

Patient Knowledge and Clinic Attendance in Adolescent Patients with Cystic Fibrosis

Traci M. Kazmerski, MD,¹ Elizabeth Miller, MD, PhD,¹ Kaleab Z. Abebe, PhD,²
Janice Matisko, RN, BSN,¹ Diane Schachner, RN, MSN,¹ and Jonathan Spahr, MD¹

Background: With the rising age of patients with cystic fibrosis (CF), strategies to improve adolescents' adherence to care are needed. This study investigates the association between knowledge of disease and objective markers of clinic attendance in an adolescent CF population.

Methods: Patients with CF aged 13–20 years completed a CF knowledge survey ($n=38$). Associations of knowledge scores with attendance measures and health status data were assessed.

Results: Higher overall knowledge scores were associated with regular evaluations by dietitians (OR 1.08, $p=0.045$) and timely completion of pulmonary function testing (PFTs; OR 1.13, $p=0.009$) and routine cultures (OR 1.12, $p=0.012$). Higher nutrition health and treatment knowledge scores were also associated with timely completion of routine PFTs (OR 1.14, $p=0.005$ and OR 1.09, $p=0.012$) and cultures (OR 1.10, $p=0.022$ and OR 1.08, $p=0.019$).

Conclusions: This is the first study demonstrating an association between adolescent disease knowledge and objectively measured adherence to recommended clinic attendance and evaluation. More adherent patients have better knowledge of their disease. Targeting adolescent CF patients' knowledge may be a modifiable factor to improve attendance and long-term management.

Introduction

NEARLY 50% OF PATIENTS with cystic fibrosis (CF) are adults.¹ As life expectancy has risen with improved care and therapies, the focus in care has shifted toward improving chronic care management.² Increasing knowledge regarding the disease process itself is theorized to influence self-care and health outcomes positively in chronic disease populations.^{3,4}

Unfortunately, numerous studies have shown that disease-related knowledge in CF is suboptimal.^{5–7} Furthermore, patients' knowledge of their chronic disease has been recognized as a factor directly associated with adherence to recommended care plans.⁸ This relationship has been demonstrated specifically among younger patients with CF and their parents. In younger children, Gudas et al. found that a higher level of knowledge of the disease was associated with better compliance as reported in a parental survey.⁹ Ievers et al. also identified a strong relationship between accurate understanding of CF treatment regimens and reported rates of adherence in children aged 6–10 years and their parents.¹⁰

Previous reports have relied on indirect means to assess adherence to treatment and have focused on younger chil-

dren and parental knowledge. With the rising number of adolescent patients with CF, elucidating the relationship between adolescent knowledge of their disease process and their adherence to both clinic attendance and treatment/preventive care is needed to guide interventions for improvement. This is especially vital, as chronically ill patients often have decreased adherence to treatment and clinic attendance during adolescence.¹¹

No studies to date have assessed knowledge of disease in an adolescent CF population and examined the relationship with objective markers of adherence to recommended care guidelines and overall health status. The purpose of this study was to address this gap in research about the potential role of adolescents' knowledge of their disease process and clinic attendance. It was hypothesized that higher levels of patient knowledge would correlate with better adherence to recommended evaluations of disease progression and clinic visit frequency.

Materials and Methods

During a 2-year period from August 2009 to September 2011, adolescent patients aged 13–22 years old with a

¹Children's Hospital of Pittsburgh of University of Pennsylvania Medical Center, Department of Pediatrics, University of Pittsburgh School of Medicine, Pittsburgh, Pennsylvania.

²Center for Research on Health Care Data Center, Division of General Internal Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania.

diagnosis of CF confirmed by positive sweat chloride and/or two disease-causing cystic fibrosis transmembrane conductance regulator (CFTR) mutations attending a pediatric CF center were offered participation in the *CF My Way*[®] program on adherence during a routine follow-up visit (not an acute illness or pulmonary exacerbation).^a As part of this program, a baseline CF knowledge assessment—a questionnaire called the My CF Knowledge (CFK)—was completed on an electronic tablet or on paper during the clinic visit.

As a quality improvement project and review of existing data, the University of Pittsburgh Institutional Review Board (IRB) deemed this study exempt from IRB approval. No informed consent was required, and participants did not receive compensation.

Measures

The CFK consists of 43 multiple-choice items that assess four sub-domains of CF knowledge: lung health (seven questions), nutrition health (14 questions), CF health (11 questions), and treatment (11 questions).³ Items were weighted evenly and an overall score was generated and reported as total percentage correct. Scores in each of the four sub-domains were also reported and reported as percentage correct. This measure has previously demonstrated good internal consistency in adolescent patients with CF ($\alpha=0.91$).¹²

Demographic information and objective measurements of disease status were obtained from query of the CF Registry for each participant. These included age, sex, percentage of predicted forced expiratory volume in one second (FEV₁), body mass index (BMI), use of nutritional supplements, and presence of a gastrostomy tube.

FEV₁ was obtained from the CFK visit and divided into mild (FEV₁ $\geq 70\%$), moderate (FEV₁ 41–69%), and severe disease (FEV₁ $\leq 40\%$) per previous accepted categorization of disease severity.¹³ BMI data were obtained during the CFK visit and divided into categories of adequate nutrition (BMI ≥ 25 th percentile), nutritional risk (BMI 10th–25th percentile), and nutritional failure (BMI ≤ 10 th percentile) per Cystic Fibrosis Foundation (CFF) consensus guidelines.¹⁴ Use of nutritional supplements (including high-calorie drinks and/or powders) and presence of a gastrostomy tube were obtained from the CFK visit.

Clinic attendance was measured via surrogate markers obtained from the CF Registry: routine completion of pulmonary function testing (PFTs), throat/sputum cultures, and evaluation by dietitians. These specific markers were chosen, as this institution routinely requires completion of these evaluations at each clinic visit regardless of disease severity. Dietitians are available for every CF clinic visit for patients in the center, and PFTs and throat cultures are routinely requested for all clinic visits. The CF Registry is particularly useful for comparing attendance among current CF center patients, as the database only contains active center patients (i.e., changes in residence, for example, result in removal from the registry database). Subjects were considered adherent to clinic attendance if the marker was recorded every 3 months per this institution's policy. Data were gathered from the 6 months prior to and following the administration of the CFK. A 7–10-day time window was considered at the

beginning and end of the 1-year data collection period before deeming subjects nonadherent.

Analyses

The associations of overall knowledge score and scores in each domain of the CFK with disease severity (FEV₁, BMI, use of nutrition supplements, and presence of a gastrostomy tube) were assessed via bivariate analyses. Associations of knowledge scores with clinic attendance measures (timely completion of pulmonary function testing and sputum/throat cultures and attendance at routine dietary visits) were examined using exact logistic regression due to small sample size. Age, sex, and disease severity (mean FEV₁ and BMI) were also examined between groups using the Mann–Whitney *U*-test or Fisher's exact test. All analyses were conducted using STATA (StataCorp LP, College Station, TX).

Results

Knowledge assessments were obtained from 38 adolescents with a confirmed diagnosis of CF, ranging in age from 13 to 20 years over a 2-year period (approximately one-third of all adolescent patients seen in the center). The mean overall CFK score for the sample was 79.6 ($SD=12.0$). The mean age was 17.1 years ($SD=1.9$; 57.9% male). Average FEV₁ for the sample was 84.0% ($SD=23\%$), with 76.3% of the sample classified as having mild disease, 18.4% with moderate disease, and 5.3% with severe disease. Average BMI for the sample was the 54.0th percentile for age ($SD=33.0$), with 73.7% of the sample classified as having adequate nutrition, 10.5% classified as nutritional risk, and 15.8% classified as nutritional failure. Approximately one in five (21.1%) of participants were using nutritional supplements, and 15.8% of participants had a gastrostomy tube in place (Table 1). This sample is representative of the adolescent patient population at this CF study center.

Disease severity (BMI and FEV₁) was not associated with overall and specific CFK scores. The presence of a gastrostomy tube or use of nutritional supplements had no significant association with overall score or any of the sub-domain scores.

Regarding clinic attendance, the primary outcome of interest, 13.2% of participants were found to be nonadherent with recommended quarterly dietary visits, 7.9% did not obtain PFTs at appropriate intervals, and 7.9% did not obtain CF throat/sputum cultures at appropriate intervals. It should be noted that different individual subjects were nonadherent with sputum cultures and PFTs and, thus, both were used as surrogates for attendance.

Overall CFK scores were calculated for both adherent and nonadherent subjects (Table 1). Using exact logistic regression models, higher overall knowledge scores were associated with regular evaluations by dietitians (OR 1.08, $p=0.045$). No sub-domain scores showed significant associations. Higher overall knowledge scores were also associated significantly with timely completion of PFTs (OR 1.13, $p=0.009$). Higher nutrition health and treatment knowledge scores were also associated with PFT completion (OR 1.14, $p=0.005$ and OR 1.09, $p=0.012$, respectively); other sub-domain scores showed no significant correlation. Higher overall scores, nutrition health, and treatment knowledge scores were associated with timely completion of routine

^a*CF My Way*[®] is a registered trademark of Novartis AG.

<i>Characteristic</i>	<i>Patient population (n=38), n (%)</i>
Age, mean years (<i>SD</i>)	17.1 (1.9)
Male	22 (57.9)
Female	16 (42.1)
<i>Health outcome measures:</i>	
FEV ₁ % predicted, mean (<i>SD</i>)	84.0 (23.0)
Mild (≥70%), <i>n</i> (%)	29 (76.3)
Moderate (41–69%), <i>n</i> (%)	7 (18.4)
Severe (≤40%), <i>n</i> (%)	2 (5.3)
BMI, percentile (<i>SD</i>)	54.0 (33.0)
Adequate (≥25th percentile), <i>n</i> (%)	28 (73.7)
Nutritional risk (10th–25th percentile), <i>n</i> (%)	4 (10.5)
Nutritional failure (≤10th percentile), <i>n</i> (%)	6 (15.8)
Use of nutritional supplements	8 (21.1)
Presence of gastrostomy tube	6 (15.8)
<i>Adherence measures:</i>	
Attendance at dietary visits—nonadherent	5 (13.2)
PFT completion—nonadherent	3 (7.9)
CF throat/sputum culture completion— nonadherent	3 (7.9)
<i>Knowledge score (CFK overall score), mean (SD)</i>	79.6 (12.0)
Nonadherent to attendance at dietary visits	68.3 (21.6)
Adherent to attendance at dietary visits	81.2 (9.3)
Nonadherent to PFT completion	59.7 (23.3)
Adherent to PFT completion	82.2 (9.7)
Nonadherent to CF throat/sputum culture completion	60.9 (25.4)
Adherent to CF throat/sputum culture completion	81.2 (9.2)

cultures (OR 1.12, $p=0.012$; OR 1.10, $p=0.022$; and OR 1.08, $p=0.019$, respectively). Other sub-domain scores showed no significant associations. No significant differences in age, sex, FEV₁, or BMI were found between nonadherent and adherent subjects (Table 2). Additional analyses looking at annual laboratory evaluation and chest radiography were not associated with CFK scores (results not shown).

This is the first study to demonstrate an association between adolescent knowledge and objectively measured adherence to recommended clinic attendance in CF. The findings suggest that adolescent patients with CF who are adherent with routine clinical follow-up (surrogates being attendance at dietary visits and completion of routine PFTs and sputum cultures) have better knowledge of their disease. Overall and selected sub-domain knowledge scores are higher in adherent patients, suggesting that knowledge of CF as a whole may be an important factor in self-management.

Unfortunately, better knowledge of disease does not translate into improved health status, as FEV¹, BMI, use of nutritional supplements, or the presence of a gastrostomy tube were not associated with greater overall knowledge. This is not surprising, as these findings are similar to previous studies showing that education had no positive effects on lung function or weight in CF.¹⁵

In this observational study, the association between higher knowledge and clinic attendance adherence demonstrated in adolescent CF patients does not imply causality. It may be that increasing patient knowledge about CF leads to increased clinic attendance, and it is also possible that routine attendance improves patient disease knowledge. The results support the possibility that targeting CF patient knowledge through innovative educational programs might help improve clinic attendance and adherence to routine evaluations.

Knowledge scores, M (SD)	Dietitian evaluation			PFT completion			Routine culture completion		
	Odds ratio	p-Value		Odds ratio	p-Value		Odds ratio	p-Value	
Overall score	1.08	0.045		1.13	0.009		1.12	0.012	
Lung health score	1.06	0.061		1.05	0.169		1.05	0.169	
Nutrition health score	1.04	0.240		1.14	0.005		1.10	0.022	
CF health score	1.05	0.134		1.05	0.289		1.06	0.160	
Treatment score	1.04	0.126		1.09	0.012		1.08	0.019	
<i>Subject characteristics,</i>									
M (SD)	Nonadherent	Adherent	p-Value	Nonadherent	Adherent	p-Value	Nonadherent	Adherent	p-Value
Age	17.2 (2.5)	17.1 (1.9)	0.619	17.8 (0.9)	17.1 (2.0)	0.386	17.8 (0.9)	17.0 (2.0)	0.316
Sex									
Male, <i>n</i>	3	19	0.654	2	20	0.621	2	20	0.621
Female, <i>n</i>	2	14		1	15		1	15	
FEV ₁ % predicted	76.0 (21.4)	84.0 (23.0)	0.353	71.7 (2.1)	84.5 (23.3)	0.234	86.7 (28.0)	83.2 (22.6)	0.999
BMI mean kg/m ²	46.8 (46.0)	54.5 (31.9)	0.730	56.0 (49.7)	53.3 (32.7)	0.871	56.7 (50.0)	53.3 (32.7)	0.829

Adherence measured via surrogate markers of clinic attendance including dietitian evaluation and PFT and routine culture completion. Subjects were considered adherent if the marker was recorded every 3 months from the 6 months prior to and following CFK administration.

While improving knowledge is certainly not the sole strategy for improving adherence,¹⁶ the association between knowledge and clinic attendance confirms prior self-management studies in younger patients that point to knowledge as an important modifiable factor. Bartholomew et al. found that an education program targeting children and their caregivers both increased CF knowledge and was associated with improved coping and self-efficacy.¹⁷ Downs et al. conducted a multi-center, randomized controlled trial to examine the effectiveness of a self-management education program on aerosol and airway clearance techniques in children with CF. Findings confirmed that disease knowledge contributes to both patients' and caregivers' capacity for self-management and adherence.¹⁸ Many have also concluded that improvements in knowledge alone are insufficient to improve self-management and suggest that greater benefit may be had from education programs that address both disease knowledge as well as problem-solving skills.^{19–21} The findings from this study suggest that targeting adolescent knowledge of their disease process may be a useful component to integrate into interventions designed to improve self-management and adherence to care.

This study's unique contribution is the focus on adolescence, specifically the relationship between CF disease knowledge and clinic attendance as an adherence measure. Adolescents with chronic disease, unlike adults who often need to be rehabilitated from illness to resume their independent lives, have no prior experience to draw on and truly need to be "habilitated" as they take responsibility for their health.²² This habilitation (or transition to independence) is often challenging; studies have found that chronically ill patients enter the adolescent years displaying reduced coping skills and adherence.^{23,24} With previous literature highlighting an association between worse adherence to respiratory medications and higher acute healthcare use in CF, finding ways to improve adherence in CF adolescents is imperative.²⁵

In light of the rising number of adolescents and adults with CF, finding novel ways to optimize this habilitation is a priority.^{26,27} Little is known about the factors that predict a successful transition of care from pediatric to adult providers.²⁸ As inadequate education can lead to poor self-management, improving patient understanding of their disease may be an important component to address as part of this transition.^{29,30}

While this study adds to the literature on CF knowledge and the relationship to adherence to routine clinical care guidelines, it is certainly not without limitations. This is a single-site study with a relatively small sample. Additionally, it included only approximately one-third of adolescent patients in the center. As it can be difficult to recruit large populations in CF due to the relatively low prevalence of CF and its severity, a larger multi-center study is warranted. The knowledge survey that was utilized was not comprehensive. It did not include questions addressing reproductive and sexual health, CF-related diabetes or liver disease, end of life care, or genetics—all of which are more prevalent topics among older CF populations and thus important to address in adolescence. Additionally, this sample was noted to have relatively overall good health as measured by FEV₁ and BMI measures, which may limit the ability to detect associations among disease severity, knowledge, and clinic attendance. Other health status measures to include in future

investigations include direct measurements of clinic attendance, rates of pulmonary exacerbations, trends in PFTs, or hospital admissions. Further demographic information to consider includes markers of socioeconomic status and parental level of education. In addition, information on nonresponders and dropouts were not available for this retrospective study.

The association between knowledge and clinic attendance found in this study does not necessarily translate to subject adherence with recommended therapies. As a cross-sectional study, it is not known whether other factors that contribute to the findings are also associated with better knowledge of the disease, and cannot imply causality. Future longitudinal studies should also include objective measures of medication and therapy compliance as well as patient-reported measures of adherence. However, it should be noted that the objective measurements of adherence to routine clinical follow-up were rather restrictive. Following the authors' institution's more aggressive approach, strict attendance at clinic visits every 3 months including dietary assessment was required to be considered "adherent." The Cystic Fibrosis Foundation recommendation is currently for annual dietary assessment. Thus, these conservative measures of attendance strengthen the findings.

In conclusion, this is the first study to demonstrate an association between patient knowledge and adherence to clinic attendance in an adolescent CF population. These findings suggest that patient knowledge of their disease may be an important modifiable factor in improving clinic attendance in adolescents with this chronic illness and should be integrated into interventions targeting self-management and adherence. Additionally, it may be important to shift attention intentionally to the deliberate education of adolescents with CF rather than rely on diffusion of information and knowledge about their disease from their parents and caregivers. Educational programs for teen populations that target knowledge of their disease should be further developed in an effort to improve self-management with the goal of influencing successful transition to adult care.

Acknowledgments

The authors would like to acknowledge Novartis AG for the use of the *CF My Way*[®] knowledge assessments, and Michael McClincy, MD, for his assistance with statistical analyses.

Author Disclosure Statement

No competing financial interests exist.

References

1. Cystic Fibrosis Foundation Annual Patient Registry Report. Bethesda, MD, 2011.
2. Yankaskas JR, Marshall BC, Sufian B, Simon RH, Rodman D. Cystic fibrosis adult care: consensus conference report. *Chest* 2004;125:1S–39S.
3. Davis MA, Quittner AL, Stack CM, Yang MC. Controlled evaluation of the STARBRIGHT CD-ROM program for children and adolescents with cystic fibrosis. *J Pediatr Psychol* 2004;29:259–267.
4. Stapleton DR, Gurrin LC, Zubrick SR, Silburn SR, Sherriff JL, Sly PD. What do children with cystic fibrosis and their

- parents know about nutrition and pancreatic enzymes? *J Am Diet Assoc* 2000;100:1494–500.
5. Henley LD, Hill ID. Errors, gaps, and misconceptions in the disease-related knowledge of cystic fibrosis patients and their families. *Pediatrics* 1990;85:1008–1014.
 6. Hames A, Beesley J, Nelson R. Cystic fibrosis: what do patients know, and what else would they like to know? *Respir Med* 1991;85:389–392.
 7. Conway SP, Pond MN, Watson A, Hamnett T. Knowledge of adult patients with cystic fibrosis about their illness. *Thorax* 1996;51:34–38.
 8. Rapoff MA. *Adherence to Pediatric Regimens*. New York: Kluwer/Plenum, 1999.
 9. Gudas LJ, Koocher GP, Wypij D. Perceptions of medical compliance in children and adolescents with cystic fibrosis. *J Dev Behav Pediatr* 1991;12:236–242.
 10. Ievers CE, Brown RT, Drotar D, Caplan D, Pischevar BS, Lambert RG. Knowledge of physician prescriptions and adherence to treatment among children with cystic fibrosis and their mothers. *J Dev Behav Pediatr* 1999;20:335–343.
 11. Coupey SM, Cohen MI. Special considerations for the health care of adolescents with chronic illnesses. *Pediatr Clin North Am* 1984;31:211–219.
 12. Kimberg CI, Marciel KK, Riekert KA, Zhang J, Quittner AL. Knowledge of disease management in adolescents with cystic fibrosis. (Abstract) NACFC. Baltimore, MD, 2010.
 13. Gee L, Abbott J, Conway SP, Etherington C, Webb AK. Quality of life in cystic fibrosis: the impact of gender, general health perceptions and disease severity. *J Cystic Fibros* 2003;2:206–213.
 14. Borowitz D, Baker RD, Stallings V. Consensus report on nutrition for pediatric patients with cystic fibrosis. *J Pediatr Gastr Nutr* 2002;35:246–259.
 15. Savage E, Beirne PV, Ni Chroinin M, Duff A, Fitzgerald T, Farrell D. Self-management education for cystic fibrosis. *Cochrane Database Syst Rev* 2011;CD007641.
 16. Everhart RS, Fiese BH, Smyth JM, Borschuk A, Anbar RD. Family functioning and treatment adherence in children and adolescents with cystic fibrosis. *Pediatr Allergy Immunol Pulmonol* 2014;27:82–86.
 17. Bartholomew LK, Czyzewski DI, Parcel GS, et al. Self-management of cystic fibrosis: short-term outcomes of the Cystic Fibrosis Family Education Program. *Health Educ Behav* 1997;24:652–666.
 18. Downs JA, Roberts CM, Blackmore AM, Le Souef PN, Jenkins SC. Benefits of an education programme on the self-management of aerosol and airway clearance treatments for children with cystic fibrosis. *Chron Respir Dis* 2006;3:19–27.
 19. Parcel GS, Swank PR, Mariotto MJ, et al. Self-management of cystic fibrosis: a structural model for educational and behavioral variables. *Soc Sci Med* 1994;38:1307–1315.
 20. Quittner AL, Kimberg C, Marciel K, Zhang J, Riekert K. Randomized, controlled trial of a behavioral adherence intervention for adolescents with cystic fibrosis: I change adherence and raise expectations (iCARE). (Abstract) *Chest* 2011;140:908A.
 21. Watson H, Bilton D, Truby H. A randomized controlled trial of a new behavioral home-based nutrition education program, “Eat Well with CF,” in adults with cystic fibrosis. *J Am Diet Assoc* 2008;108:847–852.
 22. Stark LJ, Quittner AL, Powers SW, et al. Randomized clinical trial of behavioral intervention and nutrition education to improve caloric intake and weight in children with cystic fibrosis. *Arch Pediatr Adolesc Med* 2009;163:915–921.
 23. DiGirolamo AM, Quittner AL, Ackerman V, Stevens J. Identification and assessment of ongoing stressors in adolescents with a chronic illness: an application of the behavior-analytic model. *J Clin Child Psychol* 1997;26:53–66.
 24. Bucks RS, Hawkins K, Skinner TC, Horn S, Seddon P, Horne R. Adherence to treatment in adolescents with cystic fibrosis: the role of illness perceptions and treatment beliefs. *J Pediatr Psychol* 2009;34:893–902.
 25. Quittner AL, Zhang J, Marynchenko M, et al. Pulmonary medication adherence and health-care use in cystic fibrosis. *Chest* 2014;146:142–151.
 26. Foster C, Eiser C, Oades P, et al. Treatment demands and differential treatment of patients with cystic fibrosis and their siblings: patient, parent and sibling accounts. *Child Care Health Dev* 2001;27:349–364.
 27. Towns SJ, Bell SC. Transition of adolescents with cystic fibrosis from pediatric to adult care. *Clin Respir J* 2011;5:64–75.
 28. Rosen DS, Blum RW, Britto M, Sawyer SM, Siegel DM, Society for Adolescent Medicine. Transition to adult health care for adolescents and young adults with chronic conditions: position paper of the Society for Adolescent Medicine. *J Adolesc Health* 2003;33:309–311.
 29. Tuchman LK, Schwartz LA, Sawicki GS, Britto MT. Cystic fibrosis and transition to adult medical care. *Pediatrics* 2010;125:566–573.
 30. Sawicki GS, Tiddens H. Managing treatment complexity in cystic fibrosis: challenges and opportunities. *Pediatr Pulmonol* 2012;47:523–533.

Address correspondence to:

Traci M. Kazmerski, MD
 Children's Hospital of Pittsburgh of UPMC
 Department of Pediatrics
 University of Pittsburgh School of Medicine
 One Children's Hospital Drive
 4401 Penn Avenue
 AOB 3rd Floor, Suite 3300
 Pittsburgh, PA 15201

E-mail: Traci.kazmerski@chp.edu

Received for publication December 13, 2014; accepted after revision April 13, 2015.