Health-Related Quality of Life Outcomes in Children and Adolescents with Congenital Heart Disease

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Objectives To compare health-related quality of life (HRQOL) in a group of pediatric patients with congenital heart disease (CHD) and healthy controls and patients with other chronic diseases, and to compare HRQOL among patients with CHD of various severity categories with one another, with controls, and with patients with other chronic diseases.

Study design In this cross-sectional survey, *t* tests were used to compare patient and proxy-reported Pediatric Quality of Life Inventory 4.0 Generic Core Scales (PedsQL) scores (including total, physical health, and psychosocial health summary scores) in children (aged 8-12 years) and adolescents (aged 13-18 years) between controls and (1) a composite CHD population; and (2) patients in each of 3 CHD severity categories: mild (no intervention), biventricle (BV; postintervention), and single ventricle (SV; postpalliation). PedsQL scores among CHD severity categories were compared by ANOVA. PedsQL scores were also compared in the CHD population and children with other chronic diseases without age stratification using *t* tests.

Results There were 1138 (children, n = 625; adolescents, n = 513) and 771 (children, n = 528; adolescents, n = 243) patient and/or proxy reporters in the CHD and healthy control groups, respectively. Total, physical health, and psychosocial health summary scores were lower in the composite CHD, BV, and SV groups compared with controls ($P < .0001$). There were significant differences among disease severity categories for all scores ($P < .01$). The composite CHD, BV, and SV groups had similar PedsQL scores as end-stage renal disease, asthma, and obesity populations.

Conclusion Children and adolescents with BV and SV CHD have significantly lower HRQOL than healthy controls and similar HRQOL as patients with other chronic pediatric diseases. Interventions targeting both physical and psychosocial domains are needed to improve HRQOL in this high-risk population. *(J Pediatr 2014;164:781-8)*.

Health-related quality of life (HRQOL) is defined as the impact of a specific illness, medical therapy, or health services policy on a patient's ability to function in various life contexts and draw personal satisfaction from physical, psychological, and social functioning (SoF) perspectives.[1] The Pediatric Quality of Life Inventory 4.0 Generic Core Scales (PedsQL) is a reliable, valid generic HRQOL measurement tool that is widely used in pediatric populations to determine HRQOL in patients with chronic diseases and healthy controls.[2] The PedsQL can be used to characterize the impact of therapies and disease processes on the HRQOL of children and adolescents with congenital heart disease (CHD) and to compare this impact with that in children and adolescents with other chronic diseases.

Given the cognitive and emotional differences between children and adolescents[3] and differences in functioning among patients with heart disease (HD) of varying severity,[4-12] individuals' HRQOL may be affected differently, depending on age and disease severity. The effects of these factors on the HRQOL of the pediatric CHD population have not been delineated in a large, geographically

BV	Biventricle	PedsQL	Pediatric Quality of Life Inventory 4.0 Generic Core Scales
CHD	Congenital heart disease	PhysHSS	Physical health summary score
EmF	Emotional functioning	PsychHSS	Psychosocial health summary score
ESRD	End-stage renal disease	ScF	School functioning
HD	Heart disease	SoF	Social functioning
HRQOL	Health-related quality of life	SV	Single ventricle
PCQLI	Pediatric Cardiac Quality of Life Inventory		

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*A list of Pediatric Cardiac Quality of Life Inventory Testing Study Consortium sites is available at www.jpeds.com (**Appendix**).

Supported by the National Institute of Child Health and Human Development (5-K23-HD048637), the American Heart Association, Pennsylvania/Delaware Affiliate (now Great Rivers Affiliate; beginning Grant-in-Aid 0465467U), the Cincinnati Children's Hospital Research Foundation, and the Children's Hospital of Philadelphia Institutional Development Fund. The authors declare no conflicts of interest.

Portions of the study were presented as a poster at the American Academy of Pediatrics' National Conference and Exhibition, San Francisco, CA, October 2-5, 2010.

0022-3476/$ - see front matter. http://dx.doi.org/10.1016/j.jpeds.2013.11.066

diverse cohort, however. Two previous studies[13,14] assessed HRQOL in pediatric patients with HD; however, relatively small HD samples were used, and findings were mixed. In the present study, we aimed to use a geographically diverse cohort to compare patient and proxy-reported PedsQL scores in children (aged 8-12 years) and adolescents (aged 13-18 years) with CHD with those of sex- and race-matched healthy controls. Within each age group, we also compared PedsQL scores among patients with CHD of various severity categories with one another and with controls to determine which severity categories within each age group might benefit from interventions targeting specific HRQOL domains. In addition, PedsQL scores among patients with CHD of various severity categories were compared to those of patients with other chronic diseases. We hypothesized that children and adolescents with CHD would have lower HRQOL than healthy controls, that HRQOL would not be significantly different between children and adolescents with mild CHD and healthy controls, and that children and adolescents with more complex CHD would have significant deficits in each HRQOL domain (total; physical health; psychosocial health; emotional functioning [EmF], SoF, and school functioning [ScF]) compared with healthy controls and patients with less complex disease. We also hypothesized that pediatric patients with complex CHD would have a similar HRQOL as those with other chronic diseases, and that pediatric patients with mild CHD would have a better HRQOL compared with those with other chronic diseases.

Methods

This study was a cross-sectional survey that used patient data originally collected to test the psychometric properties of the Pediatric Cardiac Quality of Life Inventory (PCQLI). The Institutional Review Board of Cincinnati Children's Hospital Medical Center approved this study and granted a waiver of consent based on its retrospective nature.

The PedsQL data used for the CHD group in this study was provided by patients with CHD and their parents to establish construct validity during the PCQLI validation study.[15-17] Patients from 7 large US pediatric cardiac centers (**Appendix**) contributed to the CHD patient population between November 2004 and December 2008. Eligibility criteria for participation in the PCQLI validation study included CHD or acquired HD, age 8-18 years, fluent in English, and attending an outpatient cardiology visit. Patients who had a significant comorbid medical condition or major developmental delay or who came to the clinic because of an acute change in clinical status were excluded. Proxy reporters were excluded if they were not fluent in English or had a major developmental delay. From the 1605 patient–parent pairs who were included in the PCQLI validation study, those patients with CHD who also had a valid self-reported or proxy-reported PedsQL score were included in the present study. The healthy population used in this study was obtained from the PedsQL healthy children database.[2,18] These normative data were obtained from the previously conducted PedsQL initial field test and a statewide State Children's Health Insurance Program evaluation in California. The PedsQL data for the chronic disease populations were obtained from a previously published database.[14]

For the PCQLI validation study, patients and proxy reporters were given the PedsQL before a routine outpatient visit. Research personnel obtained patient and parent demographic information and patient clinical information (diagnostic, surgical, and medical history) for the CHD population through parent report and chart review. The original diagnostic category was divided into 2 subgroups: biventricle (BV) CHD and single ventricle (SV) CHD. CHD severity categories included mild CHD (patients with CHD who had not undergone surgical or catheter-based intervention), BV CHD (patients with CHD with BV physiology who had undergone surgical or catheter-based intervention), and SV CHD (patients with CHD with SV physiology who had undergone surgical palliation). The number of cardiac surgeries (defined as sternotomy or thoracotomy) and number of cardiac catheterizations were reported for the BV CHD and SV CHD groups only. The time since the most recent hospitalization was reported for all CHD disease severity categories. For the healthy control population, patient sex and race data for the child and adolescent groups were obtained from the PedsQL healthy children database.

The PedsQL, developed by Varni et al, was used to assess HRQOL and has well-documented reliability and validity in the pediatric HD and healthy populations.[2,13,14,18] The PedsQL includes 23 items that evaluate physical functioning and psychosocial functioning. The physical health summary score (PhysHSS) includes physical functioning, and the psychosocial health summary score (PsychHSS) includes EmF, SoF, and ScF. The total score incorporates all of these domains. There are separate forms for children aged 8-12 years and adolescents aged 13-18 years as well as their proxy reporters; the content of the items in each form are the same except for appropriate changes to pronouns and language modifications to accommodate developmental differences. All answers are based on a 5-point Likert scale (0, not a problem; 1, almost never a problem; 2, sometimes a problem; 3, often a problem; 4, almost always a problem). Items are reverse-scored and linearly transformed to a scale of 0-100, with higher transformed scores indicating a better HRQOL.

The number of cardiac surgeries, number of cardiac catheterizations, and time since the most recent hospitalization were compared in the BV CHD and SV CHD groups using the Wilcoxon rank-sum test. Patient and proxy-reported PedsQL scores (total score, PhysHSS, PsychHSS, EmF, SoF, and ScF) were compared between the composite pediatric CHD group and sex- and race-matched healthy controls in 2 age groups (children [age 8-12 years] and adolescents [age 13-18 years]) and between patients in each of 3 CHD severity categories and healthy controls in both age groups using *t* tests. Patient and proxy-reported PedsQL scores (total score, PhysHSS, PsychHSS, EmF, SoF, and ScF) were