Adolescents With Cystic Fibrosis: Family Reports of Adolescent Health-Related Quality of Life and Forced Expiratory Volume in One Second

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ABSTRACT. Purpose. To assess the relationship between adolescent and parent reports of adolescent health-related quality of life (HRQL) and between adolescent pulmonary function (forced expiratory volume in 1 second as percent of predicted) and reporter perceptions of adolescent health.

Methods. Twenty-four adolescents with cystic fibrosis (CF), their mothers, and their fathers completed the Child Health Questionnaire during routine CF clinic visits at 2 urban hospitals. Patients were between the ages of 11 and 18 years (mean age: 14.2 years) and were predominantly male (75%). The best measure of forced expiratory volume in 1 second as percent of predicted for the year of the study was also collected for each adolescent.

Results. Adolescent pulmonary function was related to the perceived adolescent physical health scales. It was not, however, associated to perceptions of adolescent emotional, social, or behavioral HRQL by any of the 3 family reporters. Associations were found between adolescent pulmonary function and self-reports of general health (0.73), role/social limitations-physical (0.47), and bodily pain (0.42). Adolescent pulmonary function was related to mother reports of adolescent general health (0.73), role/social limitations-physical (0.73), bodily pain (0.55), and physical functioning (0.70). Father perceptions of adolescent health were associated to adolescent pulmonary function on general health (0.54), role/social limitations-physical (0.60), and physical functioning (0.64). Associations between adolescent and parent perceptions of adolescent HRQL were also health scale-specific. Mother and child reports of adolescent HRQL were related on adolescent behavior problems (0.71) and role/social limitations attributable to behavior (0.48), role/social limitations attributable to physical (0.62), bodily pain (0.69), physical functioning (0.69), family activities (0.45), and general health (0.66). Associations were found between father and adolescent reports on perceived adolescent behavior problems (0.66); self-esteem (0.65); and role/social limitations attributable to physical (0.49), general health (0.61), and perceived mental health (0.48).

Conclusions. Results demonstrate the need to include multiple informants and comprehensive, multidimensional measures of HRQL, in addition to pulmonary function, when assessing health in adolescents with CF. Pediatrics 2001;107(5). URL: http://www.pediatrics.org/cgi/content/full/107/5/e70; cystic fibrosis, health-related quality of life, pediatrics, pulmonary function, forced expiratory volume in 1 second.

ABBREVIATIONS. CF, cystic fibrosis; FEV₁, forced expiratory volume in 1 second; HRQL, health-related quality of life; CHQ, Child Health Questionnaire; PCHQ, Parent Child Health Questionnaire; SD, standard deviation.

Pulmonary function is one of the traditional parameters of health status for patients with cystic fibrosis (CF). Forced expiratory volume in 1 second (FEV₁) is considered a reliable pulmonary function surrogate indicator for disease progression and projected survival.1 As such, it has been widely used in clinical work and research on children, adolescents, and adults with CF.2– 4 Although pulmonary function tests, along with other physical parameters, are commonly used to monitor health status, biomedical measures by themselves do not provide the patient or clinician adequate information about the impact of disease on the daily functioning of adolescents with CF.

CF may have considerable impact on a person’s health-related quality of life (HRQL), defined as the “subjective and objective impact of dysfunction associated with an illness, injury, medical treatment, and health care policy.”5 Conceptualization of HRQL supports a subjective, multidimensional, and comprehensive model of health. This represents an expanded view of the traditional medical model that assesses health primarily through physical outcomes.

There is growing interest in the inclusion of HRQL outcome measures to “evaluate differential changes in morbidity and the relative efficacy of medical interventions.”5 Although HRQL assessments may be useful to patients and professionals when making clinical decisions, evaluating efficacy of treatments, or when planning psychosocial interventions, HRQL has been neglected in clinical practice and research on children and adolescents with CF.

Only a few studies have examined the relationship between biomedical factors and HRQL in children or adolescents with CF.2,4,6,7 Some of these studies report a strong correlation between HRQL and disease severity, as measured by FEV₁6,7 whereas others have found that no such relationship exists.2,4 In addition, little is known about the relationships between different family members as sources of information concerning the health of adolescents with CF. Mothers have been the primary reporters of child...
and adolescent health for research on families and CF, although fathers have been reported to share responsibility for their child’s care and adolescents have been found to be competent reporters of their own health. Past research on child and parent perceptions of HRQL in adolescents with CF has shown that parent and child reports of child health are not interchangeable. Fathers and adolescents have been underrepresented in the CF literature; however, each may offer a unique perspective of adolescent health.

The average life expectancy for a patient with CF has greatly increased over the last 3 decades. This increase in expected lifespan brings with it new and different challenges for children with CF and their families. Perceived adolescent health status is especially important as these children and their parents negotiate normal issues of adolescent development, such as gaining independence from parents, identity formation, and increased involvement with peers. The purpose of this study was to investigate the relationship between adolescent, maternal, and paternal perceptions of adolescent HRQL, and the relationship between reporter perceptions of adolescent HRQL and health status as measured by pulmonary function (FEV\textsubscript{1} % predicted) in adolescents with CF.

METHODS

Participants

Twenty-four adolescent, mother, and father triads were selected, based on complete family responses, from a larger convenience sample that included 32 adolescents, 32 mothers, and 25 fathers. English-speaking adolescents with CF ages 11 to 18 years and their parents were recruited from consecutive routine visits to the CF clinics at 2 Northeast medical centers. Adolescents demonstrated a wide range of pulmonary function (FEV\textsubscript{1} % predicted).

Study Design

Approval was obtained from the institutional review board at each hospital before patients and their accompanying parent(s) were recruited at 2 medical centers in Massachusetts. A complete explanation of the study was presented to the adolescent and the accompanying parent. Individual parent consent and adolescent assent forms were signed. Each participant was given a separate, self-addressed envelope in which to return the completed questionnaire to the investigator. An additional questionnaire and mailing envelope was brought home to any nonaccompanying parent for completion. The child and parent reports of the Child Health Questionnaire (CHQ) are self-administered. The importance of completing the measure independently was stressed. It was also explained that psychological support would be available, through each CF clinic, should participation in the study result in any psychological or emotional discomfort for the patients or their parents.

Measures

CHQ

The CHQ contains 75 items measuring 10 health concepts. These include physical functioning, bodily pain, role/social limitations attributable to physical condition, general health perceptions, role/social limitations attributable to emotions, role/social limitations attributable to behavior, mental health, behavior problems, self-esteem, and limitations in family activities. The scale scores range from 0 to 100 with higher scores indicating better function. It is a well-validated and reliable generic measure.\textsuperscript{12}

Brief descriptions of the CHQ scales are as follows:\textsuperscript{12}

- Physical functioning measures the presence and extent of physical limitations attributable to health-related problems.
- Role/social limitations-physical measures the extent of limitations in school-related activities and activities with friends.
- General health measures overall health and illness.
- Bodily pain/discomfort measures both the intensity and frequency of general pain and discomfort.
- Role/social limitations-emotional-behavioral measures limitations in the kind, amount, and performance of schoolwork and activities with friends because of emotional or behavioral difficulties.
- Self-esteem measures the following dimensions of self-esteem: satisfaction with school and athletic ability, looks/appearance, ability to get along with others and family, and life overall.
- Mental health measures the frequency of both negative and positive states.
- General behavior measures the frequency of behavior problems and ability to get along with others.
- Family-limitations in activities measures the frequency of disruption in usual family activities.

Parent Child Health Questionnaire (PCHQ)

The PCHQ consists of 73 items that measure the same child health concepts as the CHQ. It also has 4 additional items that measure the impact of the child’s disease on parent’s self-reported physical health, self-reported mental health, parent’s emotion, and time spent on activities related to the care of the child’s medical condition.

FEV\textsubscript{1}

Pulmonary function, assessed by the FEV\textsubscript{1} % predicted was obtained from each patient’s medical chart at the completion of the study. The best measure of FEV\textsubscript{1} % predicted for the year of the study was used for each patient.

Statistical Methods

Independent t tests were calculated on demographic data to examine site differences. Because no site differences were found, the data were pooled for analysis. Pearson’s product moment correlations were calculated between adolescent pulmonary function (FEV\textsubscript{1} % predicted) and adolescent, maternal, and paternal reports of HRQL. Pearson’s correlations were also calculated between adolescent self-health reports and mother and father reports of adolescent HRQL. All statistical analyses used SPSS, Version 6.1 (SPSS, Chicago, IL).

RESULTS

Of the 39 families that signed the initial consent forms, 6 families (15%) failed to return completed questionnaires after agreeing to participate. Additionally, 8 of the remaining 33 fathers (24%) failed to return completed forms, whereas only 1 of the remaining adolescents (3%) and only 1 of the remaining mothers (3%) failed to participate. Total questionnaire return was 25 for fathers (64%), 32 for mothers (82%) and 32 for adolescents (82%). The present study is based on 24 complete mother, father, and adolescent triads. Of the adolescents, 18 were male (75%) and 6 were female (25%). Their mean age was 14.6 years (standard deviation [SD] = 2.2). The mean FEV\textsubscript{1} % predicted was 75.9 (SD = 27.3). Mean FEV\textsubscript{1} % predicted for males was 78.9 (n = 18; SD = 27.7), whereas the mean for females was 66.9 (n = 6; SD = 26.3). No significant gender differences were found for age or pulmonary function.

Table 1 presents the correlations between adolescent FEV\textsubscript{1} % predicted and adolescent, mother, and father reports of adolescent HRQL. The physically oriented health scales are presented first. Adolescent pulmonary function was related to reporter perceptions of adolescent physical health and functioning by all 3 reporters. Pulmonary function was not found
to relate to any reports of adolescent emotional, social, or behavioral health. Table 2 presents the associations between adolescent and parent reports of adolescent health.

**DISCUSSION**

This study yields results that have important implications for theoretical, clinical, and research considerations. Theoretically, the findings from this study support a conceptualization of health that reaches beyond the traditional medical model, toward a conceptualization of health that includes emotional, behavioral, and social dimensions. This broader conceptualization seems to be of particular importance when caring for children and adolescents with chronic progressive conditions, such as CF, and requires additional study to see whether it can be generalized to other chronic progressive diseases. The low correlations between adolescent pulmonary function and reporter perceptions of adolescent social, emotional, and behavioral health demonstrate the need for the inclusion of multidimensional measures of HRQL in clinical assessment and research on adolescents with CF. These HRQL measures should complement, and be used in addition to, traditional biomedical parameters of health. This broader assessment will be useful to clinicians and researchers when prescribing treatments, making clinical decisions, and developing research protocols for families living with CF and perhaps with other chronic progressive diseases.

The findings confirm a moderate to strong relationship between adolescent pulmonary function and reporter perceptions of adolescent physical health. This association is stronger than past reports of pulmonary function and HRQL in adolescents and adults with CF. This finding may be attributed to a greater sensitivity of the CHQ and PCHQ to detect physically oriented HRQL in adolescents with CF. The availability and use of psychometrically sound measures to assess HRQL in children continues to be an important consideration for future research. Child- and adolescent-specific HRQL measures should be used to assess the health and functioning aspects of daily living for children. Previous research has suggested that adult measures of quality of life do not have clinical or practical utility when applied to a pediatric CF population. Replication of the present study, using the CHQ, and additional studies, using other psychometrically sound, child-focused measures of HRQL, are warranted. The CHQ is a generic HRQL measure that also can be used to assess health outcomes in children and adolescents across a variety of conditions, as well as with healthy children. The use of CF-specific HRQL measures may contribute additional assessment and intervention information. Research into the relationship between generic and disease-specific HRQL measures for children and adolescents with CF is also warranted.

The strong associations between adolescent pulmonary function and adolescent self-perceptions of physically oriented health are consistent with previous research. These findings represent a partial contradiction to earlier reports of denial of physical symptoms as a protective strategy for psychosocial adaptation to CF. The adolescents in this study reported moderate to strong relationships between their pulmonary function and self-reports of general health, pain, and limitations in activities at school and with friends because of physical health problems. This demonstrates an awareness of physical symptoms and contradicts previous reports of denial of symptoms in adolescents and adults with CF. Adolescent pulmonary function, however, was not related to adolescent self-reports of physical functioning or emotional or behavioral health. This inconsistent pattern may be because of protective denial, or it may represent a psychosocial or emotional resiliency not explained by protective denial. The use of denial as a form of coping and adaptation in adolescents with CF deserves additional investigation. Previous research has reported that female adolescents with CF rely more heavily on denial than do adolescent males with CF. Therefore, our findings may be biased by the high male to female ratio of adolescents in this study. Future research may need to examine the relationship between perceived HRQL and the gender-specific use of denial. This finding also supports the use of multidimensional measures of HRQL to further examine the relationships between different domains of HRQL and denial in adolescents with CF.

Additional findings also emphasize the importance of including multiple informants, specifically adolescents and fathers, in clinical assessments and research on HRQL in adolescents with CF. Both
mother and father reports of adolescent HRQL were strongly related to adolescent self-reports of general health and behavior problems. Closely related child and parent perceptions of adolescent general health may be because of the continued high level of parental involvement around issues of general health when a child has CF.

Parent and adolescent reports of perceived adolescent behavior problems were also strongly associated. This is in contradiction to previous reports. The strong associations between parents and adolescents may be attributable to parental and child involvement in treatments for CF and possible behavioral problems around treatment issues in adolescence. Some studies have reported an increase in behavior problems in children and adolescents with CF, although these studies have included only parent reports. A closer look at child and parent reports of behavior problems in children and adolescents with CF is needed. Self-reports of adolescents should be included in research on behavior. Additional research should also examine the relationship between self-reported behavior problems and perceived HRQL in adolescents with CF.

In addition, associations were found between parent and adolescent perceptions of adolescent HRQL on 8 of the remaining health scales.Mother and father reports, however, were related to adolescent self-reports on different health scales. This suggests that mothers, if they are sole respondents for families living with CF, do not fully represent the family perceptions of adolescent HRQL. Father reports of adolescent HRQL were strongly related to adolescent perceptions of self-esteem and moderately related to adolescent self-reports of mental health. Mother and adolescent reports, however, were strongly associated on bodily pain, role/social limitations attributable to physical pain and physical functioning, and moderately related on perceptions of limitations in family activities and role/social limitations attributable to behavior. These scale-specific results may be attributed to role-specific parenting, whereby mothers may be more involved with the adolescents’ physical care and fathers may be more involved with the adolescent around nonphysical aspects of health, such as mental health and self-esteem. Future research may want to examine parental roles, or time spent in various parenting activities, as they relate to perceived HRQL in parents and adolescents with CF. Future analyses may also want to examine family interreporter differences in perceived adolescent HRQL. This was not an outcome directly examined in this study.

Neither maternal nor paternal responses were correlated with the adolescents’ reports of role/social limitations attributable to emotion. This result may represent an increase in parental focus on the adolescents’ physical limitations when an adolescent has CF. It may also be because adolescents with CF may not want to burden their parents with the emotional limitations that they may feel as children and parents deal with the physical implications of the disease. This finding could also be attributable to typical adolescent development. Adolescence is a time when children begin to turn to peers and other key people, outside of their family, for emotional support.

The inconsistent pattern of association seen in this study between adolescent and parental perceptions of the adolescent’s health supports including multiple informants when assessing HRQL in adolescents with CF. In particular, it underscores the particular need for including fathers and adolescents when assessing child mental health and self-esteem and adolescent self-reports of perceived emotional limitations. Adolescent and father perceptions may add another dimension to our understanding of how adolescents with chronic disease, and their families, perceive quality of life and the impact of disease on it. Future research may also include peer, sibling, and teacher perceptions of adolescent health. A study examining health care professionals’ perceptions of adolescent health would also be beneficial. The treating professionals’ perceptions of adolescent HRQL will be important to study because their perceptions may significantly affect the psychosocial interventions or therapeutic options offered to the adolescent and family.

CONCLUSION

The results of this study provide support for the use of comprehensive, multidimensional measures of HRQL, in addition to pulmonary function, when doing research, clinical assessments, or measuring the efficacy of treatments and procedures in adolescents with CF. Furthermore, it suggests that adolescent and parent reports of adolescent HRQL are not interchangeable. Fathers and adolescents provide their own unique perspective of adolescent health status. Every effort should be made to include them, in addition to maternal reports, when assessing HRQL in adolescents with CF.

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