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Journal of Clinical & Translational Endocrinology

journal homepage: www.elsevier.com/locate/jcteCystic fibrosis-related diabetes: The patient perspective[☆]Kelly A. Mason^{a,*}, Brynn E. Marks^{b,c}, Colleen L. Wood^d, Trang N. Le^e^a University of Virginia, Department of Pediatric Endocrinology, PO Box 800386, Charlottesville, VA 22908, USA^b Children's National Hospital, Division of Endocrinology and Diabetes, 111 Michigan Avenue NW, Washington District of Columbia 20010, USA^c George Washington University School of Medicine & Health Sciences, Department of Pediatrics, Ross Hall, 2300 Eye Street, NW, Washington DC 20037, USA^d Billings Clinic, Pediatric Subspecialties Department, Pediatric Diabetes and Endocrinology, 2800 Tenth Avenue North P.O. Box 37000, Billings, MT 59107, USA^e Virginia Commonwealth University, Department of Internal Medicine and Pediatrics, Division of Pediatric Endocrinology, Diabetes, and Metabolism, 2305 N. Parham Road Suite 1, Richmond, VA 23229, USA

ARTICLE INFO

Keywords:

Cystic fibrosis-related diabetes

Patient

Perspective

Understanding

Perception

Quality of life

ABSTRACT

Cystic fibrosis-related diabetes (CFRD) affects nearly 20% of adolescents and 40–50% of adults. However, the impact on patients and their families is poorly understood. Here, we examine how patients perceive CFRD and identify gaps in our understanding of the patient experience.

Despite its relatively high prevalence, data suggest that many individuals are not aware of the possibility of developing CFRD or compare it to other types of diabetes. Annual oral glucose tolerance testing (OGTT) may serve as an opportunity to provide education and prepare individuals for the possibility of developing abnormalities in glucose tolerance.

Many cite lack of awareness of CFRD as the most difficult part of the diagnosis. While factors such as older age and a strong support system promote acceptance, most individuals view the diagnosis negatively and struggle to balance the demands of diabetes with other obligations, including airway clearance, nebulizer therapies, supplementation nutrition, and administration of vitamins and medications. Relatively few people with CFRD monitor their blood glucoses consistently, which is attributed to time constraints or an attempt to avoid pain. In addition, many feel that they are not prone to hypoglycemia and are not concerned with long-term complications, anticipating that they will succumb to their pulmonary disease before these become problematic. The adolescent period presents unique challenges for adherence as children work to develop autonomy.

Factors that promote CFRD adherence include incorporating management into daily CF routines and the support of knowledgeable providers to help develop an individualized approach to management. Diabetes technology has the potential to reduce treatment burden and improve glycemic control, but data in CFRD are limited, and additional study is needed.

Given that CFRD is associated with a decline in health-related quality of life, it is critical that providers understand patients' perspectives and address gaps in understanding and barriers to management.

Introduction

Cystic fibrosis-related diabetes (CFRD) has become more prevalent with time [1] and impacts nearly half of adults living with cystic fibrosis (CF). It is associated with reduced lung function, poor nutritional status, and increased mortality. Despite the fact that it is the most common comorbidity in CF, many individuals are unaware of the potential for developing CFRD, feeling surprised and devastated upon learning of the

diagnosis. Those who are aware of the association between CF and CFRD tend to frame their knowledge of CFRD based on the experiences of friends and family members with other types of diabetes. Annual oral glucose tolerance testing (OGTT) serves as an important screening tool, but may also be an opportunity to provide education and prepare individuals for the possibility of developing abnormalities in glucose tolerance.

Upon learning of the diagnosis, individuals report concern about the

[☆] Disclosure summary: KAM, BEM, CLW, and TNL receive support from the Cystic Fibrosis Foundation. BEM has received investigator initiated research funding from Dexcom, Inc and Tandem Diabetes Care.

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<https://doi.org/10.1016/j.jcte.2021.100279>

Received 8 August 2021; Received in revised form 11 November 2021; Accepted 13 November 2021

Available online 18 November 2021

2214-6237/© 2021 The Authors.

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lack of professional advice, potential need for injections and dietary modifications, and ability to balance the demands of CFRD with CF management. Factors that promote acceptance include older age, a strong support system, and the ability to incorporate CFRD management into established CF routines. Those who are younger with a more active lifestyle view the diagnosis as a disruption and tend to have more difficulty with adjustment. Many view the diagnosis negatively and struggle to balance the demands of diabetes management with CF care. In fact, relatively few monitor their blood glucose levels regularly. Understanding factors that promote or impede adherence is critical to reducing treatment burden.

It is important for providers to recognize the impact of CFRD on individuals and their families. By identifying factors that affect patient understanding and adherence, providers may help patients and families develop ways to reduce the negative impact of CFRD on overall health and quality of life.

Awareness and understanding of CFRD

As noted above, the incidence of CFRD has increased as life expectancy has improved. Despite the fact that CFRD now affects nearly half of adults with CF, many studies have shown that the association between CF and CFRD is still not well recognized. In fact, only 50% of individuals participating in semi-structured interviews were even aware that CFRD was possible [2,3]. Furthermore, many individuals use experiences of family members, friends or acquaintances with other types of diabetes to frame their knowledge of CFRD [2,4,5]. For example, in a study by Dashiff et al., up to 40% of adolescents with CFRD compared it to other types of diabetes. Thirty percent of parents in the same study, could not explain the exact cause of CFRD nor its unique physiology, but did report that it was ‘different than regular diabetes’ or ‘not as bad as’ type 1 diabetes mellitus (T1DM) or type 2 diabetes mellitus (T2DM) [5]. Adult data are limited, though responses from one study suggest that those with normal or impaired glucose tolerance perceive CFRD as similar to T1DM and T2DM, whereas those with CFRD recognize that it is distinct from other types of diabetes, primarily due to differences in dietary recommendations [4].

Many individuals who have CFRD themselves or have children with CFRD recognize the association between CFRD and ‘blood sugars’ [2,5] and/or the pancreas, with some reporting an association with insulin [5] (Table 1). Others feel that there is a hereditary predisposition or increased risk owing to dietary habits [4]. Some believe that CFRD would happen even in the absence of CF [5]. Still others feel that CFRD occurs as a result of poor CF care [2].

Screening

Although annual OGTT is recommended to screen for CFRD [6], only 67% of eligible children and 37% of eligible adults underwent screening in 2019 [7]. Frequently cited reasons for neglecting this screening, which must be performed fasting, include lack of time or appointment availability, lack of symptoms, and fear of needles [4]. It is important to address these barriers, by means of increased education, communication

Table 1
Perceptions regarding the cause of CFRD (2,4,5).

Related to:
• Blood sugar
• Pancreas
• Insulin
• Insufficient insulin
Requires dietary change
Occurs as a result of not taking care of CF
Hereditary predisposition
Eating well protective
Insulin used to improve nutrition
Diagnosis unrelated to CF

and process improvement, as screening provides important diagnostic information and may be used as an opportunity to educate and prepare patients and families for the possibility of CFRD [2].

Diagnosis

The diagnosis of T1DM is often unexpected and, for many, is their first experience with a chronic medical condition. On the other hand, those with CFRD are often diagnosed during adolescence or adulthood after living with CF for many years. While some individuals may be aware of the potential for CFRD, many are not and cite this as the most difficult part of the diagnosis [3,5]. Periodic education on the potential for developing CFRD may help some individuals prepare for the diagnosis [2,4]. Others, however, may perceive the diagnosis as adding to an already complex, time-consuming medical regimen involving airway clearance, inhaled therapies and antibiotics, enzyme replacement and caloric supplementation [2,5].

While few individuals report feeling a sense of relief once CFRD is identified as the cause of previously unexplained symptoms, studies demonstrate that many individuals with CFRD describe their diagnosis negatively [3,5], feeling ‘shocked’, ‘devastated’ or ‘annoyed’. This reaction tends to be more prevalent amongst younger individuals with more active lifestyles who resent the diagnosis of diabetes and view it as a disruption (Table 2). In fact, some individuals diagnosed during adolescence recall rebelling by intentionally going against provider recommendations [3].

Parents of children diagnosed with CFRD report concerns about learning to administer injections, fear of complications such as severe hypoglycemia, and worry about the impact of CFRD on their child’s CF. In general, these negative feelings do not wane. Over time, parents continue to worry about their child’s ability to manage diabetes independently, endorsing concern about their child being away from home and fear that their child could die [5].

Those who are older and have a consistent routine tend to have less difficulty adjusting to the diagnosis of CFRD [3]. For this group, applying the coping mechanisms and routines developed over years of caring for their CF seems to facilitate adjustment to CFRD. In fact, many adults with CFRD participating in semi-structured interviews report eventually “learning to live” with it despite the “extra hassle” [3]. Other factors that facilitate acceptance of the diagnosis include the support of others at home and improvement of symptoms in response to treatment [3]. Some individuals tend to view CFRD as less significant compared to their CF, which limits the impact of the diagnosis [2,3]. For others, recognizing the importance of CFRD upon transitioning to insulin therapy promotes acceptance and adherence [3] (Table 2). Ultimately, despite initial concerns about injections [2,3] and frustrations with another chronic illness, most report a desire to continue with their lives and learn to minimize the impact [3].

Management

The management of CFRD requires attention to blood glucose levels and carbohydrate intake along with insulin injections and awareness of

Table 2
Factors contributing to adjustment (3–4).

Factors that Promote Adjustment to CFRD	Barriers to Adjustment to CFRD
Older age	Younger age
Living with others/support	Living alone
Structured CF routine	More active lifestyle
Preparation for the diagnosis	Lack of preparation
Coping mechanisms learned from CF	Time constraints
	• Effort put into CF care
	• Other life obligations
Improvement in symptoms	
Minimizing CFRD compared to CF	

the impact of medications and exercise on glucose control [6,8]. It is recommended that those with CFRD who are on insulin monitor glucose levels at least 3 times per day and strive to optimize glycemic control to reverse the catabolic effects of insulin deficiency. Management should also involve quarterly clinic visits with ongoing diabetes self-management education provided by a multidisciplinary team with expertise in CFRD [8].

Factors that promote and impede adherence

Many individuals struggle to balance the demands of CFRD management with other obligations [9]. In fact, studies reveal that relatively few people with CFRD monitor their blood glucoses consistently. In one study, only 50% of individuals monitored their glucoses daily, some monitoring as little as every 14 days [9]. Some cite forgetting or attempting to avoid pain as reasons for inconsistent monitoring [2,9] (Table 3). In addition, some parents feel that they are inflicting pain upon their child by checking their blood glucose levels and avoid monitoring due to child resistance. Many individuals who are accustomed to performing CF care at home report embarrassment at having to manage their CFRD in public settings [2]. Others report time constraints due to the effort put into their pulmonary regimen or other obligations [2,9]. Some rationalize the lack of monitoring, reporting that they are not prone to hypoglycemia [9] or that glucose levels would be inaccurate due to frequent snacking [2]. Still others endorse lack of concern about long-term complications of diabetes, anticipating that they will succumb to their pulmonary disease before these become problematic [9].

On the other hand, those with a family, job, or travel tend to be motivated to care for their CFRD to avoid potential complications [4] (Table 3). Although some individuals with CFRD lack concern about long-term complications, many worry about the impact on their CF [5,9]. This, along with recognition of the health benefits associated with improved glycemic control, tends to improve patient adherence [2]. Another factor promoting adherence is the ability to incorporate CFRD management into the daily pulmonary regimen [2,9] and, as opposed to individuals with other types of diabetes, those with CFRD generally find it easier to develop such routines [9].

Adolescence

The treatment of adolescents with CFRD presents unique challenges. Children with chronic conditions, such as CF [10] and T1DM [11,12], tend to become more responsible for self-management as they age due, in part, to a decline in parental involvement. Adherence often becomes challenging for adolescents as they work to develop this autonomy [13].

In addition to difficulties adhering to a complex CF regimen, the introduction of CFRD as a “new” chronic disease layered upon CF can be disconcerting and upsetting [2,3]. Interestingly, in one study, adolescents and parents perceived CFRD management as simpler than CF management. In this study, many adolescents were able to manage their

CFRD independently, though they were still working to become proficient in their CF care. Parents felt that, by managing their CFRD, adolescents would develop the confidence necessary to take a more active role in their CF management [2]. In another study, however, parents reported an overall lack of confidence in their adolescent’s ability to manage their CFRD [5].

Glycemic control throughout adolescence has not been well-studied in CFRD but tends to decline in those with T1DM [11,12,14]. Given that CFRD requires attention during times when adolescents are away from their parents [2], it is important to identify factors that improve adherence to CFRD management.

Data from the T1DM population demonstrate that authoritative parenting, as defined by high levels of warmth and moderate levels of parental control, is associated with improved hemoglobin A1c levels, better adherence, and improved quality of life [15]. Similarly, support of adolescent autonomy improves competence in both CFRD and CF management [5]. Many parents feel that supervision [2] and assistance [5] facilitate adolescent CFRD management. While some adolescents find these helpful, others report that disease-related communication facilitates self-management. Such discussions should focus on providing an understanding of CFRD and explaining the rationale behind specific disease-related tasks. To foster these discussions, providers should offer on-going education tailored to the unique needs of the family, providing methods for promoting adolescent autonomy such as encouraging opinions, providing choices, praising initiative, and fostering problem-solving skills [5]. This becomes particularly important as adolescents begin the transition process with the ultimate goal of successfully transferring to an adult CF program [16].

Diabetes technology

As noted above, many individuals diagnosed with CFRD worry about the need for injections and relatively few monitor their blood glucoses consistently. Diabetes technology has the potential to improve glycemic control and decrease treatment burden. Indeed such technology has been associated with improved treatment satisfaction in T1DM [17], but data in CFRD are limited.

Continuous glucose monitors (CGM) are minimally invasive devices that can potentially replace fingersticks by use of a subcutaneous sensor that measures and reports glucose values every 5–15 min [18]. These devices are FDA approved and widely used for T1DM and have been associated with improvements in glycemic control, health-related quality of life (HRQoL), and treatment satisfaction in this population [19,20]. Perceptions of CGM technology within the CF population are overwhelmingly positive. In fact, 81% of respondents agreed with at least 5 of 7 positive statements about CGM use in one study [21]. Furthermore, the overall survey score among individuals with CF was higher than the score previously recorded among those with T1DM, suggesting a more positive perception within the CF community. In one survey, 75% of the 51 respondents with CFRD reported using a CGM. Despite such positive perceptions, CGM use was discontinued by 19%, mostly commonly due to cost and increased worry about blood glucose levels [21].

In the same study, 29% of those with CFRD reported using insulin pump therapy [21]. As compared to injection based therapy, insulin pumps more closely mimic physiologic insulin secretion and have been associated with improved glycemic control and psychosocial benefits in individuals with T1DM [22,23]. Perceptions of insulin pumps within the CF community were less positive, with only 22% of respondents agreeing with at least 5 of 7 positive statements referencing pump use. As seen with CGM systems, insulin pumps were discontinued by nearly a third of respondents, most commonly attributed to pain and skin irritation [21].

Diabetes technology has the potential to simultaneously optimize glycemic control and reduce treatment burden for those with CFRD. However, strategies to improve device uptake and sustain use are

Table 3

Factors contributing to treatment adherence (2, 3, 5, 9).

Factors that Promote Adherence	Barriers to Adherence
Travel, career, family	Forgetting
Worry about long-term diabetes complications	Lack of concern about long-term diabetes complications
Incorporating CFRD into CF routine	Younger age
Improvement in symptoms/lung function	Lack of immediate effects
Adverse event related to CFRD	Pain
Switching from oral agent to insulin	Child resistance
	Time constraints
	Not prone to hypoglycemia
	Embarrassment treating in public

needed to maximize the benefits of such technology.

Psychosocial effects

In a longitudinal observational study of individuals with CF aged 14–48 years, CFRD was noted to negatively impact more than half of the HRQoL domains assessed through the Cystic Fibrosis Quality of Life Questionnaire (CFQ-R) [24] including social functioning, body image, chest symptoms, emotional responses, and interpersonal relationships [25].

A separate survey compared HRQoL scores among people with CFRD to those with impaired glucose tolerance or normal glucose tolerance [26]. The treatment burden score was highest among individuals with CFRD treated with insulin and there was a trend towards similar findings in the physical functioning, social and role functioning, and body image domains. These findings were not observed in individuals with CFRD who were not treated with insulin, suggesting that the increased burden of CFRD can be attributed to insulin therapy. Interestingly, when compared to T1DM, CFRD had less impact in the domains of treatment burden and worry about diabetes [27]. The reason behind the differences in treatment burden scores was not evaluated, though it may be related to the fact that some individuals with CFRD are treated with intermittent insulin therapy during illness or only one injection per day. Other factors may include the ability of those with CFRD to incorporate diabetes management into their existing CF routine, or due to minimizing CFRD compared to CF. The differences in scores related to worry about diabetes may be due to differences in insulin regimens, or to the lower risk of diabetic ketoacidosis in CFRD, or less concern about long-term complications.

The rate of depression in CF is higher than that of the general population [28]. Whether CFRD increases the risk of depression within the CF population warrants further study. In a retrospective cross-sectional study of 209 adults with CF, the odds of depression were noted to be 2.3 times greater among individuals with CFRD (48%) compared to those without (28%) [29]. Another study found no difference in the rates of depression in children with CFRD and their parents when compared to those without CFRD, though it should be noted that the number of participants with CFRD was very small [30]. Neither study identified an increased risk of anxiety in individuals with CFRD [29,30]. However, additional study is needed to determine whether the duration or severity of CFRD and/or treatment regimen impacts the rates of anxiety and depression.

Role of providers

Understanding how individuals conceptualize CFRD is critical to providing high-quality care. As previously noted, many individuals with CF are either unaware of the association with CFRD or frame their knowledge of CFRD based on the experiences of friends or family members with other types of diabetes. Annual OGTT provides an opportunity for education and may help patients and families prepare for the possibility of developing CFRD in the future.

For those who ultimately develop CFRD, care should involve collaboration between members of a multidisciplinary team including the pulmonologist, registered dietician with knowledge of nutrition guidelines for CFRD, and an endocrinologist with an understanding of CF, amongst others. Providers should work with patients to develop management strategies tailored to their unique lifestyles [9] and identify ways to incorporate CFRD management into an individual's CF regimen to reduce treatment burden [5]. Education must be ongoing, providing individuals with an understanding of the importance of glycemic control on multiple health-related outcomes including growth, nutrition, and pulmonary function. Providers should strive to understand barriers to management, confront problematic behavior in a compassionate manner [4], and partner with patients to identify ways to improve CFRD management and quality of life.

Future directions

We are beginning to understand the impact of CFRD on individuals and their families, but additional studies are needed to better understand how patients perceive all aspects of CFRD including understanding and awareness, barriers to screening and management, and psychosocial impact. Such studies should evaluate the age at which to begin education on screening rationale and the unique physiology of CFRD as well as who patients prefer to provide such information. Some may prefer that this education be provided by a trusted member of the pulmonary team whereas others may prefer to meet with an endocrinologist so they have established care should abnormalities in glucose tolerance occur in the future. Studies are also needed to better understand why patients forego annual screening and identify novel ways to overcome these barriers. More detailed studies assessing the impact of diabetes technology and various insulin regimens on HRQoL are also needed. Finally, additional investigation is necessary to better characterize the association between CFRD and mental health in patients and their families.

Much of the information available thus far comes from semi-structured interviews. One benefit of such studies is that they allow participants to speak freely within a common set of topics. However, data gathered from such interviews are not adjusted for variables that may impact perception such as CFRD duration or treatment regimen and alternate forms of study should be explored.

Funding

This work was supported by the Cystic Fibrosis Foundation Envision CF: Emerging Leaders in CF Endocrinology II Program [grant numbers MASON19GEO, MARKS19GEO, WOOD19GEO].

CRedit authorship contribution statement

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