

Children with Congenital Heart Defects Intellectual Functioning and Family Impact

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Logo

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DEPARTMENT OF PSYCHOLOGY

2019

Doctoral Dissertation in Psychology
Department of Psychology
University of Gothenburg
2019-05-24

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Printed by BrandFactory AB, Gothenburg 2019
ISBN 978-91-7833-396-7 (Tryck)
ISBN 978-91-7833-397-4 (PDF)
ISSN 1101-718 X avhandling/Göteborgs universitet, psykologiska inst.

DOCTORAL DISSERTATION IN PSYCHOLOGY

Abstract

Ryberg, C. (2019). *Children with Congenital Heart Defects: Intellectual functioning and Family Impact*. Department of Psychology, University of Gothenburg, Sweden.

The purpose of the present doctoral thesis was to investigate intellectual functioning and the influence of the children with congenital heart defects in their families. We analyzed how the severity of the heart defect, the child's age, and the socio-economic status of the child's family were related to the intellectual functioning of the children and to the impact on the family. A scale from a screening tool, the PedsQL 3.0 Cardiac module, was also tested to identify children and adolescents at risk of intellectual problems. A psychological model for understanding the development of children was used in discussing how physical and psychosocial factors affect each other and how parents, healthcare professionals, and the child's environment influence children's development.

This thesis is based in three empirical studies. Studies I and II deal with intellectual functioning in children with CHD in Sweden, and suggest a screening instrument to detect early intellectual difficulties in children. Children with a broad spectrum of congenital heart defects, different ages, different intellectually functioning and from families with different socio-economic backgrounds were investigated. Study I found out that children with CHD treated with surgery or by catheter interventions as a group performed within the normal range on overall intellectual functioning and identified severity of the heart diagnosis and SES as important factors related to increased risk for lower FSIQ. Study II found that the use of the Self-report and the Proxy-reports of PedsQL Subscale Cognitive Problems was valid as a screening tool for identifying children who need to undergo further cognitive evaluation. Study III investigate the influence of a child with CHD in the family and showed that in families of children with CHD mothers reported higher levels of negative impact than fathers, that severity of the CHD was significantly related to parental stress for both fathers and mothers, that mothers with low and medium SES reported higher stress than mothers in the high SES groups and that the strongest predictor for negative impact for both mothers and fathers was the presence of multiple risk factors. Our results show that children and parents of children with CHD are a heterogenous group and that we need longitudinal studies to help us understand how children with congenital heart defects develop over time and how their families experience the impact of having a child with CHD.

Key words: Intellectual functioning, self-reports, proxy reports, congenital heart defects, neurodevelopment, congenital heart defects, mothers and fathers of children with congenital heart defects.

List of papers

This thesis consists of a summary and the following three papers, which are referred to by their roman numerals:

- I. Ryberg, C., Sunnegårdh, J., Thorson, M., & Broberg, M. (2016). Intellectual Functioning in Children with Congenital Heart Defects Treated with Surgery or by Catheter Interventions. *Frontiers in Pediatrics*, 4(113). doi:10.3389/fped.2016.00113.
- II. 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? *Frontiers in Pediatrics*, 4(127). doi:10.3389/fped.2016.00127.
- III. Ryberg, C., Sunnegårdh, J., & Broberg, M. (Submitted). The Impact of Children with Congenital Heart Defects on their Families according to Mothers' and Fathers' reports.

To my mother and father

Svensk sammanfattning

Syftet med föreliggande doktorsavhandling var att undersöka intellektuellt fungerande och påverkan i familjen av barn med medfött hjärtfel. Vi analyserade om svårighetsgraden på hjärtfelet, barnets ålder samt socioekonomisk status i barnets familj var relaterade till a) intellektuellt fungerande hos barnen och b) påverkan i familjen. En delskala ur ett screeningsverktyg, PedsQL 3.0 Cardiac module, prövades också för att identifiera barn och ungdomar med risk för intellektuella problem. En psykologisk modell för att förstå utveckling hos barn används som stöd i avhandlingen för att diskutera hur fysiska och psykosociala faktorer kan påverka varandra och hur föräldrar, vårdpersonal, och barnets omgivning influerar barns utveckling. Modellen belyser det komplexa system inom vilket barnets utveckling äger rum och betonar föräldrarnas och omgivningens roll i att främja utveckling hos barn i allmänhet och i detta fall barn med medfött hjärtfel.

Medfött hjärtfel (CHD) är den vanligaste medfödda missbildningen hos människor och innebär ett betydande och globalt hälsoproblem (Dolk, Loane, gran, och grupp, 2011, Seghaye, 2017; Van der Linde et al, 2011). Det ökade antalet överlevande barn med CHD har ökat medvetenheten om vikten av att förbättra barnens långsiktiga möjligheter, till exempel när det gäller att kunna utveckla sin fulla potential och få ett gott liv (Lundell, 2005). Den neuropsykologiska utvecklingen, inklusive intellektuellt fungerande, är ett område med stor betydelse för framtidsutsikterna för barn med CHD. Studier har visat att många barn med CHD har lägre intellektuell funktion än genomsnittet, men det är oklart vilka faktorer som är relaterade till lägre intellektuell funktion i denna grupp. Forskning om riskfaktorer för nedsatt intellektuell funktion är angeläget givet den inverkan som intellektuell funktion har i vardagen för barn.

Barn tillbringar en stor del av sin tid i skolan där deras prestationsförmåga kontinuerligt utvärderas. Prestationsförmågan inverkar på barns självförtroende och tilliten till förmågan att klara sig själv; och kan vara en källa till oro hos föräldrar när det gäller barnets framtida möjligheter i samhälle. Föräldrar behöver information som hjälper dem att ha positiva förväntningar på vad barnen kan förväntas klarar av men också information om när och var de kan söka professionellt stöd. Det finns en allmän oro för att många barn med CHD har intellektuella problem som gör det svårt för dem att klara sina studier, behålla ett jobb och försörja en familj. Tidig identifikation av barn som löper risk för utveckling av låg intellektuell förmåga och svårigheter att klara skolan är av stor betydelse för att kunna implementera lämpliga åtgärder och stödja positiv utveckling hos barnet. Både föräldrar och professionella är

därmed i behov av vetenskaplig kunskap om faktorer av betydelse för intellektuellt fungerande och familjepåverkan av barn med CHD.

Avhandlingen bygger på tre empiriska studier; två studier handlar om intellektuell funktion hos barn med CHD i Sverige och en studie undersöker påverkan i familjen av att ha ett barn med CHD. I de första två studier var det övergripande syftet att analysera hur psykosociala och socioekonomiska faktorer påverkar den intellektuella funktionen hos barn med CHD i Sverige. Studierna beskriver intellektuell funktion hos barn med ett brett spektrum av medfödda hjärtdiagnoser och föreslår ett screeninginstrument för att tidigt kunna upptäcka intellektuella svårigheter hos barn med hjärtfel, vid olika åldrar och från familjer med olika socioekonomisk bakgrund. Syftet i den sista studien var att undersöka hur föräldrar upplever påverkan av deras barn med CHD i familjen; även här undersöks barn med ett brett spektrum av medfödda hjärtfel, i olika åldrar, olika intellektuellt fungerande och i familjer med olika socioekonomisk bakgrund.

I studie I beskrivs och jämförs intellektuell funktion hos barn med medfött hjärtfel i förhållande till diagnosens svårighetsgrad, barnets ålder, och socioekonomisk status (SES) i familjen. Barn med CHD som hade genomgått kirurgi eller kateterbehandling testades med hjälp av Wechsler intelligensskalor för att bedöma fullskala IQ (FSIQ). FSIQ analyserades sedan i förhållande till ålder (3-, 5-, 9- och 15-år), diagnosens svårighetsgrad (mild, måttlig och svår) och SES (låg, medel och hög). Resultaten visade att barn med svår CHD hade signifikant lägre FSIQ än barn med mild och måttlig CHD och att barn från familjer med låg SES hade lägre FSIQ än barn från familjer med medel eller hög SES. Dessa faktorer bör beaktas vid planering av interventioner och uppföljningsprogram för barn med CHD.

I studie II utvärderades om en delskala (kognitiva problem) i ett screeningsverktyg för pediatrik livskvalitet (PedsQL 3.0, Cardiac module) kunde användas för att identifiera barn och ungdomar med risk för intellektuella svårigheter. PedsQL-data samlades in med självskattning (i åldrarna 5, 9 och 15 år) och proxy-rapporter från föräldrarna vid 3, 5, 9 och 15 års ålder. Data från den korta screeningen (5 items) relaterades till uppnått testresultat på FSIQ (Wechsler intelligensskalor). Resultatet visade en stark association mellan föräldrarnas skattning av kognitiva problem och faktiskt resultat på IQ-test. Detta visar att det är möjligt att identifiera barn med kognitiva problem så tidigt som tre år genom proxy-rapporter av kognitiva problem. Sammantaget visar studierna på behovet av ett multidisciplinärt uppföljningsprogram samt behov av tillförlitliga instrument för att upptäcka barn i riskzonen för intellektuella problem.

I studie III undersöktes hur familjen påverkas av barn med medfött hjärtfel enligt föräldrarnas rapporter. Positiv och negativ påverkan i familjen analyserades i relation till fäder och mödrars rapporter och i relation till barnets ålder, svårighetsgrad av diagnosen, intellektuell fungerande hos barnet med medfött hjärtfel och SES i familjen. Resultaten visade att mödrar rapporterade mer negativ inverkan i familjen än fäder, att SES var associerade med negativ inverkan enligt mödrar men inte fäder och att svårighetsgrad i diagnosen var relaterad till påverkan i familjen enligt både mödrar som fäder. Ett oväntat resultat var att föräldrar till barn med genomsnittlig och låg intellektuell funktion rapporterade mer positiv påverkan av barnet i familjen än vad föräldrar till barn med hög intellektuell funktion gjorde. Dessutom rapporterade både mödrar och fäder att den kumulativa risken, dvs att ha två eller flera riskfaktorer, var signifikant associerad med negativ påverkan i familjen. Socioekonomisk status, diagnosens svårighetsgrad och barnets intellektuella funktion har visat påverka familjen på olika sätt. Därför bör dessa riskfaktorer beaktas vid planering av interventioner och uppföljningsprogram för familjer med barn med CHD.

Studierna har vissa svagheter såsom avsaknad av kontrollgrupper och att svarsfrekvens var proportionell högre i gruppen med mest komplex CHD jämfört med de med mindre allvarlig svårighetsgrad av hjärtfel. Våra studier var tvärsnittsstudier, därför behöver resultaten tolkas med försiktighet. Vi behöver longitudinella studier som hjälper oss att förstå hur barn med medfött hjärtfel utvecklas över tid, hur deras familjer upplever påverkan av att ha ett barn med CHD.

Abbreviations

AHA	American Heart Association
BUP	Child and Youth Psychiatric Center
CHD	Congenital Heart Defect
DD	Developmental Disorders or Disabilities
DSM-5	Diagnostic Statistical Manual of Mental Disorders, fifth ed.
DSA	Developmental Systems Approach
FSIQ	Full Scale Intelligence Quotient
HLHS	Hypoplastic Left Heart Syndrome
HRQOL	Health Related Quality of Life
ID	Intellectual Disability
IF	Intellectual Functioning
IQ	Intellectual Quotient
PedsQL	Pediatric Quality of Life Inventory
SCC	Satisfaction with Care
SES	Socioeconomic Status
WISC-IV	Wechsler Intelligence Scale for Children- 4th revision
WPPSI-III	Wechsler Preschool and Primary Scale of Intelligence- 3th revision

Acknowledgements

I would like to start by thanking all the children and parents who have participate in this study for their patience and invaluable contribution when answering tests and questionnaires, adding more time to the time they already spend in hospitals and at doctors' appointments. I have enormous admiration and gratitude for these children and their families for teaching and helping me understand how they and their families deal with the impact of being born with a heart disease.

I would also like to thank all financial contributors that have made this study possible. A main contributor of this work was The Swedish Children's Heart Association (*Hjärtebarnsförbundet*). This association has not only helped me with financing research but also with encouragement and trust in my work, helping me meet parents and children every year in their annual activities. As well I am grateful to *Sahlgrenska Universitetssjukhuset*, which has provided me with support and has made it possible to carry on my research at The Queen Silvia Children's Hospital of Gothenburg in the pediatric cardiology department.

An enormous gratitude to my supervisors Malin Broberg and Jan Sunnegårdh for their endless patience in guiding me through the long and many times heavy process of learning how to conduct research. Thank you for always being there when I needed a supporting hand and guidance.

A very important contribution to this project was given by the invaluable work of my colleagues Elisabet Björklund, Azarnosh Leufrèn and Maria Leinonen working at the participating hospitals, Skaraborg Hospital Skövde (SkaS), North Älvsborg County Hospital (NÄL) and Southern Älvsborg Hospital (SÄS). My colleagues not only engaged in collecting data for the project, but also contribute in enriching discussions during the process of data gathering.

Last but not least, I want to Thank Maria Thorson for the years of assistance and engagement in this project as well as my colleagues Lotta Leffler and Sandra Buratti for their invaluable work with testing and analyzing data, resulting in this thesis and two published articles. Thank you for being part of a research team that was always happy to meet children with heart diseases and their families.

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Introduction

This doctoral thesis investigates intellectual functioning (IF) in children with congenital heart defects (CHD) in Sweden and factors that influence IF in these children. Furthermore, it explores the impact that children with CHD have in their families according to parents' reports. I use a psychological model to support the discussion on how physical and psychosocial factors influence each other and how parents, health care professionals, and the wider society play important roles in the development of these children. The model helps us understand the complex system within which child development takes place and draws attention to the important roles that parents and the environment play in promoting the development of children with CHD.

CHD, a common cause of congenital malformations, represents a major worldwide health problem (Dolk, Loane, Garne, & group, 2011; Seghaye, 2017; van der Linde et al., 2011). In Sweden, CHD is one of the leading causes of death in full term babies and small children (Lundell, 2005). In developed countries like Sweden, these heart defects can be diagnosed before birth, allowing parents the possibility to terminate the pregnancy or to prepare for special care of their child at birth. Because these children now live longer due to advances in health care, it has been suggested that research should examine what special needs these children have and what interventions are needed to improve the children's opportunities so they can develop to their full potential and live long and good lives (Lundell, 2005).

To help children with CHD develop to their full potential, more research in neuropsychological development, including IF, is needed. Although studies have shown that children with CHD are at risk for reduced intellectual function, it is still not clear what physical and psychosocial factors impact IF in this group. Research on risk factors for impaired IF has become of interest because of the impact IF has on children's everyday lives. Children spend a great deal of their time at school where their performance is continually evaluated, affecting children's self-esteem and trust in their own abilities. Parents need information that helps them form positive and realistic expectations about their children, the impact they have in their families, and prepare them to seek professional support when needed.

The dissertation is based on three empirical studies. Two studies examine intellectual function in children with CHD in Sweden and a one examines the impact the children have on their families according to parents' reports. In the first two studies, the overall purpose was to analyze how physical and psychosocial factors affect the intellectual function of children with CHD in Sweden. These two studies describe intellectual function in children

with a wide range of congenital heart diagnoses and suggest a screening instrument for early detection of intellectual difficulties in children with heart failure at different ages and in families with different socioeconomic backgrounds. Study three investigates how parents of CHD children experience the impact of their children in their families in relation to the same risk-factors; i.e., child age, severity of CHD, and SES in the family.

The outline of the present thesis is as follows. I begin by defining CHD, its incidence in different populations, and the expected risk for intellectual difficulties or developmental disorders in relation to the severity of the CHD diagnosis. I discuss the impact that children with congenital heart defects have on their families addressing both the positive and the negative impacts, differences between fathers and mothers on reported impact, and introduce some interventions reported in the literature to promote family functioning. Later, concepts such as intelligence and cognitive functions are defined since these terms are often used indiscriminately in the literature. I present theories of cognitive development fundamental to understanding children's IF. To provide a research context, I review previous studies on children with CHD treated either by surgery or by catheter interventions. Finally, I introduce a model from the perspective of developmental science that addresses factors involved in the development of social and cognitive competence. The model is meant to help identify and understand individual differences in intellectual development in children in general and in children born with medical risks in particular, emphasizing the fundamental importance of family resources and interactions in children's development. Such models are of great help in assessing risks as well as protective factors for children's development and can be used to guide the design of prevention and interventions programs within medical and psychological pediatric health practices.

Background

Definition and Prevalence of Congenital Heart Defects (CHD) in Children

The four major cardiovascular diseases affecting children are acquired heart disease, arrhythmias, systemic hypertension, and congenital heart defects (CHD). CHD is the most prevalent (Delamater & Jent, 2009; Dolk et al., 2011; Seghaye, 2017; van der Linde et al., 2011). The American Heart Association (AHA) describes CHD as structural defects in the heart and major blood vessels present at birth; these defects develop soon after conception and often before the mother is aware that she is pregnant. Defects range in severity from simple, such as “holes” between the chambers of the heart, to very severe malformations, such as complete absence of one or more chambers or valves (AHA, 2017).

CHD can now be detected during fetal development; studies of fetuses done with ultrasound have shown that congenital heart defects are more common in the fetus than after birth and that the disease prospects are more serious in the fetal stage than in live births. Studies of spontaneous abortion fetuses have shown severe CHD, often in association with chromosome disorders (Sunnegårdh, 2000). The cause of CHD is unknown in most cases, yet it is believed to be a result of the combined influence of genetically-determined predispositions, chromosomal malformations, and environmental factors.

There are different ways to classify CHDs. One classification is based on the anatomical defects of the heart, grouping the different heart defects in two subtypes: acyanotic and cyanotic.

Acyanotic CHDs, the most frequent types, involve holes in different parts of the heart compartments thereby routing the fully oxygenated blood back into the lungs rather than other parts of the body (e.g., ventricular septal defects, atrial septal-atrial ventricular canal defects, patent ductus arteriosus, coarctation of the aorta, and valvular lesions).

Cyanotic CHDs are associated with lesions that obstruct the normal blood flow, reducing as a consequence the oxygenation level of the blood (e.g., cardiomyopathy, hypoplastic left heart syndrome, pulmonary atresia, tetralogy of Fallot tricuspid atresia, and transposition of the great arteries) (Bernstein, 2004 cited in (Delamater & Jent, 2009).

CHD and Developmental Disorders or Disabilities

In this thesis, the classification of diagnosis severity was shaped by Professor Jan Sunnegårdh (Jan Sunnegårdh, 2011). The classification considers anatomical defects or diagnoses as well as the probability that these and the treatment will lead to further medical complications for the child. This division concurs with studies showing that the complexity of the heart disease is often associated with the risk for impairments in cognitive, behavioral, or physical functions. The presence of developmental disorders or disabilities (DD) is reported to be higher in children with severe CHD compared to the general population (Marino, et al., 2012).

A Scientific Statement of the AHA (2012) has established a detailed classification of comorbidity that reflects how the different heart diagnoses are related to the level of risk for children with CHD to present intellectual difficulties or developmental disorders or disabilities (DD). This classification places children with CHD into four main groups (similar groupings as used in the present studies). In our studies, however, we have used only the first three groups (i.e., the fourth group was excluded from the study; this will be explained later). Below is an explanation of each group.

Children with mild forms of CHD. Although this group has the highest incidence, few children in this group show signs of DD (AHA, 2012). Children in this group have diagnoses such as atrial septal defect, ventricular septal defect, or isolated semilunar valve disease. These children are usually treated once with surgery or by catheter intervention with little risk for further complications and little or no need for rehabilitation. Children in this group are seldom followed-up medically with respect to their CHD and are not part of other rehabilitation or follow-up programs at the hospital.

Children with complex forms of moderate two-ventricle CHD. According to the Scientific Statement of the AHA (2012), these children are at higher risk for DD. Children in this group have diagnoses such as coarctation of the aorta, complex semilunar valve disease, atrioventricular septal defect, ventricular septal defect with comorbidities, tetralogy of Fallot, and total anomalous pulmonary venous connection. This group is equivalent to the moderate severity group in our studies and includes children treated with surgery and/or by catheter interventions. These children are followed-up regularly by their home hospital since they are at risk for further complications and sometimes in need of further rehabilitation. These children, although part of medical follow-ups, are not usually part of follow-up rehabilitation programs unless they present specific needs. If this is the case, the attending pediatrician refers

them to specialists (i.e., no multidisciplinary assessment is done on a routine basis).

Children with severe two-ventricle or palliated single-ventricle CHD. According to Scientific Statement of the AHA (2012), these children are at high risk for DD. A minority of the children in this group are expected to perform within the normal range in all developmental aspects. Children in this group are those with diagnoses such as transposition of the great arteries, truncus arteriosus, interrupted aortic arch, tetralogy of Fallot/pulmonary atresia with major aortopulmonary collateral arteries, pulmonary atresia with intact ventricular septum, hypoplastic left heart syndrome, and tricuspid atresia. This group is equivalent to the severe CHD group in our studies. Children with complex heart defects have a more uncertain long-term prognosis (i.e., serious cardiac complications are common) and these children often need rehabilitation measures. Children in this group have the possibility to be part of a follow-up program in one of the two cardiac centers for children in Sweden, either in Gothenburg or Lund. However, since many of these children come from remote areas, their families often find it difficult to consistently be involved in rehabilitation or follow-up programs.

Children with genetic disorders or syndromes co-occurring with CHD. Nearly all of the children in this group have DD. Children in this group include those with diagnoses such as Down syndrome, 22q11 deletion, Noonan syndrome, Williams syndrome, and multiple congenital anomalies such as CHARGE syndrome (Marino et al., 2012). After discharge from the hospital, these children are referred to the habilitation center near their home. Habilitation centers have specialist multidisciplinary teams that offer support, knowledge, and means to access their facilities and services. Children in this group with comorbid conditions were excluded from the studies in the present thesis because of the known impact on intellectual functioning.

Prevalence and Survival

The Global Burden of Disease study reported that the leading cause of death among younger children in North America, Australia, Europe, East Asia, and most countries in Latin America and Caribbean between 1990 to 2013 was preterm birth complications and congenital anomalies (Collaboration, 2016). Prevalence of CHD varies significantly over time and shifts among populations. The total reported CHD prevalence has increased from 0.6 per 1,000 live births in 1934 to 9.1 per 1,000 live births in 1995, with Asia having the highest prevalence (9.3 per 1,000 live births) and Africa having the lowest (1.9 per 1,000 live births) (van der Linde et al., 2011).

Economic wealth in a country is highly related to the number of live born children with CHD. The highest reported CHD birth prevalence (8.0 per 1,000 live births) is reported in high-income countries, decreasing (7.3 per 1,000 live births) in upper-middle income countries (6.9 per 1,000 live births), and in lower-middle income countries. Data from low-income countries are not available (van der Linde et al., 2011). Sweden has been successful in decreasing mortality rates among newborn babies with congenital heart defects and reflects the international rate of eight children with CHD per 1,000 live born babies (Lundell, 2005).

Survival rates are related to the complexity of the heart disease. It is expected that 95% of children with mild forms of CHD will live past their twenties. For children with moderate forms of CHD, the long-term survival rate (>20 years) decreases to 90% and for children with the most severe forms of CHD, the expected survival past 20 years decreases to 80% (Marino et al., 2012; Said, Driscoll, & Dearani, 2015; Warnes, 2005).

Pediatric Cardiac Care in Sweden

The health care available to children in Sweden is considered to be among the best in the world (Wettergren, Blennow, Hjern, Söder, & Ludvigsson, 2016). Sweden's relatively good economy has made it possible to sustain a social insurance system that supports families and children with free primary and specialized health care (Wettergren et al., 2016).

As Sweden has about 10 million inhabitants spread over a large area, to ensure high quality care certain pediatric health care services have been centralized. After pediatric heart surgery was centralized to two centers in Sweden in 1993, pediatric mortality rate for open heart surgery went from 9.5% (1988 – 1991) to 1.9% (1995 – 1997) (Lundström, Berggren, Björkhem, Jögi, & Sunnegårdh, 2000). Together, the Nordic countries – Finland, Denmark, and Sweden – have decreased mortality rates within 30 days of surgery from 10% in 1991 to 1% in 2010 (Lindberg, 2012). Mortality rates continue to decrease in children with CHD, an improvement that has brought into focus the long-term consequences for these children. In the western world, the fastest growing patient population is of adults with CHD (Lindberg, 2012), a trend that will require society to address new challenges associated with this group's long-term survival, challenges that include attending not only to their physical needs but also to their cognitive and psychosocial needs as well as understanding family aspects of bringing up a child with CHD, gaps in knowledge that this thesis will eventually address.

Family-Centered Care

Swedish health care has developed in many ways since modern health practices were introduced in the 1930s. An important aspect of this development has been the shift of the responsibility placed on different actors, moving from public responsibility towards more parental responsibility and participation (Hallberg, Lindbladh, Petersson, Råstam, & Håkansson, 2005). Today, parents take on greater responsibility for their children's health care while public health care are more focused on providing more attention to families as a whole. This new approach emphasizes the psychosocial aspects of care and public care providers support families by helping them make well-informed health decisions rather than by telling them what to do (Hallberg et al., 2005). This shift towards a more family-centered care aims to empower families in health care by teaching them how to access knowledge and support their decision-making, strategies that have had many positive effects. In Sweden, health care professionals have worked with a family-centered care approach, where the development of a partnership between professionals and the family of the sick child is a fundamental task (Coyne, Holström, & Söderbäck, 2018).

Although many advances have been done in the care of sick children, levels of anxiety have not significantly decreased in parents of children with chronic diseases such as CHD (Utens & Levert, 2015). A study on the impact of infants' severe congenital heart disease on the family (Werner, Latal, Valsangiacomo Buechel, Beck, & Landolt, 2014) reported that parents of children diagnosed with a severe CHD who had undergone bypass surgery within the first year of life most frequently described not wanting to have more children and having to live a life full of ups and downs (like a "roller coaster"). Furthermore, the study indicated that the type of CHD (cyanotic vs. acyanotic) or the number of medications did not predict the impact of the child's disease on the family. However, the presence of a genetic disorder and low levels of perceived social support have more negative disease impacts, suggesting that good social support networks probably mediated the daily stressors related to the child's CHD, resulting in lower impact on the family (Werner et al., 2014).

Child-Centered Care

Over the past decade, child-centered care has become part of pediatric care. The need for a concept that placed children and their interests at the center of health care thinking and practices was raised by the UN Convention on Children's rights in 1989 (*Convention on the rights of the child*, 1989). This concept considers children and adolescents as active participants in procedures and

decision-making regarding their health care (Ford, Campbell, Carter, & Earwaker, 2018). Child-centered care gives the child a voice and promotes his/her engagement. In addition, the relationship with the family has to be considered in the child's care; a family-centered care focuses on how to encounter the family with a sick child and on practices that view the family as a unit (Coyne et al., 2018). Child-centered care has also been associated with the term person-centered care, which emphasizes empowerment and respect for autonomy (Coyne et al., 2018) and is mainly used in adult care settings. Child-centered care and person-centered care, however, share the same values and highlight the principles of the person's/child's rights to dignity and respect.

At a conceptual level, there are more similarities than differences in all these terms, since they all highlight the respect for the patient and the importance of working towards a good partnership between health care professionals, children, and families. Different clinical environments might determine which approach to take; family-centered care might be more appropriate in the context of new-born with CHD. In addition, when more surgical procedures are needed as the child becomes older, child-centered care might be more appropriate, improving satisfaction and health care outcomes. Furthermore, since the majority of children with CHD are now expected to become adults, become independent, and be transferred to adult care (Grown Up Congenital Heart Disease, GUCH), person-centered care will be the right approach.

Satisfaction with Care

Medical care and how parents perceive the care they receive have been studied by the Department of Public Health Sciences at the Karolinska Institute in Sweden (Lawoko & Soares, 2004). This study compares satisfaction with care (SCC) between parents of children with CHD and parents of children with other diseases, reporting that parents of children with CHD were more satisfied with their children's medical care and waiting periods for treatment than parents of children with other diseases. Furthermore, Lawoko (2004) reported that the disease severity had little impact on the SCC for parents of children with CHD and parents of children with other diseases as SCC was mostly determined by parental and child characteristics. For example, increased financial burden, high levels of distress, feelings of hopelessness, and social isolation among parents were associated with lower SCC, whereas higher age of children and their parents, better social interaction, and better knowledge about clinical procedures were associated with higher SCC (Lawoko & Soares, 2004).

Furthermore, parental perceptions of their child's care are highly important for how the child adapts to their own disease. Clinical experience as well as studies have shown that children with CHD are influenced not only by the severity of their cardiac disease and the type of medical care received (Testone & Terrier, 2003) but also by how parents and relatives perceived the medical care of the child and how they are able to focus on the psychological needs of their children (Daliento, Mapelli, & Volpe, 2006).

Parents of Children with CHD

Giving birth to a child normally brings demands and new challenges for any parent. When the child is affected by a CHD, those demands and challenges may be accentuated by worry and extra care-taking demands (Montis & Tumbarello, 2011). Distress symptoms, common in all parents after the birth of a child, often intensify when the child has a chronic illness (Lawoko & Soares, 2002). Fear of potential loss of the child, worry about the child's future psychological well-being, and fear about the impact that the child might have on the family are among the main concerns reported by parents and children with CHD (Du Plessis et al., 2018; Simeone et al., 2018; Van Horn, DeMaso, Gonzalez-Heydrich, & Erickson, 2001).

Parents of children with CHD have shown higher risk for developing mental health problems following cardiac surgery of their child (Woolf-King, Anger, Arnold, Weiss, & Teitel, 2017) and discharge from hospitals (Fischer et al., 2012). Mothers often report higher levels of stress than fathers (Gardner, Freeman, Black, & Angelini, 1996; Goldberg, Morris, Simmons, Fowler, & Levison, 1990; Lawoko & Soares, 2006). In terms of family impact, distress in families of children with chronic conditions is often reported as marital dissatisfaction (Berge, Patterson, & Rueter, 2006); however, the divorce rate has not been found to be higher in families of children with CHD than in control families (Sabbeth & Leventhal, 1984). As a group, parents of children with CHD report higher levels of quality of life than parents of children with other chronic diseases, with the exception of parents of children with severe cardiac diagnosis such as HLHS. Parents of children with HLHS have reported more stress and personal strain as well as higher levels of demand and challenges resulting in feelings of lowered parenting competence (Brosig, Mussatto, Kuhn, & Tweddell, 2007).

It is important to point out that the majority of parents of children with CHD create families that function as well as any other families and that a majority of parents are able to cope with the stressors presented by the child's condition (Hearps et al., 2014). Studies have reported that most parents

of children with CHD are capable of coping with the demands of having a sick child (Rodriguez & Patterson, 2006; Visconti, Saudino, Rappaport, Newburger, & Bellinger, 2002). However, a group of parents, often of children with severe CHD, have shown an elevated risk for psychological problems and symptomatology (Hearps et al., 2014; Helfricht, Latal, Fischer, Tomaske, & Landolt, 2008). Predictors that help identify parents at risk need to be studied further. Parents' education and available social support systems both at society levels and within families are factors often mentioned as determinants of good adaptation to the demands of having a child with CHD (Hearps et al., 2014; Helm et al., 2018; March & Kein-Malpass, 2018; Visconti et al., 2002).

Families of Children with CHD

Families of children with CHD have often been described as receiving different kind of impacts from the disease. The term impact has often been used in the literature to describe negative feelings or stress. In the instrument used in the present study, both the negative and positive impact as well as the impact on siblings were reported by parents of children with CHD.

Positive impact in families of children with CHD

Positive emotions in parenting, such as joy and happiness, are related to the interactions parents have with a specific child (Kurtz-Nelson & McIntyre, 2017) and are fundamental in the development of children. According to Fleck (2015), the experience of positive impact of the child is associated to feelings of enjoyment and pride that the child brings to the family and feelings of competence concerning taking care of the child (Fleck et al., 2015). In studies of children with developmental delay, positive feelings and optimism serves as a protective factor for parents experiencing high levels of stress (Kurtz-Nelson & McIntyre, 2017).

Although positive feelings are common in parents, in the case of children with CHD sometimes the disease itself might bring positive effects on the family. A study addressing the psychosocial outcomes after cardiac surgery in pre-schooled children reported that some parents described a positive "value" of having a child with CHD in the family (Brosig et al., 2007). Although parents worried about their child's future, they also reported that family members were closer and more understanding because of what they had gone through with their child and that they were thankful for the support of relatives and friends; furthermore, most parents reported that they had learned so much

about their child's condition making them feel good about themselves (Brosig et al., 2007). Some parents have described their child as having more "compassion, tolerance and maturity" than their peers and reported having gone through a re-evaluation of what is important, helping them appreciate life (Kratz, Uding, Trahms, Villareale, & Kieckhefer, 2009).

Parents of children with CHD have been described as fluctuating between fearing for their children's vulnerable state and celebrating their child's strength due to their survival (Lee & Rempel, 2011). This has been described as "the embrace of the paradox" where parents must contain and struggle with their internal process of loving their child as he/she is and wanting to get rid of the illness, between maintaining hope, being positive, and finding solutions while receiving negative information about their child's condition (Larson, 1998). The embrace of the paradox has been reported in studies of parents of children with CHD (Lee & Rempel, 2011; Wray et al., 2018) where parents report the tension of being grateful and celebrating their child's recovery after surgery while not feeling they have the right to talk about their fears and anxieties during the process. In studies of parents of children with intellectual disabilities, "the embrace of the paradox" has also been addressed, adding that sometimes accepting conflicting thoughts without being preoccupied by those thoughts by letting positive emotions co-occur with negative emotions during stressful events, help parents access their psychosocial coping resources (Boström, Broberg, & Hwang, 2009; Folkman, 2008).

Some studies have addressed factors that mitigate and even benefit parents during the adaptation process of living with a child with CHD. These factors include the positive feelings resulting from the experience of managing their situation, strengthening their experience as parents, being able to participate in training and information forums, and accessibility of advice (e.g., contact with cardiac liaison nurses and contact with other "heart parents and families") (Wray et al., 2018). In the specific case of fathers, it has been shown that the more they participate in the additional care or the medical treatment of their children, the more they experience assured effects of being positively engaged with their infants (Darke & Goldberg, 1994). The perception of having good social support has been reported to help parents maintain their motivation towards coping, facilitating them to mobilize their cognitive and behavioral efforts to better adaptation (Tak & McCubbin, 2002).

Negative impact in families of children with CHD

Most parents find that affection comes during the days after the birth of their child. However, when the birth is associated with complications and

medical procedures, it might bring disillusionment and negative feelings, especially in mothers (Roseth, Bongaardt, Lyberg, Sommerseth, & Dahl, 2018). According to Fleck (2015), this negative response is associated with stress, anxiety, and anger. In the case of families of children with CHD, parents either already know that their child has a heart condition or will be diagnosed almost immediately after birth.

Negative impact is associated with the medical condition of the child and includes the psychological and emotional consequences in parents related to the health condition. Negative impact could be stress, anxiety, depression, and hopelessness, sometimes including behaviours such as social isolation, which is frequently reported in parents of children with CHD (Fleck et al., 2015; Gregory, Prouhet, Russell, & Pfannenstiel, 2018; Kaugars, Shields, & Brosig, 2017; Wray et al., 2018).

Stress is often addressed in studies of parents having children with CHD (Soulvie, Desai, White, & Sullivan, 2012), reporting sometimes significantly higher stress scores than norm groups, and even reaching clinically significant levels (Uzark & Jones, 2003). Parents of children with CHD often express experiencing anxiety, depression, and trauma during the diagnosis of their child, hospital stays for surgery, and during transitions from hospital to home (Woolf-King, Arnold, Weiss, & Teitel, 2018). A study of parents of children with CHD the first year of life reported that parents with CHD showed significantly higher parenting stress than parents of healthy children, especially because parents experienced their infant as more demanding and difficult to soothe, intensifying their feelings of incompetence (Golfenshtein, Hanlon, Deatrck, & Medoff-Cooper, 2017).

A review of the literature reported severity of the heart disease and socioeconomic status to be associated with negative impact on the family; uncertainty of the child's condition with severe diagnosis such as HLHS was highly related to stress in parents as well as to social isolation and financial instability (Jackson, Frydenberg, Liang, Higgins, & Murphy, 2015). Studies on parental stress seem to agree that for most families of children with CHD – independently of severity of the heart diagnosis – the hospitalization periods are highly stressful (Jackson et al., 2015; Vrijmoet-Wiersma, Ottenkamp, van Roozendaal, Grootenhuis, & Koopman, 2009). Furthermore, prematurity in children with CHD is highly related to parental stress, since these children are at even higher risk for presenting neurodevelopmental problems, which in turn might increase concerns, stress, and total burden for parents and their families (Kaugars et al., 2017).

Impact on siblings in families of children with CHD

Impact of CHD on siblings have been scarcely described in the literature. However, there is an agreement in the literature that the consequences for parents and siblings of children with severe CHD are permanent and interfere with all aspect of life for these families (Brosig et al., 2007; Caris et al., 2018). A meta-analysis of siblings of patients with chronic diseases reported that parents tend to rate the impact of the disease more negatively than siblings do (Sharpe & Rossiter, 2002) and that siblings reported fewer problems adapting than parents do (Caris et al., 2018). However, internalizing problems such as anxiety and depression were often associated with siblings when the disease was severe, interfering with every day activities when the disease was life-threatening (Sharpe & Rossiter, 2002; Vermaes, VanSusante, & Bakel, 2012). Siblings reporting negative adjustment to the disease have shown to be younger (Guite, Lobato, Kao, & Plante, 2004), which has been hypothesized to depend on the closer age to their sibling being diagnosed, the more acute the crisis is in the family, and because younger children have not yet developed coping strategies to help them deal with their sibling's disease (Guite et al., 2004).

Family interventions for families of children with CHD

Family interventions for families of children with CHD should help parents balance their emotions of the perceived positive and negative impact of the child with CHD. Negative impacts in families do not necessarily imply lower positive impacts. However, reporting high levels of negative impact and low levels of positive impact might indicate risk for adverse consequences for parents of children with CHD and their families. Parents with high levels of stress might experience higher levels of negative feelings and a decrease of positive feelings, affecting parent-child interactions (Kurtz-Nelson & McIntyre, 2017). Parent-child interactions are one of the most influential factors for child development as they are essential in developing the social and intellectual competence of children (Guralnick, 2006, 2012).

Health care professionals need to be aware of the impact CHD has on family members and family relations, including how parents cope with stress and knowledge about their adaptation process to the illness (Ahn, Lee, & Choi, 2014). This awareness might help develop family psychosocial interventions that can be incorporated to health care routines. Results from studies measuring psychosocial outcomes in families of children with CHD have emphasized the importance of asking about parental stress and family functioning as well as behavioral and learning difficulties within the context of routine

pediatric cardiology follow-up programs (Brosig et al., 2007; Woolf-King et al., 2018). Children with CHD in families that perceive good health care and social support as well as support from their family and friends have shown better outcomes in their physical and psychological health (Tak & McCubbin, 2002). Furthermore, these families are more prone to access resources and function more effectively (Wray & Maynard, 2006), consequently reducing the level of cost for society.

Integrating parents and other family members in psychosocial interventions (i.e., family-based interventions) is the most often recommended strategy in the literature in promoting psychological well-being of children with CHD (Ahn et al., 2014; Brosig et al., 2007; Bruce, Lija, & Sundin, 2013; Landolt, Ystrom, Stene-Larsen, Holmstrom, & Vollrath, 2014; Smith, Swallow, & Coyne, 2015; Sood et al., 2018; van der Mheen et al., 2018; Woolf-King et al., 2018; Wray & Maynard, 2006). However, very few interventions have been designed to meet the needs of families of children with CHD.

In Europe, the first evidenced-based intervention program initiative was The Congenital Heart Disease Intervention Program – School (CHIP-S) (McCusker, Doherty, Molloy, Casey, Rooney, Mulholland, Sands, Craig, & Steward, 2012). This intervention was designed to support and improve mental health and to reduce stress in parents of preschool children with CHD. In collaboration with a research team in the Netherlands, this program has been extended to include a module for children with CHD and his/her siblings, resulting in the CHIP-Family (CHIP-F) (van der Mheen et al., 2018). The CHIP-F consists of a parent module based on the CHIP-S protocol, and a child module that allows children with CHD to participate with a sibling or a friend. The child module uses cognitive behavioral therapy protocols in a relaxed and fun way to help children regulate their emotions, promote good self-esteem, develop problem solving skills, and establish positive thinking (van der Mheen et al., 2018). This program is currently under evaluation.

Health-related quality of life (HRQOL)

Since the concept of quality of life is a very broad concept often used in many contexts, health-related quality of life (HRQOL) has become a more used concept within health care. HRQOL refers to how people's lives are affected by a disease and its treatment (Sand, 2015). HRQOL instruments have become important measurements in clinical practice. The health component of these instruments indicates quality of life related to health (Theunissen et al., 1998). For the World Health Organization (WHO), health involves components of physical as well as psychological and social functions (WHO, 1948).

Most parents of children with CHD report satisfaction with care and with the support received by their families. However, parents caring for children with severe CHD report feeling isolated and having lower possibilities for self-development, implying decreased quality of life (Goldbeck & Melches, 2006). Parents of children with severe CHD experience higher levels of stress related to the amount of surgical procedures their child have to experience and in many cases uncertainty of the prognosis. Furthermore, some parents experience financial burden since the possibility to hold a full-time job is affected (Connor, Kline, Mott, Harris, & Jenkins, 2010).

Quality of life in children is described as children's ability to function in different contexts, such as family, school, and peer groups, while deriving personal satisfaction (Marino, Cassedy, Drotar, & Wray, 2016). HRQOL in children with CHD has been reported to be lower than in healthy children in areas such as motor functioning and autonomy as well as in their global positive emotional functioning (Krol et al., 2003)

Although patient self-reports are the standard method for measuring HRQOL, it is often the case in pediatric care that the child is too young, too sick, or too cognitively impaired to independently complete a HRQOL questionnaire. In these cases, proxy-reports are provided by the parents. Studies looking at the parent-proxy reports of their children's HRQOL have demonstrated good feasibility, reliability, and validity for children aged 2 to 16 (Theunissen et al., 1998; Varni, Limbers, & Burwinkle, 2007).

Unlike in health care, HRQOL terms and instruments are rarely used in psychology. In Study II, we attempted to understand if the subjective self and proxy ratings of cognitive problems of children and parents using HRQOL instruments were related to objective assessments of intelligence measured by psychological test instruments such as the Wechsler Scales of Intelligence. A study by Miatton (2008) looked at parental views of cognitive skills of their child with CHD and compared these results to cognitive measures finding accuracy and usefulness of the parental questionnaire on cognitive functioning of their child (Miatton, De Wolf, Francois, Thiery, & Vingerhoets, 2008). Whereas Miatton (2008) looked at children from 6 to 12 years old, we broaden the search by looking at children with CHD from 3 to 15 years old.

Socioeconomic status of the family

Many studies have reported on the relationship between socioeconomic status and parental factors influencing outcomes in children with CHD (Bellinger et al., 1995; Forbess, Visconti, Bellinger, Howe, & Jonas, 2002; Forbess, Visconti, Hancock-Friesen, et al., 2002; Wernovsky et al., 2000). Low income

environments have been associated with higher levels of stress in families, and in cases of continuous stress this negatively influences cognitive performance and health in children (Deary, Weiss, & Batty, 2010; Nisbett, Aronson, et al., 2012).

Low socioeconomic status of families (SES) is related to higher levels of stress in parents of children with CHD (Helfricht et al., 2008; Soulvie et al., 2012). Factors such as mother's education (Alton, Taghados, Joffe, Robertson, & Dinu, 2015), being a single parent (DeMaso, Campis, et al., 1991), and parental social support (Visconti et al., 2002) are also related to levels of stress in parents of children with CHD. Family environment and mothers' educational level have been found to have important repercussions in children's cognitive development in breastfeeding mothers, even more than the nutritional effect (Gibbs & Forste, 2014; Quigley et al., 2012). Parent's characteristics and parenting practices have important impacts on child development. Factors such as parents' sensitivity to the child – e.g., talking to children when they are small, reading to them (Gibbs & Forste, 2014), and making books and magazines available – are important factors as shared learning experiences with their children outside their home (Nisbett, Blair, et al., 2012).

Cumulative risk in families

When children are exposed to the effect of cumulative risk factors, consequences on their development are much more adverse than when confronted with a single risk factor (Gutman, Sameroff, & Robert, 2003; Sameroff, Seifer, Baldwin, & Baldwin, 1993). This might explain why socioeconomic factors such as poverty, stress, and low education have such strong impact on child development, burdening both children and their families. Cumulative risk theory has proven to be effective in predicting behavioral problems from early childhood to adolescence (Appleyard, Egeland, Van Dulmen, & Sroufe, 2004).

Cumulative risk is usually calculated by dichotomizing each risk factor exposure as 0 when the risk is not present or as 1 when the risk is present, and then adding the dichotomous score to reach a score of multiple risk or total risk index (Evans, Li, & Whipple, 2013). The cumulative effect of a severe CHD and social stress as the result of unemployment or financial burden has been shown to bring negative effects in the quality of life of families of children with CHD (Goldbeck & Melches, 2006).

Follow-up Program for Children with CHD in Sweden

A multidisciplinary follow-up and rehabilitation program for children with severe CHD was introduced at the Queen Silvia Children's Hospital in Gothenburg in the spring of 2008. The program aimed to reach a complete and multidisciplinary follow-up and rehabilitation for children and adolescents with complex heart defects from diagnosis to transfer to the adult health care (GUCH). The multidisciplinary team includes pediatric cardiologist, specialist cardiology nurse, physiotherapist, occupational therapist, nutritionist, speech pathologist, social worker, psychologist, and specialist dentist. A similar team was also formed in Lund, the other specialized center in pediatric cardiology in Sweden.

Although children with moderate and severe CHDs have been followed medically for many years, the follow-up rehabilitation program in Gothenburg and Lund includes only children with the most severe diagnoses, such as single ventricle, and only children living in the Gothenburg and Lund areas. Because many children with CHD live outside Gothenburg or Lund, they are followed-up medically at their local hospitals. Therefore, these children and their families are unable to take advantage of the multidisciplinary and individualized work available to children and families enrolled in the follow-up and rehabilitation program in the two specialized pediatric centers.

The need for national multidisciplinary follow-up and rehabilitation programs for children with CHDs was recognized by the Children's Heart Association in Sweden. During the autumn of 2016, a group of pediatric cardiologists, child neurologists, specialist nurse, psychologist, and a representative from the Children's Heart Association developed a draft with the aim to initiate work to create a national program for the monitoring and rehabilitation of children with severe CHD. The preliminary work in developing the guidelines for a national follow-up and rehabilitation program began in the autumn 2016 by reviewing both the American and the Norwegian guidelines. This review made it clear that children with CHD represent a group with increased risk for cognitive as well as motor development problems, learning difficulties such as delayed reading and writing development, attention and concentration difficulties, and poorer psychological health compared to control groups (Areias et al., 2014; Husler, Dubowy, Knobl, Meyer, & Schölmerich, 2007; Marino et al., 2012; Marino et al., 2016; Pike et al., 2012; Polito et al., 2015; Sarrechia, De Wolf, et al., 2015; Schaefer et al., 2013; von Rein et al., 2015).

The group addressed the need for a multi-professional team. Therefore, at the beginning of February 2017, a working group consisting of diverse professions from pediatric care units within the country was invited by

the Children's Heart Association to generate the guidelines for a national follow-up and rehabilitation program for children with severe CHD in Sweden. This recent development means that most of the children in the present thesis have not been part of a comprehensive multi-professional follow-up rehabilitation program.

The proposed program suggests a structure for screening assessment and intervention regarding several psychological aspects such as cognitive/intellectual, emotional, and behavioral development. Many studies have suggested that regular developmental screening for children with CHD needs to be taken seriously. While pediatric nurse practitioners play an important role in educating parents about the potential developmental risks to children with CHD, a team of different experts working with these children and their families is needed to address questions about parental stress, family functioning, and behavioral expectations for the child, all this in the context of routine medical/cardiac follow-ups (Brosig et al., 2007; Soto et al., 2011; Soulvie et al., 2012). The follow-up and rehabilitation program have the potential to form an important contribution to children with CHD and their families, addressing possible risks in a timely manner and with the right professionals.

CHD is a serious medical condition; however, the long-term impact on the child's development and everyday life varies greatly. One of the focuses of the present thesis is intellectual functioning in children with CHD. This thesis addresses some of the factors that can help our understanding of the large range of individual variations. In the scientific literature on neurodevelopment of children with CHD, many terms and functions are used to describe children's intellectual functioning. In the following section, the different terms and concepts will be discussed and compared.

Definition of Terms and Concepts to Describe Intellectual Functioning

This section aims to address the confusion found in the literature, which often uses different terms indiscriminately (Karsdorp, Everaerd, Kindt, & M., 2007; Khalil et al., 2014), making it hard to draw conclusions from studies of children with CHD. In the present thesis, I have chosen to use the term intellectual functioning (IF) since IF is what psychologists first assess when questions on school maturity arise (i.e., the child's intellectual resources, ability to reason, how the child plans, thinks, and communicates, and how the child interacts with others and resolves problems). When children are tested, many factors are measured and observed, among others the child's ability to cooperate in the test situation, curiosity, difficulties in understanding instructions,

distractibility, and fine motoric ability. All these functions are fundamental in being able to function in school and often easy to recognize in the test situation.

Studies often use the term neurodevelopmental disorders in evaluating children's intellectual performance. The problem with the term neurodevelopmental disorders is that it involves conditions detected at early development and includes the concept of developmental delays (i.e., slow development in cognition or in physical skills). If children do not catch up in their development even with changed circumstances, the developmental delay will most probably lead to a diagnosis of intellectual or developmental disability (Marino et al., 2012). Since most studies using the neurodevelopmental term are cross-sectional, they do not reflect the child's problem during the course of development. Some of the children reported in these studies might show neurodevelopmental delays at early ages due to the impact of CHD, surgery, and other perioperative factors. However, these children might recover after treatment, later showing normal intellectual functioning. Other studies on neurodevelopmental outcomes in children with CHD aim to measure only specific cognitive functions such as visuospatial skills, executive functioning, working memory, and processing speed. These studies often yield confusing results due to small sample sizes gathered at specific or wide range of ages and restricted to one type or a few types of diagnosis.

Neurodevelopmental disorders

Severe developmental and intellectual difficulties are diagnosed according to the Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5). In the DSM-5, the American Psychiatric Association defines neurodevelopmental disorders as a set of conditions with start at early development, often before the child enters school; these developmental deficits range from specific limitations in learning and executive functions to global deficits in social skills or intelligence.

Neurodevelopmental disorders often co-occur. For example, many children with attention-deficit/hyperactivity disorder (ADHD) also have specific learning difficulties. For some disorders, the clinical presentation includes symptoms of excess (e.g., excessive or repetitive behavior) as well as deficits (e.g., restricted interests or insistence on restricting similarity), resulting in developmental delays.

Intellectual disability and adaptive functioning

According to DSM-5, the term Intellectual Disability (ID) describes individuals having deficits in general mental abilities such as reasoning, problem solving, planning, abstract thinking, academic learning, and learning from experiences. General mental abilities are typically measured with individually administered psychometric tests that are culturally adapted and that are assessed by a clinical psychologist. Most tests have a population mean of 100 with SDs of 15 (100 ± 15). Individuals with ID will present scores of approximately two standard deviations (SD) below the population mean on these tests.

A diagnosis of ID implies that the intellectual deficits should cause impairments in adaptive functioning, limiting individuals' ability to live independently, to participate in social life, and to fulfil academic or occupational responsibilities. Impairment in daily adaptive function limits the individual in reaching age-related social standards in his/her surroundings and most individuals with ID require support and special curricula such as special education.

Learning disability

Although affecting learning abilities, ID is not the same as a learning disability. Learning disabilities are restricted to specific types of learning – i.e., academic learning. Learning disability refers to difficulties with school performance in areas such as reading, writing, or mathematics. In contrast, IDs affect three different types of learning: academic learning, experiential learning, and social learning. Children with learning disabilities have difficulties with only one type of learning (i.e., academic learning), whereas children with IDs have trouble with all three types of learning (Reynolds, Zupanick, & Dombeck, 2013).

Intellectual functioning and intelligence

Although determined by many factors, IF primarily refers to intelligence – the ability to reason, plan, think, and communicate. These abilities allow individuals to solve problems, to learn, and to use good judgment. One measure of intelligence is the Intelligence Quotient (IQ). IQ tests are designed in such a way that 97.5% of the population would score at or above 70. If an individual has an IQ score below 70, it is likely that the individual meets the criteria for ID and will require help such as compulsory special education.

Since the last century, psychologists have developed tests that aim to measure the trait called intelligence. Tests were constructed to measure the ratio of mental age to chronological age multiplied by 100. The general intellectual ability reflected in the full-scale scores of major tests of intellectual ability or IQ was created with the view that cognitive ability is a multidimensional trait. Many of the different abilities tested in IQ tests were positively correlated to each other, which many interpret as reflecting an underlying general cognitive ability or *g*, which is measured by the full-scale scores on the major tests of intellectual ability or IQ. Therefore, IQ tests have generally been viewed as a measure of intellectual ability (Dickens, 2008).

The term intelligence involves cognitive abilities evaluated by intelligence tests. It is sometimes used synonymously with cognition, which refers to the process by which individuals acquire knowledge from the environment. During cognitive processing, various mental processes are used such as perception, memory, abstract thinking, reasoning, and problem solving as well as integrative and controlling functions of these abilities, the executive function, which integrates planning and strategic thinking (Sparrow & Davis, 2000).

Intelligence tests are frequently used as part of an evaluation battery to assess development, to make a clinical diagnosis, or to further investigate the presence of developmental disabilities. Test results often contribute to decisions regarding entitlement for early interventions, special education, or decisions related to the compulsory education system (Williams, Sando, & Soles, 2014).

Testing intelligence in children with CHD

Most studies show that the results of intellectual testing in children with CHD are within ± 1 SD of the mean value; however, in tests involving motor skills results often end up between -1 to -2 SD from the expected mean (Snookes et al., 2010). These reduced motor abilities resulting from CHD and post-operative recovery might negatively influence the normal development of motor and spatial skill, which in turn affects performance on overall intellectual functioning in these children (Karsdorp et al., 2007).

Most psychologists believe that intelligence is multidimensional and that intelligence can be influenced by social factors such as poverty, emotional disturbance, brain dysfunction, and genetic or congenital physical illnesses and disabilities (Lezak, 1995). This might partly explain why children with CHD, who as a group perform at population mean levels, show an overrepresentation of learning difficulties. In a study of 117 children with CHD

tested with The Wechsler Intelligence Scale for Children, the scores were in the average range, and only 13% had a total IQ scores below 85 and 2% below 70 (Spijkerboer, Utens, Borgers, Verhulst, & Helbing, 2008). However, it was reported that as many as 33% of the group had repeated one or several school years and 14% received special education services. The same study considered school-related behavioral outcomes in relation to IF and found that teachers reported significantly more somatic complaints, such as being tired, compared to normative groups among children with CHD, and that children with poorer IF were reported as having more behavioral and emotional problems (Spijkerboer et al., 2008).

Limitations in adaptive ability decreases the individual's potential to become independent, participate in social life, and satisfy academic or occupational responsibilities. If CHD indeed affects adaptive ability, children with CHD will need help in at least one domain of adaptive functioning (e.g., abstract thinking, social behaviors, and practical behaviors), as deficiencies in these areas will probably limit their ability to perform in social settings or at school.

In children with CHD, poor academic performance has been associated with severity of diagnosis (Karsdorp et al., 2007). The more severe the heart diagnosis, the higher the risk for lower IF (Ortinou, Inder, Lamberth, et al., 2012). A large study (1770 children with CHD born between 1996 and 2009 and evaluated with the Bayley Scales of Infant Development) found lower intellectual functioning and poorer motor skills compared to the general population. Furthermore, children with the most severe heart disease had lower functioning than children with milder forms of CHD (Gaynor et al., 2015).

Psychomotor functioning in children with CHD

Studies on children with CHD with different levels of severity of cardiac diagnosis often address the common denominator of low results in fine and gross motor coordination areas, which alters the psychomotor development of the child (Porcayo-Mercado, Pliego-Riviero, Aguarre-Pérez, & Rucuardo-Garcell, 2013) and probably decreases chances of performing well on intellectual assessments. A study conducted by the Norwegian Institute of Public Health examined the occurrence of developmental impairments in three-year-old children with various levels of CHD, aiming to identify predictors associated with developmental impairments. They reported that children with CHD had three times higher probability to present gross motor and communication impairments in comparison to controls (Brandlistuen et al., 2011). Although children with isolated CHD often presented normal intellectual scores, they

showed more motor and language difficulties, with motor difficulties being the most frequently found morbidity (Hovels-Gurich et al., 2006; Miatton, De Wolf, François, Thiery, & Vingerhoets, 2007).

Other studies on cognitive performance in school-aged children with CHD report large deficits in sensorimotor functioning and language, implying that the children had reduced skills when asked to imitate hand and finger positions, suggesting that their ability to perform motor tasks is compromised by poor hand-eye coordination and lower levels of accuracy in fine visomotor skills, often resulting in impulsive strategies compared with healthy controls (Miatton, De Wolf, Francois, Thiery, & Vingerhoets, 2007). These results make us wonder why difficulties at school are common while the great majority perform within average norms on IF. A study on academic proficiency in children with CHD reported that these children often show lower educational achievements and that the rate of special education was almost threefold greater than the general school population (Mulkey et al., 2014). But are there other factors in addition to severity of CHD related to intellectual functioning in children with CHD? What are the factors related to IF in children in general and are these important for understanding individual differences in children with CHD? To find answers to these questions, I will turn to developmental psychology and theories specific to the development of cognition as well as theories of overall child development.

Cognitive and Developmental Theories: Risk and Protective Factors in Children with CHD

Developmental theories deal with how people grow and mature. Most theories within this discipline focus on understanding development occurring during childhood as this is the period during which most noticeable, and probably most important, changes occur in children's lives. Developmental theories describe change over time in one or more areas of behavior or psychological activity such as reasoning, awareness, or social behavior (Miller, 2002). Most parents expect that their children will reach certain abilities at certain ages, so-called developmental milestones. The most common milestones are when a baby begins to crawl, sit, walk, jump, build a tower with blocks, and hold a pen to draw. Examples of milestones in the social and emotional domains are the child responding to his/her name, smiling, and responding to interaction with others, distinguishing between friends and strangers, expressing different emotions such as irritation for a lost toy, imitating simple actions, and showing anxiety of separation from a prime caregiver. Within the cognitive domain, it is expected that the child will learn new things such as recognizing familiar

faces, responding to familiar sounds, trying to imitate facial expressions, understanding and responding to words, and developing concepts (McLeod, 2012).

Parents are often aware of their child's accomplishments in comparison to other children. Parents are usually the first detectors of their child's intellectual and cognitive development and have been found to be accurate in their concerns about delays in their child's development. Miatton et al. investigated the association between parental proxy-reports and estimated IQ (using a short version of the Wechsler Scales of Intelligence) for children between 6 and 12 years old with CHD: the more cognitive problems the parents reported, the lower the children's IQ (Miatton et al., 2008).

Early cognitive developmental theories

In 1936, Jan Piaget developed one of the first recognized theories of cognitive development in children. Piaget viewed intelligence as an active process where the child interacted with his/her surrounding to understand and to adapt to the world. For Piaget, cognitive development was reached by a progressive reorganization of different mental processes resulting from biological maturation and environmental experience. Piaget developed some basic concepts that are still important in understanding IF in children. He argued that children were not less intelligent than adults but they simply thought differently because of their limited experience (Jerlang et al., 2006).

Lev Vygotskij (1934), another influential theorist on cognitive development, highlighted the fundamental role of social interaction in the development of intelligence. Whereas Piaget believed that development was necessary to prepare for learning, Vygotskij argued that it was learning that facilitated development, i.e., learning preceded development. For Vygotskij, social development was fundamental in early cognitive abilities such as language and social communication, which he believed constituted the foundations for learning (Jerlang et al., 2006; McLeod, 2007).

In 1948, Edward Tolman questioned behaviorists in their assumption that people and animals were passive absorbers of information. He argued that humans had an active role in learning, developing a cognitive view of learning that has become more accepted in modern psychology. Tolman began using the term cognitive map, defining it as an internal representation of external environmental features. He argued that individuals acquire large number of cues or signals from the environment every day that help them build mental images called cognitive maps (McLeod, 2013).

These early theories are commendable since they have succeeded in emphasizing the importance of the connections established between individuals and their surroundings, including relationships between parents and their child, for healthy intellectual development (Sameroff & Mackenzie, 2003). All these early theories argue that IF or cognition, our understanding of the world, and problem-solving abilities in general depend on experiences with others. But what about genes? Is cognition or IQ not determined to a great extent from conception?

Nature vs. nurture

It is well known that people's lives are shaped by contributions and interactions between both nature and nurture. Nature refers to the process of biological maturation and inheritance, while nurture refers to the impact of the environment and learning through experiences (McLeod, 2012). Diverse aspects such as physical, social, emotional, and cognitive development influence each other during this process. Developmental theories help us understand not only normal child development but also processes of delayed development in one or several domains.

Nature. The development of the central nervous system and the cardiovascular system occur simultaneously in early gestation; therefore, the high incidence of structural abnormalities in the brain in children with CHD is not surprising. Genetic factors that affect both systems will influence each other: CHD may alter cerebral flow and oxygen delivery, resulting in secondary effects of the emerging fetal nervous system (Wernovsky, 2006). Genetic factors such as genetic comorbidities are associated with neurodevelopmental problems in children with CHD. Children with CHD and genetic comorbidity constitute around one-third of all newborns with CHD. Although there is significant variability in outcome (e.g., intelligence functioning or full scale IQ [FSIQ]) within specific genetic disorders, the FSIQ tend to be significantly lower in children with comorbid conditions compared to children with CHD alone (Gaynor et al., 2015; Latal, 2016; G. Wernovsky, 2006). In the studies included in the present thesis, children with CHD and genetic comorbidity were excluded.

Nurture. Pre-operative factors such as hospitalization required after birth, mechanical intubation or ventilation, and invasive interventions such as balloon atrial septostomy carry risks for the central nervous system. Often these patients have subnormal oxygen saturation, potentially compromising the delivery of the oxygen to the brain (Wernovsky, 2006). Neonates with critical congenital cardiac disease were shown to suffer from diminished blood flow and hence decreased oxygen to the brain (Licht et al., 2004).

Intraoperative factors and their impact on children undergoing surgery for CHD have been widely researched in relation to neurodevelopmental outcomes. Studies looking at the relationship between intraoperative variables related to vital organ support (such as safe conduct of CPB, i.e., deep hypothermic circulatory arrest (DCHA) versus low-flow bypass, regional cerebral perfusion, pH management, hematocrit, and degree of inflammation) and neurodevelopmental consequences have reported no improved neurodevelopmental outcomes with procedural modifications (Hirsch et al., 2012). No relationship has been found between vital support strategies and results on Psychomotor Development (PDI) and Mental Development (MDI) scores of the Bayley scales of Infant Development-II or neurodevelopmental outcomes measured by neuropsychological testing (Bellinger et al., 2011; Newburger et al., 2012).

Post-operative factors often imply invasive monitoring, mechanical ventilation, and risk for complications such as seizures. Seizures are a marker for early central nervous system injury and have been reported to be associated with lower scores on developmental tests in children (Bellinger et al., 1995; Bellinger et al., 1999; Bellinger, Rappaport, Wypij, Wernovsky, & Newburger, 1997).

Socioeconomic factors related to neurodevelopmental outcomes in children with CHD reflect the interwinding of environmental and genetic factors (Wernovsky, 2006). However, new findings and theoretical developments in the field of intelligence agree that the influence of genes is not as determinant as previously thought (Nisbett, Blair, et al., 2012). The shared environment (i.e., the environment shared by siblings in the same family) account for almost all variation on IQ in families at the lowest socioeconomic levels (Turkheimer, Haley, Waldron, D'Onofrio, & Gottesman, 2003).

Are biological or genetic factors more important? The definitive answer is that both are important especially if a child is exposed to cumulative risk factors. When biological risk factors in the child (e.g., a congenital heart disease, prematurity, and/or disability) co-occur with social hardship, then the probabilities for difficulties in social and cognitive development increase (Guralnick, 2013). What could be the specific risk and protective factors for intellectual functioning in children with CHD?

Contemporary models of development

Children with CHD are at risk for presenting neurodevelopmental delays and disabilities (Gaynor et al., 2015; Marino et al., 2012; Snookes et al., 2010). However, the range of the problems can vary widely. Today, no single congenital cardiac diagnosis or a specific treatment can predict the consequences

of CHD (Mussatto et al., 2015). Two contemporary models of child's development could be useful in understanding development in children with CHD: Sameroff's transactional model (Sameroff & Mackenzie, 2003) and Guralnick's developmental systems model (DSA) (Guralnick, 2013). Both these models recognize the importance of consistent trade-offs between the child, the parents/caregivers, the environment, and the influences between these factors on developmental growth. They also address the biological aspects, especially the genetic ones, constantly interacting with environmental events influencing development in children (Guralnick, 2011).

In the transactional model, the development of children is a product of continuous dynamic interactions between them and the experiences provided by their family within the social context they live in. The transactional model emphasizes the bidirectional effects of the children and the environment promoting cognitive as well as socio-emotional development (Sameroff & Mackenzie, 2003). In the developmental systems model, the parent-child transactions are also seen as a core feature. Parent-child transactions have proven to be one of the most important factors influencing children's development and are necessary for promoting optimal child development (Guralnick, 2012).

In an attempt to combine the psychological theoretical context, I will use Guralnick's developmental systems approach (DSA) as a way to understand the development in children with CHD and the important role of their parents and families in this process.

OBS! INTRODUCE FIG 1 THE DEVELOPMENTAL SYSTEMS APPROACH (ADAPTED FRÅN GURALNICK, 2011)

The developmental systems approach (DSA)

Child social and cognitive competence

DSA (Figure 1) aims to address the mechanisms that are fundamental in promoting development in children and was originally intended to address issues related to children being at risk for developmental delays and disabilities (Guralnick, 2012). In their process to accomplish and demonstrate their social and cognitive competence, children rely on developmental resources (domains of cognition, motor, language, socio-emotional, and sensory-perceptual) as well as on organizational processes (executive function, motivation, metacognition, emotion regulation). Children's social and cognitive competence depends on their own social and cognitive abilities and developmental capacities, which in cases of pre-term children or children with severe congenital diseases are at risk of having insufficient developmental resources. A meta-analysis reviewing studies on behavior problems and cognitive functioning in children with CHD (Karsdorp et al., 2007) reported that children with more severe diagnoses were at higher risk for impaired cognitive functioning, specifically with respect to intellectual functioning, compared with children with less severe diagnoses. More severe cognitive impairments were attributed to an accumulation of risk factors (Karsdorp et al., 2007).

For child factors such as prematurity, lower birth weight (<2.5 kg), longer length of hospitalization, abnormal brain maturation, and abnormal cardiac anatomy, overall morbidity in the first year of life is associated with low PDI (Psychomotor index) and MDI (Mental Development index) scores of the Bayley scales of Infant Development-II (Gaynor et al., 2015; Marino, 2013; Newburger et al., 2012). Failure to thrive is common in infants with complex heart defects such as hypoplastic left heart syndrome, and children with poor growth may be at risk for worse surgical and neurodevelopmental outcomes (Slicker et al., 2013). Limited feeding capabilities of the child and the need for supplemental tube feeding have shown negative association with developmental outcomes in children (Mussatto et al., 2014; Mussatto et al., 2015). Therefore, the ability of the child to take full oral feedings has been positively related to cognitive as well as language and motor skills in children with CHD (Mussatto et al., 2014; Werner et al., 2014). These risk factors interact with protective factors and shape the child's capacity to develop social and cognitive abilities. For preterm children and probably also for children born with a cardiac disease, the risk factors constitute an impact sufficient

enough to reduce children's global levels of social and cognitive capacity in relation to full term and healthy babies (Stephens & Vohr, 2009).

Developmental resources

Developmental resources are those abilities, skills, and knowledge organized in domains of cognition, language, motor, social-emotional, and sensory-perceptual development in the child. These areas are often developed by goal-oriented activities meant to increase children's competence. An example of this is the growth of vocabulary during childhood. A growing vocabulary will increase children's syntactic competence and their ability to understand the intentions of others. This example shows how developmental resources are clearly interdependent and are combined and re-combined between and within domains becoming an intrinsic part of a system supporting children's social and cognitive competence (Guralnick, 2011).

A review study of neurocognitive consequences of surgery in children with CHD found that although intelligence scores typically fall within the normal range, there was a high prevalence of deficits in language as well as gross and fine motor functions (Miatton, De Wolf, François, Thiery, & Vingerhoets, 2006). This result suggests these children have difficulties integrating more complex forms of social and cognitive competence.

Organizational processes

Organizational processes relate to those mental processes organized to interact with each other in such a way that allow the child to function at different cognitive levels, e.g., within goal directed activities. Guralnick (2011) identified several high cognitive processes as organizational processes: executive functioning, metacognition, social cognition, motivation, and emotion. These processes are interrelated and require regulation and contribute to children's development and competence.

However, to meet cognitive and social-emotional challenges (e.g., teaching babies to appropriately modulate their emotions), the caregiver must adapt to the child's developmental level and capabilities. Therefore, in cases of ill or pre-term babies, where the child's interaction and response to parent's are different, difficult, or sometimes negative, it becomes more difficult for parents and caregivers to understand and match their infant's needs and effectively promoting their development (Sameroff & Mackenzie, 2003). According to Guralnick (2012), families with children with medical conditions or

preterm babies will need to adjust to these conditions, increasing the risk for strained family patterns of interaction.

Family patterns of interaction

Family patterns of interaction include the adjustments that families often do in their environment to adapt to their child's unique needs with the focus on supporting their child's development. This component requires parental attention to the child's developmental initiatives and involves parents' willingness to adjust to the child's specific changing developmental patterns, supporting their child's development in the best way possible. Family patterns of interaction can take the following forms.

Parent child transactions consisting of a series of "relationship processes". These processes slowly develop over time through repeated sequences of positive and matched parent-child exchanges and involves sensitive-responsiveness, affection, and engagement of parents towards their child. According to Guralnick (2012), parent-child transactions during the first three years of the child's life are the most influential factors for child development. Parents need to be accessible to understand how children use their developmental resources and organizational processes to work towards social and cognitive competence (Guralnick, 2011).

Parents of children at risk more often develop poor parent-child transactions compared to parents with minimal risk. Pre-term children (or children with congenital diseases that often require early surgery and intensive care) may be more likely to present characteristics that constitute stressors and affect components of the family pattern of interactions, especially parent-child transactions (Guralnick, 2012). A study of 246 children with CHD ages 8 to 18 and their parents found that children and adolescents with greater severity of the heart disease from low socioeconomic status (SES) families were at greater risk for cognitive problems. The study reported that parents of children and adolescents from lower socioeconomic families reported greater cognitive problems in their children (Limbers, Emery, & Uzark, 2013). Other studies have reported similar association of SES and Intellectual functioning in children with CHD after corrective surgery (Forbess, Visconti, Hancock-Friesen, et al., 2002).

Family orchestrated child experiences include parents' ability to create or organize experiences that generate a stimulating environment for their child. This includes quality time with the child, participating together in activities in the community and in learning activities. Families with children at risk have lower exposure to these activities. Environmental factors such as

visits to hospitals or interventions such as surgery, medication, or rehabilitation can influence one or more components in family members and their resources, leaving no time or economical means for some families for planned activities, negatively affecting family's patterns of interactions (Guralnick, 2012).

Psychosocial and environmental factors influence intellectual outcomes in children with CHD (Latal, 2016). Children with CHD often present gross motor problems and problems with coordination and slower processing speed, limiting their ability to participate in some leisure activities (Latal, 2016). As a result, children already at biological risk may also experience less optimal family environments that increase the risk of stressful interactions and further decrease of optimal developmental.

Health and safety provided by the family include the parents' responsibility to provide a healthy and safe environment for their children. Factors such as making sure children have good nutrition, preventive care such as vaccinations and routine controls, as well as protecting children from unsafe neighborhood and domestic violence are included in this area. This area becomes complicated for parents of sick children. Clinical experience shows that many parents suffer from guilt concerning failure to protect their child from being ill.

Failure to thrive is common in infants with complex heart defects such as hypoplastic left heart syndrome. Children with poor growth may be at risk for worse surgical and neurodevelopmental outcomes (Slicker et al., 2013). Knowledge and awareness of the child's heart disease has been addressed as a protective factor contributing to better health outcomes of children with CHD (Daily et al., 2015). Families that engage in understanding the child's medical condition might also feel more capable of making informed decisions and protecting their child.

Family resources

All families have resources that contribute to child development and these enable parents to adjust to their child's characteristics and the daily stressors involved in a family. Family resources include both individual characteristics of the parents and the material resources in the family.

Individual characteristics of parents

Individual characteristics of the parents include their physical and mental health as well as their intellectual ability. Notably, these characteristics include

parents' attitudes towards child rearing and how they address their children's specific needs (Guralnick, 2011).

Parental factors such as parental anxiety, maternal post-traumatic stress, and overprotection are some of the factors often revealed as significantly contributing to behavioral problems in children with CHD. Maternal stress influences development of the child during fetal and early postnatal life which in turn influences intellectual functioning in children with CHD (Grizenko, Fortier, Gaudreau-Simard, Jolicoeur, & Joobar, 2015; Nilsson, Nilsson, Ostergren, & Rasmussen, 2004). Parents' perceptions of their children's level of illness have been shown to correlate with objective measures of postoperative morbidity (Franck, McQuillan, Wray, Grocott, & Goldman, 2010) such as learning problems and limited intellectual functioning. Family factors such as parenting style and stressors such as strained economy and limited social networks can further moderate intellectual functioning and school outcomes in children with CHD (Gaynor et al., 2015; Gaynor et al., 2007; McCusker et al., 2007; McCusker et al., 2010; Newburger et al., 2012).

A study examining mental health and coping styles in both mothers and fathers of infants born with a severe congenital heart defect assessed parents of 70 infants born with severe congenital heart defect. Questionnaires examined psychological functioning and coping strategies of the parents. Results showed higher levels of clinical psychological distress in mothers compared to fathers of children with CHD and differences between parents in coping styles. Moreover, the extent of distress in both parents was not predicted by the severity of the cardiac disease or demographic factors, but by coping styles, knowledge, subjective worry, and family functioning, results that demonstrate the importance of these variables (Doherty et al., 2009).

However, there are also factors associated with positive adaptation, resilience factors. Resilience refers to positive outcomes despite experiencing adverse situations. Resilience in parents is often associated with the way they respond to demanding or stressful situations, including the cognitive ability to assess difficult circumstance and to respond with mobilizing skills towards better understanding and positive problem-solving (Olsson, 2008). Mother's education has shown to be significantly related to better functional outcomes in children with CHD. Higher maternal education was significantly associated with better functional abilities in a study of 4.5-year-old children who had undergone cardiac surgery (Alton et al., 2015). Maternal perceptions of their child's disease also play an important role in the child's adaptation to the disease. In one study, maternal perceptions were potent predictors of emotional adjustment, with approximately 33% of the variability in adjustment accounted for by maternal perceptions, while medical severity accounted for less than 3% of the variability. That is, severity of illness appears less critical to

successful adaptation than the quality of the mother-child relationship (De-Maso et al., 1991).

Material resources

Material resources include not only economical means but also social support. Social support can take many forms such as extended family, support networks, advice on rearing children, and babysitting possibilities.

Childhood illnesses and disabilities occurring in families from lower socioeconomic status and with social problems may further impact parent-child transactions negatively due to high care-taking demands and lost time at work, a situation that may result in higher parental stress, symptoms of depression or anxiety, relational problems, and less sensitive parenting, in addition to the vulnerability as the result of the child's medical condition (Guralnick, 2013). Medical conditions often include symptoms such as pain, feeding difficulties, difficulties with medication, as well as the child being connected to monitors and intravenous devices that make it difficult for parents to comfort their child.

The relation between SES and intellectual functioning is well investigated in the normal population (Camargo-Figuera, Barros, Santos, Matijasevich, & Barros, 2014; Erdem, van Lenthe, Prins, Voorham, & Burdorf, 2016; Hart et al., 2005; Julvez et al., 2014; Olsson & Hwang, 2003) as well as in children with CHD (Bergvall & Cnattingius, 2008; Gaynor et al., 2015; Limbers et al., 2013). Intellectual outcomes in children with CHD are often shown to be moderated by maternal education (Newburger et al., 2012), comorbidity, and SES (Forbess, Visconti, Hancock-Friesen, et al., 2002; Spijkerboer et al., 2008).

Protective factors include higher SES, which may be related to the ability to act on health guidance and treatment proposals (Schaefer et al., 2013). Parents' willingness and ability to learn, get involved, and to be with their child during the treatment process by cooperating with physicians and nurses increase the children's sense of security and better understanding of the medical procedure they experience. Having a child admitted for surgery at the cardiac care unit surrounded by nurses and specialist doctors gives many parents a sense of worthlessness. For the child, however, what really matters is that parents are with them, supporting them, and motivating them to go through the process.

Clinical experience shows that parents who stimulate their children in creative ways (e.g., reading interesting stories, singing to them, borrowing books at the library that describe with pictures and age appropriate

language the procedures that the child goes through, joking, and laughing) increase the likelihood their children will experience better intellectual development. Parents can actively interact with their children through mobilizing family resources, e.g., inviting relatives and friends, playing games, and talking to other patients within the ward. When parents are unable to invite the child's friends to visit because of high risk of infections, many organize on-line visits through Skype. Sometimes, teachers visit the child with cards and pictures from their classmates. All these interactions allow the child to use his/hers social and cognitive resources to maintain intellectual development. This is shown in our studies where children with severe CHD and high SES were less impacted in their intellectual functioning as children with severe or less severe CHD but low SES. This result is probably because families with high SES had better resources in terms of coping strategies, confidence, and competence to be active in introducing family interactions that allowed them to continue having positive parent-child transactions even in the hospital setting.

Stressors threatening parent-child transactions

The DSA model considers that particular characteristics of the child (e.g., the presence of illness) will increase the risk for stressors, placing parent-child transactions at risk. Stressors in the form of child's characteristics create increasingly adverse effects over time on family patterns of interaction and in the child's cognitive and social competence (Guralnick, 2011, 2012). Parents with a child with CHD requiring surgery as a newborn might experience difficulties adjusting to and understanding the child's signals and needs. These difficulties may in turn make parents hold back and feel insecure in interaction with their child and consequently increase the risk for delaying the development of a secure relationship and high quality parent-child transactions (Goldberg, Simmons, Newman, Campbell & Fowler, 1991; Guralnick, 2012).

Feeding difficulties and low weight gain are often factors addressed as sources of great stress for parents of children with CHD (Tregay et al., 2017). Children with CHD might show poor nutritional and inadequate feeding ability after having neonatal surgery (Medoff-Cooper, Naim, Torowicz, & Mott, 2010; Tregay et al., 2017). Feeding problems in children with CHD can result in significant differences in how infants with CHD interact with their mothers during feeding compared with healthy infants (Lobo, 1992). Feeding problems have been shown to be related to stress in mothers of young children with CHD since it interferes with attachment development and social interactions during infancy (Goldberg et al., 1990; Hill et al., 2014). Some studies have reported that mothers of infants with CHD show lower

levels of positive affect and engagement compared to mothers of non-CHD infants (Gardner et al., 1996). Feeding difficulties have a double component of complexity for parents: they create insecurity among parents regarding their ability to provide for and feed their child and they emphasize the uncertainty of the child's condition, since the child's poor weight gain increases the risk for negative outcomes and eventually failure to thrive (Rempel, Harrison, & Williamson, 2009; Rempel, Ravindran, Rogers, & Magill-Evans, 2013).

Stressors in families of ill children can be of such magnitude that they there is a risk parents will develop insensitive responses to their child. According to Guralnick (2012), anxiety and depression in parents can foster low-sensitive responses and perceptions of vulnerability of their child under long periods, interfering with parent-child transactions (Guralnick, 2012). These interactions, as a primary part of family patters of interactions, are fundamental to support children's' social and cognitive competence.

Summary of Studies

General and Specific Aims

General Aim

The purpose of the present doctoral thesis was to investigate intellectual functioning and the influence of the children with congenital heart defects in their families. We analyzed how the severity of the heart defect, the child's age, and the socio-economic status of the child's family were related to the intellectual functioning of the children and to the impact on the family. A scale from a screening tool, the PedsQL 3.0 Cardiac module, was also tested to identify children and adolescents at risk of intellectual problems. A psychological model for understanding the development of children was used in discussing how physical and psychosocial factors affect each other and how parents, healthcare professionals, and the child's environment influence children's development.

Study I

The aim of Study I was to investigate intellectual functioning in children with CHD treated with surgery or by catheter techniques at the Queen Silvia Children's Hospital of Gothenburg. Intellectual functioning was investigated in relation to severity of the heart diagnosis, the child's age, and the socioeconomic status of the families. The aims of the study were:

1. To examine if children with severe have lower intellectual functioning than children with mild and moderate CHD.
2. To investigate if older children with CHD have lower intellectual functioning than younger children with CHD.
3. To explore if children with CHD living in families with low SES have lower intellectual functioning than children in families with higher SES.
4. To analyze if there is an interaction effect of severity of diagnosis and SES, i.e. if children with low SES and severe CHD have lower intellectual functioning compared to children with high SES and severe CHD.

Study II

The aim of Study II was to evaluate whether the PedsQL Cognitive Problems subscale from the cardiac module could be used as a screening tool for identifying children with CHD who need to undergo more extensive cognitive assessment. The specific aim was:

1. To study the association between self- as well as proxy reports on the Cognitive Problems Scale (five items) from the PedsQL (Cardiac Module 3.0), for children and adolescents with CHD in four different age groups (3-, 5-, 9-, and 15 year- old), with their actual cognitive performance on standardized cognitive test, i.e., The Wechsler Scales of Intelligence (Swedish versions).

Study III

The aim of Study III was to investigate the impact reported by parents of children with CHD treated with surgery or by catheter interventions. Using the Family Impact Questionnaire, we investigated the differences in impact reported by fathers and mothers. Positive and negative subscales of the Family Impact questionnaire were analyzed in relation to the parents' gender, the child's age, and severity of the diagnosis, the socioeconomic status of the family and the intellectual functioning of the child. We also investigate the cumulative impact of these factors in parents reports. The specific aims were:

1. To investigate if mothers of children with CHD report more negative and less positive impacts than fathers.
2. To examine if parents of younger children with CHD report more negative and less positive impacts than parents of older children.
3. To explore if parents of children with severe CHD report more negative and less positive impacts than parents of children with mild CHD.
4. To investigate if parents of children with CHD in families with low SES report more negative and less positive impacts than parents in families with high SES.
5. To understand if parents of children with CHD and low intellectual functioning report more negative and less positive impacts than parents of children with CHD and high intellectual functioning.
6. To analyze whether parents of children with CHD who experience an accumulation of risk factors (low SES, low child age, more severe heart diagnosis, and low intellectual functioning), report higher levels of negative and less positive impacts than families with no or only one risk factor.

Methods

Study I

The sample for study I consisted of 228 children who completed testing with the Wechsler Scales of Intelligence. Of these, 111 (48.7%) were girls and 117 (51.3%) were boys. Participants were recruited over a seven-year period (2008-2015). Participants were families living in the Västra Götaland Region (VGR) with children treated with surgery or catheter interventions for CHD at Queen Silvia Children's Hospital in Gothenburg, Sweden. We excluded children with chromosomal defects and disabilities known to influence intellectual functioning (Fig. 2)

Children in the study were tested with the Swedish versions of the Wechsler Scales of Intelligence (Wechsler, 2005, 2007) to assess general intellectual functioning. The Wechsler Preschool and Primary Scale of Intelligence, Third Edition (WPPSI-III Swedish version) was used to test the 3- and 5-year-old children, and the Wechsler Intelligence Scale for Children, Fourth Edition (WISC-IV Swedish version) was used to test the 9- and 15-year-old children. Only core subtests were used to compute the Full-Scale IQ (FSIQ). Ages were chosen to match the follow-up program for children with CHD at Queen Silvia Children's Hospital in Gothenburg (Table 1).

The children had a wide range of cardiac diagnoses and were divided into three groups (Table 1). The configuration of the three groups was designed by pediatric cardiologist, Jan Sunnegårdh (Sunnegårdh, 2011), and is based on the child's severity of the cardiac diagnosis, and on the risk that the child had for further complications:

The mild group – children usually treated once with surgery or by catheter intervention with little risk for further complications and who were, in most cases, no longer followed-up (e.g., atrial septal defect (ASD), ventricular septal defect (VSD), persistent ductus arteriosus (PDA), coarctation of the aorta (CoA), and pulmonary stenosis (PS)).

The moderate group – children who had been treated with surgery and/or catheter intervention and were followed-up regularly since there were risks for further complications (e.g., transposition of the great arteries (TGA), tetralogy of Fallot (ToF), complete AV-defect (CAVSD), total anomalous pulmonary venous drainage (TAPVD), and aortic stenosis (AS)).

The severe group – children with complex heart defects for whom long-term prognosis was uncertain and serious complications were not uncommon (e.g., univentricular heart lesions, pulmonary atresia (PA) with VSD, major aortopulmonary collaterals (MAPCA), and heart transplantation (Htx)). These groups were further sub-divided according to age.

Table 1. Distribution of included children according to age and severity of the CHD (see also fig. 2. Study population).

Severity of CHD	3 years	5 years	9 years	15 years	N	%
Mild	19	17	19	18	73	32.0
Moderate	20	17	24	17	78	34.2
Severe	26	19	18	14	77	33.8
N (%)	65(28.5)	53(23.2)	61(26.8)	49(21.5)	228	100.0

The Hollingshead “Four Factor Index of Social Status” was used to determine socioeconomic status index (SES) for each family. This index weighs education, occupation, and employment status to determine a composite score of social status (Adams & Weakliem, 2011). Answers from 17 families were missing and for 154 families both parents answered the questionnaire. From our sample of 228 children, 366 parents (202 mothers and 164 fathers) answered the SES questions; the mean score of SES was 43.6 (SD = 12.5).

Analysis of variance (ANOVA) and post hoc tests, Bonferroni were performed to explore differences of intellectual functioning (FSIQ) of children with CHD in relation to age (3-, 5-, 9-, and 15-year-olds), the severity levels of their heart defect (mild, moderate, and severe) and SES (low, medium, and high).

Study II

The sample for Study II overlapped with sample 1 since it involved 184 children with CHD that had completed testing with the Wechsler Scales of

Intelligence and 316 parents (173 mothers, 141 fathers). Both children and their parents completed self-reports and proxy-ratings on the Cognitive Problems Subscale of PedsQL Cardiac Module 3.0. Of these, 96 (52.2%) were girls and 88 (47.8%) were boys.

In study II we used PedsQL Cardiac Module 3.0, subscale Cognitive Problems (five items) to measure perceived cognitive problems (Tahirovic, Begic, Tahirovic, & Varni, 2011). The scale includes a self-report version as well as a parental report-version. The questionnaire has been extensively validated and reliability tested internationally (Karen Uzark, Jones, Burwinkle, & Varni), including on a Swedish sample (Peter Sand, Kljajić, & Sunnegårdh, 2013). Self-reports were gathered from the 5-, 9-, or 15-year-olds, but not for 3-year-olds as they are too young for self-reports.

The distribution of children according to age was: 3-year-olds, $n = 56$ (30.4%); 5-year-olds, $n = 34$ (18.5%); 9-year-olds, $n = 53$ (28.8%); and 15-year-olds, $n = 41$ (22.3%). The distribution of children according to severity of the CHD was: mild group, $n = 61$ (33.2 %); moderate group, $n = 72$ (39.1 %); and severe group, $n = 51$ (27.7 %); and the mean score of SES was 44.0 ($SD = 12.3$).

Due to the variability of the 5- and 9-year-olds' reading skills, the questions were read out loud by the test leader for these two age groups. The 5-year-olds indicated their answer by pointing on a scale with different "smiley faces", and the 9-year-olds pointed out their answers using a five-point Likert scale ranging from 0 (never a problem) to 4 (almost always a problem). The 15-year-olds as well as the parents completed the form by reading it themselves and rating their answers on the five-point Likert scale. Both parents were asked to complete the form separately and were specifically told not to discuss their answers with each other. For 72% of the patients, we had the rating for both parents on the PedsQL Cognitive Problems subscale, so we used the mean value of these two values in our analyses. The intra-class correlation for the two parental measures was .86. For 22% of the patients, we only had the mother's ratings. For 6% of the patients, we only had the father's rating, so for these patients we just used this one value. When calculating the final scores, the items are reversed and linearly transformed according to following formula provided with the PedsQL: $0=100$, $1=75$, $2=50$, $3=25$, and $4=0$. Thus, the higher the score correlated with the less perceived cognitive problems.

ANOVAs were calculated to investigate differences between groups on the descriptive measures. The relevant variables all met the normality assumption, and homogeneity assumptions were also checked. When the homogeneity assumptions were not met, the Games-Howell post hoc test was used instead of the Bonferonni correction. Correlations were conducted for the different predictors and the outcome variable. Finally, multiple linear

regression analyses were conducted to investigate the unique contribution of the different predictors in the model. In the final regression analyses, SES, severity of diagnoses, self-ratings and proxy-ratings were entered as predictors with FSIQ as the dependent variable. Gender was not included since no association was found between this variable and the dependent variable for any of the age groups. Hierarchical regressions were computed since they allow for evaluation of variance accounted for by the different blocks of predictors.

Study III

The sample for Study III also overlapped with sample 1, since it involved parents of children that had been tested with Wechsler Scales of Intelligence. The size of study III consisted of 356 parents of 211 children who answered to The Family Impact Questionnaire (Donenberg & Baker, 1993). The Family Impact Questionnaire (FIQ) was answered by 158 fathers (44.4%) and 198 mothers (55.6%).

The FIQ is a 50-item questionnaire that evaluates parents' perceptions of the "child's impact on the family compared to the impact of other children same age have on their families". It has been used in studies measuring parental stress in families of preschoolers with developmental delays (Eisenhower, Baker, & Blacher, 2005). Parents respond to items on a four-point Likert-type scale going from *not at all*, to *very much*, rating the impact the child with CHD has in the family - from 0 (not at all) to 3 (very much) - compared with the impact other children the same age has on their families (Eisenhower et al., 2005).

In our study parents answered four sub-scales (36 items) of the six common subscales of the FIQ: Impact on social relationships (11 items), Negative feelings towards parenting (9 items), Positive feelings towards parenting (7 items) and Impact on siblings (9 items). Items on finances (7 items) and marriage (7 items) were not included. By combining Impact on social relationships with Negative feelings towards parenting a scale measuring Negative Impact (20 items) was created. We explored family impact of CHD by examining parents reports on two sub- scales: Negative Impact (20 items) that measure stress and feeling of frustration and anxiety in parents, and Positive impact (9 items) that evaluates parents' satisfaction and feelings of joy and pride (Fleck et al., 2015). Higher scores in the sub-scale Negative Impact indicate negative perceived impact or stress while in the Positive Impact sub- scale, higher values imply more positive perception of the child's impact on the family.

For study III, the distribution of children according to severity of the CHD was: mild group, n = 68 (32.2 %); moderate group, n = 76 (36 %); and severe group, n = 67 (31.8 %). The distribution of children according to age was as follow: 3-year-olds, n = 63 (29.4%); 5-year-olds, n = 50 (23.7%); 9-year-olds, n = 57 (27%); and 15-year-olds, n = 42 (19.9%). And from the sample of 356 parents of 211 children (158 fathers and 198 mothers) answered the SES questions; the means SES was 43.7 (SD = 12.5).

Analyses of variance (ANOVA) was used for group comparisons with eta square to analyze effect size interpreting and Bonferroni for post hoc testing. For between groups comparisons of FIQ we used answers of parents in the Negative and Positive impact scales in relation to their children's age (3-, 5-, 9-, and 15 years olds), the severity levels of children's heart defect (mild, moderate and severe), child's FSIQ (low, average and high) and SES of the families (low, medium and high). We used independent samples t-tests to compare mean differences in total scores between fathers' and mothers' reports on Negative impact and Positive impact scales. When less than 20 % of the answers in each scale was missing, we replaced missing values with the mean value of the individual scale's case. In our study of 353 parents participated (158 fathers and 198 mothers), one father and one mother had more than 20% of missing values on the Negative impact scale, in the Positive scale one father and two mothers had more than 20 % of missing values, therefore, these cases were excluded by SPSS.

OBS! FIG. 2 INTRODUCE FIG 2 IN PDF OF STUDY POPULATION

Main findings

Study I

For the 228 children in the study, the total mean score in the Full-Scale IQ (FSIQ) was 100.8 (SD = 14.5), i.e., within normal range. No significant differences were found between boys and girls.

When exploring intellectual functioning in relation to severity of CHD we found that children in the severe CHD group had significantly lower FSIQ than children in the mild CHD group and the moderate CHD group. More children with severe CHD had FSIQ below -1 SD (32.5%) than children with milder forms of CHD.

Investigating intellectual functioning in relation to age we found that FSIQ was significantly lower for 9-year-olds compared to the 3-year-olds). When analyzing intellectual functioning in relation to SES we found that children from families with low SES had significantly lower intellectual functioning compared with both the medium SES and the high SES group. The high SES group also had significantly higher FSIQ than the medium SES group. When children were divided into categorical IQ-groups (low, average, and high intellectual functioning), a higher proportion of children from families with low SES had FSIQ below -1 SD (34.1%) compared to children from families with average (14.7%) or high SES (2.9%).

We found no interaction effect between severity of diagnosis and SES for FSIQ. Using ANOVA to control for the effect of SES on FSIQ, we found that the effect size for severity of diagnosis on FSIQ decreased from .06 to .05 after controlling for SES, but the differences in FSIQ between the severity groups remained significant. Both SES and severity of CHD diagnosis had significant main effects on FSIQ. When children were divided into categorical IQ groups (low, average, and high intellectual functioning), a larger proportion of children both diagnosed with severe CHD and living in families with low SES performed in the low range of intellectual functioning than children diagnosed with severe CHD living in families with medium and high SES. Among children from low SES families and a severe CHD, 8 of 20 (40%) had low IQ (-1 or -2 SD), compared to those from medium SES families 13 of 43 (30%) and from high SES families 1 of 4 (25%).

Study II

There was a significant difference between the different age groups in the proxy reports. This effect was of a medium size and the Games-Howell post hoc comparison showed that the parents of the pre-school children reported significantly less cognitive problems than the parents of the school children.

When looking at correlations between the predictors and FSIQ were calculated we found out that for the 3-year-olds, there was a negative relation between severity of diagnosis and FSIQ, a result that indicated that the severity of diagnosis was associated with poorer intellectual functioning. No such relationship, however, was found for the older children. When it comes to the association between self-report and FSIQ, there was only a significant positive relationship for the 15-year-olds, indicating that the less perceived cognitive problems, the better their intellectual functioning. For the proxy-report, we found a positive correlation for all age groups, indicating that the less cognitive problems the parent's perceived, the higher the intellectual functioning of the children.

To test the unique contribution and to further investigate the variance accounted for by self- and proxy-reports on the dependent variable, hierarchical regression analyses were conducted separately for all age groups. SES predicted a significant amount of the variance in each of the four regression analyses (7%, 7%, 13%, and 20%, respectively). Severity of diagnosis, however, was only significantly associated with intellectual functioning for the 3-year-olds; children with a more severe diagnosis had a significantly lower FSIQ than children with a mild diagnosis.

When controlling for SES and severity of diagnosis, there still existed an association between the proxy-ratings and the FSIQ for the 3-year-olds, 9-year-olds, and the 15-year-olds. For the 3-year-olds, this association accounted for 13% of the variance and for the 9-year-olds it accounted for 22% of the variance. For the 15-year-olds, the self- and proxy-reports of Cognitive Problems explained 27% of the variance in the dependent variable.

To further investigate a way to use the Cognitive Problem subscale on PedsQL as a screening tool for detecting cognitive problems in children, a cut-off score was assessed. To prove a cut-off score that would be appropriate when using PedsQL Cognitive Problem subscale, the children were divided into two groups. Since children achieving a FSIQ score of below 85 very often experience learning difficulties and to have an inclusive cut-off score criteria, children scoring below 90 in FSIQ were compared to children achieving a score of 90 and above on the FSIQ. Because children having a score above 90 hardly experience learning difficulties, they should have a mean

value of 73.7 on the self-reports at the age of 15 with a FSIQ above 90. For the proxy-reports, these values range from 75.5 to 85.3 for the children with a FSIQ above 90, depending on age group. Thus, a possible inclusive cut-off score for both the 15-year-olds self-reports and the proxy reports for all age groups were proposed to be 80. If the score falls below 80 on the Cognitive Problem subscale, the child should be formally evaluated.

Study III

Looking at the Impacts Fathers and Mothers of Children with CHD reported we find out that on the Negative Impact Scale, mothers scored significantly higher than fathers, indicating higher levels of negative impact in mothers than in fathers. On the Positive Impact Scale, mothers' scores and the fathers' scores were similar.

The fathers' and mothers' answers revealed no relation between positive or negative impact and the age of the child. However, Severity of CHD was significantly associated with fathers' and mothers' reports on negative impact. No relation was found between SES of the family and fathers' reports of negative impact. But, a significant relation was found for mothers. Mothers in the Medium SES group reported significantly higher negative impact than mothers in the High SES group. For the Positive Impact Scale, significant relations were found for fathers but not for mothers. Fathers in the Low SES group reported significantly higher positive impact than fathers in the High SES group.

We found no relation between intellectual functioning of the child and negative impact in either fathers or mothers. However, both fathers and mothers reported a relation between intellectual functioning of the child and positive impact.

Both fathers and mothers in the High-Risk groups reported significantly higher negative impact than fathers and mothers in the Low Risk groups.

General Discussion

I will start the discussion by setting the dissertation's results in the context of the described theory and in the context of my clinical experience working with children with CHD and their families at The Queen Silvia's Children Hospital in Gothenburg, Sweden.

Children with congenital heart defects (CHD) begin their lives with a longer hospital stay after birth compared to healthy born infants. They are thoroughly examined. If diagnosed during the neonatal period, they are inspected for further symptoms or complications. Some will undergo surgery only a few hours or days after birth, but others will wait for further medical evaluations. For many, the CHD will be repaired and neither the child nor the parent will have to stay in contact with the cardiac care unit. For others, the CHD will require further surgery and catheter interventions and more visits to hospitals and more contact with doctors and care professionals, resulting in higher levels of anxiety for both the children and their families. Many times, children will experience long stays at hospitals, making them lose valuable play time, time to move around and discover, and time at school in contact with their friends. The cumulative effect of all these factors in children with CHD is indirectly addressed in this thesis through the measure of intellectual functioning and the measure of the impact of CHD on these families.

How can the results of the three empirical studies be understood in light of the literature presented in the background and how can the results of the three studies guide clinical practice and future research? The finding that most children with CHD had normal intellectual functioning is a result of the high quality pediatric cardiac care available in Sweden. In addition to the actual specialized health care, the overall welfare state contributes in decreasing stress levels in families compared to other countries where the same support is not available. When a child is sick, the national health insurance in Sweden allows one parent to stay at home and care for the child with pay. In cases of life threatening diseases, both parents are allowed to stay at the hospital to help with the care of their child with economic compensation that equals about 80% of their income (Wettergren et al., 2016). This gives a sense of security to parents taking care of their sick children. Although centralization means that cardiac surgery is only performed at two centers in Sweden and many parents must travel when cardiac surgery is needed, this centralization is associated with decreased mortality rates in the whole country. Furthermore, parental satisfaction with cardiac care has shown higher levels of satisfaction compared to parents of children with other diseases (Lawoko & Soares, 2004).

Nevertheless, parental satisfaction with pediatric cardiac care does not always reflect the emotional impact that the disease has on the family. Our results on family impact according to mother's and father's reports revealed that the more severe the heart disease, the more stress families experience.

Described in terms of the DSA model presented in the background section, these factors could be seen as reducing the stress experienced by parents by empowering them to be active in the care of their child. Because state welfare eases the economic burden of the disease, parents can spend time with their children. In addition, national welfare allows families to mobilize their resources that promote active parental engagement with children, creating internal schemas of security for their children and thereby maximizing possibilities for the child's normal development. However, especially for those parents whose children have a severe CHD, family patterns of interaction might be compromised. The relationship process in parent-child transactions are, at least temporally, set aside since surgical procedures and longer stays at hospitals are needed for the child's survival. Parents will not only be stressed about their child's situation, but also experience difficulties establishing parent-child relations. The emotional rhythm needed for parent-child exchanges to create the shared psychological state is disturbed. Children waking from anesthetics and heavy medications will not be able to fully participate in the relationship process. In addition, stressed parents will have difficulties adapting their interactions and responses to their child's behavior.

Our studies have shown the cumulative impact of low SES in severe and less severe forms of CHD. According to the DSA model, low SES might increase stressors that influence family resources and family patterns of interactions. Furthermore, low SES parents might have more limited abilities or cognitive skills. These limitations make it difficult for the parents to cope with a sick child's needs and as a result they might become, for example, over-protective, limiting their children's possibilities to use and develop their own social and cognitive resources. In addition, in situations of increased stress and few available protective factors and coping strategies, some parents develop characteristics that result in low cooperation with doctors and care professionals by questioning and rejecting the help that is offered. In my clinical experience, such behavior patterns and emotional reactions in parents are often mirrored in children: the children are afraid to trust health care providers working at the care unit, sometimes resulting in oppositional behavior, refusal of examinations and treatments, and even in depressive symptoms.

Intellectual functioning in children with CHD

Study I consisted of a sample of Swedish children with CHD treated with surgery or by catheter interventions. We found that as a group our sample performed within the normal range on overall intellectual functioning, a result that contradicts some earlier studies such as the ones presented in a literature review by Amianto (2011). In this literature review, some studies reported that CHD in children was related to lower intellectual functioning; however, other studies have found normal performance in these children not only with respect to general intelligence, but also with respect to academic results, learning abilities, and visuospatial abilities (Amianto et al., 2011a; Massaro, El-Dib, Glass, & Aly, 2008).

Although 65% of children in the present study performed within the normal range, 17% had scores -1 or -2 SD below the mean and 18% had scores +1 SD above the mean. When comparing subgroups of children with CHD, some children are clearly more at risk than others in terms of intellectual functioning. As hypothesized, intellectual functioning in children with CHD was related to severity of diagnosis, age, and SES.

In this study, we recruited similarly sized diagnosis groups with diverse severity levels to avoid focus on specific heart defects, a limitation of many other studies (Andropoulos et al., 2012; Bergemann et al., 2015; Walter Knirsch et al., 2012; Newburger et al., 2012). The results show that children with severe CHD had significantly lower intellectual functioning than children with mild or moderate CHD. This finding corresponds with studies reporting that children with milder forms of CHD, such as ventricular septal defect, present fewer neurodevelopmental problems than children with more severe forms of CHD (Gaynor et al., 2010; Sarrechia, Miatton, et al., 2015).

The fact that children with more severe forms of CHD present higher risk for neurodevelopmental problems suggests that cognitive problems could be related to intraoperative factors and to the surgery procedures themselves (Massaro et al., 2008). Although some studies have evaluated the risks that specific surgery techniques confer on children's intellectual functioning, no clear relation has been proven (Pizarro et al., 2014). However, genetic comorbidities and neurological status before surgery have been shown to be significant (W. Knirsch et al., 2010). Brain development during fetal and early postnatal life is influenced by environmental conditions such as maternal stress, and this psychosocial strain in turn influences intellectual functioning (Grizenko et al., 2015; Nilsson et al., 2004). A study measuring brain size in infants with CHD found that although brain size in these children was smaller than in healthy term infants, cerebral growth rates were comparable with the cerebral growth rates of the controls (Ortinou, Inder, Lamberth, et al., 2012).

In addition, no significant differences were evident in neurodevelopmental outcomes in pre-term born infants with CHD compared to term-born infants (Ortinou, Inder, Lambeth, et al., 2012; M. von Rhein et al., 2014).

Studies have highlighted the long-term effects of preoperative status (Heinrichs et al., 2014; Majnemer et al., 2009) and of the surgical procedures, which are determined by the severity of the heart defect (Sarajuuri et al., 2012; von Rhein, Dimitropoulos, Buechel, Landolt, & Latal, 2012; Wernovsky, 2006). In Study I, the SD in the severe group was wider than in the mild and moderate groups – i.e., the difference in intellectual functioning between the lowest and highest performance in the severe group was larger than in the mild or moderate groups. This finding has also been observed in previous studies (Bellinger et al., 2009; Bergemann et al., 2015; von Rhein et al., 2015).

Unrestricted as well as very restricted age groups are well-known problems in many previous studies (Mackie et al., 2013; McCusker et al., 2007; Pizarro et al., 2014). To overcome this problem, we tested children at well-defined ages. Results showed that children in the older groups (9- and 15-year-olds) had significantly lower intellectual functioning compared to the 3-year-olds. Three-year-old children were assessed with WPPSI-III and the 9- and 15-year-olds were assessed with WISC-IV, a choice that probably influenced the results. Although good correlations have been established between these instruments, the WPPSI-III usually produces slightly higher scores than the WISC-IV. In addition, as children become older and progress through school, the demands on their intellectual functioning increase. Although intellectual functioning is one of the best predictors of school performance, there are specific cognitive functions that influence learning (e.g., attention and memory). These specific cognitive factors are not targeted in the FSIQ of the Wechsler scales.

Deficits in specific cognitive factors were shown in one study of children with CHD tested at age 5 and 10; despite stable intelligence scores, the risk for cognitive deficits increased with age (Heinrichs et al., 2014). Difficulties with specific cognitive functions – e.g., attention, working memory, and processing speed – may not impact general performance in a test situation such as the Wechsler Scales of Intelligence, but may become evident in everyday situations as demands in school increase with age. The clinical experience is that children with CHD more often require special education and learning interventions when they are older. Future research should target specific cognitive domains such as attention, working memory, and executive function in children with CHD in relation to FSIQ and school performance.

Our results showed a significant relation between SES and intellectual functioning in children with CHD. Children from families with low SES had significantly lower intellectual functioning compared with both the

medium SES and the high SES groups. This finding corresponds with studies carried out in healthy populations in which intellectual functioning is related to socioeconomic status (Buckingham, 2013; Turkheimer et al., 2003). Results in our study showed that the high SES group also had significantly higher FSIQ compared to the medium SES group. Compared to children from families with average or high SES, more children from families with low SES had FSIQ below -1 SD. Although it is important to interpret this result cautiously since our sample had very small subgroups, this result corresponds with previous studies that show that parental education (McCusker et al., 2007), environmental processes (Alton et al., 2015; Gaynor et al., 2015), and parental stress (Majnemer et al., 2008) are related to intellectual functioning in children with CHD (Limbers et al., 2013).

Low intellectual functioning can adversely influence many aspects of children's lives (Limbers et al., 2013), affecting not only school functioning and education (Deary, 2012) but also emotion regulation (McClure, Halpern, Wolper, & Donahue, 2009) and health (I. J. Deary, 2012). Clearly, these children need strategic interventions. Studies reporting results of interventions aimed to promote intellectual functioning have shown advantages not only for intellectual functioning but also for self-perceived health development, emotional adjustment, and resilience in children with CHD; these types of interventions have been shown to reduce levels of anxiety in mothers, to increase good mental health in parents, and to promote good family functioning (McCusker, Doherty, Molloy, Casey, Rooney, Mulholland, Sands, Craig, Stewart, et al., 2012; McCusker et al., 2010).

In Study I, no interaction effect was found between severity of diagnosis, age, and SES, probably because of the relatively small sample size. However, when using a nonparametric test, children simultaneously exposed to severe CHD diagnosis and to low SES were found to more often perform in the low range of intellectual functioning. This finding also corresponds with previous studies showing that children with severe heart defects and lower SES are at greater risk for low intellectual functioning (Limbers et al., 2013; Sarrechia, De Wolf, et al., 2015).

A limitation of Study I was the absence of a control group. Although believed to be reliable, norm data have limitations – e.g., it is impossible to determine whether the groups are comparable on important background variables. Another limitation was the use of only one dependent variable, FSIQ. The response rate in the severe group was much higher than the response rate in the mild group. We do not have background data on the non-responders and there is a risk of response-bias. It is common that parents with higher education more often agree to participate in scientific studies compared to parents with lower education. It might also be the case that parents of children with more

difficulties at school were more inclined to participate because they wanted a cognitive evaluation of their child and more support from the school. We know that FSIQ in our mild CHD group was normally distributed, and the FSIQ scores in the severe CHD group were skewed toward lower values on the FSIQ.

Looking closer at the severity of diagnosis and SES relationship, we found that 23% of children in the mild group had high SES, whereas only 7% of children in the severe group had high SES. FSIQ in the mild group could be systematically higher because parents with high IQ and higher education more often agreed to participate or FSIQ in the mild group could be lower than expected because parents who were worried about their child more often agreed to participate. The moderate and severe groups' participation level was much higher and the risk for systematic response bias lower, although it is possible that the most fragile families, such as families with very ill children and very low socioeconomic status, did not participate. More studies using larger samples are needed to confirm the results of the present study.

A final limitation was the use of two measurements: the WPPSI-II and WISC-IV. The WIPPSI is usually considered to produce FSIQ scores higher than the WISC, which may account for the higher scores among the 3-year-olds. Because this is a cross-sectional study, effects should be interpreted with caution. The differences between age groups could be due to many factors. The uncertainty of the results, in part, are due to factors such as older and less effective surgical techniques used in the older children and the fact that different tests were used in younger and in older children.

In summary, children with CHD treated with surgery or catheter interventions in Sweden, as a group, performed well on FSIQ, although we identified severity of the heart diagnosis and SES as factors related to increased risk for lower FSIQ in children with CHD. Providing parents with accurate information on the risks of lower intellectual functioning, supporting schools with psycho-educational advice, and introducing follow-up and intervention programs for children as early as possible are important steps for improving the outcomes for children with CHD. Therefore, children with severe CHD and children from low SES families should be assessed for further interventions and included in follow-up and intervention programs.

Families of children with CHD need truthful information, including the risks to child development as a consequence of the disease. In the meantime, families need social support and help finding coping strategies to deal with the special situation of having a child with CHD. In addition, both parents and health care professionals need to be aware of the essential role that family resources and family patterns of interaction play in the child's development. Helping parents adjust and moderate stressors resulting from surgical procedures, medication, unfamiliar environment, etc. might improve their

attitudes towards health care professionals, making them feel competent in their role as parents. By feeling confident, parents might engage in parent-child transactions and family-orchestrated experiences as well as feeling they are protecting their child. All these aspects are fundamental in the long-term development of the social and cognitive competence of the child.

Screening instrument for further cognitive assessment

To identify those children with CHD who might need to undergo more extensive cognitive assessments, Study II evaluated whether the PedsQL Cognitive Problems subscale from the cardiac module could be used as a screening tool. This evaluation was done by investigating the association between children with CHD self-reports of their cognitive problems as well as their parents' reports (proxy-reports) of their children's cognitive problems and their children's actual FSIQ (measured using the Wechsler Intelligence Scales). In recent years, many studies have investigated the intellectual functioning of children with CHD (Amianto et al., 2011; Karsdorp, Everaerd, Kindt, & Mulder, 2007). This study adds to the literature by also looking at how children themselves and their parents rate their experienced cognitive problems and how these subjective ratings are associated with the more objective measures of FSIQ. In addition, this study found that the screening tool PedsQL Cognitive Problems subscale was useful for identifying children and adolescents who need further cognitive interventions.

When looking at the children's self-report, there was a strong association between the 15-year-olds' reports of cognitive problems and their FSIQ. These results are in line with previous empirical results and metacognitive theory (Lai, 2011). This type of self-evaluation demands that the child has developed certain metacognitive skills. In this particular case, the children needed to have a metacognitive component referred to as *cognitive knowledge* – i.e., knowledge about themselves as learners and the factors that affect their cognition (Flavell, 1979). The metacognitive ability of cognitive knowledge can be evident in children as young as six, but often these skills consolidate in adolescence (Schraw & Moshman, 1995). Therefore, it is not surprising that we found an association only for the 15-year-olds since this group is the only group where most of the children should have fully developed this type of metacognitive ability. However, it is likely that some of the 9-year-olds had this type of cognitive knowledge although this did not affect the results on a group level. This assumption is supported by the correlation of .27, albeit not significant, between the self-reports of 9-year-old and their FSIQ.

Regarding the proxy-reports, our results showed a moderate to strong correlation between the parental rating of cognitive problems and the FSIQ for all age groups. This result agrees with results from Miatton (2008), but our study adds to their results by showing that this correlation also exists for children as young as 3 and 5. This result suggests that parents have a good understanding of their very young children's cognitive problems (i.e., as early as 3 years old). However, when controlling for other factors in the regression, the association between the proxy-reports and the FSIQ was not significant for the 5-year-olds. This inconsistency could be a power issue due to the low number of participants ($n = 34$) in this age group compared to the other groups. This is evident when considering the high beta value of the proxy-report predictor for the 5-year-olds compared with the other age groups.

This study lends support to the idea that both the self-report and the proxy-reports of PedsQL Subscale Cognitive Problems can be used as a screening tool for identifying children who need to undergo further cognitive evaluation. Regarding the self-reports, the children need to be 15 years old for the screening tool to be valid. For both the self- and the proxy-reports, the suggested cut-off value for the PedsQL Cognitive Problem Subscale is 80; if a score is below 80, then a more formal evaluation with standardized tests is warranted.

Using the PedsQL Subscale Cognitive Problems as a screening tool is both economically sound and a time-saving alternative compared to more standardized cognitive testing procedures. In addition, not all clinics have the trained staff to perform more standardized evaluations, so this type of screening tool can help these clinics identify patients who need further evaluations. Using a screening tool also enhances the possibility of testing more children from an early age. This early detection means that appropriate resources and interventions can be set in place as early as possible for children with CHD, which in turn leads to better development for the child. Cognitive abilities not only affect learning but also affect many daily functions such as emotion regulation (McClure, Halpern, Wolper, & Donahue, 2009) and health (Deary, Pattie, & Starr, 2013). Earlier studies have shown that it is mainly children with a severe cardiac diagnosis (Wright & Nolan, 1994) who suffer from cognitive impairments, so performing extensive testing on children with milder forms of cardiac diagnosis could prove to be an insufficient and costly procedure. In these circumstances, it is highly beneficial to use the Cognitive Problems Subscale to screen children in the mild groups to identify those few who may experience cognitive problems and need to undergo more extensive evaluations.

A limitation of this study is that measured IQ in younger children is slightly unstable and does not necessarily reflect the intellectual

functioning the child will have as an adolescent (Wechsler, 2005, 2007). According to leading researchers in the field, IQ becomes stable around early adolescents (Deary, Pattie, & Starr, 2013). Although we should expect a higher variability in FSIQ in children below the age of eight, our study still found a moderate association between the younger age groups and proxy-reports, suggesting that a valid relationship exists between these measurements and the parents' ratings of their children's ability.

Another limitation is that the distribution was slightly biased for the children with different cardiac diagnosis severity. Although the number of children from the different diagnosis groups was somewhat equal, in real life there are more children with mild forms of CHD than children with a severe cardiac diagnosis. In our study, 80% of the families with a child suffering from a severe diagnosis who were contacted agreed to participate in the study. However, only 44% of the children with a moderate cardiac diagnosis and 31% with mild cardiac diagnosis agreed to participate. Because cognitive impairments are more prevalent among the children with more severe cardiac diagnoses, it is fortunate that this group is the most well represented. Although there exists a distribution bias in this study, this bias is unlikely to significantly impact the results.

This study revealed how accurately the children assessed their own cognitive competence and how accurately parents assessed their children's cognitive competence. Children's development, as hypothesized in the DSA model, is supported by constantly increasing social and cognitive competence. Younger children have not developed the capacity to assess their own capabilities, which they will be able to do, according to our results, by the time they are 15 years old. By this age, children's development is sufficient to control and organize cognitive skills. Processes such as metacognition, executive function, and social cognition will develop and help prepare the child to start understanding their own capabilities. Parents, on the other hand, have good knowledge of their child's cognitive capabilities. Cognitive capabilities, even at the age of 3, were accurately assessed by parents. These results help us realize that parents of children with CHD had the intellectual ability, confidence, and competence to describe their child accurately; that is, family resources in the form of personal characteristics of parents were well developed.

Results from this study are important because they confirm the fundamental role that parents have in their children's development. Understanding developmental resources in the form of abilities and skills as early as possible will help parents assess the needs of their child in time. Health care professionals working with parents of children with CHD need to support and facilitate resources that help families access possibilities for early detection and interventions for those children at risk. So far, these possibilities have

been given to parents and children with severe CHD, although only a few resources have been in place to detect children at risk, even with milder forms of CHD. However, it is the spirit of the follow-up program for children with CHD in Sweden to fulfil this need. As mentioned earlier, parents need accurate information about their child's risk for developmental problems in order for them to find help in assessing the problem and getting early interventions. By confirming the important role that parents have in detecting their child's cognitive abilities, we might help them feel confident in asking for assessments and interventions that can promote their child's social and cognitive competence.

Impact of children with CHD in their families

In families of children with CHD, mothers reported higher levels of negative impact than fathers (Study III). This finding is consistent with our hypothesis that mothers of children with CHD report more negative impacts than fathers, a finding that is consistent with studies reporting that mothers of children with CHD have higher levels of stress than fathers (Goldberg et al., 1990). Mothers of children with CHD have shown higher scores than fathers in studies measuring distress dimensions such as anxiety, depression, and somatization (Lawoko & Soares, 2002). Compared to fathers, mothers experience more worry associated with having a child with CHD (Casey et al., 2010) as well as more worries associated with having healthy young children (Kwok & Wong, 2000). We found no significant differences between fathers' and mothers' reports on the Positive Impact Scale, a finding that can be interpreted as both mothers' and fathers' experiencing similar levels of positive impact associated with a child with CHD. In studies of children with developmental delays and behavioral problems (Baker, Blacher, Crnic, & Edelbrock, 2002; Baker et al., 2003), mothers and fathers often report similar views and similar degrees of stress. According to Baker (2003), parents with children with delays more frequently discuss the child's difficulties with each other and with professionals, actions that can result in higher parental agreement. Optimistic appraisals on children's resilience could be a way for parents to compensate for the vulnerability associated with the uncertainty of their children's lives when parenting children with severe CHD such as hypoplastic left heart syndrome (Lee & Rempel, 2011).

A previous study has noted that parents of healthy children and parents of children with CHD both experience distress around the birth of their child; however, for parents of healthy children, distress tends to disappear over time, whereas for parents of children with CHD distress tends to intensify with the awareness that the child is chronically ill and might have to undergo heart

surgery (Lawoko & Soares, 2002). We found no significant relation between a child's age and negative impact, which contradicted our hypothesis that mothers and fathers of younger children with CHD report more negative impacts in their families than parents of older children with CHD. While some studies address stress in parents of young children associated with more demanding medical contact and surgical activities, other studies report that parental stress increases with the child's age, especially if the child with CHD also has developed behavioral problems (Visconti et al., 2002). These problems can make limit-setting difficult when children are older, a situation that often results in higher levels of stress in parents (Uzark & Jones, 2003).

Because the children in our study had a wide range of cardiac diagnoses, we divided them into three severity groups based on the risk the child had for further complications due to their cardiac diagnosis (i.e., mild, moderate, and severe CHD). We found that severity of the CHD was significantly related to parental stress for both fathers and mothers. Both fathers and mothers of children with severe CHD reported higher levels of negative impact than parents of children with mild or moderate forms of CHD. This finding confirms our hypothesis that parents of children with severe CHD report more negative impacts than parents of children with mild CHD. Our results are in line with studies comparing children with severe diagnoses that report stronger psychological impacts on parents caring for children with severe CHD (Montis & Tumbarello, 2011). When comparing severe diagnoses such as single ventricle physiology (SV) with less severe diagnosis such as biventricular (BV) physiology (Torowicz, Irving, Hanlon, Sumpter, & Medoff-Cooper, 2010), studies found that infants with SV physiology were more difficult to soothe compared to infants with BV physiology or healthy controls, and mothers of infants with SV physiology were more stressed than mothers of infants with BV physiology. In addition, our results are supported by studies finding that significant variation exists within different diagnostic groups of children with CHD, where parents of children with severe conditions such as HLHS report more negative impacts on the family than parents of children with milder forms of CHD such as TGA (Brosig et al., 2007). However, our results contradict studies that found that parental stress is not related to the severity of the child's CHD (Goldberg et al., 1990; Uzark & Jones, 2003). For example, one study found that mothers of children with serious heart conditions such as cyanotic-complex had fewer unscheduled visits to a general practitioner and fewer visits to emergency wards than mothers of children who had milder forms of CHD (Casey et al., 2010). In addition, one study found that a child's severity of CHD diagnosis did not significantly predict a parent's psychosocial status (i.e., anxiety, depression, and somatization) (Lawoko & Soares, 2006).

SES in families of children with CHD has been found to be associated with increased stress in parents (Visconti et al., 2002). However, the hypothesis that parents of children with CHD in families with low SES experience more negative impacts compared to parents in families with high SES was only partially confirmed. In our study, mothers with low and medium SES reported higher stress than mothers in the high SES groups, although this relation was not found in fathers. Socioeconomic factors in mothers of children with CHD such as marital status (i.e., being a single parent) have been reported to be related to higher stress scores (DeMaso, Campis, et al., 1991; Fleck et al., 2015). Casey (2010) found a strong association between health care demands and psychosocial factors, confirming higher frequency of multiple admissions in young children with CHD in lower SES groups (Spencer, Lewis, & Logan, 1993). Previous studies found that low SES, often related to mother's education, was significantly related to functional and mental outcomes and the psychosocial well-being of children with CHD (Alton et al., 2007; Alton et al., 2015; Larsen et al., 2010). In our study, fathers with low SES scored significantly higher on the Positive Impact Scale than fathers in the medium and high SES groups, but we did not find any studies confirming these results. A literature review of pediatric psychology research on fathers' roles found that fathers were underrepresented in research (Phares, Lopez, Fields, Kamboukos, & Duhig, 2005), pointing to the importance of our results. According to Phares et al. (2005), although studies of chronically ill children have shown similarities in responses between mothers and fathers, parents may play different roles in dealing with child-related issues. Therefore, pediatric research should include both mothers and fathers. Parents of children with severe CHD have shown ambivalence about experiencing their children as more vulnerable versus the joy of feeling that their children are more special due to their survival (Larson, 1998; Lee & Rempel, 2011). Since mothers report higher levels of stress, it is possible to argue that fathers' positive perceptions of the child's impact on the family is an attempt to find a balance within the family.

Some studies have shown that children with severe CHD have lower intellectual functioning compared to the general population (Gaynor et al., 2015). Studies on children with intellectual disabilities or developmental delay have reported more negative impacts on parents compared to parents with children with typical development (Boström, Broberg, & Hwang, 2010). Our hypothesis that parents of children with CHD with low intellectual functioning report more family stress than parents of children with high intellectual functioning was not confirmed. On the contrary, for both fathers and mothers, intellectual functioning was negatively associated with positive impact. Fathers of children with average IQ as well as mothers of children with low and average IQ reported significantly higher positive impact than fathers and

mothers of children with the high IQ. Larson (1989) discusses the opposite positions that parents often report concerning the impact the child has on the family, which can be interpreted as a way to balance their ambivalent perceptions (Larson, 1998). This finding was also confirmed in a study by Boström et al. – parents rated high negative impact when having a child with intellectual disabilities or developmental delays and at the same time as they reported similar high levels of positive impact as the control group (2010). One interpretation could be that as long as parents experience a balance between negative and positive impact, they are not at high risk for adverse psychological effects such as anxiety and depression; however, if negative experiences are common and positive experiences are rare, there is an increased risk for psychological symptoms and need for support.

In the present study, the strongest predictor for negative impact for both mothers and fathers was the presence of multiple risk factors. Both the fathers and mothers reported that an accumulation of risk factors was related to negative impact. This analysis can have important clinical implications as it should direct the clinical eye towards the approximately 30% of families who have an accumulation of risk factors and therefore are at high risk of experiencing negative impacts. It might be wise to evaluate the presence of risk factors when meeting families in clinical settings.

This study was limited by the absence of a control group of parents of healthy children. We relied both on norm data and comparisons found in the existing literature about parents of children with CHD or chronic diseases. This limitation could imply a risk since our data are limited by unknown background variables. The response rate was proportionally higher in the group with severe diagnoses, which could imply a response bias and as a consequence an overestimation of the results of the negative impact in our sample. This is a cross-sectional study, so the results must be interpreted carefully. Each group had its unique and particular characteristics and therefore inferences must be handled carefully, especially inferences over time. Longitudinal studies of fathers and mothers are needed to understand the impact of having a child with CHD and the development of negative and positive factors in the family. Finally, since the fathers and mothers reported the impact of their child together, the likelihood is high that the parents influenced each other's views on the child's impact on the family. However, in spite of these limitations, our study gave us a good perspective of the fragile situation parents of a child with CHD experience and the importance of addressing preventive interventions for these parents and their families.

This study adds to the understanding of how CHD impacts families and how parents of children with CHD influence their children's development. Using the DSA model to evaluate our findings, we could confirm that ill

children do create stressors that affect family resources. These stressors might with time interfere with the child's social and cognitive competence.

Although clearly there are positive emotions involved in giving birth to a child, it is not uncommon that parents of children with CHD also experience what we called a negative impact, which is sometimes described as distress, which includes components of stress and depression. The mixed emotions that giving birth to a child with CHD results in parents feeling worry, sorrow, and disappointment as well as feelings of pride and joy of having a baby survivor and fighter. All these mixed feelings or emotions might interfere with family patterns of interactions that in turn might compromise the child's social and cognitive competence.

Developing the processes of interplay between parent and child, which are often established in the context of family routines, are interrupted by the presence of surgical interventions and stays at care units in hospital. The important conversations between parents and children and those give-and-take exchanges between them are needed to develop a secure attachment, but these interactions are often stifled by tubes and machines keeping the child alive. Families with short-term stays in pediatric cardiac care units are more likely to start family routines that might develop a continuous and stable communication process between parent and child, interactions that are important in developing the child's social and cognitive competence. These parents can more quickly initiate a safe home environment and make child-specific adjustments necessary using their sensitivity and warm interactions. For families spending longer periods in care units or coming back for more surgery or catheter interventions, this reintegration into the home environment will be more difficult. The longer a child is in a care unit, the higher the levels of distress among parents and the more difficult it will be for them to establish synchronous relationships with their child. Children and parents staying at the hospital experience mixed emotions; they are positive because they know that the intervention is needed for the child to become better, but they are also stressed because there is always uncertainty and risk associated with every intervention. Furthermore, hospital environments are far from the ideal for promoting parent-child transactions. Families are constantly interrupted. The need to take measurements, take samples, and make assessments as well as the need for staff to check on the child's status make it difficult for parents and child to engage in sensitive-responsive interactions.

Results in our three studies show that although the majority of parents succeeded in promoting child's social and cognitive competence even in cases of children with severe CHD, there were some families that needed support to develop family resources and family patterns of interaction in order to promote their child's social and cognitive competence.

Clinical Implications

During this study, work was started to establish a national multi-professional follow-up program for children. The working group proposed a follow-up program that covered several developmental domains, such as the cognitive and psychomotor domains. The recommended psychological follow-up procedure included a structure for screening, assessment, and intervention regarding cognitive/intellectual, emotional, and behavioral development of the child. The goal was to introduce a systematic multidisciplinary follow-up rehabilitation program for children with CHD that helps professionals identify those children at risk who, in addition to the follow-up of their heart disease, also need multidisciplinary interventions to improve their development in full.

The first two empirical studies in the present thesis are an important source of knowledge for addressing the dimensions of intellectual functioning in this population. Study I reveal the large differences in intellectual functioning in the different severity groups of children with CHD and the important repercussion of SES factors in combination with the severity levels of the heart disease. Study II provides good results in using a time-effective, cost-effective, as well as reliable screening tools for detecting cognitive problems.

Based on the two first studies, we suggest the following procedure for ensuring children with CHD with compromised intellectual functioning are followed-up. The contact nurse at the pediatric unit of each regional hospital introduces the PedsQI-heart module to parents of children and adolescents who have CHD. This module will be used to screen for cognitive difficulties that can influence school performance. This introduction could be done at a regular medical follow-up visit. If the score on the cognitive section (five items of the PedsQI-heart module) is under 80, the child will be referred to the psychologist at the hospital for further evaluation. The psychologist performs a basic psychological evaluation using the Wechsler Intelligence Scales. If parents report behavioral issues, learning difficulties, or results that fall under -1 SD from the mean, contact with the school is suggested in order to discuss pedagogic strategies. If suspicion of neuropsychiatric problems arises, a referral will be sent to the Childs Psychiatric Center (BUP) for further evaluation and intervention. If results are under -2 SD from the mean, special compulsory education for the child will be suggested (preferably after a second evaluation).

Study III emphasizes inclusion of parents and families in pediatric cardiac care. We know that having a child with CHD increases the family's need for information. This information, which goes beyond the child's cardiac diagnosis and medication, has to address understanding the child's cues

so parents can respond to their child's needs, specifically developmental needs, and important aspects of the child's life such as addressing behavioral problems and the development of social skills.

Interventions designed to address medical information as well as the emotional and psychological strains that parents, siblings, and extended family undergo during the treatment process of the child are fundamental for securing the best health care for children with CHD and their families. Study III will become an important complement to the follow-up and rehabilitation program for children with CHD since it focuses on the emotional impact that children with CHD have on their families, raising awareness of the psychosocial concerns and needs of families. Moreover, Study III reveals there is a need for family interventions in pediatric cardiac care.

Theoretical contribution

The introduction of cognitive developmental theories is meant to bring up awareness of the risk and protective factors that play a role in the development of children with CHD. The theoretical framework model designed by Guralnick (2011) – Development Systems Approach (DSA) – addresses the mechanisms that interact to influence a child's social and cognitive competence. This model recognizes that biological factors interact with environmental and parental factors in child development.

This study adds to the existing literature regarding the cumulative effects of severity of the heart defect and the importance SES factors on intellectual functioning and family impacts. These two factors not only influence children's intellectual performance but also parental stress in perceived negative impact on the family.

Because our studies do not include data on factors related to either nature or nurture, no conclusions can be drawn regarding why children from families with low SES showed lower intellectual functioning than children with the same severity of CHD from families with higher SES. One possible explanation could be that intellectual functioning is highly related to SES (Turkheimer et al., 2003). In addition, when stressors accumulate in families (e.g., economic stress and stress related to the medical and care for the child), risks may exceed the resources available to regulate the stress and increase the risk for negative processes, including less optimal parent-child interaction, placing the already medically-vulnerable child in a developmental context that may not be as supportive as needed.

Clinical practice in pediatric cardiology has been successful in increasing survival rates through the years for children with CHD. In Sweden,

children with moderate and severe CHD have been followed medically and controlled, offering the best medical solutions for their health. However, the results of the present thesis provide support to broaden the focus of pediatric care from pure medical to habilitation interventions directed to improve social and cognitive competence.

These interventions should promote the best possibility for development and health for children with CHD so these children have the same possibilities for development as healthy children. Families, preschools, schools, and health caregivers need guidance and support to give the child the best possible developmental context, not only concerning physical development but also psychological and socio-emotional development. Advice and support can be directed towards understanding the child's developmental needs that result from the child's cognitive and social abilities and ability to organize and use these capacities. Guidance might also include bringing up the importance of family resources including the capacity of parents to interact with their child, competence of health care professionals in the child's specific needs, and the material and social resources available to the family. All these resources, however, are often susceptible to stressors in the child's and family's environment when the child is born with a CHD. This research might help us better educate parents, teachers, and health care providers of the important role that each one has in promoting the child's social and cognitive competence.

Methodological Challenges

To investigate the intellectual functioning in children, we could simply ask parents or teachers about their perception of the child's intellectual performance. However, this method might be subject to bias from the informant and without the careful assessment of the different aspects or cognitive abilities that involve the integrated concept of intellectual functioning. On the other hand, intellectual functioning is not only a matter of cognitive abilities associated with IQ; intellectual functioning is influenced by personality traits, motivation, and environmental factors that promote or limit the child's ability to resolve problems, take in information, learn, communicate, and socialize at school or in everyday life.

We decided to use The Wechsler Scales of Intelligence as these are comprehensive psychological instruments for assessing general intellectual ability (i.e., Full Scale IQ) in children. While these instruments are reliable measures of intellectual functioning, they do not fully answer the question why children with CHD present learning difficulties or require special education

more often than other children (Spijkerboer et al., 2008, Mulkey et al., 2014). To address this question, we would have needed to gather information from teachers and parents as well as observations on the behavior of the child while learning and functioning in daily situations.

The task involved in testing children and gathering data from a large sample of children and their parents at four different locations brought a series of challenges regarding keeping the fidelity of the method. While all 15-year-olds wanted to be tested without their parents present, none of the three-year-old children wanted their parents to leave the room. This was the case for almost all 5-year-olds as well, but it was not the case for all 9-year-olds, who sometimes wanted their parents to wait outside the test room.

Although the majority of testing was done at the Queen Silvia's Children's Hospital (73%), it required help from several test leaders. The test leaders, who were from different hospitals, were clinical psychologists with a great deal of experience in testing children using the Wechsler Intelligence Scales. The psychologists testing at these hospitals meet every semester to discuss the tests and the gathering of data. In addition, during these meetings the psychologists discussed problems with specific test situations. At the Queen Silvia's Children Hospital, an additional four test leaders with less experience were employed during the spring 2015. To increase fidelity of the method, test leaders were closely supervised by the project leader (Carmen Ryberg). The project leader trained all test leaders in the use of tests, emphasizing the importance of staying faithful to the instructions and communicating with the project leader. Furthermore, test leaders were subjected to supervision at all times, which included the project leader doing stick controls to see how test leaders tested, giving them feedback, answering all questions concerning testing technique, and assessing scoring.

Gathering information from parents was also challenging. We wanted to gather information from these children's parents separately. Often children were accompanied by one parent to their testing session. This parent answered questionnaires while waiting their child to be tested. This was a positive way of keeping the parent's focus on questionnaires instead of interfering with the child during testing. Parents were asked both in writing and verbally not to discuss answers with their partners. This was explained to parents by mentioning that we were interested in studying how mothers and fathers independently perceived issues concerning their child. The parent who accompanied the child to the testing session received a kit that included questionnaires to give to the other parent along with an envelope; the completed kit was to be sent later by post. This method of gathering data is not without problems. Parents at home had the possibility to ask or discuss answers, which is not uncommon among parents. To avoid this problem, we could have chosen to interview

parents separately. Interviews would have been a good way to understand the challenges and emotions within the family when raising a child with CHD.

Future research

Research is needed that focuses on risk factors that compromise the social and cognitive competence in children with CHD. In addition, studies are needed that focus on both short-term and long-term consequences of interrupted family patterns of interactions because of surgical procedures. A fundamental requirement is understanding consequences of the difficulty parents have establishing relationships during the first year of the child's life, as the child's optimal development might be affected. These studies might help us suggest measures in pediatric health care that encourage not only parent-child interactions but also caregiver relationships, all this with the goal to reach an optimal development for children with CHD.

Cross-sectional and longitudinal studies are necessary to understand the social consequences now and in the long-term development of the disease in children. To establish better interventions, care providers and researchers need a better understanding of how socioemotional connectedness with parents, the peer network, and the community the child grows up in are affected by the CHD.

We also need longitudinal studies to have an accurate picture of the cognitive development of children with CHD. Furthermore, more research is needed on specific cognitive functions that interfere with these children's school activities, learning, and possibilities to obtain an education and to become an independent adult. Future research should target specific cognitive domains such as attention, working memory, and executive function in children with CHD in relation to FSIQ and school performance. Future studies should also focus on why there is an overrepresentation of children with cognitive impairments among the children with more severe diagnoses.

Longitudinal studies of experiences and psychological symptoms in fathers and mothers separately are needed to understand the differences in perceived impact of having a child with CHD in the family and how they evolve over time. It is important to study how the child-parent relationship is affected by the disease and what repercussions this might bring to the development of the child. Furthermore, we need to understand the developmental trajectory of families to detect potential resilient factors that can be used by parents of a child with CHD. In addition, it is important to evaluate studies in Sweden that include family interventions directed to promote competence in

children with CHD and emotional wellbeing such as the family intervention program CHIP.

We need research that examines family resources and evaluates results from family interventions. By using family screenings and follow-up assessments, we might be able to understand the specific characteristics and needs that families have so we can better adjust and establish more specific interventions for families. That is, interventions need to be evaluated and fine-tuned to meet the cultural and social needs of specific populations.

Last, but not least, it is necessary to evaluate the consequences of the high survival rate in the CHD population. Questions will need to be answered. What are the long-term repercussions for children with CHD when they become adults? What are the emotional, social, and cognitive costs for children growing up with CHD? What are the emotional costs for families raising children with CHD? What are the future possibilities for children being born with CHD? What are the resilient factors that many children have shown even with a CHD? How can we improve the pediatric cardiac care for children with CHD and their families?

Final Words

Clinical practice in pediatric cardiac care involves understanding both the medical risks of the child and the psychosocial consequences of the disease. For a psychologist, this means not only listening, understanding, and assessing concerns such as school difficulties, behavioral problems, and family concerns, but also suggesting interventions to address these problems. Many parents of children with CHD are worried that their child will not survive the disease and they are worried about what will become of their child if they do survive the disease. They worry that their child might not have access to the resources that he/she needs to be successful at school and work and become an independent adult with a good life.

Many studies show that children with CHD have higher incidence of cognitive impairments and poor academic results compared to healthy controls (Amianto et al., 2011b; P. A. Karsdorp et al., 2007). Therefore, demands for future health care practitioners will include the consideration of cognitive and developmental issues for these children. Parents need scientific information not only about the structural or functional heart problem of their child but also about the social and cognitive risks associated with the disease. By giving parents more complete information on risks and possibilities for their child, we might help them understand their child in a way that will help them provide their child with the resources he/she needs to develop into

intellectually as well as emotionally competent adults. In addition, families need to be informed about family and parental factors that might stimulate their child's social and cognitive competence (as discussed in Guralnick's model) and to seek advice from professionals when needed. By screening, assessing, and counselling parents on the risk factors as well as the resiliency factors that can limit or promote their children's possibilities for development, we might take one step further towards better pediatric cardiac care for children with CHD and their families.