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Quality of Life is Diminished in Patients with Tetralogy of Fallot with Mild Residual Disease: A Comparison of Tetralogy of Fallot and Isolated Valvar Pulmonary Stenosis

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Abstract

The objective of this study is to compare quality of life (QOL) in patients with mild pulmonary insufficiency (PI) after Tetralogy of Fallot (TOF) repair or after balloon dilation for isolated valvar pulmonary stenosis (VPS). A cross-sectional study of patients with TOF ($n = 12$) and VPS ($n = 19$), ages 8–18 years, who underwent cardiac magnetic resonance (CMR) and cardiopulmonary exercise test (CPET) was conducted. Patients with genetic syndromes were excluded. The groups were matched by severity and duration of PI using propensity scores. PI was greater than mild if the regurgitant fraction by CMR was $>20\%$. Health status and QOL assessment included Child Health Questionnaire Child Self-Report (CHQ-CF87), Child Health Questionnaire Parent Report (CHQ-PF50), and Pediatric Cardiac QOL Instrument (parent and patient). QOL scores were compared between groups. Due to propensity matching, the groups had, at worst, mild PI and normal right ventricular ejection fraction on CMR. Parental perception of QOL was significantly worse in TOF as compared to VPS in the domains of general perception of health ($P = 0.03$), physical functioning (PF; $P = 0.004$), and family cohesion ($P = 0.048$). There were no differences in self-reported QOL between groups. There was no association between QOL and right ventricular function on CMR or percent-predicted maximal oxygen consumption on CPET in both groups. Parent-perceived QOL, in the domains of general health perception, PF, and family cohesion, is significantly reduced in patients with TOF with mild residual disease suggesting that the psychosocial impact of congenital heart disease may be significant even with successful repair and satisfactory medical status.

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Compliance with Ethical Standards

Research Involving Human Participants and/or Animals All procedures performed in studies involving human participants were in accordance with the Ethical Standards of the Institutional and/or National Research Committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from all individual participants included in the study.

Conflict of interest The authors declare that they have no conflict of interest.

Keywords

Congenital heart disease; Health status; Quality of life; Pulmonary stenosis; Tetralogy of Fallot

Introduction

Although long-term survival after surgical repair for Tetralogy of Fallot (TOF) is comparable to the general population, patients experience significant morbidity primarily associated with residual pulmonary insufficiency (PI). In the long term, PI results in right ventricular dilation and ultimately in right ventricular dysfunction with a resultant progressive decline in exercise tolerance [1–4]. These morbidities result in repeated procedures and re-interventions to preserve right ventricular function. Patients also face challenges with cognitive function, education, and employment as adults [5, 6]. Psychosocial morbidities [7–9], neurodevelopmental morbidities [10, 11], and diminished physical functioning (PF) [12] can have a negative cumulative effect on quality of life (QOL) [13]. As an important multidimensional measure of well-being, QOL reflects an individual's ability to function in social, emotional, psychological, and physical domains as well as personal satisfaction [13–15]. QOL has been shown to be diminished in patients with CHD as compared to the general population [16–18].

Our objective was to evaluate the health status and QOL of patients with mild residual disease, defined as mild PI with preserved ejection fraction. We therefore sought to investigate health status and QOL in children and adolescents with repaired TOF as compared to patients with valvar pulmonary stenosis (VPS) status post balloon dilation matched for age and residual disease burden. We hypothesized that health status and QOL would be comparable in these groups.

Methods

Study Population and Data Collection

We conducted a single-center cross-sectional study of subjects with VPS ages 8–18 years at The Children's Hospital of Philadelphia (CHOP) from June 2009 to February 2010 as previously described [19]. This study was approved by the Institutional Review Board for the Protection of Human Subjects at CHOP (IRB #6372). All subjects provided written informed consent prior to participation. VPS subjects status post pulmonary valve balloon dilation identified from the cardiac databases at our institution underwent one day of testing that included a protocol-based cardiac magnetic resonance (CMR), cardiopulmonary exercise test (CPET), and completion of health status and QOL questionnaires by patients and their parent-proxy reporters. Patient health status was measured using the Child Health Questionnaire Child Self-Report (CHQ-CF87) and the Child Health Questionnaire Parent Report (CHQ-PF50), while QOL assessment included a disease-specific instrument, the Pediatric Cardiac QOL Instrument (PCQLI) [20]. The VPS cohort was found to have mild residual disease, defined as mild residual PI after balloon dilation with preserved ventricular ejection fraction.

TOF subjects were drawn from a cohort with repaired TOF enrolled in a cross-sectional study between January 2005 and February 2009, for which they underwent the same CMR, CPET, and QOL protocol [21]. Testing was performed within 3 months in the TOF group. Patients with genetic syndromes were excluded from this analysis. Subjects with VPS and TOF were matched by duration and severity of PI [years from initial procedure and pulmonary regurgitant fraction (RF) on CMR, respectively] using propensity score matching. This resulted in a group of TOF subjects with mild disease, given that the VPS group had on average mild residual PI after balloon dilation [19, 22]. In this study, the health status of the TOF group and VPS group was compared with healthy control population using published normative data available for the CHQ-PF50 [23, 24]. QOL in the VPS and TOF groups was compared with two CHD control groups based on the severity of disease using published data for mild [isolated bicuspid aortic valve (BAV) with no aortic stenosis or insufficiency who have not undergone intervention] and severe CHD (patients with single ventricle who have undergone Fontan operation) [20, 25].

Health Status and Quality of Life Instruments

The CHQ is a comprehensive self-report instrument that measures psychosocial and physical well-being which has been validated for the general population of children ages 5–18 years by the Health Institute at New England Medical Center [23]. This tool provides qualitative assessment of overall health status across multiple domains. The Child Health Questionnaire Parent Form 50 (CHQ-PF50) comprises 50 items and is completed by a parent-proxy reporter. Overall well-being is measured by psychosocial (PsS) and physical (PhS) health summary scores and 12 individual health concepts are assessed by subscale scores, including PF, bodily pain/discomfort, role/social limitations—both physical and emotional/behavioral, self-esteem, mental health, behavior, general health perceptions, family activities, family cohesion, and parental impact—both time and emotional. The PhS is generated from the weighted average of subscale scores of PF, role/social—physical, general health, and bodily pain. The PsS is derived from the subscale scores of self-esteem, mental health, behavior, and role/social—emotional behavioral. Both summary measures include subscales evaluating time and emotional impact on the parent. A higher score reflects better overall physical and mental health status. The Child Health Questionnaire Child Form 87 (CHQ-CF87) is completed by the child and contains 87 questions assessing the health concepts corresponding to the CHQ-PF50 subscales with the exception of the parental impact subscales and distinction of the role/social—emotional and role/social—behavioral subscales. Summary scores are not generated [23].

PCQLI

The Pediatric Cardiac QOL Inventory (PCQLI) is a reliable and validated instrument of disease-specific QOL in pediatric heart disease [13, 15, 20]. The PCQLI has both child and parent-proxy forms: child (8–12 years), parent of child (parent-proxy), adolescent (13–18 years), and parent of adolescent (parent-proxy) forms. Disease impact and psychosocial impact subscale scores are generated with a maximum of 50 points each. A total PCQLI score is generated from the sum of subscale scores and higher scores represent better health-related QOL [20]. Significant differences in PCQLI QOL scores have been demonstrated between patients with mild CHD not requiring surgical intervention, moderate CHD with

biventricular physiology, and severe CHD (patients with single-ventricle physiology after Fontan operation) [20, 25].

Cardiac Magnetic Resonance—CMR studies were performed using a standard imaging protocol and read by an experienced physician blinded to the patients' clinical information, as previously described [19]. PI was measured as pulmonary RF and considered mild if it is <20%, moderate if 21–40%, and severe if greater than 40%. Other CMR variables included indexed end-systolic and end-diastolic RV volumes and RV ejection fraction. RV ejection fraction was considered normal if it is greater than 50% [26, 27].

Cardiopulmonary Exercise Test—Patients completed an exercise protocol as previously described [19]. The percent of predicted maximum $\dot{V}_{O_2}$ (%m $\dot{V}_{O_2}$) was calculated for each patient, according to normative values for age, gender, and body size. Percent-predicted m $\dot{V}_{O_2}$ was considered abnormal if less than 85% predicted [19]. Oxygen pulse was calculated as the m $\dot{V}_{O_2}$ divided by the peak heart rate and indexed for body surface area (ml/ O_2 /beat/ m^2) and used as a surrogate of stroke volume.

Statistical Analysis

The groups were matched by RF and duration of PI using propensity scores. Continuous variables are presented as median with interquartile ranges (IQRs) or as mean and standard deviation (SD) as appropriate. Categorical variables are described using frequencies and percentages. The Mann–Whitney U test for continuous variables and χ^2 test for categorical variables were performed to compare health status and QOL between the VPS and TOF groups. Linear regression was used to examine the relationship between the outcome variables, QOL and m $\dot{V}_{O_2}$. Normative data for the CHQ-PF50 were used to compare VPS and TOF subscale scores with published norms via paired sample t tests. In addition, paired sample t tests were used to compare PCQLI scores of the VPS and TOF groups with two CHD control groups: mild CHD (isolated BAV without history of intervention) and severe CHD (single ventricle who have undergone Fontan palliation) [18, 25]. An α of <0.05 was considered significant and all analyses were conducted using SAS©9.2.

Results

Patient Characteristics

Matching for severity and duration of PI identified 12 subjects with TOF and 19 with VPS. The groups were comparable in age, race, and gender (Table 1). By study design, subjects had comparable PI, measured as RF. There was mild residual PI and less than mild RV dilation with preserved right ventricular function in both groups. Both the percent-predicted maximal oxygen consumption and the oxygen pulse were comparable in the groups (Table 1). All TOF patients underwent cardiac surgical repair and required at least one catheterization procedure (Table 2). VPS patients did not require cardiac surgery and underwent transcatheter pulmonary valve balloon dilation.

Health Status and QOL Comparisons to Normative Populations

VPS Group Comparisons to Normative Sample—In the VPS group, the *Parent (CHQ-PF50)* health status scores were comparable to a normative sample, as there were no significant differences in summary or subscale scores (Table 3). VPS patients had statistically lower psychosocial impact and total PCQLI scores than patients with mild CHD. There was no difference in patient disease impact scores between the VPS and mild CHD groups. Parent-proxy psychosocial score was also significantly lower in the VPS compared to the mild CHD group, with no difference in parent-proxy disease impact or total scores (Table 4). Patients in the VPS group had statistically higher patient disease impact scores than those with severe CHD. There was no difference in patient psychosocial impact or total scores. The parent-proxy disease impact scores and parent-proxy total scores in the VPS group were also significantly better than those in the severe CHD group, with no difference in parent-proxy psychosocial impact scores (Table 5).

TOF Group Comparisons to Normative Sample—In the TOF group, the *Parent (CHQ-PF50)* physical and psychosocial summary scores were comparable to the normative healthy sample. However, lower scores in several subscales were apparent including general health perception ($P = 0.0001$) and family cohesion ($P = 0.035$; Table 3). TOF patients had better bodily pain scores compared to the normative sample. TOF patients had PCQLI psychosocial impact scores that were significantly lower than patients with mild CHD. There was no difference in patient disease impact or total PCQLI scores between TOF and mild CHD patients. Parent-proxy psychosocial impact and total PCQLI scores were also lower in TOF compared with patients with mild CHD. There was no difference in parent-proxy disease impact scores (Table 4). Compared with severe CHD patients, TOF patients had significantly better patient and parent-proxy disease impact scores. No difference was found in patient psychosocial impact or total scores between TOF and severe CHD patients. Parent-proxy psychosocial impact scores, however, were significantly lower in TOF patients compared with severe CHD patients. There was no difference in total parent-proxy scores between TOF and severe CHD groups (Table 5).

Comparison of VPS with TOF Groups

Parent-Proxy Health Status (CHQ-PF50)—Although parent-proxy physical and psychosocial summary scores were similar among patients with TOF and VPS, certain subscales were significantly lower in TOF as compared to VPS, including parent-proxy-reported general health perception, PF, and family cohesion. Subscale scores in the major psychosocial domains including global behavior, global health, behavior, mental health, self-esteem, social limitations (behavioral and physical), parental impact, and family factors were comparable between the groups (Table 6).

Child Health Status (CHQ-CF87)—Self-reported mental health scores were better in patients with TOF compared to VPS. All other self-reported QOL scores were comparable between the groups (Table 7).

Parent-Proxy and Child QOL (PCQLI)—Parents of patients in the TOF group reported similar overall QOL as those in the VPS group, as evident by the parent-proxy PCQLI total

score. There were no group differences in parent-reported PCQLI disease impact or psychosocial impact scores. There were no significant group differences in any of the patient-reported PCQLI measures of QOL (Table 7).

Discussion

In this study, we sought to investigate the differences in health status and QOL in children and adolescents with TOF and VPS with mild residual disease, defined as mild PI with preserved ejection fraction. In comparison to patients with VPS matched for similar residual disease burden, *parent-proxy*-reported QOL scores in the domains of general health perception, PF, and family cohesion were significantly reduced in patients with repaired TOF. In TOF with mild residual disease, parent-proxy-reported QOL scores in the domains of general health perceptions and family cohesion were lower compared to healthy children. Furthermore, parental psychosocial QOL was significantly lower in TOF than mild and severe CHD groups. Although parent-proxy-reported health status in VPS with mild residual disease was comparable to healthy children, their psychosocial QOL scores were significantly lower in VPS compared to those with mild CHD.

Even in the face of mild residual disease, parent-proxy-reported general health perception was significantly lower in TOF as compared to VPS. Furthermore, parent-proxy psychosocial impact was lower in TOF than those with mild and severe CHD. This suggests that in TOF, the parent-perceived QOL is lower than the severity of residual disease. Alternatively, our finding might suggest that having a child that underwent cardiac surgery might, in and of itself, result in impaired QOL when compared to parents of children that underwent nonsurgical procedures (such as balloon dilation for VPS). Similarly to our results, Mellion et al. demonstrated that parent-reported QOL scores in adolescents with mild CHD (such as VPS) were higher than in those with other biventricular CHD such as repaired TOF [18]. Other studies in TOF have demonstrated that parents perceive their children to have diminished self-esteem and general health [10]. In addition, the parent-reported QOL is diminished in children and adolescents with TOF and comparable to chronically ill children [28]. Contrary to our findings, Uzark et al. demonstrated that parent-perceived QOL in children and adolescents with CHD was similar to healthy children; however, the study found an incremental decrease in parent-perceived QOL with increasing severity of CHD [29].

Parent-reported PF was worse in the TOF group as compared to VPS. Other studies have also shown that adults with repaired TOF report worse physical health status despite self-reported QOL that is similar to healthy peers [30, 31]. Diminished PF has also been shown in adolescents with repaired TOF; however, worse PF was associated with RV systolic dysfunction as well as with concurrent symptoms and number of operations [10, 32]. We found worse parent-reported PF in TOF patients despite preserved right ventricular function, and while the aerobic capacity was low in the TOF group, it was not associated with reported PF. These findings suggest that factors aside from clinical status may play a role in parent-reported PF. One possible explanation is that parents of children with CHD generally underestimate their child's exercise capacity [33]. We did not find an association between

reported PF and aerobic capacity on CPET in this study, even though abnormal oxygen consumption was apparent in the TOF group.

The assessment of parental perception of their child's QOL is important as health care utilization is typically associated with parent-proxy QOL [34]. Familial factors beyond the degree of heart disease may drive parental perceptions of QOL. Adverse family relationships have been shown to negatively impact psychosocial QOL [17], whereas positive family characteristics, such as high family cohesion, increased expressiveness, and less intrafamilial conflict, have been shown to favorably influence parent-proxy QOL in CHD [17]. We found that the parent-proxy-reported family cohesion score was significantly worse in the TOF compared to the VPS group. Parental well-being and ability to cope with their child's heart disease play a significant role in family cohesion, where worse family cohesion is associated with higher parental psychological distress [35]. Our findings suggest that interventions to strengthen family welfare and family support that integrate both the child's individual needs as well as parental needs may improve psychosocial QOL.

Parental stress is also associated with QOL and higher levels of parental psychological distress have been associated with more severe heart defects at primary diagnosis [36, 37]. The initial diagnosis is a stressful time for parents of children with CHD and diagnosis is increasingly more common in the prenatal period [38, 39]. During prenatal counseling, parents of children with CHD place a greater importance on QOL of their future child, including the number of lifetime interventions and physical limitations than their fetal cardiologist [40]. Parental perceptions of their child's CHD and anticipated QOL formed during prenatal counseling may influence their expectations, psychological preparation, and coping with their child's illness. It is possible that despite the mild residual disease after surgical repair in this study group, psychological stress is higher in parents of children with TOF compared to VPS. This may be due to the parental perception of illness severity at initial diagnosis, given that patients with TOF undergo open heart surgery and possibly more future re-interventions than patients with VPS. Parental stress is associated with parental perception of illness and is greater in parents of children with repaired single-ventricle heart disease when compared to those with two-ventricle repairs [7]. Conversely, parental stress can also be unrelated to the severity of CHD, suggesting the need for practitioners to consider measures to address parental stress irrespective of underlying CHD diagnosis [41].

To our knowledge, QOL in VPS patients after balloon dilation has not been previously evaluated. We found that parent-proxy-reported QOL in VPS with mild residual disease after balloon dilation was similar to that in a normative healthy population. However, when compared with a national sample of patients with mild CHD (BAV) who did not require surgical intervention, both parent and patient QOL scores particularly with respect to psychosocial impact were lower. Our findings suggest that although VPS patients are in many cases considered to be on the mild end of the spectrum of CHD, the need for intervention such as balloon dilation might have a detrimental impact on their QOL, particularly from a psychosocial standpoint.

Risk factors aside from underlying disease such as lack of psychosocial support may lead to worse parental psychological distress and poor parent and child well-being [35]. The

availability of social support and resources to families with a child affected by CHD has been associated with better QOL possibly due to promotion of resilience and improved adaptation in patients with heart disease [42]. Furthermore, parents of children with CHD report worse QOL for themselves than parents of healthy children, and parents' own mental status, coping abilities, and self-perceived QOL may moderate the perception of their child's QOL [43, 44].

This study was limited by its small sample size, but significant differences that point to diminished general perception of health, PF, and family cohesion even in the face of mild disease were apparent. In both groups, the QOL data were acquired prospectively as part of research study protocols lending strength to our findings. Furthermore, we used both generic and disease-specific instruments to provide a multidimensional assessment of QOL. Because we matched patients with TOF to those with VPS and mild disease by duration and severity of PI, the resulting groups have mild residual disease as dictated by VPS so they are not generalizable to TOF patients with more severe disease.

Conclusions

Parent-reported QOL is affected in children with CHD, even in the context of mild residual disease. Furthermore, in certain domains, QOL is worse in TOF patients than in those with VPS in the context of mild residual disease. It is possible that having a relatively more severe disease (TOF) with open heart surgery early in life is the main driver of these differences. Our findings should alert health care providers that QOL might be diminished even in the face of mild residual disease. Strategies to improve family cohesion, manage parental stress, and promote coping may result in improved QOL.

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Abbreviations

QOL	Quality of life
TOF	Tetralogy of Fallot
VPS	Isolated valvar pulmonary stenosis
CMR	Cardiac magnetic resonance
CPET	Cardiopulmonary exercise test
RF	Regurgitant fraction
PI	Pulmonary insufficiency

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Table 1

Subject demographics, CMR, and CPET variables

	VPS	TOF	<i>P</i> values
Number of subjects	19	12	
Age at testing	15.8 ± 3.5	15.6 ± 2.4	0.70 *
Males (%)	63	67	0.72
Caucasian (%)	75	94	0.14
Years of exposure to PI	14 ± 4.1	14 ± 2.7	1.0 *
CMR			
Pulmonary regurgitant fraction (%)	17.9 ± 11.8	25.9 ± 11.3	0.05 *
RV end-diastolic volume (m_1/m_2)	94.1 ± 29.9	110.4 ± 24.6	0.14
RV end-systolic volume (m_1/m_2)	34.9 ± 16	44.8 ± 18.9	0.15
RV ejection fraction (%)	64.7 ± 7	60.9 ± 8.8	0.21
CPET			
Indexed oxygen pulse [(ml/O ₂)/(beat/m ²)]	6.5 ± 1.7	6.1 ± 1.5	0.57
Percent-predicted mV _{O₂}	82 ± 10	86 ± 6	0.19

* Subjects recruited to the VPS study were of similar age as those recruited as part of the TOF study. By study design, subjects were matched by duration and severity of pulmonary insufficiency

Table 2

Cardiac interventions

	TOF <i>n</i> = 12
Cardiac surgeries ^a	12
Complete TOF repair	12
Palliative procedure (BT shunt)	3
Reoperation for residual lesion	4
Cardiac catheterization procedures ^a	
Diagnostic catheterizations	
Preoperative	10
Postoperative	10
Interventional catheterizations	3

BT Blalock–Taussig^aTotal number of procedures in group

Table 3

CHQ-PF50: comparison of VPS and TOF with CHQ-PF50 normative sample

	Normative sample (n = 96)	VPS (n = 19)	P values*	TOF n	P values†
Physical functioning	96.5 ± 12.8	94.6 ± 19	0.589	11	88.9 ± 9.6 0.059
General health perceptions	72.8 ± 14.6	70.9 ± 15	0.606	11	53.6 ± 17 0.0001*
Bodily pain	81.8 ± 17.9	84.2 ± 21.2	0.605	11	94.6 ± 9.3 0.022*
Family activities	88.7 ± 19.5	90.1 ± 13.2	0.765	12	88.9 ± 13.9 0.974
Family cohesion	73.5 ± 22.1	80 ± 22.1	0.243	12	58.3 ± 31.2 0.035*
Role/social-physical	92.1 ± 21.6	94.7 ± 16.7	0.621	12	91.7 ± 20.7 0.948
Role/social-emotional/behavioral	92.3 ± 20.7	91.8 ± 21.6	0.924	12	88.9 ± 20.7 0.591
Parental time impact	85.4 ± 20.9	92.9 ± 16.7	0.143	12	97.2 ± 9.6 0.056
Parental emotional impact	77.3 ± 21.4	71.9 ± 25	0.330	11	77.3 ± 16.2 0.996
Self-esteem	76.8 ± 15.1	81.5 ± 15.7	0.220	12	83.7 ± 10.4 0.129
Mental health	76.6 ± 12.9	80 ± 11.3	0.287	12	79.9 ± 11.3 0.399
Behavior	76.3 ± 17.6	83.1 ± 14.5	0.117	11	77.3 ± 16.6 0.862
Physical summary score	53 ± 8.8	52.1 ± 10.4	0.693	11	49.6 ± 7.9 0.884
Psychosocial summary score	50 ± 8.9	50.7 ± 4.8	0.740	11	52.9 ± 7.9 0.298

Values are mean ± standard deviation

* P values comparing VPS to normative sample,

† P values comparing TOF to CHQ-PF50 normative sample

Table 4

PCQLI: comparison of VPS and TOF with mild CHD

	Mild CHD (<i>n</i> = 54)	VPS <i>n</i>	<i>P</i> values [*]	TOF <i>n</i>	<i>P</i> values [†]	
Parent						
Disease impact	41.2 ± 9.2	19	42.6 ± 6.7	12	41 ± 6.2	0.929
Psychosocial impact	41.1 ± 8.2	19	33.1 ± 8.1	9	26.7 ± 5.6	<0.0001
Total score	82.4 ± 16.8	19	75.6 ± 13.1	9	66.7 ± 11.3	0.009
Patient						
Disease impact	41.5 ± 7.4	18	39.3 ± 10.5	11	43.2 ± 5.3	0.469
Psychosocial impact	41.5 ± 8.1	18	30 ± 9	10	31.2 ± 6.9	0.0004
Total score	83 ± 14.2	18	69.4 ± 18.6	10	74.3 ± 9.9	0.069

Results reported as mean ± SD

* P values comparing VPS to mild CHD,

† P values comparing TOF to mild CHD

Table 5

PCQLI: comparison of VPS and TOF with severe CHD

	Severe CHD (n = 318)	VPS n	P values [*]	TOF n	P values [†]
Parent					
Disease impact	31.1 ± 8.9	19	42.6 ± 6.7	12	41 ± 6.2 0.0002
Psychosocial impact	33.4 ± 9.5	19	33.1 ± 8.1	9	26.7 ± 5.6 0.037
Total score	64.6 ± 17.4	19	75.6 ± 13.1	9	66.7 ± 11.3 0.717
Patient					
Disease impact	31.2 ± 8.3	18	39.3 ± 10.5	11	43.2 ± 5.3 <0.0001
Psychosocial impact	33.9 ± 9.3	18	30 ± 9	10	31.2 ± 6.9 0.36
Total score	65.2 ± 16.4	18	69.4 ± 18.6	10	74.3 ± 9.9 0.082

Results reported as mean ± SD

^{*}P-values comparing VPS to severe CHD.

[†]P-values comparing TOF to severe CHD

Table 6

Summary of CHQ-PF50: comparison of VPS with TOF

Variables	VPS	TOF	<i>P</i> values
Global health	93 (85–100)	85 (60–100)	0.180
Physical functioning	100 (100)	89 (78–100)	0.010 *
Role/social–emotional/behavioral	100 (100)	100 (83–100)	0.476
Role/social–physical	100 (100)	100 (100)	0.626
general health perceptions	75 (58–83)	58 (43–67)	0.016 *
Bodily pain/discomfort	100 (70–100)	100 (80–100)	0.221
Family activities	96 (79–100)	94 (81–100)	0.627
Family cohesion	85 (60–100)	60 (30–85)	0.048 *
Parental time impact	100 (89–100)	100 (100)	0.251
Parental emotional impact	75 (50–100)	75 (58–92)	0.695
Self-esteem	83 (75–92)	83 (77–90)	0.932
Mental health	80 (75–90)	80 (70–89)	0.902
Behavior	85 (71–96)	73 (64–93)	0.298
Global behavior	85 (60–100)	85 (60–85)	0.169
Changes in health	Same 79%	Same 64%	0.824
	Better 21%	Better 27%	
	Worse 0%	Worse 9%	
Physical summary score	55 (50–57)	51 (47–54)	0.169
Psychosocial summary score	52 (48–55)	53 (46–59)	0.561

* Results reported as median (interquartile range)

Table 7

CHQ-CF87 and PCQLI: comparison of VPS with TOF

Subscales (abbreviation)	VPS	TOF	P values
Global health (GGH)	85 (60–100)	85 (60–100)	0.842
Physical functioning (PF)	100 (93–100)	96 (96–100)	0.913
Role/social limitations: emotional (RE)	100 (78–100)	100 (100)	0.057
Role/social limitations: behavioral (RB)	100 (100)	100 (100)	0.311
Role/social limitations: physical (RP)	100 (100)	100 (100)	0.39
General health perceptions (GH)	70 (53–82)	69 (61–74)	0.730
Bodily pain/discomfort (BP)	80 (60–100)	80 (70–90)	0.778
Family activities (FAs)	100 (63–100)	96 (90–100)	0.640
Family cohesion (FC)	85 (60–100)	60 (30–85)	0.066
Self-esteem (SE)	77 (68–96)	79 (73–85)	0.541
Mental health (MH)	72 (63–80)	82 (73–88)	0.036*
Behavior (BE)	82 (68–88)	76 (73–88)	0.846
Global behavior (GBE)	85 (60–100)	85 (60–100)	0.909
Changes in health (CH)	Same 58%	Same 75%	0.633
	Better 37%	Better 25%	
	Worse 5%	Worse 0%	
PCQLI			
Patient			
Disease impact	41 (35–47)	44 (39–47)	0.380
Psychosocial impact	31 (29–33)	32 (28–35)	0.648
Total patient score	73 (67–79)	78 (66–81)	0.486
Parent			
Disease impact	44 (39–49)	40 (38–46)	0.360
Psychosocial impact	31 (28–38)	29 (24–29)	0.126
Total parent score	76 (64–83)	69 (61–76)	0.115

* Results reported as median (interquartile range)