CONGENITAL HEART DISEASE

# Is the severity of congenital heart disease associated with the quality of life and perceived health of adult patients?

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**Objective:** To explore whether the severity of congenital heart disease is associated with the quality of life and perceived health status of adult patients.
**Design:** Descriptive, cross sectional study.
**Setting:** Adult congenital heart disease programme in one tertiary care centre in Belgium.
**Patients:** 629 patients (378 men, 251 women) with a median age of 24 years.
**Main outcome measures:** Disease severity was operationalised in terms of initial diagnosis (classification of Task Force 1 of the 32nd Bethesda Conference), illness course (disease severity index), and current functional status (New York Heart Association (NYHA) class, ability index, congenital heart disease functional index, and left ventricular ejection fraction). Quality of life was measured by a linear analogue scale, the satisfaction with life scale, and the schedule for evaluation of individual quality of life. Perceived health status was also assessed with a linear analogue scale.
**Results:** Scores derived from the disease severity classification systems were weakly negatively associated with quality of life and health status, ranging from −0.05 to −0.27. The NYHA functional class and ability index were consistently associated with quality of life and perceived health.
**Conclusions:** This study showed that the severity of congenital heart disease is marginally associated with patients' quality of life and perceived health. Functional status was more related to patients' assessment of their quality of life than was the initial diagnosis or illness course.

Congenital heart diseases comprise a wide spectrum of heart defects with varying levels of severity. The type of heart defect may affect the progress of the disease, the prognosis, and patients' ability to carry out normal functions. Indeed, some conditions, such as mild valvar disease or a small ventricular septal defect, do not require specific treatment or specialised follow up.[1] Conversely, more complex defects such as a univentricular heart or transposition of the great arteries require surgical treatment and lifelong follow up care in a tertiary care centre specialised in paediatric or adult congenital heart disease.[1] In general, more severe heart defects are associated with worse outcomes.[2]

In addition to the medical problems, many patients with congenital heart disease are facing specific psychosocial, educational, and behavioural challenges and issues. In this respect, the feeling of being different from peers, social impediments due to physical restrictions, and problems obtaining employment and insurance are often reported.[3] In many patients, congenital heart disease can be considered to be a chronic condition. This is particularly so when a patient's daily life is impeded by the long term nature of congenital heart disease, when there is a prevailing uncertainty with respect to the course of the illness and its prognosis, and when the patient experiences symptoms of the illness and restrictions in the level of activity.[4] It is assumed that these barriers influence patients' quality of life.[5] It is, however, uncertain whether the quality of life is associated with the severity of the congenital heart disease.

Few studies have attempted to examine the relation between disease severity and the quality of life of adults with congenital heart disease.[6–8] A potential relation between severity and quality of life may also be inferred from a series of studies that have applied the same instrument in different populations of heart defects.[9–12] In these studies, quality of life was measured in terms of either subjective health status,[6 9–12] the emotional response to health problems,[8] or external life conditions, interpersonal relationships, and internal psychological states.[7] Patients were categorised according to the severity of their disease, based on the initial diagnosis,[7 9–12] the New York Heart Association (NYHA) functional class, the ability index of Somerville, and criteria such as cyanosis,[6] arrhythmia, cardiac failure, and residual defects.[8]

Some associations were found between functional class and aspects of subjective health status. Although most studies have defined quality of life in terms of health status, there is more evidence that quality of life and health status are conceptually different issues.[13 14] It is therefore not appropriate to use a subjective assessment of health status as a measure of quality of life. Moreover, the respective studies measured disease severity in various ways. Indeed, disease severity is a problematic concept with respect to congenital heart disease, since several issues such as initial diagnosis, the course of the illness, functional status, or prognosis may reflect the severity of the disease in each patient. A comprehensive assessment of disease severity in congenital heart defects therefore requires an evaluation of the respective aspects. An indicator of severity is not only the heart defect itself or patients' functional class but also the number of surgical and interventional procedures or the expected course duration.

The absence of a comprehensive assessment of disease severity and the various approaches to quality of life investigations make the available study results inconclusive. This study therefore focused on exploring the association between the various indicators of congenital heart disease

**Abbreviations:** CHD-TAAQOL, congenital heart disease-TNO/AZL adult quality of life; NYHA, New York Heart Association; Q1, first quartile; Q3, third quartile; SEIQoL-DW, schedule for the evaluation of individual quality of life-direct weighting; SF-36, 36 item short form health survey

severity and on both the quality of life and the perceived health of adult patients, based on a sound conceptual foundation.

## PATIENTS AND METHODS

### Study population

During a two year period, all adult (≥ 18 years old), literate, Dutch speaking patients who visited the outpatient clinic for congenital heart disease at the University Hospital of Leuven, Belgium, were asked to participate in the study. Patients were excluded if it was their first visit to the outpatient clinic at the centre, if they were assessed as having learning disabilities during the clinical interview, or if they were referred for or in follow up after percutaneous closure of an atrial septal defect or a patent foramen ovale. Informed consent was provided orally.

Seven hundred and sixteen patients with congenital heart disease met the inclusion criteria. However, 66 (9.2%) of these patients refused to participate in the study, eight (1.1%) felt too emotionally distressed to participate because the cardiologist just told them that they needed a reoperation or that pregnancy was contraindicated, and 13 (1.8%) were not included for practical reasons (for example, further technical investigations, taxi was waiting), yielding a sample of 629 patients. Table 1 summarises the demographical and clinical characteristics of the study sample. Three hundred and seventy eight male (60.1%) and 251 (39.9%) female patients were included. The median age was 24 years. The most prevalent primary diagnoses were tetralogy of Fallot and ventricular septal defect. Table 1 describes only the six most prevalent heart defects, occurring in more than 5% of the patient sample.

### Variables and measurement

#### Disease severity

Various components of congenital heart disease severity were measured: initial diagnosis; illness course; and current functional status (table 2).[1 15–18] To determine the need for specialised care, Task Force 1 of the 32nd Bethesda Conference of the American College of Cardiology categorised congenital heart diseases into three classes, mild, moderate, and severe heart defects,[1] based on the initial diagnosis or specific types of operations (table 2).

With respect to health services research, the disease severity index was developed to reflect the course of the illness.[15] The disease severity index encompasses three categories. ''Low severity'' was reserved for patients who had undergone a maximum of one cardiovascular operation or one catheterisation procedure. Patients who had undergone more than one cardiovascular operation or catheterisation were placed in the ''moderate severity'' group. Lastly, patients with persistent cyanosis, < 92% oxygen saturation at rest, or single ventricle physiology were placed in the ''high severity'' group (table 2).

To assess the current functional status, three classification methods were used: the NYHA functional class,[16] the ability index,[17] and the congenital heart disease functional index, which was developed by the department's research group for a previous study.[18] The ability index and the congenital heart disease functional class are specifically designed for adults with congenital heart disease, whereas the NYHA classification was initially developed for patients with heart failure and later on adapted for patients with angina.[16] A fourth indicator for patients' functional status was the left ventricular ejection fraction, as measured by echocardiography. This parameter is traditionally used by cardiologists as an indicator of patients' functioning.

**Table 1** Demographic and clinical characteristics of 629 adult patients with congenital heart disease

| Variable | Number |
|---|---|
| Sex (n = 629) | |
| Men | 378 (60.1%) |
| Women | 251 (39.9%) |
| Median age (years) (n = 629) | 24 |
| First quartile | 20 |
| Third quartile | 29 |
| Range | 18–66 |
| Marital status (n = 626) | |
| Unmarried (living with parents) | 346 (55.3%) |
| Living alone, divorced, or widowed | 60 (9.6%) |
| Married or cohabiting | 220 (35.1%) |
| Employment status (n = 629) | |
| Student | 175 (27.8%) |
| Blue collar worker | 149 (23.7%) |
| White collar worker | 189 (30.0%) |
| Independent | 37 (5.9%) |
| Unemployed or looking for work | 25 (4.0%) |
| Housewife | 12 (1.9%) |
| Retired | 5 (0.8%) |
| Not able to work or disabled | 17 (2.7%) |
| Other | 20 (3.2%) |
| Primary medical diagnosis | |
| Tetralogy of Fallot | 112 (17.8%) |
| Ventricular septal defect | 108 (17.2%) |
| Coarctation of the aorta | 89 (14.1%) |
| Congenital stenosis of aortic valve | 65 (10.3%) |
| Pulmonary valve stenosis (congenital) | 48 (7.6%) |
| Complete transposition of great vessels | 37 (5.9%) |

#### Quality of life

Although the concept of quality of life is often discussed in the biomedical literature, there is still no consensus on its definition, conceptualisation, or measurement. In preparation for this study, we evaluated the different conceptualisations of quality of life.[28] For this evaluation, we identified six critical conceptual pitfalls regarding this concept.[28] Firstly, it is argued that quality of life may not be interchanged with health status or functional abilities. Secondly, it is more likely that quality of life relies on a subjective appraisal than on objective parameters. Thirdly, quality of life research is often characterised by a poor distinction between indicators and determinants of quality of life. Fourthly, quality of life can change over time but is not highly fluctuating. Fifthly, although quality of life is mostly expressed negatively, for example, in terms of impediments, several issues can contribute to a good quality of life. This illustrates that quality of life should be approached positively as well. Lastly, the assessment of overall quality of life is preferred above health related quality of life. By focusing on health related quality of life, investigators may substantially overestimate the impact of health related factors and, conversely, may seriously undervalue the effect of non-medical phenomena.[19]

Relying on published concept analyses of quality of life[20–22] and considering the pitfalls described in the literature,[28] it is indicated that quality of life should be defined and measured in terms of life satisfaction. Therefore, we defined quality of life as ''the degree of overall life satisfaction that is positively or negatively influenced by people's perception of certain aspects of life important to them, including matters both related and unrelated to health''.

##### *Linear analogue scale*

The overall quality of life was measured with a linear analogue scale. This is a vertical, graded, 10 cm line, ranging from the ''worst imaginable quality of life'' (score of 0) to the ''best imaginable quality of life'' (score of 100). The use of this linear analogue scale allows patients to give their own rating of their overall perceived quality of life. Linear

**Table 2** Classification systems to categorise patients with congenital heart disease according to the severity of the disease

| Classification system | Prevalence |
|---|---|
| Initial diagnosis (n = 629) | |
| Task Force 1 of the 32nd Bethesda conference of the American College of Cardiology[1] | 164 (26.1%) |
| • Mild (for example, isolated aortic valve disease, closed ASD without residua) | 365 (58.0%) |
| • Moderate (for example, coarctation of the aorta, tetralogy of Fallot) | 100 (15.9%) |
| • Severe (for example, Fontan operation, Eisenmenger, double outlet ventricle) | |
| Illness course (n = 629) | |
| Disease severity index[15] | |
| • Low: maximum 1 cardiovascular operation or 1 catheterisation procedure | 404 (64.2%) |
| • Moderate: more than 1 cardiovascular operation or catheterisation | 203 (32.3%) |
| • High: persistent cyanosis, <92% oxygen saturation at rest, or single ventricle physiology | 22 (3.5%) |
| Current functional status (n = 627) | |
| NYHA functional class[16] | |
| • Class I: patients with cardiac disease but without resulting limitation of physical activity; ordinary physical activity does not cause undue fatigue, palpitation, dyspnoea, or anginal pain | 511 (81.5%) |
| • Class II: patients with cardiac disease resulting in slight limitation of physical activity; they are comfortable at rest; ordinary physical activity results in fatigue, palpitation, dyspnoea, or anginal pain | 85 (13.6%) |
| • Class III: patients with cardiac disease resulting in major limitation of physical activity; they are comfortable at rest; less than ordinary activity causes fatigue, palpitation, dyspnoea, or anginal pain | 26 (4.1%) |
| • Class IV: patients with cardiac disease resulting in inability to carry on any physical activity without discomfort; symptoms of heart failure or the anginal syndrome may be present even at rest; if any physical activity is undertaken, discomfort increases | 5 (0.8%) |
| Ability index[17] (n = 628) | |
| • Class 1: normal life (full time work or school, pregnancy poses no health risk) | 524 (83.4%) |
| • Class 2: able to work (intermittent symptoms, interference with life, pregnancy possible) | 78 (12.4%) |
| • Class 3: unable to work (limitation of all activities, pregnancy poses health risk) | 24 (3.8%) |
| • Class 4: extreme limitation (dependent, almost house bound) | 2 (0.3%) |
| Congenital heart disease functional index[18] (n = 628) | |
| • Class 1: no surgery, good clinical status, medical follow up not strictly necessary | 24 (3.8%) |
| • Class 2: with or without surgery, functionally perfect, postoperative normalisation of clinical condition, medical check up every 3 to 5 years, competitive sports permitted | 221 (35.2%) |
| • Class 3: with or without surgery, functionally good, medical restrictions, medical check up every 1 to 2 years, recreational sports permitted | 355 (56.5%) |
| • Class 4: with or without surgery, moderate functional status, functioning at own pace, medical check up every year | 23 (3.7%) |
| • Class 5: with or without palliative surgery, bad functional status, cyanosis, medical check up every 6 to 12 months | 5 (0.8%) |
| Left ventricular ejection fraction | Median: 62 (IQR: 15) |

ASD, atrial septal defect; IQR, interquartile range; NYHA, New York Heart Association.

analogue scales are widely used in quality of life research, particularly in cancer populations.[23] It can be used to measure a variety of symptoms and aspects of functioning, as well as overall quality of life.[23 24] We used the linear analogue scale in this study because of its advantages of being easy to use[25] and less burdensome for respondents.[26] This simplicity and ease of use may result in high response rates and operational efficiency of the study.[24] Despite their simplicity, such analogue scales have been shown in several studies to be valid, reliable, and responsive to changes in clinical conditions.[23 24 27] We explored some lines of evidence on validity, reliability, and responsiveness of the linear analogue scale for quality of life in adults with congenital heart disease, indicating good psychometric properties for the use in this population of patients. Details of the psychometric evidence are described elsewhere.[28]

### *Satisfaction with life scale*

Because the quality of life was defined in terms of life satisfaction, the satisfaction with life scale[29] was used as a second indicator of quality of life. This instrument comprises five statements and seven response categories, ranging from "strongly disagree" to "strongly agree". An aggregate score can be obtained by summing the scores of the individual items. The minimum score of life satisfaction is 5 and the maximum score is 35. The validity and reliability have been extensively assessed in previous studies and indicate good psychometric properties.[30] For their use with patients with congenital heart disease, we also found this instrument to be psychometrically sound.[28]

### *Schedule for the evaluation of individual quality of life-direct weighting*

To explore the determinants of quality of life, we used the schedule for the evaluation of individual quality of life-direct weighting (SEIQoL-DW).[31] The SEIQoL-DW was developed to examine quality of life from an individual perspective. It consists of three successive steps: firstly, the respondent nominates the five areas that are most important for his or her quality of life; secondly, the actual status of each specified area is rated from 0 to 100 on a visual analogue scale; and, thirdly, the relative importance of each selected area is quantified relative to each other area with the use of a five segment disk. The use of SEIQoL-DW overcomes the problem of predetermined questions, which assumes that each person's quality of life is influenced by the same determinants and that different aspects of life are equally important for all people.[32] We evaluated validity, reliability, and responsiveness of the SEIQoL-DW for use in adults with congenital heart disease.[33] Although the SEIQoL-DW cannot

be regarded as a measure of quality of life itself, it is a valid and reliable instrument to explore determinants for patients' quality of life.[33] Responsiveness of the SEIQoL-DW in patients with congenital heart disease may be problematic.[33] The use of the SEIQoL-DW allows the calculation of a single index by summing the products of the rated level and applying weights for each of the five areas. This index ranges from 0 to 100. Note, however, that the SEIQoL-DW index cannot be regarded as a quality of life index but rather as an aggregate score of the most important determinants.

### Health status

Because there is a substantial difference between quality of life and self perceived health, we also measured health status in this study. Subjectively perceived health status was measured with a linear analogue scale ranging from the ''worst imaginable health state'' (score of 0) to the ''best imaginable health state'' (score of 100). This linear analogue scale is part of the EuroQol instrument and previous studies have reported its good validity and reliability.[34] Also in adults with congenital heart disease, this linear analogue scale has been shown to be valid, reliable, and responsive.[28]

### Procedure

After the advanced clinical nurse practitioner and the cardiologist saw the patients during their scheduled visit at the outpatient clinic, an independent researcher approached the patients to explain the purpose, procedure, and time required to participate in the study. Patients were instructed on how to fill out the questionnaires after giving oral informed consent. The researcher stayed with the patient to provide clarification if needed and to ensure that patients filled out the questionnaires independently, without assistance from accompanying people. The questionnaires were checked for completeness and patients were asked to complete missing data if necessary. The data collection procedure lasted about 15–20 minutes. This study was approved by the local ethics committee and has therefore been performed in accordance with international ethical standards.

One cardiologist scored the NYHA, ability index, and congenital heart disease functional index based on data from the clinical examination. Interrater reliability was therefore not an issue in this study. The cardiologist was blinded as to other outcomes. Data on left ventricular ejection fraction were retrieved from the medical record.

### Statistical analysis

The data were analysed with SPSS 9.0 (SPSS Inc, Chicago, Illinois, USA). Nominal level data were expressed in percentages. After having been checked for normality, medians and first (Q1) and third quartiles (Q3) were calculated because continuous variables were not normally distributed. Spearman's correlation coefficients were calculated to evaluate the relation between the severity of the congenital heart disease and quality of life. Cyanotic and acyanotic patients were compared by the Mann-Whitney U test. All tests were two sided with a level of significance set at $p \leqslant 0.05$. The Bonferroni correction was applied to adjust for the inflating type I error in multiple testing.

Since the satisfaction with life scale is an ordinal level instrument, summation of the scores of the individual items is not appropriate.[35] Yet, in the descriptive statistics, we calculated an overall score by summing the items scores to allow comparison with published data. For the inferential statistics, however, we transformed the ordinal scale into a probability scale by means of ridit analysis.[35] A ridit was calculated for each patient, representing an aggregate score over all items for that patient.

## RESULTS

### Disease severity

According to the initial diagnosis based on the Task Force 1 classification, a majority of the patients were classified as having moderate congenital heart disease (table 2). In contrast, the disease severity index classified 64.2% as having a mild heart defect. The NYHA classification and the ability index were fairly comparable with 81.5% and 83.4% of patients, respectively, in class I. The congenital heart disease functional index placed 56.5% of the patients in class 3 (table 2). The median left ventricular ejection fraction was available for 491 patients. The median ejection fraction was 62%.

Frequency distributions indicate that these classification schemes measure different indicators of disease severity. This was confirmed by the correlation coefficients between the respective classifications, which ranged from 0.25–0.49, except for the relation between NYHA and ability index ($r_s = 0.86$).

### Quality of life and perceived health

Overall, the quality of life of adults with congenital heart disease was good. The median scores on the linear analogue scale and the satisfaction with life scale were 80 (Q1 = 75, Q3 = 87) and 28 (Q1 = 24, Q3 = 30), respectively. The median SEIQoL-DW index was 79.04 (Q1 = 69.56, Q3 = 87.20). Patients perceived their health to be good, the median linear analogue scale score being 80 (Q1 = 70, Q3 = 90).

Since we defined quality of life in terms of life satisfaction, we assumed that the linear analogue scale and the satisfaction with life scale are highly interrelating. We found a correlation coefficient of 0.52 ($p < 0.001$). According to the cut off boundaries for small (0.1–0.3), moderate (0.3–0.5), and large correlations (> 0.5),[36] both scales can be regarded as highly interrelating. On the other hand, the expected low to moderate correlation with the SEIQoL-DW ($r_s = 0.42$, $p < 0.001$) and the linear analogue scale of health status ($r_s = 0.37$, $p < 0.001$) was confirmed. The correlation between the SEIQoL-DW and the linear analogue scale of health status was even lower ($r_s = 0.31$, $p < 0.001$).

### Relation between disease severity and quality of life

Scores derived from the disease severity classification systems indicated a weak and negative association with quality of life parameters and perceived health (table 3). The correlations were significant only for parameters reflecting functional status, such as NYHA, ability index, and congenital heart disease functional index. The highest correlations with functional status were found for satisfaction with life and perceived health.

Comparison of the 20 patients with cyanotic conditions versus the 609 patients with acyanotic conditions showed that there was no significant difference (Bonferroni correction: $p < 0.0125$) in quality of life parameters. However, health status was perceived to be significantly lower ($U = 4036$, $p = 0.01$) by cyanotic patients (median 72.5, Q1 = 60.5, Q3 = 80) than by acyanotic patients (median 80, Q1 = 70, Q3 = 90).

## DISCUSSION

This is the first study that comprehensively explored the association between the severity of congenital heart disease and quality of life, as well as the subjectively perceived health of a large sample of adults with congenital heart disease. The measurement of quality of life was built on a conceptual basis. Quality of life was defined and measured in terms of life satisfaction. This is in contrast to most quality of life reports, which measure quality of life as subjective health

**Table 3** Spearman's test correlation matrix for severity of congenital heart disease versus quality of life and health status

| Classification system | LAS quality of life | SWLS | SEIQoL-DW | LAS health status |
|---|---|---|---|---|
| Task Force 1 | −0.05 | −0.06 | −0.08 | −0.10 |
| Disease severity index | −0.09 | −0.09 | −0.05 | −0.12 |
| Functional status | | | | |
| NYHA | −0.20* | −0.28* | −0.18* | −0.27* |
| Ability index | −0.18* | −0.25* | −0.13* | −0.24* |
| Congenital heart disease functional index | −0.11 | −0.07 | −0.15* | −0.20* |
| Left ventricular ejection fraction | 0.10 | 0.07 | 0.06 | 0.04 |

*Bonferroni correction: $p<0.0025$.
LAS, linear analogue scale; SEIQoL-DW, schedule for the evaluation of individual quality of life-direct weighting; SWLS, satisfaction with life scale.

status. It has, however, been suggested that such an approach may be flawed, since quality of life and health status are two related, albeit distinct, concepts.[13 14] Indeed, the correlation between the linear analogue scale of quality of life and the linear analogue scale of health status in this study was 0.37 (95% confidence interval 0.30 to 0.44). Furthermore, a clear distinction was made between indicators (linear analogue scale, satisfaction with life scale) and determinants (SEIQoL-DW) of quality of life. An appropriate assessment of quality of life needs to include one or more indicators of quality of life itself and may not be limited to the measurement of possible influential factors such as physical functioning, symptoms, perceived health, and mood.

We found a weak, negative association between the severity of congenital heart disease, quality of life, and perceived health. Results showed that heart disease severity had a detrimental impact on patients' lives only when it was measured in terms of poor functional status. This means that the initial diagnosis or the course of the illness does not influence quality of life or perceived health. The finding that correlations between functional status parameters and quality of life or perceived health were low indicates that patients with more severe conditions do not experience the congenital heart disease having a major effect on the overall perception of their life situation.

The achievement of optimal ventricular function has traditionally been the mainstay of outpatient and inpatient treatment for patients with congenital heart disease. This is largely based on the assumption that quality of life will follow ventricular performance. The present study refutes this assumption, as no significant associations were found between left ventricular ejection fraction and quality of life or health status. Note, however, that data on left ventricular function were excluded for patients with a transposition of the great arteries who had undergone an atrial switch repair (Mustard or Senning), since the right ventricle is the systemic ventricle in these patients.

## Comparison with the literature

Empirical evidence on the relation between the severity of congenital heart disease and quality of life or subjective health status is scarce.[6–8 37] Using different conceptualisations and methods impedes the comparability of the results. None the less, some similarities can be observed. Ternestedt and colleagues[7] found that patients with tetralogy of Fallot rated their quality of life higher than patients with atrial septal defect. This indicates that more severe heart defects are not necessarily associated with worse quality of life. Furthermore, no association between quality of life and NYHA class was found.[7]

This study confirmed the findings of Lane and colleagues[6] that subjective health status in patients with cyanotic conditions is lower than in those with acyanotic conditions.

The correlations between perceived health status and NYHA and between health status and ability index were also found by Kamphuis and colleagues.[8] Correlation coefficients cannot, however, be directly compared because Kamphuis and colleagues used the 36 item short form health survey (SF-36) and we used the linear analogue scale to assess perceived health status.

The present study was also in line with the results of a series of articles published by Meijboom and colleagues. Their studies in patients with atrial septal defect,[11] ventricular septal defect,[10] tetralogy of Fallot,[9] and transposition of the great arteries[12] indicated that subjectively perceived health status in the four diagnostic categories was fairly even.

## Methodological limitations

In the absence of a de facto standard for the classification of the severity of congenital heart defects, various indicators of congenital heart disease severity were explored: initial medical diagnosis, the course of the illness, and functional status. The different angles of the respective classification systems are expressed by the disparities in frequency distribution (table 2) and by the limited interrelation. The validity of the classification schemes for use in quality of life research is uncertain. Only NYHA class and the ability index have previously been used in quality of life studies of congenital heart disease. Although the NYHA functional class is often used in clinical research on congenital heart disease, it purports to categorise patients with heart failure. If the classification system is merely based on the content of the NYHA class then it cannot be considered a valid tool for the categorisation of patients with heart defects. Nonetheless, the NYHA class was consistently associated with quality of life and health status. Regarding the congenital heart disease functional index, no evidence on reliability and validity is available to date. Interrater variability was precluded since classification schemes were filled out by one cardiologist.

Although a prognosis of life expectancy may also reflect the severity of a disease, this component was not included in this study, since individual prognosis cannot be predicted. Certain patients with congenital heart disease die of sudden death. So far, the risk of sudden death cannot be stratified.

The ideal means of assessing overall clinical and functional status is cardiopulmonary exercise testing, including the measurement of peak oxygen uptake. We did not include such exercise testing in the present study, which can be considered a methodological limitation. However, in a related study, we explored the association between maximum exercise capacity and peak oxygen uptake versus quality of life and self perceived health in a sample of 36 patients with tetralogy of Fallot or transposition of the great arteries. Both quality of life and perceived health were not significantly related to maximum exercise capacity and peak oxygen uptake, with correlation coefficients ranging from 0.06–0.26

(data on file). These findings should, however, be interpreted with a measure of caution because of the very small sample size. Therefore, we did not include these results in the present paper.

Although the sample of this study was large, it is not necessarily representative of the population of patients with congenital heart disease. This is because the eligible patients were recruited from the outpatient clinic of our centre. It should be noted that many patients born with heart defects are treated in the first years of life and do not need regular follow up care at a tertiary care centre. Patients with mild congenital heart diseases were underrepresented in this sample (26.1%), whereas this group accounts for 51% of the congenital heart disease population.[38] However, whether this underrepresentation has affected the results of this study is doubtful, since the severity of the congenital heart disease, in terms of initial diagnosis, does not influence patients' quality of life or perceived health. Furthermore, we did not include patients with mental retardation. Although inclusion of this group of patients would be of the utmost relevance, because they may be regarded as the most severely handicapped, they were excluded because self report by questionnaires requires intact intellectual abilities.

In this study, we evaluated only linear relations. Future research should therefore also investigate non-linear associations.

Recently, a disease specific instrument for adults with congenital heart disease was developed: congenital heart disease-TNO/AZL adult quality of life (CHD-TAAQOL).[39] Although we used this instrument in our sample,[28] we did not use it to assess the relation with disease severity because the CHD-TAAQOL does not result in a single aggregate score.

## Conclusion

This study showed that the severity of congenital heart disease is marginally associated with patients' quality of life. Patients' assessment of their quality of life relates more to functional status than to the initial diagnosis or the course of the illness. Stronger associations were found between perceived health and functional status. Patients with cyanotic heart defects had lower perceptions of the status of their health than did acyanotic patients. The findings of this study are crucial to the development of key strategies to enhance the quality of life of this patient population and to provide appropriate counselling.

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