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Development of a Cystic Fibrosis Primary Palliative Care Intervention: Qualitative Analysis of Patient and Family Caregiver Preferences

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Abstract

To prevent or mitigate chronic illness burden, people with cystic fibrosis (pwCF) and their family caregivers need primary (generalist-level) palliative care from the time of diagnosis forward. We used qualitative methods to explore their preferences about a screening-and-triage model ("*Improving Life with CF*") developed to standardize this care. We purposively sampled and interviewed 14 pwCF and caregivers from 5 *Improving Life with CF* study sites. Thematic analysis was guided by *a priori* codes using the National Consensus Project's Guidelines for Quality Palliative Care. Participants included 7 adults and 2 adolescents with CF (3 with advanced disease), 4 parents, 1 partner (7 women; 5 people of color). Few were familiar with palliative care.

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Illness burden was described in multiple domains, including physical (e.g., dyspnea, pain), psychological (e.g., anxiety), and social (e.g., family well-being; impact on work/school). Most preferred survey-based screening with care coordination by the CF team. Preferences for screening approaches varied. PwCF and caregivers experience illness burden and are receptive to a CF-team delivered primary palliative care screening-and-triage model with flexible processes.

Keywords
cystic fibrosis, primary palliative care, qualitative research, patient-centered research, program development, quality of health care, caregiver burden

Introduction

Approximately 31,000 children, adolescents, and adults of varying races and ethnicities in the United States live with cystic fibrosis (CF), a genetic disease characterized by progressive organ dysfunction due to abnormal functioning of the cystic fibrosis transmembrane conductance regulator (CFTR) protein (1). Common manifestations include recurrent infections of the lungs and sinuses, pancreatic insufficiency, gastrointestinal dysmotility, CF-related diabetes, arthropathy, and bone disease (2–4). People with CF (pwCF) may have distressing symptoms, such as breathlessness, constipation, pain, and anxious or depressed mood, and may experience progressive impairment of physical or psychosocial functioning (5,6). Caregiver distress may also be very high (7). Recently, the advent of disease-modifying therapy—the CFTR modulators—has extended life expectancy, helped control symptoms, prevent or reduce complications, and made treatment burden more manageable for many pwCF (5–10). However, at this time, there remains no cure for CF, and the current life expectancy of pwCF is 53 years (11). Illness burden can be high for both pwCF and their families, and the need for effective strategies to sustain quality of life continues (8,9).

Palliative care mitigates illness burden and is recognized as an essential component of CF care by clinicians, pwCF and their family caregivers (10,12). A comprehensive plan of care aligned with the principles and practices of palliative care is appropriate from the time of CF diagnosis forward (6). Although access to specialists in palliative care is limited, best practices in primary (generalist-level) palliative care (13) could be implemented by the CF team in multidisciplinary CF clinic settings. Variation exists across and within practices, however (14,15). While prior work in non-CF populations has examined models of palliative care emphasizing both primary and specialty level services (16), few studies have explored the applicability and features of these models in CF (17). In one study of pwCF, family caregivers, and CF providers, respondents perceived that palliative care for CF was unique due to the life-long, fluctuating and progressive nature of CF, the close relationships forged between pwCF and their care teams over time, and the multiple care transitions throughout the lifespan, such as moving from pediatric to adult-centered care, and for some, to lung transplant care (12). Therefore, it is crucial to ensure that palliative care services are designed and implemented to address the quality of life concerns of pwCF and their families across diverse settings.

To provide a framework for addressing these gaps in daily clinical practice, The Cystic Fibrosis Foundation (CFF) convened a committee to develop consensus guidelines for models of CF palliative care delivery (18). The committee used the National Consensus Project's Clinical Practice Guidelines for Quality Palliative Care (NCP guidelines) (19) as a starting point and endorsed the NCP framework for high-quality primary palliative care that includes: structures and processes that promote patient-centered and family-focused care; interventions that address the physical, psychological, social, and spiritual sources of distress or burden; care that is culturally-sensitive, ethical and legal; and competent care at the end of life (20). The committee evaluated screening tools (18,21) and proposed a model of primary palliative care for CF that incorporates regular screening in CF care settings to identify concerns that could be addressed by CF care teams. The model aims to ensure access to palliative care interventions for pwCF and their families as the need arises.

Although the CFF palliative care guidelines build upon established NCP guidelines, information to help guide their implementation in CF-specific settings is limited and the opinions of pwCF about potential care processes and interventions that could impact their care experience have yet to be explored. The present qualitative study and its parent study afforded a starting point for addressing these important gaps.

With support from the CFF, our team has designed and implemented a multi-site pragmatic trial to extend and evaluate this model of primary palliative care for CF: the "*Improving Life with CF*" study (20). The project includes screening-and-triage workflows, best practice treatment guidelines for high frequency problems, patient education, and provider training including on-demand webinars and a 6-hour curriculum on best practices using a "train-the-trainer" approach.

During the early design of the model, we interviewed pwCF and family caregivers of pwCF with the goal of better understanding their:

1. Prior experiences with palliative care, advance care planning, and goal-setting discussions;
2. Perceptions of the illness burden associated with CF, including the applicability of the key domains of the

NCP guidelines for Quality Palliative Care in identifying sources of distress or burden, and,
3. Preferences for the varied components of a palliative care screening-and-triage intervention (e.g., process of initial screening, follow-up care, and models of screening).

Herein, we present the results of a qualitative analysis of the interview transcripts, which further informed both the design and ongoing prospective evaluation of feasibility and outcomes of the *Improving Life with CF* intervention.

Material and Methods

Overview of Study Design

We conducted a qualitative study incorporating purposive sampling of a diverse group of pwCF and family members of pwCF. Participants were recruited from five CF centers that were participating in the *Improving Life with CF* palliative care intervention. These centers are located in Manhattan and in the New York City suburban area, Boston, and Atlanta. Participants completed a 1-hour semi-structured interview consisting of 15 questions and probe follow-up questions. The interview guide was developed by the study Principal Investigators (PIs; A.G. and L.D.) with input from additional members of the study team including a palliative care expert (R.P.) and a qualitative researcher (M.B.). Questions were organized by 5 content areas: (1) Concepts and principles guiding the program; how the CF community understands palliative care, (2) Concerns or conditions that might be addressed by a screening program aimed at improving quality of life and illness burden, (3) Process of screening, (4) Process of follow-up (i.e., triage), and (5) Scenarios for feedback on 2 different screening models: Clinic-based versus home-based screening (see Supplemental Appendix: Semi-structured Interview Guide: Topics and Outline).

Table 1. Participant Characteristics.

Characteristic	N=13
Persons with CF	8
Age 12–17 years	2
Age ≥18 years	6
CFTR modulator use	6
Advanced CF lung disease (FEV_1 <40% predicted)	3
Lung transplant recipient	1
Caregivers of persons with CF	5
Parent of child ages 0-11 years	2
Parent of adolescent ages 12-17 years	1
Parent of adult	1
Partner/spouse	1
Caregiver of patient with advanced lung disease	1
Caregiver of patient using CFTR modulator therapy	2
Participant sex	
Male	6
Female	7
Participant Race/Ethnicity	
Non-Hispanic White	8
Non-Hispanic Black	2
Hispanic	2
Asian	1

Study Sites and Purposive Sampling Technique

PwCF and family caregivers were recruited separately (i.e., not as related dyads) from each of the 5 participating study sites: Massachusetts General Hospital, Mt. Sinai Hospital, Northwell Health, Stony Brook University Medical Center, and Children's Healthcare of Atlanta. Each site was asked to recruit at least 2 participants based on the primary demographics of the patient population managed by the practice (e.g., Northwell Health is located in a highly urban area with a relatively larger population of Hispanic pwCF and therefore was asked to identify potential Hispanic participants; Children's Healthcare of Atlanta was asked to identify potential participants in the younger age categories, as well as parents of children and adolescents).

Adolescents and adults with CF, parents of a child or adult with CF, and partners of an adult with CF were recruited. We purposively sampled participants to obtain a rich diversity of experiences and identified pwCF who varied in terms of age, disease severity, race, and ethnicity. Our approach was designed to minimize homogeneity among a small number of participants (21). Selection of participants was guided by the literature (17,22). We considered possible sources of variation in patient and family experience related to differences in clinical characteristics and outcomes, adaptation to CF and illness burden, and disparities associated with race/ethnicity, gender, timely diagnosis, and access to newly-available disease-modifying therapies, as well as underrepresentation in research (17,22–25). Our sampling elements included: (1) patient age, (2) patient disease and treatment characteristics (specifically, use of CFTR modulators such as elexacaftor/tezacaftor/ivacaftor, advanced CF lung disease status, receipt of organ transplantation); (3) participant sex, and (4) race/ethnicity.

Our sampling approach aimed to identify and interview as many as 16 subjects, a number suggested in prior studies to be associated with data saturation (26). We used one of our study objectives: assessing the extent to which the NCP palliative care domains were applicable to a broad range of CF-specific experiences, as a standard to judge whether our sample size was adequate. The standard was achieved in the study.

Recruiting and Interviewing

Local Institutional Review Board (IRB) approval or exemption was obtained at each study site. Candidates were approached by the site PIs, and if interested, consented and enrolled. Sites scheduled a 1-hour, remote interview, either by telephone or via Microsoft Teams' audio (with video disabled) for each participant. Interviews were conducted between August and October 2020 by the same qualitative researcher (M.B.) across all 5 sites. In accordance with site-specific IRB requirements, interviews were audio-recorded using either Microsoft Teams or a digital audio recorder, and demographic, medical, and treatment characteristics were collected at the time of enrollment or interview.

Analysis

Qualitative researchers from the study team (M.B. and J.P.) developed a codebook containing *a priori* (i.e., deductive) codes based on the 5 content areas of the interview guide (Supplemental Appendix). For each code, we included a definition, sample quote, and where applicable, notes containing inclusion and exclusion criteria and rules for allowing transcript text to be double-coded to multiple codes. The initial codebook was reviewed by the research team (A.G., L.D., S.D., and R.P.). Several themes were expanded to include positive and negative statements relevant to each domain, and guidelines for double coding passages of text were further refined. Two researchers co-coded 2 transcripts while discussing and making modifications to the codebook to ensure further clarity.

After finalizing the codebook and database, the 2 coders independently coded the same 5 transcripts, which yielded a kappa of 0.81 with 97% agreement. Thereafter, the remainder of the transcripts were coded independently by both coders using NVivo. The final kappa score for all 13 transcripts was 0.80 with 98% agreement (27). Participants' demographics, CF disease and treatment characteristics, and quantified responses to select open-ended questions (e.g., average number of NCP palliative care domains identified per respondent, preferences for palliative care screening) were summarized with frequencies using NVivo software analytics and Microsoft Excel.

Results

Sites identified 16 individuals for participation in the study, 16 were screened for eligibility, one declined participation, 15 provided consent, one withdrew from the study, and 14 were interviewed. The final sample included 7 adults with CF, 2 adolescents with CF, and 4 parents and 1 partner of pwCF; the sample demographics are presented in Table 1. One interview was unusable due to technical problems with the recording.

The study interview guide covered 5 content areas (Supplemental Appendix) and information obtained from participants was summarized for each of these categories, including: (a) Knowledge about palliative care, goals discussions, and advance care planning; (b) Problems and sources of burden (i.e., concerns or conditions that might be addressed by a screening program aimed at improving quality of life and illness burden [this was further subcategorized *a priori* into the 8 NCP domains for quality palliative care]); (c) Initial process of screening; (d) Triage and follow-up care, and (e) Scenarios for gaining feedback on 2 different screening models: clinic-based versus home-based.

As noted, an aim of our study was to analyze "(b) Problems and sources of burden" and these were grouped into 8 defined subcategories based on the NCP domains: (1) *structure and process* (a domain which explores approaches and essential processes when delivering palliative care at the systems level), (2) *physical aspects of care*, (3) *psychological and psychiatric aspects of care*, (4) *social aspects of care*, (5) *spiritual, religious, and existential aspects of care*, (6) *cultural aspects of care*, (7) *end-of-life care*, and (8) *ethical and legal aspects of palliative care, including goals of care*.

Qualitative Analysis

Knowledge About Palliative Care, Goals Discussions, and Advance Care Planning. The 8 participants who reported familiarity with palliative care either equated it with end-of-life care or referenced experiences with older family members without CF, rather than their own personal experiences. For example, one female parent participant stated: "…Actually, [we] just recently [heard about palliative care] because we lost my mother-in-law…she was just about to go into palliative care and then she passed away." Those unfamiliar with palliative care tended to be younger or healthier pwCF or the parents of younger children who had not been engaged in palliative care discussions. When discussing concerns, values, and preferences, we again saw an indication that these discussions were not being introduced early in the disease trajectory; there was a difference between those who were younger and healthier, and those with more advanced disease. When discussing goals of care and advance care planning, most participants focused on short-term priorities relevant to their stage of illness, such as "doing what was needed" to stay healthy and maintaining daily treatment regimens. An adolescent male stated: "My goals are to always take my medication for a better life, a better future." Few participants (N = 5) were familiar with advance care planning and few had discussed end-of-life care preferences. However, one male with advanced CF, who had undergone lung transplant described a conversation he had with his provider: "I wouldn't want to go on life support again, at least not in my current condition. So, we've had that conversation as far as treatment preferences [when prognosis is short]."

Problems and Sources of Burden. Participants discussed an average of 3 symptoms (range 2-5) considered "most important" to the quality of life of pwCF and their family caregivers. Table 2 describes specific concerns aligned with the NCP domains. For example, all participants described physical symptoms related to CF, most frequently pain, fatigue, shortness of breath, and gastrointestinal issues, which they may not disclose to their providers or discuss with others. As one adult Hispanic female with CF stated:

> When I usually get pain and stuff like that, I don't really try to reach out to anyone, just because I've been dealing with it for

Table 2. Symptoms and Burdens Identified by Participants Categorized by National Consensus Project's (NCP) Domains.

NCP domains	Response frequency (N=13)	Specific symptoms mentioned	Sample quotes
Structure and process	1	n/a	"I think part of it is kind of helping people understand the impact of what it means to have the disease [CF] and how it can impact your life and how it can impact your loved one's life…trying to see the big picture…Not freaking out about it, but seeing the big picture so you can try to break it down in smaller pieces and attack each piece as…a smaller task to get to your ultimate goal, so you're not overwhelmed." **-Adult White Male, Partner of Person with Advanced CF Lung Disease**
Physical symptoms	13	Pain, gastrointestinal symptoms, cough, sleep disturbance, dyspnea, fatigue, sinus problems, side effects of elexacaftor/ tezacaftor/ivacaftor and other medications, sudden fever, and infections	"I think one is pain and there is definitely shortness of breath and just in terms of symptoms, specific to CF I would say not having an appetite to eat food." **-Adult Asian Female with CF** "I feel it would be important to screen for the digestive issues, based on what we've gone through and what the boys have gone through, the majority of their issues, at this point, have been GI involved and a lot of times the GI issues at this age don't get as much attention as it probably needs." **-Adult White Male, Parent of Children with CF** "One thing that impacts my life the most is like just pretty much feeling tired, fatigue and everything. That's actually the main thing, just feeling tired a whole lot." **-Adult White Male with CF**
Psychological and psychiatric	8	Anxiety, stress, stigma, family's mental health, trauma, fear of mortality	"I mean, I can't always speak for my son, but I know that he does worry about his mortality even though he says he doesn't. And deep conversations, it does come up. But we haven't had those really deep kinds of discussions about the future." **-Adult White Female, Parent of Adult with CF** I think [it's] relevant to ask - how you've been feeling about yourself within the last 2 weeks, have you ever had certain thoughts like are you doing well? Have you been worried about yourself or others? Yeah, I believe that's helpful." **-Adolescent Black Male with CF**
Social aspects of care	13	Relationship with siblings, family planning, romantic relationships, social support, family's needs, money, schoolwork, treatment costs, career planning, life transitions	"Our stress right now is college.. She's a senior, so we're a little nervous about the whole thing, about her going away and stuff like that. It would be nice for me as a parent to have some kind of counseling through this time of our lives." **-Adult White Female, Parent of Adult with CF** "My top would be anxiety/depression, mental health and awareness, being that CF can

(continued)

Table 2. (continued)

NCP domains	Response frequency (N=13)	Specific symptoms mentioned	Sample quotes
			become a very lonely illness in that we're not allowed to sit with other people with CF." **-Adult White Female with CF**
Spiritual, religious, and existential	3	Religion, spirituality	"I do [value spiritual aspects of care]. My family, we're very religious." **-Adult White Female with CF**
Cultural aspects of care	2	CF not well known in country of origin, families play a strong support role in certain cultures	"First of all, where I come from [in Africa], I never heard of CF in my life. It was something completely new to me. So, when I needed to go do the sweat test for my baby, you know, I don't know what it is." **-Adult Black Female, Parent of Child with CF** "In my [South Asian] tradition, they [doctors] know what's going on, they know a lot of people support me, they know that I have support." **-Adult Asian Female with CF**
Care of the patient nearing the end of life	0	n/a	n/a
Ethical and legal aspects of care (including end of life)	2	Advance care planning, end-of-life discussions	Interviewer: Do you think it's a good idea for them to ask you about … end of life care, you know [things] that some people might find sensitive? Participant Response: "I've spoken about end-of-life stuff." **-Adult Hispanic Male with CF**

> a very long time, but maybe something that they [providers] could do is like maybe also reach out to see if we're okay, if we're stable at the moment.

The majority of participants (N = 8) also discussed emotional distress; anxiety and depression were most frequently reported, and family members discussed caregiver distress and burden.

All participants discussed the importance of addressing social factors, and these varied widely by participant demographics. Life-stage transitions and education milestones were commonly highlighted as important by adolescents and parents of younger children. Family caregivers cited financial planning, family well-being, and caregiver mental health as important contributors to wellbeing. One parent of 2 young children with CF stated: "[Providers should] be mindful of treatment burdens, not only on the patient." Another parent described the emotional difficulties she experienced in the early years of caring for a baby with CF: "I just used to cry. You know, wake up in the morning, take care of her, I needed support."

Adults with CF discussed career planning, intimate relationships, and family-building concerns as important influences on well-being, while younger participants discussed school and transitioning to college. While less salient, perspectives regarding spirituality revealed that some participants valued the positive impact on well-being. Three participants specifically discussed their need for spiritual support, but most participants acknowledged that while they themselves may not require support in this area of their lives, others might find it helpful. The relevance of cultural aspects of palliative care were discussed by 2 participants who were not born and raised in the United States, indicating that values, customs, and belief systems may be an important factor among underrepresented pwCF requiring future exploration. One participant had never heard about CF in her country of origin and the other explained that her care team recognized that strong social support was common in her "tradition."

Although concerns relevant to the NCP domain, *competent care at the end of life*, were raised, they focused on advance care planning and we chose to code the content as reflecting the *ethical/legal* domain because none of our subjects had end-stage disease. One of the 2 participants who discussed the need for advance care planning was a lung transplant recipient who stated that he found these conversations about end-of-life care helpful, and expressed the desire to have discussed these topics much earlier in his care:

> I've always been very appreciative when my doctors or my team talked to me about end-of-life things because, for instance, something that I found out recently…I always thought that at the end, I would end up kind of just like suffocating to death almost. And for whatever reason, it never crossed my mind—even though I heard of palliative care, but I didn't really think—and I talked to my docs, and they basically explained to me that that's not how it works and that'll never happen to me. And that took a huge load off my shoulders. So that was really important to know that, and I wish I knew that maybe 20 years ago.

We asked participants about the topics that might be included in a screening program—such as concerns about disease progression, romantic relationships, spiritual needs, or practical problems (i.e., financial, school, or work-related)—or if there are other concerns that are too personal or sensitive to discuss. Most participants stated that they were open to being asked about anything, qualifying this with statements such as "a provider can ask about something, but a person could then choose not to discuss an uncomfortable topic." However, there were 2 participants who felt that end-of-life care was too sensitive to discuss openly. For example, one adult female with CF stated:

> Talking about issues like end [of-life] care and those kinds of things, a year ago the doctor did bring it up with me several times, and even the social worker talked with me, but I was just the kind of person who could not bring myself to talk about it. I didn't come to terms with it.

Overall, participants expressed an openness toward discussing most topics, and were willing to engage in conversations about sensitive issues, as long as providers first asked for permission about whether it was acceptable to discuss this.

Initial Process of Screening. While specific preferences for screening methods to address these topics varied, in general, participants supported the idea of regular screening in CF centers to identify palliative care needs, emphasizing that oftentimes, pwCF may not spontaneously report a problem or desire for intervention, as stated by one parent:

> You know, unless it's actively bothering them [his children with CF], it would probably not be something we would necessarily bring up, but screening for those sorts of [palliative care] issues, I think, would allow our care team and the social workers to decide how we need to treat those emotional issues, like see a psychologist and [access] self-care…

Ten participants specifically felt that a screening questionnaire would be helpful to initiate the process, stating, for example, that it would allow people to "address problems without being confrontational" or as one participant stated: "Maybe some people are more afraid to bring something up in a conversation, so maybe they're more apt to circle it on a piece of paper." Another participant perceived that a screening questionnaire could address a broad range of topics that would otherwise be overlooked:

> I think that [screening] could help…me personally. If I saw something like that…on the list, maybe it's something I haven't realized…[is] bothering me. And then I see it right there in front of me, and I'm like, 'You know what? …that may be [something that] subconsciously has been bothering me'.

However, 2 participants did not believe that using a screening questionnaire was a good idea. One perceived it as impersonal and expressed the belief that participants may not be forthcoming or elaborate in their responses (e.g., "I don't think people are always honest on questionnaires") and the other perceived it as prying (e.g., "I've been in situations, and there are things [that I] don't want to talk about, [that are not] very comfortable and for me, I'm an extremely introverted person"). The process of regular screening and follow-up interventions was acceptable to most participants. When asked who should initiate screening, most stated that screening should be offered by either a social worker or a physician, with a limited number of participants stating that it should be the provider with whom they had the closest relationship. Some participants felt strongly that they would only want to discuss sensitive topics with specific members of the care team. For example, one adult male with CF stated: "I wouldn't want my medical doctors, like my lung doctors, asking me about God and stuff."

Triage and Follow-up Care. Participants expressed that, once a problem was identified, follow-up by a CF care team member was key. Most participants perceived that referral to specialty care was appropriate for problems that the CF care team could not address, but that follow-up and monitoring by the CF team was critical: "I guess [the CF team could] provide their expert opinion, and let the patient know that this really isn't their field of expertise, give them a good resource, you know where they can get help for that, but still…keep up-to-date and be involved the best they can". Two parents expressed the need for the primary CF team to remain involved in care delivery and coordination, as one mother of an adult with CF stated, so that care needs didn't "fall through the cracks." One salient preference was for the care team to remain involved and coordinate their care, to continue to offer resources and intervene on their behalf with specialists. One parent suggested that once an initial screening occurs, the care team should be prepared to offer resources and referral to specialists before the visit:

> [If it's] emotional stress, depression and anxiety [that's detected], [the provider could say],'hey, have you reached out to these [resources]…here's your…list of mental health providers in your area that are covered under your insurance'.… Just having that little bit of information ahead of time, before you address it with the patient or family [would be helpful].

Table 3. Perceived Barriers and Facilitators of Home-Based and Clinic-Based Screening.

	Clinic-based screening	Home-based screening
Facilitators	• Immediate action/planning • Interpersonal/face-to-face • "Down time" during the clinic visits • "You can't push it off" • Clinic time is "CF Time"	• More time to respond/think about responses • Telehealth has become integrated into daily life • "Better for introverts" • Overcomes time constraints in clinic visits • Avoids travel to clinic • More efficient • Fewer scheduling conflicts
Barriers	• Lengthens the clinic visit • Overwhelming/anxiety levels "already high" during clinic visits • Clinic visits are "too rushed"	• People will forget to complete screening questionnaires or procrastinate • Problems will take longer to be addressed • Lacks personal connection • Too busy at home—"things get pushed aside/not a priority"

There was variation regarding how this should be done and by whom. Most participants felt that the CF physician (N = 6) or social worker (N = 3) should be responsible for triage and follow-up interventions to address clinical problems. Several participants (N = 3) stated that follow-up depended on the specific problem that was identified in the initial screen (i.e., emotional problems should be addressed by a social worker, and physical problems should be managed by a physician: "If it's emotional [in nature], maybe the social worker, but if it's actual physical pain, then they should let the doctor know"). Several participants mentioned other members of the CF team (e.g., nurses or nurse practitioners) as the preferred person to oversee follow-up and triage. Overall, while preferences for who should conduct screening varied, consistency and accountability with follow-up were perceived as important (i.e., the provider who initially reaches out should be responsible for follow-up).

Scenarios for Feedback on 2 Different Screening Models: Clinic-Based Versus Home-Based. To assess preferences for clinic versus home-based screening models, we described both types of scenarios to participants, including different processes for initiating the screening and follow-up care. Most participants preferred clinic-based (N = 8) versus home-based screening (N = 3) because it was perceived to be more personal (i.e., enabled face-to-face interaction with providers), allowed for dedicated time for this task, and facilitated immediate management of problems (Table 3). As one parent explained:

> I know myself, my schedule at work, there's a lot of school paperwork and things that I have to fill out and I forget, and it's just so crazy—so when I go to the CF center, it's like my priority is that visit, speaking with the doctor, speaking with the care team, it's all about…my daughter, and her care, and just that time.

However, some preferred home-based screening because it was perceived to be more focused and would allow more time to think about their responses, with the increasing acceptability of telehealth also noted. Others perceived several barriers to screening during clinic visits: anxiety associated with the clinic visit, the extra time required for screening, and lengthening the duration of an already long visit. For example, an adult male with CF explained:

> When I'm going into the hospital [CF clinic], I want to see the doctor. I want to hear about my status, and I want to get…[-right] out of there. [I want to] find out how I'm doing and what are the next steps, if any…and I want to go home. I don't want to be sitting around the hospital. So, to be able to sit at home and do it, I'm more comfortable. I will take my time. I'll be able to answer more thoughtfully.

Perceived barriers to home-based screening included lack of time, concerns about forgetting to complete the questionnaires, and the lack of comfort and personal connection when discussing problems remotely. One participant compared the 2 options as follows:

> You don't have the ability to forget [about screening in the CF clinic] or just get busy with other things. …It will allow the doctor or whoever to go over [it] with you in-person, so you get that 'face-to-face'; you get that interpersonal experience that you don't always get via phone call or email.

Several participants reported that if they were to complete the questionnaires at home prior to an upcoming visit, they would then prefer to discuss the responses face-to-face at the clinic, which represents a hybrid of the 2 proposed models.

Discussion and Conclusion

This qualitative study assessed perceptions about palliative care and obtained feedback about the new primary palliative care screening-and-triage model planned for testing in the *Improving Life with CF* study. Many pwCF and family members we interviewed were unfamiliar with palliative care, goals discussions, and advance care planning. Those who were familiar with these concepts often equated them with end-of-life issues. Our study builds upon prior work which seeks to address misconceptions equating palliative care with end-of-life and hospice care (28) by introducing palliative care earlier, prior to the onset of advanced disease. Knowledge deficits about palliative care and advance care planning could be addressed by age-appropriate education throughout the lifespan, for example, as part of a transition program that prepares adolescents with CF for adult care.

PwCF and their family caregivers most frequently endorsed physical, psychological/psychiatric, and social aspects of care as important for improving quality of life, while other domains such as cultural, spiritual, and ethical/legal were perceived as highly relevant by a subset of participants. Our findings are consistent with the evidence-based NCP guidelines which established 8 domains for assessment and the provision of quality palliative care, and suggest that these domains are relevant for pwCF and their families (17,29–31). Nonetheless, participants' specific concerns and care preferences varied, and they perceived that the fluctuating nature of the disease requires management of a wide range of symptoms and problems throughout the illness trajectory (19), reinforcing the need for a CF-specific approach to palliative care (6). Our study highlights the potential for unmet needs to be continuously reassessed through regular screening, adding to preliminary work indicating the acceptability and potential effectiveness of an early intervention approach to primary palliative care in pwCF. Such a strategy would allow services offered in primary CF care settings to be tailored to changes over the life course, such as changes in clinical status, quality of life, disease progression, function, the onset of new comorbidities, and age-related decline.

Most participants were comfortable with CF team members broaching sensitive topics, and several suggested that the optimal approach would be to screen broadly while allowing pwCF and caregivers to accept or decline to speak about those topics that make them uncomfortable. This underscores the need to understand communication preferences in the CF community. To this end, some of the *Improving Life with CF* study sites have chosen to simultaneously participate in the CFF Partnership Enhancement Program (32,33).

Use of a screening questionnaire was generally acceptable and most participants preferred implementation by a CF clinician with whom the person with CF feels comfortable. Once a management plan is initiated, oversight by the CF care team was uniformly perceived as crucial. Even after referral to specialty care, follow-up is important to ensure the plan is working, and that important concerns are not overlooked. Flexibility with the screening approach was also considered to be important; while most people preferred clinic-based screening, others perceived that it was easier to complete questionnaires at home. Some participants suggested a hybrid approach—that is, completing the questionnaires at home and then discussing the results in clinic. This approach was incorporated into the *Improving Life with CF* intervention by some of our participating sites.

The primary palliative care screening-and-triage intervention developed through the *Improving Life with CF* project includes both foundational elements concordant with the CFF Models of Palliative Care Delivery guidelines (18) (e.g., routine screening for unmet needs in multiple domains using the Integrated Palliative Care Outcome Scale (34) and Brief Assessment Scale for Caregivers (35,36)) and modifiable, site-specific elements. The needs assessment is therefore built on the strengths and resources of each CF center and tailored to patient and caregiver preferences. Our project also includes resources to support the self-management of common symptoms. The salience of these symptoms was confirmed through the current work. Resources for clinicians (see https://improvinglifewithcf.org/) also are available and aim to improve psychosocial, integrative, and medical interventions. We are currently testing these components, and others, through a pragmatic trial of our new primary palliative care model in 5 CF centers managing nearly 1000 pwCF.

Limitations and Strengths

There are limitations of this study, including the small sample size, and the lack of information on participants' socioeconomic status, educational level, and insurance coverage, all of which could potentially reduce the generalizability of our findings to other groups of pwCF and their family caregivers. We sought to mitigate these limitations by using a purposive sample to obtain a broad range of views. We believe that our purposive sample's focus on disease characteristics, age, race/ethnicity, and status as a pwCF or family caregiver has enabled us to identify a comprehensive and rich range of experiences and palliative care needs. We also hoped that the sample would offer feedback about all the key domains identified as significant by the NCP guidelines (Table 2) (19), and this was confirmed in our analysis. The latter was the standard we applied to determine whether our sample size was adequate. A final limitation, which we could not address through sampling and the study design, is that the introduction of highly effective CFTR modulators at a younger age might decrease the burden of disease or shift it to older age, making some of the study findings less applicable for the future. However, even with improvements in disease activity, palliative care needs in CF will likely remain significant. Strengths of our study include the multisite design; an interdisciplinary study team; and application of a rigorous analytic approach designed to minimize individual-level coder subjectivity.

Conclusions

While pwCF and family caregivers participating in our study were largely unfamiliar with concepts related to palliative care, most perceived that regular screening and access to generalist-level palliative care offered by CF professionals was important to address symptoms and concerns related to living with a serious chronic illness. Most participants expressed that screening via a questionnaire was acceptable, although opinions about the optimal process varied. Flexibility is key, with needs assessment tailored to individual preferences and close attention to follow-up by CF clinicians. There are 3 main practice implications from this work: (1) Palliative care should be introduced early and be clearly defined, (2) the nature of CF makes regular screening important as symptoms, needs, and preferences of those living with CF and their families may change over time, and (3) CF clinicians should address the need for palliative care education and interventions in a manner acceptable to pwCF and family members, with ongoing involvement and coordination by the care team when specialty services are needed.

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Ethical Approval

Ethical approval to report this case was obtained from Partners Human Research (2020P001075), Mount Sinai School of Medicine (20-00845), Feinstein Institutes for Medical Research Northwell Health (20-0439), Stony Brook University (IRB2020-00294), and Emory University (0000054959).

Statement of Human and Animal Rights

All procedures in this study were conducted in accordance with the Partners Human Research's (2020P001075), Mount Sinai School of Medicine's (20-00845), Feinstein Institutes for Medical Research Northwell Health's (20-0439), Stony Brook University's (IRB2020-00294), and/or Emory University's (0000054959) approved protocols.

Statement of Informed Consent

Verbal informed consent was obtained from the participants for their anonymized information to be published in this article.

Data Sharing Statement

The dataset for this study, including the study protocol, statistical analysis plan, and individual de-identified participant data underlying the results reported in publication (text and tables), will be made accessible 4 months from initial request to investigators who provide a methodologically sound proposal, as determined by the study's lead investigators. The dataset will be provided after its de-identification, in compliance with relevant regulatory and privacy laws, data safeguards, and requirements for patient consent and anonymization.

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Supplemental Material

Supplemental material for this article is available online.

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