QUALITY-OF-LIFE IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS MANAGED IN BOTH REGIONAL OUTREACH AND CYSTIC FIBROSIS CENTER SETTINGS IN QUEENSLAND

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Objectives To assess the health-related quality-of-life (HRQOL) of children/adolescents with cystic fibrosis (CF) and compare HRQOL in children managed by cystic fibrosis outreach service (CFOS) with those treated in a cystic fibrosis center (CFC). To compare HRQOL of children with CF in Queensland with previously published HRQOL data from the United States and examine the relationship between HRQOL scores and pulmonary function.

Study design Participants were children/adolescents with CF and their parents managed by the Royal Children's Hospital Queensland at a CFC or CFOS. Two HRQOL surveys were used: PedsQL™ and Cystic Fibrosis Questionnaire (CFQ).

Results There were 91 CFC and 71 CFOS participants with similar demographics. PedsQL™ total summary score was statistically higher in CFOS, $P = .05$. There was no significant difference in CFQ scores between groups. Queensland parents reported lower HRQOL for their children compared with US parents ($P<.01$) despite similar pulmonary function. Declining pulmonary function correlated with worse CFQ scores in adolescents, $P < .05$.

Conclusions Children living in regional Queensland reported as good as or slightly better HRQOL compared with children attending a CFC. Parent proxy HRQOL scores were generally low suggesting a reduced perception of HRQOL by parents for their children. *(J Pediatr 2006;148:508-16)*

The life expectancy of patients with cystic fibrosis (CF) continues to increase, which is thought to be due to multiple reasons: earlier diagnosis with neonatal screening, improved treatment of respiratory infections, improved nutritional health, and centralized treatment in a tertiary cystic fibrosis center (CFC) with a multidisciplinary approach to treatment.[1-3]

In Queensland, most children with CF living in the urban area within driving distance of the capital, Brisbane, are treated by a CF multidisciplinary team in a tertiary CFC. Children are reviewed at least 3 times a year and have full access to the multidisciplinary team. Children with CF living in regional or rural areas across Queensland and Northern New South Wales are managed by their local pediatrician or general practitioner and local hospital, and they also attend outreach clinics visited by the cystic fibrosis outreach service (CFOS). The CFOS travels to seven sites in Queensland with the greatest distance being 1700 km from the tertiary center. The CFOS varies, although it usually includes a pediatric respiratory physician, physiotherapist, dietician, and cystic fibrosis nurse. Regional staff, such as pediatricians, physiotherapists, dieticians, and clinical nurses, are invited to attend the clinics. Outreach clinics occur twice per year except for one site, which has one clinic and two telehealth clinics per year.

Traditionally clinical outcomes such as spirometry and anthropometric data are used to assess patients' progress and the effectiveness of therapy, but these fail to assess the impact the disease has on the patients' every day social and physical functioning, emotional well-being, and their perception of their chronic illness. The US CF Foundation recommends that the quality-of-life of patients also be included in outcome parameters and in assessing treatment outcomes.[4]

People living in rural and remote Australia have been shown to have poorer health compared with those living in metropolitan zones, with higher mortality rates and lower life expectancy.[5] There is however, a paucity of health-related quality-of-life (HRQOL) information for children living in rural or regional Australia. Spurrier et al found poorer

ARIA	Accessibility/Remoteness Index of Australia	CFQ	Cystic Fibrosis Questionnaire
CF	Cystic fibrosis	FEV_1	Forced expiratory volume in 1 second
CFC	Cystic fibrosis center	HRQOL	Health-related quality-of-life
CFOS	Cystic fibrosis outreach service	SES	Socioeconomic status

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HRQOL using The Child Health Questionnaire in children from lower socioeconomic contexts in Australia,[6] and Cameron et al found reduced HRQOL using the same tool in children with diabetes living in regional compared with urban areas in Victoria (Australia).[7,8]

We hypothesized that HRQOL may be reduced in children with CF from regional areas as was found in children with diabetes. This study was designed using both a generic HRQOL measure, the PedsQLTM,[9] and a disease-specific HRQOL measure, the Cystic Fibrosis Questionnaire (CFQ),[10,11] to examine HRQOL in urban and regional children and adolescents with CF and the correlation between the HRQOL and disease severity as measured by change in pulmonary function, and to enable comparison of HRQOL with published data from the United States[10,12] and Australia.[13]

METHODS

The participants were children and adolescents, and their parents, living in Queensland and Northern New South Wales who were treated by the Royal Children's Hospital CF team in a tertiary (CFC) or outreach setting (CFOS). Children were between 2 and 19 years of age and had a proven diagnosis of CF based on genetic and/or sweat testing and compatible clinical features. Geographical location was categorized according to the Accessibility/Remoteness Index of Australia (ARIA).[14] ARIA classification uses road distances to calculate remoteness and accessibility to general service centers, and it is grouped into five categories: Highly Accessible, Moderately Accessible, Accessible, Remote, and Very Remote.

Demographic details were collected from medical records or available pathology databases including the neonatal screening records. CF genotypes for all children were grouped as homozygous for the ΔF508 mutation (ΔF508/ΔF508), heterozygous ΔF508 mutation (ΔF508/other), no ΔF508 mutation (other/other), or not available. Socioeconomic status (SES) was available for a cohort of 6- to 13-year-olds as collected on the CFQ. It was derived from the highest level of maternal education and categorized into grade 12 or less, completed grade 12/diploma/trade, and professional/college degree.

Approval was obtained from the Royal Children's Hospital and District Ethics committee. Parents and children >8 years of age gave written consent for the study.

HRQOL Questionnaires

Two HRQOL surveys were administered; a generic HRQOL measure, PedsQL^{TM}, and a disease-specific HRQOL measure, the CFQ. Both have been previously validated and tested for reliability.[10,15]

PedsQL™

The PedsQL^{TM} is a generic measure comprising parallel child self-report and parent proxy-report formats. Child self-report includes questionnaires for ages 5 to 7, 8 to 12, and 13 to 18 years. Parent proxy-report includes questionnaires for ages 2 to 4 (toddler), 5 to 7 (young child), 8 to 12 (child), and 13 to 18 years (adolescent), and it assesses parents' perceptions of the child's HRQOL.[15] Both the child self-report and the parent proxy-report comprise 23 items, measuring four multidimensional generic scales (Physical, Social, Emotional, and School), based on a 4-week recall. A 5-point response scale is used for all but the young child self-report (ages 5 to 7 years), which only has a 3-point response scale. The PedsQL^{TM} was scored according to the instructions available on the website,[16] with higher scores representing a better HRQOL score. Any missing item was given the mean score of items completed for that domain if more than half of the answers for that specific domain were available; otherwise they were left blank and recorded as missing.[16] Results are categorized into four generic core scales (mean of the sum of the items for each Physical functioning, Emotional functioning, Social functioning, and School functioning) and three summary scores: Psychosocial Health Summary Score (mean of the sum of the items of Emotional functioning, Social functioning, and School functioning), Physical Health Summary Score (same as Physical functioning scale score) and Total Scale Score (mean of the sum of all the items in all the scales). The Total Scale Score is suitable as a summary score for the primary analysis of HRQOL outcome in clinical trials and group comparisons.[17]

CFQ

The CFQ is a disease-specific HRQOL measure consisting of four versions. A self-report CFQ-Teen/adult (≥14 years of age), CFQ-Child interview version (6-11 years of age), CFQ-Child self-report (12-13 years of age) and a CFQ-Parent (proxy) version used in conjunction with the CFQ-Child version (6-13 years of age). Each version of the CFQ is different, but all are composed of broad domains including physical symptoms, emotional functioning, vitality, social functioning, and school functioning plus domains specific to CF such as body image, eating disturbances, treatment burden, respiratory symptoms, weight, and digestive symptoms. The number of items for each domain varies according to the version of the questionnaire, and all are based on a 2-week recall. The CFQ was scored according to the instructions in the manual,[10] with higher scores representing a better HRQOL. Any missing item was given the mean score of items completed for that domain if more than half of the answers for that specific domain were available, otherwise they were left blank and recorded as missing.[10] Results were categorized into each domain according to the CFQ version.

Data Collection

The PedsQL^{TM} was administered before the CFQ as per the recommended administration guidelines.[9] Patients and parents completed surveys in the waiting room of a routine clinic visit or had surveys mailed to them, thus simul-