



Gender differences in treatment adherence among youth with cystic fibrosis: Development of a new questionnaire

Joän M. Patterson^{a,*}, Melanie Wall^b, Jerica Berge^c, Carlos Milla^d

^a Division of Epidemiology & Community Health, University of Minnesota, School of Public Health, 1300 South 2nd Street, Suite 300, Minneapolis, MN 55454, United States

^b Division of Biostatistics, University of Minnesota, School of Public Health, 420 Delaware Street SE, Minneapolis, MN 55455, United States

^c Family Medicine & Community Health, University of Minnesota, Medical School, 420 Delaware St. SE, Minneapolis, MN 55455, United States

^d Center for Excellence in Pulmonary Biology, Stanford University, 770 Welch Road; Suite 350, Palo Alto, CA 94304, United States

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Abstract

Background: Some prior studies have reported that girls with cystic fibrosis (CF) experience higher morbidity and mortality compared to boys. In this study, the authors compared boys' and girls' perceptions of disease-related strains and resources associated with living with CF, and the relationship of these factors to CF treatment feelings and behaviors.

Methods: All 10–21 year olds with CF at the Minnesota Cystic Fibrosis Center were invited by mail to complete a new self-report survey (Living with CF Questionnaire — LCFQ). Of these 177 youth, 58% (49 boys and 54 girls) returned surveys.

Results: Exploratory and confirmatory factor analyses revealed nine factors in the LCFQ. Partial support was found for hypothesized gender differences in these factors. Compared to boys, girls reported significantly more illness-related strains and worries, including emotional strains, greater treatment discouragement, lower self-esteem, and lower adherence to some aspects of the CF treatment regimen (coughing, eating high-fat foods, taking meds/pills).

Conclusions: Living with CF appears to have a greater emotional impact on adolescent girls compared to boys. These gender differences may contribute to the poorer pulmonary function observed among girls with cystic fibrosis during the adolescent years.

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1. Introduction

Cystic fibrosis (CF) is the most common inherited life-shortening, multisystemic disease among Caucasians. The majority of affected patients develop chronic lung disease characterized by airway obstruction and bronchial mucus plugging, which leads to recurrent infections and declining pulmonary function over time. Most patients also experience severe destruction of the exocrine pancreas, which produces malabsorption and malnutrition. Improvements in nutritional and pulmonary therapies and treatment at specialized treatment centers have resulted in increased life expectancy;

the predicted median age of survival in 2005 was 36.5 years in the U.S. compared to 14 years in 1969 [1,2]. However, these improved therapies require regular, lifelong adherence to a complex routine in order to be effective. A typical treatment regimen includes daily chest physiotherapy, a variety of inhaled medications, oral doses of antibiotics and pancreatic enzyme supplements, regular aerobic physical activity, as well as following a high-calorie/high-fat diet.

Most of the morbidity and mortality associated with CF is due to pulmonary complications. Persistent airway inflammation, chronic infection and inspissation of secretions leads to an irreversible and progressive obstructive process [3]. Both male and female patients develop this progressive decline in pulmonary function; however, the age of onset tends to occur at the ages of 12–14 years for girls and 19–20 years for

* Corresponding author. Tel.: +1 612 624 1394; fax: +1 612 624 0315.

E-mail address: jasu@umn.edu (J.M. Patterson).

boys [4,5]. The relative risk for survival is significantly lower for females compared to males for all ages from 2 to 20 years; and this gender gap does not narrow over time [2]. The reasons for these gender-based differences in outcomes have been perplexing issues that have yet to be fully explained. It is clear that nutritional status, airway microbiology, and pulmonary function are strong predictors of later mortality, although reports of gender differences in these factors have been inconsistent [4–6].

Behavioral explanations related to gender differences in treatment adherence, which in turn affects nutritional status and pulmonary function, and differences in desired and received support from family and friends may be factors associated with this gender gap. Based on our review of the literature and conversations with health care providers in a specialized CF treatment center, we postulate the following behaviors as possibly associated with increased morbidity among girls:

1.1. Inadequate fat and calorie intake

Studies of treatment adherence have shown that girls are less likely to adhere to a high fat, high calorie diet compared to boys [7,8]. Girls are more likely than boys to engage in dieting behaviors in response to the cultural unacceptability of female obesity [9], which may be related to the tendency of some girls with CF to embody the feminine cultural norm of thinness as an indicator of attractiveness [10]. Girls with CF are actually more likely to perceive their smaller stature as positive and have higher body satisfaction than girls without CF [9].

1.2. Suppression of coughs

Coughing aids in removing mucus, thereby being an important mechanism to facilitate the clearance of abnormal secretions. Youth with CF may be self-conscious about the impact of their coughing on others, and some may resort to suppressing it as a way to avoid negative attention. Cough suppression may be more characteristic of girls, who generally pay more attention to socially acceptable behaviors and fitting in with peers. Compared to boys, adolescent girls have greater public self-consciousness, which is largely attributable to the greater attention they pay to social relationships, including how others view them [11]. Coughing is less likely to fit girls' notions of "feminine;" whereas, boys generally are encouraged to cough and spit during intense sports participation and therefore, may not experience the same social discomfort [10].

1.3. Failure to take prescribed medications and enzymes regularly

Girls are more likely than boys to have periods of denial regarding their disease during which they discontinue important components of their treatment regimen, such as

skipping medications and enzymes [10]. Here again, because they are more aware of how others perceive them, girls may feel self-conscious about standing out as different during a developmental period when they want to fit in and be like their peers [11,12].

1.4. Reduction in physical activity

It is normative for healthy adolescent girls to decrease their levels of physical activity at puberty [13–15], and this gender difference is evident in girls with CF as well [16]. Although there has been increased athletic involvement among girls in the last two decades, the societal images of femininity still do not generally encourage physical activity or exertion for girls and may contribute to girls' decline in physical activity at puberty [10,17]. Boys with CF, on the other hand, are more likely to engage in sports and exercise as central to the cultural masculine identity and as an unconscious way to deny the presence of any limitation in their life due to CF [17]. The decline in physical activity for girls with CF may also be related to malnutrition [16]. Inadequate energy intake causes impaired muscular function, and impaired muscular function, including respiratory muscle, leads to a reduction in physical activity. Girls with CF have a poorer pattern of weight gain than males even before puberty [1].

1.5. Skipping chest physiotherapy

This has been reported as the most disliked component of treatment and associated with the poorest adherence [18]. Miller et al. [17] suggest that gender differences in treatment adherence may be related to girls' greater passivity and acceptance of life circumstances, which translates into a sense of powerlessness and lower expectations of self.

In contrast, the cultural masculine identity encourages boys to take control of their lives and to be more action-oriented.

1.6. Quality of social relationship patterns

It is also expected that parents and others may socialize and respond to boys and girls with CF differently. Given the small stature that is associated with CF, it is plausible that adults (and perhaps peers) may perceive girls with CF as weak and fragile, and not capable of vigorous physical activity. Girls may be subtly discouraged to participate, or at least not encouraged. In contrast, males with CF, even though small in stature and size relative to their peers, may strive to increase their body mass as a way to defy their illness and preserve their masculine identity [10,17]; and they may be encouraged by adults to participate in strenuous sports activities, as a way to compensate for their physical differences. Family support and functioning patterns, as well as a family's reaction to the illness, have been shown to affect CF treatment adherence [19–21]. It is possible that there are gender differences in how parents and others respond to the care and support needs of

youth with CF, and these responses are likely to affect the way youth perceive and manage their chronic disease and ultimately, their survivability.

1.7. Perceptions of strain and support

Developmental psychologists have reported that adolescent girls have greater public and private self-consciousness compared to boys [11,22]. Girls are more attuned to how others view them (public) and to their own inner feelings, albeit unbeknown to others (private). Being self-conscious about body image, diet, and treatment requirements may contribute more strain for girls and negatively affect their self-esteem [23]. Qualitative studies suggest that non-adherence increases girls' sense of guilt [10,24] and contributes to lower self-esteem and self-efficacy. Furthermore, girls with CF are reported to worry more about the future and the implications of their illness for life choices [10,17]. In contrast, boys with CF and other chronic illnesses have been reported to be less passive and more action-oriented, attempting to take charge of their illness, having a higher sense of controllability, living more in the present, and being less concerned about the future [10,17].

These seven factors: (a) inadequate fat and calorie intake, (b) suppression of coughs, (c) failure to take prescribed medications and enzymes regularly, (d) reduction in physical activity, (e) skipping chest physiotherapy, (f) quality of social relationship patterns, and (g) perceptions of strain and support warrant further investigation for their possible relationship to increased morbidity among girls with CF.

We used stress theory [25–27] to develop an integrative framework for these findings from the literature and clinical observations of youth living with CF. Stress theory purports that adaptation to stressful life events is affected by the balance between two major sets of factors.

On one side, there is the overall accumulation of demands being experienced at any point in time. These demands include stressor events and ongoing strains, including the strains of living with a chronic illness, getting along with family members and peers, particularly related to the developmental stage of youth. On the other side, there are capabilities for managing these demands. Capabilities include (a) tangible (e.g., money) and intangible (e.g. social support) resources that a person has or is able to access and (b) coping behaviors — what is actually done to manage the demands. This balance between demands and capabilities is strongly influenced by perceptions or “meanings” that are given to both the demands and to the capabilities. Through primary appraisal, stressors or strains can be exaggerated or minimized. Similarly, through secondary appraisal, some kinds of capabilities can emerge, such as believing in oneself as capable or believing that my health condition is not as bad as that of another person. One way of conceptualizing outcomes — or adaptation — using a stress framework is to focus on an individual's biologic functioning. So, in the case of CF, biologic functioning as measured by pulmonary function and/or nutritional status can

be examined in relationship to personal, family and friend sources of strain and support as well as positive and negative coping behaviors. It is expected that the interaction of strains, resources and coping will influence CF treatment feelings and behaviors, which are related to biologic health outcomes. Fig. 1 shows our conceptual model for this study. Although not actually assessed in this study, we recognize that there are bidirectional effects between health outcomes and treatment adherence and the other psychosocial factors in our model.

1.8. Study purpose and hypotheses

The purpose of this study was two-fold. First we examined the reliability and validity of a new set of scales derived from the Living with Cystic Fibrosis Questionnaire (LCFQ) designed to assess psychosocial factors and treatment adherence among youth with CF as described in our literature review. Second, we examined hypothesized differences between boys and girls with CF on the LCFQ scales and the relationship of these scale scores to treatment adherence. We hypothesized that compared to boys, girls would: (a) be more non-adherent to the CF treatment regimen; (b) experience higher levels of stress (due to illness-related strains and strains with parents and their peers), and greater self-care discouragement, which all would be related to treatment non-adherence; and (c) seek more support and report higher levels of family and peer support, which would be associated with better treatment adherence.

2. Methods

Following approval from the University of Minnesota Institutional Review Board, all youth between the ages of 10 and 21 years ($n=177$) who were being followed at the Minnesota Cystic Fibrosis Center were sent a self-report questionnaire in July 2002, along with a letter to their parents explaining the study, and consent and assent forms. Parents of minors and youth who were 18 years or older provided informed consent; youth younger than 18 years provided informed assent. Reminder postcards were sent if the questionnaire was not returned in four weeks.

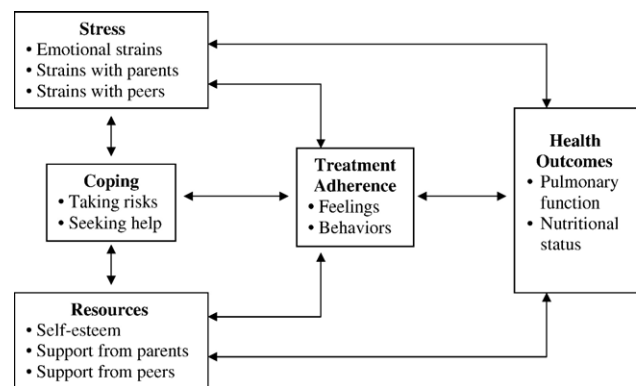


Fig. 1. Conceptual model showing relationship between strains, resources, adherence feelings/behaviors and health outcomes for youth with CF.

2.1. Measures

2.1.1. Living with CF Questionnaire (LCFQ)

A questionnaire was designed with the goal of measuring the different family, peer, and personal factors expected to be associated with living with CF as reviewed in the introduction and as shown in Fig. 1. Although, in retrospect, some of our hypothesized variables bear a relationship to some of the scales in the health-related quality of life Cystic Fibrosis Questionnaire–CFQ [28,29], our conceptualization and data collection preceded the dissemination and availability of the U.S. version of CFQ. For each questionnaire item, respondents indicated how often it occurred, choosing from the following descriptive categories: never (0), less than once a year (0.5), a few times a year (5), at least once a month (12), at least once a week (52), at least once a day (365). (Note: the number in parentheses represents the numeric value we assigned to each category for analytic purposes.) Due to high skewness in the response scale, a square root transformation was done to all variables prior to all analyses including scale and index construction.

2.1.2. Self-esteem

We included one established scale to assess self-esteem, which provided a measure of convergent validity with the psychological constructs in our conceptual model. The six-item Rosenberg [30] self-esteem inventory is a Likert-scaled measure of positive or negative attitudes toward the self. Excellent reliabilities and validities for this scale have been reported extensively in the literature [30,31]. For our sample, Cronbach's alpha for the self-esteem scale was 0.77.

2.2. Analysis

Exploratory principal component analysis with promax rotation and confirmatory factor analysis using maximum likelihood, guided by our conceptual model, were used to develop the scales.

Means and standard deviations of scales and items from the LCFQ, stratified by gender, were examined. Two-sample *t*-tests, adjusted for age, were used to test for differences in the scales between genders. With samples of size 49 and 54 in boys and girls respectively, there is 0.80 power to detect differences at the 0.05 level with effect size differences in the scales of at least 0.55. In other words, if the true means between boys and girls are different by at least half a standard deviation (assuming the standard deviations are the same), we have 0.80 power of detecting it. Furthermore the sample size provides 0.80 power to detect true correlations of size greater than 0.35 at the 0.05 level.

3. Results

3.1. Participants

Of the 103 youth who returned completed questionnaires (58% response rate), 49 were boys and 54 were girls. The mean

age for boys was 13.9 years (SD=4.22); for girls, it was 16.0 years (SD=3.95). Respondents were compared to non-respondents using the CF Center's clinically-recorded measures of their forced expired volume in 1 s (FEV₁) and their allometric index (AI) — the ratio of weight to predicted weight-for-height. Overall there were no significant differences between respondents and non-respondents in the parameters of interest.

However, when examined by gender and age group, for both boys and girls older than 16 years, the non-respondents had higher FEV₁ than the respondents (95.8% predicted versus 81.5% predicted for boys and 103.7% predicted versus 81.1% predicted for girls; $p=0.02$ for both comparisons). In addition, non-respondent girls younger than 12 years had a lower AI than the respondents in that age group (95% versus 106%, $p=0.015$).

Of the 103 respondents, 100 (96.2%) were Caucasian, two (1.9%) were Asian/Oriental/ Pacific Islander, and one youth specified "other" race. Of the 103 youth, 69 lived with two parents, 19 lived with their mothers only, one lived with father only, five lived with friends most of the time, five with a spouse/partner most of the time, three youth lived alone; for one, the living situation was not reported.

3.2. Measurement using the Living with Cystic Fibrosis Questionnaire (LCFQ) CF overall non-adherence index

Motivated by the different behaviors described in the Introduction related to possible gender differences in treatment adherence, the following items were included in the LCFQ: You skip taking your meds/pills, You skip doing chest physical therapy treatments, You skip eating the high-fat foods recommended for you, You limit your vigorous physical activity, and You feel embarrassed when you cough around others. Note that cough suppression is measured by embarrassment related to it and not what was actually done to adhere to treatment recommendations. Because of the distinct mechanistic way each type of non-adherence may relate to pulmonary function or nutritional status, it is of interest to consider each non-adherence item separately. In addition, a total non-adherence index was created by summing the five equally weighted items. CF overall non-adherence is conceived as an index and not a scale [32,33], i.e., it is not representative of some underlying trait (hence factor analysis or Cronbach's alpha on the items is not appropriate), but instead, it is completely defined by the items used to construct it. The CF overall non-adherence index has face validity in that it contains an item related to each of the CF treatment recommendations described in the introduction.

3.3. Living with CF scales

Development of the CF scales began by using exploratory factor analysis (EFA) with the 31 LCFQ items. The promax-rotated results are shown in Table 1. There were 11 eigenvalues greater than one, and these 11 dimensions explained 72% of the variability in these data. Due to the relatively small sample

Table 1
Results of exploratory factor analysis^a

	F1	F2	F3	F4	F5	F6	F7	F8	F9	F10	F11
1. You get into hassles/fights with your parents	0.76										
2. Your parent(s) nag you too much about doing your treatments	0.75										
3. Your mother protects you too much		0.76									
4. Your father protects you too much		0.81									
5. Your father expects you to exercise harder than you think you can			0.76								
6. Your mother expects you to exercise harder than you think you can			0.64								
7. Your parents disagree about how to manage your CF				0.80							
8. You receive understanding & support from your mother					0.90						
9. You receive understanding & support from your father				–0.46	0.52						
10. Kids make fun of all the pills you take							.84				
11. You try to hide your health situation from other kids											
12. Kids tease you because you are smaller than they are								.42			
13. Your friends show that they care about you					0.62						
14. You worry about the future						0.74					
15. You feel like you are under a lot of pressure and stress						0.66					
16. You feel lonely						0.70					
17. You feel sad						0.79					
18. You worry about being too thin							.81				
19. You worry about not having enough muscle						0.41	.53				
20. You worry about being fat								.77			
21. You feel bad about the way you look							.43	.54			
22. You are too tired to exercise									0.89		
23. You have a hard time keeping up in sports									0.80		
24. You stay at home when you don't really want to						0.73					
25. You drink beer, wine, or hard liquor dichotomized — Ever/Never										0.71	
26. You smoke cigarettes — dichotomized Ever/Never										0.84	
27. You talk with an adult about your problems											.68
28. You talk with an adult about your feelings about CF											.89
29. You talk to a friend about having CF											.40
30. You feel motivated to take care of yourself because of someone who cares about you											.71
31. You get so sick of all you have to do to take care of yourself that you want to give up				0.74							

^a All values of promax-rotated factor loadings greater than 0.40 are shown.

size and thus expected large variability, only factor loadings from the exploratory model greater than 0.4 are shown.

Forcing a confirmatory factor model guided by the EFA and our conceptual framework, we obtained the scales and items found in Table 2 and described below.

3.3.1. Parent–youth strains

We combined the first four factors (Table 1) found from the EFA (while separating off the negatively loaded question related to receiving understanding from your father and the self-discouragement question) to make this family system scale. Empirical evidence supporting this combination were the correlations found between the four factors measured with simple structure ranging from 0.28 to 0.69, and a well-fitting second order factor analysis that considered the seven items as measures of four factors and the four factors as measures of one shared underlying factor (Chi-square=5.8, $df=11$, p -value=0.88).

Cronbach's alpha for these seven items is 0.73. Studies of treatment adherence for chronic disease in children suggest that these parent–youth strains reflect the developmental stage of adolescents when youth are making an effort to differentiate and be more independent, with less control from their parents [21,34].

3.3.2. Parent support

Factor five from the EFA was split into two factors in the CFA, one for parent support and a separate one for friend support matching our conceptual framework. The parent support scale is comprised of two items, one asking about mother and one about father. Cronbach's alpha for this scale is 0.51.

3.3.3. Friend support

This scale was a single item: Your friends show that they care about you. While the EFA found this item loading on the factor with the parent support items, this item is not overly correlated with the parent support factor in the CFA ($r=0.39$). Thus, separating it from the parent support items is not empirically problematic and motivated by our conceptual framework, it is of interest to consider parents and friends separately.

3.3.4. Peer relationship strains

This scale has three items: Kids make fun of all the pills you take, You try to hide your health situation from other kids, Kids tease you because you are smaller than they are. The empirical evidence for this scale, based only on the EFA shown in Table 1 is weak. The two teasing items load on factors seven and eight which are also measured by questions 18–21 related to worries

Table 2
Results of confirmatory factor analysis^a

	Parent– youth strains	Parent support	Peer relationship strains	Friend support	Personal illness strains worries	Risk taking behavior	Seeking support	Self-care motivation	Self-care discouragement
You get into hassles/fights with your parents	0.46								
Your parent(s) nag you too much about doing your treatments	0.54								
Your mother protects you too much	0.75								
Your father protects you too much	0.60								
Your father expects you to exercise harder than you think you can	0.37								
Your mother expects you to exercise harder than you think you can	0.58								
Your parents disagree about how to manage your CF	0.40								
You receive understanding & support from your mother		0.35							
You receive understanding & support from your father		0.97							
Kids make fun of all the pills you take			0.27						
You try to hide your health situation from other kids			0.86						
Kids tease you because you are smaller than they are			0.50						
Your friends show that they care about you				1					
You worry about the future					0.66				
You feel like you are under a lot of pressure and stress					0.82				
You feel lonely					0.85				
You feel sad					0.69				
You worry about being too thin					0.31				
You worry about being fat					0.41				
You feel bad about the way you look					0.66				
You are too tired to exercise					0.40				
You have a hard time keeping up in sports					0.52				
You stay at home when you don't really want to					0.65				
You drink beer, wine, or hard liquor dichotomized — ever/never						0.32			
You smoke cigarettes — dichotomized Ever/Never						0.83			
You talk with an adult about your problems							0.77		
You talk with an adult about your feelings about CF							0.58		
You talk to a friend about having CF							0.39		
You feel motivated to take care of yourself because of someone who cares about you								1	
You get so sick of all you have to do to take care of yourself that you want to give up									1

^a Estimated standardized factor loadings are presented for all loadings not fixed to zero. Values of 1 are given for scales made up of only one item.

about appearance (not unexpected as peer teasing is certainly correlated with worries about appearance). The item related to trying to hide your health situation from other kids does not have a large loading with any of the 11 factors in the EFA, although it does have a 0.23 and 0.17 loading (not shown) with factors seven and eight respectively. Examination of just the three peer relationship strains items finds one dimension to explain 53% of the variability suggesting they share a common underlying dimension. Thus, based on theoretical grounds and some empirical evidence, the peer relationship strain scale was based on the three items. Cronbach's alpha for this scale is 0.54.

3.3.5. Illness strains and worries

This scale was comprised of items 14–24 (Table 1) related to factors six, seven, eight and nine from the EFA. These 11 items have a Cronbach's alpha of 0.85.

Additional analyses of just these 11 items found three eigenvalues greater than one, suggesting three factors, albeit moderately to highly correlated with each other in the promax rotation.

Thus, following our literature review, we also examined three subscales within this overall scale: *emotional strains* with a Cronbach's alpha of 0.84 (You worry about the

future, You feel like you are under a lot of pressure and stress, You feel lonely, You feel sad); *appearance worries* with a Cronbach's alpha of 0.55 (You worry about being too thin/fat — two items, You worry about not having enough muscle, You feel bad about the way you look); and *physical strains* with a Cronbach's alpha of 0.64 (You are too tired to exercise, You have a hard time keeping up in sports, You stay at home when you don't really want to).

3.3.6. Risk taking behavior

The two substance use items loaded together in the EFA (factor 10) and are included the same way in the CFA to make up the risk-taking behaviors factor: You drink beer, wine or hard liquor, You smoke cigarettes. Rather than using the original frequency scaling for these two items due to sparseness of response (less than 8% of respondents gave a response other than Never to cigarette smoking and 32% for alcohol use), both items were dichotomized as “never” versus “ever” and combined to make a scale resulting in values of 0, 1, or 2. The odds ratio between the two dichotomous variables was 7.6 (1.5–39.0, 95% CI).

3.3.7. Seeking support

This scale is derived from factor 11 in the EFA and contains the three support-seeking items: You talk with an adult about your problems, You talk with an adult about your feelings about CF, and You talk to a friend about having CF. Cronbach's alpha for this scale is 0.58. Note that the question related to self-care motivation also loaded on factor 11 but is considered on its own as a distinct single item scale for theoretical reasons.

3.3.8. Self-care motivation and self-care discouragement

Despite the suggestion from the EFA that the item, “You feel motivated to take care of yourself because of someone who cares about you,” loaded on the same factor as the three seeking support items, and the item “You get so sick of all you have to do to take care of yourself that you just want to give up” loaded on the same factor as mother and father's expectation of exercise, we treated each as separate single item scales for theoretical reasons. Both self-care motivation and discouragement represent multifaceted concepts and are representative of an internalized state of passivity and giving up, or conversely, of actively taking charge of the illness situation [10,17,24].

With the addition of some correlated error terms in the CFA model, the RMSEA was 0.079 (95% CI, 0.07–0.09); and the CFI was 0.75 with a Chi-square to degree of freedom ratio of 1.64. All estimated factor loadings from the CFA (Table 2) are statistically significant at the 0.01 level.

As a validity check of the different scales, we compared the correlations for each of them with the well developed self-esteem scale. Convergent validity of the strains scales and the self-care discouragement scale is supported by the expected significant negative correlations found between them and the self-esteem scale for both girls ($r = -0.30$ to -0.41) and boys

($r = -0.25$ to -0.43). See Table 3. For boys, the non-adherence index was negatively correlated with self-esteem ($r = 0.35$, $p < .01$); for girls, the correlation was in the expected direction ($r = -0.25$) but not significant. Finally, the convergent validity of the self-care motivation question is supported by the expected moderate positive correlation with self-esteem ($r = 0.35$, $p < .01$) for boys; and the positive correlation for girls ($r = 0.19$), although not significant.

3.4. Gender comparisons for LCFQ

Age-adjusted means for the LCFQ scales and items were compared for boys and girls in our sample (see Table 4). Girls scored higher than boys on all of the scales and items, including experiencing more strains as well as seeking and receiving more support. The gender differences were significant for illness strains and worries (including subscales for emotional strains and physical strains), friend support, and seeking support. Similarly, girls scored higher than boys on all of the adherence factors, and the differences were significant for self-care discouragement and the CF overall non-adherence index (including three of the five specific items: Embarrassed to cough, Skip high-fat foods, and Skip meds/pills). It is further noted that for both boys and girls, the most frequent non-adherence behavior was skipping chest physical therapy, but there was no significant gender difference in this behavior.

It should be noted that the means for three of the positive factors (parent support, friend support, and self-care motivation) are all considerably higher than the means for all of the negative factors, implying that the positive factors occur more frequently than the negative factors. Of the two items comprising the parent support scale, scores were significantly higher (p -value $< .01$) for support from mom compared to support from dad as reported by both boys and girls. However, there were no significant differences between boys and girls with CF on the parent support scale. The gender comparison for the self-esteem scale revealed an opposite pattern: boys scored significantly higher than the girls in our sample.

3.5. LCFQ intercorrelations and associations with age

As shown in Table 3, increasing age was significantly associated with (a) increases in risk taking behaviors for both boys ($r = 0.49$, $p < .000$) and girls ($r = 0.63$, $p < .000$) and (b) greater non-adherence for boys ($r = 0.36$, $p < .01$) and girls ($r = 0.33$, $p < .01$). Age also was associated with less self-care motivation for boys ($r = -0.28$, $p < .05$), but not for girls.

For girls, all of the strain scales (illness, with parents, with peers) were significantly associated with self-care discouragement and overall non-adherence ($r = 0.41$ to 0.51 , $p < .001$ to $.000$). For boys, self-care discouragement was not associated with any of the strains scales and non-adherence was only associated with illness strains ($r = .51$, $p < .000$) and peer strains ($r = 0.32$, $p < .05$). All of the strains scales were significantly intercorrelated for boys and even more strongly for girls. Self-

Table 3
Correlations between measurements (girls on upper triangle, boys on lower triangle)

	1	2	3	4	5	6	7	8	10	9	11	12
Strains												
1. Parent–youth strains		.56****	.51****	-.01	-.01	-.32**	.12	.03	-.01	.41***	.49****	-.05
2. Illness strains and worries	.31*		.47****	-.06	-.16	-.41***	.11	.14	-.18	.52****	.51****	.20
3. Peer relationship strains	.36**	.38**		.21	.16	-.33**	-.22	-.08	-.04	.51****	.41***	-.08
Resources												
4. Parent support	-.03	-.02	-.21		.55****	.16	.25	-.08	.42****	-.14	.02	-.16
5. Friend support	.07	-.06	-.28*	.28*		.09	.14	.03	.33**	-.09	.10	-.05
6. Self esteem	-.32*	-.43**	-.20	-.02	.06		.18	-.04	.19	-.30*	-.25	.02
Behaviors												
7. Seeking support	.38**	.18	-.03	.23	.15	-.04		.22	.37**	-.11	.22	.23
8. Risk taking behaviors	.24	.14	.48****	-.07	-.01	-.26	-.03		.12	.07	.38**	.63****
Adherence factors												
9. Self-care motivation	-.13	-.10	-.17	.23	.09	.35**	.39**	-.23		-.11	-.00	-.01
10. Self-care discouragement	.21	.13	.16	.08	.04	-.25	-.06	.04	.14		.43****	-.11
11. CF overall non-adherence index	.19	.51****	.32*	-.14	-.11	-.35**	-.18	.26	-.15	.30*		.33**
Age												
12. Age in years	-.12	.20	.19	-.08	-.12	-.16	-.20	.49****	-.28*	-.05	.36**	

*= $< .05$.
 **= $< .01$.
 ***= $< .001$.
 ****= $< .000$.

care discouragement was associated with non-adherence for both boys ($r=0.30, p<.05$) and girls ($r=0.43, p<.001$).

For girls, having support from parents and friends was significantly associated with self-care motivation ($r=0.42$ and 0.33 , respectively) but support was not associated with self-care discouragement or non-adherence. None of these associations were significant for boys. The behavior of seeking support was significantly associated with self-care motivation for both boys and girls ($r=0.39$ and 0.37 respectively, $p<.01$). Although in the expected direction, seeking support was not significantly associated with having support from parents or friends for either boys or girls. However, seeking support had an unexpected positive association with parent–youth strains, but only for boys ($r=0.38, p<.01$).

4. Discussion

4.1. Living with CF Questionnaire (LCFQ)

The CF overall non-adherence index and the scales and items derived from our exploratory and confirmatory factor analyses of the LCFQ show promise in assessing important psychosocial factors among youth living with CF. Using both empirical and conceptual considerations, the confirmatory factor analysis resulted in acceptable alpha reliabilities (.73–.85) for two of the strains scales — parent–youth strains and illness strains/worries. Of the three subscales within illness strains/worries, emotional strains also had an acceptable alpha (.84); however, physical strains and appearance worries had lower alpha coefficients (.64 and .55). Seeking support and parent support and peer relationship strains also had lower alpha coefficients in the .51 to .58 range, perhaps related to the small number of items in these scales. However, these scales with lower alpha coefficients

Table 4
T-test comparison of age-adjusted means for boys and girls for self-esteem and Living with CF scales and items

	Boys ($n=49$) Mean	Girls ($n=54$) Mean
Strains		
Parent–youth strains	2.76	3.54
Illness strains & worries	2.04	3.70**
Emotional strains	2.61	4.72*
Physical strains	1.37	3.10**
Appearance strains	1.98	3.12
Peer relationship strains	1.91	2.18
Resources		
Parent support	10.64	11.79
Mom support	12.34	13.94
Dad support	8.94	9.64
Friend support	8.81	12.41*
Self esteem ^a	3.29	2.93***
Behaviors		
Seeking support	2.49	3.91*
Risk taking behaviors	0.29	0.35
Adherence factors		
Self-care motivation	8.32	8.69
Self-care discouragement	3.32	6.31*
CF overall non-adherence index	10.90	20.71***
Embarrassed to cough	1.08	3.58**
Limit physical activity	0.87	2.04
Skip high-fat foods	1.37	3.32*
Skip chest physical therapy	5.27	7.04
Skip meds/pills	2.32	4.73*

* $p<.05$.
 ** $p<.01$.
 *** $p<.001$.

^a The metric for the self-esteem scale is different from all the other scales and items in this table.

did hold up with significant loadings in the confirmatory factor analysis thus providing empirical evidence over and above the conceptual justification for their combination.

4.2. Hypothesized gender differences in living with CF

When we examined gender differences using the CF overall non-adherence index of the LCFQ, we found partial support for our hypothesis that adolescent girls would show more non-adherent behaviors and feelings in managing their CF than boys. These differences are most evident for embarrassment about coughing, skipping meds/pills and skipping high-fat foods, all behaviors that may be more visible to peers and others and may contribute to self-consciousness among girls. Our findings are consistent with Walters' [35] report that girls with CF have poorer nutritional habits and body self-image compared to boys. Although, girls limited their physical activity more than boys, the difference only approached statistical significance ($p=0.065$). Of all the treatment behaviors, the most frequently skipped for both boys and girls was chest physical therapy, which is consistent with the DiGirolamo et al. [18] finding that physical therapy is the most disliked aspect of the CF treatment regimen. However, there was no significant gender difference in this behavior. Also related to the treatment regimen, girls reported significantly more discouragement about treatment than boys, indicating that they get so sick of taking care of themselves that they just want to give up. On the positive side, however, there was no gender difference in self-care motivation. These different results provide support for the importance of maintaining discouragement and motivation as two separate items in our model.

Emotional strains that emphasize sad and worrisome feelings were the most frequently reported component of the illness strains and worries scale. The emotional strains subscale included worry about the future, which supports the Willis et al. (2001) finding as more characteristic of girls with CF. Although girls reported more appearance strains than boys, as would be normatively expected, this gender difference only approached significance. Girls did report greater physical strains than boys, and they were more likely to limit physical activity compared to boys. Although we did not examine the relationship between physical activity and nutritional status, it is possible that suboptimal weight and height contributed to limitations in and worries about physical activity for girls, which would be consistent with the findings of Selvadurai et al. [16] Not surprisingly, experiencing more illness strains and worries was associated with greater discouragement, but only for girls.

Strains in girls' relationships with their peers also were associated with greater discouragement, which is consistent with what others have reported about girls being more attuned to their social context [11,22]. Consistent with more discouragement, worry and strains, girls reported significantly lower scores on self-esteem than boys, which corroborates the findings of Sawyer et al. [23] More strains with parents were associated with poorer self esteem for both boys and

girls, which is consistent with findings of Graetz et al. [20] that non-supportive behaviors from parents were associated with poorer psychological adjustment in their sample.

Consistent with our third hypothesis, girls did report that they sought support from others more often than boys did, and girls had higher scores than boys for having support from friends, again suggesting that emotional concerns and needs are more salient for girls as suggested by Willis et al. [10] However, there were no significant gender differences in perceived support from parents. Interestingly, both boys and girls reported receiving more support from their mothers than from their fathers, which may reflect the family system dynamic where mothers are more involved in chronic illness management than fathers [36,37]. It is important to note that for both girls and boys, their scores for parent and friend support were three to four times higher than their scores for the various strain scales. How well this greater emphasis on support versus strains represents all youth with CF cannot be determined given the self-selection of our sample from a regional CF treatment center acknowledged to have particularly good medical outcomes.

We were able to show that psychological strains at the personal, family, and friend level were all associated with reduced treatment adherence. It is, of course, possible that poorer adherence contributes to more strains and worries, particularly illness strains and worries. Both Willis et al. and Williams [10,24] suggest from their qualitative studies that non-adherence increases a sense of guilt, particularly for girls. These thoughts and behaviors may create a kind of vicious cycle for girls, each reinforcing the other. Parent–youth strains were associated with non-adherence, although it was only significant for girls. It is quite probable that non-adherence also increases the strain between parents and their youth with CF, with parents pushing harder and nagging more as they worry about the consequences of their youth not complying with treatment requirements. Parallel to normative tension between parents and their independence-seeking adolescents, this, too, can become an escalating vicious cycle with non-adherence increasing parents' tendency to monitor more and nag, which in turn may reduce an adolescent's motivation to follow through with the treatment regimen. In this study, we did not collect data on parents' perceptions of their youth's behaviors, which is an important next step in this research that would contribute additional understanding to the emergence and nature of these parent–youth issues.

Having or seeking support had no significant association with adherence for either boys or girls. However, seeking support was associated with more self-care motivation for both genders. The significant positive association between seeking support and parent–youth strains for boys was surprising. Perhaps boys who are not getting along with their parents seek support outside the home, although seeking support was not associated with having more friend support.

It is unclear from these data how self-care motivation relates to adherence since the associations were low and non-

significant. Furthermore, there was no significant association between self-care motivation and self-care discouragement. Only self-care discouragement was related to non-adherence for both boys and girls.

Even though smoking and alcohol use were risks taken very infrequently by our sample, risk taking behaviors were significantly associated with non-adherence for girls, and approached significance for boys. This gender difference may reflect the older age of girls in this sample where some experimentation with alcohol may be normative. Age and alcohol use were correlated 0.65 for girls and 0.51 for boys. Strains in peer relationships were significantly related to risk taking for boys, but not for girls, suggesting boys may cope differently with relationship strains than girls.

5. Conclusions

In sum, we found partial support for our first hypothesis that girls would be less adherent to the CF treatment regimen (overall adherence, coughing, high-fat foods, meds/pill) and experience greater self-care discouragement. In addition, our second hypothesis that girls would report more frequent strains than boys was partially supported with illness strains and worries (including emotional and physical strains) occurring more frequently for girls. Illness strains and worries had an understandable and strong relationship to non-adherence for both genders. Non-adherence was associated with strains with peers for both genders, but strains with parents was significant only for girls. We found partial support for our third hypothesis in that girls were more likely than boys to seek support and report support from friends; however contrary to our hypothesis, neither parent or friend support had a significant relationship with better adherence. Although not hypothesized, self-esteem was significantly higher for boys and it was associated with better adherence, but only for boys. Considering the higher scores for self-care discouragement and emotional strains and the lower self-esteem score, CF appears to have a greater emotional impact on girls.

6. Limitations

Our study findings are limited by having a relatively low response rate of 58%, and hence our sample does not necessarily represent the population of youth with CF in the U.S. Although there were only modest differences in clinical measures between respondents and non-respondents (with older non-respondents surprisingly having *worse* FEV₁ scores), we do not know how non-respondents would respond to the psychosocial factors in the LCFQ. Therefore, our results should be interpreted with this potential sample bias in mind. There is a need to replicate these findings with a larger sample, more representative of the range of biopsychosocial functioning among youth with CF. However, despite this sample limitation, the CF overall non-adherence index and the scales and items derived from the LCFQ offer

promise in assessing psychosocial factors that may contribute to understanding gender differences in CF morbidity and mortality. Additional psychometric information about the LCFQ scales could be obtained by examining associations with the health-related quality of life CF Questionnaire.

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