behaviors that facilitated or impeded daily treatments were reported in terms of the previous couple of weeks. Item stems were generated to reflect these recall windows and items were separated on the measure to facilitate adolescents' recall of that particular context. In addition, demographic questions were included to query the timing of the most recent illness and period of emotional distress.

Pilot Testing the Instrument Using Cognitive Interviews

Once items were generated for the preliminary measure, an additional four participants were recruited for cognitive interviews, which helped clarify the wording of the items and ensure that adolescents' were interpreting the questions as intended. The first participant was a 17-year-old female of Hispanic descent who had a current FEV_1 % predicted of 91% and a body mass index percentile (BMI%) of 16 (BMI = 18.4). The second participant was a 17-year-old boy of Hispanic descent who had an FEV_1 %

predicted of 70% and a BMI% of 75 (BMI = 23.4). The third participant was a 13-year-old female with an FEV_1% predicted of 70 and a BMI% of 62 (BMI = 20). The final participant was also a 13-year-old female with an FEV_1 % predicted of 65% and a BMI% of 22 (BMI = 17.3).

Cognitive probes were designed to understand how adolescents interpreted each question. Particular emphasis was placed on their comprehension of similar terms, such as "*nagging*", "*pushing*" and "*reminding,*" to determine if teens viewed these as distinct concepts or different ways of saying the same thing. Another emphasis was to explore which recall window teens used when answering questions. Typically, each item was followed by an open-ended question designed to elicit a description of the context in which the behavior occurred and the definitions of key words. The questions addressing *nagging*, *pushing*, and *reminding,* asked teens to contrast these three terms and identify differences among them. If teens' were confused by an item, they were asked how to change the wording to clarify the meaning. The cognitive interviews revealed some important information regarding youths' responses, including, confusing wording, appropriate recall windows, and how well teens could differentiate between similar, yet distinct phrases, such as nagging, pushing and reminding.

Participants reported that several phrases were confusing; these phrases and subsequent revisions are listed in Table 3.1 along with other changes to the measure. The modifications reflected the suggestions provided by the teens. There were also differences among youth in the time periods used to recall behaviors. One youth, in particular, considered every behavior she could remember and indicated that she did not see the reference term "*in the last two weeks*" that preceded the questions. This term was underlined in the final measure. No other difficulties with the recall period were reported; in fact, questions based on discrete but distant events (i.e., hospital stays, periods of sickness) were easily identified and described by participants. Moreover, when asked which behaviors and settings they were considering when deciding how to respond to these items, teens described the most recent hospital stay or sickness. Finally, there were two negatively worded items, *Didn't have high-calorie foods at home* and *Didn't take my CF seriously*, that teens viewed as confusing; these items were dropped from the measure.

The cognitive probes also evaluated how well teens separated different terms for treatment reminders. During the structured interviews, it was apparent that adolescents referred to treatment reminders in several different ways. Some discussed them as friendly and helpful, while others described them as constant, unnecessary, coercive, and annoying. Because reminders were the most frequently mentioned behavior during the structured interviews, four family and four friend questions were written to adequately capture these different behaviors. During the cognitive interviews, each participant was asked to describe how they interpreted, *nagging* versus *pushing*, versus *reminding*. Participants described nagging as incessant or poorly timed reminders, such as providing a reminder after the teen had already completed his/her treatment. They described pushing as continuous reminders when the teen did not want to do the treatments. The findings indicated that these behaviors should be separate items in the final questionnaire.

Similar to the structured interviews, adolescents were reluctant to label annoying treatment-related behaviors from family and friends as "unsupportive." For example, when asked why she labeled "*Nagged me about my treatments*" as supportive, one teen responded, "*They do it because they care…but it's annoying.*" In essence, she was expressing two perceptions of her parents' behavior: "*they care for me*," and "*I don't like it when they constantly remind me about treatments.*" This duality highlighted some of the complexities of understanding individual perceptions about supportive and unsupportive behaviors. It appeared that these perceptions were influenced by several factors, including the timing of reminders, teens' willingness to do the treatments, and the quality of adolescents' relationship with their families and friends.

A number of strategies were discussed to address this duality, such as changing the wording of the questions and including an additional dichotomous category that allowed youth to indicate if the behavior was annoying. Ultimately, these strategies were not implemented because the wording reflected the teens' experiences and an additional category would substantially increase respondent burden. This duality embedded in some unsupportive behaviors appears to be an important contributor to teens' perceptions of disease-specific support. If these perceptions are shown to influence disease management outcomes, it will be important to investigate other factors that contribute to youths' perceptions of support.

Chapter 4

Validation of the PASS-CF

The PASS-CF was included in the initial assessment of a multi-center randomized trial addressing the effectiveness of a brief adherence intervention conducted during clinic visits. The iCARE (I change adherence and raise expectations) study, funded by the Cystic Fibrosis Foundation, Novartis Pharmaceuticals, and Genentech, Inc., also included measures of treatment adherence, HRQoL, and health outcomes. This study used a cluster, randomized controlled trial design to examine the effects of implementing this intervention on patient adherence as measured by self report and pharmacy refill histories. Twenty-two participating CF Centers were randomized to either active treatment or a control condition that provided ongoing monitoring of patients' treatment adherence through pharmacy refill data. Data for this study included 85 participants from 7 of the 22 centers who had completed the Baseline assessment as of April 22, 2010. The demographic information for these participants is listed in Table 4.1. Unfortunately, pharmacy refill data was not yet available for inclusion in this dissertation.

Measures

Treatment Adherence Questionnaire-CF (TAQ-CF). The TAQ-CF provided a measure of self-reported treatment adherence. This measure asked teens to report how often and for how long they engaged in 15 treatment-related behaviors. Frequency was rated on a six-point scale ranging from "*Not at all,"* to *"3 or more times a day"* and responses were scored in terms of number of occurrences per week. For example, *3 or more times a day"* was scored as 21, while *"3 times a week"* was scored as 3. Duration was also rated on a six-point scale, with anchors representing 5-minute intervals between

0 and 25 minutes. Responses were scored in terms of the number of minutes reportedly spent in the treatment activity (i.e., 0, 5, 10, 15, 20, 25). The measure has been shown to be reliable and valid (Ievers et al., 1999). For the purposes of this study, composite scores were created by averaging treatments with similar administration. Inhaled aerosol treatments included inhaled antibiotics and medications used to open the airways and thin mucus. Oral medications included vitamin supplements and oral antibiotics. Nutrition included meals, snacks and nutritional supplements. Adherence to enzymes, airway-clearance, and exercise were measured separately.

Cystic Fibrosis Questionnaire-Revised (CFQ-R). The CFQ-R is a well-established measure of HRQoL (Palermo et al., 2008; Quittner et al., 2005). It has separate, developmentally appropriate versions for patients ages six through adulthood. The 50 item Teen/Adult form (ages 14 and older), as well as the 35-item Child self-report version (ages 12-14), were used in this study. The Teen/Adult form contains 12 subscales and the Child form contains 8 subscales. A standardized score is calculated for each subscale (range 0-100), with higher scores representing better HRQoL. Five of these scales are included on both measures and were used in this study: Respiratory Symptoms, Physical Functioning, Treatment Burden, Emotional Functioning, and Social Functioning. Four of these five scales have been shown to have strong internal consistency, with the Treatment Burden scale demonstrating lower internal consistency and test-retest reliability (Palermo et al., 2008). Data from this study is consistent with previous reports, with adequate internal consistency (Cronbach's α = .59-.91) for all scales except Treatment Burden (Teen/Adult, α = .27; Child α = .29). Although the

Treatment Burden scale showed low internal consistency, it measured an important aspect of disease management and was retained in the analyses.

Cystic Fibrosis Foundation Registry. Participants' health information, including their pulmonary function (FEV_1 % predicted), BMI, and number of pulmonary exacerbations and/or hospitalizations, was provided by the Cystic Fibrosis Foundation Registry. The CFF Registry was started over 40 years ago to track the health outcomes of children and adults with CF. Data for each patient is regularly entered by their healthcare team into the national database after each clinical encounter (Cystic Fibrosis Foundation, 2009). Health data from the last clinic visit was extracted from the Registry data and exacerbations were counted for one year prior to enrollment in the study. Exacerbations followed a zero-inflated count distribution that suggested a majority of participants (68%) did not have an exacerbation in the last year. For computational convenience, this variable was recoded to indicate whether an adolescent had at least one exacerbation in the last year.

Analytic Approach

The PASS-CF asked about the frequency and perceived supportiveness of 24 behaviors provided by family members and 27 behaviors provided by friends. Exploratory analyses were conducted to identify problematic items, determine the factor structure of supportiveness ratings, and identify which behaviors were most predictive of adolescent health outcomes. Although the analyses were exploratory, there were specific patterns that were hypothesized a priori that were consistent with either the perceived or functional models of support.

Perceived Support Model. The perceived support definition led to a model in which individual questions were used as indicators of a single unobserved construct, defined as an individual's perceptions of the support they received from family and friends (Figure 1.1). These perceptions were hypothesized to relate most strongly with measures of emotional functioning, but also relate to disease management. This model was evaluated in two steps. First, exploratory factor analyses (EFA) were conducted to understand the latent structure of the items. These analyses used a weighted-least squares estimator that was robust to the deviations from normality often seen in ordered categorical data, similar to that observed on the PASS-CF (Muthén & Muthén, 1998). Next, scree plots were used to identify the number of salient factors by plotting the eigenvalues of the inter-item correlation matrix across the number of factors. The eigenvalues are an index of how much variability between items is accounted for by each factor. The plots help visually identify discontinuities in the pattern of eigenvalues, which suggest when to stop adding factors to the model. The second step used the composite scores derived from the EFAs (i.e., average of items with factor loadings ≥ .60) to correlate with the outcome measures. Although it is difficult to directly compare the models derived from both types of responses, it was expected that the supportiveness ratings would be well-represented by a single latent variable, whereas the frequency ratings were not expected to produce a coherent factor structure.

Functional Support Model. The second model, which focused on specific support behaviors, indicated that each behavior listed on the PASS-CF would contribute uniquely to the prediction of the outcome variables (Figure 1.2). This model was not expected to yield general latent constructs; consequently, the relationship between each behavior and outcomes were explored using bivariate correlations. Spearman's rho was used because the item-level data were categorical. Items were expected to be more strongly correlated with outcomes that were directly related to the behavior assessed by the item. For example, reminders about exercise were expected to increase the amount of exercise and general reminders about treatments were expected to increase treatment adherence. The EFAs were run using Mplus 5.21 (Muthén & Muthén, 1998); all other analyses were run using SAS 9.2 (SAS Institute Inc., 2009).

Results of the Validation

Missing Data. Rates of missing data in this study were generally low. The missing data for each measure were as follows: CFQ-R = 4%; TAQ between 0% and 8%, when counting responses of "Not Applicable" as missing; Health outcomes = 14%; PASS-CF family frequency ratings = 0% - 4%; PASS-CF family supportiveness ratings = 5% - 7%; PASS-CF friend frequency ratings = 0% - 5%; and PASS-CF friend supportiveness ratings = 6% - 11%. Due to the moderate to low amounts of missing data, the relatively small sample size in relation to the number of parameters being estimated, and the exploratory nature of the analyses, missing data were handled using list-wise deletion.

Descriptive Statistics. In order to identify questions with limited variability, items were ranked according to how often teens endorsed the "*always*" category for behaviors of family (Figure 4.1) and friends (Figure 4.2), as well as by how often teens endorsed the "*very supportive*" category for these behaviors (Figure 4.3 and Figure 4.4). Behaviors were subdivided by the recall period associated with their item stems: "*during the last two weeks…*," "*the last time I was not feeling well…*," and "*the last time I was feeling down…*"

For families, the top five most *frequent* weekly behaviors were 1) *treated me like a normal person*, 2) *showed me they cared about my health*, 3) *helped me remember to do my treatments*, 4) *helped me with my treatments (mixing medications, cleaning nebulizer)*, and 5) *helped me remember to do my treatments.* The top five most *supportive* weekly behaviors were 1) *treated me like a normal person*, 2) *showed me they cared about my health*, 3) *helped me remember to do my treatments*, 4) *pushed me to do my treatments*, and 5) *spent time with me*. These ratings suggested that teens value their family's concern for their health, treatment reminders, and active encouragement to do their treatments.

For friends, the most *frequent* weekly behaviors were 1) *complained about my CF*, 2) *left me out of activities because of my CF*, 3) *told others about my CF without permission*, 4) *nagged me about exercising*, and 5) *distracted me from doing my treatments.* The most *supportive* weekly behaviors were 1) *accepted my CF*, 2) *showed me they care*, 3) *told others about my CF without permission*, 4) *did sports or exercised with me*, and 5) *made sure I had enough to eat*. These ratings suggested that adolescents value being accepted by their friends, but that the most frequent friend behaviors tend to be less supportive.

Similar to findings from the formative research, teens appeared to be reluctant to endorse behaviors as unsupportive, especially if they were provided by friends. Consequently, the supportiveness ratings for family resembled a 4-point instead of a 5-point scale and ratings for friends resembled a 3-point scale. Categories were collapsed accordingly for all subsequent analyses. There were also items that showed little variability in terms of their frequency ratings. Limited variability was defined as more than 80% of responses in one category. Items limited in this way included questions 1 and 8 for family behaviors, and questions 3, 7, 16, and 17 for friend behaviors. These items were dropped from all analyses of both frequency and supportiveness ratings.

Independent groups *t*-tests were used to examine item response differences between males and females and between younger (age < 14) and older (age ≥ 14) adolescents for both family and friend behaviors (Tables 4.2 & 4.3). For family behaviors, there were few differences between males and females. More differences were found between younger and older teens. Consistent with increasing independence as teens get older, they reported that family members spent less time with them during treatments, but increased how often they "pushed" them to complete treatments. Older adolescents also reported that time spent with family members during treatments was less supportive than did younger adolescents.

For friend behaviors, there were few gender or age differences. Compared to females, males reported more treatment-related help from friends and more willingness to plan activities around treatments. Conversely, compared to males, females perceived friends' inquiries about their health and having someone who listens during times of emotional difficulty as more supportive. Compared to younger teens, older teens reported fewer discussions about CF and less nagging about treatments. They also rated personal discussions about their illness as less supportive than younger teens. These findings suggest that when compared with younger teens, older teens may be receiving less treatment related support from both family and friends.

Perceived Support Model. The perceived support model suggested that teens' perceptions of support is the principal mechanism linking supportive behaviors to important outcomes. This model also suggested that perceptions of support could be represented by a simple factor structure. Because the supportiveness ratings directly assessed adolescents' perceptions of support, it was expected that these ratings would follow this model better than the frequency ratings. Support for this hypothesis was evident in the scree plots (Figures 4.5 & 4.6). For family behaviors, supportiveness ratings showed a strong first factor and a clear discontinuity between the first and second factor, while the frequency ratings did not show a strong first factor and did not have a clearly defined discontinuity, suggesting no clear factor structure for these ratings. The ratings of friend behaviors were similar, with the supportiveness ratings showing a clear two-factor solution while the frequency ratings did not show a clear factor structure. The first and second factor for friend behaviors differed in terms of the questions recall periods, with the first factor reflecting items with a two-week recall and the second factor reflecting items tied to the last period of sickness and/or feeling down. Supports during these periods of distress were among the least common of the friend behaviors (Figure 4.4), which may have contributed to a two, instead of a one-factor solution.

The factor loadings for supportiveness ratings of family and friend behaviors were generated using a weighted least squares estimator with a promax oblique rotation (Tables 4.4 & 4.5). These loadings suggested two questions were not consistent with the one-factor model for family behaviors (items 4 & 7) and two questions were not consistent with the two-factor model for friend behaviors (items 9 & 10). Thus, these four items were not included in the calculation of composite scores. Using the information from these EFA analyses, composite scores were created by averaging the items with factor loadings greater than .60. If adolescents indicated that a behavior never occurred in the last two weeks, that item was dropped from their composite. As predicted, all three composite scores showed high internal consistency (Family Factor-1, Cronbach's α = .94; Friend Factor-1, α = .95; Friend Factor-2, α = .92). They were also moderately correlated with one another (*r'*s = .50 -.67), suggesting that teens perception of support are relatively consistent across supportive behaviors from family and friends.

These three composite scores were then correlated with outcomes (Table 4.6). It was expected that these composites would be more strongly related to emotional outcomes, such as the CFQ-R Emotional Functioning scale, than to specific disease management behaviors. This hypothesis was not supported. There were no significant relationships with the CFQ-R Emotional Functioning scale, but teens' perceptions of family support were related to improved enzyme and airway clearance adherence and to longer exercise times. Additionally, their perceptions of friend support were related to the frequency of aerosol treatments, as well as more time spent doing airway clearance and aerosol treatments. The family composite and the first friend composite were also related to teens' report of Physical Functioning on the CFQ-R. None of the composites were related to other scales on the CFQ-R or to health outcomes.

In sum, the supportiveness ratings were consistent with the perceived support model in producing simple factor structure with strong internal consistency, but inconsistent in terms of the relationships among composite scores and outcomes. Neither perceived support from family nor friends was related to emotional outcomes, but there were relationships among perceived support from family and friends and measures of treatment adherence, as well as the Physical Functioning scale of the CFQ-R.

Functional Support Model. The functional support model suggested that the specific supportive or non-supportive behaviors directly influenced the outcomes. The scree plots from the EFAs confirmed the prediction that frequency ratings would not produce a simple latent structure. The relationships among individual behaviors and outcomes were explored using simple bivariate correlations (Tables 4.7-4.12).

It was expected that the correlations would be higher for behaviors that closely matched the outcome of interest. For example, reminders about treatments would be more closely related to adherence to those treatments than to emotional functioning. There was some evidence for stronger relationships among behaviors matched to outcomes, such as the relationship between treatment reminders from family and enzyme adherence (r = .35), the relationship between friends' naggings about exercise and time spent exercising (r = .43), and the relationship between friends' naggings about eating and BMI% (r = .37). However, the general pattern of relationships was one of weak or inconsistent correlations among support behaviors and outcomes.

Despite the relative sparse set of relationships, there were a few informative patterns. First, a number of support behaviors were related to better enzyme adherence, while other components of disease management appeared to be easily interrupted by family and friends. Families' reminders, encouragement, rewards for treatments, and flexibility were related to enzyme adherence (r = .22 to .35), as were friends naggings to exercise (r = .24) and encouragements to eat (r = .24). In contrast, adherence to airway clearance, aerosol medications, and oral medications were negatively related to family and friend behaviors (Tables 4.7 & 1.10).

The second pattern showed differences in how family and friends both facilitated and interfered with treatments. Family behaviors were related to better enzyme adherence, but most of the relationships with health outcomes indicated interference with disease management (Table 4.9). Friend behaviors were more consistently related to better treatment outcomes than family behaviors (Tables 4.9 & 4.12). In particular friends' acceptance the illness, encouragement, flexibility, and treatment reminders were related to fewer pulmonary exacerbations (r = -.22 to -.33) and better nutrition (r = .23 to .37), but poorer adherence to oral medications (r = -.23 to -.30). Moreover, the presence of friends during treatments appeared to interfere with treatment adherence (r = .22 to .30). This data pattern also highlighted the benefits and costs of individual behaviors. For example, families' naggings about treatment appeared to improve outcomes at the cost of emotional functioning. Nagging was related to fewer pulmonary exacerbations (r = -.29), but was also related to lower scores on the CFQ-R Emotional Functioning scale (r = -.27).

The final pattern was the consistent relationships among family support behaviors during periods of increased distress (i.e., illness or sadness) and HRQoL (r =.22 to .38), which suggested support may be more appreciated when matched with teens' current needs. In general, these patterns suggested complex relationships among supportive and non-supportive behaviors and outcomes. These complexities highlighted the importance of considering the contexts of disease management tasks and support behaviors. For example, relationships differed according to disease management tasks, whether family members or friends provided the support, and according to the needs of adolescents with CF. The implications of these patterns will be discussed in Chapter 5.

Chapter 5

Summary and Conclusions

Successfully managing cystic fibrosis (CF) requires substantial daily time and effort to complete the treatments necessary to maintain health (Modi & Quittner, 2006a). Similar to other chronic illnesses, successful management is particularly challenging during adolescence, as youth transition to adult roles and relationships with family and friends change (Zindani et al., 2006). There is evidence that changes in family relationships, such as decreased parental supervision, negatively influences treatment adherence (Modi et al., 2008), while support from friends has been shown to facilitate improved management (Shroff Pendley et al., 2002). Adolescents with CF face the additional challenge of not being able to access support from those who best understand their struggles—other youth with CF. It is surprising that given the changes in family and peer relationships and the significant decline in treatment adherence during adolescence, no study to date has examined the relationship between social support and disease management in adolescents with CF.

This project was designed to construct and validate a disease-specific measure of social support that was sensitive to the context of CF. The formative research revealed approximately 47 distinct behaviors provided by families and 44 behaviors provided by friends. These behaviors fell into five general categories: *treatment related, unhelpful behaviors, emotional support, relational support,* and *tangible support.*

Examination of the most frequently endorsed behaviors suggested both similarities and differences between the behaviors provided by families and friends. The most frequent behaviors included treatment reminders, nagging about treatments, and helping youth improve their mood when feeling down. There were also clear differences between families and friends; most notably, differences between the types and amount of tangible and relational support. Consistent with previous literature (Graetz et al., 2000; La Greca et al., 1995; Shroff Pendley et al., 2002), families appear to provide more tangible support relative to friends, while friends seem to provide more relational support than families. These findings were also consistent from the descriptive results of the initial validation of the PASS-CF, which showed differences in the types of behaviors provided by family and friends. Family provided more support during times of physical or emotional distress and friends were more engaged with exercise than family members. Interestingly, the most frequent friend behaviors were less-supportive (e.g., complained about CF, left teen out of activities because of CF, told others about teen's CF without their permission), highlighting the complexities of teen's relationships with peers and the importance of including non-supportive behaviors in the study of social support.

The formative research also revealed that adolescents were often reluctant to rate behaviors from family and friends as unsupportive. Although some behaviors, such as nagging, were clearly described as annoying or unwanted, teens also recognized and appreciated that these behaviors were well-intentioned and motivated by concerns for their health. These findings were consistent with previous work in type-1 diabetes (La Greca et al., 1995) that found that some adolescents rated unsupportive behaviors, such as nagging, as supportive. This ambivalence surrounding teens' perceptions of what is "supportive" highlighted the importance of separating perceptions from the actual provision of support and suggested that future research would benefit from better understanding both the functional role of specific support behaviors and the contributing factors to teens perceptions of those behaviors.

Information from the formative research was used to create a measure of disease-specific social support for CF. The most frequently mentioned behaviors were included in the final version of the PASS-CF, with 24 items for family and 27 items for friends. Each item asked participants to report the frequency of the behavior and rate its supportiveness. By including both frequency and perceived support, the measure facilitated examination of the different mechanisms through which support may influence disease management: through teens' perceptions or through the functional roles played by specific behaviors.

Previous research on social support has suggested there is a link with disease management, but has not been clear about which aspect of support was important for improved management, resulting in inconsistent findings (Gallant, 2003; Nausheen et al., 2009). Two measurement models appeared to underlie the definitions used in previous research, each with a distinct mechanism through which social support influences disease management. The perceived support model (Figure 1.1) suggested that supportive and non-supportive behaviors indirectly affect disease management through teens' perceptions of the support. The functional support model (Figure 1.2) suggested that these behaviors directly affect management. Findings from the validation study were consistent with the measurement component of these two models and provided partial support for relationships with treatment adherence, HRQoL, and health outcomes.

Results from the exploratory factor analyses were consistent with the measurement components of the two models of support. The perceived supportiveness model implied a single latent variable that influenced adolescents' supportiveness ratings of individual behaviors. In contrast, the functional support model suggested that each behavior was uniquely related to disease management and that these relationships could not be adequately summarized by latent factors. Consistent with these predictions, teens' supportiveness ratings were well-defined by a simple factor structure (i.e., one or two factors); in contrast, the frequency ratings did not yield a coherent latent structure. This finding supported the assertion that teens' perceptions of support were distinct from their reports of the frequency of supportive and non-supportive behaviors.

Beyond the possibility of different measurement models, the perceived support and functional support models also suggested different relationships with health outcomes. The perceived support model emphasized teens' perceptions as the link between support and outcomes. It was expected that the composites of teens' supportiveness ratings would relate broadly to improved disease management, but relate most strongly with outcomes also influenced by social-cognitive processes, such as the CFQ-R Emotional Functioning scale. In contrast, the functional support model emphasized the functional role of specific behaviors and suggested that the strength of the relationship between support behaviors and specific outcomes would be tied to the match between the behavior and outcomes. For example, treatment reminders were expected to influence enzyme adherence more strongly than reminders to exercise. Results supported some of these hypothesized relationships. The pattern of results also highlighted the complexity of the relationships among perceived support, functional support, treatment adherence, HRQoL, and health outcomes. These complex relationships point to a number of challenges and future directions in studying social support in CF.

As predicted, the supportiveness composites for family and friends were related to measures of treatment adherence. Perceived support from family was related to better enzyme adherence and more time spent exercising, while perceived support from friends was related to better adherence to enzymes, airway clearance, and aerosol medications. The composites were also related to the Physical Functioning domain of the CFQ-R. These relationships suggested that teen's perceptions were related to elements of disease management. However, these data did not directly test how perceptions influence management, which may be a promising direction for future research.

Contrary to expectations, the perceived support composites were not related to emotional functioning. These findings were contrary to previous work by Graetz and colleagues (2000), who found relationships among unsupportive behaviors and self-reported symptoms of internalizing (e.g., depression, anxiety) and externalizing (e.g., acting out, conduct problems) behaviors. The discrepancy between these results may have been due to differences in definitions of support and the measures that were used in these studies. Graetz and colleagues created separate composite scores for supportive and unsupportive behaviors by multiplying frequency and supportiveness ratings. This approach was different from the one used to test the perceived support model for the PASS-CF. Because teens' perceptions of supportive and non-supportive behaviors on the PASS-CF were not sufficiently distinct to yield a separate factor, they were included together in the composite scores of the supportiveness ratings. Moreover, the CFQ-R Emotional Functioning scale did not directly measure internalizing or externalizing behaviors, which were the primary outcomes in the Graetz study.

The partial support found for the perceived support model was consistent with previous studies that have reported inconsistent relationships between social support and disease management (Gallant, 2003; Nausheen et al., 2009). Although the models used to validate the PASS-CF helped clarify the link between definitions and measurement of social support, the continued inconsistency in findings suggest further work is needed to understand how perceptions relate to emotional, behavioral, and health outcomes.

Similar to the perceived support model, results from exploratory analyses provided mixed support for the functional support model. This model predicted stronger relationships among behaviors and outcomes that were closely related, such as friends' naggings about exercise and time spent exercising, or families' reminders about treatments and better adherence to pancreatic enzymes. There were also a number of family and friend support behaviors that were related to outcomes, providing evidence that specific behaviors provided by family and friends influence disease management. Unfortunately, the most striking pattern seen in the results was the paucity of strong relationships. This sparse pattern of relationships highlighted the challenges of understanding the role of family and friends in disease management in CF.

There were several trends in the results that suggest possible future directions. When interpreting these trends, it is important to note that because of the relatively small sample and the large number of statistical parameters that were estimated, these trends may contain a number of spurious and undetected relationships. The results were also cross-sectional and thus, could not determine the causal relationships among support behaviors and outcomes. Some of these challenges will be addressed when the iCARE study completes recruitment of nearly 700 adolescents at 22 CF centers. The study will also provide longitudinal data on disease management and health outcomes, as well as pharmacy refill data on treatment adherence. As with all exploratory studies, these findings should also be confirmed with additional studies other than the iCARE project.

The first pattern of relationships suggested differences in how specific support behaviors relate to disease management activities. The disease management activities in CF are numerous and distinct. For example, airway clearance is an uncomfortable and time consuming (i.e., 20 minutes twice a day) treatment designed to clear mucus from the lungs and prevent bacterial infections, while enzymes are oral medications taken with every meal and snack and lead to digestive symptoms when not taken. Results suggested that enzyme adherence was more amenable to change than other management tasks. In contrast, airway clearance seemed to be difficult to influence in a positive direction, but was easily interrupted by non-supportive behaviors. These finding have important clinical implications. Helping teens experience success in managing one treatment may improve self-efficacy and increase the chance of success with other treatments (Marks, Allegrante, & Lorig, 2005). An initial focus on more malleable treatment behaviors, such as enzyme adherence, may facilitate greater success for teens than focusing on more challenging behaviors, such as airway clearance.

Differences in relationships among social support and disease management illustrated the need for precisely defined links between support behaviors and management tasks. Different activities present unique barriers to treatment which may require unique types of support. Unfortunately, most questions on the PASS-CF were not precisely linked to specific treatment tasks. With few exceptions, youth were asked to report on classes of behaviors, such as treatment reminders, and not consider the context of the behaviors (e.g., reminders about airway clearance versus reminders about enzymes). The support behaviors included on the PASS-CF were more precise than previous studies of support in CF; yet, they may still have been too general to capture the relationships among management activities and specific supports. This reduced specificity may partially explain the general absence of strong relationships among support behaviors and outcomes. Given the large number of treatments in CF, achieving this level of specificity would have required scores of additional items and significantly increasing the time required to complete the measure. To overcome this challenge, future studies would benefit from focusing on one or two management activities and carefully articulating the link between specific supportive behaviors and those activities.

Precisely defining these links may also require accessing the perspectives of family and friends. By relying solely on teens' recall of supportive behaviors, the PASS-CF may not have captured the full range of behaviors provided by family and friends. There were likely supports that were provided by others but not recognized by the teens. The recognition of support by the recipient has been shown to influence functional and emotional outcomes. Research on young-adult couples has suggested that instrumental support that was provided, but not noticed by the recipient (i.e., invisible support), reduced the amount of negative affect when compared to support that was noticed (Bolger, Zuckerman, & Kessler, 2000; Shrout, Herman, & Bolger, 2006). Using daily diaries and multiple respondents, the authors found that instrumental support provided by partners was related to better management of significant life stressors, regardless of whether or not it was noticed by the recipient. However, support that was noticed resulted in more anger, more depressed mood, and more anxiety. In essence, the support being provided had a cost (increased mood disturbance) and a benefit (better management of the stressor) that was moderated by whether the recipient noticed the support. This work suggests that there may be important differences between adolescents' report of behaviors they noticed versus the actual provision of support by family and friends. Such differences have been shown to be important in predicting functional and emotional outcomes in healthy young-adult populations. It is not known if the same would be true for adolescents with CF. The use of repeated measurements from daily diaries and multiple informants (e.g., parents, friends, teens) would further clarify the relationships among actual support provision, perceived support and disease management. Daily diaries have been successfully used to better understand the family context of patients with CF. In particular, the Daily Phone Diary has been well-validated in CF (Modi & Quittner, 2006b), suggesting that this form of assessment may be useful in studying social support in CF.

A second set of future directions was informed by the pattern of relationships among outcomes and supportive and non-supportive behaviors from both family and friends. Unsupportive family behaviors were related to greater treatment burden and lower social and emotional functioning. Importantly, they were also related to a number of poor health outcomes, including worse nutritional status and lung function. Interestingly, higher frequencies of nagging by family members were associated with fewer pulmonary exacerbations, but also worse emotional functioning. These relationships were consistent with teens' reported ambivalence about parental reminders during the individual formative interviews—they recognized the importance of the reminders, but were annoyed by them. These findings were also consistent with the work of Bolger and colleagues (Bolger et al., 2000; Shrout, Herman, & Bolger, 2006) that was discussed previously. There appear to be costs and benefits to support provided by family. It is not known if the same pattern applies to friends or if the “invisible support” described by Bolger and colleagues also influences treatment behaviors.

Similar to family support, there were complexities in the relationships among support from friends, disease management and health outcomes. Friends’ engagement in the teens’ illness and reminders about treatments were positively related to health outcomes, such as nutrition and fewer pulmonary exacerbations, but the presence of friends during treatments interfered with adherence to airway clearance and aerosol medications. These findings are consistent with research in adolescent type-1 diabetes that showed a positive relationship between peer involvement and improved metabolic control (Shroff Pendley et al., 2002). The findings also support the assertion made by Shroff Pendley and colleagues that friends likely both facilitate and interfere with disease management. Reminders and encouragement from friends appeared to be beneficial, but the presence of friends during treatment times may have been distracting, thus, interfering with adherence.

There were also differences between support behaviors provided by family and friends. Friends appeared to be adept at influencing teens’ nutrition, while families’ efforts in this domain were generally counterproductive. Similarly, nagging from friends appeared to be helpful and was perceived as supportive, which contrasts with the more complex relationships among nagging from family members and emotional and health outcomes that were previously discussed. These findings emphasized the importance of considering the context of support behaviors in relation to health outcomes. Similar behaviors (e.g., being present during treatments, treatment reminders, nagging) may have different outcomes depending on whether they were provided by family or friends.

The third and final pattern was the consistent relationship between emotionally supportive behaviors, especially during times of illness or sadness, and youths' report of their respiratory and physical symptoms. These results highlighted the power of support during times of distress. Family behaviors during these times were rated by teens as among the most supportive, and showed some of the strongest factor loadings in the EFA of family supportiveness ratings, representing four of the top six items in that analysis. Social support seemed to be most effective when it met the needs of the recipient, which is consistent with previous literature (Decker, 2007). The strong ratings of supportiveness during times of distress indicated that the match between supportive behaviors and needs contributed to individuals' ratings of supportiveness. The match was also consistent with teens' descriptions of the differences between reminding and nagging during the qualitative interviews. They described reminders as helpful when the reminder coincided with a forgotten treatment, but annoying when the reminder was provided when not needed because the treatment had been completed. It may be that these were the same behaviors, but were interpreted by adolescents according to their perceived needs. This is another example of the importance of separating the provision of specific behaviors from adolescents' perceptions of support. Such separation could be achieved by including multiple informants and measuring how behaviors change across time as measured by daily diaries.

In sum, the results from the initial validation of the PASS-CF provided partial support for the perceived and functional support models. They also illustrated the complexities of studying social support in a medically complicated illness, such as CF. The questionnaire represented an important step in examining disease-specific social support in CF. However, the questions may still be too broadly worded to capture the relationships among support behaviors and disease management. It will also be important to assess the perspective of family and friends in future studies to better separate the actual provision of support from what teens noticed. The role of family and friends in the management of a complex and burdensome chronic illness like CF continues to be an important, clinically relevant research question. This study replicated and built on the work of previous investigators (Bearman & La Greca, 2002; Graetz et al., 2000; La Greca & Bearman, 2002) by clarifying the link between the definition and measurement of support, and by highlighting the complexities of linking support behaviors to disease management. It also suggested several promising paths forward in the study of social support in CF. Findings from this line of research have the potential to inform clinical interventions with friends and family of youth with CF, ultimately facilitating better management of the condition.

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Figures

Figure 1.1 Perceived support model.

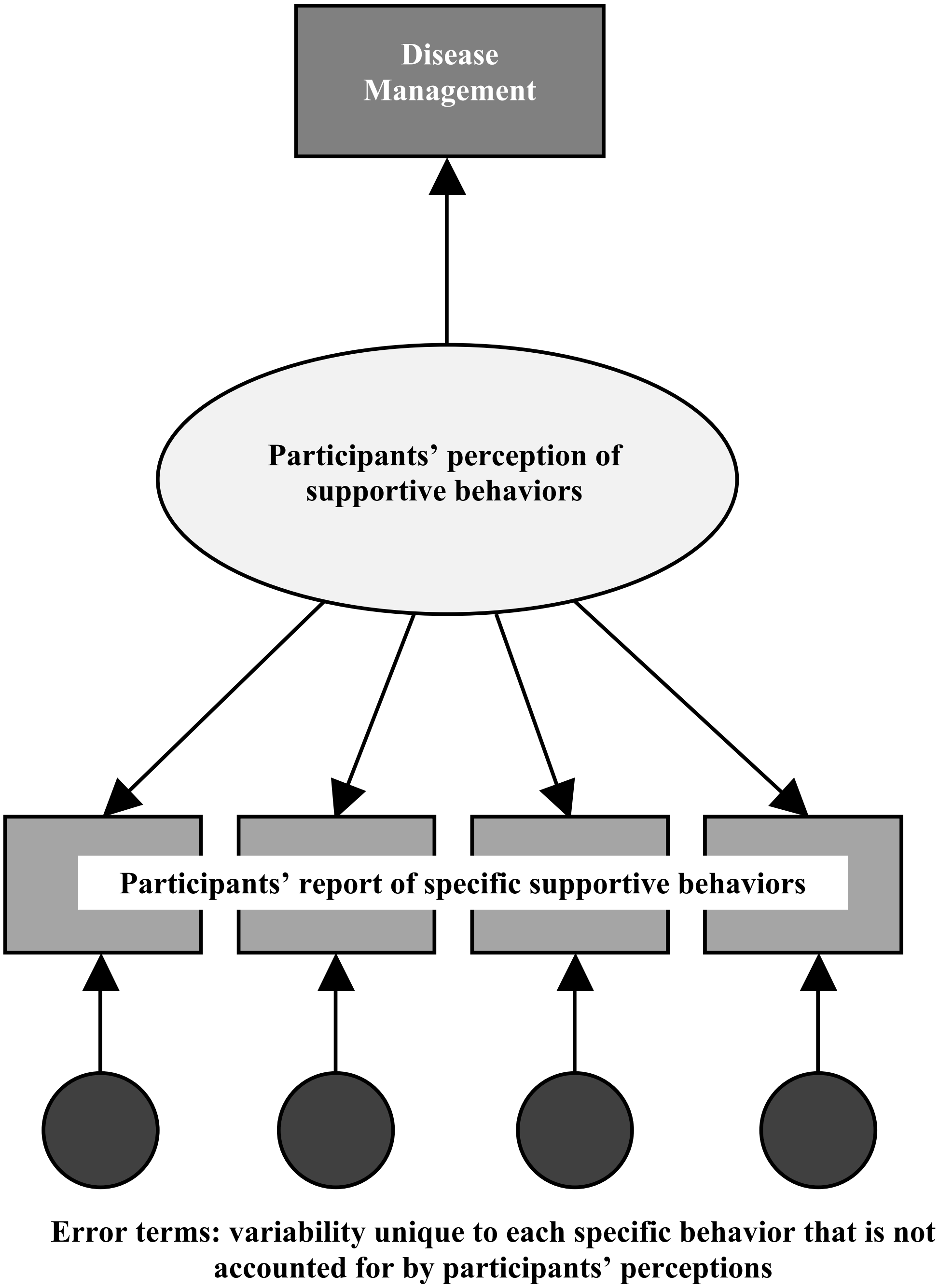


Figure 1.2 Functional support model.

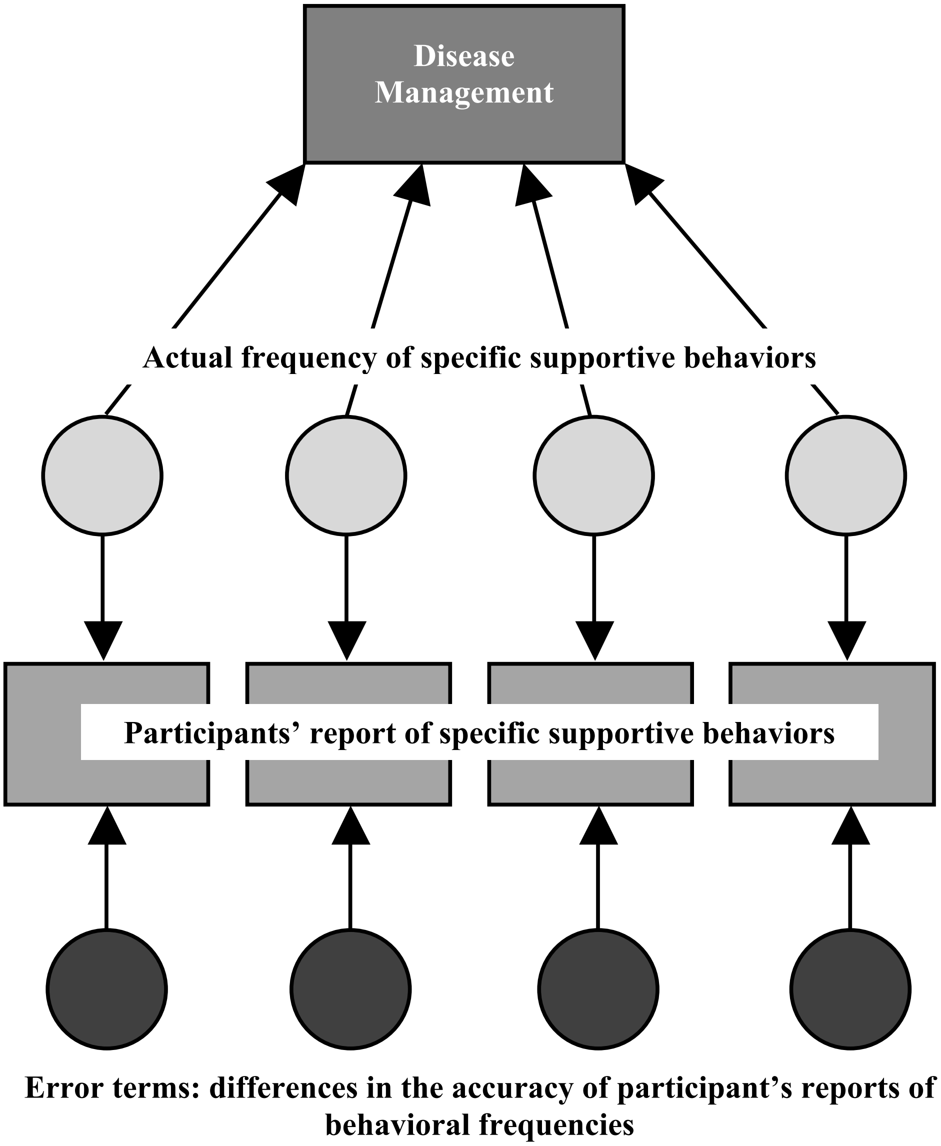


Figure 4.1 Item Characteristics of Family Frequency Ratings

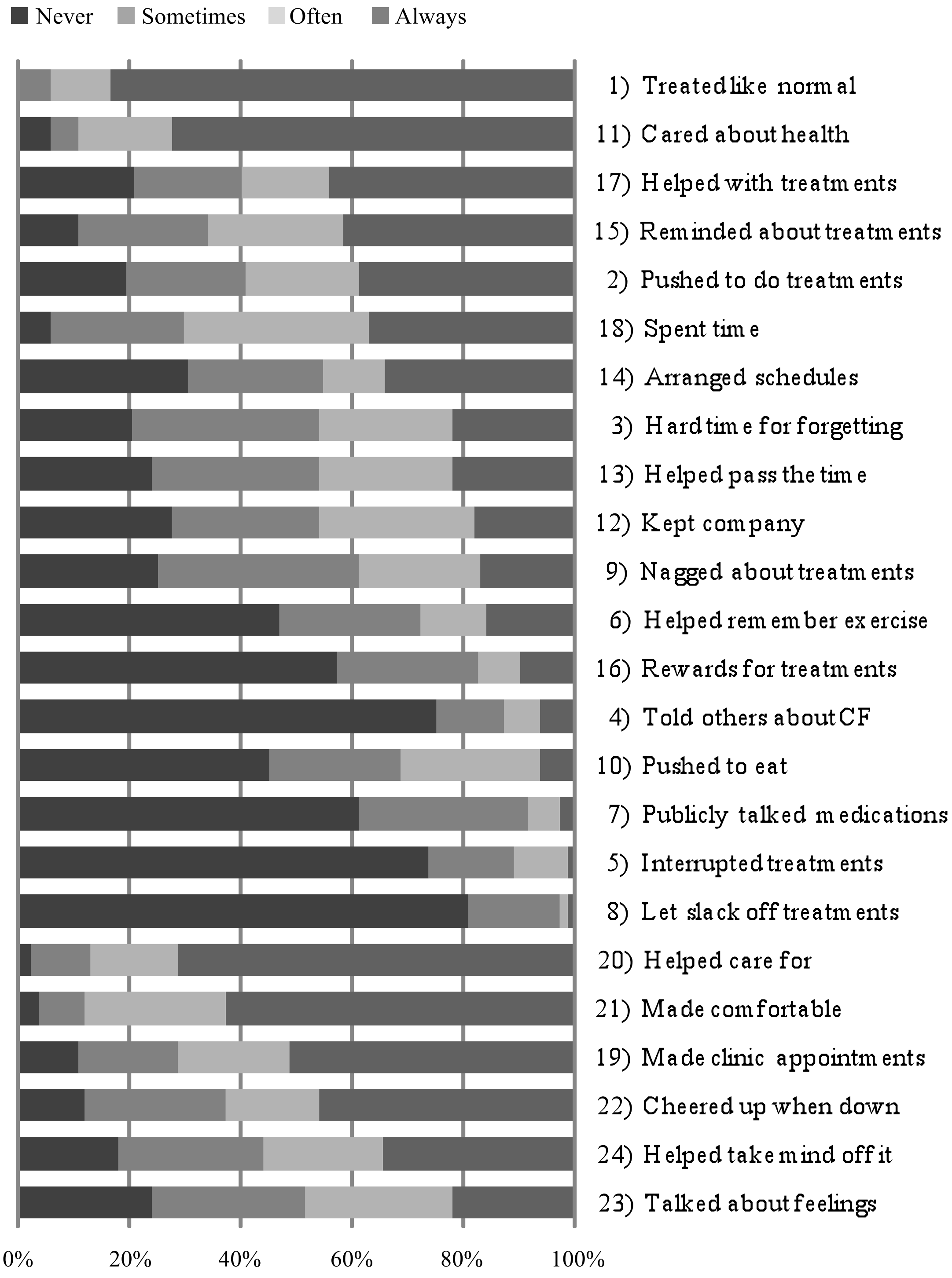


Figure 4.2 Item Characteristics of Friend Frequency Ratings

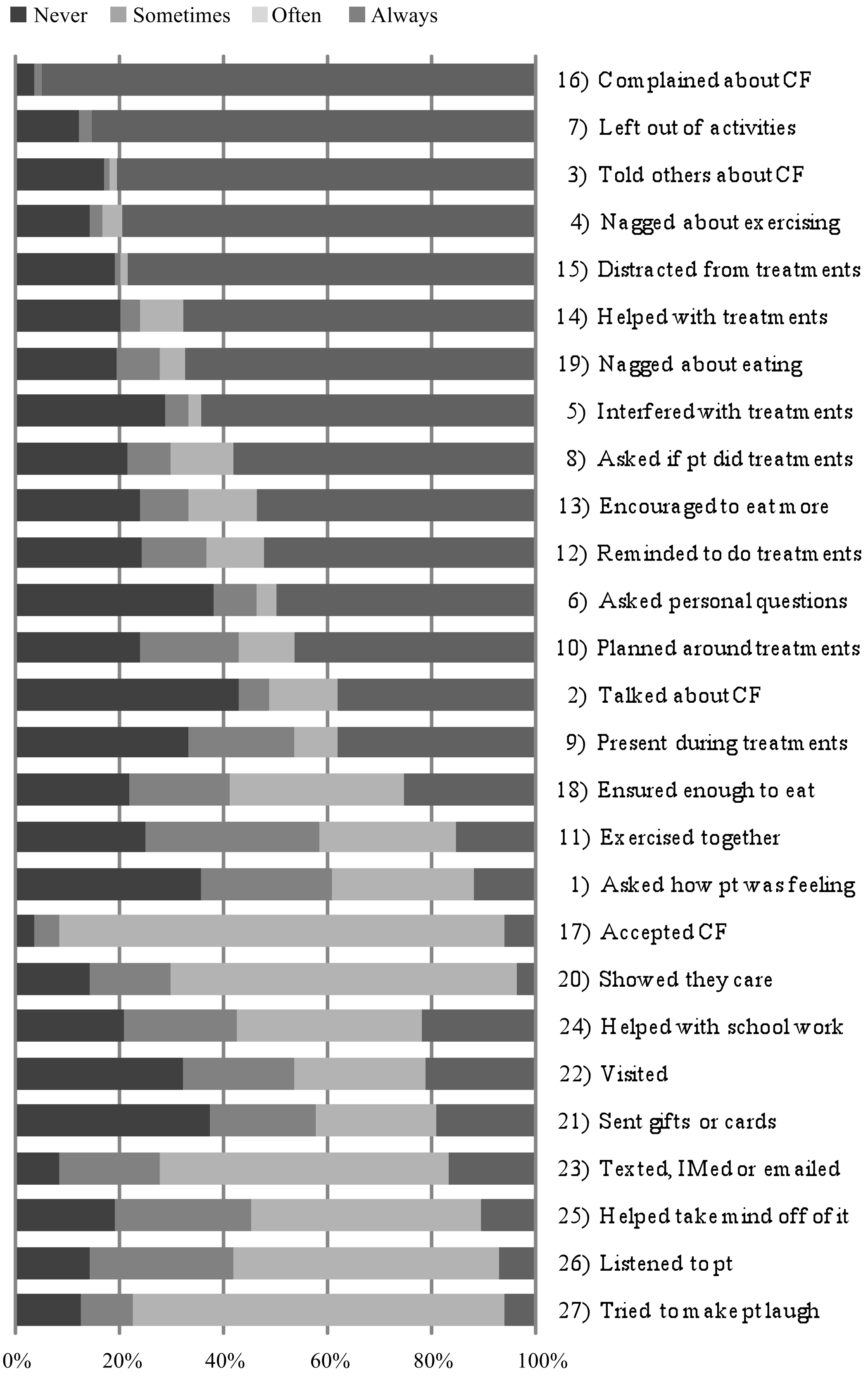


Figure 4.3 Item Characteristics of Family Supportiveness Ratings

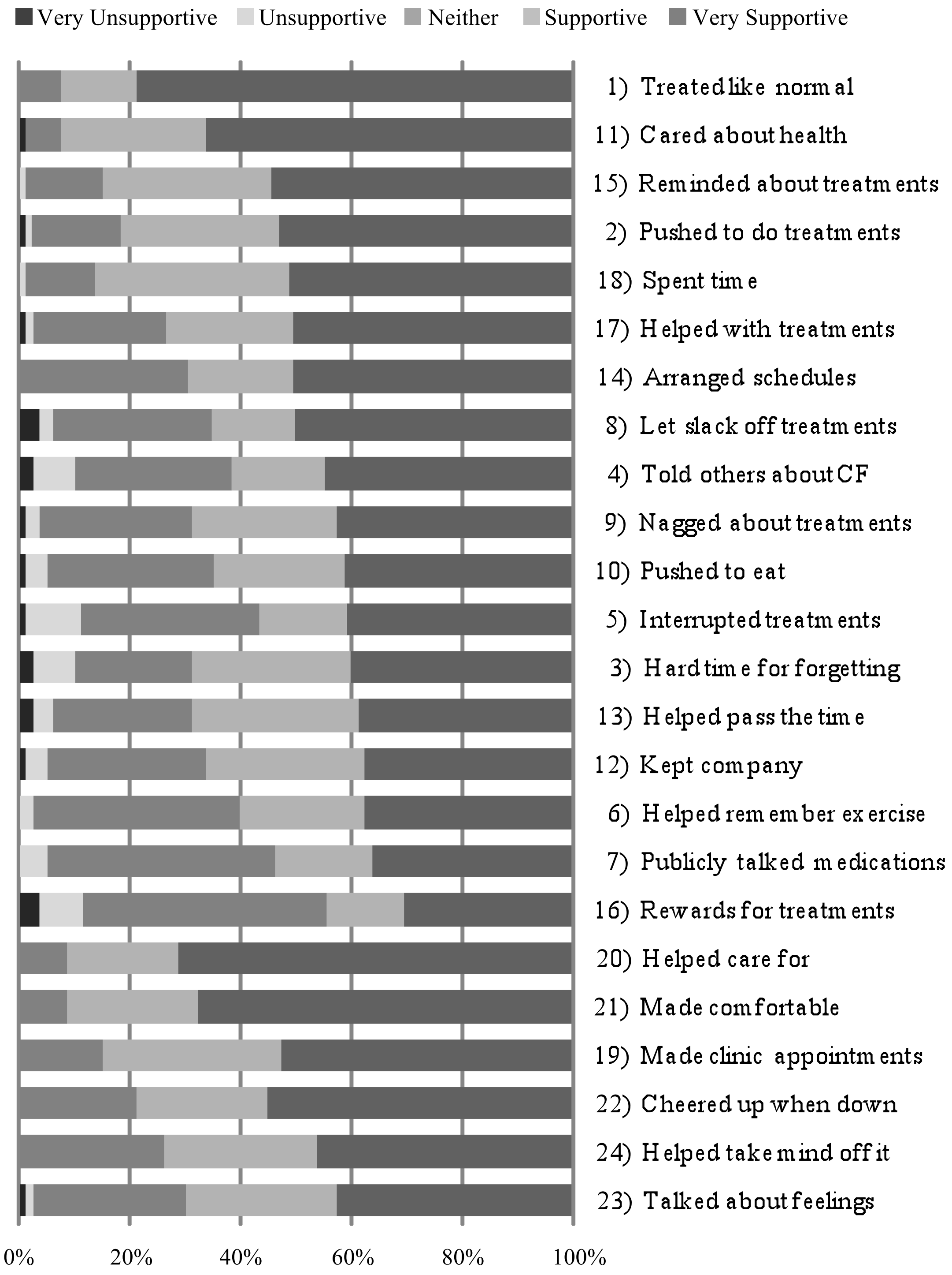


Figure 4.4 Item Characteristics of Friend Supportiveness Ratings

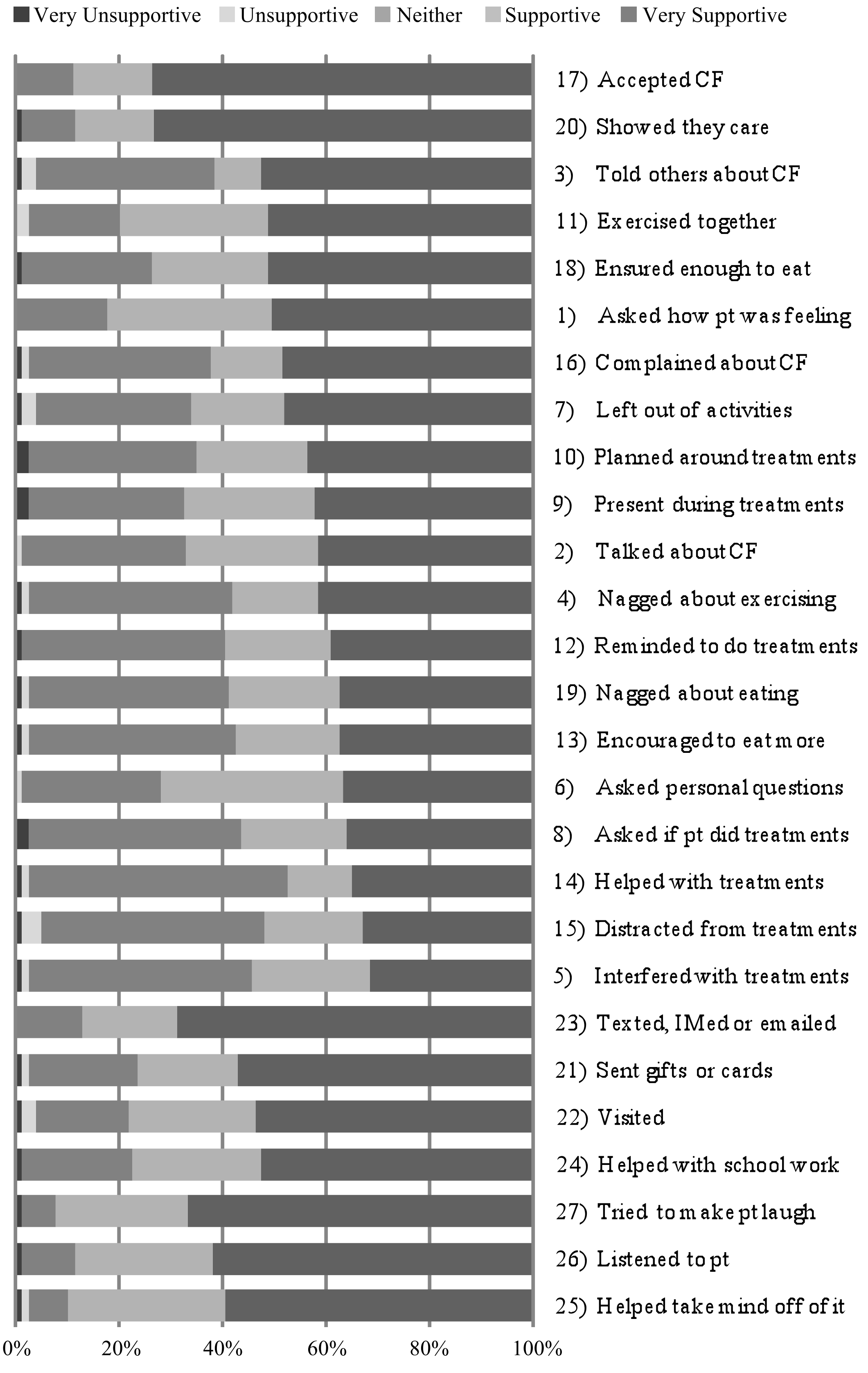


Figure 4.5 Scree Plots for Family Behaviors.

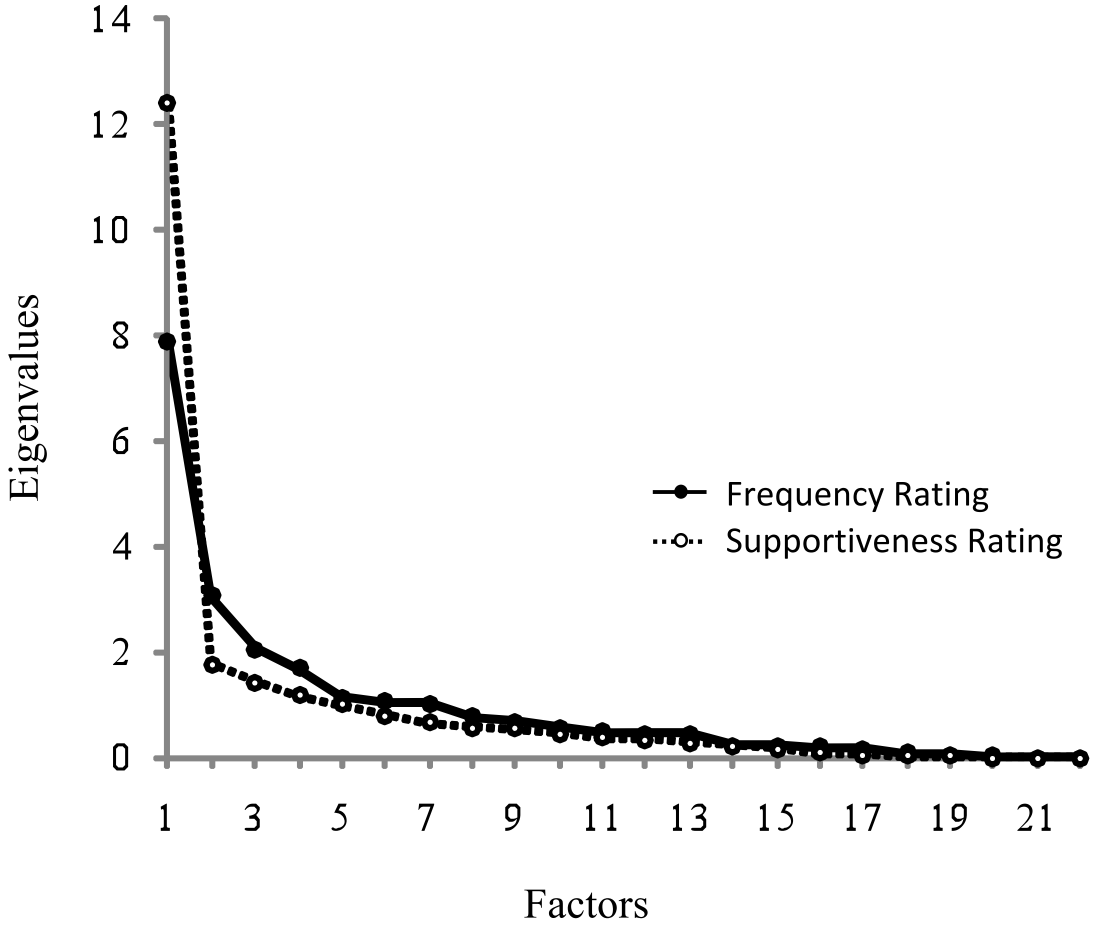


Figure 4.6 Scree Plots for Friend Behaviors.

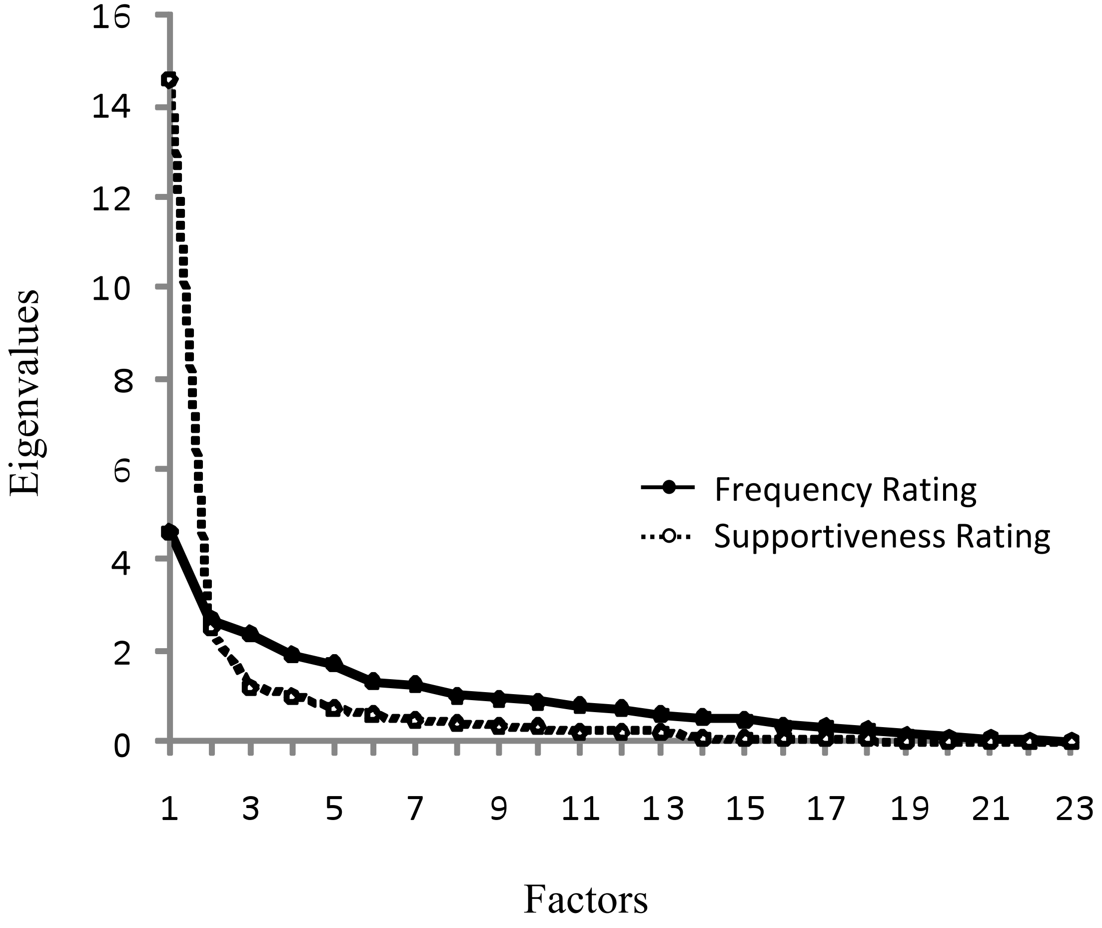


Table 2.1 Categories of Support Behaviors Provided by Family Members

	Frequency	Supportiveness
Treatment related	**144**	**2.64**
• Provided treatment reminders	78	2.63
• Prepared medications	19	2.82
• Helped do treatment	16	2.80
• Monitored treatments	12	2.20
• Provided rewards for treatment	8	2.57
• Helped problem solve	7	2.80
• Provided information	2	3.00
• Gave encouragement	2	2.00
Unhelpful behaviors	**104**	**-0.99**
• Treatment related	45	-0.47
• Nagged about treatments	15	-0.69
• Pressured teen to do treatments	12	0.75
• Asked teen to do activities which interfered with treatments	9	-1.20
• Didn't monitor	7	-1.00
• Withheld information	2	-1.00
• Relational	35	-1.09
• Talked about CF in public, or told others about the illness	10	-1.00
• Tried to control teens behavior	7	-1.29
• Were intrusive	6	-1.40
• Engaged in unwanted conversation	4	-0.75
• Didn't visit	3	0.00
• Conflict	14	-1.85
• Health related	9	-1.44
• General	5	-2.75
• Emotional	10	-1.25
• Were insensitive	8	-1.43
• Were overprotective	5	-1.40
• Withdrew from teen	2	0.00

	Frequency	Supportiveness
Tangible	**93**	**2.53**
• Provided comfort items	32	2.47
• Helped navigate medical system	18	2.40
• Provided transportation	13	2.58
• Accommodated diet	13	2.50
• Provided care when ill	9	2.86
• Facilitated physical activity	6	2.60
• Provided money	2	3.00
Emotional	**80**	**2.55**
• Positive Attention	43	2.49
• Visited or Called	28	2.54
• Showed interest	10	2.29
• Made time for teen	5	3.00
• Helped improve mood	27	2.37
• Engaged in conversation	13	2.17
• Provided laughter	4	2.75
• Encouraged activity	3	2.50
• Other	7	--
• Did activities with teen	12	2.42
• Aligned with teen	3	3.00
• Helped normalize CF	3	2.33
• Gave physical affection	2	3.00
• Showed empathy	2	2.50
Relational	**56**	**2.44**
• Allowed autonomy	12	2.55
• Showed flexibility	12	2.67
• Provided distractions from illness	11	2.10
• Helped with decision making	5	2.50
• Kept others informed	4	2.50

Frequency = Number of times a category was endorsed by teens; Ratings of supportiveness were on a 7-point scale (-3 *Very Unsupportive* to 3 *Very Supportive*)

Table 2.2 Categories of Support Behaviors Provided by Friends

	Frequency	Supportiveness
Emotional	**102**	**2.27**
• Showed concern	47	2.16
• Visited or called when ill	17	2.31
• Asked about well being	8	1.60
• Other ways of showing concern	6	2.33
• Sent gift or card when ill	6	1.83
• Helped improve mood	36	2.23
• Provided laughter	9	2.57
• Provided activity	9	1.78
Relational	**95**	**2.24**
• Accompanied during physical activity	31	2.31
• Sport	10	2.33
• Exercise	7	2.17
• Bicycling	5	2.75
• Outdoor activity	5	2.00
• Swimming	4	2.50
• Walking	3	2.00
• Sense of belonging	18	2.43
• Showed acceptance	10	2.29
• Regularly keeps in touch	8	2.57
• Managed social networks	15	1.64
• Flexible schedule	14	2.31
• Provided distraction from illness	12	2.50
• Respected privacy	2	2.50
Treatment related	**86**	**2.21**
• Provided treatment reminders	32	2.31
• Monitored health and adherence	19	2.18
• Used a directive style	15	1.58
• Told adults when teen was not feeling well	10	2.63
• Provided encouragement	6	2.60
• Helped problem solve	2	2.50
Unhelpful behaviors	**82**	**-1.30**
• Treatment Related	43	-0.84
• Scheduled activities that interfere with treatment	17	-1.38
• Nagged about treatments	13	0.33
• Impatience with treatment	5	-0.33
• Discouraged doing treatments	4	-2.25
• Engaged in risky behavior	2	-3.00
• Were critical of teen	14	-2.07
• Made discouraging comments	6	-1.83
• Fought	3	-1.67
• Teased	3	-2.33
• Gossiped about teen	2	-3.00
• Were intrusive	11	-1.70
• Asked unwanted questions	5	-1.50
• Engaged in unwanted conversations	4	-1.75
• Gave unwanted attention	2	-2.00
• Were insensitive	9	-1.63
• Were unavailable	6	-1.00
Tangible	**38**	**2.09**
• Provided food	10	1.86
• Helped with school work	9	1.88
• Helped doing treatments	8	2.63
• Shared food	5	2.33
• Went out to eat	4	1.50
• Provided transportation	1	3.00

Frequency = Number of times a category was endorsed by teens; Ratings of supportiveness were on a 7-point scale (-3 *Very Unsupportive* to 3 *Very Supportive*)

Table 2.3 Most Common Support Behaviors Provided by Family.

Supportive or Unsupportive Behaviors		Ratings of Supportiveness (7-point scales)^a	Reported Frequency (in weeks)^b
	n	Label (mean)	Label (mean)
Treatment related			
● Reminded the teen about treatments.	20	Very Supportive (2.62)	Twice a week (2.36/week)
○ Prepared medications and treatments.	11	Very Supportive (2.73)	Every other Day (3.45)
● Helped do treatments.	5	Very Supportive (2.77)	Daily (5.74)
○ Provided rewards for completing treatments.	5	Very Supportive (2.50)	Every two weeks (0.52)
● Monitored treatments and health.	5	Very Supportive (2.66)	Daily (7.00)
Unhelpful behaviors			
○ Was intrusive, controlling, or overprotective.	11	A Little Unsupportive (-1.04)	Once a month (0.31)
● Nagged about treatments.	8	Neutral (-0.46)	Twice a week (1.94)
○ Engaged in conflicts and arguments about health outcomes and treatment.	8	A little Unsupportive (-1.41)	Every two months (0.12)
● Interfered with treatment by providing competing activities.	5	A Little Unsupportive (-1.20)	Twice a week (2.00)
Tangible Support			
○ Provided comfort items when teen was ill or in the hospital.	12	Supportive (2.48)	--
○ Provided transportation to clinic.	10	Very Supportive (2.57)	--
○ Helped navigate medical system.	9	Very Supportive (2.56)	--
○ Accommodated teen's dietary needs.	9	Supportive (2.44)	Twice a week (1.89)
Emotional support			
○ Provided positive attention (visiting when ill, show interest in activities).	16	Very Supportive (2.67)	Twice a week (2.62)
● Helped to improve mood when feeling down by talking, doing activities, or telling jokes.	15	Supportive (2.47)	Every two months (0.10)
Relational support			
○ Allowed for appropriate autonomy in decision-making and activities	8	Very Supportive (2.50)	Every two months (0.19)
● Accompanied teen when doing physical activities, such as outdoor activities, sports and exercise.	8	Supportive (2.44)	Every two weeks (0.63)
● Provided distractions from illness	8	Supportive (2.13)	Every two weeks (0.56)
● Were flexible in scheduling activities to accommodate treatments.	6	Very Supportive (2.50)	Once a week (1.33)

^{a} 7-point scale: -3 *Very Unsupportive* to 3 *Very Supportive*; ^{b}Frequencies were not reported for behaviors that occurred exclusively in the context of illness, clinic visits or hospitalizations; Shaded bullets indicate behaviors provided both by family members and friends.

Table 2.4 Most Common Support Behaviors Provided by Friends.

Supportive or Unsupportive Behaviors		**Ratings of Supportiveness** (7-point scales)^a	**Reported Frequency** (in weeks)^b
	n	Label (mean)	Label (mean)
Treatment related			
● Reminded the teen about treatments.	15	Supportive (2.08)	Every two weeks (0.41/week)
○ Encouraged teen to do treatments	11	Supportive (1.98)	Every two weeks (0.49)
● Monitored treatments and health.	10	Supportive (1.95)	Every two months (0.15)
○ Inform parent or teacher when teen is ill.	7	Very Supportive (2.57)	--
● Helped with treatments.	6	Very Supportive (2.61)	Every two weeks (0.35)
Unhelpful behaviors			
● Interfered with treatment by providing competing activities.	11	Unsupportive (-1.07)	Every two weeks (0.38)
● Nagged about treatments.	9	A Little Supportive (1.02)	Weekly (0.76)
○ Engaged in conversations which the teen viewed as intrusive.	8	Unsupportive (-1.56)	Every four months (0.07)
○ Was critical of the teen (teasing, arguing, spreading rumors)	7	Unsupportive (-1.95)	Every two months (0.10)
Tangible Support			
○ Helped ensure there was ample food for the teen (went out to eat, shared food, purchased food).	11	Supportive (2.09)	Monthly (0.77)
○ Helped with school work when teen was ill or in the hospital.	7	Supportive (1.79)	--
Emotional support			
○ Showed concern (visiting or sending cards when ill, asking how the teen is feeling).	19	Supportive (2.14)	Every two weeks (0.54)
● Helped to improve mood when feeling down by talking	18	Supportive (2.28)	Every two months (0.17)
Relational support			
● Accompanied teen when doing physical activities, such as outdoor activities, sports and exercise.	20	Supportive (2.35)	Every two days (2.61)
○ Help manage the teen's social network (informing friends of condition, making introductions).	9	Supportive (1.72)	Every two months (0.13)
○ Showed they accept the teen by calling or visiting.	8	Very Supportive (2.56)	Every two weeks (0.60)
● Provided distractions from illness	8	Very Supportive (2.56)	Every two months (0.09)
● Were flexible in scheduling activities to accommodate treatments.	6	Very Supportive (2.50)	Once a week (1.33)

[a] 7-point scale: -3 *Very Unsupportive* to 3 *Very Supportive*; [b]Frequencies were not reported for behaviors that occurred exclusively in the context of illness, clinic visits or hospitalizations; Shaded bullets indicate behaviors provided both by family members and friends.

Table 3.1 Changes to the PASS-CF Recommended by Cognitive Interviews.

Issue identified through cognitive interviews	**How it was addressed in the final measure**
1) Identified behaviors that occurred outside the recall window (two weeks, last clinic visit, last hospitalization, last period of illness, last period of feeling down or depressed).	Underlined the word that defined recall window (two weeks or last)
2) Struggled to understand two negatively worded items (*Didn't have high-calorie foods at home; Didn't take my CF seriously*)	These items were dropped from the questionnaire.
3) Unsure whether *not feeling well* meant being physically ill or feeling depressed.	Changed *not feeling well* to *feeling sick*.
4) Confused by the term *feeling down*.	Changed the item stem to read…*feeling down or sad*.
5) Unsure how to respond to questions about friends with CF when they do not have friends with CF.	Added a checkbox indicating they do not have friends with CF.
6) Identified complex cognitions about frequent treatment reminders (nagging). Many adolescents viewed it as a sign that their parents cared, but found the behavior to be annoying unhelpful. They consequently, indicated that the behavior was supportive or neither, despite indicating that it was unhelpful.	An additional response option was considered that would allow the youth to indicate if they found the behavior to be annoying. However, given the length of the questionnaire, and two response options per item, this change was not included in the final measure.

Table 4.1 Demographics of iCARE Participants.

Sample size	*n* = 85
Age	14.34 (2.51)
Gender	45% female
Health Outcomes	
FEV_1 % Predicted	88.18 (22.62)
Mild ($FEV_1 > 70\%$)	79%
Moderate ($40\% < FEV_1 < 70\%$)	20%
Severe ($FEV_1 < 40\%$)	1%
BMI (percentile)	50.62 (27.34)
Percent with one or more pulmonary exacerbations in the last year	32%
Time since last clinic visit (in months)	1.61 (2.00)
Time since last illness (in days)	38.79 (81.69)
Time since last feeling down (in days)	29.02 (25.24)
Family Support Network	
Network size	5.87 (3.48)
Family composition	
Parents	40%
Siblings	29%
Grandparents	12%
Extended Family	14%
Rating of closeness	3.45 (0.59)
Friend Support Network	
Network size	7.17 (2.94)
Percent with ≥ 1 friend that knows diagnosis	91%
Percent of network that knows diagnosis	85%
Rating of closeness	3.41 (0.53)

Table 4.2 Gender Differences in Frequency and Support Ratings.

Abbreviated Item Stem	**Family** *Difference[male – female] (StdErr)*		**Friends** *Difference[male – female] (StdErr)*	
	Frequency	Supportiveness	Frequency	Supportiveness
17) Helped with treatments	-.76 (.26)	--		
4) Told others about CF	--	.48 (.25)		
6) Helped remember exercise	--	.50 (.21)		
10) Planned around treatments			.63 (.27)	--
14) Helped with treatments			.60 (.27)	--
1) Asked how pt was feeling			--	-.39 (.18)
26) Listened to pt			--	-.30 (.15)

*Only statistically significant (*p* < .05) values are listed

Table 4.3 Age Differences in Frequency and Support Ratings.

Abbreviated Item Stem	**Family** *Difference[older – younger]* *(StdErr)*		**Friends** *Difference[older - younger]* *(StdErr)*	
	Frequency	Supportiveness	Frequency	Supportiveness
2) Pushed to do treatments	.54(.25)	--		
3) Hard time for forgetting	.70 (.22)	--		
12) Kept company	-.51 (.23)	--		
14) Arranged schedules	-.70 (.27)	--		
17) Helped with treatments	-.62 (.26)	--		
20) Helped care for	-.34 (.17)	--		
12) Kept company	--	-.43 (.21)		
13) Helped pass the time	--	-.52 (.21)		
14) Arranged schedules	--	-.57 (.19)		
22) Cheered up when down	--	-.45 (.18)		
2) Talked about CF			-.58 (.30)	--
4) Nagged about exercising			-.51 (.24)	--
6) Asked personal questions			--	-.36 (.18)

*Only statistically significant (*p* < .05) values are listed

Table 4.4 Factor Loadings for Family Supportiveness Ratings.

Abbreviated Item Stem	**First Factor**	**Second Factor**
20) Helped care for	**0.91**	--
22) Cheered up when down	**0.91**	--
21) Made comfortable	**0.89**	--
12) Kept company	**0.89**	--
13) Helped pass the time	**0.88**	--
24) Helped take mind off it	**0.88**	--
11) Cared about health	**0.84**	--
15) Reminded about treatments	**0.82**	--
18) Spent time	**0.82**	--
9) Nagged about treatments	**0.80**	--
2) Pushed to do treatments	**0.80**	--
3) Hard time for forgetting	**0.79**	--
14) Arranged schedules	**0.76**	--
19) Made clinic appointments	**0.75**	--
23) Talked about feelings	**0.72**	--
10) Pushed to eat	**0.70**	--
17) Helped with treatments	**0.65**	--
6) Helped remember exercise	**0.65**	--
16) Rewards for treatments	**0.61**	--
5) Interrupted treatments	**0.61**	--
7) Publicly talked medications	0.55	--
4) Told others about CF	0.47	--

Table 4.5 Factor Loadings for Friend Supportiveness Ratings.

Abbreviated Item Stem	**First Factor**	**Second Factor**
6) Asked personal questions	**0.99**	-0.31
4) Nagged about exercising	**0.91**	-0.09
2) Talked about CF	**0.86**	0.01
15) Distracted from treatments	**0.84**	-0.06
14) Helped with treatments	**0.84**	0.15
13) Encouraged to eat more	**0.81**	0.16
8) Asked if pt did treatments	**0.81**	0.23
5) Interfered with treatments	**0.79**	-0.09
19) Nagged about eating	**0.79**	0.09
1) Asked how pt was feeling	**0.70**	0.13
12) Reminded to do treatments	**0.68**	0.29
18) Ensured enough to eat	**0.64**	0.37
11) Exercised together	**0.63**	0.23
10) Planned around treatments	0.58	0.46
9) Present during treatments	0.56	0.44
25) Helped take mind off of it	0.03	**0.97**
27) Tried to make pt laugh	0.04	**0.95**
26) Listened to pt	-0.05	**0.94**
21) Sent gifts or cards	-0.11	**0.91**
22) Visited	-0.07	**0.86**
24) Helped with school work	0.13	**0.74**
23) Texted, IMed or emailed	0.25	**0.69**
20) Showed they care	0.27	**0.61**

Table 4.6 Correlations among Supportiveness Composites and Outcomes.

	TAQ-CF					
	Airway Clearance	**Aerosol**	**Exercise**	**Nutrition**	**Oral Meds**	**Enzymes**
	Frequency (Number per week; 6-point scale: none-3x daily)					
	Mean = 9.73 (StDev = 6.10)	7.79 (3.90)	8.61 (6.78)	6.03 (3.30)	7.09 (3.97)	19.23 (5.26)
Family Factor-1	.24	--	--	--	--	.34
Friend Factor-1	--	.21	--	--	--	--
Friend Factor-2	--	.27	--	--	--	.24
	Duration (6-point scale; 0-25 minutes)					
	18.82 (6.58)	10.20 (4.79)	22.64 (4.11)			
Family Factor-1	--	--	.30			
Friend Factor-1	.29	.34	--			
Friend Factor-2	.28	.31	--			

	CFQ-R				
	Respiratory Symptoms	**Physical**	**Treatment Burden**	**Emotional**	**Social**
	76.02 (17.93)	86.26 (16.71)	63.55 (19.31)	80.70 (13.36)	76.20 (15.09)
Family Factor-1	--	.33	--	--	--
Friend Factor-1	--	.25	--	--	--
Friend Factor-2	--	.27	--	--	--

	Health Outcomes		
	Pulmonary Exacerbation	**Fev_1 % Predicted**	**BMI %ile**
	32%	88.85 (22.65)	50.12 (27.17)
Family Factor-1	--	--	--
Friend Factor-1	--	--	--
Friend Factor-2	--	--	--

Table 4.7 Correlations among Family Frequency Ratings and Adherence.

	TAQ-CF					
	Airway Clearance	**Aerosol**	**Exercise**	**Nutrition**	**Oral Meds**	**Enzymes**
	Frequency (Number per week; 6-point scale: none-3x daily)					
2) Pushed to do treatments	-.23	--	--	--	--	--
3) Hard time for forgetting	--	--	--	--	--	--
5) Interrupted treatments	--	-.25	--	--	--	--
6) Helped remember exercise	--	--	--	.26	--	.24
9) Nagged about treatments	--	--	--	--	--	--
10) Pushed to eat	--	--	--	--	--	.22
12) Kept company	--	--	--	--	--	--
13) Helped pass the time	--	--	--	--	--	--
14) Arranged schedules	--	--	--	--	--	.32
15) Reminded about treatments	--	--	--	--	--	.35
16) Rewards for treatments	.25	--	--	--	--	.23
17) Helped with treatments	--	--	--	--	--	--
	Duration (6-point scale; 0-25 minutes)					
2) Pushed to do treatments	--	--	--			
3) Hard time for forgetting	--	--	--			
5) Interrupted treatments	--	--	--			
6) Helped remember exercise	--	--	--			
9) Nagged about treatments	--	--	--			
10) Pushed to eat	--	--	--			
12) Kept company	--	--	--			
13) Helped pass the time	--	--	--			
14) Arranged schedules	--	--	.30			
15) Reminded about treatments	--	--	--			
16) Rewards for treatments	--	--	--			
17) Helped with treatments	--	--	--			

Table 4.8 Correlations among Family Frequency Ratings and HRQoL.

	CFQ-R				
	Respiratory Symptoms	**Physical**	**Treatment Burden**	**Emotional**	**Social**
2) Pushed to do treatments	--	--	-.29	--	--
3) Hard time for forgetting	--	--	--	--	--
4) Told others about CF	--	--	--	--	-.28
5) Interrupted treatments	--	--	--	--	--
6) Helped remember exercise	--	--	--	--	--
7) Publicly talked medications	--	--	--	--	--
9) Nagged about treatments	--	--	--	-.27	--
10) Pushed to eat	--	--	--	--	--
11) Cared about health	--	--	--	--	--
12) Kept company	--	--	--	--	--
13) Helped pass the time	.22	--	--	--	--
14) Arranged schedules	--	.25	--	--	--
15) Reminded about treatments	--	--	--	--	--
16) Rewards for treatments	.23	.24	--	--	--
17) Helped with treatments	--	--	--	--	--
18) Spent time	--	.34	--	.24	--
During last period of not feeling well					
19) Made clinic appointments	--	.30	--	.22	--
20) Helped care for	--	.38	--	.24	.28
21) Made comfortable	--	.23	--	--	--
During last period of feeling down or depressed					
22) Cheered up when down	.23	--	--	--	--
23) Talked about feelings	.25	.32	--	--	--
24) Helped take mind off it	.32	.27	--	--	--

Table 4.9 Correlations among Family Frequency Ratings and Health Outcomes.

	Health Outcomes		
	Pulmonary Exacerbation	**Fev_1 % Predicted**	**BMI %ile**
2) Pushed to do treatments	--	--	-.23
3) Hard time for forgetting	--	--	--
4) Told others about CF	--	--	-.27
5) Interrupted treatments	.23	-.24	--
6) Helped remember exercise	--	--	--
7) Publicly talked medications	--	--	--
9) Nagged about treatments	-.29	--	--
10) Pushed to eat	--	--	-.23
11) Cared about health	--	--	--
12) Kept company	--	--	--
13) Helped pass the time	--	--	--
14) Arranged schedules	--	--	--
15) Reminded about treatments	--	--	--
16) Rewards for treatments	--	--	--
17) Helped with treatments	--	--	--
18) Spent time	--	--	--
During last period of not feeling well			
19) Made clinic appointments	--	--	--
20) Helped care for	--	--	--
21) Made comfortable	--	--	--
During last period of feeling down or depressed			
22) Cheered up when down	--	--	--
23) Talked about feelings	--	--	--
24) Helped take mind off it	--	--	--

Table 4.10 Correlations among Friend Frequency Ratings and Adherence.

	TAQ-CF: Frequency					
	Airway Clearance	**Aerosol**	**Exercise**	**Nutrition**	**Oral Meds**	**Enzymes**
	Frequency (Number per week; 6-point scale: none-3x daily)					
4) Nagged about exercising	--	--	--	--	--	.24
5) Interfered with treatments	--	--	--	--	--	--
8) Asked if pt did treatments	--	--	--	--	-.25	--
9) Present during treatments	-.22	-.23	--	--	--	--
10) Planned around treatments	--	--	--	--	-.30	--
11) Exercised together	--	--	--	--	--	--
12) Reminded to do treatments	--	--	--	--	-.23	--
13) Encouraged to eat more	--	--	--	--	--	.24
14) Helped with treatments	-.30	--	--	--	--	--
15) Distracted from treatments	--	--	--	--	--	--
18) Ensured enough to eat	--	--	--	--	--	--
19) Nagged about eating	--	--	--	--	--	--
	Duration (6-point scale; 0-25 minutes)					
4) Nagged about exercising	--	--	.43			
5) Interfered with treatments	-.25	--	--			
8) Asked if pt did treatments	--	--	--			
9) Present during treatments	--	--	--			
10) Planned around treatments	--	--	.28			
11) Exercised together	--	--	--			
12) Reminded to do treatments	--	--	--			
13) Encouraged to eat more	--	--	--			
14) Helped with treatments	--	--	--			
15) Distracted from treatments	--	--	--			
18) Ensured enough to eat	--	--	--			
19) Nagged about eating	--	--	--			

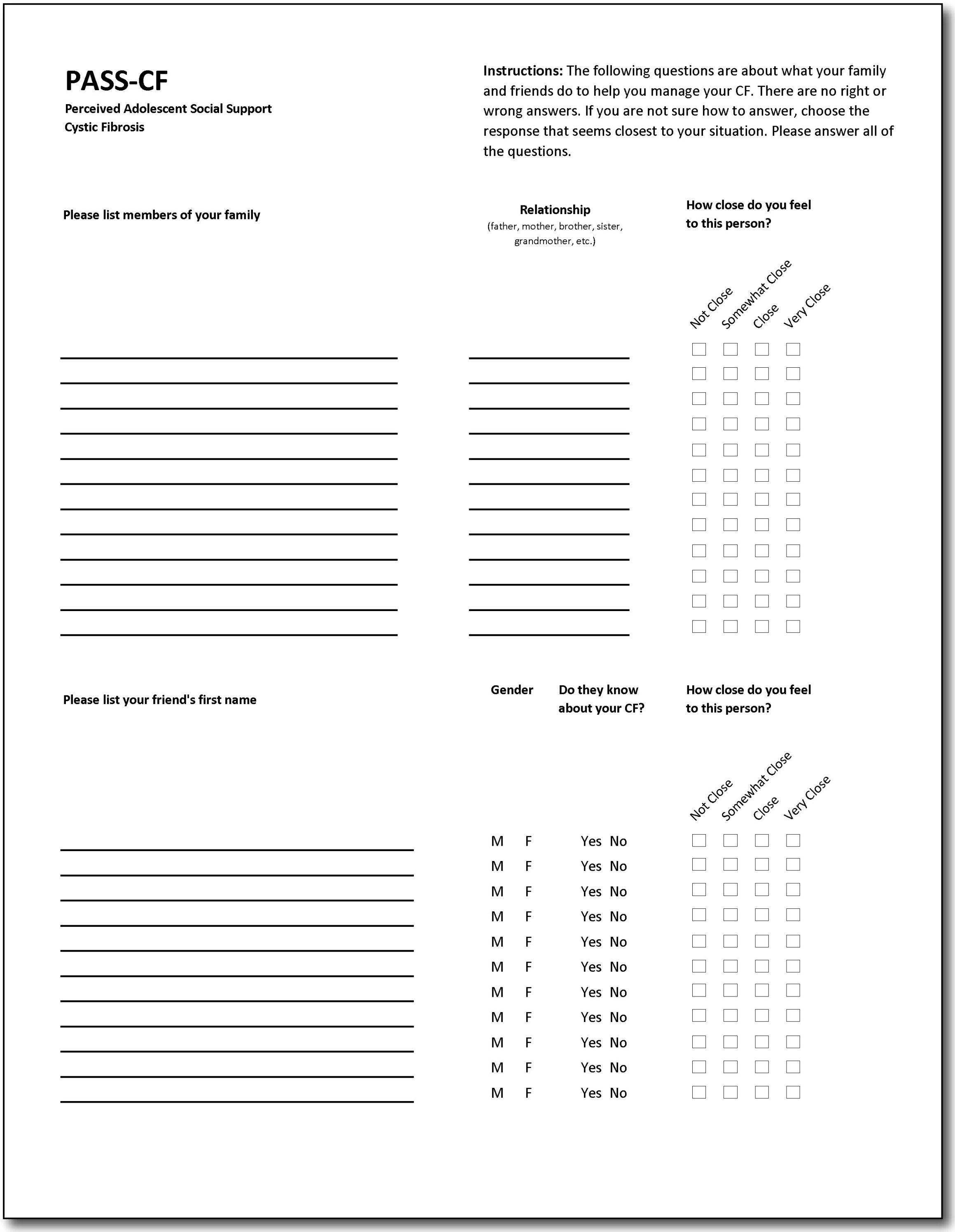
Table 4.11 Correlations among Friend Frequency Ratings and HRQoL.

	CFQ-R				
	Respiratory Symptoms	**Physical**	**Treatment Burden**	**Emotional**	**Social**
1) Asked how pt was feeling	--	--	--	--	-.22
2) Talked about CF	--	--	--	--	--
4) Nagged about exercising	--	.23	.25	.22	.25
5) Interfered with treatments	--	--	--	--	--
6) Asked personal questions	--	--	.25	--	-.25
8) Asked if pt did treatments	--	-.23	--	--	--
9) Present during treatments	--	--	--	--	--
10) Planned around treatments	--	--	--	--	--
11) Exercised together	--	--	--	--	--
12) Reminded to do treatments	--	--	--	--	--
13) Encouraged to eat more	--	--	--	--	--
14) Helped with treatments	--	--	--	--	--
15) Distracted from treatments	--	--	--	--	.27
18) Ensured enough to eat	--	--	--	--	--
19) Nagged about eating	--	--	--	--	--
20) Showed they care	--	--	--	--	.23
During last period of not feeling well or was in the hospital					
21) Sent gifts or cards	--	--	--	--	--
22) Visited	--	--	--	--	--
23) Texted, IMed or emailed	--	--	--	--	--
24) Helped with school work	--	--	--	-.25	--
During last period of feeling down or depressed					
25) Helped take mind off of it	--	--	--	--	--
26) Listened to pt	--	--	--	--	--
27) Tried to make pt laugh	--	--	--	--	--

Table 4.12 Correlations among Friend Frequency Ratings and Health Outcomes.

	Health Outcomes		
	Pulmonary Exacerbation	**Fev$_1$ % Predicted**	**BMI %ile**
1) Asked how pt was feeling	--	--	--
2) Talked about CF	-.33	--	--
4) Nagged about exercising	-.28	--	--
5) Interfered with treatments	--	--	--
6) Asked personal questions	--	--	--
8) Asked if pt did treatments	--	--	.28
9) Present during treatments	--	--	--
10) Planned around treatments	-.22	--	--
11) Exercised together	--	--	--
12) Reminded to do treatments	-28	--	.37
13) Encouraged to eat more	--	--	.24
14) Helped with treatments	--	--	.23
15) Distracted from treatments	--	--	--
18) Ensured enough to eat	--	--	--
19) Nagged about eating	--	--	.37
20) Showed they care	--	--	--
During last period of not feeling well or was in the hospital			
21) Sent gifts or cards	--	--	--
22) Visited	--	--	--
23) Texted, IMed or emailed	--	--	--
24) Helped with school work	--	--	--
During last period of feeling down or depressed			
25) Helped take mind off of it	--	--	--
26) Listened to pt	--	--	--
27) Tried to make pt laugh	--	--	--

Appendix A



PASS-CF

Perceived Adolescent Social Support
Cystic Fibrosis

Instructions: The following questions are about what your family and friends do to help you manage your CF. There are no right or wrong answers. If you are not sure how to answer, choose the response that seems closest to your situation. Please answer all of the questions.

Please list members of your family	**Relationship** (father, mother, brother, sister, grandmother, etc.)	**How close do you feel to this person?**			
		Not Close	Somewhat Close	Close	Very Close
________________	__________	☐	☐	☐	☐
________________	__________	☐	☐	☐	☐
________________	__________	☐	☐	☐	☐
________________	__________	☐	☐	☐	☐
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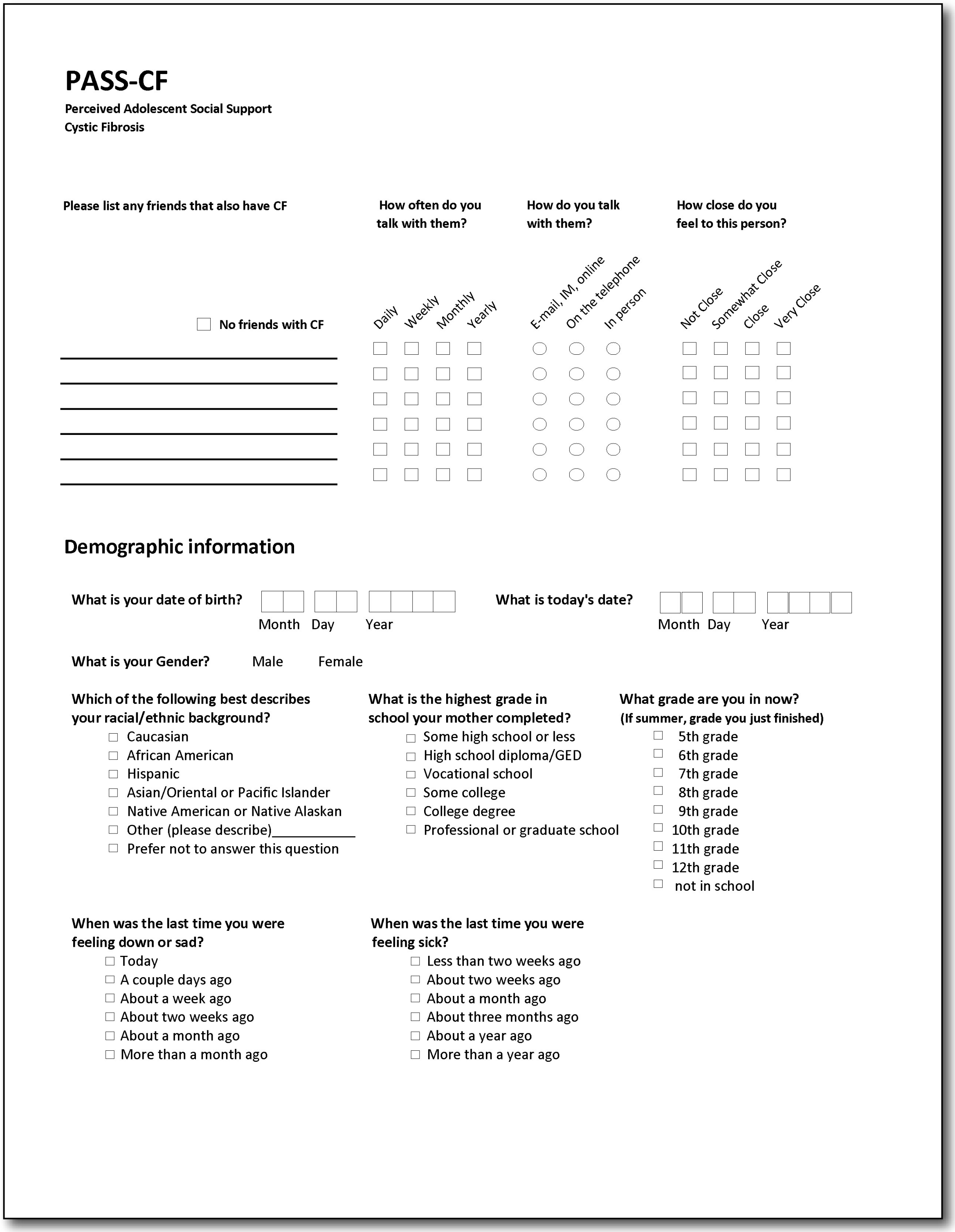
________________	__________	☐	☐	☐	☐
________________	__________	☐	☐	☐	☐

Please list your friend's first name	**Gender**	**Do they know about your CF?**	**How close do you feel to this person?**			
			Not Close	Somewhat Close	Close	Very Close
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐
________________	M F	Yes No	☐	☐	☐	☐



PASS-CF

Perceived Adolescent Social Support
Cystic Fibrosis

☐ **No friends with CF**

Please list any friends that also have CF	**How often do you talk with them?**				**How do you talk with them?**			**How close do you feel to this person?**			
	Daily	Weekly	Monthly	Yearly	E-mail, IM, online	On the telephone	In person	Not Close	Somewhat Close	Close	Very Close
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐
____________	☐	☐	☐	☐	○	○	○	☐	☐	☐	☐

Demographic information

What is your date of birth? ☐☐ Month ☐☐ Day ☐☐☐☐ Year

What is today's date? ☐☐ Month ☐☐ Day ☐☐☐☐ Year

What is your Gender? Male Female

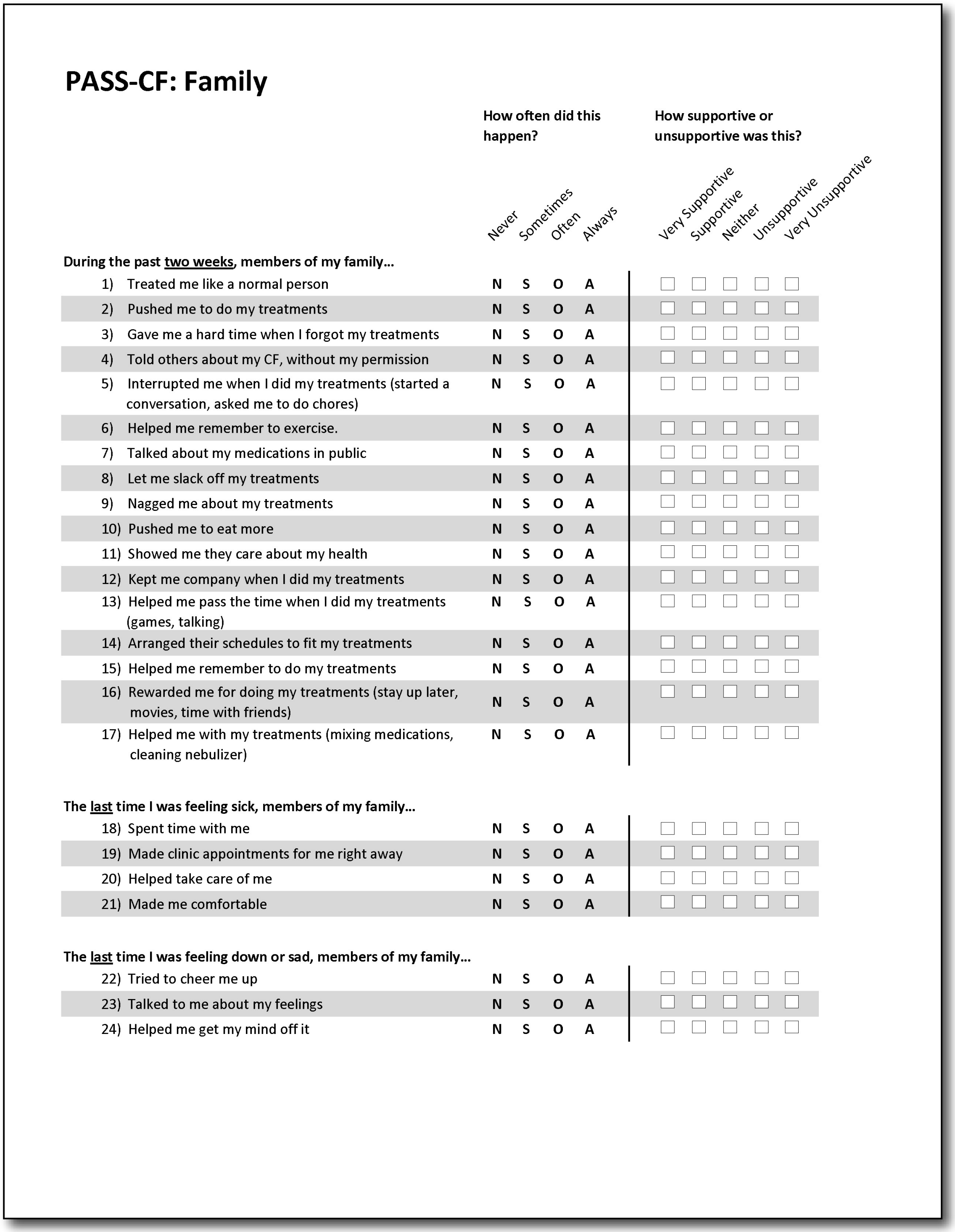
Which of the following best describes your racial/ethnic background?
- ☐ Caucasian
- ☐ African American
- ☐ Hispanic
- ☐ Asian/Oriental or Pacific Islander
- ☐ Native American or Native Alaskan
- ☐ Other (please describe)___________
- ☐ Prefer not to answer this question

What is the highest grade in school your mother completed?
- ☐ Some high school or less
- ☐ High school diploma/GED
- ☐ Vocational school
- ☐ Some college
- ☐ College degree
- ☐ Professional or graduate school

What grade are you in now?
(If summer, grade you just finished)
- ☐ 5th grade
- ☐ 6th grade
- ☐ 7th grade
- ☐ 8th grade
- ☐ 9th grade
- ☐ 10th grade
- ☐ 11th grade
- ☐ 12th grade
- ☐ not in school

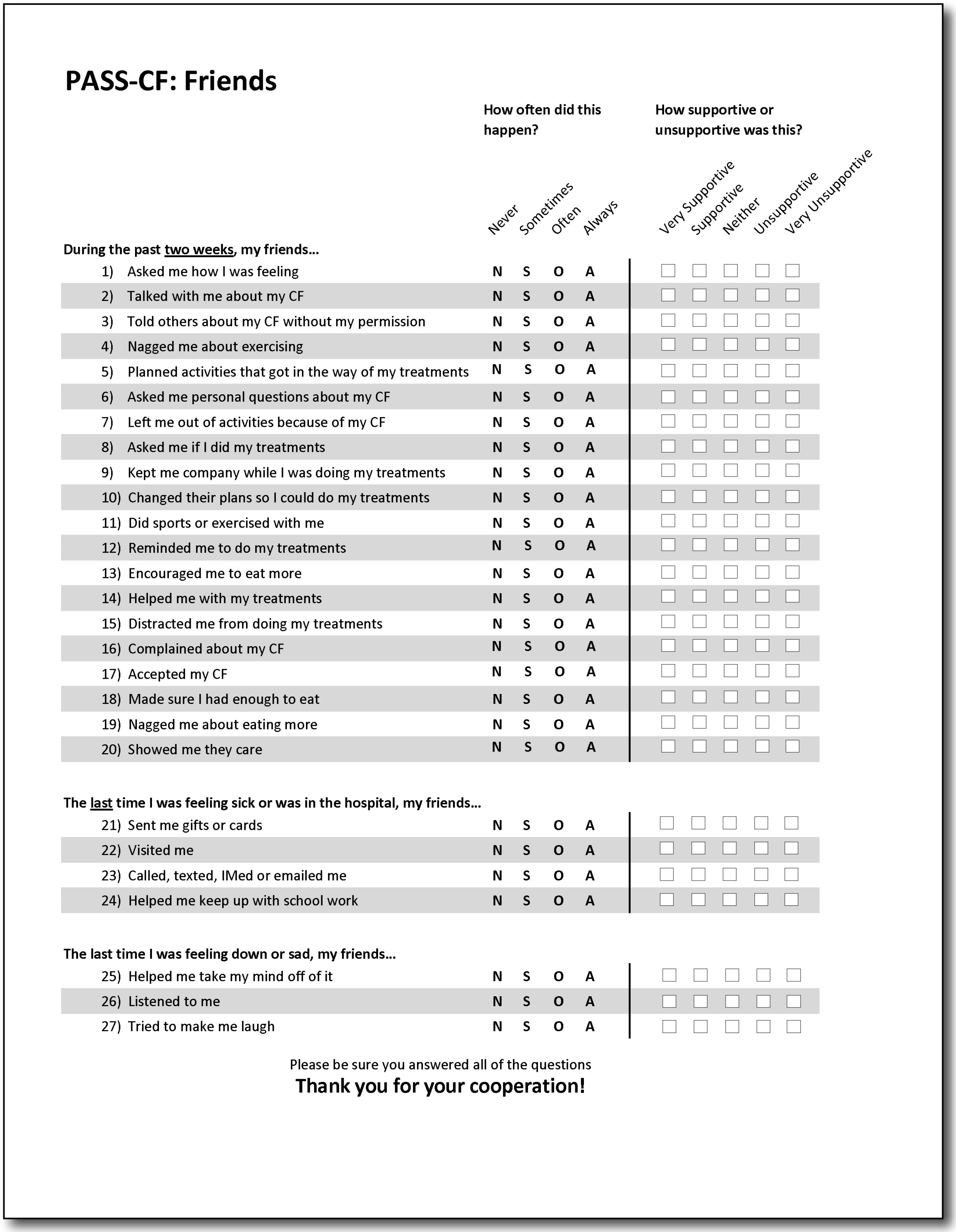
When was the last time you were feeling down or sad?
- ☐ Today
- ☐ A couple days ago
- ☐ About a week ago
- ☐ About two weeks ago
- ☐ About a month ago
- ☐ More than a month ago

When was the last time you were feeling sick?
- ☐ Less than two weeks ago
- ☐ About two weeks ago
- ☐ About a month ago
- ☐ About three months ago
- ☐ About a year ago
- ☐ More than a year ago



PASS-CF: Family

	How often did this happen?				How supportive or unsupportive was this?				
	Never	Sometimes	Often	Always	Very Supportive	Supportive	Neither	Unsupportive	Very Unsupportive
During the past <u>two weeks</u>, members of my family…									
1) Treated me like a normal person	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
2) Pushed me to do my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
3) Gave me a hard time when I forgot my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
4) Told others about my CF, without my permission	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
5) Interrupted me when I did my treatments (started a conversation, asked me to do chores)	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
6) Helped me remember to exercise.	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
7) Talked about my medications in public	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
8) Let me slack off my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
9) Nagged me about my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
10) Pushed me to eat more	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
11) Showed me they care about my health	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
12) Kept me company when I did my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
13) Helped me pass the time when I did my treatments (games, talking)	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
14) Arranged their schedules to fit my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
15) Helped me remember to do my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
16) Rewarded me for doing my treatments (stay up later, movies, time with friends)	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
17) Helped me with my treatments (mixing medications, cleaning nebulizer)	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
The <u>last</u> time I was feeling sick, members of my family…									
18) Spent time with me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
19) Made clinic appointments for me right away	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
20) Helped take care of me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
21) Made me comfortable	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
The <u>last</u> time I was feeling down or sad, members of my family…									
22) Tried to cheer me up	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
23) Talked to me about my feelings	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
24) Helped me get my mind off it	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐



PASS-CF: Friends

	How often did this happen?				How supportive or unsupportive was this?				
	Never	Sometimes	Often	Always	Very Supportive	Supportive	Neither	Unsupportive	Very Unsupportive
During the past __two weeks__, my friends…									
1) Asked me how I was feeling	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
2) Talked with me about my CF	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
3) Told others about my CF without my permission	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
4) Nagged me about exercising	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
5) Planned activities that got in the way of my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
6) Asked me personal questions about my CF	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
7) Left me out of activities because of my CF	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
8) Asked me if I did my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
9) Kept me company while I was doing my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
10) Changed their plans so I could do my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
11) Did sports or exercised with me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
12) Reminded me to do my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
13) Encouraged me to eat more	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
14) Helped me with my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
15) Distracted me from doing my treatments	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
16) Complained about my CF	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
17) Accepted my CF	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
18) Made sure I had enough to eat	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
19) Nagged me about eating more	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
20) Showed me they care	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
The __last__ time I was feeling sick or was in the hospital, my friends…									
21) Sent me gifts or cards	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
22) Visited me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
23) Called, texted, IMed or emailed me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
24) Helped me keep up with school work	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
The last time I was feeling down or sad, my friends…									
25) Helped me take my mind off of it	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
26) Listened to me	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐
27) Tried to make me laugh	**N**	**S**	**O**	**A**	☐	☐	☐	☐	☐

Please be sure you answered all of the questions

Thank you for your cooperation!