PEDIATRIC ALLERGY, IMMUNOLOGY, AND PULMONOLOGY
Volume 27, Number 2, 2014

DOI: 10.1089/ped.2014.0327

Family Functioning and Treatment Adherence in Children and Adolescents with Cystic Fibrosis

Robin S. Everhart, PhD,[1] Barbara H. Fiese, PhD,[2] Joshua M. Smyth, PhD,[3] Adrienne Borschuk, MS,[1] and Ran D. Anbar, MD[4]

Background: Cystic fibrosis (CF) is one of the most challenging pediatric illnesses for families to manage. There is, however, limited research that considers the associations between family functioning and treatment adherence in children and adolescents with CF.
Methods: Nineteen children with CF (mean age = 12.42 years, mean forced expiratory volume in one second (FEV_1) = 90.9% predicted) and their families participated in the study. Caregiver and child participants completed interview-based assessments and were then videotaped during a family mealtime.
Results: Mean scores on several domains of family functioning fell in the ''unhealthy'' range. Better family functioning was found among older children. Better family functioning was also associated with better adherence to antibiotic treatment and worse adherence to enzymes.
Conclusions: Findings suggest that family functioning may be an important correlate of treatment adherence in children and adolescents with CF. Future research should replicate these findings in larger samples of children and adolescents with CF.

Introduction

Cystic fibrosis (CF) is a genetically transmitted disease that affects 1 in 3,700 children born in the United States every year, and is one of the most challenging pediatric illnesses for families to manage.[1] Oppositional child behaviors, difficulty swallowing pills, managing therapies in the context of daycare and school, and lack of time have been identified as barriers to treatment compliance in CF.[2–4] As highlighted by a family systems perspective, the child's environment plays an important role in child health outcomes and child well-being.[5] The association between family functioning and treatment adherence has been studied extensively in some pediatric conditions, including diabetes and asthma.[6–8] There is limited, but encouraging, evidence linking treatment adherence with the family context in families with a child with CF.[9,10]

In considering family functioning, observational studies of mealtimes can serve as a microcosm for how families regulate child behavior, manage their affect, communicate, and interact with one another.[11,12] Family mealtimes are often a more difficult routine for families with CF to manage because of the need to comply with increased nutritional demands.[13] Families with a young child with CF score significantly lower than families without a child with a pediatric illness on multiple dimensions of family functioning.[1,14–16] Research has focused exclusively on how families with a toddler, preschooler, or school-age child manage such challenges at the dinner table.[17]

The purpose of this exploratory, mixed-methods study was to determine the level of family functioning among families with a child with CF between the ages of 8 and 19 years from an observational measure of family functioning, and to determine the association between family functioning and treatment adherence. We expected that the level of family functioning would decrease as the child's age increased. Second, we expected that lower family functioning would be related to lower levels of adherence. The aims of our study highlight the lack of existing research and knowledge on treatment adherence in children and adolescents with CF. As the disease is progressive, adherence to medical regimens is critical for a longer life expectancy and better quality of life among patients with CF.[18]

Methods

All relevant Institutional Review Boards approved this study. Nineteen families with a child with CF were enrolled

[1]Department of Psychology, Virginia Commonwealth University, Richmond, Virginia.
[2]Family Resiliency Center, Department of Human and Community Development, University of Illinois at Urbana–Champaign, Champaign, Illinois.
[3]Department of Biobehavioral Health, The Pennsylvania State University, State College, Pennsylvania.
[4]Department of Pediatrics, State University of New York Upstate University Hospital, Syracuse, New York.

Table 1. Mean Scores on the Mealtime Interaction Coding System (MICS) and Associations with Socioeconomic Status (SES; n = 16 Families)

MICS subscales	*Mean*	*Standard deviation*	*Range*	*Number (%) of families in ''unhealthy'' range*	*Correlation with SES (ρ)*
Task accomplishment	4.94	0.85	2–6	1 (6)	0.43
Communication	4.63	1.31	1–6	6 (38)	0.72**
Roles	5.06	0.68	4–7	2 (13)	0.56*
Affect regulation	4.50	1.10	2–6	6 (38)	0.75**
Interpersonal involvement	4.69	1.35	1–7	5 (31)	0.61*
Behavior control	4.94	0.77	3–6	3 (19)	0.55*
General functioning	4.88	0.89	3–6	3 (19)	0.56*

Scores on MICS scales below 5.0 are considered ''unhealthy.''
*$p < 0.05$, **$p < 0.01$.

(53% boys, mean (M) = 12.42 years, standard deviation (SD) = 4.13 years). Mean forced expiratory volume in one second (FEV_1) was 90.9% predicted (SD = 19.38%; range 52–135%). Fifteen of the 19 children were prescribed at least one antibiotic (e.g., azithromycin); three children were taking tobramycin only. Seventeen children were prescribed at least one pancreatic enzyme (e.g., pancrelipase). All children were prescribed at least one aerosolized medication (e.g., dornase alfa). The majority (95%) of primary caregivers was the child's biological mother (M = 40.58 years, SD = 7.25 years), and 95% of these had at least a high school degree or equivalent. Hollingshead scores ranged from 20 to 58 (M = 39.47, SD = 12.63). All families were Caucasian.

Families were recruited through a pulmonary clinic at a medical center in a mid-size city. Inclusion criteria included a clinical diagnosis of CF, child age between 8 and 19 years, child living at home, no hospitalizations for CF in the last 6 months, and no lung transplant. Written informed consent was obtained from caregivers. Parents completed a background questionnaire and the Treatment Adherence Rating Scale (TARS),[19] a 16-item measure of how well the patient followed the prescribed CF regimen in the past 2 weeks. Socioeconomic status (SES) was measured by the Hollingshead Four Factor Index,[20] which determines SES from an average of parent educational level (1 = ''less than 7th grade'' to 7 = ''professional degree'') and occupation type (1 = ''unemployed'' to 9 = ''higher executive'').

Following completion of the questionnaire, families were instructed on the use of the video camera and asked to call the researchers to retrieve the video camera after completion of the meal. Families were asked to rate how typical the meal was for their family on a scale from 1 = ''not at all typical'' to 4 = ''very typical.'' Families were compensated for their time. Pulmonary Function Tests (PFT) from the last 3 months were obtained from the pulmonary clinic for each patient enrolled in the study, and these were reviewed by a board-certified pulmonologist.

The Mealtime Interaction Coding System (MICS)[21,22] was used to assess family functioning from the videotaped mealtime observation. Family functioning was coded by trained research assistants along six dimensions (task accomplishment, communication, affect management, interpersonal involvement, behavior control, roles) and overall family functioning on a scale ranging from 1 = ''very unhealthy'' to 7 = ''very healthy.'' Clinical cutoff scores have been developed for each dimension of the MICS.[21] Scores from 1 to 4 are considered ''unhealthy'' family functioning and scores from 5 to 7 are considered ''healthy.''[16]

Statistical analyses

Data analyses were performed using SPSS v20.0 (IBM Corp., Armonk, NY). Spearman correlations (ρ) were used to determine associations between SES, lung functioning, MICS scores, and TARS scores. Student's t-test analyses were used to assess for differences in MICS scores across child age dichotomized as school age (8–12 years) and adolescent (13–19 years). Spearman correlations were used to determine associations between family functioning and TARS scores.

Results

MICS ratings

Average ratings on each of the MICS dimensions and number of families falling in the ''unhealthy'' range for each scale can be found in Table 1. After enrolling in the

Table 2. Mean Standard Deviation MICS Scores Across Older and Younger Age Groups

	Younger age group (6–12 years) n = 9	*Older age group (13–19 years)* n = 7	t	df	p
Task accomplishment	4.78 (1.09)	5.14 (0.38)	−0.84	14	0.42
Communication	4.22 (1.56)	5.14 (0.69)	−1.44	14	0.17
Interpersonal involvement	4.44 (1.74)	5.00 (0.58)	−0.81	14	0.43
Behavior control	4.78 (0.97)	5.14 (0.38)	−0.94	14	0.37
Roles	4.89 (0.60)	5.29 (0.29)	−1.17	14	0.26
Affect regulation	4.11 (1.27)	5.00 (0.58)	−1.71	14	0.10*
Overall family functioning	4.67 (1.11)	5.14 (0.38)	−1.07	14	0.30

*$p \leq 0.10$.

study, three families declined to complete a mealtime taping. Demographics and disease-related indicators (e.g., severity) for families that completed the mealtime tape did not differ from families that did not complete a taping. All families rated the mealtime as typical (nine families) or very typical (seven families). Mean scores for all scales but Roles were <5.0 and in the ''unhealthy'' range. Family SES was associated with all subscales of the MICS except for Task Accomplishment (see Table 1).

Family functioning and child age

Table 2 presents mean MICS scores by dichotomized child age (8–12 years vs. 13–19 years). Although no significant differences at $p<0.05$ were found, mean levels of family functioning were higher on all scales among the older age group. There was a trend for scores on Affect Regulation to differ by child age ($t(14)=-1.71$, $p=0.10$), such that lower levels of family functioning related to affect regulation were found among families with a younger child with CF.

Family functioning and treatment adherence

Spearman correlations were conducted between the subscales of the MICS and parent report of the TARS (Table 3). The association between the Pancreatic Enzymes subscale and Roles subscale of the MICS was significant ($p=-0.53$, $p=0.04$). Several trends also emerged between other subscales of the MICS (e.g., Task Accomplishment, Interpersonal Involvement) and the Pancreatic Enzymes subscale, as well as between subscales of the MICS (Behavior Control, Overall) and the Antibiotics subscale. Better scores on family functioning were associated with worse scores on the Enzymes subscale and better scores on the Antibiotics subscale. Scores on the TARS were not significantly correlated with Hollingshead scores.

Discussion

The current study used a novel approach to understand family functioning and treatment adherence among children and adolescents with CF. Consistent with other studies,[15] family functioning was in the ''unhealthy'' range on all scales except the Roles subscale. Families may be more task-oriented at the expense of more emotional and interactional aspects of family functioning at mealtime.[15] Further, only one family scored in the ''unhealthy'' range on Task Accomplishment, suggesting that families with a child with CF may be more focused on mealtime tasks and activities during meals. Lower levels of family functioning were also present among lower SES families. SES may be associated with parenting behaviors, psychological stress, and disease burden, which may impact family functioning.[16,23,24]

Despite that fact that adolescence is often marked by attempts at increasing autonomy and challenging parental authority,[25] we found that family functioning in CF may improve as a child matures. As children carry the CF diagnosis from birth, families are required to respond to the burden of caring for an ill child and the demands of CF at an earlier point in the child's development than in other pediatric conditions. Despite our limited sample size, the magnitude of associations between family functioning and both enzyme and antibiotic adherence ($\rho=0.4$–0.5) suggest that family-level variables may be central to these two

TABLE 3. ASSOCIATIONS BETWEEN FAMILY FUNCTIONING (MICS), TREATMENT ADHERENCE RATING SCALE, AND CHILD AGE

TARS subscales	*Mean*	*Standard deviation*	*Task accomplishment* ρ, p	*Communication* ρ, p	*Interpersonal involvement* ρ, p	*Behavior control* ρ, p	*Roles* ρ, p	*Affect regulation* ρ, p	*Overall* ρ, p	*Child age* ρ, p
Airway clearance	4.47	0.54	0.15, 0.57	0.11, 0.69	0.16, 0.55	0.34, 0.19	0.15, 0.59	0.18, 0.51	0.36, 0.17	−0.30, 0.21
Aerosolized medications	4.63	0.57	0.09, 0.74	−0.09 0.73	0.10, 0.71	0.20, 0.45	0.00, 1.0	−0.02, 0.95	0.24, 0.38	−0.39, 0.10
Pancreatic enzymes	4.56	0.61	−0.49, 0.05*	−0.23, 0.39	−0.44, 0.09*	−0.43, 0.10	−0.53, 0.04**	−0.41, 0.12	−0.43, 0.10	−0.43, −0.10
Nutrition	4.48	0.77	−0.19, 0.49	−0.13, 0.64	−0.11, 0.69	−0.20, 0.49	−0.12, 0.66	−0.25, 0.37	−0.11, 0.71	−0.11, 0.71
Antibiotics	4.75	0.39	0.43, 0.15	0.45, 0.12	0.48, 0.10	0.51, 0.08*	0.27, 0.38	0.39, 0.19	0.53, 0.06*	−0.15, 0.59
TARS total	4.57	0.46	−0.24, 0.36	−0.10, 0.71	−0.10, 0.72	−0.02, 0.96	−0.27, 0.32	−0.15, 0.59	0.00, 1.0	−0.45, 0.06*

TARS, Treatment Adherence Rating Scale.
*$p<0.10$; **$p<0.05$.

components of adherence. Interestingly, the direction of associations between family functioning and these two aspects of adherence differed; future research is needed to confirm these findings in larger samples. Our findings might reflect the fact that mealtimes are often equated with illness burden for families, which is when most children take enzymes.[15] On the other hand, families may find it easier to plan ahead and adhere to an acute course of antibiotics versus chronic therapies.

The primary limitation of this study was its small sample size, which restricted the statistical power of this study, as well as the ability to generalize study findings. Although this study conceptualized adherence as an outcome measure, it is quite possible that adherence is predicting other outcome variables. Future research should utilize longitudinal samples to determine causality between family functioning and adherence, and to assess developmental considerations in associations with adherence. As SES-related disparities in CF outcomes have also been documented,[26] future studies should incorporate other measures of SES (e.g., income, insurance status) in understanding differences in family functioning by SES.

In conclusion, findings from this exploratory study advance our understanding of adherence in CF within the family context. Collectively, findings suggest that family functioning may be an important correlate of treatment adherence. Healthcare providers and clinicians may also wish to highlight the importance of the family in their treatment planning with adolescents with CF.

Acknowledgments

The authors wish to acknowledge the physicians, respiratory therapists, and staff at State University of New York (SUNY) Upstate for their assistance with the project, as well as the families that participated in the study.

Author Disclosure Statement

No competing financial interests exist. This study was funded in part by a traineeship grant from the Cystic Fibrosis Foundation to the first author (EVERHA08A0).

References

1. Mitchell MJ, Powers SW, Byars KC, Dickstein S, Stark LJ. Family functioning in young children with cystic fibrosis: Observations of interactions at mealtime. J Dev Behav Pediatr 2004; 25:335–346.
2. 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; 5:507–515.
3. Modi AC, Quittner AL. Barriers to treatment adherence for children with cystic fibrosis and asthma: what gets in the way? J Pediatr Psychol 2006; 31:846–858.
4. Patterson JM, Wall M, Berge J, Milla C. Associations of psychological factors with health outcomes among youth with cystic fibrosis. Pediatr Pulmonol 2009; 44:46–53.
5. Kazak AE, Rourke MT, Crump TA. Families and other systems in pediatric psychology. In: Roberts MC (ed.). Handbook of Pediatric Psychology. New York: Guilford, 2003:159–175.
6. Lewin AB, Heidgerken AD, Geffken GR, et al. The relation between family factors and metabolic control: the role of diabetes adherence. J Pediatr Psychol 2006; 31: 174–183.
7. Wiebe DJ, Berg CA, Korbel C, et al. Children's appraisal of maternal involvement in coping with diabetes: enhancing our understanding of adherence, metabolic control, and quality of life across adolescence. J Pediatr Psychol 2005; 30:167–178.
8. Drotar D, Bonner MS. Influences on adherence to pediatric asthma treatment: a review of correlates and predictors. J Dev Behav Pediatr 2009; 30:574–582.
9. Patterson JM, Budd J, Goetz D, Warwick WJ. Family correlates of a 10-year pulmonary health trend in cystic fibrosis. Pediatrics 1993; 91:383–389.
10. Quittner AL, Tolbert VE, Regoli MJ, Orenstein DM, Hollingsworth JL, Eigen H. Development of the role-play inventory of situations and coping strategies for parents of children with cystic fibrosis. J Pediatr Psychol 1996; 21: 209–235.
11. DeLambo KE, Ievers-Landis CE, Drotar D, Quittner AL. Association of observed family relationship quality and problem-solving skills with treatment adherence in older children and adolescents with cystic fibrosis. J Pediatr Psychol 2004; 29:343–353.
12. Fiese BH. Family Routines and Rituals. New Haven, CT: Yale University Press, 2006.
13. Crist W, McDonnell P, Beck M, Gillespie CT, Barrett P, Mathews J. Behavior at mealtimes and the young child with cystic fibrosis. Dev Behav Pediatr 1994; 15:157–161.
14. Stark LJ, Jelalian E, Powers SW, et al. Parent and child mealtime behavior in families of children with CF. J Pediatr 2000; 136:195–200.
15. Janicke DM, Mitchell MJ, Stark LJ. Family functioning in school-age children with cystic fibrosis: an observational assessment of family interactions in the mealtime environment. J Pediatr Psychol 2005; 130:179–186.
16. Spieth L, Stark LJ, Mitchell M, et al. Observational assessment of family functioning at mealtime in preschool children with cystic fibrosis. J Pediatr Psychol 2001; 26: 215–224.
17. Janicke DM, Mitchell MJ, Quittner AL, Piazza-Waggoner C, Stark LJ. The impact of behavioral intervention on family interactions at mealtime in pediatric cystic fibrosis. Child Health Care 2008; 37:49–66.
18. DiMatteo MR, Giodani PJ, Lepper HS, Croghan TW. Patient adherence to medical treatment outcomes: a meta-analysis. Med Care 2002; 40:784–811.
19. Quittner AL, Drotar D, Ievers CE. Treatment Adherence Rating Scale: unpublished measure, 1998.
20. Hollingshead A. Four-Factor Index of Social Status. New Haven, CT: Department of Sociology, Yale University, 1975.
21. Dickstein S, Hayden LC, Schiller M, Seifer R, San Antonio W. Providence Family Study mealtime family interaction coding system. Adapted from the McMaster Clinical Rating Scale. East Providence, RI: E.P. Bradley Hospital, 1994.
22. Epstein NB, Baldwin LM, Bishop DS. The McMaster Family Assessment Device. J Marital Fam Ther 1983; 9: 171–180.
23. Bornstein M, Bradley R. Socioeconomic Status, Parenting, and Child Development. Mahwah, NJ: Lawrence Erlbaum, 2003.

24. Holmbeck GN, Bruno EF, Jandasek B. Longitudinal research in pediatric psychology: an introduction to the special issue. J Pediatr Psychol 2006; 31:995–1001.
25. Allison BN, Schultz JB. Parent–adolescent conflict in early adolescence. Adolescence 2004; 39:101–119.
26. Schechter MS, McColley SA, Regelmann W, et al. Socioeconomic status and the likelihood of antibiotic treatment for signs and symptoms of pulmonary exacerbation in children with cystic fibrosis. J Pediatrics 2011; 159:819–824.e811.

Address correspondence to:
Robin S. Everhart, PhD
Department of Psychology
Virginia Commonwealth University
P.O. Box 842018
Richmond, VA 23284

E-mail: reverhart@vcu.edu

Received for publication January 3, 2014; accepted after revision February 24, 2014.