



# Assessment of Quality of Life in Young Patients with Single Ventricle after the Fontan Operation

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**Objectives** To assess self-reported quality of life (QOL) in a large multicenter cohort of adolescent and young adults surviving Fontan.

**Study design** Cross-sectional. The Pediatric Quality of Life Inventory (PedsQL) was administered to 408 survivors of Fontan ages 13-25 years enrolled in the Pediatric Heart Network Fontan Follow-up Study. Subjects also completed either the Child Health Questionnaire (age <19 years) or Short Form Health Survey (age ≥19 years). PedsQL data were compared with matched controls without a chronic health condition. Correlations between the measures were examined.

**Results** Mean PedsQL scores for subjects receiving Fontan were significantly lower than those for the control group for physical and psychosocial QOL ( $P < .001$ ). Overall, 45% of subjects receiving Fontan had scores in the clinically significant impaired range for physical QOL with 30% in the impaired range for psychosocial QOL. For each 1 year increase in age, the physical functioning score decreased by an average of 0.76 points ( $P = .004$ ) and the emotional functioning score decreased by an average of 0.64 points ( $P = .03$ ). Among subjects ≥19 years of age, the physical functioning score decreased by an average of 2 points for each year increase in age ( $P = .02$ ). PedsQL scale scores were significantly correlated with conceptually related Child Health Questionnaire ( $P < .001$ ) and Short Form Health Survey scores ( $P < .001$ ).

**Conclusions** Survivors of Fontan are at risk for significantly impaired QOL which may decline with advancing age. Routine assessment of QOL is essential to inform interventions to improve health outcomes. The PedsQL allowed QOL assessment from pediatrics to young adulthood. (*J Pediatr* 2016;170:166-72).

**Trial registration** [ClinicalTrials.gov](http://ClinicalTrials.gov): NCT00132782.

Advances in medical and surgical care have dramatically improved the life expectancy of children with congenital heart disease (CHD), including children born with complex single ventricle defects who undergo the Fontan operation. Although the Fontan surgery does not provide anatomic correction, it provides separate pulmonary and systemic circulations supported by the single ventricle, allowing an increasing number of patients to reach adulthood and the opportunity for a productive life. However, greater knowledge of psychosocial morbidity and the potential impact of the Fontan operation on overall quality of life (QOL) and functioning are essential to improving outcomes in this patient population.

QOL is a multidimensional construct which includes physical, psychological, and social functioning, consistent with the World Health Organization's definition of health.<sup>1</sup> Furthermore, the World Health Organization has emphasized that QOL is a subjective experience, defined as "individuals' perceptions of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards, and concerns."<sup>2</sup> QOL measurement, however, has been fraught with many conceptual and

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Supported by the National Heart, Lung, and Blood Institute (HL068269, HL068270, HL068279, HL068281, HL068285, HL068292, HL068290, HL068288). This work is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health/National Heart, Lung, and Blood Institute. J.V. holds the copyright and the trademark for the PedsQL and receives financial compensation from the Mapi Research Trust, which is a nonprofit research institute that charges distribution fees to for-profit companies that use the Pediatric Quality of Life Inventory. He did not receive compensation for this project. The other authors declare no conflicts of interest.

Portions of the study were presented as an abstract and poster at the American Heart Association Scientific Sessions, Chicago, IL, November 17, 2014.

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<http://dx.doi.org/10.1016/j.jpeds.2015.11.016>

CHD	Congenital heart disease
CHQ-87	Child Health Questionnaire
Fontan 1	Pediatric Heart Network Fontan Cross-Sectional Study
Fontan 2	Fontan Follow-up Study
PedsQL	Pediatric Quality of Life Inventory
QOL	Quality of life
SF-36	Short Form Health Survey

methodological issues.<sup>3</sup> It is often incorrectly used as a generic label to describe a range of physical, health status, and psychosocial variables, including objective observations by others reflecting their standards. Past studies have often relied on parental report, are limited by small sample size/single center design, are restricted to a specific age-group, or use different measures across age-groups.<sup>4-10</sup> No longitudinal follow-up of QOL has been reported in patients with Fontan physiology as they move through childhood to young adulthood, perhaps in part reflecting the lack of an available self-report QOL measurement instrument that spans the age range of this growing population of survivors of Fontan.

Our aims were to describe self-reported QOL in a large multicenter cohort of adolescent and young adult patients following the Fontan operation as measured by the Pediatric Quality of Life Inventory (PedsQL), and to examine the correlation between the PedsQL and related conceptual functional domains (physical and psychosocial/mental functioning) as measured on the Child Health Questionnaire (CHQ-87) in patients <19 years of age and the Short Form Health Survey (SF-36) in patients 19 years of age and older.

## Methods

The Pediatric Heart Network Fontan Cross-Sectional Study (Fontan 1) characterized a multi-institutional cohort (7 sites) of 546 survivors after the Fontan procedure, ages 6-18 years at enrollment.<sup>11</sup> From this original cohort, patients who were alive with a Fontan circulation were approached for enrollment in the Fontan Follow-up Study (Fontan 2) from 2009-2011<sup>12</sup> and are the subjects of this study. Changes in functional health status from Fontan 1 to Fontan 2 as well as predictors of these changes in this patient cohort have been previously reported.<sup>12</sup> The protocol was approved by each center's institutional review board and informed consent was obtained.

To assess QOL, patients completed the PedsQL 4.0 generic core scales<sup>13</sup> and the PedsQL cardiac module.<sup>14,15</sup> The 23-item PedsQL core scales encompass physical, emotional, social, and school/work functioning. Items are linearly transformed to a 0- to 100-scale, so that higher scores indicate better QOL. Parallel child, teen, and young adult versions of the PedsQL have been developed with similar content and wording across forms to facilitate evaluation of differences across and between age groups as well as longitudinal tracking. To create a psychosocial health summary score, the mean of the emotional, social, and school/work functioning scales is computed. The reliability and validity of the PedsQL generic core scales have been demonstrated in healthy and patient populations.<sup>13,16-18</sup> The PedsQL cardiac module has 6 scales related to symptoms, treatment barriers (for patients on medications), perceived physical appearance, treatment anxiety, cognitive problems, and communication. Formatting and scoring are the same as the PedsQL generic core scales. Validity and reliability for 8- to 18-year-olds has been demonstrated.<sup>14</sup>

Patients <19 years old completed the CHQ-87, the same measure administered to patients 10-18 years of age in Fontan 1. The CHQ-87 assesses functional health status across a range of areas related to physical, mental, and social domains.<sup>19</sup> The scale domain scores range from 0-100, with higher scores indicating better function.

In patients  $\geq 19$  years of age, functional health status was measured using the SF-36 (version 2). The 36-item SF-36 is a multipurpose, short form health survey. It yields an 8-scale profile of functional health and well-being scores as well as psychometrically based physical and mental health component summary measures.<sup>20</sup> The reliability and the validity of the SF-36 have been established.<sup>21,22</sup>

## Data Analyses

Descriptive statistics for demographic and clinical variables are reported as means and SD or medians with IQR for continuous variables and frequencies/proportions for categorical variables. Patient characteristics and PedsQL scale and summary scores were compared between adolescents (<19 years old) and young adults ( $\geq 19$  years of age) using appropriate statistical tests (Student *t* test for means, Wilcoxon ranked-sum test for medians, or Fisher exact test for frequencies). PedsQL Scale and summary scores were also compared with an age-, sex-, and race/ethnicity-matched healthy children sample from the PedsQL database. The healthy comparison group was comprised of 342 subjects from the PedsQL healthy children database, which is composed of children and young adults without a chronic health condition<sup>16,17</sup> and included 242 subjects <19 years old and 100 subjects 19-25 years of age. Comparisons were repeated after adjustments for minor differences in age and sex and showed no significant differences from unadjusted values. Adjusted values are reported. For each individual scale and the psychosocial health summary score, we report the frequency of scores greater than 1 SD below the general pediatric population sample mean, the cut-off score for significantly impaired QOL.<sup>16</sup> Individual item analysis with ranking was performed to identify the most significant problems or lowest mean scores within PedsQL core and cardiac module scales. Unadjusted linear regression models were used to test the association between each of the PedsQL scale scores and age. Regression analyses were used to test the association of PedsQL Scales with income, maternal education, and sex. Spearman correlation coefficients were calculated to test the association between the PedsQL scores and the corresponding CHQ-87 and SF-36 scores. Effect sizes were designated as small (0.10-0.29), medium (0.30-0.49), and large ( $\geq 0.50$ ).

## Results

Of 427 subjects enrolled in Fontan 2 from November 2009 to May 2011, 408 (96%) completed the PedsQL. In addition to the PedsQL, 255 adolescents <19 years old completed the CHQ-87, and 153 young adults 19-25 years of age completed

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