

**Living with a congenital heart disease:
Adolescents' and young adults' experiences**

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To my "heart sisters" - I did it for you!

ABSTRACT

Adolescents and young adults with a congenital heart disease (CHD) belong to a new and in several respects unexplored population within health research. They represent a diversity of heart defects requiring different surgical procedures and interventions that create different physiological and anatomical structures, with various impacts on life. Extensive improvements in congenital cardiology and heart surgery have created new groups of children surviving into adolescence and adulthood. One of these new groups comprises adolescents and young adults living with a surgically palliated univentricular heart. The overall aim in this thesis was to illuminate how adolescents and young adults with a CHD experience life and their life situation. Furthermore, the thesis aims to describe the impact of a CHD on health perception, sense of coherence, quality of life and satisfaction with life in adolescents and young adults with a CHD or, in particular, a surgically palliated univentricular heart.

A multi-method approach was used, consisting of in-depth interviews and surveys. The first two studies involved adults with various kinds of CHD. Study I involved six in-depth interviews, analysed with the phenomenological-hermeneutical method. Study II consisted of statistical analysis of EQ-5D questionnaire data from 1435 adults included in the GUCH registry. The last two studies involved adolescents and young adults living with a surgically palliated univentricular heart. Seven in-depth interviews in Study III were complemented by a survey in Study IV using the Sense of Coherence scale and the modified study-specific Essence of Existence questionnaire. The 33 responses were analysed with a combination of content analysis and descriptive statistics.

The results show that adolescents and adults with a CHD in most cases perceive their health to be good, and the same as their peers. Pain or discomfort and anxiety or depression were found to be prevalent health problems, but adults with a CHD reported less pain or discomfort than a general population. It was further shown that symptoms may occur even if the adult with a CHD reports himself/herself to be asymptomatic. Despite limitations in everyday life, adolescents and young adults with a surgically palliated univentricular heart experience satisfaction with their lives and see themselves as exceptional, strong and healthy. This indicates that there is no direct association between the severity of the heart defect and the experience of satisfaction with life. The findings suggest that the existential maturity they had developed, along with their experienced happiness over "being me", may promote a stronger sense of coherence.

The conclusion from this thesis was that, through a process of adaptation, the CHD becomes integrated and a normal part of life. However, adolescents and young adults could at the same time be seen as "walking a fine line" when balancing the aspects of health and disease that co-exist in the life of a person with a CHD. Thus, the concept of "health within disease" (as opposed to "health within illness") emerges to describe this population, since they experience themselves most of the time as healthy.

Key words: congenital heart disease, health, hermeneutics, interviews, quality of life, sense of coherence

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LIST OF PAPERS

This thesis is based on the following papers, referred to in the text by their Roman numerals I-IV.

- I Berghammer M., Dellborg M., Ekman I. Young adults experiences of living with congenital heart disease.
International Journal of Cardiology 2006; 110:340-347.

- II Berghammer M., Karlsson J., Ekman I., Eriksson P., Dellborg M. Self-reported health status (EQ-5D) in adults with congenital heart disease.
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- III Berghammer M., Brink E., Rydberg A., Dellborg M., Ekman I. Committed to life - adolescents and young adults experiences from living with Fontan circulation.
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- IV Berghammer M., Rydberg A., Ekman I., Hanseus K., Karlsson J. Sense of coherence, health perception and satisfaction with life in adolescents' and young adults' living with Fontan circulation.
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ABBREVIATIONS

CHD	Congenital heart disease
EQ-5D	EuroQol five-dimension questionnaire
EQ-VAS	EuroQol visual analog scale
GUCH	Grown-up congenital heart disease
HRQoL	Health-related quality of life
NYHA	New York Heart Association classification system
QoL	Quality of life
SOC	Sense of coherence
TCPC	Total cavo-pulmonary connection

DEFINITIONS

GUCH/ACHD	GUCH is the abbreviation of grown-up congenital heart disease. In Europe this term is more commonly used for adult persons with a congenital heart disease. In USA the abbreviation ACHD (adult congenital heart disease) is more common.
Univentricular heart	Univentricular heart is a comprehensive term of different complex heart defects where the surgical end result leads to a pulmonary circulation not passing a venous or subpulmonary ventricle.



INTRODUCTION

Adolescents and young adults with a congenital heart disease (CHD) belong to a new and in several respects unexplored population within health research; they constitute a new field in the health care system (Warnes, 2005; Warnes, Williams, Bashore et al., 2008b). Only fifty years ago, infant mortality in untreated CHD was 60–70% (Somerville, 1998) but since then there have been extensive achievements in cardiovascular medicine (Marelli & Gurvitz, 2011). The achievements obtained during recent decades include advances in diagnosis, medical management, surgical interventions and nursing care, which have resulted in decreased mortality and improved survival for children with a CHD. Almost 95% of the children whose CHD is either surgically repaired or palliated nowadays reach adulthood (Moons, Bovijn, Budts et al., 2010). Even children born with very complicated heart defects now have the prospect of an adult life (Brickner, Hillis, & Lange, 2000b; Warnes, 2005). Thus, a completely new spectrum of patients is now reaching adulthood.

Even if the initial heart defect is usually surgically repaired or palliated in early childhood, there may be a need for long-term follow-up (Warnes, et al., 2008b). Not all consequences and functional aspects of a repaired or palliated heart 15, 20 or 25 years after the intervention are fully established as yet. A European follow-up survey of a cohort of adults with a CHD reported a low mortality rate during the five years of the survey, but a large proportion of the adults in the study experienced substantial morbidity (Engelfriet, Boersma, Oechslin et al., 2005). It would appear that health-related problems worsen over time which indicates the importance of long-term follow-up in order to maintain health and avoid lapses in care (Yeung, Kay, Roosevelt et al., 2008).

That a CHD affects many aspects of life has been shown both in clinical practice and earlier research. Such aspects as health, quality of life (QoL), health-related QoL (HRQoL) and psychosocial concerns have been shown to be of importance. The heart defect is deeply connected with identity and a negative impact on sense of self, body image and view of the future because of the heart defect have been reported (Kovacs, Sears, & Saidi, 2005; Tong, Sparacino, Messias et al., 1998). In contrast are the findings of a positive impact of the heart defect: the association with feelings of being healthy (Ternstedt Östman, 1989) and even that a better QoL is experienced compared to a healthy age- and gender-matched population (Moons, Van Deyk, De Bleser et al., 2006). The population of adolescents and adults is largely comprised of individuals living mainly without consequences from their CHD, to individuals with severe consequences and limitations in daily life (Gatzoulis, Webb, & Daubeney, 2010; Moons, et al., 2010).

Knowledge about how life is experienced from an insider's perspective is a prerequisite for individualized care with a person-centred focus (Ekman, Swedberg, Taft et al., 2011; Ekman, Wolf, Olsson et al., 2012). Since most children with a CHD are now able to reach adulthood there is a need for lifetime management. The chosen care interventions should be tailored in order to support and strengthen the individual's capacity throughout the whole life span, focusing on seeing the possibilities and promoting health. When providing person-centred care, including the needs of the whole person, it is essential to understand the impact of the heart defect on daily life and to establish which aspect are of greatest concern for adolescents and young adults with a CHD. The main intention of this thesis is therefore to investigate what it means to live with a CHD, in particular a surgically palliated univentricular heart, with a focus on the individual's health and overall life situation.

BACKGROUND

Congenital heart disease

Congenital heart disease (CHD) is a comprehensive term encompassing over a hundred different heart malformations, and the incidence of a CHD in live-born children is 0.8%, almost one per 100 (Gatzoulis, et al., 2010). A CHD is the most common malformation of all congenital malformations (Dolk, Loane, Garne et al., 2011) and in Sweden approximately 800–1000 children are born every year with some kind of CHD (SWEDCON, 2011). Over the past several decades, advances within cardiovascular medicine have significantly lowered mortality rates for children and adolescents with a complex CHD (Marelli, Mackie, Ionescu-Ittu et al., 2007; Warnes, Liberthson, Danielson Jr et al., 2001). Survival rates into adulthood for children with a surgically repaired CHD have previously been estimated to be above 85% (Perloff, 1991) and were shown to be over 90% in a recent analysis by Moons and co-workers (2010).

The different malformations have a variable anatomy with highly varying impact on the circulatory system. They can be classified as minor, significant and complex defects with a wide range in severity: at one extreme there are heart malformations that can self-heal, while at the other extreme are complex malformations that require surgical repair. The complex heart malformations frequently require early and sometimes acute surgical or catheter-based correction (Wren, Richmond, & Donaldson, 2000). Often the surgical repair is needed during the child's first year and, thereafter, surgical correction is performed several times throughout life in order to maintain functional circulation. It is well appreciated that most adults who have their lives transformed by surgical interventions received reparative and not curative surgery (Gatzoulis, et al., 2010).

To monitor and detect potential medical complications and assess the need for further interventions, it is important to have regular follow-up of children living with a CHD (Gatzoulis, et al., 2010; Warnes, 2005; Warnes, et al., 2001). In the future, many adults will present different haemodynamic and cardiac problems from those currently seen, due to the impact of new and innovative surgical procedures (Warnes, et al., 2001). North American and European guidelines have recommended ongoing cardiac surveillance of adults with a CHD (Deanfield, Thaulow, Warnes et al., 2003; Warnes, Williams, Bashore et al., 2008a). The follow-up is dependent on the complexity of the heart defect and of the interventions being performed, indicating that there is a greater need for regular follow-up for those children with a more complex heart defect. However, the following up of those who have a less complex defect is also beneficial, given the general risk of secondary morbidity in adults with a CHD (Engelfriet, et al., 2005; Warnes, 2005).

Univentricular heart

Univentricular heart is a comprehensive term encompassing various complex heart defects for which the common surgical end result is pulmonary circulation that does not pass a pumping ventricle. Previously inoperable groups include children born with highly complex malformations such as pulmonary atresia or hypoplastic left heart syndrome, where either the right or left ventricle is underdeveloped and a normal physiology with two ventricles cannot be restored (Warnes, 2005). From the 1980s to the 1990s, these children could be saved by the construction of a univentricular circulation, which leads the unsaturated, returning venous blood directly to the lungs. This condition is unique in having the most complicated circulation and completely different haemodynamics (Rydberg, Teien, Karp et al., 1998).

The pioneering Fontan procedure, first described by Fontan and Baudet (1971) has led to an increased survival into adulthood for children born with a univentricular heart or single ventricle physiology (van den Bosch, Roos-Hesselink, van Domburg et al., 2004; de Leval & Deanfield, 2010). A Fontan circulation is created through multiple surgical palliations, in which an adequate blood flow and a near normal venous pressure are achieved despite the absence of the pumping action of the right (subpulmonary) ventricle (Fontan & Baudet, 1971). The Fontan physiology is based on the principle that the systemic venous blood return is passed directly to the pulmonary circulation (de Leval & Deanfield, 2010; Pike, Evangelista, Doering et al., 2007). The multiple surgical procedures are performed at various developmental stages early in life, gradually creating the changed circulation and the haemodynamics (Müller, Christov, Schreiber et al., 2009). Since the Fontan procedure was introduced, several modifications have improved the surgical technique, the most recent modification being a total cavopulmonary connection (TCPC) (van den Bosch, et al., 2004; de Leval & Deanfield, 2010; Rydberg, et al., 1998). The Fontan procedure has provided a satisfactory surgical palliation and an improved life expectancy; adolescents and young adults now living with Fontan circulation belong to the first generation of patients with this condition surviving into adulthood (van den Bosch, et al., 2004; de Leval & Deanfield, 2010). The term univentricular heart will be used in this thesis, regardless of the underlying anatomical defect and of the surgical procedure performed (TCPC or classic Fontan).

Growing up with a congenital heart disease

Many children with minor forms of CHD have a normal life expectancy and a lifestyle with few, if any, limitations. For children living with a moderately severe CHD, the long-term outlook varies tremendously, depending on the outcome of the previous surgical interventions and the haemodynamic situation. Those with more complex forms of CHD require regular follow-up, may have limited exercise capacity and are sometimes unable to maintain full-time employment. For

example, individuals with a univentricular heart are usually cyanosed, they have to undergo several multiple surgeries and may still be future candidates for cardiac transplantation (Wren, et al., 2000).

A complex CHD is nowadays increasingly diagnosed before the child is born, mainly through prenatal echocardiography. If it is not detected then, a complex CHD is usually noticed within the first week after birth, presenting with symptoms of cyanosis, heart failure or even shock. It can also be detected during post-natal examination as a heart murmur, weak femoral pulse or low saturation; the clinical manifestations depending on the defect. In complex CHDs, the main treatment is surgical, in which the heart defect is either palliated or corrected, aiming to create as well-functioning circulation as possible. The surgical interventions are performed at different ages, depending on the severity and complexity of the heart defect (Gatzoulis, 2010). Before their operation many of these children present difficulties managing daily life, lacking enough energy to eat, grow and play. They often present symptoms associated with the heart defect, such as shortness of breath, tiredness or cyanosis.

In the scientific statement by Marino and fellow researchers (2012), it was declared that children with a CHD have an increased risk for developmental disorder, disabilities or developmental delay. For many of the children with a CHD, in particular those with a complex CHD, life is associated right from the start with several hospital stays, frequent medical check-ups with painful examinations and, for many, multiple surgical interventions (Birkeland, Rydberg, & Hägglöf, 2005). The many interruptions to normal life influence the whole family and, during recent years, there has been an increased interest in evaluating the emotional, psychological and behavioural aspects and outcomes associated with growing up with a CHD.

A CHD in itself could also be seen as a stressor, both for the child as well as for the parents, because it is connected with life-challenging events early in the child's life, for which the outcome is difficult to predict at first. Studies have shown that parents of a child with a CHD experience feelings of distress – anxiety, hopelessness and somatic complaints – to a greater extent than parents of healthy children or parents of children with other diseases (Lawoko & Soares, 2002). In a comparison of children's perceptions with those of their parents, Uzark and co-workers (2008) found significant differences between the parents' and the child's perception, as well as between children with a CHD and healthy children, and also within the group of children with a CHD. Compared with healthy children, an overall lower QoL was reported across all age groups of children with a CHD, who registered especially low scores in the domains of physical and psychosocial functioning. That the perception differs between the child and parent has also been demonstrated (Fredriksen, Diseth, & Thaulow, 2009). They found that children with a CHD reported a higher incidence of behavioural and emotional problems compared to parental reports, indicating that the children experienced more

problems than the parents were aware of. However, it was also found that the behaviour problem scores in children with cardiac disease were significantly lower than in healthy children. Even if children with a CHD, unlike their peers, have to go through many life-challenging events and report behavioural and emotional problems, studies show that they also regard themselves as just like their healthy peers (Shearer, Rempel, Norris et al., 2012). Strategies for mastering the CHD have been reported and the children learn from an early age to either situate their CHD in the foreground or background according to their needs.

Growing up with a CHD may present a psychological burden for the child, with the early awareness of the risk of a shortened life. The uncertainty can lead to deep existential concerns, for which the child – and the whole family – may need intensive psychosocial support at certain periods (Birkeland, et al., 2005; Kovacs, et al., 2005). Studies have shown that these individuals face many psychosocial challenges during adolescence and young adulthood, for example feelings of being different from others and the many concerns regarding their body image, such as how to deal with their scar (Claessens, Moons, de Casterle et al., 2005; Horner, Liberthson, & Jellinek, 2000; Tong, et al., 1998). Tong and co-workers (1998) found that adolescents with a CHD experienced many dilemmas during this period. These concern how to manage the CHD, for example, the dilemma of whether to see oneself as normal or not, whether to disclose the CHD or not to others, social interaction versus social isolation and the challenge of uncertainty. Horner and co-workers (2000) argued that early adulthood is a more difficult period than adolescence, both in terms of physical health, relationships, work and their uncertain future. Kovacs and co-authors (2005) have focused on biopsychosocial experiences in adults with a CHD, because having a CHD may be associated with an increased risk of psychological difficulties. Areas of functioning that they claimed might be particularly affected include neurocognitive functioning, body image, social and peer relationships, and mild delays in developmental functioning. Therefore, they concentrated on three domains: psychological factors, social factors and QoL factors. Possible psychological factors include neurocognitive deficits occurring after surgery; social factors concern a delayed progression into full adulthood due to overprotection or impaired peer relationships and the last factor, health-related QoL, reflects the impact of physical illness on overall well-being.

Rijen and her co-workers (2003) evaluated the psychological function of adults with a CHD via interviews in addition to a questionnaire and found that adults with a CHD compared favourably with the general population. Many studies concerning adults with a CHD report a similarly positive picture. Even when many stressors are experienced during life, studies have also shown that adolescents and young adults with a CHD perceive their lives as normal and themselves as healthy (Ternstedt, Wall, Oddsson et al., 2001), regard themselves as strong and resistive (Zahmacioglu, Yildiz, Koca et al., 2011) and mainly consider their QoL to be good (Moons, Van Deyk, Marquet et al., 2009; Ternstedt Östman,

1989). Nevertheless, it has been stated that, along with the medical treatment, there is a need to focus on psychosocial consequences within this population, which would benefit from structured psychosocial programs (Birkeland, et al., 2005; Kovacs, Saidi, Kuhl et al., 2009; Uzark, et al., 2008).

Grown-up congenital heart disease

Grown-ups with congenital heart disease (GUCH) constitute a new and growing population (Moons, et al., 2010; Perloff, 1991; Thilen, 2005). Marelli and co-workers (2007) showed that the prevalence rates of severe CHD increased between 1985 and 2000, with a significantly higher increase in adults. The number of adults will continue to grow given that more and more children will become adults, and the adult population is very likely to exceed the number of children with a CHD (Brickner, Hillis, & Lange, 2000a; Brickner, et al., 2000b; Gatzoulis, et al., 2010). This implies that the profile of this patient population will change, not only because of advancing age, but also with improved survival of patients with complex anomalies. In addition, with the impetus to perform lasting repair at an earlier age and with changing surgical procedures, there will be changes in the anticipated disease patterns (Warnes, et al., 2001). In Marelli and co-workers Canadian population (2007), in the year 2000 the overall number of adults exceeded the number of children with CHD. The number of adults and children with a severe CHD were nearly equal. As a result of these observed demographical changes over the last two decades, the mean age for the total population living with a CHD has increased, most notably among those with a severe CHD. The median age of the Canadian adult CHD population was 40 years in the year 2000 and 29 years for the subset of adults with a severe CHD. From national calculations, there are approximately 30 000 adults living with a CHD in Sweden (Marelli, et al., 2007; SWEDCON, 2011) and these young adults have a variety of different heart defects. Many adolescents and young adults with a CHD face the prospect of future operations or other long-term complications, for example arrhythmias (Engelfriet, et al., 2005; Gatzoulis, et al., 2010).

To generalize and describe the typical person who has grown up with CHD is difficult because the whole population encompasses a wide variety of initial heart defects and of palliative or corrective surgical interventions performed (Gatzoulis, et al., 2010). The various heart defects may represent a difference in impact on life but, regardless of the anatomical structure and the complexity of the defect, the existential aspects of living with a CHD are likely to cover universal feelings and experiences that override the differences. Some of these feelings concern the aspects connected with longevity and the uncertainty of one's future. Despite the existential aspects of living, it is shown that these individuals in many cases are able to live a relatively normal life in comparison with their peers, gaining employment and considering themselves to have a good life with a good perceived health and QoL (Moons, Van Deyk, et al., 2006; van Rijen, et al., 2003).

PERSPECTIVES AND VIEWPOINTS

Adolescents and adults with a CHD have a long history of health care and of being patients through many episodes of hospital stays, when either surgical procedures, examinations of different kinds or routine check-ups have been performed throughout their lives. Today the word *patient* has often been given a medical contextual meaning of someone who is sick, has a disease or is under treatment, even if the word was not originally connected with disease or being sick. The origin of the word patient comes from the Latin word *pati*, meaning suffer or endure, but was not connected with disease. Instead the focus was on the person who was suffering and had to endure it (Eriksson, 2006).

This thesis is based on the assumption that adolescents or adults with a CHD, even those with a surgically palliated univentricular heart, are seen as people or subjects with capabilities, who create meaning and knowledge from their life experiences. Given that the health-care context is not the focus of the thesis, the participating adolescents and young adults will be referred to here as adolescents, young adults or adults, rather than as patients.

Disease and illness

In order to understand these experiences it is of interest to set them in the context of existing definitions of disease and illness. The medical anthropologist Eisenberg (1977) stated that disease is a deviation of the anatomical structure of an organ, whereas illness is the experience of a change in the person's own condition and social function. Kleinman (1995) also makes a distinction between disease and illness, in which disease is defined as a dysfunction of a biological or physiological process, and illness incorporates the psychosocial experience and the meaning of experienced symptoms and disability (if present). Disease and illness do not stand in a one-to-one relationship to each other, since disease may occur in the absence of illness, meaning that an individual might have a disease without knowing it or without having symptoms; equally, an individual might experience illness and symptoms without having a disease. Illness is defined as a dynamic process in which the "sick" person is part of a social network living and responding to symptoms and disability. Adolescents and adults who have grown up with a CHD are aware of their CHD from the start. If heart-related symptoms then occur, even if the symptoms are a new experience, these individuals are already aware of their CHD. Within this context, it is logical to apply the definition of disease as a deviation of the anatomical structure or of an organ. However, a CHD is not necessarily experienced as an illness, because it depends on other factors and concerns the subjective and psychological dimensions of life. In this thesis, a CHD is thus considered and referred to as a long-term disease. This reflects the fact that, even if the CHD is surgically repaired or palliated during childhood, people with a CHD still have to take account of their heart defect on some level throughout life.

The same separation of disease as a biological dysfunction and illness as a subjective and emotional experience, was made by Twaddle and Hessler (1987). They also introduced a third dimension: sickness (or *unhealth*), which they defined as the social phenomenon of being unhealthy, seen as an identity that is given to the individual by others on the basis of failure in performing expected activities. Having a CHD or a univentricular heart does not have to include the state of unhealth according to their definition; nor are subjective feelings of being unhealthy a necessary feature of the disease.

The impact of a CHD is fundamentally connected with its consequences for daily life, and how it is integrated into life could be seen as an ongoing process, contributing to the subjective meaning of living with a CHD. This indicates that, through this process, different people living with a disease (the CHD) may or may not perceive it as an illness and, within one individual, this meaning can change over the course of a lifetime.

This is line with the reasoning by Toombs (1992) regarding the meaning of illness. Toombs focused particularly on the illness aspect and what the illness means for the specific person. She argued that a long-term illness not only involves the current context, but also considerations about the future, for example, a potential deterioration in the person's condition. The lived experience represents the reality of the illness and is unique; the illness is subjectively experienced and Toombs argued that a person's whole existence is involved, not only the biological body. She maintained that, during an illness, one's view of the world and oneself changes, influenced by the attitude of other people and usually it occurs a shift from being healthy into having an illness. When applying Toombs definition of illness to the context of living with a CHD, a person with a CHD who does not have any symptoms and who functions well can be seen as healthy, despite the heart defect.

Health

There is debate about whether it is possible for someone who has a disease or is experiencing illness to achieve health. According to Toombs (1992), health is more than the absence of disease, and health can be consistent with disease and suffering, with the exception of unbearable suffering. The individual living with a disease is not focusing on the reality of physical signs or symptoms; rather, the effect of these signs or symptoms upon daily life becomes the important evaluating factor.

It is important for adolescents and adults with a CHD, as for most people, that their biological function is as intact as possible, but an intact biological function is not in itself enough to feel healthy. In caring science, the focus is not only the biological body and function, but rather the focus is based on the person's experience of disease, illness and health. Health is the ultimate goal of caring and the importance of health are also stated in Swedish Health Care Law, the target for

healthcare is stated to be a good health (HSL 1982:763). The main concerns in caring are health and existence, in particular how life is affected by a disease, a long-term illness or when experiencing unhealth. The goal in caring science includes helping the individual to maintain health and relief from suffering as well as improving health by alleviating the effects of a disease (Dahlberg & Segesten, 2010).

There are various definitions of health; one example is the biostatistical or biological theory developed by Boorse (2004), in which health is a one-dimensional construct involving the biological body. It focus on normality, where health is only possible in the absence of disease. According to this definition, a CHD is not consistent with health. Another example is the holistic theory of health, focusing on positive aspects; for example, Nordenfelt (1995, 2007) defined health as the ability to realize vital goals. A person has health as long as individual goals that are meaningful to the person are realizable. The salutogenic theory developed by Antonovsky (1987) also has a holistic ground, where health is evaluated as a resource for the individual. Health is seen as a continuum, with the anchoring points *dis-ease* and *ease*, where the individual strives towards the ease end of the continuum. According to Nordenfelt's definition, a CHD would be consistent with health, as long as the vital goals are realized or, according to Antonovsky, as long as a movement on the continuum could occur.

One other widely known definition of health is that of the World Health Organization (WHO), in which health is defined as "*a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity*" (WHO, 2010). This definition has been debated, since the definition points to an ideal state rather than a realistic one. It has been revised and expanded during recent years, towards a multidisciplinary concept expressing positive life values, where health is seen as a resource instead of a goal. Health is according to WHO seen as a resource for daily life, not the objective of living; it is a positive concept, emphasizing social and personal resources as well as physical capabilities. This means that it is interpreted differently between individuals, constantly in flux according to the particular life situation and integrated into the individual's life.

Eriksson (1995, 2002) defined health as a dynamic process, constantly in movement and concerning all aspects of human life. According to this definition, the human being is seen as a whole, consisting of body, soul and spirit; an inner balance, balance in relation to others and in life in general have to be achieved in order to have health. Eriksson (1989) have defined health according to three dimensions: *health as behaviour*, *health as being* and *health as becoming*. *Health as behaviour* is connected with actions or behaviour promoting health, *health as being* is connected with the balance in one's inner state, and *health as becoming* is connected with growing towards health. The stable balance between motion and stillness, also called the life rhythm, is connected with "being capable of" (Dahlberg & Segesten, 2010). In order to understand the health experience of adolescents and adults with a CHD, health is viewed from the standpoint of Eriksson's definition of health, in which health is a concept including the whole person's existence.

CENTRAL CONCEPTS

For people with a CHD, health has been shown to be of central importance (Ternstedt Östman, 1989) and two concepts that have shown to be closely connected to health are sense of coherence (SOC) and quality of life (QoL). It is therefore important to consider how the concepts of SOC and QoL can be viewed and applied within the setting of adolescents and adults with a CHD.

Sense of coherence

The sense of coherence (SOC) concept, which includes the salutogenic model, was developed by Antonovsky (1979). The salutogenic model focuses on a person's resources and capacities to maintain health, instead of focusing on the pathogenic perspective (Antonovsky, 1979, 1987). The underlying intention with the SOC concept was to place a focus on the resilience factors that keep people healthy, in order to be able to explain why some people can stay healthy when going through stressful life events whereas others cannot. Antonovsky (1987) argued that the different resilience factors had one main thing in common: the ability to make stressors in life manageable. Within the context of adolescents and adults with a CHD, one fundamental stressor in life is their CHD which could be connected with the prospect of an uncertain future.

Antonovsky (1979, 1987) viewed SOC as a crucial determinant of an individual's position on the health continuum. He suggested that the SOC concept is both universal and a stable, generalized and long-term way of looking at the world and one's own life. He further maintained that a person with a stronger SOC is more likely to have more ability in handling stressful situations in a more effective way. In order to measure individuals' overall capacity to cope with stressful life situations and to explain the factors that maintain health, Antonovsky (1987) developed the SOC scale. The SOC scale has been widely used and SOC scores have a demonstrated association with health and QoL.

Quality of life

In the context of the dramatically improved survival rates of adolescents and young adults with a CHD in recent decades, there is increased interest in how these individuals experience their QoL. There is no unitary definition of QoL that can be used within all contexts and for all circumstances, and separate disciplines view and define QoL differently: in sociology the focus is on well-being, including both objective and subjective dimensions; in psychology the focus is on perceived well-being and satisfaction; philosophy focuses on life values and in medicine the focus is mostly on normality and function (Polit & Beck, 2012). A starting point for the QoL concept is multidisciplinary and in general associated with positive values such as happiness, success, wealth, health and satisfaction (Lindström, 1994). QoL can semantically be given the meaning the "essence of existence".

WHO (1995) also refers to QoL as a multidimensional construct in which the respondent simultaneously evaluates several dimensions. The WHO definition of QoL – “*individuals perceptions of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns*” – encompasses QoL as a subjectively viewed concept, including both positive and negative facets of life. The overall judgement of QoL is affected by the person’s physical health, psychological state, level of independency, social relationships and their relation to other salient features in the environment (Bowling, 2001).

In her analysis of the different definitions of QoL, Ferrans (1990) concluded that they can be grouped into five broad categories; *normal life, happiness or satisfaction, achievement of personal goals, social utilities* and *natural capacity*. *Normal life* definitions focus on the ability of the individual to function on a similar level as healthy people; *happiness or satisfaction* definitions focus on an individual’s feelings of happiness or satisfaction; *achievement of goals* definitions focus on the ability to fulfil personal goals; *social utility* definitions focus on the ability to lead a socially useful life and *natural capacity* definitions focus an individual’s psychological and mental capacities. Moons and co-workers (2006) added two other conceptualizations; *utility* and *satisfaction with life*, where *utility* refers to health state valuations while *satisfaction with life* concerns to what degree the overall QoL is positively evaluated.

QoL is commonly used as a generic label to describe physical and psychosocial variables (Gill & Feinstein, 1994), covering a variety of concepts such as functioning, health status, perceptions, life conditions, behaviour, happiness, lifestyle and even symptoms (Bowling, 2001; Simko, 1999). The central terms in QoL are needs, resources and objectives of life, and both material and non-material measures, including both objective conditions and subjective perceptions, are used (Fayers & Machin, 2007). QoL can further be seen as the capability to fulfil one’s needs, improving one’s resources to reach life objectives, in other words, developing strategies and skills to manage life (Nordenfelt, 2007).

There is a growing consensus that QoL is determined by the subjective appraisal of one’s living conditions (Fayers & Machin, 2007; Gill & Feinstein, 1994) since objectively measured indicators of living conditions have only been found to account for about 15% of an individual’s QoL (Day, 1996). Within this thesis, QoL is seen as the subjective perception of overall satisfaction with life, including all aspects concerning the essence of existence. The essence of existence definition of QoL was developed by Lindström (1993a) and is a frequently used QoL model within the Nordic countries and within the setting of children with a CHD (Bratt, Östman-Smith, Axelsson et al., 2012; Ekman-Joelsson, Berntsson, & Sunnegårdh, 2004; Mellander, Berntsson, & Nilsson, 2007). The definition is primarily intended for the setting of children and adolescents. The essence of existence definition of QoL takes into account life as a whole, using both subjective and objective measures. It presupposes necessary internal and external resources for a good life

for the individual and it is outlined from a holistic context model including living circumstances and resources for a good life, in order to cover a child's life situation (Lindström & Eriksson, 1993a). It is built on three spheres: external, interpersonal and personal, where QoL is defined as satisfaction with life focusing on the areas of life that are of importance, with the initial focus on the life situation of children and their families. The important areas are: health, functioning, psychological support, family and social domains.

Within the field of adolescents and adults with a CHD, Moons and co-authors (2005) have tried to construct a sound conceptual definition of QoL, based on a review of already existing definitions. According to them, QoL could be defined as *“the degree of overall life satisfaction that is positively or negatively influenced by an individual's perception of certain aspects of life that are important to them, including matters both related and unrelated to health”*. This definition refers to general life satisfaction and it suggests that the only indicator of QoL is overall life satisfaction. Other variables are instead seen as determinants with either a positive or a negative impact (Moons, Van Deyk, Budts et al., 2004). This way of viewing QoL emphasizes the importance of the subjective approach to QoL.

QoL means different things to different people and most people have an intuitive sense of what the concept stands for (Fayers & Machin, 2007). It is concluded that QoL has more to do with subjective well-being (de Weerd et al., 1991) and life satisfaction (Meeberg, 1993; Simko, 1999) than with objective measures. This is exemplified by Albrecht and Devlieger's (1999) construct of the disability paradox, in which a high QoL can be experienced even by those who have a disease. The paradox is demonstrated by physically disabled people who experience a good QoL, in contrast to most external observers' assumption that they live an undesirable life. The disability paradox, therefore, further addresses the disparity between objective conditions and subjective experiences.

In the context of people with a disease, the main focus is on those aspects of QoL that are affected by the disease, and the term health-related quality of life (HRQoL) is used instead. HRQoL is seen as a demarcation of the concept QoL and it refers to the physical, psychological and social functioning of the individual and to the impact of the disease and treatment on ability and daily functioning. It concerns how people perceive aspects of life that relate to their health or how their health affects the value of those aspects (Bowling, 2005; Fayers & Machin, 2007). Bowling distinguished HRQoL from overall QoL with the argument that HRQoL focuses on the impact of a perceived health state and, when health status deteriorates, the perspective on life, roles, relationships and experiences will change. According to Fayers and Machin (2007) HRQoL can include varying relevant aspects. It usually covers functioning and well-being but could include all, or some, of the following aspects: general health, physical functioning, physical symptoms, emotional functioning, cognitive functioning, role functioning, social well-being and functioning.

PREVIOUS KNOWLEDGE

Within the whole population of children, adolescents and adults with a CHD, the research focus has shifted during the last decade, from studying survival rates towards a deepened interest in issues concerning long-term follow-up of health, quality of life, functioning in daily life, as well as psychosocial concerns. One group that has been of special interest in recent years is adolescents and adults living with a surgically palliated univentricular heart. Adolescents and young adults living with Fontan circulation belong to the first generation living with this pioneering, surgically created circulation and there are still many unanswered questions surrounding what will characterize their experience of adulthood. The research effort presented in this thesis focuses particularly on the subjective experience of young people living with a CHD, including living with a univentricular heart, and what impact the heart defect has on their lives. Therefore, the summary of the literature will highlight studies investigating health, quality of life and some of the different aspects of life that are affected by having a heart defect.

Perception of health and quality of life

Among adolescents and young adults with a CHD, studies have showed both comparable and contradictory results concerning health perception, QoL and HRQoL. One partial explanation could be that the studies have been performed at different times over a period when surgical and medical treatment was making constant and radical progress. Differences in sample sizes, age range and types of heart defect could have contributed further. In order to put the findings into context, comparisons with either healthy control groups, a general population or adolescents and adults having congenital heart defects have been made in the studies mentioned below.

It is shown that adults with a CHD may experience their health differently over time. The major adjustment after surgical intervention in childhood is usually successful; however, the experienced restrictions related to the heart disease may increase with time (Ternstedt, et al., 2001). In terms of overall QoL and satisfaction with life, adults with a CHD have reported even better scores than healthy controls (Moons, Van Deyk, et al., 2006; Silva, Vaz, Areias et al., 2011).

When measuring HRQoL the dimensions of general health and physical function are typically the most affected (Gratz, Hess, & Hager, 2009; Kamphuis, Ottenkamp, Vliegen et al., 2002; Lane, Lip, & Millane, 2002; van Rijen, Utens, Roos-Hesselink et al., 2005). In a study of young adults with complex CHD compared with the general population, it was shown that this group only experienced limitations in the physical dimensions of HRQoL and subjective health status (Kamphuis, et al., 2002). Lane and co-workers (2002) also found that adults with CHD had significantly poorer levels of physical functioning and lower overall general health perception. Those with an inoperable or a cyanotic condition

and those with a surgically cured condition (for example an atrial septal defect) reported the poorest QoL. Loup and co-workers (2009) investigated HRQoL, anxiety and depression in adults with CHD. They found that these adults reported excellent scores, comparable to the general population and with no differences between their initial diagnosis. Also Immer and his co-workers (2005) showed that HRQoL was similar to an age- and gender- matched general population, but they found the main restrictions not only in the physical function but also in the emotional aspect which is in contrast to other findings. Even for adults with minor defects, not in need of surgical correction but with social impediments (for example in relation to work or leisure time), physical function and general health is affected (Fekkes, Kamphuis, Ottenkamp et al., 2001). For those adults without social impediments health status were found to be comparable with the general population and better scores were found in the dimension of bodily pain, indicating that healthy people report a higher occurrence of bodily pain.

Concerning children, adolescents and young adults who have undergone the Fontan procedure, significantly lower health scores have been reported in comparison with their siblings (Manlhiot, Knezevich, Radojewski et al., 2009) as well as in comparison with healthy controls (McCrindle, Williams, Mital et al., 2007), namely that lower physical activity levels were found to be associated with lower general health. Pike and co-workers (2011) have showed that this group of young people living with a surgically palliated univentricular heart experience residual and sequelae from the multiple surgical procedures and the natural course of their disease. They experience many different symptoms, disease-related stressors and limitations and their primary worries are about health. QoL scores for the specific population of adolescents and adults with a univentricular heart have been shown to be similar to a healthy population (Overgaard, Schrader, Lisby et al., 2011; Saliba, Butera, Bonnet et al., 2001), but other studies have also shown that these adolescents and young adults report lower physical health and are more depressed than a healthy population (Pike, Evangelista, Doering et al., 2011). In the same study, Pike and co-workers also showed that, despite lower levels of physical health, no differences in QoL were found in comparison with the healthy population.

Impact of heart defect severity on health and quality of life

One commonly asked question has been whether the heart defect affects perceived QoL, with the underlying assumption being that the more severe the defect, the more difficulties experienced and therefore a lower QoL can be expected. Ternstedt & co-workers (2001) compared patients with an atrial septal defect (ASD), which is considered a minor defect, to patients having the more complex defect tetralogy of Fallot (TOF), 20 and 30 years after surgery. Unexpectedly, patients with the minor defect reported their QoL to be lower than patients with the more complex defect. However, when focusing on health status, other studies have shown either no differences between different diagnosis groups (van Rijen,

et al., 2005) or significantly lower health status for adults with a cyanotic heart defect compared with adults with an acyanotic defect, which is a less severe defect (Lane, et al., 2002; Moons, Van Deyk, De Geest, Gewillig et al., 2005; Simko & McGinnis, 2005). Cyanotic patients also presented higher disability scores (Simko & McGinnis, 2005). That the severity of the heart defect had an effect on health status but only a marginal impact on overall QoL and life satisfaction, is also found by Vigl and co-workers (2011). They showed that patients with a moderate or severe heart defect reported significantly lower scores in the physical dimensions of HRQoL. For the mental dimensions of HRQoL, socio-demographic factors showed a stronger association, than did severity of the heart defect.

Impact of other factors on health and quality of life

It has been suggested that SOC is an important determinant of health (Eriksson & Lindström, 2006) and of QoL (Eriksson & Lindström, 2007). Moons and Norekvål (2006) have speculated that SOC could be a mediating factor in improving QoL for adults with a CHD and Neuner and co-workers (2011) recently showed, in a study of adolescents with CHD, that SOC is an independent predictor of QoL. It has further been shown by Luyckx and co-workers (2012) investigating individual and contextual determinants of QoL in adolescents and found that SOC, perceived health status and parental support positively predict QoL over time. Parental support and SOC were interwoven resources and parental support was found to have a positive influence on SOC.

Impact of a congenital heart disease on daily life

The focus on what it means to live with a CHD has become of increasing interest during recent years. A CHD involves an uncertainty surrounding the prognosis and course of the illness, the signs and symptoms and how physical activity restrictions may intrude on daily life (Moons, Van Deyk, De Geest, et al., 2005). Earlier research has found that adolescents and adults experience feelings of being different and not being like others (Claessens, et al., 2005; Gantt, 1992); the findings were interpreted as a normalization process, where striving towards being normal was in central focus. Feelings of being different are shaped by both the individual's personality and the attitudes of the health care professionals and can, in turn, influence the care that is provided (Claessens, et al., 2005). This is reflected in the findings by Tong and co-workers (1998), who interpreted growing up with a CHD as facing different dilemmas during adolescence and early adulthood. The participating adolescents and young adults were grappling with either regarding themselves as normal, equal to everybody else, or perceiving themselves as different, deficient and less than perfect. Studies have also concluded that young adulthood is a more difficult time than adolescence, mainly due to the many new dimensions to their lives, such as relationships, work and future (Horner, et al., 2000).

Social functioning has been indicated to be of great relevance for adolescents and adults with a CHD, more than other aspects of living with a CHD. A diminished social function has further been shown to be predictive of depression and anxiety (Kovacs, et al., 2009). Higher levels of psychological stress are seen in adults with a CHD compared to a normal population (Simko & McGinnis, 2005), which has been explained as a response to the additional stressors in their lives, for instance their uncertain life expectancy. Horner and co-workers (2000) have shown that many of the adults are concerned over a shorter life expectancy, disability and aspects of starting a family, including problems with childbearing. The shorter life expectancy is associated with existential worries, which have been shown to be closely tied to body image. Adolescents and adults with a CHD may be reluctant to accept surgical interventions that leave scars because of fear and anxiety over intimate relationships, as reported by Rietveld and colleagues (2002). Horner and co-workers (2000) found that the adults in their study functioned well in daily life, but psychiatric evaluations revealed that many of them met symptomatic criteria for psychiatric diagnoses.

In conclusion, previous knowledge has demonstrated that a CHD affects many aspects of life where psychosocial aspects has been shown to be of great concern. Diverse results have been shown regarding QoL and health perception, but the overall QoL are in most respects good. Instead has physical limitations and perception of general health recurrently shown lower scores. However, more knowledge regarding what it means to live with a CHD or a univentricular heart needs to be established in order to provide well-suited care.

RATIONALE FOR THE STUDY

That a CHD plays an important role both in the person's own life as well as for the whole family is known, but how the specific consequences of living with a CHD are managed has not been well studied. Adolescents and young adults with a CHD have a diversity of heart defects, requiring different surgical procedures and interventions that create both unique physiology and anatomical structures with different impacts on life. While many of these individuals are able to live a relatively normal life despite their heart defect, others face diverse obstacles throughout life.

However, not all consequences of a CHD have been documented; therefore there is a current need to increase our understanding of how people with a CHD experience and manage life. There is a need to expand the current view of what it means to live with a CHD and to deepen our understanding of its nature. A combination of different approaches are needed; registry-based data with a large sample size need to be complemented by in-depth interviews with a small group of informants to give a wider description of their lives and to explore different aspects.

The extensive improvements in congenital cardiology and heart surgery during the last decades have created new groups of children surviving into adolescence and adulthood. One group is adolescents and young adults living with a surgically palliated univentricular heart. Univentricular heart is a comprehensive term covering various complex heart defects where the surgical end result leads to a pulmonary circulation not passing a venous or subpulmonary ventricle. Today these complex heart defects are palliated by multiple surgical procedures performed over a time span of several years, including repeated interventional and diagnostics procedures as well as multiple open-heart surgery procedures. These adolescents and young adults represent the first generation living with this condition and how these individuals experience life is barely explored. The achievements and progress within this field have added years to their lives and it is therefore necessary to further consider and emphasize the impact of all interventions being made. What does the experience of single or multiple interventions and surgeries mean to the individual in the long run? In order to evaluate the meaning of adding years to a person's life within this group, more tailored study designs focusing on their lived experience are needed.

The insights derived from this thesis could facilitate our understanding of how life and adjustments to life are influenced by the repeated palliative procedures or by one successful repair and, furthermore, what adjustments are needed from the healthcare professionals. The foremost rationale is to create a solid fundament of knowledge from the perspective of adolescents and adults that will increase understanding and be useful in care settings. This deeper knowledge is important as a basis for strengthening the capacity of the individual and to provide health care within a salutogenic approach, enabling more directed clinical interventions.

AIM

Overall aim

To interpret and illuminate how adolescents and young adults with a congenital heart disease (CHD) experience life and their life situation. Furthermore, the thesis aims to explore and describe the impact of a CHD and the influence on health perception, sense of coherence, quality of life and satisfaction with life in adolescents and young adults living with a CHD or, in particular, a surgically palliated univentricular heart.

Specific aims

Study I

To explore and gain a deeper understanding of young adults' experiences of living with a congenital heart disease in order to enhance the quality of care provided by the health care system.

Study II

The aim was twofold; firstly to evaluate and compare the reported health status and health perception of adult patients with a CHD included in the GUCH registry and, secondly, to investigate what variables influenced the patients' health status and health perception.

Study III

To illuminate and gain a deeper understanding of adolescents' and young adults' experiences of living with a surgically palliated univentricular heart (a Fontan circulation).

Study IV

To explore the life situation of adolescents' and young adults' living with Fontan circulation and to investigate what impact their condition (the univentricular heart) has on health, quality of life, sense of coherence and satisfaction with life.

METHODS

The overall methodological approach in this thesis is hermeneutical since all four studies are interpretations of experiences and self-reported data on what it means to live with a CHD and, in particular, a surgically palliated univentricular heart.

Methodological perspective and viewpoints

Hermeneutics focuses on understanding people's life-world. According to the French philosopher Ricoeur (1976), both explanation and understanding are needed in order to interpret a person's life-world, since understanding encompasses explanation and explanation develops understanding. The intention is both to explain and understand. The dialectic movements between explanation and understanding can be compared with the circular movements in the hermeneutical spiral. These movements consist of an ongoing circulation between the parts and the whole, in which the interpretation of each part builds on the other parts, leading to a final comprehensive understanding and interpretation (Patton, 2002). Pre-understanding is of essential importance; without this, we are not able to understand at all. It means that the comprehension of a phenomenon increases as the data unfold (Morse, 2003; Polit & Beck, 2012) and when adding one piece to another, understanding develops step by step. New findings contribute to understanding, and a new pre-understanding emerges in coming interpretations.

In order to capture, explain and understand the phenomenon of living with a CHD, in particular a surgically palliated univentricular heart, a multi-method approach has been used (Morse, 2003). According to the steps of how knowledge develops, an initial understanding was given in Study I of adults' experiences of living with a CHD, stimulating new questions regarding how health is perceived. This in turn led to Study II, which evaluated health status and reported health perception of a broader sample of adults with a CHD, giving a wider description of their reported health status. Differences in reported health status were found, with a higher occurrence of health problems in the group of adults with a surgically palliated univentricular heart, which raised further questions regarding how they experienced their life situation and what impact the univentricular heart had on their life. This led to Study III, which gave a deeper understanding of what it means to live with a univentricular heart from the perspective of adolescents and young adults. The final study (Study IV) complemented the picture of how these adolescents and young adults experienced their life situation.

Design

This thesis employs a multi-method design, containing both quantitative and qualitative analyses with interviews, instruments and questionnaires. This design aims to provide a comprehensive picture of human behaviour and experience (Morse, 2003). The different data sets were treated independently during the data

collection but, as a whole, the findings complement each other. By combining different approaches, the perspective of the adolescents and young adults became the central focus, which facilitated a broader comprehensive understanding of life experiences among those living with a CHD or a surgically palliated univentricular heart.

Both inductive (Study I and III) and deductive approaches (Study II and IV) were used. In three of the studies (Study I, III and IV), the design contained both an explorative and descriptive element. The aim was to interpret and understand the meaning of living with a CHD (Study I) or a univentricular heart (Study III) and further to describe the respondents' experiences. In Study IV the aim was to describe the life situation of adolescents and young adults as well as their perception of sense of coherence, health, QoL and overall satisfaction with life. In Study II, a correlational, retrospective, cross-sectional design was used in order to investigate how adults report their health status, their general health perception and what variables influenced their health status (Table 1).

Table 1. Overview of the research design for all the studies in the thesis.

Study	I	II	III	IV
Focus	What it means to live with a CHD	How health status and general health are reported	What it means to live with a surgically palliated univentricular heart	Experienced life situation when living with a surgically palliated univentricular heart, including health, QoL and SOC
Design	Explorative Descriptive	Correlational Retrospective Cross-sectional	Explorative Descriptive	Explorative Descriptive
Data collection	Qualitative: In-depth interviews	Quantitative: Questionnaire EQ-5D The GUCH registry	Qualitative: In-depth interviews	Qualitative & Quantitative: Questionnaires SOC scale, Study-specific Essence of Existence questionnaire
Sample*	20 adults with CHD	1435 adults with CHD	13 adolescents and young adults with a surgically palliated univentricular heart	34 adolescents and young adults with a surgically palliated univentricular heart
Data analysis	Phenomenological - hermeneutical method	Statistical analysis	Phenomenological - hermeneutical method	Content analysis & descriptive statistics

*The sample sizes consist of the eligible participants for each study

The quantitative analyses were used to sort, compare, generalize and describe the different groups of patients, their perception of health or the frequency of symptoms (Study II) and the perception of health, QoL, SOC and overall life situation (Study IV). In Study IV a combination of both qualitative and quantitative analysis were performed due to the data. The qualitative analysis; the phenomenological hermeneutical method was used in two of the studies in order

to gain insight into what it means to live with a CHD (Study I) or a surgically palliated univentricular heart (Study III). The lived experience cannot be shared but *the meaning of the experiences* can and in order to learn more about a person's life-world we must search for meanings through interpretation. Interpretation involves both understanding and explaining, since they are interwoven.

In the final study (Study IV) a qualitative content analysis was used, focusing on the content in order to describe the life situation of these individuals. Content analysis was originally used in theology and in mass media research in order to analyse text and interpret individual responses to texts (Krippendorff, 2004). Content analysis focuses on the characteristics of language as communication, with attention to the content or contextual meaning of the text. The text may be transcriptions of taped interviews, articles, books or diaries. According to Hsieh and Shannon (2005), a directed content analysis can be used when the existing theory or prior research about a phenomenon is incomplete or would benefit from further description. In order to better understand adolescents and young adults living with a surgically palliated univentricular heart, content analysis was used when analysing the written responses on the study-specific Essence of Existence questionnaire.

Selection, participants and setting

The four studies include different study populations, different selection criteria and also different settings.

Table 2. Inclusion criteria for the four studies.

Study no	Inclusion criteria
I	20–40 years old Having a CHD Member of the Swedish Childrens Heart Association
II	A patient at the GUCH clinic in Gothenburg EQ-5D data available
III & IV	Diagnosis of univentricular heart Surgical palliation, according to Fontan/ TCPC procedure, performed before 1995 at the selected paediatric cardiac surgery unit in Sweden

Study I

The underlying intention was to gather rich and informative data in order to explore and describe the phenomenon of living with a CHD. It could be assumed that people who are members in an association or a support group have considered more profoundly the impact of the heart defect and evaluated their experiences of living with a CHD more thoroughly compared to non-members.

Meeting others in the same situation has been shown to be supportive and empowering (Davison, Pennebaker, & Dickerson, 2000; Halding, Wahl, & Heggdal, 2010). For this reason, invitations were sent to adult members of the Swedish Childrens Heart Association (Hjärtebarnsförbundet), which is a nationwide non-profit association for children, adolescents and adults born with a CHD and their families. The age range 20–40 years old was chosen with the intention that the reported experiences should illuminate young adulthood.

Alongside the inclusion criteria, the study sample was partly strategically selected from five geographical locations in Sweden (Malmö, Lund, Umeå, Stockholm and Göteborg) within a radius up to 200 km. The selected cities were chosen because together they represented the residence area of the majority of potential participants. Out of twenty letters sent, one declined to participate and twelve did not respond at all. After a reminder letter, the final study population consisted of seven adults. One of the seven adults who accepted participation could not be interviewed due to sickness, which therefore gave six adults in the study. The participants comprised five women and one man, in the age range 22–39 years, with a mean age of 31 years. The participants had various congenital heart defects and had been operated either once or up to three times: one participant had had a heart transplant and one a heart and lung transplant. Four of the participants were married or lived with a partner and two lived alone. Three of the participants had children. All but one were working full-time.

Study II

The underlying intention was to achieve a broader sample in order to describe the population and perform correlational analyses between different groups. Therefore the Swedish GUCH registry was used. This is a nationwide registry covering all adult patients with a CHD who have their medical follow-ups at a GUCH clinic in Sweden. The data in the registry can be accessed as a national database or viewed separately clinic by clinic. For the present study, only the GUCH clinic in Gothenburg was chosen. This GUCH clinic is the second largest in Sweden, with over 2000 patients registered in 2010 (SWEDCON, 2011). At the time of the study, a total of 1435 patients were included in the registry and, of those, 1274 patients had valid EQ-5D data available of whom 1144 patients had valid EQ-VAS data available. Thus, a total of 1274 patients were included in the study. The study population had a mean age of 37 years, 51% were men and 51% were single, while 48% were married or lived with a partner.

Study III and IV

These two studies involved the same study population, originally based on a survey of children with a univentricular heart, where surgical palliation was performed according to the Fontan or the TCPC procedure. The survey was performed in 1995 with the aim of evaluating QoL for the child and the total life situation for the whole family, via a questionnaire completed by the parents. Recruitment for the survey was based on children from the two collaborating

paediatric cardiology units, served by one operating paediatric cardiac surgery unit in Sweden. For the present studies, the same participants were identified, but now as adolescents and young adults and thus they were asked directly to participate in an interview (Study III) and/or to respond to the questionnaires (Study IV).

In Study III, performed in 2010, participants were selected from only one of these two paediatric cardiology units. The sample consisted of thirteen adolescents and young adults and, of those thirteen, four were lost to follow-up due to missing addresses and could therefore not be reached, one was excluded due to cognitive difficulties and one had died. Therefore the remaining study population comprised seven adolescents or young adults (two men and five women) aged 17–32 years, with a mean age of 22 years. All of the participants either studied or worked full time, two of them were married or lived with a partner and two had children. Six of the participants had a heart defect that had been surgically palliated with a modified Fontan procedure (TCPC), while a classic Fontan procedure had been performed on the seventh participant at the age of 10.6 years after two previous surgical interventions. The mean age when the Fontan procedure was performed was 4.5 years (age range 1.6–12 years). One participant had undergone one previous surgical heart palliation, whereas for three participants this was their third, and for the remaining three it was their fourth surgical intervention.

In Study IV, all adolescents and young adults with univentricular heart, palliated with the Fontan or TCPC procedure before 1995, were identified from the same hospital records as in the 1995 study. The initial sample consisted of 48 participants and all but three had undergone a surgical palliation according to the current TCPC procedure in 1993 or 1994 at the same paediatric cardiac surgery unit. Those three had undergone a classic Fontan palliation in 1992 or 1993. After the TCPC procedure, one patient had a valvular prosthesis implantation and two patients had a revision from previous lateral tunnel TCPC to an external conduit TCPC. Three out of the initial sample had died by the time of the follow-up study in 2011. Of the remaining 45 eligible participants, three declined, three others had cognitive difficulties and five were lost to follow-up due to missing addresses, which gave a remaining study population of 34 participants.

Data collection

The data collection methods varied from interviews (Study I and III), clinical variables and registry data (Study II) to different instruments and questionnaires (Study II and IV). In order to evaluate health status and general health perception in addition to investigating what variables influenced health status, data from the Swedish GUCH registry were collected and the validated EQ-5D questionnaire was used (Study II). In order to evaluate and describe the participants' overall life situation and the impact of their univentricular heart, Study IV employed a study-specific modified version of the Essence of Existence questionnaire in

addition to the validated Sense of Coherence (SOC) scale. In order to gain a deeper understanding of the experiences of living with a CHD and further interpret what it means to live with a CHD or a univentricular heart, in-depth interviews were conducted (Study I and III).

In-depth interviews (Study I and III)

The purpose with performing in-depth interviews in the context of congenital heart disease was to elucidate the illness experience of living with a CHD and to interpret what it means to live with a CHD (Study I) or in particular a surgically palliated univentricular heart (Study III). In an interview, the situation is shared between the interviewer and the interviewee and, according to Ricoeur (1976), an interview is created jointly between the narrator (interviewee) and the listener (interviewer), where the interviewee becomes the co-investigator of the study (Ekman & Skott, 2005; Van Manen, 1990). The interview is a mediation built on communication, referentiality and self-understanding; this indicates that telling a story and then interpreting the text also can reveal meanings of which the interviewee is not aware.

It was considered important to establish personal contact with each respondent. Face-to-face interviews were therefore seen as important because they allow the interviewer to interpret the interviewee's reactions and thus to be sensitive and adaptable to what the interviewee wants to talk about. It is the task of the interviewer to provide a permissive atmosphere so the interviewee feels safe and secure (Kvale & Brinkmann, 2009). The art of interviewing also involves keeping the questions open and being sensitive to the meaning of the phenomenon, not only to oneself but also to the interviewee in order to be orientated to the substance of the topics that emerge. According to Kvale and Brinkmann (2009), a qualitative interview is a conversation form that aims to understand dimensions of the interviewee's life-world. The goal of the interviews was to encourage the interviewee to reflect over the phenomenon of living with a congenital heart disease (Study I) or living with a univentricular heart (Study III).

Interview procedure

Interviews focussing on the interviewee's personal history were conducted and the interviews were audio-recorded. Each interview was initiated with an opening question such as (Study I): "Can you tell me how you experience your life situation today?" and (Study III): "Can you tell me how your univentricular heart influences your life today? The interviewees were encouraged to speak freely and additional questions were asked in order to clarify the circumstances in their story.

Each interview lasted from 30 to 120 minutes and was transcribed verbatim. It was either conducted in the person's own home or in a secluded room at the hospital, according to the interviewee's own wishes. During the interviews, the

interviewer wrote field notes and after every interview the interviewer wrote a summary of the interview, noting spontaneous thoughts and feelings from the interview situation. The interviews transcripts were validated against the tapes.

Due to technical difficulties with the tape recorder, two interviews were unfortunately immediately erased during rewinding (III). In these two cases, where a back-up tape recorder had not been used, a second telephone interview was made a week later. After that technical hitch, an additional back-up tape recorder was used in the following interviews.

The Swedish GUCH registry (Study II)

The Swedish GUCH registry is a nationwide database, covering detailed longitudinal information on all adult patients attending one of the seven GUCH clinics throughout Sweden (Thilén, 2007). The registry was created in 1992 in order to follow up adult patients with a CHD. The underlying aim was to serve as a knowledge base about adults with a CHD to assure the quality of the care provided to them and to create conditions for national guidelines, in order to identify patients with a high risk for complications. Data are collected every consecutive visit, which facilitates comparisons and the ability to feed back to paediatric cardiology units with a long-term perspective. Since 2009, the GUCH registry is part of the newly created Swedish registry for congenital heart disease (SWEDCON), which aims to cover congenital heart disease in all ages, including the surgical interventions being made. SWEDCON makes it possible to follow up the patient throughout the whole life span and the registry brings together the knowledge from both paediatric cardiology and GUCH, including congenital heart surgery both for children, adolescents and adults (SWEDCON, 2011).

Because the registry-based study (Study II) was performed in 2009 and based on data from 2005 to 2008, the original GUCH registry (for adults) was used. At their first visit to the GUCH clinic, patients are asked for their consent to be included in the GUCH registry. They are given oral and written information about the registry, including information about the option to refuse permission to use their data for research purposes. Before a patient is included, a signed informed consent is obtained. The data are organized according to the patient's main diagnosis, which is based on the anatomical structural defect. In the GUCH registry there are 11 different main diagnosis groups: shunt lesions, aorta valve lesions, aorta anomalies, transposition (TGA), tetralogy of Fallot (TOF) /right ventricle anomalies, mitral valve anomalies, tricuspid valve anomalies, pulmonary valve anomalies, truncus arteriosus, single ventricle (SV) (all types) and other.

In order to evaluate the patients' perception of their health state and perceived general health, the validated generic EuroQol five-dimension questionnaire (EQ-5D), was chosen and added to the registry in 2005. Additional data in the registry are age, gender, marital status, housing, education and employment. Medical data such as the patient's diagnosis, catheterization or catheter interventions, type

of surgery, need of a pacemaker or of medication. Also included are physiological data, such as electrocardiogram, echocardiogram, and the patient's physical function scored by a cardiologist, using the New York Heart Association classification system (NYHA). If the patient is experiencing any symptoms and reports this spontaneously to the cardiologist, it is documented in the registry.

Questionnaires (Study II and IV)

EQ-5D (Study II)

Health status and general health perception were measured in Study II with the generic EQ-5D questionnaire. EQ-5D is a standardized instrument for measuring health outcomes and has become a well-used instrument, applicable to a wide range of health conditions (Brooks, 1996; The EuroQol Group, 1990). EQ-5D provides a simple descriptive profile of health, including a global generic cardinal index of health suitable for cross-national comparisons.

EQ-5D consists of the EQ visual analog scale (EQ-VAS), the EQ-5D self-classifier and a standard set of socio-demographic questions (1990). The self-classifier used comprises five dimensions: mobility, usual activity, self-care, pain/discomfort and anxiety/depression, where each dimension has three severity levels: no problem, moderate (some) problems and severe (extreme) problems. (Brooks, 1996). EQ-VAS is a rating scale (also called the "health thermometer") from 0 to 100 where the endpoint 0 represents the worst imaginable health and 100 represent the best imaginable health.

EQ-5D was originally developed in 1990 by the EuroQol group (1990) as a simple generic measurement which would capture physical, social and mental functioning. Physical function is encompassed within the "mobility" and "self-care" dimensions, social function is covered by the "usual activity" dimension and mental function is assessed with the "anxiety/depression" dimension (Brooks, 1996). The respondent is asked to choose the most appropriate statement regarding the experienced health state. In order to improve EQ-5D, there are now two available versions, where the newer version has five response levels instead of three. Therefore the EQ-5D instrument was renamed according to its two existing versions: EQ-5D-3L (the original version) and EQ-5D-5L (the new version). The original version, now the EQ-5D-3L, was used in the GUCH registry (Study II).

The study specific Essence of Existence questionnaire (Study IV)

The first version of the Essence of Existence questionnaire was developed in the middle of the 1980s by a group of child health specialists led by Bengt Lindström at the Nordic school of public health (NPH) in Gothenburg, Sweden. The aim was to evaluate health and QoL in children and the total life situation of their families and to create a Nordic standard of children's life conditions. The questionnaire was mainly for completion by the parent or for the parent and the child together. The 60-item questionnaire was pilot-tested, modified, reformulated and translated.

ed into all the Nordic languages (Lindström, 1994; Lindström & Eriksson, 1993a). The questionnaire is constructed from a holistic context model including living circumstances and resources for a good life within the following three spheres: *external sphere*, *interpersonal sphere* and *personal sphere* (Lindström, 1994; Lindström & Eriksson, 1993a). The model was also developed to be suitable for disabled children (Lindström & Eriksson, 1993b).

When the survey of children with a surgically palliated univentricular heart started in 1995, Lindström and co-workers' questionnaire (1993a, 1993b) was used, since it enabled assessment in a general population and in a population of children with chronic diseases. In order to collect disease-specific data in the survey, modifications were made, such as adding open-ended questions concerning the impact of the heart defect on daily life and on the overall life situation, in order to reflect the specific circumstances for these children and their families. Some existing questions were removed. This modified questionnaire consisted of 50 questions, ten of which were heart defect-related. With the addition of the heart defect-related questions, it provided a more detailed framework for describing the life situation for these children and their families.

For Study IV, part of the modified version from the 1995 survey was used, because the same respondents were involved, but now as adolescents and young adults. Since the original questionnaire was developed more than twenty years ago (Lindström, 1994), some of the questions needed updating and reformulation. In order to suit the current circumstances in 2011, the questionnaire was further adapted in collaboration with a researcher at NPH to include both semi-structured and open-ended questions. These study-specific adaptations were made in order to capture the overall life situation from the adolescent and young adult perspective instead of from the parental perspective. In order to direct the questions to adolescents and young adults, minor adjustments were made, such as: *how do you perceive your health?* instead of *how do you perceive the health of your child?*

The study-specific questionnaire consisted of 28 questions, of which 14 were taken from the modified questionnaire used in the 1995 child survey. The other questions were not considered suitable, since they concerned either the parental situation or the perspective of a child. Included in the *external sphere* were questions regarding occupation (work vs. school), working or studying part-time vs. full-time, disability pension and satisfaction with work, education, housing and financial circumstances. The heart defect-related questions in this sphere concerned the impact of the heart defect on the work situation, finances and education. The *inter-personal sphere* covered questions regarding the social network, the family situation, independent life and satisfaction with the family situation, friends and relatives, and leisure activities. The heart defect-related questions in this sphere concerned limitations experienced as a result of the heart defect, the experience of living a normal life in comparison with others and the impact of

the heart defect on the family situation and leisure activities. *The personal sphere* involved questions regarding activity, self-esteem, concomitant diseases, psychosomatic symptoms, satisfaction with general health, QoL and satisfaction with life in general. The heart defect-related questions in this sphere concerned the occurrence of heart-related symptoms, the impact of the heart defect on health and QoL, and obstacles faced in life because of the heart defect.

Layout of the study specific Essence of Existence questionnaire:

The layout of the questionnaire was divided into four sections (see Appendix); Section 1 contained seven questions on demographic and socio-economic data.

Section 2 contained six questions with five or more response alternatives about self-esteem, life satisfaction, health perception and overall QoL. Self-esteem was measured with six dichotomous indicators, where the items were scaled from 1 to 7 and summed into a total score. According to the original version, a score below 30 was considered as low self-esteem. The indicators were: dependent/independent, passive/active, lonely/not lonely, worried/calm, sad/happy and anxious/confident. Nine questions were used to measure satisfaction with different aspects of life (housing, work, financial situation, education, health, family situation, leisure activities, friends and ability to influence one's life situation), where the items were rated from 1 to 5. QoL and perception of health were measured with global ratings with five response alternatives. The question regarding normal life could be answered by three response alternatives: yes, no or don't know.

Section 3 contained 10 questions regarding the heart defect, its impact on daily life including any limitations, living an independent life, whether symptoms or other diseases were experienced and whether the respondent had been sufficiently informed about the heart defect and its consequences. These questions had three or more response alternatives and space for other comments. The questions on the experience of being limited by the univentricular heart, living an independent life and being sufficiently informed could be answered by three fixed responses: yes, no or partly. The influences of the heart defect on different aspects of life were evaluated by a rating from 1 to 5. The question "Obstacles in life" concerned an overall judgment of the difficulty of handling daily life in terms of level of disability. The following response alternatives were used: (I am) not at all disabled/slightly disabled/moderately disabled/seriously disabled/very seriously disabled. For some of the questionnaire items, follow-up questions were included in order to get more details regarding the respondents' individual situation.

Section 4 consisted of five questions concerning health care received, to capture the adolescent and young adult perspective on their needs as patients. These responses are not presented in Study IV. The final question was open-ended, asking the respondent to share aspects of their experiences of living with a univentricular heart (for the entire study-specific questionnaire, see Appendix).

Sense of Coherence scale

The concept of sense of coherence (SOC) has three components: comprehensibility, manageability and meaningfulness and is defined as a global orientation that expresses the extent to which one has a pervasive, enduring though dynamic feeling of confidence that a) the stimuli deriving from one's internal and external environments in the course of life are structured, predictable, and explicable; b) resources are available to meet the demands posed by these stimuli; and c) these demands are challenges, worthy of investments and engagement (Antonovsky, 1987, 1993). The SOC scale originally contained 29 items, from which a 13-item version was developed; the latter was used in Study IV. Both these scales have been translated and adapted into several languages and cultures. The Swedish version of the Sense of Coherence scale has been adapted, translated and tested by Langius and co-workers (1992).

The 13-item Sense of Coherence scale is built on the conceptualization of the three components *comprehensibility*, *manageability* and *meaningfulness*. The component *comprehensibility* consists of 5 items, *manageability* consists of 4 items and *meaningfulness* consists of 4 items. The respondents mark their response on a 7-point scale with two anchoring verbal responses "very seldom or never" and "very often". The total sum ranges from 13 to 91 and a higher score indicates a stronger SOC. The total scores are divided into low, medium and high SOC scores (Antonovsky, 1993; Langius, Björvell, & Antonovsky, 1992).

Procedure

In Study I, the Swedish Childrens Heart Association was helpful in identifying and contacting potential informants. The researcher sent the study information to the Association's chancellery staff and from there the information letter was sent to 20 potential participants. The study information included the purpose of the study and what would be required of them as participants. Responses were forwarded to the organisation, who then informed the interviewer. Of the twenty potential informants, three responses were received, two positive and one negative. After one month a reminder letter was sent, which resulted in five additional positive replies. The interviewer then contacted the potential participants, all of whom gave their informed consent to participate in the study. The interviews commenced after these seven participants had given their consent to participate. They decided the time and location for the interviews themselves; all but one interview was conducted in the interviewee's own home. One person could not be interviewed due to sickness, thus a total of six interviews were performed.

Before starting to collect the data for Study II, a formal approval from the administrator of the registry was obtained. The registry contains data from all patients living in the western region in Sweden who receive specialist care at the GUCH clinic in Gothenburg. Study II consists of data from all the patients' visits between

the years 2005 and 2008, but for the calculations of EQ-5D, data from the last visit only were selected. The self-administered questionnaire EQ-5D is routinely sent to the patient together with the notification of their next appointment with the cardiologist or the nurses. The patient is asked to fill out the questionnaire at home and bring it when visiting the GUCH clinic. After the clinic visit, the completed EQ-5D questionnaire is registered in the database, together with other information from the visit.

For Study III, all the seven potential participants from one of the two collaborating paediatric cardiology units in the 1995 survey, received an information letter about the study. The letter stated that participation was voluntarily and could be withdrawn at any time. In those cases where the participants were under the age of eighteen, an informed consent was also signed by the parent or guardian. All the potential participants who were contacted gave their verbal and written consent.

In Study IV, all the 34 potential participants from the 1995 survey from both collaborating paediatric cardiology units received an information letter about the study and the two questionnaires – the study-specific Essence of Existence questionnaire and the SOC scale – together with the consent form. Seventeen responses were received during the first month, of which three stated they declined. Three reminder letters were sent out to the non-responders at monthly intervals, which resulted in four completed questionnaires. One more reminder letter was then sent and supplemented with a telephone call, which resulted in four completed questionnaires. Finally, three weeks later, two more completed questionnaires were received.

Data analysis

Phenomenological hermeneutical analysis (Study I and III)

An approach inspired by Ricoeur's phenomenological hermeneutics (Ricoeur, 1976), further developed and described by Lindseth and Norberg (2004) was used in order to reveal the experiences and what it means to live with a CHD or a univentricular heart. The phenomenological hermeneutic analysis has the goal of gaining understanding and knowledge from the interviewee's perspective, through dialectic movement between explaining parts of the interview text and understanding the text as a whole (Lindseth & Norberg, 2004; Ricoeur, 1976). The interpretation of the text focuses on what unfolds about people's experiences in the world, i.e., what the text "talks about", and the main purpose is to obtain a valid and common understanding of the meaning of the text. The method assumes that interpretation of the interview texts can reveal meanings of which the interviewees may not be consciously aware (Ekman, Lundman, & Norberg, 1999; Ekman & Skott, 2005; Eldh, Ehnfors, & Ekman, 2004; Lindseth & Norberg, 2004).

The analysis followed the method described by Lindseth and Norberg (2004). They presented the phenomenological-hermeneutical method as consisting of: *naïve reading*, *structural analysis* and *interpreted whole*. Dialectic movements are involved between parts of the text and the whole. The dialectic movements have an existential overtone and involve being close to the text while also dissociating from it, in other words, being focused on what the text says but at the same time on the understanding it gives (Ricoeur, 1976).

In the *naïve reading*, the interviews were read several times in order to become familiar with the text and reach an initial understanding. *Structural analysis* was then performed, in which the interviews were read through systematically and “meaning units” were identified and excerpted from the whole text. Meaning units refer to sequences of the text that have a meaning of their own and are thus relevant to the aim of the studies in this thesis. These meaning units were then brought together, sorted into groups according to meaningful connections, condensed and grouped into themes and subthemes. Patterns of similarities formed themes, which can be defined as a thread of meaning that penetrates text sequences.

In the *interpreted whole*, the texts were read again and we reflected on the themes pertaining to what it means to live with a CHD or to be a young person living with a surgically palliated univentricular heart. The *interpreted whole* phase is built on the assumption that understanding and explanation of the details of the text contribute to the understanding of the whole of the text and vice versa. In this manner, the meaning of the text becomes appropriated, i.e., a deepened understanding is reached, which is also called comprehensive understanding. The interpretation was guided by the researchers’ pre-understanding based on extensive experience from meeting children, adolescents and young adults with a CHD and/or a univentricular heart, as well as knowledge of previous research. The pre-understanding was constantly reflected upon and reconsidered during the entire data analysis and interpretation process.

Content analysis (Study IV)

Since prior research about the phenomenon of living with a univentricular heart would benefit from an extended description, a directed content analysis described by Hsieh and Shannon (2005) was used. This method presupposes that variables are defined beforehand, and the initial step of the content analysis in Study IV was to identify key concepts or variables as initial coding categories (Potter & Levine-Donnerstein, 1999). Next, operational definitions for each category were determined for all the areas covered in the study-specific questionnaire. The analysis further consisted of focusing on the answers within the predefined categories (items), by analysing the manifest content and presenting it in a descriptive manner and as frequencies. The analyses of the open ended questions reveal and extend the understanding of how adolescents and young adults with a univentricular heart experience their life situation.

Analysis of EQ-5D (Study II)

When analysing the data, the patient's last visit with EQ-5D data available was selected and extracted. The score obtained from each dimension on the self-classifier is a one-digit number: 1 (no problems), 2 (moderate problems), or 3 (severe problems). Combining the five dimensions results in a five-digit number that describes the person's state of health; for example, 11111 denotes a person with no health problems in any of the five dimensions. The five-digit number can be converted to a single summary index by applying a formula weighting all levels in each dimension using an index tariff. An index value of 1 represents full health and 0 corresponds to death, which indicates that a negative value represent a state worse than being dead (The EuroQol Group, 1990).

Statistical analysis (Study II and IV)

The statistical software SPSS (version 15 and 18–19) was used for the statistical analysis (SPSS Inc., 291 Chicago, IL, USA). Descriptive statistics were used to characterize the sample and to display frequencies, proportions, means and medians (Study IV) and frequencies of EQ-5D data (Study II). For exploring relationships, non-parametric tests were used: Spearman's rank correlation coefficient to explore associations between SOC and health, SOC and QoL (Study IV); the Mann-Whitney U test to compare EQ-VAS scores and EQ-5D index values between two independent groups (Study II); the Kruskal-Wallis one-way analysis of variance to compare EQ-VAS and EQ-5D index values between all the diagnosis groups (Study II) and a multiple stepwise logistic regression to find predictors for EQ-5D index (1.0) and EQ-VAS (85–100) (Study II). For all tests, statistical significance was set at a p-value of <0.05.

Ethical considerations

The thesis was designed to follow the ethical guidelines and principles in line with the World Medical Association Declaration of Helsinki (2000), namely that the research should contribute to welfare, respect the autonomy of the participant, have an intention of doing good or not harming the participant and follow the principle of justice. One keystone in the principles is that the people invited to participate may choose to participate or decline of their own free will. The ethical considerations were taken into account for the thesis as a whole as well as for each separate study.

There was no formal application to the ethical board for Study I since that was not required at that time. The GUCH registry study (Study II) conformed to the ethical guidelines of the Declaration of Helsinki in that a priori approval was obtained from the institution's human research committee. The registry is approved by the ethical committee and by the Swedish data inspection board. For Study III and IV ethical approval was obtained from the ethics committee (Dnr 09-024M and 2011-246-32M).

In the studies involving direct (Study I and III) or indirect (Study II and IV) contact with the participants, they were informed in advance about the study and the aim. They were guaranteed confidentiality and when presenting the findings they were guaranteed anonymity. The potential risks for those participating in the studies are estimated to be low. However, there was a potential risk in Study I and III of arousing strong feelings in the interviewees about either their current life situation or about painful memories from childhood in connection with the heart defect. When planning the study, contact was established with an experienced counsellor, so that, if the participant felt a need for further counselling after the interview, it could be arranged. After each interview (Study I and III), the participant was asked if he or she felt a need for further therapeutic counselling regarding any feelings that emerged as a result of the interview, but this was not requested by any of the participants. Similarly, when filling out the questionnaire (Study IV) there was a potential risk of being reminded or overwhelmed by memories and feelings regarding one's life situation.

Nonetheless, all participants in these studies were born with a CHD, which could imply that they had already developed different strategies of handling painful event or memories, including feelings of existential concern, during childhood. To live with a CHD or in particular a univentricular heart is strongly connected with the knowledge that life is vulnerable and that no guarantees can be given for life expectancy, which was also shown in the interviews. All participants mentioned aspects of life expectancy. Especially the adolescents and adults with a univentricular heart were acutely aware of having survived due to improvements in surgical techniques, but this was connected with gratitude rather than any apparent fear. The interviewees might therefore have seen the interview study as an opportunity to share their experience, which could in fact be beneficial and empowering. When participating in an interview study, the relationship between the interviewer and interviewee is crucial. The interviewee is in a position of dependence to the interviewer, which makes it even more important to try to create a warm and permissive climate, to be structured and clear, but at the same time kind and sensitive (Kvale & Brinkmann, 2009).

RESULTS

The main findings in this thesis are summarized below:

Study I

- Adults with a CHD demonstrated ambivalence about how they should perceive themselves and whether they were healthy or not. They also expressed ambivalent feelings regarding whether they should disclose or not disclose their heart defect and regarding the gratefulness expected of them but at the same time not feeling grateful.

Study II

- As a group, adults with a CHD perceive their health to be good, in line with general populations.
- Most problems were reported according to the EQ-5D dimensions pain/discomfort and anxiety/depression, the same pattern as found in general populations.
- A difference in reported health status was found according to the diagnosis group: some respondents in particular diagnosis groups experienced significantly more health problems and reported a lower general health perception.

Study III

- Adolescents and adults with a surgically palliated univentricular heart experience themselves as exceptional, strong and healthy.
- A commitment to life was found and interpreted as connected with the fact that they survived against all odds.

Study IV

- A majority of the adolescents and adults with a surgically palliated univentricular heart experienced symptoms, but they still considered their health to be good or very good and the majority were also satisfied or very satisfied with their QoL.
- Adolescents and adults with a surgically palliated univentricular heart experienced a moderate SOC.
- The perception of health, QoL and self-esteem were all positively associated with SOC.

The results and findings from the different studies are presented both comprehensively and separately, study by study.

Demographic and clinical variables

Variables that could be compared across all studies are presented in Table 3.

Table 3. Characteristics of the study population in the studies.

CHARACTERISTICS	STUDY I	STUDY II*	STUDY III	STUDY IV
PARTICIPANTS	Adults with various types of CHD	Adult patients included in the GUCH registry	Adolescents & adults with Fontan circulation	Adolescents & adults with Fontan circulation
Number of participants	6	1274	7	20
Median age	30.5	33.0	22.0	23.0
Age range (years)	22-39	15-87	17-32	18-32
Gender (% women)	85	49	87	55
OCCUPATION (%)				
Employed	84.0	58.6	28.5	50.0
Studying	16.0	15.0	71.4	50.0
FAMILY SITUATION (%)				
Married/ cohabiting	66.0	47.9	28.5	20.0
Living alone	34.0	51.1	57.0	57.0
Living with parents	-	18.7	14.0	35.0

*Study II and IV: Additional variables were included, therefore the total sum is not 100 percent

The overall study population in this thesis consists of young people, and only one of the studies involved adults over the age of 40 years (Study II). The median age was below 34 years in all studies. The skewed age distribution for this growing population of adults with a CHD is clearly exemplified by the GUCH registry data (Study II), as shown in Figure 1.

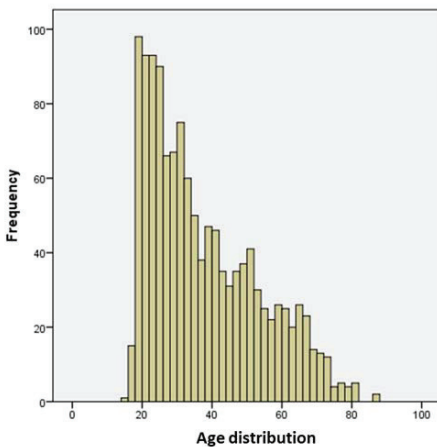


Figure 1. The age distribution of the 1435 patients included in the GUCH registry (Study II).

The peak in this population is found in the lower ages, between 20 and 30 years, exemplifying that adults with a CHD form a young population. This is further illustrated by variables such as family situation and occupation, where a higher proportion of the participants in Study III and IV were still studying and a lower proportion were married or lived with a partner.

Health and quality of life

Health was an important topic for the participants and was self-rated in EQ-5D (Study II) and in the Essence of Existence study-specific questionnaire (Study IV); in addition, both health and QoL were brought up during the interviews (Study I and III). In Study I the adults found it difficult to know how to perceive themselves: they were ambivalent about whether they were sick or healthy. They were also in two minds about whether they should tell others about their CHD; on the one hand they wanted people to know but on the other hand they wanted to be just like others and live a normal life without a disease. In contrast, adolescents and adults with a surgically palliated univentricular heart were not ambivalent and perceived themselves as healthy (Study III).

In order to establish what variables predicted an EQ-5D index value of 1.0 and an EQ-VAS score of 85–100, a multiple stepwise logistic regression was performed (Study II). Independent predictors of an EQ-5D index value of 1.0 were symptoms ($p<0.0001$), age ($p<0.0001$), NYHA class ($p<0.0001$) and gender ($p<0.0005$). The regression model further showed that symptoms ($p<0.0001$), age ($p<0.0001$) and NYHA class ($p<0.0001$) were independent predictors of an EQ-VAS score of 85–100.

As a group, patients with a CHD reported a good health state and a positive general health perception (Study II), and so did adolescents and adults with a surgically palliated univentricular heart (Study IV), of whom 80% considered their health to be good or very good and 90% were satisfied or very satisfied with their QoL. The EQ-5D results (Study II) showed that the health state profile of full health (11111) was reported by 52.3% and the mean EQ-5D index value for the whole population was 0.87.

Table 4 shows the distribution of reported problems in EQ-5D for the study population as a whole (Study II) and most of the adults experienced no health problems. If any health problems were reported, this was mainly as moderate rather than severe problems. The most commonly reported health problems were reported in the pain/discomfort dimension and the anxiety/depression dimension.

Table 4. Distribution of patients reporting no problems, moderate problems or severe problems in the EQ-5D health dimensions (all patients in Study II, $n=1274$).

EQ-5D	Mobility	Self-Care	Usual activities	Pain/discomfort	Anxiety/depression
No problems % (n)	88,8 (1131)	96,3 (1227)	84,0 (1070)	68,1 (867)	70,3 (895)
Moderate problems % (n)	10,8 (138)	3,1 (40)	13,8 (176)	29,9 (381)	27,5 (350)
Severe problems % (n)	0,4 (5)	0,5 (7)	2,2 (28)	2,0 (26)	2,3 (29)

Sense of coherence, self-esteem and satisfaction with life

The SOC scale was used in Study IV with adolescents and young adults with univentricular heart. It was found that the total mean score for SOC was 61.5 (range 32–87), median 63.0 (Study IV). Fourteen of the adolescents (70%) scored in the moderate range (46–74) and the other six were evenly spread in the high range (75–91) or in the low range (13–45). Significant correlations were found between SOC and QoL as well as between SOC and self-esteem. It was found that a higher SOC score was positively associated with more positive health perception.

In Study IV, satisfaction with life in relation to various dimensions – housing, work, financial situation, education, health, family situation, leisure activities, friends and ability to influence one’s life situation – showed that most of the participants reported high satisfaction, from 50 to 100%, in the different dimensions. For example, fourteen (70%) were satisfied or very satisfied with their health.

Symptoms and limitations in daily life

The analyses of the GUCH registry data (Study II) showed that 30% reported symptoms and that those who reported symptoms also presented a significantly lower EQ-5D index value (mean 0.78) than those who did not report symptoms (mean 0.91). Patients with a univentricular heart were among those who reported symptoms most frequently (Study II) which was also evident in more detail in the responses to the study-specific Essence of Existence questionnaire (Appendix 1). General and heart-related symptoms were reported by 80% of the study population, and the most frequently reported symptoms in Study IV were breathlessness and tiredness, mostly occurring during stress, hard physical activity or overstrain; most frequently reported symptoms in the registry-based study (Study II) were chest pain and palpitations.

Limitations in daily life, either partial or more common, were described by 90% of the adolescents and young adults with surgically palliated univentricular heart (Study IV) and the limitations were mainly physical. That this group experiences more limitations than others was also shown in the EQ-5D results (Study II), in which patients with a single ventricle reported the highest level of problems in almost all dimensions (Table 5). Table 5 presents percentages of reported problems according to the different diagnosis groups (Study II). However, as shown in the pain/discomfort dimension, patients with tricuspid valve malformation reported a higher number of problems.

A high occurrence of reported problems was found among patients with a univentricular heart (Study II), which to some extent could be explained by the responses about symptoms in Study IV. At the same time, 70% in Study IV rated themselves as not being disabled or disabled to a small extent due to physical restrictions.

Table 5. Reported problems in the five dimensions, presented as percentages within each diagnosis group.

N	Diagnosis group	EQ-5D				
		Mobility	Self-care	Usual activities	Pain/discomfort	Anxiety/depression
122	Aortic valve lesions	9.0	2.5	12.3	23.8	22.1
126	Aortic anomalies	4.0	0.8	11.1	21.4	21.4
97	Pulmonary valve anomalies	11.3	3.1	11.3	23.7	28.9
26	Mitral valve anomalies	3.8	3.8	15.4	34.6	38.5
26	Tricuspid valve anomalies	-	-	11.5	42.3	30.8
92	Fallot/ RV anomalies	17.4	4.3	18.5	28.3	32.6
89	Transposition	5.6	1.1	6.7	28.1	25.8
17	Single ventricle, all types	29.4	5.9	47.1	41.2	41.2
5	Truncus arteriosus	20.0	-	40.0	20.0	20.0
542	Shunt lesions	14.4	5.5	18.6	36.7	31.2
132	Other	6.6	2.3	17.3	37.9	37.1
1274	Total population	11.2	3.6	16.0	31.9	29.8

Daily life experiences

The participants' interpretations differed over how they experienced life and what it means to live with a CHD or, in particular, a surgically palliated univentricular heart. They reported that being born with a CHD involves special circumstances and unique experiences that had contributed to various adjustments throughout life and the person's psychosocial development had been shaped in relation to others in different directions. The following interpretation indicated that the participants in Study III experienced their mere existence as unique whereas the participants in Study I were struggling to be normal and not feel different (Table 6). Table 6 gives a comprehensive overview of the themes and subthemes interpreted during the analysis process (Study I and III) and Figure 2 shows the comprehensive understanding in Study I.

Balancing life between being healthy or not (Study I)

Living with a CHD was interpreted as ambivalence. The participants were born with different types of heart defects and these had been repaired differently. The participants found it difficult to know how they should perceive themselves, which was complicated by the fact that their CHD was invisible to others. All of the participants had an inner wish to live a normal life, which for them meant being healthy; this was related to feelings of suffering and grief. They sometimes experienced themselves as living in two worlds: in one world they were sick and

Table 6. Themes and subthemes from the structural analysis in Study I and III.

Study I	Study III
Living with Illness Accepting or not accepting the situation Feelings of suffering Thoughts and fears about the future Wish for a normal life/to live beyond one's own capacity Security/insecurity Expected gratefulness – not being grateful	Happiness over being me Feeling proud and mature Being humble and accepting other people Feeling healthy and special Feeling normal and the same as one's friends Belief in oneself
Having the disease Like – unlike Sick – healthy Tell – no tell Invisible handicap	Focusing on possibilities Dealing with physical restrictions Mastering medical procedures Trying to control the heart defect
	Being committed to life Holding on to life and creating meaning Living with uncertainty Making the most out of life

in the other world they lived a healthy life. This was associated with trying to find a balance between these extremes, aptly described by Ternulf-Nyhlin as “walking a fine line” (1991). They were also exceeding the limitations they were born with, living in some respects beyond their capacity in order to be normal and like others.

A representation of their reported experiences is shown in Figure 2, consisting of a frame with a core inside. It also illustrates the interpreted themes. The frame symbolizes the theme “Having the disease” and is not changeable, always constant, and becomes the person’s armour, which is easily visible. Despite the differences in the manifestation of the heart disease, the interviewees’ narratives were concordant in this regard. Inside the frame is the core symbolizing “Living with illness”. The core is the essence of living with the illness and is held within the person; it represents feelings, emotions and thoughts and cannot be seen from the outside. The core is changeable; it can either be large or small depending on how the person feels. For a person with a CHD who feels well and does not experience any heart symptoms, the core is small and it is possible to cope with life; if the person is feeling ill, the core is bigger. Sub-themes associated with the core were found to cover different meanings.

Feeling exceptional, strong and healthy (Study III)

Living with a surgically palliated univentricular heart was interpreted as being committed to life. The adolescents and adults had gone through many difficult and painful examinations and surgical interventions, experienced limitations

and restrictions and suffered from many hospital stays. With this in mind, they experienced themselves as exceptional, strong and healthy. The participants knew they existed thanks to innovative surgical procedures and advanced care and they created meaning around their heart defect that focussed on possibilities instead of the challenging medical procedures and other obstacles.

When facing life, they always knew that they had been through the most difficult ordeal – survival against the odds – and that this had made them strong. These experiences contributed to their development of an existential maturity that was integral to their efforts to make the most out of life. In the context of their experiences, a profound “happiness over being me” was revealed, along with a feeling of being committed to life.

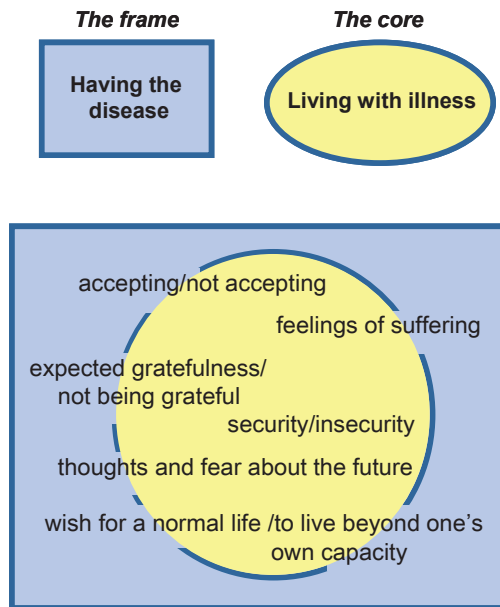


Figure 2. The Living and Having model.

DISCUSSION

Reflection on the findings

The experience of health

Health was a central aspect in this thesis and findings showed that both adults with a CHD (Study I and II) as well as adolescents and young adults with a univentricular heart (Study III and IV) regarded themselves as healthy, even experiencing good or excellent health. It may be considered incompatible to have a complex heart defect, surgically palliated with several procedures and giving rise to physical limitations, and yet still experience good health or be satisfied with one's QoL. An ambivalence concerning the view of oneself was also described: *I think I see myself as healthy....* (Study I). Adolescents and adults with a surgically palliated univentricular heart were, on the other hand, confident in the way they viewed themselves: *I am healthy and the same as my friends...* (Study III).

That the experience of health and QoL differed between adults with different heart defect and is not directly associated with the severity of the CHD, has also been shown by Ternstedt (1989). She focused on adults who had been operated as children and described these people's views of life. It was found that those with a more severe CHD reported a higher QoL, than those having a less severe defect.

Claessens and co-workers (2005) have earlier shown that adolescents and adults with a CHD view themselves as different from others without a disease and they make efforts to fit in and be normal. This was interpreted as a process of normalization. In children with a CHD, Bjorbækmo and Engelsrud (2008) found that the children perceived themselves as having the same capacity as healthy children, but at the same time they were aware of their physical limitations, which could be seen as a way of managing the CHD and as a way to normalize. The children were ambivalent in that they wanted others to take their limitations into consideration, but at the same time they did not want others to know about the limitations. This is similar to the ambivalence shown in Study I, in which the participants wanted to tell others about their CHD but at the same time keep it hidden. Their way of normalizing was expressed in their efforts to exert themselves, living over their capacity in order to be as good as others.

In spite of the ambivalence found in Study I, in combination with the three other studies in this thesis a strong sense of being healthy despite the CHD emerges. It is pertinent to bear in mind that the participants were all born with a heart defect, in some cases a very complex heart defect. Adolescents and adults with a CHD do not know any other life and their CHD is an integral part of their personality. Their physical limitations, symptoms and signs may be so well integrated that they are not even reflected upon as distressing.

The experience of symptoms

In the analysis of which variables could explain and influence the perception of health status and perceived health state (Study II), the strongest independent predictor was symptoms, followed by age and NYHA classification. Thirty percent of the study population experienced symptoms of some kind. It is understandable that symptomatic patients report a lower health state and experience more problems, given that symptoms might influence daily life. It was also found that symptoms were not always reported at the clinic visits, since some of the apparently asymptomatic patients reported health problems on EQ-5D.

The discrepancy between symptoms reported at the clinic visit versus when completing the self-assessment questionnaire is interesting and may have clinical implications. It may reflect fear of painful examinations and worry about the consequences of reporting symptoms. Experienced symptoms might be interpreted as a sign of a worsened state, which could indicate the need for future surgical interventions and therefore they are not disclosed. On the other hand, failure to report symptoms might reflect a neglect of those symptoms, connected with strategies of mastering the CHD in the way it is integrated into daily life. Once they become accustomed to their own signs and symptoms, they may not even recognize them as symptoms or strange sensations any more, but rather as a natural part of life and not worth mentioning at the clinic visit.

This integration of symptoms is further supported in the comparison of EQ-5D data between this study population and published results from a general population and from a healthy control group. Adults with a CHD reported lower levels of problems in the EQ-5D pain/discomfort dimension, than the healthy population did. One might assume that people with a disease of some kind would report and experience a higher level of pain and discomfort than healthy people do. Yet the same tendency has also been reported by Rijen and co-workers (2005), studying adults with various malformations, and by Saliba and co-workers (2001), who compared a group of adolescents and adults with a univentricular heart with a healthy group. The findings could to some extent be explained in terms of how the questions were phrased, whether they presented response options or were open-ended so that the respondent could answer freely. In Study IV of this thesis, in which more open-ended questions were used, it was found that most of the adolescents and young adults with a univentricular heart experienced some kind of symptoms, but not necessarily bodily pain.

Growing towards health

From the outsider perspective and with a problem-focused approach, adolescents and adults might be seen both as disabled and as not being able to experience good health. This thesis has shown that they do not necessarily view themselves in that way. Throughout life, the heart defect is integrated and a balance is achieved in the person's inner state. Bjorbækmo & Engelsrud (2008) interviewed children

with a CHD and found that they learn even during childhood how to adjust and respond to different situations. They are aware of their limitations but at the same time they see themselves as having the same capacities as other children. This could be linked with Eriksson's definition of health as *becoming* (1995), meaning that the individual grows towards health. Eriksson's health concept suggests that people adapt throughout life to their limitations. This indicates that their view of themselves does not necessarily undergo a shift when they get an illness, as described by Toombs (1992).

Toombs' statements regarding the changed view of the world and of oneself, could in this context more accurately be described as a changed view from being sick (when born with a CHD) towards being healthy as a result of the many surgical procedures that have been performed. Even if they face various kinds of limitations during their childhood, they are not aware of them because they have never experienced "normal" health. Their only reference to a normal level of functioning is embedded in their disease.

This conclusion has also been drawn by Pike and co-workers (2011), who found QoL to be comparable between adolescents and adults living with Fontan circulation. Physical health and functional status deficits were found but they concluded that these were not perceived as a "true" disability or limitation such as a paralysis. Therefore, good health can be experienced, despite the impact of the heart defect.

"Health within disease"

According to the definitions by both Eisenberg (1977) and Kleinman (1995), adolescents and adults with CHD can be considered to have a disease but at the same time experience good health. The applicability of Moch's concept "health within illness" (1989), is tied to her view of illness as not something to get rid of, but rather something to get in touch with. If we substitute the word *disease* for *illness*, giving "health within disease", the findings from this thesis can be well understood. Thus, people living with a CHD can experience themselves as being healthy, and, indeed, the adolescents and young adults with a uni-ventricular heart in Study III firmly and consistently expressed their experience of good health and their perception of themselves as exceptional, strong and healthy. Zahmacioglu and co-workers (2011) reported similar findings to those in Study III. They studied adolescents with a single ventricle, in whom the surgical palliation according to the Fontan method was performed in their early years, and these adolescents also expressed feelings of being resilient and strong.

In order to provide a conceptual definition of "health within illness", Moch (1998) described the following components: *an opportunity, increased meaningfulness, connectedness/relatedness and awareness of self*. She described illness as an opportunity to increase the meaningfulness of life through connectedness or relatedness with the environment and awareness of self during a state of compromised well-

being. When applying the concept to adolescents and adults living with a CHD or a surgically palliated univentricular heart, the first component, *an opportunity*, could be understood on a more abstract and existential level, as making the best out of the life one has. *Increased meaningfulness* follows the same theme by creating meaning and reasons for having the heart defect and what possible benefits can be drawn. One of the participants expressed it as "...I have got this heart defect because I can handle it..." (Study III). The third component, *connectedness/relatedness* could be understood as the actual experiences that increase meaningfulness, for example, communication with others or the relationship with health care professionals. The fourth component, *awareness of self*, could be understood as self-knowledge and increased control over one's own body, in line with Heggdal's concept of *body knowledge* (2003).

Body knowledge

Heggdal (2003) studied the process of learning to master a long-term illness over time, in terms of gradually developing mastery strategies in order to integrate the illness as a part of life. This process is centred on the concept of body knowledge and it is defined as *a basic psychosocial process for developing personal knowledge about one's own body, coping, health and well-being*. The body knowledge process is both a process of learning, coping and recovery, and a social process in the sense that a person is engaged in dialogue with others regarding the meaning of bodily changes, symptoms and their alleviation and how health can be promoted. A social context characterized by knowledge, along with understanding and hope, function as facilitating factors for health transitions (Halting & Heggdal, 2011; Heggdal, 2003).

The body-knowledge process was originally developed based on data from groups of adults with different non-congenital diseases (Heggdal, 2003). However, most parts of the body-knowledge process would be applicable to adolescents and young adults with a CHD. The biggest difference is the time span of the experience of disease, which will be longer within the CHD context, in which the mastery process starts during childhood and continues to develop through adolescence to adulthood. A well-rooted body knowledge could be another part of the explanation for why young people with a CHD experience good health and their knowledge of their bodies, including their capacities, has been demonstrated even in childhood (Bjorbækmo & Engelsrud, 2008).

For adolescents and adults with a CHD their body knowledge starts to grow during childhood, when an integration of their disease begins and the CHD becomes part of their normal state of being. Another group with many similarities is young people living with cystic fibrosis (CF) and for both these populations, recent advances in treatment have resulted in higher survival rates. Caizer and her co-workers (2008) have explored the idea of acceptance in adolescents with CF and found that those with a higher trust in their ability to live with the CF and to manage negative consequences presented lower scores in the areas of

anxiety, depression and functional disability. Therefore they argued that accepting the limitations and thereafter readjusting one's life goals may have a positive effect upon well-being in adolescents and young adults with CF. Although adolescents and young adults with a CHD experience limitations and obstacles throughout their lives, by the time they reach adolescence or young adulthood, they have strategies for handling different situations and they are well aware of what their bodies can perform. This can be seen in the findings from Study III, in which the participants regarded themselves as having a normal life, just like their peers, having adapted to their physical limitations over the years. All participants experienced physical limitations. For example, one of the young women was only able to run at most 500 meters, but she did not consider this to be a limitation; rather, it was explained with the words ... *"I don't like sports anyway"....*

Existential maturity

It has been shown that the experience of illness can accelerate personal growth (Moch, 1989). Having a heart defect could be considered to be a life crisis, in that it is integrated in life and involves existential as well as future concerns, such as *how long will I live, how long will my heart function, do I need re-operations and what consequences will that have on life?* Through a life crisis, growth can be achieved. Living with a CHD can accelerate growth, as shown in the interview studies (Study I and III) in which the participants regarded themselves as more mature than other people and reported that the CHD had forced them to face existential concerns early in life. This was interpreted as feelings of suffering (Study I) and as a development of an existential maturity (Study III).

Other studies have indicated the related phenomenon of post-traumatic growth; for example, Engvall and co-workers (2011) reported that adolescents and young adults who had received a cancer diagnosis in childhood experienced personal growth due to their experiences of going through cancer. Post-traumatic growth cannot be directly applied to the context of adolescents and adults with a CHD, given that they have lived with their CHD their whole lives; in this context, there is no *pre* and *post* in the same way as when a person born healthy gets a cancer diagnosis.

In order to put the experienced maturity and growth within this group into context, the term existential maturity is appropriate, interpreted in this thesis as the maturity that adolescents and adults with a CHD have developed as a consequence of directly experiencing the uncertainty and fragility of life. The awareness of having survived can be seen as strengthening the development of existential maturity in these adolescents and adults. Early in life they become aware that life cannot be taken for granted, and so do their parents. Their whole childhood is to some extent connected with the uncertainty of life and yet at the same time it is evidence of their own vitality; having survived is a great achievement that gives them an inner strength.

Being committed to life

One of the most surprising findings within this thesis was the ability of the adolescents and young adults with a univentricular heart to interpret their experiences and their view of themselves in such a positive light (Study III). They expressed a deeply rooted happiness over “being me” and over being born with a heart defect. It could be speculated that this exceptional happiness is merely a strategy for coping with the disease, suggested by Zahmacioglu and co-workers (2011). They investigated the psychological framework of adolescents with a surgically palliated univentricular heart and also reported that these adolescents felt strong and resilient. They interpreted their findings in terms of psychological defence mechanisms and did not emphasize the existential aspects of living with a surgically palliated univentricular heart.

Casier and co-workers (2011) also measured acceptance and well-being in adolescents and young adults with CF and found that acceptance has positive effects on the adjustment to chronic illness, beyond the effects of socio-demographic and disease-related variables. Acceptance is not particularly studied within this thesis but the findings in Study III, of participants managing life positively and seeing possibilities in daily life, could be explained as coping or a defence mechanism. However, this cannot be the full explanation. One all-embracing aspect stands out: the awareness of actually having survived. The participants in Study III shared a commitment to life and were fully aware that their survival was due to improvements in surgical techniques, management and medical treatment. Their strong feeling of being committed to life helped them and strengthened them further in their ability to balance expectations and hurdles in life.

Sense of coherence

It has been suggested that growing up with a CHD can positively influence SOC due to the experiences associated with having a heart defect (Moons & Norekvål, 2006). The many experiences when growing up with a CHD could enhance comprehensibility, manageability and meaningfulness, which would positively influence SOC development. When investigating the life situation and SOC among adolescents and young adults with a univentricular heart (Study IV), scores in the higher range (75–91) were expected on the SOC scale. Most of the SOC scores were instead found in the moderate range (46–74), with only a few of the participants (n=3) scoring in the high range (75–91) or in the low range (13–45).

In the comparison of the SOC results with another Swedish study with healthy adolescents, the present study showed a slightly lower mean value: 61.3 points compared to 66 points (Myrin & Lagerström, 2008), but the comparison with healthy adolescents by Nio (2010) found instead that adolescents with a CHD scored higher than the healthy group. When comparing the SOC mean values with other published studies of adolescents with different types of CHD, Study IV revealed a higher mean value. For example, Neuner and co-workers (2011)

found lower scores, with a mean value of 48.5 in comparison to the mean value of 61.3 in Study IV. Nio (2010) also found lower scores in comparison with the data in Study IV, with a mean of 45.03 for all adolescents. Another aspect of the study by Nio (2010) was an examination of the influence of other factors on SOC, such as co-morbidities and numbers of hospitalizations. Co-morbidities were associated with a lower total SOC score and lower scores on the manageability dimension, but manageability scores were higher for those adolescents with six to ten hospitalizations than those with less than five. This phenomenon was then interpreted as an ability to manage the CHD and that the inevitable stress of a hospitalization could promote manageability. Even though the total SOC score in Study IV was in the moderate range, it was higher than the results of both Nio and Neuners.

Since the sample in Study IV is small, it is not possible to draw any definitive conclusions, but it could be noted that Study IV consists of adolescents and young adults with a univentricular heart, a condition requiring many surgical interventions with several hospitalizations over the years which could indicate a higher SOC as according to Nio.

The findings from Study III revealed that the participating adolescents and young adults experienced themselves as being unique and having an existential maturity as a result of their experiences. The strong sense of being committed to life due to the fact of having survived might be another part of the explanation. To explain the diversity between the divergent findings from different countries is beyond the scope of this thesis, but the phenomenon of differences between findings from different countries has been acknowledged elsewhere, for example in the review by Kovacs and colleagues (2005).

Antonovsky (1987) argued that SOC potentially influences a person's health, but that it could also be the other way around: health could have an impact on SOC (Eriksson & Lindström, 2006). In a study by Apers and co-workers (2012), it was investigated how SOC and the different domains of general and disease-specific health influenced one another. They found that SOC was a predictor of future perceived health over time, both of general health and disease-specific health, in individuals with a CHD. Neuner and co-workers (2011) also found SOC to be a predictor of QoL over time. Since the sample was small in Study IV, no advanced statistical analyses were conducted. However, the findings did show a tendency for both health and QoL to be associated with higher SOC scores, even if this was not statistically established.

Antonovsky (1987) suggested that the experiences from childhood and adolescence could build a foundation for the more definitive development of SOC in adulthood. SOC could be characterized as a personal way of thinking, being and acting with an inner trust, which leads to identifying and using the resources at one's disposal (Lindström & Eriksson, 2005). It can be seen as a lifelong process and is likely to vary during adolescence, due to different developmental stages and the transition into adulthood. To see the development as a lifelong pro-

cess might be an explanation for the total SOC score (in Study IV) standing in contrast to the interviewees' positive outlook on life (in Study III). SOC is stabilized throughout adulthood, but the experiences from childhood and adolescence foster a provisional ability to handle potential stressors later in life. Crucial is the focus on the resources that surround a person, which could reduce inner stress and allow a focus on the person's capabilities. Salutogenesis can be helpful in order to understand health, illness and disease and further be of interest in the development of health-promotion activities. Antonovsky (1987) saw SOC as a salutary resource rather than a coping strategy and its contribution could be part of the reason why those living with a CHD experience good health.

Homo capax

The observation that individuals living with severe conditions experience good health has been highlighted earlier by Lindsey (1996), who explored the experience of feeling healthy when living with a chronic condition. She stressed the need for a paradigm shift in nursing care in order to promote health and meaning, in other words, there has to be a movement away from the problem-oriented approach towards an approach focusing on capability despite the chronic condition. The inherent resources and capabilities of individuals with a CHD pervade the findings in this thesis. The work of Ricoeur could help in understanding the results presented.

According to Ricoeur (2011), people's capabilities are not all the same, but all people have a will, a sense of self and abilities. He viewed human beings as *homo capax* (the capable human), which is in parallel with the vulnerable human being. Growing up with a CHD includes suffering or having to go through many different challenging surgical procedures and examinations, long hospital stays involving operations, pain and medications of different kinds. Ricoeur (2011) underscored that suffering is not only pain or other severe symptoms; indeed, the utmost suffering is to be rejected as a capable human being – a person. People with a CHD have been through many life-challenging events and have experienced suffering but, in spite of their experiences, the participants in the present studies demonstrated vitality and a capability in mastering life and finding ways through it, which reflect Ricoeur's description of *homo capax*.

The concepts of person and *homo capax* imply a state that must be captured and strived for, a state built on relationships and partnership with other people (Ricoeur, 2011). Others who have a great significance for adolescents and young adults with a CHD, besides family and friends, are the health care professionals who have become a natural and important part of their lives over the years. These adolescents and young adults have been in contact with the health care setting for all their lives, undergoing extensive surgical procedures, interventions, examinations and regular follow-up visits. The health care professionals thus have a great opportunity to foster in young children with a CHD a sense of capability that will grow through adolescence and into adulthood.

Methodological considerations

The participants in Study I were interpreted as ambivalent in the way they perceived themselves and their CHD, and in their encounters with the health care setting. These findings differed from Study III, in which the participants viewed themselves as exceptional, strong and healthy. One explanation for the different perceptions of health and of themselves may have been the time that elapsed between the two studies. We know that health care professionals are of great importance for people living with a CHD and the participants in Study I were operated in another medical era than the participants in Study III. During these years, the treatment and management of a CHD has advanced, including significant developments in paediatric heart surgery, which may partly explain the diversity in the findings across the different study periods.

Moreover, the prognosis for long-term outcomes in children with a complex heart defect, such as a univentricular heart, has been established during these years. Since the participants in Study I were older, they had had their surgeries at a time when a univentricular heart was considered an inoperable condition. The remarkable medical progress during these years could have influenced the professionals in the way they interact with their patients, because they had no insight at first into long-term outcomes. Adolescents and adults living with a surgically palliated univentricular heart belong to the first generation facing adulthood, and there are still many questions to explore further.

Another diversity found in the results is the calculations of the EQ-5D data (Study II) compared to the interviews in Study III and the results from the questionnaire in Study IV. The first explanation could be the difference in sample size and in distribution. The GUCH registry contains a wider range of diagnoses of varying severity in comparison with the other studies. The result from the EQ-5D data revealed a difference between adults with a univentricular heart and the other diagnosis groups, indicating that this group perceived their health as lower and also experienced a higher occurrence of problems according to the EQ-5D dimensions. When further exploring the experiences of adolescents and young adults with a univentricular heart through in-depth interviews, it was unexpectedly found that this group experienced themselves as healthy and just like others. Study IV showed results in the same vein, although these respondents also experienced limitations and symptoms of various kinds, which they regarded as just a normal part of life, something they were used to. They perceived both their health and QoL to be good or even very good, and they were to a large extent satisfied with life.

Finally, the diversity in these findings could be a reflection of how the phenomenon was investigated (approached inductively or deductively) and the impact of the chosen design (explorative, descriptive or cross-sectional and retrospective), methods (qualitative and quantitative) and techniques (interviews) and questionnaires (EQ-5D, the study-specific Essence of Existence questionnaire and

the SOC scale). All these choices influence what questions are asked. Another observed diversity was the overrepresentation of women in the two interview studies, which both had a small sample size. In Study III, all of the potential participants meeting the inclusion criteria accepted and consented to participate. The overrepresentation of women in Study I might reflect that women are more likely to participate in interview studies. A gender difference was also found when analysing the registry data (Study II), namely that women were more likely to report symptoms, compared to men. However, it is already known that women in general have a tendency to report more symptoms than men. In another CHD population, Engelfriet and Mulder (2009) showed that a larger proportion of women with a CHD were symptomatic and more often had functional limitations due to the CHD than men did.

Sampling and participants

In qualitative research studies, the aim is mainly to discover meaning and not to generalize to a target population. According to Polit and Beck (2012), a critical step is to select the setting with high potential for information richness. They also addressed the issue of sample size, pointing out that a smaller number of participants can generate a large amount of data for analysis. In contrast, a quantitative research study seeks to select a sample that will make it possible to achieve statistical validity and to generalize from the results. Sample sizes are in general larger in quantitative studies and a general recommendation is to use the largest sample possible (Polit & Beck, 2012).

In Study I, the sample size of 20 participants was dictated by the simple fact that the number of potential participants in the chosen geographical areas at the time consisted of only 20 adults. The age range of 20–40 years was chosen with the intention of getting some variety in the sample but also based on the likelihood that, by that age, they would have reflected over their life situation. In Study I, the underlying intention was to get rich and informative interview data, which is why potential participants were contacted through the Swedish Childrens Heart Association. This choice could be viewed, on the one hand, as a positive strategy of getting individuals who could be expected to have reflected more over their situation. This would probably also include having discussed the consequences of having a heart defect with others in the same situation. On the other hand, it could be viewed negatively, since it excludes those who are not members of the selected organization. It may be that people who join an association are mainly those with more negative consequences of their heart defect.

The Swedish Childrens Heart Association started originally as an association for parents who had a child with a CHD. Such parents joined the association mainly in order to get support from others while bringing up their child or when their child died after complications due to the heart defect. The majority of the adolescents and adults in this association are members due to their parents' choice many years in the past. By the time they reach adolescence or adulthood

it has probably become a normal part of their lives; they have grown up with the association through different camps and meetings with their peers. They are members regardless of the consequences of their heart defect and, during the interviews (Study I), they all mentioned being connected to others with a CHD as a great asset in their lives. They found it beneficial and empowering to meet others in the same situation and they had shared many fun moments together. It is impossible to know whether divergent results would have been obtained by interviewing other adults. Nonetheless, it is worthy of note that other qualitative interview studies in the same period showed similar results (Claessens, et al., 2005; Tong, et al., 1998).

In qualitative studies there are no fixed rules for sample size. The key issue is to generate enough in-depth data to illuminate categories, patterns and other dimensions of the phenomenon under study (Polit & Beck, 2012). In three of the studies (Study I, III and IV), a purposive sampling approach was used, which in Study III and IV involved the selection of special or unique cases. These two studies have their origin in the original child survey from 1995, which guided the present sampling. All those who had been operated according to the Fontan or the TCPC procedure before 1995, at one of the two centres in Sweden performing heart surgery for children, were invited to participate in Study IV, now as adolescents and young adults. The child survey in 1995 was a collaboration project between two centres and, for the interview study (Study III), all potential and available adolescents and adults at the collaborating centre were invited to participate. In Study IV, all former participants in the child survey were asked to fill out the two questionnaires.

The one guiding principle in selecting the sample within a phenomenological study is that all participants must have experienced the phenomenon and must be able to articulate these experiences (Polit & Beck, 2012). A small sample size can always be seen as a limitation, but it is important to bear in mind the distinction between qualitative and quantitative sampling methods. In qualitative studies, the sample size is usually small, non-random and intensively studied. The sample was small in Study III and IV, however, it consisted of the total population of patients operated in that period (before 1995) from the two collaborating centres. Of the 34 available participants, 20 participated (58.8% response rate), which is considered a good response rate.

In Study II, a consecutive sample was obtained, consisting of all adult patients (n=1435) included in the GUCH registry at the GUCH clinic in Gothenburg. One obvious limitation is that it only included adults from one single GUCH centre. However, during the planning of the study, the inclusion of other centres was discussed. No other analysis of the EQ-5D data from the registry has been made and this study could thus be seen as a base for further multicentre studies. In order to describe the population accurately, the original diagnosis categories in the GUCH registry were used. This was also problematic, since the groups were

different in size as a consequence of congenital heart defects occurring at different developmental stages during fetal evolution. Therefore, some of the statistical analysis has to be interpreted with caution. An alternative way to describe the population would have been to use the classification of Task Force 1 of the 32nd Bethesda conference (Warnes, et al., 2001), but the original classification, according to the different heart defects, was considered to provide a more detailed and descriptive overview of the population and therefore to be more relevant.

Validity and reliability

Validity refers to whether an instrument measures what it is intended to measure, while reliability refers to the degree of consistency or dependability with which an instrument measures an attribute. Objectivity in quantitative research is also important and objectivity of the data refers to when two independent researchers will produce similar judgements or conclusions (Polit & Beck, 2010).

Rigour and trustworthiness in qualitative research

Lincoln and Guba (1985) have argued that trustworthiness is analogous to validity and reliability within quantitative research. Within this term, they outlined four criteria that are necessary to develop trustworthiness: *Credibility*, *Dependability*, *Confirmability* and *Transferability*. These four criteria are highly connected to each other. *Credibility* refers to confidence in the truth of the data and the interpretation of it, which is seen as an overriding goal of qualitative research. According to Lincoln and Guba it involves two aspects: conducting the study in a way that enhances the credibility of the findings and taking steps to demonstrate it. In order to enhance credibility during the interviews (Study I and III), the statements were followed up by additional questions. In addition, a detailed description of the interviewees and the method used was produced. The interpretation was based on the original source, the interview text, which is illustrated by citations exemplifying the subthemes and themes. In addition, credibility was strengthened by the richness of the data from the in-depth interviews.

Dependability refers to stability of data over time and conditions if the study were replicated in the same (or similar) context and by the same researcher. Dependability is closely related to credibility, just as validity is to reliability in quantitative research (Polit & Beck, 2012). *Confirmability* refers to objectivity and congruence between two or more independent observers about the accuracy of the data and its relevance and meaning. The criterion is concerned with the data genuinely representing the information provided by the participants. The findings must represent the participant's voice. Confirmability was established in the present studies through the participation of the authors and strengthened by an ongoing interpretative process in collaboration between them. Two of the researchers in Study I and three of them in Study III participated through the entire analysis – from the naïve understanding through the structural analyses and the interpreted whole. *Transferability* refers to the potential for extrapolation, in other

words, to what extent the findings can be applied to other settings or groups. This is established through sufficiently descriptive data such that the readers could evaluate the applicability to other contexts. Some of the findings in both Study I and III could probably be applied in contexts where a serious disease is present at birth.

Validity and reliability in quantitative research

The three quantitative measurements used in this thesis – EQ-5D (Study II), the SOC scale (Study IV) and the study-specific Essence of Existence questionnaire (Study IV) – are all well-established questionnaires. Both validity and reliability has been widely tested for EQ-5D and it has been proven that EQ-5D has both good validity (Brazier, Jones, & Kind, 1993) and reliability (van Agt, Essink-Bot, Krabbe et al., 1994). This is also the case for the SOC scale (Langius, et al., 1992) and for the original Essence of Existence questionnaire (Lindström, 1994; Lindström & Eriksson, 1993a).

Other forms of validity are relevant in Study IV. *Face validity* is a subjective assessment concerning whether the items in a questionnaire appear to be measuring what they claim to measure. *Content validity* refers to whether the items of a scale include all aspects of the attribute to be measured and to what extent the questionnaire reflects the specific domain of interest (Bowling, 2005). These two validity types were tested in Study IV. A limitation with these validity tests is that they can only be seen as a preliminary step in evaluating an instrument or questionnaire.

Advantages and disadvantages with the questionnaires used

The advantages and disadvantages with the questionnaires are the following: EQ-5D (Study II) is a short, generic instrument that is well used and validated (Brooks, 1996). The main argument for using it is that it is an easy-to-complete self-assessment tool that can be used to calculate quality-adjusted life years (QALY) when doing cost-effectiveness analysis (Brooks, 1996; The EuroQol Group, 1990). It is well suited as a complement to other disease-specific instruments, as originally intended (Brooks, 1996), and it has been shown to discriminate between different groups (van Agt, et al., 1994). The disadvantages are that EQ-5D is a rather blunt questionnaire for capturing health problems and, when using it within a particular group (such as adults with a CHD in Study II) and, in order to obtain richer data, the addition of a disease-specific instrument would have been beneficial. However, another benefit with using EQ-5D in Study II is that it provides an overview of the studied population's health status and how they perceived their health state.

The Essence of Existence questionnaire (Study IV) captures relevant aspects of life, including physical, social and mental components. The disadvantage of using a modified version of an instrument is that it was not thoroughly tested for relia-

bility and validity. In this case the intention was to measure health and QoL in the target population, with the inclusion of their life situation. The study-specific questionnaire is designed to be disease specific and is an adaptation of the original Essence of Existence questionnaire. It would be justifiable to use it within the setting of CHD, given that the questions added are of relevance for all groups of people with a CHD. Even though the study-specific questionnaire is not a validated instrument for the measurement of QoL and health, it nonetheless provides a tool for learning more about this group's life situation, which therefore justifies its use.

The Sense of Coherence scale (Study IV) has been psychometrically evaluated in several studies (Langius, et al., 1992; Lindström & Eriksson, 2005) and has been found to have acceptable validity and reliability. A Swedish study investigated whether the SOC construct is applicable to adolescents (Hagquist & Andrich, 2004) and in a Rasch analysis they found that it discriminated adolescents from the general population. They further concluded that the results were consistent with Antonovsky's conception of the SOC scale as a measure of one global factor. When constructing the SOC scale, Antonovsky (1987) emphasized that the components constitute one factor and that he did not recommend scoring each one separately. The SOC scale is built on a theoretical model that explains successful coping faced with different stressors. In Study IV, the use of the SOC scale provides valuable information about these adolescents' and adults' perspective on life and how they manage these stressors.

CONCLUSIONS

The conclusions from this thesis are:

- Adults with a CHD can experience ambivalence about how they should perceive themselves, how to interact with the healthcare system and report their health status and whether to perceive their health to be good. They can be seen as “walking a fine line” to balance the aspects of health and disease that coexist in the life of a person with a CHD.
- Unreported symptoms and discomfort may occur even if the adult with a CHD reports himself or herself to be asymptomatic.
- A low health status was predicted by self-reported symptoms, low physical functioning, age and gender.
- A low general health perception was predicted by self-reported symptoms, a low physical function and age.
- Pain/discomfort and anxiety/depression were found to be prevalent health problems among adults with a CHD, but they report less pain/discomfort than a general population, which indicate adaptation.
- Despite many limitations in their everyday lives, adolescents and young adults living with a surgically palliated univentricular heart experience satisfaction with their lives and view themselves as exceptional, strong and healthy.
- There is no direct association between the severity of the heart defect and the experience of satisfaction with life.
- Adolescents and young adults with a CHD develop an existential maturity due to their unique experiences and this, along with the experienced happiness over “being me”, might promote a sense of coherence.
- The CHD becomes an integrated and normal part of life.
- The concept of “health within disease” (as opposed to “health within illness”) emerges and seems to be more useful when considering adolescents and adults with a CHD.

IMPLICATIONS FOR CARE

In the care context of adolescents and adults with a CHD it is important to acknowledge that many of them experience themselves as healthy and living a normal life, just like their healthy peers. However, these young people have been through many challenging episodes that have influenced them and their families. Adolescents and young adults with a CHD cannot all be treated according to the same regime because one heart defect can differ from another, not only anatomically but also physiologically, as well as in the diverse surgical repair procedures that are used. All these different factors affect daily life. It is important to provide care based on the practical guidelines from the American College of Cardiology/American Heart Association (Warnes et al., 2008) and the European Society of Cardiology Working Group on Grown Up Congenital Heart Disease (Baumgarten et al., 2010).

What this indicates is the importance of creating a *person-centred care*. A person-centred care approach is initiated by the health professionals through active listening to the patient's story. To listen means being focused and being present, but also acknowledging the patient as an active partner with resources and self-esteem. By providing every patient with a contact nurse, continuity could be established, which could facilitate seeing the patient as an active partner. The health professional resources can be seen as assets to the patient, including being the patient's medical advisor, providing guidance in aspects of the CHD. The patient is seen as a capable person, not only in relation to the type of heart defect, but as a partner and an expert in what it means to live with the CHD. Health professionals need to take into consideration these adolescents' and young adults' extensive experience of being in a health care context and living with the CHD. The follow-up by the health care professionals has become a natural element in their lives since childhood and, for many of them, it continues throughout the whole lifespan; their care has to be characterized by lifetime management, in which other aspects of life than medical problems are also considered.

FUTURE RESEARCH

The results from this thesis suggest the following avenues for further research:

- Develop and validate a disease-specific quality of life instrument for adults with CHD, which should be used in complement with the EQ-5D instrument. The next step is to implement and evaluate the instrument into the care of adults with CHD combined with a tailored and person centered care/health plan.
- Follow-up of quality of life and health status on a regular basis and provide feedback at every measurement.
- Implement and evaluate a useful symptom assessment instrument in order to systematically evaluate the occurrence of symptoms more in detail.
- Since a CHD affects the whole family it is also of importance to highlight the partner perspective or even the parental perspective, in order to provide support.
- Evaluate the influence of other factors on sense of coherence in individuals living with CHD.
- Create a GUCH - heart school with a person-centred care approach, as part of an interventional study, with the main goal to increase knowledge and to relieve symptoms. This will be performed in the setting of a focus group in which participants can meet others with CHD.

SVENSK SAMMANFATTNING

Denna avhandling bidrar till förnyad kunskap och en ökad förståelse om hur det är att vara ungdom/ung vuxen och leva med ett medfött hjärtfel, med särskilt fokus på hur det är att leva med enkammarhjärta. Avhandlingen belyser unga människors erfarenheter och innefattar mätningar av hälsa och känsla av sammanhang beskrivet med hjälp av intervjuer och olika frågeformulär.

Avhandlingen omfattar fyra studier där *Studie 1* gav kunskap om hur vuxna med olika typer av medfödda hjärtfel upplever sin livssituation. Hur hälsa upplevs besvarades inte av denna studie, vilket därför ledde till *Studie 2*. Här besvarades frågan hur vuxna med medfödda hjärtfel rapporterar sitt hälsostatus och skattar sin generella hälsa med hjälp av hälsoskattningsinstrumentet EQ-5D. I denna studie framkom att vuxna med enkammarhjärta rapporterade en stor andel hälsoproblem. I *Studie 3* efterfrågades därför upplevelsen av att leva med ett opererat enkammarhjärta vilket gav en djupare förståelse kring vad det innebär att leva med ett enkammarhjärta. I *Studie 4* utforskades olika dimensioner av livet närmare, däribland dessa personers känsla av sammanhang, samt upplevelse av hälsa och livskvalitet vilket gav en kompletterande bild av hur livet med ett enkammarhjärta kan vara.

I avhandlingen har olika metoder och tekniker använts i de olika studierna. *Studie I* (sex vuxna med medfött hjärtfel) och *Studie III* (sju ungdomar/unga vuxna med enkammarhjärta) innefattar intervjuer analyserade med en kvalitativ tolkande metod (Fenomenologisk-hermeneutisk metod). *Studie II* (1435 patienter inkluderade i GUCH-registret, vilket är ett register för vuxna med medfödda hjärtfel) innefattar kvantitativ metodik där olika statistiska analyser har använts för att mäta hälsostatus med frågeformuläret EQ-5D, inklusive mätning av personernas skattning av sin generella hälsa med EQ-VAS. *Studie IV* (34 ungdomar/unga vuxna med enkammarhjärta) innefattar en kombinerad kvantitativ och kvalitativ metodik, där det studiespecifika frågeformuläret "Essence of Existence" har använts och där svaren analyserats med deskriptiv statistik kompletterat med innehållsanalys av öppna frågor. Även dessa personers känsla av sammanhang mättes med Sense of Coherence (SOC) skalan.

I *Studie I* framkom huvudtemat ambivalens i synen på sig själv, sin sjukdom och till sjukvården. Personerna upplevde att de fick gå en slags balansgång i hanteringen av sjukdomen och förhållandet till sjukvården. I *studie II* framkom att vuxna med medfödda hjärtfel skattar sin hälsa högt. Det som framförallt påverkade hälsostatus var förekomst av självrapporterade symtom, högre ålder, lägre funktionsstatus och att vara kvinna. Vad som påverkade den generella hälsan var även här förekomst av självrapporterade symtom, lägre funktionsstatus och högre ålder. Vissa diagnosgrupper skattade lägre hälsa, bland annat patienter med enkammarhjärta och kvinnor oavsett diagnosgrupp rapporterade mer pro-

blem än män. Det framkom även att de patienter som inte spontant rapporterar om sina symtom vid läkarbesöket ändå angav problem då de besvarade formuläret EQ-5D. Det visar att de patienter som uppfattas asymtomatiska även kan ha symtom. I *studie III* framkom huvudtemat "Att vara speciell, stark och frisk". Ungdomar och unga vuxna med enkammarhjärta upplever ett slags "åtagande till livet" vilket leder till att de fokuserar på möjligheterna istället för de svårigheter de möter. Det resulterar även i en glädje att vara den de är. I *studie IV* framkom hur ungdomar och unga vuxna med opererat enkammarhjärta upplever begränsningar i sin vardag med återkommande symtom, men att många samtidigt är nöjda och känner en tillfredsställelse med de flesta dimensioner i livet.

Avhandlingen visar att de flesta ungdomar och unga vuxna med medfödda hjärtfel upplever god hälsa och känner sig i stort som friska jämnåriga. Symtom är vanligt förekommande men upplevs inte alltid som besvär. Att vara född med medfött hjärtfel innebär att man inte känner till något annat liv och hjärtfelet blir en naturlig del av livet. Det kan innebära att den enskilde anpassar sig till symtom, vilket gör att dessa inte alltid uppfattas som något onormalt. Avhandlingens resultat kan ge en ökad förståelse för hur det är att leva med ett medfött hjärtfel/samt ett enkammarhjärta och vilka implikationer det kan ha i det fortsatta livet. Det är av betydelse att en personcentrerad vård erbjuds dessa patienter eftersom ett medfött hjärtfel inte är det andra likt och två personer med samma typ av hjärtfel kan uppleva det på olika sätt. Vården bör därför anpassas sig till vars och ens resurser och förutsättningar och inte enbart utgå från olika typer av hjärtfel. En fördjupad kunskap kan leda till ett mer individbaserat omhändertagande samt anpassade stödåtgärder tex. i form av riktade interventioner.

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