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Health-Related Quality of Life in Children and Adolescents with Cystic Fibrosis: Convergent Validity with Parent Reports and Objective Measures of Pulmonary Health

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Abstract

Objective—This study examined the convergent validity of health-related quality of life (HRQOL) reported by patients with cystic fibrosis compared with their parents' reports and objective pulmonary measures across three time points.

Methods—Ninety-two children (8–13 years) and adolescents (14–18 years) with CF and their parents completed Cystic Fibrosis Questionnaires to examine concordance with Wisconsin Chest X-Ray (WCXR) scores and pulmonary function tests, e.g., forced expiratory volume at one second (FEV1), and parent-child/adolescent concordance across multiple HRQOL domains. Concordance was analyzed relative to patient age and gender.

Results—Parent reports were closely aligned with WCXR scores, while patient reports were more closely aligned with FEV1. Adolescents and parents of both age groups had more HRQOL domains concordant with pulmonary health measures than did child self-reports. Parent-child concordance was inversely related to child age, particularly with male adolescents. Children generally reported better HRQOL than parents. Male adolescents and their parents were more likely to have significantly discordant HRQOL scores than female adolescents and their parents. Male and female adolescents reported higher HRQOL than their parents reported for all but Vitality and Health Perception domains. Younger male children showed concordance with their parents on five of seven domains.

Conclusions—Parent-child/adolescent discordance on HRQOL was consistent with normative child development expectations. Findings underscore the value of enlisting perspectives from parents as well as children regarding HRQOL.

Patient-reported outcome (PRO) measures inform medical interventions and evaluate their effectiveness.¹ Health-related quality of life (HRQOL) instruments represent PROs that

measure perceived health of patients with cystic fibrosis (CF) and provide clinicians and scientists valuable disease-specific information across multiple functional dimensions.² However, when patients are children, it is unclear whether children or parents are more appropriate and reliable informants. Although children with CF are typically encouraged to recognize symptoms and increasingly assume responsibilities for self-care, child development theory and research suggest their emergent cognitive and psychosocial capacities still put them in need of parent proxies for medical care, decision-making, and research.³ This study compared the concordance of parent proxy-reports and child self-reports on HRQOL for children with CF relative to objective pulmonary measures across three time points.

Parent-Child Concordance

Research across clinical populations, e.g., asthma, attention deficit hyperactivity disorder, cancer, central nervous system problems, chronic pain, CF, dermatitis, diabetes, epilepsy, eye or ear problems, heart disease, obesity, rheumatoid problems, and healthy pediatric samples shows mixed findings concerning domains of function on which parents and their children agree. Some studies show greater parent-child concordance for observable physical versus psychosocial HRQOL domains.⁴⁻⁷ Others report higher levels of parent-child agreement for psychosocial domains.⁸⁻⁹ Yet another study of healthy children and their parents shows significant parent-child discordance on physical and psychosocial domains. Parents tend to perceive their children's HRQOL to be higher than children's reports.¹⁰ However, children with CF report higher overall HRQOL compared to their parents' proxy appraisals; while parents of children with CF tend to report greater treatment burden than their children report.¹¹⁻¹² Objective measures of pulmonary health and frequency of pulmonary exacerbations are associated with HRQOL for patient-reported illness-specific symptoms, general health, physical functioning, and pain.¹¹⁻¹⁴ One study shows no relationship between changes in pulmonary status and changes in patient-reported HRQOL over time,¹⁴ whereas another shows changes in patient-reported respiratory symptoms and weight congruent with changes in related HRQOL.¹⁵

Gender and age also may play a role in children's and adolescents' perceptions about HRQOL. Research shows that female adolescents with CF¹⁶ as well as those in the general population¹⁷ tend to report lower HRQOL than their male counterparts. These gender differences in HRQOL have been associated with more severe pulmonary disease among female adolescent and adults with CF.¹⁸ Additionally, differences between children and adolescents on self-reported HRQOL have been documented in normative samples.¹⁷ For example, younger children are more likely to report digestive problems than adolescents. By contrast, body image may more likely to become an issue during adolescence than childhood. Similarly, age differences have been found in CF populations. Children with CF in late childhood (ages 8 to 12 years) report less favorable HRQOL in the HRQOL domains of Psychosocial, Social, Emotional, and School.¹⁹ However, a study of pain shows no associations between child age, gender and parent-child concordance.²⁰ Thus, the influence of age and gender of perceived HRQOL remains unclear.

Given the conflicting reports from previous research, designed a study to test the following hypotheses: (a) parent reports would have greater concordance with objective pulmonary measures than child reports; that parent-child concordance would (b) be higher on domains associated with observable symptoms, and (c) show adolescent gender differences. Based on theories of child development we predicted that parent-child concordance would (b) be inversely related to child age.

METHODS

Design

This study was conducted within the Wisconsin Newborn Screening (NBS) Project, a longitudinal evaluation of the benefits and risks of NBS for CF.²¹ The enrollment period for the original study, conducted from 1985–1998, involved random assignment of newborns into either an early diagnosis or control group. Control group participants were diagnosed by traditional methods or through un-blinding the child's screening results at age 4 years per study protocol. Parents of 145 children with CF, confirmed by sweat chloride levels of ≥ 60 mmol/L, provided written informed consent to enroll their children into the randomized controlled trial (RCT). Six children were excluded as not meeting the criteria of having a NBS specimen collected and analyzed within 28 days after birth. Patients received regular evaluations and treatment through a study protocol based on CF standard of care. Details about the RCT can be found elsewhere.^{21–23}

Previously enrolled children with CF between 8 and 18 years were invited to participate in the current HRQOL study. Parents of minors and adult patients provided written informed consent. Patients age 14 to 17 provided assent in addition to parental written informed consent. From 2002–2006, we collected data for each patient yearly at the beginning of three regularly scheduled clinical evaluations at one of two CF Centers. Mean time between first and second evaluations was 1.1 years (SD=0.3) and between second and third was 1.0 year (SD=0.3). Health Sciences Institutional Review Boards at UW-Madison, Medical College of Wisconsin, and Children's Hospital of Wisconsin approved both studies (#1983–270 and #1998–105).

HRQOL Concordance Sample

Measures

Pulmonary Health: Pediatric pulmonologists and radiologists blinded to patient study groups independently scored chest radiographs using the Wisconsin Chest Radiograph System.²⁴ Total Wisconsin Chest X-Ray (WCXR) scores were derived from six components: hyperinflation, peribronchial thickening, bronchiectasis, nodule/branching opacities, large round/ill-defined opacities, and atelectasis. Scores ranged from 0 (normal/no disease) to 100 (severe disease). PFTs were routinely performed every six months. Pediatric Alternate Spirometry System (PASS)²⁵ provided quality assurance for PFT data. Analyses included FEV1 data collected closest to the dates of completed HRQOL measures. We used baseline pulmonary data, not associated with pulmonary exacerbations. All outcome measures were subject to rigorous quality control.^{24–25}

Health-Related Quality of Life: Four versions of the Cystic Fibrosis Questionnaire (CFQ) measure disease-specific HRQOL.^{27–28} CFQ-Child for ages 6–11 (interview) and ages 12–13 (self-report) contains 35-items within eight domains. CFQ-Teen/Adult for ages 14 years (self-report) contains 48-items within twelve domains. CFQ-Parent (perceptions of child's HRQOL) contains 44-items within eleven domains. In this study, parents and children ages 8–13 years completed seven domains common to CFQ-Parent and CFQ-Child. Parents and children ages 14 years completed ten domains common to CFQ-Parent and CFQ-Teen/Adult. CFQ-Parent does not include Social, therefore this domain was excluded from analysis. Four-point Likert scales rate the difficulty, frequency, or applicability of items. Other items use true-false responses. Raw scores were standardized from 0 (poor) to 100 (excellent) QOL. Ceiling effects have been noted for the Eating, Digestive, and Body Image domains for both child versions.^{4, 27}

Statistical Analyses

CFQ-Child data for ages 6–11 and 12–13 years were combined because they contain the same questions. Mixed-effects Tobit regression analysis measured cross-sectional concordance between parent and child HRQOL reports, as well as relationships between HRQOL and pulmonary health (WCXR, $n=88$; FEV1, $n=89$). CFQ domain scores were dependent variables in each model and treated as left-censored at 0 and right-censored at 100. We used Mixed-effects Tobit regression to compensate for ceiling effects on all but CFQ Vitality, and floor effects on Weight.

We assessed parent-child reporting concordance by including indicators for parent versus child report, CFQ-Child versus CFQ-Teen/Adult version, and their interaction in the model. Differences in relationships between pulmonary health and HRQOL between parent and child reports were assessed by including three-way interactions between reporter type, patient CFQ version, and pulmonary health. Each model included random intercepts for correlations between multiple parent and child observations for each patient. Gender differences were assessed by including additional interactions with patient gender in each model. We analyzed each CFQ domain and pulmonary health measure separately. Because they were non-normally distributed, WCXR scores were included as their base-2 log to ease interpretation of the results.

We selected a p -value of <0.012 for statistical significance to address multiple comparisons and preserve an overall statistical significance level of 0.05. This value was determined by assessing the average correlation between the eleven CFQ domains and using this to modify a standard Bonferroni adjustment.²⁸ Reported 95% confidence intervals were adjusted for multiple comparisons.

RESULTS

Sample Characteristics

The current study included 92 patients (mean age=13.3 years; $SD=2.7$) who participated in the original RCT. For this analysis, patient and parent participants in both the early diagnosis group and the control group were combined. As shown in Table 1, patients were predominately white (96%) and male (59%); parents were well-educated and most were married. Based on a comparison of data found in the Cystic Fibrosis Registry for Clinical characteristics of patients in this study were similar in demographic and clinical characteristics as patients with CF throughout the United States.^{27, 29}

Internal Consistency

In our study, internal consistency within CFQ domains was measured using Cronbach's alpha. Except for Treatment (0.48), reliability coefficients on CFQ-Child domains were good (0.71–0.79). Most CFQ-Teen/Adult domains had favorable reliability (0.71–0.94). Eating (0.59) and Digestion (0.59) were marginal. Most of the CFQ-Parent domains had favorable reliability (0.73–0.94). The Digestion (0.64) and Treatment Burden (0.61) domains were marginal. These results were similar to previous reports.^{4, 26, 27}

HRQOL Concordance Sample

All 92 patients completed the first CFQ; 73 (79%) completed the second, and 70 (76%) completed the third. Most (97%) completed at least two CFQs; 85% completed all three. Analyses were restricted to time points for which both parent and child reports were available. Eighty-eight patients had valid WCXR data and eighty-nine had valid PFT data for at least one of three times, while 10.7% of potential WCXR scores and 16.7% of potential PFT scores were missing. CFQ data were missing due to parent or child attrition.

Reasons for missing pulmonary data included no WCXR when the CFQ was completed or vice versa, inadequate or no PFT to match CFQ or vice versa. Table 1 shows sample characteristics.

The current study sample was more likely to be white than the non-responders from the larger RCT sample ($\chi^2=4.8$, $p=0.0282$). None of the four African-American children who were participants in the larger RCT study took part in this study. Parents of children in the full RCT sample who did not take part in the current study had less education than those of children who did so (Mother's Education: $\chi^2=12.5$, $p=0.0058$; Father's Education: $\chi^2=15.2$, $p=0.0017$). The genotype of the current sample differed from that of the non-responders ($\chi^2=9.3$, $p=0.0096$), primarily because 3/4 of the RCT participants who had a genotype other than DF508/DF508 or DF508/Other did not participate in the current study. The current sample was more likely to have the DF508/DF508 (55% vs 44%) or DF508/Other (40% vs 35%) genotype, and less likely to have another type (4% vs 21%).

Pulmonary Function Tests

The coefficients presented for FEV1 can be interpreted as the expected difference in the CFQ domain score between two patients whose FEV1 scores differed by one point. For example, the results in Figure 1, Panels 1a and 1b indicate that a patient with a FEV1 score of 90% would be expected to have a parent-reported Weight score that is 1.5 points higher than a patient with an FEV1 score of 89% and a parent-reported Weight score that is 15 points higher than a patient with an FEV1 score of 80%. FEV1 is related to similar Child-CFQ and Parent-CFQ domains as was WCXR with one exception: Body Image. Both Child-CFQ and Parent-CFQ were positively related to Physical Functioning (Child Coeff=0.37, 95% CI: 0.10, 0.64; Parent Coeff: Coeff=0.43, 95% CI: 0.16, 0.69) and Respiratory Symptoms (Child: Coeff=0.18, 95% CI: 0.01, 0.35; Parent Coeff=0.20, 95% CI: 0.03, 0.37). In addition, FEV1 was positively associated with the parent-report of their children's Weight (Coeff=1.54, 95% CI: 0.71, 2.38) and Health Perceptions (Coeff=0.50, 95% CI: 0.20, 0.80), but not Body Image. However, Child self-report of Body Image (Coeff=0.53, 95% CI: 0.12, 0.95) was positively related to FEV1. These results stand in contrast to those using WCXR, in which parent-report of child Body Image was related to pulmonary health, but child report of Body Image was not. The CFQ-Child did not include Health Perceptions or Weight domains.

Chest Radiographs

Figure 1 displays HRQOL relationships with objective measures of pulmonary health. The coefficients presented for WCXR shown in Panels 1c and 1d can be interpreted as the expected difference on the CFQ domain score between two patients when one patient's WCXR score is double that of the other. For example, the results for the Weight domain indicate that the parent-reported Weight score of a patient who has a WCXR score of 10 will be 23.3 points lower than that of a patient with a WCXR score of 5. WCXR was negatively related to the child self-report (N=50) on the Physical Functioning (Coeff: -7.30, 95% CI: -11.58, -3.02) and Respiratory Symptoms (Coeff: -4.42, 95% CI: -7.34, -1.49) domains. Among parents of patients who completed the CFQ-Child (N=50), WCXR was negatively related to parent-reported HRQOL on these measures, as well as Weight (Coeff: -23.32, 95% CI: 36.70, -9.93), Body Image (Coeff: -8.08, 95% CI: -14.85, -1.31), and Health Perceptions (Coeff: -7.23, 95% CI: -12.04, -2.41) domains. CFQ-Child version does not include patient report of Health Perceptions or Weight. Because of this, only the relationship between WCXR and Body Image differed significantly between the CFQ-Child and CFQ-Parent ($p<0.001$).

Among both patients who completed the CFQ-Teen/Adult and their parents (N=56), WCXR was negatively related to HRQOL on Weight (Teen Coeff: -13.89, 95% CI: -25.76, -2.02; Parent Coeff: -17.32, 95% CI: -29.26, -5.37), Health Perceptions (Teen Coeff: -6.30, 95% CI: -10.72, -1.88; Parent Coeff: -7.05, 95% CI: -11.48, -2.62), Physical Functioning (Teen Coeff: -4.28, 95% CI: -8.34, -0.21; Parent Coeff: -6.42, 95% CI: -10.48, -2.36), Respiratory Symptoms (Teen Coeff: -3.74, 95% CI: -6.43, -1.06; Parent Coeff: -4.61, 95% CI: -7.30, -1.93), and Vitality (Teen Coeff: -2.79, 95% CI: -5.35, -0.23; Parent Coeff: -2.80, 95% CI: -5.35, -0.24). Though the relationship between WCXR and the parent-report on the Physical Functioning domain was stronger than the relationship between WCXR and adolescent self-report ($p < 0.001$), both teens and parent reports are similarly related to the teen's pulmonary health.

Figures 2 shows these analyses of HRQOL and WCXR scores by the gender of the child. The results are largely similar for male and female children. Despite these similarities, there is one domain on which male and female children differ: female children's parent-reports of Eating Disturbance were significantly related to WCXR (Coeff: -9.12, 95% CI: -17.06, -1.19). This was not true for male children's parent-reports or either group's self-reports, and the difference between the relationship between WCXR and the CFQ-Parent and the female CFQ-Child is statistically significant ($p < 0.05$).

Male and female adolescent self-reports were similar on most domains, though WCXR was related to the Physical Functioning (Coeff: -5.42, 95% CI: -9.97, -0.87) and Vitality (Coeff: -6.48, 95% CI: -11.00, -1.96) domains only among female adolescents. Both male and female adolescent self-reports were negatively related to WCXR. Parent-reports showed few differences between parents of male and female adolescents, though the relationship between Weight and WCXR was slightly stronger for parents of male adolescents (Male = -19.92, 95% CI: -32.27, -7.56; Female = -13.45, 95% CI: -26.21, -0.70), while like female adolescents, only parents of female adolescents showed a relationship between Vitality and WCXR (Coeff = -3.30, 95% CI: -6.08, -0.52). Overall, there were few differences by gender.

There were more differences between WCXR and FEV1 on their relationship with adolescent CFQ reports. Though FEV1 was positively related to both adolescent and parents of adolescent HRQOL on the Weight (Teen Coeff = 2.17, 95% CI: 1.28, 3.07; Parent Coeff = 1.89, 95% CI: 1.01, 2.76), Health Perceptions (Teen Coeff = 0.57, 95% CI: 0.26, 0.88; Parent Coeff = 0.53, 95% CI: 0.22, 0.84), Physical Functioning (Teen Coeff = 0.57, 95% CI: 0.29, 0.85; Parent Coeff = 0.42, 95% CI: 0.14, 0.69), and Eating Disturbance (Teen Coeff = 0.70, 95% CI: 0.21, 1.19; Parent Coeff = 0.50, 95% CI: 0.01, 0.98). In addition, adolescent self-report of Body Image (Coeff = 0.58, 95% CI: 0.15, 1.02) and Respiratory Symptoms (Coeff = 0.23, 95% CI: 0.06, 0.40) were positively related to FEV1, though parent reports on these measures were not. Adolescent reports were more strongly related to FEV1 than parent reports on all of these domains except Health Perceptions.

Figures 3 shows these analyses of HRQOL and FEV1 by the gender of the child. For CFQ-Child, only the Respiratory Symptoms domain showed child gender difference for the relationship between FEV1 and HRQOL. Female child self-report (Coeff: 0.21, 95% CI: 0.04, 0.39) and parent-report of their male children's HRQOL (Coeff: 0.19, 95% CI: 0.00, 0.39) on Respiratory Symptoms were significantly aligned with FEV1. Relationships between FEV1 and HRQOL did not differ by gender of the adolescent for the adolescent-reported HRQOL on most domains. However, the relationship between parent-report for female children and FEV1 was slightly stronger on Eating Disturbance (Coeff: 0.52, 95% CI: 0.01, 1.03) and Respiratory Symptoms (Coeff: 0.19, 95% CI: 0.01, 0.36) than that for male children, so that these relationships were only significant for female adolescents.

Parent-Child Concordance

The coefficients in Figure 4 represent the difference between the patient's self-reported HRQOL and his/her parent-reported HRQOL. A positive coefficient indicates that a patient self-reported better HRQOL than did his/her parent. Of the seven domains common to both the CFQ-Child and CFQ-Parent, patient self-reports and parent-reports were similar on five domains. Children reported higher/better HRQOL for Digestive Symptoms (Coeff: 12.06, 95% CI: 5.33, 18.79) and Body Image (Coeff: 25.88, 95% CI: 16.19, 35.57) than their parents did for them. Both male and female children reported better HRQOL on Body Image (Male: 23.33, 95% CI: 11.34, 35.31; Female: 30.11, 95% CI: 14.40, 45.81), however, only females reported better HRQOL than their parents reported for them on Digestive Symptoms (Coeff: 20.52, 95% CI: 9.56, 31.49) and Eating Disturbance (Coeff: 18.19, 95% CI: 0.47, 35.91). Males reported worse HRQOL on Emotional Functioning (Coeff: -7.36, 95% CI: -13.44, -1.27) than their parents reported for them. The difference in discordance between the male-female child CFQs on the Emotional Disturbance domain was significant ($p < 0.05$).

More differences were present between CFQ-Teen/Adult and CFQ-Parent reports. Of the ten domains common to both CFQ-Teen/Adult and CFQ-Parent, adolescent and parent reports differed on eight domains. Adolescents consistently reported their HRQOL as higher than their parents' reports on Weight (Coeff: 23.28, 95% CI: 7.10, 39.45), Body Image (Coeff: 16.02, 95% CI: 6.95, 25.09), Digestive Symptoms (Coeff: 20.24, 95% CI: 13.66, 26.82), Eating Disturbance (Coeff: 14.17, 95% CI: 2.14, 26.19), Physical Functioning (Coeff: 9.99, 95% CI: 4.75, 15.24), Treatment Burden (Coeff: 7.98, 95% CI: 2.72, 13.23), Emotional Functioning (Coeff: 6.02, 95% CI: 1.39, 10.66), and Respiratory Symptoms (Coeff: 4.50, 95% CI: 0.25, 8.74). Despite this, there were many differences by gender, which showed that most of these differences were driven by male adolescents. Adolescent males reported higher HRQOL on Weight (Coeff: 35.53, 95% CI: 14.29, 56.77), Body Image (Coeff: 24.47, 95% CI: 11.98, 36.96), Digestive Symptoms (Coeff: 19.53, 95% CI: 9.56, 31.49), Eating Disturbance (Coeff: 18.42, 95% CI: 2.48, 34.37), Physical Functioning (Coeff: 14.76, 95% CI: 7.52, 22.00), Treatment Burden (Coeff: 10.07, 95% CI: 2.98, 17.17), and Respiratory Symptoms (Coeff: 7.31, 95% CI: 1.60, 13.01). Adolescent females reported significantly higher HRQOL on Digestive Symptoms (Coeff: 20.94, 95% CI: 11.42, 30.46) and Emotional Functioning (Coeff: 7.67, 95% CI: 0.94, 14.40) than their parents.

DISCUSSION

This study extended previous research^{4–20} by documenting parent-child discordance relative to gender, age, and objective health measures of patients with CF over time. Our hypothesis that parent-reports of their child's HRQOL would have greater concordance with objective health measures than the child's self-report was partially supported. Domains most strongly associated with both objective measures included Weight, Physical, Health, and Respiratory. Overall, parent reports were more closely aligned with WCXR, while patient reports were more closely aligned with PFT, particularly in adolescents. These findings might be explained by the type of information yielded by PFTs versus chest radiographs. PFTs are sensitive to the patient's current pulmonary status which the child/teen subjectively experiences, whereas WCXR scores provide information about the patient's overall pulmonary health, which parents are more likely to observe. Across age groups and respondents, objective measures of health were more strongly associated with physical aspects of HRQOL than psychosocial domains. Lack of associations between objective pulmonary measures and self-report measures on Emotional and Treatment Burden suggest factors other than pulmonary health, e.g., adaptation, influence those aspects of HRQOL. Overall, these results are consistent with previous studies showing patients with CF report better HRQOL than their parent proxies.^{11–12}

Contrary to previous findings⁴⁻⁷ and our hypothesis, children's age group was more salient to parent-child concordance than observable vs. non-observable HRQOL domains, e.g., Digestive (observable) and Body Image (non-observable) showed similar levels of disagreement between parents and their children or adolescents. Our hypothesis that parent-child HRQOL concordance would be inversely related to child age was supported by results showing that parents and adolescents differed on eight of ten common domains, while parents and younger children differed on only two of seven common domains. The most striking parent-adolescent difference was on Weight, whereas the greatest parent-child difference was on Body Image. Children and adolescents self-reported better HRQOL than did their parents. Findings suggest parents are more concerned about nutrition-related domains than their children.

Our hypothesis that significant gender differences would emerge with regard to parent-child agreement was supported. Female adolescents demonstrated better agreement with their parents than male adolescents for seven of ten domains. However, male and female adolescents reported better HRQOL than their parents reported for all domains. This pattern was not evident for younger children, with males demonstrating better agreement with their parents on five of seven domains. Interestingly, despite previous reports and our findings that patients tend to self-report better HRQOL than their parents, younger male children in this study reported worse HRQOL than their parents on the Eating Disturbances, Physical Functioning, Respiratory Symptoms, and Emotional Functioning. This finding warrants further study of gender differences in coping with CF in middle childhood. However, gender was not a significant variable associated with objective pulmonary measures. The significant association between parent proxy reports of Eating Disturbance in their daughters, but not sons, and WCXR suggests parents, most of whom were mother in this study, may watch their daughters' eating habits more closely than their sons. Finally, our finding of no significant gender differences on HRQOL patient self-reports also differs from previous findings that show females tend to report overall lower HRQOL than males.^{16,18,29}

Consistent with previous studies, these results confirm the utility of the CFQ, a disease-specific QOL instrument, for measuring perceptions of health status, particularly respiratory symptoms and physical functioning across informants and age groups.¹¹ Our findings also highlight the importance of considering patient developmental level and gender when interpreting HRQOL data. We agree with Upton and colleagues⁴ that "parents and children contribute different but valid information" to the assessment of HRQOL. A comprehensive appraisal of HRQOL in research and clinical practice should include both patient and parent reports.

Limitations include using different CFQ versions with different items and domains, thus limiting the comparisons for some domains. The small size of the subsamples could have affected some results, e.g., lack of significant relationships between FEV1 and Respiratory on the CFQ Parent reports of female children. Most patients (69%) had normal PFT results at the first data collection which also could have affected the relationship between the results. Sample homogeneity regarding racial/ethnic background and relatively good health of patients could have affected results.

Findings from this study also point to several areas that merit additional investigation. The lack of significant associations between objective health measures and patient-reported emotional well-being and perceptions of treatment burden suggest that other factors may influence these subjective experiences. Future studies might combine qualitative interviews with HRQOL questionnaires to better elucidate adaptation strategies that patients and family employ to preserve their psychological well-being in the face of progressive health conditions, like CF, that require increased treatment demands over time. Some of the mixed

finding within the QOL literature may be attributed to the mix of generic and disease-specific measures used in studies. In this study CFQ, a disease-specific questionnaire, was particularly useful in detecting issues salient to the CF population, such as parents' focus on their younger children's nutrition and eating disorders in young girls. Additional research is needed to advance the science of HRQOL through the development of disease specific instruments for patients and their parents that are sensitive to differences in gender and the child development.

CONCLUSIONS

Parent-child/adolescent discordance on HRQOL was consistent with normative child development expectations. Findings underscore the value of enlisting perspectives of parents as well as their children regarding HRQOL. Additional research is needed to continue to refine HRQOL instrumentation to better identify factors that contribute to QOL within and across clinical populations.

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Figure 1. Relationship between CFQ and WCXR/FEV1
 Panel a and c: Black: Child (WCXR, N=50; FEV1, N=50)
 Panel a and c: Light Grey: Parent (WCXR, N=50; *FEV1, N=50)
 Panel b and d: Dark Grey: Teen (WCXR, N=55; FEV1, N=56)
 Panel b and d: White: Parent (WCXR, N=55; FEV1, N=56)

Figure 2. Gender Differences in Relationship between CFQ and WCXR
 Panel 2a: Black: Male Child (N=31); Light Grey: Parent (N=31)
 Panel 2b: Black: Female Child (N=19); Light Grey: Parent (N=19)
 Panel 2c: Dark Grey: Male Teen (N=32); White: Parent (N=32)
 Panel 2d: Dark Grey: Female Teen (N=24); White: Parent (N=24)

Figure 3. Gender Differences in the Relationship between CFQ and FEV1

Panel 3a: Black: Male Child (N=31); Light Grey: Parent (N=31)

Panel 3b: Black: Female Child (N=19); Light Grey: Parent (N=19)

Panel 3c: Dark Grey: Male Teen (N=30); White: Parent (N=30)

Panel 3d: Dark Grey: Female Teen (N=25); White: Parent (N=25)

Figure 4. Discordance between Parent and Child CFQ Reports

Panel 4a: Black: Child (N=53); White: Teen CFQ (N=61)

Panel 4b: Black: Male Child (N=33); Light Grey: Female Child (N=20)

Panel 4c: Dark Grey: Male Teen (N=32); White: Female Teen (N=29)

Table 1

Sample Characteristics

Demographic Characteristics		CF Risk Factors	
	N	%	
Child			
Gender			Pseudomonas Status at Time 1
Male	54	58.7%	Nonnuroid
Female	38	41.3%	Absent
Race/Ethnicity			Intermittent
White	88	95.7%	Persistent
Hispanic	2	2.2%	Mucoid
Native American	1	1.1%	Absent
Missing	1	1.1%	Intermittent
Center			Persistent
Madison	47	51.1%	Genotype
Milwaukee	45	48.9%	DF508/DF508
Parent			DF508/Other
Reporting Parent			Other
Mother	77	83.7%	Meconium Ileus
Father	15	16.3%	Yes
Education			No
Less than High School	6	6.5%	Pancreatic Sufficiency
High School Degree	28	30.4%	Sufficient
Some College	31	33.7%	Insufficient
College Degree	27	29.3%	WCXR Score Group at Time 1
			Mild Disease
			Moderate
			Moderate-Severe
			Severe
			Missing
			FEV1 Score Group at Time 1
			Normal
			Mild Disease

Demographic Characteristics		CF Risk Factors	
N	%	N	%
		Moderate	6 6.5%
		Moderate-Severe	4 4.3%
		Severe	1 1.1%
		Missing	7 7.6%