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PREVALENCE OF UNMET PALLIATIVE CARE NEEDS IN ADULTS WITH CYSTIC FIBROSIS

Elizabeth T. Trandel, BA^a, Joseph M. Pilewski, MD^a, Elisabeth P. Dellon, MD, MPH^b, Kwonho Jeong, MS^a, Jonathan G. Yabes, PhD^a, Laura T. Moreines, MSN, CRNP^c, Robert M. Arnold, MD, FAAHPM^a, Zachariah P. Hoydich, BS^a, Dio Kavalieratos, PhD^a

^aUniversity of Pittsburgh

^bUniversity of North Carolina at Chapel Hill

^cYale New Haven Health System

Abstract

Background—Physical and emotional burdens impair quality of life (QoL) in many adults with cystic fibrosis (CF). Palliative care (PC) improves QoL in other serious illnesses, yet the full array of palliative needs amenable to PC are unknown in CF.

Methods—We surveyed 164 adults with CF using the Supportive Care Needs Survey-34 (SCNS-34) to assess unmet PC needs across five domains, the Edmonton Symptom Assessment System (ESAS) to assess symptom burden, and the Cystic Fibrosis Questionnaire—Revised (CFQ-R) to assess CF-specific QoL. We assessed associations between SCNS-34 domain scores and respondent characteristics, including symptom burden and FEV₁.

Results—Median age was 29 years; 56% of respondents were male. Median FEV₁ was 57% predicted. 78% of respondents reported 1 unmet PC need; physical & daily living (72%) and psychological (66%) needs were most prevalent. Symptom burden was correlated with all SCNS-34 domains scores, and strongly correlated with the physical ($r=0.79$) and psychological ($r=0.72$) domain scores. FEV₁ was moderately inversely correlated with the physical domain score ($r=-0.41$). Forty-four of the 45 inverse correlations between SCNS-34 domain scores and CFQ-R domain scores were significant. Patient-reported anxiety and depressive symptoms were significantly associated with higher scores in five and four SCNS-34 domains, respectively.

Conclusions—Adults with CF have substantial unmet PC needs. Patient-reported symptom burden is more strongly associated with reporting unmet PC needs than FEV₁. Routine screening of unmet PC needs, using tools such as the SCNS-34, may enable CF care teams to optimize the provision of primary and specialist PC.

Correspondence concerning this article should be addressed to Dr. Dio Kavalieratos, Section of Palliative Care and Medical Ethics, Department of Medicine, University of Pittsburgh, 230 McKee Place, Suite 600, Pittsburgh, PA 15213.

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Keywords

Palliative care; needs assessment; patient-reported outcomes; quality of life

1. Background

Cystic fibrosis (CF) is a complex, chronic disease in which many adults with CF report suffering from persistent physical and psychological concerns, such as dyspnea, fatigue, anxiety, and depression.^{1–5} These individuals also cope with immense, life-long treatment burden and are required to navigate decisions around complex and high-risk treatments from a young age.⁶ These factors significantly impair quality of life (QoL) of individuals with CF.^{1–6} Palliative care is an overall approach to care, and a clinical specialty, that has been shown in a recent meta-analysis to improve important patient-centered outcomes, such as QoL and symptom burden.⁷

Whereas palliative care has been shown to improve outcomes in other serious illnesses, palliative care research specific to CF is very limited.⁸ When developing palliative care interventions for CF, caution should be paid to the disease's unique factors, such as its trajectory, difficult prognostication, early diagnosis, and expanding portfolio of treatment options. While previous research in CF has examined specific needs likely amenable to palliative care—such as symptoms, transplant-related concerns, and needs at the very end of life—the full spectrum of needs in CF remains unknown.^{1,9,10} Furthermore, no holistic, validated measure of palliative care needs exists for CF.

The literature on the role and efficacy of palliative care in CF is small, but growing.^{2,11–14} One recent pilot randomized clinical trial demonstrated feasibility, acceptability, and patient-perceived benefits of CF-specific longitudinal specialist palliative care, including improvements in physical symptoms, QoL, and mood.^{11,13} However, at present, specialist palliative care is rarely used in CF, and when people with CF do receive such specialist palliative care services, it is often in the immediate dying process.¹⁵ As research in lung cancer has shown that palliative care may be more effective in early-stage disease, utilization only at the immediate end of life may preclude patients and their families from receiving its full benefits.¹⁶

To aid in developing CF-specific palliative care services for those most in need, there is a need to understand the complete spectrum of unmet needs in CF and to recognize which patients most require additional support to address suffering. The goals of this study were to define the prevalence of unmet palliative care needs in adults with CF, to determine which clinical and demographic factors are associated with reporting increased unmet needs, and to begin the validation process of a palliative care needs assessment tool in CF.

2. Methods

2.1 Study design and participants

We recruited a convenience sample of English-speaking patients, aged 18 years, at an academic, multidisciplinary adult CF program that currently provides longitudinal care to

over 250 adults. Participants were approached for voluntary participation by a research assistant during scheduled clinic visits between December 2014 and August 2015. There was no incentive to participate. Survey data were collected on tablet computers and directly stored in REDCap, a secure, web-based research data capture application. These data were originally collected through a quality improvement (QI) project to assess quality of care, which led to the development of an embedded specialty palliative care intervention.¹³ While we provided the CF care team members with the results, we did not specifically track clinical follow-up based on this screening. The original QI project was approved by the UPMC Quality Improvement Review Committee. We subsequently received approval from the University of Pittsburgh Institutional Review Board (#PRO16070142) to de-identify the dataset and perform cross-sectional analyses, including the present analysis.

2.2 Measures

2.2.1 Measures: Supportive Care Needs Survey Short Form 34 (SCNS-34)—

We assessed unmet palliative care needs using the Supportive Care Needs Survey Short Form 34 (SCNS-34), a comprehensive needs assessment tool originally designed to measure the perceived needs of patients with cancer; it is well validated in oncology.¹⁷ As has been done in other conditions, such as cardiovascular diseases, we minimally reworded three items to be relevant to CF g., “fears about the cancer spreading” to “fears about my CF getting worse”).¹⁸

Respondents indicate their level of need for help for 34 items across five domains (psychological, health system & information, physical & daily living, patient care & support, and sexuality) using a five-point Likert scale (see complete list of items in Appendix). An answer of 1 is defined as having no need, 2 as having a need that was satisfied, and 3, 4, or 5 as having some degree of distress and recent unmet need. A standardized Likert summated score (potential range: 0–100) was calculated for each SCNS-34 domain; higher scores represent higher levels of unmet need.¹⁹

2.2.2 Measures: Edmonton Symptom Assessment System (ESAS)—

The Edmonton Symptom Assessment System (ESAS) assesses symptom burden by evaluating the severity of nine symptoms (pain, tiredness, nausea, depression, anxiety, drowsiness, shortness of breath, appetite, and feeling of wellbeing), each scored from 0 (no symptom) to 10 (worst possible symptom).²⁰ An ESAS total distress score was calculated as a sum of participants’ raw responses to the nine items (potential range: 0–90).

2.2.3 Measures: Cystic Fibrosis Questionnaire—Revised—

The Cystic Fibrosis Questionnaire—Revised (CFQ-R) is a validated, disease-specific instrument that measures the impact of CF on nine domains of health-related QoL (physical, emotional, social, role/school, vitality, body image, eating, treatment burden, and health perceptions) and three symptom scales (weight, respiratory, and digestion).^{21,22} Standardized summated scores were calculated for each of the nine HRQoL domains (potential range: 0–100), with higher scores indicating better HRQoL.

2.2.4 Measures: Demographic and Clinical Variables—We accessed the electronic medical record (EMR) for demographic variables (e.g., age, gender, race) and clinical measures (e.g., body mass index (BMI), FEV₁ at time of visit, number of hospitalizations in prior 12 months). We accessed the EMR’s “problem list” to determine prior clinical diagnoses of depression and/or anxiety, using any listed problem with “anxiety” or “depression” in the description.

2.3 Statistical Analysis

Domain-level standardized scores were calculated for the five SCNS-34 domains and the nine CFQ-R domains. We calculated the Cronbach’s alpha to assess the reliability of each of the five SCNS-34 domains for this population. We used Pearson’s correlations to determine associations between SCNS-34 domain scores and symptom burden, CFQ-R domain scores, and FEV₁ and Wilcoxon rank-sum test to compare SCNS-34 domain scores between categories of binary variables such as gender, clinical diagnosis of anxiety or depression, and patient-reported presence of anxiety or depressive symptoms. Lastly, we conducted a series of multivariable linear regression analyses to determine the adjusted association of various demographic and clinical factors on each of the five SCNS-34 domain scores. In each multivariable regression model, we included the following variables that could be justified clinically or theoretically as predictors of SCNS-34 domains: FEV₁, age, number of pulmonary exacerbations in prior 12 months, BMI, sex, clinical diagnosis of depression, and clinical diagnosis of anxiety. We did not model the CFQ-R or ESAS in multivariable regressions, as the directionality of the association between the SCNS-34 domains with these variables is unclear. All statistical analyses were performed using Stata/SE 15.1 for Windows (StataCorp LLC., TX). Significance was assessed at the $p < 0.05$ level.

3. Results

3.1 Demographics

We surveyed 164 patients from our adult CF clinic (71% of clinic population at time of data collection; see Table 1). The median age was 29 years (range: 18,66), and 56% of participants were male. The median FEV₁ was 57% predicted (range: 16%, 110%). Twenty-three percent of this sample had a clinical diagnosis of depression, and 17% had a clinical diagnosis of anxiety. Forty-six percent of respondents reported any symptoms of depression (ESAS score $\geq 1/10$), with 18% reporting moderate-to-severe depressive symptoms (ESAS score $\geq 4/10$). Fifty-four percent of respondents reported any symptoms of anxiety, with 26% reporting moderate-to-severe anxiety symptoms. We considered only respondents who reported moderate-to-severe symptoms. ESAS total distress score ranged from 0 to 75 out of a possible 90, with a median score of 15/90 (mild symptom burden). The nine standardized summated CFQ-R domain scores ranged from 48.27 (vitality) to 81.91 (eating).

3.2 Prevalence of Domain-Level Unmet Palliative Care Needs

Overall, 78% of participants reported at least one unmet palliative care need. Across the five need domains, 72% of participants reported one or more unmet physical need, 66% reported one or more unmet psychological need, 41% reported one or more health system &

information need, 30% reported one or more patient care & support need, and 20% reported one or more sexuality need.

SCNS-34 domain-level mean standardized summated scores ranged from 36.07 (physical & daily living) to 14.38 (sexuality) (see Table 2). While score ranges to categorize severity have not been established, the scores for both the psychological and physical domains are comparable to those for patients with a recent cancer diagnosis and inpatients with cardiovascular disease, and higher than those for survivors of cancer.^{18,23,24}

Using Cronbach's alpha, reliability was found to be acceptable for all domains except sexuality (alphas: health system & information=0.95, psychological=0.95, physical & daily living=0.91, patient care & support=0.89, sexuality=0.80). Within the sexuality domain, "being given information about sexual relationships" appears to weaken the reliability. Removing this item results in an increased alpha of 0.96.

3.3 Prevalence of Individual Unmet Palliative Care Needs

Three specific palliative care needs were indicated as unmet by at least half of respondents, and ten by at least one-third of respondents (see Table 3). The two most prevalent needs were in the physical & daily living domain: "lack of energy/tiredness" (65%) and "feeling unwell a lot of the time" (52%). The third most prevalent, "fears about my CF getting worse" (50%), was in the psychological domain. As seen in Table 3, the ten most prevalent needs were all either physical or psychological.

Prevalence of the remaining 24 unmet needs ranged from 31% ("keeping a positive outlook") to 6% ("being given information about sexual relationships") (see Appendix). The first need outside of either the physical or psychological domains, "being informed about whether your CF is under control," was the 15th most prevalent (25%). This need was in the health system & information domain.

Each need in the psychological and physical & daily living domains was endorsed as unmet by at least one-fifth of participants. Conversely, each need in the patient care & support and sexuality domains was endorsed as unmet by less than one-fifth of participants.

3.4 Correlations between SCNS-34 Domain Scores and Respondent Characteristics

Several clinical and demographic characteristics were significantly, although generally weakly, correlated with specific SCNS-34 domain scores (all $p < 0.05$; see Table 4). FEV₁ was moderately inversely correlated with the physical & daily living domain score and weakly inversely correlated with the psychological domain score. Number of pulmonary exacerbations in prior 12 months was weakly correlated with the physical and psychological domain scores. Female gender was associated with reporting significantly higher level of need in the physical & daily living domain ($p = 0.019$; see Table 5).

In comparison, symptom burden (as measured by the ESAS total score), was significantly correlated with all five SCNS-34 domain scores (all $p < 0.05$; see Table 4). Symptom burden was, as hypothesized, strongly correlated with the physical & daily living and psychological domain scores.

We examined the relationship between depression and anxiety and SCNS-34 domain scores in two distinct ways: by clinical history as documented in the EMR, or by way of patient-reported presence of anxiety or depressive symptoms, using the ESAS. A clinical history of depression was significantly associated with a higher SCNS-34 domain score in four of the five domains (all $p < 0.05$; see Table 5), while a history of anxiety was significantly associated with higher scores in the psychological and health system domains (both $p < 0.05$).

In comparison, current, patient-reported moderate-to-severe depressive symptoms were significantly associated with a higher score in all five SCNS-34 domains (all $p < 0.001$; see Table 5). Current, moderate-to-severe anxiety symptoms were significantly associated with a higher SCNS-34 domain score in four domains. These results demonstrate that patient-reported presence of symptoms of depression or anxiety is perhaps a stronger sentinel for present unmet palliative care need than a clinical history of these diagnoses.

Each of the nine CFQ-R domain scores—with higher scores indicating better health—was inversely correlated with all of the five SCNS-34 domains scores (see Table 4; all $p < 0.05$), with one exception: the SCNS-34 sexuality domain score was not significantly correlated with the CFQ-R treatment burden domain score. Again, the strongest correlations were in the physical and psychological domains.

3.5 Multivariable Linear Regression Analyses

Our multivariable analyses were generally consistent with the bivariate analyses in directionality and significance. FEV₁ was inversely associated with the SCNS-34 physical domain score ($b = -0.37$, 95% CI = $[-0.53, -0.20]$), age was associated with the physical ($b = 0.48$, 95% CI = $[0.09, 0.87]$) and sexuality ($b = 0.56$, 95% CI = $[0.26, 0.87]$) domain scores, and number of pulmonary exacerbations in the past 12 months was associated with the physical domain score ($b = 2.19$, 95% CI = $[0.53, 3.85]$) (all $p < 0.05$). A clinical diagnosis of depression was associated with four of the five SCNS-34 domain scores: psychological ($b = 14.28$, 95% CI = $[5.63, 22.94]$), health system & information ($b = 10.25$, 95% CI = $[2.85, 17.64]$), patient care & support ($b = 8.31$, 95% CI = $[0.93, 15.69]$), and sexuality ($b = 8.96$, 95% CI = $[2.29, 15.63]$) (all $p < 0.05$). Some variables were statistically significant in the correlation analyses, yet lost significance at $p < 0.05$ in the multivariable analyses: FEV₁ was no longer significantly associated with the SCNS-34 psychological domain score, a clinical diagnosis of anxiety no longer with the psychological or health system & information domain scores, number of pulmonary exacerbations no longer with the psychological domain score, and female gender no longer with the physical & daily living domain score. The direction of these associations did remain congruent with the correlation analyses. Additionally, the associations that were not significant in the correlation analyses did not reach significance in the multivariable analyses. Full results are available in the Appendix.

4. Discussion

To our knowledge, this is the first study to assess the full breadth of palliative care needs in adults with CF and to examine the relationship between these needs and respondent characteristics. We found that adults with CF have substantial unmet palliative care needs

and that self-reported symptom burden is more strongly associated with reporting unmet needs than objective clinical or demographic factors.

Our study demonstrated substantial unmet palliative care needs in adults with CF, especially physical and psychological needs. More than 75% of participants reported at least one unmet palliative care need overall, with 72% reporting one or more physical & daily living need and 66% reporting one or more psychological need. Comparatively, only 20% reported one or more unmet sexuality-related needs. These data show that the unmet needs of adults with CF are different than those of other lung diseases, such as lung cancer, as prevalence of physical needs is higher and prevalent of health system & information needs is lower in CF.²⁵ The most prevalent needs—“lack of energy/tiredness,” “feeling unwell a lot of the time,” “fears about my CF getting worse,” and “not being able to do the things you used to do”—are all core foci of palliative care. These data justify the use of palliative care services in CF, as palliative care has been shown to improve similar needs in other conditions.⁷

We found that FEV₁—a clinical measure of pulmonary function—was moderately inversely correlated with the SCNS-34 physical & daily living domain. This finding is intuitive: as pulmonary disease progresses, FEV₁ decreases and activities of daily living require greater effort. However, because it was not broadly correlated with the unmet need domains, FEV₁ is unlikely to be a valuable proxy for unmet palliative care needs broader than physical needs, such as psychological, existential, or informational needs. Beyond FEV₁, we found either small or no correlations between unmet need and other objective demographic information, including age, BMI, and number of recent pulmonary exacerbations.

Our findings align with research documenting the high prevalence of depression and anxiety in adults with CF.^{4,5} Our data demonstrate that over half of individuals attending regular CF clinic visits report depression and nearly half report anxiety. The demonstrated association between such symptoms and unmet palliative care needs—including needs beyond the psychological domain—reinforces the importance of prior consensus recommendations calling for systematic mental health screening and treatment in CF care.²⁶ As our data were gathered at regular CF clinic visits, the prevalence of symptoms of depression and anxiety among patients who do not regularly attend clinic may be different, perhaps higher.

Our data also underscore the importance of systematically screening for patient-reported outcomes (PROs) beyond anxiety and depression in CF care. In other diseases, systematically collecting and monitoring PROs has been shown to improve a broad range of outcomes, including clinician awareness of symptoms, symptom management, patient-clinician communication, patient satisfaction, QoL, and even survival.^{27–29} Yet, as our data show, adults with CF may suffer from a variety of other burdens, including physical symptoms and existential distress. Given the correlation between symptom burden and other palliative needs, symptom burden may appear to be an appealing proxy; yet, we caution that proxies lack the nuance needed to identify and action individual palliative needs. As such, we suggest that current screening efforts be expanded to capture the larger spectrum of CF-related burdens amenable to palliative intervention. Such a holistic screening approach can guide the provision of primary palliative care by the CF care team—including basic symptom management and beginning discussions about suffering, prognosis, and goals—

while allowing for identification of the subset of patients at highest need for specialty-level palliative care. Yet to minimize response fatigue, further research is needed to identify a parsimonious set of measures that allow for complete yet efficient capture of palliative needs in CF that align with existing screening efforts.

Our study is the first to begin the process of validating the SCNS-34 in adults with CF. We found strong inverse correlation between the SCNS-34 and the CFQ-R, therefore demonstrating convergent validity between the SCNS-34 and a widely-used and well-validated CF-specific HRQoL tool. We also demonstrate concurrent validity, as the SCNS-34 is associated with variables with which we would expect it to be associated, such as ESAS total distress score and current presence of anxiety and depressive symptoms. We were unable to assess predictive validity in this analysis due to its cross-sectional nature. While the validity of the SCNS-34 in CF remains to be explored, we provide preliminary evidence that it is a useful tool to screen for unmet palliative care needs in CF.

There are limitations to this study. The predictive and discriminant validities of the SCNS-34 remain unknown in CF, yet we show the instrument displays promising convergent and concurrent validity. The ESAS is also not a CF-specific tool, and may overlook relevant symptoms, such as coughing. Additionally, because we determined prior clinical history of depression and anxiety using the EMR's "problem list," our estimates may be either understated due to underreporting, or overstated due to inclusion of more mild manifestations of anxiety/depression such as "adjustment disorder with anxiety." Our convenience sampling strategy may have introduced selection bias within our site, yet this concern is minimized given that 71% of our clinic's population participated. Because our data were collected at outpatient visits, there may be a subset of patients who were not surveyed because they either are frequently inpatient or do not attend regular clinic visits. Finally, because this was a cross-sectional study, it does not evaluate how unmet needs, symptom burden, or HRQoL change during disease progression, nor can causality be determined.

This study demonstrates that adults with CF experience a diverse array of unmet palliative care needs—particularly in highly-symptomatic individuals—and suggests that palliative care services may be beneficial in CF care. Furthermore, as subjective PROs are more strongly associated with reporting unmet needs than other clinical or demographic factors, screening for unmet palliative care needs at CF clinics may allow clinicians to determine which patients most require further primary and specialty palliative support.

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Appendix

Table 1.

SCNS-34 Needs

SCNS-34 Domain	SCNS-34 Need
Psychological (10)	Anxiety
	Feeling down or depressed
	Feelings of sadness
	Fears about my CF getting worse
	Worries that the results of treatment are beyond your control
	Uncertainty about the future
	Learning to feel in control of your situation
	Keeping a positive outlook
	Feelings about death and dying
	Concerns about the worries of those close to you
Physical (5)	Pain
	Lack of energy/tiredness
	Feeling unwell a lot of the time
	Work around the home
	Not being able to do the things you used to do
Health System & Information (11)	Being given written information about the important aspects of your care
	Being given information (written, diagrams, drawings) about aspects of managing your illness and side-effects at home
	Being given explanations of those tests for which you would like explanations
	Being adequately informed about the benefits and side-effects of treatments before you choose to have them
	Being informed about your test results as soon as feasible
	Being informed about whether your cystic fibrosis is under control
	Being informed about things you can do to help yourself to get well
	Having access to professional counselling (eg, psychologist, social worker, counsellor, nurse specialist) if you, family, or friends need it
	Being treated like a person, not just another case
	Being treated in a hospital or clinic that is as physically pleasant as possible
Having one member of hospital staff with whom you can talk to about all aspects of your condition, treatment and follow-up	
Patient Care & Support (5)	More choice about which doctor you see
	More choice about which hospital you attend
	Reassurance by medical staff that the way you feel is normal
	Hospital staff attending promptly to your physical needs
	Hospital staff acknowledging, and showing sensitivity to, your feelings and emotional needs
Sexuality (3)	Changes in sexual feelings
	Changes in sexual relationships
	Being given information about sexual relationships

Table 2.

Prevalence of Unmet Palliative Care Needs in Adults with CF (N=164)

Overall Rank	Prevalence	Need	Domain
1	65%	Lack of energy/tiredness	Physical & daily living
2	52%	Feeling unwell a lot of the time	Physical & daily living
3	50%	Fears about my cystic fibrosis getting worse	Psychological
4	43%	Not being able to do the things you used to do	Physical & daily living
5	40%	Anxiety	Psychological
6	39%	Uncertainty about the future	Psychological
7	38%	Worry that the results of treatment are beyond your control	Psychological
8	37%	Pain	Physical & daily living
9	35%	Work around the home	Physical & daily living
10	34%	Concerns about the worries of those close to you	Psychological
11	31%	Keeping a positive outlook	Psychological
12	30%	Feeling down or depressed	Psychological
13	29%	Learning to feel in control of your situation	Psychological
14	28%	Feelings of sadness	Psychological
15	25%	Being informed about whether your cystic fibrosis is under control	Health system & information
16	24%	Feelings about death and dying	Psychological
17	21%	Being informed about things you can do to help yourself to get well	Health system & information
18	19%	More choice about which doctor you see	Patient care & support
19	18%	Being treated in a hospital or clinic that is as physically pleasant as possible	Health system & information
20	17%	Having one member of hospital staff with whom you can talk to about all aspects of your condition, treatment and follow-up	Health system & information
21 (tie)	16%	Being treated like a person, not just another case	Health system & information
21 (tie)	16%	Being informed about your test results as soon as feasible	Health system & information
21 (tie)	16%	Changes in sexual relationships	Sexuality
21 (tie)	16%	More choice about which hospital you attend	Patient care & support
26 (tie)	16%	Being adequately informed about the benefits and side-effects of treatments before you choose to have them	Health system & information
26 (tie)	16%	Hospital staff attending promptly to your physical needs	Patient care & support
27	15%	Having access to professional counselling (eg, psychologist, social worker, counsellor, nurse specialist) if you/, family, or friends need it	Health system & information
28 (tie)	13%	Being given explanations of those tests for which you would like explanations	Health system & information

Overall Rank	Prevalence	Need	Domain
28 (tie)	13%	Reassurance by medical staff that the way you feel is normal	Patient care & support
28 (tie)	13%	Hospital staff acknowledging, and showing sensitivity to, your feelings and emotional needs	Patient care & support
28 (tie)	13%	Changes in sexual feelings	Sexuality
32	9%	Being given written information about the important aspects of your care	Health system & information
33	8%	Being given information (written, diagrams, drawings) about aspects of managing your illness and side-effects at home	Health system & information
34	6%	Being given information about sexual relationships	Sexuality

Table 3.

Multivariable linear regression models predicting SCNS-34 domain scores (N=164)

Coeff [95% CI] (p-value)	Psychological	Physical & Daily Living	Health System & Information	Patient Care & Support	Sexuality
FEV¹	-0.14 [-0.30, 0.03] (0.100)	-0.37 [-0.53, -0.20] (0.000)*	0.00 [-0.14, 0.13] (0.945)	-0.03 [-0.17, 0.11] (0.698)	0.04 [-0.08, 0.17] (0.508)
Age	0.17 [-0.22, 0.56] (0.384)	0.48 [0.09, 0.87] (0.017)*	0.17 [-0.17, 0.50] (0.326)	0.26 [-0.07, 0.60] (0.122)	0.56 [0.26, 0.87] (0.000)*
Number of pulmonary exacerbations	1.59 [-0.07, 3.25] (0.061)	2.19 [0.53, 3.85] (0.010)*	0.53 [-0.89, 1.94] (0.465)	0.56 [-0.86, 1.97] (0.439)	1.03 [-0.25, 2.31] (0.113)
BMI²	0.49 [-0.50, 1.49] (0.330)	0.12 [-0.88, 1.11] (0.818)	0.48 [-0.37, 1.33] (0.267)	0.51 [-0.34, 1.35] (0.240)	-0.02 [-0.79, 0.74] (0.955)
Female	4.88 [-2.45, 12.22] (0.190)	5.38 [-1.95, 12.71] (0.149)	2.88 [-3.39, 9.14] (0.366)	2.86 [-3.39, 9.12] (0.367)	2.18 [-3.47, 7.83] (0.448)
Diagnosis of Depression	14.28 [5.63, 22.94] (0.001)*	7.50 [-1.15, 16.16] (0.089)	10.25 [2.85, 17.64] (0.007)*	8.31 [0.93, 15.69] (0.028)*	8.96 [2.29, 15.63] (0.009)*
Diagnosis of Anxiety	3.68 [-6.06, 13.42] (0.457)	1.86 [-7.88, 11.60] (0.707)	3.81 [-4.51, 12.13] (0.367)	1.72 [-6.59, 10.03] (0.683)	2.86 [-4.64, 10.36] (0.453)
Constant term	13.60 [-8.04, 35.23] (0.216)	33.92 [12.29, 55.55] (0.002)	-0.51 [-18.98, 17.97] (0.957)	-3.01 [-21.46, 15.44] (0.748)	-9.62 [-26.28, 7.04] (0.256)

* p-value<0.05

¹ Forced expiratory volume in one second, as measured at time of visit

² Body mass index

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HIGHLIGHTS

- Unmet physical and psychological palliative needs are prevalent in adults with CF
- Symptoms are a stronger predictor of needs than physiologic measures (e.g., FEV₁)
- Unmet palliative care needs are associated with lower quality of life in CF
- The SCNS-34 is a clinically useful tool to evaluate palliative care needs in CF

Table 1.

Sample Characteristics

Characteristic	n (%) ¹ [N=164]
<i>Social Demographics</i>	
Age	
Median (range)	29 (18, 66)
18 – 29	84 (51%)
30 – 39	55 (34%)
40 – 49	14 (9%)
50+	11 (7%)
Racial Background	
Caucasian	160 (98%)
African American	1 (1%)
Other or multiple races	3 (2%)
Ethnic Background	
Non-Hispanic	163 (99%)
Hispanic or Latina/o	1 (1%)
Gender	
Male	92 (56%)
Female	72 (44%)
Relationship Status	
Single	101 (62%)
Partnered/Married	63 (38%)
Work or school status	
Attending school outside the home	21 (13%)
Looking for work	6 (4%)
Working full or part time	71 (43%)
Full time homemaker	12 (7%)
Not attending school or working due to my health	41 (25%)
Not working for other reasons	13 (8%)
Education	
Some high school or less	4 (2%)
High school diploma/GED	51 (31%)
Vocational School	2 (1%)
Some college	32 (20%)
College degree	58 (35%)
Professional or graduate degree	17 (10%)

Characteristic	n (%) ¹ [N=164]
Religious Preference	
Protestant Christian	30 (18%)
Roman Catholic	44 (27%)
Other Christian	30 (18%)
Jewish	2 (1%)
Buddhist	1 (1%)
Agnostic/Atheist/No religion	21 (13%)
Other	36 (22%)
Importance of religious or spiritual life	
Not at all important	33 (20%)
Not too important	36 (22%)
Fairly important	46 (28%)
Very important	49 (30%)
Finances (at end of month)	
Money left over	75 (46%)
Just enough money to make ends meet	68 (41%)
Not enough money to make ends meet	21 (13%)
<i>Clinical Characteristics</i>	
FEV₁%²	
Median (range)	57 (16, 110)
Mild (FEV ₁ ≥ 80%)	42 (26%)
Moderate (50 – 79%)	58 (35%)
Severe or Very severe (< 49%)	64 (39%)
Transplant status³	
Ineligible	158 (96%)
Pre-transplant	5 (3%)
Post-transplant	1 (1%)
Pancreatic insufficiency	126 (77%)
CF-related diabetes	64 (39%)
Diagnosis of depression	37 (23%)
Diagnosis of anxiety	28 (17%)
Pulmonary exacerbations⁴ in prior 12 months, Median (range);	0 (0,21)
Hospitalizations in prior 12 months, Median (range)	1 (0, 36)
Outpatient CF clinic visits in prior 12 months, Median (range)	4 (0,10)

Characteristic	n (%) ¹ [N=164]
BMI ⁵ , Median (range)	21.4 (15.2, 43.0)
<i>Patient-Reported Symptoms</i>	
Current depressive symptoms	
Any depression	75 (46%)
Mo derate-to-severe depression	30 (18%)
Current anxiety symptoms	
Any anxiety	87 (54%)
Moderate-to-severe anxiety	42 (26%)
Symptom burden	
No symptom burden	18 (11%)
Mild	100 (61%)
Moderate	44 (27%)
Severe	2 (1%)
CFQ-R standardized domain scores	
Physical	60.82 ± 31.34
Emotional	65.41 ± 16.41
Social	68.63 ± 20.78
Role/school	74.75 ± 24.57
Vitality	48.27 ± 13.64
Body image	60.82 ± 31.34
Eating	81.91 ± 25.35
Treatment burden	56.84 ± 20.61
Health perception	58.13 ± 25.60

¹Percentage totals might exceed 100% due to rounding off errors.

²Forced expiratory volume in one second, as measured at time of visit

³Patients ineligible for lung transplantation were either evaluated and deemed ineligible, or were not yet clinically eligible for transplant evaluation at time of data collection. Pre-transplant patients were either actively on the transplant list or were evaluated and deferred.

⁴Increase in respiratory symptoms (e.g., increased cough, sputum production, shortness of breath) accompanied by an acute decrease in lung function, resulting in hospital admission

⁵Body mass index

Table 2.

Mean SCNS-34 Domain-Level Standardized Likert Summated Scores in Adults with CF

Domain	# items	Mean Domain Score \pm SD [N=164]
Physical & daily living	5	36.07 \pm 26.02
Psychological	10	29.28 \pm 24.21
Health system & information	11	19.69 \pm 19.87
Patient care & support	5	18.63 \pm 19.74
Sexuality	3	14.38 \pm 18.58

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Table 3.

Most Prevalent Unmet Palliative Care Needs in Adults with CF

Overall Rank	Prevalence	Need	Domain
1	65%	Lack of energy/tiredness	Physical & daily living
2	52%	Feeling unwell a lot of the time	Physical & daily living
3	50%	Fears about my cystic fibrosis getting worse	Psychological
4	43%	Not being able to do the things you used to do	Physical & daily living
5	40%	Anxiety	Psychological
6	39%	Uncertainty about the future	Psychological
7	38%	Worry that the results of treatment are beyond your control	Psychological
8	37%	Pain	Physical & daily living
9	35%	Work around the home	Physical & daily living
10	34%	Concerns about the worries of those close to you	Psychological

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Table 4.

Pearson's Correlations Coefficients between SCNS-34 Domain Scores and Respondent Variables in Adults with CF

	Psychological	Physical & Daily Living	Health System & Information	Patient Care & Support	Sexuality
FEV1	-0.18*	-0.41*	-0.02	-0.04	-0.02
Continuous ESAS Score	0.72*	0.79*	0.40*	0.40*	0.37*
CFQ-R Domain Scores					
Physical Functioning	-0.58*	-0.80*	-0.34*	-0.37*	-0.27*
Emotional Functioning	-0.65*	-0.60*	-0.44*	-0.39*	-0.26*
Social Functioning	-0.59*	-0.59*	-0.41*	-0.44*	-0.32*
Role/school	-0.63*	-0.78*	-0.40*	-0.44*	-0.29*
Vitality	-0.53*	-0.66*	-0.32*	-0.31*	-0.26*
Body Image	-0.58*	-0.80*	-0.34*	-0.37*	-0.27*
Eating	-0.40*	-0.49*	-0.31*	-0.35*	-0.22*
Treatment Burden	-0.35*	-0.37*	-0.17*	-0.19*	-0.12
Health Perceptions	-0.68*	-0.81*	-0.43*	-0.42*	-0.35*
Age	0.09	0.20*	0.10	0.15	0.28*
Number of pulmonary exacerbations in past 12 months	0.22*	0.30*	0.09	0.09	0.15
BMI	0.01	-0.10	0.09	0.10	0.06

* p-value<0.05

Table 5.

Median SCNS-34 Domain Scores for Binary Demographic Characteristics in Adults with CF

	Median [IQR]	Psychological	Physical & Daily Living	Health System & Information	Patient Care & Support	Sexuality
Gender	M (n=92)	25.0[5.0,40.0]	30.0[10.0,55.0]	14.8[0.0,27.3]	17.5[0.0,25.0]	0.0[0.0,25.0]
	F (n=72)	30.0[10.0,50.0]	40.0[27.5,55.0]	23.9[1.1,27.3]	20.0[0.0,30.0]	8.3[0.0,25.0]
	p-value [‡]	0.054	0.019 *	0.179	0.175	0.168
Clinical diagnosis of depression	No (n=127)	25.0[5.0,40.0]	35.0[10.0,55.0]	13.6[0.0,25.0]	10.0[0.0,25.0]	0.0[0.0,25.0]
	Yes (n=37)	40.0[25.0,60.0]	40.0[30.0,55.0]	25.0[15.9,36.4]	25.0[0.0,30.0]	25.0[0.0,25.0]
	p-value [‡]	0.001 *	0.063	0.001 *	0.038 *	0.023 *
Clinical diagnosis of anxiety	No (n=136)	25.0[5.0,40.0]	35.0[10.0,55.0]	14.8[0.0,25.0]	15.0[0.0,25.0]	0.0[0.0,25.0]
	Yes (n=28)	33.8[18.8,55.0]	42.5[30.0,60.0]	25.0[12.5,33.0]	25.0[0.0,30.0]	25.0[0.0,33.3]
	p-value [‡]	0.031 *	0.076	0.019 *	0.157	0.102
Moderate-to-severe depressive symptoms in ESAS	No (n=134)	25.0[5.0,37.5]	30.0[10.0,45.0]	13.6[0.0,25.0]	10.0[0.0,25.0]	0.0[0.0,25.0]
	Yes (n=30)	60.0[45.0,70.0]	57.5[45.0,75.0]	30.7[15.9,45.5]	27.5[20.0,45.0]	25.0[0.0,41.7]
	p-value [‡]	<.001 *	<.001 *	<.001 *	<.001 *	<.001 *
Moderate-to-severe anxiety symptoms in ESAS	No (n=122)	21.3[5.0,35.0]	30.0[10.0,45.0]	13.6 [0.0, 5.0]	10.0[0.0,25.0]	0.0[0.0,25.0]
	Yes(n=42)	57.5[37.5,70.0]	60.0[40.0,70.0]	25.0[13.6,36.4]	25.0[10.0,40.0]	16.7[0.0,33.3]
	p-value [‡]	0.004 *	<.001 *	<.0001 *	<.001 *	0.063

[‡]Wilcoxon rank-sum test

* p-value<0.05