


Quality of life in adults with congenital heart disease

Mascha Kamphuis

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PROEFSCHRIFT

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VII. "When we speak about active ageing, we certainly do not mean only physical aspects, however important they may be. What is equally important is social and mental well-being, which are essential when adding more life to years and not merely years to life." (1999, Brundtland, World Health Organization)

VIII. Het bepalen van zoektermen bij een literatuuronderzoek op het gebied van kwaliteit van leven wordt ernstig bemoeilijkt door het ontbreken van een universeel erkende definitie van het begrip.

IX. "Denial is often considered maladaptive, but the helpful nature of denial should not be overlooked." (Zeltzer, 1980)

X. Zonder kritiek kom je niet verder, zonder positieve kritiek zeker niet.

XI. De vermoeidheid na het sporten, die wijst op verlies van lichamelijke energie, gaat samen met geestelijke leegheid, die weer ruimte biedt voor nieuwe inspanningen.

XII. "Remember that the best relationship is one in which the love for each other exceeds the need for each other." (Dalai Lama)

XIII. Hoe moeilijker het pad, hoe mooier het (voor-)uitzicht.

Stellingen

behorend bij het proefschrift
'Quality of life in adults with congenital heart disease'
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13 november 2002

I. Volwassenen met een complexe aangeboren hartafwijking hebben in het algemeen een goede kwaliteit van leven, hun (ervaren) fysieke toestand is echter een belangrijk punt van zorg. (dit proefschrift)

II. Hoewel volwassenen met een milde aangeboren hartafwijking geen fysieke of medische reden hebben om zich beperkt te voelen, kan onjuiste informatie of gebrek aan informatie aanleiding geven tot onnodige restricties in het dagelijks leven. (dit proefschrift)

III. Ter voorkoming van onnodige restricties bij patiënten met een aangeboren hartafwijking is het noodzakelijk dat bedrijfsartsen en verzekeringsmaatschappijen beter geïnformeerd worden over de huidige prognose en toekomstperspectieven van deze groep. (dit proefschrift)

IV. Het is van belang om patiënten met een aangeboren hartafwijking op jonge leeftijd loopbaanbegeleiding te bieden, zodat eventuele fysieke beperkingen en adviezen voor de hoogst mogelijke opleiding meegenomen kunnen worden in beslissingen voor de toekomst. (dit proefschrift)

V. Een afsluitend kindercardiologisch consult voor patiënten met een aangeboren hartafwijking op de leeftijd 16-18 jaar is een belangrijk moment om de adolescent optimaal te informeren over zijn/ haar aandoening en te verwachten restricties in het dagelijks leven en tevens om met die gegevens een beslissing te nemen over de noodzaak tot het vervolgen van cardiale controle. (dit proefschrift)

VI. Samenwerking tussen de kindercardioloog en de (congenitaal) cardioloog is cruciaal voor een goede zorg van de volwassene met een aangeboren hartafwijking.

Forever is our today
Who waits forever anyway?
Queen: 'Who wants to live forever'

Aan PaMa

Contents

Chapter 1

Introduction and outline of the thesis

Introduction

Advances in medical and surgical treatment of congenital heart disease have led to reduced mortality in patients with a range of cardiovascular abnormalities. Mortality rates used to be about 85% in the 40's but have fallen to 15% nowadays.[1,2] This has resulted in an increase in the number of adults alive with congenital heart disease. In the Netherlands, accurate statistics on prevalence rates are lacking. It is estimated that at least 70,000 adults with different types of congenital heart diseases, including those with a mild defect, are alive in the Netherlands. This number increases with about 1400 cases each year.[3] It is known that many of the patients with corrected or palliated congenital heart disease will suffer from medical complications (such as arrhythmia, cardiac failure, conduit obstruction, or valve insufficiency).[4]

Mortality and morbidity are the conventional measures for the effects of treatment and prevention in medicine. In chronic diseases however (such as congenital heart disease) mortality and morbidity show little variance and the patients' priorities may lie elsewhere. It is therefore increasingly important to address the patients' health-related quality of life and to focus on several psychosocial issues in addition to biochemical or physical outcomes.

Screening and monitoring of such issues may allow identification of psychosocial problems in the individual in order to better plan and allocate resources for research, training and health care.[5] Moreover, compliance with treatment will increase if this is associated with improved health-related quality of life. Patients' satisfaction with the medical consultation may also increase if physicians address their emotional wellbeing as well as functional health status.[6] A recent study in daily clinical oncology practice has shown that routine, standardised health-related quality of life assessment is a simple, efficient, effective and acceptable means of increasing communication.[7]

In 1996, the Leiden Center for Child Health and Pediatrics (TNO[a]-LUMC[b]) started a study in patients with mild congenital heart disease. In this study, a questionnaire was used comprising validated measurements (the TNO-AZL[c] Adult Quality of Life (TAAQOL)- questionnaire[8], and the Short Form-36 items[9,10] (SF-36)), as well as questions concerning disease-specific health-related quality of life and disease-related limitations.[11]

[a] TNO : Netherlands Organization for Applied Scientific Research, Prevention and Health
[b] LUMC : Leiden University Medical Center, Willem-Alexander Department of Pediatrics
[c] AZL : Academic Hospital Leiden, now known as Leiden University Medical Center

This study (Chapter 3) was then extended and the questionnaire further developed. Health-related quality of life (Chapters 4 to 6) and several psychosocial aspects, such as difficulties in daily life, disease-related knowledge and employment (Chapters 7 and 8) were evaluated in 2 groups of adults with a congenital heart defect: patients with previously operated complex congenital heart disease, as well as patients with mild congenital heart disease. Data were compared with each other, and with those of reference groups from the general population. In addition to the introduction of this thesis, Chapter 2 presents a literature review on health-related quality of life studies in congenital heart disease.

Definitions

Health-related quality of life

In the methodological literature, consensus about the definition of health-related quality of life is growing. Health-related quality of life is considered a multi-dimensional construct, including domains of physical, social, cognitive and emotional functioning of the patient,[12-16] in concordance with the definition of health by the World Health Organization.[17]

Literature invariably shows considerable disagreement between patients and proxies (such as the parent, physician, nurse or others) in their ratings of the patient's health-related quality of life.[18,19] Therefore, it is commonly proposed that the perception of the patient is crucial to the concept of health-related quality of life. In some situations however (if the patient is a child or mentally disabled), proxy ratings are the only ratings possible.[18,19]

In addition, various authors have suggested a second level of subjectivity to the concept of health-related quality of life: the value that the patient assigns to the perception of functioning.[14,15,20-24] We are interested not only in the patient's perception of the ability to climb the stairs (subjective health status); we also want to know the personal feeling of that ability or disability (health-related quality of life). Thus, the concept of health-related quality of life involves a double subjectivity.

In summary, the health-related quality of life instrument used for this study (the TAAQOL[8]) includes the following characteristics: 1. it comprises several scales measuring the main dimensions of health, 2. the individual is the respondent, and 3. patients were asked not only to report their level of

functioning but also how they feel about their (dys)functioning. Furthermore, although the phenomenon to be measured is subjective in nature, the questionnaire meets the scientific criteria such as reliability, validity, sensitivity and standardisation.

Study groups

In this thesis, 3 groups of patients were studied.

Group 1: patients with previously operated complex congenital heart disease

Patients with previously operated complex congenital heart disease were approached, excluding those after anatomical corrective surgery according to the segmental analysis.[25] All 78 included patients had undergone a non-anatomical correction, such as venous switch for transposition of the great arteries, a correction with the use of allogenous tissue (e.g. Rastelli correction, insertion of a mechanical valve), or partial or complete cavopulmonary connection. The main common characteristic of this group was that postoperatively none of the participants had an anatomically normal heart as described by the segmental analysis.[25] It was expected that this group would experience medical problems and limitations.

Group 2A and 2B: patients with mild congenital heart disease.

None of them had needed an operation or intervention.

2A: 94 patients with a "hemodynamically insignificant" defect, who all were still under cardiac surveillance in the Leiden University Medical Center at the time of study.

2B: 82 patients with a "hemodynamically insignificant" defect and those with a spontaneously resolved congenital heart lesion, who were selected from the archives and who were not all under cardiac surveillance in the Leiden University Medical Center anymore. For this group it was expected that patients would not experience heart-related problems, and therefore this group was considered as a control group in comparisons with group 1.

Reference groups

In addition, for some of the measures a formal *reference group* from the general population was also available.

Outline of the thesis

Chapter 2 presents a literature review on health-related quality of life studies in congenital heart disease. Special attention is paid to the conceptual description of health-related quality of life, as well as to the measurements used to assess health-related quality of life, and the resulting outcome of the included studies.

Chapter 3 presents the study, in which the first draft of the congenital heart disease questionnaire was evaluated in patients with mild congenital heart disease. In this study, social impediments experienced by these patients are examined, and their health-related quality of life and health status are compared with those of the general population.

In **Chapter 4** the data of health-related quality of life and subjective health status in adults with a previously operated complex congenital malformation are compared with those of the general population. In addition, these data are related to indices of physical status, which may increase our understanding of the concept of health-related quality of life.

Chapter 5 focuses on patients with mild congenital heart disease, and studies the need to continue their cardiac follow-up into adult life. This study considered other factors than hemodynamical data, such as health-related quality of life and subjective health status (as compared with reference data), difficulties in daily life, and cardiac status (i.e. regularity of follow-up, current diagnosis, and antibiotic regimen).

Chapter 6 reports on the development of a congenital heart disease-specific instrument for measurement of health-related quality of life, based on the format of the TAAQOL. Disease-specific instruments are more responsive to change for example when an intervention is done. To date, no such instruments for congenital heart disease have been described.

In order to improve patient education, the study in **Chapter 7** evaluates difficulties in daily life, and satisfaction with level of knowledge about their disease for both patient groups.

Chapter 8 describes job participation, career and actual job problems in adults with complex congenital heart disease, compared with those of adults with mild congenital heart disease and reference groups. With this information, career counselling may be improved.

Finally, all results and conclusions are summarised and placed in perspective in **Chapter 9**.

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Chapter 2

Quality of life studies for congenital heart disease: a review of definition, measures and results

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S.P. Verloove-Vanhorick, E. Verrips

Submitted

Abstract

Definitions and measures of health-related quality of life were evaluated in studies on congenital heart disease patients and the outcome of their health-related quality of life was summarised.
In total, 69 papers were studied in detail. In 46% (32) of these no description of health-related quality of life was provided. Moreover, given definitions varied widely. Seven research-groups conceptually defined health-related quality of life as a self-reported multidimensional construct, and 7 (partly other) research-groups used a *validated* self-report multidimensional measure. Four studies focussed on the patients' appreciation of their health status. In general, most patients with a congenital heart disease (even those with complex disease) felt they were healthy. However, subgroups (such as older or cyanosed patients) appear to do worse.
In conclusion, health-related quality of life in research with congenital heart disease patients is seldom defined clearly and measures often aim at different targets. Therefore, these studies should be compared with great caution. Health-related quality of life outcome is surprisingly positive in most studies that measured congenital heart disease patients' own perception. Coping strategies might explain this.

Introduction

In medicine, the effects of therapy and prevention of disease are conventionally measured in terms of mortality and morbidity. In chronic diseases, however, mortality and morbidity show little variance. Moreover, the patient's focus may lie elsewhere. In evaluation studies in chronic conditions, health status and health-related quality of life are therefore important additional measures of outcome. Furthermore, increasingly it has become the objective of health care in Western Countries, to optimise health-related quality of life. Screening and monitoring of health-related quality of life may allow identification of psychosocial problems in the individual and better planning and allocation of resources for research, training and health care.[1] Moreover, compliance with treatment will improve if treatment is associated with improvement in health-related quality of life and patients' satisfaction with the medical consultation may also increase if physicians address their general wellbeing as well as functional status.[2] A recent study in daily clinical oncology practice has shown that routine, standardised health-related quality of life assessment is a simple, efficient, effective and acceptable means of increasing communication.[3]

Defining health-related quality of life

In the methodological literature, consensus about the definition of health-related quality of life is growing. Health-related quality of life is considered a multi-dimensional construct, including domains of physical, social, cognitive and emotional functioning of the patient,[4-8] in concordance with the definition of health by the World Health Organization.[9]

Furthermore, it is commonly proposed that the perception of the patient is crucial to the concept of health-related quality of life. In most psychobiological models of health behaviour, it is implicit that it is the perception of the patients and not (necessarily) the objective truth that is important.[10] Therefore, the phenomenon is defined as subjective in the sense that the patient's perspective is central, and not a clinical test or the perception of the doctor or a close relative. Literature invariably shows considerable disagreement between patients and proxies (such as the parent, physician, nurse or others) in their ratings of the patient's health-related quality of life.[11,12] In some situations however (the patient is a child or mentally disabled), proxy ratings are the only ratings possible.[11,12] And although clinically these

perspectives may be important, these must be considered as concepts different from health-related quality of life.

Moreover, various authors have suggested a second level of subjectivity to the concept of health-related quality of life: the psychological value that the patient assigns to the perception of functioning.[6,7,13-17] We are interested not only in the patient's perception of the ability to climb the stairs, we also want to know the personal feeling concerned with that ability or disability. Thus, the concept of health-related quality of life involves a double subjectivity: firstly, the patient's perception of functioning, and secondly, the patient's evaluation of that functioning.

The above mentioned theoretical considerations have implications for the way health-related quality of life has to be measured. Firstly, instruments should include several scales measuring the main dimensions of health-related quality of life. Secondly, the individual is the preferred respondent. Thirdly, individuals must be asked not only to report their level of functioning but also how they feel about their functioning. Furthermore, although the phenomenon to be measured is subjective in nature, its measurement should meet scientific criteria such as reliability, validity, sensitivity and standardisation.

The growing importance of health-related quality of life is also reflected in research in patients with congenital heart disease. A rapidly growing number of studies in this field seems to focus on health-related quality of life. Similar to the medical literature in general,[6] the question arises how rigorously health-related quality of life has been defined and measured in these studies.

The aim of the present review is therefore, to evaluate definitions and measures of health-related quality of life in studies on patients with congenital heart disease. Moreover, results and conclusions of these studies with regard to health as judged by the patients are summarised.

Methods

Selection of references

We conducted a computer search using PubMed (NCBI), Current Contents and Psych-info (Winspirs-Silver Platter) databases (Biomedical, Social-Behavioural and Psychology/ Psychiatry Sciences). The search covered the years 1966 to November 2001. Since 'quality of life' is the primary focus of this

review, this term was used in combination with 'heart defects, congenital'. Both search terms were searched as Mesh heading and as free text in any field including abstracts.

Although some publications were not categorised by Mesh as 'quality of life' study, they concerned measurement of the patients' opinion of health. No efficient search term was available for such studies, and therefore, these were collected from personal files which resulted from extensive pilot searches and snowball techniques (i.e. review of the references of articles thus obtained).[18] Moreover, to further check and complete this search, the Spilker Quality-of-Life-Bibliography and updates were evaluated which cite all articles up to 1994 that are said to describe or employ quality of life instruments[19-23] as well as the Cochrane Library database.

All search hits were captured in a computer database, excluding doubles. The resulting references (abstracts) were evaluated to judge if the paper reprint should be studied. Selection criteria for this detailed study were A. Studying congenital heart disease, and B.1. Suggesting health-related quality of life as an outcome measure (in title or abstract), and/ or B.2. Studying the patients' own perception of their health, and/ or B.3. Studying different dimensions (such as physical, mental, and social). Publications were excluded if they concerned editorials or letters, or were not written in English or German. To restrict the search, publications on the following patient-groups were not considered: patients with cardiomyopathy, and patients after cardiac transplantation. In addition, studies with mentally retarded patients (e.g. Down syndrome) were not included, since this warrants a specific approach for the questionnaire used.

All publications that met the selection criteria A, and B1, B2 or B3 were studied by paper reprint. Of these, all studies that focused on the patients' health-related quality of life as self-reported were included for detailed evaluation of the health-related quality of life definition, the used measures and results (Tables 1 and 2).

Results

General characteristics of the search

The computer searches resulted in 150 hits. After reading the abstracts, 64 articles were selected for studying in further detail, since they met both inclusion criteria A and B.[24-87] The selection was completed with 5 papers from personal files that focused on the patient's opinion of their health, and either did not specifically mention health-related quality of life[10,88-90] or comprised also patients with non congenital heart diseases.[91]

Studies originated from research centers all over the world, most frequently from the USA (15), Germany (11), and Japan (8).

Of the 69 selected studies, 23 had focused on the patients' health self-report (Tables 1 and 2). Of these, two dissertations were found by searching the database Psych-info.[41,74] Two publications originated from the database Current Contents.[36,54] All others were traced by searching PubMed.

Conceptual definition

Of those 57 studies that suggested health-related quality of life as an outcome measure, 32 (56%) showed no definition or explanation of the concept of health-related quality of life at all. Many publications only used the term 'quality of life' in the abstract and studied solely medical outcome. Studies that described health-related quality of life, used a diversity of descriptions: medical outcome (e.g. re-operations/ residual defects/ lung function/ growth/ exercise testing), New York Heart Association functional class, Somerville index, work/ education/ social life/ marital status, and complaints/ symptoms.

Publications with 'quality of life' in the title in which no definition was given, used descriptions that indicated what was measured, e.g. 'an overall picture of the life of the child',[31] 'family and personal life',[37] or 'personal health assessment'.[58]

In total, 7 research-groups conceptually defined health-related quality of life as a self-assessed, multidimensional construct (Tables 1 and 2).[36,38,41,48,49,62,63,74,76] In addition, the publication by Simko et al. completely focussed on defining the concept of health-related quality of life,[73] which was further evaluated in a congenital heart disease group in the dissertation mentioned.[74] Van Doorn et al. stated that there was 'no universally accepted definition', but used 'age specific validated questionnaires to measure the patients' current health status'.[91]

Four research-groups (in 5 studies) defined health-related quality of life according to all our requirements (multidimensional, self-report, and subjective in two respects).[36,41,48,49,76]

Measures

In total, 11 research-groups with 15 publications included a self-reported questionnaire,[10,36,38,41,51,57-59,62,63,68,74,76,90,91] of which 10 research-groups used a multidimensional construct (Tables 1 and 2).[10,36,38,41,51,62,63,68,74,76,91] Of these, 7 research-groups used a validated questionnaire that referred to previous validating studies (Table 1).[36,41,62,63,68,74,76,91]In 5 publications the second level of subjectivity had been measured using a questionnaire.[36,41,51,76,91]

All other studies that focused on the patients' opinion of their health, used (semi-structured) non-validated interviews, and described several problems and subjects as indicated by the patients (Table 2).[39,43,48,49,78,86,88,89]

Outcome

In total, 23 studies focussed on health as judged by the patients themselves (Tables 1 and 2). Some of these studies used well-validated questionnaires and reference groups, others included a single question about general health, or used interviews. Mainly mixed groups of patients with congenital heart disease have been studied, both operated and unoperated patients.

For a valid judgement of conclusions, only those studies were considered that used a validated measure (Table 1). Their outcome could be compared with a reference group and could therefore be placed in perspective.

When generalising these conclusions, it appeared that most patients with a congenital heart disease, even those with a complex malformation (transposition of the great arteries, tetralogy of Fallot, or univentricular atrioventricular connection) felt they were healthy.[36,41,57-59,62,63,68,74,76,90,91] The majority of patients with a congenital heart disease experienced no limitations and their health-related quality of life did not differ from or was even better than that of norm groups.

Studies in which health judgement was compared with medical outcome showed that the severity of the heart disease was not necessarily congruent with health-related quality of life,[10,76] and that cardiac functional status is irrespective of the presence of anatomical or electrophysiological sequelae.[90]

However, those studies that evaluated health-related quality of life more specifically per age group or diagnosis, indicated impaired health-related quality of life below the age of 15 years,[91] above the age of 23 years[68] or 35 years,[76] and in cyanosed patients.[68,74] In two studies by Immer at al. in patients with complex congenital heart disease, results showed worse outcome in the older age group (16-20 years) than in the younger group (8-16 years).[48,49]

Discussion

The present study identified publications that measured health-related quality of life in patients with congenital heart disease and evaluated these studies on the description of a definition of health-related quality of life and the instrument used. Results concerning health-related quality of life as judged by the patients themselves were summarised.

Only few studies in patients with congenital heart disease described what was meant by health-related quality of life in the way we had defined it,[36,41,48,49,76] or used a validated health-related quality of life-instrument that measured the concept according to our definition (multidimensional, self-report, and subjective in two respects).[36,41,76,91]

The finding that quality of life studies aimed at many different and often wrong targets, was also found in previous reviews on health-related quality of life measures in medical literature [6] and in children.[14]

Main point of discussion is that no gold standard exists for the concept of health-related quality of life.[92] For the definition we used, we took several landmark publications into account. One of the key factors of this definition is that it should be considered multidimensionally,[4-8] in concordance with the definition of health by the World Health Organization, that 'health is a state of complete physical, mental, and social well-being, and not merely the absence of disease or infirmity'.[9] Such an approach is most valuable for developing an intervention to improve a specific impaired aspect of life.

Furthermore, it is commonly proposed that the perception of the patient is crucial to the concept of health-related quality of life.[6,7,13-17] Only then it is a concept different from clinically well-known and generally accepted measures such as blood pressure, exercise tests, or laboratory values. In that way, it will add to all such outcome measures that already exist.

By not defining the concept of health-related quality of life, or by using several different measurements in publications, correct interpretation of results is impossible and comparison of results between studies will remain difficult. During the years more health-related quality of life instruments have been developed, and nowadays the diversity of them makes it hard to choose the appropriate one and to compare between studies. The choice of an instrument will mainly depend on the research question. It has to be stressed that at least extensive description of the definition and measurement should take place.

Considering the studies that have measured health-related quality of life as judged by the patients themselves, comparison is difficult with the different instruments and patients included. However, trying to do so and focussing solely on those studies that used a validated instrument, we may conclude that patients with various congenital heart diseases, even those with a complex lesion (such as tetralogy of Fallot, univentricular atrioventricular connection, or transposition of the great arteries) appear to feel healthy.[58,59,68,76] Although this seems to be a surprising finding, it confirms the distinction between medical outcome and health-related quality of life. It is suggested that limitations may become accepted and the value of life changes by the development of coping mechanisms.[62,68,76] This phenomenon was also found in a study on extremely low-birth-weight infants at adolescence.[93] The influence of coping can be confirmed by findings such as a better health-related quality of life outcome in patients than in the general population,[36,62] and no relation between the severity of the disease and health-related quality of life outcome.[10,76,90]

Some studies, however, found worse health-related quality of life in cyanosed patients and in patients with complex disease.[49,68,74] In addition, studies indicated worse health-related quality of life at older age, although the limit changed per study.[49,68,76] Perhaps these patient groups could benefit from learning coping mechanisms. It would be relevant to study such mechanisms in further detail and to gain insight in possible ways to optimise the patients' wellbeing.

All reviewed studies focussed on relatively young patients. Since studies on medical outcome have shown a decreasing physical condition in patients with complex congenital heart disease after the age of 30, the number of reported problems might increase with age. Furthermore, as a result of medical developments, nowadays it is possible to treat patients with much more complicated congenital heart diseases than in the past. It is to be expected that

a new generation of patients will experience more medical problems than nowadays. It remains unclear what effect this might have on the results of their health-related quality of life.

Remarks and study limitations

Some methodological issues may have influenced the results of this review. In general, only a selection of studies is published, and results are only partly presented in publications. In addition, it was not possible to evaluate the studies in all languages, which can cause the 'Tower of Babel bias'.[18,94]

Moreover, since it seemed that some quality of life studies were not categorised by Mesh terms as such, these studies were collected from personal files. It is therefore not certain if all such studies were traced. However, with extra control from the Spilker Quality-of-Life-Bibliography and updates,[19-23] the Cochrane Library databases, and the extensive personal files, it is assumed that no important paper has been omitted.

Conclusion

Health-related quality of life in research with congenital heart disease patients is seldom defined clearly and applied measurements aim at different targets. Therefore, studies suggesting health-related quality of life as an outcome measure should be viewed and compared cautiously. Although it is important to use a multidimensional construct and to focus on how the patients feel about their functioning when studying health-related quality of life, at least extensive description of the definition and applied instrument is necessary.

The positive outcome for health-related quality of life in congenital heart disease in studies that measured patients' own perception seems surprising. However, subgroups (such as older or cyanosed patients) appear to do worse. Perhaps these patient groups could benefit from learning coping mechanisms. With information on coping mechanisms, counselling of patients and parents could be improved.

Table 1 Characteristics of studies, included because patients' perception was questioned, VALIDATED measures

Reference 1st author publ. year country	Patient characteristics sample, diagnosis age (mean)	Description QoL a. multidimensional b. patients' perception c. double subjectivity	Measurement 1. paper/pencil 2. interview: face to face or by phone	Measurement QoL a. multidimensional b. patients' perception b1- multidimension b2- general health c. double subjectivity	Results
MøjenLaane[63] '97 Sweden	n= 51 spont. closed VSD n= 83 norm 2-8 (5) yr.	a, b mainly judgement parent	1, method of Lindström*	a, b1, b2 overall score and per domain	'No difference compared with norm (but lower satisfaction with family network).'
MøjenLaane[62] '97 Sweden	n= 164 several CHD n= 301 norm 2-12 (6) yr.	a, b mainly judgement parent	1, method of Lindström*	a, b1, b2 overall score and per domain	'There were no statistically significant differences between the CHD groups (spontaneously cured, after surgery, associated malformations) and the controls for overall QoL. A trend existed for higher subjective experience of QoL in the total group as well as in all the subgroups'
Van Doorn[91] '00 UK	n= 19 Mitral valve replaced (all but 3 concerned CHD) norm from literature 10-25 (14) yr.	none '..no universally accepted definition exists..'	1, CQOL* (9-15 yr.) 1, SF-36* (>16yr.)	a, b1, c +judgement parent overall score a, b1, b2 score per domain	Younger patients: 'All patients reported impaired QoL.' Older patients: 'All but two perceived their health as norm.'
Fekkes[36] '00 Netherlands	n= 94 mild CHD n= 350 norm 15-30 (22) yr.	a, b, c	1, TAAQOL* 1, SF-36*	a, b1, b2, c score per domain a, b1, b2 score per domain	'Patients with social restrictions: QoL comparable to general population. Patients without social restriction: better QoL than general population.'

*validated, CHD= congenital heart disease, CQOL= Child Quality of Life questionnaire, QoL= quality of life, SF-36= Short Form-36 items, TAAQOL= TNO-AZL Adult Quality of Life, VSD= ventricular septal defect

Table 1 (continued) Characteristics of studies, included because patients' perception was questioned, VALIDATED measures

Reference	Patient characteristics	Description QoL	Measurement	Measurement QoL a. multidimensional	Results
1st author publ. year country	sample, diagnosis age (mean)	a. multidimensional b. patients' perception c. double subjectivity	1. paper/pencil 2. interview: face to face or by phone	b. patients' perception b1- multidimension b2- general health c. double subjectivity	
Saliba[68] '01 France	n= 67 Univentricular heart norm from literature 17-49 (21) yr.	none	1, Dukes*	a, b1, b2 score per domain	'The Duke scores of adults with univentricular heart were similar to the normal population. Cyanosis predicted a worse score for physical and perceived health measures. Patients younger than 23 years scored better for almost all health and dysfunction measures'
Simko[74] '00 USA	n= 124 several CHD n= 124 norm 18-59 yr.	a, b	1, SIP*	a, b1, b2 overall score and per domain	'Adults with CHD did not see themselves as having any physical limitations, their perception of themselves and their individual expectations are normal.'
Meijboom					'The majority patients considered their health as: good/ excellent for...
'93[90]	n= 104 ASD	none	1, CBS-survey*	b2	87%
'94[57]	n= 109 VSD	none	1, CBS-survey*	b2	84%
'95[58]	n= 77 4F	none	1, CBS-survey*	b2	82%
'96[59]	n= 58 TGA norm from literature ± 20 yr.	none	1, CBS-survey*	b2 percentages	88%.' 'The personal health assessment is as good as that of the Dutch population.'

*validated, ASD= atrial septal defect, CBS= central database of Statistics Netherlands, CHD= congenital heart disease, QoL= quality of life, SF-36= Short Form-36 items, SIP= Sickness Impact Profile, TGA= transposition of the great arteries, 4F= Tetralogy of Fallot, VSD= ventricular septal defect

Table 1 (continued) Characteristics of studies, included because patients' perception was questioned, VALIDATED measures

Reference	Patient characteristics	Description QoL	Measurement	Measurement QoL a. multidimensional b. patients' perception b1- multidimension b2- general health c. double subjectivity	Results
1st author publ. year country	sample, diagnosis age (mean)	a. multidimensional b. patients' perception c. double subjectivity	1. paper/pencil 2. interview: face to face or by phone		
Greenwood[41] '00 USA	n= 80 several CHD n= 80 norm group 25-41 (35) yr.	a, b, c	1,QoL-model: SF-36*, QoL inventory*, Rosenberg*, Social Support*, Pearlin Mastery Scale*	a, b1, b2, c score per domain	'Patients see their functional status below that of peers. On the other hand, patients appear to have a similar level of life satisfaction.'
Ternestedt[76] '01 Sweden	n= 32 4F, ASD >25 yr. and >35 yr.	a, b, c +judgement cardiologist	1+2, Kajando/ Ternestedt*	a, b1, b2, c overall score and per domain	'The 4F group rated their QoL higher than the ASD group, but both groups had lower figures than at the 20 year follow-up. Fewer patients with 4F than the ASD group considered that their lives were affected by the heart disease.'

*validated, ASD= atrial septal defect, CHD= congenital heart disease, QoL= quality of life, SF-36= Short Form-36 items, 4F= Tetralogy of Fallot

Table 2 Characteristics of studies, included because patients' perception was questioned, NOT validated instruments

Reference 1st author publ. year country	Patient characteristics sample, diagnosis age (mean)	Description QoL a.multidimensional b.patients' perception c.double subjectivity	Measurement 1. paper/pencil 2. interview: face to face or by phone	Measurement QoL a.multidimensional b.patients' perception b1- multidimension b2- general health c.double subjectivity	Results
Kendall[10] '01 UK	n= 57 several CHD 11-16 yr.	none	1	a, b1, b2 percentages per subject	'The majority of the adolescents appear to regard their state of health as good/ average (96%), and themselves as not/ little different from peers (70%). Their perception of health was most strongly related to the degree to which the condition disrupted social relationships and not to the complexity of the disease.'
Gersony[38] '93 USA	n= 1681 AoS, PS, VSD norm from literature/ institute adults	a, b other: NYHA-class, interval since recent examination, insurance	1 or 2 (by phone)	a, b1, b2 descriptions and percentages	'Self-perception of health status was similar to that of the general population'
Kaemmerer[51] '94 Germany	n= 146 several CHD >18 yr.	none	1	a, b1, b2, c percentages per subject	'60% of the patients felt not healthy or somewhat impaired. No relation with the ability index of Somerville was found. For 40% of the patients the greatest burden of disease were the physical limitations.'

AoS= aortic stenosis, CHD= congenital heart disease, NYHA= New York Heart Association functional class, PS= pulmonary stenosis, QoL= quality of life, VSD= ventricular septal defect

Table 2 (continued) Characteristics of studies, included because patients' perception was questioned, NOT validated instruments

Reference 1st author publ. year country	Patient characteristics sample, diagnosis age (mean)	Description QoL a. multidimensional b. patients' perception c. double subjectivity	Measurement 1. paper/pencil 2. interview: face to face or by phone	Measurement QoL a. multidimensional b. patients' perception b1- multidimension b2- general health c. double subjectivity	Results
All studies using interviews					
Hallidie-Smith[43] '77 UK	n= 27 large VSD 6-16 yr.	none	2	a, b1	'All the patients remarked that they were symptom free and felt completely unrestricted both at work and in their leisure time.'
Immer[48] '94 Switzerland	n= 169 several CHD 8-16 yr.	a, b, c +judgement physician	2	a, b1, c descriptions + percentages per domain and for an overall score	'85% of the total group experience their QoL as normal. Impairment is most often caused by additional non-cardiac malformations or very severe heart disease. For the complex group only 17% experience a normal QoL.'
Immer[49] '98 Switzerland	n= 112 several CHD 16-20 yr.	a, b, c +judgement physician	2	a, b1, c descriptions + percentages per domain and for an overall score	'In 87% of the total group the QoL is normal. For patients with complex CHD (4F, TGA) a worse outcome was found than at younger age.'
Tong[78] '98 USA	n= 9 several CHD 13-25 yr.	none	2 semi-structured protocol	a, b1, c descriptions	Results cannot be summarised. This concerns an explorative study about the adults' subjective experiences and dilemmas; several subjects and concerns were raised.

CHD= congenital heart disease, QoL= quality of life, TGA= transposition of the great arteries, 4F= Tetralogy of Fallot, VSD= ventricular septal defect

Table 2 (continued) Characteristics of studies, included because patients perception was questioned, NOT validated instruments

Reference 1st author publ. year country	Patient characteristics sample, diagnosis age (mean)	Description QoL a.multidimensional b.patients' perception c.double subjectivity	Measurement 1. paper/pencil 2. interview: face to face or by phone	Measurement QoL a.multidimensional b.patients' perception b1- multidimension b2- general health c.double subject	Results
Mair[89] '92 UK	n= 38 Fontan 13-46 (22) yr.	none	2	b2 or parent percentages	'The majority of patients (89%) felt they were in good/ excellent condition.'
Doucet[88] '81 Canada	n= 25 several CHD 18-30 (24) yr.	none	2 semi-structured protocol	a, b1, c descriptions	Results cannot be summarised. This concerns an explorative study about the adults' subjective experiences and dilemmas; several subjects and concerns were raised.
Ghisla[39] '83 Germany	n= 39 4F 18-50 (25) yr.	none	2	a, b1, b2, c percentages per domain	'77% of all patients say to feel healthy and 72% of patients say to have the same chances for work as healthy people. All patients that experience restrictions have neurological or cardiological complexities.'
Wimmer[86] '90 Austria	n= 19 PH/ inoperable CHD Intervention study with nifedipine 1-14 yr.	none	2	b2 descriptions	'Considerable importance was attached to statements made by the patients/ parents, as to whether there was a subjective improvement with an increase in performance and well-being. Succes of the treatment was determined by an improved QoL.'

CHD= congenital heart disease, 4F= Tetralogy of Fallot, PH= pulmonary hypertension, QoL=quality of life

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Chapter 3

Health-related quality of life in young adults with minor congenital heart disease

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Abstract

The present study examined the social impediments experienced by young adults with minor congenital heart disease and compared the health status and health-related quality of life of these congenital heart disease patients with a matched group (age, sex) from the general population. Young adults with minor congenital heart disease, not in need of surgical correction, were sent a questionnaire.

Results showed that a substantial part of the congenital heart disease patients experienced social impediments, i.e. at school (19%), with medical examinations for job or insurance (19%), during free time (15%), in choosing (13%) or performing a job (9%) and taking out a life insurance policy (8%). Those congenital heart disease patients that experienced social impediments reported a health status and health-related quality of life comparable with the general population. Remarkably, congenital heart disease patients without social impediments reported a better health-related quality of life for 6 of the 12 scales, when compared with the general population.

This last result might be explained by coping mechanisms resulting in a higher appreciation of health status, which is expressed in a higher health-related quality of life.

Introduction

Traditionally, morbidity and mortality are the most widely used measures of medical outcome. Due to improved health care and treatment these measures insufficiently capture the full impact of medical interventions. Many diseases are not fatal any longer but may yet have a severe impact on a person's life. During the last decades these improved medical interventions led to a growing interest in assessing functional limitations and well-being of patients with several kinds of disease after treatment. Not only the medical results should be considered, but also the subjective satisfaction with life as judged by a patient should be used to reflect the impact of diseases on the lives of individuals.[1-5] Gradually, health status and quality of life are developing into standard outcome measures, in addition to morbidity and mortality.

Health status or functional status mainly focuses on a person's functioning. In general three domains are distinguished: physical, psychological and social functioning. Health-related quality of life is distinguished from health status in a way that health-related quality of life focuses on the emotional impact of limitations in health status, in addition to health status *per se*. In measuring health status and health-related quality of life, both may be proxy-reported or self-reported. Either way, it is generally agreed that what counts is the patients' subjective perspective, and that such subjectivity can be measured in a scientific, e.g. reliable, reproducible and valid way.

In paediatric cardiology the late effects of early diagnosed congenital heart disease have become of more interest. Recent research has focused on the quality of life of congenital heart disease patients and their psychosocial situation several years after (surgical) treatment. Kaemmerer et al. examined psychosocial problems of adolescents and adults with congenital heart disease after surgical correction.[6] The results showed that almost 60% of the patients did not feel healthy, and that they experienced impairments. Patients indicated that their congenital heart disease negatively affected their choice of occupation and their performance of sports.

Utens et al. assessed the behavioural and emotional problems in children and adolescents, at least nine years after surgical correction for congenital heart disease.[7] Children and adolescents obtained significantly higher problem scores on the Child Behavior Checklist and the Youth Self Report than same aged peers from reference groups.

Many studies focused on patients with congenital heart disease after surgical correction.[6-12] However, there are also patients with minor congenital heart disease who have not undergone any treatment. Patients with minor congenital heart disease are not in need of surgical treatment, because the defect is not expected to get worse or to give any major complaints. The prognosis of these patients is excellent. Although their minor defect could be operated, the risks for an operation are too high in comparison with the expected (minimal) improvement of their health. The present study focuses on these adolescents and young adults, ranging from 15 till 30 years of age. Although no surgical correction relating to their heart disease was needed, the question remains whether their condition has any impact on their daily life now they have reached adulthood.

Therefore, the present study had two objectives:

1) Which social impediments were and are experienced in daily life by people with minor congenital heart disease in relation to their heart disease, and which future social impediments do they expect?

2) How is the health status and health-related quality of life of those people with minor congenital heart disease who experienced social impediments in comparison to those patients who experienced no social impediments and how do health status and health-related quality of life of both groups differ from the general population?

Methods

Design

Two groups of subjects were approached. In August 1996, young adults with minor congenital heart disease (n= 148), not in need of surgical correction, between 15 and 30 years of age, who visited the cardiology department of the Leiden University Medical Center in the preceding year, were asked to participate in the study. Those agreeing to participate were sent a set of questionnaires. In addition, questionnaires were mailed to a sample of 2800 Dutch households drawn at random from the national telephone registry. From each household one person aged 15 years or older was asked to complete the questionnaires. Non-responders were sent reminder letters 2 months and 3 months following the initial mailing. This national study included also the norming of the Dutch language version of the SF-36 Health Survey.[13]

The study was approved by the "Medical Ethical Committee" of the Netherlands Organization for Applied Scientific Research (TNO) and informed consent was obtained from all participating patients or their parents.

Questionnaires

QOL-CHD

The Quality of Life-Congenital Heart Disease questionnaire (QOL-CHD) was only sent to the group of patients with a congenital heart disease. The QOL-CHD, a disease specific quality of life questionnaire was specifically tailored to patients with congenital heart disease. Main goal of this questionnaire was to make an inventory of the social impediments for patients with congenital heart disease.

The questionnaire comprises 2 scales: The first 13-item scale measures the social impediments that patients have experienced in the past and present. Patients are asked if they experienced limitations in relation to their congenital heart disease, for example at home, at school, when choosing a career, at work or getting a drivers license. The reliability of this scale, measured by Cronbach's alpha, was $\alpha = 0.77$. The second 8-item scale measures the social impediments that patients expect for the future, for example in daily life, choice of career, sports or family planning. Reliability for this scale: $\alpha = 0.73$. In addition the QOL-CHD contains six questions to make an inventory of those social impediments that are imposed by the patient's doctors. Patients are asked if their doctor imposed social impediments in relation to their congenital heart disease for daily life, school, choice of career, sports, free time activities and social life.

The items concerning these aspects were derived from the literature about congenital heart disease[6,14-17] and based on the experiences in clinical practice.

Health status: SF-36

Both the group of patients with a heart disease and the general population sample were asked to complete the SF-36. The SF-36 is a widely used instrument to measure health status; it discriminates well between healthy and less healthy people.[18-22]

The questionnaire measures functional health status and includes eight scales: physical functioning, role functioning (physical), role functioning (emotional), social functioning, bodily pain, mental health, vitality, and general health perceptions. The range for all scales is 0 to 100, with higher values indicating better functioning.

Health-related quality of life: TAAQOL

Both samples were asked to fill in the TNO-AZL Adult Quality of Life questionnaire (TAAQOL). The TAAQOL is a validated, generic health-related quality of life questionnaire developed by researchers from TNO and the Leiden University Medical Center. Health-related quality of life is defined as health status weighted by emotional impact of problems in health status.[23-25] The TAAQOL comprises 12 scales: gross motor functioning, fine motor functioning, cognitive functioning, sleeping, pain, social functioning, limitations of daily activities, sexuality, vitality, happiness, depressive moods, and aggressiveness. The TAAQOL measures the emotional impact of self-reported functional problems.

In all scales, except in the scales concerning vitality, happiness, depressive moods and aggressiveness, each item consists of two questions. First the frequency of occurrence of a specific complaint or limitation during the last month is asked. If such a problem has occurred, the subjective appraisal of this problem is assessed. An example is given in Figure 1.

Figure 1 Item-example of the TAAQOL (scoring in parentheses)

Did you have difficulty in the last month with .....

walking up the stairs? ❑ no (1) ❑ a little ❑ some ❑ a lot

How much did that bother you?

❑ not at all (2) ❑ a little (3) ❑ quite a lot (4) ❑ very much (5)

Each item is encoded into one single score ranging from 1 to 5. A score of 1 is given when there is no limitation, a score of 2 when there is a limitation (i.e. a little, some, a lot) but when the person is not bothered by the limitation; a score of 3 when there is a limitation and the person experiences this limitation "a little" negatively; a score of 4 when there is a limitation and the

person experiences this limitation "quite a lot" negatively; and a score of 5 when there is a limitation and the person experiences this limitation "very much" negatively. This encoding of the scores allows for a weighting of functional problems by their emotional impact.

In the scales measuring vitality, positive moods, depressive moods and aggressive moods, the items measure only the frequency of a specific complaint or limitation during the last month. In the items in these domains it is not asked how much the person is bothered, because items in these domains already imply a positive or negative emotional state. In these scales, each item score is ranging from 1 to 4. An example is presented in Figure 2.

Figure 2 Item-example of the TAAQOL (scoring in parentheses)

In the last month, did you feel				
energetic?	❑ no (1)	❑ a little (2)	❑ quite (3)	❑ very (4)

For all TAAQOL-scales, items are added up and linearly transformed to 0 to 100 scales with higher values indicating a better quality of life.

Statistical analysis

Scale scores for both the SF-36 and the TAAQOL were obtained by adding item scores within scales, and transforming crude scale scores linearly to a 0 to 100 scale, with higher scores indicating a better health status and quality of life.

Based on the results from the QOL-CHD, the congenital heart disease patients were divided into two groups: patients who had experienced no social impediments in relation to their heart disease and patients who had experienced one or more social impediments in relation to their heart disease. SF-36 and TAAQOL scale scores of both groups were compared with each other and with the scores from the general population sample. This was done by means of analysis of variance with posthoc analysis (Tuckey HSD) to obtain the significance levels of the differences between the three groups. All three groups were included in every analysis.

Respondents

One hundred and ten patients with minor congenital heart disease agreed to participate in the study (73%). Ninety-four patients completed at least one of the two questionnaires (64%). The age of the patients ranged from 15 to 30 years, with a mean age of 22 years and 3 months. The percentage of women in the sample was 58%.

The group of 94 congenital heart disease patients contained patients with the following diagnoses: Ventricular septal defect (VSD) (44); Aortic stenosis (AoS)/ insufficiency/ aortic valve anomalies (20); Atrial septal defect of secundum type (ASDII) (21); Pulmonary stenosis (PS) (4); and combined anomalies (PS/ AoS, VSD/ PS, VSD/ AoS) (5).

Response rate for the general population sample was 63% (n= 1771). From this sample only the respondents within the age range of the congenital heart disease group were included in the analysis (15 to 30 yr.). The mean age of this group (n= 350) was 23 years and 10 months. The percentage of women in this sample was 57%.

Results

Social impediments

In the QOL-CHD, the congenital heart disease patients were asked if they had experienced any social impediments in different aspects of their daily life. In Table 1 the social impediments indicated by the respondents are presented.

Although no reference data are available several topics deserve attention because of the high percentage of social impediments mentioned: at school (19%), and during free time activities (15%). Several older respondents experience drawbacks in choosing (13%) or performing a job (9%), different kinds of medical examinations in relation to getting a job or insurance (19%), and taking out a life insurance policy (8%).

Table 1 Congenital heart disease- related social impediments experienced by patients in the past and present

		Experienced social impediments	
Social impediments	**n**	**Yes (n)**	**%**
At school	93	18	19
Free time	93	14	15
Attending lessons	93	9	10
At home	93	7	8
Contact with peers	93	7	8
Going on holiday	93	7	8
Items relevant for patients older than 18 years			
Medical examination for job or insurance	78	15	19
Choice of career	78	10	13
Performing job	78	7	9
Taking out a life-insurance policy	78	6	8
Selection procedure for job	78	6	8
Getting a job	78	4	5
Getting driver's license	78	4	5

The QOL-CHD also addressed questions concerning social impediments in relation to their heart disease which patients possibly could expect in the future. These questions regarded choice of school, choice of profession, career, daily life, sports, free time activities, living on your own and family planning. Patients could indicate in which field they expected social impediments in the future. The results are presented in Table 2.

Table 2 shows that a substantial number of the congenital heart disease patients expect limitations in relation to their heart disease in the field of sports (26%), and to a lesser degree with regard to career (11%) and family planning (10%).

The respondents were also asked if there were any social impediments that were imposed by a physician in relation to their cardiac condition. These results are presented in Table 3.

Several respondents mentioned imposed social impediments with regard to daily life (8%), choice of career (5%) and sports (23%). Many of those impediments are unnecessarily imposed. For the group of mild congenital heart disease patients involved in this study only the subgroup of patients with aortic stenosis should be advised not to participate in intensive sports that require heavy exercise (like power-lifting). Of those 21 patients that are limited in (intense) sports only 12 are diagnosed with aortic stenosis. The other nine patients have other diagnoses (mainly VSD) that do not require any limitation on the exercise of sports.

Also the limitations in free time activities like diving or going to the sauna are unnecessarily imposed because these activities are allowed for all patients with mild congenital heart disease involved in the study.

Table 2 Congenital heart disease- related social impediments that patients expect for the future

		Social impediments expected by the patient	
Possible social impediments	**n**	**Yes (n)**	**%**
Sports	93	24	26
Career	93	10	11
Family planning	93	9	10
In daily life	93	6	6
Choice of profession	93	5	5
Free time	93	5	5
Living on your own	93	2	2
Choice of school	93	1	1

Table 3 Social impediments imposed by the patient's doctor (n= 93)

Field of social impediments	Imposed social impediments in this field		Specified impediments (n)
	Yes (n)	%	
Sports	21	23	No sports (1), no intense sports (9), no top-class sport (7), no competitive sports (3)
Daily life	7	8	No coffee or cigarettes (1), no heavy exercise (lifting) (3), careful with infections/wounds (2)
Choice of career	5	5	Intense physical profession is not advised (pilot/nurse/stock-broker/building-worker) (5)
Free time activities	4	4	No diving (2), sauna (1), lifting (1)
At school	1	1	No intense exercise with physical education (1)
Social life	1	1	-

Health status

To evaluate the differences in health status of the congenital heart disease patients with and without social impediments in comparison with each other and with the general population, the SF-36 scale scores of these three groups were considered. The SF-36 scale scores for each of these groups are shown in Table 4.

As can be seen in Table 4, the group of congenital heart disease patients with no impediments reports a health status that is comparable to the general population for most scales of the SF-36. For the scale 'bodily pain' this congenital heart disease group even indicates a better score. The other group of congenital heart disease patients, those with social impediments, reports a similar health status for 6 of the 8 SF-36 scales when compared with both the general population or with the congenital heart disease group with no impediments. However, for the two scales 'physical function' and 'general health' the patients with social impediments report a worse health in comparison with both the general population and the congenital heart disease group without impediments ($p < 0.05$).

Table 4 Mean SF-36 scale scores for adolescents with congenital heart disease with and without social impediments and for the general population, 95% confidence intervals for difference (95% CI), and P-value of ANOVA differences of mean Higher scores indicate better health status

Scale	Group 1 Mean CHD-patients without social impediments n= 44	Group 2 Mean CHD-patients with social impediments n= 43	Group 3 Mean general population n= 350	95% CI difference group 1 and group 2	95% CI difference group 1 and group 3	95% CI difference group 2 and group 3	group 1 vs. group 2 p-value	group 1 vs. group 3 p-value	group 2 vs. group 3 p-value
Physical function	95.9	88.9	93.9	1.5 – 12.4	-2.1 – 6.1	-9.1 –(-0.8)	.01	.49	.01
Role physical	92.0	90.5	88.3	-11.0 – 14.2	-5.6 – 13.1	-7.3 – 11.8	.95	.61	.85
Bodily pain	89.4	82.2	82.2	-1.8 – 16.3	0.5 – 14.0	-6.8 – 6.8	.15	.03	1.00
General health	84.2	71.4	79.2	4.1 – 21.6	-1.5 – 11.5	-14.4 –(-1.2)	.01	.17	.02
Vitality	73.9	66.6	71.6	-0.8 – 15.3	-3.7 – 8.3	-11.0 – 1.1	.09	.63	.14
Social function	94.3	86.9	89.5	-1.1 – 15.9	-1.5 – 11.1	-9.0 – 3.8	.10	.17	.61
Role emotional	85.6	86.5	85.7	-15.7 – 13.9	-11.1 – 10.8	-10.4 – 12.0	99	1.00	.99
Mental health	82.1	77.7	79.5	-3.0 – 11.8	-2.9 – 8.1	-7.4 – 3.8	.35	.62	.72

CHD= congenital heart disease

Health-related quality of life

To evaluate the differences in health-related quality of life of the congenital heart disease patients with and without social impediments in comparison with each other and with the general population, the TAAQOL scale scores of these three groups were considered. The TAAQOL scale scores for each of these groups are shown in Table 5.

It is quite remarkable that the congenital heart disease patients without social impediments report a better health-related quality of life than the general population for a substantial number of scales: 'social functioning', 'daily activities', 'vitality' and 'depressive moods' ($p < 0.05$); and for the scales 'pain' and 'sexuality' ($p = 0.05$) (Table 5). The congenital heart disease patients with social impediments report a health-related quality of life equal to that of the general population for most scales. Only for the scale 'vitality' do the congenital heart disease patients with social impediments report a worse health-related quality of life than the general population ($p < 0.05$). When both congenital heart disease groups are compared with each other, the group with no social impediments reports a better health-related quality of life for the scales 'pain', 'vitality' and 'depressive moods' ($p < 0.05$). For the other 9 scales there are no significant differences between these two groups.

Discussion

The first objective of this study was to determine which social impediments were and are experienced by people with minor congenital heart disease in relation to their heart disease. The results of the QOL-CHD questionnaire showed that they experienced a lot of social impediments in daily life. Almost one out of every five patients with minor congenital heart disease has experienced social impediments at school in relation with their heart disease. One out of every eight patients experienced social impediments with the choice of a career, and one out of every five patients experienced social impediments in passing medical examinations for job qualification or insurance applications. Other studies confirm the results that people with congenital heart disease find obstacles in obtaining employment, health insurance and life insurance.[6,26,27]

Table 5 Mean health-related quality of life scores (TAAQOL) for adolescents with congenital heart disease with and without social impediments and for the general population, 95% confidence intervals for difference (95% CI), and P-value of ANOVA differences of mean
Higher scores indicate better quality of life

Scale	Group 1 Mean CHD-patients without social impediments n= 44	Group 2 Mean CHD-patients with social impediments n= 43	Group 3 Mean general population n= 350	95% CI difference group 1 and group 2	95% CI difference group 1 and group 3	95% CI difference group 2 and group 3	group 1 vs. group 2 p-value	group 1 vs. group 3 p-value	group 2 vs. group 3 p-value
Gross motor	97.0	92.6	93.7	-2.4 – 11.2	-1.8 – 8.4	-6.3 – 4.0	.28	.28	.86
Fine motor	100	99.0	97.9	-2.4 – 4.4	-0.4 – 4.7	-1.5 – 3.7	.77	.13	.58
Pain	88.1	78.1	81.0	0.3 – 19.4	0.0 – 14.2	-10.1 – 4.4	.04	.05	.63
Sleeping	85.1	77.1	79.5	-3.0 – 19.0	-2.1 – 14.2	-10.3 – 6.4	.20	.19	.84
Cognitive functioning	91.6	82.1	84.0	-0.9 – 19.9	-0.1 – 15.4	-9.7 – 6.0	.08	.06	.85
Social functioning	96.0	90.7	89.0	-2.6 – 13.3	1.1 – 12.9	-4.3 – 7.7	.26	.02	.79
Daily activities	95.0	90.7	84.1	-5.8 – 14.4	3.4 – 18.4	-1.0 – 14.2	.58	.01	.10
Sexuality	98.0	92.4	90.9	-4.1 – 15.3	0.0 – 14.2	-5.8 – 8.8	.37	.05	.88
Vitality	75.8	59.2	68.0	6.2 – 26.8	0.1 – 15.5	-16.5 –(-1.0)	.01	.04	.02
Happiness	74.4	70.9	73.3	-5.9 – 13.0	-5.8 – 8.3	-9.5 – 4.8	.65	.91	.73
Depressive moods	88.2	78.9	81.5	1.1 – 17.5	0.6 – 12.8	-8.8 – 3.6	.02	.03	.59
Aggressiveness	90.9	82.7	86.5	-0.4 – 16.9	-2.0 – 10.9	-10.3– 2.7	.07	.24	.36

CHD= congenital heart disease

Concerning the social impediments that patients expect for the future, one out of every four patients expects limitations in sports, one out of every 10 patients expects being limited in their career or family planning.

A substantial number of patients mentioned that their physician imposed social impediments, especially with regard to sports. Many of these social impediments are unnecessarily imposed. The group of patients with mild congenital heart disease contains several subgroups with different diagnoses. Only one group, those patients with aortic stenosis should be advised not to participate in intensive sports that require heavy exercise. However, the results indicated that many patients with other diagnoses were unnecessarily limited by their physician, especially in the field of sports. Some other free time activities like diving or going to the sauna were also unnecessarily imposed because these activities are allowed for all patients with mild congenital heart disease involved in the study.

The second objective was to determine health status and health-related quality of life of people with minor congenital heart disease in comparison with the general population. The sample of congenital heart disease patients was divided into two groups: the patients that experienced no social impediments in relation to their congenital heart disease, and those who experienced one or more social impediments in relation to their congenital heart disease. Both congenital heart disease groups were also compared with each other.

The SF-36 was used to measure health status, and the TAAQOL to assess health-related quality of life. The congenital heart disease patients with social impediments reported no differences for most of the SF-36 scales when compared with the general population, but they indicated less optimal scores for the scales 'physical function' and 'general health'. However, these differences are small. These patients also reported no differences for most health-related quality of life-scales in comparison with the general population. Other studies have shown that people with more severe heart diseases have a less good physical condition than the general population.[10,28] In our study, the congenital heart disease patients that experienced social impediments reported a less good health status than the general population for two of the eight scales, i.e. 'physical function' and 'general health'.

These results conform to the results of studies with patients with more severe congenital heart disease. However, these differences in health status are only partially manifested in a less good health-related quality of life for this group, i.e. the scale 'vitality'. For the other health-related quality of life-scales there is no difference with the general population.

When we look at the other group of congenital heart disease patients, those who did not experience social impediments, we see a different image. This group of congenital heart disease patients reported a health status comparable with the general population. However, this group of congenital heart disease patients also reported a better health-related quality of life on 6 out of the 12 TAAQOL-scales in comparison with the general population. These differences are puzzling. Health-related quality of life was measured by using the TAAQOL, focusing mainly on the subjective appraisal of health status. An explanation could be that patients with minor congenital heart disease who experience no social impediments feel relieved because of their rather good health status and therefore appreciate their health more positively. This could result in the higher appreciation of their functioning regarding 'social functioning', 'daily activities', 'sexuality', 'pain', 'vitality' and 'depressive moods'. Møyen Laane et al. also found a trend for a higher subjective experience of quality of life for different subgroups of children with congenital heart disease.[29] They suggest that this may represent development of coping mechanisms and revision of values of life. Other researchers suggest that people with congenital heart disease want to prove that they can overcome the limitations of their heart disease and perform as good, or even better than people with a completely normal heart.[30] Several studies indicate that children and adolescents with congenital heart disease perform better at school and reach a higher educational level than people from the general population.[6,31]

From our study it is concluded that minor congenital heart disease generally does not influence health-related quality of life negatively. Considering the results on health status it can be concluded that minor congenital heart disease does not influence health status for most patients with mild congenital heart disease. These results could be used when educating patients with minor congenital heart disease and their parents about their prognosis. Most children and young adults with minor congenital heart disease can have a life which is not bothered by a poor health status or poor health-related quality of life. However, when entering adulthood they may find some obstacles from society in their way.[32] The education for these patients should be

directed towards overcoming these obstacles. However, it is also important to inform physicians about the social impediments for patients with different diagnosis of minor congenital heart disease. Too many patients with a congenital heart disease diagnosis that requires no social impediments are limited in performing sports by their physician. Furthermore, information should be directed towards institutions like insurance-companies and employers that no additional health risks are to be expected for this category of individuals with congenital heart disease.

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Chapter 4

Health-related quality of life and health status in adult survivors with previously operated complex congenital heart disease

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Abstract

Objective: To examine the impact of previously operated complex congenital heart disease on health-related quality of life and subjective health status and to determine the relation between these parameters and physical status.
Design: Cross sectional; information on medical follow-up was sought retrospectively.
Setting: Patients were randomly selected from the archives of the paediatric cardiology department, Leiden University Medical Center, Leiden, The Netherlands, and approached irrespective of current cardiac care or hospital of follow up.
Patients: Seventy eight patients with previously operated complex congenital heart disease (now aged 18 to 32 years) were compared with the general population.
Main outcome measures: Health-related quality of life was determined with a specifically developed questionnaire (TNO-AZL Adult Quality of Life (TAAQOL)) and subjective health status was assessed with the 36 item short form health survey (SF-36). Physical status was determined with the objective physical index, Somerville index and New York Heart Association functional class.
Results: Health-related quality of life of the patients was significantly worse than that of the general population in the dimensions gross motor functioning, and vitality ($p < 0.01$). Correlations between health-related quality of life and physical status were poor. Patients had significantly worse subjective health status than the general population in the dimensions physical functioning, role functioning physical, vitality, and general health perceptions ($p < 0.01$). Correlations between subjective health status and physical indices were weak.
Conclusion: Adult survivors with previously operated complex congenital heart disease experienced limitations only in the physical dimensions of health-related quality of life and subjective health status. Objectively measured medical variables were only weakly related to health-related quality of life. These results indicate that, when evaluating health-related quality of life, dedicated questionnaires such as the TAAQOL should be used.

Introduction

Advances in surgery and intensive care have increased the life expectancy of children with complex congenital heart disease. Therefore, it is important to assess the long-term results of treatment of these patients, not only in medical terms (for example, morbidity and mortality) but also in terms of health-related quality of life and subjective health status. Since a longer life does not necessarily mean a better life, knowledge of the emotional response to health status problems, may allow identification of areas of daily life that need to be improved (for example, by medical care or by provisions in social or infrastructural care management). Moreover, patients' satisfaction with the medical consultation may increase if physicians assess a patient's general wellbeing, as well as functional status.[1]

Some studies have explored the subjective health status of patients with congenital heart disease in adulthood[2-10] but none have examined health-related quality of life using a clear definition. In addition, correlating health-related quality of life with the patient's physical condition may increase our understanding of the concept of health-related quality of life.[11]

Therefore, in a group of patients with previously operated complex congenital heart disease, the aim of the study was, firstly, to compare patients' health-related quality of life and subjective health status with those of the general Dutch population (age and sex matched) and, secondly to relate health-related quality of life and subjective health status to physical status.

Methods

The study was approved by the local medical ethical committee and informed consent was obtained from each participant. A cross sectional design was used and information on medical follow up was searched for retrospectively.

Definitions

Health-related quality of life

Although there is no universal definition for health-related quality of life, there is growing consensus that it should be assessed multidimensionally, including physical health, psychological state, and social relationships.[12-15] Moreover, quality of life and health status are distinct constructs[12,16,17]; quality of life is determined not only by the patient's health status problems but also by the emotional response to these problems. To measure quality of life according to this definition, quality of life questionnaires have been developed for children (TNO-AZL Child Quality Of Life (TACQOL)- questionnaire[14,18]) and for adults (TNO-AZL Adult Quality of Life (TAAQOL)-questionnaire[19]). Both questionnaires have been validated in the general population as well as in patients with various chronic diseases, such as minor congenital heart disease.[14,18-21] Figure 1 shows an example of the format of the questionnaires.

Health-related quality of life is used since non-health factors such as income or housing (which may also contribute to quality of life) were not taken into account. For clinicians *health-related* quality of life is most relevant.

Figure 1 Example of the format of the health-related quality of life questionnaire: TNO-AZL (adult/child) Quality Of Life (TAxQOL)

Did you have difficulty in the last month with ....

walking up the stairs? ❑ no ❑ a little ❑ some ❑ a lot

How much did that bother you?

❑ not at all ❑ a little ❑ quite a lot ❑ very much

The 36 item short form health survey (SF-36) questionnaire is widely used and thoroughly validated.[22,23] It measures subjective health status, posing such questions as "What are your limitations?" or "Do you feel healthy?" in contrast to "How do you feel about certain limitations?" in the TAAQOL.

Diagnosis

In the present study, patients with previously operated complex congenital heart disease were approached, excluding those who had undergone anatomical corrective surgery. All patients included in this study had undergone a non-anatomical correction- that is, partial or complete cavopulmonary connection, venous switch for transposition of the great arteries, or a correction with the use of allogenous tissue (for example, Rastelli correction, insertion of a mechanical valve). The main common characteristic was that postoperatively none of the participants had an anatomically normal heart as described by the segmental analysis,[24] as opposed to patients after anatomical corrective surgery (for example, operated ventricular or atrial septal defects, coarctation of the aorta, patent ductus arteriosus, uncomplicated tetralogy of Fallot, total anomalous pulmonary venous connection, and uncomplicated pulmonary or aortic stenosis).

Patient selection

To prevent selection bias, patients were selected from the archives of the department of paediatric cardiology, Leiden University Medical Center, Leiden, the Netherlands, which has complete information on patients seen from 1950 onwards. From these files, 4383 patients were born between 1968 and 1982. Of these, in the year 1999, 2280 were randomly selected. Then, 251 patients with operated complex congenital heart disease were included and further studied irrespective of current cardiac surveillance.

The latest medical information and home addresses were obtained from the most recent medical file, general practitioners or local authorities.

Among the 251 included patients, 123 (49%) had died. Others (n= 41) were excluded for the following reasons: they were not living in the Netherlands or did not speak Dutch n= 28 (11%), they had learning disabilities n= 12 (5%), or they had participated in a pilot study n= 1 (0.5%). One patient was lost to follow up. The remaining 86 patients were invited to participate in the study; of these, 78 patients agreed to participate (91%). Table 1 lists the diagnoses and gives the number of patients per diagnosis or operation.

Table 1 Diagnosis of 78 patients previously operated on for complex congenital heart disease (numbers of patients is given in parentheses)

Main diagnosis	Main operation
1. systemic right ventricle (39)	
TGA (22)	Senning (12) or Mustard (10)
TGA + PS (5)	Senning (3) or Mustard (2) with PS repair
TGA + VSD + PS (6)	Senning (5) or Mustard (1) with VSD closure and PS repair
L-TGA + VSD + PS (2)	VSD closure with PS repair (2)
L-TGA + VSD + ASD (1)	VSD closure and ASD closure (1)
L-TGA + VSD (1)	VSD closure (1)
L-TGA + PS (1)	Tricuspid valve replacement (1)
AV-discordance + DORV + PS (1)	Tricuspid valve replacement and conduit (1)
2. conduit or mechanical prosthesis (23)	
PA + VSD (11)	Rastelli (3); correction with other conduit (8)
AoS (4); AVSD (1); MI (1);	Mechanical prosthesis (6)
TGA + VSD + PS (4)	Rastelli (4)
TGA + VSD (1)	ASO with conduit (1)
TrArt (1)	Correction with conduit (1)
3. univentricular atrioventricular connection (11)	
TA (8); DILV (2); hypoplastic LV (1)	Fontan (11)
4. palliative operation- shunt (5)	
AVSD (1)	Glenn shunt (1)
DILV (1)	Blalock-Taussig shunt (1)
Univentricular heart (1)	Modified Blalock-Taussig shunt (1)
TA (1)	After Glenn shunt: AV shunt axillary (1)
TA + TGA (1)	After banding AP: PH (1)

AoS= aortic stenosis; AP= pulmonary artery; ASD= atrial septal defect; ASO= arterial switch operation; AV= atrioventricular; AVSD= atrioventricular septal defect; CHD= congenital heart disease; DILV= double inlet left ventricle; DORV= double outlet right ventricle; L-TGA= congenitally corrected TGA; LV= left ventricle; MI= mitral insufficiency; PA= pulmonary atresia; PH= pulmonary hypertension, PS= pulmonary stenosis; TA= tricuspid atresia; TGA= simple transposition of the great arteries; TrArt= common truncus arteriosus; VSD = ventricular septal defect

Measurements

Questionnaires on health-related quality of life and subjective health status were completed.

Health-related quality of life

The TAAQOL is a Dutch questionnaire developed by TNO and the Leiden University Medical Center. Psychometric performance (reliability and validity) of the TAAQOL is satisfactory.[19] It consists of 45 questions, divided into 12 domains. Each domain contains two to four questions (the actual number per domain is given in parentheses): gross motor functioning (4), fine motor functioning (4), pain (4), sleeping (4), cognitive functioning (4), social functioning (4), daily activities (4), sexual activity (2), vitality (4), happiness (4), depressive moods (4) and aggressiveness (3). For each item, the frequency of occurrence of a health status problem is assessed. If such a problem is reported, the emotional reaction to this problem is also determined. Figure 1 shows an example of the format of the question. The reference period is formulated as "the last month". The two questions of the domain sexual activity were not appropriate for the present study since the occurrence of problems was compared with one month previously ("Were you less sexually active than a month ago?"). Because such questions are only relevant when an intervention has taken place, this domain was not considered. Scores of each subscale are normalised to a scale ranging from 0 to 100, with higher scores representing better quality of life.[19]

Subjective health status

The SF-36 was used to measure subjective health status.[22] This questionnaire is based on 35 items divided into eight domains (the number of questions per domain is indicated in parentheses): physical functioning (10), role functioning physical (4), bodily pain (2), general health perceptions (5), vitality (4), social functioning (2), role functioning emotional (3) and mental health (5). Scores range from 0 to 100, with higher scores indicating better subjective health status.

TAAQOL and SF-36 data of the Dutch general population are available.[19,22]

Physical status

Medical data were collected from the medical records. If clinical information was older than one year, patients were re-examined at the Leiden University Medical Center (by MK and JO or HV).

Physical status was scored by means of three indices determined from these data. Firstly, to measure objectively the severity of the congenital heart disease, the presence of cyanosis, arrhythmia, cardiac failure and residual defects was combined into an "objective physical index". Because each variable counted for one point, a score of 0 to 4 was possible, with a higher score indicating a more serious cardiac condition. Secondly, the index of Somerville[25] and thirdly, the New York Heart Association (NYHA) functional class[26] were determined (Table 2). In both, a higher score indicates more complaints or limitations.

Table 2 Ability index of Somerville and the New York Heart Association (NYHA) functional class for patients with cardiac disease

Somerville index

1. Normal life: full time work or school, pregnancy manageable.
2. Able to work (part time): interference with life (socio/community imposition).
3. Unable to work: limitations on all activities, pregnancy risk.
4. Extreme limitation: dependent, almost housebound.

NYHA classes

I. No limitations of physical activities. Ordinary activity does not cause undue anginal pain, dyspnoea, fatigue, or palpitations.

II. Slight limitations of physical activities. Comfortable at rest, but ordinary activity results in anginal pain, dyspnoea, fatigue, or palpitations.

III. Marked limitations of physical activities. Comfortable at rest, less than ordinary activity causes anginal pain, dyspnoea, fatigue, or palpitations.

IV. Unable to carry on any physical activities without discomfort. Symptoms of cardiac insufficiency or the anginal syndrome may be present even at rest. If any physical activity is undertaken discomfort is increased.

Statistical analyses

Sample size was based on results of the pilot study[19] in which a mean difference of 11 units (on the 0 to 100 scale) was found between patients and the general population on the SF-36 vitality scale. Similar effect sizes are reported in the American SF-36 manual for similar conditions. The sample size (n= 80) was chosen to have 80% power for such a difference. With this sample size there is at least an 80% power to detect a correlation coefficient of 0.30 or more.

The participants' health-related quality of life (as measured by the TAAQOL) and their subjective health status (as measured by the SF-36) were compared with those of a random sample of the general Dutch population using

analysis of variance with adjustment for age and sex. The sample of the general Dutch population comprised 831 persons for the TAAQOL and 361 for the SF-36.[19] After comparison, the data were summarised as mean values and 95% confidence intervals.

Within the group of patients, the overall association between health-related quality of life and subjective health status on the one hand and the objective physical index, the Somerville index, and the NYHA class on the other was quantified using canonical correlation analysis to reduce the number of statistical tests. With this method, the maximum association between two sets of variables is quantified.[27] When the canonical correlation was significant, pairwise Spearman rank correlations were used to quantify the association between the various indices.

Since the distribution of a number of these variables was skewed, all statistical analyses were performed on rank transformed data. A probability value of $p \leq 0.01$ was considered significant because many statistical tests were done. SPSS for Windows version 10.07 (SPSS Inc, Chicago, Illinois, USA), was used to perform statistical calculations.

Results

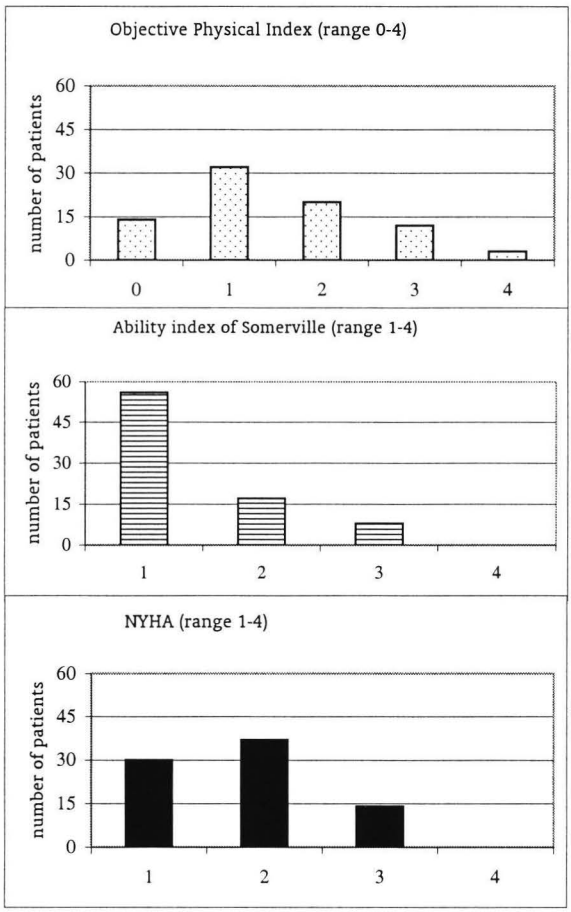
Patient characteristics

There was an overall response rate of 91%. Medical data were available for all 78 participants and questionnaires for 76 of them. There were 44 men and 34 women, with an average age of 24.3 (range 18 to 32) years. At the time of the study, 46 participants had a job for more than 12 h/week (59%; two patients were receiving a partial disability benefit), 15 were receiving a full or partial disability benefit (19%), 11 were still at school (14%), three were housewives (4%), and three (4%) were on sick leave or unemployed. Participants lived with a partner n= 34 (43%), with their parents n= 31 (40%), had their own house n= 7 (9%) or lived in a student (or other) house n= 6 (8%).

For n= 47 (60%) of the participants the main surgery had been performed before the patient was three years old. Fifty participants (64%) had undergone a previous palliative procedure (such as Rashkind septostomy or banding of the pulmonary artery). The mean age at first operation or intervention was 2.2 (range 0 to 24) years; 45 patients (58%) had had an intervention soon after birth.

Figure 2 shows the physical status of the patients. Mean score for the objective physical index is 1.48, for the Somerville index 1.41, and for the NYHA class 1.8. Most patients scored 1 in the objective physical index n= 31 (40%), indicating that they had at least one of the physical complications. According to the Somerville index, n= 9 (11%) of the participants were unable to work and were limited in all activities (score 3). For the NYHA, n= 36 (46%) of the participants were in class II and were thus experiencing slight limitations in physical activities.

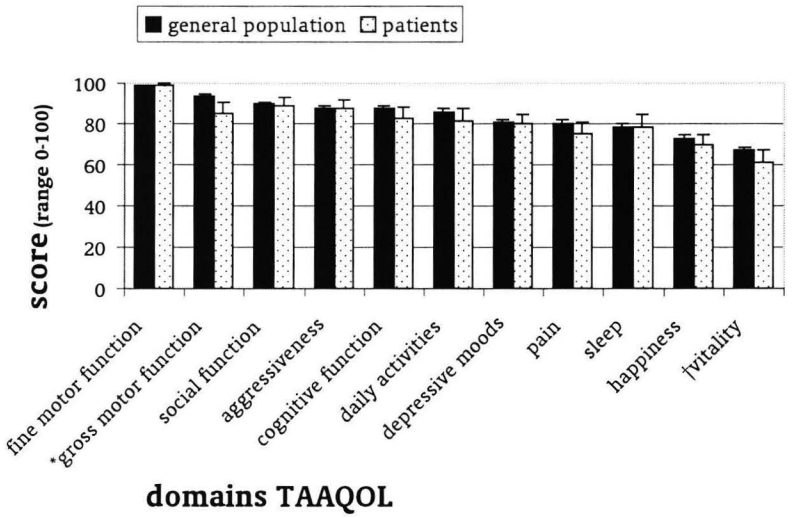
Figure 2 Outcome of physical status in the three indices (n= 78)



Health-related quality of life: patients compared with the general population

Figure 3 gives the average TAAQOL scores of the patients and of the general population sample. The patients had lower health-related quality of life in almost all dimensions and scored significantly lower on gross motor functioning ($p < 0.001$) and vitality ($p < 0.01$).

Figure 3 Health-related quality of life (TAAQOL) in 76 patients with previously operated complex congenital heart disease is compared with that of the general population (n= 831): mean scores and 95% confidence interval *$p < 0.001$ †$p < 0.01$



Health-related quality of life related to physical status: within the patient group

Several health-related quality of life dimensions correlated significantly with the objective physical index, the Somerville index and the NYHA class (Table 3). Canonical correlation analysis showed that the first canonical correlation was significant ($r = 0.768$); in particular the NYHA class was the most important correlate of gross motor functioning. The pairwise Spearman correlations (Table 3) showed the following pattern: health-related quality of life correlated weakly with the objective physical index but significantly for gross motor functioning and cognitive functioning. The strongest relations are between the TAAQOL dimensions and the NYHA class.

Table 3 Spearman's *r* correlation for medical indices correlated with the TAAQOL and the SF-36 for 76 patients previously operated on for complex congenital heart disease *p< 0.001 †p< 0.01

Questionnaire	Objective physical index	Somerville index	NYHA class
TAAQOL			
Gross motor function	-0.33†	-0.53*	-0.77*
Daily activities	-0.23	-0.46*	-0.45*
Vitality	-0.12	-0.42*	-0.47*
Sleep	-0.25	-0.38*	-0.43*
Social functioning	-0.10	-0.30†	-0.35†
Pain	-0.03	-0.24	-0.36*
Cognitive functioning	-0.30†	-0.33†	-0.32†
Depressive moods	-0.17	-0.24	-0.31†
Fine motor function	-0.18	-0.22	-0.29†
Happiness	-0.19	-0.06	-0.22
Aggressiveness	-0.03	-0.15	-0.19
SF-36			
Physical functioning	-0.42*	-0.64*	-0.87*
Mental health	-0.36*	-0.16	-0.2
Social functioning	-0.24	-0.54*	-0.45*
Role function physical	-0.28	-0.48*	-0.42*
General health	-0.25	-0.41*	-0.47*
Vitality	-0.25	-0.31†	-0.45*
Bodily pain	-0.22	-0.32†	-0.45*
Role function emotion	-0.24	-0.20	-0.22

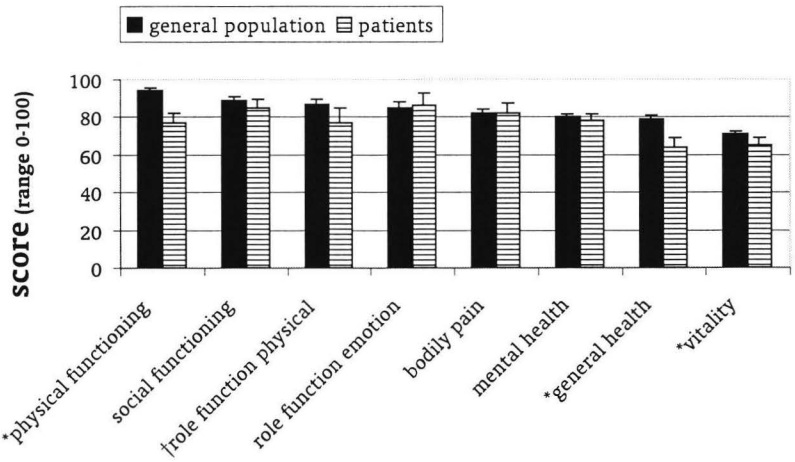
Subjective health status: patients compared with the general population

The SF-36 dimensions role functioning physical ($p< 0.01$), physical functioning, vitality and general health perception ($p< 0.001$) were significantly lower in patients than in the general population (Figure 4).

Subjective health status related to physical status: within the patient group

Almost all SF-36 dimensions correlated significantly with the three physical indices (Table 3). Canonical correlation analysis showed that the NYHA class was the strongest correlate of the SF-36 dimensions and of physical functioning in particular (r= 0.864). The patterns for pairwise Spearman correlations were the same as those for health-related quality of life, with the strongest relations between subjective health status dimensions and the NYHA class (Table 3).

Figure 4 Subjective health status (SF-36) in 76 patients with previously operated complex congenital heart disease is compared with that of the general population (n= 361): mean scores and 95% confidence interval *p<0.001 †p<0.01



Discussion

This study evaluated health-related quality of life and subjective health status in adults with previously operated complex congenital heart disease and determined the relation between these parameters and physical status.

Patients compared with the general population

Comparison between the patients and the general population revealed significantly lower scores in the patient group for the physical dimensions only: gross motor functioning and vitality (in the TAAQOL) and physical functioning, role functioning physical, general health perceptions, and vitality (in the SF-36). Thus, both physical function and appraisal of physical dysfunction were significantly lower in the patient group.

There was no significant difference in any of the scores for the non-physical dimensions (social functioning, role functioning emotional, daily activities, depressive moods, etc.) between patients and the general population. Some studies on social, psychological and behavioural outcome in children and adolescents with heart disease have reported problems in these fields,[5,28-32] whereas others show normal social behaviour and successful educational and occupational achievements in this group of patients,[8,9,33-36] thus supporting the promising outcome of the present study. These latter studies indicate that, over time, patients may adapt to their disease and that denial may even help to normalise functioning.

To improve the patients' long term quality of life, physical aspects must be taken into account.

First, appropriate exercise should be encouraged to avoid patients imposing unnecessary restrictions on themselves,[37] sometimes caused when patients receive insufficient information from the specialist.[38] In addition, parents should be informed that overprotection can have a negative impact on the physical activities of the child.[39] Counselling should, therefore, be based on a thorough clinical assessment together with the use of specific recommendations.[39-41] Another way to improve physical capacity is to offer exercise training. For example, in adult male patients with chronic heart failure, specific cardiac training has been shown to improve physical capacity and quality of life,[42,43] as in healthy subjects.[44] In addition, training in children with various congenital heart diseases has been shown to improve physical abilities.[45-47] However, in patients with previously operated complex congenital heart disease the value of exercise training should be further studied, focusing on long term effects in adulthood.[48]

Prevention of postoperative complications may also avoid poor outcome of the physical dimension of health-related quality of life, but this topic is beyond the scope of the present study.

Relation to physical status

Relating physical indices to the dimensions of health-related quality of life and subjective health status showed weak correlations. The objective physical index correlated less with health-related quality of life and subjective health status than the index of Somerville and the NYHA class. This means that variables such as cyanosis, cardiac failure, or arrhythmia (the best indicators for the objective severity of the disease) do not correlate with the patients' health-related quality of life or subjective health status. This is a surprising finding, but other reports on these correlations show the same pattern: factors other than the complexity of the disease (e.g. parental attitudes and perceptions, mother's concern, and social support) play a more important role in psychosocial outcome.[9,33,49,50]

Correlations between all the quality of life dimensions and the Somerville index were significantly weaker than the correlations between the quality of life dimensions and the NYHA class. Thus, although the Somerville index was specifically devised to take into account the adjustment and perception of patients with congenital heart disease,[51] its usefulness for this purpose was not confirmed in the present study.

The NYHA class correlated significantly and relevantly with the dimensions gross motor functioning (TAAQOL) and physical functioning (SF-36) (Spearman's $r= 0.77$ and $r= 0.87$, respectively). When focussing solely on these dimensions, the NYHA class can indeed provide a good estimate of health-related quality of life and subjective health status. However, when evaluating the patients' overall health-related quality of life and subjective health status, parameters other than those routinely used in a physically oriented cardiological setting should be included.

Remarks and study limitations

When measuring long term outcome of patients with previously operated congenital heart disease, mortality has to be considered. In the unbiased group of 251 selected patients, 123 (49%) had died and could therefore not participate in the study; at least nine of these latter patients were over 18 years old when they died. In addition, two of the participants died from cardiac causes during the study period. Mortality in the age group 18 to 32 years is, therefore, at least 8.1% (that is, 11 of 135 patients died). Although the

study found good health-related quality of life in many aspects, the considerable risk of dying after the age of 18 years has to be taken into account. In addition, studying only surviving patients implies a positive selection bias.

The overall response in this study was 91%. There was no significant difference between the participants and non-participants for sex or residence. Similarly, there was no significant difference in response between the subdiagnosis groups. However, there was a significant difference between age groups. The response rate in the 24 to 32 year age group was lower than in the 18 to 24 year age group (77% and 95%, respectively; $p= 0.02$). Communication with some of the non-participants indicated that older patients may have more family or work responsibilities and thus less time to participate in a study; how this may have influenced our results remains, however, unclear.

Conclusions

Patients with previously operated complex congenital heart disease experienced limitations only in the physical dimensions of health-related quality of life and subjective health status. Objective medical variables correlated only weakly with health-related quality of life and can therefore not be used to assess health-related quality of life. Wagner and colleagues[52] reported that "the routine use of health status measures may enhance patients' care". The dedicated health-related quality of life instrument used in the present study (TAAQOL) is well validated[19] and valuable in patient care since it measures the patients' own health related feelings and problems in daily life. It provides more relevant information than just measuring functioning with subjective health status.[12,16,17] These results indicate that, when evaluating health-related quality of life, dedicated questionnaires such as the TAAQOL should be used.

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Chapter 5

The need for cardiac follow-up in adults with mild congenital heart disease

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Abstract

Objective: To discuss the need for continuing cardiac surveillance in adults with hemodynamically insignificant congenital cardiac disease.
Methods: In 82 patients with mild congenital cardiac malformations, aged from 17 through 32 years, we investigated the subjective health status, the health-related quality of life, any difficulties encountered in daily life, the regularity of follow-up, current diagnosis, and antibiotic prophylaxis.
Results: The subjective health status, and the health-related quality of life, did not differ from those of the general population. Nevertheless, patients experienced unnecessary difficulties with choice of sport, obtaining insurance cover, and education. After clinical re-evaluation, diagnosis and antibiotic regimes had to be changed in 9 patients (11%).
Conclusions: Patients with a mild congenital cardiac malformation consider themselves to be in good health. So as to fine tune the diagnosis, and update the information supplied to the patients, we suggest that at least a cardiological assessment be carried out at the age of 16 to 18 years. In this way, patients might be protected from unnecessary difficulties, such as restrictions for sport or the charging of unjustifiably high rates for insurance.

Introduction

Patients with a hemodynamically insignificant congenital cardiac malformation, for example, a small ventricular septal defect, or minimal valvar pulmonary stenosis, have no relevant physical limitations. A recent publication showed percentages for the need for follow-up beyond 16 years based on the risks for progression.[1] It seemed that, for certain malformations, follow-up is not necessary. We have now extended this study, taking into account factors other than hemodynamic data. Thus, we evaluated whether the subjective health status, and health-related quality of life, as assessed by such patients differed from those of the general population, and whether these patients experienced unnecessary difficulties in their daily life. In addition, we investigated the cardiac state with regard to regularity of cardiac follow-up, current diagnosis, and use of prophylaxis against endocarditis. This information will be helpful when considering the need for continuing cardiac surveillance, which will improve the health care provided for the individual patient.

Methods

The local medical ethical committee approved the study. All participants gave informed consent.

Selection of patients

To avoid bias in selection, patients were randomly selected from the archives of the Department of Paediatric Cardiology at Leiden University Medical Center, and approached irrespective of current cardiac surveillance (Table 1).[2] In total, we selected randomly 249 patients, aged from 17 to 32 years, with mild congenital cardiac malformations, including those with a spontaneously resolved malformation. An operation or intervention had not been necessary in any patient, as the malformation was accepted as "hemodynamically insignificant". Exclusion criterions, with the numbers of those excluded indicated in parentheses, were participation in a previous study (43),[3] mental retardation (39), and the inability to speak Dutch or residence outside the Netherlands (18). We found that five patients had died from non-cardiac causes, while six patients were lost to follow-up. The final sample of 138 patients was invited to participate in the study.

Questionnaire

All patients completed a questionnaire, encompassing the following topics:

Subjective health status: measured with the widely used Short Form-36.[3-5] Scores for the 8 domains, namely physical functioning, general perceptions of health, physical role functioning, bodily pain, vitality, social functioning, emotional role functioning, and mental health, covered by 35 items, range from 0 to 100, with higher scores indicating better subjective health status.

Health-related quality of life: measured with the well validated questionnaire (the TAAQOL questionnaire) devised by the Netherlands Organization for Applied Scienctific Research together with the Academic Hospital of Leiden, the latter now known as the Leiden University Medical Center.[3,5,6] Scores for the 11 domains, namely gross motor functioning, fine motor functioning, pain, sleeping, cognitive functioning, social functioning, daily activities, vitality, happiness, depressive moods, and aggressiveness, covered by 43 items, ranged from 0 to 100, with higher scores again representing better quality of life.

Health-related quality of life, and subjective health status, are distinct concepts, since health-related quality of life involves the appraisal of limitations in the subjective health status.[5,7-10] For both questionnaires, comparative data are available from the Dutch general population.[3,4,6]

Difficulties in daily life: measured with the 'difficulties in daily life' questionnaire, which contains 26 items, and has been described previously.[2]

Cardiac state

Data regarding medical follow-up and current diagnosis were collected from most recent medical files. Participants whose latest cross-sectional colour echocardiogram had been performed more than one year ago were re-examined at the Leiden University Medical Center. In addition, the use of endocarditis prophylaxis was evaluated.

Statistics

Sample size was based on the results of our previous study.[3,5]

The subjective health status, as assessed using Short Form-36, and health-related quality of life assessed using the Dutch questionnaire, were compared with those of a random sample of the general Dutch population, using analysis of variance adjusted for age and gender. Since the distribution of a number of these variables was skewed, analyses were performed on rank-transformed data. The sample of the general Dutch population comprised 361 subjects who had completed Short Form-36, and 831 who had filled in the quality of life questionnaire for adults.[3] After comparison, data were summarised using mean values and 95% confidence intervals.

A probability value of $p \leq 0.01$ was considered significant.

Results

Patients (Table 1)

Of 138 invited patients, 82 agreed to participate in the study, giving a response of 59%. Of the patients, 31 were male. The overall age ranged from 17 to 32 years, with a mean of 24.6 years.

Table 1 Diagnosis after re-examination, and cardiac follow-up in 82 adults with a mild congenital heart lesion

		Cardiac follow-up			
Diagnosis after re-examination		**Withdrawn**	**Discharged**		**Actual**
A. *Persisting lesions*				change[1]	
Ventricular septal defect	20	7	4	1[1]	8
Pulmonary stenosis	7	2	1	-	4
Atrial septal defect	6	2	1	3[1]	-
Aortic stenosis	6	2	-	-	4
Bifoliate aortic valve	3	1	-	1[1]	1
Abnormal pulmonary venous drainage	2	1	1	-	-
Mitral valvar prolapse	1	-	-	-	1
Total	45	15	7	5[1]	18
		change[2]		change[2]	
B. *Spontaneous resolution*	37	4[2]	30	3[2]	-

[1] in 5 patients the malformation appeared not to have been resolved at cardiac re-examination (Table 3); [2] in 7 patients the malformation appeared to have been resolved at cardiac re-examination (Table 3)

Subjective health status, and health-related quality of life

These factors did not differ significantly from those of the general population.

Difficulties in daily life (Table 2)

In total, 9 patients (11%, 95% confidence interval 5-20%) experienced difficulty in at least one of the items.[2] Four of these patients had withdrawn from follow-up, 4 were still under cardiac care, and 1 had been discharged.

Table 2 Difficulties in daily life related to the congenital cardiac malformation in 80 adults with a mild congenital heart lesion

Items	n	(%)	
Did you ever feel restricted in the choice of			
a sport?	5	(6)	2 PS*, 2 VSD*, 1 AoS
a house?	1	(1)	1 AoS
Did you ever give up on a sport?	2	(3)	1 PS*, 1 VSD*
Were you ever excluded from			
an educational course?	1	(1)	1 ASD*
a job after medical examination?	1	(1)	1 AoS
sport after medical examination?	1	(1)	1 PS*
Where you ever prevented from			
taking out a health insurance policy?	3	(4)	2 PS*, 1 VSD
Did you ever had to pay higher premium than other			
people for a mortgage?	1	(1)	1 AoS
a life insurance?	2	(3)	2 AoS
a health insurance?	1	(1)	1 VSD
Total patients, who experience difficulties for at least one of the items	9	(11)	3 VSD*(2), 3 AoS, 2 PS*, 1 ASD*

*Unnecessary difficulties. Abbreviations: AoS= aortic valve stenosis, ASD= atrial septal defect, PS= pulmonary stenosis, VSD= ventricular septal defect

Cardiac state (Tables 1 and 3)

Of the participating sample, 45 patients had been discharged (55%), 19 had withdrawn from medical follow-up (23%), and 18 were still under cardiac supervision (22%). After updating medical assessment in 67 patients, of whom 26 required an additional cross-sectional colour echocardiogram, the diagnosis needed revision in 12 patients (15%). In total, the regime chosen for prophylaxis against endocarditis required revision in 9 of the 39 participants who needed such prophylaxis, either because of the change in diagnosis, or because they had forgotten about it (23%, 95% confidence interval 11-39%).

Table 3 Patients with changes of diagnosis at clinical re-evaluation and consequences for endocarditis prophylaxis

Patient	Gender	Year of birth	First diagnosis		Latest cardiac control		Withdrawn/ Discharged	Diagnosis before re-evaluation	Diagnosis after re-evaluation	Change in endocarditis regimen
			Year	Age	Year	Age				
1.	F	1969	1975	6	1975	6	D	VSD		stop
2.	F	1969	1969	0	1984	15	D	VSD		all forgotten
3.	F	1972	1976	4	1992	20	W	VSD	spontaneous	stop
4.	F	1973	1973	0	1987	12	D	VSD	closure	stop
5.	F	1976	1976	0	1987	11	W	VSD		stop
6.	F	1980	1981	1	1987	7	W	VSD		all forgotten
7.	F	1969	1984	15	1988	19	W	MVP with MI	resolved MVP	stop
8.	M	1969	1973	4	1988	19	D	resolved AS	AV bicuspid	start
9.	F	1978	1979	1	1993	15	D	closed VSD	VSD	start
10.	M	1974	1974	0	1978	4	D	closed ASD	ASD (most likely)	no change
11.	F	1980	1984	4	1988	8	D	closed ASD		no change
12.	M	1978	1983	5	1993	15	D	closed ASD		no change
13.	F	1977	1977	0	1986	9	W	PS	PS-no change	start, all
14.	M	1972	1972	0	1996	24	D	VSD	VSD-no change	forgotten

Abbreviations: AS= aortic valve stenosis, ASD= atrial septal defect, AV= aortic valve, D= discharged, MI= mitral insufficiency, MVP= mitral valvar prolaps, W= withdrawn from medical follow-up, PS= pulmonary stenosis, VSD= ventricular septal defect

Discussion

The subjective health status, and the health-related quality of life, in our adults with mild congenital cardiac malformations did not differ significantly from these values as measured in the general population. Previous studies on quality of life in patients with a mild defect had similarly shown no difference, or even a better quality of life, when compared with the general

population.[3,11,12] It has been suggested that this might represent mechanisms for coping, and revision of values of life.

Despite this overall good result, however, nine patients (11%) had experienced at least one difficulty in their daily life because of their cardiac disease, mainly involving sport or insurance. Because of the relatively young age of the patients, some items are not applicable to all, such as trying to obtain life insurance or employment. The percentage of patients with such difficulties, therefore, might increase with advancing age.

In a previous study of patients having mild congenital cardiac malformations, at least half of those who were under cardiac surveillance, and truly had a cardiac malformation, had experienced difficulties.[3] It is not surprising that these patients experience more difficulties than those discharged years ago, or those whose malformation had resolved in the meantime, the latter group being included in the present study.

Data on the extent of experiencing difficulties in daily life are not available for the general population. We judged if these difficulties encountered in our patients were legitimate, therefore, according to several guidelines.[13-16] It seemed that some difficulties could be avoided in certain patients with mild malformations. In our study, this involved 5 such patients (Table 2).

To prevent these difficulties, the medical state, and the information provided to the patients, should be updated with re-assessment.[13,15,16] Restrictions might have been imposed on the patient at an early age, since clinical outcome was unclear at that time. In other patients, advice about possible restrictions should be reinforced, since patient recall is disappointing.[17] A "health passport", which summarises information on diagnosis, operation, exercise, family, and career planning, may therefore be helpful, and might prevent patients from withdrawal from cardiac follow-up.[18]

No consensus exists among employers and insurance companies concerning their policy, largely because of lack of information about the individual malformations and their consequence.[19,20] Another means of avoiding difficulties, therefore, is to provide clear and consistent guidelines to insurance companies and employers.

With regard to the cardiac state, two-fifths of the overall group of patients had withdrawn from follow-up despite the fact that they still possessed a malformed heart. The same proportion was found in previous studies for patients with ventricular septal defect.[21,22]

At cardiac re-examination, the diagnosis needed revision in 12 patients (15%). Some of the changes could be explained by inaccurate investigations in the past due to technical limitations, but changes such as subsequent closure of a ventricular septal defect are well known. The incidence of spontaneous closure of ventricular septal defects after the age of 10 years ranges from 6% to 20%.[21,23-26] In the current study, spontaneous closure of the ventricular septal defect was registered in at least 9 of 30 patients (30%) after the age of 10 years. Following closure of the defect, these patients could stop using endocarditis prophylaxis. After re-examination, however, four additional patients needed to re-start their prophylaxis.

Our results could have been affected by bias introduced by non-response, there being a significantly higher response from women, from patients living closer to the hospital, and from those with a resolved malformation. During cardiac re-examination, several patients asked questions, or expressed worries that they lived with for several years. It is possible, therefore, that patients with relatively more difficulties or questions joined the study. This might have resulted in a shift towards a more negative outcome of health-related quality of life and difficulties in daily life. The outcome of the entire group of patients with mild malformations, therefore, might be more positive than suggested by our present study.

With regard to the need for continuing follow-up, our study showed that a selection of patients with a mild malformation considered themselves to be in good health, but experienced unnecessary difficulties. In addition, their diagnosis can change with consequences for endocarditis prophylaxis and insurance. To prevent these problems, we suggest an evaluation of these patients at least at the age of 16 to 18 years. At this age, patients can understand the implications of their cardiac malformation, and be reminded of antibiotic prophylaxis. It can then be decided whether further follow-up is needed.

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Chapter 6

Development and validation of a disease-specific module for health-related quality of life and congenital heart disease

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Abstract

In this study, the development and reliability and validity of a congenital heart disease-specific (CHD) health-related quality of life module is presented: the CHD-TAAQOL (TNO-AZL Adult Quality Of Life). Health-related quality of life was defined as health status weighted by emotional response to problems in health status. The CHD-TAAQOL belongs to the generic health-related quality of life - instrument (the TAAQOL questionnaire). Items were based on a pilot study, interviews with patients, expert meetings and literature.

The CHD-TAAQOL was tested in 156 patients with mild or complex congenital heart disease. Reliability, scale structure, convergent validity and criterion validity were calculated.

The CHD-TAAQOL module consists of three scales (number of items in parentheses): 'Worries' (10), 'Symptoms' (9), 'Impact cardiac surveillance' (7). Cronbach's alfa ranged from 0.77 to 0.82. Scale structure was confirmed by principal component analysis, corrected item-scale and interscale correlations. Convergent validity was good. Criterion validity showed significantly lower scores for complex congenital heart disease patients than for patients with mild congenital heart disease. Overall, 55% of reported health status problems were associated with negative emotions, which is an argument for assessing health-related quality of life as a concept distinct from health status.

The CHD-TAAQOL module together with the generic TAAQOL may be used to assess group differences for disease-specific health-related quality of life in congenital heart disease patients. Testing of reliability and validity of the CHD-TAAQOL shows satisfactory results. However, to detect changes in health-related quality of life over time, further research has to be performed.

Introduction

The mean value for the prevalence of congenital heart malformations at birth is 7.7 per 1,000 live births.[1] In the Netherlands, each year approximately 1600 children with congenital heart disease are born.[2] Because of medical improvements, especially in operative techniques and postoperative treatment, nowadays at least 85% of those neonates will likely survive to adulthood. This results in a growth of the population of adults with congenital heart disease with 5% every year.[3] Studies on mortality and morbidity in patients with different congenital heart diseases have been performed.[4-7] To assess the influence of disease and treatment on daily life of patients with chronic disease, health-related quality of life in addition to mortality and morbidity as an outcome measure has been used increasingly during the last decades.[8,9]

Health-related quality of life can be measured with generic instruments. These instruments are designed for use across a wide range of clinical conditions and for comparison with the general population. Their broad applicability is derived from their coverage of the complete spectrum of function, disability and distress. Different types of generic instruments for adults are available, such as the SF-36 (Short Form- 36 items) and the TAAQOL (TNO/AZL Adult Quality Of Life questionnaire).[10-12]

In disease-specific instruments, items specific for the disease group or function group not covered by the generic instrument are included. Disease-specific instruments are therefore more responsive to change for example when an intervention is done.[13] When assessing quality of life preferably disease-specific instruments should be used in addition to generic instruments.[13-15] To date, no disease-specific instrument for congenital heart disease has been described.

In the present study, the following basic principles for the development of a disease-specific instrument were applied, derived from a publication by Sprangers et al.:[16] 1. Items must cover relevant areas of quality of life not (sufficiently) covered by the core (generic) questionnaire 2. The format should be compatible with the core instrument. 3. The items should preferably employ the same time frame as the core instrument. 4. It should be easy to understand and to respond to and be brief: the number of items in the instrument should not exceed that of the core questionnaire. 5. It should comprise different item scales. 6. It should exhibit adequate levels of reliability and validity, including responsiveness to clinically important changes in health status over time.

As a complement to the already validated generic core instrument (TAAQOL), the Congenital Heart Disease (CHD)-TAAQOL instrument was developed, which is described in the present study. Secondly, its scale structure, reliability, convergent validity and criterion validity was tested.

Materials and Methods

The study was approved by the local medical ethical committee. All patients gave informed consent. A cross sectional design was used and data on medical follow-up was searched for retrospectively or updated by medical examination.

Patient selection

The archives of the department of paediatric cardiology at the Leiden University Medical Center contained information on 4383 patients born from 1968 to 1982. Out of these patients, 2280 were randomly selected. All patients with complex or mild congenital disorder were sorted out. Those 500 patients were further studied for most recent medical information by contacting hospitals, general practitioners and local authorities.

Of the entire group, 128 patients had died before the study started. Exclusion criteria were (number of patients in parentheses): mental retardation (51), not speaking Dutch/ not living in the Netherlands (46), participated in a previous study (44).[12] Seven patients were lost to follow-up. The final sample of 224 patients was invited to the study.

Instruments

Core instrument: TAAQOL

In the TAAQOL, health-related quality of life was measured based on the following definition: health-related quality of life is health status weighted by the emotional response to problems in health status.[11,12] Figure 1 shows an example of the format of the items in the questionnaire. The first question refers to health status; the second question refers to the emotional impact in case of a health status problem. Scores of the two questions are combined in one score for each item (combined item score). The questionnaire and format have been validated in the general population, as well as in different disease groups, as part of the development of a series of quality of life-instruments for various age groups and respondents.[11,12,17-27]

Figure 1 TAAQOL format

Did you have difficulty in the last month with ....

walking up the stairs? ❑ no ❑ a little ❑ some ❑ a lot

How much did that bother you?

❑ not at all ❑ a little ❑ quite a lot ❑ very much

The TAAQOL includes 45 items, divided into 12 different scales. Each scale contains 2 to 4 items (in parentheses): gross motor function (4), fine motor function (4), pain (4), sleeping (4), cognitive function (4), social function (4), daily activities (4), sexuality (2), vitality (4), happiness (4), depressive moods (4) and aggressiveness (3). Scores for each scale range from 0 to 100, with higher scores representing better quality of life. With the TAAQOL, convergent validity was tested.

SF-36 (Short Form- 36 items)

Apart from the health-related quality of life questionnaires, participants were asked to fill in the SF-36, an internationally well-known questionnaire for subjective health status.[10]

This questionnaire was based on 35 items representing 8 multi-item scales (with the number of questions in parentheses): physical function (10), role limitations due to physical health problems (4), bodily pain (2), general health perceptions (5), vitality (4), social function (2), role limitations due to emotional health problems (3) and mental health (5). Scores range from 0 to 100, higher scores indicating better subjective health status. With this questionnaire convergent validity was tested.

NYHA class

By studying medical data, the New York Heart Association functional class (NYHA class) was determined. The NYHA class is a classification for severity of the disease based on specific cardiac complaints, rated from 1 to 4, with higher score for worse status.[28] With this variable criterion validity was evaluated.

Disease-specific instrument: CHD-TAAQOL, the development

The TAAQOL-format (Figure 1) was used to develop the disease-specific instrument for health-related quality of life in congenital heart disease patients. In a previous study, the first concept of the disease-specific questionnaire was created.[12] The items were based on literature and clinical experience of paediatric cardiologists and psychologists. Main goal of this version was to make an inventory of the impact of the cardiac defect on daily life for adults with congenital heart disease. Items important to the patients and remarks by the patients in this study were taken into account during the drafting of the next version of the disease-specific instrument. The expert advice from 3 psychologists, a paediatric cardiologist and a cardiologist specialised in congenital heart disease was used. By individual interviews with 4 congenital heart disease patients selected from the outpatient clinic, clarity, utility and completeness of the questionnaire were evaluated.

Testing of the CHD-TAAQOL was then performed in a group of 156 patients with mild or complex congenital heart disease. Items relevant for less than 10% of the patient group were excluded. Items that did not influence strength of Cronbach's alfa were also excluded. Finally, items that did not fit in the factor structure were excluded, only if this not influenced strength of Cronbach's alfa.

Since the number of items in the instrument should not exceed the number of questions of the core questionnaire,[16] specific scales were chosen to be included in the CHD-TAAQOL. Literature on disease-specific health-related quality of life[16,29-31] and the advice of experts gave insight in the importance of different scales taking account the age of the patients in this study. The following scales were found to be important and relevant for a disease-specific instrument in this study population: 'Symptoms', 'Impact cardiac surveillance' and 'Worries'.

Reliability and validity testing of the CHD-TAAQOL

SPSS for windows, version 10.07 (2000), was used to perform statistical calculations.

To test scale structure

Reliability was evaluated by calculating Cronbach's alfa for each scale.

Scale structure was evaluated by means of several procedures. Principal Component Analysis with varimax rotation was done on the combined-item scores. The item-rest and item scale correlation coefficients were calculated. Finally, the relationship between the different CHD-TAAQOL scales was evaluated with Spearman correlation coefficients.

To test convergent validity

To investigate convergent validity of the CHD-TAAQOL, the relationship with the TAAQOL- and SF-36- scales was investigated. We hypothesised correlations on the basis of expected problems in patients with congenital heart disease and the formulated items in the different questionnaires. With the 'Symptoms' scale (CHD-TAAQOL) physical function is questioned, but more extensively than in the TAAQOL and SF-36. We therefore expected the 'Symptoms' scale in the CHD-TAAQOL to be moderately correlated ($r > 0.5$) with the gross motor function- and vitality- scales in the TAAQOL and the physical function-, role limitation physical-, vitality- and general health perceptions-scales in the SF-36. For the scale 'Impact cardiac surveillance' large correlations were not expected, although the questionnaire format evaluates frequency of cardiac surveillance, which could be correlated to physical condition. The 'Worries' scale was expected to associate moderately with both physical condition as well as mental state.

To test criterion validity

Studies on health-related quality of life are based on the assumption that health problems may have a negative impact on health-related quality of life. Consequently, instruments based on assessing health-related quality of life should be able to make this impact visible. Therefore, correlations were calculated between the CHD-TAAQOL scales with the NYHA class. We expected moderate correlation ($r > 0.5$) between the NYHA class and the scales 'Symptoms' and 'Impact cardiac surveillance'.

In addition, criterion validity of the CHD-TAAQOL was evaluated by comparing scale scores of the group of patients with mild congenital heart disease with those of the group of patients with complex congenital heart disease by means of ANOVA with ranked scores and correction for age and sex.

To test conceptual validity

The distinction between health status problems and the (negative) emotional impact in case of a health status problem was evaluated by calculating the number of health status problems and the percentages of such problems that led to a negative feeling. The total number of health status problems was computed by dichotomising every health status item into 'no problem' (never) vs. 'a problem' (a little/ some/ a lot).

Results

Patients

Of the 224 selected patients, 160 agreed to participate (71%). Four patients did not return their questionnaire, so results were based on a sample of 156 patients. This resulted in an overall response rate of 70%, with a higher response in the group of patients with complex congenital heart disease (88%) compared with the group of patients with a mild congenital heart disease (58%).

Diagnoses of the participants are listed in Table 1. All 156 participants were aged between 17 and 32 years with a mean of 24.5 years. The group consisted of 73 men and 83 women.

Table 1 Participating patients: main congenital cardiac disease groups

Mild lesions (80)			**Complex lesions (76)**	
Spontaneous resolution	36		Systemic morphologically right ventricle	38
Persisting lesions	44			
Ventricular septal defect		19	Conduit or mechanical prosthesis	22
Pulmonary stenosis		7		
Atrial septal defect		6	Univentricular atrioventricular connection- Fontan	11
Aortic stenosis		6		
Bifoliate aortic valve		3	Palliative operation- shunt	5
Abnormal pulmonary venous drainage		2		
Prolapsing mitral valve		1		

Disease-specific instrument: CHD-TAAQOL, the development

After re-evaluation and adaptation of the first version of the questionnaire[12] according to the patients' remarks and an expert team, 4 patients with congenital heart disease were interviewed about this second version. Then, some formulations were simplified. This resulted in a questionnaire consisting of three scales, with the number of items in parentheses: 'Symptoms' (11), 'Impact cardiac surveillance' (12) and 'Worries' (17). This revised questionnaire was send to the participating sample of 156 patients.

The analyses of these data showed only 11 missings in answers on items were reported. Items relevant for less than 10% of the patients were excluded. These were the following 4 items: syncope ('Symptoms'), heart catheterisation ('Impact cardiac surveillance'), exercise test ('Impact cardiac surveillance') and hobbies ('Worries'). Five items did not influence strength of Cronbach's alpha and were excluded as well: transpiration ('Symptoms'), other investigations ('Impact cardiac surveillance '), using antibiotics ('Impact cardiac surveillance'), to see the general practitioner ('Impact cardiac surveillance'), sexuality ('Worries'). To optimise factor structure, the following 5 items were excluded, all from the scale 'Worries': income, household, going on holidays, the possibility to raise children and cardiac surveillance. Removal of these items did not have substantial impact on the strength of Cronbach's alfa. After excluding these items, the final version (Congenital Heart Disease- TNO-AZL Adult Quality of Life questionnaire, CHD-TAAQOL) consisted of the following three scales, with the number of items in parentheses: 'Symptoms' (9), 'Impact cardiac surveillance' (7) and 'Worries' (10), altogether 26 items. Items are shown in Table 2. It took the participants about 5-10 minutes to answer the CHD-TAAQOL.

Reliability and validity testing of the CHD-TAAQOL

Scale structure

Cronbach's alfa were 0.77, 0.78, 0.82 for the 'Symptoms', 'Impact cardiac surveillance' and 'Worries' scales, respectively.

Principal Component Analysis with varimax rotation explained 44% of the variance and reproduced the scale structure very well: all but two items ('Symptoms': looking pale and 'Worries' exercise/ sports) loaded higher on their own factor than on other factors. Of all factor loadings, 85% was above 0.50. Details are shown in Table 2.

Table 2 Rotated factor matrix (only factor loadings > 0.2 are mentioned)

Items	factor 1	factor 2	factor 3
Symptoms			
short of breath <100m walking		**0.74**	
short of breath 1-5 km walking		**0.80**	
orthopneu		**0.65**	0.22
nycturia		**0.59**	
oedema		**0.41**	
palpitations	0.36	**0.57**	0.21
dizziness	0.31	**0.38**	
cyanosis during exercise	0.22	**0.41**	0.23
looking pale	**0.50**	0.27	
Impact cardiac surveillance			
ECG		0.26	**0.76**
echocardiography			**0.70**
taking blood		0.34	**0.67**
X-ray			**0.66**
anti flu vaccination	-0.21	0.32	**0.51**
to specialist for heart	0.24		**0.65**
admission to hospital for heart	0.23		**0.55**
Worries about..			
education	**0.59**		
job	**0.73**		
career	**0.66**		
living independently	**0.59**	0.21	
being home alone	**0.50**	0.44	
exercise/ sports	0.50	**0.52**	
having friends	**0.62**		
relationship	**0.69**		
having children	**0.33**		0.28
general health	**0.61**	0.20	0.26

bold: highest correlation

Table 3 shows that the scale specificity was good for all but two items ('Symptoms': looking pale and cyanosis during exercise), as the item-rest correlation coefficients with their own scale of all but these two items were higher than the corresponding item-scale correlations with any other scale. Twenty-one out of 26 items (81%) had a corrected item-scale correlation higher than 0.40. The correlation between the scales showed that no scales shared more than 25% of their variance, indicating no clear relation between the scales, which confirms the multi-dimensionality of the instrument (Table 3).

Table 3 Item-scale and interscale correlations of the CHD-TAAQOL

	Symptoms	Impact cardiac surveillance	Worries
Number of items	9	7	10
Corrected item-scale correlation			
Item-rest correlation higher than correlation with other scales	7	7	10
Item-rest correlations higher than 0.40	6	6	9
Item-rest correlations higher than 0.60	2	1	2
Correlation between scales			
Symptoms	-		
Impact cardiac surveillance	0.41	-	
Worries	0.48	0.25	-

Convergent validity

Spearman correlation coefficients between the CHD-TAAQOL and the TAAQOL and SF-36 are presented in Table 4. Overall, most CHD-TAAQOL scales demonstrated moderate correlations with the TAAQOL and SF-36 scales in the hypothesised direction. The correlation between the scale 'Worries' and daily activities (TAAQOL) and social function (SF-36) on the other hand were present but had not been predicted. The expected correlations between 'Worries' and gross motor function, vitality and happiness (TAAQOL) and physical function, role limitation physical, vitality and general health (SF-36) were not found.

Table 4 Spearman correlation coefficient between the TAAQOL and SF-36 with the CHD-TAAQOL

TAAQOL	Symptoms	Impact cardiac surveillance	Worries
Gross motor function	***0.61**	0.27	*0.40
Fine motor function	0.24	-0.03	0.28
Cognitive function	0.46	0.19	0.42
Sleep	0.45	0.13	0.38
Pain	0.45	0.17	0.28
Social functioning	0.28	0.19	0.43
Daily activities	0.48	0.10	**0.51**
Vitality	***0.58**	0.13	*0.38
Happiness	0.14	0.02	*0.38
Aggressiveness	0.23	-0.08	0.28
Depressive moods	0.40	0.18	***0.62**
SF-36			
Physical function	***0.67**	**0.52**	*0.45
Role limitation physical	***0.504**	0.20	*0.45
Role limitation emotional	0.21	0.03	0.39
Social functioning	0.47	0.25	**0.54**
Pain	0.45	0.10	0.26
Mental health	0.26	0.13	***0.61**
Vitality	***0.501**	0.12	*0.495
General health	***0.62**	0.40	*0.499

*Correlation hypothesised **bold:** moderate correlations (>0.5)

Criterion validity

To evaluate criterion validity, correlations between the CHD-TAAQOL scales and the NYHA class were calculated. Spearman correlation higher than 0.5 are shown for 'Symptoms' and 'Impact cardiac surveillance' with NYHA class (-0.56 and -0.55 respectively).

In addition, criterion validity was evaluated by comparing the CHD-TAAQOL scale scores between patients with mild congenital heart disease and patients with complex congenital heart disease. ANOVA analyses with correction for age and sex showed that patients with complex congenital heart

disease score significantly lower on all scales compared with the group of mild congenital heart disease patients, indicating a worse quality of life. Mean scores (standard deviation) of the 'Symptoms', 'Impact cardiac surveillance' and 'Worries' scales in mild congenital heart disease patients were 92.1 (8.9), 97.8 (5.6) and 89.8 (11.7) respectively, whereas they were 86.4 (13.2), 85.4 (9.7) and 84.4 (14.4) in patients with complex congenital heart disease (significance p= 0.00, p= 0.00 and p= 0.01, respectively).

Conceptual validity

A total of 1175 problems were reported of which 649 (45%) were associated with a negative emotional response (Table 5). Overall 19% to 77% (depending on the scale involved) of the reported health status problems led to negative emotional reactions to such problems.

Table 5 Total number of problems and number of problems that elicited a negative emotion

Scale	#P	#Pneg	#Pneg%
Impact cardiac surveillance	331	63	19%
Symptoms	385	234	61%
Worries	459	352	77%
Total	1175	649	55%

#P = total number of problems
#Pneg = number of problems, with negative emotional reaction
#Pneg% = % with negative emotional reaction

Discussion

In the present study, the development, and reliability and validation of a congenital heart disease-specific quality of life instrument, the CHD-TAAQOL, was described.

With the development, patients' opinions were taken into account, as well as the advice of experts. Item generation and item reduction resulted in a instrument consisting of three scales: 'Symptoms' (9), 'Impact cardiac surveillance' (7) and 'Worries' (10), altogether 26 items.

The scales show good reliability. The CHD-TAAQOL instrument may be used to assess group differences for disease-specific health-related quality of life in patients with congenital heart disease. However, when the value of a scale for an individual is of interest, the CHD-TAAQOL cannot be used safely; for use in the clinical situation much higher levels of Cronbach's alfa are mandatory.

The scale structure of the CHD-TAAQOL scales was confirmed by principal component analysis and was supported by high item-rest correlations. Almost every item-rest correlation (the correlation with its own scale) was higher than the item correlation with the other scales. In addition, the low to moderate inter-scale correlations support the assumption that health-related quality of life should be defined with multiple independent scales.[8,25,32]

Convergent validity was evaluated by relating CHD-TAAQOL scales to TAAQOL- and SF-36- scale scores. Almost all hypothesised correlations were found: 'Symptoms' was correlated with gross motor function and vitality in the TAAQOL and the physical function-, role limitations physical-, vitality- and general health perceptions- scales in the SF-36. In addition, 'Impact cardiac surveillance' was moderately correlated ($r > 0.5$) with the physical function scale in the SF-36, most likely due to the first part of the CHD-TAAQOL question, regarding frequency of cardiac surveillance (Figure 1). The scale 'Worries' was correlated almost $r = 0.5$ to the physical scales in the SF-36 (physical function, role limitations physical, vitality and general health perceptions). These correlations were as predicted. No correlation was found between 'Worries' and gross motor function, and vitality in the TAAQOL. Mental scales however, as depressive moods (TAAQOL) and mental health (SF-36) were stronger correlated to the scale 'Worries' than to these physical scales. This assumes that 'Worries' obviously relates more to mental status than to physical health. It could not be explained why the correlation between 'Worries' and Happiness in the TAAQOL, turned out not to be relevant. The 2 unpredicted correlations ('Worries' with daily activities in the TAAQOL and 'Worries' with social function in the SF-36) could, however, be explained. Firstly, the scale 'Worries', also contains items on different daily activities (such as education, exercise/ sports). Secondly, the scale 'Worries' contains items on social life (such as relationships, having friends). Therefore, these correlations are well understood.

Evaluation of criterion validity was performed by correlating the CHD-TAAQOL with the NYHA class, which represents the severity of the disease. These correlations were all in the expected direction. In addition, differences between the group of patients with mild and the group with complex congenital heart disease showed significantly lower scores in all of the CHD-TAAQOL scales for the severely diseased group. These results demonstrate that the CHD-TAAQOL can detect differences in health-related quality of life between healthy and less healthy subjects.

The distinction between health status problems and the emotional reactions to such problems is mentioned in different publications[33,34] and appears to be relevant in the present study: overall 55% of the problems in health status elicit a negative feeling in the patients. This percentage varies greatly among the different scales. In other words, there is more to health-related quality of life than just measuring health status. This phenomenon is an argument for assessing health-related quality of life as a concept distinct from health status.

Overall, the instrument meets the criteria described by Sprangers et al..[16] The CHD-TAAQOL covers areas of quality of life not (sufficiently) covered by the core instrument (criteria 1), the format is compatible with the core instrument (criteria 2), it is easy to understand and the number of items does not exceed that of the core instrument (criteria 4), it comprises different item scales (criteria 5) and exhibits adequate levels of reliability and validity (criteria 6).

Two exceptions, however, have to be discussed. For the scale 'Impact cardiac surveillance' a different time frame was chosen ('the last year'), since it was meaningless asking for impact of surveillance only in the last 4 weeks (criteria 3). In addition, no longitudinal study with the CHD-TAAQOL was performed, so no conclusions can be drawn considering sensitivity to change (criteria 6). Future research should give final conclusions on that subject.

Remarks

Although the items of the instrument were chosen on the basis of, among others, the previous study[12] and interviews with patients, for the choice of the scales of the CHD-TAAQOL the patients' preference was not reviewed. Experts decided on the scales to be included and literature on disease-specific questionnaires was taken into account. Because of this selection, various

subjects that could also be relevant to the patients (such as side effects of medication) were not included in the instrument. Whether the module (CHD-TAAQOL together with the TAAQOL) is sufficient to be used in a study depends on the research question.

Although the selection method was chosen in such a way that selection bias was prevented, the response percentage in the patients with mild congenital heart disease was 58%. Non-response bias could have affected the results. Non-participants lived further away from the hospital than participants. Furthermore, the response percentage of participating women (66%) and men (49%) was different. Since many participants had to visit the hospital for cardiac check-up, travel distance and availability of time could have played a role in participating in the study. Also, the response percentage in the group of patients whose cardiac malformation had resolved was higher (74%) than that in the group with minor actual malformation (50%). During cardiac re-examination, most patients asked questions or expressed worries that they had lived with for several years. It is possible that patients with relatively low health-related quality of life and with more restrictions or questions joined the study. This could have resulted in negative selection bias, which might have influenced calculations of criterion validity.

Conclusions

The CHD-TAAQOL questionnaire together with the generic TAAQOL may be used to assess group differences for disease-specific health-related quality of life in patients with congenital heart disease. Testing of reliability and validity of the TAAQOL-CHD shows satisfactory results. However, to detect changes in health-related quality of life over time, further research has to be performed.

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Chapter 7

Disease-related difficulties and satisfaction with level of knowledge in adults with mild or complex congenital heart disease

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Abstract

Objectives: To evaluate difficulties in daily life, and satisfaction with level of knowledge about their disease, in patients with congenital cardiac disease in order to improve counselling.
Methods: A self-administered questionnaire was completed by 80 patients with mild, and 76 with complex congenital cardiac disease. They were aged from 17 to 32 years.
Results: Even patients with only mild malformations experienced difficulties related to their disease, but being found in only 11%, these were significantly less than those uncovered in 87% of those with complex disease ($p < 0.001$). Those patients with complex malformations frequently felt restricted in choices because of their disease in areas such as sport (59%), employment (51%), and education (34%). Other difficulties reported were: paying a higher premium for life insurance (29%), having to give up on a sport (28%), and being excluded from a job (18%). Depending on the item, between one-fifth and two-thirds of participants reported gaps in knowledge, most frequently for "causes of congenital cardiac disease", "future consequences", and "family planning". For 53% of those with mild anomalies, and 93% of severely affected patients, the cardiologist is the most important source of information.
Conclusions: A minority of adults with mild and a majority of those with complex congenital cardiac disease report difficulties in daily life. A substantial number of these patients feel that they have an inadequate level of knowledge about their disease. Our results suggest the need for a specific programme of counselling.

Introduction

Since the first operation for correction of a congenital cardiac malformation was performed in 1945, technologies have developed, and survival of patients has increased, so that now there is a substantial number of adults with congenital cardiac disease.[1]

Consequently, restrictions related to the disease, and future perspectives, have become increasingly important. Few previous studies have evaluated difficulties in daily life of such adults with congenital cardiac disease. These studies were mainly focussed on employability and insurability.[2-10] Difficulties in other areas of life, such as sport or having children, have hardly been studied.

Understanding of the chronic illness is associated with less distress and confusion, more satisfaction with medical care, better compliance with treatment, and an improved emotional state.[11] With new laws, and increasing emancipation of patients, the demand for education increases. New sources of information, such as the Internet, have changed the needs of adults with congenital cardiac disease. The level of knowledge of such patients with congenital cardiac anomalies has also been examined in only a few studies. Some evaluated what the patient was supposed to know according to the researchers or physicians,[11-18] while others focussed on the patients' satisfaction with their own level of knowledge.[9,10,16,19,20]

We carried out our cross-sectional study in patients with mild and in patients with complex malformations with the following objectives:

- to assess difficulties in daily life related to the cardiac anomaly,
- to assess the satisfaction of the patients with level of knowledge about their disease, and
- to assess the sources of information concerning their health used by these patients.

Methods

All participating patients gave written informed consent. The study was approved by the local medical ethical committee.

Selection of patients

There is information on 4383 patients with congenital cardiac disease, born between 1968 and 1982, in the archives of the Department of Paediatric Cardiology at Leiden University Medical Center. Of these patients, 2280 were randomly selected. Only those with mild or complex malformations were included in the study, resulting in a sample of 500 patients.

Patients with mild anomalies had not been submitted to surgery, either because their cardiac anomaly was insignificant, or because it had disappeared spontaneously.

Patients with complex malformations had undergone operative procedures that had not led to an anatomically normal heart as described by segmental analysis.[21]

Information on this sample of 500 patients was received from hospitals, general practitioners, and local authorities. Of those 500 patients, 128 had died before the study started. Additional criterions for exclusion were: mental retardation in 51 patients, not speaking Dutch or not living in the Netherlands in 46, joined a previous study in 44.[22] Another 7 patients were lost to follow-up. This left 224 patients who were invited to participate in the study. Their diagnoses and characteristics are given in Tables 1 and 2.

Table 1 Participating patients: main congenital cardiac disease groups

Mild lesions (80)			**Complex lesions (76)**	
Spontaneous resolution	36		Systemic morphologically right ventricle	38
Persisting lesions	44			
Ventricular septal defect		19	Conduit or mechanical prosthesis	22
Pulmonary stenosis		7		
Atrial septal defect		6	Univentricular atrioventricular connection- Fontan	11
Aortic stenosis		6		
Bifoliate aortic valve		3	Palliative operation- shunt	5
Abnormal pulmonary venous drainage		2		
Prolapsing mitral valve		1		

Medical data

Information on the medical history was searched for in the medical files. If clinical information was older than one year, patients were re-examined to confirm their cardiac diagnosis.

Questionnaire

All participants completed a self-administered questionnaire. It encompassed the following topics.

Difficulties in daily life

We included 26 items based on previous studies,[7-9,19,23] our pilot study,[22] and clinical practice. The items covered different subjects such as life/ health insurance, education, employment, sport, and other daily activities (Table 3). If a difficulty was noted, it was specifically asked whether this was due to the cardiac disorder. Results are presented in total numbers and percentages of patients who felt restricted because of their congenital heart disease (Table 3).

Satisfaction level of knowledge about disease

Patients were asked if they felt that they had sufficient knowledge concerning 9 different subjects (Table 4). The subjects were again based on our pilot study.[22] At the end of the questionnaire, an open question was added: "What subject concerning your heart disease would you like to know more about?".

Sources of information

We asked which sources of information were used to obtain information about personal health.

Statistics

Frequencies were run with the SPSS 10.0 package. The group of patients with complex malformations was compared with that with mild disease using the Mann-Whitney U test for comparison between groups, and the Kruskall-Wallis test for comparison of ordinal data from multiple groups.

Table 2 Characteristics of patients

	Mild lesions (80)	Complex lesions (76)
age: mean, median, range	24.6, 25, 17-32 yr.	24.3, 24.5, 18-32 yr.
male	30 (38%)	43 (57%)
medical care >1 year ago	67 (84%)	6 (8%)
highest education finished		
primary education	21 (26%)	32 (42%)
continued education	59 (74%)	44 (58%)
daily life		
employed (more than 12 hours/week)	61 (76%)	43 (57%)
+disablement benefit partial	-	2 (3%)
student	13 (16%)	11 (15%)
+disablement benefit fully	-	4 (5%)
disablement benefit, no activities	-	10 (13%)
on sick leave (whiplash injury/pregnancy)	-	2 (3%)
housewife	2 (3%)	1 (1%)
+disablement benefit fully	2 (not related to CHD)	1 (1%)
unemployed	1 (1%)	2 (3%)
+disablement benefit partial	1 (not related to CHD)	-

CHD= congenital heart disease

Results

Patients

Of the 224 selected patients, 160 agreed to participate (71%). Of these, 4 patients did not return their questionnaire, so results were based on a sample of 156 patients. The response rate in the group of patients with complex disease (88%) was higher compared with that in the group of patients with a minor

malformation (58%). Diagnoses and characteristics of the participants are listed in Tables 1 and 2.

Difficulties in daily life (Table 3)

The difficulties identified as directly related to the congenital malformation are listed in Table 3. In total, 11% of the patients with mild malformations reported one or more difficulties compared with 87% in patients with complex anomalies. All differences in percentages per item between those with mild and complex anomalies were highly significant ($p < 0.001$).

In the group with complex anomalies, only 10 patients did not mention any difficulties related to the disease. These were 7 patients with a systemic morphologically right ventricle, accounting for 18% of the sample, one with a conduit or mechanical prosthesis, accounting for 5%, one with a functionally univentricular heart after a Fontan procedure, 9% of this group, and one of the five who had undergone a palliative shunt procedure.

For each subject, we present most relevant difficulties for the group of patients with complex lesions due to this congenital malformation.

Life insurance: Overall, 27 patients (36%) had problems with taking out a life insurance policy: 5 were rejected, 14 had to pay a higher premium, and 8 experienced both problems.

Employment: Besides 14 patients (18%) who were excluded from a job, 12 patients (16%) had given up on a job because of their cardiac disease. Six of them were excluded after medical examination. Since 6 patients were both rejected and had given up, altogether 20 patients (26%) had been restricted in employment. In addition, two patients were excluded from a possible promotion. Of the group, 45 (59%) were in paid employment, compared with 61 (76%) of those with mild malformations ($p < 0.001$).

Education: Altogether, 6 patients had either given up on an education or had been rejected because of their cardiac anomaly (8%).

Sport: In total, 21 patients had given up on sport (28%), with the highest percentages in patients who had undergone a palliative shunt procedure or else those with a functionally univentricular heart. Only 1 patient was rejected after medical examination.

Table 3 Difficulties in daily life related to the congenital cardiac malformation. All differences between those patients with mild and complex lesions were highly significant: $p < 0.001$

	Mild lesions		Complex lesions	
Items	**n**	**(%)**	**n**	**(%)**
Did you ever feel restricted in the choice of ...				
an educational course?			26	(34)
a job?			39	(51)
a sport?	5	(6)	45	(59)
a hobby?			9	(12)
a house?	1	(1)	15	(20)
a holidays destination?			20	(26)
Did you ever give up ... an educational course?			4	(5)
a job?			12	(16)
a sport?	2	(3)	21	(28)
a hobby?			2	(3)
a holiday?			7	(9)
Did your physician ever advised you not to have children?			11	(14)
Did you ever thought it was better not to have children?			22	(29)
Were you ever excluded from... an educational course?	1	(1)	4	(5)
a job?			14	(18)
possible promotion?			2	(3)
a job after medical examination?	1	(1)	6	(8)
sport after medical examination?	1	(1)	1	(1)
Where you ever prevented from ...			6	(8)
applying from a driving licence?				
taking out a mortgage policy?			6	(8)
taking out a life insurance policy?			13	(17)
taking out a health insurance policy?	3	(4)	3	(4)
Did you ever had to pay higher premium than				
other people for ... a mortgage?	1	(1)	6	(8)
a life insurance?	2	(3)	22	(29)
a health insurance?	1	(1)	7	(9)
Total patients, who experience difficulties for at least one of the items	**9**	**11%**	**66**	**87%**

Children: Of the patients, 22 (29%) think that they "better not have children", while 11 (15%) were "advised not to have children by their physician". More than one-quarter of patients with functionally univentricular heart were advised not to have children.
Other subjects: Restriction in the choice of their holiday destination was reported by 20 patients (26%), 6 patients were rejected for a driving licence (8%), and 2 gave up on a hobby (3%). Altogether, 15 patients felt restricted in the choice of a house (20%). Six of them were also rejected for mortgage and life insurance, and these factors could have played a role in the restriction.

Satisfaction with level of knowledge about disease (Table 4)

Depending on the subject, from one-third to three-fifths of the patients with mild malformations indicated insufficient knowledge, with the item "causes of your cardiac disease" as the most important to know more about, with 61% wanting to know more. This is followed by "future consequences", with 55% looking for more information. Some additional remarks from those with mild anomalies were "Will the defect increase with pregnancy?", "How long do I need medical care?", and "What about the anaesthesia at the dentist?".

Between one-fifth and two-thirds of those with complex malformations reported problems. The items "future consequences" and "family planning" were the items most identified as the topics for more information, identified by 64% and 51%, respectively. These items were also mentioned in the answers to the open question by about one-fifth of the responders. Remarks from the patients with complex malformations were: "Is there an association for patients with congenital heart disease?", and "Do you have information on heart transplantation, living together, insurance?".

For all subjects, except for "future consequences" and "consequences for family planning", patients with mild anomalies made greater demands for thorough information than those with complex lesions. This difference is significant for 4 of the items (Table 4).

Table 4 Satisfaction with level of knowledge

	percentages of patients answering 'not enough'		
Do you know enough about ...	**Mild lesions**	**Complex lesions**	**significance p< 0.01**
future consequences of your cardiac disease?	55	64	
consequences of your cardiac disease for family planning?	45	51	
causes of your cardiac disease?	61	43	Yes
what kind of cardiac disease you have?	51	37	
consequences of your cardiac disease for occupation/ career?	39	32	
consequences of your cardiac disease for exercise/ sport?	45	30	Yes
consequences of your cardiac disease for your education?	39	26	Yes
things to do to remain healthy?	36	24	
consequences of your cardiac disease for spending free time?	35	20	Yes
Percentage of patients who feel that their level of knowledge is insufficient	**35-61%**	**20-64%**	

Sources of information

For 53% of those patients with mild lesions, and 93% of those severely affected, the cardiologist is the most important source of information concerning their health. In addition, general practitioners inform about one-third of both groups. Parents and friends also play an important role in the dissemination of information in from one-tenth to one-fifth. Television was the source of information for 10% of those with mild, and 15% of those with complex anomalies. For patients with complex malformations, the patient's association was the source of information for 18%, and the Internet for 7%. Other sources that were mentioned were magazines or papers, the psychologist, and the library.

Discussion

Our study has shown that a minority of patients with mild lesions and a majority of those with complex anomalies, experience difficulties in daily life directly related to their cardiac disease. In addition, a substantial number of patients felt that they were insufficiently informed about their disease.

Difficulties in daily life

Studies that have focussed on difficulties in daily life for patients with congenital cardiac disease are few. Data published has mainly concerned problems related to employment or availability of insurance.[2-10] In our study, nearly one-third of patients with complex malformations reported difficulties in employment, and just over half felt restricted in the choice of a job because of their cardiac disease. Previous studies concluded that between one-tenth and two-fifths of the patients, depending on the lesions studied, were not able to have the job they wanted. The percentage mentioned in the study by Ghisla et al.,[5] describing a group of patients with complex malformations (tetralogy of Fallot) accounting for over two-fifths, is comparable to that found in our study. For life insurance, just over one-third of our patients with complex malformations were either rejected for cover, or had to pay higher premium than usual. Other groups have reported about one-third of patients being rejected for life insurance.[4,7] It has to be taken into account that not all patients had applied for life insurance yet because of the age range chosen for the study and, therefore, this proportion may well increase with advancing age. This might apply for all other items as well.

Level of knowledge about disease

In our study, from one-fifth to two-thirds of the patients, depending on the item, felt themselves insufficiently informed. Other studies have also shown a substantial percentage of patients wanting more information about their disease. Mühler et al.,[9] and Otterstad et al.,[19] reported that one-third and three-fifths of the patients, respectively, felt insufficiently informed. Wright et al.[10] found proportions between one-third and three-quarters depending on the item. Differences may be explained by the use of slightly different items. Percentages for the items: "..consequences of your cardiac disease for family planning", "..causes of your cardiac disease?/ what kind of cardiac disease do you have?" and "..consequences of your cardiac disease for exercise/ sport?" in our study, nonetheless, were remarkably similar to those found by Wright et al.[10] We can, therefore, conclude that over a period of 14 years, the need by patients for knowledge has not decreased, and their education has still to be improved.

A more recent study showed that a high percentage of 96.8% of patients was satisfied with the information given about their congenital cardiac malformation.[16] This cannot be explained by better objective understanding, since the study also identified important gaps in their knowledge. The high percentage of satisfaction might be the result of more attention for counselling, with the help of a nurse practitioner who is working in the University Hospital. Another explanation may be that the reported high satisfaction is biased by the fact that the researcher questioned the patient directly. Although it is not possible to prove which argument is true, the results of that study emphasise the difference between true understanding of the illness and satisfaction with knowledge about the disease. And both need to be improved.

How to prevent difficulties and improve satisfaction with level of knowledge?

To prevent unnecessary difficulties, and to close gaps in knowledge, counselling should be optimised. Subjects, such as vocational choice, should be discussed at an early age. Specific advice should be given to the patients. They should be encouraged to apply for insurance from several companies, or to seek assistance from an independent insurance agent. Job training or career counselling can be offered. In case of doubts about advice, re-assessment has to take place.[24-26] Moreover, all information should be reinforced, since the recall of the patients is disappointing.[17] Regular follow-up and a "health passport" in which information about diagnosis and operation, exercise prescription, family and career planning is summarised, could prove particularly helpful.[27]

Methodological issues

The sample of patients included in our study may not be fully representative for the overall population of patients with congenital cardiac disease. Only three-fifths responded from the group of patients with mild lesions. Non-participants lived further away from the hospital than participants, but this is not likely to have influenced the results in this study. Since the percentage of participating women and men responding was different, the bias of those not responding may have affected the results.

During cardiac re-examination, it was observed that most patients asked questions, or expressed worries, that they had lived with for several years. It is possible that patients with relatively more difficulties or questions joined the study, influencing results towards more difficulties and gaps in knowledge in the group of patients with mild disease.

In conclusion, if in future we are to prevent difficulties and improve satisfaction with level of knowledge about their disease, we must develop a dedicated programme of counselling for adults with congenital cardiac disease.

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Chapter 8

Employment in adults with congenital heart disease

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"What This Study Adds" Box

Paid employment is important in daily life, not only in terms of earnings and social status, but it is also valued for its social support and social distraction. Although various studies have shown that the majority of patients with congenital heart disease is able to work, it is unclear whether patients experience handicaps and which factors are related to reduced job participation. Such information could contribute to the improvements of vocational counseling and employment prospects of patients with congenital heart disease.

The present study shows that patients with complex congenital heart disease have reduced job participation, as compared with patients with mild congenital heart disease and the general population. Many receive disablement benefit, or experience career problems or job handicaps for mobility. Career counseling focusing on physical abilities and level of education may help to prevent or reduce these job-related problems.

Abstract

Objective: To evaluate job participation, career and actual job problems in adults with complex congenital heart disease, compared with those of adults with mild congenital heart disease and reference groups.
Design: Cross sectional study.
Setting: Patients were randomly selected from the Archives of Pediatric Cardiology of the Leiden University Medical Center.
Patients and main outcome measures: In total, 76 patients with complex and 80 with mild congenital heart disease (aged 17 to 32 years) completed a self-report questionnaire on employment and handicaps, with reference data available (response 70%).
Results: In the study-groups, 59% (45) of patients with complex congenital heart disease had a paid job compared with 76% (61) of those with mild congenital heart disease. Patients with complex disease older than 25 years had a significantly lower job participation (64%) than the general population (83%). Multiple logistic regression showed that group of congenital heart disease and level of education were significantly and independently related to job participation (odds ratios 4.8, 99%CI 1.2-19.6, and 4.7, 99%CI 1.3-17.2 respectively). Of the complex congenital heart disease patients, 55% (42) experienced disease-related career problems, in contrast to only one patient with mild disease. Both congenital heart disease groups showed more job-related handicaps for mobility than the reference group. However, in the mild congenital heart disease group, these could be attributed to additional non-cardiac diseases.
Conclusions: Patients with complex congenital heart disease have reduced job participation, as compared with patients with mild congenital heart disease and the general population. Many receive disablement benefit, or experience career problems or job handicaps. Career counseling focusing on physical abilities and level of education may help to prevent or reduce these job-related problems.

Introduction

Advances in medical and surgical treatment of congenital heart disease have led to the survival of increasing numbers of young adults with a range of residual cardiovascular abnormalities. Employment prospects are a major concern for patients and their parents. Paid employment is important in daily life, not only in terms of earnings and social status, but it is also valued for its social support and social distraction.[1] Moreover, a recent report has revealed an association between unemployment and depression in cyanosed congenital heart disease patients.[2] Although various studies have shown that the majority of patients with congenital heart disease is able to work,[3-7] it is unclear whether patients experience handicaps and which adaptations would be helpful. Such information could contribute to the improvements of vocational counseling and employment prospects of patients with congenital heart disease.

Therefore, the aim of this study was to evaluate, in adults with previously operated complex congenital heart disease: job participation, career-related problems, and job handicaps. These data were compared with those of patients with mild congenital heart disease. For job participation and job handicaps, data were also compared with a reference group from the general population.

Methods

The study was approved by the local Medical Ethical Committee and informed consent was obtained from each participant. It concerns a cross sectional study. Information on cardiac diagnosis was searched for retrospectively; however, if this information dated from more than one year since, patients were re-examined.

Patient selection

Selection procedure

In total, 80 patients with previously operated complex and 80 with mild congenital heart disease had to participate in the study to reach sufficient power. Our procedure to select these patients aimed at a representative sample of congenital heart disease patients from the entire Dutch population and not just those who currently visited the Leiden University Medical Center.

Therefore, patients were selected from the Archives of the Department of Pediatric Cardiology (Leiden University Medical Center) which has complete information on patients seen from 1950 onwards. From these files, 4383 patients were born between 1968 and 1982. All patient files were numbered. By using a random numbering list, the files were studied. Patients were categorised in a certain congenital heart disease group and then excluded (death, mental retardation, etc.) or included for further study. After having included 30 patients, their latest medical information and home addresses were obtained from most recent medical file, general practitioners or local authorities, since the Archives contained only information till childhood. It was important to find out if the patient was still alive and still part of the category included in this study: complex previously operated or mild congenital heart disease. If the patient could indeed be included according to our criteria, the patient was invited to participate in the study. This process continued until in total a sample of 80 patients for each patient group agreed to participate.

Therefore, 2280 files from the Archives needed to be studied. This selection included 500 patients with previously operated complex or mild congenital heart disease. From these 500 patients, 128 patients had died. Others were excluded for the following reasons (number of patients in parentheses): mental retardation (51), not speaking Dutch/ not living in the Netherlands (46), or having participated in a previous study (44)[8]. Seven patients were lost to follow-up. The remaining 224 patients (86 with complex, and 138 with mild congenital heart disease) were invited to participate in the study. Of these, 160 agreed to participate, of which 156 returned the questionnaire.

Definitions of the two patient groups concerned the following.

Group 1: patients with previously operated complex congenital heart disease

Patients with previously operated complex congenital heart disease were approached, excluding those after anatomical corrective surgery according to sequential segmental analysis (a proven practical system for the classification of congenital heart disease based on the anatomical construction of the heart).[9] All included patients had undergone a non-anatomical correction, i.e. venous switch for transposition of the great arteries, a correction with the use of allogenous tissue (e.g. Rastelli correction, insertion of a mechanical valve), or partial or complete cavopulmonary connection. The main common characteristic of this group was that, postoperatively, none of the participants

had an anatomically normal heart as described by the segmental analysis,[9] as opposed to patients after anatomical corrective surgery (e.g. operated ventricular or atrial septal defects, coarctation of the aorta, patent ductus arteriosus, pulmonary or aortic stenosis, uncomplicated tetralogy of Fallot, and total anomalous pulmonary venous connection).

Table 1 Participating patients: main congenital heart disease groups (number of patients in parentheses)

Complex CHD n= 76	
Main diagnosis	**Main operation**
1. systemic right ventricle (38)	
TGA (21)	Senning (11) or Mustard (10)
TGA + PS (5)	Senning (3) or Mustard (2) with PS repair
TGA + VSD + PS (6)	Senning (5) or Mustard (1) with VSD closure and PS repair
L-TGA + VSD + PS (2)	VSD closure with PS repair (2)
L-TGA + VSD + ASD (1)	VSD closure and ASD closure (1)
L-TGA + VSD (1)	VSD closure (1)
L-TGA + PS (1)	Tricuspid valve replacement (1)
AV-discordance + DORV + PS (1)	Tricuspid valve replacement and conduit (1)
2. conduit or mechanical prosthesis (22)	
PA + VSD (11)	Rastelli (3); correction with other conduit (8)
AoS (4); MI (1);	Mechanical prosthesis (5)
TGA + VSD + PS (4)	Rastelli (4)
TGA + VSD (1)	ASO with conduit (1)
TrArt (1)	Correction with conduit (1)
3. univentricular atrioventricular connection (11)	
TA (8); DILV (2); hypoplastic LV (1)	Fontan (11)
4. palliative operation- shunt (5)	
AVSD (1)	Glenn shunt (1)
DILV (1)	Blalock-Taussig shunt (1)
Univentricular heart (1)	Modified Blalock-Taussig shunt (1)
TA (1)	After Glenn shunt: AV shunt axillary (1)
TA + TGA (1)	After banding AP: PH (1)

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Table 1 (continued) Participating patients: main congenital heart disease groups (number of patients in parentheses)

Mild CHD	**n= 80**
Main diagnosis	
1. Current congenital heart disease (44)	
Small VSD (19)	
Mild PS (7)	
Small ASD (6)	
Mild AoS (6)	
AoV-bicuspid (3)	
Abnormal lung vein drainage (2)	
MVP (no regurgitation) (1)	
2. Congenital heart disease resolved (36)	

AoS= aortic stenosis; AoV= aortic valve; AP= pulmonary artery; ASD= atrial septal defect; ASO= arterial switch operation; AV= atrioventricular; AVSD= atrioventricular septal defect; CHD= congenital heart disease; DILV= double inlet left ventricle; DORV= double outlet right ventricle; L-TGA= congenitally corrected TGA; LV= left ventricle; MI= mitral insufficiency; MVP= mitral valvar prolapse; PA= pulmonary atresia; PH= pulmonary hypertension, PS= pulmonary stenosis; TA= tricuspid atresia; TGA= simple transposition of the great arteries; TrArt= common truncus arteriosus; VSD= ventricular septal defect

Group 2: patients with mild congenital heart disease

Patients with a mild congenital heart disease and those with a spontaneously resolved congenital heart lesion were included. None of them had needed an operation or intervention, as the malformation was diagnosed as "hemodynamically insignificant". For this group the hypothesis was that they would not experience heart-related problems in daily life. Therefore, for comparison with complex congenital heart disease patients, this group was considered as a control group.

In addition, *reference groups* were available for comparison with the patient data of job participation and job handicaps. For job participation, patients were compared with the general population concerning the overall percentages employed and the percentages employed divided into different age groups, as available from the central database of Statistics Netherlands (Source: CBS, Statistics Netherlands 1999). For job handicaps, patient data were compared with data of a non-impaired healthy reference group selected in a study by Andries et al. (n= 185).[10]

Questionnaire

Participants completed a questionnaire at home consisting of the following subjects.

Job participation and general data (i.e. age, sex, and education). Primary education concerned education and vocational training for 12 to 16 years olds, whereas higher education concerned secondary education and university.

Career-related problems, evaluated with 5 questions as part of the 'Difficulties in daily life'-module which covered different subjects, i.e. life and health insurance, education, employment, sports, and other daily activities.[11] For employment, the questions concerned: feeling restricted in the choice of a job, being excluded from a job, having given up on a job, being excluded from a job after medical examination, and not being promoted. If a problem was noted, it was specifically asked whether this resulted from the cardiac disorder.

Job handicaps, evaluated with a questionnaire that was developed and validated in other research projects within the TNO (Netherlands Organization for Applied Scientific Research) Vocational Handicap Research Program.[10,12,13] This questionnaire comprised three different parts: daily life activities (for all respondents), work activities and adaptations (for those presently working), and reasons to stop working and adaptations (for those with previous working experience).

Job handicaps were measured by comparing job demands and patient/ worker (dis)abilities, as well as adjustment at work. Therefore, firstly, difficulties with daily life activities were evaluated for 18 different activities divided into 5 domains (number of activities in parentheses): mobility (7), communication (3), mental abilities (3), hand/ arm movements (3), and physical

power (2). Then, it was assessed if these activities were performed at work, causing difficulties or not. By comparing daily life activities with the same activities done at work, the following classification for job handicaps was determined:

a. no difficulty: the activity caused no difficulty at all or was not applicable at work,
b. adaptation: the activity caused difficulty in daily life, but not at work due to certain adaptations having been made, or
c. job handicap: the activity caused difficulty both in daily life and at work, not withstanding adaptations having been made.

The division for this classification in the 2 groups of patients was described in percentages, for all domains together and for each domain specifically.

Statistical analyses

To examine the impact of the group of congenital heart disease (complex or mild), age, sex, and level of education on job participation, multiple logistic regression analyses were performed. These analyses were applied to all patients except those who were studying, since their reason for not having a job was going still to school, which is not related to the disease. Patients were categorised as 'working' if they had a paid job for more than 12 hours a week.

For job handicaps, chi-square analyses were used for comparison between patients with complex and patients with mild disease, and the non-impaired healthy reference group with regard to the overall and domain-specific percentages. Therefore, the percentages for no difficulty, adaptation, and handicap were dichotomised as follows: no difficulty vs. other, adaptation vs. other, and handicap vs. other.

Non-response analyses concerning age, sex, group of disease, and distance to the hospital was also performed using chi-square tests.

The level of significance was set at $p \leq 0.01$.

Results

Patients

Of the 224 selected patients, 160 agreed to participate. Four patients did not return their questionnaire, so analyses were based on a sample of 156 patients. This resulted in an overall response rate of 70%, with an 88% response in the

group of patients with complex congenital heart disease vs. 58% for the group of patients with mild congenital heart disease.

Table 1 lists the diagnoses and presents the number of patients per diagnosis or operation. All 156 participants, 73 men and 83 women, were aged between 17 and 32 years with a mean of 24.5 years (Table 2).

Table 2 Characteristics of patients

	Mild lesions (80)	**Complex lesions (76)**
age: mean, median, range	24.6, 25, 17-32 yr.	24.3, 24.5, 18-32 yr.
male	30 (38%)	43 (57%)
medical care >1 year ago	67 (84%)	6 (8%)
highest education finished		
primary education	21 (26%)	32 (42%)
continued education	59 (74%)	44 (58%)
daily life		
employed (more than 12 hours/week)	61 (76%)	43 (57%)
+disablement benefit partial	-	2 (3%)
student	13 (16%)	11 (15%)
+disablement benefit fully	-	4 (5%)
disablement benefit, no activities	-	10 (13%)
on sick leave (whiplash injury/pregnancy)	-	2 (3%)
housewife	2 (3%)	1 (1%)
+disablement benefit fully	2 (not related to CHD)	1 (1%)
unemployed	1 (1%)	2 (3%)
+disablement benefit partial	1 (not related to CHD)	-

CHD= congenital heart disease

Non-response analyses between participants and non-participants within the group of patients with mild congenital heart disease showed the following significant differences. Participants in this group lived closer to the hospital than non-participants and the response percentage of participating women (50 of 76; 66%) was significantly higher than that for men (30 of 62; 48%). In addition, the response in the group of patients whose mild cardiac lesion had resolved (36 of 49; 73%) differed significantly from that in the group with current mild disease (44 of 89; 49%).

Job participation

A total of 45 patients with complex disease had a paid job (59%), compared with 61 in the mild group (76%), which is significantly different. These percentages of job participation were similar to the reference group of the general population (66%). However, in the age group 25 to 29 years, job participation in the complex group (64%) was significantly lower ($p < 0.01$) than that in the general population (83%).

Twenty patients (of which 17 with complex congenital heart disease) received disablement benefit (fully or partial). For those with disablement benefit, the following numbers of complex congenital heart disease diagnosis were found: systemic right ventricle 3 of 38 (8%), conduit or mechanical prosthesis 6 of 22 (27%), univentricular atrioventricular connection 4 of 11 (36%), and palliative operation 4 of 5 (80%).

Multiple logistic regression on the sample excluding students (Table 3) showed that having a complex congenital heart disease and having finished only primary education, increased the risk for unemployment significantly ($p < 0.01$) and independently. Female gender and age had no significant influence.

Patients with complex disease worked in wide range of (sometimes physically demanding) occupations, e.g. welder, driver, electrician, waiter, cook, construction worker, nurse, or computer worker.

Table 3 Determination of factors concerned with employment, based on multiple regression analysis. Showing odds ratios for 'not working', in the sample excluding students, n= 131

	% not working	odds ratio	99% confidence interval
mild*	9%	4.8	1.2 - 19.6
complex	31%		
higher education*	12%	4.7	1.3 - 17.2
primary education	36%		
male*	14%	3.5	0.9 - 13.4
female	25%		
age group 17-24 years*	20%	1.0	0.3 - 3.7
age group 25-32 years	20%		

*reference category

Career-related problems

For patients with a complex congenital heart disease, the following problems were found: feeling restricted in the choice of a job (39; 51%), being excluded from a job (13; 17%), having given up on a job (12; 16%), being excluded from a job after medical examination (7; 9%), and not being promoted (2; 3%). Considering all these problems together, 42 patients (55%) stated to have experienced at least one problem in their working career as a result of their heart disease. Main reasons to stop working were physical disabilities, tiredness, and emotional problems.

Only one patient with a mild aortic stenosis had to choose another job. He wanted to join the army, which was not possible with this defect. No other cardiac-related problem was mentioned in the mild congenital heart disease group.

Job handicaps

For job handicaps, no significant differences were found between the group of patients with a complex disease and that with a mild disease. However, the reference group had a significantly ($p < 0.01$) lower percentage of handicaps (7; 4%) for the domain mobility than the group of patients with a complex disease (5; 14%), and the group with a mild disease (7; 12%). No

significant differences were found for the other domains at work. In contrast to the patients with complex disease, all 7 mild congenital heart disease patients who experienced a handicap suffered from a non-cardiac disease, such as knee or shoulder problems, or asthma. The diagnoses of the 5 patients with complex disease who experienced handicaps were systemic right ventricle (3), and conduit or mechanical prosthesis (2).

Specific adaptations

For those patients with complex disease who had to stop working or wanted another job in relation to their heart disease, mostly the work was physically too strenuous, patients had emotional problems, or problems with transport. Only 2 patients mentioned specific adjustments for disease-related handicaps at work, but 10 patients would have appreciated adaptations (12 of those 44 who answered the questionnaire; 27%).

Most important of these adaptations were more flexibility in working hours, reduced time-pressure demands, adapted working hours, and increased freedom to organize one's work. In the reference group, 15% wanted at least one of these adaptations. Patients with mild disease did not mention any specific adaptation related to their heart defect.

Discussion

This study evaluated job participation, career-related problems, and job handicaps for adults with complex congenital heart disease, compared with those of adults with mild congenital heart disease and reference groups.

Job participation

The majority of patients with complex disease participated in economic life. However, the percentage of working patients with complex congenital heart disease is significantly lower than in the group of patients with mild disease (58% versus 76%, respectively) and than in the general population in the age group 25 to 29 years (64% versus 83%, respectively). Excluding two studies that presented extreme percentages,[2,14] and focusing solely on the studies with patients with complex congenital heart disease, the percentages of job participation ranged from 71%-84%[5,6,15-17]; this is somewhat higher than in the present study.

The percentage of complex congenital heart disease patients that received disablement benefit (26%) is higher than reported in other studies: 10-15%.[5,6,15] This is probably due to the high level of social security in the Netherlands compared with other countries.

Multivariate analyses showed a relation between group of congenital heart disease and education and job participation; this is in accordance with a previous study.[12] Although cause and consequence cannot be predicted from this cross sectional study, it seems plausible to assume from these results that having a complex congenital heart disease increases the risk for unemployment and that stimulating patients to complete the highest possible educational level will improve their job perspectives.

Career-related problems

More than half of the patients with complex congenital heart disease experienced problems in their working career. Previous studies have reported similar problems;[3,15,18,19] however, because of differences in diagnosis groups studied, and questions asked, specific comparison is difficult. In our study, patients with complex disease revealed significantly more career-related problems than patients with mild congenital heart disease.

Job handicaps

For job handicaps, significant differences between both patient groups and the reference group were found for mobility. None of the handicapped patients with complex congenital heart disease had another chronic disease, in contrast to the group of patients with a mild disease of which all handicapped patients had a non-cardiac chronic disease. Therefore, it is assumed that for the patients with a complex congenital heart disease, handicaps were related to the cardiac defect. As a specific reason to quit the job, physical problems were frequently reported. Most important specific adaptations mentioned concerned working time and workload.

The results of this study allow us to make recommendations specifically for the group of patients with complex congenital heart disease. Career counselling and utilisation of job adjustments taking into account physical abilities are important in this patient group to prevent unemployment and dependency on disablement benefits, and to reduce work-related problems. Using existing guidelines for counselling might be helpful.[20] The patients

mentioned specific adaptations mainly concerning working time and workload. Creating possibilities to do work at home could also reduce or prevent job-related problems.

Another important issue is to encourage patients to complete higher education, since this is associated with higher level of job participation.
Patients should also be made aware that questioning medical health during an interview for a job is prohibited according to the Dutch Medical Examinations Act (1997). In addition, in the Netherlands specific financial advantages are available for employers who hire persons with a handicap. In the USA, the National Rehabilitation Act (1973) prohibits discrimination because of a pre-existing condition.[21,22]

Celermajer et al. found inconsistencies in job policy that might be due to lack of appropriate guidelines for the outcome of adults with a congenital heart disease.[23] This could also cause unnecessary problems and even unemployment. Therefore, informing employers and company physicians about this outcome is recommended.

Advising patients about these issues is an important task for the paediatric or congenital cardiologist dealing with adult patients.

Remarks and study limitations

The results of this study might have been affected by selection bias. The difference in response percentages between patients with complex (88%) and mild disease (58%) can be explained. Most patients with mild disease had their latest medical examination more than one year previously and had to visit the hospital for update cardiac examination, thus involving more effort than just answering the questionnaire. This could also explain why participants in the mild group lived closer to the hospital than non-participants. In addition, it might be that more patients with the possibility to make this visit were unemployed. This could then explain the difference in the response percentage of participating women (66%) and men (49%), assuming that more men have a job than women do. Therefore, job participation in the mild group, might still be underestimated. The response in the group of patients whose mild cardiac lesion had resolved (74%) differed from that in the group with actual mild congenital heart disease (50%). During cardiac re-examination, it was observed that most patients asked questions or expressed worries that they had lived

with for several years. Possibly, patients with relatively more difficulties or questions might have joined the study, influencing results towards more difficulties in this participating group of patients with mild disease than in the entire group. As for the group of patients with mild disease, it might also be assumed that a negative selection of complex congenital heart disease patients with relatively more unemployment, and more difficulties or questions than in the total group took place. This influence on employment will, however, only be minor, since only few participants had to visit the hospital for re-examination and therefore arguments for time efforts do not count.

Studying only surviving patients implies a positive selection bias. This applies only for the group of patients with complex disease, since no cardiac death was reported in the group of patients with mild disease. The present study focused on relatively young patients. Since studies on medical outcome have shown a decreasing physical condition in patients with complex congenital heart disease after the age of 30 years, most likely the number of reported problems would increase with age. Furthermore, medical developments allow treating more patients with more complicated congenital heart diseases than in the past. It is expected that a new generation of patients will experience even more career-related problems and more unemployment than the current study shows.

Conclusions

Patients with complex congenital heart disease have reduced job participation as compared with patients with mild congenital heart disease and the general population. Many of them receive disablement benefit, or experience career problems or job handicaps (mainly concerning mobility). Career counselling focusing on physical abilities and level of education may help to prevent unemployment and dependency on disablement benefits, and to reduce job-related problems.

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Chapter 9

Summary and conclusions

Introduction (Chapter 1)

Advances in medical and surgical treatment have led to increased survival in patients with congenital heart disease. Therefore, it has become increasingly important to address these patients' health-related quality of life and to focus on several psychosocial issues.

The aim of this thesis was to describe the long-term outcome in congenital heart disease in terms of health-related quality of life and other psychosocial issues such as employment, disease-related knowledge and difficulties in daily life. With this information, methods to optimise health-related quality of life can be sought for, and patient education and career counselling can be improved.

In 1996, the Leiden Center for Child Health and Pediatrics (TNO[a]- LUMC[b]) started a study in patients with mild congenital heart disease. In this study, a questionnaire was used comprising validated measurements (the TNO-AZL[c] Adult Quality of Life (TAAQOL)- questionnaire, and the Short Form-36 (SF-36) as well as questions concerning disease-specific health-related quality of life and disease-related limitations. This study (Chapter 3) was then extended and the questionnaire further developed. Health-related quality of life and several psychosocial aspects, such as employment, disease-related knowledge, and limitations in daily life were evaluated in different groups of adults with a congenital heart defect (Chapters 4 to 8).

[a] TNO : Netherlands Organization for Applied Scientific Research, Prevention and Health
[b] LUMC : Leiden University Medical Center, Willem-Alexander Department of Pediatrics
[c] AZL : Academic Hospital Leiden, now known as Leiden University Medical Center

Summary of the results

Chapter 2 reviewed definitions and measures of health-related quality of life in studies with congenital heart disease patients and summarised outcome of health-related quality of life in these patients.

In total, 69 papers were studied in detail. In 46% (32) of these studies no description of health-related quality of life was provided. Moreover, given definitions of health-related quality of life varied widely. Seven research-groups conceptually defined health-related quality of life as a self-reported

multidimensional construct, and 7 (partly other) research-groups had used a *validated* self-report multidimensional measure. Four studies focussed on the patients' own perception of their health-related quality of life.

In general, most patients with a congenital heart disease (even those with complex disease) felt they were healthy. However, subgroups (such as older patients or patients with cyanosis) appeared to do worse.

It was concluded that health-related quality of life in research with congenital heart disease patients is seldom defined clearly and measures often aim at different targets. Therefore, these studies should be compared with great caution. Health-related quality of life outcome is surprisingly positive in most studies that measured congenital heart disease patients' own perception. Coping strategies might explain this.

Chapter 3 presented the study, in which the first draft of the congenital heart disease questionnaire was used in 94 young adults with mild congenital heart disease, who were all currently under cardiac surveillance in the Leiden University Medical Center. Social impediments experienced by these patients were studied and their health-related quality of life (measured with the TAAQOL) and health status (measured with the SF-36) were compared with a matched group (age, sex) from the general population.

A substantial part of the congenital heart disease patients experienced impediments, for example at school (19%), with medical examinations for getting a job or insurance (19%), during free time (15%), in choosing (13%) or performing a job (9%), and taking out a life insurance policy (8%).

Those congenital heart disease patients that experienced social impediments reported a health-related quality of life and subjective health status that was comparable with the general population. Congenital heart disease patients without restrictions reported even a better health-related quality of life for 6 of the 12 TAAQOL-scales than the general population. This last result might be explained by coping mechanisms.

It was concluded that having a mild congenital heart disease generally does not influence health-related quality of life negatively. However, in adulthood, patients with mild congenital heart disease may encounter obstacles in society. The information and education for the patients and their parents should be directed towards overcoming these obstacles.

Chapter 4 described the impact of previously operated complex congenital heart disease (n= 78) on health-related quality of life (measured with the TAAQOL) and subjective health status (measured with the SF-36) and determined the relation between these measures and physical indices.

Health-related quality of life of the patients was significantly worse than that of the general population in the domains gross motor functioning and vitality ($p < 0.01$). Correlations between health-related quality of life and physical indices were poor. Patients had a significantly worse subjective health status than the general population on the domains physical functioning, role functioning physical, vitality, and general health perceptions ($p < 0.01$). Correlations between subjective health status and physical indices were weak.

These results indicate that these patients need specific attention for their physical quality of life. Objectively determined physical indices are only weakly related to health-related quality of life. Therefore, when evaluating health-related quality of life, dedicated questionnaires such as the TAAQOL should be used.

Chapter 5 focussed on 82 adults with hemodynamically insignificant congenital heart disease and studied their health-related quality of life and subjective health status (as compared with the general population), difficulties in daily life, and cardiac status (i.e. regularity of follow-up, current diagnosis, and antibiotic regimen) in order to discuss the need for continuing their cardiac surveillance.

The patients' health-related quality of life and health status did not differ from those of the general population. Nevertheless, patients experienced unnecessary difficulties with choice of sport, education or obtaining insurance cover. After clinical re-evaluation, diagnosis and antibiotic regimen had to be changed in 9 participants (11%).

It was concluded that patients with a mild congenital heart lesion consider themselves to be in good health. However, to fine tune diagnosis and update the patients' information, at least a cardiological assessment at age 16 to 18 is suggested. In this way, patients might be protected from unnecessary difficulties, such as restrictions for sport or unjustifiable high rates for insurance.

In **Chapter 6**, the development, and reliability and validity of a congenital heart disease-specific health-related quality of life module was presented: the CHD-TAAQOL (Congenital Heart Disease-TAAQOL).

The CHD-TAAQOL belongs to the generic health-related quality of life-instrument (TAAQOL), in which health-related quality of life was defined as health status weighted by emotional response to problems in health status. Items for the CHD-TAAQOL were based on the pilot study (Chapter 3), interviews with patients, expert meetings and literature.

Then, the CHD-TAAQOL was tested in 156 patients with complex or mild congenital heart disease. Reliability, scale structure, convergent validity (correlation with the TAAQOL- and SF-36- domains), criterion validity (impact of disease) and concept validity (the difference between health status and the emotional response) were calculated. The CHD-TAAQOL module consists of three scales (items): 'Symptoms' (9), 'Impact Cardiac Surveillance' (7), and 'Worries' (10). Cronbach's alfa ranged from 0.77 to 0.82, which is sufficient for the use in groups. Scale structure was confirmed by principal component analysis, corrected item-scale and interscale correlations. Convergent validity was good. Criterion validity showed significantly lower scores for complex congenital heart disease patients compared with patients with mild congenital heart disease. Overall, only 55% of reported health status problems were associated with negative emotions, which is an argument for assessing health-related quality of life as a concept distinct from health status.

It was concluded that the CHD-TAAQOL module together with the generic TAAQOL can be used to assess group differences for disease-specific health-related quality of life in congenital heart disease patients. Testing reliability and validity of the CHD-TAAQOL shows satisfactory results. However, to detect changes in health-related quality of life over time, further research has to be performed.

Chapter 7 evaluated in adults with mild or complex congenital heart disease (n= 156) difficulties in daily life, and satisfaction with level of knowledge about their disease.

It showed that 11% of patients with mild malformations experienced difficulties related to their disease, which is significantly less ($p< 0.001$) compared with the percentage (87%) of difficulties for adults with complex disease. Those patients with complex malformations frequently felt restricted in choices because of their disease in areas such as sport (59%), employment

(51%), and education (34%). Other difficulties reported were: paying a higher premium for life insurance (29%), having to give up on a sport (28%), and being excluded from a job (18%). Depending on the item, between one-fifth and two-thirds of participants reported gaps in knowledge, most frequently for "causes of congenital cardiac disease", "future consequences", and "family planning". For 53% of patients with mild anomalies, and 93% of complex congenital heart disease patients, the cardiologist is the most important source of information.

In conclusion, a minority of adults with mild, and a majority of those with complex congenital heart disease report difficulties in daily life. A substantial number of these patients feel that they have an inadequate level of knowledge about their disease. Our results suggest the need for a specific programme of counselling.

Chapter 8 compared job participation, career and actual job problems in adults with complex congenital heart disease (n= 76), with those of patients with mild congenital heart disease (n= 80) and reference groups, using a self-report questionnaire.

In the study-groups, 59% (45) of patients with complex congenital heart disease had a paid job compared with 76% (61) of those with mild congenital heart disease ($p< 0.01$). Patients with complex disease older than 25 years had a significantly lower job participation (64%) than the general population (83%). Multiple logistic regression showed that severity of disease and level of education were significantly ($p< 0.01$) and independently related to job participation (odds ratios 4.8, and 4.7, respectively).

Of the complex congenital heart disease patients, 55% (42) experienced disease-related career problems, in contrast to only one patient with mild disease. Both congenital heart disease groups showed more job-related handicaps for mobility than the reference group ($p< 0.01$). However, in the mild congenital heart disease group, these could be attributed to additional non-cardiac diseases.

It was concluded that patients with complex congenital heart disease have reduced job participation as compared with patients with mild congenital heart disease and the general population. Many receive disablement benefit, or experience career problems or job handicaps. Career counselling focusing on physical abilities and level of education may help to prevent or reduce these job-related problems.

Considerations

Study design and generalisability

In order to prevent selection bias, our procedure to select patients aimed at a representative sample of congenital heart disease patients from the entire Dutch population and not just those who currently visited the Leiden University Medical Center. Indeed it seemed that many of the participating patients had moved to other areas in the Netherlands and currently had their check-ups in other hospitals or were not under cardiac surveillance anymore. However, due to non-response bias, the participating sample (especially that of patients with mild congenital heart disease) might have been a negative selection of patients with relatively more unemployment, and more difficulties or questions than in the total group.

Moreover, within the samples several diagnosis groups were included. It is likely that between these groups, differences in outcome occur. Small sample sizes per diagnosis group precluded specification of the results per diagnosis. Therefore, it is important to continue this type of research in specific diagnosis groups.

Definition and instrument used to assess health-related quality of life

The instrument used for the measurement of health-related quality of life was based on the principle that health-related quality of life differs from health status. Therefore, the TAAQOL-questionnaire firstly questioned health status, followed by a question concerning the emotional impact associated with a health status problem. Indeed it was found that only part of the health status problems resulted in a negative affective response to those problems (for the TAAQOL, 47% to 81% of all problems were associated with a negative emotion). The distinction between these two concepts is therefore justified.

General conclusions

The following can be concluded from the studies described in this thesis.

The concept of health-related quality of life in research with congenital heart disease patients is seldom defined clearly and measures often aim at different targets. These studies should be compared with great caution. Moreover, measures for health-related quality of life are only weakly related to medical variables. Therefore, a dedicated and validated questionnaire should be used in future research on health-related quality of life.

A reliable and valid disease-specific health-related quality of life instrument is presented (CHD-TAAQOL), which can be used in addition to the generic instrument (TAAQOL).

Patients with complex congenital heart disease experience a reduced physical health-related quality of life, and have obvious problems with career and employment. The studied group of patients with mild congenital heart disease however, feels well but experiences unnecessary difficulties in daily life. Both groups will need specific attention in order to overcome these problems.

The above conclusions lead to the following recommendations.

Recommendations for clinical practice

- Since the *appreciation* of functioning differs from functioning per se (Chapter 2 and 4), implementation of quality of life- questionnaires at medical consultations should be considered, to facilitate treatment decisions, and for increasing satisfaction with consultation and patient compliance.
- Specifically for patients with mild congenital heart disease, knowledge about the cause and consequences of their cardiac defect is experienced as insufficient (Chapter 3, 5 and 7). Therefore, more attention is needed for this lack of knowledge. The implementation of the use of a "health passport" in paediatric cardiology could be helpful for improvement of patient remembrance.

Recommendations for clinical practice (continued)

- We recommend a routine consultation in patients with mild congenital heart disease at age 16 to 18 years as an ideal opportunity to discuss daily life issues and to try to prevent many of the reported problems in later life (Chapter 3, 5 and 7).
- Career counselling should be focused on the patients' physical abilities, in order to prevent or reduce unemployment, career problems and handicaps (Chapter 8). In addition, the child should be advised to follow the highest possible education.

Recommendations for future research

- To improve the patients' health-related quality of life, it is recommended to develop and evaluate a rehabilitation/ sports program for adults with complex congenital heart disease (Chapter 4). In addition, a study on the determinants of health-related quality of life is recommended (Chapter 2 and 3). More knowledge about the influence of different variables such as personality characteristics, coping, social support, or self-esteem will help to evolve theories about health-related quality of life and then to develop interventions to improve the patients' health-related quality of life.
- Health-related quality of life research in specific groups of congenital heart disease patients using well-defined and validated measures (such as the TAAQOL and the CHD-TAAQOL) is important (Chapter 2 and 6) in order to compare such studies.

Chapter 10

Nederlandse samenvatting:

Kwaliteit van leven bij volwassenen met een aangeboren hartafwijking

Introductie (Hoofdstuk 1)

Door ontwikkelingen in de medische zorg en chirurgische behandeling is de overleving van patiënten met een ernstige aangeboren hartafwijking toegenomen. Daarom wordt het steeds belangrijker om de gezondheidsgerelateerde kwaliteit van leven te bepalen en tevens diverse psychosociale aspecten te evalueren.

Het doel van dit proefschrift is om bij volwassenen met een aangeboren hartafwijking de gezondheidsgerelateerde kwaliteit van leven te bepalen en daarnaast andere psychosociale aspecten zoals de werksituatie, beperkingen in het dagelijks leven, en ziektegerelateerde kennis te beschrijven. Hiermee kunnen methoden gezocht worden om de kwaliteit van leven te optimaliseren en de patiënteninformatie en -educatie te verbeteren.

Definities

Subjectieve gezondheidstoestand

De subjectieve gezondheidstoestand werd gemeten met de goed gevalideerde vragenlijst SF-36 (Short Form-36 items). Deze vragenlijst wordt door de patiënt zelf ingevuld en vraagt naar het functioneren in diverse domeinen (zoals fysiek, emotioneel, sociaal etc.).

Gezondheidsgerelateerde kwaliteit van leven

Hoewel een algemeen geaccepteerde definitie van gezondheidsgerelateerde kwaliteit van leven ontbreekt, werd aan de hand van diverse publicaties de volgende omschrijving geformuleerd: het gaat om de door de persoon waargenomen problemen in diens gezondheidstoestand (voor diverse domeinen), gecombineerd met de affectieve reactie op dergelijke problemen.

De vragenlijst om gezondheidsgerelateerde kwaliteit van leven te meten in dit proefschrift, is de TAAQOL (TNO[a]-AZL[b] Adult Quality of Life), een goed gevalideerde vragenlijst. Deze vragenlijst wordt ingevuld door de patiënt zelf en vraagt in eerste instantie naar het functioneren en bij disfunctioneren vervolgens naar de beleving daarvan.

[a] TNO: Nederlandse Organisatie voor Toegepast Natuurwetenschappelijk Onderzoek, te Leiden
[b] AZL: Academisch Ziekenhuis Leiden, tegenwoordig Leids Universitair Medisch Centrum (LUMC)

In 1996 startte het Leiden Center for Child Health and Pediatrics (TNO-PG- LUMC, afdeling Kindergeneeskunde) een studie met patiënten met een milde aangeboren hartafwijking. In deze studie werd de eerste versie van de vragenlijst voor aangeboren hartafwijkingen gebruikt. Deze vragenlijst bestond uit gevalideerde instrumenten, zoals TAAQOL en de SF-36, maar ook uit nieuwe ziektespecifieke onderdelen. Deze studie (Hoofdstuk 3) werd later uitgebreid en de vragenlijst werd verder ontwikkeld. Kwaliteit van leven en andere psychosociale aspecten, zoals werk, kennis en sociale beperkingen werden vervolgens geëvalueerd in verschillende groepen volwassenen met een aangeboren hartafwijking (Hoofdstukken 4 tot en met 8).

Patiëntengroepen
In dit proefschrift worden 3 patiëntengroepen bestudeerd.

Groep 1: 78 patiënten met een eerder geopereerde complexe aangeboren hartafwijking (17 tot 32 jaar). Al deze patiënten zijn aan hun hart geopereerd, maar het eindresultaat leverde geen anatomisch gecorrigeerd hart op zoals dat beschreven is door de segmentale analyse. Voorbeelden zijn: patiënten na Mustard of Senning operatie voor transpositie van de grote vaten, of na een Fontan operatie voor tricuspidalis atresie.

Groep 2: patiënten met een milde aangeboren hartafwijking (17 tot 32 jaar). Deze patiënten hebben geen van allen een operatie nodig (gehad). Voorbeelden zijn: patiënten met een klein ventrikel septum defect, of milde aorta stenose.
2A: 94 patiënten met een 'hemodynamisch niet significant' defect, die ten tijde van het onderzoek onder controle waren in het LUMC.
2B: 82 patiënten, met een 'hemodynamisch niet significante' of spontaan verdwenen aangeboren hartafwijking, die geselecteerd werden uit het archief en niet allen meer onder controle waren in het LUMC. Het was te verwachten dat deze groep geen hartgerelateerde problemen zou ondervinden en daarom werd deze groep ook wel als controlegroep afgezet tegen groep 1.

Referentiegroepen

Daar waar deze beschikbaar waren, werden de patiëntendata vergeleken met gegevens van de algemene populatie.

Samenvatting van de resultaten

In **Hoofdstuk 2** wordt een literatuuroverzicht gepresenteerd over de literatuur betreffende kwaliteit van leven bij aangeboren hartafwijkingen. In dit overzicht ging de speciale aandacht uit naar de definiëring van het concept kwaliteit van leven en de gebruikte meetmethoden. Tevens werden de uitkomsten samengevat.

In totaal werden 69 publicaties die kwaliteit van leven als uitkomst suggereerden, in detail bestudeerd. Hiervan gaven 32 (46%) geen beschrijving van het concept. Daarnaast waren de definities van kwaliteit van leven zeer variabel. Zeven onderzoeksgroepen definieerden kwaliteit van leven als een multidimensioneel samengestelde parameter. Bij zeven (deels andere) onderzoeken werd een goed gevalideerde vragenlijst gebruikt. Vier onderzoeken richtten zich op de affectieve reactie van de patiënt op de eigen gezondheidstoestand.

De uitkomsten van de onderzoeken lieten zien dat de patiënten met een aangeboren hartafwijking (zelfs bij een complexe hartafwijking) zich in het algemeen goed voelen. Bepaalde subgroepen, zoals cyanotische of oudere patiënten, lijken zich echter slechter te voelen.

Uit dit overzicht valt te concluderen dat kwaliteit van leven in de literatuur zelden duidelijk gedefinieerd wordt en dat de onderzoeken zich richten op diverse verschillende doelen. Daarom is voorzichtigheid geboden bij het vergelijken van de uitkomsten van dergelijke onderzoeken. In het algemeen is de kwaliteit van leven uitkomst voor patiënten met een aangeboren hartafwijking opmerkelijk positief. Dit kan verklaard worden door coping strategieën (de wijze waarop je met je problemen omgaat).

In **Hoofdstuk 3** worden de resultaten beschreven van het onderzoek waarin de eerste versie van de vragenlijst voor patiënten met een aangeboren hartafwijking werd gebruikt voor jong volwassenen met een milde afwijking (n= 94). Bij deze patiënten werd bepaald welke beperkingen zij hebben ervaren in het dagelijks leven. Tevens werden hun gezondheidsgerelateerde kwaliteit van leven (gemeten met de TAAQOL) en subjectieve gezondheidsstatus (gemeten met de SF-36) vergeleken met een gematchte groep (leeftijd en geslacht) uit de algemene populatie.

Een substantieel deel van de patiëntengroep liep tegen beperkingen aan, zoals op school (19%), bij medisch onderzoek voor werk of een verzekering

(19%), in de vrije tijd (15%), bij het kiezen (13%) of uitvoeren (9%) van werk of bij het afsluiten van een levensverzekering (8%).

De groep die dergelijke beperkingen ondervond rapporteerde een gezondheidsgerelateerde kwaliteit van leven en subjectieve gezondheidsstatus vergelijkbaar met de algemene populatie. Patiënten zonder deze beperkingen bleken zelfs een betere gezondheidsgerelateerde kwaliteit van leven (voor 6 van de 12 TAAQOL schalen) te rapporteren dan de algemene populatie. Dit kan verklaard worden door coping mechanismen.

De conclusie van dit onderzoek luidt dat een milde aangeboren hartafwijking de gezondheidsgerelateerde kwaliteit van leven in het algemeen niet negatief beïnvloedt. Er zijn echter wel een aantal beperkingen waar deze mensen tegenaan kunnen lopen. Informatie en educatie voor de patiënt en ouders moeten gericht zijn op het voorkomen van deze beperkingen.

In **Hoofdstuk 4** wordt een groep patiënten (n= 78) met een eerder geopereerde complexe aangeboren hartafwijking beschreven. Hun gezondheidsgerelateerde kwaliteit van leven (gemeten met de TAAQOL) en subjectieve gezondheidsstatus (gemeten met de SF-36) werden vergeleken met die van de algemene populatie. Tevens werd de relatie tussen deze maten en de objectieve gezondheidstoestand bepaald.

De gezondheidsgerelateerde kwaliteit van leven van de patiënten was significant slechter dan die in de algemene populatie voor de domeinen grove motoriek en vitaliteit ($p < 0,01$). Correlaties tussen de gezondheidsgerelateerde kwaliteit van leven en de objectieve gezondheidstoestand waren laag. Patiënten hadden een significant slechtere subjectieve gezondheidsstatus dan de algemene populatie voor de domeinen fysiek functioneren, rolbeperkingen fysiek, vitaliteit en algemene gezondheidsperceptie ($p < 0,01$). De correlaties tussen de subjectieve gezondheidsstatus en de objectieve gezondheidstoestand waren eveneens laag.

Deze resultaten benadrukken dat er speciale aandacht moet zijn voor de fysieke aspecten van de gezondheidsgerelateerde kwaliteit van leven van deze patiënten. Omdat de relatie tussen kwaliteit van leven en objectieve gezondheidstoestand laag is, zullen tevens specifieke kwaliteit van leven vragenlijsten (zoals de TAAQOL) gebruikt moeten worden om een compleet beeld van de patiënt te krijgen.

In **Hoofdstuk 5** wordt de noodzaak van cardiologische vervolgcontrole voor volwassenen met een milde aangeboren hartafwijking (n= 82) bediscussieerd. Hiervoor werden de gezondheidsgerelateerde kwaliteit van leven en subjectieve gezondheidsstatus vergeleken met de algemene populatie. Tevens werden de beperkingen in het dagelijks leven en de cardiale status (regelmaat van medische controle, huidige diagnose en antibiotica regime) geëvalueerd.

De gezondheidsgerelateerde kwaliteit van leven en subjectieve gezondheidsstatus van de patiënten verschilden niet van die van de algemene populatie. Het bleek echter dat de patiëntengroep onnodige beperkingen ondervond met bijvoorbeeld de keuze van sport, de hoogte van verzekeringspremies en deelname aan onderwijs. Na medische herevaluatie bleek dat de diagnose en het antibiotica regime gewijzigd moest worden bij 9 patiënten (11%).

Concluderend voelen de patiënten met een milde aangeboren hartafwijking zich goed gezond. Echter, om de diagnose te bevestigen en daarbij tevens de juiste informatie te verstrekken is het belangrijk deze groep te controleren tenminste op de leeftijd van 16 tot 18 jaar. Op deze manier kunnen volwassen patiënten beschermd worden tegen onnodige beperkingen bij sport, of voor onrechtvaardig hogere premies voor verzekeringen.

In **Hoofdstuk 6** wordt de ontwikkeling van een ziekte-specifieke kwaliteit van leven vragenlijst beschreven: de Congenital Heart Disease- TNO-AZL Adult Quality of Life (CHD-TAAQOL). Tevens werden betrouwbaarheid en validiteit van deze vragenlijst bepaald.

De CHD-TAAQOL hoort bij de generieke kwaliteit van leven vragenlijst, de TAAQOL. Items zijn gebaseerd op eerder onderzoek (Hoofdstuk 3), interviews met patiënten, expert meetings en literatuur. De CHD-TAAQOL werd getest in 156 patiënten met een milde of complexe aangeboren hartafwijking.
De betrouwbaarheid van de schalen, schaalstructuur, convergentie (relatie met TAAQOL en SF-36), criterium (effect van ziekte) en concept (verschil tussen gezondheidsstatus en de affectieve reactie) validiteit werden berekend.

De CHD-TAAQOL module bestaat uit 3 schalen (aantal items tussen haakjes): Symptomen (9), Impact cardiale controle (7) en Zorgen (10). De Cronbach's alfa was 0,77 tot 0,82, wat goed is voor gebruik op groepsniveau. De schaalstructuur werd bevestigd met principale component analyses, gecorrigeerde item-schaal en interschaal correlaties. De convergentie validiteit was goed. De criterium validiteit liet significant lagere scores zien voor

patiënten met een complexe hartafwijking vergeleken met die van patiënten met een milde hartafwijking. In totaal was slechts 55% van de gerapporteerde gezondheidsproblemen geassocieerd met negatieve emoties, hetgeen bevestigt dat gezondheidsgerelateerde kwaliteit van leven niet uitwisselbaar is met subjectieve gezondheidsstatus.

Men kan hieruit concluderen dat de CHD-TAAQOL module samen met de TAAQOL gebruikt kan worden om groepsverschillen in gezondheidsgerelateerde kwaliteit van leven te meten bij patiënten met een aangeboren hartafwijking. De testen van betrouwbaarheid en validiteit van de CHD-TAAQOL laten bevredigende resultaten zien.

In **Hoofdstuk 7** worden de beperkingen in het dagelijks leven en de tevredenheid met het kennisniveau over de ziekte bij patiënten met een milde (n= 80) of complexe (n= 76) aangeboren hartafwijking geëvalueerd.

Het bleek dat 11% van de patiënten met een milde afwijking ooit beperkingen gerelateerd aan de hartafwijking had ondervonden. Dit was significant minder ($p< 0,01$) dan het percentage patiënten met een complexe afwijking dat beperkingen meldde (87%). Deze laatste groep gaf beperkingen aan in de keuzes voor sport (59%), werk (51%) en scholing (34%). Andere gerapporteerde beperkingen waren: het betalen van hogere premies voor een levensverzekering (29%), een sport opgeven (28%) en afgewezen worden voor een baan (18%).

Afhankelijk van het onderwerp, gaf 20% tot 64% van alle patiënten aan onvoldoende kennis te hebben over hun hartafwijking, met name wat betreft de onderwerpen: oorzaken van de hartafwijking, gevolgen voor de toekomst en familieplanning. Voor 53% van de patiënten met een milde afwijking en 93% van de patiënten met een complexe afwijking was de cardioloog de belangrijkste bron van informatie.

Concluderend kan gesteld worden dat een minderheid van de volwassenen met een milde aangeboren hartafwijking en een meerderheid van volwassenen met een complexe afwijking beperkingen rapporteert in het dagelijks leven. Een substantieel deel van alle patiënten geeft een inadequaat kennisniveau aan. Deze resultaten suggereren de behoefte aan een speciaal voorlichtingsprogramma voor volwassenen met een aangeboren hartafwijking.

In **Hoofdstuk 8** worden vergelijkingen gemaakt tussen volwassenen met een complexe aangeboren hartafwijking (n= 76), volwassenen met een milde afwijking (n= 80) en de algemene populatie, voor wat betreft het percentage werkenden, en de ervaren problemen in de carrière of op de werkplek (handicaps).

In de onderzoeksgroepen had 59% van de patiënten met een complexe hartafwijking een betaalde baan vergeleken met 76% van de patiënten met een milde afwijking ($p < 0,01$). Voor patiënten ouder dan 25 jaar was het percentage in de complexe groep (64%) significant lager dan het percentage in de algemene bevolking (83%). Multipele regressie analyse liet zien dat de ernst van de ziekte en het onderwijsniveau significant ($p < 0,01$) en onafhankelijk gerelateerd zijn aan de kans op werk (odds ratios van 4,8 en 4,7 respectievelijk).

Van de patiënten met een complexe aangeboren hartafwijking had 55% (42/76) ziekte-gerelateerde problemen in de carrière, tegenover slechts 1% (1/80) van de patiënten met een milde afwijking.

Beide patiëntengroepen hadden meer werkgerelateerde handicaps voor mobiliteit dan de referentiegroep ($p < 0,01$). Dit was in de groep met een milde aangeboren hartafwijking zeer waarschijnlijk te wijten aan diverse niet cardiale aandoeningen.

Uit dit onderzoek valt te concluderen dat volwassenen met een complexe aangeboren hartafwijking minder participeren op de arbeidsmarkt dan patiënten met een milde afwijking en de algemene populatie. Velen ontvangen een WAO-uitkering, ervaren problemen in de carrière, of hebben handicaps op het werk. Om deze problemen te voorkomen of te verminderen is het van belang om patiënten met een aangeboren hartafwijking op jonge leeftijd loopbaan begeleiding te bieden, zodat eventuele fysieke beperkingen en adviezen voor de hoogst mogelijke opleiding meegenomen kunnen worden in beslissingen voor de toekomst.

Conclusies van dit proefschrift (Hoofdstuk 9)

De volgende conclusies kunnen uit de beschreven onderzoeken getrokken worden.

Het concept gezondheidsgerelateerde kwaliteit van leven in onderzoek bij aangeboren hartafwijkingen is zelden duidelijk gedefinieerd en meetmethoden voor kwaliteit van leven richten zich op diverse verschillende doelen. Tevens blijkt dat kwaliteit van leven slecht gerelateerd is aan medisch fysieke indices. Daarom is het belangrijk specifieke en gevalideerde vragenlijsten te gebruiken voor onderzoek van gezondheidsgerelateerde kwaliteit van leven.

Een betrouwbare en gevalideerde ziekte-specifieke vragenlijst werd gepresenteerd (CHD-TAAQOL), die gebruikt kan worden tezamen met de algemene vragenlijst, de TAAQOL.

Patiënten met een complexe aangeboren hartafwijking ervaren een verminderde fysieke kwaliteit van leven en hebben evidente problemen op de arbeidsmarkt. De bestudeerde groep patiënten met een milde afwijking voelt zich goed, maar ervaart onnodig beperkingen in het dagelijks leven. Beide groepen verdienen extra aandacht om deze problemen te voorkomen.

Bovenstaande conclusies leiden tot de volgende aanbevelingen.

Aanbevelingen voor de klinische praktijk

- Daar de *beleving* van functioneren evident anders is dan het functioneren op zich (Hoofdstuk 2 en 4), dient het afnemen van een kwaliteit van leven vragenlijst bij een medisch consult te worden overwogen, voor het bepalen van het beleid, en het verhogen van de 'patient compliance' en de tevredenheid van de patiënt met het consult.
- Vooral bij patiënten met een milde aangeboren hartafwijking wordt de kennis omtrent de aard en consequenties van de afwijking als onvoldoende ervaren (Hoofdstuk 3, 5 en 7). Daarom dient hieraan meer aandacht te worden besteed. Een "health passport" zou hierbij een middel kunnen zijn om als geheugensteun te dienen.

Aanbevelingen voor de klinische praktijk (vervolg)

- Consultatie van alle patiënten met een milde aangeboren hartafwijking op de leeftijd van 16 tot 18 jaar wordt door ons aanbevolen. Dit biedt een ideale mogelijkheid om de jongvolwassen patiënt goed voor te lichten, waardoor diverse problemen voorkomen kunnen worden (Hoofdstuk 3, 5 en 7).
- Bij loopbaan planning van patiënten met een complexe hartafwijking dient rekening gehouden te worden met de fysieke mogelijkheden van de patiënt. Het volgen van de hoogst haalbare opleiding dient te worden gestimuleerd (Hoofdstuk 8).

Aanbevelingen voor nader onderzoek

- Om de kwaliteit van leven van de patiënt met een complexe aangeboren hartafwijking te verbeteren, is het van belang dat onderzoek gedaan wordt naar de mogelijkheden en de effectiviteit van een revalidatie/ sportprogramma voor deze groep patiënten (Hoofdstuk 4). Daarnaast zal een onderzoek naar de determinanten van kwaliteit van leven (zoals coping, self esteem, sociale steun) ertoe bijdragen dat interventies voor het verbeteren van de kwaliteit van leven ontwikkeld kunnen worden (Hoofdstuk 2 en 3).
- Kwaliteit van leven onderzoek bij specifieke groepen aangeboren hartafwijkingen met goed gedefinieerde en gevalideerde instrumenten (zoals de TAAQOL en de CHD-TAAQOL) is belangrijk om vergelijking mogelijk te maken (Hoofdstuk 2 en 6).

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Curriculum vitae

De auteur van dit proefschrift werd geboren op 19 november 1970 te Groningen. Zij behaalde haar eindexamen VWO-B in 1989 aan het Christelijk Lyceum te Alphen aan den Rijn. In hetzelfde jaar begon zij met de studie Geneeskunde in Utrecht. Na het behalen van het artsexamen in 1997 was ze gedurende korte tijd werkzaam in het Psychiatrisch Ziekenhuis cluster Jeugd 'De Jutter', in Den Haag. Hier hield ze zich bezig met de somatiek van adolescenten (12 tot 18 jaar) en deed ze tevens ervaring op in de psychiatrische diagnostiek. Vervolgens was ze werkzaam als AGNIO op de afdeling Kindergeneeskunde van het Groene Hart Ziekenhuis te Gouda. In 1998 diende zich de kans aan om een multidisciplinair wetenschappelijk onderzoek op te zetten en uit te voeren bij het Leiden Center for Child Health and Pediatrics (TNO-PG en LUMC, afdeling Kindergeneeskunde; te Leiden). In 2000 werd het vervolgens dankzij de afdeling Cardiologie mogelijk dit onderzoek voort te zetten, dat in 2002 resulteerde in dit proefschrift.
Per juni 2002 is zij werkzaam bij Maatzorg, OKZ (Ouder en Kind Zorg), regio Delft om zich verder te specialiseren in de richting van de Jeugdgezondheidszorg.