Health-related quality of life and everyday life for children with right ventricular outflow tract anomalies. Perspectives from children and parents.

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Perspectives from children and parents

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Health-related quality of life and everyday life for children with right ventricular outflow tract anomalies

Perspectives from children and parents

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LUND
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DOCTORAL DISSERTATION

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Abstract:

Background: Children with right ventricular outflow tract (RVOT) anomalies typically undergo surgical repair during their first months of life and thereafter they will need one or more reoperations through their life.

Aims: The overall aim was to describe health-related quality of life (HRQoL) for children with congenital heart disease (CHD) with a special focus on children with RVOT anomalies and to explore children's and their parents' experiences of everyday life during assessment that could result in a new heart surgery.

Methods: Two quantitative studies with HRQoL assessment for 337 children with CHD and 97 children with RVOT anomalies were performed. Another two qualitative interview studies were conducted, in which nine children with RVOT anomalies and their parents were interviewed at three time points (at start of the preoperative assessment, at the assessment decision and around 11 months after the decision).

Results: Children with three or more surgeries had lower HRQoL than children with fewer heart surgeries. Children with RVOT anomalies rated their lowest HRQoL for cognitive function, as did their parents. The agreement for HRQoL between children with RVOT anomalies and their parents was strong for 13 of 22 items. The thematic analysis of the interviews with children resulted in three themes and eight subthemes: *Me and my heart disease* explores the children's experiences of their heart disease; *Being me* explores their sense of self; *Being placed in someone else's hands* describes how the assessment was a safety net for the child, at least until the decision for heart surgery. The interviews with parents resulted in five themes: *The heart surgery keeps my child alive* illuminates' parents' experiences related to the assessment: *Everyday struggles* illuminates the struggles parents had to face on behalf of their child in everyday life; *Unconditional love, Trust in life* and *Togetherness* illuminates the ways in which the parents gained inner strength in their everyday lives.

Conclusion: By inviting both the child and their parents to participate in the child's care, individually tailored support can be identified and given.

Key words: HRQoL, everyday life, children, parents, CHD, RVOT anomalies, heart surgery, thematic analysis

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May your choices reflect your hopes and not your fears

Nelson Mandela

This thesis is dedicated to the children and their parents who so generously shared their thoughts and feelings.

Table of Contents

Abstract

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Populärvetenskaplig sammanfattning

Det föds 750–1000 barn/år med medfödda hjärtfel i Sverige. Variationen är stor när det gäller svårighetsgraden av hjärtfel, allt från enkla till medelsvåra till komplicerade. En tredjedel av dessa barn behöver behandlas med hjärtoperationer eller kateterbehandling. En grupp av barn som tidigt i livet behöver hjärtopereras och som även senare i livet vanligtvis behöver genomgå ytterligare hjärtoperationer är de med förträngning av höger kammares utflöde. De behöver också livslång uppföljning med utredningar bestående av ultraljud, magnetröntgen och arbetsprov för att bedöma hjärtats funktion och behovet av ytterligare operationer. Det har visat sig i tidigare studier att barn som behöver flera hjärtoperationer har en lägre hälsorelaterad livskvalitet och att deras föräldrar upplever mer stress och ångest än föräldrar till barn som behöver annan kirurgisk behandling.

Denna avhandling syftar till att beskriva hälsorelaterad livskvalitet hos barn med medfött hjärtfel, med fokus på barn med förträngning i höger kammares utflöde och barnens föräldrar. Avhandlingen syftar även till att belysa såväl barnens som föräldrarnas upplevelser av barnens utredning inför en eventuell ny operation och hur de upplever det dagliga livet.

Avhandlingen bygger på två kvantitativa och två kvalitativa studier. Studie 1 är en registerstudie med 337 barn i åldrarna 9–17 år med olika typer av medfödda hjärtfel och som besvarat det hälsorelaterade livskvalitetsformuläret DCGM-12. Resultatet visade att de barn som opererats tre eller flera gånger hade lägst hälsorelaterad livskvalitet. Det var också dessa barn vars hälsorelaterad livskvalitet var jämförbar med barn med andra kroniska sjukdomar (astma, epilepsi, diabetes, cerebral pares, artrit och cystisk fibros). Däremot hade de barn som genomgått färre än tre operationer eller ingen alls och barn med enkammarhjärta, bättre hälsorelaterad livskvalitet än barn med andra kroniska sjukdomar. Majoriteten av barnen gick i vanlig skola (88 %). Det var barn som opererats tre eller fler gånger samt barn med enkammarhjärta som hade behov av mer hjälp i skolan. En majoritet av barnen (87 %) skattade sin fysiska kapacitet som jämförbar med jämnårigas.

I studie 2 besvarade 97 barn i åldrarna 8–18 år med hjärtfel med förträngning av höger kammares utflöde och deras föräldrar tre hälsorelaterade livskvalitetsformulär, PedsQL Hjärtmodul, PedsQL 4.0 och DCGM-12. Resultatet visade att både barn och föräldrar skattade lägst hälsorelaterad livskvalitet när det gällde barnens kognitiva funktion. För mer än hälften av frågorna i PedsQL Hjärtmodul var överenstämmelsen stark mellan hur barn och föräldrar skattade barnens hälsorelaterade livskvalitet, även

för de yngre barnen i åldrarna 8–12 år. Nästan hälften av barnen skattade att de *nästan alltid* eller *ofta* hade problem med andfåddhet vid träning och en mindre andel barn (22 %) behövde *nästan alltid* eller *ofta* vila mer än sina vänner. Det var också en mindre andel av barnen (10 %) som ansåg att deras hjärtfel orsakade dem olika typer av problem eller svåra känslor.

I studie 3 intervjuades nio barn i åldrarna 9–17 år med hjärtfel med förträngning i höger kammares utflöde vid tre tillfällen under drygt ett års tid. Intervjuerna gjordes under och efter en utredning avseende behovet av en ny hjärtoperation. Av de nio barnen var det fyra som behövde en ny hjärtoperation. Resultatet från intervjuerna visade att även om barnen kände av sitt hjärtfel, oftast genom trötthet i olika grad, hade de anpassat sig och de hade ibland svårt att berätta hur de påverkades av sitt hjärtfel eftersom det blivit en så naturlig del av deras liv. De hade också vant sig vid tanken på att någon gång i framtiden behöva göra en ny hjärtoperation, ibland till den grad, att de hade svårt att berätta om hur det påverkade deras liv. När de beskrev sig själva var det i positiva termer som *glad* och *självständig* och deras berättelser visade att de också levde ett meningsfullt liv där inte hjärtfelet dominerade. Barnen tyckte det var ok att genomgå en utredning och kände sig trygga med det. Rädslan för operationen och vad som skulle kunna hända kom först när de fick veta beslutet om operation. För en del barn uppstod det missförstånd i kommunikation och vid den information de fick i samband med beslutet om operation som ledde ibland till svåra situationer och mer oro för de barnen.

I studie 4 intervjuades barnens föräldrar (de blev intervjuade vid samma tillfällen som deras barn i tredje studien). Resultatet visade att föräldrarna upplevde att det kändes tryggt att barnen utreddes även om svåra minnen av barnens tidigare hjärtoperationer väcktes till liv hos dem. De behövde hantera en oro för sina barn som de beskrev som mer än vanlig föräldraoro och de behövde också strida för sina barns rätt i skolan. Föräldrarna upplevde att de behövde vara starka för både sig själva och sina barn och att de hade lärt sig navigera genom svårigheter och utmaningar. De hämtade egen inre kraft genom det speciella kärleksband de kände till sitt ”hjärtbarn”, som skapades i en tid när de riskerade att mista sitt nyfödda barn. De hämtade också kraft genom att skapa sociala nätverk bestående av familj och vänner, genom sin tro samt deras stora tillit till vården och personalen de lärt känna där.

Sammanfattningsvis så visar resultatet på upplevelser av stor tacksamhet men samtidigt visar det också på att det finns förbättringsområden i vården att arbeta vidare med. Genom att mäta barnens hälsorelaterade livskvalitet kan eventuella problemområden upptäckas tidigt och stödåtgärder kan sättas in. De missförstånd i kommunikationen som uppstod kan förebyggas genom att involvera och bjuda in barn och föräldrar till ökad delaktighet i barnets vård. Många av de svårigheter som barn och föräldrar upplevde i samband med utredning och hjärtoperation kan därigenom förhoppningsvis undvikas för barn och deras familjer. Genom att använda ett barncentrerat perspektiv i vården underlättas ökad delaktighet och då kan ett mer riktat stöd ges till varje enskilt barn och dess föräldrar.

List of Papers

Paper I

Svensson B, Idwall, E, Nilsson F, Liuba P. Health-related quality of life in children with surgery for CHD: a study from the Swedish National Registry for Congenital Heart Disease. *Cardiology in the Young* 2017; 27:333-343. doi: 10.1017/S1047951116000585.

Paper II

Svensson B, Idwall E, Nilsson F, Liuba P. Health-related quality of life in children with earlier surgical repair for right ventricular outflow tract anomalies and the agreement between children and parents. *Front Cardiovasc Med.* 2020; 7:66. doi: 10.3389/fcvm.2020.00066.

Paper III

Svensson B, Liuba P, Wennick A, Berghammer M. "The only thing I wonder is when I will have surgery again": everyday life for children with right ventricular outflow tract anomalies during assessment for heart surgery. *Cardiology in the Young* 2022:1-6. doi: 10.1017/S1047951122000907.

Paper IV

Svensson B, Liuba P, Wennick A, Berghammer M. "I dread the heart surgery but it keeps my child alive" - Experiences of parents of children with right ventricular outflow tract anomalies during the assessment for cardiac reoperation. *Congenital Heart Disease* 2023: 18:3, pp. 349-359. doi;10.32604/chd.2023.028391.

Abbreviations

ACHD	Adult congenital heart disease
ASD	Atrial septal defect
CAT	Common arterial trunk
CCC	Child-centred care
CHD	Congenital heart disease
DCGM-12	DISABKIDS Chronic Generic Module - short version
DCGM-37	DISABKIDS Chronic Generic Module - long version
DORV	Double outlet right ventricle
FCC	Family-centred care
HCP	Health care professional
HRQoL	Health-related quality of life
MRI	Magnetic resonance imaging
NYHA	New York Heart Association
PedsQL	Pediatric Quality of life Inventory
PVR	Pulmonary valve replacement
QoL	Quality of life
RVOT	Right ventricular outflow tract
SWEDCON	Swedish national registry for congenital heart disease
TA	Thematic analysis
TOF	Tetralogy of Fallot
UVH	Univentricular heart
VSD	Ventricular septal defect

Introduction

Children with CHD have varying health care needs depending on their heart disease and its severity. Those with severe RVOT anomalies typically undergo surgical repair during their first months of life and thereafter many of them will need one or more reoperations at some point in their lives. They must go through several multifaceted assessments during follow-up to assess the optimal time for reoperation and they may lack or have relatively vague symptoms that are difficult to interpret in these assessments. Determining the optimal time for reoperation is important, due to the risk of serious complications which can occur later in life, if the reoperation is delayed (Gatzoulis et al. 2000).

This continuous assessment that may culminate in a reoperation can affect these children in many ways. They already have a history of surgery and frequent hospital visits, often with experiences of painful procedures, such as injections and postoperative pain. To our knowledge there is a gap in the literature concerning how these children rate their HRQoL and how they experience the everyday life, with the knowledge that reoperation will be needed at an unknown time in the future.

Numerous studies have shown that HRQoL research is needed and that the increased research into HRQoL measurement and the qualitative studies of everyday life for children with CHD and their parents can provide important knowledge about children's well-being and the experiences of living with a CHD (Helm et al. 2017). Results from such research can in turn be used to improve the care of children with CHD (van der Watt et al. 2017). A previous study has also shown that the communication between children and health care professionals (HCPs) can be facilitated when HRQoL measurement is used (Varni et al. 2005).

The ambition of this thesis was to explore and improve the understanding of how children with RVOT anomalies experience their everyday life and how they rate their HRQoL. Additionally, it was important to explore how their parents experience everyday life, as well as how they rate their child's HRQoL. A child-centred care (CCC) approach with the child's interests and needs in focus enables both children and their parents to participate in the child's care. This is of great benefit to this group of children and could strengthen them in relation to their health care throughout life.

Background

Children with congenital heart disease

CHD is the most common congenital malformation and occurs in 9 per 1000 live births (Liu et al., 2019). In Sweden, 750–1000 children per year are born with CHD, according to the latest annual report from the Swedish national registry for congenital heart disease, SWEDCON (SWEDCON, 2021). Approximately one third of children with CHD need surgical or transcatheter intervention (SWEDCON 2023). There is a great variation in complexity of CHD, ranging from mild forms, such as an asymptomatic ventricular septal defect (VSD) without any need for treatment, to moderate-to-severity CHD malformations such as tetralogy of Fallot (TOF) with symptoms and a need for treatment, to the most complex forms, including hypoplastic left heart syndrome with severe symptoms and a need for repeated operations leading to univentricular circulation (Marino et al. 2012).

For children with mild CHD with a need for treatment, usually one single cardiac surgery or catheterization is sufficient, but children with complex CHD often need more than one surgery or catheterization. One group of children with CHD needing multiple reoperations throughout their lifetime is children with RVOT anomalies.

Children with RVOT anomalies

Of all children with CHD, approximately 20% belong to the heterogeneous group *children with RVOT anomalies* (Arunamata and Goldstein 2022). The most common cyanotic heart disease, TOF (7–10%), belongs to this group as well as double outlet right ventricle (DORV) and pulmonary stenosis (Skoglund et al. 2014). Common arterial trunk (CAT) may also be included in this group due to the need for early implantation and repeated need of valved conduits between the right ventricle and the pulmonary artery.

These children's first surgery is performed early in life and is the first step of a lifelong cardiac care (Arunamat and Goldstein 2022). Commonly they need reoperation with pulmonary valve replacement (PVR) later in life. Once a conduit is inserted, later reoperations are necessary because the conduits undergo degenerative changes and secondary worsening of the valve function. For that reason, optimising of the timepoint for reoperation is important, and delaying it may lead to late complications, including arrythmias, heart failure and death (Gatzoulis et al. 2000).

Thorough preoperative assessment, including clinical examination, echocardiography, magnetic resonance imaging (MRI), computed tomography (CT) and exercise test, needs to be performed whenever the indication for reoperation is suspected. MRI is recognized as an important assessment tool in the decision making for PVR (Geva et al. 2010), although the optimal timing of repeated MRI remains debatable (Mohamed et al. 2020).

Thus, children with RVOT anomalies will face a life journey including surgical and transcatheter interventions and hospital stays with assessments, probably knowing that they will need reoperations in the future but with no certainty about when these operations will be needed.

Health-related quality of life

The reasons for using HRQoL assessment in children with chronic illness in research and in comprehensive care are manifold. The assessment can provide useful information regarding the child's state of health and can be used to identify different levels of morbidity. It may also contribute to increased understanding of different aspects of illness from the children's perspective (Drotar 1998; Marino et al. 2010a). HRQoL, health and functional status are overlapping concepts but are not interchangeable with each other (Drotar 2004). Quality of life (QoL) is a broader concept that includes HRQoL: "QoL includes physical and psychological characteristics and limitations (physical health, social functioning, and psychological status) that, taken together, describe a child's ability to function in relevant situational contexts (eg, family, school, peer) and to derive personal satisfaction from doing so" (Drotar 2004).

HRQoL refers to QoL that relates to health issues and is defined as "the influence of a specific illness, medical therapy, or health services policy on the ability of patients to both function in and derive personal satisfaction from various physical, psychological, and social life contexts" (Costello et al. 2015). This definition of HRQoL is appropriate in this thesis, since it focuses both on the children's experiences of their own HRQoL and their parents' experiences of their child's HRQoL and the impact of the heart disease on various aspects of their lives.

Previously, the assessment of children's HRQoL relied on the parent's report based on the assumption that children lack the cognitive and verbal skills to report their own HRQoL (Upton et al. 2008). Other studies have shown that children from five years of age appear to be capable of reliably self-reporting when using an age-appropriate HRQoL questionnaire (Varni et al. 2007) suggesting that the child self-report should be considered as the standard assessment and the parent report as a complement. The only exceptions should be if the child is too ill or otherwise unable to provide a self-report (Varni et al. 2007).

HRQoL in children with CHD

Evaluation of HRQoL in children with CHD has become increasingly important due to a "paradigm shift in clinical research from short-term mortality prevention to long-term morbidity assessment" (Marino et al. 2010a). The advances in medical treatment for children with CHD have resulted in a significantly improved survival to adulthood (Marino et al. 2010a). However, the children who survive may suffer from morbidity related to the heart disease and surgical treatment. These morbidities may originate from neurodevelopmental dysfunction (O´Brien et al. 2013) or psychological maladjustment (Latal et al. 2009) and can be assumed to have a negative impact on the individual's HRQoL.

In line with this, previous research has shown that children with CHD have lower HRQoL than healthy children (Uzark et al. 2008) and that children with complex CHD had lower HRQoL than children with mild CHD (Marino et al. 2010a; Sand et al. 2013). On the other hand, there are also studies showing that children with CHD have rated a higher HRQoL than healthy controls (Aguilar-Alanis et al. 2021; Reinar et al. 2019) and that HRQoL was affected less than expected by different types of surgical techniques (Heusch et al. 2017).

Identified risk factors for impaired HRQoL are the need for future surgery, severe forms of heart disease (Marino et al. 2010a; Uzark et al. 2008) and neurodevelopmental impairment (Latal et al. 2009). Studies have shown that HRQoL increased six months after surgery compared to preoperative ratings, for children with correctable CHD, such as VSD and atrial septal defect (ASD) (Dai et al. 2023). Additionally, 8-year-old children who had undergone palliative surgical repair reported lower HRQoL than children who had undergone a complete repair (Derridj et al. 2022).

Very little is known about HRQoL for children with RVOT anomalies. One study was found about QoL for adults with RVOT anomalies, and the result showed that their perception of their health declined over time but only for those adults with no reinterventions (Skoglund et al. 2014). Children with TOF have shown comparable HRQoL to healthy children (Kwon et al. 2011) but also a predominance of worse psychosocial health than in healthy controls (Neal et al. 2015). Furthermore, children with TOF have an increased risk of impaired neurodevelopment (Bellinger et al. 2015) and compared to children with valvular pulmonary stenosis, defined as a mild residual disease, parents reported worse QoL for children with TOF in some domains (Bhatt et al. 2017). Children with CAT, another type of RVOT anomaly, also have lower HRQoL than children with mild CHD (O´Byrne et al. 2015).

Given the results of these studies focusing on children with diagnoses within the spectrum of RVOT anomalies, there is a need to further investigate HRQoL in this group of children.

Everyday life and chronic conditions

Everyday life can be affected by emotional, physical, and social challenges for children with chronic medical conditions who have a long experience of various kinds of medical issues (Fayed et al. 2014). A chronic medical condition is defined as a health problem that lasts three months or more, affects a child's normal daily activity and requires frequent hospital visits (Mokkink et al. 2008). With the paradigm shift to long-term morbidity assessment related to advanced diagnostics and treatment, resulting in more children with CHD surviving to adulthood, CHD should be considered as a chronic illness (Connor et al. 2010). Parallel to this, interest in everyday life research has grown in recent years (Svensson et al. 2020). The focus has shifted from survival to what it means to live with a CHD and how the CHD affects their daily activities (Dodge-Khatami 2016). In contrast to survey measurement with fixed-choice questions, face-to-face interviews can reveal other aspects of children's and parents' experience of living with CHD (Wei et al. 2015). Interviews may provide a more nuanced picture of their experiences (Jordan et al. 2014).

Everyday life for children with CHD

Previous research has shown that repeated surgeries during infancy and adolescence are a burden för children with CHD (Andreasen et al. 2014) and that children may be afraid of invasive procedures and feel uncertain about the future (Chong et al. 2018). Research has also shown that living with CHD can leave adolescents feeling limited in their physical activity and with a wish to fit in (Shearer et al. 2013).

Research on everyday life has also shown that adolescents with complex CHD lacked knowledge of their heart disease (Lee and Kim 2012) and that there is a need for disease-specific knowledge so the adolescents can "become a manager of CHD in their lives" (Burstrom et al. 2017). They lacked not only knowledge about the disease but also strategies for how to communicate about the disease to others (Birks et al. 2006). This need is also recognized as a call for action for HCPs to receive comprehensive training so they can support children with CHD (van der Watt et al. 2017). One review study showed that living with CHD involves a balancing act that generates both possibilities and struggles in everyday life (Svensson et al. 2020). Another study on adolescents with CHD who had undergone at least two surgeries, showed that they could balance their lives with CHD by situating the CHD in the foreground or in the background as it suited their needs (Shearer et al. 2013). This ambivalence, also shown in young adults, might lead them to hide their symptoms from HCPs and sometimes even from themselves (Berghammer et al. 2006).

Everyday life for parents of children with CHD

Previous research has shown that parents of children with CHD report more stress and anxiety than parents of healthy children or children with other diseases

(Golfenstein et al. 2017; Lawoko and Soares 2006; Wei et al. 2015), and that they could be in need of psychosocial interventions to strengthen their sense of empowerment and their ability to better act as their child's caregivers (Gramzlo et al. 2020). Parents of children with CHD face a lifetime of adjusting to the challenges of the disease, and especially stressful events may be follow-up visits or repeated interventions (Mussatto 2006).

The parents face challenges during hospital visits for their child's heart surgery and they try to maintain the essential elements of their daily lives (Kosta et al. 2015) even if they experience a major stress for instance during the waiting time before the surgery (Sjostrom-Strand and Terp 2019). Even if most mothers have a positive relationship with their infant with CHD, a considerable part of the mothers' struggle with bonding to their infant after the heart surgery (Jordan et al. 2014). Home care after their child's heart surgery also has a great impact on the parents' emotional and everyday life (Ni et al. 2019). As with studies using HRQoL measurement, studies that investigate experiences of everyday life often provide important learning on how to improve care based on an understanding of the children's and the parents' experiences (Harvey et al. 2013).

Rationale

Children with RVOT anomalies often have their surgical repair early in life. This is followed by a lifelong contact with cardiac healthcare which, among other aspects, consist of awareness that they may need future surgeries with an uncertain timing. Later in life they may have a need for a PVR and once a conduit is replaced, later reoperations are necessary. Previous studies have shown that children with CHD awaiting further surgery are a risk group for impaired HRQoL.

To the best of our knowledge, there are no previous studies of how children with RVOT anomalies and their parents experience this form of repeated assessment that may culminate in a heart surgery. Nor has a representative group of children with RVOT anomalies rated their HRQoL.

Through the assessment of HRQoL and everyday life studies, we intended to increase current understanding of these children's HRQoL and how they experienced their everyday life during the assessment that could end up in a reoperation. This understanding can pave the way for how HCPs could improve the care and provide the support that these children and their parents need.

Aims

The overall aim for this thesis was to describe HRQoL for children with CHD, with a focus on children with RVOT anomalies, and to explore children and their parents' experiences of everyday life during assessment that could culminate in a reoperation.

The specific aims of the studies were:

Paper I
To describe the relationship between the number of surgeries for CHD in children and HRQoL, NYHA, cognitive function, physical activity, and psychosocial ability using data from the SWEDCON registry. Another goal was to compare the HRQoL of children with CHD with a reference population of children with other chronic diseases.

Paper II
To comprehensively describe self-reported HRQoL in children with RVOT anomalies, to evaluate which questionnaire best identifies HRQoL problems and to assess the agreement between self-reported and parent-reported HRQoL.

Paper III
To explore how children diagnosed with RVOT anomalies experience their heart disease and their everyday life during the assessment and after the decision on whether to perform a new cardiac surgery.

Paper IV
To illuminate how parents of children with RVOT anomalies experienced their child's heart disease and their everyday life during the assessment and after the decision on whether to perform a reoperation.

Method

Study design

In this thesis, both quantitative methods (studies I and II) and qualitative methods (studies III and IV) were used (Table 1). Quantitative methods, using numerical data and statistical analysis generate broad data (Braun and Clarke 2013). Qualitative methods using interview material as data (Braun and Clarke 2013) answer questions that concern experiences from the perspective of the participants (Hammerberg et al. 2016). Quantitative and qualitative methods complement each other and enable a holistic view of these children's HRQoL and everyday life.

Study I, with a retrospective cross-sectional design, contributed by giving an overall picture of self-reported HRQoL for a large population of children with CHD. Study II, with a prospective cross-sectional design, contributed with self-reported and parent-reported HRQoL for the selected group of children with RVOT anomalies.

In studies III and IV, a longitudinal design with a constructionist approach was used, where interview data were collected at three time points to gain a deeper understanding of how everyday life is experienced by children with RVOT anomalies and their parents during and after an assessment for heart surgery.

Table 1. Study overview

Paper	Paper I	Paper II	Paper III	Paper IV
Design	Retrospective registry study	Prospective cross-sectional with descriptive data	Explorative inductive qualitative	Explorative inductive qualitative
Focus	HRQoL related to number of heart surgeries	HRQoL and the agreement between child self-reports and parent reports	How children experience everyday life	How parents experience everyday life
Datacollection	HRQoL questionnaire DCGM-12 cognitive function, psychosocial ability, physical activity, NYHA	HRQoL questionnaires DCGM-12 PedsQL Cardiac Module PedsQL 4.0	Individual interviews at three timepoints	Individual interviews at three timepoints
Study population	Children with CHD (n=337)	Children with RVOT anomalies and their parents (n=97)	Children with RVOT anomalies (n=9)	Parents of children with RVOT anomalies (n=9)
Analysis	Statistical	Statistical	Thematic	Reflexive thematic

CHD = congenital heart disease, DCGM-12 = DISABKIDS Chronic Generic Module - short version, HRQoL = health-related quality of life, PedsQL = Pediatric Quality of Life Inventory, RVOT = right ventricular outflow tract.

The Swedish context

In 1993, paediatric cardiac surgery in Sweden was centralized to two tertiary referral centres, at Skåne University Hospital in Lund and Queen Silvia Children's Hospital in Gothenburg (Lundström et al. 2000). In connection with the amendment of the Health Care Act (2017:30) in 2018, the previous system of national healthcare was replaced with national highly specialized care (The National Board of Health and Welfare 2023). Children with CHD are thus jointly cared for by the two tertiary referral centres and the paediatrics departments at the local hospitals throughout Sweden. In the capital of Sweden, the Karolinska University Hospital perform complete cardiac investigation including catheterization. Several of the larger paediatric departments perform non-invasive investigation such as MRI. On average, 450 children undergo cardiac surgery per year in Sweden. For children with RVOT anomalies, the preoperative assessment is performed whenever the indication for reoperation is suspected; the assessment mainly consists of clinical examinations, echocardiography, MRI, CT and exercise test. Evaluation of the possible need for reoperation is commonly performed by the local hospital in collaboration with the specialized paediatric cardiac centres. For elective procedures, all data are collected and discussed at a surgical meeting attended by paediatric cardiologists, cardiac surgeons and radiologists.

Study population

Study I

The sample consisted of 337 children between 9 and 17 years of age with CHD. Children attending the outpatient clinic at the Children's Heart Centre in Lund, Skåne University Hospital and answering the questionnaire DCGM-12 between 2009 and 2014 were included (Table 2). The completed questionnaires were retrieved from the SWEDCON registry.

Study II

The sample consisted of 97 children between 8 and 18 years of age with various diagnoses of RVOT anomalies (Table 2, 3) and was obtained from the SWEDCON registry (n=172) and from surgical records from the Children's Heart Centre in Lund (n=26). The search was performed in September 2015. Exclusion criteria were genetic disorders (Down syndrome, trisomy 18, chromosome disorder for chromosomes 4 and 13, Wolf Hirschhorn syndrome), heart surgery/intervention within three months, univentricular heart (UVH) and moving abroad after surgery. The response rate was 50% (n=97).

Table 2. Participant characteristics in the studies.

Characteristics	Paper I Children with CHD	Paper II Children with RVOT anomalies and their parents	Paper III Children with RVOT anomalies	Paper IV Parents of children with RVOT anomalies
Number of participants	337	97	9	9
Children				
Median age	14			
Mean age		12.9*	13.8	13.8*
Age range	9-17	8-18*	9-17	9-17*
Gender, girls N (%)	131 (39)	38 (39)*	6 (67)	6 (67)*
Parents				
Mothers N (%)		62 (64)		7 (78)
Fathers N (%)		16 (17)		2 (22)
Both N (%)		12 (12)		
Unspecified N (%)		7 (7)		
Diagnoses N (%)				
Aortic anomalies	39 (12)			
Aortic valve anomalies	43 (13)			
ASD	35 (10)			
AVSD	11 (3)			
CAT	6 (2)	1 (1)	1 (11)	1 (11)
CCTGA	2 (1)			
DORV		14 (14)	2 (22)	2 (22)
Mitral valve anomalies	17 (5)			
PAVSD		5 (5)		
PAVSD with MAPCA		9 (9)		
PDA	8 (2)			
Pulmonary valve anomalies	31 (9)		2 (22)	2 (22)
TGA	25 (7)			
TGA, PS			1 (11)	1 (11)
TOF	36 (11)	68 (70)	3 (33)	3 (33)
Tricuspid valve anomalies	2 (1)			
UVH	18 (5)			
VSD	58 (17)			
Others	6 (2)			
Number of surgeries and the UVH group N (%)				
0	140 (42)			
1	117 (35)	37 (38)	3 (33)	3 (33)
2	31 (9)	36 (37)	3 (33)	3 (33)
≥3	31 (9)	24 (25)	3 (33)	3 (33)
UVH**	18 (5)			

* Refers to the children. ** Children with UVH are not included in the number of surgeries. ASD = atrial septal defect, AVSD = atrioventricular septal defect, CAT = common arterial trunk, CCTGA = congenitally corrected transposition of the great arteries, DORV = double outlet right ventricle, MAPCA = major aorto pulmonary collateral arteries, PAVSD = pulmonary atresia with ventricular septal defect, PDA = persistent ductus arteriosus, PS = pulmonary stenosis, RVOT = right ventricular outflow tract, TGA = transposition of the great arteries, TOF = tetralogy of Fallot, UVH = univentricular heart, VSD = ventricular septal defect.

Table 3. Diagnoses (studies II–IV) within the group of RVOT anomalies

Right ventricle outflow tract anomalies
CAT (common arterial trunk)
DORV (double outlet right ventricle)
PAVSD (pulmonary atresia with ventricular septal defect)
PAVSD with MAPCA (PAVSD and major aorto pulmonary collateral arteries)
PS (pulmonary stenosis)
Pulmonary valve anomalies
TGA (transposition of the great arteries)
TOF (tetralogy of Fallot)

Studies III and IV

The samples consisted of children between 9 and 18 years of age with various diagnoses of RVOT anomalies (Table 2, 3) who had undergone previous surgery and an MRI to assess the need for PVR. They were recruited from two Swedish University Hospitals, one of which was a tertiary referral centre. Exclusion criteria were children unable to speak and understand the Swedish language or unable to participate in the interview situation due to cognitive impairment. A total of 15 children and their parents were invited to participate, of whom six parents declined participation.

Data collection

The data were collected from registry data (study I), generic questionnaires (studies I and II), disease-specific questionnaires (study II) and interviews (studies III and IV).

SWEDCON registry

Study I

SWEDCON is a registry for children and adults with congenital heart disease and consists of four parts: fetal heart diagnostics, paediatric cardiology, adult congenital heart disease (ACHD) and congenital heart surgery. The intention is to be able to follow up patients from childhood to adulthood and thus obtain as complete information as possible about the normal course of and treatment results for various congenital heart defects. The measurement of HRQoL for children with CHD has been added to the SWEDCON registry at the outpatient clinics since 2009 (SWEDCON 2023).

The paediatric part of the SWEDCON registry consists of individual-based data about medication, diagnosis, type and number of surgeries and catheter interventions, types of cardiac investigations and NYHA class. When HRQoL data for study I was collected, it consisted of DCGM-12, cognitive function, physical activity, and psychosocial ability. Cognitive function was measured by the child's need for extra support in school, and physical activity was measured by more or less

than three hours of physical activity per week (including activity that does not raise the pulse). Since 2014, physical activity is registered in a physiotherapy module, and since 2017, cognitive function is one of several domains in the HRQoL questionnaire PedsQL Cardiac Module (SWEDCON 2023). A validation study showed that data from the SWEDCON registry is reliable for use in research (Bodell et al. 2017).

DCGM-12

Studies I and II

DCGM-12 is a generic self-report questionnaire, validated for use in children from 8 to 17 years with chronic disease; it is a short form of DCGM-37, which was developed by the European DISABKIDS group (The European DISABKIDS group 2006). A generic measurement can be used to compare with other chronic conditions (Marino et al. 2010a). The DCGM-12 questionnaire includes 12 items measuring mental (4 items), social (4 items), and physical (4 items) impact on the individual's health condition (only 10 items are used when those concerning medication are excluded). The response for each item is graded using a five-point scale indicating frequency of behaviours or feelings as 1 = never, 2 = seldom, 3 = quite often, 4 = very often and 5 = always. A higher score (max 100) indicates a better HRQoL. Percentiles for self-perceived HRQoL are integrated in the questionnaire from a reference population of European children of the same age and gender with other chronic diseases. This enables a comparison with children with asthma, arthritis, dermatitis, diabetes, cerebral palsy, cystic fibrosis and epilepsy. The questionnaires are available in an age-appropriate self-report version and a proxy-report version for significant others (The European DISABKIDS group 2006).

PedsQL 4.0 and PedsQL Cardiac Module

Study II

PedsQL questionnaires are problem based and consist of self-report for children with age-appropriate questions and proxy-report for significant others. A five-point response scale is used (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem, 4 = almost always a problem). The items were reverse scored and linearly transformed to a 0 to 100 scale (0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0), so higher scores indicated better HRQoL (Uzark et al. 2003).

PedsQL 4.0 is a generic questionnaire and consists of 23 items in four domains: physical functioning (8 items), emotional functioning (5 items), social functioning (5 items), and school functioning (5 items). A physical health summary score can be generated by responses given in the domain physical functioning. To generate a psychosocial health summary score, the mean is computed as the sum of the items divided by the number of items in the domains emotional, social and school functioning (Varni et al. 2001). The PedsQL Cardiac Module is a disease-specific

questionnaire. A disease-specific questionnaire can detect differences between subgroups (Marino et al. 2010a). The PedsQL Cardiac Module has 27 items in six domains: heart problem (7 items), treatment (5 items), perceived physical appearance (3 items), treatment anxiety (4 items), cognitive problems (5 items), and communication (3 items) (Uzark et al. 2003).

Interviews

Studies III and IV

The interviews were performed individually based on an interview guide. An interview guide helps the interviewer to perform the interviews with different participants more systematically by defining in advance the issues to be explored (Patton 2015). The guide was developed based on a literature review focusing on the research aim and adapted as an age-appropriate version for children and an adult version for the parents. The semi- structured interview (interview guide approach) is the most common method of collecting qualitative data (Braun and Clarke 2013).

Central issues in the interview guide were (1) the children's and parents' experiences of everyday life during and after the period when the child went through an assessment and received a decision about whether heart surgery was necessary, (2) the experiences of the child's CHD and heart surgeries, (3) the impact of the child's CHD and heart surgeries on everyday life, and (4) thoughts about the child's leisure, schooldays and future. Interviewing children is different to interviewing adults but can successfully be done from as young as preschool age. By interviewing children, their own thoughts and interpretations are heard instead of only relying on an adult interpretation of the children's lives (Acker et al. 2003).

Children in this interview study were aged from 9 to17 years at the first interview. The youngest child (aged nine) was given the possibility to make drawings as well as giving verbal responses during the interview. All interviews, except for the first interview with one child, were conducted with the child alone. Three interviews per participant (in total 54 interviews) were conducted (Figure 1 and Table 4) and the interviews lasted from 47 to 129 minutes per child (median 101 minutes) and from 86 to 156 minutes per parent (median 135 minutes). The whole study lasted an average of 14.6 months for children who had undergone a reoperation and 15.4 months for those who did not need any reoperation at this assessment.

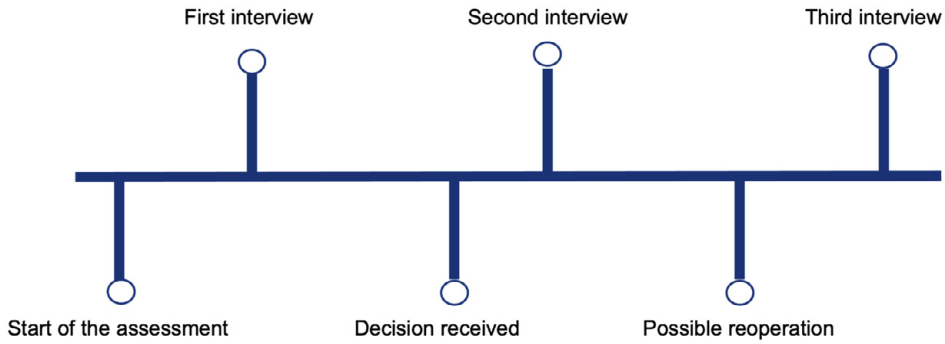

Figure 1. Timeline showing the three interviews during each child's assessment

Table 4. The main characteristics of the interviews (studies III and IV)

	First interview	Second interview (after the decision for or against surgery)	Third interview (children who needed surgery)	Third interview (children who did not need surgery)
Number of interviews (child+parent)	18	18	8	10
Time point (mean) for interviews	At the start of the assessment	3.9 months after interview 1	10.7 months after interview 2 (6.7 months after surgery)	11.5 months after interview 2
Setting	14 interviews were performed in the participants' homes except for two children and their parents who were interviewed in the hospital	16 interviews were performed in the participants' homes except for one child and one parent who were interviewed by telephone	All interviews were performed in the participants' homes	All interviews were performed in the participants' homes
Opening question	Child: What has your day in school been like? Parent: Can you tell me about your child (the childs name)?	The last time I was here…and now you have received a decision	The last time I was here…and now you/your child has gone through surgery. Can you tell me what it has been like?	A year has passed since last time I was here…how have you/your child been and what has happened?

Procedure

In study I, prior to collecting HRQoL data from the SWEDCON registry, formal approval from the steering committee for the SWEDCON registry was provided. Prior to study II, an information letter about the study was sent to the participant's local hospital.

In study II, the family's postal address was obtained from the child's medical journal. The three questionnaires were sent out by post, together with an information

letter and consent form. The information letter included instructions about how to complete the questionnaire and how to administer the questionnaire to the child. The children and their parents completed the questionnaires at home and returned them in the enclosed envelope. The families received a reminder call, giving them an opportunity to ask questions about the study, one to two weeks after receiving the study information. If no response was received after three to four weeks, a reminder letter was sent.

In studies III and IV, the children and their parents received oral and written information about the study from the two nurses working at the clinics at the two University Hospitals. After obtaining written informed consent from the parents and from children over 15 years of age, the first author contacted them to provide more information and to arrange a time and date for the first interview.

Data analysis

Study I

Statistical analyses were performed using SPSS version 21.0, and R version 3.1.2 (R Core Team 2014). The data are presented as median and 10^{th} to 90^{th} percentile. As these data were skewed, non-parametric tests were used. To assess differences, Kruskal-Wallis with Monte Carlo p values were based on 10000 samples and the Mann-Whitney exact U-test was used. Results with a p value < 0.05 were considered statistically significant. Bonferroni post-hoc tests were used to correct for multiple comparisons.

The DCGM-12 was transformed from a raw score to a range of 0–100 (The European DISABKIDS group 2006). Thus, a "standard raw score", calculated as ((point−12)/48) X 100 (if 12 questions were answered) or ((point−10)/40) X 100 (if 10 questions were answered). For each of the four groups with children with biventricular CHD and for the one group with children with UVH, the confidence interval for the expected quantiles was found by simulating 1000 replicates with as many observations as those present in a specific group. The 2.5^{th} and 97.5^{th} bootstrapped empirical percentiles for each expected quantile were then used to provide the confidence intervals for the expected pattern. The observed quantiles of the DCGM-12 score were also compared with the expected distribution using a Kolmogorov-Smirnov test. Internal consistency was assessed using Cronbach's alpha (studies I and II).

Study II

Statistical analyses were performed using SPSS version 23.0, and R (R core team R 2016). The data are presented as mean and standard deviation. Child and parent

agreement was measured at three score levels: total, domain and item level. At the item level in the PedsQL Cardiac Module, the children were divided into two age groups (8–12 and 13–18 years of age). A strong agreement between children and parent was defined as no significant difference in mean score combined with a large correlation assessed with the Wilcoxon test and Spearman correlation. For effect size (Cohen 1988), small effect size was defined as r = 0.10–0.29, medium as r = 0.30–0.49, and large as r = 0.50–1.0. To compare the total score in the child report in PedsQL Cardiac Module, PedsQL 4.0 and DCGM-12 a linear mixed effects model (nlme package, version 3.1-137) was used to account for repeated measures (Pinheiro et al. 2018). Post-hoc comparisons were made using the package multcomp version 1.4-8 (Holthorn et al. 2008), adjusting for multiplicity.

The DCGM-12 was transformed as described above in study I. In addition, the responses were transformed so that 5 (*always*) indicated better HRQoL and 1 (*never*) indicated lower HRQoL in items 3–8 and 11–12. To identify the items where the children reported the lowest HRQoL, the answers "*never"* or "*seldom"* and "*always"* or "*often"* were merged depending on the item's value range. In PedsQL Cardiac Module and PedsQL 4.0, the domain score was computed as the sum of items divided by the number of items answered. If more than 50% were missing, the domain sum was not computed (Varni et al. 2001). To identify the items children reported most problems with in the PedsQL Cardiac Module and PedsQL 4.0, the frequencies of 3 (*often a problem)* and 4 (*almost always a problem*) were merged and ranked. Dropout analysis was performed using Fisher's exact test concerning age, gender and diagnosis (TOF).

Studies III and IV

A thematic analysis (TA) approach was used to analyse the transcribed child interview data (study III) and a reflexive TA for the transcribed parent interviews data (study IV). The analyses were guided by the six phases of TA (Braun and Clarke 2006) (Table 5). An inductive approach was used at the semantic level and latent level in study III and at the latent level in study IV.

With the experience of conducting TA in study III, the analysis in study IV could be developed into what Braun and Clarke identified as reflexive TA, which is a further development of TA into three parts, coding reliability, codebook and reflexive approaches (Braun and Clarke 2019). They describe reflexive TA thus: "The themes are creative and interpretive stories of the data, produced at the intersection of the researcher's theoretical assumptions, their analytic resources and skill, and the data themselves" (Braun and Clarke 2019). In the first TA (study III), two authors coded independently, and the final themes were a result of this collaboration, with the first author (also the interviewer) having the main responsibility. In the second (study IV), reflexive TA was created by the first author alone, since a single coder is usual in

reflexive TA (Braun and Clarke 2022). The codes and the themes were discussed regularly with the other authors during the whole procedure.

Table 5. The phases of the thematic analysis as described by Braun and Clarke (2006, 2019)

Phase	Description of the process
1. Familiarizing with your data	Transcribing data, reading and re-reading the data, noting down initial ideas.
2. Generating initial codes	Coding interesting features of the data in a systematic fashion across the entire data set, collecting data relevant to each code.
3. Searching for / generating themes*	Collating codes into potential themes, gathering all data relevant to each theme.
4. Reviewing themes	Checking if the themes work in relation to the coded extracts and the entire data set, generating a thematic map of the analysis.
5. Defining and naming themes	Ongoing analysis to refine the specifics of each theme, and the overall story the analysis tells, generating clear definitions and names for each theme.
6. Producing the report	The final opportunity for analysis. Selection of vivid, compelling extract examples, final analysis of selected extracts, relating back from the analysis to the research question and literature.

* Used in reflexive thematic analysis

Conceptual framework

The conceptual framework chosen in this thesis consists of an interpretative paradigm and a child-centred perspective. A child-centred perspective approach is influenced by the interpretative theoretical orientation (Sommer et al. 2010), which is attuned with the interpretative paradigm. This approach is built on fundamental beliefs about the child perspective, for example, viewing children as social actors who can affect their surroundings.

Interpretative paradigm

A paradigm refers to beliefs and assumptions about the world and is more general than a theory (Malterud 2016). The overall paradigm in this thesis is an interpretative paradigm, which means that the emphasis is to understand the participants, both their HRQoL (studies I and II) and their everyday life experiences (studies III and IV). The direction in which the result can be used originates in the researcher's disciplinary domain (Malterud 2016). In this thesis the domain is nursing, which mean that the result is intended to contribute to improved care for children with CHD, especially RVOT anomalies, awaiting heart surgery. The combination of quantitative and qualitative methods was used in order to get a holistic picture of this group of children with RVOT anomalies. This is achieved by describing their HRQoL in a wider perspective and by illuminating their experiences of everyday life with a deep perspective.

A constructionist approach was used, meaning that the individual's worldview is constructed by various systems of meaning that we all exist in. There is not one

truth, but rather, the truth or knowledge changes when the worldview or discourse changes (Braun and Clarke 2013). A constructionist approach within an interpretative paradigm indicates that the interpretations are socially constructed (Crotty 2010). This means that the participants' experiences (studies III and IV) were used to construct realities obtained via interviews, which is taken-for-granted knowledge (Terry et al. 2017).

The interpretative paradigm includes both ontological and epistemological approaches. Subjectivity can be understood at an ontological level, which concerns how human beings, in this case the interviewed children and their parents, experience their world, and at an epistemological level, which concerns how the author describes and reflects upon these experiences (Malterud 2016). In reflexive TA, the researcher's subjectivity is a part of the analysis and is acknowledged in the reflexivity (Braun and Clarke 2022). The interviewer in studies III and IV has a long experience within paediatric cardiology and has as an extensive nursing experience of interacting with children and their parents in different care situations, both during inpatient and outpatient care.

Child-centred perspective

A child-centred perspective is an essential concept in this thesis, as the focus is on children's experiences. It should be noted that a child-centred perspective consists of two perspectives: a *child perspective* and the *children's perspective* (sometimes called the *child's perspective*), and these perspectives are represented throughout this thesis, from the study design to the results.

The *child perspective* is the adult voice about the child and is essential for the children's perspective, meaning that the adult must believe in the child's capacity in order to be able to see it. "Child perspectives direct adults' attention towards an understanding of children's perceptions, experiences and actions in the world" (Sommer et al. 2010). In this thesis, the child perspective is generated by the parent reports in study II and by the parent interviews in study IV. The intention with having individual interviews and analysing them separately was based on the importance of highlighting both the children's perspective and the child perspective.

In contrast the *children's perspective* indicates that the child's own voice expresses the experience. According to Sommer et al. (2010), "Children's perspectives represent children's experiences, perceptions and understanding in their life world." This perspective is elicited by child reports in studies I and II and by the individual interviews in study III, where the analysis separated the children's responses from their parents' responses.

The interest for the child-centred perspective has grown in Scandinavia but also in other parts of the world, and this perspective has become important both in research and in child-related professions (Sommer et al. 2010). A global foundation for this

focus is the United Nations Convention on the Rights of the Child (1989), which became Swedish law in 2020 (Government offices of Sweden 2018:1197). Another foundation is the Swedish government agency known as Barnombudsmannen (*the Child Ombudsman*) whose mission is to protect children's rights and interests. Finally, the Scandinavian welfare model contributes with its democratic values, where the family and the society share responsibility for fostering and caring for children. These societal foundations are some of the factors that have given the children the status of citizens and at the same time facilitated the growing interest in the child-centred perspective (Sommer et al. 2010). The children and parents who participated in these studies live their lives in this societal context.

Child-centred perspective in the care of children

Child-centred care (CCC) means that children's interests are in focus and can be seen as an extension of the concept of family-centred care (FCC) which is the most common model for nursing care (Carter et al. 2016). However, the perspective of the ill child is not so prominent in the FCC concept (Mikkelsen and Frederiksen 2011). Furthermore, there is no solid evidence that FCC is appropriate in child health care (Shields et al. 2012). In a previous review (Coyne et al. 2018), three concepts of centeredness– FCC, CCC and person-centred care (PCC) were compared. Both similarities and differences were found. The most important difference is that the focus in FCC is on the family as a unit, whereas in PCC and CCC the focus is on the individual level. Additionally, in CCC the perspective is on the individual child as an independent actor who belongs to a family, this is comparable to PCC, where the individual is an adult person (Coyne et al. 2018). The conceptual difference in centeredness needs to be considered in health care. For example, in neonatal care, FCC maybe the most appropriate concept to deliver the best care to the child and the parent, but for older children, CCC might be the best option (Coyene et al. 2018). A CCC approach gives the child the right to participate in health care matters and decisions about his or her care (Coyne et al. 2016).

Ethical considerations

The studies were approved by the Ethics Committee for Human Research at the Lund University (approval number 2011/749; 2014/66) and were conducted in accordance with the Declaration of Helsinki developed by the World Medical Association in 2013.

In studies II, III and IV an information letter and an age-appropriate information letter with informed consent for two caregivers and the child were sent by post (study II) or handed over by the nurses (studies III and IV). The fact that participation was voluntary and that participants could drop out at any time was emphasized, together with a declaration that the child's further treatment was not

affected in any way by non-participation or dropping out of the study. It was the child who decided whether to participate, so even if the parents wanted it, if the child hesitated or said no, they were excluded from the study. The interviewer was not involved in the children's care.

Only one parent was interviewed and the parent themselves were given the choice of which one it should be. All interviews took place in the children's homes, except that for two children and two parents the first interview took place in the hospital and for one child and one parent the second interview was by phone. Interviewing children in their home may balance to some extent the inherent power balance between the child and the adult interviewer, especially if the child is allowed to choose the place for the interview (Coad et al. 2015). In these studies, the interviewer asked in both the child's and the parent's presence where the child would like the interview to take place.

There are other circumstances to consider when entering the child's home, such as the other family members present at the interview and what impact that may have on the child. The interviewer is also a guest in the child's home (Coad et al. 2015). The interviews ended by asking whether the participants wanted to tell the interviewer anything else and asking how they felt after having participated in the interview. The rationale for doing this was to identify any participants who were affected negatively by the interview and needed support. The parents received a contact number to a psychologist in case they or the child needed support.

None of the participants reported any negatively feelings, either for themselves or on their child's behalf, in connection with the interview situation. However, some of the parent stated that the interviews had had a positive effect, mostly for their child but also for them. A common statement from both children and parents was that by participating in the studies they helped others and that was satisfying.

Results

The results in study I describe the HRQoL assessment of children with CHD and in study II the HRQoL assessment of children with RVOT anomalies.

The results in studies III and IV illuminate the experiences of everyday life for children with RVOT anomalies and their parents during an assessment for a possible reoperation.

HRQoL assessment

HRQoL in children with CHD

Children's self-report in comparison to the number of surgeries and to children with other chronic conditions (study I)

HRQoL was measured in children with CHD using DCGM-12 in relation to the number of cardiac surgeries they had undergone. The median of the overall total score was 95 (10^{th}–90^{th} percentile 75–100). Children with biventricular CHD (95%) were further categorized into four groups by number of surgeries, and children with UVH (5%) were grouped separately. The group of children with three or more surgeries had significantly lower HRQoL ($p<0.001$) compared to the children in the other four groups (Figure 2). The group of children with three or more cardiac surgeries had comparable HRQoL to children with asthma, arthritis, dermatitis, diabetes, cerebral palsy, cystic fibrosis and epilepsy. The groups of children with fewer or no cardiac surgeries ($p<0.001$) and the group of children with UVH ($p=0.047$) had higher HRQoL than children with other chronic conditions in the DISABKIDS reference group (Figure 3).

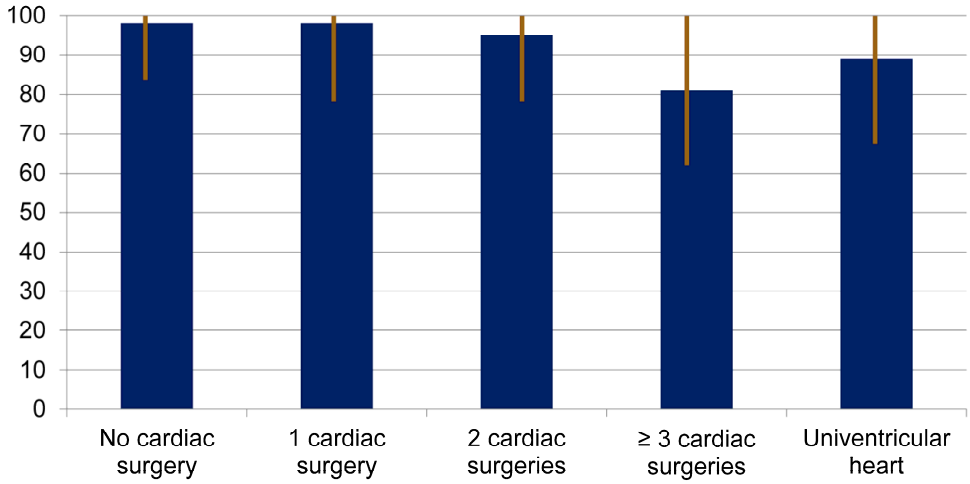

Figure 2. Children with biventricular CHD categorized according to number of surgeries; children with univentricular heart are grouped separately (rightmost column). Data are shown as median and 10th–90th percentiles. A DCGM-12 total score of 100 indicates the highest possible HRQoL.

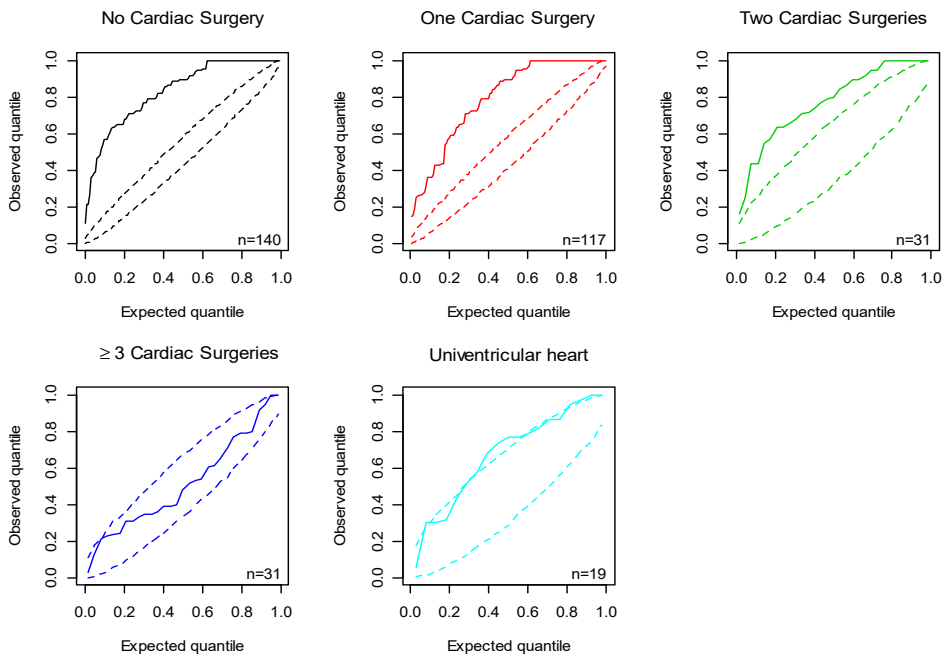

Figure 3. The distribution of the DCGM-12 total score in relation to the reference score for children with other chronic diseases. The solid line denotes total score for children with CHD; the area within the dotted lines includes total score for children with other chronic diseases.

Supplementary HRQoL variables (study I)

Besides DCGM-12, at that time in the SWEDCON registry, there were supplementary HRQoL variables such as NYHA class, cognitive function, psychosocial ability, and physical activity. The majority of respondents were in NYHA class I (87%) and attended regular school (88%). Most of the children in NYHA class II had undergone at least two surgeries or had UVH. Nearly all children in NYHA class III had undergone three or more surgeries or had UVH. Children who had undergone three or more surgeries and children with UVH needed more help in school (cognitive function) than children with no surgeries or only one surgery ($p<0.001$) (Figure 4). There was no significant difference between the groups of children concerning psychosocial ability and physical activity.

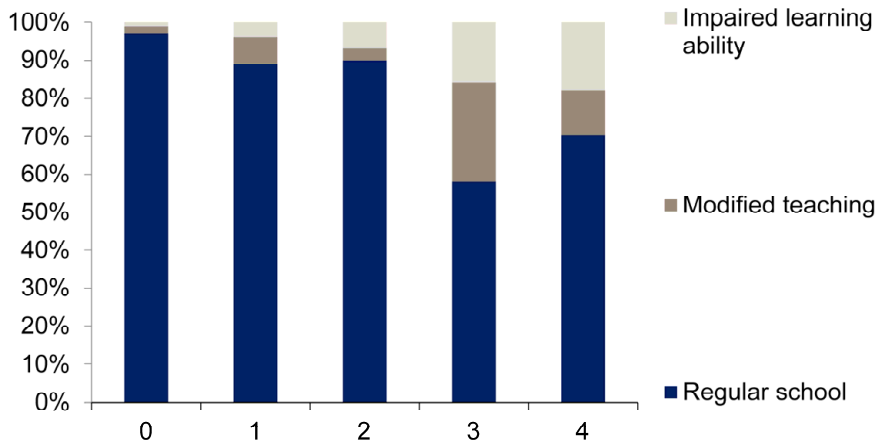

Figure 4. Proportion of children (%) at different cognitive functioning levels (measured by level of help needed in school) in the five groups of children. 0 = no surgeries, 1 = one surgery, 2 = two surgeries, 3 = three or more surgeries, 4 = univentricular heart.

HRQoL in children with RVOT anomalies

Child self- report and parent report in DCGM-12 (studies I and II) and in PedsQL (study II)

In study I, the median for DCGM-12 total score was 95 (10^{th}–90^{th} percentile 75–99) compared to 88 (55–100) in study II. The corresponding HRQoL for the parents (study II) was 90 (48–100). The mean total score for the child report in PedsQL Cardiac Module (study II) was significantly lower than in PedsQL 4.0 ($p \leq 0.012$) and DCGM-12 ($p \leq 0.001$). In PedsQL Cardiac Module, the children and the parents reported the lowest mean score in the cognitive problems domain. They had the highest mean score in the physical appearance domain. In PedsQL 4.0, the lowest mean score was reported by both children and parents in the school functioning

domain. The children had the highest mean score in the physical functioning and social functioning domains. The parents had the highest mean score in the physical functioning domain (Table 6). Higher score means better quality of life.

Table 6. Mean score and standard deviations (SD) for the child and parent reports in PedsQL Cardiac Module, PedsQL 4.0, and DCGM-12. 100 =highest HRQoL

	Mean	SD	Mean	SD	p	r
PedsQL Cardiac Module	**Child (N=93)**		**Parent (N=90)**			**N=88**
Heart problems	76	18	77	20	>.3	0.80***
Physical appearance	79	24	80	23	>.3	0.49**
Treatment anxiety	74	29	76	29	>.3	0.65***
Cognitive problems	67	26	62	29	0.065	0.83***
Communication	74	27	71	31	>.3	0.61***
Total score	74	19	74	20	>.3	0.73***
PedsQL 4.0	**Child N=90**		**Parent N=89**			**N=85**
Physical functioning	82	19	79	22	0.067	0.84***
Emotional functioning	76	22	72	23	0.033*	0.82***
Social functioning	82	20	78	24	0.055	0.78***
School functioning	72	22	70	23	>.3	0.80***
Psychosocial[a]	77	20	73	21	0.049*	0.85***
Total score	78	18	75	21	0.040*	0.86***
DCGM-12	**Child N=91**		**Child N=91**			**N=86**
Total score	81	21	81	23	>.3	0.78***

The highest possible score is 100. a: aggregated emotional functioning + social functioning + school functioning. Column p: p-value from Wilcoxon test for mean rank comparison between child and parent report. Column r: Spearman correlation between child and parent report. (*p<0.05, **p<0.01, ***p<0.001.)

The agreement between child and parent reports (study II)

In PedsQL Cardiac Module and PedsQL 4.0, the children reported significantly higher HRQoL than their parents reported at the domain level (Table 6). In PedsQL Cardiac Module, the agreement between the children and their parents was strong in all domains except physical appearance, and in PedsQL 4.0 the agreement was strong in the domains physical, social and school functioning. In PedsQL Cardiac Module, the agreement was also calculated at the item level between children in the two age groups (8–12 and 13–18 years of age) and their parents. There was strong agreement for 13 of 22 items in both age groups, but the items differed between the younger and older children. For the younger age group, the correlation was medium between children and parents (r = 0.364–0.497) for two items in the heart problems, perceived physical appearance and communication domains. Children reported significantly higher HRQoL than their parents for one item and lower for two items. For the older age group, the correlation was medium (r = 0.452–0.499) for three items in the treatment anxiety domain and for one item in the perceived physical appearance domain. Children reported significantly higher HRQoL than their parents for four items and lower for one item.

Items for the most frequently reported problems in PedsQL and DCGM-12 (study II)

Regarding the result for items for specific concerns in both PedsQL Cardiac Module and PedsQL 4.0, three of six items were related to cognitive problems and school functioning. In PedsQL Cardiac Module at item level, nearly half of the children reported *almost always* or *often* having problems with breathlessness when participating in sports, and 22% reported *almost always* or *often* having problems due to having to rest more than their friends (Table 7). In DCGM-12, a total of 10–12% of children reported that their heart disease caused them different kinds of troubles or negative feelings (Table 8).

Table 7. Items for specific concerns by child self-report in PedsQL Cardiac Module and PedsQL 4.0.

PedsQL Cardiac Module N=90	**Child self-report N (%)**
Almost always or often problem with…	
A1 I get out of breath when I do sports activity or exercise	39 (43)
A7 I have to rest more than my friends	20 (22)
E2 I have troubling solving math problems	23 (25)
E5 It is hard for me to remember what I read	18 (19)
E4 It is hard for me to pay attention to things	17 (18)
F3 It is hard for me to explain my heart problem to other people	16 (17)
PedsQL 4.0 N=90	**Child self-report N (%)**
Almost always or often problem with…	
A2 It is hard for me to run	17 (19)
A8 I have low energy	16 (18)
B5 I worry what will happen to me	15 (17)
D1 It is hard to pay attention in class	15 (17)
D2 I forget things	18 (20)
D3 I have trouble keeping up with my schoolwork	14 (16)

Table 8. Items for specific concerns by child self-report in DCGM-12.

DCGM-12 Condition: heart disease N=90	**Child self-report N (%)**
Never or seldom	
Do you feel like everyone else even though you have your condition?	9 (10)
Always or often	
Is your life ruled by your condition?	9 (10)
Does your condition bother you when you play or do other things?	10 (11)
Are you unhappy because of your condition?	11 (12)
Do you feel different from other children/adolescents?	10 (11)

Experiences of everyday life

The analysis of the interviews resulted in three main themes and eight subthemes for the children (study III) and in five main themes for the parents (study IV) (Figure 5). The findings are aggregated and presented under the following headings: *The safe assessment and the dreaded reoperation, Adaptation of struggles and limitations* and *Sources of strength.*

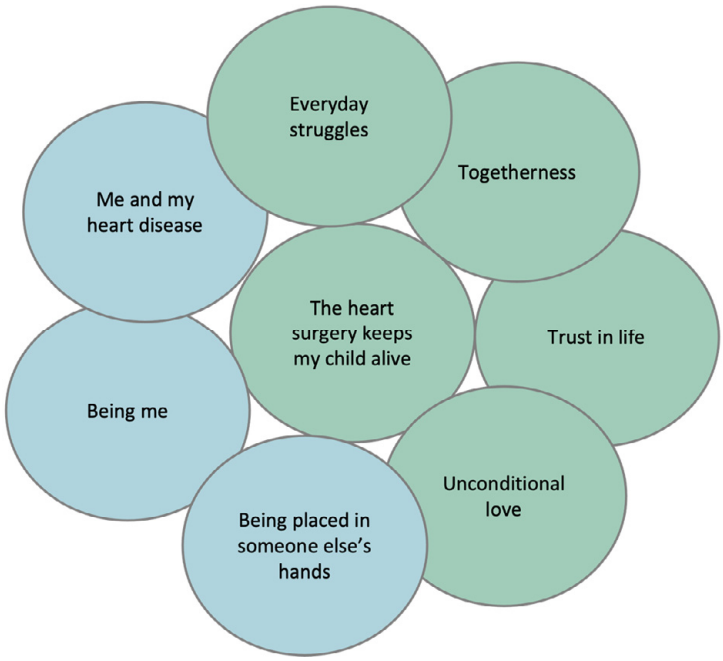

Figure 5. Thematic map of the children's and parents' main themes. Blue circles represent the children's three main themes and green circles represent the parents' five main themes.

The safe assessment and the dreaded reoperation

The duality of emotions

Children with RVOT anomalies (study III) and their parents (study IV) considered the assessment for reoperation as a safety net, although fear and worries were awakened when they considered the reoperation. The parents' memories from the child's first surgery were easily awakened and could be felt as if etched in their history. There were two aspects to these memories: in one way parents feared that they might lose their child in a new surgery and in another way the fact that their child had survived one or more surgeries earlier in life gave some comfort and confirmed the strength of their child. Children expressed their fear of surgery both in tears and words.

The decision

The decision to operate or to postpone surgery was in most cases delivered to parents by phone or letter and they in turn had to pass the information further to their children. This was a harder task for the parents when the decision was surgery since they sometimes had to answer difficult question from the child, such as whether the child was going to die during surgery, and at the same time handle their own fears. Undergoing assessment was not a psychological strain for the children; the fear only came with the decision to have surgery. Their reactions differed depending on their symptoms and how they felt about undergoing yet another surgery, but also depending on their personality. The reactions ranged from simply surrendering themselves to the surgery to, at the other extreme, starting to prepare their own funeral.

The surgery affects life

For the parents, the waiting time between the decision and the surgery was hard and, in their view, too long. Both the children and the parents gave accounts of unexpected clinical situations in connection with the surgeries that did not turn out as planned, for example when a complication made life worse instead of better, or when the surgery involved implantation of a biological valve for a child who feared receiving human tissues. Some of these accounts may reflect misunderstanding or misinformation and lapses in communication prior to the surgery. Despite these setbacks, some time after the surgery the parents experienced a new strength in themselves and in their children and the children expressed a fighting spirit; they had not given up.

The need for future surgery

For children who did not need surgery, the feeling of relief banished their worries about having to undergo a new surgery, even if this fear never left them completely. They were so accustomed to the thought of a future surgery that they sometimes found it hard to explain how it affected their lives. The decision not to operate gave the parents a confirmation that their child's heart was in good shape, and they could put aside the thoughts of surgery, at least for now. At the same time, they were aware that the child's heart valve had lasted so much longer than they were initially told, which meant that every day was a day closer to the future surgery.

Adaptation due to struggles and limitations

Children's limitations because of the heart disease

The children struggled more or less with a daily tiredness that intensified during physical activity. The sports at school were a challenge and the lessons that followed could also be difficult. Some had to give up their favourite leisure activity, such as football, and if they ran out of energy, they might need to rest for a whole day. They

also commented that this tiredness meant that they had days when they had to go home directly after school to rest instead of going out with friends. Sometimes it was difficult for them to interpret their tiredness, whether it was their heart disease, normal adolescence, the winter darkness or just having a bad day. Thoughts such as who they would have been without the heart disease appeared every now and then, as well as a silent struggle to be like everyone else. They had to manage their fear of heart surgery, and they succeeded in putting this fear aside until the surgery was a fact.

Parents' struggles in everyday life

The parents' struggles were in different directions. One was to fight for their child's rights in the school world. Another direction was within themselves, and it could involve managing anxiety far beyond the usual parent worries, and they needed to hide this anxiety from their child. They had to be strong both for their child and for themselves even if they sometimes felt powerless as parents. They reported the need of a huge inner strength, for example when they had to calm down their child after a panic attack at the hospital. For this kind of situation they had no instruction manual but instead they had to trust their intuitive feelings about how to handle such situations.

The adaptation process

The children had learned to adapt to their heart disease, finding ways to ease the limitations it imposed. The most common adaptation was to rest and take a short break, but also to manage their strength so it would last longer; alternatively, they developed ways of showing their lack of energy without expressing it in words. The other side of the adaptation process was that they got so used to the limitations that they rarely experienced them as limitations anymore, to the extent that they found it hard to describe in what ways they were affected by their heart disease. The parents, like the children, had developed the strategy of suppressing their worries and anxiety concerning future reoperation until it became a reality. This strategy may have been facilitated by their child's inner strength and well-being thus far.

Sources of strength

Togetherness and a strong sense of self

The parents gained strength from their unconditional love to their child, and from a special bond that was created from the time when they could have lost their newborn child. They described a particular closeness to their 'child of the heart'. This love was expressed in how the parents described their child and, together with the child's joie de vivre and fighting spirit, the parents gained inner strength. The children also described their sense of self in positive terms, such as happy, open-minded, and independent. They gave accounts of having a meaningful life with a "being me" where the heart disease did not rule over their lives. Their spare time was filled with

activity, for example, football, dance, listening to music or just relaxing. The scar could sometimes symbolize for them that they had survived, which made them special and strong, because not everyone survives such surgery.

The children reported that they had a strong social network, both at home and in school, and the most important things in life were family, friends and having fun. The parents emphasized the family as the most important source of strength, along with the importance of building social networks both for themselves and for their children. Togetherness was their way through difficulties and talking to others was the most common way to feel less alone. Another form of togetherness for some of the parents was having faith in God.

Confidence in life

The parents' well-being was dependent on the child's well-being, so when the child was well, the parents were well. The parents experienced a trust in life, and with that came an acceptance of how life had become, but also an insight that life is fragile and cannot be taken for granted. Trust and gratitude to the HCPs facilitated this process. The experience of feeling well treated by HCPs gave the parents a feeling of comfort and security. Children had similar experiences: they felt well taken care of and spoke of good memories from their hospital stays. The children and their parents shared a trust and belief in a bright future where the heart disease was not an obstacle.

Discussion

General discussion of the findings

The most important results from the four studies are discussed below, both separately and together.

HRQoL in children with CHD

The results in study I showed that, although children with CHD have a high HRQoL overall, children with biventricular CHD who had undergone three or more surgeries and children with UVH had significantly lower HRQoL than the children with fewer surgeries. There was no significant difference between children who had had none, one or two surgeries. Another study showed that patients aged 12–26 years with more than one cardiac surgery had lower HRQoL than those who had only one operation (Silva et al. 2011). Some of the differences between our study and the study by Silvia et al. (2011) are the different age span of the participants and that HRQoL was assessed with two different questionnaires.

Measuring HRQoL in children with CHD remains challenging, mostly due to the wide age range and their development during childhood and adolescence. Additional challenges include the wide spectrum of CHD complexity (from mild to complex) and treatment (pharmacological, transcatheter or surgical) and the extensive range of outcomes (Marino et al. 2008). All these variables, together with the differences in data collection and different kind of questionnaires, may explain some of the discrepancy in the results of HRQoL studies for children with CHD (Latal et al. 2009).

The finding that children with biventricular CHD and multiple surgeries as well as children with UVH present lower HRQoL is not surprising. This negative impact of heart surgery on HRQoL has been shown in several studies (Latal et al. 2009; Landolt et al. 2009; Andreasen et al. 2014), including the impact of an increased number of surgeries (Francis et al. 2023). The result in study I confirm that a higher number of surgeries, probably related to more severe cardiac disease, involving many hospitals stays with more or less painful procedures, impact the children's HRQoL negatively.

HRQoL agreement between children and parents

The agreement at the domain level between children and their parents in study II was strong in both physical and emotional domains. This is in contrast with earlier studies that showed stronger agreement in the domains that reflect observable behaviour (Eaglesen et al. 2013; Patel et al. 2017; Uzark et al. 2003, 2008). However, the result in study II that children self-reported higher HRQoL than their parents reported for them, agrees with other studies (Niemitz et al. 2017; Kwon et al. 2011; Berkes et al. 2010). Despite this, it is difficult to draw further conclusions about differences and similarities between studies concerning parent–child agreement because the statistical methods differ. However, there is one common conclusion, namely that the agreement results show the need for both child self-report and parent report in the evaluation of HRQoL in children with CHD (Uzark et al. 2008; Patel et al. 2017; Eagleason et al. 2013).

HRQoL in clinical follow-up

The results in study II suggested that the PedsQL Cardiac Module could best identify the children's problems and is therefore useful in clinical follow-up. Earlier research has indicated that there is a need for HRQoL assessment as part of the clinical decision-making for children with CHD (Amodeo et al. 2022; Abassi et al. 2020; Eagleson et al. 2013; Francis et al. 2023). One study, in which children with various forms of CHD filled out the generic PedsQL 4.0 questionnaire during an outpatient visit, showed that 30% of the children received interventions such as exercise tests, education and counselling according to the result of the HRQoL assessment (Uzark et al. 2013). A similar result was shown in a study where children with TOF had responded to the same questionnaire. In that study, 38% of the children had received similar interventions (such as education and counselling) according to the results of the HRQoL assessment (Siebrasse et al. 2019).

Cognitive function in children with CHD

Earlier research has shown that there is a need for follow-up and screening according to the increased neurodevelopmental risk in children with TOF (Bellinger et al. 2015; Holland et al. 2017; Miatton et al. 2007). This has been confirmed in the studies in this thesis. In study I, children with biventricular CHD who had undergone three or more heart surgeries and children with UVH needed significantly more help in school than children with less complex CHD. In study II, both children and parents gave the lowest rating in cognitive function (PedsQL Cardiac Module) and school functioning (PedsQL 4.0) with strong agreement at the domain level. In study IV, the parents of children with RVOT anomalies described their struggles to assert their children's rights in school.

One previous review study showed that children with chronic illness (including heart disease) have a poorer school experience than children without chronic illness

(Lum, et al. 2017). For children with TOF who filled in PedsQL 4.0, the most commonly placed referrals were school intervention and neurodevelopment testing, according to the result of the HRQoL assessment (Siebrasse et al. 2019). This strengthen the importance of using HRQoL assessment as a screening tool in follow-up for cognitive impairment which has been demonstrated in previous studies (Buratti et al. 2016; Daliento et al. 2006).

Children's and parents' adaptation to the heart disease

For families with a chronically ill child, the focus in earlier research has changed from family functioning in terms of limitations and mental illness, to instead focusing on the family's adaptation to the challenges posed by the child's chronic illness (Jackson et al. 2015).

The children's and their parents' narratives in studies III and IV revealed patterns of how they had adapted to the challenges and difficulties originating from the child's CHD, including the treatment. This ability to adapt is an important finding in studies III and IV. The children sometimes described symptoms of the heart disease having a huge impact, such as having to rest every day after school, but at the same time they shrugged off the experience as barely worth mentioning. Furthermore, both children and parents showed adaptative behaviour in relation to their awareness of the need for future surgeries, at least until it became an inescapable reality. One way to understand this adaptation is by the construct *acceptance* in illness identity, which is strongly related to adaptive functioning and the ability to maintain a coherent identity (Oris et al. 2018). Illness identity is defined as "the degree to which a chronic health condition, such as heart disease is integrated into someone's identity" (Van Bulck et al. 2018) and is based on four constructs: *engulfment*, *rejection*, *acceptance* and *enrichment*.

Acceptance can also lead to a satisfaction with self (Shearer et al. 2013). The fact that children in study II rated the highest HRQoL on the physical appearance scale (PedsQL Cardiac Module) and described themselves in positive terms in study III could be seen in the light of the *acceptance* construct in illness identity. This has also been shown for young adults (17–32 years of age) with UVH, where they felt happiness over "being me" and also felt committed to life (Berghammer et al. 2015). However, Marino et al. (2012) emphasized that children with CHD need help from HCPs to maximize their adaptive behaviour, since adaptive behaviour is one of the factors that are essential for the child's self-esteem.

Heart disease and the child's identity

The result of the most reported problems in study II was that approximately 10–12% of the children *always* or *often* felt that their heart disease ruled their lives or reported that they felt unhappy because of the heart disease (items in DCGM-12). This result could be related to the construct *engulfment* in illness identity. Engulfment indicates

to what degree the disease dominates the person's identity (Van Bulck et al. 2021), and the results in study II show that a clear majority of these children did not feel that their heart disease dominated their lives.

This is consistent with the results in study III, where the children gave accounts of having a meaningful life with a "being me", in which the heart disease did not feature in their sense of self. A previous study of young adults with CHD showed that "living with illness" was a fluctuating condition which could claim a large or a small part of the person in question, depending on their emotional state (Berghammer et al. 2006), which suggests that sometimes the person and sometimes the disease was in control. In the same way, there were moments for the children in study III when the heart disease took over, such as when they received the decision that a new heart surgery was needed. Some of them felt they drowned in frightening feelings and a fear of death or felt that strong symptoms took over and ruled their everyday life. Engulfment has also been found to be related to stronger feelings of having an illness, as in the study by Oris et al. (2016) with adolescents with diabetes.

Parenting concerns

The parents in study IV needed a huge inner strength since they experienced an anxiety far beyond the usual parental worries and felt sometimes extreme powerlessness, with no instruction manual for how to be a parent to their child. This has been shown in previous research, where parents of children with hypoplastic left heart syndrome experienced "extraordinary parenting" when they needed to safeguard their child's precarious survival (Rempel et al. 2007). The need parents expressed could be exemplified by the Parenting and Childhood Chronicity model, initially developed with parents of children aged 15 months to 16 years with various chronic conditions in order to describe the often invisible work that is required to raise a child with a chronic health condition (Ray 2002). The parents in study IV and in the Rempel et al. study (2007), have in common the worry and anxiety beyond usual parenting, but the studies also have some differences. The children in study IV were aged 9–17 years whereas in the study by Rempel et al. (2007) the children were younger than five years. In contrast to the Rempel et al. study, the parents in study IV did not in general fear for their child's survival in everyday life. That was more of a challenge that arose at the time of the decision to perform a reoperation.

The most important thing in life

The friends clearly have an impact in children's life, as shown in study II, where children had the highest HRQoL score for the domain social functions (PedsQL 4.0). This is in line with the finding in study III that the most important aspects in life for those children was their family and friends. One earlier study has shown that children and young adults between 12 and 26 years of age had a better self-reported HRQoL regarding social relationships than the healthy reference population.

Furthermore, that study indicated that a positive family setting and social support made it easier for the children to adapt to the heart disease (Silva et al. 2011).

The parents in study IV had over the years managed to successfully create sources of strength. One source of strength, the importance of family, was in line with the children's narratives in study III, but the parents also described a social network of colleagues, friends, or parents in a similar situation. Similar results have been reported in earlier research, where parents of children with CHD reported that their psychosocial status improved with an improved social network (Lawoko et al. 2006) and that families were brought closer together because of their child's CHD (Wei et al. 2015). The parents in study IV expressed the value of meeting other parents in a similar situation, but few of them had established that kind of relationship outside the hospital. HCPs have an important role in mediating contact with, for example, patient associations.

The findings in study IV illuminate the close bond between the parent and child, as well as the parent's trust in life. It is likely that this close bond originates from the time of the child's birth when the parents needed to face the fear of losing their child. This close bond has been described previously, where the early attachment between mother and infant was established despite the hospital stay for heart surgery, and the bonding was a sign of a closer relationship (Jordan et al. 2014). Later in life, parents' warmth was associated with a better QoL in adolescents with CHD (Im et al. 2018). During a hospital stay for the child, these parents are vulnerable. One study showed that the majority of the parents had some trauma symptoms, which in turn could impair their ability to bond (Franish-Ray et al. 2013). The HCP can facilitate this bonding by limiting the time the infant is separated from the parents and by protecting the infant from pain (Daliento et al. 2006).

Child-centred care

The parents in study IV had established trust for their HCP over time and were in general terms satisfied with the child's care. Lawoko et al. (2006) showed that satisfaction with the child's care was more important than the child's clinical severity, when it comes to parents' long-term psychosocial morbidity. Despite being satisfied with the care their child received, the parents in study IV talked about difficulties related to shortcomings in communication with HCPs, which their children also reported in study III. Most likely, these difficulties could be remedied with a CCC perspective in the child's care. A CCC approach means that children's interests are in focus and that they are allowed to participate in their own care. Essential for CCC is a recognition that the children's viewpoint is not always the same as that of their parents or HCPs. This approach can facilitate the child's participation and additionally includes both the child's perspective and the child perspective, which means that the parents' participation is also integrated, even if the HCP needs to embrace especially the child's perspective (Quaye et al. 2019).

Methodological considerations

In both quantitative and qualitative research, the research quality needs to be evaluated. In quantitative studies, reliability and validity demonstrate the quality of the methods (Polit and Beck 2021). In qualitative studies, trustworthiness is shown by credibility, dependability, confirmability, and transferability (Lincoln and Guba 1985).

Studies I and II

Validity

The validity of a quantitative study concerns the extent to which the conclusions are credible or not (Polit and Beck 2021). A threat to validity is the participants' confounding characteristics. CHD is a heterogeneous disease and there are different ways to categorize it, usually into three groups: *mild, moderate* (repaired biventricular CHD) and *complex* (palliated UVH) (Marino et al. 2010b). A limit of this way of categorizing is the variation in complexity, from no symptoms to requiring multiple surgeries, within the same diagnosis. To avoid this variation in complexity, the children in study I were divided into groups related to the number of surgeries they had undergone, which implies both a complexity in the diagnosis and how much health care they have had in their lives. In order to differentiate between children with biventricular CHD and children with UVH, those with UVH were grouped separately.

The uneven sample distribution in study I, with a large proportion (42%) having no surgeries, may have influenced the result. This could be a sample bias (Polit and Beck 2021) but probably it is representative of children who visit the outpatient clinics, since the majority of children with CHD do not need surgical treatment. The population of children with CHD consists both of children with mild CHD (when surgery is not needed) and complex CHD, as shown in previous research (Bratt et al. 2015; Uzark et al. 2008). The sample in study II consisted of children with different diagnoses, which could be a confounding factor. However, to secure the sample, they were selected from the group with RVOT anomalies in the SWEDCON registry (which is based on EPCC codes) (Franklin et al. 2002) and finally determined by paediatric and ACHD cardiologists responsible for the registry.

Procedure

The age range 8–18 years was determined based on the recommendation that younger children (5–7 years of age) need adult support to be able to answer the questionnaire (Uzark et al. 2003; Varni et al. 2001). For children from 8 years, the reliability of the self-reports is good if the questionnaires are constructed for different age groups (Riley 2004), which the questionnaires in studies I and II are.

The data collection consisted of questionnaires with closed-ended multiple-choice questions. The questionnaires were collected either from a register (Study I) or sent out by post to the children and their parents (study II). Sending questionnaire by post is cost-effective for reaching respondents who are geographically dispersed (Polit and Beck 2021), as was the case with the participants in study II. The shortcoming with this approach is that there is no way to get an overview of how the surveys are filled out. However, regarding the concern that parents may have an influence on the child's self-report, a recent study has shown that there was no difference in mean score (PedsQL 4.0) according to whether the questionnaire was administered in the clinic or by post (Derridj et al. 2022). Furthermore, no differences were found in an earlier study concerning agreement between child and parent depending on where the questionnaires were filled in (at home or in the clinic) (Upton et al. 2008). In order to optimize the circumstances, an information letter stressing that the child should fill out the questionnaire independently, was sent out. Furthermore, those few questionnaires where it was stated that the parents had responded on behalf of the child were excluded from the study.

Validity and reliability for Questionnaire

The concept of validity for questionnaires means to what extent the questionnaire measures what the intention was to measure. *Internal validity* refers to whether the results are reliable based on the methods used, and *external validity* concerns to what extent the measurement has wider validity. *Reliability* concerns to what extent the scores are free from measurement error. A reliable questionnaire is dependable and thus trustworthy. (Polit and Beck 2022).

The questionnaires PedsQL Cardiac Module, PedsQL 4.0 (study II) and DCGM-12 (studies I and II) are well-established instruments. Reliability and validity have been proven for PedsQL Cardiac Module (Sand et al. 2013; Uzark et al. 2003) and for PedsQL 4.0 (Petersen et al. 2009; Varni et al. 2001) and both have previously been used in studies for children with CHD (Moreno-Modina et al. 2020; Raj et al. 2019; Uzark et al. 2008; Wang, et al. 2014). The DCGM-12 was derived from the original 37-item DCGM-37 and also has proven reliability and validity (Bullinger et al. 2002; Carona et al. 2014; The European DISABKIDS group 2006). To our knowledge, DCGM-12 has not been used for children with CHD earlier. DCGM-12 became relevant for these studies since it was the HRQoL assessment used in the SWEDCON registry when study I was performed.

The level of agreement has been an issue for discussion since parallel measurement (child and parent versions consisting of the same questions) became available (Creemens et al. 2006). Level of agreement can be used as a validation for a new questionnaire but also to understand the difference between child and parent report (Patel et al. 2017; Varni et al. 2007), which was the intention for the assessment of agreement in study II. The higher total score in DCGM-12 than in PedsQL Cardiac Module and PedsQL 4.0 in study II and lower percentage of respondents reporting

difficulties in DCGM-12 could be related to the absence of measurement on domain levels and that, in contrast to PedsQL, it is not a problem-based questionnaire. The results in study II indicate that problems perceived by the children were more easily detected with the disease-specific questionnaire, a result that supports the superiority of disease-specific questionnaires (Marino et al. 2010a).

The response rate in study II was 50%; however, the dropout analysis showed no differences between the groups regarding gender, age or diagnosis. The analysis was calculated on the largest group, children with TOF. In order to comprehensively describe HRQoL in this complex group of children and their parents, three questionnaires were chosen. It is possible that fewer questionnaires would have increased the response rate by putting less burden on the participants. Another reflection whether allowing the parents to choose how they wanted to complete the questionnaires would have increased the response rate. However, in a study where the parents could choose between completing a follow-up questionnaire online, by post or by telephone, the overall response rate was 57%. The parents had chosen the option that best suited them, and the authors believed that without that possibility, the response rate would be even lower (Pulham et al. 2019). According to Public Health Agency of Sweden, the response rate for different kinds of survey investigation has decreased over time and not only for the national population health survey. For 2022, the response rate for that survey was 37.9% (Public Health Agency of Sweden).

Studies III and IV

Credibility

Credibility in qualitative research refers to the confidence and truth in the findings and can be described as a twofold task (Lincoln and Guba 1985). The first is to perform the study in such a way that the findings and interpretations are credible and the second is to demonstrate this credibility.

Lincoln and Guba (1985) speak about "prolonged engagement", "persistent observations" and "building trust." In this thesis, "prolonged engagement" was facilitated by the author's long experience of nursing care in paediatric cardiology, ensuring a thorough knowledge of the context. Furthermore, the three interviews with each participant in studies III and IV spanned a period of over a year, which enhanced the conditions for a prolonged engagement. The fact that all 18 participants took part in all three interviews strengthened the richness in the data. During the interviews, probe questions were used to gain more depth but also to clarify what the participants really meant, and this was illustrated in the findings with quotes. To enhance the "persistent observation" aspect, the analysis was performed from the transcribed interview text and with a constant focus on the aim, so the distinction between relevant and irrelevant data was possible. The "building

trust" aspect was enhanced by the ethical considerations through the whole process, from the recruitment, with a special emphasis on the children being addressed directly to the written reports. The reports of the findings using quotations were written with the participants' anonymity in mind first and foremost.

Dependability and credibility

Dependability and credibility are linked together in the same way as reliability and validity (Lincoln and Guba 1985). By demonstrating credibility, the dependability is established. To further ensure dependability, only one interviewer conducted all the interviews. In addition, rigor was enhanced by using Braun and Clark's 15-point checklist of criteria for good thematic analysis (Table 9) (Braun and Clarke 2006).

Table 9. A 15-point checklist of criteria for good thematic analysis (Braun and Clarke, 2006)

Process	Number	Criteria
Transcription	1	The data have been transcribed to an appropriate level of detail and the transcripts have been checked against the tapes for "accuracy".
Coding	2	Each data item has been given equal attention in the coding process.
	3	Themes have not been generated from a few vivid examples but instead the coding process has been thorough, inclusive and comprehensive.
	4	All relevant extracts for each theme have been collected.
	5	Themes have been checked against each other and back to the original data set.
	6	Themes are internally coherent, consistent, and distinctive.
Analysis	7	Data have been analysed – interpreted, made sense of – rather than just paraphrased or described.
	8	Analysis and data match each other- the extracts illustrate the analytic claims.
	9	Analysis tells a convincing and well-organized story about the data and topic.
	10	A good balance between analytic narrative and illustrative extracts is provided.
Overall	11	Enough time has been allocated to complete all phases of the analysis adequately, without rushing a phase or giving it a once-over-lightly.
Written report	12	The assumptions about, and specific approach to thematic analysis are clearly explicated.
	13	There is a good fit between what you claim you do, and what you show you have done – i.e., the described method and reported analysis are consistent.
	14	The language and concepts used in the report are consistent with the epistemological position of the analysis.
	15	The researcher is positioned as active in the research process; themes do not just "emerge".

Confirmability

Confirmability refers to the researcher's objectivity but, according to Braun and Clarke, no such thing as objectivity could exist in TA. The researcher in TA, and specially in reflexive TA, plays an active part in the process of producing knowledge. However, this requires that a reflexive journal is created for the researcher's reflections and insights (Braun and Clarke 2022). For studies III and IV, a reflexive journal was created at the beginning of the interviews and kept throughout data collection and analysis, to facilitate a questioning, reflexive

thinking. Reflexivity can be considered as a disciplined self-reflection throughout the research process (Braun and Clarke 2022). According to Patton (2015), quality control, created directly after the interview, is important. Questions such as where the interview took place, whether there was anybody else around, how the interviewee reacted to the questions, how well the questions were asked, and the interviewer's feelings and thoughts about the situation and the interaction and what was revealed, enhanced a reflexive quality (Patton 2015). In this thesis, the interviewer's reflections, feelings and thoughts were audio-recorded after each pair of interviews with child and parent and transcribed later together with the interviews.

Transferability

Transferability refers to whether the findings can be applied to other contexts or groups of people. In order to facilitate the reader's evaluation of the transferability (Lincoln and Guba 1985) studies III and IV aimed to richly describe the contexts, the participants characteristics, data collection and the process of analysis. The sample size in studies III and IV was mainly dependent on the relatively small paediatric population with various forms of complex RVOT anomalies and how many of these children would be called for an assessment of their need for reoperation within the time frame of this thesis. According to Braun and Clarke (2022), a smaller participant group is defined as 10 interviews or fewer. In this thesis, only nine children participated, but this generated 27 interviews with a matching number of parent interviews. The qualitative aspects of the data are the most important, namely that the data should be rich and complex to enable a deep and nuanced analysis (Terry et al. 2017).

Conclusion

Children with CHD requiring multiple surgeries have lower HRQoL than children with fewer surgeries (study I) and children with RVOT anomalies and their parents reported the lowest HRQoL in the cognitive domain and the highest in the physical appearance domain (study II). Seen through an adaptation lens, the children with RVOT anomalies and their parents in studies III and IV experienced going through the assessment and evaluation for cardiac surgery as a safety net, even if it aroused their fear and anxiety.

Study I
Children with a higher number of cardiac surgeries, probably related to the severity of cardiac disease and more hospital care, had lower HRQoL, lower cognitive function (measured by the child's need for extra support in school) and a higher NYHA classification than children with a lower number of cardiac surgeries.

Children with CHD reported higher HRQoL than children with other chronic diseases, except for children with biventricular CHD who had undergone three or more cardiac surgeries.

Study II
Problems concerning cognitive function should be considered by HCPs, as both children and parents reported the lowest HRQoL in this domain.

The variations in the level of agreement between children with RVOT anomalies and their parents in the assessment of the child's HRQoL indicate that both child self-report and parent report need to be taken into account.

Study III
The children's symptoms, their experiences during the assessment, their future surgeries, and how their heart disease affects their everyday life could be better understood as elements of their adaptation to the heart disease.

Their fear of heart surgery was aroused at the beginning of the assessment but was manageable for the children until it became an inescapable reality. By inviting the children to participate in their own care, shortcomings in communication can be avoided and tailored support can be achieved.

Study IV

The parents of children with RVOT anomalies were grateful for the assessment and had learned to navigate through the fears it aroused. They still experienced several stressful situations during the assessment process that need to be addressed. By understanding the parents' experiences and their sources of inner strength, the HCP can better aid the parents to cope with the experiences.

Clinical implications and future research

Previous research has shown the clinical utility of using HRQoL assessment for children, and the findings in this thesis confirm the benefits of including HRQoL assessment in the clinical follow-up for children with CHD. The variations in the level of agreement between child self-report and parent report emphasized the need to include both these reports. According to Varni et al. (2007), the child self-report should be considered as the standard assessment, which can be used as a benchmark if prioritization is needed.

Understanding the children's and parents' experiences can facilitate improvements in the child's care. For this to take place, the HCPs need to find out the child's and parents' anxieties and fears but also their adaptation strategies in their everyday life, as well as their strategies regarding the assessment prior to a possible reoperation. This could be done by inviting the child and parents to participate from the start of the assessment. A CCC approach, including the child's perspective and a child perspective, could facilitate this participation and the opportunity to tailor individual support for both the child and the parent. With participation in a CCC approach, the care pathway can be established and the information and communication gaps that these children and parents experienced can be prevented. The newer technology telemedicine, which became invaluable during the pandemic, has been shown to be a significant tool for families with chronically ill children when they need to establish contact with the HCP. Physical distance can be bridged by the use of telecommunication devices (Provenzi et al. 2020), and telemedicine can perhaps be an opportunity to bridge the gaps in communication between the families, the local hospital and the operating centre and a way to give the families more insight into the decision process.

Future research could also include a potentially interesting study to explore how younger children (under 8 years of age) with RVOT anomalies rate their HRQoL and how they experience their everyday life. To develop a tailored improvement in the children's care, a participatory study design should probably be considered.

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References

Abassi, H., Huguet, H., Picot, M-C., Vincenti, M., Guillaumont, S., Auer, A., Werner, O., De La Villeon, G., Lavastre, K., Gavotto, A., Auquier, P., Amedro, P. (2020). Health-related quality of life in children with congenital heart disease aged 5 to 7 years: a multicentre controlled cross-sectional study. *Health Qual Life Outcomes.* 18(1), 366. doi: 10.1186/s12955-020-01615-6.

Acker, J et al. (2003). Interviewing children and adolescents. In Holstein J. J. and Gubrium J. F., eds. *Inside Interviewing.* Thousands Oak: Sage Publications. pp.32-54.

Aguilar-Alaniz, E., Reyes-Pavon, R., Van-der-Ende, J., Felix-Orta, F. J., Delgado-Servin-de la Mora, I., Aroca-Peinado, A., Polo-Lupez, L., Moreno-Salas, J. C. (2021). Quality of life of children and adults following cardiac surgery for congenital heart disease: a Mexican cohort. *Arch Cardiol Mex.* 91(1), pp.34-41. doi: 10.24875/ACM.20000107.

Amodeo, G., Ragni, B., Calcagni, G., Piga, S., Giannico, S., Yammine, M. L., Drago, F., Ciofo degli Atti, M. L., Rossi, A., De Stasio, S., Grimaldi Capitello, T. G. (2022). Health-related quality of life in Italian children and adolescents with congenital heart diseases. *BMC Cardiovasc Disord.* 22(1), 173. doi: 10.1186/s12872-022-02611-y.

Andresen, B., Andersen, M.A., Lindberg, H., Døhlen, G. Fosse, E. (2014). Perceived health after percutaneous pulmonary valve implantation: in-depth interviews of patients and next-of-kin. *BMJ open.* 4:e 005102. doi:10.1136/bmjopen-2014-005102.

Arunamata, A. and Goldstein, B. H. (2022). Right ventricular outflow tract anomalies: neonatal interventions and outcomes. *Semin Perinatol.* 46(4) 151583 doi: 10.1016/j.semperi.2022.151583.

Bellinger, D. C., Rivkin, M. J., DeMaso, D., Robertson, R. L., Stopp, C., Dunbar-Masterson, C., Wypij, D., Newburger, J. W. (2015). Adolescents with tetralogy of Fallot: neuropsychological assessment and structural brain imaging. *Cardiol Young*. 25, pp.338-347. doi:10.1017/S1047951114000031.

Berghammer, M., Dellborg, M., Ekman, I. (2006). Young adults experiences of living with congenital heart disease. *Int J Cardiol.* 110(3), pp.340-7. doi: 10.1016/j.ijcard.2005.08.006.

Berghammer, M., Brink, E., Rydberg, A. M., Dellborg, M., Ekman, I. (2015). Committed to life: adolescents and young adults experiences of living with Fontan circulation. *Congenit heart Dis*. 10(5), pp.403-12. doi: 10.1111/chd.12244.

Berkes, A., Varni, J. W., Pataki, I., Kardos, L., Kemény, C., Mogyorósy, G. (2010). Measuring health-related quality of life in Hungarian children attending a cardiology clinic with the Pediatric Quality of Life Inventory ™. *Eur J Pediatr.* 169(3), pp.333-47. doi: 10.1007/s00431-009-1059-0.

Bhatt, S. M., Goldmuntz, E., Cassedy, A., Marino, B. S., Mercer-Rosa, L. (2017). Quality of life is diminished in patients with tetralogy of Fallot with mild residual disease: a comparison of tetralogy of Fallot and isolated valvar pulmonary stenosis. *Pediatr Cardiol.* 38, pp.1645-1653. doi: 10.1007/s00246-017-1709-3.

Birks, Y., Sloper, P., Lewin, R., Parsons, J. (2006). Exploring health-related experiences of children and young people with congenital heart disease. *Health Expect.* 10(1), pp.16-29. doi: 10.1111/j.1369-7625.2006.00412.x.

Bodell, A., Björkhem, G., Thilén, U., Naumburg, E. (2017). National quality register of congenital heart disease-Can we trust the data?. *J Congenit Cardiol*. 1(11). doi:10.1186/s40949-017-0013-7.

Bratt, E-L., Luyckx, K., Goossens, E., Budts, W., Moons, P. (2015). Patient-reported health in young people with congenital heart disease transitioning to adulthood. *J Adolesc Health*. 57(6), pp.658-65. doi: 10.1016/j.jadohealth.2015.07.021.

Braun, V. and Clarke, V. (2006). Using thematic analysis in psychology. *Qual Res Psychol*. 3(2), pp.77-101. doi.org/10.1191/1478088706qp063oa.

Braun, V. and Clarke, V. (2013). *Successful Qualitative Research: a practical guide for beginners.* London: SAGE Publications Ltd.

Braun, V. and Clarke, V. (2019). Reflecting on reflexive thematic analysis. *Qual Res Sport, Exerc Health*. 11(4), pp.589-597. doi:10.1080/2159676X.2019.1628806.

Braun, V. and Clarke, V. (2022). Conceptual thinking for thematic analysis. *Qual Psychol*. 9(1), pp.3-26. ISSN: 2326-3601. doi.org/10.1037/qup0000196.

Bullinger, M., Schmidt, S., Petersen, C., The DISABKIDS group. (2002). Assessing quality of life of children with chronic health conditions and disabilities: a European approach. *Int J Rehabil Res*. 25(3), pp.197-206. doi: 10.1097/00004356-200209000-00005.

Buratti, S., Ryberg, C., Broberg, M., Sunnegårdh, J. (2016). Do self- and proxy reports of cognitive problems reflect intellectual functioning in children and adolescents with congenital heart defects? *Front Pediatr*. 28(4). doi: 10.3389/fped.2016.00127.

Burström, Å., Bratt, E-L., Frenckner, B., Nisell, M., Hanséus K., Rydberg, A., Öjmyr-Joelsson, M. (2017). Adolescents with congenital heart disease: their opinions about the preparation for transfer to adult care. *Eur J Pediatr.* 176(7), pp.881-889. doi: 10.1007/s00431-017-2917-9.

Carona, C., Silva, N., Moreira, H., Canavarro, M. C., Bullinger, M. (2014). Does the small fit them all? The utility of Disabkids-10 Index for the assessment of pediatric health-related quality of life across age-groups, genders, and informants. *J Child Health Care.* 19(4), pp.466-77. doi: 10.1177/1367493514522867.

Carter, B., Bray, L., Dickinson, A., Edwards, M., Ford, K. (2016) *Child-centred nursing: promoting critical thinking*. London: SAGE Publication Ltd.

Chong, L. S. H., Fitzgerald, D. A., Craig, J. C., Manera, K. E., Hanson, C. S., Celermajer, D., Ayer, J., Kasparian, N. A., Tong, A. (2018). Children's experiences of congenital heart disease: a systematic review of qualitative studies. *Eur J Pediatr*. 177(3), pp.319-336. doi: 10.1007/s00431-017-3081-y.

Coad, J., Gibson, F., Horstman, M., Milnes, L., Randall, D., Carter, B. (2015). Be my guest! Challenges and practical solutions of undertaking interviews with children in the home setting. *J Child Health Care.* 19(4), pp.432-43. doi: 10.1177/1367493514527653.

Cohen. J. W. (1988). *Statistical power for behavioral science*. Hillsdale, NJ: Lawrence Erlbaum Associates.

Connor, J. A., Kline, N. E., Mott, S., Harris, S. K., Jenkins, K. J. (2010). The meaning of cost for families of children with congenital heart disease. *J Pediatr Health Care.* 24(5), pp.318-25. doi: 10.1016/j.pedhc.2009.09.002.

Costello, J. M., Mussatto, K., Cassedy, A., Wray, J., Mahony, L., Teele, S. A., Brown, K. L., Franklin, R. C., Wemovsky, G., Marino, B. S. (2015). Prediction by clinicians of quality of life and adolescents with cardiac disease. *J Pediatr.* 166(3), pp.679-83. doi: 10.1016/j.jpeds.2014.11.061.

Coyne, I., Hallström, I., Söderbäck, M. (2016). Reframing the focus from a family-centred to a child-centred care approach for children's healthcare. *J child Health Care.* 20(4), pp.949-502. doi: 10.1177/1367493516642744.

Coyne, I., Hallström, I., Söderbäck, M. (2018). Centeredness in healthcare: A concept synthesis of family-centered care, person-centered care and child-centered care. *J Pediatr Nurs.* 42, pp.45-56. doi: 10.1016/j.pedn.2018.07.001.

Creemens, J., Eiser, C., Blades, M. (2006). Characteristics of health-related self-report measures for children aged three to eight years: a review of the literature. *Qual Life Res.* 15(4), pp.739-54. doi: 10.1007/s11136-005-4184-x.

Crotty, M. (2010). *The foundations of social research: meaning and perspective in the research process.* London: Thousand Oaks: New Delhi: SAGE Publications.

Dai, W-S., Lin, W-H., Lin, C-H., Chen, Q., Cao, H. (2023). Postoperative health-related quality of life in children with congenital heart disease: a short term follow-up study. *J Cardiothorac Surg.* 18(1), 17. doi: 10.1186/s13019-023-02110-x.

Daliento, L., Mapelli, D., Volpe, B. (2006). Measurement of cognitive outcome and quality of life in congenital heart disease. *Heart.* 92(4), pp.596-74. doi: 10.1136/hrt.2004.057273.

Derridj, N., Bonnet, D., Calderon, J., Amedro, P., Bertille, N., Lelong, N., Goffinet, F., Khoshnood, B., Guedj, R. (2022). Quality of life of children born with a congenital heart defect. *J Pediatr.* 244, pp.148-153. doi: 10.1016/j.jpeds.2022.01.003.

Dodge-Khatami, A. (2016). Advances and research in congenital heart disease. *Transl Pediatr.* 5(3), pp.109-111. doi:10.21037/tp.2016.05.01.

Drotar, D. (1998). *Measuring health-related quality of life in children and adolescents: implications for research and practice.* New York: Lawrence Erlbaum Associates, Inc, Publishers.

Drotar, D. (2004). Validating measures of pediatric health status, functional status, and health- related quality of life: key methodological challenges and strategies. *Ambul Pediatr.* 4, pp.358-64. doi: 10.1367/A03-101R.1.

Eagleson, K. J., Justo, R. N., Ware, R. S., Johnson, S. G., Boyle, F. M. (2013). Health-related quality of life and congenital heart disease in Australia. *J Paediatr Child health*. 49(10), pp.856-64. doi: 10.1111/jpc.12296.

Fayed, N., de Camargo, O. K., Elahi, I., Dubey, A., Fernandes, R. M., Houtrow, A., Cohen, E. (2014). Patient-important activity and participation outcomes in clinical trials involving children with chronic conditions. *Qual Life Res*. 23(3), pp.751-7. doi: 10.1007/s11136-013-0483-9.

Francis, J., Prothasis, S., George, J., Stoica, S. (2023). Patient-reported outcome measures in congenital heart surgery: a systematic review. *Cardiol Young.* 33(3), pp.337-341. 10.1017/S1047951123000057.

Franich-Ray, C., Bright, M. A., Anderson, V., Northam, E., Cochrane, A., Menahem, S., Jordan, B. (2013). Trauma reactions in mothers and fathers after their infant's cardiac surgery. *J Pediatr Psychol.* 38(5), pp.494-505. doi: 10.1093/jpepsy/jst015.

Franklin, R., Jacobs, J. P., Tchervenkov, C., Béland M. (2002). European Paeditric cardiac code short list crossmapped to STS/EACTS short list with ICD-9 & ICD-10 crossmapping. *Cardiol Young*. 12, pp. 23-49.

Gatzoulis, M.A., Balaji, S., Webber, S.A., Siu, S.C., Hokanson, J.S., Poile, C., Rosenthal, M., Nakazawa, M., Moller, J.H., Gillette, P.C., Webb, G.D., Redington, A.N. (2000). Risk factors for arrhythmia and sudden cardiac death late after repair tetralogy of Fallot: a multicentre study. *Lancet.* 356, pp.975-81.

Geva, T., Gauvreau, K., Powell, A. J., Cecchin, F., Rhodes, J., Geva, J., del Nido, P. (2010). Randomized trial of pulmonary valve replacement with and without right ventricular remodeling surgery. *Circulation.*14(122), S201-8. doi: 10.1161/CIRCULATIONAHA.110.951178.

Golfenshtein, N., Hanlon, A. L., Deatrick, J. A., Medoff-Cooper, B. (2017). Parenting stress in parents of infants with congenital heart disease and parents of healthy infants: the first year of life. *Compr Child Adolesc Nurs.* 40(4), pp.294-314. doi: 10.1080/24694193.2017.1372532.

Government offices of Sweden. (2018). *Lag om Förenta nationernas konvention om barnets rättigheter (2018:1197).* Available from: https://www.riksdagen.se/sv/dokument-lagar/dokument/svensk-forfattningssamling/lag-20181197-om-forenta-nationernas-konvention_sfs-2018-1197. [accessed April 2023].

Gramszlo, C., Karpyn, A., Demianczyk, A. C., Shillingford, A., Riegel, E., Kazak, A. E., Sood, E. (2020). Parent perspectives on family-based psychosocial interventions for congenital heart disease. *J Pediatr*. 216, pp.51-57. doi: 10.1016/j.jpeds.2019.09.059.

Hammarberg, K., Kirkman, M., de Lacey, S. (2016). Qualitative research methods: when to use them and how to judge them. *Hum Reprod.* 31(3), pp.498-501. doi: 10.1093/humrep/dev334.

Harvey, K. A., Kovalesky, A., Woods, R. K., Loan, L. A. (2013). Experiences of mothers of infants with congenital heart disease before, during, and after complex cardiac surgery. *Heart Lung.* 42(6), pp.399-406. doi: 10.1016/j.hrtlng.2013.08.009.

Helm, P. C., Kempert, S., Körten, M-A., Lesch, W., Specht, K., Bauer, U. M. M. (2017). Congenital heart disease patients´and parents´perception of disease-specific knowledge: health and impairments in everyday life. *Congenital Heart Dis*. 13(3), pp.377-383. doi: 10.1111/chd.12581.

Heusch, A., Kahl, H. J., Hensel, K. O., Calaminus, G. (2017). Health-related quality of life in paediatric patients with congenital heart defects: association with the type of heart defect and the surgical technique. *Qual Life Res.* 26, pp.3111-3117. doi: 10.1007/s11136-017-1653-y.

Holland, J. E., Cassidy, A. R., Stopp, C., White, M. T., Bellinger, D. C., Rivkin, M. J., Newburger, J. W., Demaso, D. R. (2017). Psychiatric disorders and function in adolescents with tetralogy of Fallot. *J Pediatr.* 187, pp.165-173. doi: 10.1016/j.jpeds.2017.04.048.

Holthorn, T., Brets, F., Westfall, P. (2008). Simultaneous inference in general parametric models. *Biom J.* 50(3), pp.346-63. doi: 10.1002/bimj.200810425.

Im, Y-M., Yun, T-J., Lee, S. (2018). Health condition and familial factors associated with health-related quality of life in adolescents with congenital heart disease: a cross sectional study. *Health Qual Life Outcomes*. 10, 16(1). doi: 10.1186/s12955-018-0841-y.

Jackson, A. C., Frydenberg, E., Liang, R. P-T., Higgins, R. O., Murphy, B. M. (2015). Familial impact and coping with child heart disease: a systematic review. *Pediatr Cardiol.* 36(4), pp.695-712. doi: 10.1007/s00246-015-1121-9.

Jordan, B., Franich-Ray, C., Albert, N., Anderson, V., Northam, E., Cochrane, A., Menahem, S. (2014). Early mother-infant relationships after cardiac surgery in infancy. *Arch Dis Child.* 99(7), pp.641-5. doi: 10.1136/archdischild-2012-303488.

Kosta, L., Harms, L., Franich-Ray, C., Anderson, V., Northan, E., Cochrane, A., Menahem, S., Jordan, B. (2015). Parental experiences of their infant's hospitalization for cardiac surgery. *Child Care Health Dev*. 41(6), pp.1057-65. doi: 10.1111/cch.12230.

Kwon, E. N., Mussatto, K., Simpson, P. M., Brosig, C., Nugent, M., Samyn, M. M. (2011). Children and adolescents with repaired tetralogy of Fallot report quality of life similar to healthy peers. *Congenit Heart Dis.* 6(1), pp.18-27. doi: 10.1111/j.1747-0803.2010.00481.x.

Landolt, M. A., Valsangiacomo, B., Latal, B. (2008). Health-related quality of life in children and adolescents after open-heart surgery. *J Pedaitr*. 152(3), pp.349-55. doi: 10.1016/j.jpeds.2007.07.010.

Latal, B., Helfricht, S., Fischer, J. E., Baursfield, U., Landolt, M. A. (2009). Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease: a systematic review. *BMC Pediatr.* 22(9). doi: 10.1186/1471-2431-9-6.

Lawoko, S. and Soares, J. J. F. (2006). Psychosocial morbidity among parents of children with congenital heart disease: a prospective longitudinal study. *Heart Lung*. 35(5), pp.301-14. doi: 10.1016/j.hrtlng.2006.01.004.

Lee, S and Kim, S-S. (2012). The life experiences of Korean children and adolescents with complex congenital heart disease: a qualitative study. *Nurs Health Sci*. 14(3), pp.398-404. doi: 10.1111/j.1442-2018.2012.00709.x.

Lincoln, Y. S., Guba, E. G. (1985). *Naturalistic Inquiry.* Newbury Park, California: SAGE Publications.

Liu, Y., Chen, S., Zühlke, L., Black, G. C., Choy, M-K., Li, N., Keavney, B. D. (2019). Global birth prevalence of congenital heart defects 1970-2017: updated systematic review and meta-analysis of 260 studies. *Int J Epidemiol*. 48(2), pp.455-463. doi: 10.1093/ije/dyz009.

Lum, A., Wakefield, C. E., Donnan, B., Burns, M. A., Fardell, J. E., Marshall, G. M. (2017). Understanding the school experiences of children and adolescents with serious chronic illness: a systematic meta-review. *Child Care Health Dev.* 43(5), pp.645-662. doi: 10.1111/cch.12475.

Lundström, N.R., Berggren, H., Björkhem, G., Jögi, P., Sunnegårdh, J. (2000). Centralization of pediatric heart surgery in Sweden. *Pediatr Cardiol.* 21(4), pp.353-7. doi: 10.1007/s002460010079.

Malterud, K. (2016). Theory and interpretation in qualitative studies from general practice: why and how?. *Scand J Public Health*. 44, pp.120-129. doi: 10.1177/1403494815621181.

Marino, B. S., Shera, D., Wernovsky, G., Tomlinson, R. S., Aguirre, A., Gallagher, M., Lee, A., Cho, C. J., Stern, W., Davis, L., Tong, E., Teitel, D., Mussatto, K., Ghanayem, N., Gleason, M., Gaynor, W. J., Wray, J., Helfaer, M. A., Shea, J. A. (2008). The development of the pediatric cardiac quality of life inventory: a quality of life measure for children and adolescents with heart disease. *Qual Life Res*. 17, pp.613-626. doi: 10.1007/s11136-008-9323-8.

Marino, B. S., Uzark, K., Ittenbach, R., Drotar, D. (2010a). Evaluation of quality of life in children with heart disease. *Progress in Pediatric Cardiology*. 29, pp.131-38. doi:10.1016/j.ppedcard.2010.06.008.

Marino, B., Tomlinson, R. S., Wernovsky, G., Drotar, D., Newburger, J. W., Mahony, L., Mussatto, K., Tong, E., Cohen, M., Andersen, C., Shera, D., Khoury, P. R., Wray, J., Gaynor, W. J., Helfaer, M. A., Kazak, A. E., Shea, J. A., Pediatric Cardiac Quality of Life Inventory Testing Study Consortium. (2010b). Validation of the pediatric cardiac quality of life inventory. *Pediatrics.* 126(3), pp.498-508. doi: 10.1542/peds.2009-2973.

Marino, B.S., Lipkin, P.H., Newburger, J. W., Peacock, G., Gerdes, M., Gaynor, J. W., Mussatto, K. A., Uzark, K., Goldberg, C. S., Johnson, W. H., Li, J., Smith, S.E., Bellinger, D.C., Mahle, W.T. (2012). Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American heart Association. *Circulation*. 126, pp.1143-1172. doi: 10.1161/CIR.0b013e318265ee8a.

Miatton, M., De Wolf, D., François, K., Thiery, E., Vingerhoets, G. (2007). Intellectual, neuropsychological, and behavioral functioning in children with tetralogy of Fallot. *J Thorac Cardiovasc Surg*. 133(2), pp.449-55. doi: 10.1016/j.jtcvs.2006.10.006.

Mikkelsen, G. and Frederiksen, K. (2011). Family-centered care of children in hospital- a concept analysis. *J Avd Nurs.* 67(5), pp.1152-1162. doi: 10.1111/j.1365-2648.2010.05574.x.

Mohamed, I., Stamm, R., Keenan, R., Lowe, B., Coffey, S. (2020). Assessment of disease progression in patients with repaired tetralogy of Fallot using cardiac magnetic resonance imaging: a systemic review. *Heart, Lung and Circulation*. 29, pp.1613-1620. doi.org/10.1016/j.hlc.2020.04.017.

Mokkink, L. B., van der Lee, J. H., Grootenhuis, M. A., Offringa, M., Heymans, H. S. A., The Dutch national consensus committee "Chronic diseases and health conditions in childhood. (2008). Defining chronic diseases and health conditions in childhood (0-18 years of age): national consensus in the Netherlands. *Eur J Pediatr.* 167(12), pp.1447-7. doi: 10.1007/s00431-008-0697-y.

Moreno-Medina, K., Barrera-Castañeda M., Vargas-Acevedo, C., Garcia-Torres, A. E., Ronderos, M., Huertas-Quiñones, M., Cabrera, S., Dominguez, M. T., Reyes, N. S., Dennis, R. J. (2020). Quality of life in children with infrequent congenital heart defects: cohort study with one-year of follow-up. *Health Qual Life Outcomes*. 18(1). doi: 10.1186/s12955-019-1265-z.

Mussatto, K. (2006). Adaptation of the child and family to life with a chronic illness. *Cardiol Young.* 16(3), pp.110-6. doi: 10.1017/s104795110600103x.

Neal, A. E., Stopp, C., Wypij, D., Bellinger, D. C., Dunbar-Masterson, C., DeMaso, D. R., Newburger, J. W. (2015). Predictors of health-related quality of life in adolescents with tetralogy of Fallot. *J Pediatr.* 166(1), pp.132-8. doi: 10.1016/j.jpeds.2014.09.034.

Ni, Z. H., Lv, H. T., Ding, S., Yao, Y. W. (2019). Home care experience and nursing needs of caregivers of children undergoing congenital heart disease operations: a qualitative descriptive study. *PloS One.* 14(3). doi: 10.1371/journal.pone.0213154.

Niemitz, M., Gunst, D. C. M., Hövels-Gürish, H. H., Hofbeck, M., Kaulitz, R., Galm, C., Berger, F., Nagdyman, N., Stiller, B., Borth-Bruhns, T., Konzag, I., Balmer, C., Goldbeck, L. (2017). Predictors of health-related quality of life in children with chronic heart disease. *Cardiol Young*. 27(8), pp.1455-1464. doi: 10.1017/S1047951117000440.

O´Brien, P., Evangelista, J-A., Green, A., Uzark, K. (2013). Health considerations for children with congenital heart disease in school. Part 2. NASN Sch Nurse. 28(1), pp.42-7. doi: 10.1177/1942602X12450738

O´Byrne, M. L., Glatz, A.C., Mercer-Rosa, L., Gillespie, M. J., Dori, Y., Goldmuntz, E., Kawut, S., Rome, J. J. (2015). Trends in pulmonary valve replacement in children and adults with tetralogy of Fallot. *Am J Cardiol*. 115(1), pp.118-24. doi: 10.1016/j.amjcard.2014.09.054.

Oris, L., Rassart, J., Prikken, S., Verschueren, M., Goubert, L., Moons, P., Berg, C. A., Weets, I., Luyckx, K. (2016). Illness identity in adolescents and emerging adults with type 1 diabetes: introducing the illness identity questionnaire. *Diabetes Care*. 39(5), pp.757-63. doi: 10.2337/dc15-2559.

Oris, L., Luyckx, K., Rassart, J., Goubert, L., Goosens, E., Apers, S., Arat., S Vandenberghe, J., Westhovens, R., Moons, P. (2018). Illness identity in adults with a chronic illness. *J clin Psychol Med Settings*. 25(4), pp.429-440. doi: 10.1007/s10880-018-9552-0.

Patel, B. J., Lai, L., Goldfield, G., Sananes, R., Longmuir, P. E. (2017). Psychosocial health and quality of life among children with cardiac diagnoses: agreement and discrepancies between parent and child reports. *Cardiol Young.* 27(4), pp.713-721doi: 10.1017/S1047951116001141.

Patton, M. Q. (2015). *Qualitative research & evaluation methods.* 4th ed. Thousands oak, California: SAGE Publications.

Petersen, S., Hägglöf, B., Stenlund, H., Bergström, E. (2009). Psychometric properties of the Swedish PedsQL, Pediatric quality of life inventory 4.0 generic core scales. *Acta Pediatr.* 98(9), pp.1504-12. doi: 10.1111/j.1651-2227.2009.01360.x.

Pinheiro, J., Bates, D., DebRoy, S., Sarkar, D. R core team. (2018). *Nlme: Linear and nonlinear mixed effects model*. *R package version 3.1-137.* Available online at https://www.R-project.org/. [accessed May 2018].

Polit, D. F., Beck, C. T. (2021). *Nursing Research: Generating and assessing evidence for nursing practice.*11th ed. Philadelphia: Wolters Kluwer Health/ Lippincott Williams & Wilkins.

Polit, D. F., Beck, C. T. (2022). *Essential of nursing research: Appraising evidence for nursing practice.* 10th ed. Philadelphia: Wolters Kluwer/ Lippincott Williams & Wilkins.

Provenzi, L., Grumi, S., Borgatti. (2020). Alone with the kids: tele-medicine for children with special healthcare needs during covid-19 emergency. *Front Psychol*. 9(11). doi: 10.3389/fpsyg.2020.02193.

Public health agency of Sweden. *Nationella hälsoenkäten.* Available from: https://www.folkhalsomyndigheten.se/folkhalsorapportering-statistik/om-vara-datainsamlingar/nationella-folkhalsoenkaten/. [accessed April 2023].

Pulham, R. A., Wray, J., Feinstein, Y., Brown, K., Pierce, C., Nadel, S., Pathan, N., Garralda, E., Ramnarayan, P. (2019). Feasibility and acceptability of methods to collect follow-up information from parents 12 months after their child's emergency admission to pediatric intensive care. *Pediatr Crit Care Med*. 20(4), e19-e207. doi: 10.1097/PCC.0000000000001892.

Quaye, A. A., Coyne, I., Söderbäck, M., Kristensson Hallström, I. (2019). Children's active participation in decision-making processes during hospitalisation: an observational study. *J Clin Nurs*. 28(23-24), pp.4525-4537. doi: 10.1111/jocn.15042.

R core team. (2014). *R: a language and environment for statistical computing.* Vienna: R foundation for statistical computing. Available from: http://www.R-project.org/. [accessed November 2015].

R core team. (2016). *R: a language and environment for statistical computing.* Vienna: R foundation for statistical computing. Available from: http://www.R-project.org/. [accessed May 2018].

Raj, M., Sudhakar, A., Roy, R., Champaneri, B., Sudevan, R., Kabali, C., Kumar, R. K. (2019). Health-related quality of life (HRQOL) in children and adolescents with congenital heart disease: a cross-sectional survey from South India. *BMJ Paeditr Open.* 3(1). doi: 10.1136/bmjpo-2018-000377.

Ray, L. D. (2002). Parenting and Childhood Chronicity: making visible the invisible work. *J Pediatr Nurs.* 17(6), pp.424-38. doi: 10.1053/jpdn.2002.127172.

Reinar, B., Oberhoffer, R., Ewert, P., Müller, J. (2019). Quality of life in young people with congenital heart disease is better than expected. *Arch Dis Child*. 104(2), pp.124-128. doi: 10.1136/archdischild-2017-314211.

Rempel, G. and Harrison, M. J. (2007). Safeguarding precarious survival: parenting children who have life-threatening heart disease. *Qual Health Res*. 17(6), pp.824-37. doi: 10.1177/1049732307303164.

Riley, A. W. (2004). Evidence that school-age children can self-report on their health. *Ambul Pediatr*. 4, pp.371-6. doi: 10.1367/A03-178R.1.

Sand, P., Kljajie, M., Sunnegårdh, J. (2013). The reliability of the Pediatric quality of life inventory 3.0 cardiac module ™ for Swedish children with congenital heart defects. *Nordic Psychology.* 65(3), pp.210-223. doi.org/10.1080/19012276.2013.824204.

Shearer, K., Rempel, G. R., Norris, C. M., Magill-Evans, J. (2013). "It's no big deal": adolescents with congenital heart disease. *J Pediatr Nurs*. 28(1), pp.28-36. doi: 10.1016/j.pedn.2012.03.03.

Shields, L., Zhou, H., Pratt, J., Taylor, M., Hunter, J., Pascoe, E. (2012). Family-centred care for hospitalised children aged 0-12 years. *Cochrane Database Syst Rev.* 17(10), CD004811. doi: 10.1002/14651858.CD004811.pub3.

Siebrasse, A., Allen, S., Lavoie, J., Snippen, J., Saudek, D. (2019). Implementation of quality-of-life assessment increases referrals for intervention in patients with tetralogy of Fallot. *Pediatr Cardiol*. 40(8), pp.1569-1574. doi: 10.1007/s00246-019-02188-y.

Silva, A. M., Vaz, C., Areias, M. E. G., Viera, D., Proença, C., Viana, V., Moura, C., Areias, J. C. (2011). Quality of life of patients with congenital heart diseases. *Cardiol Young*. 21(6), pp.670-6. doi:10.1017/S1047951111000576.

Sjostrom-Strand, A. and Terp, K. (2019). Parents´ experiences of having a baby with a congenital heart defect and the child's heart surgery. *Compr Child Adolesc Nurs.* 42(1), pp.10-23. doi: 10.1080/24694193.2017.1342104.

Skoglund, K., Berghammer, M., Eriksson, P., Svensson, G., Thilén, U., Dellborg, M. (2014). Decline in self-reported health (EQ-5D) over time after surgical reconstruction of the right ventricular outflow tract: a longitudinal cohort study of 103 patients. *Congenit Heart Dis*. 10(2), e54-9, doi: 10.1111/chd.12208.

Sommer, D., Pramling Samuelsson, I., Hundeide, K. (2010). *Child perspective and children's perspectives in theory and practice.* London: Springer.

Svensson Kofod, M., Wahlberg, A., Gislason, G. H. (2020). Chronic Paradoxes: a systematic review of qualitative family perspectives on living with congenital heart defects. *Qual Health Res*. 30(1), pp.119-132. doi: 10.1177/1049732319869909.

SWEDCON Årsrapport (2021). Nationellt register för medfödda hjärtsjukdomar. Available from: https://www.ucr.uu.se/swedcon/arsrapporter. [accessed April 2023].

SWEDCON (2023) Nationellt register för medfödda hjärtsjukdomar Available from: https://www.ucr.uu.se/swedcon. [accessed April 2023].

Terry, G., Hayfield, N., Clarke, V., Braun, V. (2017). Thematic analysis. In: Willig, C. and Rogers, W. S., eds. *The SAGE handbook of qualitative research in psychology*. 2nd ed. London: SAGE Publications Ltd.

The European DISABKIDS Group. (2006). *The DISABKIDS questionnaires: Quality of life questionnaires for children with chronic conditions*. Lengerich: Pabst Science Publishers.

The National Board of Health and Welfare. (2023). Hjärtkirurgi på barn och ungdomar. Available from https://www.socialstyrelsen.se/kunskapsstod-och-regler/regler-och-riktlinjer/nationell-hogspecialiserad-vard/oversikt/hjartkirurgi-barn-och-ungdomar [acessed may 2023].

Upton, P., Lawford, J., Eiser, C. (2008). Parent-child agreement across child health-related quality of life instruments: a review of the literature. *Qual Life Res*. 17(6), pp.865-913. doi: 10.1007/s11136-008-9350-5.

Uzark, K., Jones, K., Burwinkle, T. M., Varni, J. W. (2003). The pediatric quality of life inventory™ in children with heart disease. *Progress Pediatr Cardiol*. 18, pp.141-148. doi:10.1016/S1058-9813(03)00087-0.

Uzark, K., Jones, K., Slusher, J., Limbers, C. A., Burwinkle, T. M., Varni, J. W. (2008). Quality of life in children with heart disease as perceived by children and parents. *Pediatrics.* 121(5), e.1060-7. doi: 10.1542/peds.2006-3778.

Uzark, K., King, E., Spicer, R., Beekman, R., Kimball, T., Varni, J. W. (2013). The clinical utility of health-related quality of life assessment in pediatric cardiology outpatient practice. *Congenit Heart Dis.* 8(3). doi: 10.1111/chd.12002.

Van Bulck, L., Goossens, E., Luyckx, K., Oris, L., Apers, S., Moons, P. (2018). Illness identity: a novel predictor for healthcare use in adults with congenital heart disease. *J Am Heart Assoc*. 22(7). doi: 10.1161/JAHA.118.008723.

Van Bulck, L., Goossens, E., Apers, S., Moons, P., Luyckx, K. (2021). Illness identity in adults with congenital heart disease: longitudinal trajectories and associations with patient-reported outcomes and healthcare use. *J Adv Nurs.* 77(12), pp.4743-4754. doi: 10.1111/jan.14949.

van der Watt, R., Pheiffer, C., Brown, S. (2017). The psychosocial themes of children with a congenital heart defect. *J Child Adolesc Ment Health*. 29(3), pp.231-244. doi: 10.2989/17280583.2017.1405815.

Varni, J. W., Seid, M., Kurtin, P. S. (2001). PedsQL™ 4.0: reliability and validity of the pediatric quality of life inventory™ version 4.0 generic core scales in healthy and patient populations. *Med Care.* 39(8), pp.800-812.

Varni, J. W., Burwinkle, T. M., Lane, M. M. (2005). Health-related quality of life measurement in pediatric clinical practice: an appraisal and precept for future research and application. *Health Qual Life Outcomes.* 16(3). doi: 10.1186/1477-7525-3-34.

Varni, J. W., Limbers, C. A., Burwinkle, T. M. (2007). How young can children reliably and validly self-report their health-related quality of life?: an analysis of 8,591 children across age subgroups with the PedsQL 4.0 generic core scales. *Health Qual Life Outcomes*. 3(5),1. doi: 10.1186/1477-7525-5-1.

Wang, Q., Hay, M., Clarke, D., Menahem, S. (2014). Associations between knowledge of disease, depression and anxiety, social support, sense of coherence and optimism with health-related quality of life in an ambulatory sample of adolescents with heart disease. *Cardiol Young*. 24(1), pp.126-33. doi: 10.1017/S1047951113000012.

Wei, H., Roscigno, C., Hanson, C. C., Swanson, K. M. (2015). Families of children with congenital heart disease: a literature review. *Heart Lung.* 44(6), pp.494-511. doi: 10.1016/j.hrtlng.2015.08.005.

Paper I

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Original Article

Health-related quality of life in children with surgery for CHD: a study from the Swedish National Registry for Congenital Heart Disease

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Abstract *Background:* As survival of children with CHD needing surgery has improved significantly, the need for follow-up in terms of health-related quality of life has become increasingly important. In this study, we sought to describe health-related quality of life in children with CHD in relation to cardiac surgery. *Methods:* A retrospective Swedish National Registry for Congenital Heart Disease survey measured using DISABKIDS chronic generic measure-short version included 337 children (age 9–17 years; 39% girls). The majority (n = 319, 95%) of children had a biventricular heart, whereas the remaining had a univentricular heart. Cardiac surgery was performed in 197 (58%) children. Health-related quality of life was expressed as total score (100 highest) and given as medians and 10–90th percentiles. *Results:* The overall total score was 95 (88–100). Children with a biventricular heart who had undergone three or more surgeries (n = 31; 9%) had the lowest total score of 81 (61–97; $p < 0.001$). Children with two or more surgeries and those with univentricular heart were classified in NYHA II more frequently than children with one or no cardiac surgery ($p = 0.005$ and <0.001, respectively). Children with three or more surgeries and those with univentricular heart needed more help at school ($p < 0.001$). Compared with children with other chronic diseases, children with CHD had a high total score except for children with three or more surgeries who had comparable total scores with children with other chronic diseases. *Conclusion:* Children with three or more cardiac surgeries and those with a univentricular heart appear to have lower health-related quality of life, cognitive ability, and NYHA classification.

Keywords: Health-related quality of life; CHD; children; registry study

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A CARDIAC MALFORMATION OCCURS IN 1% OF neonates and represents the most common type of congenital disease. Nearly one-third of children with CHD need corrective or palliative treatment via either surgery or catheterisation.[1] With advances in surgical and postoperative care, the large majority of children nowadays reach adulthood. Therefore, long-term follow-up with regard to health-related quality-of-life measurement has become increasingly important.

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Defining quality of life as a concept for children is challenging, particularly in view of the developmental stages.[2] Nevertheless, research focussing on health-related quality of life for children with CHD has increased during the last decade.[3] Health-related quality of life is defined "as the influence of a specific illness, medical therapy, or health services policy on the ability of patients to both function in and derive personal satisfaction from various physical, psychological, and social life contexts".[4]

Previous research in the field of CHD has yielded different results. Kwon et al[5] showed that the health-related quality of life in 20 children with tetralogy of Fallot, which typically requires at least one surgery, was comparable with healthy children and was higher

compared with other chronically ill children. Similar findings were obtained by Mussatto and Twedell et al[6] that health-related quality of life of 182 children with complex CHD was higher than in those with non-cardiac chronic diseases. Uzark et al[7] reported that children with various types of CHD (n = 347) perceived their health-related quality of life to be lower than healthy children. In the same study, children with more than one cardiac surgery and need for additional surgery and those with untreatable or palliated CHD had lower physical functioning and cognitive function scores. In a multicentre study by Marino et al[8] on 1605 children, those with repaired biventricular heart disease had lower health-related quality of life than children with mild CHD, with no previous surgical or catheter-based interventions. According to the same study, an increased number of cardiac surgeries appears to decrease the health-related quality of life. This finding could not be replicated by Mussatto and Twedell,[6] who found no association between the number of open-heart operations and quality of life in children.

The discrepancy in results could be explained by differences in the data collection questionnaire, cohort size, or selected diagnoses.[9] A generic questionnaire such as the Pediatric Quality of Life Inventory (PedsQL) 4.0 generic core scales provides the advantage of comparing cardiac children with healthy and/or chronically ill children.[5,6,10] Disease-specific questionnaires for children with CHD such as the PedsQL 3.0 cardiac module and Pediatric Cardiac Quality of Life Inventory (PCQLI) have the ability to distinguish between subgroups within a certain disease.[7,8,11] DISABKIDS questionnaires are available as both generic and disease-specific versions for children with chronic conditions and have been used in studies on children with diabetes,[12] cancer,[13] and previous major trauma.[14] To the best of our knowledge, there are no published studies using DISABKIDS questionnaires in children with CHD.

Since 1994, paediatric cardiac surgery in Sweden has been centralised to only two tertiary referral centres, in Lund and Gothenburg. Approximately 600 cardiac surgeries are performed every year at these two hospitals[1] and a number of variables including health-related quality of life are stored in the Swedish National Registry for Congenital Heart Disease, which was started in 2009. In the paediatric population, the health-related quality of life is assessed using the DISABKIDS chronic generic measure-short version. As a complement to this generic short questionnaire, other measures related to health-related quality of life such as NYHA classification, cognitive function, physical activity, and psychosocial ability are added to the Swedish National Registry for Congenital Heart Disease.

The overall aim of this study was to describe the relationship between the number of surgeries for CHD in children and the DISABKIDS chronic generic measure-short version-based health-related quality of life, NYHA, cognitive function, physical activity, and psychosocial ability using data from the Swedish National Registry for Congenital Heart Disease. Another goal was to compare the health-related quality of life of children with CHD with a reference population of children with other chronic diseases.

Material and methods

The Swedish National Registry for Congenital Heart Disease

The paediatric part in the Swedish National Registry for Congenital Heart Disease contains information about medical data including diagnosis, medication, type and number of surgeries and catheter interventions, types of cardiac investigations, physical function as NYHA class and physical activity, psychosocial data as cognitive function and psychosocial ability, and health-related quality-of-life data.[1] The health-related quality-of-life data, physical activity, and psychosocial data are collected at seven hospitals, whereas other data are collected at 30 hospitals across the country.[1] Information about the Swedish National Registry for Congenital Heart Disease is given to children and their parents during their visit at the outpatient clinics. Those who agree to participate in the registry are asked to complete DISABKIDS chronic generic measure-short version and to answer additional questions regarding NYHA, cognitive function, physical activity, and psychosocial ability. In this study, the degree of registry completeness was 91%.

Data collection

Children who attended the outpatient clinic at Pediatric Heart Center of the Skåne University Hospital in Lund between February, 2009 and September, 2014 were included. All data (detailed below) were retrieved from the Swedish National Registry for Congenital Heart Disease. Inclusion criteria were as follows: history of CHD, between 9 and 17 years, and completion of DISABKIDS chronic generic measure-short version. The registry variables included health-related quality of life, demographics, cardiac diagnosis, and previous cardiac surgeries including information on type, number, and age at surgery, NYHA, cognitive function, physical activity, and psychosocial ability. Children with a biventricular

heart were divided into four groups based on the number of cardiac surgeries: no cardiac surgery, one cardiac surgery, two cardiac surgeries, and three or more cardiac surgeries. Children with a univentricular heart were grouped separately. Cardiac surgery was defined as an operation on the heart or the intrathoracic great vessels through a sternotomy or thoracotomy in either curative or palliative purpose.

DISABKIDS chronic generic measure-short version

The DISABKIDS chronic generic measure-short version is a self-reported questionnaire earlier validated for use in children from 8–17 years of age with chronic disease[2] and was developed by "The DISABKIDS group Europe", a cooperation project between Germany, Sweden, France, the Netherlands, Austria, and the United Kingdom.[15] The DISABKIDS chronic generic measure-short version assesses health-related quality of life of children with disabilities in relation to subjective and social well-being with special emphasis on health care and medical treatment.[2] The questionnaire includes 12 items – 10 items excluding medications – measuring mental (four items), social (four items), and physical (four/two items) impact on the health condition.[15] Each question considers the last 4-week period, and the response for each item is graded using a five-point scale, indicating frequency of behaviours or feelings as 1 = never, 2 = seldom, 3 = quite often, 4 = very often, and 5 = always. The higher the score (maximum 100), the higher the self-perceived health-related quality of life. Percentiles for self-perceived health-related quality of life are integrated in the DISABKIDS chronic generic measure-short version from a reference population of children of the same age and gender with other chronic conditions. This enables a comparison with children with asthma, arthritis, dermatitis, diabetes, cerebral palsy, cystic fibrosis, and epilepsy.[15]

NYHA, cognitive function, physical activity, and psychosocial ability

Questions about these variables were addressed to children and their parents together. NYHA[16] ranged from (1–4) "capacity like healthy peers" to "symptom at rest"; cognitive function ranged from (1–4) "regular school" to "severe impairment learning ability"; psychosocial ability[17] ranged from (1–4) "no significant conflicts with friends" to "permanent contact problems"; and physical activity ranged from (1–3) "no extra physical activity" to "more than 3 hours physical activity per week". Moreover, two additional options were added to the variables NYHA, cognitive function, and psychosocial ability: "not classifiable/mainly limited by non-cardiac conditions" and "unknown". For the variable physical activity, "unknown" was added.

Statistical analysis

Analyses for group comparison and descriptive statistics were performed using SPSS version 21.0 (SPSS IBM, New York, United States of America). The remaining analyses were carried out using R version 3.1.2.[18] The DISABKIDS chronic generic measure-short version total score is given as median and as a measure of dispersion at the 10 to 90th percentiles. As these data were skewed, non-parametric tests such as the Kruskal–Wallis with Monte Carlo p values based on 10,000 samples and exact Mann–Whitney U-test when appropriate were used to assess the differences. The total score from The DISABKIDS chronic generic measure-short version were transformed from a raw score to a range of 0–100. Thus, a "standardised raw score" = ((point – 12)/48) × 100 (if 12 questions were answered)) or = ((point – 10)/40) × 100 (if 10 questions were answered)). A p value <0.05 was considered to be statistically significant. Post-hoc tests were Bonferroni-adjusted for multiple comparisons.

In order to adjust for confounding factors, a model for the health-related quality of life in the different groups was made. Transformed scores, 100 – standardised score, were analysed using a quasi-Poisson model with identity link to allow for remaining over-dispersion and to correct for the variance structure. Estimates were then converted back to the standardised scale. All pairwise interactions were assessed; interactions with $p < 0.05$ were retained.

For each of the four groups with children with a biventricular heart and for the one with a univentricular heart, the confidence interval for the expected quantiles was found by simulating 1000 replicates with as many observations as those present in a specific group. The 2.5th and 97.5th bootstrapped empirical percentiles for each expected quantile were then used to provide the confidence intervals for the expected pattern in Figure 2. The observed quantiles of the DISABKIDS chronic generic measure-short version score were also compared with the expected distribution using a Kolmogorov–Smirnov test.

Results

In total, 337 children who completed the DISABKIDS chronic generic measure-short version were included. The median age (10–90th) was 14 years (9–17 years) and 39% were girls.

The median time between surgery and DISABKIDS chronic generic measure-short version was 10 years (6–14 years). The main demographic surgical and pharmacological characteristics are detailed in Table 1. The majority (n = 319, 95%) had a biventricular heart. Of these, 140 (42%) children had no cardiac surgery, 117 (35%) had one cardiac surgery, 31 (9%) had two surgeries, and 31 (9%) had three or more cardiac surgeries. In total, 18 (5%) children had univentricular heart surgeries (Table 1). The main cardiac diagnoses are shown in Table 2.

The DISABKIDS chronic generic measure-short version

The overall total score was 95 (75–100) with no difference in gender ($p > 0.3$). When children with biventricular heart and univentricular heart were grouped together, those with three or more cardiac surgeries had a lower total score than those with no, one, or two cardiac surgeries ($p < 0.001$; Fig 1a). When children with univentricular heart were grouped separately, the total score remained significantly lower for those with biventricular heart and three or more surgeries ($p < 0.001$; Fig 1b). Children with a univentricular heart had a lower total score than those with one or no surgery ($p = 0.005$, Fig 1b). The total score for children with three or more cardiac surgeries was comparable with that for children with other chronic diseases; however, the total score for children with no, one, or two cardiac surgeries ($p < 0.001$) and for those with univentricular heart ($p = 0.047$) was higher compared with children with other chronic diseases (Fig 2). Among those with cardiac surgery, there was no difference ($p > 0.3$) in total score between children undergoing their first surgery before 1 month of age

Table 1. Demographic, pharmacological, and surgical characteristics of the study groups.

	No cardiac surgery (n = 140)	1 cardiac surgery (n = 117)	2 cardiac surgeries (n = 31)	⩾3 cardiac surgeries (n = 31)	UVH (n = 18)	Total (n = 337)
Age (median (10–90th percentiles))	14 (9–17)	14 (9–16)	14 (9–17)	14 (9–16)	15 (9–16)	14 (9–17)
Boys (n (%))	80 (57)	82 (70)	17 (55)	16 (52)	9 (50)	204 (61)
Number of children with 1st surgery within 1st month (n (%))		32 (27)	9 (29)	14 (45)	14 (78)	69 (20)
Number of children on medications (n (%))	23 (16)	29 (25)	5 (16)	13 (42)	17 (94)	87 (26)

UVH = univentricular heart

Table 2. Main cardiac diagnoses in the study groups.

	No cardiac surgery (n (%))	1 cardiac surgery (n (%))	2 cardiac surgeries (n (%))	⩾3 cardiac surgeries (n (%))	Total (n (%))
Aortic anomalies	5 (4)	22 (19)	9 (29)	3 (10)	39 (12)
ASD	21 (15)	14 (12)	0	0	35 (10)
VSD	37 (26)	19 (16)	2 (6)	0	58 (17)
PDA	7 (5)	1 (1)	0	0	8 (2)
Aortic valve anomalies	32 (23)	8 (7)	0	3 (10)	43 (13)
Tetralogy of Fallot	0	14 (12)	11 (35)	11 (35)	36 (11)
Congenitally corrected transposition	0	0	1 (3)	1 (3)	2 (1)
Mitral valve anomalies	15 (11)	2 (2)	0	0	17 (5)
Pulmonary valve anomalies	19 (14)	7 (6)	2 (6)	3 (10)	31 (9)
TGA	0	18 (15)	3 (10)	4 (13)	25 (7)
Tricuspid valve anomalies	1 (1)	1 (1)	0	0	2 (1)
Truncus	0	0	2 (6)	4 (13)	6 (2)
AVSD	0	8 (7)	1 (3)	2 (6)	11 (3)
UVH	0	0	4*	8*	12 (4)
HLHS	0	0	0	6*	6 (2)
Others	3 (2)	3 (3)	0	0	6 (2)
Total	140	117	31	31	337

ASD = atrial septal defect; AVSD = atrioventricular septal defect; HLHS = hypoplastic left heart syndrome; PDA = persistent ductus arteriosus; TGA = transposition of the great arteries; UVH = univentricular heart; VSD = ventricular septal defect
*Only included in the UVH group

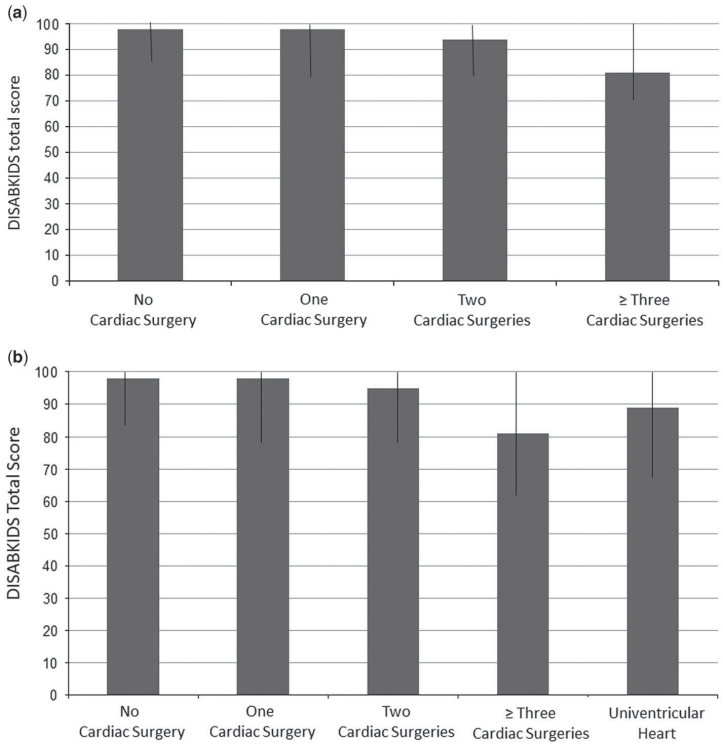

Figure 1.
(a) The DISABKIDS chronic generic measure-short version total score in children grouped according to the number of surgeries. (b) Children with univentricular heart are grouped separately. Data are shown as median (10–90th percentiles).

and those with surgery beyond 1 month of age. The results of the items included in the DISABKIDS chronic generic measure-short version are detailed in Table 3. Although we could not observe any interaction between NYHA and cardiac surgeries, we studied children classified in NYHA I (Table 4) and found that children with three or more surgeries had lower total scores than those with none or one surgery ($p > 0.001$) or those with two surgeries ($p = 0.002$). The internal consistency for the DISABKIDS chronic generic measure-short version expressed by Cronbach's α was 0.84.

NYHA, cognitive function, physical activity, and psychosocial ability

Data on NYHA, physical activity, cognitive function, and psychosocial ability were unavailable for five and nine children, respectively. The majority of respondents were classified in NYHA I ($n = 290$, 87%) and attended regular school ($n = 293$, 88%). The groups with two ($p = 0.005$), three, or more surgeries ($p < 0.001$) and univentricular heart ($p < 0.001$) had a larger number of children in NYHA II than those with no or one cardiac surgery (Fig 3). Nearly all children in NYHA III were among those with three or more surgeries and those with a univentricular heart ($p < 0.001$). Children with three or more surgeries and those with a univentricular heart needed more help in school than children with no or one cardiac surgery ($p < 0.001$). No significant differences between the groups of children were found in terms of "physical activity" and "psychosocial ability" (Fig 3).

Adjusted model

In total, 38% of the deviance for the total score in DISABKIDS chronic generic measure-short version was explained in the quasi-Poisson regression model (Tables 5 and 6). Variables that predicted lower total score were "NYHA", "number of cardiac surgery",

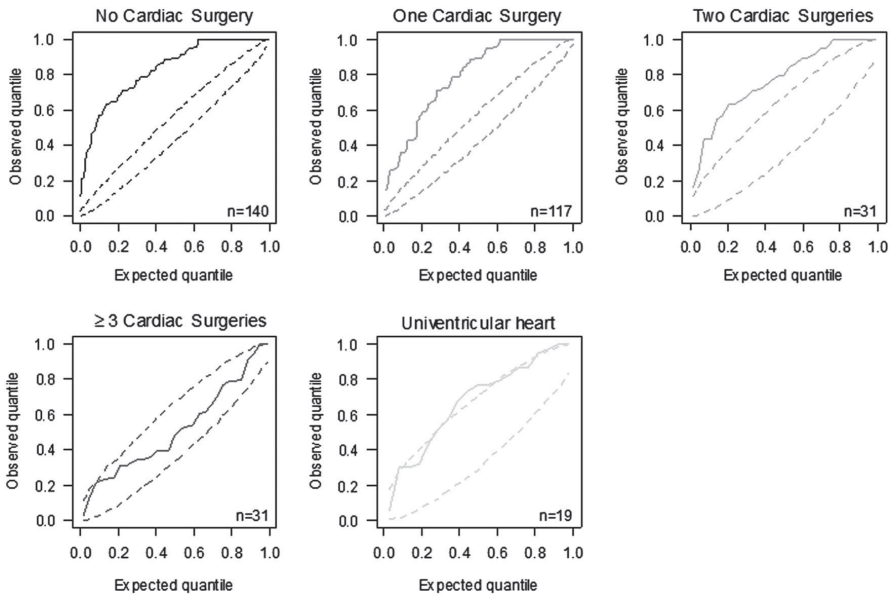

Figure 2.
The distribution of the DISABKIDS chronic generic measure-short version total score in relation to the reference score for children with other chronic disease. The full line denotes total score for children with CHD; the area within the dotted lines includes total score for children with other chronic disease.

"cognitive function", "psychosocial ability", and "physical activity and gender".

Discussion

In a retrospective survey of a nationwide registry for CHD including a relatively large cohort from a tertiary referral centre for paediatric cardiac surgery, we found that children with a biventricular heart and three or more cardiac surgeries and children with a univentricular heart had lower health-related quality of life, were more frequently classified in NYHA II or III, and needed help more often in school. To the best of our knowledge, this is the first registry-based study using DISABKIDS chronic generic measure-short version to assess health-related quality of life of children with CHD.

In our cohort, the majority of cardiac surgeries were performed on bypass ("open-heart"), and nearly one-third were relatively complex, including corrective or palliative surgeries in children with tetralogy of Fallot, transposition of the great arteries, and aortic valve disease. Importantly, the distribution of these surgeries (detailed in Table 2) resembled the overall distribution of paediatric cardiac surgeries at our centre. In a comprehensive review of previous randomised, case-controlled, and cohort studies, Latal et al[9] noticed that children with open-heart surgery for CHD are at risk for impaired quality of life. The authors' call for future self-report studies addressing the quality of life in children and adolescents with cardiac surgery remains important. Other aspects related to cardiac surgery deserve attention too. In the multi-centre study by Marino,[8] both the disease severity and the frequency of cardiac surgeries were associated with lower health-related quality of life. The main finding in our study indicating lower quality of life in children with three or more cardiac surgeries is thus in line with previous studies showing an impaired health-related quality of life in children with more complex heart disease.[8–11,19] On the other hand, as suggested in a previous study by Ternestedt et al 20 and 30 years after surgery for atrial septal defect and tetralogy of Fallot,[20] the complexity of CHD is not necessarily congruent with certain aspects related to the health-related quality of life.

Comparison with children with other chronic diseases

In our study, 306 children (92%) had a higher total score compared with children of the same age and

Table 3. Items included in the DISABKIDS chronic generic measure-short version.

	No cardiac surgery (median (10–90th percentiles))	1 cardiac surgery (median (10–90th percentiles))	2 cardiac surgeries (median (10–90th percentiles))	⩾3 cardiac surgeries (median (10–90th percentiles))	UVH (median (10–90th percentiles))
1. Do you feel like everyone else though you have your condition?	5 (4–5)	5 (3–5)	5 (3–5)	4 (3–5)	5 (3–5)
2. Are you free to lead the life you want even though you have your condition?	5 (4–5)	5 (4–5)	5 (4–5)	4 (3–5)	5 (3–5)
3. Is your life ruled by your condition?	1 (1–3)	1 (1–3)	1 (1–2)	2 (1–4)	2 (1–3)
4. Does your condition bother you when you play or do other things?	1 (1–3)	1 (1–3)	1 (1–3)	2 (1–4)	2 (1–4)
5. Are you unhappy because of your condition?	1 (1–2)	1 (1–2)	1 (1–2)	1 (1–3)	1 (1–2)
6. Does your condition get you down?	1 (1–2)	1 (1–2)	1 (1–3)	1 (1–3)	1 (1–3)
7. Do you feel lonely because of your condition?	1 (1–1)	1 (1–1)	1 (1–2)	1 (1–3)	1 (1–3)
8. Do you feel different from other children/adolescents?	1 (1–2)	1 (1–3)	1 (1–2)	2 (1–4)	1 (1–3)
9. Do you think that you can do most things as well as other children/adolescents?	5 (4–5)	5 (4–5)	5 (4–5)	4 (3–5)	5 (3–5)
10. Do your friends enjoy being with you?	5 (5–5)	5 (5–5)	5 (4–5)	5 (4–5)	5 (4–5)
11. Does taking medication bother you?	1 (1–5) n = 23	1 (1–3) n = 29	1 (1–5) n = 5	2 (1–4) n = 13	1 (1–3) n = 17
12. Do you hate taking your medicine?	1 (1–3) n = 23	1 (1–3) n = 29	1 (1–3) n = 5	1 (1–4) n = 13	1 (1–4) n = 17

UVH = univentricular heart
Each item is graded using a five-point scale, indicating frequency of behaviours or feelings as 1 = never; 2 = seldom; 3 = quite often; 4 = very often; 5 = always. 5 is the highest total score for items 1, 2, 9, and 10. For the other items, 1 is the highest total score
Data are shown as median (10–90th percentiles)

Table 4. DISABKIDS chronic generic measure-short version total score for children in NYHA I in the study groups.

NYHA I	n (%)	Total score	p value
No cardiac surgery (0)	134 (46)	98 (85–100)	0–3 < 0.001
1 cardiac surgery (1)	102 (35)	98 (79–100)	1–3 < 0.001
2 cardiac surgery (2)	24 (8)	97 (84–100)	0–2 = 0.002
⩾3 cardiac surgery (3)	20 (7)	83 (63–100)	
UVH (4)	10 (3)	95 (72–100)	

UVH = univentricular heart
The total score is shown as median (10–90th percentiles)

gender with other chronic diseases; this is comparable with results from previous studies, although a different questionnaire was used.[5,6] The reference population in the DISABKIDS questionnaire is composed of European children with asthma, arthritis, dermatitis, diabetes, cerebral palsy, cystic fibrosis, and epilepsy. The differences between healthcare systems in European countries may have an impact on the result; however, in another study,[12] Swedish children with diabetes responded to DISABKIDS chronic generic measure-long version and the median of total score was 80. Importantly, the total score in the long version is comparable with that of the short version.[15] Children born with diseases, similar to the children in our study, may rate their health-related quality of life independent of their functional or health state because they have no perception of another health state.[21] The difference between

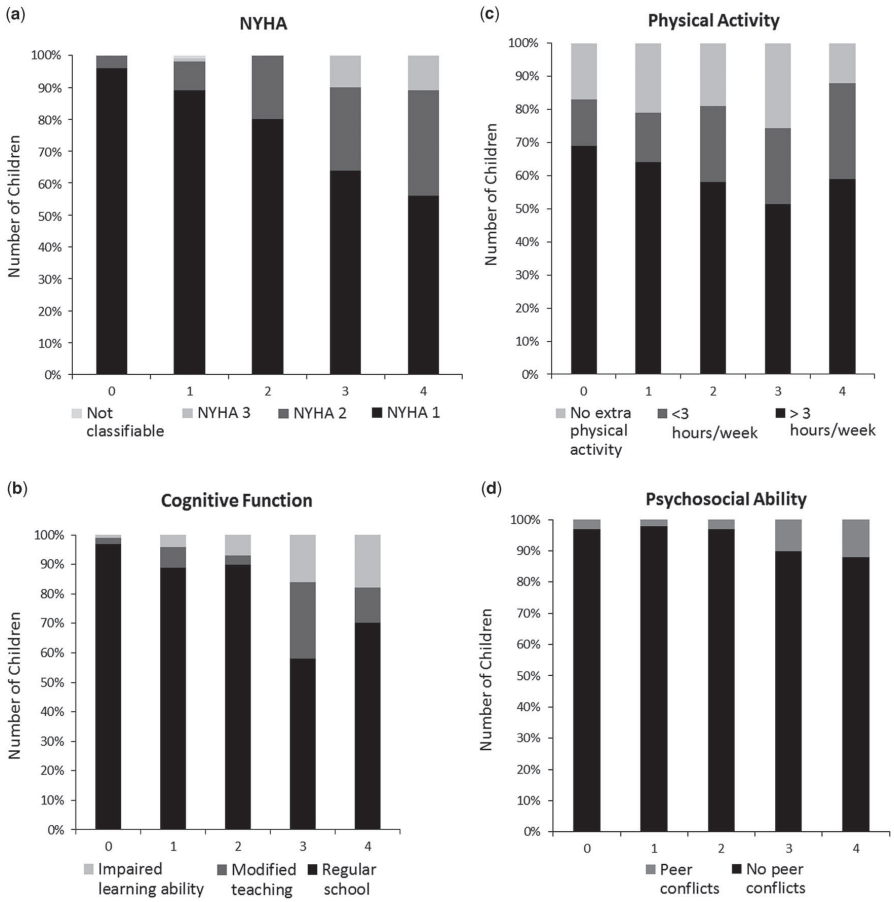

Figure 3.
*Supplementary variables in the Swedish National Registry for Congenital Heart Disease: 0 = no cardiac surgery, 1 = one cardiac surgery, 2 = two cardiac surgeries, 3 = three or more cardiac surgeries, 4 = univentricular heart. For the variable "cognitive function", the answer options 3–4 are merged; for the variable "psychosocial ability", 2–4 are merged. (**a**) NYHA; (**b**) Cognitive function; (**c**) Physical activity; (**d**) Psychosocial ability.*

acquired and congenital disease could have had an impact on the results of our study.

NYHA

NYHA is a subjective index of heart-related physical capacity and was earlier used as a surrogate in quality-of-life research.[3] A study by Berghammer et al[22] showed that NYHA was a predictor of self-reported health status according to the generic Euro QoL-5 Dimension questionnaire responded by adults with CHD. In our study, a decreased total score was associated with NYHA II and III, suggesting a possible link between decreased health-related quality of life, NYHA classification, and the complexity of heart disease. As children in NYHA I with three or more surgeries had lower total score than the rest of children in NYHA I, it is conceivable that the complexity of the heart disease rather than NYHA accounts for the decreased health-related quality of life.

Cognitive function

The association between CHD and cognitive function has been assessed in several studies, with results suggesting a link between severity of CHD and impaired cognitive function.[7,11,19,23] In our study, cognitive ability was estimated by the need for

Table 5. Estimates from the quasi-Poisson model.

	Estimate	SE	p value	n
Intercept*	94.06	1.32	<0.001	
NYHA class				
NYHA II	−11.4	2.55	<0.001	33
NYHA III	−17.1	8.07	0.03	5
Psychosocial ability				
Peer conflict	−9.81	3.79	0.01	11
Age				
10 years old	0.398	1.65	>0.3	44
11 years old	4.71	1.31	<0.001	31
12 years old	3.16	2.34	0.17	8
13 years old	3.26	2.06	0.11	11
14 years old	−0.063	1.97	>0.3	51
15 years old	1.91	1.54	0.21	51
16 years old	0.836	1.62	>0.3	44
17 years old	−0.307	1.81	>0.3	34
Cognitive ability				
Adaptation of teaching	−2.25	2.38	0.34	21
Impaired learning ability	−6.73	3.28	0.04	17
Cardiac surgery				
1 cardiac surgery	0.372	0.794	>0.3	111
2 cardiac surgery	−0.741	1.37	>0.3	29
⩾3 cardiac surgery	−7.28	2.42	0.002	30
UVH	−1.72	2.6	>0.3	15
Physical activity and gender				
>3 hours/week physical activity and girl	−0.119	0.91	>0.3	70
No extra physical activity and boy	−2.68	1.66	0.107	39
No extra physical activity and girl	−1.76	1.87	0.34	23
<3 hours/week physical activity and boy	−9.02	2.92	0.002	19
<3 hours/week physical activity and girl	1.23	0.749	0.1	33

UVH = univentricular heart

*Intercept represent: boys (n = 198), age = 9 years (n = 50), no cardiac surgery (n = 139), NYHA I (n = 286), >3 hours/week physical activity and boy (n = 140), no conflicts with peers (n = 313), regular school (n = 286)

Table 6. Analysis of variance table of deviances.

Parametric terms	df	F	p value
NYHA class	2	11.448	<0.001
Physical activity	2	5.725	0.003
Gender	1	0.017	>0.3
Physical activity and gender	2	5.625	0.004
Psychosocial ability	1	6.718	0.01
Age	8	4.153	<0.001
Cognitive ability	2	2.434	0.089
Cardiac surgery	4	2.576	0.037

df = degree of freedom; F = observed F-statistic

R^2 (adjusted) = 0.381, n = 324

aid at school, which is a fairly rough measure of cognitive impairment. Further research is needed to better understand the link between health-related quality of life, neurodevelopment, and psychosocial factors.[23]

Physical activity and gender

In the study by Mueller et al,[24] boys reported higher scores than girls in one item, physical well-being. In this study, 168 children with tetralogy of Fallot responded to self-reported KINDL-R quality-of-life questionnaire. This is consistent with our result showing that boys with <3 hours of physical activity a week had a decreased total score.

There were no significant differences in total score between children with one, two, or no cardiac surgery. This finding can indicate that fewer surgeries have no impact on total score or that DISABKIDS chronic generic measure-short version is not sufficiently sensitive to discriminate between these groups. A drawback with this questionnaire is that there is no possibility to divide the result into domains, as with a longer questionnaire. The shorter time required to complete the questionnaire is clearly an advantage.

In order to achieve a better understanding at a group level for children with CHD, categorising is necessary because of the mixed spectrum of CHD with both mild and complex diagnoses. Categories in previous research were severity of CHD[19], separate diagnoses,[20] or cardiac surgery[8]. In our study, children were categorised based on the number of cardiac surgeries, as this illustrates the complexity of

CHD and enables rough delineation between mild and complex CHD within the same diagnosis.

Study limitations

The overall cohort size, the relatively low number of children with two or more surgeries, and children with univentricular heart may have influenced the results. Although the rate of response in our study was very high, the non-responders may choose to do so for various reasons including different perception of the utility of long-term health data acquisition. In addition, there are other potential ways to assess cognitive function and physical ability, such as intelligence test and exercise test, respectively, which were not used in our study. Future studies may be important to interrogate the correlation of these indices to those derived from the questionnaire.

Conclusion

Children with CHD estimated their health-related quality of life to be higher than children with other chronic diseases, except for children with a biventricular heart and three or more cardiac surgeries. Children with higher number of cardiac surgeries, probably coincident with more severe cardiac disease, appear to have lower health-related quality of life, cognitive ability, and NYHA. Thus, health-related quality-of-life variables in the Swedish National Registry for Congenital Heart Disease could be used to identify children at risk for lower health-related quality of life. A validated instrument for health-related quality of life in children with CHD is important for future "interventional" studies aimed to improve the health-related quality of life in risk patients.

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Conflicts of Interest

None.

Ethical Standards

The study was approved by the Ethics Committee for Human Research at the Lund University (#2011/749).

References

1. About Swedcon: Swedish National Registry for Congenital Heart Disease 2015, Retrieved November 2015 from http://www.ucr.uu.se/swedcon.
2. Bullinger M, Schmidt S, Petersen C, The DISABKIDS group. Assessing quality of life of children with chronic health conditions and disabilities: a European approach. Int J Rehabil Res 2002; 25: 197–206.
3. Bratt EL, Moons P. Forty years of quality-of-life research in congenital heart disease: temporal trends in conceptual and methodological rigor. Int J Cardiol 2015; 195: 1–6.
4. Costello JM, Mussatto K, Cassedy A, et al. Prediction by clinicians of quality of life for children and adolescents with cardiac disease. J Pediatr 2015; 166: 679–683.
5. Kwon EN, Mussatto K, Simpson PM, Brosig CB, Nugent M, Samyn MM. Children and adolescents with repaired tetralogy of Fallot report quality of life similar to healthy peers. Congenit Heart Dis 2011; 6: 18–27.
6. Mussatto K, Twedell J. Quality of life following surgery for congenital cardiac malformations in neonates and infants. Cardiol Young 2005; 15: 174–178.
7. Uzark K, Jones K, Slusher J, Limbers CA, Burwinkle TM, Varni JW. Quality of life in children with heart disease as perceived by children and parents. Pediatrics 2008; 121: e1060–e1067.
8. Marino BS, Tomlinson RS, Wernovsky G, et al. Validation of the pediatric cardiac quality of life inventory. Pediatrics. 2010; 126: 498–508.
9. Latal B, Helfricht S, Fischer JE, Bauersfeld U, Landolt MA. Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease: a systematic review. BMC Pediatr 2009; 9: 6. doi:10.1186/1471-2431-9-6.
10. Eagleson KJ, Justo RN, Ware RS, Johnson SG, Boyle FM. Health-related quality of life and congenital heart disease in Australia. J Pediatr Child Health 2013; 43: 856–864.
11. Sand P, Kljajic M, Sunnegårdh J. The reliability of the pediatric quality of life inventory 3.0 cardiac module for Swedish children with congenital heart defects. Nord Psychol 2013; 9: 1–14.
12. Chaplin JE, Hanas R, Lind A, Tollig H, Wrammer N, Lindblad B. Assessment of childhood diabetes – related quality-of-life in West Sweden. Acta Paediatr 2009; 98: 361–366.
13. Sandeberg M, Johansson E, Björk O, Wettergren L. Health-related quality of life relates to school attendance in children on treatment for cancer. J Pediatr Oncol Nurs 2008; 25: 265–274.
14. Janssens L, Gorter JW, Ketalaar M, Holtslag HR. Health-related quality-of-life measures for long-term follow-up in children after major trauma. Qual Life Res 2008; 17: 701–713.
15. The European DISABKIDS Group. The DISABKIDS Questionnaires Quality of Life Questionnaires for Children with Chronic Conditions-Handbook. Pabst Science Publishers, Lengerich, Germany, 2006.
16. The Criteria Committee for the New York Heart Association. Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels, 9th ed. Little Brown and Company, Boston, Massachusetts, 1994: 253–255.
17. Birkeland A, Rydberg A, Hägglöf B. The complexity of the psychosocial situation in children and adolescents with heart disease. Acta Pædiatr 2005; 94: 1495–1501.

18. R Core Team 2014, R: a language and environment for statistical computing. R Foundation for Statistical Computing, Vienna. Retrieved November 2015 from http://www.R-project.org/.
19. Tahirović E, Begić H, Nurkić M, Tahirović H, Varni JW. Does the severity of congenital heart defects affect disease-specific health-related quality of life in children in Bosnia and Herzegovina? Eur J Pediatr 2010; 169: 349–353.
20. Ternestedt BM, Wall K, Oddsson H, Riesenfield T, Groth I, Schollin J. Quality of life 20 and 30 years after surgery in patients operated on for tetralogy of Fallot and for atrial septal defect. Pediatr Cardiol 2001; 22: 128–132.
21. Davis E, Waters E, Mackinnon A, et al. Paediatric quality of life instruments: a review of the impact of the conceptual framework on outcomes. Dev Med Child Neurol 2006; 48: 311–318.
22. Berghammer M, Karlsson J, Ekman I, Eriksson P, Dellborg M. Self-reported health status (EQ-5D) in adults with congenital heart disease. Int J. Cardiol 2013; 165: 537–543.
23. Marino BS, Uzark K, Ittenbach R, Drotar D. Evaluation of quality of life in children with heart disease. Prog Pediatr Cardiol 2010; 29: 131–138.
24. Mueller GC, Sarikouch S, Beerbaum P, et al. Health-related quality of life compared with cardiopulmonary exercise testing at the midterm follow-up visit after tetralogy of Fallot repair: a study of the German competence network for congenital heart defects. Pediatr Cardiol 2013; 34: 1081–1087.

Paper II

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Health-Related Quality of Life in Children With Earlier Surgical Repair for Right Ventricular Outflow Tract Anomalies and the Agreement Between Children and Their Parents

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Background: Children diagnosed with right ventricle outflow tract (RVOT) anomalies require surgical repair early in life, reoperations and lifelong follow-up. The aim is to comprehensively describe their health related quality of life (HRQoL) and to assess the agreement in this regard between children and parents.

Methods and Results: Child- and parent-reported HRQoL was assessed in 97 children aged 8–18 years using three different HRQoL questionnaires. The mean age was 12.9 ± 3 years. The mean total score for the child report was lower in the PedsQL Cardiac Module than in the PedsQL 4.0 and DISABKIDS ($p \leq 0.001$). The mean score for each domain in PedsQL Cardiac Module ranged between 67 (cognitive function) and 79 (physical appearance), and between 72 (school function) and 82 (physical and social function) in PedsQL 4.0. Nearly half of the children reported problems with shortness of breath during physical activity. In the PedsQL Cardiac Module the child-parent agreement was strong for 13 of 22 items.

Conclusion: HRQoL problems as perceived by children with RVOT anomalies are best identified with the PedsQL Cardiac Module and relate mostly to cognitive and physical functioning. The agreement findings suggest the need to take into account both child- and parent reports in the assessment of HRQoL.

Keywords: complex right ventricular outflow tract anomalies, HRQoL, children, agreement, PedsQL questionnaire

INTRODUCTION

Children With Right Ventricular Outflow Tract Anomalies

Children with RVOT anomalies include a heterogeneous group of congenital heart defects with need for early treatment and a multifaceted clinical assessment during follow-up. The surgical repair is usually performed during the first months of life and many of the children will eventually need one or more reoperations before adult age, often due to a leaking pulmonary valve or re-stenosis of the reconstructed outflow tract (1). Once a conduit is implanted, later re-interventions are necessary since conduits undergo degenerative changes and secondary worsening of the valve function. Delayed pulmonary valve replacement will result in adverse right ventricle remodeling

with subsequently increased risk for cardiac events in later life, such as ventricular arrhythmias, heart failure and death. Optimizing time for re-intervention is therefore essential in order to avoid such complications (1). The relatively vague clinical symptoms in these children make it difficult to integrate the children's clinical status in the decision-making in terms of re-intervention and follow up. It has recently been suggested that health-related quality of life (HRQoL) can be useful when it comes to individualizing treatment decision making in children with congenital heart disease (CHD) (2). Furthermore assessment of HRQoL in pediatric patients can facilitate communication between children and health care professionals and hidden morbidities can be identified (3).

Heath-Related Quality of Life (HRQoL)

HRQoL has been defined as "the influence of a specific illness, medical therapy, or health services policy on the ability of patients to both function in and derive personal satisfaction from various physical, psychological, and social life contexts" (2). During the past years, it has been proposed that HRQoL measurement could aid in medical care and improve communication among children, parents and healthcare professionals (4). Previous studies indicated that children with congenital heart disease (CHD) may have lower HRQoL than healthy children (5) and that children with severe heart disease have lower HRQoL than children with mild or moderate heart disease (6). Cardiopulmonary bypass and reoperations are risk factors for impaired HRQoL (7). In children with CHD we have previously reported a negative correlation between the number of surgeries and HRQoL total score using DISABKIDS chronic generic measure-short version (8). Children with tetralogy of Fallot, a common type of RVOT anomaly, need at last one surgery during their lifetime. Previous study observed comparable HRQoL between children with tetralogy of Fallot and healthy children (9). However, other studies have indicated a psychosocial impact of CHD even after successful repair and a satisfactory clinical status for children with tetralogy of Fallot (10) and worse psychosocial health than for healthy controls (11). Decreased HRQoL has also been reported in children with truncus arteriosus, another type of RVOT anomaly (12).

Child- and Parent-Reports in HRQoL Measurement

Previously the assessment of HRQoL mainly relied upon parents' proxy reports, due to the belief that children lack the cognitive and linguistic skills to self-report HRQoL (13). Children from five years of age seem capable of reliably self-reporting HRQoL with an age-appropriate questionnaire (14). Such questionnaires provide the possibility of considering child-report as the standard in the assessment of HRQoL in the pediatric population. The parent-report may provide another perspective on the child's HRQoL (13). The level of agreement between children and parents has been an issue for discussion since parallel measurement (child and parent versions consist of same questions about the child's HRQoL) became available (15). Measurement of agreement is not only used as a validation for a new instrument but also to understand the difference between child-and parent- report (16). Traditionally, family-centered care (FCC) approach has been the gold standard in pediatric health care setting. Nowadays more child-centered care (CCC) approach by taking into account the child's perspective is recommended (17).

Previous research indicated that conduit operation at higher age is a risk factor for late mortality (18), and that higher age at first conduit operation is associated with a reduced risk of reintervention. These findings illustrate the importance of further research concerning optimal timing for pulmonary valve replacement. The clinical status is generally important in establishing the need and timing for reoperation. Since symptomatology in these patients is often vague, HRQoL assessment might provide an important complement in the decision making for pulmonary valve replacement. The aims of the present study were to comprehensively describe the self-reported HRQoL in children with right ventricular outflow tract anomalies, to evaluate which questionnaire best identifies HRQoL problems and to assess the agreement in this regard between children and their parents. In this study, an extended assessment with three earlier validated questionnaires was used to measure HRQoL using both child and parent-report.

MATERIALS AND METHODS

We conducted a cross-sectional study using a descriptive design with three questionnaires, two generic: the Pediatric quality of life Inventory (PedsQL 4.0), DISABKIDS chronic generic measure-short version and a disease-specific questionnaire for children with cardiac diseases: the PedsQL Cardiac Module. The DISABKIDS chronic generic measure-short version has been used as a patient related outcome in the Swedish national registry for congenital heart disease since the beginning of 2009 (19). PedsQL Cardiac Module has been available in the Swedish national registry for congenital heart disease since 2017.

Study Population

The sample ($n = 198$) was obtained from the Swedish national registry for congenital heart disease (19) ($n = 172$) and from the surgical list ($n = 26$). The search was performed in September 2015 and children between 8 and 18 years old (born between 1997 and 2007) with various diagnoses concerning right ventricular outflow tract anomalies (**Table 1**) were included. Exclusion criteria were Mb Down, Trisomy18, chromosome disorder for chromosomes 4 and 13, Wolf Hirschhorn syndrome, heart surgery/intervention within three months, single ventricle and moving abroad after surgery.

Data Collection

The three questionnaires, PedsQL 4.0, PedsQL Cardiac Module, and DISABKIDS chronic generic measure-short version child- and parent-report, together with an age-appropriate information letter, compliance and non-compliance form and health form were sent out by post. The information letter included detailed instruction about how to complete the PedsQL and how to administer the PedsQL to the child. The children and their parents completed the questionnaires at home and returned them

TABLE 1 | Surgical characteristics and diagnoses in the study population.

	8–12 years *N* (%)	13–18 years *N* (%)	Total sample *N* (%)
Number (N) of patients	47 (48)	50 (52)	97
***N of catheterizations**	18 (38)	19 (38)	37 (38)
1	10 (56)	11 (58)	21 (57)
2	3 (17)	1 (5)	4 (11)
≥3	5 (27)	7 (37)	12 (32)
N of cardiac surgery			
1	19 (40)	18 (36)	37 (38)
2	14 (30)	22 (44)	36 (37)
≥3	14 (30)	10 (20)	24 (25)
Surgery with conduit	17 (36)	22 (44)	39 (40)
Without conduit	30 (64)	28 (56)	58 (60)
Age at first surgery			
< 1 month	12 (26)	7 (14)	19 (20)
1–3 months	9 (19)	7 (14)	16 (17)
3 months–1 year	23 (49)	26 (52)	49 (51)
> 1 year	3 (6)	10 (20)	13 (13)
Age at last surgery		%50	
< 1 year	25 (53)	17 (34)	42 (43)
1-7 years	15 (32)	15 (30)	30 (31)
8-17 years	7 (15)	18 (36)	25 (26)
Cardiac defects			
Tetralogy of Fallot	31 (66)	35 (70)	66 (68)
Fallot with PA	0	3 (6)	3 (3)
DORV (Fallot)	5 (11)	1 (2)	6 (6)
Acyanotic Fallot	0	2 (4)	2 (2)
PAVSD+MAPCA	8 (17)	1 (2)	9 (9)
TRUNCUS Arteriosus	0	1 (2)	1 (1)
DORV	3 (6)	5 (10)	8 (8)
PAVSD	0	2 (4)	2 (2)

**Diagnostic catheterization included; PA: pulmonary atresia, DORV: double outlet right ventricle, VSD: ventricular septal defect, MAPCA: major aorta-pulmonary collateral arteries.*

in the enclosed envelope. After some weeks if no response had been obtained the first author telephoned the families following the instruction in the information letter. One reminder letter was sent out.

PedsQL 4.0 and PedsQL Cardiac Module

PedsQL questionnaires include parallel child- and parent-report. PedsQL 4.0 consists of 23 items in four domains: physical functioning (eight items), emotional functioning (five items), social functioning (five items), and school functioning (five items). A physical health summary score is the same as the domain of physical functioning. To generate a psychosocial health summary score the mean is computed as the sum of the items divided by the number of items in the domains emotional, social, and school functioning (20). The PedsQL Cardiac Module is a disease-specific module and has 27 items in six domains: heart problem (seven items), treatment (five items), perceived physical appearance (three items), treatment anxiety (four items), cognitive problems (five items), and communication (three items) (21).

Both the PedsQL 4.0 and PedsQL Cardiac Modules were developed through focus groups and cognitive interviews and have been tested for reliability and validity (6, 20–22). Child- and parent-report used in this study includes the ages of 8–12 and 13–18 years. The difference between the various age scales is developmentally appropriate language (21). The instruction consists of how much of a problem each item has been during the last month. A five-point response scale is used (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem, 4 = almost always a problem). The items were reverse-scored and linearly transformed to a 0 to 100 scale (0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0), so higher score indicated better HRQoL (21).

DISABKIDS Chronic Generic Measure-Short Version

The DISABKIDS chronic generic measure-short version consists of child- and parent-report questionnaires earlier validated for use in children from 8 to 17 years of age with chronic disease (23) and was developed by "The DISABKIDS group Europe", a cooperation project between Germany, Sweden, France, the Netherlands, Austria, and the United Kingdom (24). The questionnaire includes 12 items−10 items excluding medications—assessing mental (four items), social (four items), and physical (four/two items) impact on the health condition. Each question considers the last 4-week period, and the response for each item is graded using a 5-point scale, indicating frequency of behaviors or feelings as 1 = never, 2 = seldom, 3 = quite often, 4 = very often, and 5 = always. A higher score (maximum 100) indicates a better HRQoL (24).

Statistical Analysis

For ease of comparisons with other previously published data in the field, our data are presented as means and standard deviation. Moreover, the central limit theorem ensures that the convergence of the distribution of the mean is rather quick for non-pathological distributions. Child and parent agreement was assessed by comparison of score means and by correlation. This was performed at three score levels: total-, domain and item level. A strong agreement between children and parent was defined as no significant difference in mean score combined with large correlation. The Wilcoxon test was used for comparison of means and Spearman correlation was used for correlation. For effect size (25): small $r = 0.10–0.29$, medium $r = 0.30–0.49$, and large $r = 0.50–1.0$. p-values less than 0.05 were considered significant (* < 0.05, ** < 0.01, *** < 0.001). Statistical analyses were performed using R (26) while simple statistics were calculated using SPSS version 23.0 (SPSS IBM, New York, United States of America). Dropout analysis was performed using Fisher's Exact test concerning age, gender, diagnoses (TOF). In order to compare the total score in child-reported PedsQL 4.0, PedsQL Cardiac Module and DISABKIDS chronic generic measure-short version a linear mixed model (nlme 3.1–137) was used to account for repeated measures (27). *Post-hoc* comparisons were made using the package multcomp 1.4–8 (28) adjusting

for multiplicity. The results were corroborated using a linear mixed model. Internal consistency for each scale and for the whole questionnaires was assessed using the Cronbachs alfa. In PedsQL 4.0 and PedsQL Cardiac Module the domain score was computed as the sum of the items divided by the number of items answered (accounts for missing data). If more than 50% were missing the domain sum was not computed (20). In our study, the frequencies of "3 = often a problem" and "4 = almost always a problem" were merged and ranked to identify the items children reported most problems with, in the PedsQL 4.0 and PedsQL Cardiac Module.

The total score from the DISABKIDS chronic generic measure-short version was transformed from a raw score to a range of 0–100 (24). Thus, a "standard raw score" = ((point −12)/48) X 100 (if 12 questions were answered)) or = ((point −10)/40) X 100 (if 10 questions were answered)). The responses were transformed so 5 (always) indicated better HRQoL and 1 (never) indicated lower HRQoL in items 3–8 and 11–12. In our study, the answers "never" or "seldom" and "always" or "often" were merged depending on the value course for the items, to identify the items that the children rated lowest HRQoL in DISABKIDS chronic generic measure-short version.

RESULTS

The response rate was 50% ($n = 97$). The mean age of this cohort was 12.9 ± 3 years; 59 (61%) were boys and 47 (48 %) were children from 8 to 12 years old. Parent responders were 64% mothers, 17% fathers, 12% both, and 7 % unknown. The diagnoses and surgical data are shown in **Table 1**. The mean time between the last surgery and responded questionnaire was 8.9 ± 4.9 years. Fifteen (17%) children were on pharmacological therapy due to their heart condition. Two child-reports and two parent-reports were excluded because the parents informed that they had completed the child-reports. The PedsQL 4.0 was completed in 90 child- and parent-reports (93%). The PedsQL Cardiac Module was completed in 93 child-reports (96 %) except for the domain treatment II ($n = 14$, 16 %) and in 90 parent-reports (93 %) except for the domain treatment II ($n = 15$, 17 %). The domain treatment II in PedsQL Cardiac Module was excluded in the results, because of few children on medication. The DISABKIDS chronic generic measure-short version was completed in 91 child-and parent-reports (94%). No significant differences ($p \geq 0.3$) were found in dropout analysis (**Table 2**) in terms of gender, age groups and diagnosis (Tetralogy of Fallot). Cronbachs α for both PedsQL 4.0 and PedsQL Cardiac Module at domain level and for the whole questionnaires varied between 0.70 and 0.95. Cronbachs α for the DISABKIDS chronic generic measure-short version was 0.9.

TABLE 2 | Drop out analysis.

	N	Dropout	*p*-value
Girls/boys	38/59	36/63	>0.3
8-12 år/13-18 år	47/50	46/53	>0.3
Fallot/others	66/31	64/35	>0.3

Frequencies and Fisher's Exact test.

TABLE 3 | Mean score and standard deviations (SD) for the child and parent reports in PedsQL cardiac module, PedsQL 4.0, and DCGM-12.

Total sample	Self		Parent			
	Mean	SD	Mean	SD	*p*	*r*
PedsQL 4.0	*N* = 90		*N* = 89			*N* = 85
Physical functioning	82	19	79	22	0.067	0.84***
Emotional functioning	76	22	72	23	0.033*	0.82***
Social functioning	82	20	78	24	0.055	0.78***
School functioning	72	22	70	23	>0.3	0.80***
Psychosocial[a]	77	20	73	21	0.049*	0.85***
Total score	78	18	75	21	0.040*	0.86***
PedsQL cardiac module	*N* = 93		*N* = 90			*N* = 87–88
Heart problems	76	18	77	20	>0.3	0.80***
Physical appearance	79	24	80	23	>0.3	0.49***
Treatment anxiety	74	29	76	29	>0.3	0.65***
Cognitive problems	67	26	62	29	0.065	0.83***
Communication	74	27	71	31	>0.3	0.61***
Total score	74	19	74	20	>0.3	0.73***
DCGM-12	*N* = 91		*N* = 91			*N* = 86
Total score	81	21	81	23	>0.3	0.78***
Median (10–90)	88	55-100	90	48-100		

*a: aggregated emotional+social+school. Column p represents p-value from Wilcoxon's test for mean rank comparison between child and parent report. *p < 0.05 and *** < 0.001. Column r: Spearman correlation between child and parent report.*

Child-Reported HRQoL

The mean total score (highest HRQoL; 100) for the child-report was significant lower in the PedsQL Cardiac Module (**Table 3**) than in PedsQL 4.0 ($p \leq 0.012$: CI=1.21-6.50) and DISABKIDS chronic generic measure-short version ($p \leq 0.001$: CI=4.17-9.42). The mean child-report score for each domain in PedsQL Cardiac Module ranged between 67 (cognitive function) and 79 (physical appearance), and between 72 (school function) and 82 (physical and social function) in PedsQL 4.0 (**Table 3**). Up to 20% of the children reported problems regarding limited physical capacity and cognitive function in the PedsQL 4.0. Nearly half of the children reported problems with breathlessness when participating in sports, a quarter reported cognitive and communication problems and 14% of the children reported problems in the PedsQL Cardiac Module regarding other people seeing their scar (**Table 4**). A total of 10% of children reported in the DISABKIDS chronic generic measure-short version that their heart disease caused them different kinds of troubles or hard feelings (**Table 5**).

Agreement Between Child- and Parent-Report for Each Questionnaire

In the PedsQL 4.0 the agreement was strong between children and parents for the domains physical-, social- and school

TABLE 4 | Items for specific concerns by child report in PedsQL cardiac module and PedsQL 4.0.

PedsQL 4.0 *N* = 90	Child self report *N* (%)
Almost always or often problem with	
It is hard for me to run	17 (19)
I have low energy	16 (18)
I worry what will happen to me	15 (17)
It is hard to pay attention in class	15 (17)
I forget things	18 (20)
I have trouble keeping up with my schoolwork	14 (16)
PedsQL cardiac module (*N* = 90–93)	
I get out of breath when I do sports activity or exercise	39 (43)
I have to rest more than my friends	20 (22)
I have troubling solving math problems	23 (25)
It is hard for me to remember what I read	18 (19)
It is hard for me to pay attention to things	17 (18)
It is hard for me to explain my heart problem to other people	16 (17)

TABLE 5 | Items for specific concerns by child report in DCGM-12.

DCGM-12 *N* = 90	Child self report *N* (%)
Never or seldom	
Do you feel like everyone else even though you have your condition?	9 (10)
Always or often	
Is your life ruled by your condition	9 (10)
Does your condition bother you when you play or do other things?	10 (11)
Are you unhappy because of your condition	11 (12)
Do you feel different from other children/adolescents?	10 (11)

function and the correlation was large in all domains (**Table 3**). Children rated significantly higher HRQoL than their parents for the domain emotional function and for total score. In the PedsQL Cardiac Module the agreement was strong between children and parents for all domains except physical appearance where the correlation was medium (**Figure 1**). In the DISABKIDS chronic generic measure-short version the agreement was strong for total score.

Agreement Between Child- and Parent-Report for Different Age Groups at Domain and Item Levels in the PedsQL Cardiac Module

Children 8-12 Years of Age

In this subgroup of children the agreement was strong between children and parents for all domains except for perceived physical appearance (**Table 6**). At item level the agreement was strong for 13 (of 22) items. The correlation was medium between children and parents (r = 0.3–0.49) for two (of seven) items in domains heart problems, for two (of three) in perceived physical appearance and for two (of three) in communication. For all other items the correlation was large ($r \geq 0.5$). Among these children the lowest HRQoL was rated for the item 'I get out of breath when I do sports activity or exercise' and children rated this significantly lower than their parents. Children rated significantly higher HRQoL than their parents for one item and lower for two items of a total of 22 items (**Table 6**).

Children 13-18 Years of Age

In this subgroup of children the agreement was strong between children and parents for all domains except communication (**Table 6**). The agreement was strong for 13 (of 22) items. The correlation was medium for three items in the domain treatment anxiety and for the item "embarrassed when other sees my body." For all other items the correlation was large. Among these children the lowest HRQoL was rated for item "I get out of breath when I do sports activity or exercise." The parents rated lowest for the item "troubles solving math problems." Children rated their HRQoL significantly higher than their parents for four items, all in the domains cognitive problems and communication, and lower for one item in the domain heart problems (**Table 6**).

DISCUSSION

Given the complexity of the studied cohort, we used three instruments to assess HRQoL. DISABKIDS chronic generic measure-short version was the first HRQoL instrument used in the Swedish national registry for congenital heart disease (19) in 2009 and later replaced by PedsQL Cardiac Module. PedsQL 4.0 has been used in previous studies (21, 29, 30). DISABKIDS chronic generic measure-short version has fewer questions, requiring thus shorter time to complete this questionnaire, but provides less detailed information regarding the physical and psychosocial functioning compared with the PedsQL questionnaires. The higher total score in the DISABKIDS chronic generic measure-short version and the lower percentage of children who reported difficulties could be related to these drawbacks, particularly in a cohort of children whose symptoms are less obvious. Our findings indicate that problems perceived by children were more easily detected with the disease-specific questionnaire PedsQL Cardiac Module, a result that supports the superiority of disease-specific questionnaires in assessing HRQoL in children with complex CHD (31). Furthermore, PedsQL questionnaires have the advantage of assessing HRQoL at both domain and item level thus enabling the opportunity to obtain more detailed and multifaceted information regarding HRQoL. Another difference between DISABKIDS chronic generic measure-short version and PedsQL is that PedsQL questionnaires are problem-based, being possibly more useful in the clinical follow up.

In a previous Swedish study on children with various types of CHD (32) a strong association was found between children and their parents' ratings of cognitive problems based on the PedsQL Cardiac Module and their intelligence score based on the

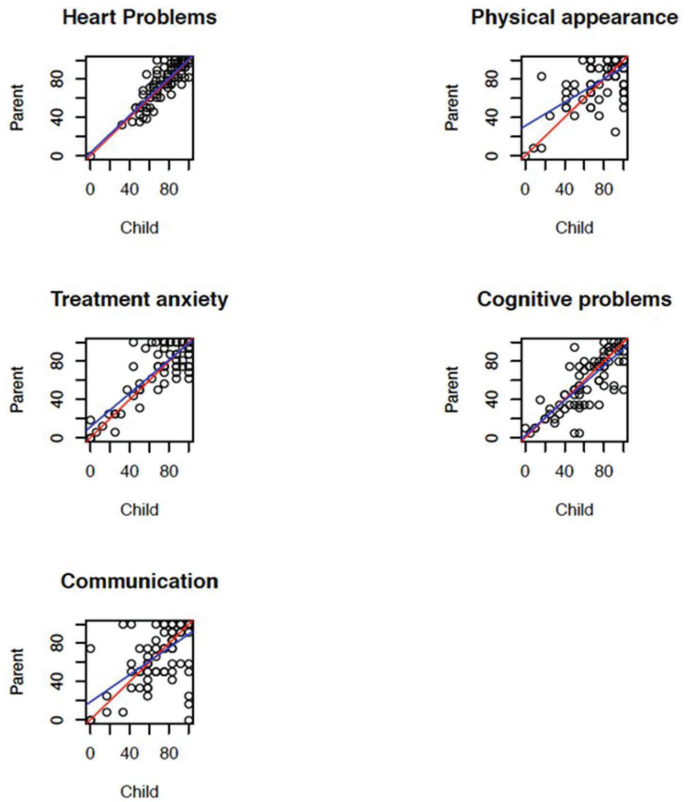

FIGURE 1 | Scatterplot with each point corresponding to a child-parent pair to show the correlation between children and parents for the domains of the PedsQL cardiac module. The red line is perfect agreement and the blue line is the robust least-squares fit.

Wechsler intelligence scales. According to Buratti the cognitive domain in PedsQL Cardiac Module could be used as a screening tool, with a cut off score of 80, to identify children needing standardized cognitive testing. In our study ∼44 % ($n = 93$) of children had a PedsQL score for the cognitive domain below 80. Importantly the lowest score was in the domains school functioning (PedsQL 4.0) and cognitive problems (PedsQL Cardiac Module).

Sixteen of 93 children in our study reported difficulties in explaining their heart problem to other people (PedsQL Cardiac Module). Children with CHD need strategies to communicate about their heart disease (33). A possible reason for these difficulties may be that children do not have the knowledge or the language to talk about their heart disease. Improved knowledge in this regard has been showed to have a positive influence on the HRQoL for adolescents with CHD (34). Healthcare professionals may have a central role by providing age-appropriate information about the heart disease.

Physical capacity is an important determinant of HRQoL. One earlier PedsQL study found a positive correlation between the ability to exercise and the HRQoL among children with Tetralogy of Fallot (9). In our study, the mean scores for the domains of physical functioning and heart problems were 82 and 79, respectively. Item analysis in PedsQL Cardiac Module showed that nearly 50% of the children reported problems due to shortness of breath during physical activity indicating lower physical functioning for these children. This finding illustrates the advantage of item analysis, which may reveal further information not apparent at domain level.

The agreement in our study was assessed on both an individual level (correlation) and a group level (comparison of means), which is recommended to obtain a full assessment of agreement

TABLE 6 | Agreement between children and parents at item level for the two age groups in PedsQL cardiac module.

PedsQL cardiac module	Self 8–12		Parents 8–12				Self 13–18		Parent 13–18			
How much of a problem has this been for you?	**Mean *N* = 41-44**	**SD**	**Mean *N* = 43-45**	**SD**	***P***	***r***	**Mean *N* = 48–49**	**SD**	**Mean *N* = 45–46**	**SD**	***p***	***r***
Heart problems	75	17	75	21	>0.3	0.813***	77	19	78	20	0.083	0.787***
1. I get out of breath when I do sports activity or exercise	43	26	54	32	0.048*	0.760***	50	34	52	34	>0.3	0.819***
2. My chest hurts or feel tight when I do sports activity or exercise	77	22	82	25	0.15	0.493**	84	23	86	22	>0.3	0.620***
3. I catch colds easily	76	27	75	30	>0.3	0.764***	70	29	78	24	0.002**	0.605***
4. I feel my heart beating fast	65	32	66	33	>0.3	0.773***	72	30	77	27	0.116	0.734***
5. My lips turn blue when I run	94	15	86	23	0.005**	0.537***	92	21	93	19	>0.3	0.510***
6. I wake up at night with troubling breathing	99	5	95	11	0.063	0.486**	97	16	95	17	0.25	0.655***
7. I have to rest more than my friends	66	35	67	32	>0.3	0.683***	71	31	68	34	>0.3	0.627***
Perceived physical appearance	83	20	82	22	> 0.3	0.374*	76	28	79	24	>0.3	0.580***
1. I feel I am not good looking	80	32	84	25	>0.3	0.364*	71	32	77	28	0.129	0.600***
2. I don't like other people to see my scars	78	32	78	31	>0.3	0.497***	78	34	81	27	>0.3	0.704***
3. I am embarrassed when others see my body	90	20	83	28	00.051	0.553***	78	33	79	27	>0.3	0.471***
Treatment anxiety	73	31	75	32	0.16	0.719***	76	28	77	27	>0.3	0.526***
1. I get scared when I am waiting to see the doctor	70	32	75	34	0.044*	0.827***	77	28	79	26	>0.3	0.666***
2. I get scared when I have to go to the doctor	75	35	77	34	0.183	0.782***	77	31	80	30	>0.3	0.499***
3. I get scared when I have to go the hospital	72	32	73	32	>0.3	0.808***	74	32	76	31	>0.3	0.452**
4. I get scared when I have to have medical treatments	73	31	74	34	>0.3	0.630***	76	33	76	32	>0.3	0.494***
Cognitive problems	66	26	64	27	> 0.3	0.848***	67	26	61	31	0.074	0.831***
1. It is hard for me to figure out what to do when something bothers me	63	32	66	30	0.156	0.691***	73	28	67	31	0.176	0.704***
2. I have troubling solving math problems	66	33	67	30	>0.3	0.841***	59	36	51	38	0.019*	0.867***
3. I have trouble writing school papers or reports.	70	30	62	31	0.051	0.672***	70	31	63	32	0.062	0.746***
4. It is hard for me to pay attention to things	64	32	59	35	>0.3	0.707***	68	31	59	34	0.040*	0.625***
5. It is hard for me to remember what I read	69	32	65	30	0.204	0.699***	65	35	64	36	>0.3	0.699***
Communication	75	24	76	30	> 0.3	0.513***	74	20	66	32	0.008**	0.740***
1. It is hard for me to tell the doctors and nurses how I feel	74	29	77	31	>0.3	0.577***	78	31	69	35	0.015*	0.642***
2. It is hard for me to ask the doctors and nurses questions	78	31	78	32	>0.3	0.458**	76	33	65	36	0.004**	0.683***
3. It is hard for me to explain my heart problem to other people	72	33	72	35	>0.3	0.385*	68	33	63	34	0.218	0.714***

*Agreement assessed by Wilcoxon signed ranks test for comparison of means and Spearman correlation. *p < 0.05, ** < 0.01, *** < 0.001.*

(35). Therefore, in our study a strong agreement was defined as a large correlation with no significant difference in mean score. The importance of using this approach can be explained as follows: although parents and children on average agree, the individual pair is not in complete agreement. Thus one contains information about the other. The picture is therefore complete only when both views are integrated. This result supports the previous findings (21, 29, 30) namely a need for both child- and parent-reports for children from 8 to 18 years of age. With the advent of the CCC approach in pediatric health care, the child's self report has become central whereas the parent report is considered n important complement. A key-element in the CCC approach is the right of especially older children to be informed and understand the diagnostic and therapeutic decision making. In a recent study (36), children with various chronic diseases reportedly felt encouraged, got insight about their health and improved their understanding of how the disease affected their life when they received feedback from health care professionals about the results of HRQoL assessment. Children with earlier corrected RVOT anomalies could substantially benefit from this since their clinical symptoms are often vague, being difficult to perceive and explain to health care providers. Applying this concept in their follow up could thus improve the understanding on their well-being and intuitively, add important information in the decision making for reoperation especially in those cases with regurgitant pulmonary valve.

Study Limitations

(1) Although the response rate was only 50%, the dropout analysis showed no differences between the groups in terms of age, gender and diagnoses. (2) The cross-sectional study design may preclude assessment of changes in HRQoL over time.

Therefore future prospective studies are important to assess these changes in children with these diagnoses. In order to mitigate the influence of time on the measured indices, we have included only patients with surgical interventions performed at least three months prior to the questionnaire. The questionnaires used in our study have excellent reproducibility over time in patients with chronic disease (21). (3) Another potential limitation is that the questionnaires were completed at home, possibly leading to biases due to potential parental support. However, as emphasized by Upton et al. (13) the place of completion (home vs. clinic) does not appear to influence the results in this regard. (4) Further studies including control population are needed in order to confirm that the difference in agreement between parents and children are specific to patients with RVOT anomalies.

CONCLUSION

Our findings indicate that HRQoL perceived by children with right ventricular outflow tract anomalies is better assessed in the disease-specific questionnaire PedsQL Cardiac Module, suggesting that the PedsQL Cardiac Module is useful in the clinical follow up of these children. Problems concerning cognitive function should be considered by healthcare professionals as both children and parent reported the lowest HRQoL in this domain. The findings regarding the agreement between children with right ventricular outflow tract anomalies and their parents suggests the need to take into account both child- and parent- reports in the assessment of HRQoL. Further longitudinal research including interviews with both children and parents is important as this approach provides deeper understanding of HRQoL over time.

DATA AVAILABILITY STATEMENT

All datasets generated for this study are included in the article/supplementary material.

ETHICS STATEMENT

This study was conducted in accordance with the ethical standard and the Helsinki Declaration and its later amendments. The study was approved by the Ethics Committee for Human Research at the Lund University (#2014/66). Informed consent was obtained from all study participants.

AUTHOR CONTRIBUTIONS

BS contributed to study design and statistical analyses, performed the study work and drafted the revised the manuscript. EI contributed to study design, review in the field of quality of life, revised the manuscript. FN was responsible for statistical analyses and reviewed the statistical part of the manuscript. PL contributed to study design and manuscript review.

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REFERENCES

1. Gatzoulis MA, Balaji S, Webber SA, Hokanson JS, Poile C, Rosenthal M, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of fallot: a multicentre study. *Lancet.* (2000) 356:975–81. doi: 10.1016/S0140-6736(00)02714-8
2. Costello JM, Mussatto K, Cassedy A, Wray J, Mahony L, Teele SA, et al. Prediction by clinicians of quality of life for children and adolescents with cardiac disease. *J Pediatr.* (2015) 166:679–83. doi: 10.1016/j.jpeds.2014.11.061
3. Varni JW, Burwinkle TM Lane MM. HRQoL measurement in pediatric clinical practice:An appraisal and precept for future research and application. *Health Quality Life Outc.* (2005) 3:34. doi: 10.1186/1477-7525-3-34
4. Marino BS, Tomlinson RS, Drotar D, Claybon ES, Aguirre A, Ittenbach R. et al. Quality of life concerns differ among patients, parents and medical providers in children and adolescents with congenital and acquired heart disease. *Pediatrics.* (2009) 123:e708–e715. doi: 10.1542/peds.2008-2572
5. Uzark K, Jones K, Slusher J, Limbers CA, Burvinkle TM Varni WJ. Quality of life in children with heart disease as perceived by children and parents. *Pediatrics.* (2008) 121:e1060. doi: 10.1542/peds.2006-3778
6. Sand P, Kljajic M Sunnegårdh J. The reliability of the pediatric quality of life inventory 3.0 cardiac module for swedish children with congenital heart defects. *Nordic Psychol.* (2013) 65:210–23. doi: 10.1080/19012276.2013.824204
7. Latal B, Helfricht S, Fischer JE, Bauersfeld U Landolt MA. Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease. *BMC Pediatr.* (2009) 9:6. doi: 10.1186/1471-2431-9-6
8. Svensson B, Idvall E, Nilsson F Liuba P. HRQoL in children with surgery for cHD: a study from the swedish national registry for congenital heart disease. *Cardiol Young.* (2016) 27:333–43. doi: 10.1017/S1047951116000585
9. Kwon EN, Mussatto K, Simpson PM, Brosig C, Nugent M Samyn MM. Children and adolescents with repaired tetralogy of fallot report quality of life similar to healthy peers. *Congenital Heart Dis.* (2011) 6:18–27. doi: 10.1111/j.1747-0803.2010.00481.x
10. Bhatt SM, Goldmuntz E, Cassedy A, Marino BS, Mercer-Rosa L. Quality of life is diminished in patients with tetralogy of fallot with mild residual disease: a comparison of tetralogy of fallot and isolated valvar pulmonary stenosis. *Pediatr Cardiol.* (2017) 38:1645–53. doi: 10.1007/s00246-017-1709-3
11. Neal AE, Stopp C, Wypij D, Bellinger DC, Dunbar-Masterson C, DeMaso DR, et al. Predictors of HRQoL in adolescents with tetralogy of Fallot. *J Pediatr.* (2015) 166:132–8. doi: 10.1016/j.jpeds.2014.09.034
12. O'Byrne ML, Mercer-Rosa L, Zhao H. Morbidity in children and adolescents after surgical correction of truncus arteriosus communis. *Am Heart J.* (2013) 166:512–8. doi: 10.1016/j.ahj.2013.05.023
13. Upton P, Lawford J Eiser C. Parent-child agreement across child HRQoL instruments: a review of the literature. *Q Life Res.* (2008) 17:895–913. doi: 10.1007/s11136-008-9350-5
14. Varni JW, Limbers CA Burwinkle TM. How young can children reliably and validly self-report their hRQoL: an analysis of 8,591 children across age subgroups with the pedsQL 4.0 generic core scales. *Health Quality Outc.* (2007) 5:1. doi: 10.1186/1477-7525-5-1
15. Cremeens J, Eiser C Blades M. Factors influencing agreement between child self-report and parent proxy-reports on the pediatric quality of life

inventory^{TM}4.0 (PedsQL^{TM}) generic core scales. *Health Quality Outc.* (2006) 4:58. doi: 10.1186/1477-7525-4-58
16. Eiser C Morse R. Can parents rate their child's hRQoL? Results from a systematic review. *Q Life Res.* (2001) 10:347–57. doi: 10.1023/A:1012253723272
17. Coyne I, Hallström I Söderbeck M. Reframing the focus from a family-centered care approach for children's healthcare. *J Child Health Care.* (2016) 20:94–502. doi: 10.1177/1367493516642744
18. Skoglund K, Svensson G, Thilen U, Dellborg M, Eriksson P. Long-term outcome after right ventricle to pulmonary artery conduit surgery and reintervention. *Scand Cardiovasc J.* (2017) 51:284–91. doi: 10.1080/14017431.2017.1343492
19. SWEDCON. *Swedish National Registry for Congenital Heart Disease.* (2009). Available online at: http://www.ucr.uu.se/swedcon (assessed November 2017).
20. Varni JW, Seid MS Kurtin PS. The PedsQL 4.0: reliability and validity of the pediatric quality of life inventory version 4.0 generic core scales in healthy and patient populations. *Med Care.* (2001) 39:800–12. doi: 10.1097/00005650-200108000-00006
21. Uzark K, Jones K, Burwinkle TM Varni JW. The pediatric quality of life inventory in children with heart disease. *Prog Pediatr Cardiol.* (2003) 18:141–8. doi: 10.1016/S1058-9813(03)00087-0
22. Petersen S, Hägglöf B, Stenlund H Bergström E. Psychometric properties of the swedish pedsQL pediatric quality of life inventory 4.0 generic core scales. *Acta Paediatr.* (2009) 98:1504–12 doi: 10.1111/j.1651-2227.2009.01360.x
23. Bullinger M, Schmidt S Petersen C. The dISABKIDS group assessing quality of life of children with chronic health conditions and disabilities: a european approach. *Int J Rehabil Res.* (2002) 25:197–206. doi: 10.1097/00004356-200209000-00005
24. The European DISABKIDS group. *The DISABKIDS Questionnaires Quality of Life Questionnaires for Children With Chronic Conditions-Handbook*. Lengerich: Germany Pabst Science Publishers (2006).
25. Cohen JW. *Statistical Power for Behavioral Sciences*. Hillsdale, NJ: Lawrence Erlbaum Associates.
26. R Core Team. *R: A Language and Environment for Statistical Computing*. Vienna: R Foundation for Statistical Computing (2016). Available online at: https://www.R-project.org/ (accessed May, 2018).
27. Pinheiro J, Bates D, DebRoy S, Sarkar D, R Core Team. *nlme: Linear and Nonlinear Mixed.* (2018). Effects Models. *R package version 3.1-137*. Available online at: https://CRAN.R-project.org/package=nlme (accessed May, 2018).
28. Holthorn T, Bretz F Westfall P. Simultaneous inference in general parametric models. *Biometrical Journal.* (2008) 50:346–63. doi: 10.1002/bimj.2008 10425
29. Berkes A, Pataki I, Kiss M, Kemeny C, Kardos L, Varni JW et al. Measuring hRQoL in hungarian children with heart disease: psychometric properties of the hungarian version of the pediatric quality of life inventory 4.0 generic core scales and the cardiac module. *Health Quality Life Outcomes.* (2010) 8:14. doi: 10.1186/1477-7525-8-14
30. Eagleson KJ, Justo RN, Ware RS, Johnson SG Boyle FM. HRQoL and congenital heart disease in australia. *J Paediatr Child Health.* (2013) 49:856–64. doi: 10.1111/jpc.12296
31. Marino BS, Uzark K, Ittenbach R Drotar D. Evaluation of quality of life in children with heart disease. *Prog Pediatr Cardiol.* (2010) 29:131–8. doi: 10.1016/j.ppedcard.2010.06.008
32. Buratti S, Ryberg C, Broberg M Sunnegårdh J. Do self- and proxy reports of cognitive problems reflect intellectual functioning in children and adolescents with congenital heart defects? *Front Pediatr.* (2016) 4:127. doi: 10.3389/fped.2016.00127
33. Birks Y, Sloper P, Lewin R Parsons J. Exploring health-related experiences of children and young people with congenital heart disease. *Health Expect.* (2006) 10:16–29. doi: 10.1111/j.1369-7625.2006.00412.x
34. Wang Q, Hay M, Clarke D Menahem S. Associations between knowledge of disease, depression and anxiety, social support, sense of coherence and optimism with hRQoL in an ambulatory sample of adolescents with heart disease. *Cardiol Young.* (2014) 24:126–33 doi: 10.1017/S1047951113000012
35. Sneeuw KCA, Sprangers MAG Aaronson NK. The role of health care providers and significant others in evaluating quality of life of patients with chronic disease. *J Clin Epidemiol.* (2002) 55:1130–43. doi: 10.1016/S0895-4356(02)00479-1
36. Petersson C, Huus K, Enskär K, Hanberger L, Samuelsson U Åkesson K. Impact of type 1 diabetes on health-Realted quality of life among 8-18-Year-Old children. *Compreh Child Adolesc Nurs.* (2016) 39:245–55. doi: 10.1080/24694193.2016.1196265

Conflict of Interest: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Paper III

Cardiology in the Young

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Original Article

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*The online version of this article has been updated since original publication. A notice detailing the change has also been published.

"The only thing I wonder is when I will have surgery again": everyday life for children with right ventricle outflow tract anomalies during assessment for heart surgery*

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Abstract

Background: Children with right ventricle outflow tract anomalies require repeated heart surgeries, thereby needing regular preoperative assessments throughout their lifetime. This situation puts a heavy burden on these children. Thus, the aim of this study was to explore how children diagnosed with right ventricle outflow tract anomalies experience their heart disease and their everyday life during the preoperative assessment and after the decision on whether to perform a new cardiac surgery. *Methods:* Individual interviews were conducted with nine children between 9 to 17 years of age on three occasions from 2014 to 2016. In total there were 27 interviews which all were analyzed with thematic analysis. *Results:* The analysis yielded three themes and eight subthemes. The theme *Me and my heart disease* concerns children's experiences of the heart disease. Almost all described symptoms and how they adapt in their everyday life. The theme *Being me* concerns the children's sense of self, where their heart disease was not prominent. The theme *Being placed in someone else's hands* describes how the assessment was more of a safety net at least until the decision of heart surgery. *Conclusion:* The children's symptoms, their experiences during the assessment, their future surgeries and how the heart disease affects their everyday life could be better understood as elements of their adaptation to the heart disease. In order to achieve individualized support based on the child's experiences and to ensure that these children are involved in their own care a child-centered approach is recommended.

Many children with complex right ventricle outflow tract anomalies such as Tetralogy of Fallot, common arterial trunk, and pulmonary atresia with ventricular septal defect require repeated heart surgeries early in life, but also later throughout their lifetime, thereby emphasising the importance of careful life-long follow-up.[1] The need for repeated heart surgeries during childhood is recognised as a heavy burden on the child.[2] Optimising the time point for re-intervention is important, since delaying re-intervention for these children can lead to complications such as ventricular arrhythmias, heart failure, and death.[3] To this purpose, thorough pre-operative assessment (henceforth named as assessment) including clinical examination, echocardiography, MRI, and exercise test need to be performed whenever the indication for reoperation is suspected. It is likely to believe that children who are going through this kind of assessment that may lead to heart surgery need extra support. According to previous research, children with complex heart disease fear for the possibility of surgery[4] and the thought of future repeated heart surgery is associated with anxiety.[2] This might have an impact on children's everyday life and in research nowadays involving children with CHD, the focus has changed from survival to how these children experience their everyday life.[5] Earlier studies have shown that they experience physical activities limitation[6,7] and feelings of isolation,[8] but no study has yet studied how children with right ventricle outflow tract anomalies experience their everyday life. To be able to provide support, studies are needed to explore how these children experience the period from assessment to decision, as well as the months thereafter. Therefore, the aim of this study was to explore how children diagnosed with complex right ventricle outflow tract anomalies experience their heart disease and their everyday life during the assessment and after the decision on whether to perform a new cardiac surgery.

Materials and methods

Study design

A longitudinal qualitative inductive study design was used since it is suited to understanding subjective experiences over time. For gathering children's experiences, interviewing is a

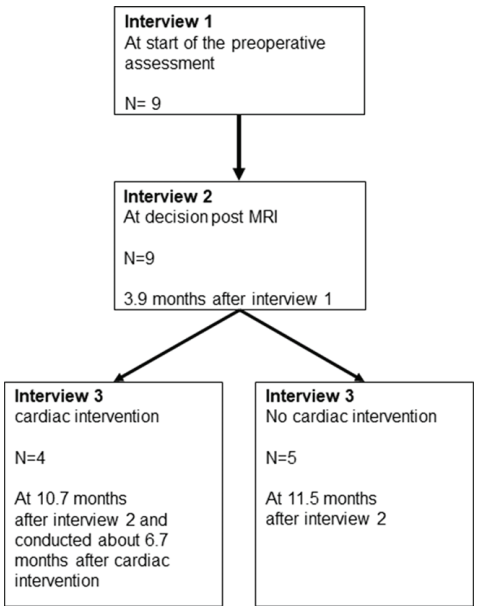

Figure 1. Mean Time points for the three interviews, the assessments stages and the number of children interviewed.

recommended method.[9] In this study, children with right ventricle outflow tract anomalies were interviewed at three chosen time points during and after the assessment stages, in total 27 interviews (Fig 1).

Participants

During the period 2014–2016, nine children (six girls) with complex right ventricle outflow tract anomalies aged between 9 and 17 years (mean age 13.8) were included in the study (Table 1). The inclusion criteria were age 9–18 years, diagnosis of complex right ventricle outflow tract anomalies, previous surgical repair, and a completed MRI. The exclusion criteria were children unable to speak and understand Swedish or unable to participate in an interview situation due to cognitive impairment. Totally 15 children and their guardians were invited to participate, where six of the guardian's declined participation for themselves and the child.

Data collection

After receiving oral and written age-appropriate information about the study, children and their guardians at two university hospitals in Sweden were invited to participate in the study by two nurses, working at the clinics. After obtaining written informed consent from the guardians and from children over 15 years of age and consent from children younger than 15 years of age, the first author (BS) contacted them to provide further information. Of the interviews, 24 were performed at the child's home, two in the hospital, and one by telephone. Children (with help from their guardians) decided the time and place for the interview. All children were given age-appropriate information explaining that their participation was voluntary and could be withdrawn at any time without giving a reason. All interviews were conducted individually, except for one first interview, when the child wanted to have his mother present. The children and their guardians received the contact information of a psychologist in case they felt a need to talk to someone further after the interview.

Interview

An interview guide was developed, based on a literature review focusing on the research aim. The main topics included in the interview concerned children's everyday life during and after the assessment, their heart disease, earlier and future heart surgery, and their future. Open-ended questions were used, and follow-up questions were asked, such as "*What did you feel?*" and "*Can you describe it?*" These follow-up questions were asked whenever the narrative needed clarification. The first interview for all children started with a presentation of the interviewer, the study, and the structure of the three planned interviews. This was followed by general questions, for instance "How does a day in school look like" and "What do you like to do in your spare-time" (Table 2). The interviewer began the second interview with "*Last time I was here . . . and now you have got a decision*," and the third interview began, for children with no need for heart surgery, with "*A year has passed since last time I was here*" and, for children who had had heart surgery, "*Since the last interview, you have been operated.*" The children were given appropriate information clarifying that there were no right or wrong answers, and they decided whether they wanted to answer each question or not. Two questions were posed at the end of each interview: "*Is there something you want to tell me that I didn't ask you?*" and "*How are you feeling now?*" All interviews were conducted by the first author and digitally recorded with the approval of the children. The interviews lasted from 47 to 129 minutes per child (mean 96 minutes).

Data analysis

A thematic analysis approach was used to analyse the interview data and the six phases of thematic analysis by Braun and Clarke guided the analysis (Table 3). Thematic analysis is a method for identifying, analysing, and reporting patterns within the data.[10] An inductive approach was used at a semantic level for both a descriptive and an interpretative perspective. The first author transcribed all the recorded interviews and when all data was generated, the analysis took place for one child at a time, including all the three interviews with the child. The first and the last author (MB) coded independently, and then assessed the results together while the third author (AW) contributed to the interpretation of the themes and subthemes. After that the whole research group discussed and approved the final analysis.

Rigor

In qualitative research, trustworthiness can be demonstrated through the credibility, dependability, and confirmability.[11] To ensure dependability, the first author conducted all the interviews after one pilot interview had been done. The fact that the first and last author coded independently and that the results included quotations from the interviews strengthens the credibility of the study. All children's experiences are described in the findings and all but one is quoted 1–3 times. Confirmability was enhanced by the first author writing a reflexivity journal for personal feelings,

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Table 1. Characteristics of the participants, previous surgical repair with transannular patch or conduit, and outcome of the latest MRI

Participants	Diagnosis	Number of operations*	Reason for MRI	Outcome
Child 1	PS, PR	2	Routine	No heart surgery
Child 2	TOF	2	Suspected problem	No heart surgery
Child 3	TGA, PS	2	Routine	No heart surgery
Child 4	DORV Fallot type	1	Routine	No heart surgery
Child 5	TOF	3	Routine	No heart surgery
Child 6	PS	1	Routine	Heart surgery
Child 7	Truncus	3	Routine	Heart surgery
Child 8	TOF	3	Suspected problem	Intervention**
Child 9	DORV Fallot type	1	Routine	Heart surgery

DORV = double outlet right ventricle, PR = pulmonary regurgitation, PS = pulmonary stenosis, TGA = transposition of the great arteries, TOF = tetralogy of Fallot.
*Number of operations before the pre-operative assessment.
**Implantation of a percutaneous stent valve via heart catheterisation.

Table 2. Interview guide for the first interview

Guide for Interview 1	Examples of age-appropriate issues
Background	Your family members?
Everyday life	How does a day in school look like? What do you like to do in your spare-time?
Wellbeing	How are you feeling? Do you need to rest more than your friends?
Self-perception	How do you look at yourself?
The heart disease	How did you find out about your heart disease? How and when do you feel your heart disease?
Hospital experience	How did you experience the MRI? Do you remember anything from your earlier surgery? What do you know about future surgery and how do you feel about it?
The future	What do think about your future? What would you like to do?

Table 3. Phases of the thematic analysis as described by Braun and Clarke (2006)

Phase	Description of the process
1. Familiarizing with your data	Transcribing data, reading and re-reading the data, noting down initial ideas
2. Generating initial codes	Coding interesting features of the data in a systematic fashion across the entire data set, collecting data relevant to each code
3. Searching for themes	Collating codes into potential themes, gathering all data relevant to each theme
4. Reviewing themes	Checking if the themes work in relation to the coded extracts and the entire data set, generating a thematic map of the analysis
5. Defining and naming themes	Ongoing analysis to refine the specifics of each theme, and the overall story the analysis tells, generating clear definitions and names for each theme
6. Producing the report	The final opportunity for analysis. Selection of vivid, compelling extract examples, related back of the analysis to the research question and literature

insights, beliefs, and observations used for highlighting pre-conceptions before and during the analysis.

Results

The data analysis yielded three themes and eight subthemes. The children's experiences of the heart disease in their everyday life are described in the theme *Me and my heart disease*. The theme *Being me* concerns children's sense of self. Their experiences during the assessment and its consequences are described in the theme *Being placed in someone else's hands*. The themes and subthemes are presented in Figure 2.

Me and my heart disease

The children's experiences of their heart disease in everyday life and how these experiences affected their ability to adapt are described in the subthemes *Limited by my heart disease* and *Adapted to my heart*. All the children had experiences of earlier heart surgery and of having a scar, which is described in the subtheme *My story of survival*.

Limited by my heart disease

Almost all the children had symptoms and experienced varying degrees of tiredness that forced them to take a rest every day, sometimes for the whole day. Physical activity reduced their energy especially, and the difference between their tiredness and lack of energy was expressed as "*I suppose I get more tired and then if I exert myself, I get out of breath much faster, yes . . . . if I get out of breath it will pass but if I get tired, I am tired the whole day*" (*Girl, 16 yrs.).* It was unusual that children felt chest pain but some of them reported that their heart was beating hard and fast during certain activities and that they sometimes felt that it would stop. Experiencing limitations was common among the children, for example, being unable to play football although it was the child's greatest wish.

Adapted to my heart

All the children had become accustomed to their heart disease, but still some of them speculated about who they would be without it.

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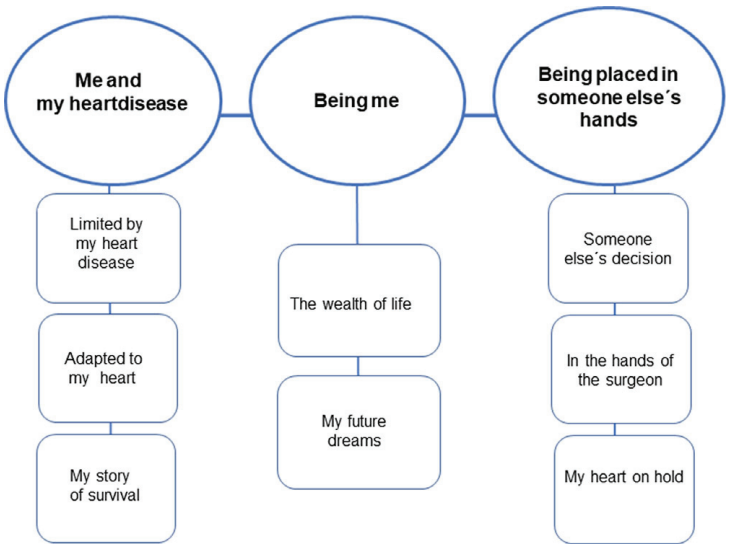

Figure 2. Thematic map over the three themes with the eight subthemes.

Sometimes they had not noticed their symptoms, including feeling breathless and needing rest, until after the surgery, when they could make a comparison. They also found their tiredness hard to interpret: was it the heart disease, or adolescence, or were they just having a bad day? The children sometimes talked about being used to the limitations resulting from their heart disease so they did not experience them as limitations. "*It has always been there without affecting me, except I can't exert myself too much*" *(Girl,17 yrs.).* The most common adaptation they learned was to rest and take short breaks, but also to manage their strength so it would last longer. Others had developed skills for showing their lack of energy without expressing it in words.

My story of survival

All the children had gone through at least one heart surgery earlier in life, but these memories were not accessible for most of the children. "*I don't remember the operation itself or afterwards or all that. I only remember that I was well taken care of and so on. But maybe it's good that I don't remember everything*" *(Girl, 17 yrs.).* They all had experiences related to their scar, which was seen as a reminder of their survival. "*But I think it is nice . . . like a story . . . something to be proud of . . . it's thanks to that that I am alive . . . otherwise I wouldn't be alive. So it's something to be proud of because not everybody survives such operations . . . that's how it is*" *(Girl, 17 yrs.).* From that perspective, the scar became a reminder of strength and power, but it could also be a reminder of exclusion and not being like others, or just a minor difference. "*I am like anyone else with a scar*"*(Girl, 11 yrs.).*

Being me

The children's narratives revealed a sense of self in which the heart disease did not feature, described in the subthemes *The wealth of life* and *My future dreams.*

The wealth of life

The children's sense of self was described in a positive way with words like *happy*, *social*, *kind*, *independent*, and *open-minded*. They described having a strong social network, both at home and in school. Almost all also expressed a steady well-being that was naturally affected by the ups and downs of life, such as success in school and getting new friends or periods of lacking energy in the winter darkness. The most important aspects of their lives were their family and friends and having fun. "*Doing what you want, in other words, avoiding what you don't want*" *(Boy, 17 yrs.).* Their spare time was filled with activities such as football, dance, biking, playing, reading, singing, listening to music, or just relaxing. All the children mentioned something or somebody that made them happy.

My future dreams

All the children easily shared their thoughts and feelings about the future and described different future dreams, which were either imaginative, "*I want to live in Monaco . . . there are a lot of car races and suchlike, so that's why . . . also because some celebrities live there*" *(Boy, 13 yrs.),* or more realistic, such as their career choices, including carpenter, doctor, nurse, professional singer, confectioner, and personal assistant. They wanted to travel to foreign places or work abroad, but some children also expressed a wish to stay close to their childhood home and city. They talked about settling down with a partner, with or without starting a family. Their stories sparkled with hope that the future was theirs for the taking.

Being placed in someone else's hands

Experiences of going through an assessment and receiving a decision about heart surgery are described in the subtheme *Someone*

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else's decision. The experiences of the heart surgery are described in the subtheme *In the hands of the surgeon*, and the subtheme *My heart on hold* describes the experiences of those who did not need heart surgery.

Someone else's decision

As soon as the assessment started, the children feared having to undergo heart surgery, but their everyday life was not markedly affected at first. To some extent, the children hoped for a decision against heart surgery, but they all knew that it would be needed someday. Irrespective of the outcome, they welcomed the assessment, which they saw as a safety net. All the children received the decision via their guardians and, where there was no need for heart surgery, the children's reactions were positive and life went on as before. Nevertheless, it only meant a temporary reprieve, since they knew that surgery would be needed, but they did not know when. The decision could give them some relief but not an assurance. "*It is very good that I do not need surgery but at the same time, I myself feel that it's heading that way, I feel worse than maybe a year ago*" *(Girl, 17 yrs.).* When the assessment resulted in a decision that heart surgery or an intervention would be needed, the children's reactions varied. One was relieved that the decision was for an intervention rather than heart surgery, while another felt that her everyday life was turned upside-down and began to prepare for her forthcoming funeral. Thus, their feelings ranged from joy, expectancy, and being offered their lives back, to one child suffering pronounced loneliness, exacerbated by the fact that the decision to perform heart surgery did not correspond to how she felt. "*They see something that I do not feel, with the heart that is, that needs to be repaired*" *(Girl, 17 yrs.).*

In the hands of the surgeon

Some children were dissatisfied with the health care professional's communication. One child had earlier got the information that the new valve would be a mechanical valve, but the day before surgery the surgeon informed her that a biological valve would be the best. This rapid change was extremely distressing as she was reluctant to receive a biological organ. After surgery and before going home, she realised what valve they had actually implanted. "*Then he said biological valve and everything burst for me and I got so miserable and I thought that I would have that person's characteristics . . . I got so afraid . . . I know I looked at myself in a mirror afterwards and thought, I still have brown eyes . . . I don't know but I got some sort of strong anxiety and everything became chaos for me"* (Girl, 17 yrs.) Despite setbacks during the surgery and the hospital stay, the children didn't give up. *"I don't know much . . . .it is sad that it has happened . . . but I can't do anything about that . . . battle on . . . yes"* (Boy, 13 yrs.).

My heart on hold

The children's experiences varied when heart surgery was not needed. Most of the time they could suppress or even forgot their worries, but on bad days, such as when they felt sad, these thoughts came back. They were so used to the prospect of heart surgery that they found it hard to say how their lives were affected, but nevertheless, the thought of future surgery was worrying. Their worries ranged from everyday life concerns, such as missed schoolwork, to matters of life and death. "*Like when they are going to operate then I will be very nervous* [whispers nervously] *. . . that . . . I might die . . . like, that nervous*" *(Girl, 9 yrs.).* One child could talk about everything, but when it came to surgery, his only response was tears. Some of the children wondered when to expect the future surgery. "*. . . mm, the only thing I wonder about my heart disease is . . . is when . . . because I would like to know when I will be operated . . . that's the only thing I want to know . . . but I don't know . . . I mean, I would like to know when, but I don't know if it would have been better to know"* (Girl, 11 yrs.).

Discussion

To our knowledge, this is the first study focusing on the everyday life experience of children with complex right ventricle outflow tract anomalies at different assessment stages and months after the medical decision was taken. Although this thorough assessment is a milestone in the follow-up, these children did not see it as a cause for concern, at least not until the decision was taken. Importantly almost all had symptoms from their heart disease but they did not always pay attention to them. They had developed strategies to manage their heart disease in their everyday life, sometimes so well that their everyday life showed no trace of the heart disease. This could be seen as an adaptation to the disease.

According to Marino et al., the definition of adaptive behavior is "an age-related construct that reflects learned skills in conceptual, practical and social arenas that are necessary for function in everyday life".[12] There might be a risk that children with CHD have difficulties adapting in their everyday life due to their underlying heart condition.[12] In our study, we could see traces of such difficulties, for instance, when the children limited themselves in their physical activity. In all the narratives, we also saw traces of adaptive behavior in their everyday life concerning their heart disease, the assessment, and future surgeries. Even though previous research has shown that future surgery is a burden for children with CHD,[2,8] the children in our study showed that it also provides them with an opportunity to adapt to this situation, as well as to the heart disease itself.

The mechanism of adaptation could partly explain why children were not concerned in the beginning of the assessment. In the study by Shearer et al., adolescents with CHD expressed feelings of comfort while being taken care of by healthcare professionals.[6] The children in our study likewise experienced feelings of being taken care of or being placed in someone else's hands when they went through the assessment. However, when informed about the medical decision, they demonstrated clear feelings of either safety or fear. In a study by Gutman et al, children with chronic kidney disease expressed wanting to be involved in treatment decision-making.[13] Based on our findings, we speculate that children with CHD would also benefit from being more involved in the treatment decision-making which could improve their feeling of safety.

The child's experiences and own perspectives can be better understood and integrated in the assessment by inviting the child to participate at an early age and by asking explorative questions regarding symptoms and their change over time. This could be achieved by using the child-centered approach. According to Söderbäck et al, a child-centered approach "includes both the adult's child perspective concerning the children's best interests in terms of care and the child's perspective with respect to his or her preferences."[14] Inviting the children to participate from the start of the assessment may also improve their hospital stay

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when the children were at their most vulnerable. Some of them had experienced deficient communication, and they expressed a wish to be invited both to participate and to communicate. This is also strengthened by Coyne and Gallagher study, where hospitalised children wanted to be included in communication exchanges and felt better when they were invited to participate.[15]

Our results reveal a contradiction in the children's descriptions about living with their heart disease. They describe its large impact, such as the need to rest every day, but at the same time shrug off the experience as barely worth mentioning. This could be understood by the chronic paradoxes of CHD, defined as "experiences and strategies that address the often lifelong and contradictory nature of living with CHD". [4] It could also be viewed by the concept of *illness identity*, defined as "the degree to which a chronic health condition, such as heart disease, is integrated into someone's identity."[16] This includes both how the disease is comprehended and how much influence the disease has on the child's self-image.

Limitations

One limitation in our study may be the small number of children (n = 9) included; however, 27 interviews were conducted in total. For a small project, five or six interviews could be enough.[17] In qualitative research, the number of participants is usually small since the aim is to give a deeper understanding of the phenomenon explored and not to generalise findings.

Conclusion

The children's symptoms, their experiences during the assessment, their future surgeries, and how the heart disease affects their everyday life could be better understood as elements of their adaptation to the heart disease. In order to achieve individualised support based on the child's experiences and to ensure that these children are involved in their own care, a child-centered approach is recommended. Furthermore, by inviting the children to participate during the assessment, the shortcomings in communication can better be avoided.

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Conflicts of interest. None.

Ethical standards. This study was approved by the Ethical Review Board at Lund University (approval no. 2014/66) and conducted in line with the World Medical Association Declaration of Helsinki.[18]

References

1. Ho JG, Schamberger MS, Hurwitz RA, Johnson TR, Sterett LS, Ebentoth ES. The effects of pulmonary valve replacement for severe pulmonary regurgitation on exercise capacity and cardiac function. Pediatr Cardiol 2015; 36: 1194–1203.
2. Andresen B, Andersen MH, Lindberg H, Døhlen G. Perceived health after percutaneous pulmonary valve implantation: in-depth interviews of patients and next-of-kin. BMJ open 2014; 4: e005102.
3. Gatzoulis MA, Balaji S, Webber SA et al. Risk factors for arrhythmia and sudden cardiac death after repair of tetralogy of Fallot: a multicentre study. Lancet 2000; 356: 975–981.
4. Chong LSH, Fitzgerald DA, Craig JC et al. Children's experiences of congenital heart disease: a systemic review of qualitative studies. Eur J Pediatr 2018; 177: 319–336.
5. Svensson MK, Walhberg A, Gislason GH. Chronic paradoxes: a systematic review of qualitative family perspectives on living with congenital heart defects. Qual Health Res 2020; 30: 1: 119–132.
6. Elissa K, Sparud-Lundin C, Axelsson ÅB, Khatib S, Bratt EL. Struggling and overcoming daily life barriers among children with congenital heart disease and their parents in the West Bank, Palestine. J Fam Nurs 2018; 24: 585–611.
7. Shearer K, Rempel GR, Norris CM, Magill-Evans J. "It's no big deal": adolescents with congenital heart disease. J Pediatr Nurs 2013; 28: 28–36.
8. Lee S & Kim S. The life experiences of Korean children and adolescents with complex congenital heart disease: a qualitative study. Nurs Health Sci 2012; 14: 398–404.
9. Patton MQ. Qualitative Research & Evaluation Methods. Sage Publications, 4th edn. USA, 2015.
10. Braun V & Clarke V. Using thematic analysis in psychology. Qual res Psychol 2006; 3: 77–101.
11. Lincoln YS & Guba EG. Naturalistic Inquiry. Sage Beverly Hills, CA, 1985.
12. Marino B, Lipkin PH, Newburger JW et al. Neurodevelopmental outcome in children with congenital heart disease: evaluation and management. A scientific statement from the American heart association. Circulation 2012; 126: 9.
13. Gutman T, Hanson CS, Bernays S et al. Child and parental perspectives on communication and decision making in pediatric CKD: a focus group study. Am J Kidney Dis 2018; 72: 4.
14. Söderbäck M, Coyne I, Harder M. The importance of including both a child perspective and the childs perspective within health care settings to provide truly child-centred care. J child health care 2011; 15: 99–106.
15. Coyne I & Gallagher P. Participation in communication and decision-making: children and young people's experiences in a hospital setting. J clin nurs 2010; 20: 2334–2343.
16. Van Bulck L, Luyckx K, Hoosens E, Oris L, Moons P. Illness identity: capturing the influence of illness on the person's sense of self. Eur J Cardiovasc Nurs 2019; 18:4–6.
17. Braun V, Clarke V, Hayfield N, Terry G. Thematic Analysis. In: Pranee Liamputtong (ed.) Handbook of Research Methods in Health Social Science. Springer, Singapore, 2019, pp. 843–860.
18. World Medical Association. Declaration of Helsinki-Ethical principles for medical research involving human subjects. 64th WMA 2013.

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Paper IV

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ARTICLE

"I Dread the Heart Surgery but it Keeps My Child Alive"—Experiences of Parents of Children with Right Ventricular Outflow Tract Anomalies during the Assessment for Cardiac Reoperation

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ABSTRACT

Background: Parents of children with complex right ventricular outflow tract (RVOT) anomalies are confronted with their child's need for heart surgery early in life and repeated reoperations later on. Preoperative assessment needs to be performed whenever an indication for reoperation is suspected. The aim was to illuminate the experiences of parents of children diagnosed with RVOT anomalies, in particular, how they experience their child's heart disease and everyday life during the assessment and after the decision on whether to perform a reoperation. **Method:** Individual interviews (n = 27) were conducted with nine parents on three occasions between 2014 and 2016 and analyzed using reflexive thematic analysis. **Results:** The analysis resulted in the following five main coexisting themes: The heart surgery keeps my child alive illuminates parents' experiences during and after the assessment and emphasizes that heart surgery, although dreaded, is central for their child's survival; Everyday struggles illuminates the different struggles parents had to face to ensure that their child would be in the best possible condition; the remaining three themes, Unconditional love, Trust in life, and Togetherness, illuminate the ways in which the parents gained inner strength and confidence in their everyday lives**. Conclusion:** Although the parents were grateful for the assessment and had learned to navigate among the fears it aroused, they experienced several distressing situations during the assessment process that should be addressed. By inviting both the parents and their child to participate in the child's care, individualized support can take into account the needs of both parents and child.

KEYWORDS

Parents; children; right ventricular outflow tract anomalies; everyday life; heart surgery; reflexive thematic analysis

1 Introduction

Parents of children with complex right ventricular outflow tract (RVOT) anomalies, such as tetralogy of Fallot (TOF), double outflow of right ventricle (DORV), and common arterial trunk (CAT), are confronted

with their child's need for surgery early in life and reoperations later on. Preoperative assessment including multiple clinical and imaging procedures needs to be performed whenever an indication for reoperation is suspected [1]. Previous research has shown that parents of children with different types of congenital heart disease (CHD) report more stress, depression, and anxiety than parents of healthy children or parents of children with other diseases [2–4]. In addition, previous research has shown that both the waiting time before heart surgery and the hospitalization for the surgery are major sources of stress for parents of younger children with CHD [5–8]. This applies to the whole family, which has to adapt to this trauma and disruption to family life during this time [8]. The repeated assessment for children with RVOT anomalies can guide the need for reoperation, so it is important to be aware of how parents experience this period of uncertainty, from the start of the assessment until they receive a decision about their child's need for further surgery. How their children experience everyday life during and after this assessment, has been described in a recently published study by our group [9] which showed that the children's fear of surgery was awoken at the start of the assessment; however, this fear had no impact on their everyday life until the decision about the need for reoperation. To the best of our knowledge, this is the first study to explore the experiences of everyday life for parents of children with RVOT anomalies during a period covering the preoperative assessment through the decision making and after reoperation, when the last interview was conducted. A better understanding of the parents' experiences is one way for health care providers to gain insights into how to improve the support given to parents [2,6]. Thus, the aim of this study was to illuminate how these parents experienced their children's heart disease and everyday life during this assessment and after the decision on whether to perform a reoperation.

2 Material and Methods

2.1 Design

An inductive design with a constructionist approach [10] was chosen to capture the parents' experiences, using semi-structured individual interviews at three different data collection times. A constructionist approach in this context means that the parents' experiences, obtained herein via interviews, are used to construct particular realities which will become the focus of research [10]. All the collected data were analyzed using reflexive thematic analysis following Braun et al. [11–13], a method that allows the researcher to interpret the meaning behind the words expressed by the interviewees. When using thematic analysis (TA), according to Braun and Clarke, it is important to decide which branch of TA, coding reliability, codebook or reflexive approach is suitable for the study [12]. A reflexive TA requires time and space for both immersion of engagement in the data and distancing [13].

2.2 Procedure and Participants

Nine parents participated in the study. Their children were 9–17 years of age (Table 1). The inclusion criteria were being a parent of a child aged between 9 and 18 years with a diagnosis of RVOT anomalies who had undergone a surgically repaire and completed an MRI examination to assess the need for pulmonary valve replacement. The exclusion criteria were parents of children unable to speak and understand Swedish or unable to participate in an interview situation due to cognitive impairment. The parents of 15 children were eligible to participate in the study. Of these, the parents of 6 children declined to participate either due to the lack of time given the child's school load or in order to spare the child from knowing about the need for eventual reoperation. All the parents were invited by two nurses at two university hospitals in Sweden. After having been given oral and written information about the study, the parents were given some time to consider their possible participation. The nine parents who gave their written informed consent to participate were then contacted by the first author (BS), who provided further information if needed. The participants had not had contact with BS outside of this study.

Table 1: The main characteristics of the participants' children

Participants	Child's age	Diagnosis	Number of previous surgeries	Decision after assessment
Father 1	17	PS, PR, RVOT conduit	2	No heart surgery
Mother 2	13	TOF	2	No heart surgery
Mother 3	11	TGA, PS	2	No heart surgery
Mother 4	9	DORV Fallot type	1	No heart surgery
Mother 5	16	TOF	3	No heart surgery
Mother 6	13	PS, transannular patch, PR	1	Heart surgery
Mother 7	17	Truncus	3	Heart surgery
Mother 8	12	TOF	3	Intervention*
Father 9	17	DORV Fallot type	1	Heart surgery

Notes: PS = pulmonary stenosis, PR = pulmonary regurgitation, TOF = tetralogy of Fallot, TGA = transposition of the great arteries, DORV = double outlet right ventricle. *Transcatheter implantation of pulmonary valve.

2.3 Interviews

A semi-structured interview guide was developed based on a literature review focusing on the research aim. Central issues in the interview guide were: (1) the parents' experiences of everyday life during and after their child went through an assessment and received a decision about whether or not heart surgery was necessary, (2) experiences of their child's heart disease and heart surgeries, (3) its impact on the whole family's everyday life, and (4) their thoughts about their child's leisure, schooldays, and future. Follow-up questions such as: How did you feel? Can you describe it? were asked to elicit in-depth responses. All interviews were conducted by the first author (BS) and digitally recorded after the approval of the participants. All three interviews (Table 2) lasted from 86 to 169 min per participant (median 135 min). At the end of the interview all participants were asked whether there was something else they wanted to tell the interviewer and asked how they felt after having participated in the interview. The rationale for doing this was to identify any participants who were affected negatively by the interview and in need of professional support. Lastly, the parents received a contact number of a psychologist in case they needed such support.

Table 2: The main characteristics of the interviews

	The first interview	The second interview	The third interview (children needed surgery)	The third interview (children not needed surgery)
Timepoint	At the start of the assessment	At the decision	At six month after surgery	At one year after interview two
Opening question	"Can you tell me about your child" (the childs name)?	"The last time I was here…and now you have received a decision"	"The last time I was here…and now your child has gone through surgery. Can you tell me what it has been like?"	"A year has passed since I last time I was here…how have you been and what has happened?"

(Continued)

Table 2 (continued)

	The first interview	The second interview	The third interview (children needed surgery)	The third interview (children not needed surgery)
Setting	Two interviews were performed in hospital and seven in the participants home	One interview was performed on the telephone and eight in the participants home	All interviews were performed in the participants home	All interviews were performed in the participants home

2.4 Data Analysis

The data were analyzed using reflexive thematic analysis according to Braun et al. [11–13], in which the researcher plays an active part in the process of producing knowledge. "The analytic process involves immersion in the data, reading, reflecting, questioning, imagining, wondering, writing, retreating, and returning" [14]. The analysis started with a phase of familiarization which was performed by BS, who had transcribed all the interviews. The first reflections were written down immediately after the interviews and further reflections were written when the interviews were in verbatim transcripts. The familiarization continued with BS reading through the whole dataset on two occasions with an open curious mind and at the same time making casual notes shaped by the research question. During the second phase, codes were generated. This was a more active phase of systematically identifying meanings in the whole dataset. The codes were at first semantic, which mean they were identified and described at a surface meaning of the data, [11] but became more latent as the coding progressed. The codes were written in a table, together with extracts from the dataset and were sorted manually. The third phase consisted of a theme development process, where the themes were developed from the generated codes. Building themes was an active analysis phase and gave meaning at the intersection of the data, the research question, and the researchers' experience and subjectivity. In the fourth and fifth phase, the themes were revised, defined, and checked against the whole dataset. A thematic map was created to show how the themes interacted and what overarching story they told. At first, the thematic map included many candidate themes which finally emerged as five coexisting themes. The sixth phase started when the report was produced and was the final test of how the themes worked, both on an individual level with the dataset but also on an overall level. The candidate themes were reworked and refined by BS in consultation with co-authors MB and AW, and in the last phase also with PL. To ease the reading of the quotes, all hesitations and repetitions were removed. Where more than just hesitations and repetitions have been removed, this is indicated in the quotes below by (…); this has only been done where it did not alter the meaning in accordance to Braun et al. [10].

2.5 Rigor

The interviewer BS has a long experience within pediatric cardiology and has, as a nurse, extensive experience of interacting with parents in different care situations. A reflexivity journal was created at the beginning of the interviews and kept throughout the whole procedure, which further facilitated a questioning, reflexive thinking. According to Lincoln et al. [15] transferability was promoted by describing the settings and the participant characteristics and dependability was enhanced by the fact that only one interviewer (BS) conducted all the interviews. In addition, rigor was enhanced by using Braun et al.'s 15-point checklist of criteria for good thematic analysis [11].

3 Results

The analysis produced five main coexisting themes (Fig. 1). The first theme, *the heart surgery keeps my child alive*, illuminates parents' experiences during and after the assessment for reoperation, which is a recurrent event in their child's life. It emphasizes that the heart is central for their child's survival and that it needs to be monitored. The second theme, *Everyday struggles,* illuminates the different struggles parents had to face in order to provide their child with the best possible conditions in everyday life. The other three themes, *Unconditional love*, *Trust in life*, and *Togetherness*, illuminate how and in what way the parents gain inner strength and confidence in their everyday life.

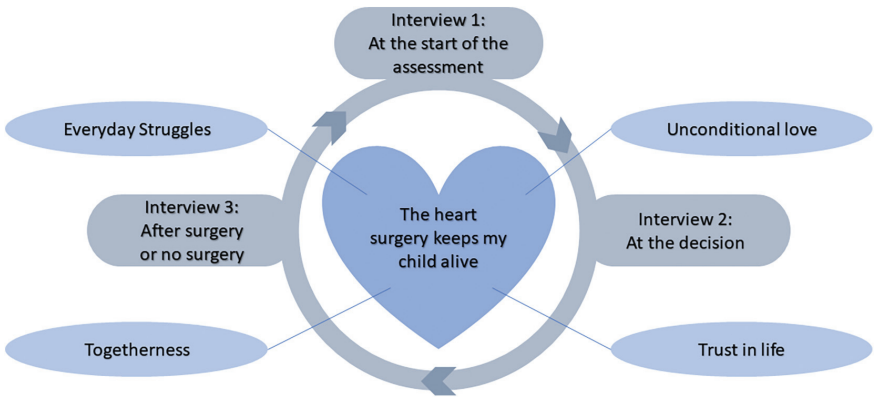

Figure 1: Thematic map of the dataset

3.1 The Heart Surgery Keeps My Child Alive

Even though the assessment was a period when both worries that the child might need a reoperation and memories of previous operations came to life, the parents found it reassuring that their child's heart was being closely monitored, since a well-functioning heart kept the child alive. The greatest fear for the parents was to lose their child or that the reoperation may lead to complications that would leave their child disabled and unable to live life as before. They were worried about how many times their child could safely be operated on. As one parent expressed it, *"It is heartbreaking that they continue to put her at risk through more operations"(M4)*, but at the same time they were aware that the surgery had improved their child's heart, which was a prerequisite for life itself. The parents of children who did not need surgery were aware that the heart valve had lasted so much longer than they were initially told, which meant that every day was a day closer to the future surgery. The outcome of the assessment, the decision that a reoperation was either necessary or not necessary, was communicated to the parents by phone, letter, or in one case at a hospital visit. The parents thought it would have been easier if they could receive the decision face to face and felt that all family members should receive the same information, especially if the decision was for heart surgery.

For those parents whose children did not feel the need of reoperation, the time leading up to the operation could be dominated by managing the child's heightening fear of dying as the time for the surgery approached. The waiting time between the decision and the surgery was also described as being on endless constant alert, including always having the phone within reach in case someone from the hospital should call. The parents commented that a shorter waiting time would have made it much easier for them. When the time for surgery arrived, the real world outside disappeared as soon as they entered the hospital world. The parents gave accounts of unexpected clinical situations in connection with the

operations, which did not turn out as planned. Some of these accounts reflected the parents' feeling of insufficient information from the health care providers before the surgery.

3.2 Everyday Struggles

Most of the parents' struggles in their everyday life took place in school, where they had to fight for their child's rights, for example by highlighting that school scheduling needed to include breaks or by repeatedly pointing out the child's need for support to be able to manage a full day. The parents' everyday life also included other kinds of struggles, such as preparing the child for a hospital visit or helping the child with endurance issues. For themselves it could involve managing anxiety far beyond the usual parental worries. The parents described needing a huge inner strength sometimes to help their child, for example when helping the child to calm down after a panic attack before an examination in the hospital or when answering difficult questions, for example, whether the child was going to die during surgery. They felt they had no instruction manual, instead they had to trust their intuitive feelings about how to handle such situations. *"I just feel it was pure instinct, that it has to be now or she will be in despair if she knows it (the MRI) won't be done (M5)"*. The parent felt they should always be there and do everything for their child, even if the situations and challenges sometimes made them feel either inadequate or powerless as a parent.

You feel so powerless, you don't quite know what to do, it's not something you can do by yourself, well of course you can support, encourage and cheer them up and all this that I do and give a lot of warmth and love and this that is needed (...) I feel inadequate, I don't know what to do, I wish I could do something, that I could just conjure, use magic, anything to be able to take this away (M4).

When the parents reflected over future surgery, they faced a new challenge compared to earlier surgeries. Their child was now older with an increased awareness and therefore needed to be prepared in a different way. At the same time, they had to take care of their own worries and protect their child from those worries. Parents appreciated it when the health care providers addressed the child directly, and they wanted help in mentally preparing their child for surgery or explaining heart disease at the child's level of understanding.

3.3 Unconditional Love

The parents' unconditional love originated from the time when they could have lost their newborn child. They remembered moments containing an emotional mix of heartfelt joy, great fear, love, and an almost overwhelming gratitude. The parents' worst memories were of the child being handed over to the anesthesiologist, when the child was baptized before the operation, or when they were not able to hold their child before the transfer to the operating table, and these memories were still, many years later, etched in their consciousness. Their feelings of gratitude were aroused when they understood that their child's heart disease could be treated and also when seeing that there were others who were worse off than themselves. When they looked back, they saw a distressing time, but their child was alive, so the outcome was good. Parents talked of the closeness they felt to the child with heart disease and how this child was dear to their heart in a different way than the child's siblings, even though they did not love this child more than their other children.

She becomes like a 'child of the heart' they are called and you become very close to each other, you do. Although I have children before who are also dear to me but it becomes a bit special precisely when they are so small from the beginning (F1).

Unconditioned love was evident in how the parents described their child's personality and their relationship. Together with that love and the child's joie de vivre, positive outlook, fighting spirit, and inner strength, the parents themselves gained inner strength and confidence. The parents expressed relief

that their child still felt so well, and it became apparent that the child's health status had an impact on the parents' wellbeing, so when their child was well, the parents were well.

3.4 Trust in Life

The parents' experienced a trust in life, and with that comes an acceptance of how everything has become as it is now. It had also given them a further insight that life is fragile and cannot be taken for granted. The parents revealed that time had shown the impossible to be possible, when getting used to the thought that their child would require several operations in life.

Being used to it did not mean that the parents were free from worries. Their best coping strategy was to postpone the anxiety until later, when it was time for surgery. Trust in and gratitude to the health care staff facilitated this procedure. The experience of feeling well treated by the health care providers was something the parents had in common, and it gave them a feeling of comfort and security. *"In addition to their professional knowledge there is a heart so they are interested, really interested in people"(F9).* The parents' feeling of trust extended to their child's future: they saw the future path as bright, even though there were worries in the far distance. Nevertheless, it was important to the parents that their child should enjoy life and find happiness in the here and now and be able to follow his or her dreams. *"But I think his best time is actually ahead of him"(M7).* This trust in life itself and in their child gave them the strength to deal with difficulties on their way through life.

3.5 Togetherness

The feeling of togetherness was an important source of strength in the parents' everyday life. Togetherness was on the whole a feasible option for coping with having a child with heart disease. Support from other people, to be able to share the burden was invaluable. Talking to others, either a partner, friend, or workmate, was the most common way of feeling less alone and being helped through difficulties. Sharing with others who were in a similar situation felt especially meaningful, since a basic understanding already existed, and at the same time it became a way of processing their own experiences. Parents expressed the importance of building a network consisting of close friends, family, and specialists around themselves but also around their child. *"Make a network, build a network around yourself to be able to handle this because you can't handle everything yourself, you can't" (M6).*

Another form of togetherness was illustrated by having faith in God, praying and knowing that others were praying for them, which was a source of strength and hope. The most important kind of togetherness was to have the family close; in the arms of the family, both the parents and their child could rest and gain strength. The child with heart disease brought the family closer, even if the journey started with navigation in unknown waters.

4 Discussion

Parents of children with complex RVOT anomalies are aware that their child will need reoperations but they live with an uncertainty of when this will happen. Since the majority of these children will undergo pulmonary valve replacement, MRI along with other imaging and clinical investigations are often used in order to best assess the need for reoperation [16]. The results of the present study showed that the parents were grateful for this assessment since it provided a thorough check-up of their child's heart. Dormant fears and new worries were awakened about whether their child would have to undergo a reoperation, but they had learned to navigate through them. They described critical situations, such as how the clinical decision after the assessment was presented to them and the period of waiting for the heart surgery after a decision for reoperation. The results also showed how the parents viewed their bond to their children, how they were coping with a range of challenges, and what they needed in order to ease the burden of having a child going through surgery while at the same time holding themselves together.

A previous study with parents to children with a specific complex CHD, namely single ventricle, showed that the parents experienced the last surgical palliation as a transition point [17]. For parents of children with complex RVOT anomalies, there is no "final surgery", as their children typically need reoperations throughout their lifetime. This need for repeated operations, that the child will never be free of the fear of surgery, is a challenge for the parents. This challenge may have contributed to an adaptation process, perhaps facilitated by their child's inner strength and well-being thus far [9]. They had adapted to and developed strategies for suppressing their worries and anxiety until the more critical situations, especially when a reoperation become a reality. A previous study showed that although mothers managed to get rid of their fear and anxiety one year after their child's surgery, they still had a feeling of lack of control [18]. In the present study, six month after surgery, the parents experienced relief and strength with traces of adaptation. We have earlier shown that their children adapted to managing their heart disease in their everyday life and could cope with the knowledge that future heart surgeries were likely [9]. An adaptation and development of strategies for managing the child's CHD is shown to be essential to coping for parents too [3], and this adaptation process may affect their need for support which could be valuable information for the health care providers.

When the parents in the present study entered the hospital for the child's heart surgery, the real world outside disappeared and their whole focus was on their child. Hospitalization in connection to surgery has been recalled in previous studies as being a situation when parents needed to employ additional strategies or support [19] and a time when parents experienced a constant level of stress during the whole hospital stay [20] and even beyond [21]. In order to ease the burden, it has been shown that providing thorough preoperative information could make the hospitalization less traumatic [22]. Such aspects of care would likely have facilitated the hospitalization period for the parents in the present study. It is also worth noting the finding in that study that parents of children undergoing heart surgery had higher levels of anxiety before their child's surgery than for parents of children undergoing other surgeries [22].

The parents unconditional love and closeness were characteristic of the parents' relationship with their child and may derive from the time of the child's birth, diagnosis, and early heart surgery, when the parents faced their worst fear: the possibility of losing their child. Unconditional love as a source of strength has not been found in previous research, but similar findings of closeness related to a stronger bonding during hospital stays have been reported [23]. The vital bonding between child and parents [19] is a prerequisite for this unconditional love and may take place in the hospital, although bonding may be more difficult in the context of a hospital stay with an infant [23]. With this in mind, health care providers should aim to facilitate and strengthen this bonding process. A previous study [24] showed that being interviewed helped mothers to infants to cope with the stress connected with their infants surgery. They suggest therefor a mental health professional to assist the mothers to tell their story.

The parents in this study gained strength from togetherness which also included the strength they drew from praying or having faith in something bigger than themselves. The fact that faith has a positive role and could be one way of gaining inner strength has been shown in previous studies with parents of children with CHD [6,19,25]. In one study, where most of the recruited parents of infants with CHD had clear religious beliefs, the parents experienced that those beliefs had a protective effect against anxiety, reduced their care burden, and further improved their quality of life [26]. In a secular country like Sweden, health care providers may need to have an open mind to consider faith as a resource that can give parents inner strength; and they should thus support and facilitate the parents in drawing on their faith.

The present study shows that there is a need for a lifelong follow-up with individualized support to meet the challenges, such as to prepare their child for heart surgery when he or she is older and has new questions and fears. This was seen to be especially hard when the child did not feel any need for reoperation, and the parents felt relieved when the health care providers turned directly to their child. A child-centered approach

[27] within health care, taking both the child perspective, meaning the parents' view of their child's experiences, and the child's own inner perspective, would be ideal. By inviting both the parents and their child to participate from the start of the assessment, the decision may be delivered according to the parents' and their child's wishes. Additionally, their individual needs during the time from the decision to the surgery may then be taken care of. Previous studies have shown that the waiting time for heart surgery is stressful for the parents [7,8,28] and also that the parents want to participate in their child's care [29]. There can be obstacles in health care to the participation of children, and in order to remove these obstacles, health care providers have to discuss and reflect upon their attitudes to children's participation in their own care [30].

5 Conclusion

Although the parents were grateful for the assessment and had learned to navigate among the fears it aroused, they experienced several stressful situations during the assessment process that must be addressed. By understanding the parents' experiences and their sources of inner strength, health care providers can better aid the parents to cope with the experience. Furthermore, by inviting both the parents and their child to participate in the child's care, an individualized support that takes into account both the parents' and the child's needs can be created.

More studies are needed to further explore the experiences of parents of children with these and other types of complex congenital heart disease in order to enable healthcare providers to tailor the way the decision making is shared with these families.

6 Limitations

The small sample size could be regarded as a limitation; however, in thematic analysis, both qualitative and quantitative aspects of the data need to be considered. The qualitative aspect is the most important, namely that the data should be rich and complex to enable a deep and nuanced analysis. Although there were few participants, the criteria were met by the rich qualitative data. The quantitative aspect is counted in number of interviews [31]. The data analyzed in this study came from 27 interviews (all nine participants being interviewed three times, with no dropouts) which is considered sufficient [13,31].

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Ethics Approval: Research involving human subjects complied with all relevant national regulations, institutional policies and is in accordance with the tenets of the Declaration of Helsinki (as revised in 2013) [32]. It was approved by the Ethical Review Board at Lund University (Approval Number 2014/66).

Conflicts of Interest: The authors declare that they have no conflicts of interest to report regarding the present study.

References

1. Ho, J. G., Schamberger, M. S., Hurwitz, R. A., Johnson, T. R., Sterrett, L. S. et al. (2015). The effects of pulmonary valve replacement for severe pulmonary regurgitation on exercise capacity and cardiac function. *Pediatric Cardiology, 36,* 1194–1203. https://doi.org/10.1007/s00246-015-1143-3
2. Wei, H., Roscigno, C. I., Hanson, C. C., Swanson, K. M. (2015). Families of children with congenital heart disease: A literature review. *Heart & Lung, 44,* 494–511. https://doi.org/10.1016/j.hrtlng.2015.08.005
3. Jackson, A. C., Frydenberg, E., Liang, R. P. T., Higgins, R. O., Murphy, B. M. (2015). Familial impact and coping with child heart disease: A systematic review. *Pediatric Cardiology, 36,* 695–712. https://doi.org/10.1007/s00246-015-1121-9
4. Golfenshtein, N., Hanlon, A. L., Deatrick, J. A., Medoff-Cooper, B. (2017). Parenting stress in parents of infants with congenital heart disease and parents of healthy infants: The first year of life. *Comprehensive Child and Adolescent Nursing, 40(4),* 294–314. https://doi.org/10.1080/24694193.2017.1372532
5. Wei, H., Roscigno, C. I., Swanson, K. M., Black, B. P., Hudson-Barr, D. et al. (2016). Parents' experiences of having a child undergoing congenital heart surgery: An emotional rollercoaster from shocking to blessing. *Heart & Lung, 45,* 154–160. https://doi.org/10.1016/j.hrtlng.2015.12.007
6. Harvey, K. A., Kovalesky, A., Woods, R. K., Loan, L. A. (2013). Experiences of mothers of infants with congenital heart disease before, during and after complex cardiac surgery. *Heart & Lung, 43,* 399–406.
7. Sjostrom-Strand, A., Terp, K. (2019). Parents' experiences of having a baby with a congenital heart defect and the child's heart surgery. *Comprehensive Child and Adolescent Nursing, 42(1),* 10–23. https://doi.org/10.1080/24694193.2017.1342104
8. Lan, S. F., Mu, P. F., Hsieh, K. S. (2007). Maternal experiences making a decision about heart surgery for their young children with congenital heart disease. *Journal of Clinical Nursing, 16,* 2323–2330. https://doi.org/10.1111/j.1365-2702.2007.02004.x
9. Svensson, B., Liuba, P., Wennick, A., Berghammer, M. (2023). "The only thing I wonder is when I will have surgery again": Everyday life for children with right ventricle outflow tract anomalies during assessment for heart surgery. *Cardiology in the Young, 33(3),* 396–401. https://doi.org/10.1017/S1047951122000907
10. Braun, V., Clarke, V. (2013). *Successful qualitative research: A practical guide for beginners*. London: Sage.
11. Braun, V., Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative Research in Psychology, 3(2),* 77–101. https://doi.org/10.1191/1478088706qp063oa
12. Braun, V., Clark, V. (2019). Reflecting on reflexive thematic analysis. *Qualitative Research in Sport Exercise and Health, 11(4),* 589–597. https://doi.org/10.1080/2159676X.2019.1628806
13. Braun, V., Clark, V. (2022). Conceptual and design thinking for thematic analysis. *Qualitative Psychology, 9(1),* 3–26. https://doi.org/10.1037/qup0000196
14. Braun, V., Clark, V. (2020). Can I use TA? Should I not use TA? Comparing reflexive thematic analysis and other pattern-based analytic qualitative approaches. *Counselling and Psychotherapy Research, 21,* 37–47. https://doi.org/10.1002/capr.12360
15. Lincoln, Y. S., Guba, E. G. (1985). *Naturalistic inquiry.* Beverly Hills, CA: Sage.
16. Mohamed, I., Stamm, R., Keenan, R., Lowe, B., Coffey, S. (2020). Assessment of disease progression in patients with repaired tetralogy of fallot using magnetic resonance imaging: A systematic review. heart. *Lung and Circulation, 19,* 1613–1620. https://doi.org/10.1016/j.hlc.2020.04.017
17. Lumsden, M. R., Smith, D. M., Twigg, E., Guerrero, R., Wittkowski, A. (2020). Children with single ventricle congenital heart defects: An interpretative phenomenological analysis of the lived experience. *Progress in Pediatric Cardiology, 59*. https://doi.org/10.1016/j.ppedcard.2020.101297
18. Menahem, S., Poulakis, Z., Prior, M. (2008). Children subjected to cardiac surgery for congenital heart disease. part 2-parental emotional experiences. *Interactive Cardiovascular and Thoracic Surgery, 7,* 605–608. https://doi.org/10.1510/icvts.2007.171066
19. Lumsden, M. R., Smith, D. M., Twigg, E., Wittkowski, A. (2019). Coping in parents of children with congenital heart disease: A systematic review and meta-synthesis. *Journal of Child and Family Studies, 28(7),* 1736–1753. https://doi.org/10.1007/s10826-019-01406-8

20. Franck, L. S., Mcquillan, A., Wray, J., Grocott, P. W., Goldman, A. (2010). Parent stress levels during children's hospital recovery after congenital heart surgery. *Pediatric Cardiology, 31,* 961–968. https://doi.org/10.1007/s00246-010-9726-5
21. Woolf-King, S. E., Anger, A., Arnold, E. A., Weiss, S. J., Teitel, D. (2017). Mental health among parents of children with chronical congenital heart defects: A systematic review. *American Heart Association, 6*, e004862. https://doi.org/10.1161/JAHA.116
22. Pomicino, L., Maccacari, E., Buchini, S. (2017). Levels of anxiety in the 24 h before and after their child's surgery: A descriptive study. *Journal of Clinical Nursing, 27,* 278–287. https://doi.org/10.1111/jocn.13895
23. Jordan, B., Franich-Ray, C., Albert, N., Anderson, V., Northam, E. et al. (2014). Early mother-infant relationships after cardiac surgery in infancy. *Archives of Disease in Childhood, 99,* 641–645. https://doi.org/10.1136/archdischild-2012-303488
24. Re, J., Dean, S., Menahem, A. (2013). Infant cardiac surgery: Mothers tell their story: A therapeutic experience. *World Journal for Pediatric and Congenital Heart Surgery, 4,* 278–285. https://doi.org/10.1177/2150135113481480
25. Wei, H., Roscigno, C. I., Swanson, K. M. (2017). Healthcare providers' caring: Nothing is too small for parents and children hospitalized for heart surgery. *Heart & Lung, 46,* 166–171. https://doi.org/10.1016/j.hrtlng.2017.01.007
26. Liu, J. F., Xie, W. P., Lei, Y. Q., Cao, H., Chen, Q. (2021). The relationship between religious beliefs and mental state, care burden, and quality of life in parents of infant patients with congenital heart disease. *Cardiology in the Young, 14,* 1–5. https://doi.org/10.1017/S1047951121004200
27. Söderbäck, M., Coyne, I., Harder, M. (2011). The importance of including both a child perspective and the child's perspective within health care settings to provide truly child-centred care. *Journal of Child Health Care, 15,* 99–106. https://doi.org/10.1177/1367493510397624
28. Kosta, L., Harms, L., Franich-Ray, C., Anderson, V., Northam, E. et al. (2015). Parental experiences of their infant´s hospitalization for cardiac surgery. *Child: Care, Health and Development, 41(6),* 1057–1065. https://doi.org/10.1111/cch.12230
29. Gramszlo, C., Karpyn, A., Abigail, C., Demianczyk, A. C., Shillingford, A. et al. (2020). Parent perspectives on family-based psychosocial interventions for congenital heart disease. *The Journal of Pediatrics, 216,* 51–57. https://doi.org/10.1016/j.jpeds.2019.09.059
30. Schalkers, I., Parsons, C. S., Bunders, J. F. G., Dedding, C. (2016). Health professionals' perspectives on children's and young people's participation in health care: A qualitative multihospital study. *Journal of Clinical Nursing, 25,* 1035–1044. https://doi.org/10.1111/jocn.13152
31. Terry, G., Hayfield, N., Clarke, V., Braun, V. (2017). Thematic analysis. In: *SAGE handbook of qualitative research in psychology.* London: Sage.
32. Association, W. M. (2013). World medical association declaration of Helsinki: Ethical principles for medical research involving human subjects. *JAMA, 310 (20),* 2191–2194. https://doi.org/10.1001/jama.2013.281053