Linköping University Medical Dissertations No. 1239

Follow-up of adults with congenitally malformed hearts with focus on individualised and computer-based education and psychosocial support

- A descriptive and interventional study

Helén Rönning

Division of Nursing Science
Department of Medical and Health Sciences
Linköping University, Sweden

Linköping 2011

Cover picture/illustration: Helén Rönning

Printed in Sweden by LiU-Tryck, Linköping, Sweden, 2011

ISBN 978-91-7393-189-2
ISSN 0345-0082

To you

Take charge of your life – design your curriculum

If you do not design your life – you will never know where the road goes

Tell me and I will forget
show me and I will remember,
share with me and I will understand

Chinese saying

CONTENTS

ABSTRACT

Background and aims: Many adults with congenitally malformed hearts are at risk for complications such as decreased function and capacity of the heart due to the heart defect and previously surgery. This advocates self-management behaviours related to medical treatments, physical activity, preventions of endocarditis, some restrictions regarding suitable employment and spare time activities, birth control and pregnancy, but also lifestyle concerns such as refraining from smoking and healthy eating. Sufficient knowledge and support are requirements for successful self-management. The overall aim of this thesis was to describe educational needs, develop a tool for assessing knowledge and to evaluate the effects of a follow-up model providing education and psychosocial support to adults with congenitally malformed hearts.

Subjects and methods: Adults (≥18 years of age) with the ten most common heart defects namely ventricular septal defect, atrial septal defect, coarctation of the aortae, aortic valve stenosis (defined as uncomplicated heart defects) and tetralogy of Fallot, complete transposition of the great arteries, congenitally corrected transposition of the great arteries, Ebstein anomaly and Eisenmenger syndrome (defined as complicated heart defects) were included in the studies. To apprehend the educational needs (I), sixteen adults with heart malformations, ranging from 19-55 years of age, were interviewed and data were analysed qualitatively using phenomenographic method. As a tool to evaluate knowledge, an instrument named Knowledge scale for adults with Congenital Malformed Hearts (KnoCoMH) was developed and psychometrically evaluated (II) in 19 + 114 adults with the ten most common heart defects average age 34 ± 13.5. A model for follow-up was described and initially evaluated (III) by 55 adults with the most common heart defects and finally tested in a randomised controlled trial (IV) with a total of 114 adults with congenitally malformed hearts (56 participants in intervention group and 58 in control group with average age 34 ± 13.5). The intervention group recived a model for follow-up with individualise and computer-based eduction and psychosocial support by a multidisciplinary team.

Results: The adults with malformed hearts described that it was important to have good educational materials and methods, and that the information should be given with respect for the individual. Tools for managing important areas in life such as the congenital heart defect, life situation, physical activity, treatment and healthcare resources should be provided. The education should

be tailored for the individual's life situation and given by two-way communication. The model for follow-up was developed based on the findings from study I and evaluated in study III and IV. The model consisted of individualised and computer-based education and psychosocial support by a multidisciplinary team. The model was evaluated with regard to knowledge (II), perceived control, symptoms of anxiety and depression at baseline and at the 3-month and 12-month follow-ups.

The KnoCoMH scale, a tool to assess knowledge, was developed and tested in study II and used as an outcome measure in Study IV. This knowledge scale consists of 46 items in four domains; General Knowledge, Medical treatment, Endocarditis prophylaxis and Contraceptives and Pregnancy and has acceptable psychometric properties for most of the knowledge domains included. The KnoCoMH was found to have acceptable psychometric properties regarding item difficulty level, internal consistency and test-retest reliability. The computer-based education used was developed in this project (published elsewhere) and consists of ten separate modules, one for each of the ten most common heart defects. Every module contains eight main areas and subheadings such as my congenitally malformed heart, cause and heredity, contraceptives and pregnancy, medical consultation and supervision, endocarditis prophylaxis, medical and surgical treatment, employment and spare time, sexual aspects and healthcare.

The results of the evaluation of the model showed a significant between-group treatment effect in general knowledge after 3-months (effect size 0.63, p= <0.01), and 12-months (effect size 0.53, p= 0.02). Knowledge regarding endocarditis also increased significantly between groups after 3-months (effect size 0.87, p= <0.01), and over time between baseline and 3-months (effect size 1.43, p=<0.001), and between baseline and 12-month (effect size 0.58, p= 0.02). Further, the intervention did neither increase nor decrease the perceived control over the heart condition or symptoms of anxiety and depression.

Conclusion: Two-way communication when given information was found to be crucial in order to enhance knowledge (I). Knowledge was seen as a tool for managing important areas in life. The KnoCoMH (II) was found to be a valid and reliable scale and can now be used to estimate knowledge in adults with congenitally malformed hearts. The model for follow-up (III) was effective in improving and maintaining knowledge (IV) about self-management in adults with heart malformation.

Keywords: congenital heart disease, education, information, instrument development, multidisciplinary team, nursing, phenomenography, psychometrics

LIST OF PAPERS

This thesis is based on the following papers, which will be referred to in the text by their roman numerals.

I. Rönning H, Nielsen NE, Swahn E, Strömberg A. Educational needs in adults with congenitally malformed hearts. *Cardiology in the Young*. 2008;18(5):473-9

II. Rönning H, Franzen Årestedt K, Nielsen NE, Swahn E, Strömberg A. Development and psychometric evaluation of the knowledge scale for adults with congenitally malformed hearts: KnoCoMH. (Submitted)

III. Rönning H, Nielsen NE, Swahn E, Strömberg A. Description and initial evaluation of an educational and psychosocial support model for adults with congenitally malformed hearts. *Patient Education and Counseling*. 2011;83:247-251.

IV. Rönning H, Nielsen NE, Swahn E, Strömberg A. Evaluation of knowledge, perceived control, symptoms of anxiety and depression related to a model for follow-up in adults with congenitally malformed hearts: a randomised control trial. (Submitted)

ABBREVIATIONS

CAS	Control Attitude Scale
EPS	Education and Psychosocial Support by a multidisciplinary team
GUCH	Grown Up Congenital Heart disease
HADS	Hospital Anxiety and Depression Scale
KnoCoMH	Knowledge scale for adults with Congenitally Malformed Hearts
KR 20	Kuder Richardsson 20
NYHA	New York Heart Association
SF36	The 36 item Short-Form health survey
SWEDCON	SWEDish registry of CONgenital heart disease
WHO	Word Health Organization

INTRODUCTION

To live with a chronic condition often endorse life-long follow-up in the healthcare system. Many adults with congenitally malformed hearts can be considered to have a chronic condition and most often in needs for regular follow-up [1]. Risks for complications are related to decreased function of the heart due to the heart defect, previous surgery, which leads to needs of self-management behaviours related to medical treatments, physical activity, preventions of endocarditis, employment and spare time, birth control and pregnancy, but also lifestyle concerns such as smoking and healthy eating [2]. One requirement for self-management is having sufficient knowledge to improve self-care [3] .

Many adults with congenitally malformed hearts lack sufficient knowledge about their heart condition [4-5]. One person expresses it like this in a paper by Verstappen et al., page 519; [5]

- I wish someone had sat down with me and said, "This is what your heart defect is. These are the consequences and what we are watching for. This is what we know and this is what we don't know. This is what we are doing to help and this is what you can do to optimise your life"

Previous studies have confirmed these needs for target education and outline the limited comprehension of knowledge in this group [6-7]. For instance, that 54% to 76% knew their congenital malformation diagnose by name [8-11]. There were knowledge deficiencies regarding medical treatment, prevention of endocarditis, birth control and pregnancy [8-11].

So far, few studies have focused on describing and evaluating educational models, materials and methods for adults with congenital malformation [6, 8-11, 12-13]. Substantial differences exist between previous studies evaluating level and content of knowledge in adults with congenitally malformed hearts. Most questions previously used have been developed for the purpose for the study. These studies are in children, parents and/or adults, often focusing on one aspect of care i.e. endocarditis prophylaxis or adherence to oral anticoagulation therapy [4, 8-9, 11-14]. When developing and evaluating interventions aiming to increase knowledge it is important to have psychometrically tested instruments sensitive for this purpose.

National and international guidelines recommend follow-up programmes that target medical and psychosocial problems among adults with congenitally

malformed hearts [15-23]. At present different programmes for this population are established in hospital outpatient clinics [24-27]. Moons et al [27] have described the current status of delivery of care in Europe 2010. Totally 50 specialist centres from 18 countries participated in this survey. Forty-seven programmes (94%) were located in a university hospital. In 94 % of the centres, cardiologists specialised in adults with congenitally malformed hearts were available and 68% had specialised nurses. The design and execution of follow-up at these centres were not presented in detail. One model used in Italy has been described by Chessa et al [24] . They have created a web site for collaboration among different centres in Italy through which patients, nurses, and physicians have access to information without login details. The outcome of this web site has not been presented yet. Another programme is Copenhagen Transition Program, an outpatient nursing clinic for adolescents with congenitally malformed hearts. This programme focusing on topics such as increasing knowledge about the heart condition, endocarditis, acute situations, nutrition, contraception, alcohol/drugs, smoking, physical activity, sleep/rest, and education. Parents are asked not to participate. The long-term outcome of this programme is also not yet described. In Canada, Reid et al [28] describe a successful transfer from paediatric to adult cardiac care in 15 specialised adult centre for congenitally malformed hearts.

Overall, there is strong consensus about the needs of further development and improvements of the care for this group [16, 22, 24-26, 28-37]. We urgently need to increase the experiences and knowledge of how to organise sufficient and successful education and psychosocial support to improve outcomes and increase the quality of care in this rapidly growing population.

BACKGROUND

ADULTS WITH CONGENITALLY MALFORMED HEARTS

Definition

Adults with congenitally malformed hearts are defined as persons who have reached the age of at least 16 and have a congenital, anatomical and physiological construction defect in the heart and/or in the big vessels [38].

Etiology

Heart anomalies are the most common form of congenital malformation and they are the most frequent cause of death among infants with birth defects [39].
The development of heart malformations takes place during the foetal stage and already at the end of the eighth foetus week the heart structure is completed [40]. The reason for developing a heart malformation is most often unknown, but both genetic and environmental factors can be the underlying causes. Only hereditary reasons for the defect are considered exceptional [41-42]. In most cases, there are multiple factors. Chromosome disorder is more common in people with heart defects than in the normal population. Harris et al found in pooled data from three large population-based registries in California, Sweden and France that 30 % of the infants with any type of congenitally malformed hearts had known chromosomal anomalies, most often Down's syndrome (11.4%). In those with atrial septal defect 27% had chromosomal anomalies, in ventricular septal defect 18%, in Tetralogy of Fallot 10% and in single ventricle 9% [43].
Extraneous factors that can cause a heart defect during the foetus stage are viral infections, diabetes mellitus in the mother, certain medicines and use of alcohol during the early stage of pregnancy [44]. In the normal population, the risk for having a child with a heart malformation is 0.8-0.9% while it increases to an average of 2-16% if one parent has a congenitally malformed heart [44-46].

The risk is twice as high if the mother has a congenital heart defect compared to if the father has it [42, 44].

Epidemiology

Adults with congenitally malformed hears is an increasing population. This is a result of improvements in diagnosis, medical treatment and heart surgery [47-48]. Since 1980 more than 85 % survive into adulthood [17, 39], compared to the previous 20 %, in the 1960´s [17]. In Sweden for instance a centralisation of paediatric heart surgery to two centres in 1993, reduced the overall 30-day mortality for open heart surgery in infants and children from 9.5% to 1.9%.[49].

The number of adults with congenitally malformed hearts in Europe is unknown [34, 37]. The reason for this is the use of different approaches to estimate the prevalence [34]. A problem with estimating the prevalence from birth or the number of births is that the subtraction of those who die or are spontaneously corrected will be missed. Those who are diagnosed after infancy or in adult age (35 %) will also be missed [50]. Survival following surgical procedures are well documented but there is still missing data on outcomes during long-term follow up [34]. According to a prospective population-based cohort study in Sweden (1992-2001) the prevalence of cardiovascular defects was 9.1 per 1,000 births [51]. Other prevalence data worldwide alter between 3 to 6 cases per 1,000 births [34] . In 2001 the ACC Bethesda Conference Task Force 1 in USA, calculated the number of survivors during three periods of surgical development and estimated the number of survivors for each period [52]. According to those data the prevalence of all forms of congenital heart defects was 3.51 per 1,000 adults. Another study from Canada, estimated the prevalence to 4.09 cases per 1,000 adults in a cohort, when using data from their national health-care system, containing all diagnoses and the use of health-care services until death [53]. They also reported a prevalence of 11.89 cases per 1,000 children which is higher than our prevalence data from Sweden (9.1 per 1,000 births) [51]. Based on 9 million inhabitants and prevalence data of 9,1 per 1,000 births, Sweden should have approximately 82,000 people born with congenitally malformed hearts. According to those data, approximately 20,000 people are in need for regular follow-ups as adults.

Sociodemographic characteristics

Adults with congenitally malformed hearts are still a young population [54-55]. In the Swedish registry of congenital heart disease [55] the mean age was 41 years (median 37), which is comparable to Canada in 2000 [53]. The Swedish population consisted of 51 % men and 49 % women and the most common heart malformations were shunt defects (Table 1), which is also comparable to the Canadian population [53]. As for marital status among those aged 20-40 in the Swedish population, 62% were single and 38% were married or cohabitant. Twelve percent were educated to a compulsory school level, 56% had finished high-school, 24% had been to university and 8% had undergone other forms of education [55].

Table 1. Main diagnoses of heart defects in the 7314 adults registered in SWEDCON 2009 [55].

Non-complicated heart defects	Atrial septal defect	18 %
	Ventricular septal defect	12 %
	Persistent foramen ovale	7 %
	Other shunt defects	6 %
	Aortic valve defects	13 %
	Aortic malformations	10 %
Complicated heart defects	Pulmonary valve defects	8 %
	Tetralogy of Fallot	7 %
	Transposition	6 %
	Marfan syndrome	3 %
	Mitral valve defect	2 %
	Tricuspid valve defects	2 %
	Other	6 %

Correction of the heart defect

Treatments among those with congenital heart defect vary due to the heterogeneous population [16-18, 56]. There are different heart defects with different associated conditions, but there are also differences among those with the same heart defects. The reason is the great improvements in diagnostic methods, medical treatment and heart surgery over the last 50 years [54]. Diagnostic methods and operation techniques in the 1960s differed greatly compared to the ones used in the 1980s. The first heart surgery with a heart and lung machine was carried out in 1954 [57-58]. This entails that a 55-year-old woman who underwent surgery in 1961 might have different complications and needs com-

pared to a woman, 33 years of age who had an operation for the same heart defect in 1980.
Available treatments for congenital heart defects are mostly corrected not curative and more operations may be needed during a person's lifetime [54, 58-59]. However nowadays treatments have developed rapidly from surgery to catherisation which is a great improvement for the population [60].

Symptoms and needs of risk reduction

Treatment of adults with congenitally malformed hearts is aimed to reduce symptoms, but also to minimise the risk and severity of late complications [54]. Adults with non-complicated heart defects have usually a normal life expectency, rarely requiring ongoing medical treatment or repeted surgery [61].

Most of the participants registered in a Swedish registry of congenital heart disease had no physical limitations according to NYHA[62] classification, see Table 2 [55]. The mortality at the end of 2009 was 3.7 % (273 cases). In 59 % cause of death was due to heart problems and in 37 % sudden death (4 % unknown) was the reason. Fortyseven percent of those who died due to heart reasonshad NYHA [62] classification III, 39 % had classification II, 32 % NYHA class I and 12 % NYHA class IV.

Table 2. NYHA[62] Functional classification of 7314 adults in SWEDCON [55]

	Explanation of the different NYHA classifications	
NYHA Class I	No limitation, ordinary physical exercise does not cause fatigue, dyspnoea or palpitations	80 %
NYHA Class II	Slight limitation in physical activity, comfortable at rest but ordinary activities result in fatigue, dyspnoea or palpitations	13 %
NYHA Class III	Marked limitation of physical activity, comfortable at rest but less than ordinary activities result in fatigue, dyspnoea or palpitations	4 %
NYHA Class IV	Unable to carry out any physical activity without discomfort. Symptoms of heart failure are present even at rest with increased discomfort during any physical activity	< 0 %
Not classified	Extracardiac limitations	3 %

In 2007, the mortality risk of among 6933 patients in the Dutch national registry CONCOR was found to be 2.8% (197 cases) [63]. Complications which was predictive to all-cause mortality was endocarditis, conduction disturbances, arrhythmias such as supraventricular and ventricular arrhythmias ,myocardial infarction, and pulmonary hypertension independent of age [63].

A cohort 2000-2005 from 58 sites in 25 countries included 2781 adults with infective endocarditis [64]. Among those, 12% had a congenitally malformed heart as a predisposing condition. Due to heterogeneity in congenitally malformed hearts, the prevalence of infective endocarditis also differs amongst them. According to ESC guidelines the risk for infective endocarditis in non-complicated heart defects as ventricular septal defects was six times higher (2 per 1000 patient-years) than in the normal population [23].

Arrhytmias among adults with congenitally malformed hearts is a common complication due to the anatomy of the heart defect itself or structural scars due to previous surgery, which are known to be substrates for arrhythmias [65-66]. High pressure or volume load in the heart or prolonged cyanosis are other common reasons for arrhythmias [65-66].

Heart failure is another problem in this population. To date data are scarce regarding the prevalence of, or methods of treating heart failure [54, 56].
This is due to the pathophysiology as a dysfunction in a congenitally malformed heart is often very different to the normal heart [54, 56]. To conclude, symptoms, late complications and needs of risk reduction among those with congenital heart defects vary due to the heterogeneous group. As long-term outcomes are not often presented, further research is required [16-18, 54, 56].

Individual knowledge about the heart condition

It is of great importance that adults with congenitally malformed hearts have knowledge about the prevalence of symptoms, late complications and needs of risk reductions [3, 54, 56]. Knowledge is required in areas such as adhering to medical treatment, awareness of symptoms and contributes to regular follow-up, prophylaxis of endocarditis, recommendations on physical activity, employment, sports and spare time and risk reduction in connection with pregnancy [16-18]. With a growing population and life expectation for many being the same as the normal population there are also requirements for risk reduction related to lifestyle changes [2]. These are for example physical activity, refraining from smoking and healthy eating.

Previous studies have confirmed the needs of increased knowledge in this group [6-7]. For instance 24% to 46% did not know the name of their heart diagnosis, and there were also requirements in other areas such as medical treatment, prevention of endocarditis, birth control and pregnancy [8-11].
Substantial differences regarding design and outcome measures exist between previous studies evaluating knowledge which leads to difficulties to compare

these results. Most questions have focused on one aspect of care in children, parents and/or adults, such as e.g. endocarditis prophylaxis or adherence to oral anticoagulation therapy [4, 8-9, 11-14].

Follow-up in the health care system

The majority of all adults with congenitally malformed hearts have been monitored on a regular basis in the hospital outpatient clinic since childhood. During the first years parents are completely responsible for all medical contacts and treatment for the child, but slowly during childhood and into adolescence these individuals have to take more responsibility for themselves [33, 67]. How this happens has to date not been that well described. The transition into adult healthcare is a need not so well developed [67-69].
During adult age, follow-up takes place every 6th month up to every 5th year depending on the heart defect and heart function. The goal with the regular follow-up is to minimise the risk and severity of late complications and reduce symptoms [54].

National and international guidelines recommend follow-up programmes target both medical and psychosocial problems among adults with congenitally malformed hearts [15-23]. At present different programmes for this population are established in hospital outpatient clinics [24-27]. Moons et al [27] described the current status of delivery of care in Europe 2010. Totally 50 specialist centres from 18 countries participated in this survey. How follow-up at these centres were designed and performed were not presented in detail.

One model presented for follow-up is the Copenhagen Transition Program[25]. This is an outpatient nursing clinic for adolescents with congenitally malformed hearts focusing on topics such as increasing knowledge about the heart condition, endocarditis, acute situations, nutrition, contraception, alcohol/drugs, smoking, physical activity, sleep/rest, and education. To our knowledge the long-term outcome of this programme is not described yet.

Few studies so far have focussed on describing and evaluating educational models, materials and methods for the group of adults with congenital malformation [6, 8-11, 12-13]. When developing and evaluating interventions aiming to increase knowledge it is important to have psychometrically tested instruments sensitive for this purpose.

Self-management

Self-management tasks focus on the individual's ability to maintain wellness [70]. The individual perspective is important and it is the person's responsibility to manage day-to-day care. Adherence to self-management behaviours improve when the person becomes involved in the care through their personal goals [70]. Self-management can be described as the individual's ability to manage symptoms, treatment, physical and psychosocial consequences, lifestyle changes inherent in living with a chronic condition and to maintain a satisfying quality of life [3].

Perceptions, motivation, learning, skills, reasoning, personality and social environment are key factors that influence behaviour changes [71]
According to Lorige and Holman self-management can be conceptualised into three tasks, medical or behavioural management, role management and emotional management [70]. Lorige and Holman have operationalised five core self-management skills to support self-management, see Figure 1.

Health professionals should provide supportive care to reach adaptation of the participant's own care plan. Agreement and partnership with health professionals, and significant others are important aspects. The participant should understand the importance of actively sharing the decision-making with health professionals and significant others, and the ability to monitor and manage signs and symptoms of the condition as well as manage the impact of the condition on physical, emotional, occupational and social functioning. The participant also needs to adopt a lifestyle that prevents risk factors and promotes health. It is important to focus on prevention and early intervention. The participant should have access to and feel confident in the ability to use support services.

There is a need for psychosocial support to achieve these self-management skills. In this thesis psychosocial support means individually targeted conselling to deal with the heart malformation.

Problem solving
- ✓ Problem definition
- ✓ Generation of possible solutions including the solicitation of suggestions from friends and healthcare professionals
- ✓ Solution implementation
- ✓ Evaluation of results

Decision-making
- ✓ Making decisions in response to changes in the heart condition

How to find and utilize resources
- ✓ Have the skill to seek different resources when needed

Form partnerships with the healthcare providers
- ✓ Have the ability to report accurately the trends and tempo of the heart condition
- ✓ Make informed choices about treatment – when to choose to discuss with healthcare providers (not only take contact when treatment needs is acute)

Taking action
- ✓ Making a short-term action plan and caring it out (having control over the heart condition and confident to behaviour changes)

Figure 1. Lorige and Holman's five core self-management skills [70].

Knowledge, learning and education

Different paradigms within philosophy, psychology and biology have different approaches to explaining the nature of knowledge [72]. There is no absolute definition of knowledge and how and when learning occurs.
The learning process is described by Marton and Booth as a change in the way one experiences oneself and the world and in emotional or intellectual behaviour [73]. In this thesis knowledge is defined as the ability to repeat and transfer information into new situations [74].

Theory of knowledge and learning

Medical educational systems are often based upon two major learning theories, behaviourism and constructivism [74-75]. Behaviourism is a technology-centred approach; learning occurs by information and repetition, rote-learning. Knowledge is the ability to repeat information but without the ability to transfer it to other situations (surface approach). Constructivism is a learner-centred approach; learning occurs by building on prior knowledge and

relevance (meaningful learning). Knowledge is the ability to repeat and transfer information to new situations, understanding is reached and deeper knowledge occurs (deep approach) [74]. Learning occurs when understanding is reached[76]. Understanding occurs when novel information is processed in the working memory and further in the long-term memory where it is organised into a schedule. Constructivism is used as a framework in this thesis [75, 77].

Patient education

At present there are different approaches for patient education. There are stress and coping theories and psychosocial theories focusing on different aspects of health behaviours [71]. This thesis focuses on the tools for education and evaluation of knowledge. This thesis uses constructivism.[75, 77] as an education theory and competence-based course design [72]. Competence-based course design means that the education is developed with content to provide the competence that is needed [72].

Tools for education

Today different tools are used in patient education such as pamphlets', web sites, video tapes or computer-based educational programs.
Studies on health information have shown the importance of relevant content and illustrations [77-78]. It is also important to understand the content and how to navigate in it. Complicated language and irrelevant content result in lost attention, and the cognitive load will be too heavy [78]. Extraneous details are known to decrease the learning process [79].

Adults with congenitally malformed hearts can be found among all academic levels, ages, using different learning styles [80]. Today there is known that text combined with pictures supports the reader's capacity to recall health educational information in brochures [81-82]. Computer-based education has another advantage; by using both channels into the working memory (by animations, (eye) and sounds (ears)) learning increases significantly [78, 83-86]. Another important issue with computer-based education is that the users can integrate existing knowledge more easily by choosing relevan content that is organised in a useful way [83] . Computer-based education tailored for the individual is a multimedia tool that has shown to significantly increase knowledge and/or management of the disease in people with chronic illness [87-91].

AIMS OF THE THESIS

The overall aim of this thesis was to describe educational needs, develop a tool for assessing knowledge and to evaluate the effects of a follow-up model providing education and psychosocial support to adults with congenitally malformed hearts.

The specific aims were:

- To describe how adults with congenitally malformed hearts experienced their educational needs.

- To develop and psychometric evaluate a knowledge scale for adults with congenitally malformed hearts.

- To describe and make an initial evaluation of a follow-up model for adults with congenitally malformed hearts, focusing on education and psychosocial support by a multidisciplinary team.

- To evaluate the effects of this model including computer-based and individualised education and psychosocial support for adults with congenitally malformed hearts.

SUBJECTS AND METHODS

Design

This thesis has an explorative design combining both qualitative (I), and quantitative descriptive (II, III), and interventional (IV) approaches in order to immerse and understand the needs of education (I, IV), tools for evaluation (II) and tools to reach knowledge and provide psychosocial support (IV) in adults with congenitally malformed hearts. Effects on psychosocial support and self-management itself were not evaluated.
An overview of the studies, I-IV and the outcomes in papers (I-IV) are described in table 3.

Table 3. Overview of the design, methods, analyses and outcome in study I-IV

	Study I	Study II	Study III	Study IV
Design	Qualitative	Descriptive	Descriptive	Randomised controlled
Participants	16 Adults with congenitally malformed hearts	19 /114 Adults with congenitally malformed hearts	55 Adults with congenitally malformed hearts	114 Adults with congenitally malformed hearts
Data collection	Interviews	Development, evaluation by observations, structured interview and psychometric tests	Description and evaluation by questions	Questionnaires
Data analyses	Phenomenography	Face validity Content validity Item-difficulty Internal consistency Test-retest reliability Discrimination ability	Descriptive statistics	Descriptive statistics Chi-square test Student t-test
Outcomes	Experiences of educational needs	Knowledge instrument for Adults with Congenitally Malformed Hearts; KnoCoMH	Initial evaluation of EPS-model	Long-term effects of EPS-model on knowledge, perceived control, symptoms of anxiety and depression

EPS= Education and psychosocial support by a multidisciplinary team

Setting, participants and procedures

Study I

A strategic selection of respondents was made in order to obtain as many different experiences of educational needs about the heart defect as possible. Our endeavour was to interview persons of both genders, with various types and severity of heart defect, aged between 18 and 55. The lower age limit of 18 was chosen since that is the age of adulthood. The upper age limit of 55 was chosen to limit the risk of age-related heart disease. Age-related heart disease, such as ischemic heart disease, can be difficult to separate from the congenitally malformed heart and it could therefore be difficult to specify a person's educational needs and the information they had experienced related to their heart defect. The selection of respondents was made from a national registry in Sweden [92]. The seleced individuals were from the South Eastern Healthcare Region catchment area with a geographical maximum restriction area of 120 kilometres from Linköping university hospital.
The respondents were sent written information about the study including a reply letter and a prepaid envelope by mail. They were asked to return the letter within a week. The respondents interested in the study, or those who had not replied after a week, received a telephone call. They were informed verbally about the study and were asked if they wanted to participate. If the respondent was interested, an interview was scheduled.

Study II, III, and IV

In study II, III and IV data collection was accomplished in 4 hospital outpatient clinics following a routine clinical visit to a physician. In study II, III and IV, consecutively selected adults with congenitally malformed hearts with the ten most usual heart defects, were included (Table 4).

Table 4. Characteristics of the participants in study I, II, III, IV

		Study I	Study II	Study III	Study IV	
PARTICIPANTS						
Adults with congenitally malformed hearts		16	19 114 (From study IV)	55 (From inter-vention group in study IV)	114	
Sex						
Men		8	12	26		
Women		8	7	29		
Age (SD)		34	34 ±13.5		34 ±13.5	
Main diagnosis					I	C
Noncomplicated heart diagnoses	Ventricular septal defect	3	1		10	6
	Atrial septal defect	1	2		2	2
	Coarctation of the aortae	2	2		9	8
	Aortic valve stenosis	2	2		8	12
Complicated heart diagnoses	Tetralogy of Fallot	1	2		11	14
	Complete transposition of the great arteries	1	2		7	7
	Congenitally corrected transposition of the great arteries		2		6	2
	Single ventricle	1	2		0	2
	Ebstein anomaly	1	2		1	3
	Eisenmenger syndrome	1	2		2	2
	Marfans syndrome	1				
	Left ventricular outflow obstruction	1				
	Pulmonary atresia	1				
		16	19		56	58

I = Intervention group. C = Control group

In study IV, baseline data were collected after enrolment and signed informed consent before the participant's visit to the physician. See Table 5 for classification of the participants according to NYHA[62].

The randomisation was done between the visit to the physician and the visit to the nurse by unpredictable allocation sequences, concealment until assignment occurred by sequentially numbered and sealed letters, prepared by a third person not involved in the randomisation. Data at the 3 and 12-month follow-ups were collected by mail and telephone interview see flow chart of the participants in Figure 2.

Table 5. NYHA[62] Functional classification of the 114 participants in study II, IV

	Explanation of the different NYHA classifications	
NYHA Class I	No limitation, ordinary physical exercise does not cause fatigue, dyspnoea or palpitations	67 %
NYHA Class II	Slight limitation in physical activity, comfortable at rest but ordinary activities result in fatigue, dyspnoea or palpitations	24 %
NYHA Class III	Marked limitation of physical activity, comfortable at rest but less than ordinary activities result in fatigue, dyspnoea or palpitations	3 %
NYHA Class IV	Unable to carry out any physical activity without discomfort. Symptoms of heart failure are present even at rest with increased discomfort during any physical activity	0 %
Not classified	Extracardiell limitations	4 %

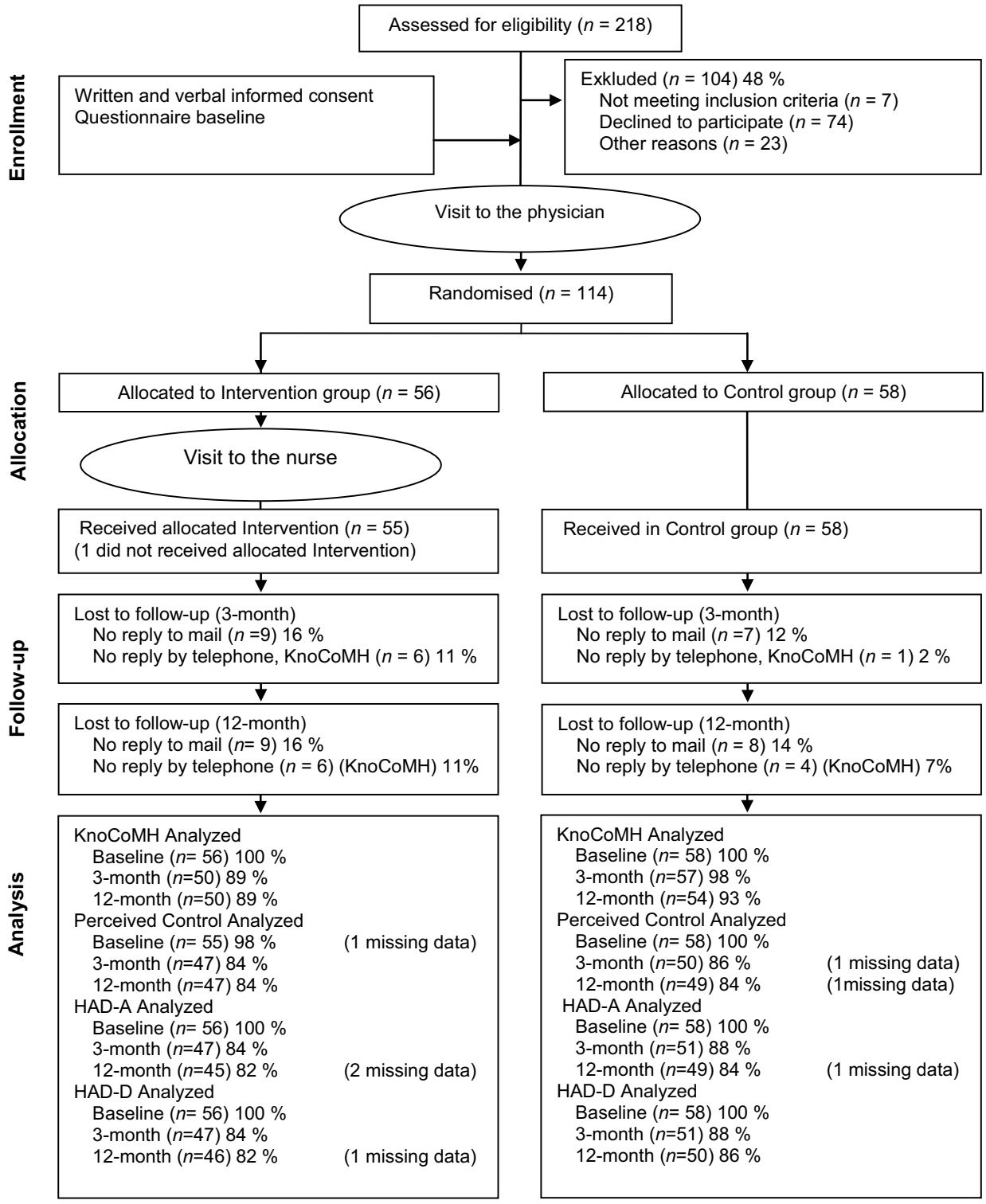


Figure 2. Flow chart of the participants in study IV, the enrolment, allocation, follow-up and analysis.

Exclusion criteria were complicating co-morbidity such as age-related coronary heart diseases, valve diseases, other life-threatening diseases, psychiatric illness or inability to read or understand Swedish.

In study IV, 3% of the participants went to the hospital outpatient clinic for a regular visit every sixth month, 50 % every year, 25 % every second year, 5 % every fifth year and 17 % had other time schedules for regular follow-ups. Thirty-three percent were on medication due to heart failure, high blood pressure, arrhythmias and/or prevention of thromboembolism. In total 68 % had undergone heart surgery and 46 % more than once. According to the physician's recommendation, 66 % of the participants should use endocarditis prophylaxis, 91 % of the females (totally *n*=51) should contact the physician if pregnant and 64 % before becoming pregnant. There were no differences in baseline characteristics between groups (Table 6).

Table 6. Socio-demographic and clinical characteristics

Patient characteristics, n (%)	Total (n=114)	Intervention (n=56)	Control (n=58)	p-value
MARITAL STATUS				
Married/cohabitant	62 (54)	31 (56)	31 (53)	ns
Single	51 (45)	24 (43)	27 (47)	ns
Widow/widower	1 (1)	1 (1)	0	ns
WORKING STATUS				
Student	17 (15)	6 (11)	11 (19)	ns
Employed	72 (63)	36 (64)	36 (62)	ns
Retirement	10 (9)	6 (11)	4 (7)	ns
Unemployed	10 (9)	4 (7)	6 (10)	ns
Other	5 (4)	4 (7)	1 (2)	ns
EDUCATION				
Compulsory school	15 (13)	6 (11)	9 (15)	ns
High school	63 (55)	33 (59)	30 (52)	ns
University	22 (19)	9 (16)	13 (22)	ns
Other	14 (13)	8 (14)	6 (11)	ns
Medical treatment at baseline, yes[3]	37 (42)	18 (32)	19 (33)	ns
Undergone heart surgery, yes[3]	78 (68)	35 (62)	43 (74)	ns
> 1 heart operation	23 (21)	9 (14)	14 (25)	ns
Smoking, yes	12 (10)	5 (9)	7 (12)	ns
Regular physical activity every week, yes	95 (83)	44 (80)	51 (86)	ns
Have been recommended endocarditis prophylaxis, yes[5]	75 (66)	37 (67)	38 (64)	ns
Only general prevention without antibiotics, yes[5]	9 (8)	6 (5)	5 (4)	ns
Have been recommended to avoid all regular physical activity, yes[5]	2 (2)	0	2 (3)	ns
Have been recommended to choose employment without physical demands yes[5]	43 (38)	20 (36)	23 (39)	ns
Have been recommended to avoid some contraceptives, yes[5]	2 (2)[6]	1 (3)[6]	1 (4)[6]	ns
Have been recommended to contact the physician if the women want to become pregnant, yes[5]	36 (64)[6]	16 (55)[6]	20 (74)[6]	ns
Have been recommended to contact the physician if pregnant, yes[5]	51 (45)[6]	26 (90)[6]	25 (93)[6]	ns

n = Number. % = Percent

NYHA= New York Heart Association.

[1]. Complicated = Congenitally malformed hearts such as complete transposition of the great arteries, congenitally corrected transposition of the great arteries, Ebstein anomaly, Eisenmenger syndrome, single ventricle, tetralogy of Fallot

[2]. Non-complicated = Congenitally malformed hearts such as aortic valve stenosis, atrial septal defect, coarctation of the aortae, ventricular septal defect.

[3]. According to medical files.

[4]. According to participants' statements.

[5]. According to responsible physician (Recommendations due to the congenitally malformed heart).

[6]. Percentage of female patients (Experimental group *n*=29, Control group *n*=27, Totally female *n*=56).

Qualitative methods

Phenomenography

Study I used a descriptive qualitative design inspired by phenomenography. Phenomenography was first used within behavioural science and pedagogy, for example, to investigate how students learn[93]. In the last decade it has been increasingly used in nursing sciences [94]. Phenomenography is used to describe different ways of experiencing a phenomenon in the surrounding world by describing the outcome space depicting different ways to experience a phenomenon [93]. Phenomena in the world can have various meanings for different people. Marton described different ways of reaching knowledge concerning other people's ways of experiencing the world, first order or second-order perspectives [95]. In the first order perspective the researcher is interested in how something really is, while in the second order perspective
the researcher is primarily interested in how things appear to people and their experiences of the world around them. Phenomenography uses the second-order perspective, the only world we can communicate about is the world as experienced. The purpose of the phenomenographic method is to describe the essential variation in the understanding of a certain phenomenon, looking for differences and similarities, and to find the underlying structure of variance [93].

Interviews

The interviews lasted between 30-90 minutes, were tape-recorded and transcribed verbatim. They were based upon open, semi-structered questions, following phenomenographic practice [96]. The sequence of the interview questions differed due to the fact that a participant's answer could inspire new questions. The goal was to achieve a conversation about educational needs. "Can you describe your experiences of the follow-up of your congenitally malformed heart?" was followed by questions regarding information they had received and how they experienced it should have been. Follow-ups could consist of a meeting with healthcare professionals in the out-patient clinic or in the hospital ward in connection with a check-up or treatment of their malformed heart from childhood until now. After each interview, a short summary of the experience during the interview was written down. This was done as a reminder of the interview situation, to be used during the analysis.

Analysis

The phenomenographic analysis seeks to understand the variation of individual experiences of a phenomenon [95]. The analysis in study I was based on the seven steps described by Dahlgren and Fallsberg [96]. The analysis was performed by the first researcher with the co-researchers as co-readers.Step 1. Familiarization: The transcribed interviews were carefully read in order to have an overview of the content. Comments were made in the margins. As a reminder of the interview situations, the summaries of the experiences during the interview were readed again. Step 2. Condensation: By reviewing the text for iteratively for statements related to the aim, the most significant statements were selected. In the material, 489 statements were identified. Step 3. Comparison: The statements were compared to find sources of variation or agreement by focusing on similarities or differences. Step 4. Grouping: The grouping was based on the previous comparison and questioning. At first five subcategories were formed, but they were divided during further analysis and the statements were later grouped into eight preliminary subcategories. Step 5. Articulation: The statements in the preliminary groups were compared and critically analysed, in order to find the central content within each group of answers. The content of each category should not be too narrow or too wide and without obvious overlapping between the categories. One main category and two descriptive categories with eight sub-categories were formed in this step. Step 6. Labeling: The categories that emerged were named in order to mirror the content. Step 7. Contrasting. The categories were compared with each other, by considering their mutual relationship. The participants described experiences of information concerning the heart defect from childhood, as adults; to those surrounding them e.g. healthcare professionals, family members, school, peers etc. The set of categories of description is sometimes referred to as an 'outcome space' [97]. The relationship between the categories can be hierarchically, linear or branched relationships [98].

Quantitative methods

Development and psychometric evaluation of the knowledge scale

Study II contained a development and psychometric evaluation of a Knowledge scale for adults with Congenitally Malformed Hearts (KnoCoMH). The development of this new knowledge scale was build on the Leuven knowledge questionnaire for congenital heart disease [8], which includes the following domains: (1) The disease and its treatment; (2) The prevention of complications, including endocarditis; (3) Physical activity and (4) Reproductive issues. With support from the literature these domains were considered to be important sources of knowledge for adults with congenital malformed hearts [19-21, 99]. Permission to use some of the items in this new scale was obtained from the constructors of the Leuven Knowledge questionnaire [8].

The study consisted of two phases 1: Development and evaluation of the initial version of the knowledge scale, and phase 2: Evaluation and refinement of the knowledge scale. During these phases Streiner and Norman's guide to developing health measurement scales was used [100].

Phase 1 - Face validity and content validity

During phase 1, face validity and content validity were assessed by the research group and 19 adults with congenitally malformed hearts (Table 4).
First the Leuven Knowledge questionnaire [8] was discussed in the research group. The group identified items not valid according to current guidelines [19-21, 99] or to the cultural context or with unclear phrasing. One item had more than one tenor; *Please mark all symptoms which may occur if your heart condition deteriorates and for which you have to contact your cardiologist*. This item was divided into two new items. Two items were not in accordance with guidelines and cultural context, one item was altered (*Consuming three or more alcoholic drinks per day is more harmful for someone with a congenital heart disease than for someone without such a disorder)* and the other one was removed (*Do you have to follow a diet? If you answer "yes", please indicate the type of diet)*.
The next step of phase 1 was to evaluate the initial version of the instrument in 19 adults with congenitally malformed hearts (Appendix A). The data collection was accomplished in two hospital outpatient clinics following a routine

clinical visit to a physician. Directly after the participant had filled in the instrument, questions were asked in a structured interview and observations were made while the instrument was completed. A formal guide was used as an interview guide. The observations aimed to evaluate tenor in the items and user-friendliness of the instrument (face validity and content validity), and were documented as comments by the researcher in the interview guide. For any item answered incorrectly, the participants were asked to explain what they thought the question was asked, and why they had responded as they did. The objective of this was to identify errors made due to misunderstandings of the questions. A short summary of experiences during the observation/interview was written down.

The results were summarised and presented to the research group who was working with the instrument again to identify not-valid items. More items with unclear tenor were found; -*Bleeding gums need extra attention* and -*You should clean your teeth at least once a day*. In the culture context, a normal healthy behaviour is to clean one's teeth every day and to prevent bleeding gums. This is not the same as having knowledge prevention of endocarditis. The items were altered with an extra phrase; (Bleeding gums need extra attention) *to prevent endocarditis*. It proved difficult to give the correct answer to the item; *Describe below, or indicate on the diagram where your heart defect is localised* , and this was much discussed. It was not identified as knowledge and thus the item was removed. Another item difficult to answer correctly and not identified as knowledge was; -*What is the main purpose of this follow-up* with where possible answers were routine check, no specific reason, personal reassurance, to detect any unexpected deterioration, to continue treatment using the latest techniques, or the doctor wanting to line his pockets. This item was removed. In the areas: The disease and its treatment, (later called General knowledge) and Reproductive issues (Contraceptives and pregnancy), the research group discussed the content of the questions and new supplementary items were added (see Appendix A; item no.8, 9, 10, 24, 27, and 28). Areas were now re-named; General knowledge (29 items (16 multiple choice questions and 1 question with 13 multiple answers to respond to)), Medical treatment (8 item (1 question with 8 multiple answers to respond to)), Endocarditis prophylaxis (13 items (6 multiple choice questions and 1 question with 7 multiple answers to respond to)), and Contraceptives and pregnancy (4 items (multiple choice questions)), a total of 54 items (Appendix A). The questionnaire was now considered as a new instrument; the Knowledge scale for adults with Congenitally Malformed Hearts; KnoCoMH.

Phase 2 - Item difficulty, internal consistency and test-retest reliability

In phase 2 the final instrument was tested among 114 adults with congenitally malformed hearts and included item difficulty, internal consistency and test-retest reliability (n: 59/58). The characteristics of the participants are described in Table 4.

Scoring was calculated by dichotomising every answer (correct/incorrect). Missing answers were dichotomised as incorrect. Items 1, 2, 3, 4, 10, 16, 22-24, 26-28 were marked individually from the medical journal and the physicians' recommendations (Appendix A). If the participant not was recommended medical treatment or endocarditis prophylaxis these areas were not marked. The area, Contraceptives and pregnancy was not marked in males.
The item difficulty index, which is the proportion of correct answers for each item, was used to identify items with low discrimination ability. Items answered correctly by more than 95 % or incorrectly by less than 5 % were considered as not useful items [100]. The internal consistency of individual items was evaluated using item-total correlation. Item-total correlation is the correlation of every individual item with the total score [100]. Items with total correlations < 0.20 should be considered for removal [100] This analysis, together with content validity (the theoretical value of the item to evaluate knowledge) was used to delete less important items [100]. KR-20 was analysed for testing the homogeneity and internal consistency of the items in the four different areas of the instrument.To evaluate the discrimination ability of the final instrument, Discrimination index was analysed [100-102]. Values below 0.20 suggests to be weak [102]. The test-retest reliability was evaluated using Pearson's correlation analysis and Intraclass correlation (ICC, two-factor mixed effects model, absolute agreement, single measures) was also used [103-105]. KR-20 was used instead of Cronbach's alpha, due to dichotomy answers [100]. KR-20 is the average of all of the possible split-half reliabilities of the scale. For best homogeneity, KR-20 should be between 0.70 and 0.90 [100].

Computer-based educational program

The computer-based education used in this thesis has been developed by the research team and evaluated by questionnaires, observations and structured interviews by a multidisciplinary team and adults with congenitally mal-

formed hearts [106]. The program consists of ten separate modules, one for each particular malformation with the same eight main areas and subheadings covering different aspects of the subject, see table 7. The malformations were ventricular septal defect, aortic valve stenosis, atrial septal defect, coarctation of the aortae, complete transposition of the great arteries, congenitally corrected transposition of the great arteries, Ebstein anomaly, Eisenmenger syndrome, single ventricle, and tetralogy of Fallot.

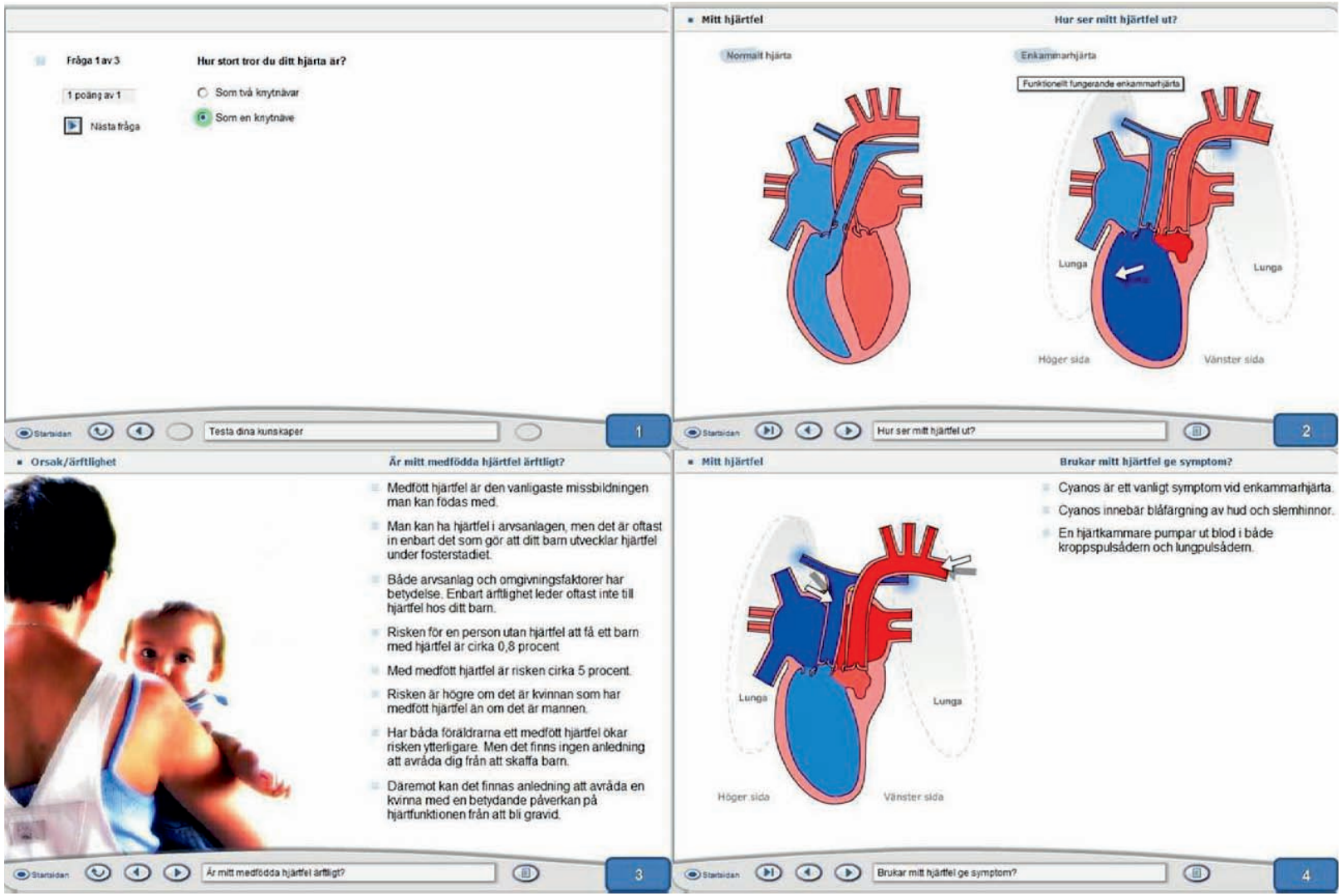
The computer-based education was developed as a complement to verbal information and not aimed to be a complete textbook. The main content and areas in the program were selected by the researchers based on the literature and clinical experience, and are meant to fit in with the users' knowledge and needs by answering frequently asked questions. Each main area ends with a self-conducted test (Picture 1).

Cognitive functions regarding content related to usability, comprehensibility and appearance were important during the development of all ten modules. The animations, narratives and text on screen were developed to be as comprehensible as possible (Picture 2). Attempts were made to construct the program to suit different learning styles. This was done either by looking at the animations and listening to the speaker or by merely reading the text on screen by means of a pop-up window. Pages without animations contained on-screen headlines and were synchronised with the speaker narratives (Picture 3) except for the subheading "Symptoms" in all modules that contained animation, speaker and headlines (Picture 4).

Every one of the ten modules consists of 55-60 minutes of information, and the program format is CD. The content of the program has been verified by the experts and the formal users but also by the literature [6, 17-21, 25, 32, 107-108]. The results of the program evaluation showed that the computer-based education was experienced as stimulating and easy to use. The appearance and quantity of the text was graded as good and the content as relevant and very useful.

Table 7: Eight main areas and their subheadings in each of the 10 modules in the computer-based educational program

Main areas/Subheadings	Main areas/Subheadings
MY CONGENITALLY MALFORMED HEART	ENDOCARDITIS PROPHYLAXIS
- The normal heart - appearance	- What is endocarditis?
- The normal heart - function	- I – risk for endocarditis?
- What does my congenitally malformed heart look like?	- When – risk for endocarditis?
- How many people have the same congenitally malformed heart?	- How to avoid endocarditis?
- Does my congenitally malformed heart give symptoms?	- What are the symptoms of endocarditis?
Test your knowledge	Test your knowledge
CAUSE AND HEREDITY	MEDICAL AND SURGICAL TREATMENT
- Why do I have congenitally heart malformation?	- Medical treatment – my congenitally malformed heart?
- Is my congenitally malformed heart hereditary?	- Can I stop the medication?
- Where can I meet other people with congenitally malformed hearts?	- Surgical treatment – my congenitally malformed heart?
Test your knowledge	- Other treatments for my congenitally malformed heart?
CONTRACEPTIVES AND PREGNANCY	*Test your knowledge*
- What to think about with regard to contraceptives	EMPLOYMENT AND SPARE TIME
- Issues when planning a pregnancy	- Can I do all types of work?
- Pregnancy	- Sport/spare time activities
Test your knowledge	- Risk for my heart when flying?
MEDICAL CONSULTATION AND SUPERVISION	- To think about – when travelling?
- Why regular check-ups?	*Test your knowledge*
- Why not operate on the congenitally malformed heart now?	SEXUAL ASPECTS AND HEALTH CARE
- Postoperative check-ups?	- Sex life/life together - effects of my congenitally malformed heart?
- When should I contact the doctor?	- My congenitally malformed heart –self-care issues?
Test your knowledge	- Alcohol, drugs, smoking and moist snuff
	Test your knowledge



Pictures 1,2,3,4 from the computer-based educational program. Picture 1: Self-conducted test. Picture 2: Animations, arrows and fingerposts of the flow in the congenitally malformed heart are presented on the screen beside a picture of the normal heart. Picture 3: Pages without animations were developed with headlines on the screen synchronised with the speaker. Picture 4: The subheading Symptoms has animation, speaker and headlines.

Instruments

Perceived control

The Control Attitudes Scale, CAS is a 4-item questionnaire with a 7-point scale (1 = not at all, 7 = very much) measuring perceptions of personal and family control in the context of cardiac disease. [109] The total score ranges from 4 to 28, higher scores indicate stronger perceptions of control over the disease. [110] The scale is translated and psychometric tested under Swedish circumstances. [111] Scores below 16 are considered to indicate low level of control [110]. Reliability coefficient, cronbach's alpha was 0.81 in this study.

Anxiety and depression

The Hospital Anxiety Depression Scale (HADS) is a well-validated 14-item questionnaire for screening symptoms of anxiety and depression in the general population as well as in somatic patient populations. [112] HADS provides separate scores for anxiety (HAD-A) and depression (HAD-D). [113-114] Score 0-7 indicates no symptoms of anxiety or depression respectively, score 8-10 indicates possible case, score 11-21 indicates probable case with symptoms of anxiety or depression. Reliability coefficient, cronbach's alpha for the anxiety score was 0.82 and 0.76 for depression score in our study.

Statistical analyses

The results in this thesis are given as frequencies, (mean, standard differences (SD), percentage, difference and mean difference between groups with 95 % confidence interval).
In study IV baseline characteristics of the control and experimental groups were compared using $\chi 2$ tests for categorical data and independent t tests for continuous variables. To test the effects of the intervention, knowledge, perceived control, and symptoms of anxiety and depression were compared between groups with "intention to treat analysis" in baseline, 3 and 12-months data.

Missing data in the CAS and HADS questionnaires were not replaced and the scale/subscale was recorded as missing for the person. If only one item were missing in the four domains respectively in the KnoCoMH, the answer was assessed as incorrect. If more than one item was missing in the same area, the area was recorded as missing for the person.
Differences between groups at baseline, after 3 and 12-months and differences over time were analysed by t-test.
Reasons for excluding 104 (48%) potential participants were denied consent, missed cases or failure to meet the inclusion criteria (Figure 1). Thirty-four (33 %) of those had a complicated heart diagnosis (11 complete transposition of the great arteries, 5 congenitally corrected transposition of the great arteries, 1 Ebstein anomaly, 16 tetralogy of Fallot and 1 unknown diagnosis), and 70 (67 %) had a non-complicated diagnosis (19 aortic valve stenosis, 6 atrial septal defect, 24 coarctation of the aortae and 21ventricular septal defect).
In total 16 participants (14 % of 114) were lost to follow-up after 3-months, 9 in the experimental group and 7 in the control group, and a total of 17 participants (15% of 114) were lost to follow-up after 12-months, 9 in the experimental group and 8 in the control group. Missing data in the instruments varied between 1.7 to 3.5 % in CAS and HADS respectively, in KnoCoMH there were no missing data (Figure 1).
Analyses of KR-20 were performed using STATA 11 for Windows (StataCorp LP, College Station, TX), all other analyses were performed using PASW Statistics 18.0 (SPSS Inc., Chicago, IL). The statistical level was set at $p < 0.05$.

Ethical aspects

All studies were approved by the Regional Ethical Review Board in Linköping (Registration No. M03-123 for study I, No M-172-05 for study II-IV) and the principles outlined in the Declaration of Helsinki were followed [115]. All participants were given verbal and written information about the study, the procedures and confidentiality and that withdrawal from the study would not affect their future care. When recipients agreed to participate, they signed a written informed consent form.
In study I, the respondents chose the location for the interview. If anxiety or other reflections arose after the interview, the respondents had the opportunity to phone the doctor responsible or the researcher. The respondents were informed that the interview would be tape-recorded and transcribed verbatim.

Interventions with computer-based education and psychosocial support were not standard care at the hospitals participating in the studies (III, IV) so no study participant was withdrawn from care as usual. Completion of the questionnaires can possibly be perceived as a violation of integrity, but the risk of causing temporary discomfort nevertheless was estimated as minimal.

RESULTS

This thesis describes and evaluates educational needs, tools for education and evaluation, and a model for follow-up, see figure 3.

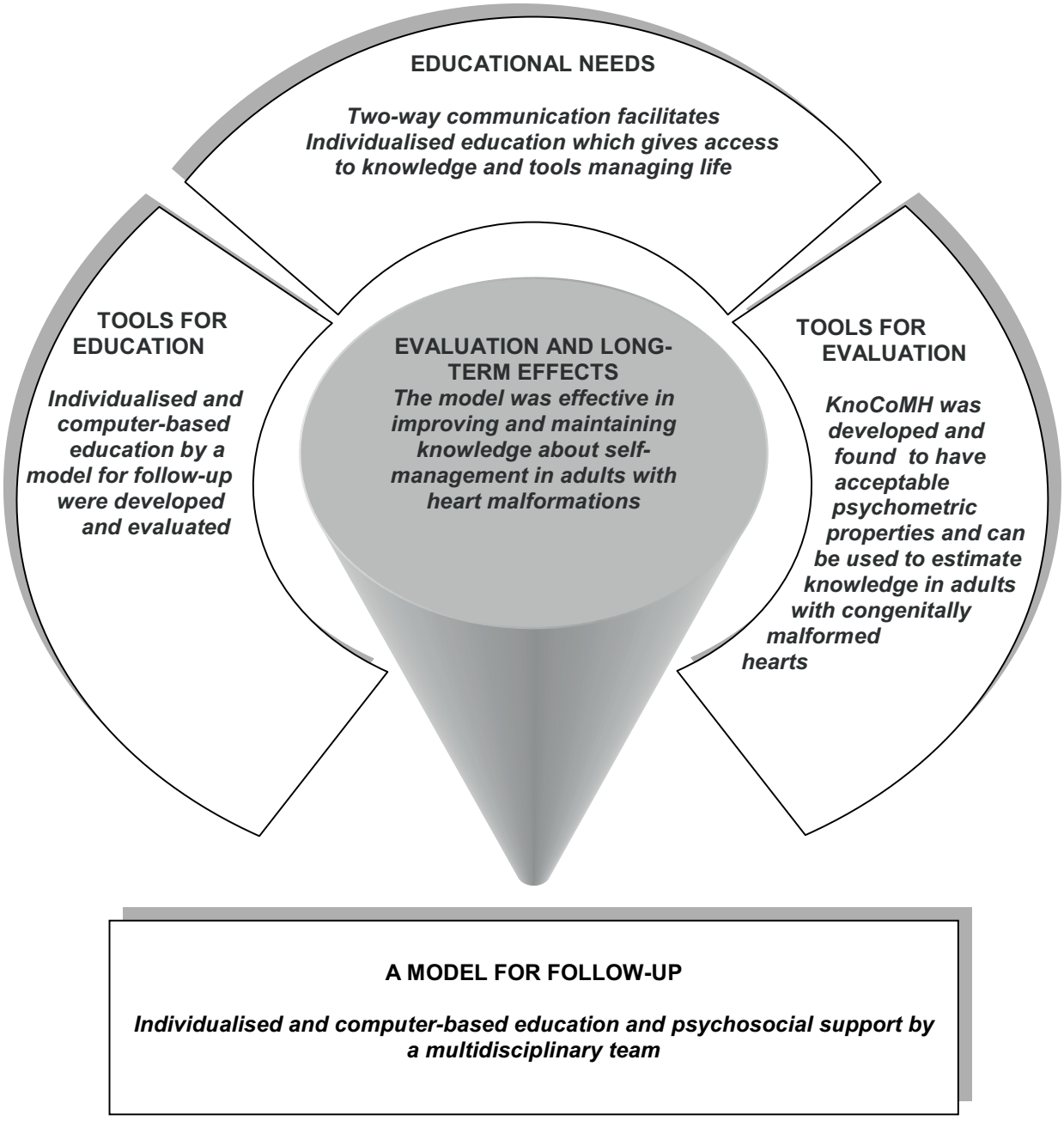


Figure 3. The main results of this thesis.

Educational needs - individualised

Communication enables individualised education

Study I described the experiences of how two-way communication is crucial for receiving individualised education, which gives access to knowledge and tools managing life. If information was provided without good communication between healthcare providers and the person with a heart defect, they could not transfer the information to apply to themselves.

- I have experienced that with heart doctors, they can be just like some little professor Calculus in the adventures of Tintin. They are extremely proficient, but a bit special and not particularly socially competent. If you face them with questions, they explode with information and then it's difficult to separate.

The respondents described that information should give easy access to knowledge through opportunities for education. It is also important to have proper educational materials and methods, and the information should be provided with respect for the individual. Tools to manage important areas in life such as the congenital heart defect, life situation, physical activity, treatments and healthcare resources should also be provided. This is facilitated if the education is tailored to the individual's life situation in a holistic approach, provided through good communication. See figure 4.

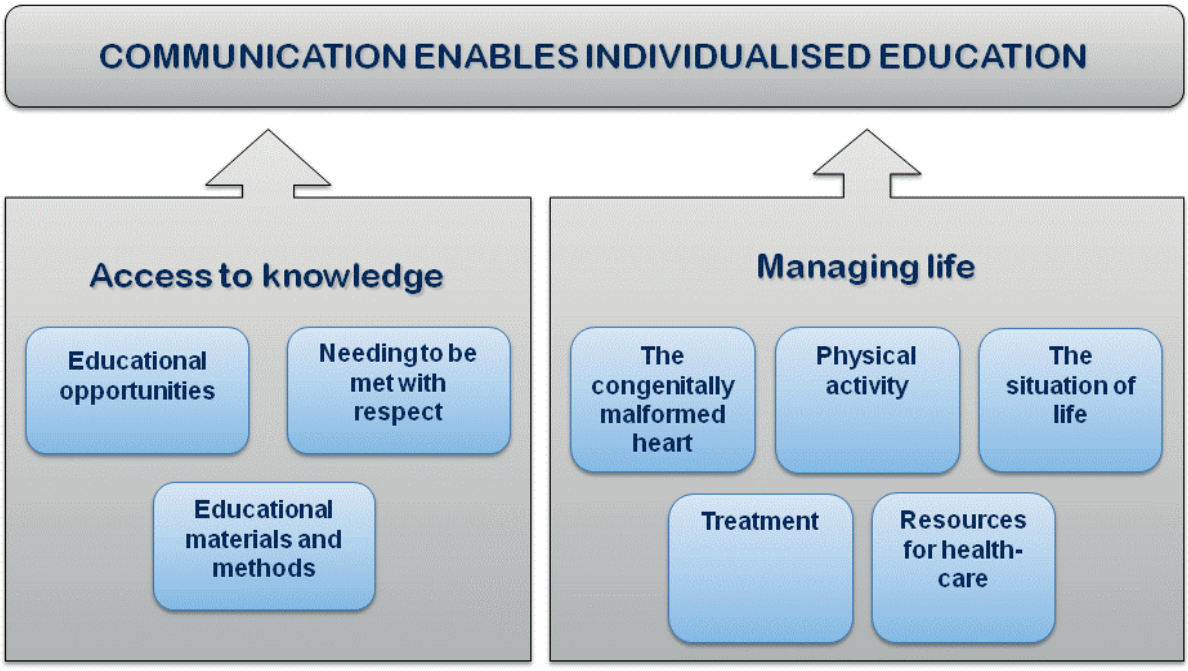


Figure 4. The main results of study I.

Access to knowledge

Educational opportunities

Education should be provided on a regular basis, be easily accessible, and adjusted to personal preferences and needs throughout the different phases during the life span. As parents receive most of the information concerning their children a completely new education is needed for young persons with congenitally malformed hearts during the transition to adult healthcare.
This should be done regardless of previous knowledge and on a regular basis more or less throughout the person's lifespan, depending on needs. The respondents described increased informational needs at deterioration, when symptoms occurred and during important stages of life e.g. when planning their career or when starting a family. Regular information sessions were mentioned as a method to increase knowledge. Other methods mentioned were opportunities to gain information during regular visits to a healthcare professional and/or during medical examinations, telephone consultations when questions arose or scheduling an extra appointment with a physician when needed. The telephone consultation could be with a physician or a nurse. Some had experiences of contacting the nurse specialised in ischemic heart disease or their physician when needed. It was experienced as an uncertainty to only have a phone number to call when necessary, and not knowing who was going to answer. Results of medical examinations should not be given by phone or mail, the respondents wanted time to ask questions face to face. Information is not always requested. There can be periods when the respondents do not wish to receive information, and other periods with increased needs of knowledge.
The respondents experienced that information should be provided at an early age, maybe around the age of 6-8 when children start school, depending on maturity. The education should start on a small scale and then build up during childhood and adolescence when the respondents start to understand that they have a heart defect and be individualised depending on their life situation in order to avoid information being provided in a universal format at one time or in acute situations. If surgery/treatment is planned, suitably individualised education should be provided irrespective of age. To be able to continue the individualised education following the transition to adult healthcare, knowledge about previous problems should be transferred.

Educational materials and methods

A number of different materials and methods were proposed or had been experienced. It was related to that education could be provided to the respondent alone or together with family members or other persons with congenitally malformed hearts. Information to teachers and other school personnel was also highlighted. No respondent had experienced formal group education together with other persons with congenitally malformed hearts. However, it was mentioned as an alternative.

The information that the respondents had received had been provided verbally. The only written material received were pictures of the heart and cards with information about endocarditis prophylaxis. The written card with information about this was experienced as useful to keep in the wallet. The respondents had not received any books or pamphlets. Some mentioned receiving copies from medical records as an alternative. It was stated that it would advantageous to receive good written information material. This would facilitate for the person with heart defect to pass on the information to people around them.

One education method mentioned was to have moving or three-dimensional pictures of the heart that could turn around in any direction or a model of a heart to open up and look into. Sequences of photos from the heart surgery together with the operation report and a meeting with the surgeon was one suggestion how knowledge could be improved. The Internet was also mentioned as a potential source of information, however some experienced education over the Internet to be dubious since it was difficult to track the origin of the information and if it was reliable. When searching the web for information the respondents experienced that they lacked knowledge about, for example, the name of their heart defect. This made it difficult to know if the information found was applicable to their situation. Most information on heart defects on the Internet was in English and despite the low mean age in the group of adults with congenitally malformed hearts, it was mentioned as a barrier that there was a lack of information in Swedish.

Needing to be met with respect

There was a need to be met with respect and kind treatment, to receive trustworthy information, feel confident and encounter an environment open to questions regarding care and treatment. The person providing information should communicate knowledge and experiences and reply to questions con-

tinuously and with an active interest to help the respondent to understand. Individualised education should be provided in a dedicated, serious, convincing and truthful way in calm surroundings, preferably not in the examination room where feelings of inferiority were experienced. Information should be facilitated by the healthcare professional using understandable language.
The respondent stated that it is respectful of healthcare professionals to admit if not having sufficient knowledge to answer a question or give education and then send the respondent to those that have the knowledge.

Managing life

The congenital heart defect

The respondents experienced that education should only contain individualised information about their specific heart defect, the aetiology, appearance, function and symptom and what had happened since childhood.
Knowledge is a source of independence and feelings security and control and helps adults with congenitally malformed hearts to manage independently in society.

Physical activity

The respondents could manage symptoms in different activities in daily life. However, education is not everything; the respondents also need to learn how to manage physical activity through their own experiences in daily life.
If the respondents did not have individualised information about how much physical activity they could do without a negative effect on the heart, they felt uncertain about physical activity and had problems managing the physical aspect of daily life. Some felt restricted and overprotected and others overexerted themselves and became very tired and symptomatic.

Life situation

Experiences of education about different life situations such as having children, heredity, length of life, work and self-care were described. The respondents wanted information to be on an individualised comprehensive level that

took the respondent's life situation with regard to family situation, work and spare time into consideration.

Treatment

Experiences of information about diagnostics and treatment such as medical examination, medication, before and after heart surgery or other procedures e.g. direct current cardioversion of supraventricular tachycardia were described. Regular medical examinations confirm that everything is all right or that something has changed. Not having adequate knowledge about medication complicates adherence and communication with healthcare professionals. Information before heart surgery or other treatment should not only consist of the preparations and the surgery/procedure, but also the postoperative and rehabilitation phase.
The respondents wanted knowledge about, for example, symptoms they could get both directly afterwards and later on, so they could manage situations that appeared, and managing life. Respondents also had thoughts about scars after surgery, which nobody had told them about, what it could be like and how to handle it.

Healthcare resources

Experiences of information about healthcare resources such as a physiotherapist, social worker and psychologist were described. Knowledge about resources could improve independence and control e.g. after heart surgery and in daily life. There were experiences of needing i.e home help or physiotherapy when returning home following heart surgery. A physiotherapist could help with exercises, a psychologist could help to handle the heart defect mentally and a social worker could provide information regarding economical and practical help.

Tools for evaluation - KnoCoMH

In study II a tool for evaluating knowledge was developed, during phase 1 and psychometricly evaluated during phase 2. Here the results from the psychometricly evaluation of the four areas in KnoCoMH (General knowledge,

Medical treatment, Endocarditis prophylaxis and Contraceptives and Pregnancy) is presented.

In domain *General knowledge*, consisting of 29 items, item difficulty index, ranged between 0.16 to 0.98 (mean 0.56) (Appendix A). Two items (item 6:2 and 6:4) were correctly answered by more than 95 % of the participants and were omitted. Item-total showed correlation between -0.21 to 0.37. Seven items (2, 6:2, 6:4, 6:12, 6:13, 20, 21) had item-total correlation < 0.30 and were omitted. Due to the clinical relevance of the items (to ensure an adequate content coverage), a total of 13 items (3, 5, 6:1, 6:7, 6:8, 7, 10, 15; 16, 22-25) were retained even if the item-total correlation was < 0.30.
The final domain General Knowledge (Appendix B and C, Swedish and English version) now includes 22 items with an internal consistency reliability of 0.68 (Appendix A). Discrimination index ranged between 0.11 to 0.68. In the test-retest, the domain showed a significant correlation (*r* 0.55) between the test occasions, with an ICC of 0.41 (Table 8).

In the domain *Medical treatment* consisting of 8 items, item difficulty index ranged from 0.16 to 0.92 (mean 0.62) (Appendix A). The item-total showed correlation between 0.22 to 0.60. Due to clinical relevance, item 4h was retained even if the item-total correlation was < 0.30. The domain had internal consistency reliability of 0.74. Discrimination index ranged between 0.22 to 1.0. The test-retest showed a significant correlation (*r* 0.50) between the test occasions, with an ICC of 0.47 (Table 8).

In domain *Endocarditis prophylaxis,* consisting of 13 items, item difficulty index ranged from 0.05 to 0.73 (mean 0.33) (Appendix A). The item-total correlation ranged from 0.28 to 0.90 and the domain had internal consistency reliability of 0.90. Discrimination index ranged between 0.16 to 1.0. The test-retest had a significant correlation (*r* 0.67) between test occasions, with an ICC 0.65 (Table 8).

In domain *Contraceptives and Pregnancy*, consisting of 4 items, item difficulty index ranged from 0.27 to 0.66 (mean 0.48) (Appendix A). The item-total correlation ranged from 0.08 to 0.52. Item 26 had item-total correlation < 0.30 and was omitted. Item 29 was retained even if item-total correlation was < 0.30. The final domain Contraceptives and Pregnancy now includes 3 items with an internal consistency reliability of 0.65. Discrimination index ranged between 0.79 to 1.0. The test-retest had a significant correlation (*r* 0.67) between test occasions, with an ICC 0.65 (Table 8).

The final instrument now consists of 46 items in four domains; General Knowledge with 22 items, Medical treatment with 8 items, Endocarditis pro-

phylaxis with 13 items and Contraceptives and Pregnancy with 3 items (Appendix B and C (Swedish and English versions).

Table 8: Test-retest KnoCoMH

Domains		Initial Test[1]		Retest[2]		Difference		
	Possible Score	Mean Score±SD	*n*	Mean Score±SD	*n*	Mean Score±SD	*r*	ICC (95% CI)
General knowledge	22	11±3.3	58	14±3.5	57	-2.9±3.2	0.55*	0.41 (0-0.67)
Medical treatment	8	5.4±1.3	19	5.6±2.0	18	-0.3±1.8	0.50*	0.47 (0.02-0.76)
Endocarditis prophylaxis	13	4.6±3.9	38	5.8±4.6	37	-1.0±3.5	0.67*	0.65 (0.42-0.80)
Contraceptives and pregnancy	3	1.6±1.0	27	1.6±0.9	26	-0.08±0.8	0.67*	0.65 (0.35-0.83)

SD=standard deviation; *n*=number; *r*= Pearson's rho; ICC= intraclass correlation coefficient; CI= confidence interval. [1]. Baseline, *n=59*. [2]. Three months later, *n*=58. *. Correlation is significant at the 0.01 level (2-tailed).

Recommendations for scoring and use

When using the KnoCoMH, scores will be calculated by dichotomising answers as correct or incorrect (Appendix B and C). Missing answers should be treated as incorrect. Twelve items will be assessed individually from the medical files and physicians' recommendations. If the participant not is recommended medical treatment, or endocarditis prophylaxis these domains should be excluded. The domain Contraceptives and pregnancy will not be included in males.

The KnoCoMH has acceptable psychometric properties for most of the knowledge domains included. It can be used for evaluating knowledge among adults with congenitally malformed hearts and its associations with other outcomes. However further studies are advisable to test construct validity, predictive validity and responsiveness.

Tools for education - a model for follow-up

In study III, a model for follow-up was described. Individualised (I) and computer-based education by a multidisciplinary team were the tools for education. This model for follow-up was also used in study IV.

The development focus of the model for follow-up was to support adults with congenitally malformed hearts in understanding their cardiac condition (meaningful learning) [77] and deliver pshychosocial support (I).

The model for follow-up will be illustrated by a case study of Charlotte 34, years of age, with tetralogy of Fallot who visited the outpatient clinic for adults with congenitally malformed hearts. This is also a description of the intervention in study IV.

Visit to the physician

Before Charlotte visits the physician, she completes an instrument to test her degree of knowledge regarding her cardiac condition; KnoCoMH (II) and a instrument to investigate symptoms of anxiety and depression, HADS, and perceived contro over the heart condition, CAS. These instruments are analysed and later used in the individual education.

During the visit to the physician, Charlotte is either only clinically evaluated and/or has an echocardiography and/or performs an exercise test. After these evaluations, the physician gives Charlotte the results/preliminary results of the tests and there is also time for Charlotte´s questions.

Visit to the specialised nurse

The next step is the visit to the nurse where Charlotte first goes through the computer-based educational program and then receives individualised (I) education and psychosocial support (I) from the nurse.

The computer-based educational program

The nurse introduces Charlotte to the computer-based educational program that relates to her congenital cardiac malformation, tetralogy of Fallot, downloaded on a CD. After the visit Charlotte keeps the CD.

Charlotte and the nurse start by looking at the cover of the CD and the list of contents on the back cover. Inside the cover, Charlotte can read the name of her cardiac malformation, if she has undergone surgery - the name and year, and whether she is recommended endocarditis prophylaxis. The name and phone number of the physician/nurse is also written on the cover. Charlotte then starts the computer-based educational program and the nurse shows her how to navigate through the main areas and subheadings (Table 7). They also look at an example of the self-conducted test (Picture 1) that ends every main

area. Now Charlotte runs the entire program herself, excluding the main area with Endocarditis Prophylaxis, as the physician did not recommend endocarditis prophylaxis to Charlotte and the nurse assists if needed. Sixty minutes later Charlotte has finished the computer-based educational program.

Individual education and psychosocial support

Now the nurse and Charlotte sit down for thirty minutes. First they look through the results of the knowledge questionnaire, KnoCoMH (II) which Charlotte filled in before her visit to the physician. They discuss the content of the computer-based educational program and the questions in the KnoCoMH from Charlotte's perspective. Through this procedure both Charlotte and the nurse gain an understanding regarding Charlotte´s knowledge, and where there are gaps. During the discussion regarding the KnoCoMH and the computer-based educational program, Charlotte's individual needs of knowledge is registered by the nurse. The next step is to evaluate the need of psychosocial support. To evaluate the psychosocial aspects 26 questions previously used by Kampuis et al [6] was included. They cover different subjects such as life/health insurance, education, employment, sport, and other daily activities (Table 9).
The questions are discussed and the nurse registers the results. If there are difficulties, the nurse asks if they are due to the malformation. Only difficulties related to the cardiac condition are registered. During this discussion, Charlotte's problem areas will appear. The discussion involve Charlotte's situation, her cardiac malformation and her new knowledge may reveal areas that she wants to change or know more about. Charlotte has always wanted to be more physically active but she is unsure about how much she can do. This becomes Charlotte's individual goal; to increase her physical activity. An individual care plan is then developed for her to reach this goal. Charlotte also gets a contact card for the nurse.

Table 9. Difficulties in daily life parts of a questionnaire previously used by Kampuis et al [6].

Did you ever feel restricted in your choice of: Educational course? Job? Sport? Hobby? House? Holiday destination?	*Were you ever excluded from:* An educational course? A job? A possible promotion? A job after medical examination? Sports after medical examination?
Did you ever give up: An educational course? A job? A sport? A hobby? A holiday?	*Were you ever prevented from:* Applying for a driving licence? Taking out a mortgage policy? Taking out a life insurance policy? Taking out a health insurance policy?
Did your physician ever advise you not to have children? *Did you ever think it was better not to have children?*	*Did you ever have to pay a higher premium than other people for:* A mortgage? A life insurance? A health insurance?

If Charlotte had any problems with the HADS questionnaire (scale score 8-10 = borderline for symptoms of anxiety/depression, >11 clinically significant anxiety/depression [116]) the nurse also addresses this with Charlotte and contacts the social worker in the multidisciplinary team. This team consists of a physiotherapist, dietician, hospital social worker, cardiologist, gynaecologist, anesthesiologist, obstretician and a pediatrician.
The nurse contacts the physiotherapist to organise an appointment with Charlotte. The physiotherapist now has the responsibility to follow up Charlotte´s goal. The physiotherapist always remains in contact with the responsible physician if any questions should occur regarding the degree of physical activity.

One month follow-up

One month after the visit to the hospital outpatient clinic the nurse telephones Charlotte as planned. The reason for this follow-up is to check if Charlotte has any questions, if the goal set has been reached and how to plan for the future. Charlotte can also choose to make contact with the nurse herself if needed. No further follow-up is planned.

Effects of individualised and computer-based education and psychosocial support

In study IV, effects such as knowledge, perceived control and symptoms of anxiety and depression were evaluatated between groups and over time at the 3 and 12-month follow-ups. The intervention group was provided with the model for follow-up, see study III. The control group received standard care including a visit to a specialised cardiologist with clinical evaluations and/or echocardiography and/or exercise tests. No structured education programme or written educational material was provided.

Knowledge

The intervention group increased their general knowledge about their heart condition after 3-months and the effect remained at the 12-month follow-up (medium effect size) (Table 10 and 11).

Table 10. Knowledge of the four areas in KnoCoMH

KnoCoMH	**Interv.** *n*	**Contr.** *n*	**Interv.** mean (SD)	**Control** mean (SD)	***Effect size***	***p-Value***
General Knowledge (Max score 22), *n*=114						
Baseline	56	58	10.9 (3.6)	11.1 (3.3)	-0.06	0.789
3-month	50	57	16.2 (3.7)	14.0 (3.5)	0.63	**0.003**
12-month	50	54	16.0 (3.6)	14.4 (3.0)	0.53	**0.020**
Medical treatment (Max score 8) *n*=37/37/42*						
Baseline	18	19	4.6 (2.3)	5.4 (1.3)	-0.61	0.189
3-month	18	19	4.7 (3.0)	5.3 (2.3)	-0.26	0.502
12-month	18	24	4.4 (2.7)	4.5 (2.5)	-0.04	0.850
Endocarditis prophylaxis (Max score 13) *n*=75						
Baseline	37	38	4.0 (3.9)	4.6 (3.9)	-0.05	0.487
3-month	34	37	9.8 (3.0)	5.8 (4.6)	0.87	**< 0.001**
12-month	32	37	8.3 (3.7)	6.9 (4.1)	0.34	0.146
Contraceptives and Pregnancy (Max score 3) *n*=56						
Baseline	29	27	1.1 (1.1)	1.6 (1.0)	-0.5	0.118
3-month	26	26	1.6 (1.1)	1.6 (0.9)	0	0.783
12-month	26	25	1.9 (0.9)	1.7 (0.8)	0.25	0.516

*In total, 37 individuals had medical treatment at baseline, 18 participants in the intervention group, and 19 in the control group. There were no treatment changes in the 3-month data. After the 12-month follow-up, 5 new participants in the control group had medical treatment. Contr.= Control group. Interv.= Intervention group. SD= Standard Deviation. *n*= Number.

A total of 37 participants in the intervention group were recommended endocarditis prophylaxis and their knowledge regarding endocarditis had increased at the 3-month follow-up (large effect size). After 12-months there were no differences between the groups (Table 10 and 11), but there was a significant difference of knowledge over time between baseline and the 12-month follow-up (Table 11) pointing out that learning occurs over time (medium effect size) (Table 10 and 11). There were no changes in knowledge in the domains medical treatment and contraceptive and pregnancy in either of the groups (Table 10 and 11).

Table 11. Differences in knowledge between 3-month follow-up/baseline and 12-month follow-up/baseline

KnoCoMH		**3-month/baseline** Mean diff. (SD) *n*	**12-month/baseline** Mean diff. (SD) *n*
General Knowledge			
(Max score 22)	Interv.	5.4 (3.3) 50	4.9 (3.8) 50
	Control	2.9 (3.2) 57	3.0 (3.5) 54
	(95 % CI)	-2.4 (-3.7 to -1.2)	-1.8 (-3.3 to -0.4)
	p-value	**< 0.001**	**0.011**
	Effect size	0.78	0.54
Medical treatment			
(Max score 8)	Interv.	0.2 (1.9) 18	-0.2 (2.7) 18
	Control	-0.1 (2.2) 19	-1.0 (2.7) 19
	(95 % CI)	-0.2 (-1.6 to 1.2)	-0.8 (-2.6 to 1.0)
	p-value	0.752	0.358
	Effect size	0.04	-0.29
Endocarditis prophylaxis			
(Max score 13)	Interv.	6.0 (4.0) 34	4.3 (3.9) 32
	Control	1.0 (3.5) 37	2.2 (3.6) 37
	(95 % CI)	-5.0 (-6.7 to -3.1)	-2.1 (-3.9 to -0.3)
	p-value	**< 0.001**	**0.021**
	Effect size	1.43	0.58
Contraceptives and Pregnancy			
(Max score 3)	Interv.	0.4 (1.2) 26	0.8 (1.2) 26
	Control	0.1 (0.8) 26	0.0 (1.0) 25
	(95 % CI)	-0.3 (-0.9 to 0.2)	-0.8 (-1.4 to -0.2)
	p-value	0.273	0.014
	Effect size	0.37	0.80

Contr.= Control group. Interv.= Intervention group. SD= Standard Deviation CI= Confidence Interval. *n*= Number.

Perceived control

There were no differences between the groups at baseline and the perceived control over the heart condition showed normal values (mean 21, SD 5.3 in control group, mean 21, SD 6.1 in intervention group). No change was found between groups over time at the 3-month follow-up (mean difference -1.0, SD

5.2 in control group, mean difference 0.3, SD 4.5 in intervention group, *p*-value 0.17 ,effect size -0.13) and the 12- month follow-up (mean difference -0.5, SD 4.9 in control group, mean difference -0.9, SD 4.6 in intervention group, *p*-value 0.68 effect size -0.28).

Anxiety and depression

There were no differences between the groups at baseline and symptoms of anxiety and depression showed normal values respectively (mean 4.6/2.0, SD 3.6/2.1 in control group, mean 4.8/2.8, SD 3.9/3.4 in intervention group).No change was found between groups over time at the 3-month follow-up (mean difference in symptoms of anxiety/depression -0.4/0.2, SD 2.2/1.7 in control group, mean difference -1.0/-0.7, SD 2.3/2.8 in intervention group, *p*-value 0.22/0.06 ,effect size -0.63/-0.53) and the 12-month follow-up (mean difference in symptoms of anxiety/depression -0.7/0.2, SD 2.5/2.5 in control group, mean difference -0.5/-0.6, SD 3.1/3.0 in intervention group, *p*-value 0.71/0.20 ,effect size -0.48/-0.32. The intervention with education and psychosocial support did not produce symptoms of anxiety and/or depression.

Perceptions of the model for follow-up

When attending the model for follow-up (III, IV), the participants also answered questions about how they perceived their knowledge, anxiety and satisfaction with the model. Before attending the model for follow-up, 47 % perceived their knowledge of their heart condition to be "good" or "excellent", three months later these numbers had increased to 89 %. Regarding symptoms of anxiety, 38% perceived "some anxiety" or "much anxiety" in connection with their heart condition before the model for follow-up. After three months this remained unchanged (39 %). Directly after attending the model for follow-up, 88 % perceived their satisfaction with the combination of the visit to the physician and the nurse to be "very good" or "excellent" but after three months this number had decreased to 75 %.

DISCUSSION

Discussion of Results

In this thesis, educational needs, the effects of tools for gaining knowledge and psychosocial support were evaluated.

Psychosocial support and knowledge are important tools for achieving self-management [3, 70]. Psychosocial support is also an important tool for supporting the process of reaching knowledge due to the opportunity to individualise the achievements depending on needs [117-118].

Educational needs and a model for follow-up

In this thesis two-way communication (I) was found to be crucial in order to enahnce knowledge when giving information. The World Health Organisation, WHO, stated in 1986 that patients should receive information and participation should be encouraged [119]. When two-way communication was facilitated there was an individualisation of the education, which was perceived to increase knowledge and provide tools for managing life (I).

According to the Swedish Healthcare Law, SFS.1982;763, all patients should be informed of their state of health and of the treatment options available and take part in the decisions regarding their care and treatment.

When information was provided without specific consideration to the individual's unique situation the respondents felt it did not apply to themselves (I). Relevance is also known to be a crucial factor for meaningful learning and adherence to self-care [70-71, 77]. Eld et al. found similar experiences of non-participation among people with heart failure when standard information was given rather than individualised information [120]. General education rather than individualised education was perceived to be a barrier for communicating the information to other people (I). Not having sufficient knowledge was also experienced (I) to be a barrier for taking part in decisions about the treatment e.g. choice of medication. In agreement with our study I, other studies have shown that it is important to have access to knowledge in order to be able to inform others e.g. family members, people at school, friends, sports coaches or general

practitioners [121-123]. The medical language was also perceived to be a barrier for access to knowledge (I). This has also been described by Birks et al among children and young people, with congenitally malformed hearts, aged between 8 and 19 years [123]. Identifying personal barriers is one of the important components in support of self-management [118]. It is difficult to come to an agreement without identifying the barriers. Difficult words could be another reason for misunderstanding and uncertainty. To avoid this, the computer-based education used in the model for follow-up (III, IV) was screened for difficult words by a licensed translator in Swedish.

Information given without good communication and knowledge on how to educate was also mentioned as a waste of time (I). Healthcare professionals are often known to focus mainly on delivering information without prioritising the communication. In addition, the person who is the object of attention is seldom invited to initiate discussions regarding the given information [124-125]. To provide self-management, education should focus on the individual´s situation and needs [70]. To improve knowledge it is also important to consider how to develop a tool for education [77-78]. The structure and amount of text and illustrations are important to avoid extraneous details which are known to decrease learning [79].

We found that adults with congenitally malformed hearts emphasised the importance of being met with respect, e.g. the way information was provided and adapted to present needs and that the way it should be provided on a regular basis was more important than who provided it. This is also known as the individual's right to autonomy, self-determination [126].
It was emphasised that truthful information provided with personal engagement and with openness to each individual situation was important.

In the development of the model for follow-up (III) the aim was to fulfil this need by individualised education, computer-based education and psychosocial support.

Education theories point out the importance of having a teaching schedule ready before starting any type of education [77]. The participants in study I perceived it to be important to have an educational plan and mutual goals which were also used in study III and IV. One strategy for evaluating a person's educational needs is to test the knowledge through an instrument developed for the population (III) and through the instrument communicate with the persons about the knowledge they have and possible barriers [117-118] for learning.

In this thesis the educational material used (III, IV) was an interactive computer-based educational program [106]. Other studies have shown that com-

puter-based education significantly improve knowledge [87]. The participants (I) perceived that information about the heart defect should contain individualised education on their specific heart defect, the cause, appearance, function, symptoms and progress since childhood, heredity, future aspects, optional work and leisure activities, self-care and length of life. This was considered when choosing the content in the individualised and computer-based education, and psychosocial support in studies III and IV.

Preparation for e.g. future heart surgery was mentioned as an important area of knowledge. Participants in study I had experienced problems with teachers in secondary school, which has also been reported in other studies on young people with congenitally malformed hearts [123]. Problematic situations mentioned were connected to the need to take some time off when visiting healthcare professionals or in connection with e.g. heart surgery. They also had financial problems and there were needs for financial advice.

Today more children with complicated heart defects reach adult age and there are more complex situations surrounding them. Healthcare resources such as a physiotherapist, social worker and psychologist were mentioned as important sources for support. The importance of a multidisciplinary team for the person with a heart defect was shown in study I. There were requirements for individualisation from a holistic approach which was the intention of the development of the model for follow-up in study III, which was then tested in study IV.

A tool for evaluating knowledge; KnoCoMH

Improvement with regard to increasingly healthy behaviours, less health care utilisation and better quality of life are some of the commonly used assessments in patient education and the methods vary depending on the outcomes measured. Knowledge itself is often evaluated in various forms of education, but rarely in patient education. The most influential component for learning at university level is the examination [72].

Evaluation of knowledge in adults with congenitally malformed hearts has previously been studied, but studies resulting in a complete instrument with scoring and comprehensive psychometric evaluation is missing [4].
The KnoCoMH (II) was developed and psychometric tested. This way of evaluating knowledge is not complete or the most sufficient way to evaluate the understanding of a phenomenon [127]. It may be stated that this type of instrument does not evaluate knowledge on the higher levels of for example the

taxonomies of Bloom[128] or Solo[129] . To do this more complex and qualitative evaluations are needed [127]. However, in research studies being able to quantify outcome measure is of importance and developing the KnoCoMH is a first step to a valid and reliable tool for evaluating the level and amount of knowledge in four domains (General knowledge, Medical treatment, Endocarditis prophylaxis and Contraceptives and Pregnancy) in relation to cardiac malformations. Ability to discriminate between people with different levels of knowledge is an important property for a knowledge test like KnoCoMH [100, 102]. After the refinement, the scale items demonstrated good variation in item difficulty. This implies that the test items cover different levels of knowledge, which gives the possibility to test progress in knowledge [100]. One exception was the domain Endocarditis prophylaxis, which had more difficult items compared to the other domains, reflected in the low mean of the item difficulty index. The consequences can be that this scale lacks discrimination ability among people with low levels of knowledge. In addition the domain Contraceptives and Pregnancy lacked the easiest and the most difficult items but in contrast to the domain Endocarditis prophylaxis, the mean of the difficulty index was higher. The reason for this is probably that this domain only includes three items.

A tool for education – individualised and computer-based education

To our knowledge this is the first initiative to use a computer-based educational program [106] for adults with congenitally malformed hearts. The strength of this program is that it is based on clinical experience, research literature and developed in close collaboration with formal users (adults with congenitally malformed hearts). According to previous studies of learning, one of the most important aspects to consider in order to reach understanding and knowledge, is relevant content and illustrations [77-78]. During the development the users' needs were the main focus (I), and which has previously been shown to be the determining factor for the usability of developed programs [130-132].

The education theory of constructivism was used with the intention to support deeper knowledge both in the individualised and computer-based education [75, 77]. The objective of the computer-based educational program was to visualise the relationships between the congenital heart defects and the normal heart, what has happened since childhood or can happen in the future. The purpose with the individualised education was to assess needs and pro-

vide individualised advice and agreements in order to emphasise the individual's role in caring for themselves [118].

Both the computer-based and individualised (I) education and psychosocial support (III-IV) in this thesis focused on developing an understanding of the heart condition. Knowledge was defined as the ability to repeat and transfer information into new situations. A model for this was developed and evaluated and knowledge by the KnoCoMH was evaluated. Whether the increased level of knowledge also led to behaviour changes was not evaluated.

Evaluation of a model for follow-up

Descriptions of the development and testing of different educational initiatives for adults with congenital malformations are scarce. As far as we know this is the first randomised trial evaluating an intervention with education and psychosocial support in this population. Our results showed that the intervention improved and retained knowledge over time. According to Cohen´s effect size index, the improvement of knowledge over time was between medium to large [133]. As this is the first study, the clinical significance of the increased knowledge on long-term effects regarding health outcomes in adults with congenitally malformed hearts is not yet known.

From the individuals' perspective, this thesis (I), in accordance with other studies, showed the importance of having access to knowledge in order to be able to inform those around e.g. family members, people at school, friends, sports coaches or general practitioners [121-123].

To our knowledge this is also the first study evaluating perceived control in adults with heart malformation. Increased knowledge about the heart condition has previously been found to decrease anxiety and improve perceived control in people with coronary heart disease. [134-135]

Our data showed low scores of anxiety and high levels of perceived control which indicates that most of the participants were in a stable condition and had adapted to living with their malformed hearts. However, since the intervention was promising in terms of increasing knowledge, further studies are needed to assess long-time effects of self-management behaviours and perceived control.

Today there are needs to increase the experiences and knowledge on how to organise sufficient and successful education and psychosocial support to improve outcomes and increase the quality of care in this growing population [136-

[139]. This thesis has contributed to an increase of this knowledge but further evaluation of long-term effects is needed.

Methodological considerations

The qualitative study

It was important to outline the individuals' views before developing education for adults with congenitally malformed hearts. To fulfill the aim a qualitative methodology was preferable. Phenomenography was chosen since the method has roots in the pedagogical area. Phenomenography can be used to describe different ways of experiencing a phenomenon in the surrounding world, to describe the variation in the understanding of a certain phenomenon, looking for differences and similarities, and to find the underlying structure of variance [95].

A strategic selection of the respondents was made in order to obtain as many different experiences of educational needs as possible. In phenomenography differences are described which should guide the selection of participants. In our study we chose different congenital heart malformations, both complicated and non-complicated, some operated and some not, people of different ages and of both genders. We considered that the strategical selection of participants was optimal according to phenomenography. Regarding generalisability, qualitative studies refer essentially to transferability whether the findings can be transferred to other settings or groups [127].

Every individual tells the truth and the truth can be different for different people. Depending on the quality of the description of the context of the data collection, for example how well the characteristics of the participants have been described, the reader can judge to which groups the results are transferable. To assess the interater reliability in our data analyses, an independent researcher not involved in the study reviewed one-third of the statements and sorted them into the category system. After modification of the category system satisfactory agreement was established. Another way of assuring credibility of the data in qualitative studies is triangulation. We used investigator triangulation in the data analysis [127].

The quantitative studies

Studies II and III were the prerequisite for a solid methodology in study IV. Study II evaluated the reliability and validity of the instrument through statistical methods. Study IV evaluated the effects of an intervention consisting of a computer-based and individualised education and psychosocial support by a multidisciplinary team. The primary outcome was knowledge measured by the instrument developed and tested in study II. The instrument was developed and tested in a Swedish population. A strength is that the KnoCoMH showed acceptable psychometric properties regarding item difficulty level, internal consistency and test-retest reliability. As randomisation was used the groups could be controlled. As for blinding the participants, the physician and the study nurse were blinded regarding the group to which the participant was randomised until after the medical consultation. This was done in order to eliminate the risk of the physician improving the way he/she provided information. To control that the results of the instrument KnoCoMH truly mirrored the knowledge of the participants they always answered the instrument at the clinic at baseline and through a telephone interview at the 3 and 12 month follow-ups.

The overall aim of this thesis was to describe educational needs, to develop a tool for assessing knowledge and to evaluate the effects of a follow-up model providing education and psychosocial support to adults with congenitally malformed hearts. Effects of psychosocial support or self-management in itself were not evaluated.

There is always a risk that the participants in the control group improve or modify their behaviour in response to the fact that they are being studied and we found some learning effects also in the control group. This is known as a non-specific effect which is difficult to control [140]. Further many adults with congenitally malformed hearts declined participation. These individuals are in the middle of life studying, making a career or starting a family. [55]

Clinical implications

The World Health Organisation has stated that patients should receive information, and that participation in self-care should be encouraged.[119] According to the Swedish Healthcare Law, SFS.1982;763, all patients should be informed of their state of health and about the treatment options available and take part in the decisions about their care and treatment.
This thesis shows that adults with congenitally malformed hearts have a need for knowledge regarding their heart condition, which also confirms previous studies underlining the importance of preparation of structured educational programmes for this population [6, 8-10]. Previously there has been a lack of knowledge regarding the design of structured programmes in order to improve required knowledge. This thesis provides a model for follow-up in the hospital out-patient clinic which was found to increase knowledge over time, but the effects on long-term outcome have to be proven.

The model for follow-up developed in this thesis is in line with the conceptual model for self-management support delevoped by Glasgow and colleges [117-118]. Figure 4 is a schematic presentation on how the model was working in the clinical setting.

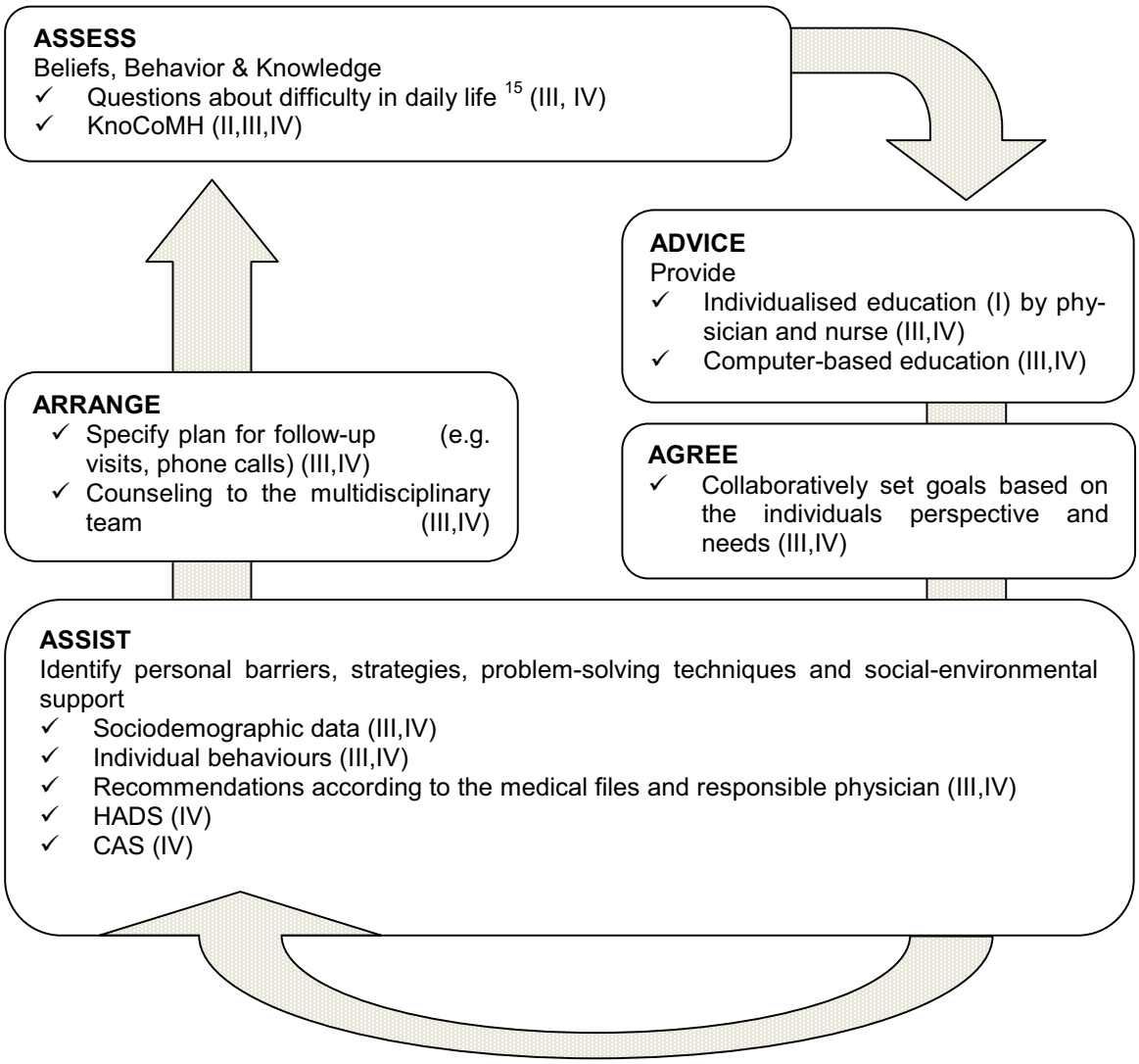


Figure 5. Schematic presentation of the model for follow-up (III, IV) [117-118].

Adults with congenitally malformed hearts need to perform self-management i.e. medical treatment, awareness of symptoms and contribute to regular follow-up, prophylaxis of endocarditis, recommendations on physical activity, employment, sports and spare time and risk reduction in connection with pregnancy [16-18]. It is known that by involving the person in the care through their personal goals a better adherence to self-management behaviours can be achieved [70].

The role of healthcare professionals is important. Support to reach adaptation of the participant's own care plan is the goal.
Glasgow and colleges have conceptualised the five A's; Assess, Advice, Agree, Assist and Arrange, with a Personal action plan [117-118]. In our model for follow-up ASSESS of beliefs, behaviour and knowledge was collected by the Kno-CoMH instrument and questions about difficulty in daily life previously developed by Kampuis et al [6]. ADVICE was provided by individualised education by physician and nurse and by the computer-based education. AGREE was reached through the individual conversation between the participants and the nurse where goals based on the individual's perspectives and needs were set. During the ASSIST, personal barriers, strategies, problem-solving techniques was identified with support from the socio demographic data, the questions about difficulty in daily life [6], and the physicians recommendations. The instruments; HADS and CAS was also used in this field (ASSIST). ARRANGE involves to plan for and arrange around the participants in hence of the individuals needs.

These different steps occurred also during the different stages in the model for follow-up. The circle with the five A's is sometimes pass once and sometimes twice or more. The importance is not to follow the circle, the important is to individualise the steps according to the individuals needs.

Research implications

There were different experiences regarding when the education should start. Most issues were that it should start as soon as possible in early childhood, build up and be adapted to the developmental stage of the child. Further research is needed in this area.

The participants in study I gave a few descriptions of what the information should contain; individualised education about their specific heart defect, the cause, appearance, function, symptoms and progress since childhood, heredity, future aspects, optional work and leisure activities, self-care and length of life. Further research is needed to evaluate if the model for follow-up fulfil this needs.
Development and further evaluation of KnoCoMH with an expanded response format is advisable. Additional testing of construct validity, predictive validity and responsiveness is needed.

Further evaluation of the participants' perception of the intervention should be studied.

Long-term effects of the model for follow-up should be further evaluated.

The use of the model for follow-up should be tested in adolescence.

KnoCoMH should be tested on adolescent.

The emerging burden of hospital admissions of adults with congenitally malformed hearts were two-three times higher than in the normal population in the Netherlands[141] - can the model for follow-up tested in this thesis change these long-term outcomes?

CONCLUSIONS

Two-way communication needs to be taken into account to enhance knowledge when healthcare practitioners give information to adults with congenitally malformed hearts. Two-way communication facilitates individualisation and respect.

The perception of when to start the education about the heart condition varied. Some wanted it to start as soon as possible in early childhood, and gradually build up, and to be adapted to the developmental stage of the child.

To having insufficient knowledge was experienced as a barrier for taking part in decisions about treatments.

There was a need to be respected as a human being, particularly in relation to how the information was provided and adapted to the participants' present needs using clear and simple language, without medical terms and Latin words.

The knowledge scale, KnoCoMH has acceptable psychometric properties regarding item difficulty level, internal consistency and test-retest reliability, and can now be used to estimate knowledge in adults with congenitally malformed hearts within the four domains: General knowledge, Medical treatment, Endocarditis prophylaxis and Contraceptives and Pregnancy.

A model for follow-up was developed focusing on individualised and computer-based education and psychosocial support by a multidisciplinary team with the purpose to be used in the hospital out-patient clinic.

The model was effective in improving and maintaining knowledge about self-management in adults with heart malformation.

SAMMANFATTNING PÅ SVENSKA (SUMMARY IN SWEDISH)

Många av de som lever med ett medfött hjärtfel behöver livslång uppföljning inom sjukvården på grund av risken för komplikationer. Komplikationer som kan förekomma beror oftast på en försämrad hjärtfunktion på grund av det medfödda hjärtfelet eller tidigare hjärtoperation. För att minska risken för komplikationer krävs en god egenvårdsförmåga i form av följsamhet till läkemedelsbehandling och sjukvårdsbesök, förebygga endokardit, val av yrke och fritidsaktiviteter, preventivmedel och graviditet, men också val av livsstil som till exempel att vara fysiskt aktiv och inte röka.

En förutsättning för att kunna utföra egenvård är bland annat att ha tillräckligt med kunskap. Tidigare forskning understryker att nivån och omfattningen av kunskap bland vuxna med medfött hjärtfel är bristfällig. Som exempel kunde endast mellan 54 till 76 % nämna sin hjärtdiagnos. Det fanns även kunskapsbrister gällande läkemedelsbehandling och förebyggande av endokardit.

En person med medfött hjärtfel beskriver det så här (Verstappen et al[5], s 519);

- Jag önskar någon hade suttit ner med mig och förklarat, "Det här är ditt medfödda hjärtfel, detta är konsekvenser av ditt hjärtfel och detta är vad vi kontrollerar och tittar efter. Detta är vad vi vet och detta vet vi mindre om. Detta gör vi för att hjälpa dig och detta kan du själv göra för att optimera ditt liv"

Det finns idag få studier som beskriver och utvärderar utbildning för personer med medfött hjärtfel och utbildningsmaterial saknas. Tidigare studier som utvärderat kunskap visar stora skillnader i hur kunskapsnivån har utvärderats. Detta gör att det är svårt att sammanställa och jämföra dessa resultat. De instrument som använts har inte genomgått en tillräcklig psykometrisk testning och/eller enbart utvärderats i vissa delar som till exempel kunskap om endokardit profylax. Inför utveckling och testning av olika metoder och modeller för att utbilda vuxna med medfött hjärtfel är det viktigt att först utveckla ett psykometriskt testat instrument som har förmågan att utvärdera kunskap inom de olika områden som kan vara viktigt för vuxna med medfött hjärtfel att ha.

Nationella och internationella riktlinjer rekommenderar uppföljning av vuxna med medfött hjärtfel riktat både mot medicinska och psykosociala problem. Idag är olika program för uppföljning etablerade runt om i värden. Mo-

ons med kollegor beskriver rådande status av vård av vuxna med medfött hjärtfel i Europa 2010. Totalt ingick 50 specialistmottagningar från 18 olika länder i denna studie. Av dessa var 47 program (94 %) placerade på Universitetssjukhus. Totalt hade 94 % av dessa progam hjärtläkare specialiserade på vuxna med medfött hjärtfel och 68 % hade även specialiserade sjuksköterskor. Hur dessa program för uppföljning var utformade presenterades inte i detalj. En modell för uppföljning i Italien har beskrivits av Chessa och medförfattare. De har tillsammans med andra kliniker skapat en hemsida på internet med syfte att kunna samarbeta mellan dessa olika centrum för uppföljning. Patienter, sjuksköterskor och läkare har tillgång till information genom webben utan inloggning för att underlätta möjligheten för lätt tillgång till information. Effekterna av denna websida är ännu inte presenterade. Ett annat program för uppföljning av medfödda hjärtfel är Copenhagen Transition Program. Detta är en sjuksköterskebaserad öppenvårdsmottagning för unga vuxna med medfött hjärtfel. Deras uppföljning fokuserar på att öka kunskap om de situationer som kan uppstå när man lever med ett medfött hjärtfel. Kunskapsområden som de jobbar med är endokardit, kunskap inför akuta situationer, kostintag, preventivmedel, alkohol och droger, rökning, fysisk aktivitet, sömn/vila och utbildning. I detta program sker uppföljningen enbart med den unga vuxna, utan föräldrarnas medverkan. Långtidseffekter av detta uppföljningsprogram är inte presenterade än. Reid med medförfattare beskriver läget i Kanada när det gäller lyckade överföringar från barn till vuxensidan för 15 specialist kliniker för medfödda hjärtfel till 47 %.

Sammantaget är det en stark konsensus om att fortsatt utveckling och förbättring av vården för medfödda hjärtfel bör ske. Idag finns ett stort behov av att dela erfarenheter och öka kunskapen om hur uppföljningen av vuxna med medfött hjärtfel kan organiseras på ett effektivt sätt, samt hur utbildning och psykosocialt stöd ska organiseras för att ge bäst ger effekt och hur man kan förbättra kvalitén av vården för denna kraftigt växande population.

Det övergripande syftet med avhandlingen var att beskriva utbildningsbehov, utveckla ett verktyg för att utvärdera kunskap, och att utvärdera effekterna av en modell för uppföljning innehållande utbildning och psykosocialt stöd för vuxna med medfött hjärtfel.

Vuxna, 18 år och äldre med de tio vanligaste medfödda hjärtfelen utgör urvalet i avhandlingen. Delarbete I är en kvalitativ studie, där 16 intervjuer har gjorts och analyserats enligt fenomenografi. Syftet var att beskriva dessa individers erfarenheter av den utbildning de fått om sitt medfödda hjärtfel för att skapa förståelse för deras utbildningsbehov.

Ett instrument som ett verktyg för att utvärdera kunskap utvecklades och testades psykometriskt i studie II med 19individer initialt och sedan 114 vuxna med de 10 vanligaste medfödda hjärtfelen.

En modell för uppföljning av vuxna med medfött hjärtfel har beskrivits och initialt testats i studie III med 55 vuxna med de nio vanligaste medfödda hjärtfelet och slutligen testats i en interventionsstudie(studie IV). Totalt ingick i studie IV, 114 vuxna med en medelålder på 34 år med medfött hjärtfel. Femtiosex deltagare lottades till gruppen som testade interventionen och 58 till kontrollgruppen.

Resultatet i denna avhandling presenterar en modell, som testats för uppföljning av vuxna med medfött hjärtfel som består av individuell och datorbaserad utbildning och psykosocialt stöd via ett multidisciplinärt team (III). Kunskap och upplevd kontroll över att leva med ett medfött hjärtfel, symptom på oro och depression utvärderades vid baseline och efter 3 och 12 månader (IV). Verktyg för tillhandahålla utbildning var i denna avhandling datorbaserad och individualiserad utbildning utvecklat utifrån de resultat som framkom i studie I. Det var viktigt att ha bra utbildningsmaterial och metoder samt att informationen var given med respekt för individen. Verktyg för att hantera viktiga delar i livet som det medfödda hjärtfelet, olika livssituationer som kan uppkomma, fysisk aktivitet, behandling och sjukvårdsresurser borde tillhandahållas. Man uppfattade att detta kunde uppnås när utbildningen var anpassad till individens hela livssituation och given genom god kommunikation. Den datorbaserade utbildningen som användes som verktyg för utbildning i modellen för uppföljningen, innehåller 10 moduler, en separat modul utvecklat för vart och ett av de 10 vanligaste medfödda hjärtfelen. Varje modul innehåller 8 huvudmenyer med undermenyer som; mitt medfödda hjärtfel, orsak och ärftlighet, preventivmedel och graviditet, läkarkontakt och kontroller, endokardit profylax, medicinsk och kirurgisk behandling, arbete och fritid, sex och samliv, samt friskvård. Verktyget för att utvärdera kunskap, KnoCoMH utvecklades och testades i studie II. Denna kunskapsenkät består av 46 frågor i fyra områden; generell kunskap, medicinsk behandling, endokardit profylax, preventivmedel och graviditet och hade psykometriska förutsättningar i de flesta områden för att kunna utvärdera kunskapsbehov.

Resultatet i studie IV visade en signifikant kunskapsökning mellan grupperna. Generell kunskap var signifikant högre efter 3 månader (effekt size 0.63, p= <0.01), och 12 månader (effekt size 0.53, p= 0.02) i interventionsgruppen. Kunskap gällande endokardit profylax ökade också signifikant mer i interventionsgruppen efter 3 månader (effect size 0.87, p= <0.01), och över tid mellan baseline och 3 månader (effekt size 1.43, p=<0.001), och mellan baseline och 12

månader (effekt size 0.58, p= 0.02). Interventionen varken förbättrade eller försämrade den upplevda kontrollen över det medfödda hjärtfelet eller symptom som oro och depression. Som konklusion kan sägas att utbildning och psykosocialt stöd via ett multidisciplinärt team var effektivt för att förbättra kunskap om egenvård hos vuxna med medfött hjärtfel. Fortsatt forskning behövs för att fastställa långtidseffekter av egenvårdsförmåga och upplevd kontroll över hjärtfelet.

ACKNOWLEDGEMENTS

These studies was carried out at the Department of Medical and Health Sciences, Division of Nursing Science, Linköping University and Department of Cardiology, Linköping University Hospital, but also at Department of Medicine, Kalmar Country Hospital, and Department of Medicine, Jönköping Country Hospital, and Department of Cardiology, Malmö University Hospital which is gratefully acknowledged.

I also wish to express my sincere gratitude to everyone who has supported me in my work with this thesis. Especially, I would like to thank the following persons:

All the adults with a malformed heart, who participated in this thesis, thank you for your time. Without your willingness to participate, this thesis had not been possible.

Eva Swahn, Professor, my supervisor. Thank you for listen to my idea there at the train station in Malmö year 2003. Thank you for believing in me from the beginning and giving me the opportunities and confidence to do this project and later on also as a PhD-student (since year 2005). Thank you for guiding me and all your support in all areas through this journey!

Anna Strömberg, Professor, my co-supervisor. Thank you for guiding me from the beginning, fist with schoolwork until I had the certifications to be a PhD-student and at the same time supporting me with applications both for grants and travelling grants to raise me up with important experiences for my coming work as a PhD-student but also as a model for how an excellent supervisor is working. Thank you for guiding me and all your support in all areas through this journey!

Niels Erik Nielsen, PhD, my co-supervisor. Thank you for guiding me from the beginning until the end with your clinical experiences from the area with heart defects. Thank you for your time to support me with this project in different ways.

Anna-Christina Ek, Professor emeritus. Thank you for accepting me as a PhD-student at the Department of Medical and Health Sciences, Division of Nursing Science, Linköping University.

Bibbi Göransson, my chief at the Department of Cardiology for believing in me from the beginning and giving me the opportunities and confidence to grow but also your support of me as a person. Tank you Bibbi! If I sometimes later on will become a chief, you will be my model for it.

Kristoffer Årestedt, PhD, *Ulf Thilén*, PhD, my co-authors and collaborators in this project. Thank you for your valuable competence and your enthusiasm for doing the manuscript as good as possible. Kristoffer, thank you for giving me time and your excellent knowledge in psychometric tests.

Philip Moons, Professor at the Centre for Health Services and Nursing Research at the Catholic University, Leuven, Belgium for the opportunities to visit you 2 weeks in year 2003 and your enthusiasm and willingness to share your excellent knowledge through the years with the PhD-project.

Katharina Höglund and *Kerstin Gustavsson*, Linköping, *Ingrid Elisson*, Kalmar, *Lena Tengvall*, Jönköping, *Marie Host* and *Linda Ternrud*, Malmö, *Karin Paulsson*, Lund, thank you all for invaluable contribution to the studies and for your friendship!

Niels Erik Nielsen, *Mats Broqvist*, *Anders Bergström*, *Bahram Shams*, *Aleksandra Trzebiatowska-Krzynska*, *Burkard von Wangenheim*, *Jan-Erik Karlsson*, *Martin Stagmo*, and *Ulf Thilen*, thank you all for valuable contribution in different ways in the studies.

Agneta Larein, and all other secretaries in the different departments and all of you I do not know the names of, thank you all for your contribution to the studies.

Sofia McGarvey, thank you for your support with translation and your friendship!

All of you who have giving me friendship and support in different ways, in the Division of Nursing Sciense and at the School of Nursing, Linköping Univer-

sity and the Department of Cardiology, Linköping University Hospital. Thank you for being there.

The last months, I have realised I have a lot of friends in my life! Tank you *Lisa, Lena, Ros-Mari, Anna, Ingela, Bibbi, Anita, Yvonne, Ulla, Birgitta, Pia, Susanna, Sofia, Marie* and *Eva*. Thank you for always being there when I needed.

The teachers in the two working team, in semester 3 and 4, in the Nursing School, Linköping University. Thank you (yes Yvonne, also semester 4 is okej to belong to☺, Inger is there you know☺), I have had a lot of fun with you and I will miss you all!

To you, *Joachim, Louise* and *Sarah*, you are the meaning in my life! Thank you for always believing in me and giving me love!

The man in my life, you know how you are. Thank you!

REFERENCES

1. Marelli AJ, Gurvitz M. From numbers to guidelines. *Prog Cardiovasc Dis.* Jan-Feb 2011;53(4):239-246.

2. Harrison JL, Silversides CK, AH. K. Health Care Needs of Adults With Congenital Heart Disease: Study of the Patient Perspective. *J Cardiovasc Nurs.* Mar 2 2011;2011:2.

3. Barlow J, Wright C, Sheasby J, Turner A, Hainsworth J. Self-management approaches for people with chronic conditions: a review. *Patient Educ Couns.* Oct -Nov 2002;48(2):177-187.

4. Van Deyk K, Pelgrims E, Troost E, et al. Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. *Am J Cardiol.* Dec 15 2010;106(12):1803-1807.

5. Verstappen A, Pearson D, Kovacs AH. Adult congenital heart disease: the patient's perspective. *Cardiol Clin.* Nov 2006;24(4):515-529, v.

6. Kamphuis M, Verloove-Vanhorick SP, Vogels T, Ottenkamp J, Vliegen HW. Disease-related difficulties and satisfaction with level of knowledge in adults with mild or complex congenital heart disease. *Cardiol Young.* May 2002;12(3):266-271.

7. Moons P, De Geest S, Budts W. Comprehensive care for adults with congenital heart disease: expanding roles for nurses. *Eur J Cardiovasc Nurs.* Feb 2002;1(1):23-28.

8. Moons P, De Volder E, Budts W, et al. What do adult patients with congenital heart disease know about their disease, treatment, and prevention of complications? A call for structured patient education. *Heart.* Jul 2001;86(1):74-80.

9. Dore A, de Guise P, Mercier LA. Transition of care to adult congenital heart centres: what do patients know about their heart condition? *Can J Cardiol.* Feb 2002;18(2):141-146.

10. Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. *Can J Cardiol.* Jul 1997;13(7):641-645.

11. Chessa M, De Rosa G, Pardeo M, et al. Illness understanding in adults with congenital heart disease. *Ital Heart J.* Nov 2005;6(11):895-899.

12. Shebani SO, Miles HF, Simmons P, Stickley J, De Giovanni JV. Awareness of the risk of endocarditis associated with tattooing and body piercing among patients with congenital heart disease and paediatric cardiologists in the United Kingdom. *Arch Dis Child.* Nov 2007;92(11):1013-1014. .

13. Veldtman GR, Matley SL, Kendall L, et al. Illness understanding in children and adolescents with heart disease. *Heart.* Oct 2000;84(4):395-397.

14. Van Damme S, Van Deyk K, Budts W, Verhamme P, Moons P. Patient knowledge of and adherence to oral anticoagulation therapy after mechanical heart-valve replacement for congenital or acquired valve defects. *Heart & Lung: The Journal of Acute and Critical Care.* 2010;In Press, Corrected Proof.

15. Nationella riktlinjer för vård, behandling och omsorg - Medfödda hjärtfel. Socialstyrelsen; 2008. http://www.socialstyrelsen.se/NR/rdonlyres/0AC586DE-CA1F-48BA-8DAD-C74F49E4012E/10014/20081024.pdf. Accessed 2009.01.19.

16. Kovacs AH, Sears SF, Saidi AS. Biopsychosocial experiences of adults with congenital heart disease: Review of the literature. *American Heart Journal.* 2005;150(2):193.

17. British Cardiac Society. Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. *Heart.* Sep 2002;88(Suppl 1):i1-14.

18. Webb CL, Jenkins KJ, Karpawich PP, et al. Collaborative care for adults with congenital heart disease. *Circulation.* May 14 2002;105(19):2318-2323.

19. Therrien J, Dore A, Gersony W, et al. CCS Consensus Conference 2001 update: recommendations for the management of adults with congenital heart disease. Part I. *Can J Cardiol.* Sep 2001;17(9):940-959.

20. Therrien J, Gatzoulis M, Graham T, et al. Canadian Cardiovascular Society Consensus Conference 2001 update: Recommendations for the Management of Adults with Congenital Heart Disease--Part II. *Can J Cardiol.* Oct 2001;17(10):1029-1050.

21. Therrien J, Warnes C, Daliento L, et al. Canadian Cardiovascular Society Consensus Conference 2001 update: recommendations for the management of adults with congenital heart disease part III. *Can J Cardiol.* Nov 2001;17(11):1135-1158.

22. Williams RG, Pearson GD, Barst RJ, et al. Report of the National Heart, Lung, and Blood Institute Working Group on research in adult congenital heart disease. *J Am Coll Cardiol.* Feb 21 2006;47(4):701-707. .

23. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J.* Dec 2010;31(23):2915-2957.

24. Chessa M, Arciprete P, Bossone E, et al. A multicentre approach for the management of adults with congenital heart disease. *J Cardiovasc Med.* Sep 2006;7(9):701-705.

25. Berg SK, Hertz PG. Outpatient Nursing Clinic for Congenital Heart Disease Patients. *Journal of Cardiovascular Nursing.* 2007;22(6):488.

26. Kafka H, Johnson MR, Gatzoulis MA. The Team Approach to Pregnancy and Congenital Heart Disease. *Cardiology Clinics.* 2006;24(4):587.

27. Moons P, Meijboom FJ, Baumgartner H, Trindade PT, Huyghe E, Kaemmerer H. Structure and activities of adult congenital heart disease programmes in Europe. *Eur Heart J.* Jun 2010;31(11):1305-1310.

28. Reid GJ, Irvine MJ, McCrindle BW, et al. Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. *Pediatrics.* Mar 2004;113(3 Pt 1):197-205.

29. Moons P, Scholte op Reimer W, De Geest S, et al. Nurse specialists in adult congenital heart disease: the current status in Europe. *Eur J Cardiovasc Nurs.* Mar 2006;5(1):60-67. .

30. Mackie AS, Pilote L, Ionescu-Ittu R, Rahme E, Marelli AJ. Health care resource utilization in adults with congenital heart disease. *Am J Cardiol.* Mar 15 2007;99(6):839-843.

31. Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJ. Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. *Eur Heart J.* Jun 2006;27(11):1324-1330. .

32. Van Deyk K, Moons P, Gewillig M, Budts W. Educational and behavioral issues in transitioning from pediatric cardiology to adult-centered health care. *Nurs Clin North Am.* Dec 2004;39(4):755-768.
33. Knauth A, Verstappen A, Reiss J, Webb GD. Transition and Transfer from Pediatric to Adult Care of the Young Adult with Complex Congenital Heart Disease. *Cardiology Clinics.* 2006;24(4):619.
34. van der Bom T, Zomer AC, Zwinderman AH, Meijboom FJ, Bouma BJ, Mulder BJ. The changing epidemiology of congenital heart disease. *Nat Rev Cardiol.* Jan 2011;8(1):50-60.
35. Dearani JA, Connolly HM, Martinez R, Fontanet H, Webb GD. Caring for adults with congenital cardiac disease: successes and challenges for 2007 and beyond. *Cardiol Young.* Sep 2007;17 Suppl 2:87-96.
36. Moons P, Meijboom FJ. Healthcare provision for adults with congenital heart disease in Europe: a review. *Curr Opin Pediatr.* Oct 2010;22(5):573-578.
37. Meijboom F, Mulder B. Problems in the organization of care for patients with adult congenital heart disease. *Arch Cardiovasc Dis.* Jun-Jul 2010;103(6-7):411-415.
38. Socialstyrelsen. State of the Art - Congenital heart diseases in adults: The Board of Health and Welfare; 2005.
39. Tennant PW, Pearce MS, Bythell M, Rankin J. 20-year survival of children born with congenital anomalies: a population-based study. *Lancet.* Feb 20 2010;375(9715):649-656.
40. Sunnegårdh J. *Barnkardiologi - en översikt*2000.
41. Perloff J. Congenital heart disease and pregnancy. *Clin Cardiol.* Nov 1994;17(11):579-587.
42. Rose V, Gold R, Lindsay G, Allen M. A possible increase in the incidence of congenital heart defects among the offspring of affected parents. *J Am Coll Cardiol.* Aug 1985;6(2):376-382.
43. Harris JA, Francannet C, Pradat P, Robert E. The epidemiology of cardiovascular defects, part 2: a study based on data from three large registries of congenital malformations. *Pediatr Cardiol.* May-Jun 2003;24(3):222-235.

44. Burn J, Brennan P, Little J, et al. Recurrence risks in offspring of adults with major heart defects: results from first cohort of British collaborative study. *Lancet.* Jan 31 1998;351(9099):311-316.

45. Driscoll DJ MV, Gersony WM, Hayes CJ, Keane JF, Kidd L, Pieroni DR, Rings LJ, Wolfe RR, Weidman WH. Occurrence risk for congenital heart defects in relatives of patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. *Circulation.* Feb 1993;87(2 Suppl):I114-120.

46. Whittemore R HJ, Engle MA. Pregnancy and its outcome in women with and without surgical treatment of congenital heart disease. *Am J Cardiol.* Sep 1982;50(3):641-651.

47. Berggren H, Olin C. [Swedish surgery of congenital heart defects. From bold pioneering interventions to world-class surgical results]. *Lakartidningen.* Aug 22-28 2005;102(34):2309-2314.

48. Freedom R, M., Lock J, Bricker J, Timothy. Pediatric cardiology and cardiovascular surgery: 1950-2000. *Circulation.* Nov 14 2000;102(20 Suppl 4):IV58-68.

49. Lundstrom NR, Berggren H, Bjorkhem G, Jogi P, Sunnegardh J. Centralization of pediatric heart surgery in Sweden. *Pediatr Cardiol.* Jul-Aug 2000;21(4):353-357.

50. Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. *Heart.* Apr 2001;85(4):438-443.

51. Cedergren MI, Kallen BA. Obstetric outcome of 6346 pregnancies with infants affected by congenital heart defects. *Eur J Obstet Gynecol Reprod Biol.* Apr 1 2006;125(2):211-216.

52. Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. *J Am Coll Cardiol.* Apr 2001;37(5):1170-1175.

53. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. *Circulation.* Jan 16 2007;115(2):163-172.

54. van der Bom T, Luijendijk P, Bouma BJ, Koolbergen DR, de Groot JR, Mulder BB. Treatment of congenital heart disease: risk-reducing measures in young adults. *Future Cardiol.* Mar 2011;7(2):227-240.

55. Socialstyrelsen. Swedish registry of congenital heart disease (SWEDCON) The Board of Health and Welfare; www.ucr.uu.se/swedcon; 2009.

56. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC). *Eur Heart J.* 2010;27:27.

57. Crafoord. Clinical studies in extracorporeal circulation with a heart-lung machine. *Acta chirurgica Scandinavica.* 1957;112(3-4):220.

58. Guleserian KJ. Adult congenital heart disease: surgical advances and options. *Prog Cardiovasc Dis.* Jan-Feb 2011;53(4):254-264.

59. Warnes CA. The adult with congenital heart disease: born to be bad? *J Am Coll Cardiol.* Jul 5 2005;46(1):1-8.

60. Meadows J, Landzberg MJ. Advances in transcatheter interventions in adults with congenital heart disease. *Prog Cardiovasc Dis.* Jan-Feb 2011;53(4):265-273.

61. Meberg A, Otterstad JE, Froland G, Lindberg H, Sorland SJ. Outcome of congenital heart defects--a population-based study. *Acta Paediatr.* Nov 2000;89(11):1344-1351.

62. AHA medical/scientific statement. 1994 revisions to classification of functional capacity and objective assessment of patients with diseases of the heart. *Circulation.* Jul 1994;90(1):644-645.

63. Verheugt CL, Uiterwaal CS, van der Velde ET, et al. Mortality in adult congenital heart disease. *Eur Heart J.* May 2010;31(10):1220-1229.

64. Murdoch DR, Corey GR, Hoen B, et al. Clinical presentation, etiology, and outcome of infective endocarditis in the 21st century: the International Collaboration on Endocarditis-Prospective Cohort Study. *Arch Intern Med.* Mar 9 2009;169(5):463-473.

65. Walsh EP, Cecchin F. Arrhythmias in adult patients with congenital heart disease. *Circulation.* Jan 30 2007;115(4):534-545.

66. Abadir S, Khairy P. Electrophysiology and adult congenital heart disease: advances and options. *Prog Cardiovasc Dis.* Jan-Feb 2011;53(4):281-292.

67. Hilderson D, Saidi AS, Van Deyk K, et al. Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. *Pediatr Cardiol.* Aug 2009;30(6):786-793.

68. Jalkut MK, Allen PJ. Transition from pediatric to adult health care for adolescents with congenital heart disease: a review of the literature and clinical implications. *Pediatr Nurs.* Nov-Dec 2009;35(6):381-387.

69. Moons P, Pinxten S, Dedroog D, et al. Expectations and experiences of adolescents with congenital heart disease on being transferred from pediatric cardiology to an adult congenital heart disease program. *J Adolesc Health.* Apr 2009;44(4):316-322.

70. Lorig KR, Holman HR. Self-Management Education: History, Definition, Outcomes, and Mechanisms. *Annals of Behavioral Medicine.* 2003;26(1):1-7.

71. Glanz K, Rimer BK. Theory at a Glance. A guide for Health Promoting Practice. . In: Institute NC, ed. Second ed. Washington, DC: NIH: National Institute of Health, U.S. Department of Health and Human Service. ; 2005: http://www.cancer.gov/PDF/481f5d53-63df-41bc-bfaf-5aa48ee1da4d/TAAG3.pdf. Accessed 2011-01-20.

72. Toohey S. *Designing Courses for Higher Education*. Berkshire, United Kingdom: The Society for Research into higher Education & Open University Press; 1999.

73. Marton F, Booth S. *Learning and Awareness*. New Jersey: Lawerence Erlbaum Associates; 1997

74. Mayer RE. *Introduction to Multimedia Learning. In: The Cambridge Handbook of Multimedia Learning*. New York: Cambridge University Press; 2005.

75. Grunwald T, Corsbie-Massay C. Guidelines for cognitively efficient multimedia learning tools: educational strategies, cognitive load, and interface design. *Acad Med.* Mar 2006;81(3):213-223.

76. Mayer RE. *The Cambridge handbook of multimedia learning*. Cambridge: Cambridge University Press; 2005.

77. Ramsden P. *Learning to teatch in higher education* Second ed. London and New York: RoutledgeFalmer, Taylor & Francis Group; 2003.

78. Sweller J. *Implications of Cognitive Load Theory for Multimedia Learning. In: The Cambridge Handbook of Multimedia Learning.* . New York: Cambridge University Press; 2005.

79. Mayer RE, Griffith E, Jurkowitz IT, Rothman D. Increased interestingness of extraneous details in a multimedia science presentation leads to decreased learning. *J Exp Psychol Appl.* Dec 2008;14(4):329-339.

80. Bass JL, Corwin M, Gozal D, et al. The effect of chronic or intermittent hypoxia on cognition in childhood: a review of the evidence. *Pediatrics.* Sep 2004;114(3):805-816.

81. Houts PS, Doak CC, Doak LG, Loscalzo MJ. The role of pictures in improving health communication: A review of research on attention, comprehension, recall, and adherence. *Patient Education and Counseling.* 2006;61(2):173.

82. Kools M, van de Wiel MW, Ruiter RA, Kok G. Pictures and text in instructions for medical devices: effects on recall and actual performance. *Patient Educ Couns.* Dec 2006;64(1-3):104-111. .

83. Mayer RE. *Cognitive Theory of Multimedia Learning. In: The Cambridge Handbook of Multimedia Learning*. New York: Cambridge University Press; 2005.

84. Mas FG, Plass J, Kane WM, Papenfuss RL. Health education and multimedia learning: educational psychology and health behavior theory (Part 1). *Health Promot Pract.* Jul 2003;4(3):288-292.

85. Mas FG, Plass J, Kane WM, Papenfuss RL. Health education and multimedia learning: connecting theory and practice (Part 2). *Health Promot Pract.* Oct 2003;4(4):464-469.

86. Huang C. Designing high-quality interactive multimedia learning modules. *Computerized Medical Imaging and Graphics.* 2005;29(2-3):223.

87. Murray E, Burns J, See TS, Lai R, Nazareth I. Interactive Health Communication Applications for people with chronic disease. *Cochrane Database Syst Rev.* Oct 19 2005(4):CD004274.

88. Beranova E, Sykes C. A systematic review of computer-based softwares for educating patients with coronary heart disease. *Patient Educ Couns.* Apr 2007;66(1):21-28. .

89. Lee TI, Yeh YT, Liu CT, Chen PL. Development and evaluation of a patient-oriented education system for diabetes management. *Int J Med Inform.* Sep 2007;76(9):655-663.

90. Strömberg A, Dahlström U, Fridlund B. Computer-based education for patients with chronic heart failure. A randomised, controlled, multicentre trial of the effects on knowledge, compliance and quality of life. *Patient Educ Couns.* Dec 2006;64(1-3):128-135. .

91. Bussey-Smith KL, Rossen RD. A systematic review of randomized control trials evaluating the effectiveness of interactive computerized asthma patient education programs. *Ann Allergy Asthma Immunol.* 2007;Jun;98(6):507-516.

92. Socialstyrelsen. GUCH - register för vuxna med medfött hjärtfel (cited 2007 March 29): The Board of Health and Welfare (Socialstyrelsen(In Swedish)http://www.socialstyrelsen.se/Amnesord/halso_sjuk/Kvalitetsregister/cirkulationsorganen/kva049.htm 1998: http://www.socialstyrelsen.se/Amnesord/halso_sjuk/Kvalitetsregister/cirkulationsorganen/kva049.htm. Accessed 29.03.2007.

93. Sjostrom B, Dahlgren LO. Applying phenomenography in nursing research. *J Adv Nurs.* Nov 2002;40(3):339-345.

94. Sjostrom B, Dahlgren LO. Applying phenomenography in nursing research. *J Adv Nurs.* Nov 2002;40(3):339-345.

95. Marton F. Phenomenography--describing conceptions of the world around us. *Instructional Science.* Jul 1981;Vol 10(2):177-200.

96. Dahlgren L, O. Fallsberg, M Phenomenography as a Qualitative Approach in Social Pharmacy Research. *Journal of Social and Administrative Pharmacy.* 1991 1991;8(4).

97. Marton F, Booth S. *Learning and Awareness*. New Jersey: Lawerence Erlbaum Associates; 1997.

98. Åkerlind G. Variation and commonality in phenomenographic research methods. *Higher Education Research & Development.* 2005;24(4):321-334.

99. Deanfield J, Thaulow E, Warnes C, et al. Management of grown up congenital heart disease. *Eur Heart J.* Jun 2003;24(11):1035-1084.

100. Streiner D, L., Norman G, R. *Health measurement scales - a practical guide to their development and use*. Fourth ed: Oxford University Press; 2008.

101. Kline E. *Classical test theory: assumptions, equations, limitations, and item analyses. In: Kline, E. (Ed.), Psychological Testing: A Practical Approach to Design and Evaluation.*: Sage, Newbury Park, pp. 97-99.; 2005.

102. McAlpine M. A Summary of Methods of Item Analysis. 2002. Retrieved 4 february 2011 from http://caacentre.lboro.ac.uk/dldocs/Bp2final.pdf. Accessed 2011-02-04.

103. Shrout PE, Fleiss JL. Intraclass correlations: uses in assessing rater reliability. *Psychol Bull.* Mar 1979;86(2):420-428.
104. McGraw KO, Wong S. Forming inferences about some intraclass correlation coefficients. *Psychological Methods.* Mar 1996;1(1):30-46.
105. Yen M, Lo LH. Examining test-retest reliability: an intra-class correlation approach. *Nurs Res.* Jan-Feb 2002;51(1):59-62.
106. Rönning. H, Nielsen. N. E, Strömberg. A, Thilen. U, Swahn. E. Development and evaluation of a computer-based educational program for adults with congenitally malformed hearts. *Accepted for publication in European Journal of Cardiovascular Nursing.* 2011.
107. Kamphuis M, Vliegen HW, Vogels T, et al. The need for cardiac follow-up in adults with mild congenital cardiac disease. *Cardiol Young.* Oct 2002;12(5):474-478.
108. Kamphuis M, Vogels T, Ottenkamp J, Van Der Wall EE, Verloove-Vanhorick SP, Vliegen HW. Employment in adults with congenital heart disease. *Arch Pediatr Adolesc Med.* Nov 2002;156(11):1143-1148.
109. Moser DK, Dracup K. Psychosocial recovery from a cardiac event: the influence of perceived control. *Heart Lung.* Jul-Aug 1995;24(4):273-280.
110. Moser DK, Riegel B, McKinley S, Doering LV, An K, Sheahan S. Impact of anxiety and perceived control on in-hospital complications after acute myocardial infarction. *Psychosom Med.* Jan 2007;69(1):10-16.
111. Årestedt K, Ågren S, Flemme I, Moser D, Strömberg A. Psychometric properties of the Swedish version of the Control Attitude Scale for patients with heart disease and their family members. *European Journal of Cardiovascular Nursing.* 2010;9:Suppl 1, 125.
112. Bjelland I, Dahl AA, Haug TT, Neckelmann D. The validity of the Hospital Anxiety and Depression Scale. An updated literature review. *J Psychosom Res.* Feb 2002;52(2):69-77.
113. Zigmond AS, Snaith RP. The hospital anxiety and depression scale. *Acta Psychiatr Scand.* Jun 1983;67(6):361-370.
114. Sullivan M, Karlsson J, Sjostrom L, et al. Swedish obese subjects (SOS)--an intervention study of obesity. Baseline evaluation of

health and psychosocial functioning in the first 1743 subjects examined. *Int J Obes Relat Metab Disord.* Sep 1993;17(9):503-512.

115. World Medical Association Declaration of Helsinki. Ethical principles for medical research involving human subjects. *Nurs Ethics.* Jan 2002;9(1):105-109.

116. Caci H, Bayle FJ, Mattei V, Dossios C, Robert P, Boyer P. How does the Hospital and Anxiety and Depression Scale measure anxiety and depression in healthy subjects? *Psychiatry Res.* May 1 2003;118(1):89-99.

117. Glasgow RE, Funnell MM, Bonomi AE, Davis C, Beckham V, Wagner EH. Self-management aspects of the improving chronic illness care breakthrough series: implementation with diabetes and heart failure teams. *Ann Behav Med.* Spring 2002;24(2):80-87.

118. Glasgow RE, Davis CL, Funnell MM, Beck A. Implementing practical interventions to support chronic illness self-management. *Jt Comm J Qual Saf.* Nov 2003;29(11):563-574.

119. WHO. A declaration in the promotion of patient´s rignts in Europe, Copenhagen: WHO; 1994 (cited 2007 March 29): http://www.who.int/genomics/public/eu_declaration1994.pdf; 1994: http://www.who.int/genomics/public/eu_declaration1994.pdf. Accessed 2007 March 29.

120. Eldh AC. *Patient partisipation - What it is and what it is not (dissertation)*. Örebro: Örebro Studies in Caring Sciences II, Örebro University; 2006.

121. Kendall L, Sloper P, Lewin RJ, Parsons JM. The views of young people with congenital cardiac disease on designing the services for their treatment. *Cardiol Young.* Feb 2003;13(1):11-19.

122. McMurray R, Kendall L, Parsons JM, et al. A life less ordinary: growing up and coping with congenital heart disease. *Coronary Health Care.* 2001;5(1):51.

123. Birks Y, Sloper P, Lewin R, Parsons J. Exploring health-related experiences of children and young people with congenital heart disease. *Health Expect.* Mar 2007;10(1):16-29.

124. Makoul G, Arntson P, Schofield T. Health promotion in primary care: physician-patient communication and decision making about prescription medications. *Soc Sci Med.* Nov 1995;41(9):1241-1254.

125. Florin J, Ehrenberg A, Ehnfors M. Patient participation in clinical decision-making in nursing: A comparative study of nurses' and patients' perceptions. *J Clin Nurs.* Dec 2006;15(12):1498-1508.
126. Beauchamp TL, Childress JF. *Principles of biomedical ethics*. New York: Oxford University Press; 2001.
127. Polit DF, Beck CT. *Nursing Research: Principles and Methods*: Lippincot Williams & Wilkins; 2004.
128. Anderson LW, Krathwohl DR. *A taxonomy for learning, teaching and assessing: A revision of Bloom's Taxonomy of educational objectives: Complete edition*. New York: Longman; 2001.
129. Biggs J, Tang C. *Teaching for Quality Learning at University* 3rd ed. Buckingham: SRHE and Open University Press; 2007.
130. Cure O. Evaluation methodology for a medical e-education patient-oriented information system. *Med Inform Internet Med.* Mar 2003;28(1):1-5.
131. Scandurra I, Hägglund M, Koch S. From user needs to system specifications: Multi-disciplinary thematic seminars as a collaborative design method for development of health information systems. *Journal of Biomedical Informatics.* 2008;41(4):557.
132. Fleisher L, Buzaglo J, Collins M, et al. Using health communication best practices to develop a web-based provider-patient communication aid: the CONNECT study. *Patient Educ Couns.* Jun 2008;71(3):378-387. .
133. Cohen J. A power primer. *Psychol Bull.* Jul 1992;112(1):155-159.
134. Moser DK, McKinley S, Riegel B, et al. The impact on anxiety and perceived control of a short one-on-one nursing intervention designed to decrease treatment seeking delay in people with coronary heart disease. *Eur J Cardiovasc Nurs.* Nov 30 2010.
135. McKinley S, Dracup K, Moser DK, et al. The effect of a short one-on-one nursing intervention on knowledge, attitudes and beliefs related to response to acute coronary syndrome in people with coronary heart disease: a randomized controlled trial. *Int J Nurs Stud.* Aug 2009;46(8):1037-1046.
136. Webb G. The long road to better ACHD care. *Congenit Heart Dis.* May-Jun 2010;5(3):198-205.

137. Dearani JA, Connolly HM, Martinez R, Fontanet H, Webb GD. Caring for adults with congenital cardiac disease: successes and challenges for 2007 and beyond. *Cardiol Young.* Sep 2007;17(Suppl 2):87-96.

138. Jacobs JP, Wernovsky G, Elliott MJ. Analysis of outcomes for congenital cardiac disease: can we do better? *Cardiol Young.* Sep 2007;17 Suppl 2:145-158.

139. Beauchesne LM, Therrien J, Alvarez N, et al. Structure and process measures of quality of care in adult congenital heart disease patients: A pan-Canadian study. *Int J Cardiol.* Dec 27 2010.

140. Finniss DG, Kaptchuk TJ, Miller F, Benedetti F. Biological, clinical, and ethical advances of placebo effects. *Lancet.* Feb 20 2010;375(9715):686-695.

141. Verheugt CL, Uiterwaal CS, van der Velde ET, et al. The emerging burden of hospital admissions of adults with congenital heart disease. *Heart.* Jun 2010;96(11):872-878.

APPENDIX

Appendix A: Discrimination ability and internal consistency
Domain: General knowledge

	Domain: GENERAL KNOWLEDGE	N	Item difficulty	Item-total[1] correlation	Item- total[2] correlation	Item[2] discrimination
1	What is the correct name of your congenitally malformed heart?	114	0.5351	0.3334	0.3546	0.68
2	How often do you have to come to the clinic for follow-up of your congenitally malformed heart?	114	0.5789	0.1275	-	
3	How has your congenitally malformed heart been treated to date?	114	0.9298	0.1295	0.1596	0.18
5	Please answer this question even if you are not currently on medication. If you experience side-effects of your medication, is it indicated that you stop taking the medication?	114	0.7281	0.1313	0.1257	0.32
6.1	When do you have to contact your cardiologist? Dizziness?	114	0.2456	0.1937	0.2507	0.46
6.2	Skin rash?	114	0.9649	-0.1119	-	
6.3	Shortage of breath?	114	0.3860	0.3488	0.3921	0.64
6.4	Diarrhoea?	114	0.9825	-0.1532	-	
6.5	Palpitations?	114	0.5000	0.3178	0.3029	0.54
6.6	Chest pain?	114	0.7105	0.3179	0.3531	0.54
6.7	Unknown fewer?	114	0.1579	0.1892	0.2604	0.29
6.8	Reduced fitness?	114	0.1930	0.2601	0.3187	0.43
6.10	Fainting?	114	0.4123	0.3303	0.3721	0.61
6.11	Tiring more quickly?	114	0.3158	0.3485	0.3653	0.54
6.12	Urinating more frequently?	114	0.9298	-0.1859	-	
6.13	Pain on urinating?	114	0.9386	-0.2125	-	
6.14	Swollen feet and legs?	114	0.2719	0.3144	0.3764	0.54

	Domain: GENERAL KNOWLEDGE	N	Item difficulty	Item-total[1] correlation	Item- total[2] correlation	Item[2] discrimination
7	If the cardiologist informs you that everything is alright, does that mean that you do not need further follow-up?	114	0.7982	0.1352	0.1256	0.25
8	Why do you have a congenital cardiac malformation?	114	0.4474	0.3712	0.3415	0.64
9	Is your congenital cardiac malformation hereditary?	114	0.3684	0.3081	0.2761	0.46
10	Did your cardiologist recommend endocarditis prophylaxis, antibiotics when visiting the dentist to extract teeth for example?	114	0.6579	-0.0103	-0.0284	0.11
15	As you have a congenital cardiac malformation, you should take antibiotics immediately if you have a temperature (without consulting a doctor).	114	0.7807	0.2909	0.3100	0.43
16	Should you go for a dental check-up at least once a year, prophylaxis to endocarditis?	114	0.1754	0.0097	0.0324	0.14
20	Smoking is more harmful for someone with a congenitally malformed heart than for someone without such a malformation.	114	0.6491	0.2187	-	
21	Consuming alcohol is more harmful for someonewith a congenitally malformed heartthan for someone without such a malformation.	114	0.2719	0.0365	-	
22	You should avoid all regular physical activity/training because of your congenital cardiac malformation?	114	0.9123	0.1883	0.2033	0.14
23	You should choose an occupation that is not too physically demanding, as you should be careful not to over-exert yourself.	114	0.6140	0.0378	0.0028	0.18
24	Could sexual activity deteriorate your cardiac condition?	114	0.7281	0.2298	0.2159	0.36
25	What is the risk that your children will have a congenital heart malformation?	114	0.1667	0.2402	0.2065	0.32
KR20				0.63	0.68	

[1]. Before item deleted [2]. After item deleted KR20= Kuder Richardson 20

Domain: Medical treatment

	Domain: MEDICAL TREATMENT	N	Item difficulty	Item-total[1] correlation	Item- total[2] correlation	Item[2] discrimination
4a	What is the name of your medication?	37	0.9189	0.4712	0.4712	0.22
4b	What dose does your medication have?	37	0.5946	0.4005	0.4005	0.67
4c	How is your medication administered ? Tablets, injection, mixture or something else?	37	0.9189	0.5340	0.5340	0.33
4d	When do you take your medication? Breakfast, lunch, afternoon, evening, night or some other time?	37	0.9189	0.4096	0.4096	0.22
4e	What is the reason for using this medication as treatment?	37	0.6757	0.4962	0.4962	0.79
4f	What is the effect or function of your medication?	37	0.5946	0.6056	0.6056	1.0
4g	What are the most common major side-effects of your medication?	37	0.1622	0.3515	0.3515	0.68
4h	Is there anything in particular you should think about when taking this medication? Is there anything you should avoid when being treated with this medication?	37	0.1892	0.2194	0.2194	0.56
KR20				0.74	0.74	

[1]. Before item deleted [2]. After item deleted KR20= Kuder Richardson 20

Domain: Endocarditis prophylaxis

	Domain: ENDOCARDITIS PROPHYLAXIS	N	Item difficulty	Item-total[1] correlation	Item- total[2] correlation	Item[2] discrimination
11	What is endocarditis?	75	0.3733	0.7510	0.7510	0.53
12	What is the most characteristic or typical sign of endocarditis?	75	0.1600	0.4480	0.4480	0.53
13	Can you get endocarditis more than once in your lifetime?	75	0.2800	0.6333	0.6333	0.79
14.1	A number of risk factors for endocarditis are listed below. Do you think these factors contribute to the onset of endocarditis? Contaminated needles (drug addicts)?	75	0.3067	0.6737	0.6737	0.74
14.2	Smoking?	75	0.4000	0.8977	0.8977	1.0
14.3	Bacteria from skin infections?	75	0.2667	0.6411	0.6411	0.74
14.4	Tooth abscesses?	75	0.2933	0.7429	0.7429	0.79
14.5	Sexual activity?	75	0.4133	0.8008	0.8008	0.95
14.6	Poor nail and skin care?	75	0.0533	0.2764	0.2764	0.16
14.7	Body piercing and tattooing?	75	0.3333	0.7695	0.7695	0.90
17	You should take endocarditis prophylaxis, (antibiotics) before every visit to the dentist.	75	0.7333	0.2839	0.2839	0.90
18	Bleeding gums need extra attention, prophylaxis to endocarditis.	75	0.2800	0.4159	0.4159	0.53
19	You should clean your teeth at least once a day, prophylaxis to endocarditis.	75	0.4533	0.3227	0.3227	0.74
KR20				0.90	0.90	

[1]. Before item deleted [2]. After item deleted KR20= Kuder Richardson 20

Domain: Contraceptives and pregnancy

	Domain: CONTRACEPTIVES AND PREGNANCY	N	Item difficulty	Item-total[1] correlation	Item- total[2] correlation	Item[2] discrimination
26	Which contraceptives are the most advisable for you to use because of your congenital cardiac malformation?	56	0.5714	0.0854	-	
27	Should you contact your cardiologist if you want to become pregnant?	56	0.4286	0.5245	0.4616	0.86
28	Should you contact your cardiologist when you are pregnant?	56	0.6607	0.4432	0.5751	1.0
29	Do you run any risk for complications because of your congenital cardiac malformation during pregnancy?	56	0.2679	0.2340	0.3128	0.79
KR20				0.53	0.65	

[1]. Before item deleted [2]. After item deleted KR20= Kuder Richardson 20

Appendix B: KnoCoMH – Swedish version

KnoCoMH; Upphovsrätt – tillgänglighet
KnoCoMH är försedd med copyright knuten till Institutionen för Medicin och Hälsa, avdelningen för Omvårdnad, Linköpings Universitet och Kardiologiska kliniken, Universitetssjukhuset i Linköping. Detta är icke vinstdrivande organisationer som garanterar tillgängligheten samtidigt som standardiseringen av innehållet, poängberäkning, skalkonstruktioner och benämning av instrumentet skyddas. Distribution av KnoCoMH samt användarstöd och instruktioner ges genom Helén Rönning. Tillåtelse att använda KnoCoMH ges rutinmässigt efter registrering, kontakt med Helén Rönning via e-post: helen.ronning@liu.se

Tillstånd. Ert projekt erhåller genom registrering tillstånd att använda KnoCoMH. Tillståndet är knutet till ert projekt och inte till någon fysisk person. *Observera att Du inte får göra ändringar i enkäten enligt regler för copyright. Endast auktoriserade formulär som finns att tillgå genom Helén Rönning är tillåtna.*

KnoCoMH. Psykometrisk testning av den svenska versionen är utförd och referensen angiven här är att betrakta som grundreferens.

- Follow-up of adults with congenitally malformed hearts with focus on individualised and computer-based education and psychosocial support. (Dissertation No. 1239) Linköping: Institution for Medical and Health Sciences, Department of Nursing science, Linköping University; 2011

Manualen refereras till som ovan.

KnoCoMH kan inte ändras på något vis för Dina studiesyften. Om KnoCoMH ingår som en del i ett större frågebatteri som skickas hem, är det önskvärd att KnoCoMH inte fylls i hemma, då risken finns att svaren inte speglar individens kunskapsnivå. Detta för att användare av KnoCoMH ska vara tillförsäkrade standardisering av innehåll och poängsättning utan undantag. Endast då kan jämförelser mellan olika studier bli korrekt.

Manual; Dikotomisering och summering av KnoCoMH

Fråga 1: Rättas utifrån journal. Frågan dikotomiseras till 0 eller 1. Om fler svarsalternativ än 1 är rätt (T.ex. har personen 2 diagnoser, både VSD och aortastenos), slås antal rätt ihop. T.ex. om personen svarat rätt på 2 av 3 diagnoser – dikotomiseras detta som rätt=1. Om personen svarat rätt på 1 diagnos av 3, dikotomiseras det som fel = 0. Om personen svarat rätt på 1 diagnos av 2, dikotomiseras detta som rätt=1.

Fråga 2: Frågan dikotomiseras till 0 eller 1. Om fler svarsalternativ än 1 är rätt (T.ex. har personen behandlats med både kirurgi och via kateterteknik), slås antal rätt ihop. T.ex. om personen svarat rätt på 2 av 3 behandlingar – dikotomiseras detta som rätt=1. Om personen svarat rätt på 1 behandling av 3, dikotomiseras det som fel = 0. Om personen svarat rätt på 1 behandling av 2, dikotomiseras detta som rätt=1.

Fråga 3: Varje fråga dikotomiseras för sig till 0 eller 1 (3a, 3b, 3c osv.). Om fler läkemedel än 1, slås antal rätt ihop under respektive fråga. T.ex. om personen svarat rätt på fråga 3a på 2 av 3 läkemedelsnamn – dikotomiseras detta som rätt=1. Om personen svarat rätt på 3a på 1 läkemedelsnamn av 3, dikotomiseras det som fel = 0. Om personen svarat rätt på 1 läkemedelsnamn av 2, dikotomiseras detta som rätt=1.

Fråga 4: Frågan dikotomiseras till 0 eller 1. Endast 1 rätt svarsalternativ finns ***(fet, kursiv text).*** Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Fråga 5: Varje fråga dikotomiseras för sig till 0 eller 1 (5.1, 5.2, 5.3 osv.). Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Fråga 6, 7 och 8: Varje fråga dikotomiseras för sig till 0 eller 1. Endast 1 rätt svarsalternativ finns ***(fet, kursiv text).*** Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Fråga 9*:* Frågan dikotomiseras till 0 eller 1. Rättas utifrån ansvarig läkares rekommendationer. Endast 1 rätt svarsalternativ finns. Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Fråga 10, 11 och 12:
Varje fråga dikotomiseras för sig till 0 eller 1. Endast 1 rätt svarsalternativ finns ***(fet, kursiv text).*** *Rätt dikotomiseras som 1. Fel dikotomiseras som 0.*

Fråga 13*: Varje fråga dikotomiseras för sig till 0 eller 1 (13:1, 13:2, osv.). Rätt dikotomiseras som 1. Fel dikotomiseras som 0.*

Fråga 14*: Frågan dikotomiseras till 0 eller 1. Endast 1 rätt svarsalternativ finns (****fet, kursiv text).*** *Rätt dikotomiseras som 1. Fel dikotomiseras som 0.*

Fråga 15*: Frågan dikotomiseras till 0 eller 1. Rättas utifrån ansvarig läkares rekommendationer. Rätt svar är JA om läkaren har rekommenderat endokarditprofylax eller allmänpreventiva åtgärder. Rätt svar är NEJ om läkaren inte har rekommenderat endokardit profylax eller allmänpreventiva åtgärder.*

Fråga 16, 17 och 18*: Varje fråga dikotomiseras för sig till 0 eller 1. Endast 1 rätt svarsalternativ finns (****fet, kursiv text).*** *Rätt dikotomiseras som 1. Fel dikotomiseras som 0.*

Fråga 19, 20 och 21*: Varje fråga dikotomiseras för sig till 0 eller 1. Rättas utifrån ansvarig läkares rekommendationer. Endast 1 rätt svarsalternativ finns. Rätt dikotomiseras som 1. Fel dikotomiseras som 0.*

Fråga 22: Frågan dikotomiseras till 0 eller 1. Endast 1 rätt svarsalternativ finns (fet, kursiv text). Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Fråga 23, 24, och 25: Varje fråga dikotomiseras för sig till 0 eller 1. Rättas utifrån ansvarig läkares rekommendationer/bedömning. Endast 1 rätt svarsalternativ finns. Rätt dikotomiseras som 1. Fel dikotomiseras som 0.

Summera de dikotomiserade frågorna i domän; Generel kunskap (22 frågor)
1, 2, 4, 5:1, 5:2, 5:3, 5:4, 5:5, 5:6, 5:7, 5:8, 5:10, 6, 7, 8, 9, 14, 15, 19, 20, 21, 22

Summera de dikotomiserade frågorna i domän; Läkemedelsbehandling (8 frågor)
3a, 3b, 3c, 3d, 3e, 3f, 3g, 3h

Summera de dikotomiserade frågorna i domän; Endokardit profylax (13 frågor)
10, 11, 12, 13:1, 13:2, 13:3, 13:4, 13:5, 13:6, 13:7, 16, 17, 18

Summera de dikotomiserade frågorna i domän; Preventivmedel och Graviditet (3 frågor)
23, 24, 25

Kunskapsenkät: KnoCoMH - För vuxna med medfött hjärtfel

Instruktion:
Detta frågeformulär fastställer vilka områden du behöver mer information om. Läs och besvara varje fråga noggrant. Endast ett svar är rätt, om inte annat anges i frågan. Vet du inte svaret på en fråga fyller du i rutan ”Vet ej”. Försök besvara alla frågor.

HJÄRTFEL OCH BEHANDLING

1. Vilket är det korrekta namnet på ditt hjärtfel?
(kryssa i de rätta svaren)

1☐ Kammarseptumdefekt (VSD) = en öppning mellan de två kammarna

2☐ Förmaksseptumdefekt (ASD) = en öppning mellan de två förmaken

3☐ Kongenitalt korrigerad transposition = vänster och höger hjärtkammare har bytt plats.

4☐ Transposition = de stora artärerna har bytt plats

5☐ Ebsteins anomali = tricuspidalisklaffen fäster längre ner i högerkammaren

6☐ Single ventricle = enkammarhjärta, endast en hjärtkammare pumpar blodet

7☐ Aortastenos = en förträngning av den vänstra hjärtkammarens klaff

8☐ Coarctatio aorta = en förträngning av aortan, stora kroppspulsådern

10☐ Fallots tetrad = en öppning mellan hjärtkamrarna och en förträngning av den högra hjärtkammarens klaff, pulmonalisklaffen och högerkammarhypertrofi

11☐ Eisenmenger = pulmonell hypertension, kvarstående förhöjt tryck i lungkretsloppet och bidirektionell shunt (en vändshunt, ett öppet hål (VSD, ASD) mellan förmak eller hjärtkammare som har flöde mellan sig både framåt och bakåt)

12☐ Annat Skriv här:……

9☐ Vet ej

2. Fram till idag, hur har ditt hjärtfel behandlats?
   (du kan kryssa i mer än ett alternativ)
   1 ☐ genom kirurgi = hjärtoperation
   2 ☐ via kateterteknik (t.ex. ballongvidgning eller slutning med paraply)
   3 ☐ pacemaker
   4 ☐ kateterbehandling av hjärtklappningsbesvär
   5 ☐ genom medicinering
   6 ☐ ingen behandling
   7 ☐ Annat: Skriv här:……..
   9 ☐ vet ej

3. Om du tar medicin, använd en ruta för varje medicin på efterföljande sidor och besvara följande frågor.

   A. Vad är namnet på din medicin?
   B. Vilken styrka har din medicin?
   C. Är medicinen i tablettform, sprutform, mixtur eller annat
   D. När tar du din medicin?(frukost, lunch, eftermiddag, kväll, till natten, när som helst)
   E. Vad är orsaken till att du behandlas med denna medicin
   F. Vilken effekt/verkan har din medicin?
   G. Vilka är de vanligaste biverkningarna du kan få av din medicin?
   H. Är det något du bör tänka på, när du behandlas med denna medicin? Är det något du bör undvika då du behandlas med denna medicin?

A. Namn: _____

B. Styrka: _____

C. Form (tablets, inj.)? _____

D. När? _____

E. Orsak? _____

F. Effekt? _____

G. Biverkning? _____

H. Tänka på? _____

A. Namn: _____

B. Styrka: _____

C. Form (tablets, inj.)? _____

D. När? _____

E. Orsak? _____

F. Effekt? _____

G. Biverkning? _____

H. Tänka på? _____

4. Var vänlig svara på denna fråga även om du inte tar medicin för tillfället. Om du upplever biverkningar av en medicin, bör du då sluta ta den medicinen?
   - 0 ☐ ***Nej, jag bör kontakta ansvarig läkare eller sjuksköterska.***
   - 1 ☐ Ja, jag bör sluta ta den medicinen direkt
   - 9 ☐ Vet ej

5. När bör du ta kontakt med din hjärtläkare?
   (du kan kryssa i mer än ett alternativ)
   - 1 ☐ ***Yrsel***
   - 2 ☐ ***Ökad andfåddhet***
   - 3 ☐ ***Hjärtklappning***
   - 4 ☐ ***Bröstsmärta***
   - 5 ☐ ***Oklar feber***
   - 6 ☐ ***Nedsatt kondition***
   - 7 ☐ ***Svimning***
   - 8 ☐ ***Ökad trötthet***
   - 10 ☐ ***Svullna ben och fötter***
   - 11 ☐ Annat: Skriv här:……
   - 9 ☐ Vet ej

6. Om din doktor informerar dig om att allting är bra, innebär det att du kan sluta gå på regelbundna kontroller?
   - 0 ☐ ***Nej***
   - 1 ☐ Ja
   - 9 ☐ Vet ej

ORSAK/ÄRFTLIGHET

7. Varför fick du ett medfött hjärtfel? Vilket av dessa påstående är sant?
   - 1 ☐ ***Det skedde en störning av hjärtats normala utveckling under min tid som foster, vilket ledde till att ett hjärtfel utvecklades.***
   - 2 ☐ Det hände någonting med hjärtat då jag var nyfödd.
   - 9 ☐ Vet ej

8. Är ditt medfödda hjärtfel ärftligt?
   - 0 ☐ ***Nej, mitt medfödda hjärtfel är inte ärftligt.***
   - 1 ☐ Ja, mitt medfödda hjärtfel är ärftligt.
   - 9 ☐ Vet ej

ATT FÖREBYGGA KOMPLIKATIONER

9. Har din hjärtläkare rekommenderat dig endokarditprofylax, antibiotika vid tandläkarbesök, som till exempel då du ska dra ut en tand?
   - 0 ☐ Nej
   - 1 ☐ Ja
   - 9 ☐ Vet ej

10. Vad är endokardit?
   - 1 ☐ En störning i hjärtrytmen
   - 2 ☐ ***En infektion i hjärtats klaffar***
   - 3 ☐ Ett förstorat hjärta
   - 4 ☐ En blockering av hjärtats blodkärl
   - 9 ☐ Vet ej

11. Vilket är det vanligaste symptomet för endokardit?
   - 1 ☐ Ont i magen
   - 2 ☐ ***Feber som varar i mer än 5 dagar***
   - 3 ☐ Förkylning
   - 4 ☐ Trötthet
   - 5 ☐ Illamående
   - 6 ☐ Hosta
   - 7 ☐ Andfåddhet
   - 9 ☐ Vet ej

12. Kan du få endokardit mer än en gång i livet?
   - 0 ☐ Nej
   - 1 ☐ ***Ja***
   - 9 ☐ Vet ej

13. Nedan listas ett antal faktorer. Kryssa i de faktorer du tror kan bidra till att man får endokardit? (du kan kryssa i mer än ett alternativ)
   - 1 ☐ ***Förorenade nålar (drogmissbruk)***
   - 2 ☐ Rökning
   - 3 ☐ ***Bakterier från hudinfektioner***
   - 4 ☐ ***Tandbölder***
   - 5 ☐ Sexuell aktivitet
   - 6 ☐ ***Dåligt skötta naglar och hud***
   - 7 ☐ ***Piercing och tatueringar***
   - 9 ☐ Vet ej

14. Eftersom du har ett medfödd hjärtfel bör du ta antibiotika omedelbart om du har feber (utan att rådgöra med en läkare).
   - 0 ☐ ***Nej***
   - 1 ☐ Ja
   - 9 ☐ Vet ej

15. Bör du gå till en tandläkare för kontroll minst en gång om året för att förebygga endikardit?
   - 0 ☐ Nej
   - 1 ☐ Ja
   - 9 ☐ Vet ej

16. Bör du ta endokarditprofylax (antibiotika) innan varje besök hos tandläkaren.
   - 0 ☐ ***Nej, enbart inför vissa ingrepp i munnen.***
   - 1 ☐ Ja
   - 9 ☐ Vet ej

17. Blödande tandkött behöver uppmärksammas noga för att förebygga endokardit.
   - 0 ☐ Nej
   - 1 ☐ ***Ja***
   - 9 ☐ Vet ej

18. Du bör borsta tänderna minst en gång per dag för att förebygga endokardit.
    0 ☐ Nej
    1 ☐ ***Ja***
    9 ☐ Vet ej

FYSISK AKTIVITET

19. Du bör undvika all regelbunden fysisk aktivitet/träning på grund av ditt hjärtfel?
    0 ☐ Nej
    1 ☐ Ja
    9 ☐ Vet ej

20. Du bör välja ett yrke som inte är alltför fysiskt krävande, eftersom du inte bör överansträng dig på grund av ditt medfödda hjärtfel.
    0 ☐ Nej
    1 ☐ Ja
    9 ☐ Vet ej

SEXUALITET OCH ÄRFTLIGHET

21. Kan sexuell aktivitet försämra ditt hjärtfel?
    0 ☐ Nej
    1 ☐ Ja
    9 ☐ Vet ej

22. Hur stor är risken att dit/dina barn kommer att ha ett medfött hjärtfel?
    1 ☐ Ingen ökad risk
    2 ☐ ***Obetydligt ökad risk***
    3 ☐ Måttligt ökad risk
    4 ☐ Betydligt ökad risk
    9 ☐ Vet ej

PREVENTIVMEDEL OCH GRAVIDITET (fråga 23-25 besvaras endast av dig som är kvinna)

23. Bör du kontakta din hjärtläkare om du vill bli gravid?
    0 ☐ Nej
    1 ☐ Ja
    9 ☐ Vet ej

24. Bör du kontakta din hjärtläkare då du är gravid?
    0 ☐ Nej
    1 ☐ Ja
    9 ☐ Vet ej

25. Har du någon risk för komplikationer på grund av ditt hjärtfel under en graviditet?
    1 ☐ Ingen ökad risk
    2 ☐ Obetydligt ökad risk
    3 ☐ Måttligt ökad risk
    4 ☐ Betydligt ökad risk
    9 ☐ Vet ej

Appendix C: KnoCoMH – English version

KnoCoMH; Copyright – availability
The copyright in KnoCoMH is held by the Division of Nursing Science, Department of Health Sciences at Linköping University and the Department of Cardiology at the University Hospital of Linköping. These are non-profit organisations that at the same time guarantee availability and protect the standardisation of the content, scoring, construction and the name of the instrument. Distribution of KnoCoMH as well as support and instructions are provided by Helén Rönning. Permission to use KnoCoMH is given by routine following registration, please contact Helén Rönning by e-mail: helen.ronning@liu.se

Permission. Following registration you are authorised to use KnoCoMH in your project. Permission is tied to your project rather than a person. *Please note that according to copyright regulations you are not permitted to make any changes to the questionnaire. Only authorised forms available from Helén Rönning are allowed.*

KnoCoMH. Psychometric tests of the Swedish version have been done according to reference:

Follow-up of adults with congenitally malformed hearts with focus on individualised and computer-based education and psychosocial support. (Dissertation No. 1239) Linköping: Institution for Medical and Health Sciences, Department of Nursing science, Linköping University; 2011

The manual also refers to the above dissertation.

KnoCoMH should not be changed to suit the aim of other studies. If KnoCoMH is one of several questionnaires that are being sent to possible study participants, it is desirable that it is not completed at home as there is a risk that the answers will not reflect the individual's level of knowledge. This will ensure KnoCoMH users standardised content and scoring without exceptions. Only then can correct comparisons between studies be made.

Manual; Dichotomising and scoring KnoCoMH

Item 1: Correct amswers should be confirmed through medical files. The question will be dichotomised separately to 1 or 0. If more than 1 answer is correct. If the person in example has 2 diagnoses, both VSD and aortic stenosis, but answer correct diagnoses in 1 of 2, the answers will be summarised and dichotomised to 1. If the person has 2 of 3 correct answers, it will be dichotomised to 1. If the person has 1 of 3 correct answers, it will be dichotomised to 0 = incorrect. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 2: Correct amswers should be confirmed through medical files. The question will be dichotomised separately to 1 or 0. If more than 1 answer is correct (If the person in example has 2 treatments, both surgery and catheterisation, but answerer correct treatment in 1 of 2, the answers will be summarised and dichotomised to 1.If the person has 2 of 3 correct answers, it will be dichotomised to 1. If the person has 1 of 3 correct answers, it will be dichotomised to 0 = incorrect. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 3: Correct amswers should be confirmed through medical files. Every question will be dichotomised separately to 1 or 0 (3a, 3b, 3c etc.). If more than 1 medical treatment, the correct answers will be summarised at every question (3a, 3b, 3c etc.). If the person in example has 2 medical treatments, both betablockers and ACE inhibitors, and answerers correct side-effects in question 3g in 1 of 2 medical treatments, the answers will be summarised and dichotomised to 1. If the person has correct answers in 2 of 3, it will be dichotomised to 1. If the person has correct answers in 1 of 3, it will be dichotomised to 0 = incorrect.Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 4: The question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 5: Every question will be dichotomised separately to 1 or 0 (5:1, 5:2, 5:3 etc.). Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.Correct answers is in **bold text.**

Item 6, 7 and 8: Every question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to 1. Incorrect answer will be dichotomised to 0.

Item 9: The question will be dichotomised separately to 1 or 0. Correct amswers should be confirmed through medical files. Only 1 answer is correct. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 10: The question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 11 and 12: Every question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to 1. Incorrect answer will be dichotomised to 0.

Item 13: Every question will be dichotomised separately to 1 or 0 (13:1, 13:2, etc.). Correct answer is **bold text.** Correct answer will be dichotomised to 1. Incorrect answer will be dichotomised to 0.

Item 14: The question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 15: The question will be dichotomised separately to 1 or 0. Correct amswers should be confirmed through medical files. Correct answer is YES if the physician was given medical advice to use endocarditis prophylaxis or general preventive measures. Correct answer is NO if the physician were not given medical advice to use endocarditis prophylaxis or general preventive measures. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 16, 17 and 18: Every question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to 1. Incorrect answer will be dichotomised to 0.

Item 19, 20 and 21: Every question will be dichotomised separately to 1 or 0. Correct amswers should be confirmed through medical files. Only 1 answer is correct. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 22: The question will be dichotomised separately to 1 or 0. Only 1 answer is correct **(bold text).** Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Item 23, 24 and 25: Every question will be dichotomised separately to 1 or 0. Correct amswers should be confirmed through medical files. Only 1 answer is correct. Correct answer will be dichotomised to1. Incorrect answer will be dichotomised to 0.

Sum up points of the dichotomised items into domain; General knowledge with 22 questions

1	*What is the correct name of your congenitally malformed heart?*
2	How has your congenitally malformed heart been treated to date?
4	Please answer this question even if you are not currently on medication. If you experience side-effects of your medication, is it indicated that you stop taking the medication?
5.1	When do you have to contact your cardiologist? Dizziness
5.2	Shortage of breath
5.3	Palpitations
5.4	Chest pain
5.5	Unknown fewer
5.6	Reduced fitness
5.7	Fainting?
5.8	Tiring more quickly
5.10	Swollen feet and legs
6	If the cardiologist informs you that everything is alright, does that mean that you do not need further follow-up?
7	Why do you have a congenital cardiac malformation?
8	Is your congenital cardiac malformation hereditary?
9	Did your cardiologist recommend endocarditis prophylaxis, antibiotics when visiting the dentist to extract teeth for example?
14	As you have a congenital cardiac malformation, you should take antibiotics immediately if you have a temperature (without consulting a doctor).
15	Should you go for a dental check-up at least once a year, prophylaxis to endocarditis?
19	You should avoid all regular physical activity/training because of your congenital cardiac malformation?
20	You should choose an occupation that is not too physically demanding, as you should be careful not to over-exert yourself.
21	Could sexual activity deteriorate your cardiac condition?
22	What is the risk that your children will have a congenital heart malformation?

Sum up points of the dichotomised items into domain; Medical treatment with 8 questions

3a What is the name of your medication?
3b What dose does your medication have?
3c How is your medication administered? Tablets, injection, mixture or something else?
3d When do you take your medication? Breakfast, lunch, afternoon, evening, night or some other time?
3e What is the reason for using this medication as treatment?
3f What is the effect or function of your medication?
3g What are the most common major side-effects of your medication?
3h Is there anything in particular you should think about when taking this medication? Is there anything you should avoid when being treated with this medication?

Sum up points of the dichotomised items into domain; Endocarditis prophylaxis with 13 items

10 What is endocarditis?
11 What is the most characteristic or typical sign of endocarditis?
12 Can you get endocarditis more than once in your lifetime?
13.1 A number of risk factors for endocarditis are listed below. Do you think these factors contribute to the onset of endocarditis?
Contaminated needles (drug addicts)?
13.2 Smoking?
13.3 Bacteria from skin infections?
13.4 Tooth abscesses?
13.5 Sexual activity?
13.6 Poor nail and skin care?
13.7 Body piercing and tattooing?
16 You should take endocarditis prophylaxis, (antibiotics) before every visit to the dentist.
17 Bleeding gums need extra attention, prophylaxis to endocarditis.
18 You should clean your teeth at least once a day, prophylaxis to endocarditis.

Sum up points of the dichotomised items into domain; Contraceptives and Pregnancy with 3 items

23 Should you contact your cardiologist if you want to become pregnant?
24 Should you contact your cardiologist when you are pregnant?
25 Do you run any risk for complications because of your congenital cardiac malformation during pregnancy?

The Instrument: KnoCoMH - For Adults with congenitally malformed hearts

Instructions:
This questionnaire allows determining the topics on which you need to be given more information about. Read and answer every question carefully. Only one answer is correct, unless stated otherwise in the question. If you do not know the answer, just mark the square "Don't know". Please answer every question.

Heart defect and treatments

1. What is the correct name of your congenitally malformed heart?
(You may enter more than one answer.)

1 ☐ Ventricular septal defect (VSD) = an opening between the two chambers.

2 ☐ Atrial septal defect (ASD) = an opening between the two atrium.

3 ☐ Congenitally corrected transposition of the great arteries = left and right chambers have switched place.

4 ☐ Complete transposition of the great arteries = switched position of the great blood vessels.

5 ☐ Ebstein anomaly = the right chamber's valve position is misplaced down into right chamber.

6 ☐ Single ventricle = "one heart chamber", only one heart chamber pumping out the blood to the body.

7 ☐ Aortic valve stenosis = a narrowing of the aortic valve of the left chamber.

8 ☐ Coarctation of the aortae = a narrowing of the aorta, the greatest artery of the vessels.

10 ☐ Tetralogy of Fallot **=** an opening between the two chambers and a narrowing of the pulmonary valve of the right chamber and hypertrophy of the right chamber.

11 ☐ Eisenmenger syndrome = pulmonary hypertension, persistent high pressure in the circulation of the lung and bidirection of an opening between the two atrium or chambers (ASD or VSD).

12 ☐ Other Write here:……

9 ☐ Don't know

2. How has your congenitally malformed heart been treated to date?
   (You may enter more than one answer.)
   1 ☐ With surgery = heart operation
   2 ☐ With catheterisation (e.g. balloon dilatation or stenting/closing with umbrella)
   3 ☐ With pacemaker
   4 ☐ With catheterisation of arrhythmias, palpitations of the heart
   5 ☐ With medical treatment
   6 ☐ No treatment

   7 ☐ Other Write here:……..

   9 ☐ Don't know

3. If you have medical treatments, use one box for each medication and answer the following questions.
   A. What is the name of your medication?

   B. What dose does your medication have?

   C. How is your medication administered? Tablets, injection, mixture or something else?

   D. When do you take your medication? Breakfast, lunch, afternoon, evening, night or some other time?

   E. What is the reason for using this medication as treatment?

   F. What is the effect or function of your medication?

   G. What are the most common major side-effects of your medication?

   H. Is there anything in particular you should think about when taking this medication? Is there anything you should avoid when being treated with this medication?

A. Name: _____

B. Dose: _____

C. Form (tablets, inj.)? _____

D. When? _____

E. Reason? _____

F. Effects? _____

G. Side-effects? _____

H. To think about? _____

A. Name: _____

B. Dose: _____

C. Form (tablets, inj.)? _____

D. When? _____

E. Reason? _____

F. Effects? _____

G. Side-effects? _____

H. To think about? _____

4. Please answer this question even if you are not currently on medication. If you experience side-effects of your medication, is it indicated that you stop taking the medication?
   - 0 ☐ ***No, I should contact my physician or nurse.***
   - 1 ☐ Yes, I should stop taken the medication directly.
   - 9 ☐ Don't know

5. When do you have to contact your cardiologist?
   (You may enter more than one answer.)
   - 1 ☐ ***Dizziness***
   - 2 ☐ ***Shortage of breath***
   - 3 ☐ ***Palpitations***
   - 4 ☐ ***Chest pain***
   - 5 ☐ ***Unknown fewer***
   - 6 ☐ ***Reduced fitness***
   - 7 ☐ ***Fainting***
   - 8 ☐ ***Tiring more quickly***
   - 10 ☐ ***Swollen feet and legs***
   - 11 ☐ Other Write here……
   - 9 ☐ Don't know

6. If the cardiologist informs you that everything is alright, does that mean that you do not need further follow-up?
   - 0 ☐ ***No***
   - 1 ☐ Yes
   - 9 ☐ Don't know

REASON/HEREDITY

7. Why do you have a congenital heart malformation? Which of those statements is true?
   - 1 ☐ ***There was an disturbance during the development of the heart during my time as a foetus, and a heart defect was developed.***
   - 2 ☐ Something happened with the heart when I was newborn.
   - 9 ☐ Don't know

8. Is your congenital cardiac malformation hereditary?
   - 0 ☐ ***No, my heart defect is not hereditary.***
   - 1 ☐ Yes, my heart defect is hereditary.
   - 9 ☐ Don't know

TO PREVENT COMPLICATIONS

9. Did your cardiologist recommend endocarditis prophylaxis, antibiotics when visiting the dentist to extract teeth for example?
   - 0 ☐ No
   - 1 ☐ Yes
   - 9 ☐ Don't know

10. What is endocarditis?
   - 1 ☐ A disruption of the heart rhythm
   - 2 ☐ ***An infection involving the heart valves***
   - 3 ☐ An enlarged heart
   - 4 ☐ A blockage of the blood vessels of the heart
   - 9 ☐ Don't know

11. What is the most characteristic or typical sign of endocarditis?
   - 1 ☐ Pain in the stomach
   - 2 ☐ ***Fever more than 5 days***
   - 3 ☐ Cold
   - 4 ☐ Feel tired
   - 5 ☐ Feel sick
   - 6 ☐ Cough
   - 7 ☐ Breathlessness
   - 9 ☐ Don't know

12. Can you get endocarditis more than once in your lifetime?
   - 0 ☐ No
   - 1 ☐ ***Yes***
   - 9 ☐ Don't know

13. A number of risk factors for endocarditis are listed below. Do you think these factors contribute to the onset of endocarditis? (You may enter more than one answer.)
   - 1 ☐ ***Contaminated needles (drug addicts)***
   - 2 ☐ Smoking
   - 3 ☐ ***Bacteria from skin infections***
   - 4 ☐ ***Tooth abscesses***
   - 5 ☐ Sexual activity
   - 6 ☐ ***Poor nail and skin care***
   - 7 ☐ ***Body piercing and tattooing***
   - 9 ☐ Don't know

14. As you have a congenital cardiac malformation, you should take antibiotics immediately if you have a temperature
    (without consulting a doctor).
   - 0 ☐ ***No***
   - 1 ☐ Yes
   - 9 ☐ Don't know

15. Should you go for a dental check-up at least once a year, prophylaxis to endocarditis?
   - 0 ☐ No
   - 1 ☐ Yes
   - 9 ☐ Don't know

16. You should take endocarditis prophylaxis, (antibiotics) before every visit to the dentist.
   - 0 ☐ ***No, only before special treatments in the mouth***.
   - 1 ☐ Yes
   - 9 ☐ Don't know

17. Bleeding gums need extra attention, prophylaxis to endocarditis.
   - 0 ☐ No
   - 1 ☐ ***Yes***
   - 9 ☐ Don't know

18. You should clean your teeth at least once a day, prophylaxis to endocarditis.
    0 ☐ No
    1 ☐ ***Yes***
    9 ☐ Don't know

PHYSICAL ACTIVITY

19. You should avoid all regular physical activity/training because of your congenital cardiac malformation?
    0 ☐ No
    1 ☐ Yes
    9 ☐ Don't know

20. You should choose an occupation that is not too physically demanding, as you should be careful not to over-exert yourself.
    0 ☐ No
    1 ☐ Yes
    9 ☐ Don't know

SEXUALITY AND HEREDITY

21. Could sexual activity deteriorate your cardiac condition?
    0 ☐ No
    1 ☐ Yes
    9 ☐ Don't know

22. What is the risk that your children will have a congenitally malformed heart?
    1 ☐ No risk at all
    2 ☐ ***Slightly increased risk.***
    3 ☐ Moderate increased risk
    4 ☐ Considerably increased risk
    9 ☐ Don't know

CONTRACEPTIVES AND PREGNANCY (Question 23-25 should only be answered by you being a woman)

23. Should you contact your cardiologist if you <u>want to become</u> pregnant?
    0 ☐ No
    1 ☐ Yes
    9 ☐ Don't know

24. Should you contact your cardiologist when <u>you are</u> pregnant?
    0 ☐ Nej
    1 ☐ Yes
    9 ☐ Don't know

25. Do you run any risk for complications because of your congenital cardiac malformation during pregnancy?
    1 ☐ No risk at all
    2 ☐ Slightly increased risk
    3 ☐ Moderate increased risk
    4 ☐ Considerably increased risk
    9 ☐ Don't know