



Research Paper

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ABSTRACT

Background: Cystic Fibrosis Related Diabetes (CFRD) is an increasingly common complication in CF. CFRD introduces an additional complex chronic disease to individuals already attending to demanding treatment regimens. An improved understanding of the reaction to and coping mechanisms surrounding CFRD may facilitate management of CFRD.

Methods: Semi-structured interviews completed by 10 children with CFRD, 10 adults with CFRD and 10 parents of children with CFRD in a single large CF Care Center were analyzed utilizing phenomenological analysis.

Results: Patients and families reported having limited knowledge of CFRD prior to diagnosis. CFRD was considered an extension of their underlying CF and successful management depended upon integration into existing routines.

Conclusions: Health care professionals caring for patients with CFRD should be informed of the specific challenges that arise in CFRD. Education in advance of CFRD diagnosis may facilitate management.

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Introduction

Cystic Fibrosis Related Diabetes (CFRD) is a common comorbidity affecting people with cystic fibrosis (CF). CFRD becomes increasingly prevalent with increasing age with an estimated prevalence of 50% in CF patients aged 30 years and greater [1–3]. As management of CF continues to advance, the proportion of patients with CF related comorbidities such as CFRD is expected to increase. Untreated CFRD is associated with worse pulmonary function, increased pulmonary exacerbation frequency, and decreased survival, while aggressive treatment of CFRD is associated with improved outcomes [4,5].

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The diagnosis of CFRD in people with CF represents the introduction of an additional chronic disease and significant additional medical burden: blood glucose monitoring, subcutaneous insulin injections and risk of hypoglycemia, attention to diet and nutrition, and additional medical appointments. The literature describing reactions and the perceived barriers to management of CFRD is limited. In previous reports, adolescents and adults recollected feelings of shock, anxiety, and relief upon the diagnosis of CFRD as well as maladaptive behaviors surrounding management requirements [6]. Some viewed their diabetes as an extension of CF, others saw CFRD as a nuisance that could be ignored when the primary challenges of CF escalated, while still others felt that having lived with CF made them better equipped to address diabetes management [7]. These data also revealed that most CF patients were unfamiliar with the possibility of developing diabetes and that many lacked the knowledge and confidence to approach this new chronic illness [6–8].

Existing reports include few participants diagnosed with CFRD as children [6], and to our knowledge there have been no studies of how children react to CFRD management. Available data on

parental reactions to their child's diagnosis with CFRD are mainly focused on adolescent autonomy [8]. We know very little about the ways in which patients and parents manage CFRD and factors that make adherence more or less difficult. Data related to such issues will help providers identify targets of intervention to improve adjustment and adherence, at diagnosis and over time. As such, the aim of the present study was to examine the reactions of children and adolescents, their parents, and adults with CF to CFRD management and factors that facilitate or impede treatment adherence.

Methods

Design

A cross-sectional, mixed-methods, descriptive study design was used. Single occasion qualitative interviews were utilized to explore participants' reactions to the diagnosis of CFRD and daily experience with the disease. The Institutional Review Board of the Children's Hospital of Philadelphia approved the study, and all subjects provided informed consent and assent as appropriate.

Participants

A convenience sample of children and adolescents with CFRD and their parents as well as adults were recruited from the Cystic Fibrosis Center and the Endocrinology clinic of the Children's Hospital of Philadelphia and the Adult CF Program at the Hospital of the University of Pennsylvania, which are tertiary care hospitals in the Northeastern United States. Adult, adolescent, and child patient participants were included if they had a diagnosis of CFRD according to accepted guidelines [9], were older than age 8 years, and spoke English. The peak age of onset of CFRD is typically in late adolescence to early adulthood and is rare in patients younger than 8 years [3,10]. Thus, including only patients greater than 8 years improved the quality of the interviews without compromising eligible participant inclusion. A child with CFRD and his/her parent(s) could both participate. Participants were excluded for mental or developmental disorders, health status, or functional impairment that would prevent completion of interviews. However, parents of such children were eligible. Two potential subjects declined to participate: one adult with CFRD and one parent completed the initial consent process and questionnaires, but did not complete the interview and were not included in the study sample. We used the standard of thematic saturation to determine the final sample size. In other words, we stopped interviewing when we determined that no new themes were emerging from the data [11].

Data collection

Semi-structured individual interviews were conducted with patients and parents by one interviewer (KM). Interview topics included the subject's initial reaction to being diagnosed or to their child's diagnosis with CFRD, their overall attitude toward CF and CFRD management, the impact of CFRD on daily life, and the methods used to cope with the daily care of CF and CFRD (see [Online Supplement](#)). Interviews were digitally recorded and transcribed verbatim. For descriptive purposes, the Cystic Fibrosis Questionnaire-Revised (CFQ-R) was used to measure each subject's health related quality of life. The CFQ-R yields a standardized scale ranging from 0 to 100, with higher scores reflecting better quality of life [12,13]. For participants with CF and the children of parent participants, clinical variables including FEV₁, BMI, and HbA1c, were

collected from the electronic medical record to characterize CF disease severity and CFRD control.

Analysis

Descriptive data were presented as mean with standard deviation and minimum and maximum if continuous or as percentages for categorical data. Visual inspection of scatterplots examining the relationship of HbA1C with CFQ-R subscales was undertaken, and pairwise correlations were performed. Two members of the study team (K.M. and V.M.) independently read two transcripts and generated ideas for an initial coding structure, and then discussed these ideas to finalize the coding structure [14]. Interview transcripts were then coded by the first author for major themes and sub-themes. Initially coding labels were applied to units of meaning within the transcribed text. Participants' comments and perceptions were sorted, aggregated, and synthesized [15]. Adult and child participant responses were analyzed together as these groups were sharing first person reactions. Parent responses were analyzed separately. Quotes were selected to illustrate key themes from the analysis ([Table 1](#)).

Results

Participants

Ten children and adolescents, ten adults and ten parents participated in the study. Nine of the child participants and one of the adult participants had a parent who participated in the study. Two of the adult participants had undergone lung transplantation. One parent whose child had both CFRD and severe autism participated, but the child was unable to participate due to developmental issues. Semi-structured individual interviews were conducted with participants either in conjunction with a previously scheduled clinic visit ($n = 19$), during an inpatient stay ($n = 7$), or over the phone ($n = 4$). One child participant and three adult participants utilized an insulin pump. Demographic data, clinical characteristics, and CFQ-R subscale results for the sample are portrayed in [Table 2](#). No relationships between CFQ-R subscales and current HbA1C were identified ([Figure 1](#)).

Prior knowledge of diabetes

Expectations and preparation prior to the diagnosis of CFRD varied widely. Many patients and parents (4/10 children, 5/10 adults, and 3/10 parents) reported having prior experience with diabetes from friends, family members, or acquaintances they knew with Type 1 or Type 2 diabetes. Knowledge about diabetes varied from knowing only that diabetes had to do with blood sugar to an understanding of complications including amputations, organ damage, neuropathy, and blindness. Five patients anticipated that with diabetes they would have to change their diet. Assumptions that one got diabetes from "not taking care of themselves" were brought up by 2 adults and 1 child.

Half of the participants were aware that diabetes as a result of CF was possible. Three parents had heard about CFRD from attending 'CF Education Nights.' Two parents knew other CF patients who had developed CFRD. Many parents recalled hearing about CFRD when their children were first diagnosed with CF.

Annual screening for CFRD helped some (3/10 children and 3/10 parents) 'warm up' to the possibility of being diagnosed with CFRD and to accept the diagnosis when it did come. One adult patient who was diagnosed prior to the initiation of CFRD screening wished that more screening had been done prior to his diagnosis.

Table 1
Illustrative quotes

Prior knowledge of diabetes	"I'll never forget going to the meeting [CF Education Night] and talking about this CF related diabetes and we're like, 'Oh no, what is that now?' It was scary to hear about that just going through the years. I wasn't happy to hear about that. I thought we would never have to worry about it." (P11) ^a "You know about Type I and Type II and you look at some the people that have it and you're like, 'Whoa, that's not going to be me.'" (A3)
Reactions to CFRD management	"At first I thought it was going to, like, be really really hard and I wouldn't know what to do at first, but then as time went by I got the feel for it and its becoming a part of my every day routine and just something I do every day now." (C5) "Because you already had a new normal. You got your new normal when he was diagnosed. So anything that came with that is all a part of the new normal because it's all related to that one thing." (P1) "I have more to do, like, more treatments, nebulizers, and enzymes. I have to worry about all that. All I have to do with my diabetes is give myself insulin and check my sugar and that's all" (C4) "I saw it as something that would hamper my lifestyle. Something that would, you know, interfere ... for the most part until then I just had to worry about taking some enzymes maybe an antibiotic. During the day it didn't really impact me ... other than a few little things here or there I was living 'a normal life.' So, that was kind of a major hurdle major for me." (A5)
Barriers to CFRD treatment adherence	"Before I go to bed I'm just super tired and I just want to go to bed and I don't want to check my sugar and I forget about it. In the morning I'm super busy, especially when I get up for school, and I have a lot of things to do and I'll be running out the door and I'll forget my sugar." (C4) "I think, like, if I get really stressed and overwhelmed with everything, like, with CF and school and everything I'll kind of not do it. Because I'm just, like, overwhelmed and I don't want to do it." (C7) "[Prandin] didn't really do anything, anything that I could see. So you couldn't really see any change then it didn't do shit was the way I saw it." (A8) "Remembering to test blood sugar can often be a task that's not easy to accomplish ... but that's not so huge in the actual care of the thing itself. Especially if you know that you have a good insulin regimen going and there's no real reason that you should be high at any point." (C6)
Factors promoting CFRD treatment adherence	"I got an ulcer on my arm ... and I was like, 'I'm not going to have these all over my body, absolutely not.' I'm very vain so I pride myself in my looks. I'm not going to walk around with ulcers on my body or lose my feet or my fingers. That's not going to happen to me because there's too much that I have planned for myself." (A7) "They really told me that you need to start really taking an aggressive stance on this. As soon as they told me it could affect lung function that's when I really started listening, 'Yeah, this needs to change.'" (A3) "A schedule that works, which is hard to develop. But if you can get a schedule that works it's much easier, because sticking to that schedule is a lot easier than sort of having to juggle it on a day to day basis assessing each situation as it comes." (C6)
Patient responsibility for CFRD management	"He decided that he was going to take [CFRD] on as one of his responsibilities. So that's ... he does it ... and I think he's more comfortable doing it. Sometimes I think he does it because he knows it bothers me to do it so he takes that on himself." (P1) "It was more or less that she didn't want me in her space. Coming up and sticking her here and doing that. You know what I mean? She's a young teenager going through, you know, changes. So, it's kind of like, "You're in my space and I don't ..." That's one of the reasons too that she has taken full control. I think, of doing it." (P3)
Role of the health care team	"Anytime I need anything, you know, a call or a text or an email away. They really help me, they support me that way." (A5)

^a A = adult participant, C = child participant, P = parent participant.

Reactions to CFRD management

The addition of "one more thing" to their existing CF routine and maintenance was the most frustrating aspect of the diagnosis (6/10 children, 6/10 adults, and 6/10 parents) due to the addition of medications, time, and the understanding of new potential complications.

Many patients and parents felt that having CF prepared them to face a diagnosis of CFRD. They were already acclimated to a routine of medications, treatments, clinic visits, and hospitalizations and were able to easily incorporate CFRD into the existing structure of CF (4/10 children, 4/10 adults, and 6/10 parents). In fact, the diagnosis of CFRD was sometimes downplayed because the addition of management of CFRD was small compared to the complex regimen for CF (2/10 children, 3/10 adults, and 2/10 parents).

Many patients (7/10 children and 4/10 adults) were upset because they didn't want to receive injections. For adults more than children (1/10 child and 7/10 adults), the diagnosis of diabetes was distressing because it was an interruption to the 'normal' routine they had developed with their CF.

Potential or realized changes in diet were distressing for patients and parents. The belief that one had to change one's diet when diagnosed with CFRD was split; more adults with CFRD believed they did have to watch their diet (1/10 children, 7/10 adults, and 3/10 parents), while a greater proportion of children with CFRD

believing that their CF diet took precedence (5/10 children, 2/10 adults, and 3/10 parents).

Factors impeding CFRD treatment adherence

Table 3 illustrates key themes related to CFRD treatment adherence. The most commonly cited reason for non-adherence to the treatment regimen for CFRD was forgetting either insulin injections or, more commonly, blood sugar measurements (9/10 children and 6/10 adults). Remembering was especially difficult when patients were hungry, not at home, or under other time demands (3/10 children and 4/10 adults). The second most common reason for non-adherence was avoidance of the pain from insulin injections or blood sugar testing (6/10 children and 3/10 adults). A recurring theme in patient interviews was the embarrassment patients faced when having to administer insulin injections in public (1/10 children and 4/10 adults). To avoid embarrassment patients did injections in the car or a restroom.

The interaction between CF and CFRD was problematic. Specifically, CFRD was difficult to care for when patients were suffering from a pulmonary exacerbation both because it was more difficult to attend to their care in general while ill, but also because medications (i.e. corticosteroids) utilized for CF made controlling their blood sugars challenging (3/10 children and 3/10 adults). The lack of

Table 2
Demographic and clinical characteristics of participants

	Child participants (N = 10)	Adult participants (N = 10)	Parent Participants ^d (N = 10)
Female, % (N)	30% (3)	30% (3)	80% (8)
White, % (N)	90% (9)	90% (9)	90% (9)
Age, years; mean (SD), min–max	14.6 (2.2) 10–17	29.5 (9.6) 19–48	47.6 (9.1) 37–64
Age of child, years; mean (SD), min–max	–	–	14.6 (2.1) 10–17
Married; % (N)	–	30% (3)	70% (7)
Household income; % (N)			
< \$20,000	–	40% (4)	10% (1)
\$20,001–\$60,000	–	10% (1)	20% (2)
\$60,001–\$100,000	–	10% (1)	40% (4)
>\$100,000	–	30% (3)	30% (3)
Age of patient ^b at CFRD diagnosis, years; mean (SD), min–max	12.5 (2.1) 10–15	19.9 (8.5) 9–35	12.7 (2.1) 10–15
Duration of patient's CFRD diagnosis, years; mean (SD), min–max	2.2 (1.5) 0–4	9.6 (4.3) 4–18	2.0 (1.6) 0–4
Patient BMI; mean (SD), min–max	20.4 (2.9) 16.0–25.8	23.5 (1.7) 20.6–25.5	20.1 (2.9) 16.0–25.8
Patient HbA1c; mean (SD), min–max	5.7 (0.6) 4.8–6.7	8.0 (1.7) 6.2–10.1	5.7 (0.6) 4.8–6.7
Patient FEV ₁ % of predicted value; mean (SD), min–max	83.4 (25.9) 38–113	67.1 (27.4) 43–130	83.4 (25.9) 38–113
Patient FVC % of predicted value; mean (SD), min–max	93.2 (20.3) 53–115	75.3 (24.5) 47–125	93.2 (20.3) 53–115
CFQ-R score ^c ; mean (SD)			
Scales ^d :			
Physical	75.7 (20.9)	74.1 (28.9)	76.2 (23)
Emotion	70.1 (21.3)	73.3 (12.9)	69.2 (12.4)
Body image	58.9 (30.6)	78.9 (21.9)	61.1 (21.5)
Eating	73.3 (32.8)	97.8 (4.7)	70 (37.5)
Treatment burden	54.4 (17.1)	58.3 (22.1)	41.7 (30.7)
Respiratory	60.2 (27.3)	72.9 (5.5)	71 (17.2)
Digestive	74.4 (30.1)	86.7 (20.2)	61.1 (17.2)

^a One parent participant had a child (not included as a child participant) with severe autism.

^b For parent participants, "patient" data refer to their children with CFRD.

^c Parent participants reported on their children's quality of life.

^d Combined scores for CFQ-child (age < 14y, n = 3) and CFQ-teen/adult (age ≥ 14y, n = 7) were combined for the "child" participants.

immediate effects from CFRD treatment was a hindrance for some patients (1/10 children and 2/10 adults).

Four patients rationalized the omission of blood sugar testing or insulin injections by saying that they snack all day so the blood reading wouldn't be accurate, that they didn't need to give insulin to lower a high blood sugar because they could 'work it off,' or that estimating the amount of insulin they needed without checking their blood sugar was 'good enough.' Two patients felt that as long as they were adherent with their insulin blood sugar monitoring was more 'flexible.'

For parents, barriers to their children completing CFRD maintenance tasks were due to time constraints, pain avoidance, and child resistance. Although not as time consuming as CF treatments, parents were frustrated by having to remind their children to check blood sugars and do insulin injections especially when trying to work around activities, school, and family events (3/10 parents). Those who were injecting their children with insulin found having to inflict pain on their child troublesome (5/10 parents). Child resistance to treatments, especially in the adolescent age group, was a challenge (3/10 parents).

Factors promoting CFRD treatment adherence

Incorporating CFRD into the daily routine was the most commonly cited strategy for adherence (6/10 children, 4/10 adults, and 1/10 parents).

Patients were strongly motivated to take care of their CFRD by either the occurrence of an adverse event attributable to their CFRD

or by the possibility of increased morbidity and mortality in the future (2/10 children and 3/10 adults). One patient noted that he was consistent with his insulin, while being non-adherent with other CF treatments, because since starting insulin he had been hospitalized less frequently. Another patient was motivated to take better control of his blood sugar in order to participate in a clinical trial for a CF medication. Two patients tried to be successful in managing their CFRD to avoid disappointing their parents or the health care team. Having parents remind them or prepare treatments was helpful for five patients. Improvement in lung function associated with better diabetes control was motivating for patients (1/10 children and 5/10 adults).

School nurse supervision of injections and blood glucose checks during the day was useful for parents to assure adherence. Parents also felt that being vigilant about organizing medications and supplies was critical to guaranteeing adherence.

Patient responsibility for CFRD management

Although many patients were still working on becoming independent with CF treatments they were able to manage diabetes on their own (9/10 children). One child who was completely dependent on her parents for both CF and CFRD care was also the youngest in our sample (10 years). All of the child participants reported that they were able to manage high and low blood glucose independently. CFRD care was seen as simpler than CF management and also required management when parents may not be present (e.g., managing hyper- or hypoglycemia). One parent felt that her child

HbA1C by CFQ-R Subscale

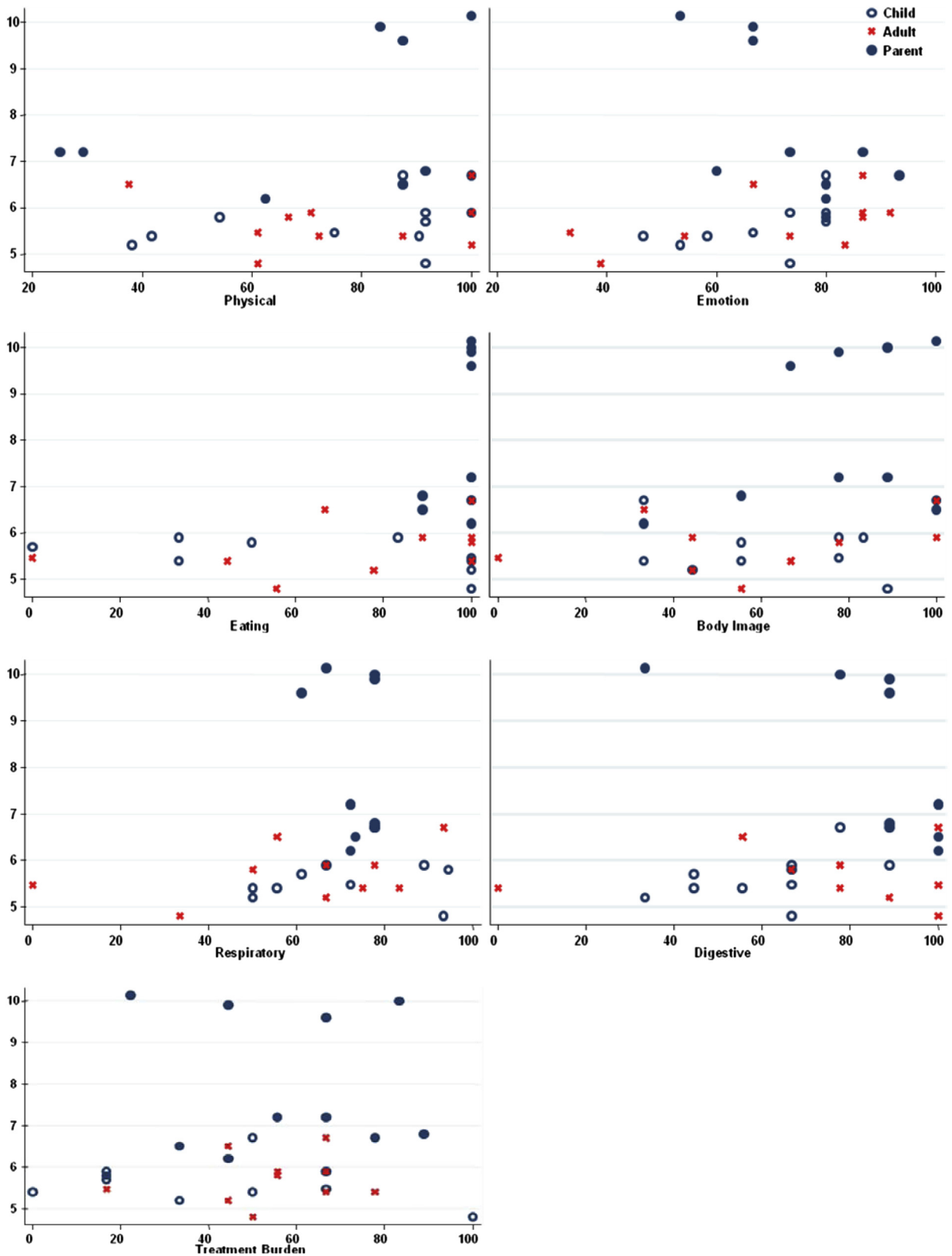


Figure 1. HbA1C and Quality of Life in CFRD. No relationships between HbA1C and various CFQ-R subscales were observed in children with CFRD (○), their parents (●), and adults with CFRD (×).

Table 3
Factors impeding and facilitating CFRD treatment adherence

Factors impeding CFRD treatment adherence	Factors promoting CFRD treatment adherence
1. Competing priorities (9/10 children, 6/10 adults)	1. Incorporating CFRD into existing routines (5/10 children, 3/10 adults, 6/10 parents)
2. Interference between CF and CFRD (3/10 children, 3/10 adults)	2. Fear of increased morbidity or mortality (2/10 children, 3/10 adults)
3. Embarrassment completing treatments in public (1/10 children, 4/10 adults)	3. Experiencing an adverse event (1/10 adults)
4. Avoidance Of pain (6/10 children, 3/10 adults)	4. Seeing positive effects from treatment (i.e. improved lung function) (1/10 children, 5/10 adults)
5. Not seeing effects of treatment (1/10 children, 2/10 adults)	5. Avoiding disappointment of parents or providers (1/10 children, 1/10 adults)
6. Minimizing importance of glucose testing (4/10 children, 4/10 adults)	6. Support or supervision of parents or other adults (5/10 children)
7. Child resistance (3/10 parents)	7. Organization of medications and supplies (1/10 parents)

wanted to take on CFRD management because having her parents give her insulin injections was an invasion of personal space.

Role of the health care team

Accessibility of the health care team was imperative for most patients. Patients valued that their health care team not only recognized how important it was for the CFRD regimen to fit into their lives, but also helped them come up with strategies to integrate it. It was important to find physicians who were knowledgeable about both CF and diabetes (3/10 adults and 1/10 parents).

Discussion

The diagnosis of CFRD represents the introduction of an additional chronic illness to patients and families already dealing with a demanding and complex treatment regimen. Patients are often diagnosed during adolescence and adulthood, after they have assimilated CF into their lives and established a care routine [3]. Adolescence is an especially tumultuous time for those with chronic disease, and patients may struggle with completing treatment recommendations in the face of social pressures and the desire for autonomy [16–18]. Little is known about how patients and their families cope with CFRD diagnosis and management and how they eventually become successful in dealing with two chronic diseases.

Many adolescent patients were able to care for their CFRD independently, and interestingly took on this responsibility before they were independent with their usual CF care. Unlike inhaled treatments and airway clearance that are typically done twice daily, insulin injections and blood sugar monitoring needs to be done several times throughout the day and often at times when parents may not be present. For adolescents going through physical changes and establishing autonomy, having their parents physically inject them was prohibitive to sharing CFRD responsibility. Many parents found that dealing with CFRD was a good introduction to the child taking on more responsibility for their health including CF. If adolescents can find success in managing CFRD they may be encouraged to take on more CF care as well.

Even for those patients who successfully integrated CFRD care into their routine, blood glucose testing was often minimized. Patients felt that blood glucose testing was optional once an insulin routine had been established. This attitude may contribute to a failure to respond appropriately to changes in glucose tolerance that arise with acute illness or changes in medications (e.g., corticosteroid use). Practitioners should be aware of this potential pitfall and focus education on the importance of continued blood glucose testing.

Dietary guidelines for CFRD are well established. However, concern was expressed regarding the dietary implications of CFRD among participants in our study [9]. This issue tended to be more

problematic for adults which may reflect education in the era prior to involvement of teams specialized in CFRD. Because of the strong emphasis placed on maintaining nutrition in patients with CF, often with a diet of high caloric intake that directly contradicts recommendations for those with Type I or Type II diabetes, practitioners should focus on dietary management and help patients and families to develop a nutrition plan that is consistent with current guidelines for CFRD. Implementation of consistent approaches to nutrition across the subspecialists caring for individuals with CF may alleviate this concern.

Our findings suggest that, for some patients, the diagnosis of CFRD signifies a major disruption to their ‘normal life.’ While CF care can often be confined to the home, diabetes care often must occur in public, which can be awkward for patients [19]. Suddenly having to deal with health maintenance tasks in public disrupts the ‘normal’ identity that patients had developed. Barriers to and facilitators of CFRD treatment adherence that we identified in this study are similar to what has been found in prior research related to adherence to the CF treatment regimen [20–22]. Not surprisingly, adhering to both the CF and CFRD regimen could be challenging; for example, completing CFRD tasks could be difficult during a pulmonary exacerbation, and some participants reported that certain CF medications interfered with blood glucose control. Given that regimen complexity and treatment burden have been associated with decreased adherence in prior research [21–23], the addition of CFRD management is a risk factor for compromised adherence to both regimens.

Consistent with prior work, only 50% of our participants were familiar with the possibility of acquiring CFRD prior to the diagnosis [6]. Additional challenges include misinformation or assumptions regarding diabetes, such as the belief that it resulted from not taking care of oneself and confusion about dietary changes. Annual CFRD screening was useful for patients to ‘warm them up’ to the possibility of CFRD [9]. This screening requirement should be an opportunity for education and preparation rather than simply another test that patients must complete. This preparation may decrease patient confusion and distress if they are eventually diagnosed with CFRD. The support of an accessible and knowledgeable health care team consisting of specialists in CFRD was perceived as instrumental for most study participants. As the issues that arise in CFRD are unique and distinct from those of Type I and Type II diabetes, many participants valued having access to health care practitioners who specifically understand the challenges and management of CFRD.

Limitations

This relatively small convenience sample may not represent the views of all patients and families with CFRD. All participants were seen at a large, urban, academic, tertiary care center. However, unlike quantitative research, in which the goal is to make statistical

inferences about the population from which a sample was drawn, the goal of qualitative research is to explore themes and assess the diversity of experiences that participants have with the phenomenon of interest [11,24]. Additionally, those who choose to participate in qualitative research may have different experiences than those who are not willing or available to participate. Some bias may have been introduced by having one author doing all the interviews, coding, and data analysis.

The lack of relationship between quality of life, including treatment burden, and HbA1C may arise for a variety of reasons. First, this study was not powered to examine this relationship. Additionally, an individual with CF may arrive at what is considered a very good HbA1C, particularly in the absence of fasting hyperglycemia, without significant effort; while individuals with more severe insulin deficiency will require more intense management. These issues are not easily represented by either duration of CFRD, total daily insulin dose (which will often reflect the large amount of calories consumed in CF), or even insulin pump use.

Conclusion

Although patients with CF and their families are experienced with chronic illness, the addition of a second illness, CFRD, may represent a significant disruption. Health care professionals caring for these patients should be informed of the specific challenges that arise with CFRD and educate patients at diagnosis on potential complications and recommendations. CFRD specific dietary recommendations should be highlighted, as diet continues to be a focus of confusion and distress amongst patients with CF and their families. Annual screening for CFRD should be seen as a chance for education and preparation for patients and families. Health care providers can best assist in the acceptance of the diagnosis of CFRD and its prescribed intervention by understanding that patients and families must incorporate CFRD into their existing routines.

References

- [1] Cystic Fibrosis Foundation Patient Registry. Annual data report 2011; 2011. Accessed 14.04.14.
- [2] Zirbes J, Milla CE. Cystic fibrosis related diabetes. *Paediatr Respir Rev* 2009 Sep;10(3):118–23. quiz 123.
- [3] Moran A, Dunitz J, Nathan B, Saeed A, Holme B, Thomas W. Cystic fibrosis-related diabetes: current trends in prevalence, incidence, and mortality. *Diabetes Care* 2009 Sep;32(9):1626–31.
- [4] Kelly A, Moran A. Update on cystic fibrosis-related diabetes. *J Cyst Fibros* 2013 Jul;12(4):318–31.
- [5] Chamnan P, Shine BS, Haworth CS, Bilton D, Adler AI. Diabetes as a determinant of mortality in cystic fibrosis. *Diabetes Care* 2010 Feb;33(2):311–6.
- [6] Collins S, Reynolds F. How do adults with cystic fibrosis cope following a diagnosis of diabetes? *J Adv Nurs* 2008 Dec;64(5):478–87.
- [7] Tierney S, Deaton C, Webb K, Jones A, Dodd M, McKenna D, et al. Isolation, motivation and balance: living with type 1 or cystic fibrosis-related diabetes. *J Clin Nurs* 2008 Apr;17(7B):235–43.
- [8] Dashiff C, Suzuki-Crumly J, Kracke B, Britton L, Moreland E. Cystic fibrosis-related diabetes in older adolescents: parental support and self-management. *J Spec Pediatr Nurs* 2013 Jan;18(1):42–53.
- [9] Moran A, Brunzell C, Cohen RC, Katz M, Marshall BC, Onady G, et al. Clinical care guidelines for cystic fibrosis-related diabetes: a position statement of the American Diabetes Association and a clinical practice guideline of the Cystic Fibrosis Foundation, endorsed by the Pediatric Endocrine Society. *Diabetes Care* 2010 Dec;33(12):2697–708.
- [10] O'Riordan SM, Dattani MT, Hindmarsh PC. Cystic fibrosis-related diabetes in childhood. *Horm Res Paediatr* 2010;73(1):15–24.
- [11] Morse JM, Field PA. *Qualitative research methods for health professionals*. 2nd ed. Thousand Oaks, CA: Sage Publications; 1995.
- [12] Modi AC, Quittner AL. Validation of a disease-specific measure of health-related quality of life for children with cystic fibrosis. *J Pediatr Psychol* 2003 Dec;28(8):535–45.
- [13] Quittner AL, Buu A, Messer MA, Modi AC, Watrous M. Development and validation of the Cystic Fibrosis Questionnaire in the United States: a health-related quality-of-life measure for cystic fibrosis. *Chest* 2005 Oct;128(4):2347–54.
- [14] Richards L. *Using NVivo in qualitative research*. Australia: Qualitative Solutions and Research Pty. Ltd.; 1999.
- [15] Graue ME, Walsh D. *Studying children in context: theories, methods, and ethics*. Sage Publication; 1998.
- [16] Zindani GN, Streetman DD, Streetman DS, Nasr SZ. Adherence to treatment in children and adolescent patients with cystic fibrosis. *J Adolesc Health* 2006 Jan;38(1):13–7.
- [17] Sawicki GS, Heller KS, Demars N, Robinson WM. Motivating adherence among adolescents with cystic fibrosis: youth and parent perspectives. *Pediatr Pulmonol* 2014 Mar 10. <http://dx.doi.org/10.1002/ppul.23017> [Epub ahead of print].
- [18] DiMatteo MR. Variations in patients' adherence to medical recommendations: a quantitative review of 50 years of research. *Med Care* 2004 Mar;42(3):200–9.
- [19] Balfe M, Doyle F, Smith D, Sreenan S, Brugha R, Hevey D, et al. What's distressing about having type 1 diabetes? A qualitative study of young adults' perspectives. *BMC Endocr Disord* 2013 Jul 25;13(1):25.
- [20] George M, Rand-Giovannetti D, Eakin MN, Borrelli B, Zettler M, Riekert KA. Perceptions of barriers and facilitators: self-management decisions by older adolescents and adults with CF. *J Cyst Fibros* 2010;9:425–32.
- [21] Modi AC, Quittner AL. Barriers to treatment adherence for children with cystic fibrosis and asthma: what gets in the way? *J Pediatr Psychol* 2006;31(8):846–58.
- [22] Bregnballe V, Schiøtz PO, Boisen KA, Pressler T, Thastum M. Barriers to adherence in adolescents and young adults with cystic fibrosis: a questionnaire study in young patients and their parents. *Patient Prefer Adherence* 2011;5:507–15.
- [23] Rapoff MA. In: *Adherence to pediatric medical regimens*. 2nd ed. New York (NY): Springer; 2011.
- [24] Knaff KA, Deatrick JA, Kodadek S. How parents manage jobs and a child's hospitalization. *MCN Am J Matern Child Nurs* 1982;7(2):125–7.

Supplement: Interview questions

1. Tell me about when you were diagnosed with CFRD and what that was like for you and your family.
Did you know anything about CFRD before you were diagnosed?
What thoughts and feelings did you have when you found out?
How did your mom/dad/child react?
2. When you were diagnosed with CFRD, did you receive all of the information you needed about it?
What did the doctors or nurses tell you about CFRD?
What questions did you have for your parents and/or the doctors and nurses?
Is there anything you wish had gone differently?
3. How did the diagnosis of CFRD affect your daily life?
Is it difficult to take care of CFRD on top of your/your child's CF regimen?
What gets in the way of taking care of your/your child's CF or CFRD?
What makes it easier to take care of your/your child's CF or CFRD?
What is the hardest part of taking care of your/your child's CF or CFRD?
4. Do you ever have symptoms of high or low blood sugar?
Does it worry you?
Do you tell your mom/dad about it?/Does your child tell you when he/she has symptoms?
Is it difficult to keep your blood sugars in control?
5. What advice would you give to other kids/adults/parents who just found out they had CFRD?
6. What advice would you give to doctors and nurses who take care of kids and teenagers with CFRD?