Research Paper

Patient and parent perceptions of the diagnosis and management of cystic fibrosis-related diabetes

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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].

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 sample study. 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.
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).
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 make sure that insulin 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

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).

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

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**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) |
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.

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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?