Title: Psychosocial risk factors for health-related quality of life in adult congenital heart disease.

ABSTRACT

Background: There is variability in the impact of adult congenital heart disease (ACHD) on health-related quality of life (HRQoL). A greater insight into the impact of ACHD may be gained from investigating HRQoL in various diagnostic groups and considering the importance of psychosocial risk factors for poor HRQoL.

Objective: We compared the HRQoL of people with ACHD with normative data from the general population and among four diagnostic groups and identified risk factors for poor HRQoL in ACHD from a comprehensive set of sociodemographic, clinical, and psychosocial factors.

Methods: We conducted a cross-sectional study with 303 participants from four diagnostic groups ("Simple", Tetralogy of Fallot, Transposition of the Great Arteries, Single Ventricle) who completed measures of illness perceptions, coping, social support, mood, and generic and disease-specific HRQoL. Data were analysed using one-sample t-tests, ANOVA and hierarchical multiple regressions.

Results: There was diminished psychosocial HRQoL in the "Simple" group compared with general population. Consistently significant risk factors for poor HRQoL included younger age, a perception of more severe symptoms due to ACHD, depression, and anxiety. Clinical factors were poor predictors of HRQoL.

Conclusions: The findings highlight the need to develop intervention studies aiming to improve HRQoL in people with ACHD and the routine assessment of illness perceptions and mood problems during key periods in people's lives. This will help address patient misconceptions that could be tackled by clinicians or specialist nurses during routine outpatient appointments and identify people in need of psychological support.

Keywords: Heart Defects, Congenital; Quality of Life; Illness Perceptions; Depression; Anxiety

INTRODUCTION

Advances in the medical and surgical management of congenital heart disease have led to an increasing number (~90%) of people surviving well into adulthood [1]. Research in ACHD has moved beyond survival and mortality towards patient-reported outcomes. ACHD can pose several challenges with respect to emotional difficulties, cognitive impairment, and compromised physical and social functioning [2], which can influence health-related quality of life (HRQoL). The impact of ACHD on HRQoL has been the focus of research throughout the past two decades. A systematic review of 31 studies revealed consistent evidence (>70% of studies reviewed) that people with ACHD experience poorer HRQoL in some physical domains compared with healthy population norms or matched controls, including poorer physical functioning and poorer perception of general health. However, their psychosocial and occupational/environmental HRQoL appeared to be comparable to the healthy population [3]. This and more recent reviews further highlighted various methodological limitations of past studies, including small sample sizes, and the lack of a robust categorisation of ACHD diagnoses [3,4].

It is important to understand the factors that may help explain why some people with ACHD experience poorer HRQoL than others. Some clinical factors have been implicated with people's HRQoL, including disease complexity (e.g. cyanosis and arrhythmias) [3,5,6]. However, past methodological limitations, limits the ability to draw clear conclusions about HRQoL in specific diagnostic groups with differing structural changes in the heart. This is an important investigation considering that recent data suggest that the relationship between structural complexity and HRQoL is not linear and that even people with mild or simple lesions may experience poor HRQoL [5,7]. Although the main research focus has been on demographic and clinical factors, evidence from the past decade have suggested that psychosocial factors may better explain the evident variability in HRQoL in ACHD [4]. These include illness perceptions [8–13], anxiety and depression [14–18], and social support [14,19–21], which have been found to be associated with HRQoL in ACHD. Qualitative studies have highlighted that adjustment to ACHD involves complex coping responses like acceptance, normalisation, and positive reframing [22–24]. Although people with ACHD

appear to have difficulty coping with their condition [25], no research has examined the impact of coping on HRQoL, with findings limited to outcomes like anxiety and depression [21]. There is limited evidence on the relative importance of these psychosocial factors for HRQoL in ACHD, after considering sociodemographic and clinical characteristics. The identification of the factors influencing HRQoL will help develop appropriate support services for adults with congenital heart disease and has been identified as a priority in the research agenda of ACHD nursing [26]. Nurses are highly engaged in the delivery of patient-centred care for people with ACHD and are ideally placed to identify and help people at risk for poor mental health outcomes and HRQoL. Thus, research on potentially modifiable psychosocial factors that impact on HRQoL will help support high quality clinical care, more efficient referral practices, and facilitate the development and delivery of psychological interventions with ACHD nurses at the forefront.

The present study aims to: a) compare the HRQoL of people with ACHD with normative data from the general population and among four distinct diagnostic groups and b) identify risk factors for poor HRQoL in ACHD from a comprehensive set of sociodemographic, clinical, and psychosocial factors.

METHODS

Design

A cross-sectional design was used; participants completed self-report demographic and psychosocial questionnaires at a single time-point.

Participants and Procedure

Participants were recruited from the Grown-Up Congenital Heart (GUCH) outpatient clinic at the Heart Hospital in London, UK. Participants were emerging adults and adults (≥16 years) with congenital heart disease, fluent in English. Exclusion criteria included: i) chromosomal conditions (trisomy21 and 22q11 deletion), ii) severe learning difficulties/mental retardation, iii) severe mental health problems, iv) patent foramen ovale diagnosis, v) any kind of surgical intervention within 6 months, vi) poor hearing/eyesight, vii) stroke history, viii) inability to attempt an exercise test (e.g. leg amputation, wheelchair-bound). Two

consultant cardiologists categorised participants into 4 diagnostic groups according to the structural complexity of diagnosis (Table 1). Individuals with more than one diagnosis were categorised according to their most structurally complex diagnosis. Given the diversity of the ACHD population and the multiple broad classifications of ACHD proposed by various organisations around the world, no gold standard currently exists. The most widely known is the classification system of the American College of Cardiology/American Heart Association (ACC/AHA), which is based on the anatomic and physiological changes in the heart [27]. Like other similar classifications, the ACC/AHA uses labels like simple/mild, moderate, and complex/severe and A, B, C, D for physiological state. These categorisations have received criticism for their inability to capture the true impact of illness on people's experiences, specifically the clinical features of the disease that explain differing psychosocial outcomes [28]. For example, in the ACC/AHA classification, TGA and SV are both categorised as complex, despite SV being associated with increased morbidity, mortality, and long-term complications (e.g. hemodynamic issues) that may not be typical in other diagnoses including TGA [29]. In addition, people often move between categories when their clinical status changes over time [27]. In view of these limitations, the present study followed an approach that allowed for clear distinction between various diagnostic groups defined by their structural changes in the heart.

Eligible participants were mailed an invitation letter, information sheet, and an interest form which participants were asked to complete and return using the provided freepost envelope. The information sheet included explanations about the purpose of the study, the role of the participant, confidentiality issues, and withdrawal rights. The interest form asked participants to indicate whether they would like to consider participating in the study in person at their upcoming outpatient appointment. Participants had at least two weeks to decide. Non-responders were sent reminder letters after 2-3 weeks, followed by a telephone call. Interested participants were sent study appointment letters for the same day of their upcoming outpatient appointment. Participants signed a consent form and completed questionnaires while waiting for or following their routine outpatient appointment.

Measures

Age, gender, marital status, educational level, and employment status were self-report. A consultant cardiologist collected clinical information from paper and electronic records using a standard form, including disease characteristics (diagnostic group, co-morbidities, arrhythmias, cyanosis in days), intervention history (interventions – surgical and catheterisation lab procedures, hospitalisation days), and current status (medication, current O_2 saturation, exercise capacity – VO_2 max, NYHA status, left/right ventricular ejection fraction – LVEF/RVEF).

Health-related quality of life was measured using generic and disease-specific questionnaires. The generic SF-36 questionnaire [30] is a 36-item measure which has eight subscales, subsequently combined to form the physical and mental component summaries (PCS, MCS). The questionnaire was scored using the official scoring software [31], whereby the eight subscales are linearly transformed into T-scores (i.e. Norm-Based Scoring – NBS), with a mean of 50 and a standard deviation of 10. The software utilises the 1998 US population norms to calculate a NBS score for each subscale and the aggregated component summaries that can range between 0 and 100, with higher scores representing better HRQoL. The SF-36 has good content, concurrent, criterion, and construct validity and good internal consistency ($\alpha \geq .78$ for subscales) [32].

The CHD-TAAQOL is a disease-specific HRQoL measure for people with ACHD [33], with 26 items comprising three subscales: symptoms, worries, and impact cardiac surveillance. Each item consists of two questions, the first about a health status problem and the second about the emotional impact of said problem. A weighted score is produced for each item. The subscales are scored are then linearly transformed, ranging from 0 to 100, with higher scores indicating better HRQoL. The scoring of the subscales wad based on the official algorithms provided by Kamphuis and colleagues [33]. The CHD-TAAQOL has good reliability (symptoms $\alpha = .77$, impact cardiac surveillance $\alpha = .78$, worries $\alpha = .82$) and construct, convergent, and discriminant validity [33].

Illness perceptions were measured with the Brief Illness Perceptions Questionnaire (Brief IPQ) [34], a 9-item scale that assesses perceptions about consequences, timeline, personal control, treatment control, identity, concern, coherence, emotional representation, and causes. Because of the congenital nature of ACHD, the causal dimension was not included in the present study. Each subscale is measured by a single item, which is rated using a continuous linear scale ranging from 0 to 10. Higher scores represent stronger perceptions in the subscale measured. The Brief IPQ has acceptable test-retest reliability and good predictive and discriminant validity [34].

Coping strategies were assessed using the Brief COPE [35], which consists of 28 items measuring 14 distinct coping strategies from adaptive to problematic. Mean scores for each subscale are obtained from the two items and they can range from 0 to 3, where higher scores indicate more use of the specific coping strategy. Considering that reliability is influenced by the number of items in a scale, the Brief COPE has minimally acceptable reliability with the values in each scale exceeding .50 and good construct validity [35].

Anxiety was assessed using the Six-item Short-Form of Spielberger State-Trait Anxiety Inventory (STAI-6) [36], which measures how a person feels at a given moment with six items (three indicating the absence of anxiety and three indicating the presence of anxiety). The three items indicating the absence of anxiety are reverse-scored. The mean score of the scale is obtained from the six items and can range between 1 and 4, where higher scores reflect higher anxiety levels. The STAI-6 has good reliability (α= .82) and concurrent validity [36].

The abbreviated version of the Center for Epidemiologic Studies Depression Scale (CES-D 10) [37] was used to measure depressive symptomatology. It consists of 10 items, two of them worded in the positive direction. The two positively worded items are reverse-scored. The summed score of the scale is obtained from the 10 items and can range between 0 and 30. The cut-off score of 10 suggested by the authors was used in the present study to indicate clinical levels of depression; a score of 0 to 10 was classified

as “without depressive symptoms” and a score of more than 10 was classified as “with depressive symptoms”. This cut-off point has high sensitivity (.85) and specificity (.80), with a reduced misclassification rate at 17.5% compared with 23.5% when a cut-off point of 8 is used [38]. The CES-D 10 has acceptable convergent validity, internal consistency (α=.78 to .79), and predictive accuracy when compared to the original 20-item measure [38].

Social support was assessed using the Multidimensional Scale of Perceived Social Support (MSPSS) [39], which is a measure of subjective assessment of social support adequacy from family, friends, and significant other using 12 items (4 items in each subscale). The mean score is calculated for each subscale and can range between 1 and 5. Higher scores indicate higher levels perceived social support. The MSPSS has construct validity and good internal consistency both at scale and subscale level (α≥ .85) across different populations [40].

The internal reliability of the measures for the current sample are provided in Supplementary File 1.

Statistical Analysis

Using G-Power [41] it was estimated that 280 participants (approximately 70 in each diagnostic group) would be needed to achieve 80% power with a medium effect size (f= .20) for generic and disease-specific HRQoL [42,43]. Missing value analysis was performed at scale level except for demographic and clinical variables (Supplementary File 2). Little’s Missing Completely At Random (MCAR) test result was p= .219, suggesting that imputation is appropriate. Missing data was imputed using Bayesian stochastic regression (chained equations, Markov Chain Monte Carlo-MCMC), except for eleven participants who had incomplete psychosocial questionnaires. One-way ANOVAs and chi-square (χ^2) tests were used to examine differences in demographic and clinical characteristics between the four diagnostic groups. One-sample t-tests were used to examine differences in scores for the PCS and MCS of the SF-36 between the total sample and norms and each of the four diagnostic groups and norms (reference mean value 50±10). A cut-off point of ≥ 0.5 standard deviation was selected to assess the percentage of participants

scoring below the norm in the PCS and MCS of the SF-36, indicating minimally important difference (MID) [42,43]. To investigate differences in HRQoL between the four diagnostic groups, one-way ANOVA with Gabriel post-hoc tests (for equal variances) or Games-Howell (for unequal variances) tests were used, based on 99% CI. The Welch test was implemented for unequal variances. To examine the factors associated with HRQoL hierarchical multiple regressions were performed. Outcome variables included generic (PCS, MCS of the SF-36) and disease-specific (three scales of the CHD-TAAQOL) HRQoL. Only variables that were found to be significantly associated ($p< 0.01$) with HRQoL outcomes in bivariate linear regressions (Supplementary File 4) were entered in the hierarchical multiple regressions. The hierarchy used to enter the variables into the regression is illustrated in Figure 1. Evaluations were conducted for singularity, multicollinearity, independence of errors, outliers, normality, linearity, and homoscedasticity that confirmed that the assumptions for regression analyses were met. Statistical analyses were performed using IBM SPSS 25 and significance was set at $p< 0.01$ and 99% Confidence Intervals to minimise family-wise error due to multiple testing. The magnitude of the relationship between variables was calculated using eta squared (η^2) [.01= small, .06= medium, .14= large] and Cohen's d (.20= small, .50= medium, .80= large) where appropriate.

Ethical Statement

Full ethical approval was granted from the Joint UCL/UCLH Ethics Committee and NRES Committee London – Bentham in Ethics of Human Research (REC reference number: 08/H0715/105). Relevant approvals were also gained from the Research & Development (R&D) department at UCLH. The investigation conforms with the principles outlined in the Declaration of Helsinki [44].

RESULTS

Sample characteristics

Among the 708 patients identified as eligible, 314 (44.4%) participated in the study. Of these, 11 had ≥50% incomplete data and were excluded from further analyses (Supplementary File 2). The final sample consisted of 303 participants. The sociodemographic details are presented in Table 2. There were age

differences between diagnostic groups (p< .001). Post-hoc comparisons indicated that the Simple group was older than the TGA (mean difference= 5.49, p= 0.007, 99% CI [0.22, 10.77]) and SV (mean difference= 8.62, p< 0.001, 99% CI [3.05, 14.18]). The ToF group was older than the SV (mean difference= 6.18, p= 0.001, 99% CI [1.32, 11.03]). The differences between groups in clinical characteristics are available in Supplementary File 3. A total of 21.5% of the sample experienced depressive symptoms, with the largest percentage observed in the Simple (28%) and SV groups (27.3%).

HRQoL comparisons with normative data

The comparisons between the ACHD sample and normative data for the overall physical (PCS) and psychosocial (MCS) HRQoL are presented in Table 3. The ToF group scored higher than the norm on the PCS. The total sample and Simple group scored lower than the norm on the MCS. The percentages of participants (total and by group) reaching the MID of 0.5 standard deviation below the norm in the PCS and MCS of the SF-36, are presented in Figure 2.

HRQoL comparisons between diagnostic groups

The comparisons between the four diagnostic groups for generic and disease-specific HRQoL are presented in Table 4. Post-hoc comparisons indicated that the SV group reported greater impact of cardiac surveillance and therefore poorer HRQoL than the Simple group (mean difference= -5.99, p= 0.003, 99% CI [-11.38, -0.60]; d= 0.55).

Factors associated with HRQoL

Generic HRQoL

The final models of the hierarchical regression for PCS and MCS are presented in Table 5 and the full hierarchical multiple regressions that include all the steps are displayed in Supplementary File 5. The final model for PCS was significant, f(22,278)= 13.48; p< 0.001 and explained 47.8% of the variance. Illness identity was the only unique predictor in this final model indicating that a perception of more severe symptoms due to ACHD was associated with poorer physical HRQoL. The final model for MCS was

significant, f(20,281)= 21.54; p< 0.001 and explained 57.7% of the variance. The unique predictors in the final model were age, self-blame, anxiety, and presence of depressive symptoms. Younger age, greater use of self-blame as a coping strategy, greater anxiety, and the presence of depressive symptoms were associated with poorer psychosocial HRQoL.

Disease-specific HRQoL

The final models of the hierarchical regression for the symptoms, impact of cardiac surveillance, and worries subscales are presented in Table 6 and the full hierarchical multiple regressions that include all the steps are displayed in Supplementary File 5. The final model for the symptoms subscale was significant, f(26,275)= 18.29; p< 0.001 and explained 59.9% of the total variance in symptoms. The unique predictors in the final model were illness identity and presence of depressive symptoms. A perception of more severe symptoms due to ACHD and the presence of depressive symptoms were associated with greater symptom impact. The final model for the impact of cardiac surveillance subscale was significant, f(19,283)= 8.57; p< 0.001 and explained 32.3% of the total variance in impact of cardiac surveillance. The unique predictors in the final model were planning and anxiety. Greater use of planning as a coping strategy and greater anxiety were associated with greater impact of cardiac surveillance. The final model for the worries subscale was significant, f(22,280)= 13.97; p< 0.001 and explained 48.6% of the total variance in worries. The unique predictors in this final step were age, illness consequences, and presence of depressive symptoms. Younger age, a perception of more consequences due to ACHD, and the presence of depressive symptoms were associated with greater worries.

DISCUSSION

The present study sought to examine differences in HRQoL between adults with congenital heart disease and normative data and among four distinct diagnostic groups and identify risk factors for poor HRQoL. Overall, the physical HRQoL for the total sample was comparable to that of the general population. However, poorer psychosocial HRQoL was observed in the Simple group. The SV group reporting greater impact of cardiac surveillance and therefore poorer HRQoL compared with the Simple group. Out of all

the demographic and clinical characteristics, only age was found to be a predictor of HRQoL in multivariate analyses, after considering psychosocial factors. Consistent significant risk factors for poor HRQoL included younger age, a perception of more severe symptoms due to ACHD, depression, and anxiety.

While greater physical and psychosocial morbidity may be expected in people with SV due to the complexity of their condition [24], people with simpler structural changes in the heart (Simple group), who require less frequent outpatient appointments and are relatively free from any disease burden, may be expected to have better HRQoL. Although unexpected, we found decreased psychosocial HRQoL in the Simple group and, unlike the other diagnostic groups, over half of people with structurally simpler diagnoses had poor psychosocial HRQoL of clinical significance. This has also been reported in previous studies that focused exclusively on people with ASD [45], CoA [46], and the surgically cured (ASD, VSD, PS, AS) [47], all of which were included in the Simple group in the present study. One possible explanation for this finding is the composition of our Simple group which included 46% people with CoA, a diagnosis which is categorised as of moderate complexity by the AHA [27]. However, over half of the other diagnoses in this group are categorised as simple by the AHA and the structural complexity of the Simple group was still lower relative to the other three groups in the study. Furthermore, a recent study with ACHD patients from 15 countries suggested that CoA may be used as a healthy comparison group along with ASD and VSD [5], supporting the categorisation in the present study. It is important to note that the Simple group in our study consisted of people that still required health monitoring at a specialist clinic as opposed to those who are physically well, free from long-term complications, and more likely to have been discharged and followed-up by their local general practitioners.

While the differences observed may be attributed to the sample composition, what they also suggest is that the relationship between structural complexity and HRQoL is not a simple linear relationship whereby decreasing structural complexity is associated with better HRQoL. In fact, there are recent evidence to suggest that even mild functional impairments may impact HRQoL via increased perceptions of stress [17].

In the present study the patients' subjective illness perceptions and emotional responses were more important for HRQoL than objective clinical characteristics corroborating the findings of previous studies [8–17,19]. These findings are also in agreement with the wider chronic illness literature about the impact of negative illness perceptions and mood problems on HRQoL in heart failure, coronary heart disease, and myocardial infarction [48–50]. Depressive symptomatology may pose an additional burden on people with ACHD, making them more vulnerable to experiencing poorer psychosocial HRQoL and limitations, while also encouraging a more negative perception in these outcomes [48]. Thus, another possible explanation for the poor psychological HRQoL in the Simple group may be related to their perceptions and mood driven by illness and treatment experiences shaping their expectations that they will be closer to the general population in HRQoL, and this may be their reference point when completing a HRQoL questionnaire. In our study, the Simple group had the highest percentage of people with depressive symptoms. Although the study did not set out to examine treatment perceptions in detail, it is possible that people with simpler diagnoses perceive that they have been "cured" early in their lives, thereby entering adulthood with unrealistic expectations and misconceptions about their functioning and their medical needs [51]; a perception reinforced further by the long periods of stability in their functioning. The use of surgical terms such as "total correction" by medical teams may further enhance these perceptions of cure [52]. The realisation later in life that their health will likely deteriorate more rapidly than anticipated and requiring further medical or surgical intervention may come as a shock, resulting in a more intense emotional response [53]. This is supported by evidence suggesting that people with less complex conditions can have unwarranted negative illness perceptions about their condition and its emotional impact which negatively influence their HRQoL [8]. An interesting avenue of future research is the in-depth exploration of treatment perceptions in relation to HRQoL in ACHD, particularly across various diagnostic groups to examine whether people form differing expectations especially following their treatment.

The findings of the present study further indicated that younger people have poorer psychosocial HRQoL and greater ACHD-specific worries (e.g. employment, family planning) compared with older individuals.

This finding supports previous studies in ACHD that have found positive relationship between age and psychosocial HRQoL [8,54]. This is unsurprising since young adulthood is a transitional life stage whereby establishing independence and social relationships, pursuing employment, and starting a family are pertinent issues as opposed to the relative life stability that characterises older adulthood [55,56]. The better psychosocial HRQoL with increasing age may be attributed to well-developed coping strategies and maturation processes as well as increased sense of coherence [57]. Research in healthy populations has indicated that HRQoL can increase with age due to adjustment in goals and personal growth [58]. The findings suggest that HRQoL in ACHD needs to be viewed within people's life stage as each is associated with different developmental tasks which can pose additional challenges to illness-related issues with which people need to cope. In relation to age and psychosocial HRQoL, it is important that the Simple group, despite being older, still demonstrated the poorest psychosocial QoL.

Limitations

This study found several risk factors for poor HRQoL in ACHD. There are inherent limitations in causality and directionality inferences based on cross-sectional data and replication of these relationships is warranted in longitudinal studies. Certain diagnoses in the Simple group might have been under-represented, as it consisted of a large percentage of CoA. This could limit generalisability in people attending their local GPs rather than specialist outpatient clinics. However, the commonest diagnoses were well represented across the diagnostic groups. A unique challenge in ACHD clinics is the number of people lost to follow-up during the transition period from paediatric to adult clinics [59], who may be in better health and have better HRQoL than those who are still followed-up at specialist clinics. Inadvertently, this might have resulted in selection bias. The present study distinguished between various diagnostic groups defined by their structural changes in the heart. The unexpected finding with regards to the impaired psychosocial HRQoL in the Simple group may reflect this categorisation. The nature of the categorisation of the diversity of ACHD is obviously important. One alternative classification system considers CoA as 'moderate' in a 3-way classification system of mild, moderate and severe [27] and using

this system may have produced different results to the present study. It is possible that the categorisation by structural complexity could be enhanced by including additional variables such as factors relating to illness and treatment experience. When studying patient-reported outcomes, a more comprehensive evaluation of the treatment history (e.g. nature of treatment, medication) along with an assessment of treatment perceptions and expectations should be considered to supplement the use of any classification based on structural complexity.

Despite these limitations, this study has addressed the limitation of previous approaches in studying ACHD as a homogeneous group by categorising participants in diagnostic groups, clearly defined in their inclusion and exclusion criteria, allowing for HRQoL comparisons based on the structural complexity of the underlying defect. Furthermore, it enabled a comprehensive evaluation of the relative contribution of psychosocial factors to HRQoL in ACHD.

Implications for nursing practice

As nursing research in ACHD has moved beyond endpoints like survival, we need to consider psychosocial factors alongside structural complexity, including the patients' perceptions and mood when investigating and addressing HRQoL. This is important considering the evidence suggesting that depression is associated with increased risk of mortality in people with ACHD [60] and that people with ACHD hold misconceptions, unrealistic expectations [51,61], and unjustifiable negative illness perceptions about their condition and its emotional impact irrespective of its complexity [8]. Specialist nurses are ideally placed to address the psychosocial needs of people with ACHD during follow-up appointments. While regular assessment of illness perceptions and screening for anxiety and depression using self-report tools may not be feasible in everyday clinical practice, it is recommended during key developmental periods. ACHD nurses could routinely enquire about specific developmental needs during outpatient appointments; finding employment, leaving the parental home, marrying, and becoming parents in young adulthood (18-30 years)[56] and maintaining employment, revising career goals, facing the prospect of

premature death and its impact on the family as people get older [55]. This will facilitate clear and personalised information and the use brief measures to assess mental health could be an effective way of identifying individuals at risk in a timely manner and referring them for professional psychological support.

There is also an urgent need for psychological interventions in ACHD targeting illness perceptions and mood involving nurses. Current programs that are underway include a self-management intervention which showed promising results for disease-related knowledge and self-management performance but not HRQoL [62] and a cardiac rehabilitation program (QUALI-REHAB) aiming to improve HRQoL in adolescents and young adults [63], and a pilot study (ACHD-CARE) is targeting psychosocial functioning, HRQoL, and resilience in people with ACHD through education, cognitive behavioural therapy, coping strategies, and peer interaction [64]. These results are highly anticipated, as they will provide important insights on the effectiveness of psychological interventions in ACHD and inform a comprehensive healthcare that includes psychosocial aspects [65].

CONCLUSION

The present study highlighted the importance of distinguishing between diagnostic groups when studying HRQoL in ACHD as the expected relationship between structural complexity and HRQoL is complex. With the inclusion of a wide range of factors, the study demonstrated the relative importance of negative illness perceptions and mood in accounting for variations in HRQoL in ACHD. The findings highlight the need to develop psychological interventions to improve HRQoL in people with ACHD and for routine assessment of illness perceptions and mood during key periods in people's lives. This will draw attention to individuals with ACHD misconceptions and unrealistic expectations that could be tackled by clinicians or specialist nurses during routine appointments and identify people in need of further psychological support.

Data availability statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

REFERENCES

1. Buber J, Valente AM. Predicting survival in adults with congenital heart disease: what are the odds? *Heart*. 2018;104(20):1643-1644. doi:10.1136/heartjnl-2018-312975
2. Tyagi M, Fteropoulli T, Hurt CS, et al. Cognitive dysfunction in adult CHD with different structural complexity. *Cardiology in the Young*. 2017;27(5):851-859. doi:10.1017/S1047951116001396
3. Fteropoulli T, Stygall J, Cullen S, Deanfield J, Newman SP. Quality of life of adult congenital heart disease patients: a systematic review of the literature. *Cardiology in the Young*. 2013;23(4):473-485. doi:10.1017/S1047951112002351
4. Moons P, Luyckx K. Quality-of-life research in adult patients with congenital heart disease: current status and the way forward. *Acta Paediatrica*. 2019;108(10):1765-1772. doi:10.1111/APA.14876
5. Moons P, Luyckx K, Thomet C, et al. Physical Functioning, Mental Health, and Quality of Life in Different Congenital Heart Defects: Comparative Analysis in 3538 Patients From 15 Countries. *Canadian Journal of Cardiology*. 2021;37(2):215-223. doi:10.1016/J.CJCA.2020.03.044
6. Casteigt B, Samuel M, Laplante L, et al. Atrial arrhythmias and patient-reported outcomes in adults with congenital heart disease: An international study. *Heart Rhythm*. 2021;18(5):793-800. doi:10.1016/J.HRTHM.2020.09.012
7. Andonian CS, Freilinger S, Achenbach S, et al. 'Well-being paradox' revisited: a cross-sectional study of quality of life in over 4000 adults with congenital heart disease. *BMJ Open*. 2021;11(6):e049531. doi:10.1136/BMJOPEN-2021-049531
8. Schoormans D, Mulder BJ, van Melle JP, et al. Illness perceptions of adults with congenital heart disease and their predictive value for quality of life two years later. *European Journal of Cardiovascular Nursing*. 2014;13(1):86-94. doi:10.1177/1474515113481908
9. Rassart J, Apers S, Kovacs AH, et al. Illness perceptions in adult congenital heart disease: A multi-center international study. *International Journal of Cardiology*. 2017;244:130-138. doi:10.1016/j.ijcard.2017.06.072
10. Riley JP, Habibi H, Banya W, Gatzoulis MA, Lau-Walker M, Cowie MR. Education and support needs of the older adult with congenital heart disease. *Journal of Advanced Nursing*. 2012;68(5):1050-1060. doi:10.1111/j.1365-2648.2011.05809.x

11. Chow PC. Quality of life, psychological resilience, personality traits and illness perception in grown-up congenital heart patients in Hong Kong. *International Journal of Cardiology Congenital Heart Disease*. 2021;6:100279. doi:10.1016/J.IJCCHD.2021.100279
12. O'Donovan CE, Painter L, Lowe B, Robinson H, Broadbent E. The impact of illness perceptions and disease severity on quality of life in congenital heart disease. *Cardiology in the young*. 2016;26(1):100-109. doi:10.1017/S1047951114002728
13. Holbein CE, Fogleman ND, Hommel K, et al. A multinational observational investigation of illness perceptions and quality of life among patients with a Fontan circulation. *Congenital Heart Disease*. 2018;13(3):392-400. doi:10.1111/CHD.12583
14. Pike NA, Evangelista LS, Doering LV, Eastwood JA, Lewis AB, Child JS. Quality of Life, Health Status, and Depression. *The Journal of Cardiovascular Nursing*. 2012;27(6):539-546. doi:10.1097/JCN.0b013e31822ce5f6
15. Gleason LP, Deng LX, Khan AM, et al. Psychological distress in adults with congenital heart disease: focus beyond depression. *Cardiology in the Young*. 2019;29(2):185-189. doi:10.1017/S1047951118002068
16. Ko JM, Tecson KM, Rashida VA, et al. Clinical and Psychological Drivers of Perceived Health Status in Adults With Congenital Heart Disease. *The American Journal of Cardiology*. 2018;121(3):377-381. doi:10.1016/J.AMJCARD.2017.10.038
17. Jackson JL, Gerardo GM, Daniels CJ, Vannatta K. Perceptions of Disease-Related Stress: A Key to Better Understanding Patient-Reported Outcomes among Survivors of Congenital Heart Disease. *The Journal of Cardiovascular Nursing*. 2017;32(6):587-593. doi:10.1097/JCN.0000000000000371
18. Westhoff-Bleck M, Briest J, Fraccarollo D, et al. Mental disorders in adults with congenital heart disease: Unmet needs and impact on quality of life. *Journal of Affective Disorders*. 2016;204:180-186. doi:10.1016/J.JAD.2016.06.047
19. Chen CA, Liao SC, Wang JK, et al. Quality of life in adults with congenital heart disease: biopsychosocial determinants and sex-related differences. *Heart*. 2011;97(1):38-43. doi:10.1136/hrt.2010.200709
20. Rometsch S, Greutmann M, Latal B, et al. Predictors of quality of life in young adults with congenital heart disease. *European Heart Journal - Quality of Care and Clinical Outcomes*. 2019;5(2):161-168. doi:10.1093/EHJQCCO/QCY046

21. Kim MY, Johnson JL, Sawatzky R. Relationship between Types of Social Support, Coping Strategies, and Psychological Distress in Individuals Living with Congenital Heart Disease. *The Journal of Cardiovascular Nursing*. 2019;34(1):76-84. doi:10.1097/JCN.0000000000000531

22. Claessens P, Moons P, de Casterlé BD, Cannaerts N, Budts W, Gewillig M. What Does it Mean to Live with a Congenital Heart Disease? A Qualitative Study on the Lived Experiences of Adult Patients. *European Journal of Cardiovascular Nursing*. 2005;4(1):3-10. doi:10.1016/j.ejcnurse.2004.12.003

23. Callus E, Quadri E, Compare A, Tovo A, Giamberti A, Chessa M. Life Experiences and Coping Strategies in Adults with Congenital Heart Disease. *La Pediatria Medica e Chirurgica*. 2013;35(5):231-240. doi:10.4081/pmc.2013.34

24. Overgaard D, King C, Christensen RF, Schrader AM, Adamsen L. Living With Half a Heart—Experiences of Young Adults With Single Ventricle Physiology. *The Journal of Cardiovascular Nursing*. 2013;28(2):187-196. doi:10.1097/JCN.0b013e3182498677

25. Ferguson M, Kovacs AH. An Integrated Adult Congenital Heart Disease Psychology Service. *Congenital heart disease*. 2016;11(5):444-451. doi:10.1111/CHD.12331

26. Goossens E, Fleck D, Canobbio MM, Harrison JL, Moons P. Development of an international research agenda for adult congenital heart disease nursing. *European Journal of Cardiovascular Nursing*. 2013;12(1):7-16. doi:10.1016/J.EJCNURSE.2011.06.009

27. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Journal of the American College of Cardiology*. 2019;73(12):1494-1563. doi:10.1016/J.JACC.2018.08.1028

28. Jackson JL, Leslie CE, Hondorp SN. Depressive and Anxiety Symptoms in Adult Congenital Heart Disease: Prevalence, Health Impact and Treatment. *Progress in Cardiovascular Diseases*. 2018;61(3-4):294-299. doi:10.1016/J.PCAD.2018.07.015

29. Jackson J, Misiti B, Bridge J, Daniels C, Vannatta K. Emotional functioning of adolescents and adults with congenital heart disease: a meta-analysis. *Congenital heart disease*. 2015;10(1):2-12. doi:10.1111/CHD.12178

30. Ware JE, Sherbourne CD. The MOS 36-Item Short-Form Health Survey (SF-36). *Medical Care*. 1992;30(6):473-483. doi:10.1097/00005650-199206000-00002
31. Saris-Baglama RN, Dewey CJ, Chisholm GB. *QualityMetric health outcomes™ scoring software 4.0*. 2010.
32. McHorney CA, Ware JE, Lu JF, Sherbourne CD. The MOS 36-Item Short-Form Health Survey (SF-36): III. Tests of Data Quality, Scaling Assumptions, and Reliability Across Diverse Patient Groups. *Medical Care*. 1994;32(1):40-66. doi:10.1097/00005650-199401000-00004
33. Kamphuis M, Zwinderman KH, Vogels T, et al. A cardiac-specific health-related quality of life module for young adults with congenital heart disease: Development and validation. *Quality of Life Research*. 2004;13(4):735-745. doi:10.1023/B:QURE.0000021690.84029.a3
34. Broadbent E, Petrie KJ, Main J, Weinman J. The Brief Illness Perception Questionnaire. *Journal of Psychosomatic Research*. 2006;60(6):631-637. doi:10.1016/j.jpsychores.2005.10.020
35. Carver CS. You want to measure coping but your protocol' too long: Consider the brief cope. *International Journal of Behavioral Medicine*. 1997;4(1):92-100. doi:10.1207/s15327558ijbm0401_6
36. Marteau TM, Bekker H. The development of a six-item short-form of the state scale of the Spielberger State—Trait Anxiety Inventory (STAI). *British Journal of Clinical Psychology*. 1992;31(3):301-306. doi:10.1111/j.2044-8260.1992.tb00997.x
37. Andresen EM, Malmgren JA, Carter WB, Patrick DL. Screening for Depression in Well Older Adults: Evaluation of a Short Form of the CES-D. *American Journal of Preventive Medicine*. 1994;10(2):77-84. doi:10.1016/S0749-3797(18)30622-6
38. Boey KW. Cross-validation of a short form of the CES-D in Chinese elderly. *International Journal of Geriatric Psychiatry*. 1999;14(8):608-617. doi:10.1002/(SICI)1099-1166(199908)14:8<608::AID-GPS991>3.0.CO;2-Z
39. Zimet GD, Dahlem NW, Zimet SG, Farley GK. The Multidimensional Scale of Perceived Social Support. *Journal of Personality Assessment*. 1988;52(1):30-41. doi:10.1207/s15327752jpa5201_2
40. Canty-Mitchell J, Zimet GD. Psychometric Properties of the Multidimensional Scale of Perceived Social Support in Urban Adolescents. *American Journal of Community Psychology*. 2000;28(3):610-617. doi:10.1023/A:1005109522457

41. Faul F, Erdfelder E, Lang AG, Buchner A. G*Power 3: A flexible statistical power analysis program for the social, behavioral, and biomedical sciences. *Behavior Research Methods*. 2007;39(2):175-191. doi:10.3758/BF03193146

42. Norman GR, Gwadry Sridhar F, Guyatt GH, Walter SD. Relation of Distribution- and Anchor-Based Approaches in Interpretation of Changes in Health-Related Quality of Life. *Medical Care*. 2001;39(10). doi:10.1097/00005650-200110000-00002

43. Norman GR, Sloan JA, Wyrwich KW. Interpretation of Changes in Health-related Quality of Life. *Medical Care*. 2003;41(5):582-592. doi:10.1097/01.MLR.0000062554.74615.4C

44. World Medical Association. World Medical Association Declaration of Helsinki: ethical principles for medical research involving human subjects. *JAMA*. 2013;310(20):2191-2194. doi:10.1001/JAMA.2013.281053

45. Ternestedt B, Wall K, Oddsson H, Riesenfeld T, Groth I, Schollin. J. Quality of life 20 and 30 years after surgery in patients operated on for tetralogy of Fallot and for atrial septal defect. *Pediatric cardiology*. 2001;22(2):128-132. doi:10.1007/S002460010178

46. Buys R, Budts W, Delecluse C, Vanhees L. Exercise capacity, physical activity, and obesity in adults with repaired aortic coarctation. *The Journal of Cardiovascular Nursing*. 2013;28(1):66-73. doi:10.1097/JCN.0B013E318239F430

47. Lane D, Lip G, Millane T. Quality of life in adults with congenital heart disease. *Heart*. 2002;88(1):71-75. doi:10.1136/HEART.88.1.71

48. Stafford L, Berk M, Reddy P, Jackson H. Comorbid depression and health-related quality of life in patients with coronary artery disease. *Journal of psychosomatic research*. 2007;62(4):401-410. doi:10.1016/J.JPSYCHORES.2006.12.009

49. Ruo B, Rumsfeld JS, Hlatky MA, Liu H, Browner WS, Whooley MA. Depressive Symptoms and Health-Related Quality of Life: The Heart and Soul Study. *JAMA : the Journal of the American Medical Association*. 2003;290(2):215-221. doi:10.1001/JAMA.290.2.215

50. Hallas CN, Wray J, Andreou P, Banner NR. Depression and perceptions about heart failure predict quality of life in patients with advanced heart failure. *Heart & Lung*. 2011;40(2):111-121. doi:10.1016/J.HRTLNG.2009.12.008

51. Harrison JL, Silversides CK, Oechslin EN, Kovacs AH. Healthcare needs of adults with congenital heart disease: Study of the patient perspective. *The Journal of Cardiovascular Nursing*. 2011;26(6):497-503. doi:10.1097/JCN.0B013E31820984C9

52. Warnes C. The adult with congenital heart disease: born to be bad? *Journal of the American College of Cardiology*. 2005;46(1):1-8. doi:10.1016/J.JACC.2005.02.083

53. Saidi AS, Paolillo J, Fricker FJ, Sears SF, Kovacs AH. Biomedical and Psychosocial Evaluation of "Cured" Adults with Congenital Heart Disease. *Congenital Heart Disease*. 2007;2(1):44-54. doi:10.1111/J.1747-0803.2007.00071.X

54. Silva A, Vaz C, Areias M, et al. Quality of life of patients with congenital heart diseases. *Cardiology in the Young*. 2011;21(6):670-676. doi:10.1017/S1047951111000576

55. Sable C, Foster E, Uzark K, et al. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. *Circulation*. 2011;123(13):1454-1485. doi:10.1161/CIR.0B013E3182107C56

56. Pinquart M. Achievement of developmental milestones in emerging and young adults with and without pediatric chronic illness--a meta-analysis. *Journal of Pediatric Psychology*. 2014;39(6):577-587. doi:10.1093/JPEPSY/JSU017

57. Moons P, Apers S, Kovacs AH, et al. Sense of coherence in adults with congenital heart disease in 15 countries: Patient characteristics, cultural dimensions and quality of life. *European Journal of Cardiovascular Nursing*. 2021;20(1):48-55. doi:10.1177/1474515120930496

58. Netuveli G, Blane D. Quality of life in older ages. *British Medical Bulletin*. 2008;85(1):113-126. doi:10.1093/BMB/LDN003

59. Wray J, Frigiola A, Bull C, Adult Congenital Heart Disease Research Network (ACoRN). Loss to specialist follow-up in congenital heart disease; out of sight, out of mind. *Heart*. 2013;99(7):485-490. doi:10.1136/HEARTJNL-2012-302831

60. Carazo MR, Kolodziej MS, Dewitt ES, et al. Prevalence and Prognostic Association of a Clinical Diagnosis of Depression in Adult Congenital Heart Disease: Results of the Boston Adult Congenital Heart Disease Biobank. *Journal of the American Heart Association*. 2020;9(9):e014820. doi: 10.1161/JAHA.119.014820

61. Wichert-Schmitt B, Oechslin E. Misperception of Survival in Adult Congenital Heart Disease and Importance of Both Anatomic and Functional Indices: Educate Your Patients! *Canadian Journal of Cardiology*. 2019;35(12):1635-1639. doi:10.1016/J.CJCA.2019.08.027

62. Lee MJ, Jung D. Development and effects of a self-management efficacy promotion program for adult patients with congenital heart disease. *European Journal of Cardiovascular Nursing*. 2019;18(2):140-148. doi:10.1177/1474515118800099

63. Amedro P, Gavotto A, Legendre A, et al. Impact of a centre and home-based cardiac rehabilitation program on the quality of life of teenagers and young adults with congenital heart disease: The QUALI-REHAB study rationale, design and methods. *International Journal of Cardiology*. 2019;283:112-118. doi:10.1016/J.IJCARD.2018.12.050

64. Kovacs AH, Grace SL, Kentner AC, Nolan RP, Silversides CK, Irvine MJ. Feasibility and Outcomes in a Pilot Randomized Controlled Trial of a Psychosocial Intervention for Adults With Congenital Heart Disease. *Canadian Journal of Cardiology*. 2018;34(6):766-773. doi:10.1016/J.CJCA.2018.02.023

65. Coleman A, Chan A, Zaidi AN. The emerging psychosocial profile of the adult congenital heart disease patient. *Current Opinion in Organ Transplantation*. 2020;25(5):506-512. doi:10.1097/MOT.0000000000000802

WHAT IS NEW

- Poor psychosocial HRQoL is evident in people with relatively simple ACHD and those of a younger age, who have negative illness perceptions, anxiety, and depression are at risk for poor HRQoL in ACHD.
- Routine assessment of misconceptions and mood by healthcare professionals during key developmental periods is needed to identify people at risk for poor HRQoL.
- There is a need for psychological interventions addressing illness perceptions and mood in ACHD.

Table 1: Description of diagnostic groups

Group	Definition
Tetralogy of Fallot (ToF)	Diagnosis of TOF, pulmonary atresia, major aorto-pulmonary collateral arteries, patients who had pulmonary valve replacement
Transposition of the Great Arteries (TGA)	Diagnosis of TGA, patients who had Mustard or Senning operations (atrial switch), including those with implantable cardioverter defibrillators and pacemakers.
Single Ventricle (SV)	Patients who had Fontan repair, total cavopulmonary connection (TCPC), and all cyanotic patients with SV physiology.
"Simple"	Structurally simpler defects, including atrial septal defect (ASD; treated surgically or percutaneously), ventricular septal defect (VSD; treated surgically), pulmonary stenosis (PS), and coarctation of the aorta (COA; treated both in childhood and adulthood and re-coarctations). COA patients with aortic valve replacement were excluded as this condition does not meet the remit of a "simple" defect.

Table 2. Demographic and clinical characteristics of the sample

Variable	*Total Sample* *n= 303*	*Simple* *n= 82*	*ToF* *n= 77*	*TGA* *n= 78*	*SV* *n= 66*	*Test Statistic*	*Sig.*	*Effect Size*
Age (*mean, S.D., range*)	33.3 (10.8)	37.2 (13.7)	34.9 (11.2)	31.8 (6.5)	28.6 (7.7)	$f(3,167.07)=10.29^{a}$	<0.001**	$\eta^2= .09$
	18-76	19-76	19-66	19-50	18-58			
Gender (*n*, %)						$\chi^2(3)=10.22$	0.017	$\varphi_c= .18$
Male	172 (56.8)	36 (43.9)	42 (54.5)	50 (64.1)	44 (66.7)			
Female	131 (43.2)	46 (56.1)	35 (45.5)	28 (35.9)	22 (33.3)			
Marital status (*n*, %)						$\chi^2(3)= 1.98$	0.576	$\varphi_c= .08$
Married/relationship	156 (51.5)	45 (54.9)	41(53.2)	36 (46.2)	34 (51.5)			
Single	147 (48.5)	37 (45.1)	36 (46.8)	42 (53.8)	32 (48.5)			
Educational level (*n*, %)						$\chi^2(3)= 3.28$	0.351	$\varphi_c= .10$
School level	202 (66.7)	51 (62.2)	51 (66.2)	60 (76.9)	40 (60.6)			
University level	101 (33.3)	31 (37.8)	26 (33.8)	18 (23.1)	26 (39.4)			
Employment status (*n*, %)						$\chi^2(3)= 2.49$	0.478	$\varphi_c= .09$
Employed	218 (71.9)	57 (69.5)	57 (74)	60 (76.9)	44 (66.7)			
Unemployed	85 (28.1)	25 (30.5)	20 (26)	18 (23.1)	22 (33.3)			
Co-morbidities no. (*mean, S.D., range*)	1 (1.1)	0.8 (1)	0.9 (1.1)	1.1 (1.2)	1.1 (1.1)	$f(3,310)= 1.40$	0.243	$\eta^2= .01$
	0-6	0-4	0-4	0-5	0-6			

Arrhythmias (*n*, %)						$\chi^2(3)= 32.98$	<0.001**	$\varphi_c= .32$
Yes	82 (27.1)	7 (8.5)	15 (19.5)	34 (43.6)	26 (39.4)			
No	221 (72.9)	75 (91.5)	62 (80.5)	44 (56.4)	40 (60.6)			
Cyanosis days (*mean, S.D., range*)	1249.6 (2468.1)	9.8 (88.3)	1544.9 (2297.3)	610.6 (1621.8)	3230.7 (3556.6)	$f(3,125.61)= 31.75^a$	<0.001**	$\eta^2= .23$
	0-17155	0-800	10-16790	4-13870	0-17155			
Interventions no. (*mean, S.D., range)*	2.4 (1.4)	1.4 (.8)	2.4 (1.2)	2.6 (1.3)	3.2 (1.6)	$f(3,160.86)= 33.17^a$	<0.001**	$\eta^2= .20$
	0-8	0-4	1-7	1-8	0-8			
Hospitalization days (*mean, S.D., range)*	40.8 (53.5)	17.2 (16.8)	46.9 (34.2)	35.5 (24.9)	69.2 (96.2)	$f(3,152.99)= 26.72^a$	<0.001**	$\eta^2= .13$
	0-800	0-100	12-200	7-140	19-800			
Medication no. (*mean, S.D., range*)	0.9 (1.3)	0.8 (1.3)	0.5 (1.1)	1 (1.3)	1.5 (1.2)	$f(3,310)= 8.05$	<0.001**	$\eta^2= .08$
	0-6	0-6	0-5	0-5	0-4			
Current O_2 saturation (*mean, S.D., range*)	95.9 (4.7)	98.2 (1.7)	97.2 (1.5)	96.2 (2.3)	91.1 (7.5)	$f(3,157.73)= 28^a$	<0.001**	$\eta^2= .32$
	60-100	93-100	92-100	82-100	60-100			
NYHA class (*n*, %)						$\chi^2(3)= 6.14$	0.105	$\varphi_c= .14$
Class I	266 (87.8)	77 (93.9)	66 (85.7)	68 (87.2)	55 (83.3)			
Class II, III, IV	37 (12.2)	5 (6.1)	11 (14.3)	10 (12.8)	11 (16.7)			
VO_2 Max (*mean, S.D., range*)	27.9 (8.3)	30.2 (8)	27.2 (8.4)	27 (8.4)	27 (7.9)	$f(3,310)= 3.12$	0.026	$\eta^2= .03$
	8-51	10-51	9-47	8-47	9-42			
Right ventricular function (*mean, S.D., range*)	57.4 (8.6)	63.4 (5.2)	55.4 (8.9)	54.2 (7.7)	55.9 (9.1)	$f(3,161.22)= 36.76^a$	<0.001**	$\eta^2= .18$

	23-75	40-73	23-73	30-68	29-75			
Left ventricular function (*mean, S.D., range*)	61 (8.6)	64.8 (7)	59.9 (7.7)	63 (8.2)	55.3 (8.7)	$f(3,310)= 21.50$	<0.001**	$\eta^2= .17$
	29-81	38-81	35-75	35-81	29-70			
Depression (*n*, %)						$\chi^2(3)= 7.38$	0.061	$\varphi_c= .16$
No depressive symptoms	238 (78.5)	59 (72)	63 (81.8)	68 (87.2)	48 (72.7)			
With depressive symptoms	65 (21.5)	23 (28)	14 (18.2)	10 (12.8)	18 (27.3)			

[a] Welch Anova; *p<.01, **p<.001; φ_c: .10= small, .30= medium, .50= large; η^2: .01= small, .06= medium, .14= large.

SV – Single Ventricle, TGA – Transposition of the Great Arteries, ToF – Tetralogy of Fallot, NYHA – New York Health Assessment.

Table 3. Physical & psychosocial HRQoL (SF-36) comparisons with norms

Scale/Subscale		*Mean (S.D.)*	*T-test*	*Sig.*	*99% CI of the Difference*	*d*
PCS						
	Total	50.9 (9.4)	*t*(301)= 1.74	0.083	-0.46, 2.34	0.09
	Simple	51.5 (10)	*t*(81)= 1.38	0.170	-1.38, 4.42	0.15
	ToF	52.8 (8.7)	*t*(76)= 2.84	0.006*	0.20, 5.44	0.30
	TGA	51 (7.5)	*t*(76)= 1.18	0.241	-1.25, 3.27	0.11
	SV	47.9 (10.8)	*t*(65)= -1.56	0.123	-5.59, 1.45	0.20
MCS						
	Total	48.2 (10.9)	*t*(301)= -2.95	0.003*	-3.47, -0.22	0.17
	Simple	45.9 (12)	*t*(81)= -3.06	0.003*	-7.57, -0.56	0.37
	ToF	50 (9.4)	*t*(76)= -0.00	0.997	-2.83, 2.82	0
	TGA	50.4 (9.1)	*t*(76)= 0.38	0.703	-2.33, 3.12	0.04
	SV	46.1 (12.2)	*t*(65)= -2.56	0.013	-7.84, 0.14	0.35

Note. Norm mean 50 (10). Positive and negative t values indicate better and poorer HRQoL respectively; *p<.01

MCS – Mental Component Summary, PCS – Physical Component Summary, SV – Single Ventricle, TGA – Transposition of the Great Arteries, ToF – Tetralogy of Fallot.

Table 4. Group comparisons in generic & disease-specific HRQoL

Scale		*Mean (S.D.)*	*Model Parameters*			
			f	*df*	*Sig.*	η^2
PCS			2.93[a]	161.15	0.035	0.03
	Simple	51.5 (10)				
	ToF	52.8 (8.7)				
	TGA	51 (7.5)				
	SV	47.9 (10.8)				
MCS			3.76[a]	161.55	0.012	0.04
	Simple	45.9 (12)				
	ToF	50 (9.4)				
	TGA	50.4 (9.1)				
	SV	46.1 (12.2)				
Symptoms			4.02	299	0.008*	0.04
	Simple	87.41 (14.22)				
	ToF	87.96 (12.28)				
	TGA	87.70 (13.04)				
	SV	80.92 (16.63)				
Impact cardiac surveillance			5.04	299	0.002*	0.05
	Simple	87.96 (10.49)				
	ToF	87.38 (9.36)				
	TGA	84.88 (10.04)				
	SV	81.97 (11.36)				
Worries			4.03	299	0.008*	0.04
	Simple	82.10 (14)				
	ToF	84.69 (13.60)				
	TGA	84.17 (15.32)				
	SV	76.79 (17.14)				

[a]Welch Anova; *p<.01

MCS – Mental Component Summary, PCS – Physical Component Summary, SV – Single Ventricle, TGA – Transposition of the Great Arteries, ToF – Tetralogy of Fallot.

Table 5. Final regression models for PCS and MCS

	PCS				MCS			
	B (SE)	***β***	***t***	***Sig.***	***B (SE)***	***β***	***t***	***Sig.***
(constant)	41.83 (13.38)		3.13	0.002*	46.91 (3.60)		13.02	<0.001**
Age	-0.01 (0.05)	-0.01	-0.20	0.840	0.17 (0.04)	0.16	4.03	<0.001**
Education level	1.30 (0.88)	0.07	1.47	0.143	-	-	-	-
Employment status	0.94 (0.95)	0.05	0.99	0.323	1.78 (0.98)	0.07	1.82	0.070
ToF	2.12 (1.20)	1.00	1.77	0.077	-	-	-	-
TGA	0.43 (1.20)	0.02	0.36	0.720	-	-	-	-
SV	2.58 (1.57)	0.11	1.64	0.101	-	-	-	-
Cyanosis days	0 (0)	-0.07	-1.10	0.274	-	-	-	-
Co-morbidities no.	-0.27 (0.49)	-0.03	-0.56	0.578	-	-	-	-
Hospitalisation days	-0.01 (0.01)	-0.07	-1.39	0.167	-	-	-	-
Current O_2 saturation	0.16 (0.13)	0.08	1.17	0.242	-	-	-	-
Medication no.	0.04 (0.42)	0.01	1.00	0.921	-	-	-	-
VO_2 max	0.02 (0.06)	0.02	0.32	0.753	-	-	-	-
Consequences	-0.43 (0.24)	-0.13	-1.83	0.068	-0.16 (0.25)	-0.04	-0.64	0.524
Personal control	0.12 (0.14)	0.04	0.84	0.403	-0.03 (0.16)	-0.01	-0.18	0.858
Treatment control	-	-	-	-	0.13 (0.15)	0.03	0.83	0.407
Identity	-2.06 (0.26)	-0.52	-7.95	<0.001**	-0.17 (0.26)	-0.04	-0.64	0.521
Concern	-0.19 (0.20)	-0.06	-0.96	0.340	-0.14 (0.20)	-0.04	-0.69	0.493
Emotional representation	0.25 (0.22)	0.08	1.12	0.264	-0.23 (0.22)	-0.07	-1.02	0.311
Positive reframing	-	-	-	-	0.82 (0.51)	0.07	1.60	0.111
Acceptance	-	-	-	-	0.62 (0.61)	0.04	1.02	0.309
Self-distraction	-	-	-	-	-0.75 (0.56)	-0.06	-1.33	0.186
Denial	-	-	-	-	-0.67 (1.05)	-0.03	-0.63	0.527
Religion	-0.69 (0.48)	-0.06	-1.43	0.154	-	-	-	-
Venting	0.53 (0.63)	0.04	0.84	0.400	-0.28 (0.70)	-0.02	-0.40	0.693
Substance use	-	-	-	-	0.00 (0.88)	0.00	0.00	0.998
Behavioural disengagement	-0.18 (0.92)	-0.01	-0.20	0.841	0.63 (1.04)	0.03	0.61	0.545

Self-blame	-	-	-	-	-2.81 (0.74)	-0.17	-3.78	<0.001**
Friends support	-	-	-	-	0.69 (0.51)	0.06	1.35	0.179
Significant other support	-	-	-	-	-0.06 (0.46)	-0.01	-0.14	0.888
Anxiety	-0.61 (0.86)	-0.04	-0.71	0.479	-2.49 (0.94)	-0.13	-2.65	0.008*
Depression	-0.08 (1.26)	0.00	-0.06	0.951	-11.14 (1.31)	-0.42	-8.51	<0.001**

Step	***R^2***	***Adjusted R^2***	***ΔR^2***	***R^2***	***Adjusted R^2***	***ΔR^2***
Step 1	.079	.069	.079**	.095	.089	.095**
Step 2	.195	.172	.116**	.345	.327	.249**
Step 3	.202	.178	.008	.465	.435	.121**
Step 4	.235	.203	.033**	.472	.438	.006
Step 5	.510	.481	.275**	.605	.577	.134**
Step 6	.515	.481	.005	-	-	-
Step 7	.516	.478	.001	-	-	-

Note. Variables were only included if they were significant in the bivariate analysis; Educational level: 0= school, 1= university; Employment status: 0= unemployed, 1= employed; ToF, TGA, SV: dummy-coded- reference group was Simple; CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms; *p<.01, **p<.001

MCS – Mental Component Summary, PCS – Physical Component Summary, SV – Single Ventricle, TGA – Transposition of the Great Arteries, ToF – Tetralogy of Fallot.

Table 6. Final regression models for symptoms, impact cardiac surveillance, and worries

	Symptoms				Impact Cardiac Surveillance				Worries			
	B (SE)	***β***	***t***	***Sig.***	***B (SE)***	***β***	***t***	***Sig.***	***B (SE)***	***β***	***t***	***Sig.***
(constant)	64.77 (18.50)		3.50	0.001*	93.07 (14.18)		6.57	<0.001**	67.03 (18.58)		3.61	<0.001**
Age	-	-	-	-	-	-	-	-	0.31 (0.07)	0.22	4.43	<0.001**
Gender	-1.47 (1.24)	-0.05	-1.18	0.239	-	-	-	-	-	-	-	-
Employment status	2.38 (1.30)	0.08	1.83	0.069	-	-	-	-	-	-	-	-
ToF	-0.32 (1.64)	-0.01	-0.19	0.847	0.38 (1.50)	0.02	0.25	0.802	2.13 (1.85)	0.06	1.15	0.250
TGA	-0.71 (1.65)	-0.02	-0.43	0.669	-1.91 (1.58)	-0.08	-1.21	0.228	1.67 (1.91)	0.05	0.87	0.384
SV	-0.42 (2.11)	-0.01	-0.20	0.841	-0.86 (1.95)	-0.03	-0.44	0.661	2.19 (2.39)	0.06	0.92	0.361
Cyanosis days	0.00 (0.00)	0.04	0.82	0.414	-	-	-	-	-	-	-	-
Co-morbidities no.	-0.62 (0.71)	-0.05	-0.87	0.385	0.14 (0.65)	0.01	0.21	0.834	-0.20 (0.67)	-0.01	-0.30	0.765
Arrhythmias	0.98 (1.56)	0.03	0.63	0.528	-3.32 (1.49)	-0.14	-2.22	0.027	-	-	-	-
Intervention no.	-	-	-	-	-0.53 (0.45)	-0.07	-1.18	0.241	-	-	-	-
Hospitalisation days	0.01 (0.01)	0.04	0.96	0.339	-	-	-	-	-	-	-	-
Current O_2 saturation	0.31 (0.18)	0.10	1.69	0.092	0.08 (0.14)	0.04	0.59	0.558	0.16 (0.18)	0.05	0.91	0.365
Medication no.	-0.07 (0.56)	-0.01	-0.13	0.896	-0.24 (0.53)	-0.03	-0.45	0.651	-	-	-	-
VO_2 max	0.14 (0.08)	0.08	1.80	0.072	0.01 (0.07)	0.01	0.21	0.832	-	-	-	-
LVEF	0.06 (0.07)	0.04	0.90	0.370	-	-	-	-	-	-	-	-

Consequences	-0.31 (0.32)	-0.06	-0.97	0.332	-0.04 (0.30)	-0.01	-0.14	0.892	-1.00 (0.38)	-0.18	-2.64	0.009*
Personal control	-0.09 (0.19)	-0.02	-0.47	0.640	-	-	-	-	-0.12 (0.23)	-0.02	-0.52	0.606
Treatment control	-	-	-	-	-	-	-	-	0.35 (0.24)	0.07	1.44	0.150
Identity	-2.62 (0.34)	-0.43	-7.64	<0.001**	-0.64 (0.33)	-0.14	-1.96	0.051	-0.89 (0.42)	-0.14	-2.15	0.033
Concern	-0.54 (0.27)	-0.11	-2.02	0.045	-0.50 (0.26)	-0.14	-1.97	0.050	-0.79 (0.32)	-0.15	-2.49	0.013
Emotional representation	0.01 (0.29)	0.00	0.02	0.987	-0.45 (0.28)	-0.13	-1.62	0.106	-0.52 (0.35)	-0.11	-1.49	0.138
Planning	-	-	-	-	-1.64 (0.59)	-0.14	-2.76	0.006*	-2.02 (0.81)	-0.12	-2.50	0.013
Self-distraction	-0.25 (0.71)	-0.02	-0.35	0.724	-	-	-	-	-0.42 (0.86)	-0.02	-0.49	0.626
Denial	-1.28 (1.34)	-0.04	-0.96	0.339	-	-	-	-	-	-	-	-
Venting	0.74 (0.89)	0.04	0.83	0.409	0.27 (0.82)	0.02	0.33	0.739	1.85 (1.08)	0.09	1.72	0.087
Substance use	-0.21 (1.15)	-0.01	-0.18	0.854	0.06 (1.07)	0.00	0.05	0.959	-1.97 (1.36)	-0.07	-1.45	0.147
Behavioural disengagement	0.37 (1.34)	0.01	0.28	0.781	-1.86 (1.18)	-0.09	-1.57	0.117	2.58 (1.54)	0.08	1.68	0.094
Self-blame	-0.61 (0.95)	-0.03	-0.65	0.518	-	-	-	-	-2.89 (1.61)	-0.13	-2.49	0.013
Family support	-	-	-	-	-	-	-	-	0.41 (0.93)	0.02	0.44	0.659
Friends support	-	-	-	-	-	-	-	-	0.81 (0.83)	0.05	0.98	0.330
Anxiety	-2.41 (1.18)	-0.10	-2.05	0.042	-2.91 (1.08)	-0.16	-2.68	0.008*	-1.62 (1.43)	-0.06	-1.13	0.261
Depression	-7.47 (1.70)	-0.22	-4.38	<0.001**	-0.12 (1.60)	-0.01	-0.08	0.939	-8.29 (2.03)	-0.22	-4.08	<0.001**
Step	***R^2***	***Adjusted R^2***		***ΔR^2***	***R^2***	***Adjusted R^2***		***ΔR^2***	***R^2***	***Adjusted R^2***		***ΔR^2***
Step 1	.094	.088		.094**	.126	.111		.126**	.023	.020		.023*

Step 2	.207	.185	.113**	.129	.112	.003	.104	.089	.081**
Step 3	.207	.182	.000	.154	.128	.024	.116	.098	.012
Step 4	.278	.246	.071**	.322	.292	.168**	.437	.414	.321**
Step 5	.581	.555	.303**	.347	.309	.025	.480	.447	.043*
Step 6	.593	.558	.011	.365	.323	.018	.487	.450	.006
Step 7	.634	.599	.041**	-	-	-	.523	.486	.037**

Note. Variables were only included if they were significant in the bivariate analysis; ToF, TGA, SV: dummy-coded- reference group was Simple; CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms; *p< .01, **p< .001

LVEF – Left Ventricular Ejection Fraction, SV – Single Ventricle, TGA – Transposition of the Great Arteries, ToF – Tetralogy of Fallot.

Figure 1. Order of entry of predictor variables in hierarchical multiple regressions

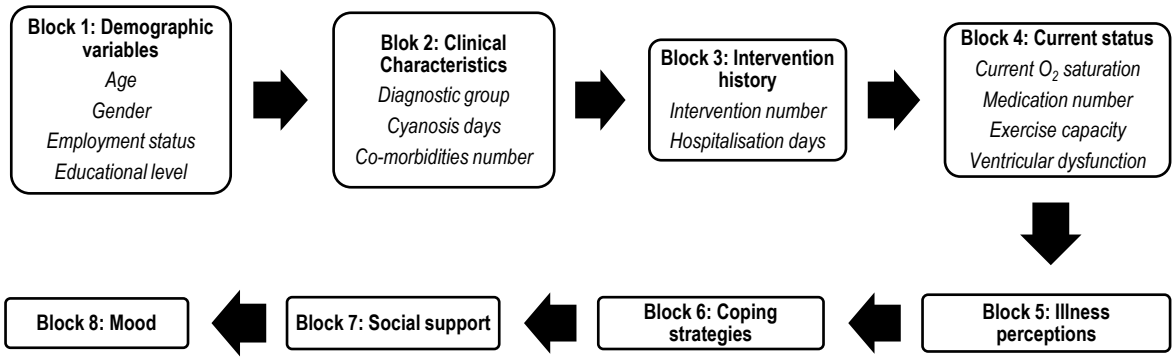

Figure 2. Percentages of participants achieving the MID below and within/above the norm on the SF-36