Need for Routine Screening of Health-Related Quality of Life in Families of Young Children with Complex Congenital Heart Disease

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Objectives To assess health-related quality of life (HRQOL) in families of young children with complex congenital heart disease (CHD), and identify the demographic, clinical, and psychosocial factors that place these children and their mothers at greater risk of vulnerability.

Study design This cross-sectional study took place from June 2015 to October 2016 at The Sydney Children's Hospitals Network Cardiac Service, Australia. Mothers of a child aged 1-5 years with either single ventricle CHD or CHD requiring neonatal biventricular repair were invited to participate. Eighty-seven mothers completed a suite of validated measures, including the Pediatric Quality of Life Inventory, which assessed the outcomes of child and maternal HRQOL.

Results Sixty percent of children with single ventricle CHD and 25% of children with biventricular repair had total Pediatric Quality of Life Inventory scores within the at-risk range. Lower child HRQOL was strongly associated with single ventricle CHD ($\beta = -0.38$; $P < .001$), physical comorbidity ($\beta = -0.32$; $P = .001$), feeding difficulties ($\beta = -0.26$; $P = .008$), and greater maternal psychological stress ($\beta = -0.18$; $P = .045$), accounting for 52% of the variance in child HRQOL. Lower maternal HRQOL was strongly associated with poorer family functioning ($\beta = 0.61$; $P < .001$), greater maternal psychological stress ($\beta = -0.23$; $P = .004$), child physical comorbidity ($\beta = -0.17$; $P = .01$), and a 'difficult' child temperament ($\beta = -0.14$; $P = .01$), accounting for 73% of the variance in maternal HRQOL.

Conclusions Lower HRQOL is common in young children with complex CHD, particularly single ventricle CHD. Several predictors of HRQOL are potentially modifiable, offering possible pathways for prevention and early intervention. Routine screening is a necessary first step toward developing models of care to improve HRQOL in this population. *(J Pediatr 2019;205:21-8).*

Young children with congenital heart disease (CHD) requiring neonatal surgical intervention experience challenges that may influence health-related quality of life (HRQOL). These factors include invasive medical procedures, extended hospitalization, potential neurologic injury, decreased exercise capacity, intense emotional responses such as fear, and uncertainty about the future. Children with the most complex cardiac conditions, particularly single ventricle CHD, generally experience poorer quality of cardiovascular circulation, greater intensity of medical care, more planned surgical procedures, greater morbidity, and poorer long-term health, compared with children requiring early biventricular repair. Caring for a child with complex CHD may also have profound physical, psychological, social, and financial consequences for parents and families.[1]

Despite recognition that HRQOL varies considerably with age,[2] most literature focuses on HRQOL in children aged >5 years with CHD or treats all children and adolescents as a collective group. Infants and young children are largely overlooked in the existing literature on HRQOL in children with CHD, limiting the capacity for prevention and early intervention strategies to bolster HRQOL in this vulnerable population. This oversight is significant, because the foundation for future health and well-being is laid during early childhood.[3] The very limited available literature examining HRQOL in young children with CHD has yielded

CHD	Congenital heart disease
HRQOL	Health-related quality of life
PedsQL	Pediatric Quality of Life Inventory
SCHN	The Sydney Children's Hospitals Network

mixed results. Some studies have found that children with CHD have impaired overall HRQOL compared with normative data,[4,5] whereas others have found no differences in HRQOL in children with CHD compared with healthy controls,[6] or even that children with CHD have better overall HRQOL and social functioning compared with population norms.[7]

The primary aim of this study was to assess HRQOL in young children with functional single ventricle CHD and planned progression to a Fontan circulation (single ventricle group), or CHD requiring neonatal biventricular surgical repair and cardiopulmonary bypass (biventricular group). Secondary aims were to examine maternal self-reported HRQOL, psychological well-being, and impact of CHD on the family, and to identify the demographic, clinical, and psychosocial factors associated with child and maternal HRQOL. Finally, the study aimed to characterize mothers' engagement with health professionals for emotional support. It was hypothesized that HRQOL would be lower for young children with CHD compared with normative data,[4] and for children with single ventricle CHD and their mothers compared with children who underwent biventricular repair and their mothers.[6,8] Lower child HRQOL was also hypothesized to be associated with hypoplastic left heart,[9] physical comorbidity,[7] greater maternal psychological stress,[6,10] and lower household income.[11]

Methods

Participants for this cross-sectional study were recruited through The Sydney Children's Hospitals Network (SCHN) Cardiac Service, which includes The Children's Hospital at Westmead and Sydney Children's Hospital at Randwick, and treats all children with complex CHD in New South Wales, Australia (Institutional Review Board Approval: LNR/15/SCHN/182). Mothers of a child aged 1-5 years with either functional single ventricle CHD with planned progression to a Fontan circulation (single ventricle group) or CHD requiring neonatal biventricular surgical repair with cardiopulmonary bypass (biventricular group) were invited to participate. The rationale for choosing a pre-Fontan single ventricle group was to maintain focus on a group of vulnerable young children that the existing literature tends to neglect, deepening our understanding of factors contributing to outcomes in this group and providing insight into how to best support families before their child's Fontan procedure. Exclusion criteria included children who had undergone the Fontan procedure, mothers aged <17 years, those with severe psychiatric conditions or cognitive impairment, and those unable to complete the questionnaire in English.

An invitation letter, information sheet, and consent form were sent to eligible mothers, with follow-up telephone calls, as appropriate. Consenting participants received a paper- or web-based questionnaire, according to preference.

Measure selection was based on recommendations of a systematic review of HRQOL models,[12] a systematic review of the CHD literature,[13] and extensive pilot work. To maximize developmental sensitivity, survey instruments were tailored to child age, with distinct validated measures for children aged 1-2 years, 2.0-3.5 years, 3.5-4.0 years, and 5 years.

HRQOL was assessed using the Pediatric Quality of Life Inventory (PedsQL).[14] Items are rated using a 5-point scale, then scores are reversed, transformed to a linear scale (0-100, with higher scores representing greater HRQOL), and the mean of each subscale calculated. The PedsQL allows comparison with a database of scores for healthy and ill children.[14,15] A cutoff of >1 SD below the normative mean indicates at-risk status and a need for clinical assessment.[16] PedsQL instruments used included the following measures. (1) The PedsQL Infant Scale (45 items)[15] is a parent-report measure assessing physical symptoms (Cronbach alpha in present sample, $\alpha = 0.81$), and physical ($\alpha = 0.81$), emotional ($\alpha = 0.88$), social ($\alpha = 0.79$), and cognitive ($\alpha = 0.94$) functioning in children aged 12-24 months. The mean of all items gives the total PedsQL score ($\alpha = 0.92$), and was used as a parent-proxy reported measure of infant HRQOL. (2) The PedsQL 4.0 Generic Core for Toddlers and Young Children (23 items)[14] is a parent-report measure for children aged 2-4 and 5-7 years assessing physical ($\alpha = 0.90$), emotional ($\alpha = 0.85$), social ($\alpha = 0.83$), and cognitive ($\alpha = 0.94$) functioning. The mean of all items gives the total PedsQL score ($\alpha = 0.85$), used as a parent-proxy reported measure of child HRQOL. (3) The PedsQL 3.0 Cardiac Module (27 items)[17] is a parent-report, disease-specific measure for children aged 2-4 and 5-7 years assessing heart problems and treatment ($\alpha = 0.89$), treatment II ($\alpha = 0.96$), perceived physical appearance ($\alpha = 0.88$), treatment anxiety ($\alpha = 0.96$), cognition ($\alpha = 0.86$), and communication ($\alpha = 0.96$).

Measures of maternal HRQOL, psychological distress and family functioning included the following metrics. (1) The PedsQL 2.0 Family Impact Module (36 items)[18] contains 8 subscales. The first 4 subscales assess parents' own physical ($\alpha = 0.93$), emotional ($\alpha = 0.92$), social ($\alpha = 0.91$), and cognitive ($\alpha = 0.96$) functioning. The mean of these 4 subscales gives the total PedsQL score ($\alpha = 0.97$), which is used as a measure of maternal HRQOL in the present study (ie, mothers' reports of their own HRQOL). The second 4 subscales assess how a child's health condition impacts their family according to 4 domains, namely, communication ($\alpha = 0.83$), worry ($\alpha = 0.82$), daily activities ($\alpha = 0.94$), and family relationships ($\alpha = 0.97$). The mean of these 4 subscales gives a family functioning summary score ($\alpha = 0.95$). The mean of all 36 Family Impact Module items from all 8 scales gives a total family impact score that encompasses both maternal HRQOL and family functioning summary scores. (2) On the Depression Anxiety Stress Scales (21 items),[19] mothers were asked to report symptoms of depression ($\alpha = 0.85$), anxiety ($\alpha = 0.78$), and stress ($\alpha = 0.86$) that they themselves had experienced over the preceding week using a 4-point scale (0-3). Higher scores indicate greater distress.

Potential predictors of HRQOL included demographic factors (12 items): child age and sex, maternal age, education, marital status, country of birth, Aboriginality, primary spoken language, number of children, rurality, gross weekly household income (compared with National average of $AUD2063), and perceived financial strain (0 [not at all] to 4 [extremely]). Child

clinical characteristics (8 items) included data from the SCHN databases, including cardiac diagnoses, surgical intervention(s), comorbidities (ie, presence of another chronic health condition or syndrome), cardiopulmonary bypass time, extracorporeal membrane oxygenation use, and length of neonatal intensive care unit stay, pediatric intensive care unit stay, and total pediatric hospital admission (consecutive and nonconsecutive days) to time of survey completion. Perceived seriousness of CHD (1 item) measured parents' perception of the seriousness of their child's CHD (from 0 [not at all serious] to 4 [extremely serious]). The Short Temperament Scale for Toddlers (30 items) or Short Temperament Scale for Children (30 items)[20] measured parents' perceptions of their child's temperament, for children aged 1-3.5 years and 3.5-8 years, respectively. The easy/difficult temperament subscale scores were used (Short Temperament Scale for Toddlers: $\alpha = 0.65$, Short Temperament Scale for Children: $\alpha = 0.76$). Scores >1 SD above the normative mean indicate a difficult temperament.[20] The Behavioral Pediatric Feeding Assessment Scale (35 items) assesses child feeding behaviors. Scores >84 indicate feeding difficulties ($\alpha = 0.65$).[21] Access to emotional support from health professionals (4 items) assesses whether mothers recalled being offered emotional support from a health professional during their child's cardiac care, perceived helpfulness of the support, difficulties accessing support, and anticipated responses to future offers of psychological referral by their cardiac team.

Statistical Analyses

IBM Statistical Package for the Social Sciences (SPSS Inc, Chicago, Illinois) 23.0 was used for analyses. Potential differences between participants and nonparticipants were examined using Pearson χ^2 tests or t tests, as appropriate. PedsQL scores were compared with normative data using 1-sample t tests. Differences in PedsQL scores between the single ventricle and biventricular groups were examined using independent samples t tests or Mann-Whitney U tests, as appropriate. Associations between child total PedsQL scores (parent-proxy report), maternal total PedsQL scores, and dichotomous variables were assessed using independent samples t tests or Mann-Whitney U tests, as appropriate. Associations between child total PedsQL scores (parent-proxy report), maternal total PedsQL scores, and continuous variables were assessed using Spearman rank or Pearson correlations, as appropriate.

Multiple linear regression was used to create 2 models, one for child total PedsQL scores, and one for maternal total PedsQL scores. After assessing multicollinearity, variables with $P < .10$ in bivariate analyses were included in multivariate modelling. Depression Anxiety Stress Scales subscales demonstrated collinearity, so only the stress subscale was included, capturing general psychological stress. Total length of hospital, neonatal intensive care unit, and pediatric intensive care unit admission demonstrated collinearity, so only total hospital admission was included. Progressive, backward elimination was used to achieve 2 final regression models incorporating variables with $P < .05$. Child and maternal age, maternal education level, and household income were tested as potential confounders, added individually to each model. A variable causing >10% change was considered a confounder and retained in the model.

Results

Of 203 eligible mothers from 201 families, 22 were uncontactable. Eighty-seven mothers completed the questionnaire, yielding a participation rate of 49% among eligible, contactable families (87 of 179). Participants and nonparticipants did not differ on child age ($P = .59$) or single ventricle/biventricular status ($P = .81$), but did differ by child sex ($P = .02$), with mothers of boys (58%) more likely to participate than mothers of girls (31%). The mean maternal age was 34.0 years ($SD = 5.8$) and the mean child age was 2.7 years ($SD = 1.2$; Table I); 30% of participants lived in regional or rural New South Wales, 48% had obtained a university degree, 60% received a gross weekly household income below the national average, and most participants (74%) had received their child's diagnosis antenatally.

Infants (aged 1-2 years) with CHD had lower total PedsQL, and psychosocial and physical summary scores, as well as lower physical and cognitive functioning, and higher social functioning compared with normative data (Table II). Children (aged 2-5 years) with CHD had lower emotional functioning scores compared with normative data, with no other differences in PedsQL scores.

Sixty percent of children with single ventricle CHD (86% of infants, 52% of children aged 2-5 years) and 25% of children with biventricular repair (48% of infants, 11% of children) had total PedsQL scores within the at-risk range, indicating a need for clinical assessment (Table III; available at www.jpeds.com). Seventy percent of the single ventricle group and 53% of the biventricular group had scores in ≥1 HRQOL domain warranting clinical assessment. Infants with single ventricle CHD had lower total PedsQL scores and physical summary and physical functioning scores compared with the biventricular group (Table IV). Children aged 2-5 years with single ventricle CHD had lower total PedsQL scores and physical summary and school functioning scores, and greater heart problems and treatment anxiety, compared with the biventricular group. Seventeen percent of children aged <3.5 years and 48% aged >3.5 years had scores indicating a difficult temperament, whereas 29% of children had scores indicating feeding problems requiring clinical investigation.

Mothers in the single ventricle group reported that their own HRQOL (single ventricle: mean, 64.87; SD, 16.83; biventricular: mean, 75.38; SD, 19.97; $P = .02$), family functioning (single ventricle: mean, 67.53; SD,19.71; biventricular: mean, 80.92; SD,23.31; $P = .01$), and total family impact (single ventricle: mean, 63.61; SD,15.51; biventricular: mean, 76.43; SD, 18.74; $P = .002$) scores were lower than for mothers in the biventricular group (Table V; available at www.jpeds.com). Mean scores indicating symptoms of depression ($P = .20$), anxiety ($P = .28$), and psychological stress ($P = .37$) experienced by mothers did not differ between groups. Moderate to severe symptoms of depression, anxiety, and stress were reported by 14%, 9%, and 12% of mothers, respectively.

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Table I. Maternal demographic and child clinical characteristics

Variables	Single ventricle group (n = 30)	Biventricular group (n = 57)	Total sample (N = 87)
Marital status			
Married or in a committed relationship	26 (87)	52 (91)	78 (90)
Not married	4 (13)	5 (9)	9 (10)
Education level			
University degree	15 (50)	27 (47)	42 (48)
No university degree	15 (50)	30 (53)	45 (52)
Gross weekly household income			
Below national average	23 (77)	29 (51)	52 (60)
Above national average	6 (20)	21 (37)	27 (31)
Area of residence			
Metropolitan	20 (67)	39 (68)	59 (68)
Rural or regional	9 (30)	17 (30)	26 (30)
Aboriginal or Torres Strait Islander			
Yes	0 (0)	0 (0)	0 (0)
No	29 (97)	56 (98)	85 (98)
Country of birth			
Australia	22 (73)	44 (77)	66 (76)
Other	8 (27)	12 (21)	20 (23)
Primary language spoken at home			
English	24 (80)	52 (91)	76 (87)
Other	6 (20)	4 (7)	10 (11)
Maternal age, years	33.6 ± 6.5	34.1 ± 5.5	34.0 ± 5.8
No. of children	2.9 ± 1.1	2.8 ± 1.1	2.8 ± 1.1
Perceived financial strain, scale 0-4	1.5 ± 1.1	1.5 ± 1.0	1.5 ± 1.0
Perceived seriousness of child's CHD, scale 0-4	3.2 ± 0.8	2.9 ± 1.1	3.0 ± 1.0
Child clinical characteristics			
Child sex			
Male	21 (70)	41 (72)	62 (71)
Female	9 (30)	16 (28)	25 (29)
Primary CHD diagnosis			
Hypoplastic left heart	9 (30)	0 (0)	9 (10)
Double outlet right ventricle	9 (30)	8 (14)	17 (20)
Transposition of the great arteries	5 (17)	29 (51)	34 (39)
Truncus arteriosus	0 (0)	6 (11)	6 (7)
Atrioventricular septal defect	0 (0)	2 (4)	2 (2)
Total anomalous pulmonary venous return	0 (0)	2 (4)	2 (2)
Aortic arch abnormality	0 (0)	6 (11)	6 (7)
Other*	7 (23)	4 (7)	11 (13)
Time of diagnosis			
Antenatal	26 (87)	38 (67)	64 (74)
Postnatal	4 (13)	19 (33)	23 (26)
Comorbidity‡			
Yes	7 (23)	12 (21)	19 (22)
No	23 (77)	45 (79)	68 (78)
Extracorporeal membrane oxygenation			
Yes	5 (17)	4 (7)	9 (10)
No	25 (83)	51 (90)	76 (87)
Current use of medication†			
Yes	19 (83)	1 (3)	20 (34)
No	4 (17)	34 (97)	38 (66)
Child age, y	2.9 ± 1.2	2.7 ± 1.2	2.7 ± 1.2
Total length of hospital admission, d§	74.4 ± 57.2	32.5 ± 33.0	47.3 ± 47.3
Total length of NICU admission, d§	14.3 ± 13.1	13.3 ± 12.0	13.6 ± 12.3
Total length of PICU admission, d§	25.4 ± 37.8	10.3 ± 12.2	15.6 ± 25.3
Cardiopulmonary bypass time, h	3.6 ± 2.2	3.2 ± 1.6	3.4 ± 1.8

NICU, neonatal intensive care unit; *PICU*, pediatric intensive care unit.
Some percentages will not add to 100% owing to missing data.
Values are n (%) or mean ± SD.
*Other primary CHD diagnoses in the single ventricle group included hypoplastic right ventricle (n = 4) and pulmonary atresia (n = 1). Other primary CHD diagnoses in the biventricular group included atrial septal defect (n = 2), aortic stenosis (n = 1), and tetralogy of Fallot (n = 1).
†Assessed for children aged 2-5 years (n = 57) using the PedsQL Cardiac Module, which is not available for infants.
‡Comorbidities included heterotaxy syndrome (n = 5), velocardiofacial syndrome (n = 3), and asthma (n = 3).
§Includes consecutive and nonconsecutive days.

After adjusting for household income, the final multiple linear regression model accounted for 52% of the variance in child HRQOL (child HRQOL; **Table VI**). The mean total PedsQL scores were 12.12 points lower for children in the single ventricle group than the biventricular group, and 11.78 points lower for children with a comorbidity than those without. For every 1-unit increase in frequency of child feeding problems, child total PedsQL scores decreased by 0.20 units, and for every

Table II. Mean PedsQL scores for children with CHD compared with normative data, assessed using the PedsQL Infant Scales (age 1-2 years) and PedsQL Generic Core Scales (age 2-5 years)*

Variables	Children with CHD (n = 87)		Normative data[21,22]		Test statistics		
	n	Mean ± SD	n	Mean ± SD	t	df	P
PedsQL Infant Scales, 0-100							
Total PedsQL score	28	76.04 ± 12.58	141	85.55 ± 8.74	−4.00	27	**<.001**
Physical summary score	28	80.14 ± 12.98	141	88.84 ± 7.68	−3.55	27	**.001**
Physical functioning	28	78.05 ± 17.17	141	90.32 ± 8.96	−3.78	27	**.001**
Physical symptoms	28	81.96 ± 14.52	141	87.54 ± 9.29	−2.03	27	.05
Psychosocial summary score	28	73.04 ± 15.88	141	83.12 ± 11.02	−3.36	27	**.002**
Emotional functioning	28	72.07 ± 15.42	141	78.60 ± 12.80	−2.24	27	.03
Social functioning	28	90.54 ± 10.48	141	84.65 ± 15.76	2.97	27	**.006**
Cognitive functioning	28	64.53 ± 27.50	141	91.14 ± 10.77	−5.12	27	**<.001**
PedsQL Generic Core Scales, 0-100							
Total PedsQL score	59	77.73 ± 17.49	10 070	81.34 ± 19.98	−1.59	58	.12
Physical summary score	59	81.78 ± 21.95	10 050	83.26 ± 15.84	−0.52	58	.61
Psychosocial summary score	59	75.16 ± 18.20	10 071	80.22 ± 16.99	−2.14	58	.04
Emotional functioning	59	68.96 ± 20.74	10 044	80.28 ± 20.08	−4.19	58	**<.001**
Social functioning	59	81.69 ± 19.38	10 036	82.15 ± 20.16	−0.18	58	.86
School functioning	37	73.78 ± 19.53	8466	76.91 ± 15.92	−0.97	58	.34

P values significant at the <.025 level are highlighted in bold typeface. A significance level of P < .025 was used to account for the multiple tests carried out.
*Lower scores indicate a poorer HRQOL.

1-unit increase in maternal psychological stress (mothers' reports of their own stress levels), the child total PedsQL scores decreased by 0.74 units. A range of factors found to be associated with poorer child HRQOL at the bivariate level were not retained in the final regression model, including diagnosis of hypoplastic left heart, greater total length of hospital admission, a difficult child temperament, poorer family functioning, and poorer maternal HRQOL (**Table VII**; available at www.jpeds.com).

After adjusting for maternal age, the final multiple linear regression model accounted for 73% of the variance in maternal HRQOL (ie, mothers' reports of their own HRQOL; **Table VI**). For every 1-unit decrease in family functioning, maternal total PedsQL scores decreased by 0.52 units. For every 1-unit increase in maternal psychological stress, maternal total PedsQL scores decreased by 1.24 units. The mean total PedsQL scores were 7.92 points lower for mothers of a child with a comorbidity than those without and 6.03 points lower for mothers who perceived their child's temperament as difficult compared with those who did not. A range of factors found to be associated with poorer maternal HRQOL at the bivariate level were not retained in the final regression model, including single ventricle surgical pathway, greater total length of hospital admission, lower household income, greater perceived financial strain, greater frequency of child feeding problems, and poorer child HRQOL (**Table VIII**; available at www.jpeds.com).

Sixty-three percent of mothers recalled being offered emotional support from a health professional involved in their child's cardiac care. Of these, 60% of mothers engaged with the support, with 97% perceiving it as helpful. Overall, 18% of mothers reported difficulty accessing professional emotional support; lack of time (50%), the belief they should cope without assistance (50%), cost (36%), and uncertainty about how to access support (29%) were common reasons. When

asked their anticipated response if offered a psychological care referral, 52% of mothers anticipated feeling well cared for, 31% interested, and 26% understood.

Discussion

As hypothesized, children with single ventricle CHD had lower parent-proxy reported HRQOL (total PedsQL scores) and poorer physical functioning than children with biventricular repair. Children aged 2-5 years with single ventricle CHD also had lower school functioning, greater heart problems, and greater treatment-related anxiety compared with the biventricular group. Factors contributing to lower HRQOL in children with single ventricle CHD may include multiple, complex surgical interventions,[22] longer hospitalization,[23] and an increased likelihood of physical limitations and neurodevelopmental impairment.[23] In this study, children with single ventricle CHD had not yet undergone the Fontan procedure, whereas children in the biventricular group had completed their major surgical interventions. This finding may influence the child's objective physical functioning and parents' perceptions of their child's well-being. Longitudinal HRQOL assessment is required to understand how HRQOL may evolve over time after cardiac surgery. We also found various clinical differences between the single ventricle and biventricular groups (eg, time of cardiac diagnosis, current use of medication).

More than one-half (53%) of children with biventricular repair and 70% of children with single ventricle CHD (59% of the total sample) scored within the at-risk range on ≥1 PedsQL domain, demonstrating the need for routine HRQOL assessment in pediatric cardiac care. Our results were substantially higher than the limited existing literature reporting on the proportion of young children in the at-risk range for HRQOL using a comparable measure. Uzark et al, for example,

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Table IV. PedsQL scores for children following a single ventricle pathway compared with children who have undergone biventricular repair*

Variables	Single ventricle group (n = 30)		Biventricular group (n = 57)		Test statistics		
	n	Median (IQR)	n	Median (IQR)		Z	P
PedsQL Infant Scales (0-100)							
Total generic PedsQL score	7	62.22 (14.74)	21	78.89 (12.50)		−2.52	**.01**
Physical summary score	7	69.74 (18.42)	21	87.50 (18.50)		−2.95	**.003**
Physical functioning	7	55.56 (22.22)	21	86.11 (18.06)		−3.75	**<.001**
Physical symptoms	7	77.50 (25.00)	21	90.00 (20.00)		−1.23	.22
Psychosocial summary score	7	61.54 (21.15)	21	78.85 (21.63)		−1.75	.08
Emotional functioning	7	60.42 (16.67)	21	72.92 (17.05)		−1.28	.20
Social functioning	7	90.00 (25.00)	21	95.00 (20.00)		−0.69	.49
Cognitive functioning	7	50.00 (38.89)	21	75.00 (38.89)		−2.82	.06
	n	Mean (SD)	n	Mean (SD)	t	df	P
PedsQL Generic Core Scales (0-100)							
Total generic PedsQL score	23	68.03 (20.47)	36	83.93 (11.94)	3.38	32	**.002**
Physical summary score	23	65.30 (23.67)	36	92.31 (12.38)	5.05	30	**<.001**
Psychosocial summary score	23	70.10 (22.05)	36	78.39 (14.69)	1.59	35	.12
Emotional functioning	23	66.68 (24.67)	36	70.42 (18.02)	0.63	57	.51
Social functioning	23	74.78 (21.56)	36	86.11 (16.70)	2.27	57	.03
School functioning	12	62.78 (22.68)	25	79.07 (15.71)	2.55	35	**.02**
	n	Median (IQR)	n	Median (IQR)		Z	P
PedsQL Cardiac Module (0-100)[†]							
Heart problems and treatment	23	50.00 (32.24)	35	89.29 (14.29)		−5.23	**<.001**
Treatment II[‡]	19	100.00 (0.00)	1	91.67 (0.00)		-	-
Perceived physical appearance	23	100.00 (8.33)	35	100.00 (0.00)		−1.43	.15
Treatment anxiety	23	75.00 (50.00)	35	100.00 (25.00)		−2.43	**.02**
Cognition or thinking	23	75.00 (41.67)	35	83.33 (40.00)		−0.74	.46
Communication	22	66.67 (77.08)	32	75.00 (33.33)		−1.28	.20

Medians and corresponding IQR are reported for non-normally distributed data. The IQR represents the difference between quartile 3 and quartile 1. P values significant at the <.025 level are highlighted in bold typeface. A significance level of P < .025 was used to account for the multiple tests carried out.
*Lower scores indicate a lower HRQOL.
†The PedsQL suite does not contain a cardiac module for infants.
‡Treatment II items can only be completed for children currently prescribed cardiac medication.

found that only 3% of children aged 2-4 years with heart disease had a PedsQL psychosocial summary score in the at-risk range (compared with 27% in our study), and did not report on the proportion of children in the at-risk range for the physical summary score.[24] This difference could be influenced by the greater inclusion of children with more complex CHD in our study, which may further suggest the vulnerability of this specific population.

The presence of single ventricle CHD, comorbidity, child feeding difficulties, and greater maternal psychological stress were associated with lower parent-proxy reported child HRQOL. Although feeding difficulties are common in

Table VI. Multiple linear regression models showing predictors of child and maternal HRQOL, measured using total PedsQL scores

Variables	B	95% CI	β	P	Model R²
Child HRQOL*					
Single ventricle pathway	−12.12	−17.71 to −6.52	−0.38	<.001	0.52
Presence of comorbidity	−11.78	−18.55 to −5.00	−0.32	.001	
Frequency of child feeding problems	−0.20	−0.35 to −0.06	−0.26	.008	
Maternal psychological stress	−0.74	−1.47 to −0.02	−0.18	.045	
Gross weekly household income[†]	3.18	−2.47 to 8.84	0.10	.27	
Maternal HRQOL					
Presence of comorbidity	−7.92	−13.99 to −1.84	−0.17	.01	0.73
Family functioning	0.52	0.38 to 0.65	0.61	<.001	
Maternal psychological stress	−1.24	−2.08 to −0.40	−0.23	.004	
Difficult child temperament	−6.03	−11.47 to −0.58	−0.14	.01	
Maternal age[†]	−0.15	−0.57 to 0.28	0.04	.50	

*Parent-proxy reported child total PedsQL scores.
†Identified confounder.

children with complex CHD[25] and have been linked to lower HRQOL in other chronically ill children,[26] only behavioral problems more generally have been associated with a lower HRQOL in young children with CHD.[10] Behavioral problems are also linked to increased parental stress,[6,10] potentially further influencing parental perceptions of child HRQOL. The association between maternal psychological stress and child emotional, behavioral, and neurocognitive outcomes is well-established,[10,27] and is likely influenced by both biological and environmental factors. Because both maternal psychological stress and child feeding difficulties are modifiable, these findings signal the importance of early identification and treatment.

With the exception of child comorbidity, the strongest correlates of maternal HRQOL (ie, mothers' reports of their own HRQOL), were psychosocial factors: poorer family functioning, increased maternal psychological stress, and difficult child temperament. This finding supports the literature identifying child clinical factors as secondary predictors of maternal HRQOL to psychosocial factors,[28] suggesting that interventions to bolster parental psychological adaptation and family functioning could improve maternal HRQOL. Participants' positive responses to future offers of psychological referral and the overwhelmingly positive experiences of those in our sample who had received emotional support demonstrate the demand for integration of psychological services in pediatric cardiac settings.

Perceiving one's child as difficult was also associated with a lower maternal HRQOL. Difficult child temperament has previously been associated with increased psychological stress in mothers of children with CHD,[29] with a bidirectional association between child temperament and maternal stress having been suggested.[30] Clinically, the focus should be on supporting mothers to respond to their child's individual behavioral style in a way that fosters secure attachment.[29]

There are limitations to this study. The sample size was moderate (and small for infants in the single ventricle group), the response rate modest, and the cross-sectional design limits causal inference. Age at Fontan procedure is older in Australia (median, 4.8 years; IQR, 3.9-5.9 years)[31] compared with the US (mean, 2.9 ± 0.9 years),[32] potentially limiting the generalizability of these results. The potential impact of using parent-proxy reports for child outcomes must also be acknowledged. Studies in older children have described cross-informant variation between parent-proxy and self-reported HRQOL[2]; however, in research involving young children with varying developmental abilities, parent-proxy report may be the best available measure. Finally, the generalizability of findings is restricted to mothers. Inclusion of fathers' perspectives in future studies will offer a more complete picture.

There are several direct clinical implications of this study. Routine HRQOL screening is a necessary first step toward identifying areas of difficulty and offering interventions to minimize early adversity effects. We recommend a tiered approach to routine HRQOL evaluation, in line with the model of psychological care proposed by Kasparian et al.[33] This process includes a pathway for universal psychoeducation and trauma-informed care, as well as screening, assessment, and intervention where indicated. Screening of HRQOL using a tool such as the PedsQL has been shown to be practical and useful in the pediatric cardiology outpatient setting,[34] and could be incorporated into annual visits to foster regular screening while minimizing burden on families. ■

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Table III. Proportion of children scoring below the clinical cutoff* for each HRQOL domain

Outcome variables	Single ventricle group (n = 30)		Biventricular group (n = 57)		Total sample (n = 87)	
	n	No. of children scoring in the at-risk range (%)	n	No. of children scoring in the at-risk range (%)	n	No. of children scoring in the at-risk range (%)
PedsQL Infant Scales						
Total generic HRQOL score	7	6 (86)	21	10 (48)	28	16 (57)
Physical summary score	7	7 (100)	21	7 (33)	28	14 (50)
Physical functioning	7	7 (100)	21	7 (33)	28	14 (50)
Physical symptoms	7	4 (57)	21	5 (24)	28	9 (32)
Psychosocial summary score	7	5 (71)	21	7 (33)	28	12 (43)
Emotional functioning	7	4 (57)	21	5 (24)	28	9 (32)
Social functioning	7	2 (29)	21	6 (29)	28	8 (29)
Cognitive functioning	7	6 (86)	21	9 (43)	28	15 (54)
PedsQL Generic Core scales						
Total generic HRQOL score	23	12 (52)	36	4 (11)	59	16 (27)
Physical summary score	23	10 (44)	36	1 (3)	59	11 (19)
Psychosocial summary score	23	9 (39)	36	7 (19)	59	16 (27)
Emotional functioning	23	11 (48)	36	12 (33)	59	23 (39)
Social functioning	23	5 (22)	36	3 (8)	59	8 (14)
School functioning	12	7 (58)	25	3 (12)	37	10 (27)
Any PedsQL subscale		21 (70)		30 (53)		51 (59)

*More than 1 SD below the normative mean.

Table V. Maternal HRQOL, maternal psychological well-being and family functioning

Measures	Total sample (N = 87)		Single ventricle group (n = 30)		Biventricular group (n = 57)		Test statistics*		
	n	Mean (SD)	n	Mean (SD)	n	Mean (SD)	t	df	P
PedsQL Family Impact Module (0-100)									
Total Family Impact	84	72.16 (18.65)	28	63.61 (15.51)	56	76.43 (18.74)	3.12	82	**.002**
Maternal HRQOL	84	71.88 (19.53)	28	64.87 (16.83)	56	75.38 (19.97)	2.39	82	**.02**
Family Functioning	84	76.45 (22.95)	28	67.53 (19.71)	56	80.92 (23.31)	2.61	82	**.01**
	n	Median (IQR)	n	Median (IQR)	n	Median (IQR)		Z	P
DASS21 (0-21)									
Depression	82	1.00 (4.00)	28	1.50 (3.00)	54	1.00 (4.00)		−1.3	.20
Anxiety	82	1.00 (3.00)	28	1.00 (3.75)	54	1.00 (2.00)		−1.1	.28
Stress	82	4.33 (4.00)	28	5.00 (4.00)	54	3.50 (4.25)		−0.9	.37

DASS21, Depression Anxiety Stress Scales, 21 items.
Medians and corresponding IQRs reported for non-normally distributed data. The IQR represents the difference between quartile 3 and quartile 1. Three mothers did not complete maternal HRQOL measures.
P values significant at the <.025 level are highlighted in bold typeface. A significance level of *P* < .025 was used to account for the multiple tests carried out.
*Compares the single ventricle and biventricular groups.

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Table VII. Bivariate analysis of demographic, clinical, psychological, and social factors associated with child HRQOL, assessed using parent-proxy reported total PedsQL scores (N = 87)

Dichotomous variables	n	Mean (SD)	t	df	P
Surgical repair pathway					
Single ventricle	30	67.52 (18.63)	3.99	41	**<.001***
Biventricular	57	82.37 (11.46)			
Diagnosis of hypoplastic left heart					
Yes	9	64.90 (20.87)	2.53	85	**.01***
No	78	78.67 (14.75)			
Time of diagnosis					
Antenatal	64	78.32 (15.66)	1.05	85	.30
Postnatal	23	74.27 (16.58)			
Presence of a comorbidity					
Yes	19	66.30 (16.17)	3.63	85	**<.001***
No	68	80.31 (14.53)			
Extracorporeal membrane oxygenation					
Yes	9	72.56 (14.40)	0.98	83	.33
No	76	78.06 (16.12)			
Child sex					
Male	62	76.81 (15.86)	−0.40	85	.69
Female	25	78.33 (16.32)			
Maternal country of birth					
Australia	66	77.81 (15.60)	−0.72	84	.47
Other	20	74.86 (17.31)			
Language primarily spoken at home					
English	76	78.16 (15.37)	−1.67	84	.10
Other	10	69.26 (18.94)			
Maternal education level					
University degree	42	79.84 (14.46)	−1.48	85	.14
No university degree	45	74.83 (16.96)			
Gross weekly household income					
Below national mean	52	74.59 (15.78)	−1.68	77	.10
Above national mean	27	80.77 (14.89)			
Child temperament					
Within normal range	63	79.66 (14.27)	2.76	83	**.007***
'Difficult'	22	69.17 (18.19)			

Continuous variables	n		r	P
Child age	87	—	0.04[†]	.69
Maternal age	86	—	0.17	.12
Perceived seriousness of cardiac condition	87	—	−0.14[†]	.19
Total time on cardiopulmonary bypass	84	—	−0.20	.07
Total days spent in NICU	85	—	−0.04[†]	.70
Total days spent in PICU	85	—	−0.25[†]	**.02***
Total length of hospital admission, days	85	—	−0.49[†]	**<.001***
Frequency of child feeding problems	85	—	−0.45	**<.001***
Maternal health literacy	82	—	0.01[†]	.94
Family functioning	84	—	0.45	**<.001***
Maternal HRQOL	84	—	0.46	**<.001***
Maternal depression	82	—	−0.32[†]	**.004***
Maternal anxiety	82	—	−0.32[†]	**.004***
Maternal stress	82	—	−0.44[†]	**.001***

NICU, Neonatal intensive care unit; *PICU*, pediatric intensive care unit.
*Significant P values (<.025) in bold.
†Spearman rank coefficient (r_s) reported for non-normally distributed data.

Table VIII. Bivariate analysis of demographic, clinical, psychological, and social factors associated with maternal HRQOL, assessed using total PedsQL scores (n = 84)

Dichotomous variables	n	Mean (SD)	t	df	P
Surgical repair pathway					
Single ventricle	28	64.87 (16.83)	2.39	82	**.02***
Biventricular	56	75.38 (19.97)			
Diagnosis of hypoplastic left heart					
Yes	9	65.69 (21.69)	1.01	82	.32
No	75	72.62 (19.28)			
Time of diagnosis					
Antenatal	62	74.64 (17.38)	2.23	82	.03
Postnatal	22	64.09 (23.33)			
Presence of a comorbidity					
Yes	18	58.96 (18.09)	3.36	82	**.001***
No	66	75.40 (18.51)			
Child sex					
Male	61	72.42 (18.52)	0.41	82	.68
Female	23	70.43 (22.37)			
Maternal country of birth					
Australia	64	72.68 (20.81)	−0.91	81	.37
Other	19	68.03 (14.06)			
Language primarily spoken at home					
English	73	72.45 (19.82)	−1.06	81	.29
Other	10	65.50 (16.53)			
Maternal education level					
University degree	40	75.63 (17.84)	−1.70	82	.09
No university degree	44	68.47 (20.55)			
Marital status					
Married	75	72.18 (19.50)	−0.42	82	.68
Not married	9	69.31 (20.76)			
Gross weekly household income					
Below national mean	49	67.24 (20.85)	−2.69	74	**.009***
Above national mean	27	79.63 (15.64)			
Child temperament					
Within normal range	62	74.76 (18.75)	2.33	82	**.02***
'Difficult'	22	63.75 (19.82)			

Continuous variables	n		r	P
Child age	84	—	0.05[†]	.64
Maternal age	83	—	0.08	.48
Perceived seriousness of cardiac condition	83	—	−0.06[†]	.60
Total time on cardiopulmonary bypass	81	—	−0.17	.12
Total days spent in NICU	82	—	−0.21[†]	.06
Total days spent in PICU	82	—	−0.23[†]	.04
Total length of hospital admission, days	82	—	−0.42[†]	**<.001***
No. of children	84	—	0.13	.24
Perceived financial strain	84	—	−0.46	**<.001***
Frequency of child feeding problems	84	—	−0.40	**<.001***
Maternal health literacy	82	—	0.07	.51
Family functioning	84	—	0.80	**<.001***
Child HRQOL	84	—	0.46	**<.001***
Maternal depression	82	—	−0.55[†]	**<.001***
Maternal anxiety	82	—	−0.45[†]	**<.001***
Maternal stress	82	—	−0.76[†]	**<.001***

*Significant P values (<.025) in bold.
†Spearman rank coefficient (r_s) reported for non-normally distributed data.