Forty Years of Quality-of-Life Research in Congenital Heart Disease: Temporal Trends in Conceptual and Methodological Rigor

Ewa-Lena Bratt PhD, RN, [1,2] and Philip Moons PhD, RN, [1,3,4]

[1] Institute of Health and Care Sciences, Gothenburg University, Arvid Wallgrens backe 1, box 457, 40530 Gothenburg, Sweden

[2] Department of Pediatric Cardiology, The Queen Silvia Children's Hospital, Rondvägen 15, 41685 Gothenburg, Sweden

[3] KU Leuven Department of Public Health and Primary Care, Kapucijnenvoer 35, PB 7001, 3000 Leuven, Belgium

[4] The Heart Centre, Copenhagen University Hospital, Blegdamsvej 9, 2100 Copenhagen, Denmark

Total word count: 4963 words

Brief title: Quality of life in congenital heart disease

Address for correspondence: Philip Moons, KU Leuven Department of Public Health and Primary Care, Kapucijnenvoer 35, box 7001, B-3000 Leuven, Belgium; Tel: +32-16-373315; Fax: +32-16-336970; email: philip.moons@kuleuven.be

Abstract

Background: The first study on quality of life (QoL) in patients with congenital heart disease was published 40 years ago. Since then, the number of QoL articles on these patients has grown exponentially. We conducted a systematic literature review of all empirical studies on QoL in patients with congenital heart disease published since 1974, with the aim of determining the range of conceptual and methodological rigor of studies and identifying temporal trends in these parameters.

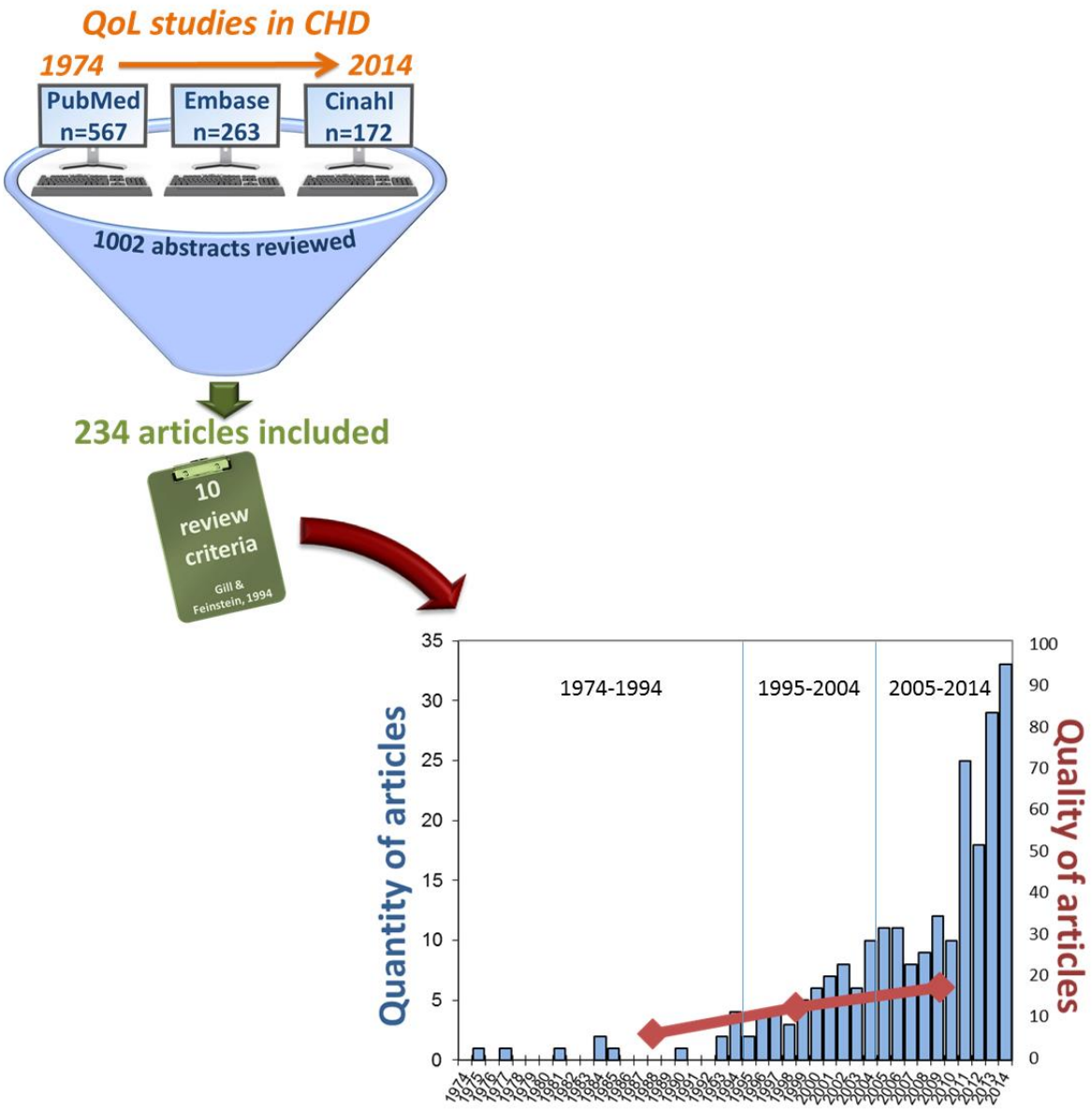
Methods: PubMed, Embase, and Cinahl were searched for empirical studies addressing QoL in children, adolescents, or adults with congenital heart disease, published between January 1, 1974, and December 31, 2014. We applied 10 review criteria that were previously developed by Gill and Feinstein in 1994 and further refined by Moons et al. in 2004. Overall, 234 articles were reviewed.

Results: We found slight but non-significant temporal improvements in conceptual and methodological rigor and in use of assessment methods. This indicates a trend toward a more professional and exacting approach in QoL assessments. However, the majority of articles still had substantial conceptual and methodological deficits. Furthermore, we observed that citation of the publications of Gill and Feinstein and Moons et al. in published QoL research is associated with higher quality scores, suggesting that these articles have a positive impact on conceptual and methodological caliber.

Conclusion: Despite 40 years of QoL research in this field, this review shows that major weaknesses in methodological rigor remain highly prevalent, which may make QoL studies inconclusive.

Key Words: Quality of Life; Review; Heart Defect, Congenital; Adolescent; Health Services Research/methods

Graphical abstract



1. Introduction

In 1974, Dr. Charlotte Ferencz published the first report on quality of life (QoL) in individuals with congenital heart disease [1]. Since then, a plethora of QoL studies have been published. The main driving force for investigating QoL in individuals with congenital heart disease is decreasing mortality and morbidity, which has resulted in prolonged life expectancy for these patients [2, 3]. Clinicians and researchers have been intrigued by the question of how do the lives of patients with a congenital heart defect "look like," and whether the heart defect has an impact on QoL.

QoL, however, is an equivocal construct [4-6]. There is no uniform definition for QoL, which yields a conceptual haziness [5]. This, in turn, results in inaccurate measurements of "true" QoL and in inconclusive study findings. To direct more attention to this phenomenon, in 1994 Gill and Feinstein evaluated the quality of existing QoL research [4]. They concluded that a large majority of QoL studies had conceptual limitations, hampering the validity of the study findings [4]. The Gill and Feinstein review is a landmark article, having been cited more than 1200 times to date (ISI Web of Science; accessed 03/19/2015).

Ten years later, Moons and coworkers [7] adopted the Gill and Feinstein methodology in evaluating studies of congenital heart disease. These investigators found that QoL studies in congenital heart disease also were lacking conceptual and methodological rigor, even more so than QoL studies in the general medical literature [4, 7]. It was striking that most researchers did not apply the same rigor to QoL studies as they did to defining and operationalizing outcomes in the area of morbidity and mortality. Indeed, successful research requires a rigorous scientific approach, in which the concepts used are properly defined and accurately documented. The methodologies applied and definitions used need to be described appropriately to allow for correct interpretation of findings and possible translation into practice.

It may be assumed, or hoped, that the articles of Gill and Feinstein [4] and Moons et al. [7] have since stimulated researchers to conduct and report their QoL studies more accurately. Hence, one would expect to observe a temporal improvement in the caliber of QoL studies, which might be a reasonable indicator of the influence of these two seminal publications. Therefore, the aims of this review were (i) to provide a description of research on QoL in congenital heart disease since the first publication from this field 40 years ago, and (ii) to determine whether temporal trends have emerged in conceptual and methodological rigor of QoL studies.

2. Methods

2.1 Search strategy

We performed a systematic literature search in PubMed, Embase, and Cinahl, including all empirical studies addressing QoL in children, adolescents, or adults with congenital heart disease. The search epoch was inclusive of January 1, 1974, through December 31, 2014. The actual search was conducted January 1, 2015. Articles on primary studies were eligible for inclusion, if they were available online (e-pub ahead of print) or in print. The search strings we employed in searching the three databases are detailed in Appendix 1 (Electronic Supplemental Material). To omit publications that were inappropriate for inclusion, a hierarchy of exclusion criteria was developed and used during the screening of the abstracts (Appendix 2: Electronic Supplemental Material).

2.2 Review criteria

To appraise the conceptual and methodological rigor of QoL studies in congenital heart disease, we applied the review criteria that were previously developed by Gill and Feinstein [4]. They developed 10 criteria to evaluate the caliber of QoL articles. These criteria were based on the following definition of QoL: *"Rather than being a description of patients' health*

status, QoL is a reflection of the way that patients perceive and react to their health status and to other, nonmedical aspects of their lives" [4]. The operationalization of the criteria was further refined by Moons et al. [7]. These criteria are extensively described in Appendix 3 (Electronic Supplemental Material).

2.3 Data selection process

A flow chart of the search and selection procedure is shown in Appendix 2 (Electronic Supplemental Material). Database searches resulted in 1002 records. After removing duplicates, the 769 remaining articles were independently screened for eligibility by both authors. For references in which there was no agreement between the two reviewers about exclusion or about the reasons for exclusion, meetings were held to reach consensus. After these consensus meetings, 535 records were excluded (Appendix 2).

Of the 234 included articles, full-text papers were independently reviewed and scored by the two authors using the 10 predefined criteria. Also here, consensus meetings were held to arrive at the final ratings of each individual article.

2.4 Data synthesis and analyses

Variables such as country where the study was conducted, age group, heart defect, sample size, and assessment tool/instrument were registered. The number of articles fulfilling each criterion was counted, and the percentage was calculated. To indicate how well individual articles performed with respect to all criteria, a summary score was calculated by summing the number of criteria an article fulfilled and dividing this sum by the number of criteria for which the article was eligible to be evaluated. The resulting value was then multiplied by 100. Hence, the main outcome measure was a summary score ranging from 0 (for articles complying with none of the criteria) to 100 (for articles complying with all criteria).

To investigate temporal trends in conceptual and methodological rigor since the publication of the first QoL article in congenital heart disease in 1974, the entire time period up to 2014 was divided into three epochs, using the publication years of Gill and Feinstein [4] and Moons et al. [7] as cutoffs between the epochs. The first period (1974-1994) is the epoch before the Gill and Feinstein review [4]. The second period (1995-2004) is the epoch after the Gill and Feinstein review [4], but before the publication of the Moons et al. article [7]. The third period (2005-2014) is the epoch after the Moons et al. publication [7]. For each period, we calculated the mean and median summary scores for each article. A one-way ANOVA was computed to investigate whether the mean summary scores for the three time periods were significantly different. A Chi-square test was performed on the proportion of studies complying with each individual criterion, over the three periods. To determine whether the publications of Gill and Feinstein [4] and Moons et al. [7] influenced the quality of the articles, we computed a Mann Whitney U test to compare the median scores before and after the respective publications. All tests were two-sided, and $P<0.05$ was used as the cutoff for statistical significance.

3. Results

3.1 Description of quality of life publications from 1974 to 2014

To date, 234 empirical studies addressing QoL in patients with congenital heart disease have been published. The number of articles grew exponentially, starting with 1-2 articles per year in the first 20 years up to almost 35 articles during 2014 (Figure 1). Most studies included patients with a mix of different diagnoses and complexities (n=106 articles; 45.3%). Of the studies that included patients with a specific diagnosis, most were on patients with transposition of the great arteries (n=26; 11.1%); single ventricle physiology (n=25; 10.7%); or Fallot/double outlet right ventricle (n=20; 8.5%) (Figure 2). Forty-five studies investigated children aged 0-9 years (19.2%); 12 studies included adolescents aged 10-18 years (5.1%);

104 studies included adult patients >19 years (44.4%); and 73 studies included patients of different age groups (31.2%). The majority of the studies were performed in the United States (n=52; 22.2%); followed by the Netherlands (n=37; 15.8%); Germany (n=31; 13.2%); United Kingdom (n=15; 6.4%); and Belgium (n=13; 5.6%) (Appendix 4: Electronic Supplemental Material). The most commonly used assessment was the Short form-36 (SF-36) (n=62; 26.5%), followed by the Pediatric Quality of Life inventory (PedsQL) (n=25; 10.7%).

PLEASE, INSERT FIGURES 1 AND 2 ABOUT HERE

3.2 Temporal trends in conceptual and methodological rigor

For all studies published from 1974 to 2014, the mean summary score was 15.2 (standard deviation: 22.1) and the median score was 0 (interquartile range: 0-25). Over the three distinct periods, a slight increase of the mean score was observed (Figure 3). This was because in 2005-2014, 15.7% of the studies had a summary score of 50 or higher compared to 5.5% of the studies in 1995-2004 and 0% of the studies in 1974-1994. The increase in mean score was, however, not statistically significant (F=2.267; p=0.106). The median summary score increased in the second period (1995-2004) but dropped again to zero during the third period (2005-2014) (Figure 3).

PLEASE, INSERT FIGURE 3 ABOUT HERE

Over the three time periods, a statistically significant increase was observed for criterion 1 (providing a definition), criterion 3 (providing reason for instrument), and criterion 4 (calculating index score) (Table 1). Still, only 23-34% of the studies fulfilled these four criteria today. Criteria 5-10 were seldom fulfilled. Indeed, in only a few studies were patients asked to give their own global rating for QoL (criterion 5), was overall QoL distinguished from health-related QoL (criterion 6), were patients invited to supplement the items that they considered to be relevant for their QoL (criterion 7), or were patients allowed to indicate which items were personally important to them (criterion 9). The two studies that fulfilled

criteria 7 and 9, however, included this score in the final ratings of QoL, resulting in a 100% score. Overall, the entire body of studies in congenital heart disease we evaluated performed worse than the studies included in the Gill and Feinstein review (Table 1). The scoring for each individual article reviewed, is presented in Appendix 5 (Electronic Supplemental Material).

PLEASE, INSERT TABLE 1 ABOUT HERE

In addition to the review criteria applied, we observed that in 1974-1994, 8 of 13 articles (61.5%) did not describe QoL in the methods or results section, but merely mentioned it in the abstract or discussion. This proportion decreased to 26 of 55 articles (47.3%) in 1995-2004; and further decreased to 19 of 166 articles (11.4%) in 2005-2014.

In order to determine whether the publications of Gill and Feinstein [4] and Moons et al. [7] may have had an impact on improving the conceptual and methodological rigor of QoL studies, we compared the scores of studies that cited or did not cite these respective articles. Five of the 221 articles (2.3%) that were published in 1995 or later cited the Gill and Feinstein publication, and 18 of 166 articles (10.8%) that were published in 2005 or later cited the Moons et al. paper. The articles that cited Gill and Feinstein [4] had a significantly higher median score (80; interquartile range: 43.75-90) than those that did not cite this paper (0; interquartile range: 0-25) ($U=64.5$; $p<0.001$). The articles that cited the Moons et al. review [7] also showed a significantly higher median score (37.5; interquartile range: 9.38-62.5) than those that did not cite it (0; interquartile range: 0-25) ($U=719.5$; $p=0.001$).

3.3 Temporal trends in measurement methods

During the first period (1974-1994), researchers predominantly used surrogate or proxy measures to assess QoL, such as employment status (38.5%); New York Heart Association (NYHA) classification (23.1%); educational level (23.1%); or symptoms (23.1%) (Table 2).

In 15.4% of the articles, the assessment method was not reported. In the second period (1995-2004), there was a trend toward the use of more comprehensive and dedicated instruments. Nonetheless, the NYHA functional class was still the most common measure (20%). The Medical Outcome Study SF-36 and the Congenital Heart Disease TNO/AZL Adult Quality of Life (CHD-TAAQOL) instruments were used in 9.1% of the studies. Still, in 18.2% of the articles, the assessment was not reported (Table 2). During the third period (2005-2014), we observed a dramatic change in assessment method used. The top 10 most frequently used assessment methods comprised only specific and dedicated QoL tools. The instruments used most frequently during this period were the SF-36 (34.3%), the PedsQL (15.1%), and the linear analogue scale (8.4%). The initially used surrogates, such as NYHA class, employment, or educational status, were not as commonly used in the third period (2005-2014). Another positive effect was a reduction in the percentage of studies that failed to specify how QoL was measured (3.6%) (Table 2).

PLEASE, INSERT TABLE 2 ABOUT HERE

4. Discussion

Over the past decades, QoL research has grown exponentially, both in the general biomedical field [5], and more specifically, in the field of congenital heart disease research. In the latter, QoL research is particularly important because it can be viewed as an indicator of the success of cardiac surgery and medical management of cardiac anomalies over the past half century. Findings of QoL research are most of all informative for afflicted patients or parents, when they have questions about future prospects with regard to their life course. For instance, parents of a newborn child with congenital heart disease want to know more about future consequences in daily life and QoL aspects. Furthermore, after prenatal diagnosis, future parents question the future QoL of their unborn child. Hence, QoL research provides guidance to health-care professionals in their medical decision making and when counseling patients

and parents. Obviously, then, this counsel should be based on the most accurate and current data [8]. In this respect, it is extremely important that clinicians be able to rely on QoL data that is conceptually and methodologically sound.

In the present review, we observed temporal improvements in the conceptual and methodological rigor in QoL studies focusing on patients with congenital heart disease. This is mainly because more articles with higher scores have been published over the past 10 years. Despite this upward trend, the majority of articles still have a low quality score. Indeed, during 2005-2014, 52.4% still failed to meet any of the quality criteria. Criteria that were fulfilled most frequently were "giving a definition of QoL" (criterion 1); "stating the domains of QoL to be measured" (criterion 2); "giving a reason for choosing the instruments they used" (criterion 3); and "aggregating the results from multiple items, domains, or instruments into a single composite score for QoL" (criterion 4). However, only one-quarter to one-third of the studies met these criteria. Criteria that were seldom fulfilled were "asking patients to give their own global rating for QoL" (criterion 5); distinguishing between overall QoL and health-related QoL" (criterion 6); "inviting patients to supplement the items that they considered to be relevant for their QoL" (criterion 7); and "allowing patients to indicate which items were important to them" (criterion 9). Hence, to achieve a better caliber of QoL studies in congenital heart disease, substantial improvement must occur.

We also investigated temporal trends in the measurement methods employed. In this respect, we noted remarkable evolution. Thirty to 40 years ago, researchers only used surrogates of QoL, such as employment status, educational level, or NYHA class. By contrast, contemporary studies mainly use dedicated QoL instruments. Furthermore, the number of articles that did not specify the assessment method decreased dramatically, from 15.4% in 1974-1994 to 3.6% in 2005-2014. This indicates a trend toward a more professional approach to conducting QoL research. This was further corroborated by the observation that increasingly more studies are describing QoL in the methods and results sections, instead of

drawing conclusions about QoL when it was not really even measured. Indeed, the proportion of studies that failed to describe QoL in the methods or results section decreased from 61.5% in 1974-1994 to 11.4% in 2005-2014.

We hypothesized that the papers of Gill and Feinstein [4] and Moons et al. [7] prompted researchers to conduct and report their QoL studies more accurately. Although only a minority of articles reviewed here cited one or both of these papers, it is likely they still had a positive impact. Indeed, the articles that did cite these papers had higher quality scores than the articles that did not cite these papers. Hence, it is reasonable to infer that these researchers learned from these papers. Therefore, the scientific community, including authors, reviewers and editors, should give more attention to conceptual and methodological QoL papers, because they may substantially advance the caliber of QoL studies.

On the basis of the Gill and Feinstein criteria [4] and the findings of the present review, we can propose some recommendations for future QoL research [9]. Following these recommendations would improve the conceptual and methodological rigor of QoL research and expand the knowledge base in this important field of research.

1. Authors are required to provide the definition of QoL they used in their study. This is imperative to make sure that readers understand what the authors mean by the term QoL. In addition, it allows reviewers and readers to check whether QoL is inappropriately interchanged with other related concepts, such as health status, for example [9].
2. Authors are required to explicitly state the domains they measured as components of QoL, since QoL is considered to be a multidimensional or multifactorial construct. The choice of assessment instrument relies on the components included [9].
3. Authors should give the reason(s) for choosing the instruments they used. Valid assessments require that the instruments employed are suitable for the intended task. This ensures that QoL is measured appropriately according to the intended goals [9].
4. Authors are required to state whether they measured overall QoL or health-related QoL.

A clear distinction between overall and health-related QoL should be expressed clearly in QoL papers [9].

5. Authors are required to explicitly state the indicators and determinants of QoL they measured in their study. Investigators need to stipulate how they measured QoL (indicators) and how they assessed influencing factors (determinants). Hence, making a clear distinction between indicators and determinants of QoL is imperative [9] .

The results of the present review should be interpreted in light of some methodological issues. The strengths of this review are that we searched three widely used databases comprising biomedical research (PubMed, Cinahl, and Embase); the search, selection process, and review were independently done by two investigators; and all QoL studies published since 1974 were eligible for inclusion. Hence, we report on 40 years of QoL research in congenital heart disease. However, some methodological limitations need to be addressed as well. First, we used the 10 criteria developed by Gill and Feinstein [4]. To the best of our knowledge, no other list of criteria for assessing the quality of QoL studies is available. If such a list were to exist, it could lead to different results. Second, some of the improvements in quality may have remained "under the radar"; in other words, undetectable using our evaluation methods. Indeed, we acknowledge that some studies that normally would have used the term QoL, alternatively, now have used the term subjective health status or perceived functional status. Hence, the impact of the Gill and Feinstein paper [4] and the Moons et al. paper [7] may be larger than we were able to demonstrate using our approach. Third, the analyses in our review were made at the article level, not at the study or project level. We recognize that some studies or research projects have led to multiple articles. Thus, results of projects may be overrepresented when they originated from research groups that are highly productive in terms of publications. However, we did not have project-level data available for analysis.

5. Conclusions

QoL research in congenital heart disease started some 40 years ago. Since then, numerous QoL studies in this area have been published. The caliber of these studies, however, was unknown up to the present date. Therefore, we were prompted to review all empirical studies published over the past 40 years in order to describe their characteristics in terms of quality and to determine whether temporal trends in conceptual and methodological rigor occurred with increasing experience in this field. We found slight but non-significant temporal improvements in conceptual and methodological rigor and in assessment methods. This indicates a trend toward a more professional approach in conducting and reporting QoL research. Furthermore, we observed that citation of the publications of Gill and Feinstein [4] and Moons et al. [7] in published QoL research is associated with higher quality scores, suggesting that these articles have a positive impact on conceptual and methodological caliber. However, we also determined that major weaknesses in methodological rigor remain present in published QoL research, and the majority of articles have substantial methodological problems. Hence, there is extensive room for improvements. We conclude that the scientific community should give more attention to conceptual and methodological papers dealing with QoL, because they may substantially advance the caliber of QoL studies. When planning future QoL studies, researchers should take the proposed requirements of this review into consideration.

Conflict of interest: None of the authors have any relationship with industry or financial associations that might pose a conflict of interest in connection with the submitted article.

Financial support: None.

References

[1] Ferencz C. The quality of life of the adolescent cardiac patient. Postgraduate medicine. 1974;56:67-73.

[2] Tennant PW, Pearce MS, Bythell M, Rankin J. 20-year survival of children born with congenital anomalies: a population-based study. Lancet. 2010;375:649-56.

[3] Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation. 2010;122:2264-72.

[4] Gill TM, Feinstein AR. A critical appraisal of the quality of quality-of-life measurements. Jama. 1994;272:619-26.

[5] Moons P, Budts W, De Geest S. Critique on the conceptualisation of quality of life: a review and evaluation of different conceptual approaches. Int J Nurs Stud. 2006;43:891-901.

[6] Meeberg GA. Quality of life: a concept analysis. J Adv Nurs. 1993;18:32-8.

[7] Moons P, Van Deyk K, Budts W, De Geest S. Caliber of quality-of-life assessments in congenital heart disease: a plea for more conceptual and methodological rigor. Archives of Pediatrics & Adolescent Medicine. 2004;158:1062-9.

[8] Donofrio MT, Moon-Grady AJ, Hornberger LK, Copel JA, Sklansky MS, Abuhamad A, et al. Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation. 2014;129:2183-242.

[9] Moons P, Jaarsma T, Norekval TM. Requirements for quality-of-life reports. European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2010;9:141-3.

Table 1 Evaluation of Conceptual and Methodological Rigor Over Three Epochs According to Predefined Criteria

Criterion	Gill and Feinstein [4]	1974-1994 (n=13)	1995-2004 (n=55)	2005-2014 (n=166)	Chi^2 (p-value)
1. Did the investigators give a definition of quality of life?	11/75 (15%)	0/13 (0%)	8/55 (15%)	45/166 (27%)	**7.75 (.02)**
2. Did the investigators state the domains they will measure as components of quality of life?	35/75 (47%)	2/13 (15%)	17/55 (31%)	44/166 (27%)	1.34 (.51)
3. Did the investigators give reasons for choosing the instruments they used?	27/75 (36%)	0/13 (0%)	6/55 (11%)	38/166 (23%)	**7.07 (.03)**
4. Did the investigators aggregate results from multiple items, domains, or instruments into a single composite score for quality of life?	27/71 (38%)	1/10 (10%)	8/46 (17%)	53/157 (34%)	**6.48 (.04)**
5. Were patients asked to give their own global rating for quality of life?	13/75 (17%)	0/13 (0%)	2/55 (4%)	15/166 (9%)	2.87 (.24)
6. Was overall quality of life distinguished from health-related quality of life?	0/75 (0%)	0/13 (0%)	0/55 (0%)	9/166 (5%)	3.83 (.15)
7. Were patients invited to supplement the items listed in the instruments offered by the investigators that they considered relevant for their quality of life?	9/71 (13%)	0/9 (0%)	1/43 (2%)	2/157 (1%)	0.40 (.82)
8. If so, were these supplemental items incorporated into the final rating?	8/9 (89%)	0/0 (0%)	1/1 (100%)	2/2 (100%)	NC
9. Were patients allowed to indicate which items were personally important to them?	6/71 (8.5%)	0/9 (0%)	1/43 (2%)	2/157 (1%)	0.40 (.82)
10. If so, were the importance ratings incorporated into the final rating?	3/6 (50%)	0/0 (0%)	1/1 (100%)	2/2 (100%)	NC

NC: Not computable. Bolded Chi^2 values indicate significant difference.

Table 2 Quality of Life Assessments Used in the Different Time Periods

1974-1994		1995-2004		2005-2014	
Instrument	**No. of studies (%) (n=13)**	**Instrument**	**No. of studies (%) (n=55)**	**Instrument**	**No. of studies (%) (n=166)**
Employment	5 (38.5)	NYHA functional class	11 (20)	SF-36	57 (34.3)
NYHA functional class	3 (23.1)	SF-36	5 (9.1)	PedsQL	25 (15.1)
Education	3 (23.1)	CHD-TAAQOL	5 (9.1)	Linear Analog Scale	14 (8.4)
Symptoms	3 (23.1)	Symptoms	5 (9.1)	CHD-TAAQOL	11 (6.6)
Perceived health status (1 item)	2 (15.4)	Pregnancies and childbearing ability	4 (7.3)	WHOQOL-BREF	9 (5.4)
Marital state	2 (15.4)	Employment	4 (7.3)	Satisfaction with Life Scale	9 (5.4)
Pregnancies and childbearing ability	1 (7.7)	Protocol of Kajandi/Lindstrom	4 (7.3)	Child Health Questionnaire	7 (4.2)
Sport and recreation	1 (7.7)	Perceived health status (1 item)	3 (5.5)	TACQOL	6 (3.6)
Insurance	1 (7.7)	Marital state	3 (5.5)	Pediatric Cardiac QOL Inventory	4 (2.4)
Active life	1 (7.7)	Education	3 (5.5)	SF-12	4 (2.4)
Normal unrestricted life	1 (7.7)	Linear Analog Scale	2 (3.6)	KINDL	3 (1.8)
Functional status	1 (7.7)	Child Health Questionnaire	2 (3.6)	Employment	2 (1.2)
Not reported	2 (15.4)	Ability index	2 (3.6)	Education	2 (1.2)
		Resumption of work	2 (3.6)	Ability index	2 (1.2)
		Medication intake	2 (3.6)	MLHFQ	2 (1.2)
		Exercise capacity	2 (3.6)	Camphor	2 (1.2)
		Sport and recreation	1 (1.8)	Kidscreen	2 (1.2)
		Pain	1 (1.8)	EQ-5D	2 (1.2)
		Sickness Impact Profile	1 (1.8)	Ulm Inventory for Children	2 (1.2)
		TACQOL	1 (1.8)	Physical Activity Questionnaire	2 (1.2)
		Satisfaction with Life Scale	1 (1.8)	Sport and recreation	1 (0.6)
		SEIQOL	1 (1.8)	NYHA functional class	1 (0.6)
		Insurance	1 (1.8)	Marital state	1 (0.6)
		Possession of a driver’s license	1 (1.8)	Protocol of Kajandi/Lindstrom	1 (0.6)
		6-minute walk test	1 (1.8)	Resumption of work	1 (0.6)
		Neurological functioning	1 (1.8)	Sickness Impact Profile	1 (0.6)
		Physical fitness	1 (1.8)	Pain	1 (0.6)
		Need for reoperation	1 (1.8)	SF-10	1 (0.6)

Active life	1 (1.8)	Early Childhood Oral Health Impact Scale	1 (0.6)
Health Utility Index	1 (1.8)	MIDAS	1 (0.6)
Psychological profile	1 (1.8)	Hopkins Symptoms Checklist	1 (0.6)
IQLC	1 (1.8)	Family Assessment Device	1 (0.6)
Quality of life questionnaire for chronic lung disease	1 (1.8)	Spanier Dyadic Adjustment Scale	1 (0.6)
Duke questionnaire	1 (1.8)	Physical Self-Perception Profile	1 (0.6)
Limitations to activity	1 (1.8)	ConQol	1 (0.6)
Not reported	10 (18.2)	LQ-KID	1 (0.6)
		SEIQOL	1 (0.6)
		TAPQOL	1 (0.6)
		Not reported	6 (3.6)

NYHA, New York Heart Association; SF-36, Medical Outcome Study Short Form-36; CHD-TAAQOL, Congenital Heart Disease TNO/AZL Adult Quality of Life; TACQOL, TNO/AZL Children's Quality of Life; SEIQOL, Schedule for the Evaluation of Individual Quality of Life; IQLC, Inventory for the Assessment of the Quality of Life in Children and Adolescents; PedsQL, Pediatric Quality of Life Inventory; WHOQOL-BREF, World Health Organization Quality Of Life – Abbreviated version; TACQOL, TNO/AZL Children's Quality of Life; SF-12, Medical Outcome Study Short Form-12; KINDL, Fragebogen für KINDer und Jugendliche zur Erfassung der gesundheitsbezogenen Lebensqualität; MLHFQ, Minnesota Living with Heart Failure Questionnaire; Camphor, Cambridge Pulmonary Hypertension Outcome Review; EQ-5D, EuroQol-5 dimensions; SF-10, Medical Outcome Study Short Form-10; MIDAS, Migraine Disability Assessment; ConQol, Congenital Heart Disease Quality of Life Questionnaire; LQ-KID, Fragebogen zur Lebensqualität bei Kindern und Jugendlichen; TAPQOL, TNO/AZL Preschool children's Quality of Life

Figure Legend

Figure 1 Number of Articles on Quality of Life in Patients with Congenital Heart Disease Published per Year

Figure 2 Heart Defects of Subjects in Quality of Life Studies

Figure 3 Temporal Trends in Conceptual and Methodological Rigor of Quality-of-Life Studies in Patients with Congenital Heart Disease

Figure 1 Number of Articles on Quality of Life in Patients with Congenital Heart Disease Published per Year

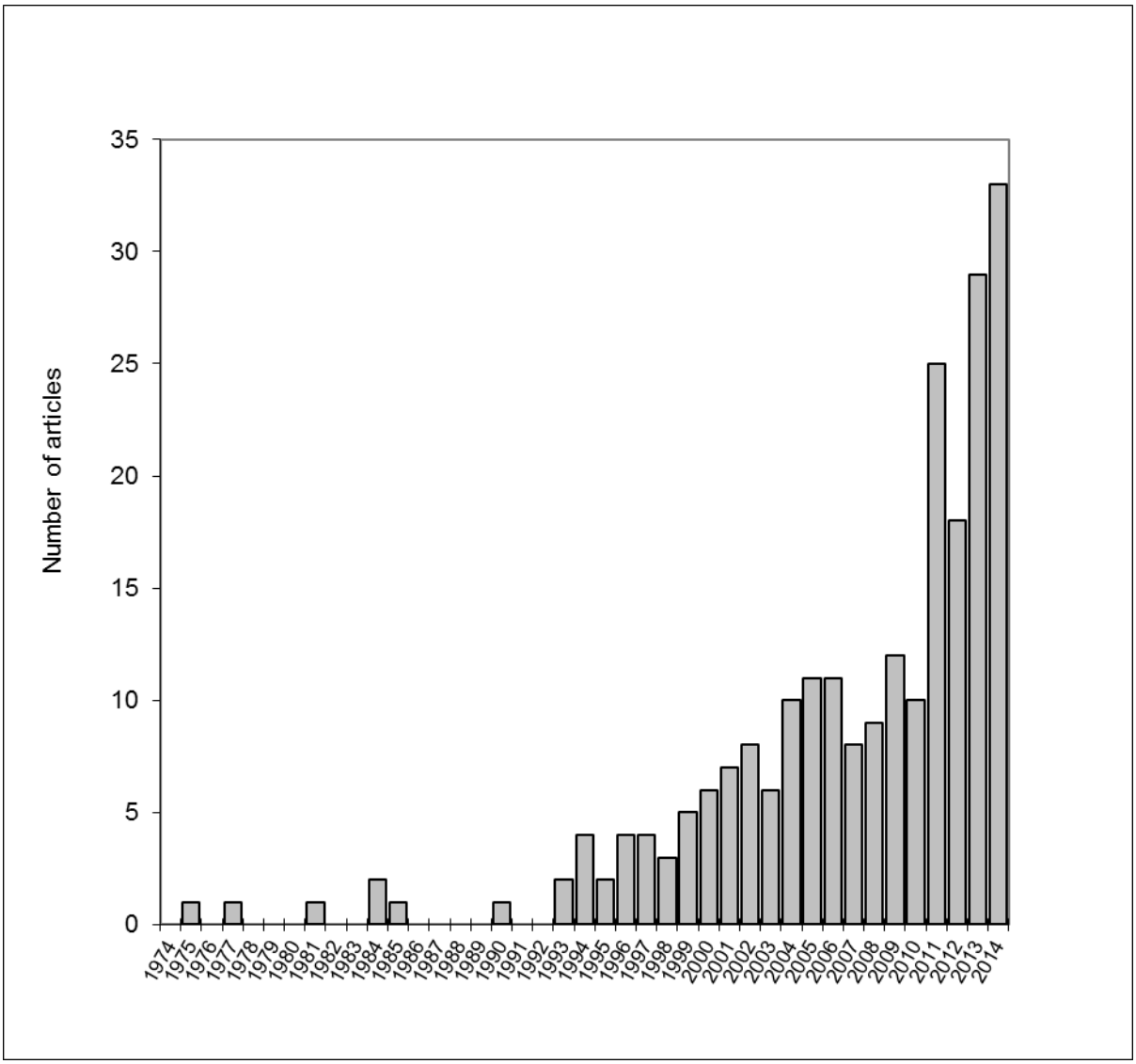
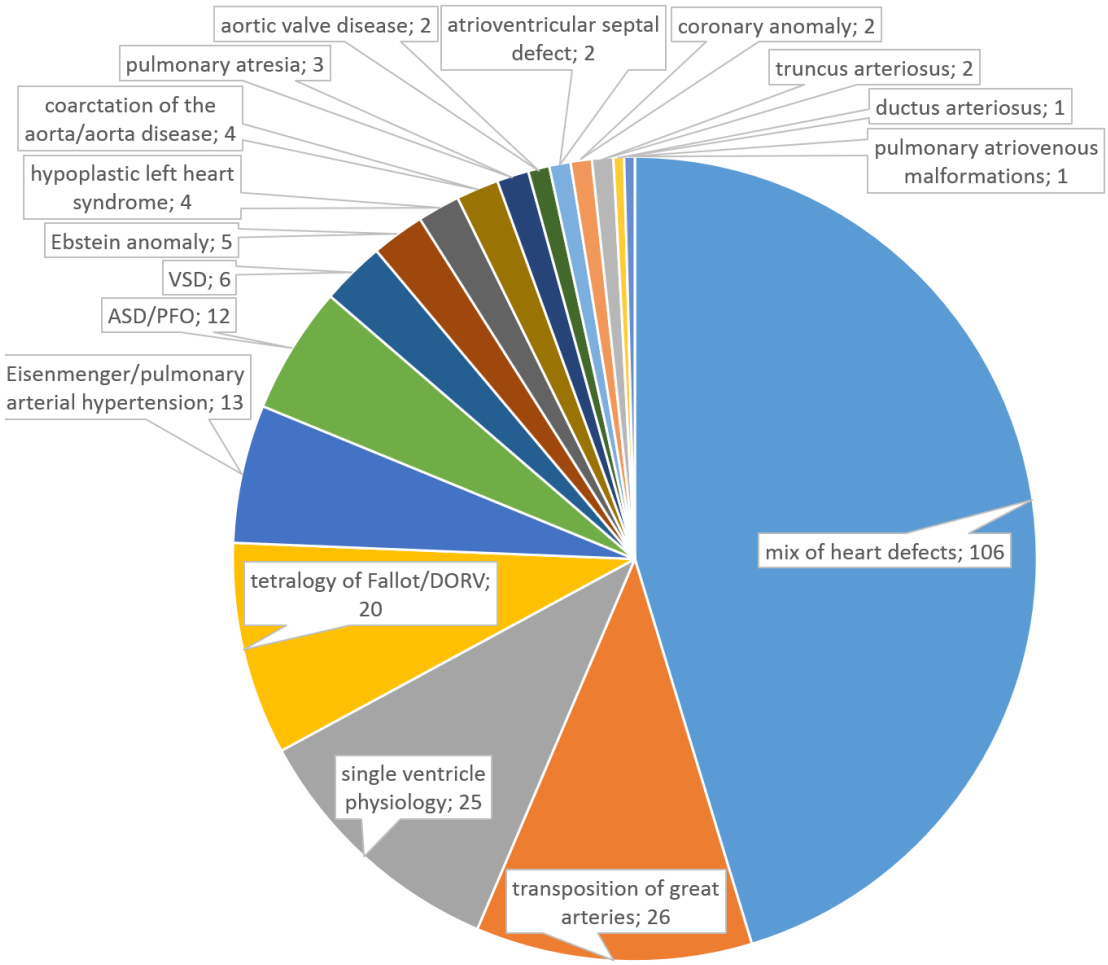
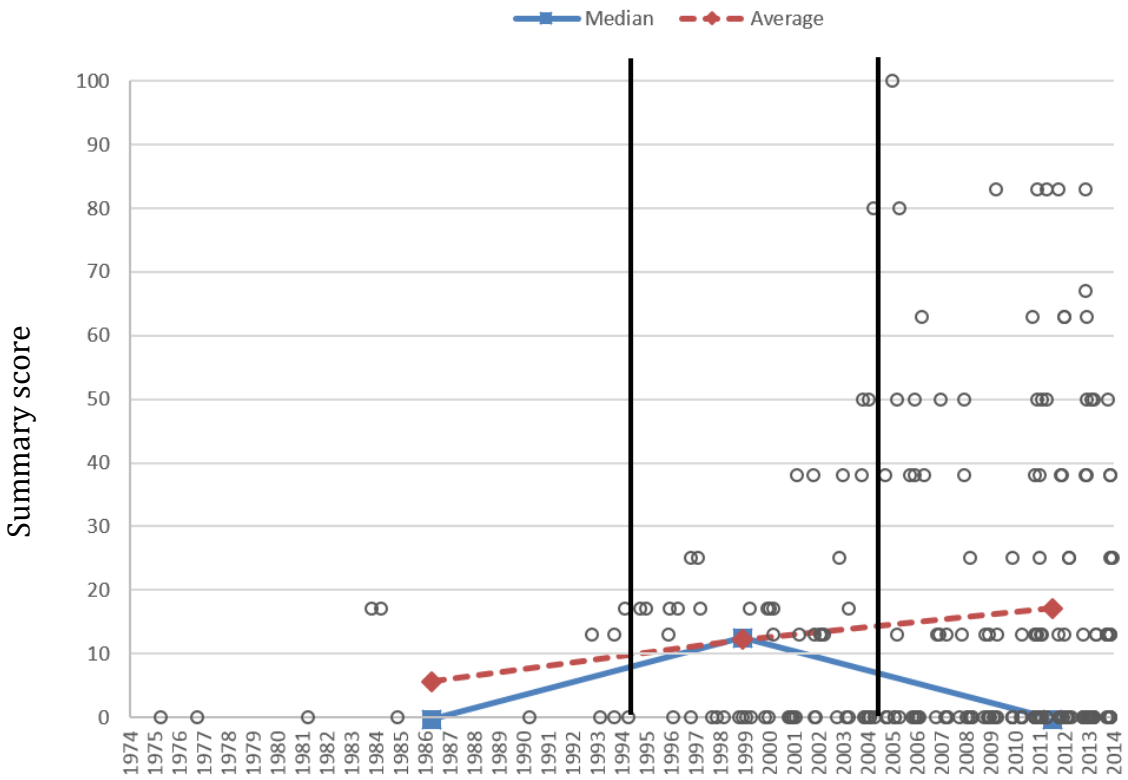


Figure 2 Heart Defects of Subjects in Quality of Life Studies



DORV, double outlet right ventricle; ASD, atrial septal defect; PFO, patent foramen ovale; VSD, ventricular septal defect;

Figure 3 Temporal Trends in Conceptual and Methodological Rigor of Quality-of-Life Studies in Patients with Congenital Heart Disease



Electronic supplemental material

Appendix 1: Search strings employed in the three literature databases.

PubMed

(("quality of life"[All Fields] OR "life quality"[All Fields]) AND ("congenital heart"[All Fields] OR "congenital cardiac"[All Fields] OR "heart defects"[All Fields] OR Fallot[All Fields] OR ("transposition"[All Fields] AND "great arteries"[All Fields]) OR ("aortic coarctation"[MeSH Terms] OR ("aortic"[All Fields] AND "coarctation"[All Fields]) OR "aortic coarctation"[All Fields] OR "coarctation"[All Fields]) OR Eisenmenger[All Fields] OR "septal defect"[All Fields] OR "septal-defects"[All Fields] OR "atrial septal defect"[All Fields] OR "ventricular septal defect"[All Fields] OR "congenital aortic stenosis"[All Fields] OR "congenital pulmonary stenosis"[All Fields] OR univentricular[All Fields] OR "single ventricle"[All Fields] OR "hypoplastic left heart"[All Fields] OR "tricuspid atresia"[All Fields] OR "pulmonary atresia"[All Fields] OR "anomalous pulmonary venous"[All Fields] OR "truncus arteriosus"[All Fields] OR "ductus arteriosus"[All Fields] OR Fontan[All Fields] OR Marfan[All Fields] OR "double outlet"[All Fields] OR "double inlet"[All Fields] OR Ebstein[All Fields] OR “anomalous aortic”[All Fields] OR “anomalous coronary”[All Fields] OR “interrupted aortic”[All Fields] OR "congenital aortic valve"[All Fields] OR "congenital pulmonary valve"[All Fields])) AND ("1900/01/01"[PDAT] : "2014/12/31"[PDAT]) AND English[lang] NOT (Editorial[ptyp] OR Letter[ptyp] OR Case Reports[ptyp] OR Review[ptyp])

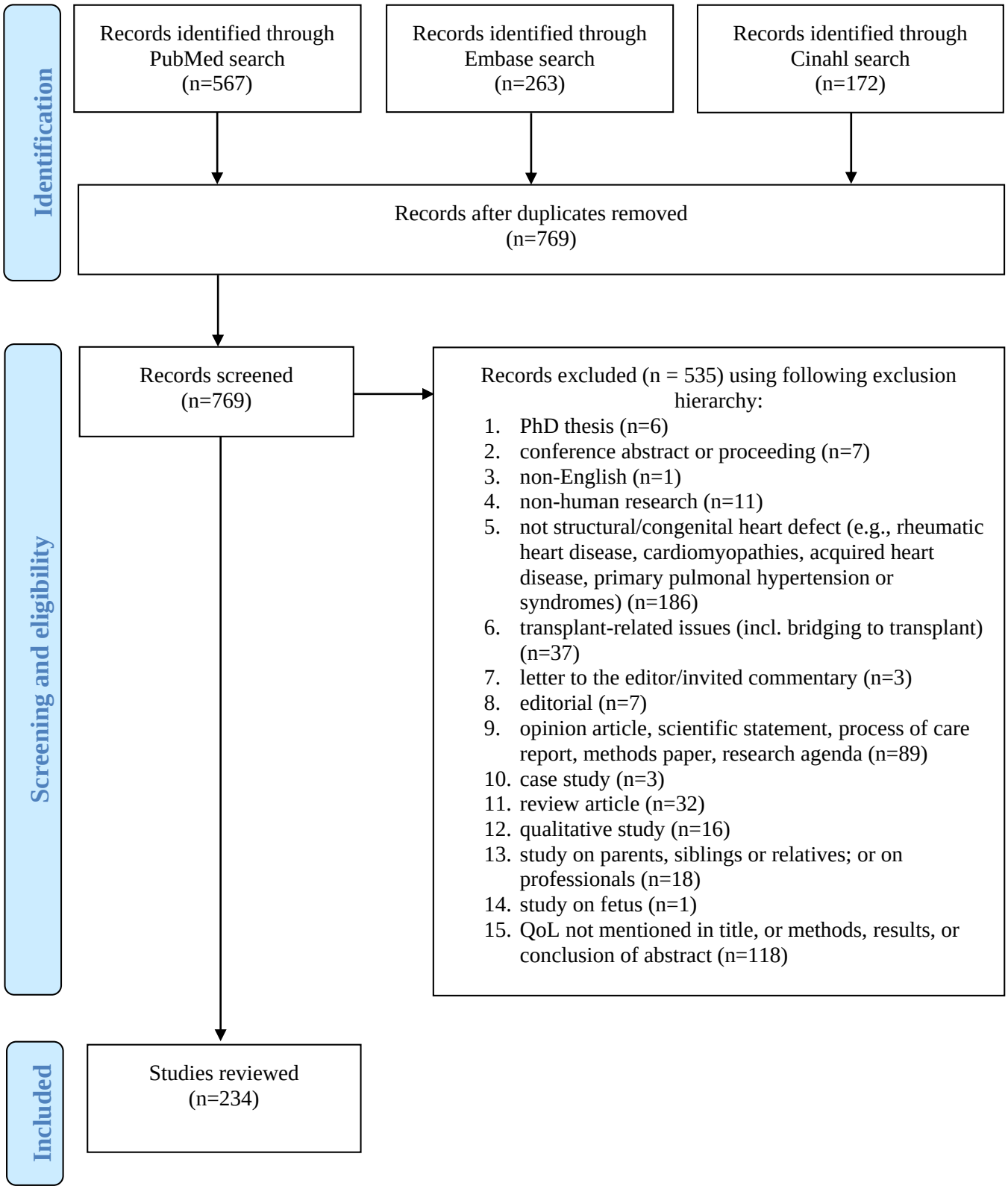
Embase

((('quality of life'/exp OR 'quality of life' OR 'life quality'/exp OR 'life quality') **AND** ('congenital heart disease'/exp OR 'congenital heart' OR 'congenital cardiac' OR heart NEAR/1 defect* OR ‘fallot’ OR transposition NEAR/1 “great arteries” OR aort* NEAR/1 coarct* OR eisenmenger OR septal NEAR/2 defect* OR congenit* NEAR/2 stenos* OR aort* AND near AND stenos* OR univentricul* OR 'single ventricle' OR 'hypoplastic left heart' OR (tricuspid OR pulmonar*) NEAR/1 atresia OR 'pulmonary vein malformation'/exp OR (anomalous AND pulmonary NEAR/1 (vein* OR venous)) OR (ductus OR truncus) NEAR/1 arteriosus OR fontan OR marfan OR double NEAR/1 (inlet* OR outlet*) OR ebstein OR anomalous NEAR/1 (aort* OR coronar*) OR interrupt* NEAR/1 aort* OR congenital NEAR/2 valve*)) NOT (‘conference abstract’/it OR ‘editorial’/it OR 'letter'/it OR 'note'/it OR 'review'/it OR 'case report'/de OR ‘erratum’/it OR ‘conference review’/it)) AND [english]/lim

Cinahl

(MH "Quality of Life") OR 'quality of life' OR 'life quality')
AND
((MH "Heart Defects, Congenital+") OR 'congenital heart' OR ‘congenital cardiac’ OR (heart N1 defect*) OR Fallot OR (transposition N1 ‘great arteries’) OR (MH "Aortic Coarctation") OR (aort* N1 coarct*) OR eisenmenger OR septal N2 defect* OR (congenit* N2 stenos*) OR (aort* N1 stenos*) OR univentricul* OR ‘single ventricle’ OR ‘hypoplastic left heart’ OR (tricuspid OR pulmonar*) N1 atresia OR (anomalous AND (pulmonary N1 (vein* OR venous))) OR ((ductus OR truncus) N1 arteriosus OR Fontan OR Marfan OR (double N1 (inlet* OR outlet*)) OR Ebstein OR (anomalous N1 (aort* OR coronar*)) OR (interrupt* N1 aort*) OR (congenital N2 valve*))
NOT
(letter OR editorial OR review OR case study OR Doctoral Dissertations OR CEU) [PT Publication Type]
LIMITERS
English language [LA Language]

Appendix 2: Flow diagram of article selection



Appendix 3: Review criteria developed by Gill and Feinstein [1] and refined by Moons et al. [2]

1. Did the investigators give a definition of quality of life?
Because a uniform definition of quality of life presently does not exist, investigators need to clarify their conceptualization of quality of life to ensure that readers have a good understanding of the term, as they define it. Therefore, investigators must provide an *explicit* definition of quality of life that serves as a basis for selecting the instruments to be used in their study. Simply referring to the wide variety of definitions, describing the components of quality of life, or citing multiple definitions without unequivocally quoting the definition underpinning the measurement is inadequate.

2. Did the investigators state the domains they will measure as components of quality of life?
Quality of life is typically considered to be a multidimensional construct, comprising multiple domains. The choice of quality-of-life instrument(s) basically relies on the components included in the instrument(s). To determine whether the selected measurement appropriately represents the desired target domain, investigators ought to stipulate explicitly which domains they consider to be significant constituents of quality of life. Just describing domains underlying a specific questionnaire is not sufficient.

3. Did the investigators give reasons for choosing the instruments they used?
Valid assessments require that the instruments used are suitable for the intended task. Since numerous quality-of-life instruments exist, investigators need to state their reasons for choosing to use a particular instrument or instruments to assess quality of life. These reasons should ensure that quality of life will be measured appropriately according to their intended goals. Just because an instrument has good psychometric properties or is widely used does not mean suitable reasons were considered for its use.

4. Did the investigators aggregate results from multiple items, domains, or instruments into a single composite score for quality of life?
It is reasonable to conclude that an effective characterization of quality of life is enhanced if investigators present a composite score that summarizes the results of multiple items, domains, or instruments. Although the richness of a profile description may be lost, an aggregated score simplifies the communication of results and permits the assessment of interrelationships between quality of life and other variables.

5. Were patients asked to give their own global rating for quality of life?
Although quality of life is principally conceptualized as a multidimensional construct, a single global rating by the patient is useful. Patients' rating of their quality of life on a 1-item scale reflects the disparate values and preferences of individual patients. Hence, such a rating serves as an overall estimate of quality of life that considers quality-of-life components deemed important by the respondent.

6. Was overall quality of life distinguished from health-related quality of life?
Health-care professionals are predominantly interested in health-related factors to be components of patients' quality of life. However, a holistic approach implies that also nonmedical phenomena emerge as being important, such as family relationships, social networks, spirituality, pet ownership, etc. Consequently, a distinction between overall and health-related quality of life should be made clear in quality-of-life articles. In this review, we considered this criterion to be fulfilled if the authors explicitly stated the difference between overall and health-related quality of life.

7. Were patients invited to supplement the items listed in the instruments offered by the investigators that they considered relevant for their quality of life?
Since there is a growing awareness that quality of life can only be affected by components that are important for an individual, an adequate measurement of quality of life should provide the possibility for respondents to indicate the domains that they judge to be important for their quality of life. Some argue that this approach is the only way that can lead to a valid measurement of quality of life, because

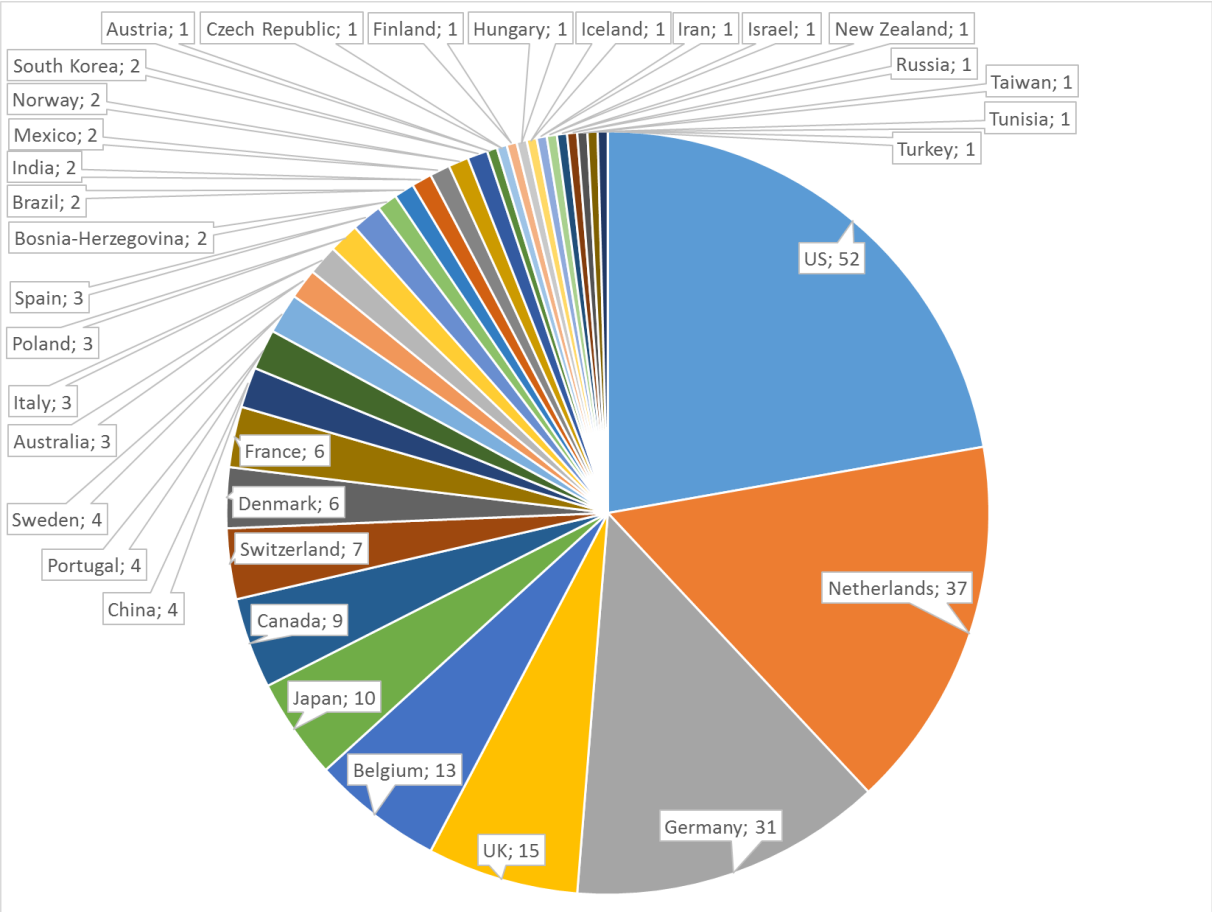
it explicitly includes the domains that are relevant for respondents.

8. If item 7 was satisfied in the research, were these supplemental items provided by the respondent incorporated into the final rating?
To take the supplemental items into consideration in the assessment of quality of life, they ought to be incorporated into the final rating. If there is no possibility of obtaining supplemental items, this criterion is not applicable.

9. Were patients allowed to indicate which items were personally important to them?
Patients need to have the opportunity to rate the importance of different items, either those specified by the investigators or those added by the patients. This offers the possibility to individually weigh the items and guards against the assumption that all items have the same importance for all patients.

10. If item 9 was satisfied in the research, were the importance ratings incorporated into the final rating?
As for the case of the supplemental items in review criterion 7, the importance rating should be incorporated into the final rating. Again, if there is no possibility of scoring the importance of different items, this criterion is not applicable.

Appendix 4. Countries where the studies were performed



Appendix 5: Caliber of quality of life studies in congenital heart disease

Year published	First author	Conceptual definition of QoL	Domains of QoL defined	Reason for choosing measurement	Score aggregated into a single index	Patient's rating on overall QoL	Distinction between overall and health-related QoL	Patient could supplement items	These items were incorporated in final rating	Patients could rate personal importance of items	Importance rate was incorporated into final rating	Summary score
1975	Taussig[3]	0	0	0	0	0	0	0	n/a	0	n/a	0
1977	Hallidie-Smith [4]	0	0	0	0	0	0	0	n/a	0	n/a	0
1981	Arciniegas [5]	0	0	0	0	0	0	0	n/a	0	n/a	0
1984	Raj-Behl [6]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Vergesslich [7]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
1985	Ishizawa [8]	0	0	0	0	0	0	0	n/a	0	n/a	0
1990	Lozano[9]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	0
1993	Gersony[10]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Miyamura [11]	0	0	0	0	0	0	0	n/a	0	n/a	0
1994	Casey [12]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Kuribayashi [13]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Meijboom [14]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Park [15]	0	1	0	0	0	0	0	n/a	0	n/a	13
1995	Benatar [16]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Meijboom [17]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
1996	Meijboom [18]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Miyamura [19]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Shibata [20]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Shyamkrishnan [21]	0	0	0	0	0	0	n/a	n/a	n/a	n/a	0
1997	Elkins [22]	0	0	0	0	0	0	n/a	n/a	n/a	n/a	0
	Moyen Laane [23]	0	1	0	1	0	0	0	n/a	0	n/a	25
	Moyen Laane [24]	0	1	0	1	0	0	0	n/a	0	n/a	25
	Schreiber [25]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
1998	Belli E [26]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Daliento [27]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Wilson [28]	0	0	0	0	0	0	0	n/a	0	n/a	0
1999	Belli [29]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Breymann [30]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Dittrich [31]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Kupilik [32]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Rosenzweig [33]	0	0	0	0	0	0	0	n/a	0	n/a	0
2000	Aeba [34]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Haas [35]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Hucin [36]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Leonard [37]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Mahle [38]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Williams [39]	0	1	0	0	0	0	0	n/a	0	n/a	13
2001	Fekkes [40]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Mair [41]	0	0	0	0	0	0	0	n/a	0	n/a	0

	Saliba [42]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Sandoval [43]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Sugimoto [44]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Ternestedt [45]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Yozu [46]	0	0	0	0	0	0	0	n/a	0	n/a	0
2002	Hovels-Gurich [47]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Kamphuis [48]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Kamphuis [49]	1	0	0	0	0	0	0	n/a	0	n/a	13
	Lane [50]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Mair [51]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Rietveld [52]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Walker [53]	0	1	0	0	0	0	0	n/a	0	n/a	13
	Yamashita [54]	0	0	0	0	0	0	0	n/a	0	n/a	0
2003	Burkhart [55]	0	0	0	1	0	0	n/a	n/a	n/a	n/a	17
	Carriço [56]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Culbert [57]	0	1	1	0	0	0	0	n/a	0	n/a	25
	Dearani [58]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Krol [59]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Simko [60]	1	1	0	1	0	0	0	n/a	0	n/a	38
2004	Burkhart [61]	0	0	0	0	0	0	0	n/a	0	n/a	0
	De Giovanni [62]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Ekman-Joelsson [63]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Jefferies [64]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Kamphuis [65]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Moons [66]	1	1	0	1	1	0	1	1	1	1	80
	Moons [67]	1	1	1	0	1	0	0	n/a	0	n/a	50
	Morgan [68]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Stephensen [69]	0	0	0	0	0	0	0	n/a	0	n/a	0
	van den Bosch [70]	0	0	0	0	0	0	0	n/a	0	n/a	0
2005	Daliento [71]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Dos [72]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Goldbeck [73]	1	1	0	1	0	0	0	n/a	0	n/a	38
	Hager [74]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Immer [75]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Irtel [76]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Moons [77]	1	1	1	1	1	1	1	1	1	1	100
	Moons [78]	1	1	0	1	0	1	1	1	1	1	80
	Rose [79]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Sairanen [80]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Simko [81]	1	1	1	1	0	0	0	n/a	0	n/a	50
2006	Andersen [82]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Boston [83]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Goldbeck [84]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Ibrahim [85]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Macran [86]	0	1	1	1	0	0	0	n/a	0	n/a	38
	Majnemer [87]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Moons [88]	1	1	1	1	1	0	0	n/a	0	n/a	63
	Reiss [89]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Schmid [90]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Spijkerboer [91]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Stulak [92]	0	0	0	0	0	0	0	n/a	0	n/a	0

2007	Bisoi [93]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Bol Raap [94]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Brosig [95]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Bruto [96]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Ebenroth [97]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Hövels-Gürich [98]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Kimmelstiel [99]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Mellander [100]	1	1	1	1	0	0	0	n/a	0	n/a	50
2008	de Koning [101]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Hoffmann [102]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Karsdorp [103]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Landolt [104]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Li [105]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Marino [106]	0	0	1	1	0	0	0	n/a	0	n/a	25
	Pilla [107]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Simons [108]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Winter [109]	0	0	0	0	0	0	0	n/a	0	n/a	0
2009	Brothers [110]	0	0	0	1	0	0	0	n/a	0	n/a	13
	da Fonseca [111]	0	0	0	0	0	0	0	n/a	0	n/a	0
	d'Udekem [112]	0	0	1	0	0	0	0	n/a	0	n/a	13
	Duffels [113]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Duffels [114]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Gratz [115]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Loup [116]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Moons [117]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83
	Müller [118]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Ovaert [119]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Vandekerckhove [120]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Vigl [121]	0	0	0	0	0	0	0	n/a	0	n/a	0
2010	Berkes [122]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Bouallal [123]	0	0	0	0	0	0	n/a	n/a	n/a	n/a	0
	Cohen [124]	1	1	0	0	0	0	0	n/a	0	n/a	25
	Dua [125]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Khan [126]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Lei [127]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Lu [128]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Martinez-Quintana [129]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Tahirovic [130]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Winter [131]	0	0	0	0	0	0	0	n/a	0	n/a	0
2011	Backer [132]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Chen [133]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Friesen [134]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Gaies [135]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Gierat-Haponiuk [136]	0	0	0	1	1	0	0	n/a	0	n/a	25
	Goldbeck [137]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Görler [138]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Hanninen [139]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Kwon [140]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Lemmer [141]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Luyckx [142]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83
	Luyckx [143]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83

	Ma [144]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Mokhles [145]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Müller [146]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Müller [147]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Neuner [148]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Overgaard [149]	1	1	1	1	1	0	0	n/a	0	n/a	63
	Silva [150]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Tahirovic [151]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Tay [152]	1	0	0	1	0	1	0	n/a	0	n/a	38
	Tay [153]	1	0	0	1	0	1	0	n/a	0	n/a	38
	Teixeira [154]	0	1	1	1	0	1	0	n/a	0	n/a	50
	Vigl [155]	1	1	1	0	0	1	0	n/a	0	n/a	50
	Yurlov [156]	0	0	0	0	0	0	0	n/a	0	n/a	0
2012	Angeli [157]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Bygstad [158]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Costello [159]	1	1	0	0	0	0	0	n/a	0	n/a	25
	Cotts [160]	1	1	1	1	1	0	0	n/a	0	n/a	63
	Czosek [161]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Ehlert [162]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Huang [163]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Knowles [164]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Lu [165]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Luyckx [166]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83
	Müller [167]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Müller [168]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Opic [169]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Pike [170]	1	1	1	1	0	1	0	n/a	0	n/a	63
	Riley [171]	1	1	0	0	0	0	0	n/a	0	n/a	25
	Schoormans [172]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Sharma [173]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Winter [174]	0	0	0	0	0	0	0	n/a	0	n/a	0
2013	Apers [175]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83
	Atz [176]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Bang [177]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Becker-Grünig [178]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Bolio-Cerdan [179]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Cha [180]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Cuypers [181]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Eagleson [182]	1	1	1	1	0	1	0	n/a	0	n/a	63
	Eslami [183]	1	1	1	0	1	0	0	n/a	0	n/a	50
	Garcia Guerra [184]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Heiberg [185]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Idorn [186]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Kahya Eren [187]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Lakhdhar [188]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Lu [189]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Mueller [190]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Müller [191]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Müller [192]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Müller [193]	0	0	0	0	0	0	0	n/a	0	n/a	0
	O'Byrne [194]	1	1	1	1	0	0	0	n/a	0	n/a	50

	Rassart [195]	1	1	1	0	1	0	n/a	n/a	n/a	n/a	67
	Ruys [196]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Sandberg [197]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Schaefer [198]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Schuuring [199]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Uzark [200]	1	1	1	0	0	0	0	n/a	0	n/a	38
	van der Bom [201]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Vis [202]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Younge [203]	0	0	0	0	0	0	0	n/a	0	n/a	0
2014	Areias [204]	1	0	0	1	0	0	0	n/a	0	n/a	25
	Bedair [205]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Bertoletti [206]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Brosig [207]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Dean [208]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Dulfer [209]	0	0	0	0	1	0	0	n/a	0	n/a	13
	Dulfer [210]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Frigiola [211]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Garcia-Guerra [212]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Gerelli [213]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Goldberg [214]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Hebert [215]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Heusch [216]	1	0	0	1	0	0	0	n/a	0	n/a	25
	Knowles [217]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Komar [218]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Komar [219]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Luyckx [220]	1	1	1	1	1	0	n/a	n/a	n/a	n/a	83
	Ly [221]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Mellion [222]	1	1	1	1	0	0	0	n/a	0	n/a	50
	Müller [223]	0	1	1	0	0	1	0	n/a	0	n/a	38
	Mussatto [224]	0	0	0	0	0	0	0	n/a	0	n/a	0
	O'Byrne [225]	1	0	0	1	0	0	0	n/a	0	n/a	25
	Opic [226]	0	0	0	1	0	0	0	n/a	0	n/a	13
	Opic[227]	0	0	0	0	1	0	0	n/a	0	n/a	13
	Raju [228]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Schoormans [229]	1	1	0	0	0	0	0	n/a	0	n/a	25
	Schoormans [230]	1	1	1	0	0	0	0	n/a	0	n/a	38
	Sistino [231]	0	0	0	0	0	0	0	n/a	0	n/a	0
	Wang [232]	0	1	0	1	0	0	0	n/a	0	n/a	25
	Werner [233]	1	1	0	1	0	0	0	n/a	0	n/a	38
	Wittlieb-Weber [234]	0	0	0	1	0	0	0	n/a	0	n/a	13
2015	Neal [235]	0	0	0	0	0	0	0	n/a	0	n/a	0
	van der Bom [236]	0	0	0	0	0	0	0	n/a	0	n/a	0

* 1974-1994 1995-2003 2004- 2014

References

[1] Gill TM, Feinstein AR. A critical appraisal of the quality of quality-of-life measurements. Jama. 1994;272:619-26.

[2] Moons P, Van Deyk K, Budts W, De Geest S. Caliber of quality-of-life assessments in congenital heart disease: a plea for more conceptual and methodological rigor. Archives of Pediatrics & Adolescent Medicine. 2004;158:1062-9.

[3] Taussig HB, Kallman CH, Nagel D, Baumgardner R, Momberger N, Kirk H. Long-time observations on the Blalock-Taussig operation VIII. 20 to 28 year follow-up on patients with a tetralogy of Fallot. The Johns Hopkins medical journal. 1975;137:13-9.

[4] Hallidie-Smith KA, Wilson RS, Hart A, Zeidifard E. Functional status of patients with large ventricular septal defect and pulmonary vascular disease 6 to 16 years after surgical closure of their defect in childhood. British heart journal. 1977;39:1093-101.

[5] Arciniegas E, Farooki ZQ, Hakimi M, Perry BL, Green EW. Results of the Mustard operation for dextro-transposition of the great arteries. The Journal of thoracic and cardiovascular surgery. 1981;81:580-7.

[6] Raj Behl P, Blesovsky A. Ebstein's anomaly: sixteen years' experience with valve replacement without plication of the right ventricle. Thorax. 1984;39:8-13.

[7] Vergesslich KA, Gersony WM, Steeg CN, Hordof AJ, Bowman FO, Malm JR, et al. Postoperative Assessment of Porcine-Valved Right Ventricular Pulmonary-Artery Conduits. American Journal of Cardiology. 1984;53:202-5.

[8] Ishizawa E, Tadokoro M, Satoh S, Suzuki Y, Haneda K, Kagawa Y, et al. Mustard procedure for simple transposition of the great arteries. The Tohoku journal of experimental medicine. 1985;145:91-6.

[9] Lozano C, Rovirosa M, Reig J, Salva JA. Surgery of atrioventricular septal defects. Review of the first 100 cases. Eur J Cardiothorac Surg. 1990;4:359-64.

[10] Gersony WM, Hayes CJ, Driscoll DJ, Keane JF, Kidd L, O'Fallon WM, et al. Second natural history study of congenital heart defects. Quality of life of patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. Circulation. 1993;87:I52-65.
[11] Miyamura H, Eguchi S, Asano K. Long-term results of the intracardiac repair of tetralogy of Fallot: a follow-up study conducted over more than 20 years on 100 consecutive operative survivors. Surgery today. 1993;23:1049-52.
[12] Casey FA, Craig BG, Mulholland HC. Quality of life in surgically palliated complex congenital heart disease. Arch Dis Child. 1994;70:382-6.
[13] Kuribayashi R, Sekine S, Aida H, Seki K, Meguro A, Shibata Y, et al. Long-term results of primary closure for ventricular septal defects in the first year of life. Surgery today. 1994;24:389-92.
[14] Meijboom F, Szatmari A, Utens E, Deckers JW, Roelandt JR, Bos E, et al. Long-term follow-up after surgical closure of ventricular septal defect in infancy and childhood. Journal of the American College of Cardiology. 1994;24:1358-64.
[15] Park I, Nakazawa M, Imai Y, Sawatari K, Momma K. Prediction of quality of life at long-term follow-up after Fontan operation by scoring risk factors. Jpn Circ J. 1994;58:646-52.
[16] Benatar A, Tanke R, Roef M, Meyboom EJ, Van de Wal HJ. Mid-term results of the modified Senning operation for cavopulmonary connection with autologous tissue. Eur J Cardiothorac Surg. 1995;9:320-4.
[17] Meijboom F, Szatmari A, Deckers JW, Utens EM, Roelandt JR, Bos E, et al. Cardiac status and health-related quality of life in the long term after surgical repair of tetralogy of Fallot in infancy and childhood. The Journal of thoracic and cardiovascular surgery. 1995;110:883-91.

[18] Meijboom F, Szatmari A, Deckers JW, Utens EM, Roelandt JR, Bos E, et al. Long-term follow-up (10 to 17 years) after Mustard repair for transposition of the great arteries. The Journal of thoracic and cardiovascular surgery. 1996;111:1158-68.

[19] Miyamura H, Takahashi M, Sugawara M, Eguchi S. The long-term influence of pulmonary valve regurgitation following repair of tetralogy of Fallot: does preservation of the pulmonary valve ring affect quality of life? Surgery today. 1996;26:603-6.

[20] Shibata Y, Abe T, Kuribayashi R, Sekine S, Seki K, Yamagishi I, et al. Surgical treatment of isolated secundum atrial septal defect in patients more than 50 years old. The Annals of thoracic surgery. 1996;62:1096-9.

[21] Shyamkrishnan KG, Singh M, Tharakan JM, Dal A. A ten-year post-surgical assessment of pulmonary hypertension in adults with patent ductus arteriosus. Indian Heart J. 1996;48:249-51.

[22] Elkins RC, Knott-Craig CJ, McCue C, Lane MM. Congenital aortic valve disease. Improved survival and quality of life. Ann Surg. 1997;225:503-10; discussion 10-1.

[23] Moyen Laane K, Meberg A, Otterstad JE, Froland G, Sorland S, Lindstrom B, et al. Quality of life in children with congenital heart defects. Acta paediatrica (Oslo, Norway : 1992). 1997;86:975-80.

[24] Laane KM, Meberg A, Otterstad JE, Froland G, Lindstrom B, Eriksson B. Does an early neonatal diagnosis of a later spontaneously closed ventricular septal defect impair quality of life? Scandinavian cardiovascular journal : SCJ. 1997;31:213-6.

[25] Schreiber C, Mazzitelli D, Haehnel JC, Lorenz HP, Meisner H. The interrupted aortic arch: an overview after 20 years of surgical treatment. Eur J Cardiothorac Surg. 1997;12:466-9; discussion 9-70.

[26] Belli E, Serraf A, Lacour-Gayet F, Prodan S, Piot D, Losay J, et al. Biventricular repair for double-outlet right ventricle. Results and long-term follow-up. Circulation. 1998;98:II360-5; discussion II5-7.

[27] Daliento L, Somerville J, Presbitero P, Menti L, Brach-Prever S, Rizzoli G, et al. Eisenmenger syndrome. Factors relating to deterioration and death. Eur Heart J. 1998;19:1845-55.

[28] Wilson NJ, Clarkson PM, Barratt-Boyes BG, Calder AL, Whitlock RM, Easthope RN, et al. Long-term outcome after the mustard repair for simple transposition of the great arteries. 28-year follow-up. J Am Coll Cardiol. 1998;32:758-65.

[29] Belli E, Lacour-Gayet F, Serraf A, Alkhulaifi AM, Touchot A, Bruniaux J, et al. Surgical management of transposition of great arteries associated with multiple ventricular septal defects. Eur J Cardiothorac Surg. 1999;16:14-20.

[30] Breymann T, Kirchner G, Blanz U, Cherlet E, Knobl H, Meyer H, et al. Results after Norwood procedure and subsequent cavopulmonary anastomoses for typical hypoplastic left heart syndrome and similar complex cardiovascular malformations. Eur J Cardiothorac Surg. 1999;16:117-24.

[31] Dittrich S, Vogel M, Dahnert I, Berger F, Alexi-Meskishvili V, Lange PE. Surgical repair of tetralogy of Fallot in adults today. Clinical cardiology. 1999;22:460-4.

[32] Kupilik N, Simon P, Moidl R, Wollenek G, Marx M, Wolner E, et al. Valve-preserving treatment of Ebstein's anomaly: perioperative and follow-up results. The Thoracic and cardiovascular surgeon. 1999;47:229-34.

[33] Rosenzweig EB, Kerstein D, Barst RJ. Long-term prostacyclin for pulmonary hypertension with associated congenital heart defects. Circulation. 1999;99:1858-65.

[34] Aeba R, Katogi T, Takeuchi S, Kawada S. Outcome of patients with cyanotic congenital heart disease undergoing a second systemic-to-pulmonary artery shunt. The Journal of cardiovascular surgery. 2000;41:23-30.
[35] Haas GS, Hess H, Black M, Onnasch J, Mohr FW, van Son JA. Extracardiac conduit fontan procedure: early and intermediate results. Eur J Cardiothorac Surg. 2000;17:648-54.
[36] Hucin B, Voriskova M, Hruda J, Marek J, Janousek J, Reich O, et al. Late complications and quality of life after atrial correction of transposition of the great arteries in 12 to 18 year follow-up. The Journal of cardiovascular surgery. 2000;41:233-9.
[37] Leonard H, Derrick G, O'Sullivan J, Wren C. Natural and unnatural history of pulmonary atresia. Heart. 2000;84:499-503.
[38] Mahle WT, Clancy RR, Moss EM, Gerdes M, Jobes DR, Wernovsky G. Neurodevelopmental outcome and lifestyle assessment in school-aged and adolescent children with hypoplastic left heart syndrome. Pediatrics. 2000;105:1082-9.
[39] Williams DL, Gelijns AC, Moskowitz AJ, Weinberg AD, Ng JH, Crawford E, et al. Hypoplastic left heart syndrome: valuing the survival. The Journal of thoracic and cardiovascular surgery. 2000;119:720-31.
[40] Fekkes M, Kamphuis RP, Ottenkamp J, Verrips E, Vogels T, Kamphuis M, et al. Health-related quality of life in young adults with minor congenital heart disease. Psychology & Health. 2001;16:239-50.
[41] Mair DD, Puga FJ, Danielson GK. The Fontan procedure for tricuspid atresia: Early and late results of a 25-year experience with 216 patients. Journal of the American College of Cardiology. 2001;37:933-9.
[42] Saliba Z, Butera G, Bonnet D, Bonhoeffer P, Villain E, Kachaner J, et al. Quality of life and perceived health status in surviving adults with univentricular heart. Heart. 2001;86:69-73.

[43] Sandoval J, Aguirre JS, Pulido T, Martinez-Guerra ML, Santos E, Alvarado P, et al. Nocturnal oxygen therapy in patients with the Eisenmenger syndrome. Am J Respir Crit Care Med. 2001;164:1682-7.

[44] Sugimoto S, Takagi N, Hachiro Y, Abe T. High frequency of arrhythmias after Fontan operation indicates earlier anticoagulant therapy. Int J Cardiol. 2001;78:33-9.

[45] Ternestedt BM, 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. Pediatr Cardiol. 2001;22:128-32.

[46] Yozu R, Shin H, Maehara T, Iino Y, Mitsumaru A, Kawada S. Port-access cardiac surgery. Experience with 34 cases at Keio University Hospital. The Japanese journal of thoracic and cardiovascular surgery : official publication of the Japanese Association for Thoracic Surgery = Nihon Kyobu Geka Gakkai zasshi. 2001;49:360-4.

[47] Hovels-Gurich HH, Konrad K, Wiesner M, Minkenberg R, Herpertz-Dahlmann B, Messmer BJ, et al. Long term behavioural outcome after neonatal arterial switch operation for transposition of the great arteries. Arch Dis Child. 2002;87:506-10.

[48] Kamphuis M, Ottenkamp J, Vliegen HW, Vogels T, Zwinderman KH, Kamphuis RP, et al. Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. Heart. 2002;87:356-62.

[49] Kamphuis M, Vliegen HW, Vogels T, Zwinderman KH, van der Wall EE, Verloove-Vanhorick SP, et al. The need for cardiac follow-up in adults with mild congenital cardiac disease. Cardiol Young. 2002;12:474-8.

[50] Lane DA, Lip GY, Millane TA. Quality of life in adults with congenital heart disease. Heart. 2002;88:71-5.

[51] Mair DD, Sim EK, Danielson GK, Puga FJ. Long-term follow-up of surgically corrected patients with common arterial trunk. Progress in Pediatric Cardiology. 2002;15:65-71.

[52] Rietveld S, Mulder BJ, van Beest I, Lubbers W, Prins PJ, Vioen S, et al. Negative thoughts in adults with congenital heart disease. Int J Cardiol. 2002;86:19-26.
[53] Walker WT, Temple IK, Gnanapragasam JP, Goddard JR, Brown EM. Quality of life after repair of tetralogy of Fallot. Cardiol Young. 2002;12:549-53.
[54] Yamashita K, Kazui T, Terada H, Washiyama N, Suzuki T, Ohkura K, et al. Total and subtotal aortic replacement for extensive aortic dissection in patients with or without Marfan's syndrome. The Japanese journal of thoracic and cardiovascular surgery : official publication of the Japanese Association for Thoracic Surgery = Nihon Kyobu Geka Gakkai zasshi. 2002;50:315-20.
[55] Burkhart HM, Dearani JA, Mair DD, Warnes CA, Rowland CC, Schaff HV, et al. The modified Fontan procedure: early and late results in 132 adult patients. The Journal of thoracic and cardiovascular surgery. 2003;125:1252-9.
[56] Carrico A, Moura C, Monterroso J, Casanova J, de Sousa R, Bastos P, et al. Patients with aortic coarctation operated during the first year of life, different surgical techniques and prognostic factors--21 years of experience. Revista portuguesa de cardiologia : orgao oficial da Sociedade Portuguesa de Cardiologia = Portuguese journal of cardiology : an official journal of the Portuguese Society of Cardiology. 2003;22:1185-93.
[57] Culbert EL, Ashburn DA, Cullen-Dean G, Joseph JA, Williams WG, Blackstone EH, et al. Quality of life of children after repair of transposition of the great arteries. Circulation. 2003;108:857-62.
[58] Dearani JA, Danielson GK, Puga FJ, Schaff HV, Warnes CW, Driscoll DJ, et al. Late follow-up of 1095 patients undergoing operation for complex congenital heart disease utilizing pulmonary ventricle to pulmonary artery conduits. The Annals of thoracic surgery. 2003;75:399-410; discussion -1.

[59] Krol Y, Grootenhuis MA, Destrée-Vonk A, Lubbers LJ, Koopman HM, Last BF. Health related quality of life in children with congenital heart disease. Psychology & Health. 2003;18:251-60.
[60] Simko LC, McGinnis KA. Quality of life experienced by adults with congenital heart disease. AACN clinical issues. 2003;14:42-53.
[61] Burkhart HM, Dearani JA, Williams WG, Puga FJ, Mair DD, Ashburn DA, et al. Late results of palliative atrial switch for transposition, ventricular septal defect, and pulmonary vascular obstructive disease. The Annals of thoracic surgery. 2004;77:464-8; discussion 8-9.
[62] De Giovanni JV. Timing, frequency, and results of catheter intervention following recruitment of major aortopulmonary collaterals in patients with pulmonary atresia and ventricular septal defect. J Interv Cardiol. 2004;17:47-52.
[63] Ekman-Joelsson BM, Berntsson L, Sunnegardh J. Quality of life in children with pulmonary atresia and intact ventricular septum. Cardiol Young. 2004;14:615-21.
[64] Jefferies JL, Noonan JA, Keller BB, Wilson JF, Griffith C, 3rd. Quality of life and social outcomes in adults with congenital heart disease living in rural areas of Kentucky. The American journal of cardiology. 2004;94:263-6.
[65] Kamphuis M, Zwinderman KH, Vogels T, Vliegen HW, Kamphuis RP, Ottenkamp J, 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 : an international journal of quality of life aspects of treatment, care and rehabilitation. 2004;13:735-45.
[66] Moons P, Marquet K, Budts W, De Geest S. Validity, reliability and responsiveness of the "Schedule for the Evaluation of Individual Quality of Life-Direct Weighting" (SEIQoL-DW) in congenital heart disease. Health and quality of life outcomes. 2004;2:27.

[67] Moons P, De Bleser L, Budts W, Sluysmans T, De Wolf D, Massin M, et al. Health status, functional abilities, and quality of life after the Mustard or Senning operation. The Annals of thoracic surgery. 2004;77:1359-65; discussion 65.
[68] Morgan JA, Peacock JC, Kohmoto T, Garrido MJ, Schanzer BM, Kherani AR, et al. Robotic techniques improve quality of life in patients undergoing atrial septal defect repair. The Annals of thoracic surgery. 2004;77:1328-33.
[69] Stephensen SS, Sigfusson G, Eiriksson H, Sverrisson JT, Torfason B, Haraldsson A, et al. Congenital cardiac malformations in Iceland from 1990 through 1999. Cardiol Young. 2004;14:396-401.
[70] van den Bosch AE, Roos-Hesselink JW, Van Domburg R, Bogers AJ, Simoons ML, Meijboom FJ. Long-term outcome and quality of life in adult patients after the Fontan operation. The American journal of cardiology. 2004;93:1141-5.
[71] Daliento L, Mapelli D, Russo G, Scarso P, Limongi F, Iannizzi P, et al. Health related quality of life in adults with repaired tetralogy of Fallot: psychosocial and cognitive outcomes. Heart. 2005;91:213-8.
[72] Dos L, Teruel L, Ferreira IJ, Rodriguez-Larrea J, Miro L, Girona J, et al. Late outcome of Senning and Mustard procedures for correction of transposition of the great arteries. Heart. 2005;91:652-6.
[73] Goldbeck L, Melches J. Quality of life in families of children with congenital heart disease. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2005;14:1915-24.
[74] Hager A, Hess J. Comparison of health related quality of life with cardiopulmonary exercise testing in adolescents and adults with congenital heart disease. Heart. 2005;91:517-20.

[75] Immer FF, Althaus SM, Berdat PA, Saner H, Carrel TP. Quality of life and specific problems after cardiac surgery in adolescents and adults with congenital heart diseases. European journal of cardiovascular prevention and rehabilitation : official journal of the European Society of Cardiology, Working Groups on Epidemiology & Prevention and Cardiac Rehabilitation and Exercise Physiology. 2005;12:138-43.

[76] Irtel TA, Vetter C, Stuber T, Kuemin A, Heimes T, Pfammater JP, et al. Impact of arrhythmias on health-related quality of life in adults with congenital cardiac disease. Cardiol Young. 2005;15:627-31.

[77] Moons P, Van Deyk K, De Geest S, Gewillig M, Budts W. Is the severity of congenital heart disease associated with the quality of life and perceived health of adult patients? Heart. 2005;91:1193-8.

[78] Moons P, Van Deyk K, Marquet K, Raes E, De Bleser L, Budts W, et al. Individual quality of life in adults with congenital heart disease: a paradigm shift. Eur Heart J. 2005;26:298-307.

[79] Rose M, Kohler K, Kohler F, Sawitzky B, Fliege H, Klapp BF. Determinants of the quality of life of patients with congenital heart disease. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2005;14:35-43.

[80] Sairanen HI, Nieminen HP, Jokinen EV. Late results and quality of life after pediatric cardiac surgery in Finland: a population-based study of 6,461 patients with follow-up extending up to 45 years. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2005:168-72.

[81] Simko LC, McGinnis KA. What is the perceived quality of life of adults with congenital heart disease and does it differ by anomaly? J Cardiovasc Nurs. 2005;20:206-14.

[82] Andersen PE, Kjeldsen AD. Clinical and radiological long-term follow-up after embolization of pulmonary arteriovenous malformations. CardioVascular and Interventional Radiology. 2006;29:70-4.
[83] Boston US, Dearani JA, O'Leary PW, Driscoll DJ, Danielson GK. Tricuspid valve repair for Ebstein's anomaly in young children: a 30-year experience. The Annals of thoracic surgery. 2006;81:690-5; discussion 5-6.
[84] Goldbeck L, Melches J. The impact of the severity of disease and social disadvantage on quality of life in families with congenital cardiac disease. Cardiol Young. 2006;16:67-75.
[85] Ibrahim R, Granton JT, Mehta S. An open-label, multicentre pilot study of bosentan in pulmonary arterial hypertension related to congenital heart disease. Canadian respiratory journal : journal of the Canadian Thoracic Society. 2006;13:415-20.
[86] Macran S, Birks Y, Parsons J, Sloper P, Hardman G, Kind P, et al. The development of a new measure of quality of life for children with congenital cardiac disease. Cardiol Young. 2006;16:165-72.
[87] Majnemer A, Limperopoulos C, Shevell M, Rohlicek C, Rosenblatt B, Tchervenkov C. Health and well-being of children with congenital cardiac malformations, and their families, following open-heart surgery. Cardiol Young. 2006;16:157-64.
[88] Moons P, Van Deyk K, De Bleser L, Marquet K, Raes E, De Geest S, et al. Quality of life and health status in adults with congenital heart disease: a direct comparison with healthy counterparts. European journal of cardiovascular prevention and rehabilitation : official journal of the European Society of Cardiology, Working Groups on Epidemiology & Prevention and Cardiac Rehabilitation and Exercise Physiology. 2006;13:407-13.
[89] Reiss N, Blanz U, Breymann T, Kind K, Bairaktaris A, Korfer R. Mechanical valve replacement of the systemic atrioventricular valve in children. ASAIO journal (American Society for Artificial Internal Organs : 1992). 2006;52:559-61.

[90] Schmid C, Debus V, Gogarten W, Loher A, Drees G, Scheld HH, et al. Pediatric assist with the Medos and Excor systems in small children. ASAIO journal (American Society for Artificial Internal Organs : 1992). 2006;52:505-8.

[91] Spijkerboer AW, Utens EM, De Koning WB, Bogers AJ, Helbing WA, Verhulst FC. Health-related Quality of Life in children and adolescents after invasive treatment for congenital heart disease. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2006;15:663-73.

[92] Stulak JM, Dearani JA, Puga FJ, Zehr KJ, Schaff HV, Danielson GK. Right-sided Maze procedure for atrial tachyarrhythmias in congenital heart disease. The Annals of thoracic surgery. 2006;81:1780-4; discussion 4-5.

[93] Bisoi AK, Murala JS, Airan B, Chowdhury UK, Kothari SS, Pal H, et al. Tetralogy of Fallot in teenagers and adults: surgical experience and follow-up. General thoracic and cardiovascular surgery. 2007;55:105-12.

[94] Bol Raap G, Meijboom FJ, Kappetein AP, Galema TW, Yap SC, Bogers AJ. Long-term follow-up and quality of life after closure of ventricular septal defect in adults. Eur J Cardiothorac Surg. 2007;32:215-9.

[95] Brosig CL, Mussatto KA, Kuhn EM, Tweddell JS. Psychosocial outcomes for preschool children and families after surgery for complex congenital heart disease. Pediatr Cardiol. 2007;28:255-62.

[96] Bruto VC, Harrison DA, Fedak PW, Rockert W, Siu SC. Determinants of health-related quality of life in adults with congenital heart disease. Congenit Heart Dis. 2007;2:301-13.

[97] Ebenroth ES, Hurwitz RA. Long-term functional outcome of patients following the mustard procedure: the next decade of follow-up. Congenit Heart Dis. 2007;2:235-41.

[98] Hovels-Gurich HH, Konrad K, Skorzenski D, Minkenberg R, Herpertz-Dahlmann B, Messmer BJ, et al. Long-term behavior and quality of life after corrective cardiac surgery in infancy for tetralogy of Fallot or ventricular septal defect. Pediatr Cardiol. 2007;28:346-54.
[99] Kimmelstiel C, Gange C, Thaler D. Is patent foramen ovale closure effective in reducing migraine symptoms? A controlled study. Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions. 2007;69:740-6.
[100] Mellander M, Berntsson L, Nilsson B. Quality of life in children with hypoplastic left heart syndrome. Acta paediatrica (Oslo, Norway : 1992). 2007;96:53-7.
[101] de Koning WB, van Osch-Gevers M, Ten Harkel AD, van Domburg RT, Spijkerboer AW, Utens EM, et al. Follow-up outcomes 10 years after arterial switch operation for transposition of the great arteries: comparison of cardiological health status and health-related quality of life to those of the a normal reference population. European journal of pediatrics. 2008;167:995-1004.
[102] Hoffmann A, Wyler F, Gunthard J, Gradel E. Late follow-up of patients who underwent palliation for complex congenital heart disease in childhood. Congenit Heart Dis. 2008;3:155-8.
[103] Karsdorp PA, Kindt M, Rietveld S, Everaerd W, Mulder BJ. Interpretation bias for heart sensations in congenital heart disease and its relation to quality of life. Int J Behav Med. 2008;15:232-40.
[104] Landolt MA, Valsangiacomo Buechel ER, Latal B. Health-related quality of life in children and adolescents after open-heart surgery. The Journal of pediatrics. 2008;152:349-55.
[105] Li J, Liu Y, Yu C, Cui B, Du M. Comparison of incisions and outcomes for closure of ventricular septal defects. The Annals of thoracic surgery. 2008;85:199-203.
[106] Marino BS, Shera D, Wernovsky G, Tomlinson RS, Aguirre A, Gallagher M, et al. The development of the pediatric cardiac quality of life inventory: a quality of life measure for

children and adolescents with heart disease. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2008;17:613-26.
[107] Pilla CB, Pereira CA, Fin AV, Aquino FV, Botta A, DalleMulle L, et al. Health-related quality of life and right ventricular function in the midterm follow-up assessment after tetralogy of fallot repair. Pediatr Cardiol. 2008;29:409-15.
[108] Simons LE, Gilleland J, McDanel AH, Blount RL, Campbell R. Initial development of the Pediatric Camp Outcome Measure. Children's Health Care. 2008;37:158-69.
[109] Winter MM, Bouma BJ, van Dijk AP, Groenink M, Nieuwkerk PT, van der Plas MN, et al. Relation of physical activity, cardiac function, exercise capacity, and quality of life in patients with a systemic right ventricle. The American journal of cardiology. 2008;102:1258-62.
[110] Brothers JA, McBride MG, Marino BS, Tomlinson RS, Seliem MA, Pampaloni MH, et al. Exercise performance and quality of life following surgical repair of anomalous aortic origin of a coronary artery in the pediatric population. The Journal of thoracic and cardiovascular surgery. 2009;137:380-4.
[111] da Fonseca MA, Evans M, Teske D, Thikkurissy S, Amini H. The impact of oral health on the quality of life of young patients with congenital cardiac disease. Cardiol Young. 2009;19:252-6.
[112] d'Udekem Y, Cheung MM, Setyapranata S, Iyengar AJ, Kelly P, Buckland N, et al. How good is a good Fontan? Quality of life and exercise capacity of Fontans without arrhythmias. The Annals of thoracic surgery. 2009;88:1961-9.
[113] Duffels MG, Vis JC, van Loon RL, Nieuwkerk PT, van Dijk AP, Hoendermis ES, et al. Effect of bosentan on exercise capacity and quality of life in adults with pulmonary arterial hypertension associated with congenital heart disease with and without Down's syndrome. The American journal of cardiology. 2009;103:1309-15.

[114] Duffels MG, Vis JC, van Loon RL, Berger RM, Hoendermis ES, van Dijk AP, et al. Down patients with Eisenmenger syndrome: is bosentan treatment an option? Int J Cardiol. 2009;134:378-83.

[115] Gratz A, Hess J, Hager A. Self-estimated physical functioning poorly predicts actual exercise capacity in adolescents and adults with congenital heart disease. Eur Heart J. 2009;30:497-504.

[116] Loup O, von Weissenfluh C, Gahl B, Schwerzmann M, Carrel T, Kadner A. Quality of life of grown-up congenital heart disease patients after congenital cardiac surgery. Eur J Cardiothorac Surg. 2009;36:105-11; discussion 11.

[117] Moons P, Van Deyk K, Marquet K, De Bleser L, De Geest S, Budts W. Profile of adults with congenital heart disease having a good, moderate, or poor quality of life: a cluster analytic study. European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2009;8:151-7.

[118] Muller J, Christov F, Schreiber C, Hess J, Hager A. Exercise capacity, quality of life, and daily activity in the long-term follow-up of patients with univentricular heart and total cavopulmonary connection. Eur Heart J. 2009;30:2915-20.

[119] Ovaert C, Thijs D, Dewolf D, Ottenkamp J, Dessy H, Moons P, et al. The effect of bosentan in patients with a failing Fontan circulation. Cardiol Young. 2009;19:331-9.

[120] Vandekerckhove KD, Blom NA, Lalezari S, Koolbergen DR, Rijlaarsdam ME, Hazekamp MG. Long-term follow-up of arterial switch operation with an emphasis on function and dimensions of left ventricle and aorta. Eur J Cardiothorac Surg. 2009;35:582-7; discussion 7-8.

[121] Vigl M, Hager A, Bauer U, Niggemeyer E, Wittstock B, Kohn FM, et al. Sexuality and subjective wellbeing in male patients with congenital heart disease. Heart. 2009;95:1179-83.

[122] Berkes A, Varni JW, Pataki I, Kardos L, Kemeny C, Mogyorosy G. Measuring health-related quality of life in Hungarian children attending a cardiology clinic with the Pediatric Quality of Life Inventory. European journal of pediatrics. 2010;169:333-47.
[123] Bouallal R, Godart F, Francart C, Richard A, Foucher-Hossein C, Lions C. Interest of beta-blockers in patients with right ventricular systemic dysfunction. Cardiol Young. 2010;20:615-9.
[124] Cohen M, Daniela M, Yalonetsky S, Gagin R, Lorber A. Psychological functioning and health-related quality of life (HRQoL) in older patients following percutaneous closure of the secundum atrial septal defect (ASD). Archives of gerontology and geriatrics. 2010;50:e5-8.
[125] Dua JS, Cooper AR, Fox KR, Graham Stuart A. Exercise training in adults with congenital heart disease: feasibility and benefits. Int J Cardiol. 2010;138:196-205.
[126] Khan AA, Tan JL, Li W, Dimopoulos K, Spence MS, Chow P, et al. The impact of transcatheter atrial septal defect closure in the older population: a prospective study. JACC Cardiovascular interventions. 2010;3:276-81.
[127] Lei BF, Chen JM, Cen JZ, Lui RC, Ding YQ, Xu G, et al. Palliative arterial switch for transposition of the great arteries, ventricular septal defect, and pulmonary vascular obstructive disease: midterm outcomes. The Journal of thoracic and cardiovascular surgery. 2010;140:845-9.
[128] Lu JC, Cotts TB, Agarwal PP, Attili AK, Dorfman AL. Relation of right ventricular dilation, age of repair, and restrictive right ventricular physiology with patient-reported quality of life in adolescents and adults with repaired tetralogy of fallot. The American journal of cardiology. 2010;106:1798-802.
[129] Martinez-Quintana E, Miranda-Calderin G, Ugarte-Lopetegui A, Rodriguez-Gonzalez F. Rehabilitation program in adult congenital heart disease patients with pulmonary hypertension. Congenit Heart Dis. 2010;5:44-50.

[130] Tahirovic E, Begic H, Nurkic M, Tahirovic H, Varni JW. Does the severity of congenital heart defects affect disease-specific health-related quality of life in children in Bosnia and Herzegovina? European journal of pediatrics. 2010;169:349-53.
[131] Winter MM, Reisma C, Kedde H, Bouma BJ, Vis JC, Luijendijk P, et al. Sexuality in adult patients with congenital heart disease and their partners. The American journal of cardiology. 2010;106:1163-8, 8.e1-8.
[132] Backer CL. 12th Annual C. Walton Lillehei Memorial Lecture in Cardiovascular Surgery: Fontan conversion--the Chicago experience. Cardiol Young. 2011;21 Suppl 2:169-76.
[133] Chen CA, Liao SC, Wang JK, Chang CI, Chiu IS, Chen YS, et al. Quality of life in adults with congenital heart disease: biopsychosocial determinants and sex-related differences. Heart. 2011;97:38-43.
[134] Friesen CL, Robertson M, Liu D, Burton H, Fleming K, Jackson S, et al. Follow-up of tetralogy of Fallot patients: tertiary centre versus satellite clinic. Cardiol Young. 2011;21:444-53.
[135] Gaies MG, Watnick CS, Gurney JG, Bove EL, Goldberg CS. Health-related quality of life in patients with congenitally corrected transposition of the great arteries. The Journal of thoracic and cardiovascular surgery. 2011;142:136-41.
[136] Gierat-Haponiuk K, Haponiuk I, Chojnicki M, Jaworski R, Bakula S. Exercise capacity and the quality of life late after surgical correction of congenital heart defects. Kardiologia polska. 2011;69:810-5.
[137] Goldbeck L, Holling I, Schlack R, West C, Besier T. The impact of an inpatient family-oriented rehabilitation program on parent-reported psychological symptoms of chronically ill children. Klin Padiatr. 2011;223:79-84.

[138] Gorler H, Ono M, Thies A, Lunkewitz E, Westhoff-Bleck M, Haverich A, et al. Long-term morbidity and quality of life after surgical repair of transposition of the great arteries: atrial versus arterial switch operation. Interact Cardiovasc Thorac Surg. 2011;12:569-74.
[139] Hanninen M, Kmet A, Taylor DA, Ross DB, Rebeyka I, Vonder Muhll IF. Atrial septal defect closure in the elderly is associated with excellent quality of life, functional improvement, and ventricular remodelling. The Canadian journal of cardiology. 2011;27:698-704.
[140] Kwon EN, Mussatto K, Simpson PM, Brosig C, Nugent M, Samyn MM. Children and adolescents with repaired tetralogy of fallot report quality of life similar to healthy peers. Congenit Heart Dis. 2011;6:18-27.
[141] Lemmer J, Heise G, Rentzsch A, Boettler P, Kuehne T, Dubowy KO, et al. Right ventricular function in grown-up patients after correction of congenital right heart disease. Clinical research in cardiology : official journal of the German Cardiac Society. 2011;100:289-96.
[142] Luyckx K, Goossens E, Missotten L, Moons P. Adolescents with congenital heart disease: the importance of perceived parenting for psychosocial and health outcomes. Journal of developmental and behavioral pediatrics : JDBP. 2011;32:651-9.
[143] Luyckx K, Goossens E, Van Damme C, Moons P. Identity formation in adolescents with congenital cardiac disease: a forgotten issue in the transition to adulthood. Cardiol Young. 2011;21:411-20.
[144] Ma ZS, Yin QY, Dong MF, Feng ZY, Wang LX. Quality of life in patients undergoing totally thoracoscopic closure for atrial septal defect. The Annals of thoracic surgery. 2011;92:2230-4.
[145] Mokhles MM, van de Woestijne PC, de Jong PL, Witsenburg M, Roos-Hesselink JW, Takkenberg JJ, et al. Clinical outcome and health-related quality of life after right-ventricular-

outflow-tract reconstruction with an allograft conduit. Eur J Cardiothorac Surg. 2011;40:571-8.

[146] Muller J, Kuhn A, Vogt M, Schreiber C, Hess J, Hager A. Improvements in exercise performance after surgery for Ebstein anomaly. The Journal of thoracic and cardiovascular surgery. 2011;141:1192-5.

[147] Muller J, Hess J, Hager A. Exercise performance and quality of life is more impaired in Eisenmenger syndrome than in complex cyanotic congenital heart disease with pulmonary stenosis. Int J Cardiol. 2011;150:177-81.

[148] Neuner B, Busch MA, Singer S, Moons P, Wellmann J, Bauer U, et al. Sense of coherence as a predictor of quality of life in adolescents with congenital heart defects: a register-based 1-year follow-up study. Journal of developmental and behavioral pediatrics : JDBP. 2011;32:316-27.

[149] Overgaard D, Schrader AM, Lisby KH, King C, Christensen RF, Jensen HF, et al. Patient-reported outcomes in adult survivors with single-ventricle physiology. Cardiology. 2011;120:36-42.

[150] Silva AM, Vaz C, Areias ME, Vieira D, Proenca C, Viana V, et al. Quality of life of patients with congenital heart diseases. Cardiol Young. 2011;21:670-6.

[151] Tahirovic E, Begic H, Tahirovic H, Varni JW. Quality of life in children after cardiac surgery for congenital heart disease. Collegium antropologicum. 2011;35:1285-90.

[152] Tay EL, Papaphylactou M, Diller GP, Alonso-Gonzalez R, Inuzuka R, Giannakoulas G, et al. Quality of life and functional capacity can be improved in patients with Eisenmenger syndrome with oral sildenafil therapy. Int J Cardiol. 2011;149:372-6.

[153] Tay EL, Peset A, Papaphylactou M, Inuzuka R, Alonso-Gonzalez R, Giannakoulas G, et al. Replacement therapy for iron deficiency improves exercise capacity and quality of life in

patients with cyanotic congenital heart disease and/or the Eisenmenger syndrome. Int J Cardiol. 2011;151:307-12.

[154] Teixeira FM, Coelho RM, Proenca C, Silva AM, Vieira D, Vaz C, et al. Quality of life experienced by adolescents and young adults with congenital heart disease. Pediatr Cardiol. 2011;32:1132-8.

[155] Vigl M, Niggemeyer E, Hager A, Schwedler G, Kropf S, Bauer U. The importance of socio-demographic factors for the quality of life of adults with congenital heart disease. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2011;20:169-77.

[156] Yurlov IA, Podzolkov VP, Zelenikin MM, Kovalev DV, Babaev GK, Putiato NA, et al. Experience with bidirectional cavopulmonary anastomosis and modified Fontan operation in patients with single ventricle and concomitant visceral heterotaxy. Interact Cardiovasc Thorac Surg. 2011;12:563-8.

[157] Angeli E, Pace Napoleone C, Balducci A, Formigari R, Lovato L, Candini L, et al. Natural and modified history of single-ventricle physiology in adult patients. Eur J Cardiothorac Surg. 2012;42:996-1002.

[158] Bygstad E, Pedersen LC, Pedersen TA, Hjortdal VE. Tetralogy of Fallot in men: quality of life, family, education, and employment. Cardiol Young. 2012;22:417-23.

[159] Costello JM, O'Brien M, Wypij D, Shubert J, Salvin JW, Newburger JW, et al. Quality of life of pediatric cardiac patients who previously required extracorporeal membrane oxygenation. Pediatric critical care medicine : a journal of the Society of Critical Care Medicine and the World Federation of Pediatric Intensive and Critical Care Societies. 2012;13:428-34.

[160] Cotts T, Malviya S, Goldberg C. Quality of life and perceived health status in adults with congenitally corrected transposition of the great arteries. The Journal of thoracic and cardiovascular surgery. 2012;143:885-90.
[161] Czosek RJ, Bonney WJ, Cassedy A, Mah DY, Tanel RE, Imundo JR, et al. Impact of cardiac devices on the quality of life in pediatric patients. Circulation Arrhythmia and electrophysiology. 2012;5:1064-72.
[162] Ehlert N, Hess J, Hager A. Shifts in exercise capacity are not reported adequately in patients with congenital heart disease. Congenit Heart Dis. 2012;7:448-54.
[163] Huang JB, Cao YT, Liang J, Lv XD. Quality of life after children undergo a radical arterial switch operation at an older age. The heart surgery forum. 2012;15:E103-7.
[164] Knowles R, Veldtman G, Hickey EJ, Bradley T, Gengsakul A, Webb GD, et al. Functional health status of adults with tetralogy of Fallot: matched comparison with healthy siblings. The Annals of thoracic surgery. 2012;94:124-32.
[165] Lu JC, Cotts TB, Dorfman AL. Diastolic function and patient-reported quality of life for adolescents and adults with repaired tetralogy of Fallot: a tissue Doppler study. Pediatr Cardiol. 2012;33:618-24.
[166] Luyckx K, Missotten L, Goossens E, Moons P. Individual and contextual determinants of quality of life in adolescents with congenital heart disease. The Journal of adolescent health : official publication of the Society for Adolescent Medicine. 2012;51:122-8.
[167] Muller J, Hess J, Hager A. Daily physical activity in adults with congenital heart disease is positively correlated with exercise capacity but not with quality of life. Clinical research in cardiology : official journal of the German Cardiac Society. 2012;101:55-61.
[168] Muller J, Hess J, Hager A. Minor symptoms of depression in patients with congenital heart disease have a larger impact on quality of life than limited exercise capacity. Int J Cardiol. 2012;154:265-9.

[169] Opic P, Utens EM, Moons P, Theuns DA, van Dijk AP, Hoendermis ES, et al. Psychosocial impact of implantable cardioverter defibrillators (ICD) in young adults with Tetralogy of Fallot. Clinical research in cardiology : official journal of the German Cardiac Society. 2012;101:509-19.

[170] Pike NA, Evangelista LS, Doering LV, Eastwood JA, Lewis AB, Child JS. Quality of life, health status, and depression: comparison between adolescents and adults after the Fontan procedure with healthy counterparts. J Cardiovasc Nurs. 2012;27:539-46.

[171] 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. J Adv Nurs. 2012;68:1050-60.

[172] Schoormans D, Mulder BJ, van Melle JP, Pieper EG, van Dijk AP, Sieswerda GJ, et al. Patients with a congenital heart defect and type D personality feel functionally more impaired, report a poorer health status and quality of life, but use less healthcare. European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2012;11:349-55.

[173] Sharma V, Burkhart HM, Cetta F, Hagler DJ, Phillips SD, Dearani JA. Fontan conversion to one and one half ventricle repair. The Annals of thoracic surgery. 2012;94:1269-74; discussion 74.

[174] Winter MM, van der Bom T, de Vries LC, Balducci A, Bouma BJ, Pieper PG, et al. Exercise training improves exercise capacity in adult patients with a systemic right ventricle: a randomized clinical trial. Eur Heart J. 2012;33:1378-85.

[175] Apers S, Moons P, Goossens E, Luyckx K, Gewillig M, Bogaerts K, et al. Sense of coherence and perceived physical health explain the better quality of life in adolescents with congenital heart disease. European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2013;12:475-83.

[176] Atz AM, Travison TG, McCrindle BW, Mahony L, Glatz AC, Kaza AK, et al. Cardiac performance and quality of life in patients who have undergone the Fontan procedure with and without prior superior cavopulmonary connection. Cardiol Young. 2013;23:335-43.
[177] Bang JS, Jo S, Kim GB, Kwon BS, Bae EJ, Noh CI, et al. The mental health and quality of life of adult patients with congenital heart disease. Int J Cardiol. 2013;170:49-53.
[178] Becker-Grunig T, Klose H, Ehlken N, Lichtblau M, Nagel C, Fischer C, et al. Efficacy of exercise training in pulmonary arterial hypertension associated with congenital heart disease. Int J Cardiol. 2013;168:375-81.
[179] Bolio-Cerdan A, Ruiz-Gonzalez S, Romero-Cardenas P, Hernandez-Morales G, Villasis-Keever MA. Prognosis of children with cardiomyopathy submitted to fontan surgery: 30 years of experience at the Hospital Infantil de Mexico Federico Gomez. Boletin Medico del Hospital Infantil de Mexico. 2013;70:151-8.
[180] Cha KS, Cho KI, Seo JS, Choi JH, Park YH, Yang DH, et al. Effects of inhaled iloprost on exercise capacity, quality of life, and cardiac function in patients with pulmonary arterial hypertension secondary to congenital heart disease (the Eisenmenger syndrome) (from the EIGER Study). The American journal of cardiology. 2013;112:1834-9.
[181] Cuypers JA, Opic P, Menting ME, Utens EM, Witsenburg M, Helbing WA, et al. The unnatural history of an atrial septal defect: longitudinal 35 year follow up after surgical closure at young age. Heart. 2013;99:1346-52.
[182] Eagleson KJ, Justo RN, Ware RS, Johnson SG, Boyle FM. Health-related quality of life and congenital heart disease in Australia. J Paediatr Child Health. 2013;49:856-64.
[183] Eslami B, Macassa G, Sundin O, Khankeh HR, Soares JJ. Quality of life and life satisfaction among adults with and without congenital heart disease in a developing country. Eur J Prev Cardiol. 2013.

[184] Garcia Guerra G, Robertson CM, Alton GY, Joffe AR, Dinu IA, Nicholas D, et al. Quality of life 4 years after complex heart surgery in infancy. The Journal of thoracic and cardiovascular surgery. 2013;145:482-8.e2.

[185] Heiberg J, Laustsen S, Petersen AK, Hjortdal VE. Reduced long-term exercise capacity in young adults operated for ventricular septal defect. Cardiol Young. 2013:1-7.

[186] Idorn L, Jensen AS, Juul K, Overgaard D, Nielsen NP, Sorensen K, et al. Quality of life and cognitive function in Fontan patients, a population-based study. Int J Cardiol. 2013;168:3230-5.

[187] Kahya Eren N, Kirdok AH, Kilicaslan B, Kocabas U, Duzel B, Berilgen R, et al. Quality of life of patients with atrial septal defect following percutaneous closure. Cardiol Young. 2015;25:42-6.

[188] Lakhdhar R, Drissa M, Drissa H. Natural history of atrial septal defect in the sixth decade : study of 5 cases. La Tunisie medicale. 2013;91:243-7.

[189] Lu JC, Ghadimi Mahani M, Agarwal PP, Cotts TB, Dorfman AL. Usefulness of right ventricular free wall strain to predict quality of life in "repaired" tetralogy of Fallot. The American journal of cardiology. 2013;111:1644-9.

[190] Mueller GC, Sarikouch S, Beerbaum P, Hager A, Dubowy KO, Peters B, et al. Health-related quality of life compared with cardiopulmonary exercise testing at the midterm follow-up visit after tetralogy of Fallot repair: a study of the German competence network for congenital heart defects. Pediatr Cardiol. 2013;34:1081-7.

[191] Muller J, Hess J, Hager A. Sense of coherence, rather than exercise capacity, is the stronger predictor to obtain health-related quality of life in adults with congenital heart disease. Eur J Prev Cardiol. 2013;21:949-55.

[192] Muller J, Hess J, Horer J, Hager A. Persistent superior exercise performance and quality of life long-term after arterial switch operation compared to that after atrial redirection. Int J Cardiol. 2013;166:381-4.
[193] Muller J, Hess J, Hager A. General anxiety of adolescents and adults with congenital heart disease is comparable with that in healthy controls. Int J Cardiol. 2013;165:142-5.
[194] O'Byrne ML, Mercer-Rosa L, Zhao H, Zhang X, Yang W, Cassedy A, et al. Morbidity in children and adolescents after surgical correction of truncus arteriosus communis. Am Heart J. 2013;166:512-8.
[195] Rassart J, Luyckx K, Goossens E, Apers S, Klimstra TA, Moons P. Personality traits, quality of life and perceived health in adolescents with congenital heart disease. Psychol Health. 2013;28:319-35.
[196] Ruys TP, van der Bosch AE, Cuypers JA, Witsenburg M, Helbing WA, Bogers AJ, et al. Long-term outcome and quality of life after arterial switch operation: a prospective study with a historical comparison. Congenit Heart Dis. 2013;8:203-10.
[197] Sandberg C, Engstrom KG, Dellborg M, Thilen U, Wadell K, Johansson B. The level of physical exercise is associated with self-reported health status (EQ-5D) in adults with congenital heart disease. Eur J Prev Cardiol. 2013.
[198] Schaefer C, von Rhein M, Knirsch W, Huber R, Natalucci G, Caflisch J, et al. Neurodevelopmental outcome, psychological adjustment, and quality of life in adolescents with congenital heart disease. Developmental medicine and child neurology. 2013;55:1143-9.
[199] Schuuring MJ, Vis JC, van Dijk AP, van Melle JP, Vliegen HW, Pieper PG, et al. Impact of bosentan on exercise capacity in adults after the Fontan procedure: a randomized controlled trial. Eur J Heart Fail. 2013;15:690-8.

[200] Uzark K, King E, Spicer R, Beekman R, Kimball T, Varni JW. The clinical utility of health-related quality of life assessment in pediatric cardiology outpatient practice. Congenit Heart Dis. 2013;8:211-8.

[201] van der Bom T, Winter MM, Bouma BJ, Groenink M, Vliegen HW, Pieper PG, et al. Effect of valsartan on systemic right ventricular function: a double-blind, randomized, placebo-controlled pilot trial. Circulation. 2013;127:322-30.

[202] Vis JC, Duffels MG, Mulder P, de Bruin-Bon RH, Bouma BJ, Berger RM, et al. Prolonged beneficial effect of bosentan treatment and 4-year survival rates in adult patients with pulmonary arterial hypertension associated with congenital heart disease. Int J Cardiol. 2013;164:64-9.

[203] Younge JO, Eindhoven JA, Utens EW, Opic P, Cuypers JA, van den Bosch AE, et al. Association between N-terminal pro-brain natriuretic peptide and quality of life in adult patients with congenital heart disease. Cardiol Young. 2013:1-7.

[204] Areias MEG, Pinto CI, Vieira PF, Castro M, Freitas I, Sarmento S, et al. Living with CHD: Quality of life (QOL) in early adult life. Cardiology in the Young. 2014;24:60-5.

[205] Bedair R, Babu-Narayan SV, Dimopoulos K, Quyam S, Doyle A, Swan L, et al. Acceptance and psychological impact of implantable defibrillators amongst adults with congenital heart disease. Int J Cardiol. 2014;181c:218-24.

[206] Bertoletti J, Marx GC, Hattge SP, Pellanda LC. Health-related quality of life in adolescents with congenital heart disease. Cardiol Young. 2014:1-7.

[207] Brosig C, Yang K, Hoffmann RG, Dasgupta M, Mussatto K. The role of psychology in a pediatric outpatient cardiology setting: preliminary results from a new clinical program. Journal of clinical psychology in medical settings. 2014;21:337-46.

[208] Dean PN, Gillespie CW, Greene EA, Pearson GD, Robb AS, Berul CI, et al. Sports Participation and Quality of Life in Adolescents and Young Adults with Congenital Heart Disease. Congenit Heart Dis. 2014.

[209] Dulfer K, Duppen N, Kuipers IM, Schokking M, van Domburg RT, Verhulst FC, et al. Aerobic exercise influences quality of life of children and youngsters with congenital heart disease: a randomized controlled trial. The Journal of adolescent health : official publication of the Society for Adolescent Medicine. 2014;55:65-72.

[210] Dulfer K, Duppen N, Van Dijk AP, Kuipers IM, Van Domburg RT, Verhulst FC, et al. Parental Mental Health Moderates the Efficacy of Exercise Training on Health-Related Quality of Life in Adolescents with Congenital Heart Disease. Pediatr Cardiol. 2014.

[211] Frigiola A, Bull C, Wray J. Exercise capacity, quality of life, and resilience after repair of tetralogy of Fallot: a cross-sectional study of patients operated between 1964 and 2009. Cardiol Young. 2014;24:79-86.

[212] Garcia Guerra G, Joffe AR, Robertson CM, Atallah J, Alton G, Sauve RS, et al. Health-related quality of life experienced by children with chromosomal abnormalities and congenital heart defects. Pediatr Cardiol. 2014;35:536-41.

[213] Gerelli S, van Steenberghe M, Murtuza B, Bojan M, Harding ED, Bonnet D, et al. Neonatal right ventricle to pulmonary connection as a palliative procedure for pulmonary atresia with ventricular septal defect or severe tetralogy of Fallot. Eur J Cardiothorac Surg. 2014;45:278-88; discussion 88.

[214] Goldberg CS, Lu M, Sleeper LA, Mahle WT, Gaynor JW, Williams IA, et al. Factors associated with neurodevelopment for children with single ventricle lesions. The Journal of pediatrics. 2014;165:490-6.e8.

[215] Hebert A, Mikkelsen UR, Thilen U, Idorn L, Jensen AS, Nagy E, et al. Bosentan Improves Exercise Capacity in Adolescents and Adults After Fontan Operation: The TEMPO

(Treatment With Endothelin Receptor Antagonist in Fontan Patients, a Randomized, Placebo-Controlled, Double-Blind Study Measuring Peak Oxygen Consumption) Study. Circulation. 2014;130:2021-30.

[216] Heusch A, Calaminus G, Kahl J, Schmidt K. Health related quality of life after corrective surgery for congenital heart disease. Klin Padiatr. 2014;226:281-6.

[217] Knowles RL, Day T, Wade A, Bull C, Wren C, Dezateux C. Patient-reported quality of life outcomes for children with serious congenital heart defects. Arch Dis Child. 2014;99:413-9.

[218] Komar M, Przewlocki T, Olszowska M, Sobien B, Podolec P. The benefit of atrial septal defect closure in elderly patients. Clinical interventions in aging. 2014;9:1101-7.

[219] Komar M, Przewlocki T, Olszowska M, Sobien B, Tomkiewicz-Pajak L, Podolec P. Is it worth closing the atrial septal defect in patients with insignificant shunt? Postepy w kardiologii interwencyjnej = Advances in interventional cardiology. 2014;10:78-83.

[220] Luyckx K, Goossens E, Rassart J, Apers S, Vanhalst J, Moons P. Parental support, internalizing symptoms, perceived health status, and quality of life in adolescents with congenital heart disease: influences and reciprocal effects. J Behav Med. 2014;37:145-55.

[221] Ly M, Roubertie F, Kasdi R, Chatti S, Vergnat M, Luu D, et al. The Modified Fontan Procedure With Use of Extracardiac Conduit in Adults: Analysis of 32 Consecutive Patients. The Annals of thoracic surgery. 2014;98:2181-6.

[222] Mellion K, Uzark K, Cassedy A, Drotar D, Wernovsky G, Newburger JW, et al. Health-related quality of life outcomes in children and adolescents with congenital heart disease. The Journal of pediatrics. 2014;164:781-8.e1.

[223] Muller J, Berner A, Ewert P, Hager A. Reduced health-related quality of life in older patients with congenital heart disease: a cross sectional study in 2360 patients. Int J Cardiol. 2014;175:358-62.

[224] Mussatto KA, Sawin KJ, Schiffman R, Leske J, Simpson P, Marino BS. The importance of self-perceptions to psychosocial adjustment in adolescents with heart disease. Journal of pediatric health care : official publication of National Association of Pediatric Nurse Associates & Practitioners. 2014;28:251-61.
[225] O'Byrne ML, Mercer-Rosa L, Zhao H, Zhang X, Yang W, Tanel RE, et al. Morbidity in children and adolescents after surgical correction of interrupted aortic arch. Pediatr Cardiol. 2014;35:386-92.
[226] Opic P, Roos-Hesselink JW, Cuypers JA, Witsenburg M, van den Bosch A, van Domburg RT, et al. Psychosocial functioning of adults with congenital heart disease: outcomes of a 30-43 year longitudinal follow-up. Clinical research in cardiology : official journal of the German Cardiac Society. 2014.
[227] Opic P, Utens EM, Ruys TP, van Domburg RT, Witsenburg M, Bogers AJ, et al. Long-term psychosocial outcome of adults with tetralogy of Fallot and transposition of the great arteries: a historical comparison. Cardiol Young. 2014;24:593-604.
[228] Raju V, Dearani JA, Burkhart HM, Grogan M, Phillips SD, Ammash N, et al. Right ventricular unloading for heart failure related to Ebstein malformation. The Annals of thoracic surgery. 2014;98:167-73; discussion 73-4.
[229] Schoormans D, Sprangers MA, van Melle JP, Pieper PG, van Dijk AP, Sieswerda GT, et al. Clinical and psychological characteristics predict future healthcare use in adults with congenital heart disease. European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2014.
[230] Schoormans D, Mulder BJ, van Melle JP, Pieper PG, van Dijk AP, Sieswerda GT, 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 : journal of the

Working Group on Cardiovascular Nursing of the European Society of Cardiology. 2014;13:86-94.

[231] Sistino JJ, Atz AM, Simpson KN, Ellis C, Ikonomidis JS, Bradley SM. The prevalence of attention-deficit/hyperactivity disorder following neonatal aortic arch repair. Cardiol Young. 2014:1-7.

[232] Wang Q, Hay M, Clarke D, Menahem S. Associations between knowledge of disease, depression and anxiety, social support, sense of coherence and optimism with health-related quality of life in an ambulatory sample of adolescents with heart disease. Cardiol Young. 2014;24:126-33.

[233] Werner H, Latal B, Valsangiacomo Buechel E, Beck I, Landolt MA. Health-related quality of life after open-heart surgery. The Journal of pediatrics. 2014;164:254-8.e1.

[234] Wittlieb-Weber CA, Paridon SM, Gaynor JW, Spray TL, Weber DR, Brothers JA. Medium-term outcome after anomalous aortic origin of a coronary artery repair in a pediatric cohort. Journal of Thoracic and Cardiovascular Surgery. 2014;147:1580-6.

[235] Neal AE, Stopp C, Wypij D, Bellinger DC, Dunbar-Masterson C, DeMaso DR, et al. Predictors of Health-Related Quality of Life in Adolescents with Tetralogy of Fallot. The Journal of pediatrics. 2015;166:132-8.

[236] van der Bom T, Winter MM, Knaake JL, Cervi E, de Vries LS, Balducci A, et al. Long-term benefits of exercise training in patients with a systemic right ventricle. Int J Cardiol. 2015;179:105-11.