“In Good Heart”: A Study of the Factors Associated with Health-Related Quality of Life in Adult Congenital Heart Disease

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ACKNOWLEDGEMENTS

"As you set out for Ithaka hope your road is a long one, full of adventure, full of discovery […]
Keep Ithaka always in your mind. Arriving there is what you're destined for.
But don't hurry the journey at all.
Better if it lasts for years, so you're old by the time you reach the island,
wealthy with all you've gained on the way, not expecting Ithaka to make you rich.
Ithaka gave you the marvelous journey. Without her you wouldn't have set out.
She has nothing left to give you now.
And if you find her poor, Ithaka won't have fooled you.
Wise as you will have become, so full of experience,
you'll have understood by then what these Ithakas mean."

Constantine P. Cavafy

And what a long and challenging road this has been!

I first wish to thank my supervisors for encouraging me to take this journey and believing that I can reach my Ithaka. To Professor Stanton Newman for his invaluable guidance throughout the years, his enthusiasm for the subject, and his inspiring and felicitous metaphors. To Dr Shashivadan Hirani for his mentoring, advice, expertise, and kind support. Thank you to all my colleagues, past and present, for reminding me that I was not alone and that each day was a step closer. Special thanks to Hayley McBain for all the support in the last few months.

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To all my friends back in Cyprus for all the encouraging calls, messages, and e-mails while we were apart and for otherwise turning my home into a B & B. I am sure there were many places around the world that you could have visited, yet you chose London every time. Thank you to my parents, for making me who I am today, for always believing in me, and for the life lessons that books could not provide; no words can describe how privileged I feel to have you as my parents. To my brothers and guardian angels, George and Mario, for still attempting to protect me from all life's troubles. Although you could not protect me from the difficulties of a PhD, your generosity, humour, and endless support have definitely helped me through the process.

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DEDICATION

I dedicate this thesis to my parents Demetris and Petroulla, who have been a source of inspiration all my life and who have wholeheartedly supported me throughout all of my Ithaka journeys.

DECLARATION

I, Theodora Fteropoulli, confirm that the work presented in this thesis is my own. Where information has been derived from other sources, I confirm that this has been indicated in the thesis.

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ABSTRACT

Aims: The overall aim of the thesis is to study the Health-Related Quality of Life (HRQoL) in Adult Congenital Heart Disease (ACHD). The thesis examines the impact of ACHD on HRQoL, the demographic, clinical, and psychosocial factors associated with HRQoL, and changes in these factors over time.

Methods: The thesis consists of three studies, including a cross-sectional, a longitudinal, and a qualitative. Three hundred and fourteen individuals, from four diagnostic groups (Simple, Tetralogy of Fallot-ToF, Transposition of the Great Arteries-TGA, & Single Ventricle-SV) were recruited from The Heart Hospital, London and completed the cross-sectional study. Clinical measures were collected by a consultant cardiologist and participants completed a range of psychosocial measures, including generic and disease-specific measures of HRQoL. One-sample t-tests were conducted to examine differences between the whole sample and diagnostic groups and general population norms and ANOVAs for differences between the four diagnostic groups in HRQoL. Hierarchical multiple regressions were employed to examine the factors associated with HRQoL. One hundred and eighty eight participants completed the longitudinal study (mean follow-up: 3.3 years). Changes in HRQoL and other psychosocial variables were explored using Hierarchical Linear Models (HLM). Semi-structured interviews were conducted with twelve participants to explore their experiences.

Results: Participants reported diminished HRQoL in some physical and psychosocial domains. The SV group reported poorer physical and psychosocial HRQoL in various domains compared with the general population norms and some of the other diagnostic groups, whilst the Simple group reported impaired psychosocial HRQoL. Illness perceptions explained the largest proportion of variance in generic (physical and psychosocial) and disease-specific HRQoL. After accounting for demographic and clinical characteristics, psychosocial factors including perceptions about the symptoms and consequences of ACHD, the use of self-blame as a coping strategy, and mood were independently associated with HRQoL. Clinical factors, HRQoL, coping, and social support remained stable over time, whilst some changes were observed in illness perceptions and anxiety. The qualitative interviews provided further insight into the physical, psychosocial, and occupational consequences of ACHD, the people's efforts to adjust to their condition and treatment and gain perspective, and the role of their social environment.

Discussion: The theoretical and clinical implications of the findings are discussed. People with ACHD may benefit from routine psychological assessment and provision of clear and timely information and interventions addressing negative illness perceptions, depression, and anxiety.

LIST OF ABBREVIATIONS

AA – Aortic Atresia
ABC – Aristotle Basic Complexity
ACHD – Adult Congenital Heart Disease
APVD – Abnormal Pulmonary Venus Drainage
AS – valvular Aortic Stenosis
ASD – Atrial Septal Defect
ASDII – Secundum Atrial Septal Defect
AVD – Atrial Valve Diseases
AVR – Aortic Valve Replacement
AVSD – Atrioventricular Septal Defect
BAV – Bifoliate Aortic Valve
BDI – Beck Depression Inventory
Brief IPQ - Brief Illness Perceptions Questionnaire
BSI – Brief Symptom Inventory
CAT – Common Arterial Trunk
CCTGA – Congenitally Corrected Transposition of the Great Arteries
CES-D – Center for Epidemiologic Studies Depression Scale
CHD – Congenital Heart Disease
CHD-TAAQOL – Congenital Heart Disease-TNO-AZL Adult's Quality of Life
CI – Confidence Intervals
CoA – Coarctation of the Aorta
ConQoL – Congenital Heart Disease Quality of Life Questionnaire
CPEX – Cardiopulmonary Exercise Testing
CSI – Coping Strategy Indicator
CT – Computerised Tomography
CVC – Cardio & Vascular Coalition
DDA – Disability Discrimination Act
DILV – Double Inlet Left Ventricle
DIV – Double Inlet Ventricle
DoH – Department of Health
DORV – Double Outlet Right Ventricle
DSM - Diagnostic and Statistical Manual of Mental Disorders
DUKE – DUKE Health Profile
ECG – Electrocardiography
ES – Eisenmenger Syndrome
FA – Framework Analysis
GBB – Giessener Complaints Questionnaire
GUCH – Grown-up Congenital Heart
HADS – Hospital Anxiety and Depression Scale

HLHS – Hypoplastic Left Heart Syndrome
HLM – Hierarchical Linear Models
HLV – Hypoplastic Left Ventricle
HRQoL – Health-Related Quality of Life
HRV – Hypoplastic Right Ventricle
ICC – Intra-Class Correlation
ICD – Implantable Cardioverter Defibrillator
IE – Infective Endocarditis
IPA – Interpretative Phenomenological Analysis
IPCCC – International Paediatric and Congenital Cardiac Code
IPQ – Illness Perceptions Questionnaire
IPQ-R – Illness Perceptions Questionnaire Revised
ISEL – Interpersonal Support Evaluation List
ISNPCHD – International Society for Nomenclature of Paediatric and Congenital Heart Disease
ISOQOL – International Society for Quality of Life Research
LAS – Linear Analogue Scale
LVOTO – Left Ventricular Outflow Tract Obstruction
MA – Mitral Atresia
MAPCAs – Major Aorto-Pulmonary Collateral Arteries
MCAR – Missing Completely at Random
MCMC – Markov Chain Monte Carlo
ML – Maximum Likelihood
MOS-SSS – Medical Outcomes Study Social Support Survey
MRI – Magnetic Resonance Imaging
MSPSS – Multidimensional Scale of Perceived Social Support
MVP – Mitral Valve Prolapse
NBS – Norm Based Scoring
NCCDPHP – National Center for Chronic Disease Prevention and Health Promotion
NHI – National Health Institute
NHLBI – National Heart, Lung, and Blood Institute
NHS – National Health Service
NICOR – National Institute for Cardiovascular Outcomes Research
NYHA – New York Heart Association
PA – Pulmonary Atresia
PAH – Pulmonary Artery Hypertension
PCQLI – Pediatric Cardiac Quality of Life Inventory
PFO – Patent Foramen Ovale
PHQ-9 – Patient Health Questionnaire-9
PPI – Public and Patient involvement
PR – Pulmonary Regurgitation
PRISMA - Preferred Reporting Items for Systematic Reviews and Meta-Analyses
PROMs – Patient-Reported Outcome Measures

PS – Pulmonary valve Stenosis
PSSS – Perceived Social Support Scale
PVD – Pulmonary Valve Diseases
PVR – Pulmonary Valve Replacement
QoL – Quality of Life
R&D – Research & Development
RACHS-1 – Risk Adjustment in Congenital Heart Surgery-1
REML – Restricted Maximum Likelihood
RVOTO – Right Ventricular Outflow Tract Obstruction
SAS – Sub-Aortic Stenosis
SCL-90-R – Symptom Checklist-90-R
S.D. – Standard Deviation
SEIQoL-DW – Schedule for the Evaluation of Individual Quality of Life-Direct Weighting
SF-36 – Short Form 36 Health Survey
SIP – Sickness Impact Profile
SOC – Sense of Coherence
SQoL – Subjective Quality of Life
SRM – Self-Regulation Model
SRV – Systemic Right Ventricle
STAI-6 – State-Trait Anxiety-6
STAI-T – State-Trait Anxiety-Trait Version
SV – Single Ventricle
SVAS – Supravalvar Aortic Stenosis
TA – Tricuspid Atresia
TAAQOL- TNO-AZL Adult's Health Related Quality of Life
TCPC – Total Cavo-Pulmonary Connection
TGA – Transposition of the Great Arteries
TOE – Transoesophageal Echocardiography
ToF – Tetralogy of Fallot
UCLH – University College London Hospitals
UIT – Uncertainty in Illness Theory
VIF – Variance Inflation Factor
VO_2 max – Peak Oxygen Uptake
VSD – Ventricular Septal Defect
WCQ – Ways of Coping Questionnaire
WHO – World Health Organization
WHOQOL-Bref – World Health Organization Quality of Life-Bref
η^2 – Eta Square
φ_2 – Cramer's V
χ^2 – Chi square

CHAPTER 1 – CONGENITAL HEART DISEASE, ITS TREATMENT, AND IMPACT

1.1. Prologue

The overall aims of the thesis are to study the Health-Related Quality of Life (HRQoL) and the factors associated with HRQoL in Adult Congenital Heart Disease (ACHD). To capture the potential changes in HRQoL and psychosocial factors over time, these were assessed on two occasions in a sample of participants. In addition, the thesis consisted of a comprehensive set of studies including both qualitative and quantitative studies so as to provide a more comprehensive understanding of HRQoL in this group of people with ACHD. This chapter provides a brief description of the heart and its function followed by a description of Congenital Heart Disease (CHD) and the emergence of ACHD as a sub-specialty. The aetiology, epidemiology, survival and mortality rates in CHD are described. This chapter also includes a description of the classification initiatives in CHD. The most common defects, their treatment, and prognosis are also presented. The chapter concludes with an overview of complications in ACHD.

1.2. The heart: structure and function

To understand CHD and its types it is important to understand the structure and function of the normal heart. The heart is a muscular, fist-sized organ weighing between 200 and 400 grams that lies in the middle to the left of the thorax and is part of the cardiovascular system. It is comprised of four chambers, the upper left and right atria and the lower ventricles, separated by a muscular wall called the septum. The atria and ventricles work together with the four valves pumping blood around the body through the arteries and veins.

Every time a heart beats, two actions take place: diastole and systole. Blood is collected in the left and right atria and a contraction pushes blood through the tricuspid and mitral valves into the respective left and right ventricles (diastole). When the ventricles are filled with blood another contraction takes place (systole). Oxygen-poor blood is pushed from the right ventricle through the pulmonary artery into the lungs, which oxygenate the blood. Oxygen-rich blood is simultaneously pumped from the left ventricle through the aortic valve to the heart and other body parts. This cycle is repeated many times, with the heart beating 60-80 times a minute during rest. Every day the heart beats approximately 100,000 times, pumping more than 7 litres of blood.

1.3. Congenital heart disease: definition and diagnosis

CHD refers to abnormalities in the structure or the function of the heart, which are present at birth. Most defects are characterised by an ill-formed heart, structural deficiencies of the valves and vessels, or small holes on the septum.

Many defects do not have obvious or even noticeable symptoms and may be diagnosed later in life. Symptoms normally depend on the severity of a specific defect. A number of typical symptoms may be observed in people with CHD:

- Cyanosis: oxygen de-saturation caused by the persistent mixing of systemic venous blood (oxygen-rich) and pulmonary blood (oxygen-poor)
- Shortness of breath
- Dizziness or fainting (syncope)
- Arrhythmias: irregular heartbeat/rhythm
- Palpitations: heartbeat abnormalities
- Fatigue

- Reduced exercise capacity

CHD may be diagnosed at any point from the antenatal period to adulthood. Antenatal diagnosis was introduced more than 30 years ago and approximately 60% of CHD is now diagnosed during the antenatal and new-born period (Knowles & Hunter, 2014). Antenatal screening is usually conducted using ultrasound (foetal echocardiography) at the mid-trimester anomaly scan and appears to be useful in planning successful postnatal management (Brown & Sullivan, 2014). Recent data for 2013-2014 suggest that in the UK, up to 45.7% of major CHD is diagnosed during the antenatal period in those undergoing intervention in infancy (National Institute for Cardiovascular Outcomes Research [NICOR], 2014). However, up to 40% of CHD may be diagnosed later in life including adulthood (Petersen, Peto, & Rayner, 2003). When CHD is suspected in the postnatal period and following an initial physical examination, diagnostic tests are carried out to confirm the presence of a cardiac defect. In the past, diagnosis was established through cardiac catheterisation. This involved a long thin tube (i.e. catheter) being inserted into an artery in the arm or leg and guided to the heart in order to map it using contrast dye and an x-ray machine. Due to recent diagnostic advances the presence of CHD may be identified through less invasive approaches, including transthoracic or transoesophageal echocardiography (TOE), electrocardiography (ECG), chest radiography, exercise stress testing, magnetic resonance imaging (MRI), and computerised tomography (CT) (Popelová, Oechslin, Kaemmerer, & Sutton, 2008).

The combination of advanced diagnostic procedures and developments in surgical and interventional techniques along with the increase in quality of care led to a large population of affected children surviving well into adulthood. The result is a

continuously increasing population of adults with CHD and the development of a new cardiovascular sub-specialty, ACHD (Perloff, 1991), which includes all adults aged 16 and above, who have successfully transitioned from paediatric to adult care (Watson & Rana, 2009). In the past, the adult population of people with CHD was referred to as Grown-Up Congenital Heart (GUCH), but this has been superseded by the term ACHD.

For the purposes of the present thesis the term ACHD is used throughout to refer to the adult population under study. The general term CHD is used within the context of the child and adolescent literature and when referring to the condition in general.

1.4. Nomenclature and classification

The inherent diversity that characterises CHD is attributed to the large number of different defects and their subtypes. A standardised system of nomenclature of CHD has only recently emerged. The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) created a comprehensive international nomenclature system by cross-mapping several existing lists from across the world. The International Paediatric and Congenital Cardiac Code (IPCCC) (Franklin et al., 2008) is the result of meticulous research and meetings between several specialists from all over the world. The aim of this initiative is to name and categorise CHD and its treatment for the purposes of risk stratification, comparison of outcomes between specialised centres, and for the overall improvement of patient care. Although the IPCCC is currently only used by individual specialist cardiologists, there is great potential for its adaptation in research.

Complexity of surgery has also been the focus of efforts to produce a standardised system. The Risk Adjustment in Congenital Heart Surgery-1 (RACHS-1) method

and the Aristotle Basic Complexity (ABC) score are two independent systems for the classification of treatment complexity. The future aim is the unification of these two systems (Jacobs et al., 2007).

The following description of the most common defects is organised in four groups (Figure 1.1). The terminology used in the thesis is based on the clinical literature and previous studies in ACHD. For the purposes of the present thesis, the term used to describe these four groups will be "diagnostic groups", with differing structural changes in the heart. This term is also used throughout the analyses described in the thesis.

1. "Simple"

The following defects were collapsed into this group:

- ventricular septal defect
- atrial septal defect
- right ventricular outflow tract obstruction (pulmonary valve stenosis)
- left ventricular outflow tract obstruction (valvular aortic stenosis, coarctation of the aorta)

2. Tetralogy of Fallot

3. Transposition of the Great Arteries

4. Single Ventricle

Figure 1.1. Description of the four diagnostic groups

The four diagnostic groups are described in the next sections. The most common long-term complications are discussed in more detail in section 1.8 (page 40).

1.4.1. "Simple"

Various defects were categorised under this diagnostic group and each is described in more detail in the following subsections. The term "Simple" was adopted to reflect the relative complexity of this group compared with the other three groups

included in the study and to relate the study to the large clinical literature and guideline reports which use the same terminology (Baumgartner et al., 2010; Billett, Cowie, Gatzoulis, Vonder Muhll, & Majeed, 2008a; Srinathan et al., 2005; Warnes et al., 2001; Warnes et al., 2008).

1.4.1.1. Ventricular septal defect

Ventricular Septal Defect (VSD) is characterised by a hole between the two heart ventricles that leads to an increase in blood flow to the lungs (Figure 1.2). It may occur as an isolated defect or as part of a more complex defect, such as Tetralogy of Fallot (see section 1.4.2, page 32). VSD is the most common acyanotic CHD, accounting for 30 to 40% of all CHD cases as an isolated defect (Baumgartner et al., 2010). It is less common in the adult population due to the frequency of spontaneous closure (40-60% of children) (Popelová et al., 2008).

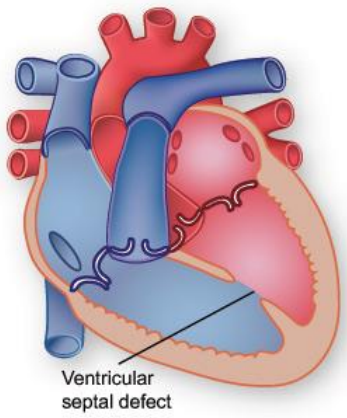

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Figure 1.2. Image representation of VSD

The natural history of VSD depends on the magnitude of the defect and the presence of pulmonary hypertension. Patients with small defects and no pulmonary

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hypertension are usually asymptomatic and do not require treatment, yet are typically at higher risk for complications (Silversides et al. 2010a). Most patients have a reparative surgery before adulthood. The most common surgical intervention is the use of a patch to cover the hole and requires an open-heart surgery and the use of cardiopulmonary bypass. The operative mortality rate for surgical repair is 1-2% and it offers a relatively good long-term outcome (Baumgartner et al., 2010). Less commonly used is the transcatheter intervention, where devices are inserted into the heart through catheters and is performed in a catheterisation lab. This procedure is performed for patients who are at high risk for surgical mortality or those with previous surgical interventions (Baumgartner et al., 2010).

The ten-year survival rate for unrepaired small VSD is 75% (Harris & Foster, 2009). The outlook for patients having the repair before the age of two is considered to be very good (Warnes et al., 2008).

1.4.1.2. Atrial septal defect

Atrial Septal Defect (ASD) is characterised by a hole between the two heart atria (Figure 1.3). In some cases the opening may develop during adulthood. This usually causes oxygen-rich blood from the left atrium to flow to the right atrium (shunt) thus increasing the pressure in the lungs (Popelová et al., 2008). ASD accounts for 9 to 11% of all CHD cases. The most common subtype (based on the location of the hole) is secundum Atrial Septal Defect (ASDII), which accounts for 75% of all ASD cases. Ostium primum ASD is less common accounting for approximately 15% of ASD cases.

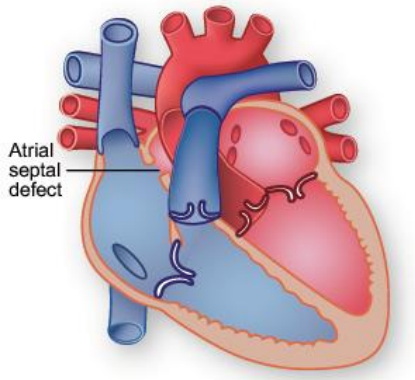

Figure 1.3. Image representation of ASD

Because of its asymptomatic nature ASD is usually diagnosed in adulthood (Baumgartner et al., 2010). A large unrepaired ASD may lead to symptoms such as reduced exercise capacity, palpitations, shortness of breath, arrhythmias, and heart failure during the fourth decade of life (Silversides et al., 2010a).

Depending on the size of the ASD, surgical closure through open-heart surgery may be required (Brickner, Hillis, & Lange, 2000). Reparative open-heart surgery has low operative mortality (1%) and patients generally have good long-term outcomes, especially if they do not experience other co-morbidities (Baumgartner et al., 2010). Transcatheter intervention is the common practice for ASDII closure and offers similar outcomes to surgical intervention (Silversides et al., 2010a).

The survival rates for unrepaired ASD beyond the age of 50-60 years do not exceed 50%, with subsequent attrition of about 6% per year (Perloff, 2009). The outlook for patients having the repair before the age of 25 is considered to be excellent, whereas patients having the repair after the age of 40 are at increased risk of long-term complications (Phillips, 2009).

1.4.1.3. Right ventricular outflow tract obstruction

Right Ventricular Outflow Tract Obstruction (RVOTO) is an obstruction in the blood flow of the right ventricle. The most common (90%) subtype is valvular Pulmonary Stenosis (PS). PS is characterised by a narrowing of the pulmonary valve and therefore a problematic blood flow to the lungs (Figure 1.4). PS is usually isolated but can also appear alongside other congenital defects. As an isolated defect, PS accounts for 10% of CHD cases (Foster & Lease, 2009).

The severity depends on blood flow rate, heart rate, and right ventricular function. Patients with mild PS may be asymptomatic, while severe PS may lead to shortness of breath, fatigue, or even chest pain (Brickner et al., 2000). Patients with mild PS may reach adulthood without requiring treatment, yet severe PS is usually treated in childhood (Popelová et al., 2008).

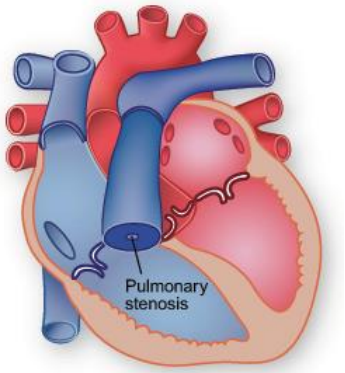

Figure 1.4. Image representation of PS

The preferred method of treatment of PS is transcatheter, with the aim to widen the narrowed part and improve blood flow. The alternative approach is surgical treatment, which is indicated for patients with associated defects and for whom transcatheter intervention is not possible (Baumgartner et al., 2010).

The survival rate for unrepaired mild PS is 95% at 20 years after diagnosis (Brickner et al., 2000). The long-term survival for repaired PS is considered excellent with rates close to the general population (97% survival at 25 years) (Popelová et al., 2008). However, long-term complications and reoperations are common in patients with PS (Warnes et al., 2008).

1.4.1.4. Left ventricular outflow tract obstruction

Left Ventricular Outflow Tract Obstruction (LVOTO) is an obstruction in the blood flow of the left ventricle. A number of subtypes may be categorised as LVOTO-specific, including valvular aortic stenosis and coarctation of the aorta.

Valvular Aortic Stenosis (AS) is the narrowing of the aortic valve, which leads to an obstruction of blood flow from the heart to the rest of the body (Figure 1.5). Depending on the position of the narrowing AS may also be Sub-Aortic (SAS) or Supravalvar (SVAS) (Aboulhosn & Child, 2006). AS accounts for 8% of all cases of CHD (Popelová et al., 2008).

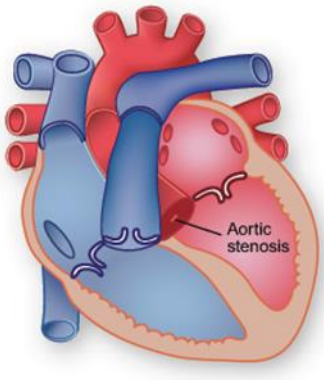

Figure 1.5. Image representation of AS

Symptoms may develop gradually in a period of 10 to 20 years and may include shortness of breath, chest pain, and syncope (Harris & Foster, 2009). Depending on

the severity and the experience of symptoms, patients may not need to undergo an intervention. Transcatheter and surgical approaches may be used as temporary palliative procedures in childhood, yet the only reparative treatment for AS is considered to be surgical Aortic Valve Replacement (AVR) (Popelová et al., 2008).

Unrepaired and asymptomatic patients are considered to have a normal life expectancy, but patients that develop symptoms in adulthood generally have reduced survival (Brickner et al., 2000). The mortality timeline for unrepaired and symptomatic patients is 5 years after chest pain symptoms, 3 years after syncope symptoms, and 2 years after heart failure symptoms develop (Brickner et al., 2000). The 25-year survival rate for repaired AS patients is 75% (Hoffman, Kaplan, & Liberthson, 2004). Owing to the progressive nature of AS, long-term complications are possible after repair (Silversides et al., 2010b).

Coarctation of the Aorta (CoA) is a narrowing (coarctation) of part of the main artery leading out of the heart (aorta) (Figure 1.6). CoA may present with other congenital heart defects (complex) as well as independently (simple) in 6 to 8% of all CHD cases (Gurvitz, 2009).

The degree of narrowing generally varies, and depending on severity, patients may not be diagnosed until adulthood. Patients usually present with symptoms such as headaches, shortness of breath, leg fatigue, and nose bleeding (Harris & Foster, 2009).

Most CoA patients typically have surgery to relieve narrowing soon after birth. Operative mortality for surgical intervention in neonates and infants is high (4-50%), while that for older children is low (0-5%) (Doshi & Syamasundar Rao, 2012).

Transcatheter approaches to treating CoA are less common (Aboulhosn & Child, 2006).

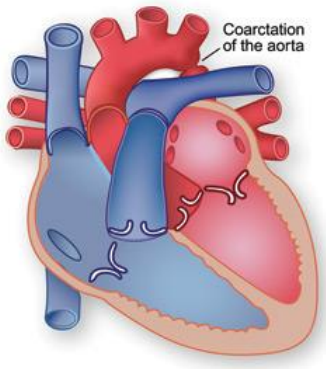

Figure 1.6. Image representation of CoA

The survival rate for unrepaired CoA is 10% at the age of 40 (Brickner et al., 2000). The 44-year survival rate for CoA repaired in childhood is approximately 73% (Popelová et al., 2008). Re-coarctation (recurring narrowing of the aorta) occurs in 5-10% of surgically treated patients. Patients with CoA are thought to be at risk of late complications (Popelová et al., 2008).

1.4.2. Tetralogy of Fallot

Tetralogy of Fallot (ToF) is characterised by the presence of four problems in the heart (Figure 1.7):

i) VSD: Hole between the two heart ventricles;

ii) Over-riding of the aorta: The artery that carries oxygen-rich blood to the body is shifted over the right ventricle and VSD, instead of exiting only from the left ventricle;

iii) Right ventricular outflow obstruction (PS): Narrowing of the valve and artery that connect the heart with the lungs;

iv) Right ventricular hypertrophy: Thickened wall of the right ventricle.

ToF is the most common *cyanotic* defect, prevalent in 10% of children after the age of 1 year (Therrien, 2009). ToF has different severity levels; the mild form does not typically cause cyanosis at birth ("pink Fallot") and it is sometimes diagnosed during adulthood (Silversides et al., 2010b). The classic form of ToF is characterised by cyanosis, and the severe form includes pulmonary atresia (under-developed pulmonary valve) (Popelová et al., 2008).

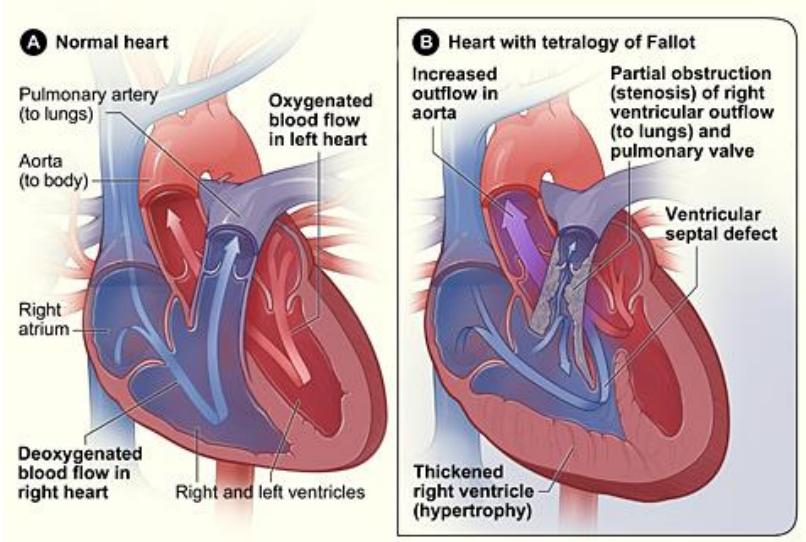

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Figure 1.7. Image representation of normal heart and heart with ToF

The majority of babies with ToF present cyanosis at birth and, if left untreated, they may experience symptoms ranging from hypoxic spells during childhood to stroke and endocarditis (see section 1.8, page 40) during adulthood (Brickner et al., 2000).

In the past, treatment for ToF involved a stepwise approach, with a temporary palliative intervention followed by a complete repair. The main goal of the palliative intervention is to increase pulmonary blood flow (Baumgartner et al., 2010). Due to

[2] NHLBI is part of the National Health Institute (NHI) and U.S. Department of Health and Human Services. The right to use images was granted through e-mail correspondence with the NHLBI. A copy of the correspondence can be found in Appendix B.

the long-term complications resulting from palliative procedures (Brickner et al., 2000) such stepwise approaches are increasingly being abandoned in favour of complete repair early in a baby's life. Typically, complete surgical repair is carried out in the first year of life (Therrien, 2009). The operative mortality for complete ToF repair is 1% (Baumgartner et al., 2010).

The survival rate for untreated ToF is very low with only 11% at age 20 years, 6% at age 30 years, and 3% at age 40 years (Brickner et al., 2000; Perloff, 2009). The survival rate after 36 years is approximately 85% for repaired patients (Warnes et al., 2008). Ten to fifteen percent of patients with ToF will eventually require re-operation and long-term complications are considered common (Silversides et al., 2010b).

1.4.3. Transposition of the great arteries

Transposition of the Great Arteries (TGA) is characterised by ventriculo-arterial discordance, with the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle (Figure 1.8). It is prevalent in 5% of all new-borns with CHD and it may present as an isolated defect or in complex forms with a VSD (Meijboom & Webb, 2009).

Cyanosis is usually evident within hours from birth and heart failure is common during the new-born period. Due to the high cyanosis-related mortality rate (90%) during the first year of life, it is generally uncommon for TGA to be left untreated (Brickner et al., 2000).

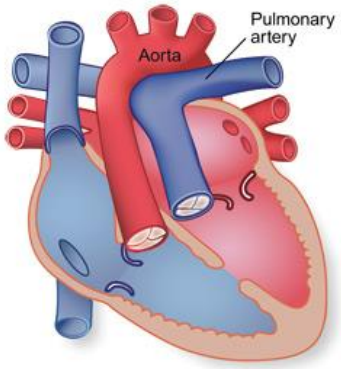


Figure 1.8. Image representation of TGA

Most patients undergo at least one operation during childhood. Critically hypoxic patients may initially undergo a transcatheter palliative procedure to improve blood mixing, yet ultimately surgical repair is required (Popelová et al., 2008). Until recently, the most common surgical procedures were the Mustard and Senning operations (atrial switch: creation of channels in atrial chambers to redirect blood flow), which were superseded by the Jatene operation (arterial switch: main arteries are switched to the right ventricles) around 1990. In complex cases of TGA with VSD and PS, the Rastelli operation (use of tubes to redirect blood flow) may be performed (Hornung, 2011). Most of the current adults with TGA have undergone one of the atrial switch operations. In the Mustard and Senning operations (using different techniques) the aim is to divert the blood and get oxygen rich blood to circulate around the body.

Atrial switch operations have a 75 to 85% survival rate at 25 years post-intervention, whereas data for the arterial switch operation are only currently beginning to emerge and suggest a 10-year survival rate of 85 to 97% (Villafañe et al., 2014). The Rastelli operation offers less favourable outcomes, with only 70% of patients surviving after 15 years (Silversides et al., 2010c). Long-term complications are considered

common after repair for TGA (Popelová et al., 2008). Arrhythmias may lead to the need for pacemaker insertion for up to 25% of patients (Meijboom & Webb, 2009).

1.4.4. Single ventricle physiology

The umbrella term "Single Ventricle" (SV) encompasses all the complex defects, which may range from cases where one of the two ventricles is missing or is underdeveloped resulting in a single functional ventricle (Hypoplastic Right Ventricle-HRV or Hypoplastic Left Ventricle-HLV) (Figure 1.9) to the closure or absence of one of the atrioventricular valves (Tricuspid Atresia-TA or Mitral Atresia-MA) (Figure 1.10). The common characteristic of all SV defects is that biventricular (two-ventricle circulation) repair is not typically feasible (Popelová et al., 2008). Patients are cyanotic from birth and with a median survival of 14 years, the prognosis for untreated SV is considered very poor (Silversides et al., 2010c).

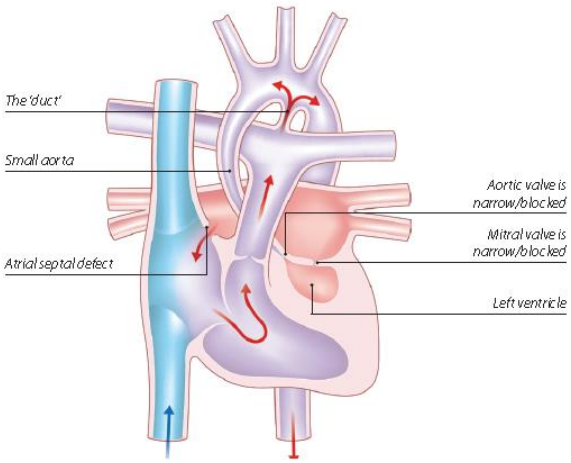


Figure 1.9. Image representation of HLV

[3] The right to use images was granted through e-mail correspondence with the British Heart Foundation. A copy of the correspondence can be found in Appendix C.

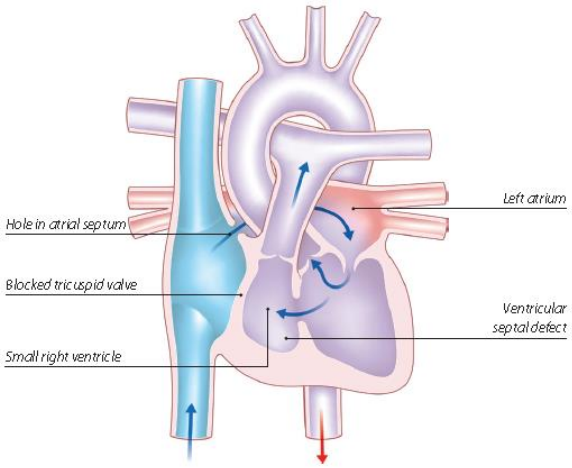


Figure 1.10. Image representation of TA

Most patients are treated in childhood with a stepwise procedure towards the Fontan operation. Depending on the level of pulmonary blood flow, patients undergo various temporary palliative procedures followed by the first stage of the Fontan operation (Gatzoulis & Swan, 2009). The second stage may involve different modifications of the Fontan procedure or the more recent Total Cavopulmonary Connection (TCPC). These procedures are generally aimed at separating systemic and pulmonary blood flow and therefore minimising cyanosis, but they are considered palliative (Gatzoulis, Swan, Therrien, & Pantely, 2005).

Research suggests that the survival rates after the Fontan operation are 86 to 94% at ten years and 82 to 87% at 15-20 years (Silversides et al., 2010c). SV defects have a multi-systemic character, which can result in serious complications (Warnes et al., 2008).

1.5. Epidemiology

CHD is the most common congenital birth defect and in the UK it is estimated that eight in 1,000 babies are born with CHD. This results in an incidence of 5,000 babies

with CHD in the UK every year (Cardio & Vascular Coalition [CVC], 2008). Recent official reports on the prevalence of CHD in the adult population are not available but estimations have indicated a population of approximately 185,000 adults in 2010, a figure that is expected to be higher at this time (Petersen et al., 2003). Moreover, the majority of people with ACHD are in the younger spectrum of adulthood (Watson & Rana, 2009).

In Europe there are approximately 1.2 million people with ACHD (Verheugt et al., 2010). Wren and O'Sullivan (2001) estimated that the adult population consists of approximately 28% complex, 54% moderate, and 18% minor defects, yet the percentages for moderate and complex defects including ToF, TGA, and SV are expected to rise (Department of Health [DoH], 2006).

1.6. Aetiology

In 80 to 90% of cases the causes of CHD still remain unclear but these cases appear to have multifactorial origin. Only 10 to 20% of cases are attributed to specific causes that have been identified (Buskens, Grobbee, Frohn-Mulder, Wladimiroff, & Hess, 1995). Some defects have a genetic basis, with higher incidence observed in babies whose parents (especially the mother) have CHD (Deanfield et al., 2003). Approximately 6 to 10% of all cases of CHD originate from chromosomal abnormalities (Buskens et al., 1995). For instance, higher rates of CHD are evident in babies with chromosomal abnormalities such as Down's syndrome (Petersen et al., 2003). Five to 10% of CHD cases are attributed to genetic disorders. Moreover, maternal diseases such as Type I diabetes, phenylketonuria, systemic lupus erythematosus and viral infections such as rubella account for about 1% of diagnosed cases of CHD (Bernier, Stefanescu, Samoukovic, & Tchervenkov, 2010). Finally,

another 1% of the cases are caused by environmental factors such as excessive maternal alcohol consumption, use of antiepileptic drugs, and use of other drugs such as lithium during pregnancy (EUROCAT, 2009). Increasing evidence suggests that maternal obesity may cause CHD and limited research indicates maternal smoking during pregnancy and air pollution as potential causal factors, yet more research is needed in these areas (Dolk, Loane, Garne, & EUROCAT, 2011).

1.7. Survival patterns and mortality rates

The medical and surgical advances of the past few decades in the diagnosis and treatment of CHD have led to increasing survival well into adulthood, especially for moderate and complex defects, including ToF, TGA, and SV. Specifically, the introduction of new diagnostic procedures, the improvement of intensive care, and the development of more efficient anaesthetic approaches are some of the factors that contributed to increased survival in CHD (Tan, 2006). In addition, the advances in surgical interventions and the increase in reparative surgeries being performed earlier in people's lives, have led to an adult population with different disease physiology and clinical outcomes than the earlier survivors (Warnes et al., 2001).

Currently, approximately 85% of people with CHD survive into adult life, compared with less than 20% fifty years ago (Deanfield et al., 2003). More recently, it has been reported that up to 90% of people with CHD may survive into adulthood (Stuart, 2012). Nonetheless, CHD leads to reduced life expectancy even with simple defects (Watson & Rana, 2009). The combined rate of perinatal mortality and pregnancy terminations in Europe is currently 0.7 per 1,000 births and is mainly attributed to complex defects, such as SV (EUROCAT, 2009).

In the adult population overall mortality rates are not very clear. However, the Euro Heart Survey, which involved 79 European centres, revealed that there is a 2.8% mortality rate after a five-year follow-up in a population of individuals with ACHD. The lowest and highest mortality rates were observed in the simple defects such as VSD and the more complex such as SV respectively (Engelfriet et al., 2005). Cardiovascular causes (chronic heart failure, sudden cardiac death) account for approximately 45% of deaths in people with ACHD (Zomer et al., 2012a), with increased mortality when late complications are present (Verheugt et al., 2010). Increasing survival and decreasing mortality rates have resulted in an adult population larger than the paediatric population, with the adult population expected to rise by 5% every year (Tan, 2006).

1.8. Long-term complications and healthcare utilisation

Most individuals with ACHD will be eventually faced with long-term complications resulting from an "ageing repaired heart" and will require further interventions due to recurring problems (Ellison et al., 2013). A study of surgeries conducted in an ACHD specialised unit in Birmingham indicated that reoperations account for 34.2% of all ACHD-related operations (Srinathan et al., 2005). Some of the most common reoperations in ACHD include AVR after previous AS surgery, Pulmonary Valve Replacement (PVR) after surgery for PS or ToF, and recurring narrowing of the aorta after surgery for CoA. Reoperations are generally less common for TGA and Fontan-operated (SV) adults (Shore & Prapa, 2011).

For these reasons, the majority of people with ACHD require long-term outpatient care by specialised cardiologists. Table 1.1 presents some of the most common long-

term complications in ACHD, their prevalence, people at risk, and current management strategies.

Many individuals with ACHD will eventually suffer from arrhythmias. As a late complication, arrhythmias occur in 18-20% of people with ACHD and are the most common cause of sudden cardiac death (52-84%) (Deal, 2009). Arrhythmias pose significant burden on individuals due to the frequent need for hospitalization and possible adverse effects of anti-arrhythmic medication (Triedman & Walsh, 2011).

Heart failure is characterized by symptoms of exercise intolerance in the presence of any abnormality in the structure and/or function of the heart (Dimopoulos, Alonso-Gonzalez, Thaulow, & Gatzoulis, 2011). Heart failure is the second most common cause of sudden cardiac death in ACHD (Deal, 2009).

Most individuals with ACHD are also vulnerable to Infective Endocarditis (IE), an infection in the heart. IE can lead to further complications, including heart failure, thus vulnerable groups are regularly monitored by blood testing (Child, Pegues, & Perloff, 2009).

Pulmonary Artery Hypertension (PAH) is defined as resting mean pulmonary artery pressures greater than 25mm Hg or exercise-induced pressures greater than 30mm Hg and is present in approximately 4% of people with ACHD (Owusu-Ansah, Lim, & Veldtman, 2009). An extreme manifestation of PAH is Eisenmenger Syndrome (ES), which occurs when pulmonary hypertension is presented with a persistent right-to-left shunt (reversal of blood flow) and cyanosis. PAH may lead to increased morbidity and mortality especially in people with complex defects (D'Alto & Diller, 2014).

Table 1.1. Common long-term complications in ACHD

Long-term complication	*Prevalence*	*People at high risk*	*Prevention*	*Treatment*
Arrhythmias	Most common complication evident in 18-20% of people with ACHD	- TGA, SV, VSD, ASD, ToF, AS	- Modifications and advancements in surgical techniques - Prophylactic arrhythmia surgery	- Anti-arrhythmic medication - Catheterisation procedures - Pacemakers - ICDs - Surgery
Heart failure	1/3 of people with ACHD experience exercise intolerance, the main feature of HF	- Cyanotic ACHD (especially those with PAH/ES) - SV (especially Fontan operated) - TGA (especially Mustard & Senning operated) - People with arrhythmias and ventricular dysfunction	- Prevention is associated with attempts to treat the underlying ACHD	- Heart (& lung) transplantation - Mechanical hearts - Palliative treatments (ICDs & pacemakers) - Medication, including diuretics (promote passing of urine), steroids, hypertension medication, anti-arrhythmic medication - Exercise training
Infective endocarditis	Accounts for 4% of admissions to a specialised ACHD service	- Previous IE - Complex cyanotic ACHD - Unrepaired CoA, VSD, PS, AS, ToF - Prosthetic valves and conduits - Surgically created shunts (blood flow pattern that deviates from the typical)	- Skin and oral hygiene - Antibiotic prophylaxis when undergoing surgical procedures, including oral surgery and natural childbirth	- Antibiotic medication - Anticoagulant medication (“blood thinners”) - Surgery
Pulmonary artery hypertension	Affects 4% of people with ACHD	- ASD, VSD, CoA, ToF, TGA, cyanotic ACHD	- Earlier repairs - Prevention is also associated with attempts to treat the underlying ACHD	- Hypertension medication - Oxygen therapy - Surgery - Heart (& lung) transplantation
High-risk pregnancy	Complications in 11% of pregnancies	- ES, TGA - Palliated or unrepaired cyanotic ACHD - Mechanical valves	- Risk assessment - Medication adjustment - Surgical repair of ACHD - Maternal and foetal monitoring - Delivery planning - Contraception	- Termination in already pregnant high-risk women - Elective delivery before 39 weeks gestation - Antibiotic prophylaxis during delivery for women with history of IE

Although pregnancy is believed to be well tolerated by women with ACHD, it is contraindicated for women with severe PAH/ES because of the high mortality risk (50%) (Warnes et al., 2008).

In addition to the above complications, ACHD may cause significant reduction in exercise capacity. Generally, competitive sports are contraindicated for cyanotic individuals (Warnes et al., 2008). The benefits of physical activity are well documented, yet the feasibility of safe exercise programs in ACHD has not been sufficiently studied. Recent trials indicate that tailored exercise training in people with ACHD can be both safe and effective (Dua, Cooper, Fox, & Graham Stuart, 2010; Findlow & Doyle, 1997; Holloway et al., 2011; Martínez-Quintana, Miranda-Calderín, Ugarte-Lopetegui, & Rodríguez-González, 2010; Thaulow & Fredriksen, 2004).

As mentioned earlier, chronic cyanosis can lead to multiple complications affecting a number of different organs. At the cardiovascular level cyanosis may lead to progressive heart failure, infective endocarditis, and syncope (Gatzoulis et al., 2005). People with cyanosis may also experience orthopaedic problems, renal dysfunction, scoliosis, gallstones, and bleeding (Petersen et al., 2003). Neurological complications have also been reported in people with cyanosis, resulting from stroke and cerebral abscesses (Gatzoulis et al., 2005). Effective management of chronic cyanosis is considered to be very important due to the increased morbidity and mortality risk.

The high morbidity in ACHD is evident in the increasing number of hospital admissions. In the UK, the proportion of all hospital admissions with a primary diagnosis of ACHD has increased from 21.5% in 1995/1996 to 27.3% in 2003/2004

in patients over 25 years (Billett, Majeed, Gatzoulis, & Cowie, 2008b). The main reasons for hospitalization are arrhythmias and interventional or diagnostic catheterization (British Cardiac Society Working Party, 2002). In addition, healthcare utilisation in people with ACHD is high, with increased number of GP visits and prescriptions compared with controls (Billett et al., 2008a).

1.9. Loss to follow-up

Despite the risk for long-term complications and the high healthcare utilisation in ACHD a significant number of people are currently not being followed-up at specialist ACHD centres and are therefore lost to follow-up (Wren & O'Sullivan, 2013). A study conducted in a single centre in Canada indicated that up to 47% of their cohort failed to receive outpatient cardiology follow-up between the age of 18 and 22. Lack of follow-up was also observed in 21% of people with severe defects (e.g. SV) (Mackie, Ionescu-Ittu, Therrien, Pilote, Abrahamowicz, & Marelli, 2009). In a Belgian study by Goossens et al. (2011) 10% of people did not receive follow-up at the recommended level of care. Both of these studies found that male sex and no prior surgery were associated with loss to follow-up. Wray, Frigiola, and Bull (2011) reported that 24% of people who had undergone repair of ToF in Great Ormond Street Hospital London since 1964 were lost to follow-up. The authors reported that 48% of late deaths occurred in this group of people. Reasons for loss to follow-up were assessed through telephone interviews and included lack of knowledge, anxiety, and ambivalence. It is likely that significant rates of people lost to follow-up also exist in other defects (Wray et al., 2011).

These data suggest that people who are currently followed-up in specialist adult clinics do not represent the whole spectrum of people with ACHD. Structured

transition from paediatric to adult clinics has been recommended in order to address this problem (Wren & O'Sullivan, 2013).

1.10. Summary

The increasing survival rates in babies born with CHD and the progression of this group into adulthood has led to the emergence of a new sub-specialty in cardiology, namely ACHD. People surviving well into their adult lives are faced with long-term complications and repeat interventions, which require them to attend regular appointments in specialist clinics. The chronic nature of ACHD poses significant challenges for people who are not only at high risk for mortality but also psychosocial consequences. The next chapter will review the evidence regarding the impact of ACHD on health-related quality of life by presenting the findings of the systematic review of the literature conducted as part of the present thesis.

CHAPTER 2 – IMPACT OF ADULT CONGENITAL HEART DISEASE ON HEALTH-RELATED QUALITY OF LIFE: A SYSTEMATIC REVIEW OF THE LITERATURE

2.1. Prologue

The first chapter provided the clinical context of the thesis by describing CHD, its treatment, and long-term complications. This chapter presents a systematic review[4] of studies that have examined HRQoL in people living with ACHD. The chapter begins with a brief overview of the increasing interest in patient-reported outcomes and especially HRQoL. Conceptualization and measurement issues in HRQoL research are then discussed followed by the rationale for the systematic review. The final section of this chapter consists of a description and discussion of the findings of the systematic review.

2.2. Background

2.2.1. Patient-reported outcome measures in chronic illness

The growing population of people surviving into adulthood resulted in the recognition of ACHD as a chronic illness. Chronic illnesses are "prolonged, do not resolve spontaneously, and are rarely cured completely" (National Center for Chronic Disease Prevention and Health Promotion [NCCDPHP], 2009). The increased survival has led to a shift of focus towards endpoints beyond mortality. People with chronic illnesses are progressively faced with not just physical but also psychosocial challenges resulting from their illness (Petrie & Revenson, 2005).

[4] The systematic review was published in February 2013 (Fteropoulli, Stygall, Cullen, Deanfield, & Newman, 2013) (see Appendix D).

This shift of focus is reflected in the widespread introduction of Patient-Reported Outcome Measures (PROMs) in evaluating the effects of healthcare. It links clearly to the UK DoH and the National Health Service (NHS) generic guidelines for the adoption of PROMs, in which the routine assessment of psychosocial outcomes and especially HRQoL in healthcare is promoted (DoH, 2008). The assessment of HRQoL outcomes is important because it is considered a major indicator of adjustment to chronic illness (Hoyt & Stanton, 2012).

2.2.2. Conceptualizing health-related quality of life

The concept of quality of life (QoL) and its definition has been the focus of a vast number of studies. The prefix "health-related" is now used to distinguish quality of life measurements carried out in the medical context of health and illness (Testa & Nackley, 1994). Nevertheless, a clear and agreed definition of health-related quality of life remains largely elusive. Although investigators disagree as to what domains or dimensions constitute the concept, there is an agreement that HRQoL is a complex, subjective, and multi-dimensional concept (Fayers & Machin, 2009).

The conceptualization of life domains that comprise HRQoL has been primarily based on the World Health Organisation's (WHO) definition of health: "*a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity*" (WHO, 1948). In the present thesis, the definition by the International Society for Quality of Life Research (ISOQOL) was adopted, whereby HRQoL is the functional consequences of a condition and its treatment on a person and encompasses physical, occupational, psychological, interpersonal, and somatic sensation elements (Schipper, Clinch, & Olweny, 1996). However, it should be noted that QoL and HRQoL are not defined well in the ACHD literature. Therefore,

the systematic review presented in this chapter aimed to be over-inclusive by studying (HR)QoL in general (see section 2.4.2.2, page 53).

2.2.3. Measuring health-related quality of life

Much as its definition, the measurement of HRQoL has been a source of disagreement. Generally, there are three approaches in measuring HRQoL, namely generic, disease-specific, and individual.

Generic measures can be used beyond the bounds of illness and disability and many were initially developed to assess HRQoL in healthy populations (Fayers & Machin, 2009). These measures may include multiple items that address various dimensions such as physical, emotional, and social functioning. The most widely used generic measure is the short-form 36 (SF-36) (Ware, Snow, Kosinski, & Gandek, 1993). One of the advantages of such generic measures is that they allow comparisons between the target patient group and normative data that has been acquired from the general population. The generic nature of such measures carries some inherent limitations, such as limited sensitivity and responsiveness to changes in HRQoL, specific to various illnesses (Guyatt, Feeny, & Patrick, 1993).

Some of the limitations of generic measures are potentially addressed by disease-specific measures, which are specific to an illness, its associated symptoms, and challenges. These measures focus on specific aspects of peoples' lives that are considered to be influenced by their illness and therefore in many cases are assumed to be more responsive to changes in the condition (Fletcher, Gore, Jones, Fitzpatrick, Spiegelhalter, & Cox, 1992). Their limitation is that they cannot be used to compare HRQoL between different illnesses and with the general population. At present, there is only one disease-specific measure for ACHD, namely the Congenital Heart

Disease-TNO-AZL Adult Quality of Life (CHD-TAAQOL) questionnaire (Kamphuis et al., 2004). Details of this measure are discussed in Chapter 5.

Individual measures of HRQoL have been developed in order to capture what is most important from the patients' perspective. In such measures, people nominate the most significant areas of their lives and rate their functioning in these areas. One such measure is the Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQoL-DW) (Hickey et al., 1996). Although these measures offer personalised insight about people's HRQoL, they are time consuming and do not allow comparisons between people because the areas of the lives that are important are determined by the individual (Wood-Dauphinee, 1999).

Generally, it is suggested that both generic and disease-specific measures are included in studies to capture comparisons to other conditions and normative data of a healthy population and also to be sensitive to the specific symptoms of the condition under study (Fletcher et al., 1992).

2.2.4. Rationale for systematic review

HRQoL has become one of the major PROMs in ACHD research (Moons, 2010). When the present study commenced, studies on HRQoL had been reviewed in four papers all of which focused in children and adolescents with CHD. Two were systematic reviews and two were overviews of the literature. The earliest systematic review was published in 2009 and focused on psychological adjustment and HRQoL in children and adolescents after open-heart surgery (Latal, Helfricht, Fischer, Bauersfeld, & Landolt, 2009). The second systematic review described and compared the HRQoL of children and young adults that were either born pre-term or born with CHD (Dahan-Oliel, Majnemer, & Mazer, 2011). Of the two overviews one

was an overview of neuropsychological, psychosocial, and HRQoL outcomes in children and adolescents with CHD (Bellinger & Newburger, 2010), while the other described the measurement of HRQoL and the status of the literature in children with CHD (Marino, Uzark, Ittenbach, & Drotar, 2010).

The reviews reported mixed findings in the literature with some of the reviewed studies indicating impaired HRQoL in children and adolescents with CHD, while others did not support this notion. There were also differences between proxy and self-reported HRQoL with parents usually reporting greater impairment compared to the children themselves (Latal et al., 2009). The difficulty of using proxy and self-reported HRQoL assessments has been identified in the overviews by Bellinger and Newburger (2010) and Dahan-Oliel et al. (2011).

Disease complexity was associated with poorer HRQoL in the review by Dahan-Oliel (2011). Latal et al. (2009) did not support this conclusion but reported that other clinical characteristics such as medication and co-morbidities were associated with impaired proxy-reported HRQoL (Latal et al., 2009). The review by Marino et al. (2010) also identified the methodological limitations of the child literature, which included small samples, variability of measures, and discrepancies between patient and proxy assessments.

One literature review which was published in 2005 focused on the biopsychosocial experiences of the adult population, including HRQoL (Kovacs, Sears, & Saidi, 2005). Similarly to the reviews in the child and adolescent literature, this study reported mixed findings on the HRQoL of people with ACHD. In addition, the impact of disease complexity on HRQoL in the adult population remained unclear.

The review by Kovacs et al. (2005) was not specifically focused on HRQoL in ACHD and identified only a small number of studies (*n*= 8). Although some of the studies included measures of disease complexity and other clinical characteristics, the review provided only a brief summary about their impact on HRQoL and did not examine these associations in detail. In addition, since the publication of the review by Kovacs et al. (2005) several new studies had been published. A more detailed examination of HRQoL in people with ACHD and the role of disease complexity and clinical factors was therefore considered necessary to gain an up to date and comprehensive understanding of this area of work.

2.3. Aims of the systematic review

The focus of the systematic review was HRQoL, thus papers that were restricted to an examination of psychosocial factors and did not include HRQoL were not considered. The literature on psychosocial variables, change over time in these variables, and their relationship with HRQoL is addressed in Chapter 3 within the context of the theoretical framework of the thesis. The aims of the systematic review were:

1) To examine the impact of ACHD on the HRQoL of patients compared with healthy controls and/or normative data.
2) To establish whether disease complexity and clinical variables are associated with HRQoL in ACHD.

2.4. Methodology

In accordance with reporting guidelines for systematic reviews, a Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) checklist (Moher, Liberati, Tetzlaff, & Altman, 2009) can be found in Appendix E. According

to the PRISMA checklist, the report consists of methodological aspects including the search strategy, eligibility criteria (e.g. participants, comparisons, outcomes, and study design), information sources, selection process, plan of analysis, synthesis of the results, and quality assessment. The report also includes a flow diagram of the selection process, results of individual studies in table form, summary of evidence, and the limitations of the included studies and the systematic review.

2.4.1. Search strategy

The electronic databases outlined below were searched from inception through to November 2011.

- Science Direct
- Pubmed
- Embase, Amed, and PsycInfo through OVIDSP
- CINAHL Plus and Medline through EBSCO

Articles published after the review completion, between November 2011 and August 2014, were also identified and the findings from these studies are summarised in the results section. The findings of newer articles identified in 2014 did not differ from the overall conclusions of the review completed in 2011. The synthesis of the findings includes the studies identified in the original search and recent studies are addressed in an addendum.

A scoping search revealed that studies in ACHD did not distinguish between QoL and HRQoL. For this reason, and to avoid excluding relevant studies, the general term *quality of life* was used to search the literature. The search did not include terms relating to specific indices of (HR)QoL (e.g. social functioning, mood, see section 2.4.2.2, page 53). Initial search terms were specific, (e.g. “quality of life AND

GUCH") but as the search did not retrieve many relevant articles, it was broadened by using various word combinations i.e. *("quality of life" OR "life satisfaction" OR "well being") AND (GUCH OR ACHD OR congenital heart disease OR congenital cardiac disease OR congenital heart defect OR congenital cardiac defect)*. Where the search yielded too many results, additional filter terms were used (e.g. *AND adult).* In addition, the bibliographies of selected articles were reviewed for relevant titles. The search strategy was conducted in a single stage and it was the same for all electronic databases. An example of the full electronic search strategy for one of the databases is provided in Appendix F.

2.4.2. Inclusion and exclusion criteria

2.4.2.1. Publication type

Articles written in English and published in peer-reviewed journals were included.

2.4.2.2. Study design and purpose

Studies were included if they used quantitative methods to measure HR(QoL), while qualitative studies, literature reviews, meta-analyses, opinion articles, commentaries, and reports were excluded. Studies were included if their purpose was to: a) describe the patients' (HR)QoL compared with normative data or healthy controls and/or b) to study associations between disease complexity or individual clinical variables and (HR)QoL. Studies were excluded if their purpose was to study associations between psychosocial variables and HR(QoL). As mentioned earlier, (HR)QoL is not well-defined in the ACHD literature. Thus, the review aimed to examine the literature on (HR)QoL in general and be over-inclusive during searches. Only studies that mentioned and described (HR)QoL were included and those that *solely* described what may be considered related components of (HR)QoL or other PROMs (e.g.

cognitive functioning, mood, social functioning) without referring to (HR)QoL were excluded. For example, a study was included if it presented data on cognitive functioning as part of its (HR)QoL measurement but a study was excluded if it described cognitive functioning independently outside the context of (HR)QoL.

2.4.2.3. Measures

Only studies using (HR)QoL measures validated in previous research with adult samples were included. These measures included validated multidimensional questionnaires and single-item measures assessing (HR)QoL. Studies that used measures of other PROMs such as social functioning, mood, or measures of psychosocial functioning were excluded (see section 2.4.2.2, page 53). Studies were excluded if they used non-validated measures such as author-designed single-item measures or question sets.

2.4.2.4. Participants

People with ACHD were defined as those aged ≥14 years. Initially age was set at ≥16 years; however, it was changed to ≥14 to reflect the increasing number of studies that include younger individuals in primarily adult samples. Such studies were included if they provided data on HRQoL using questionnaires designed for adults (see section 2.4.2.3). Furthermore, when studies with mixed samples distinguished between paediatric and adult samples in terms of (HR)QoL assessment and reporting, only the adult data was included in the systematic review. Studies that were exclusively on paediatric samples and assessed (HR)QoL using paediatric measures were excluded as the review was focused on adults. Studies were excluded if they consisted solely of Marfan syndrome samples. Marfan syndrome is a hereditary connective tissue disorder that can affect the heart among other body

organs in 80% of cases (Child, 1997), hence it is not considered to be a congenital heart defect exclusively.

2.4.3. Assessment of relevance

Relevance was assessed based on the article title, then on the abstracts, and finally by reviewing full-text articles. An initial review of the titles was conducted by the researcher and articles were excluded if the condition under study was not relevant (i.e. other medical conditions such as coronary heart disease). Articles were retained for abstract assessment when there was uncertainty about their relevancy based on title alone. The abstracts were then assessed using the eligibility criteria by the researcher along with a member of the supervisory team. When there was uncertainty about the relevance of articles these were discussed and cross-checked by the whole supervisory team using the inclusion and exclusion criteria. Overall, there was agreement in the assessment of article relevance and there were no grey areas in deciding the relevance of articles because the search was intended to be over-inclusive and it was clear whether the articles were on the topic or not.

2.4.4. Analysis plan

A quantitative meta-analysis was not conducted as part of this review. A narrative synthesis was considered appropriate due to the anticipated heterogeneity among studies in terms of participant characteristics, outcome measures and study design. The extracted information was tabulated for ease of synthesis of the data.

2.4.5. Data extraction and synthesis

A data extraction form (Appendix G) was used to record general article information (title, author(s), and source), sample characteristics (target population, inclusion/exclusion criteria, and demographic information for the ACHD group and

the normative data/control group), study characteristics (aim(s), design, (HR)QoL definition, measures, and statistical analysis), and main findings.

For the purpose of describing the findings, the scales or subscales of each (HR)QoL instrument used in the studies were categorised into three broad domains, namely physical, psychosocial, and environmental/occupational HRQoL and related subdomains (Table 2.1). Therefore, Table 2.1 represents the measurement rather than the operationalization of (HR)QoL in the studies. The categorisation was conducted by the researcher and cross-checked by the supervisory team. Physical HRQoL included all scales or subscales measuring concepts relating to physical functioning and physical health, while psychosocial HRQoL included those that measured concepts relating to psychological and social functioning, and mental health. Environmental/occupational HRQoL included all scales and subscales measuring physical and home environment, access to healthcare, daily activities, work, and financial resources.

Table 2.1. Categorisation of scales and subscales into domains of HRQoL for the systematic review data synthesis

Measure	*Physical HRQoL*									*Psychosocial HRQoL*							*Environmental/ occupational HRQoL*
	Physical functioning	Role physical	Pain	General health	Symptoms	Gross motor functioning	Fine motor functioning	Cognitive functioning	Sleep	Psychological/ mental health	Role emotional	Vitality	Happiness	Sexual functioning	Social functioning		Environment & daily activities
SF-36	✓	✓	✓	✓						✓	✓	✓			✓		
WHOQOL-Bref	✓									✓					✓		✓
DUKE	✓		✓	✓						✓					✓		
SIP	✓								✓	✓							✓
TAAQOL			✓			✓		✓		✓		✓	✓	✓	✓		✓
SQoL										✓					✓		✓
CHD-TAAQOL					✓												
LAS				✓													
SEIQoL-DW				✓													
GBB					✓												

WHOQOL-Bref=World Health Organization Quality of Life- Bref, DUKE=DUKE Health Profile, SIP=Sickness Impact Profile, TAAQOL=TNO-AZL Adult's Health-Related Quality of Life, SQoL=Subjective Quality of Life, LAS=Linear Analogue Scale, GBB=Giessener Complaints Questionnaire.

2.4.6. Quality assessment

To assess the quality of each study a checklist was created based on the Quality Index proposed by Downs & Black (1998). The checklist assessed the quality of a study in four areas: reporting, external validity, bias, and selection bias (Appendix H). Each item on the scale was given a score of 2, 1 or 0. The quality of each study was calculated by summing up the score points. The best possible score was 36, with 0-9= poor quality, 10-18= low quality, 19-27= moderate quality and 28-36= high quality.

2.5. Results

2.5.1. Identified articles

A total of 431 potential articles were identified. The details of the article selection process are presented in Figure 2.1. After exclusions based on title relevance, the abstracts of 201 articles were examined. After further exclusions, the full-texts of the remaining 58 articles were examined in detail for inclusion in the systematic review. In addition, examination of the bibliographies of these articles resulted in 3 relevant articles. A total of 31 articles were included in the systematic review.

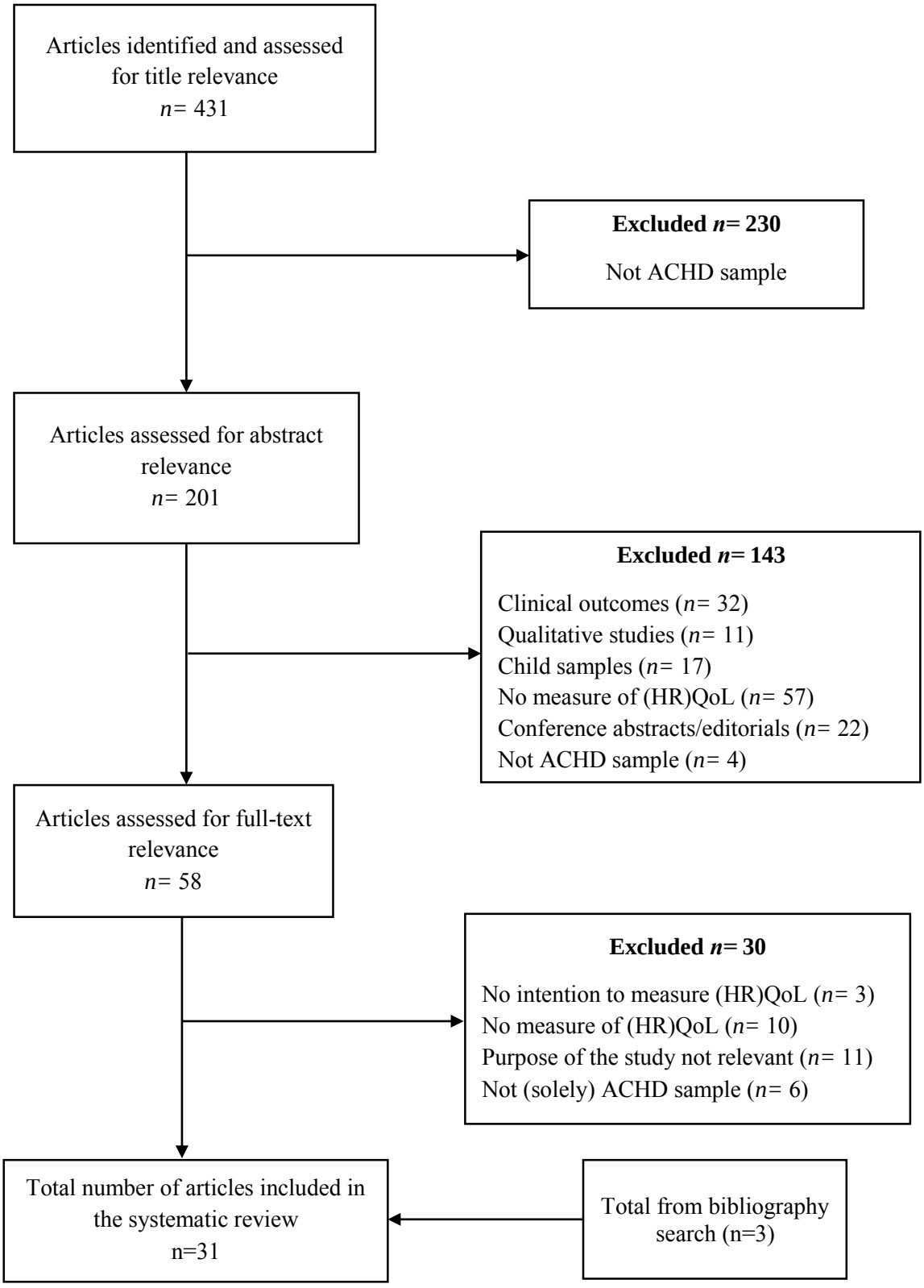


Figure 2.1. Flow chart of article selection for the systematic review

2.5.2. Article characteristics

The characteristics of all reviewed studies are presented in Table 2.2. Twenty-six studies examined HRQoL in people with ACHD in comparison with normative data or healthy controls and 22 examined the relationship between disease complexity or clinical variables and HRQoL. The reviewed articles were published between 2001 and 2011. The majority of studies originated from Europe (*n*= 23, 74%). The remaining originated from North America (*n*= 6, 19%) and Asia (*n*= 2, 7%).

2.5.3. Study quality

The methodological quality of all studies is reported in Table 2.2. The majority of studies (*n*= 27, 87%) were of moderate quality, while the remaining (*n*= 4, 13%) were low. The main reasons for low quality were poor reporting and insufficient sample size. Most studies failed to report whether the sample size resulted from power calculations, thus it is not known whether the studies had sufficient power to answer the research questions. Some studies did not clearly describe the participant selection process, making it difficult to assess whether the individuals that took part were a representative sample of the population they were recruited from. Of particular note is the study by Ternestedt et al. (2001) in which the statistical analysis was not described. It was therefore difficult to evaluate the validity of the findings. This study and those by van den Bosch et al. (2004), Cohen, Daniela, Yalonetsky, Gagin, and Lorber (2010), and Jefferies, Noonan, Keller, Wilson, and Griffith (2004) were rated as low quality due to poor methodological reporting and small sample size.

Table 2.2. Characteristics of the studies included in the systematic review

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	*Diagnosis or patient group*			
1	Bol Raap et al. (2007)	Retrospective 13yrs follow-up (QoL cross-sectional)	*N*= 28 (Follow-up *N*= 25) Mean age (at surgery): 34 (19-50) Gender: 15 male, 13 female	All VSD	Normative data *N*= 4410	TAAQOL	Moderate (22)
2	van den Bosch et al. (2004)	Cross-sectional	*N*= 22 for (HR)QoL assessment (total *N*= 36) Mean age (at operation): 12 (2-34) Gender: 18 male, 18 female	21 TA, 9 DILV, 6 other	Normative data	SF-36	Low (18)
3	Bruto et al. (2007)	Prospective (cross-sectional)	*N*= 912 Mean age: 30 (11) Gender: 474 male, 438 female	Obstructive lesions (79 CoA, 67 AS), shunt lesions (99 VSD, 55 ASD, 45 AVSD), 9 complex acyanotic, complex cyanotic (169 ToF, 87 TGA) and other	Normative data	SF-36	Moderate (22)
4	Chen et al. (2011)	Prospective (cross-sectional)	*N*= 289 Mean age: 33.2 (10.6) Gender: 105 male, 184 female	*86 ASDII, 64 VSD, 60 ToF	Controls age- and gender-matched	WHOQOL-Bref-Taiwan	Moderate (27)
5	Cohen et al. (2010)	Cross-sectional	*N*= 27 Mean age: 68.6 (5.7, 60-87) Gender: 10 male, 17 female	All ASDII	Controls age- and gender-matched *N*=27 Mean age: 69.9(±8.2, 60-78) Gender: 10 male, 17 female	CHD-TAAQOL only symptoms subscale	Low (17)

*Only conditions diagnosed in more than 10% of the sample were listed and compared
S.D.= Standard Deviation, DILV=Double Inlet Left Ventricle, AVSD=Atrioventricular Septal Defect.

Table 2.2 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
6	Daliento et al. (2005)	Cross-sectional	*N*= 54 Mean age: 32 (4) Gender: 24 male, 30 female	All ToF	Normative data	SF-36	Moderate (22)
7	Ebenroth & Hurwitz (2007)	Longitudinal 10yrs follow-up	*N*= 35 Mean age: 25.4 (19-37) Gender: 23 male, 12 female	All TGA	Normative data	SF-36 only physical component	Moderate (24)
8	Gratz et al. (2009)	Cross-sectional	*N*= 564 Median age: 24 (14-37) Gender: 309 male, 255 female	32 cyanotic, 31 Fontan circulation, 98 TGA with arterial switch, 38 TGA no arterial switch, 96 ToF, 47 Ebstein anomaly, 33 PS/PR, 66 left-heart obstruction AS/CoA, 62 isolated shunt, 61 other	Normative data and controls *N*=53 Median age: 25 (14-57) Gender: 33 male, 18 female	SF-36	Moderate (24)
9	Hager & Hess (2005)	Prospective (cross-sectional)	*N*= 149 Median age: 23.8 (14-59.8) Gender: 89 male, 60 female	13 SV, 47 TGA, 9 CCTGA, 32 ToF, 7 CoA, 3 AS, 6 PS, 7 Ebstein's anomaly, 5 VSD, 3 ASD, 14 other	Normative data	SF-36	Moderate (22)
10	Immer et al. (2005)	Cross-sectional	*N*= 233 [(HR)QoL-*N*= 154] Mean age: 35 (16) Gender: n/a	125 ASD, 34 AVD/PVD, 21 CoA, 17 ToF/TGA (cyanotic), 7 AVSD	Normative data age- and gender-matched	SF-36	Moderate (22)
11	Irtel et al. (2005)	Cross-sectional	*N*= 67 Median age: 25 (16-62) Gender: 39 male, 28 female	32 TGA, 35 ToF	Normative data age- and gender-matched	SF-36	Moderate (24)

PR=Pulmonary Regurgitation, CCTGA=Congenitally Corrected TGA, AVD=Atrial Valve Diseases, PVD=Pulmonary Valve Diseases.

Table 2.2 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
12	Jefferies et al. (2004)	Cross-sectional	*N*= 32 Mean age: 27 (9) Gender: 15 male, 17 female	17 acyanotic (3 ASD, 2 MVP, 2 Marfan, 3 VSD, 3 AS, 2 CoA, 2 PS) & 15 cyanotic (4 ToF, 3 TGA, 5 SV, 2 Down Syndrome, 1 TA)	Normative data from two studies age-matched	SF-36	Low (17)
13	Kamphuis et al. (2002a)	Cross-sectional	*N*= 82 with mild CHD Mean age: 24.6 (17-32) Gender: 31 male, 51 female	20 VSD, 7 PS, 6 ASD, 6 AS, 3 BAV, 2 APVD, 1 MVP, 37 spontaneous resolution	Controls *N*= 361 (SF-36) *N*= 831 (TAAQOL)	TAAQOL & SF-36	Moderate (25)
14	Kamphuis et al. (2002b)	Cross-sectional	*N*= 78 with complex CHD Mean age: 24.3 (18-32) Gender: 44 male, 34 female	Surgery type: 39 SRV, 23 conduit or mechanical prosthesis, 11 SV, 5 palliative operation shunt	Controls age- and gender-matched *N*= 361 (SF-36) *N*= 831 (TAAQOL)	TAAQOL & SF-36	Moderate (25)
15	Lane et al. (2002)	Cross-sectional	*N*=276 Median age: 31 (16-85) Gender: 41.7% male, 58.3% female	Treatment groups: 68 cured, 105 corrected, 23 palliated, 70 medical, 10 inoperable	Normative data	SF-36	Moderate (24)
16	Loup et al. (2009)	Cross-sectional	*N*= 153 Mean age: 26 (11) Gender: 94 male, 59 female	43 ToF, 59 TGA, 51 VSD	Normative data age- and gender-matched	SF-36	Moderate (21)
17	Lu et al. (2010)	Cross-sectional	*N*= 62 Median age: 28.5 (14-69) Gender: 37 male, 25 female	All ToF	Normative data	SF-36	Moderate (22)

MVP=Mitral Valve Prolapse, BAV=Bifoliate Aortic Valve, APVD=Abnormal Pulmonary Venus Drainage, SRV=Systemic Right Ventricle.

Table 2.2 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
18	Mokhles et al. (2011)	Cross-sectional	*N*= 509 Mean age: 19 (0-66) (SF-36 ≥14yrs) Gender: 301 male, 208 female	170 AVD, 152 ToF, 63 PA/ PS or VSD, 51 TGA with PA or PS, 26 CAT, 26 PA/PS with intact septum, 3 AA with biventricular heart	Normative data	SF-36	Moderate (22)
19	Moons et al. (2004)	Cross-sectional	*N*= 89 Median age: 24 (20- 26.5) Gender: 52 male, 37 female	76.4% simple TGA, 23.6% complex TGA	N/A	LAS & CHD-TAAQOL QoL determinants	Moderate (22)
20	*Moons et al. (2006)	Cross-sectional	*N*= 404 Median age: 13 (18-56) Gender: 221 male, 183 female	79 VSD, 68 ToF, 62 CoA, 42 AS, 31 PS, 20 TGA, 17 mixed AVD, 17 ASDII, 15 mitral insufficiency, 12 SV, 7 DORV, 34 other	Controls age- and gender-matched *N*= 404 Median age: 23 (18-58) Gender: 221 male, 183 female	LAS & CHD-TAAQOL QoL determinants	Moderate (24)
21	*Moons et al. (2005)	Cross-sectional	*N*= 629 Median age: 24 (18-66) Gender: 378 male, 251 female	112 ToF, 108 VSD, 89 CoA, 65 AS, 48 PS, 37 TGA, 170 other	N/A	LAS & SEIQoL-DW	Moderate (23)
22	Müller et al. (2011)	Cross-sectional	*N*= 58 Median age: 27.9 Gender: 28 male, 30 female	23 PS group (9 DILV, 5 TGA, 5 CCTGA+VSD, 1 TA, 1 PA, 1 HLHS, 1 AVSD), 35 ES group (21 VSD, 9 ASD, 6 AVSD)	Normative data	SF-36	Moderate (23)

*Studies with common sample.

PA=Pulmonary Atresia, CAT=Common Arterial Trunk, AA=Aortic Atresia, DORV=Double Outlet Right Ventricle, HLHS=Hypoplastic Left Heart Syndrome, ES=Eisenmenger Syndrome.

. **Table 2.2** ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
23	Rietveld et al. (2002)	Cross-sectional	*N*= 82 Mean age: 30.2 (17-77) Gender: 40 male, 42 female	13 mild CHD (small VSD, ASDII, mild PS), 44 moderate (CoA, ToF, AS), 25 severe (Fontan, Rastelli, Mustard, inoperable)	N/A	SF-36	Moderate (19)
24	Rose et al. (2005)	Cross-sectional	*N*= 111 Mean age: 33 (12) Gender: 56 male, 55 female	21 ToF, 16 ASD, 12 VSD, 11 AS, 10 CoA, 7 TGA, 6 aortic insufficiency, 5 PR, 4 PDA, 4 ES, 15 other	Normative data	WHOQOL-Bref & GBB	Moderate (19)
25	Saliba et al. (2001)	Cross-sectional	*N*= 67 Enrolled= 89 Mean age: 22.7 (17-49) Gender: 42 male, 47 female	35 TA, 13 MA, 24 DIV, 17 other	Normative data	Duke Heart Profile	Moderate (22)
26	†Simko & McGinnis (2003)	Cross-sectional, case-control	*N*= 124 Mean age: 26.4 Gender: 54 male, 70 female	23 ToF, 21 VSD, 11 CoA, 20 TGA, 6 SV, 12 ASD, 7 TA, 10 AS, 14 PS	Controls age-, gender-, race- & socio-economic status-matched *N*= 124 Mean age: 26.5 Gender: 54 male, 70 female	SIP	Moderate (21)
27	†Simko & McGinnis (2005)	Cross-sectional	*N*= 124 Mean age: 26 (8.5) Gender: 54 male, 70 female	cyanotic (23 ToF, 20 TGA, 6 SV, 7 TA) & acyanotic (21 VSD, 11 CoA, 12 ASD, 10 AS, 14 PS)	Controls age- and gender-matched *N*= 124 Mean age: 26.5 Gender: 54 male, 70 female	SIP	Moderate (23)

†Studies with common sample.
DIV=Double Inlet Ventricle.

Table 2.2 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
28	Ternestedt et al. (2001)	Longitudinal (20 & 30yrs post-surgery)	*N*= 26 Mean age: 20yrs: 28.7, 30yrs: 38.7 Gender: 15 male, 11 female	¥12 ToF, 14 ASD	N/A	SQoL	Low (13)
29	Vandekerckhove et al. (2009)	Cross-sectional	*N*= 39 Median age: 19.9 (15.8-28.1) Gender: 29 male, 10 female	24 simple TGA, 7 TGA+VSD, 4 Taussig-Bing anomaly, 4 TGA/VSD with arch obstruction	Normative data	TAAQOL	Moderate (19)
30	Winter et al. (2008)	Cross-sectional	*N*= 47 Mean age: 35 (21-69) Gender: 20 male, 17 female	All SRV (31 TGA, 16 CCTGA)	Normative data age- and gender-matched	SF-36	Moderate (22)
31	Winter et al. (2010)	Cross-sectional	*N*= 133 Mean age: 38 (18-75) Gender: 69 male, 64 female	43 CoA, 42 TGA, 36 Marfan, 12 ES	Normative data age- and gender-matched	SF-36	Moderate (22)

¥Diagnostic groups age- and gender-matched at operation and follow-up.

2.5.4. Study design and sample demographics

Most studies (*n*= 29, 94%) were cross-sectional and 2 were longitudinal. All but four studies (*n*= 27, 87%) included a comparison group of either normative data or healthy controls. The number of participants ranged from 22-912 and more than half of the studies (*n*= 17, 55%) had fewer than 100 participants. Age ranged from 14-85 years and gender was roughly balanced within the studies. The majority (*n*= 23, 74%) included people with a range of diagnoses, 7 (23%) included people with a single diagnosis and 1 (3%) categorised people in terms of the treatment they had received.

2.5.5. (HR)QoL instruments

A variety of instruments were used (Table 2.2). Generic instruments included the SF-36 (*n*= 19), TAAQOL (*n*= 4), SIP (*n*= 2), WHOQOL-Bref (*n*=2), Duke Health Profile (*n*= 1), and Subjective Quality of Life (SQoL, *n*= 1). Single-item generic QoL was assessed by the Linear Analogue Scale (LAS, *n*= 2). Disease-specific and individual measures included the CHD-TAAQOL (*n*= 2) and the Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQoL-DW, *n*= 1) respectively.

2.5.6. The impact of ACHD on HRQoL

Table 2.3 presents the findings of the 26 studies that compared the HRQoL of people with ACHD with either normative data or healthy controls.

Table 2.3. Studies that examined HRQoL in people with ACHD in relation to a comparison group

Authors (Year)	*Physical HRQoL*									*Psychosocial HRQoL*							*Environmental/ occupational HRQoL*
	Physical functioning	*Role physical*	*Pain*	*General health*	*Symptoms*	*Gross motor functioning*	*Fine motor functioning*	*Cognitive functioning*	*Sleep*	*Psychological/ mental health*	*Role emotional*	*Vitality*	*Happiness*	*Sexual functioning*	*Social functioning*	*Environment & daily activities*
Bol Raap et al. (2007)			=			=	=	↓	↓	=		=	=	=	=	=
van den Bosch et al. (2004)	↓	↓	=	↓						↓	=	=			=	
Bruto et al. (2007)	↓	=	↑	↓						=	=	=			=	
Chen et al. (2011)	↓									↓					=	↑
Cohen et al. (2010)					↓											
Daliento et al. (2005)	↓	=	=	=						=	=	=			=	
Ebenroth & Hurwitz (2007)	↓	=	=	↓								↓			=	
Gratz et al. (2009)	↓	=	=	↓						=	=	=			=	
Hager & Hess (2005)	↓	=	=	↓						=	=	↓			=	
Immer et al. (2005)	=	=	=	=						=	=	=			=	

Table 2.3 ***Continued***

Authors (Year)	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*
	Physical functioning	***Role physical***	***Pain***	***General health***	***Symptoms***	***Gross motor functioning***	***Fine motor functioning***	***Cognitive functioning***	***Sleep***	***Psychological/ mental health***	***Role emotional***	***Vitality***	***Happiness***	***Sexual functioning***	***Social functioning***	***Environment & daily activities***
Irtel et al. (2005)	↓	↓	↓	↓						↓	↓	↓			↓	
Jefferies et al. (2004)	↓	↓	=	↓						=	=	=			=	
Kamphuis et al. (2002a)	=	=	=	=		=	=	=	=	=	=	=	=	=	=	=
Kamphuis et al. (2002b)	↓	↓	=	↓		↓	=	=	=	=	=	↓	=	=	=	=
Lane et al. (2002)	↓	↓	↓	↓						↓	↓	↓			↓	
Loup et al. (2009)	=	=	=	=						=	=	=			=	
Lu et al. (2010)	=	=	↓	↓						=	=	=			=	
Mokhles et al. (2011)	↓	=	↑	↓						=	↑	=			=	
Moons et al. (2006)				↑												
Müller et al. (2011)	↓	↓	=	↓						=	=	↓			=	

Table 2.3 ***Continued***

Authors (Year)	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*
	Physical functioning	***Role physical***	***Pain***	***General health***	***Symptoms***	***Gross motor functioning***	***Fine motor functioning***	***Cognitive functioning***	***Sleep***	***Psychological/ mental health***	***Role emotional***	***Vitality***	***Happiness***	***Sexual functioning***	***Social functioning***	***Environment & daily activities***
Rose et al. (2005)	↓			↓	↓					↓					=	
Saliba et al. (2001)	=		=	=						=					=	
Simko & McGinnis (2003)	↓								↓	↓						↓
Vandekerckhove et al. (2009)			=			=	=	=	=	↓		↓	=	=	=	=
Winter et al. (2008)	↓	↓	↑	↓						=	=	=			=	
Winter et al.(2010)	↓	↓	↓	↓						=	=	=			=	
Total no.	**17↓**	**8↓**	**4↓**	**15↓**	**2↓**	**1↓**	**4=**	**1↓**	**2↓**	**7↓**	**2↓**	**7↓**	**4=**	**4=**	**2↓**	**1↓**
	5=	**10=**	**14=**	**5=**		**3=**		**3=**	**3=**	**16=**	**14=**	**13=**			**21=**	**4=**
			3↑	**1↑**							**1↑**					**1↑**

Note. ↓ means poorer HRQoL, ↑ means better HRQoL, = means similar HRQoL (in relation to comparison group), blank means not measured.

2.5.6.1. Physical HRQoL

Twenty-two studies examined *physical functioning*. Seventeen (77%) studies reported poorer physical functioning in people with ACHD than the comparison group, whereas 5 (23%) studies reported no differences. Risk factors for poorer functioning included younger age (Bruto, Harrison, Fedak, Rockert, & Siu 2007; Mokhles et al., 2011) and female gender (Chen et al., 2011).

Eighteen studies examined *role limitations due to physical problems*. Eight (44%) studies found greater role limitations due to physical problems in people with ACHD than the comparison group, whereas 10 (56%) studies found no differences.

Twenty-one studies examined *pain*. Four (19%) studies reported greater pain and 3 (14%) reported less pain in people with ACHD than the comparison group, while fourteen (67%) studies reported no differences.

Twenty-one studies examined *general health perception*. Fifteen (71%) studies suggested poorer and one (5%) study suggested better general health perception in people with ACHD than the comparison group, while 5 (24%) studies found no differences.

Both studies that examined *symptoms* reported more cardiac-related (e.g. shortness of breath) symptoms in people with ACHD than the comparison group.

Four studies examined *gross motor functioning*. One (25%) study indicated poorer gross motor functioning in people with ACHD than the comparison group whereas 3 (75%) found no differences.

All (*n*= 4) studies that examined *fine motor functioning* reported no differences between people with ACHD and the comparison group.

Four studies examined *cognitive functioning*. One (25%) study reported poorer cognitive functioning in people with ACHD than the comparison group, while 3 (75%) reported no differences.

Five studies examined *sleep*. Two (40%) studies reported poorer sleep functioning in people with ACHD than the comparison group, whereas 3 (60%) found no differences.

2.5.6.2. Psychosocial HRQoL

Twenty-three studies examined *psychological/mental functioning*. Seven (30%) studies indicated compromised psychological/mental functioning in people with ACHD, whereas 16 (70%) reported no differences in comparison with the general population or healthy controls.

Seventeen studies examined *role limitations due to emotional problems*. Two (12%) studies reported greater and 1 (6%) study reported fewer role limitations due to emotional problems in people with ACHD than the comparison group, whereas 14 (82%) studies reported no differences.

Twenty studies examined *vitality*. Seven (35%) studies indicated less vitality in people with ACHD than the comparison group, while 13 (65%) indicated no differences.

All (*n*= 4) studies that examined either *happiness* or *sexual functioning* indicated no differences between people with ACHD and the comparison group.

Twenty-three studies examined *social functioning*. Two (9%) studies reported reduced social functioning in people with ACHD than the comparison group, while 21 (91%) reported no differences.

2.5.6.3. Environmental/occupational HRQoL

Six studies examined *environment and daily activities*. One (17%) study reported poorer and one (17%) reported better environmental circumstances in people with ACHD than the comparison group, while 4 (66%) reported no differences.

2.5.7. The relationship between disease complexity and clinical variables and HRQoL in ACHD

HRQoL was most commonly explored in relation to cyanosis (*n*= 9, 41%), followed by diagnosis (*n*= 7, 32%), functional status (*n*= 7, 32%), exercise capacity (*n*= 6, 27%), ventricular dysfunction (*n*= 5, 23%), arrhythmias (*n*= 4, 18%), and treatment type (*n*= 2, 9%). Table 2.4 presents the findings of the reviewed studies that examined the relationship between clinical variables and HRQoL.

Table 2.4. Studies that examined associations between clinical variables and HRQoL

Clinical variable *Authors (Year)*	*Physical HRQoL*									*Psychosocial HRQoL*							*Environmental/ occupational HRQoL*	
	Physical functioning	*Role physical*	*Pain*	*General health*	*Symptoms*	*Gross motor functioning*	*Fine motor functioning*	*Cognitive functioning*	*Sleep*	*Psychological/ mental health*	*Role emotional*	*Vitality*	*Happiness*	*Sexual functioning*	*Social functioning*	*Environment*	*& daily activities*
CYANOSIS																	
Bruto et al. 2007	Y	Y	---	Y						Y	---	Y			Y		
Jefferies et al. 2004	Y	Y	---	Y						---	---	Y			---		
Lane et al. 2002	Y	Y	Y	Y						Y	---	Y			Y		
Saliba et al. 2001	Y		---	---						---					---		
Gratz et al. 2009	Y	---	---	Y						---	---	---			---		
Simko & McGinnis 2005	---								---	---						Y	
Daliento et al. 2005	---	---	---	---						---	---	---			---		
Loup et al. 2009	---	---	---	---						---	---	---			---		
Moons et al. 2005				---													
DIAGNOSIS																	
Gratz et al. 2009	---	---	---	Y						---	---	---			---		
Irtel et al. 2005	---	---	---	---						Y	---	Y			---		
Müller et al. 2011	Y	---	---	---						---	---	---			Y		
Saliba et al. 2001	---		---	Y						---					---		
Simko & McGinnis 2005	Y								Y	Y						Y	
Winter et al. 2010	Y	Y	Y	Y						---	---	---			---		
Ternestedt et al. 2001										Y					Y	Y	
EXERCISE CAPACITY																	
Gratz et al. 2009	Y	---	---	Y						---	---	---			---		

Table 2.4 ***Continued***

Clinical variable / *Authors (Year)*	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*	
	Physical functioning	*Role physical*	*Pain*	*General health*	*Symptoms*	*Gross motor functioning*	*Fine motor functioning*	*Cognitive functioning*	*Sleep*	*Psychological/ mental health*	*Role emotional*	*Vitality*	*Happiness*	*Sexual functioning*	*Social functioning*	*Environment & daily activities*	
Hager & Hess 2005	Y	---	---	Y						---	---	---			---		
Irtel et al. 2005	Y	Y	Y	Y						---	Y	---			---		
Müller et al. 2011	Y	Y	---	Y						---	---	Y			---		
Rose et al. 2005	Y			Y	---					---					---		
Ebenroth & Hurwitz 2007	---	---	---	---								---			---		
FUNCTIONAL STATUS																	
Chen et al. 2011	Y									---					---	Y	
Ebenroth & Hurwitz 2007	Y	Y	---	---								---			---		
Kampuis et al. 2002b	Y	Y	Y	Y		Y	Y	Y	Y	Y	Y	Y	---	Y	Y	Y	
Moons et al. 2005				Y													
Rietveld et al. 2002	---	---	---	---						---	---	---			---		
Cohen et al. 2010					---												
Daliento et al. 2005	---	---	---	---						---	---	---			---		
VENTRICULAR DYSFUNCTION																	
Lu et al. 2010	Y	Y	Y	Y						---	---	---			---		
Moons et al. 2005				---													

Table 2.4 ***Continued***

Clinical variable Authors (Year)	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*
	Physical functioning	*Role physical*	*Pain*	*General health*	*Symptoms*	*Gross motor functioning*	*Fine motor functioning*	*Cognitive functioning*	*Sleep*	*Psychological/ mental health*	*Role emotional*	*Vitality*	*Happiness*	*Sexual functioning*	*Social functioning*	*Environment & daily activities*
Daliento et al. 2005	---	---	---	---						---	---	---			---	
Ebenroth & Hurwitz 2007	---	---	---	---								---			---	
Irtel et al. 2005	---	---	---	---						---	---	---			---	
ARRHYTHMIAS																
Bruto et al. 2007	---	---	---	Y						---	Y	---			---	
Irtel et al. 2005	Y	Y	Y	Y						Y	Y	Y			Y	
Loup et al. 2009	---	---	---	---						---	---	---			---	
Daliento et al. 2005	---	---	---	---						---	---	---			---	
TREATMENT TYPE																
Loup et al. 2009	---	---	---	Y						Y	---	Y			---	
Lane et al. 2002	Y	Y	Y	Y						Y	Y	Y			Y	

Note. Y means association with HRQoL, --- means no association with HRQoL, blank means not measured.

Cyanosis

The majority of studies (*n*= 5, 63%) reported poorer physical functioning in cyanotic compared with acyanotic individuals, while the remaining (*n*= 3, 37%) reported no differences. One study (100%) reported poorer environmental circumstances for cyanotic than acyanotic individuals. There were mixed findings regarding the relationship between cyanosis and the following domains: role limitations due to physical problems, general health perception, and vitality. In all other domains, the general pattern of findings indicated that there were no HRQoL differences between cyanotic and acyanotic individuals.

Diagnosis

The majority of studies (*n*= 3, 60%) reported poorer general health perception in people with complex than those with simpler diagnoses, while the remaining (*n*= 2, 40%) reported no differences. One study (100%) reported poorer sleep functioning in people with complex than those with simpler diagnoses. Mixed findings were evident regarding the relationship between diagnosis and physical functioning and environmental/occupational HRQoL. Of note, the study by Ternestedt et al. (2001) reported better HRQoL in the more complex (ToF) compared with the simpler (ASD) diagnosis. In all other domains, the majority of studies indicated that there were no differences in HRQoL in terms of diagnosis.

Exercise capacity

The findings indicated that there was no association between exercise capacity and most HRQoL domains except for physical functioning and general health. The majority of studies (*n*= 5, 83%) reported that poorer exercise capacity was associated

with poorer physical functioning and poorer general health perception, while the 1 study (17%) reported no significant associations.

Functional status

Functional status was measured by the following indexes: New York Heart Association (NYHA), Ability, Warnes, and Somerville. The majority of studies (*n*= 3, 60%) reported that people who had poor functional status had poorer physical functioning compared with those who had good functional status, while 2 (40%) reported no differences. One study (100%) indicated that people who had poor functional status had poorer gross motor, fine motor, cognitive, sleep, and sexual functioning, than those who had good functional status. Two studies (100%) reported poorer environmental HRQoL in people who had poor functional status compared with those who had good functional status. Mixed findings were reported regarding the relationship between functional status and role limitations due to physical problems. In all other domains, the findings suggested that there were no differences in HRQoL in terms of functional status.

Ventricular dysfunction

The general pattern of findings suggested that there was no association between ventricular dysfunction and HRQoL.

Arrhythmias

There were mixed findings regarding the association between arrhythmias and general health perception and role limitations due to emotional problems. In all other domains, the findings indicated that there were no differences in HRQoL between individuals who experienced arrhythmias and those who did not.

Treatment type

Two studies examined the relationship between treatment type and HRQoL. One study reported better general health perception, better psychological/mental health, and greater vitality in people with TGA who were treated with modern surgical techniques (arterial switch) compared with those treated with older techniques (Mustard & Senning). There were no differences between the two groups in physical functioning, role limitations due to physical problems, pain, role limitations due to emotional problems, and social functioning. Contrary to expectations, the second study reported that patients who had curative surgery had poorer psychological/mental health and social functioning compared with patients who had corrective surgery (further operation possibility) and those who did not have surgery. In addition, patients who had palliative surgery (further operation possibility) and patients who did not have surgery were similar in terms of HRQoL. Patients who were deemed inoperable had poorer HRQoL than all other groups, except that they had similar social functioning and vitality when compared to patients who had curative surgery.

Disease complexity

A few studies used broader categorisations of disease complexity, such as the Task Force 1 (Table 2.5). The majority of studies (n= 4, 67%) indicated no differences in HRQoL between the groups, where broad disease complexity categorisations were used.

Table 2.5. Studies that examined associations between disease complexity categorisations and HRQoL

Authors (year)	*Categorisation*	*Association with HRQoL*
Bruto et al. (2007)	Diagnosis, native cyanosis, surgery type (obstructive, shunt, complex)	No
Moons et al. (2004)	Diagnosis (simple, complex)	No
Moons et al. (2005)	Diagnosis and need for follow-up appointments (Task Force 1: mild, moderate, severe)	No
Rietveld et al. (2002)	Diagnosis (cardiologist: mild, moderate, severe)	No
Chen et al. (2011)	Diagnosis and need for follow-up appointments (Task Force 1: mild, moderate, severe)	Physical HRQoL
Kamphuis et al. (2002b)	Cyanosis, arrhythmias, heart failure, residual defects (Objective physical index: score 0-4)	Gross motor & cognitive functioning

2.5.8. Summary of studies published after the review completion

An additional 21 studies were identified after the completion of the systematic review. The characteristics of the newer studies are presented in Table 2.6. All studies were cross-sectional and the number of participants ranged between 24 and 2360. Fourteen (67%) studies had a sample size smaller than n= 100. Compared with the studies that were included in the published systematic review most of which consisted of mixed samples, the majority 14 (67%) of newer studies tended to focus on a single diagnosis, while 6 (29%) included people with various diagnoses, and 1

(4%) included two diagnoses. The majority of studies (*n*= 18, 86%) used the SF-36 to measure HRQoL. Other measures used were the CHD-TAAQOL (*n*= 1), the EQ-5D (*n*= 1), the psychological General Well-Being Index (*n*= 1), and the WHOQOL-BREF (*n*= 1).

Table 2.6. Characteristics of the studies published after the review completion

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	*Diagnosis or patient group*			
1	Angeli et al. (2012)	Cross-sectional	*N*= 62 Mean age: 24 (7, 16-58) Gender: 32 male, 30 female	All SV	Not reported	SF-36, CHD-TAAQOL	Low (13)
2	Berghammer et al. (2013)	Cross-sectional	*N*= 1274 Median age: 33.1 (15-87) Gender: 647 male, 627 female	126 aortic anomalies, 122 aortic valve lesions, 97 pulmonary valve anomalies, 26 mitral valve anomalies, 26 tricuspid valve anomalies, 92 ToF/RV anomalies, 89 TGA, 17 SV, 5 truncus arteriosus, 542 shunt lesions, 132 other	Normative data	EQ-5D	Moderate (26)
3	Buys et al. (2014)	Cross-sectional	*N*= 73 Mean age: 27.3 (7.9, 16-46) Gender: 53 male, 20 female	All ToF	Normative data *N*= 1742	SF-36	Moderate (22)
4	Buys et al. (2013)	Cross-sectional	*N*= 103 Mean age: 28.7 (6.3, 18-50) Gender: 70 male, 33 female	All COA	Normative data *N*= 1742	SF-36	Moderate (21)
5	Bygstad et al. (2012)	Cross-sectional	*N*= 53 for (HR)QoL assessment (total *N*= 55) Median age: 32.2 (18.4-60) Gender: All male	All ToF	Controls age-matched *N*= 33.5 (19.9-57.1)	SF-36	Moderate (21)

Table 2.6 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
6	Callus et al. (2014)	Cross-sectional	*N*= 143 Mean age: 39.1 (14.6, 16-73) Gender: 63 male, 80 female	18 conotruncal defects, 6 aortic valve anomalies, 1 endocardial cushion defects, 88 septal defects, 1 pericardial anomalies, 7 great vessel anomalies, 1 coronary anomalies, 15 SV, 1 pulmonary venous return anomalies, 3 other	Normative data	Psychological General Well-Being Index	Moderate (23)
7	Cotts et al. (2012)	Cross-sectional	*N*= 25 Mean age: 42.6 (16) Gender: 11 male, 14 female	All CCTGA	Normative data & Controls with mild ACHD (11 ASD, 14 VSD) *N*= 25 Mean age: 39.7 Gender: 11 male, 14 female	SF-36	Moderate (22)
8	Enomoto et al. (2013)	Cross-sectional	*N*= 72 Mean age: 26.1 (5.5, 18-39) Gender: 41 male, 31 female	13 simple (10 ASD/VSD, 2 AVD, 1 other), 39 moderate (17 ToF, 4 VSD+ valve problems/obstruction, 4 Ebstein's, 3 AVSD, 8 other), 20 complex (5 CCTGA, 4 TGA, 4 TA, 2 PA, 2 truncus arteriosus, 1 other)	Normative data	SF-36	Moderate (24)
9	Frigiola et al. (2014)	Cross-sectional	*N*= 34 Mean age: 37.4 (11.8, 18.1-57.6) Gender: 17 male, 17 female	All ToF	Normative data	WHOQOL-Bref	Moderate (27)

Table 2.6 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	*Diagnosis or patient group*			
10	Hanninen et al. (2011)	Cross-sectional	*N*= 67 Mean age: 68 (60-86) Gender: 28 male, 39 female	All ASD	Normative data age-matched	SF-36	Moderate (19)
11	Hickey et al. (2012)	Cross-sectional	*N*= 396 for (HR)QoL assessment (total *N*= 840) Median age: 29.5 (18-64.9) Gender: 511 male, 329 female	All ToF	Normative data age- and gender-matched	SF-36	High (29)
12	Idorn et al. (2013)	Cross-sectional	N= 60 Mean age: 20.7 (17.8-27.4) Gender: 28 male, 32 female	All SV (Fontan)	Age- and gender-matched controls *N*= 172 Mean age: 21.7 (18.4-28.7) Gender: 84 male, 88 female	SF-36	Moderate (26)
13	Kahya Eren et al. (2013)	Cross-sectional	N= 69 Mean age: 39.7 (14.2) Gender: 18 male, 51 female	All ASD	Age-, gender-, and socioeconomic status-matched controls *N*= 69 Mean age: 40.2 Gender: 18 male, 51 female	SF-36	Moderate (24)
14	Knowles et al. (2012)	Cross-sectional	N= 224 Median age: 33 (18-60) Gender: 112 male, 112 female	All ToF	Normative data	SF-36	Moderate (24)
15	Müller et al. (2013b)	Cross-sectional	N= 546 Median age: 26.9 (16-71) Gender: 310 male, 236 female	85 simple, 154 moderate, 307 complex	Normative data age- and gender-matched	SF-36	Low (17)

Table 2.6 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
16	Müller et al. (2013c)	Cross-sectional	N= 56 Median age: Senning: 18.9 (17.7-20), ASO: 18.8 (17.1-19.5) Gender: 40 male, 16 female	All TGA (28 Senning, 28 ASO)	Normative data age- and gender-matched	SF-36	Moderate (21)
17	Müller et al. (2014)	Cross-sectional	N= 2360 Mean age: 28.6 (11.2, 14-75) Gender: 1302 male, 1058 female	112 cyanotic, 149 Fontan, 106 TGA Rastelli, 214 TGA Senning & Mustard, 59 TGA ASO, 399 ToF, 128 Ebstein, 144 PS/PR, 206 CoA, 295 AS, 287 shunt, 261 other	Normative data	SF-36	Moderate (21)
18	Opić et al. (2014)	Cross-sectional	N= 79 Mean age: 27.5 (3.4, 20-35) Gender: 53 male, 25 female	48 ToF, 31 TGA	Normative data	SF-36	Moderate (24)
19	Pike et al. (2012)	Cross-sectional	N= 54 Mean age: 25.6 (9, 15-50) Gender: 26 male, 28 female	All SV	Age-, gender-, ethnicity-, marital status-, educational level-, and geographic region-matched controls *N*= 66 Mean age: 24.5 Gender: 26 male, 40 female	SF-36	Moderate (26)
20	Riley et al. (2012)	Cross-sectional	N= 99 Mean age: 37.2 (17-67) Gender, 54 male, 45 female	30 simple, 55 moderate, 14 complex	Normative data	SF-36	Moderate (22)

Table 2.6 ***Continued***

	Authors (year)	*Design*	*Participants*		*Comparison group*	*(HR)QoL measure*	*Study quality*
			N, Mean age (S.D., range), gender	***Diagnosis or patient group***			
21	Ruys et al. (2013)	Cross-sectional	*N*= 18 for (HR)QoL assessment (total *N*= 30) Median age: 22 years (20-25) Gender: 17 male, 13, female	All TGA ASO	Normative data age- and gender-matched and control group of Mustard patients (*N*= 58, Median age: 27, Gender: 40 male, 18 female)	SF-36	Low (17)

A summary of the findings from these recent studies regarding the impact of ACHD on HRQoL can be seen in Table 2.7. In terms of physical HRQoL, out of 20 studies examining physical functioning, 9 (45%) reported poorer physical functioning and 1 (5%) study reported better physical functioning in people with ACHD than the comparison group, while 10 (50%) reported no differences. Seventeen studies examined role limitations due to physical problems, with 5 (29%) reporting greater role limitations in people with ACHD than the comparison group and 12 (71%) reporting no differences. Out of 17 studies examining bodily pain, 3 (18%) found less pain and 1 (6%) found greater pain in people with ACHD than the comparison group, whereas 13 (76%) reported no differences. Seventeen studies examined general health perception and 11 (65%) reported poorer general health perception in people with ACHD than the comparison group and 6 (35%) found no differences. In terms of psychosocial HRQoL, out of 21 studies examining psychological/mental health, 3 (14%) found poorer and 1 (5%) found better psychological/mental health in people with ACHD than the comparison group, while 17 (81%) found no differences. Out of 16 studies examining role limitations due to emotional problems, 1 (6%) reported greater and 1 (6%) reported fewer role limitations in people with ACHD than the comparison group, while 14 (88%) found no differences. Seventeen studies examined vitality and 6 (35%) reported less and 1 (6%) reported more vitality in people with ACHD than the comparison group, while 10 (59%) found no differences. Out of 18 studies examining social functioning, 2 (11%) found better social functioning in people with ACHD than the comparison group and 16 (89%) found no differences. In terms of environmental/occupational HRQoL 1 (100%) study found better environmental circumstances in people with ACHD than the comparison group. Overall, the pattern of findings regarding the impact of

ACHD on HRQoL remained unchanged with the inclusion of newer studies (Table 2.7).

Since the majority of the recent studies focused on a single diagnostic group rather than mixed samples, their inclusion enabled the examination of HRQoL in various diagnostic groups compared with the general healthy population. Three studies focused on people with diagnoses categorised in the Simple group (CoA: *n*= 1, ASD: *n*= 2). Together, the findings from these studies reported no differences in any HRQoL domain between people with ACHD and the comparison group. Interestingly, the study by Buys et al. (2013) which focused on people with CoA found poorer general health in the physical domain and poorer psychological/mental health and lower vitality in the psychosocial domain compared with normative data.

Five studies focused on people with ToF and the majority found poorer physical functioning, poorer general health, and poorer vitality in this group compared with the general population but no differences in other physical and psychosocial domains. One of these studies examined environmental/occupational HRQoL and found better HRQoL in people with ToF compared with normative data. Three studies focused on people with TGA and the majority found no differences in any domain of HRQoL between this diagnostic group and the comparison group. Of note, the findings regarding role limitations due to emotional problems were mixed across the three studies. Three studies focused on people with SV and the majority found poorer physical functioning, greater role limitations due to physical problems, and poorer general health in this group compared with the general population but no differences in other physical and psychosocial domains.

Table 2.7. Summary of findings from studies published after the systematic review completion

Authors (year)	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*	
	Physical functioning	***Role physical***	***Pain***	***General health***	***Symptoms***	***Gross motor functioning***	***Fine motor functioning***	***Cognitive functioning***	***Sleep***	***Psychological/ mental health***	***Role emotional***	***Vitality***	***Happiness***	***Sexual functioning***	***Social functioning***	***Environment & daily activities***
Angeli et al. (2012)	=	=	=	↓						↓	=	↓			=	
Berghammer et al. (2013)	=	=	↑							=					=	
Buys et al. (2014)	↓	=	=	↓						↓	=	↓			=	
Buys et al. (2013)	=	=	=	↓						↓	=	↓			=	
Bygstad et al. (2012)	↓	=	=	↓						=	=	=			=	
Callus et al. (2014)				=						=		=				
Cotts et al. (2012)	=	=	=	=						=	↓	=			=	
Enomoto et al. (2013)	↓	↓	=	↓						=	=	=			=	
Frigiola et al. (2014)	=									=					↑	↑
Hanninen et al. (2011)	=	=	=	=						=	=	=			=	
Hickey et al. (2012)	↓	↓	↑	↓						=	=	↓			=	

Table 2.7 ***Continued***

Authors (year)	*Physical HRQoL*									*Psychosocial HRQoL*						*Environmental/ occupational HRQoL*
	Physical functioning	***Role physical***	***Pain***	***General health***	***Symptoms***	***Gross motor functioning***	***Fine motor functioning***	***Cognitive functioning***	***Sleep***	***Psychological/ mental health***	***Role emotional***	***Vitality***	***Happiness***	***Sexual functioning***	***Social functioning***	***Environment & daily activities***
Idorn et al. (2013)	↓	↓	=	↓						=	=	=			=	
Kahya Eren et al. (2013)	=	=	=	=						=	=	=			=	
Knowles et al. (2012)	↓	=	↑	↓						=	=	↓			=	
Müller et al. (2013b)	=									=						
Müller et al. (2013c)	=	=	=	=						=	=	=			=	
Müller et al. (2014)	↓									=						
Opić et al. (2014)	=	=	↓	↓						↑	=	↓			↑	
Pike et al. (2012)	↓	↓	=	↓						=	=	=			=	
Riley et al. (2012)	↓	↓	=	↓						=	=	=			=	
Ruys et al. (2013)	↑	=	=	=						=	↑	↑			=	
Total no. (including initial systematic review studies)	**26↓ 10= 1↑**	**13↓ 22=**	**5↓ 27= 6↑**	**26↓ 11= 1↑**	**2↓**	**1↓ 3=**	**4=**	**1↓ 3=**	**2↓ 3=**	**10↓ 33= 1↑**	**3↓ 28= 2↑**	**13↓ 23= 1↑**	**4=**	**4=**	**2↓ 37= 2↑**	**1↓ 4= 2↑**

Very few studies (*n*= 2) with mixed diagnostic samples investigated HRQoL differences between diagnostic groups. Two studies (100%) reported poorer physical functioning in people with complex than those with simpler diagnoses. One study (100%) found greater role limitations due to physical problems and poorer social functioning in people with complex than those with simpler diagnoses. In all other domains, the majority of studies indicated that there were no differences in HRQoL in terms of diagnosis.

Two studies used broader categorisations of disease complexity (i.e. Task Force 1). Enomoto et al. (2013) reported poorer physical functioning in moderate and severe groups compared with the mild and poorer general health perception in the severe group compared with the moderate group. Müller, Hess, and Hager (2013b) found poorer physical functioning but better psychological/mental health with increasing complexity.

2.6. Discussion

This review is the first to have systematically examined the evidence regarding HRQoL in ACHD. A total of 31 studies were systematically reviewed and an additional 21 studies were identified after the review completion. Although the review findings on the HRQoL in people with ACHD were mixed, some initial inferences can be made.

In the majority of studies there was a lack of conceptualization of HRQoL. There was also an evident variability in the measurement of HRQoL between the studies. This was reflected in the use of generic, disease-specific, and individual measures of HRQoL. However, the majority of studies used the generic SF-36 measure. In order to facilitate the synthesis of the findings the scales/subscales of different measures

were categorised in three broad HRQoL domains: physical, psychosocial, and environmental/occupational and related sub-domains. The systematic review followed a different approach to previous reviews in that it provided a summary of the literature by examining the weight of evidence in various HRQoL domains. This coupled with the larger number of studies reviewed, provided a clearer picture on the magnitude of evidence.

2.6.1. HRQoL comparisons with normative data or healthy controls

The first objective of this review was to investigate the HRQoL of people with ACHD in comparison to normative data or healthy controls. The general pattern of findings indicated that people with ACHD experience reduced physical functioning, poorer general health, and greater symptoms than the comparison group of either normative data or healthy controls. In some domains of physical HRQoL (i.e. role limitations due to physical problems, pain, sleep, gross/fine/cognitive/motor functioning) as well as psychosocial and environmental/occupational HRQoL the majority of studies indicated no differences between people with ACHD and the comparison group. This pattern of findings in the published systematic review was supported by the findings of recent studies in ACHD identified after the review completion.

The majority of recent studies focused on a single diagnostic group compared with older studies, the majority of which examined mixed samples. This likely reflects the increased recognition of the need to distinguish between diagnostic groups due to the heterogeneity of the ACHD population. Most of the older studies with mixed samples did not distinguish between diagnostic groups when examining HRQoL possibly due to their limited sample sizes that precluded such analyses. The review

of recent studies provided some insight into HRQoL in various diagnostic groups compared with the general healthy population. The pattern of findings indicated that people with ToF and SV appear to experience poorer HRQoL than the comparison group but only in certain domains pertaining to physical HRQoL (physical functioning, general health). On the other hand, the findings indicated that there were no differences in HRQoL between people in the Simple and TGA diagnostic groups and the comparison group. These findings suggest that certain groups (i.e. ToF, SV) may be vulnerable to experiencing reduced physical HRQoL. This finding is in line with the findings of the systematic review regarding the ACHD population as a whole, which appears to experience diminished HRQoL in certain physical domains but not psychosocial and environmental/occupational domains.

It was difficult to compare the findings of the present systematic review with the literature review by Kovacs et al. (2005). The latter identified only 8 studies conducted in ACHD and did not distinguish between different domains of HRQoL. The authors concluded that there were mixed findings with some studies reporting poorer HRQoL and others reporting normal HRQoL in people with ACHD. Mixed findings were also reported in reviews conducted in child populations (Bellinger & Newburger, 2010; Latal et al., 2009; Marino et al., 2010). Dahan-Oliel et al. (2011) reported similar findings to the present systematic review, indicating that young adults with CHD experience impaired physical HRQoL.

The findings of the present study however, suggest that these individuals are mostly concerned with the physical limitations and practical issues arising from their condition. A chronic condition like ACHD introduces significant physical challenges with individuals experiencing cardiac-related symptoms and impaired physical

functioning. However, the physical impact of ACHD does not appear to extend to people's ability to carry on with their typical role activities, considering the number of studies indicating lack of impact on the role physical subscale. Although the evidence on cognitive, fine motor, gross motor, and sleep functioning were limited to a few studies, the general pattern of findings indicated that people with ACHD were not impaired in these domains.

These findings are likely to reflect the impaired physical state of people with ACHD. Continuous cardiac surveillance coupled with unexpected health deterioration can have a significant physical burden. However, these findings also raise an important issue regarding expectations. Given that HRQoL is patient-reported, it is possible that the findings reflect a perceived lack of understanding leading people to develop unrealistic expectations and misconceptions about their physical functioning. In addition, the somewhat unclear disease course in ACHD may leave people feeling uncertain and insecure about their condition and prognosis (Claessens et al., 2005).

The evident lack of impairment in psychosocial HRQoL across most of the reviewed studies suggests that despite experiencing significant physical limitations, people with ACHD find ways to cope. Some authors proposed that these individuals successfully adjust to their condition because they develop adaptive coping strategies early in their lives. One such coping mechanism is normalization, whereby people accept and incorporate their illness and treatment in their daily lives from an early age (Deatrick, Knafl, & Murphy-Moore, 1999). This form of normalization has been reported in qualitative studies with children and adolescents with CHD but also adults (Berghammer, Dellborg, & Ekman, 2006; Claessens et al., 2005). In the study by Claessens and colleagues (2005), people with ACHD struggled to live a "normal"

life, adjusting their daily life to their functioning and matching their lifestyle to that of their healthy peers.

Other authors proposed that people with ACHD cope with the stressors of living with a chronic condition because of an increased sense of coherence. For example, Moons and Norekvål (2006) attributed the lack of HRQoL impairment in their earlier study (Moons et al., 2006) to an increased sense of coherence. They argued that growing up with ACHD has made people understand, manage, and find meaning in their experiences. This helps them gain perspective which facilitates better psychosocial adjustment.

Some authors have argued that lack of psychosocial impairment in people with ACHD can be attributed to a change in internal standards, values, and priorities (i.e. response shift) (Schwartz & Sprangers, 1999). The response shift phenomenon has been typically proposed as an explanation of changes in HRQoL over time in longitudinal studies. However, some authors argued that response shift may also be evident in cross-sectional studies. Specifically Moons et al. (2009) argued that individuals who grow up with ACHD may develop different internal values from healthy individuals. In one of the studies included in the present systematic review, younger individuals reported poorer HRQoL compared to their peers, whereas older individuals reported normal HRQoL compared to their healthy counterparts (Bruto et al., 2007). This may signify that response shift is taking place over time leading older individuals to redefine of their standards, values, and priorities and therefore modify their expectations of HRQoL.

Although a limited number of studies examined environmental/occupational HRQoL, the evidence indicated a lack of impairment in this HRQoL domain. This

finding suggests that people with ACHD do not generally have problems with work, access to healthcare, and their environment. It is unsurprising that these individuals did not experience problems with access to healthcare considering that the samples in the reviewed studies mostly consisted of individuals monitored regularly in specialist ACHD clinics. On the other hand, many guideline reports have suggested that people with ACHD may face occupational problems, including career and insurance problems (Perloff, 1991). The lack of impairment in occupational HRQoL in the reviewed studies can be attributed to its assessment in already employed individuals. However, there was a limited number of studies examining this particular HRQoL domain; hence, more studies are needed to establish whether people with ACHD experience limitations in the occupational domain of their lives.

2.6.1.1. Changes in HRQoL over time

It is worth noting that because the reviewed studies were mostly cross-sectional, it was not possible to examine whether changes in HRQoL occur over time. Only two longitudinal studies were identified in the systematic review. Ternestedt et al. (2001) reported mean scores in HRQoL for two groups (ToF, ASD; *n*= 26) at two follow-ups, specifically 20 and 30 years after surgery. However, they did not provide a description of any statistical analysis conducted to study changes over the two time points and the results and discussion sections lacked clarity about changes in HRQoL. Thus it was not possible to extract the relevant information. The study by Ebenroth and Hurwitz (2007) reported minimal changes over time in a sample of 35 people with TGA, with lower energy at a 10-year follow-up. Two additional longitudinal studies were identified after the review completion. One study consisted of mostly people with ToF (*n*= 103) and indicated a decrease in general health

perception at a 3-year follow-up but no changes in the subscales of the EQ-5D, including mobility, self-care, usual activities, pain/discomfort, and anxiety/depression (Skoglund et al., 2014). Another study with a mixed sample (*n*= 349) reported that social inhibition (feeling unpleasant among other people) decreased while negative emotions increased over a period of 10 years (van Rijen et al., 2005a). The evident lack of studies examining changes in HRQoL in ACHD point to the need of more research that includes longitudinal assessments of HRQoL.

2.6.2. Relationship between disease complexity and clinical variables and HRQoL

The second objective of the review was to study the relationship between disease complexity as well as clinical variables and HRQoL. The findings indicated no differences in HRQoL between groups in studies that used broad disease complexity categorisations such as the Task Force 1 by Warnes et al. (2001). Two of the most recent studies however, found that physical HRQoL diminished with increased disease complexity. Taking into account older and more recent studies, the findings with regards to disease complexity using broad categorisations were mixed. These findings are in agreement with a recent meta-analysis of the impact of disease complexity on emotional functioning in adolescents and adults with CHD (Jackson, Misiti, Bridge, Daniels, & Vannatta, 2015). The authors concluded that this heterogeneity in findings across studies may be due to the failure of broad categorisations (e.g. Task Force 1: simple, moderate, complex) in capturing the true impact of illness on people's experiences. The main criticism is that broader categorisations do not sufficiently differentiate between the most complex SV and the comparatively less complex TGA diagnoses because both are categorised as

"complex", although the TGA group do not typically experience the same morbidity and mortality risk as the SV group (Jackson et al., 2015).

However, the same meta-analysis suggested that individual clinical variables need to be explored in order to explain heterogeneity across studies in emotional functioning. In the present systematic review there was an evident impact of individual clinical variables on physical HRQoL, whereby being cyanotic, having a more complex diagnosis, experiencing reduced exercise capacity and poorer functional status were associated with a poorer general health perception and greater physical limitations. This finding is in agreement with the review by Dahan-Oliel et al. (2011) who found that disease complexity was associated with poorer physical HRQoL in adolescents and young adults with CHD. There was either a lack of association or mixed findings in other physical subscales as well as psychosocial and environmental/occupational HRQoL. This finding is in agreement with previous reviews in children and adolescents (Latal et al., 2009; Marino et al., 2010). The review by Kovacs et al. (2005) was inconclusive regarding the impact of disease complexity on HRQoL in ACHD. The findings of this review suggest that disease complexity and clinical factors may be associated with certain domains of physical HRQoL but not psychosocial or environmental/occupational HRQoL.

These findings, coupled with the variability in HRQoL differences between people with ACHD and the general population discussed in the previous section, suggest that clinical factors alone may be a poor explanation for variability in HRQoL in ACHD and that other psychosocial processes may help explain some of this variability. The limited ability of clinical factors in explaining people's HRQoL emphasizes the importance and supports the adoption of biopsychosocial approaches

to health and illness research (Skevington, Lofty, & O'Connell, 2004). This approach was advocated in Engel's seminal work, who in an attempt to unify the three perspectives, namely biological, psychological, and social, proposed the biopsychosocial model (Engel, 1977). In Engel's model, the individual is the focal point and therefore illness is viewed as a human experience with its inherent psychosocial as well as biological parameters (Figure 2.2). All three parameters are thought to interact to influence health.

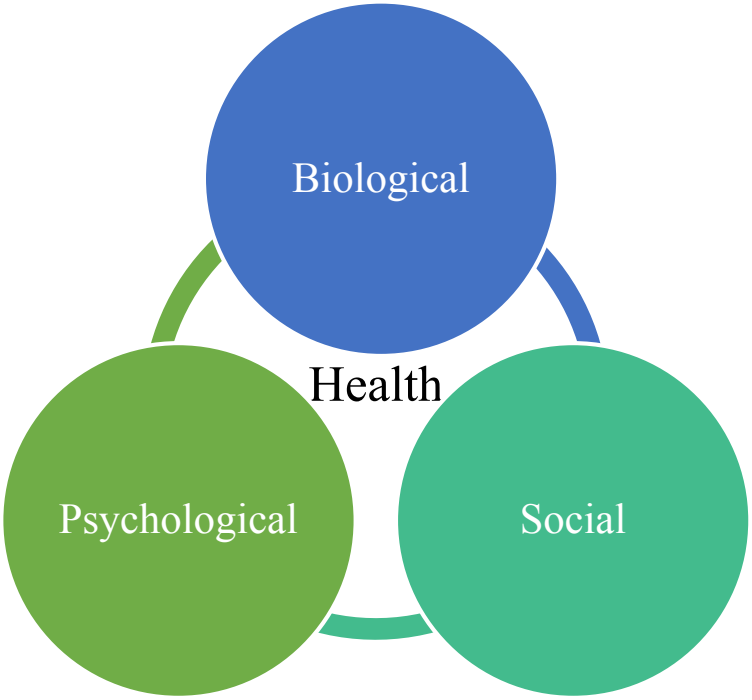


Figure 2.2. The biopsychosocial model

It has been increasingly recognised that various factors may influence an individual's HRQoL; these do not only include biomedical or clinical variables but also psychological and interpersonal variables (Ferrans, Zerwic, Wilbur, & Larson, 2005; Wilson & Cleary, 1995). Therefore, studying the collective effects of these variables can provide further insight about the HRQoL of people with ACHD.

2.6.3. Limitations of the studies reviewed and implications for future research

There are number of methodological issues that are apparent in the research reviewed in the systematic review. One difficulty is that by definition the ACHD population is heterogeneous. This is exacerbated in some studies with a lack of distinction between diagnostic groups when examining differences between the sample and the comparison group. Furthermore, while some studies focused on a single diagnosis, the majority included various diagnoses. In general, the heterogeneity makes for large variability and difficulty in comparing studies. This heterogeneity coupled with the different nomenclature used in the studies makes it difficult to draw conclusions regarding HRQoL problems in specific diagnostic groups. While this limitation was addressed to some extent by more recent studies that focused on a single diagnostic group, there is a need for a more clear distinction between various diagnostic groups in future studies with mixed samples.

The quality assessment of the reviewed studies indicated that lower quality was mainly due to poor reporting and insufficient sample sizes. Over half of the studies reviewed included a sample of less than 100 participants and it was not reported whether it was informed by power calculations. Studies were also characterised by poor reporting of methodology, including recruitment and statistical analysis. In addition, the vast majority of studies failed to define QoL and HRQoL. They also used various instruments to measure (HR)QoL. It has been argued that different conceptualizations and measures of (HR)QoL are likely to lead to diverse results (Testa & Nackley, 1994).

The lack of longitudinal assessments was evident in the literature with only four studies assessing changes in HRQoL over time. Longitudinal studies are important

for establishing whether HRQoL changes over time and the factors that may influence such change.

2.6.4. Limitations of the systematic review

In addition to the limitations of the reviewed studies discussed in the previous section, the limitations of the systematic review need to be addressed. The review was limited to articles written in English and published in peer-reviewed journals. It is acknowledged that the use of translation services would have resulted in the inclusion of articles published in other languages; however, the funds for such services were not available at the time. The search strategy was broad and was conducted in a single step using the pre-determined terms. A staged search strategy of each facet (i.e. population, outcomes etc.) might have resulted in a more flexible and comprehensive search.

As mentioned earlier, the systematic review attempted to provide some initial inferences on the impact of ACHD on HRQoL based on the magnitude of evidence. However, it should be noted that there were mixed findings across studies. An effort was made to identify possible reasons for the mixed findings, for example certain differences in methodologies, sample size, and patient characteristics; however, no systematic patterns were observed in these characteristics that could help explain why some studies reported poorer, better, or equal HRQoL in relation to the comparison group.

It was not within the scope of the review to examine the relationship between psychosocial variables and HRQoL in ACHD. As discussed earlier, some of the variability in HRQoL may be explained by underlying psychosocial processes. These relationships remain largely unstudied in the ACHD population. However, research

interest in such associations is rapidly increasing and this literature is addressed in Chapter 3.

2.7. Summary

This systematic review is the first that attempted a synthesis of evidence regarding HRQoL in ACHD. The review also highlighted the limitations of previous research. These limitations are important in terms of implications for future research, as a growing number of studies are being published every year in this rapidly evolving field. Despite the mixed findings and methodological limitations evident in previous research, the magnitude of evidence indicated that people with ACHD face certain physical limitations compared to the general healthy population, and that some clinical factors that reflect disease complexity are associated with HRQoL. The role of psychosocial factors has begun to attract significant research interest with an increasing number of studies examining the relationship between these factors and HRQoL in ACHD. The next chapter will focus on psychosocial factors and their relationship with HRQoL guided by previous research in ACHD and the theoretical framework used in the present study.

CHAPTER 3 – THE ROLE OF PSYCHOSOCIAL FACTORS IN RELATION TO HEALTH-RELATED QUALITY OF LIFE IN ADULT CONGENITAL HEART DISEASE

3.1. Prologue

The previous chapter described the current state of the literature regarding HRQoL in ACHD by presenting the findings of a systematic review. This indicated that people with ACHD experience impaired physical HRQoL, while their psychosocial and environmental/occupational HRQoL appeared to be similar to the general population. In addition, although some clinical factors were found to be associated with HRQoL they did not account for much of the variability in HRQoL. It is therefore possible that psychosocial factors may help explain some of the variation in HRQoL. This chapter describes the theory that was used to guide the selection of some of the psychosocial variables included in the present study. The role of psychosocial factors in relation to HRQoL in ACHD and other chronic illnesses across the lifespan is described by drawing evidence from both quantitative and qualitative studies. The chapter also addresses the literature with regards to longitudinal changes in psychosocial factors.

3.2. Rationale for theoretical framework

Theories in health psychology are typically concentrated in explaining behaviour by mapping the relationship between clinical, psychological, and social factors. Therefore, much of the focus has been the prediction of behaviour and the psychosocial processes through which behaviour change occurs. These theories provided the basis for many interventions aiming to alter unhealthy behaviours in

patients and healthy populations (Michie, Johnston, Francis, Hardeman & Eccles, 2008). Some examples of such theories include the Health Belief Model, the Theory of Planned Behaviour, the Theory of Reasoned Action, the Social Cognitive Theory, the Transtheoretical Model, the Stress-Coping Model etc. (Michie, et al., 2005).

These theories are not specifically focused on explaining HRQoL but rather consider it as an outcome which may be influenced by modifying cognitions, emotions, and behaviour. In fact, previous attempts to incorporate HRQoL in a theoretical model have been directed towards conceptualizing HRQoL rather than the processes which may influence HRQoL (Bakas et al., 2012; Taillefer, Dupuis, Roberge, & LeMay, 2003). Examples include the conceptual model by Wilson and Cleary (1995) and its subsequent revision by Ferrans et al. (2005). Although this model helps conceptualize HRQoL, it lacks a description of how the relationship between various psychosocial factors may help explain HRQoL outcomes.

The use of theory in studying the factors associated with HRQoL helps identify patients at risk for poor HRQoL and the psychosocial factors which are amenable to change and hence can be targeted in future interventions aiming to improve HRQoL. One model that has been used to study outcomes in already ill populations is the Self-Regulation Model (SRM) of health and illness proposed by Leventhal, Meyer, and Nerenz (1980), which was selected as the basis for the present study. There are several reasons that motivated the selection of this particular model. Firstly, it has been utilised more extensively in previous studies that examined HRQoL in cardiac populations as well as various other chronic illnesses (Hagger & Orbell, 2003). Secondly, the processes that underlie the model regarding the relationship between the variables are clearly defined. Thirdly, it describes psychosocial variables which

are amenable to change and can be therefore targeted in future interventions (McAndrew et al., 2008). Finally, it has been developed and validated in many chronic illnesses throughout the years, which resulted in the development of validated measures for its core components.

Although the purpose of the study was not to test the validity of the assumptions underlying the SRM or an attempt to test the SRM, it was used to guide the selection of some of the psychosocial variables that were considered as likely to influence HRQoL in ACHD. This framework is described in the following section.

3.3. The self-regulation model

As mentioned earlier, the SRM of health and illness was proposed by Leventhal and colleagues (1980) and is part of the wider group of self-regulation theories. Self-regulation broadly refers to efforts made by people to alter their own responses in relation to the achievement and maintenance of personal goals (Maes & Karoly, 2005). In the context of health and illness, referral to self-regulation implies the capability of an individual to act in order to prevent unhealthy consequences or to restore one's health and well-being when illness or health problems have already occurred (Purdie & McCrindle, 2002).

According to the SRM, when people are faced with a symptom, an illness, or a threat to their health, they create mental representations (i.e. illness perceptions) in order to make sense of and manage their problem. Three basic assumptions underlie the model (Diefenbach & Leventhal, 1996):

1) People are active problem-solvers that try to make sense of their condition and symptoms based on various sources of information.

2) The illness representation guides the choice of coping procedures and their evaluation in terms of their effectiveness to produce the desired outcomes.
3) The illness representation is individualised and may not be in line with the medical reality.

People form parallel cognitive and emotional representations that derive from internal (i.e. somatic cues or symptoms) or external (i.e. experience or doctor's advice) stimuli. The cognitive and emotional representations lead to the choice of coping procedures to manage the health threat and emotional reactions respectively, which in turn are evaluated in relation to the expected outcomes (Leventhal, Diefenbach, & Leventhal, 1992).

Leventhal, Leventhal, and Contrada (1998) defined coping procedures as *"the cognitive and behavioural actions people take (or do not take) to enhance health and to prevent, treat (i.e. cure or control), and rehabilitate from illness"* (p. 722). During evaluation, if the coping procedures do not produce the desired outcome, the illness representation may be re-defined (Figure 3.1).

The SRM assumes that individual and social factors influence the process of illness representation. The history of prior illnesses stored in memory plays a significant role in the emotional and illness representation as well as the choice of coping procedures. According to Diefenbach and Leventhal (1996), these memories are automatically retrieved when a person forms a representation of their current experience. Moreover, a new ambiguous experience is interpreted based on the somatic self of an individual. Thus, people determine whether a symptom is novel or it is common part of the self. The authors also argue that people are continuously interacting with and are influenced by their social context and are active recipients of

social information. Hence, illness perceptions may be influenced by the medical encounter, family members as well as by social comparison (Leventhal et al., 1998).

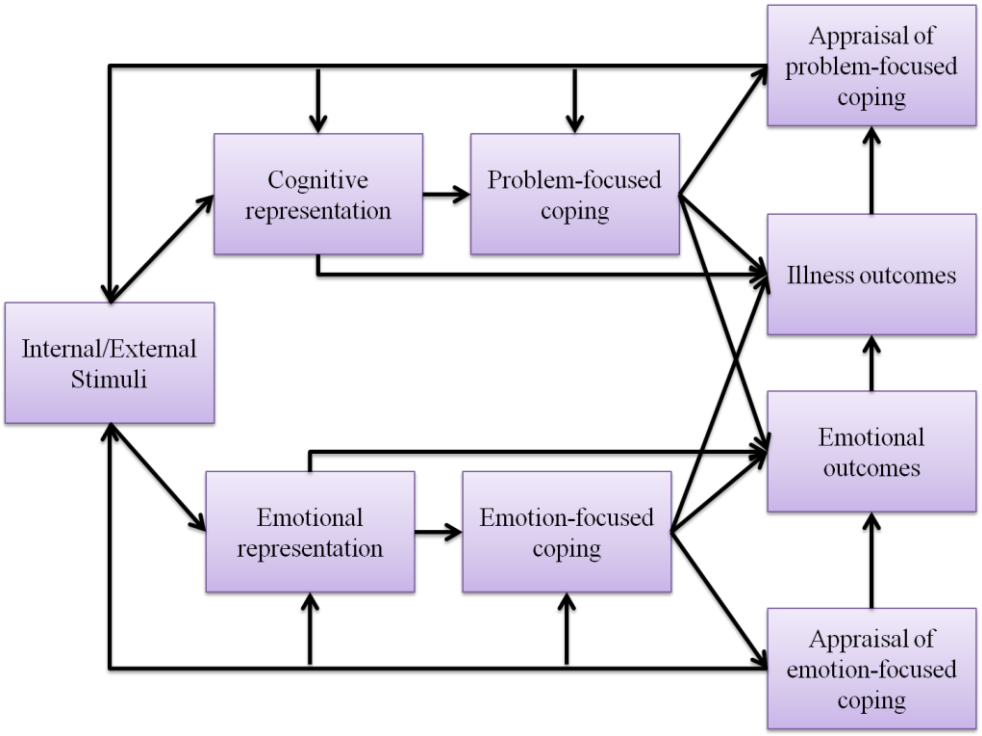


Figure 3.1. The self-regulation model [adapted from Hagger & Orbell (2003)]

Illness perceptions have at least five attributes or dimensions (Leventhal et al., 1998):

1) Identity, which represents the disease label and its associated symptoms.
2) Timeline, which defines whether the disease is acute, cyclic, or chronic.
3) Alleged causes such as genes, infection, stress, and fate.
4) Physical, social, and economic consequences (suppositional or real).
5) Perceived controllability/curability, which is the extent to which the disease can be controlled and cured.

Although these five dimensions are widely acknowledged as the cornerstone of how people perceive their illness, it has been argued that some may be more or less

relevant for specific group of patients and can therefore vary between illnesses (Heijmans & de Ridder, 1998).

The SRM has been used to predict objective clinical outcomes as well as various illness adjustment outcomes including adherence to medication, and self-management behaviours. However, the most common outcome used in previous studies across various conditions is HRQoL (Hagger & Orbell, 2003) which is also the focus of the present thesis. In this study, the cognitive representation being investigated are illness perceptions. The SRM can also integrate mood as part of the emotional representation. Although anxiety and depression can be important outcomes in their own right, their influence, and especially that of depression, on both clinical outcomes and HRQoL has been well supported by a wealth of studies in cardiac populations (e.g. Gaynes, Burns, Tweed, & Erickson, 2002; Hallas, Wray, Andreou, & Banner, 2011; Stafford, Berk, Reddy, & Jackson, 2007). The role of coping strategies will also be investigated within the SRM. Finally, the role of perceived social support will be examined, as the SRM can potentially integrate this well-established concept. Evidence regarding the role of these factors in HRQoL and changes in these factors over time will be discussed next.

3.4. The role of illness perceptions in HRQoL

Evidence suggests that illness perceptions may influence HRQoL directly or indirectly through coping. While some studies have provided support for the mediating role of coping, much of the research has indicated that illness perceptions have a direct association with outcomes (Hagger & Orbell, 2003).

Illness perceptions have been studied in populations such as heart failure, coronary heart disease, myocardial infarction, cancer, arthritis, diabetes, asthma, HIV, and

epilepsy. Findings in myocardial infarction (French, Lewin, Watson, & Thompson, 2005), coronary heart disease (Stafford, Berk, & Jackson, 2009) as well as other medical conditions (Petrie, Jago, & Devcich, 2007) have suggested that illness perceptions are significant predictors of HRQoL. A meta-analysis by Hagger and Orbell (2003) indicated that physical functioning, social functioning, and psychological well-being are strongly associated with having weaker perceptions of illness identity (i.e. fewer symptoms attributed to the illness) and weaker perceptions of illness consequences. Having a strong perception of control over one's illness was also found to be associated with positive outcomes (Hagger & Orbell, 2003).

In ACHD, very few studies have examined illness perceptions and their association to HRQoL. In a cross-sectional study of 99 people with ACHD, Riley and colleagues (2012) reported that strong perceptions about a negative illness impact (i.e. illness consequences) and perceptions of a cyclical illness timeline (i.e. illness unpredictability and experience of symptoms in cycles) was associated with poorer physical HRQoL. In addition, strong perceptions about a negative illness impact, strong perceptions of an emotional impact, and a cyclical illness timeline were associated with poorer psychosocial HRQoL. However, illness perceptions did not remain significant independent predictors of HRQoL when entered into a multiple regression model that included anxiety and depression.

In a longitudinal study of 845 people with ACHD, Schoormans and colleagues (2014) measured illness perceptions at baseline and HRQoL two years later. They found that negative illness perceptions were predictive of poor physical and psychosocial HRQoL in the two-year follow-up, independent of age, gender, disease complexity, and functional status. Specifically, strong perceptions about negative

illness consequences and about a negative emotional impact of the illness predicted poor psychosocial HRQoL. Strong perceptions about negative illness consequences, a cyclical timeline, and weak perceptions about curability predicted poor physical HRQoL. Surprisingly, weak perceptions of a negative emotional impact predicted poor physical HRQoL. A weak perception of a coherent illness understanding also predicted impaired physical HRQoL.

Other studies in ACHD have examined the relationship between one particular aspect of illness perceptions, namely sense of coherence, and HRQoL. In a cross-sectional study of 546 people with ACHD, Müller et al. (2013b) found that a strong sense of coherence was associated with both physical and psychosocial HRQoL. Sense of coherence was associated with better HRQoL in both cross-sectional (Apers et al., 2013b; Wang, Hay, Clarke, & Menahem, 2014) and longitudinal studies (Apers et al., 2013c; Luyckx, Missotten, Goossens, & Moons, 2012) in adolescents with CHD. In the longitudinal study by Apers et al. (2013c), sense of coherence at baseline predicted increases in generic and disease-specific HRQoL over a period of 9 months after accounting for demographic characteristics and disease complexity.

Research on illness perceptions in ACHD and adolescent CHD has been limited to very few studies, which have suggested that illness perceptions were significant predictors of HRQoL in these populations. These studies have also suggested that illness perceptions may be better predictors of HRQoL than demographic characteristics and clinical status. The limited number of studies in ACHD on this topic highlights the need for more research to enable conclusions regarding these associations.

3.5. The role of coping in HRQoL

In order to successfully adjust, people with ACHD will need to develop adequate coping strategies to help them manage the physical, cognitive, and psychosocial implications of living with ACHD and its treatment regimen. Coping can have two main functions, which are tackling the problem (i.e. problem-focused) and controlling the emotions (i.e. emotion-focused) (Folkman, Lazarus, Gruen, & DeLongis, 1986). For example, problem-focused coping may include efforts like planning and problem solving whereas emotion-focused coping may include avoidance and positive re-appraisal.

Most research on coping in ACHD has been qualitative. Studies involving adolescents and adults with CHD indicated that they use coping strategies such as normalization (Berghammer et al., 2006; Claessens, et al., 2005; Lee & Kim, 2010; Shearer, Rempel, Norris, & Magill-Evans, 2013), denial or acceptance (Berghammer et al., 2006; Horner, Liberthson, & Jellinek, 2000; Overgaard, King, Christensen, Schrader, & Adamsen, 2013; Zahmacioglu, et al., 2011), and avoidance or approach (Birks, Sloper, Lewin, & Parsons, 2007; Chiang, Chen, & Chen, 2011; McMurray, et al., 2001; Tong et al., 1998).

Denial, which is characterised by denying the reality of an event, has been reported as a way of coping with stressful experiences such as early operations in a qualitative study of adolescents with complex CHD (Zahmacioglu et al., 2011). However, another qualitative study with a mixed sample of adolescents with CHD found that denial was generally followed by a form of normalization, with adolescents reporting acceptance of their condition, an awareness of their limitations and adjustment of their activities or even avoidance of physically demanding situations (McMurray et

al., 2001). Normalization has also been reported in qualitative studies with people with ACHD (Berghammer et al., 2006; Claessens et al., 2005). Claessens et al. (2005) interviewed individuals with ToF, TGA, and SV and found that normalization was central for their successful adjustment to ACHD. Another qualitative study that included people with complex ACHD indicated that these individuals were accepting of their condition (Horner et al., 2000). Positive reframing is another coping strategy that has been reported in a previous qualitative study with people with complex ACHD (Overgaard et al., 2013). The development of such coping strategies as acceptance and normalization seemed to derive from the need for self-monitoring especially as people get older and recognise their role in managing their illness and treatment regimen (Chiang et al., 2011; Tong et al., 1998).

A cross-sectional study reported lower use of active problem solving in people with ACHD (van Rijen et al., 2004). In this study, women appeared to engage more in avoidance coping and less in seeking social support than the norms. Men on the other hand used more social support and less venting of negative emotions than the norms. Reduced problem solving in people with ACHD has also been suggested by other authors (Enomoto et al., 2013). In another study by Eslami, Macassa, Sundin, Khankeh, and Soares (2014) people with ACHD engaged in more palliative reaction coping (attention diversion) than healthy controls. Evidence also suggests that the adoption of coping strategies is independent of disease complexity (Enomoto, et al., 2013; Eslami et al., 2014; van Rijen et al., 2004).

No studies have investigated the relationship between coping strategies and HRQoL in ACHD, yet the role of coping in other outcomes has been previously demonstrated. In a study of adolescents with CHD, task-oriented coping involving an

active approach to problem solving, explained 30% of the variance in resilience (Lee, Kim, & Choi, 2014). The authors concluded that active problem solving was a significant predictor of positive adaptation in adolescents with CHD.

Research in other cardiac populations such as heart failure has suggested that denial, self-distraction, and self-blame coping strategies are predictive of poor HRQoL (Graven & Grant, 2013; Klein, Turvey, & Pies, 2007). Although small, the impact of coping strategies on HRQoL was evident in a number of chronic illnesses in a review by de Ridder and Schreurs (1996). Another review by the same authors that focused on psychological interventions targeting coping in chronic illnesses suggested coping to be effective in influencing positive HRQoL outcomes (de Ridder & Schreurs, 2001).

3.6. The role of mood in HRQoL

In ACHD variable prevalence rates of clinically significant levels of anxiety and depression have been reported based on self-report measures and clinical interviews. Popelová, Slavík, & Skovránek (2001) reported that clinically significant depression was evident in 34% of the sample studied using Zung's self-report questionnaire. Other studies that used the Beck Depression Inventory (BDI) indicated a 9-32% prevalence of moderate to severe depression (Bang et al., 2013; Kourkoveli, et al., 2014; Kovacs, et al., 2009b). A study by Bromberg, Beasley, D'Angelo, Landzberg, & DeMaso (2003) reported that 27% of the sample exhibited clinical levels of depression using the Brief Symptom Inventory (BSI). Studies that used the Hospital Anxiety and Depression Scale (HADS) reported that 22-41% had at least moderate levels depression (Cohen et al., 2010; Eslami, Sundin, Macassa, Khankeh, & Soares, 2013; Riley et al., 2012). Pike et al. (2012) used the Patient Health Questionnaire

depression module (PHQ-9) and found that moderate to severe depression was evident in 32% of the sample studied. Müller, Hess, & Hager (2012) reported a prevalence of 9% of depressive symptoms in people with ACHD using the Center for Epidemiologic Studies Depression scale (CES-D). When assessed by clinical interviews based on the Diagnostic and Statistical Manual of Mental Disorders (DSM) criteria, prevalence rates of depression ranged between 27% and 33% (Bromberg et al., 2003; Kovacs et al., 2009b).

Studies that used the HADS self-report measure for anxiety indicated that moderate to severe anxiety levels were prevalent in 41-63% of the sample (Cohen et al., 2010; Eslami et al.; 2013; Riley et al., 2012). Kovacs et al. (2009b) used the State-Trait Anxiety Inventory-Trait version (STAI-T) and reported that 34% of the sample exhibited elevated anxiety. Assessments of anxiety by clinical interviews based on the DSM indicated prevalence between 9% and 26% (Bromberg et al., 2003; Kovacs et al., 2009b).

Qualitative studies have also reported feelings of anxiety and depression in people with ACHD (Berghammer et al., 2006; Claessens et al., 2005; Horner et al., 2000). Berghammer et al (2006) found that people with ACHD felt more anxious during periods when their condition was more noticeable, for example when they felt sick or tired. They also found that people with ACHD were fearful about their future, that their health would worsen, and that they would experience a shortened life. Fear about the future has also been reported by Horner and colleagues (2000) who found that people with ACHD worried about sudden death or a progressive decline in their health and functioning. The authors also noted that during their young adulthood people with ACHD became increasingly focused on their physical symptoms which

caused symptoms of depression and anxiety and were worried about family planning. Cornett and Simms (2014) interviewed a sample of individuals with ACHD and found that they faced a number of emotional challenges, including depression and shame about their limitations, psychological trauma due to earlier operations, and fear about the future because of uncertainty.

Studies that compared levels of anxiety and depression in people with ACHD and either healthy controls or normative data indicated mixed findings. Studies that used the HADS reported either higher (Eslami et al., 2013), lower (Cox, Lewis, Stuart, & Murphy, 2002), or equal (Cohen et al., 2010; Loup, et al., 2009) levels of mood disorders in people with ACHD compared with norms or healthy controls. One study reported higher levels of psychological distress as measured by the Symptom Checklist-90-R (SCL-90-R) in people with ACHD compared with norms (Brandhagen, Feldt, & Williams, 1991). A study that used the CES-D found that people with ACHD had lower levels of depression than their healthy counterparts (Müller et al., 2012). Using the STAI, Müller, Hess, and Hager (2013a) indicated that people with ACHD had higher state anxiety levels but equal trait anxiety levels compared with healthy controls. Overall, the evidence suggests that disease complexity is not associated with anxiety and depression (Eslami et al., 2013; Kovacs et al., 2009b; Loup et al., 2009; Müller et al., 2012, Müller et al., 2013a; Popelová et al., 2001).

An association between mood and HRQoL in ACHD has been suggested in a number of cross-sectional studies. In general, lower levels of anxiety, depression, and psychological distress have been found to be associated with better HRQoL (Chen et al., 2011; Pike et al., 2012; Riley et al., 2012). Riley and colleagues (2012)

studied the HRQoL of 99 people with ACHD and found that poor HRQoL in the physical and psychosocial domains was not associated with disease complexity but with higher anxiety and depression levels.

Similar relationships have been reported in another study of a larger sample of 879 adolescents and adults with CHD, in which both anxiety (Müller et al., 2013a) and depression (Müller et al., 2012) scores correlated to all domains of HRQoL. In the latter, stronger associations were observed between depression and the mental health sub-domain of HRQoL; hence higher levels of depression were associated with poorer mental health (Müller et al., 2012). Using regression analysis, Rietveld and colleagues (2002) found that lower scores of trait anxiety predicted better overall HRQoL.

The association between higher levels of anxiety and depression and poor HRQoL has been reported in studies with various ACHD. In the study by Pike et al. (2012), poorer HRQoL was strongly predicted by higher depression levels in a sample of 54 people with SV. Higher anxiety and depression levels were associated with poorer HRQoL in a study of 27 people with ASD (Cohen et al., 2010). Chen et al. (2011) studied the determinants of HRQoL in 289 people with various ACHD using regression analysis. The authors found that higher levels of psychological distress were predictive of poorer HRQoL in all domains including physical, psychological, social, and environmental after accounting for demographic characteristics and disease complexity.

3.7. The role of the social environment in HRQoL

It is believed that the social environment can enhance the people's efforts to cope with their illness and the stress it may cause thus serving as a "buffer". The presence

of social support may minimise the experience of stress in the first instance and influence the adoption of positive health behaviours (Heaney & Israel, 2008).

It has been argued that social support consists of two types, namely structural and functional support (Lett et al., 2005). Structural support refers to size and frequency of contact with the patient's network of people. Measures include number of people in a patient's network and frequency of contact. Functional support refers to the nature of support. House (1981) categorised functional support based on the nature of supportive behaviours and includes: a) instrumental: provision of practical assistance b) emotional: provision of empathy, love, and caring c) informational: provision of advice and information that a person may use to solve problems and d) appraisal: provision of constructive feedback and affirmation. There is also a distinction between the actual received support and perceived support by the person who receives it (Lett et al., 2005).

Social support however, might not always be beneficial. Shinn, Lehmann, and Wong (1984) argued that the impact of negative support, characterised by hostility, conflict or even incompatible support to an individual's needs might be more prominent than the effects of positive social support. Other authors have characterised positive and negative social support as a "double-edged sword", whereby there is an interaction between the two (Revenson, Schiaffino, Majerovitz, & Gibofsky, 1991).

Evidence has suggested that people with ACHD received adequate amount of support from their environment. Knowles and colleagues (2012) reported no differences in support levels between people with ACHD and their siblings and Pike and colleagues (2012) reported that these individuals did not differ in perceived social support from norms. Another study suggested higher levels of social support

in people with ACHD compared with normative data (Rose et al., 2005). On the contrary, lower levels of instrumental, informative, problem-oriented, emotional, and total social support were reported in people with ACHD than norms in a study of 342 patients (van Rijen et al., 2004). However, in the same study, fewer discrepancies between the preferred and received support were observed in people with ACHD rather than norms. A qualitative study by Cornett and Simms (2014) also found that people with ACHD received emotional support from their family but this was somehow complicated by their need to protect their family members from their own feelings and thoughts. Qualitative studies have also highlighted the challenges faced by people with ACHD regarding disclosure of their condition to their social environment (Berghammer et al., 2006; Claessens et al., 2005; Cornett & Simms, 2014). These studies have found that although people with ACHD were comfortable disclosing their condition to close friends and family they tended to hide it from people outside their close circle. Non-disclosure in people with ACHD was prominent in situations where they would meet new people due to fear of being judged or rejected (Cornett & Simms, 2014).

A number of studies have found an association between perceived social support and HRQoL in ACHD. In a cross-sectional study of 54 people with ACHD, higher levels of social support were found to be a significant independent predictor of better overall HRQoL after accounting for depression and functional status (Pike et al., 2012). Rose and colleagues (2005) used structural equation modelling to study the determinants of HRQoL in 111 people with ACHD. The authors found that social support rather than clinical factors was a significant determinant of psychological

and social HRQoL. Therefore, a higher level of social support was strongly associated with better psychological and social HRQoL.

Higher levels of social support were also associated with better HRQoL in all domains in the cross-sectional studies by Areias et al. (2013) and Wang et al. (2014) of mixed adolescent and young adult CHD cohorts. Other studies found that adolescents and adults with CHD who reported greater perceived social support, reported better HRQoL in all domains including physical, psychological, social, and environmental compared with those who reported poorer perceived social support (Coelho et al., 2013; Silva et al., 2011; Teixeira et al., 2011). Luyckx and colleagues (2014) conducted a longitudinal study of 429 adolescents (mean age: 15.75) to study the determinants of HRQoL using structural equation modelling analyses. They found that baseline parental support predicted relative improvements in HRQoL over a 9-month period. Although peer support did not predict longitudinal HRQoL, it was a significant predictor of HRQoL at both time points. These findings provide further support to the notion that higher levels of social support, and especially parental support, influence positive HRQoL outcomes in CHD.

3.8. The dynamic process of self-regulation

The SRM postulates that the self-regulation process is dynamic and thus influenced by changes in the illness experience, such as new symptoms or the introduction of a new treatment (Leventhal et al., 1998). There is currently a lack of longitudinal research in ACHD examining changes over time in the psychosocial factors described above. Similarly, research in other chronic illnesses is limited. This section describes the evidence from the wider chronic illness literature on longitudinal changes in illness perceptions, coping strategies, mood, and social support.

Regarding illness perceptions, a study in people recovering from myocardial infarction found that perceived controllability and curability decreased whereas illness identity and consequences remained stable over a period of twelve months (Petrie & Weinman, 1997). The authors argued that perceptions pertaining to the impact of cardiac disease remained stable over time because these are well-defined to the point that they become lay beliefs. On the other hand, perceptions about cure and control decreased because people's initial perceptions of their myocardial infarction as an acute episode changed into a more chronic view of the illness (Petrie & Weinman, 1997). Another study in people with osteoarthritis found that those who experienced progression of disability reported decreased perceived controllability and coherence, a more negative illness identity, increased perceived consequences and a more negative emotional representation in the six-year follow-up compared with those who did not experience progression of disability (Bijsterbosch et al., 2009). These findings suggest that changes in illness perceptions over time are associated with changes in illness progression as health deterioration can bring about new symptoms and physical limitations enhancing a more negative perception of the illness. Lawson et al. (2008) found that illness identity, consequences, and controllability remained stable over a period of two years in newly diagnosed people with diabetes. The authors also found that emotional representation decreased while illness coherence increased over the two-year follow-up. The authors argued that the overall stability in illness perceptions in their study may be because people developed their illness beliefs early in their illness experience and these were not challenged by changes in health and treatment. Taken together, the findings from the above studies suggest that illness perceptions change over the course of a chronic illness and these changes are associated with illness progression.

In addition to illness perceptions, long-term changes in coping may occur especially in the context of chronic illness (Lazarus, 1993). Limited research in cardiac populations has indicated that coping strategies in people with coronary heart disease (van Elderen, Maes, & Dusseldorp, 1999) and people recovering from myocardial infarction (Kristofferzon, Löfmark, & Carlsson, 2005) remained stable over a period of twelve months. Similarly, a review in stroke found that the coping strategies adopted by people remained stable over time (Donnellan, Hevey, Hickey, & O'Neill, 2006). The observed stability of coping over time may reflect the adoption of habitual or automatic behaviours by people with chronic illnesses (de Ridder & Schreurs, 2001). On the other hand, a study in breast cancer has reported significant changes in coping six months after diagnosis, more specifically an increase in detachment and a decrease in seeking social support, spirituality, and wishful thinking (Danhauer, Crawford, Farmer, & Avis, 2009). These findings can potentially be explained by the nature and length of treatment as some people with cancer may need to undergo more frequent treatment such as chemotherapy and/or radiation therapy following diagnosis which may influence their coping process compared with people with cardiac disease or stroke.

With regards to mood, there has been one study in ACHD that examined changes over time. van Rijen et al. (2005b) examined the development of psychopathology (including anxiety and depression) in 220 people with ACHD over a period of ten years. The analyses were based on categorization in trajectories of psychopathology and indicated that although the majority (80.9%) remained stable, 8.2% exhibited an increase and 10.9% exhibited a decrease in psychopathology over a period of ten years. Older adults (28-32) more often were free of emotional problems compared

with younger adults (20-27). The authors noted that emotional problems decreased over time despite the higher levels observed in young adulthood and concluded that people with ACHD recover from early emotional problems as they grow older. However, as the authors stated their analysis provided only a rough idea about psychopathology in people with ACHD over time and thus repeated measures analyses are required in future studies.

Social support on the other hand has been generally viewed as a trait-like characteristic that remains stable over time (Hoyt & Stanton, 2012). Though, there has been evidence that social support may change over the course of a chronic illness depending on the illness and treatment demands. Research in cardiac disease has indicated that social support changes from the time of hospitalization to the early period of recovery from myocardial infarction. Specifically, up to 14.5% of people reported either reduced or increased support over a period of a month (Leifheit-Limson et al., 2012). The authors argued that these changes in social support may be related to situational factors in the early period of myocardial infarction. Experiences like hospitalization and recovery following discharge from hospital may have altered people's perceptions of social support as they can influence people's support needs, availability, or quality of social support (Leifheit-Limson et al., 2012). In a five-wave longitudinal study of people with rheumatoid arthritis, Strating, Suurmeijer, and van Schur (2006) found that satisfaction with emotional support remained stable from T1 to T4 but decreased over a period of eight years between T4 and T5. This change may also be related to the changing support needs of people with rheumatoid arthritis over the course of their illness (Strating et al., 2006).

The evidence above suggest that psychosocial factors including illness perceptions, coping strategies, mood, and social support can potentially change over the course of a chronic illness due to changes in health and treatment. The apparent lack of longitudinal research in ACHD point to the need for more longitudinal studies to examine how these psychosocial factors change over time.

3.9. Summary

This chapter highlighted the importance of various psychosocial variables in explaining variation in HRQoL in ACHD within the context of the SRM. The evidence reviewed in this chapter indicated that illness perceptions, coping, mood, and the social environment can significantly influence HRQoL. There are, however, limited studies that have examined the relative importance of these factors for HRQoL in ACHD and a lack of longitudinal research examining changes in these factors over time in ACHD. There is a need for further research in order to establish if and how these psychosocial variables impact on the HRQoL of people with ACHD and whether these factors change over time. The next chapter provides an overall summary of the literature and the rationale for the present thesis.

CHAPTER 4 – SUMMARY OF THE LITERATURE, RATIONALE AND AIMS OF THE PRESENT THESIS

4.1. Summary of the literature

Chapter one highlighted the growing population of people with ACHD. It described CHD and its treatment and the long-term complications that affect people with ACHD.

Chapter two described the impact of ACHD on HRQoL by presenting the findings of the systematic review of the literature. Despite the evident variability of the findings in the reviewed studies, strong evidence suggested that ACHD can have a significant impact on certain aspects of physical HRQoL. However, this impact did not appear to extend to psychosocial and other domains of HRQoL in most of the studies. In addition, clinical factors were found to have limited ability to explain variability in HRQoL. These findings suggest that it may be that psychosocial factors may help explain some of the variability not detected in the studies.

Chapter three described the usefulness of the SRM in explaining and predicting variation in HRQoL in chronic illness. The chapter also reviewed the evidence in ACHD and other chronic illnesses regarding the impact of psychosocial variables on HRQoL and changes in these factors over time.

4.2. Rationale for the present study

There was a lack of agreement and clarity on the concept of HRQoL across the studies reviewed in Chapter 2. In addition, studies used one of the three types of measures (generic, disease-specific, or individual) to assess HRQoL. It has been increasingly recognised that the use of both generic and disease-specific measures

may offer more insight into HRQoL of the population under study (Fletcher et al., 1992). Generic measures offer the opportunity to compare HRQoL across various diagnostic groups and the general population, whereas disease-specific measures provide more detailed information regarding specific life domains that may be influenced by an illness. The present thesis utilises both generic and ACHD-specific instruments the measurement of HRQoL.

The majority of the reviewed studies had small sample sizes and were not informed by power calculations. Insufficient sample sizes are generally limited in their ability to detect small effects and in some cases may lead to misinterpretations (Vandenbroucke et al., 2007). The present thesis addresses this limitation by including a large and adequately powered sample.

One limitation of previous studies identified in the systematic review was the lack of clear distinction between various diagnostic groups that limits the ability to draw conclusions regarding the HRQoL in these groups. The review also identified a number of different clinical factors that were used across studies to examine their association with HRQoL. The present study includes four clearly defined diagnostic groups but also includes a wide range of clinical measures to study the impact of ACHD on HRQoL.

In Chapter 3 it was evident that in ACHD very little research has investigated whether individual differences in HRQoL can be explained by psychosocial variables. A more comprehensive approach to studying the factors associated with HRQoL in ACHD is currently lacking. The present study attempts to address this gap in the literature by examining a broad range of factors that may potentially

influence HRQoL in ACHD, including demographic and clinical characteristics and psychosocial constructs drawn from the SRM and previous research.

At present, there is also a lack of longitudinal research examining HRQoL in ACHD over time with only four studies reporting such findings (section 2.6.1.1, page 96). These studies had limitations in that they either focused on one diagnostic group or had small sample sizes. None of the studies included measures of both generic and disease-specific HRQoL. Furthermore, these studies used traditional methods of repeated measures analysis, which are prone to various statistical bias (see section 5.6.8.4, page 165). The variation in follow-up years among participants was not controlled for in the analysis, thus the findings of these studies may have been a result of measurement bias. Furthermore, as evidenced in Chapter 3 (section 3.8, page 119) studies have yet to examine changes in psychosocial factors, including illness perceptions, coping strategies, mood, and social support in ACHD. The present thesis seeks to add to the literature by including a follow-up study to examine whether HRQoL and other psychosocial variables in ACHD change over time using a more appropriate statistical method, thereby eliminating some of the bias associated with traditional repeated measures analysis methods.

Furthermore, most qualitative studies in congenital heart disease reviewed within Chapter 3 have focused on children and adolescents and very few on adults. However, with the rapidly increasing population of adults compared with younger people with ACHD (Tan, 2006), there is a need to study their experiences of living and adapting to ACHD over the life course. Previous qualitative studies in ACHD did not specifically focus on HRQoL and included predominantly younger adults with moderate and complex ACHD (i.e. ToF, TGA, SV). The thesis seeks to add to

the literature with the inclusion of a qualitative study and the adoption of a mixed methods approach. The qualitative study was designed to address aspects of the quantitative component of the thesis, thus enabling a more detailed understanding about HRQoL in ACHD.

4.3. Aims and objectives

The aims of the present thesis were defined as detailed below:

- Aim 1a: To examine the impact of ACHD on HRQoL by comparing the HRQoL of people with ACHD with normative data from the general population.
- Aim 1b: To examine differences in HRQoL between four diagnostic groups.
- Aim 2: To investigate the factors associated with HRQoL in ACHD by using a set of demographic and clinical measures and psychosocial constructs.
- Aim 3: To examine whether HRQoL and other psychosocial variables, including illness perceptions, coping strategies, mood, and social support, change over time and whether diagnostic group influences change.
- Aim 4a: To explore the experiences of people on how ACHD had influenced their HRQoL in physical, social, occupational, and psychological domains over the years.
- Aim 4b: To explore how people adjust to living with ACHD.

4.4. Hypotheses

The hypotheses formulated below were based on past literature outlined in Chapter 2 and the theoretical framework of the present thesis described in Chapter 3.

- People with ACHD will report significantly poorer HRQoL compared with the general population norm, predominantly in physical domains of HRQoL.

- Poorer HRQoL will be more pronounced in the SV group when compared with the general population and when compared with the Simple group.
- Psychosocial variables including illness perceptions, coping strategies, mood, and social support will account for a significant amount of variance in HRQoL over and above that accounted for by demographic and clinical characteristics.

No hypotheses were formulated for research questions #3 and #4 as these investigations were exploratory or of a qualitative nature.

4.5. Using a mixed methods approach

The present thesis uses a mixed methods approach by including a qualitative study. This approach has become an equally important research methodology to quantitative and qualitative approaches alone (Johnson, Onwuegbuzie, & Turner, 2007). There have been several definitions of mixed methods research. Johnson et al. (2007) synthesized 19 different definitions and in turn, suggested the following definition:

"Mixed methods research is the type of research in which a researcher or team of researchers combines elements of qualitative and quantitative research approaches (e.g., use of qualitative and quantitative viewpoints, data collection, analysis, inference techniques) for the broad purposes of breadth and depth of understanding and corroboration." (p. 123)

Therefore, the combination of both quantitative and qualitative methods can provide a deeper understanding of the phenomenon in question (Johnson et al., 2007). The application of mixed methods in health research is considered as a patient-centred

approach by some researchers (O'Cathain, Murphy, & Nicholl, 2007). Quantitative approaches usually include large samples and statistical analyses to investigate research questions. Although statistical approaches minimize researcher bias, they generally lack an in-depth understanding of a phenomenon, which qualitative approaches may offer (Ritchie & Lewis, 2003). For this reason, it is believed that mixed methods research may help "offset" each of the two methods' weaknesses and bias (Bryman, 2006). Another benefit from using mixed methods is triangulation, which enables the researcher to compare the results and examine similarities and differences from the two approaches (Curry, Nembhard, & Bradley, 2009).

There are several typologies of mixed methods designs, which generally depend on the timing of each method and whether one of the two methods (quantitative or qualitative) is predominant (Curry et al., 2013). The present study utilised a concurrent embedded design, whereby the quantitative approach was dominant, and the qualitative approach was supplemental or embedded within the quantitative study (Creswell & Plano Clark, 2011). The data collection for the qualitative study took place after the cross-sectional study but concurrently with the longitudinal study. The purpose of this mixed-methods approach was *"complementarity"*, defined by Greene et al. (1989) as using quantitative and qualitative methods to *"measure overlapping but also different facets of a phenomenon, yielding an enriched, elaborated understanding of that phenomenon."* Therefore, the purpose of the qualitative study within this thesis was to enhance understanding and maximise the interpretation of the findings of the quantitative study (Onwuegbuzie & Leech, 2006).

4.6. Structure of subsequent chapters

The analyses presented in Chapters 6 to 10 will address each of the aims described above. A discussion of the findings is included immediately after each set of analyses and a general discussion will integrate the findings and discuss the implications. The next chapter will describe the methodology of the quantitative component of the thesis. The methodological details of the qualitative study along with the analysis and findings are presented in its respective chapter (Chapter 10).

CHAPTER 5 – METHODOLOGY

5.1. Prologue

This chapter describes the methodology of the quantitative component of the thesis including the study design, recruitment strategy and procedures, the measures utilised, and the statistical analysis plan.

5.2. Study design

A cross-sectional design was used to answer research questions #1 and #2 presented in the previous chapter. Within-subjects repeated measures design was used to answer research question #3. Qualitative methods were used to answer research question #4 (described in Chapter 10).

5.3. Ethical approval

Full ethical approvals for both the cross-sectional and longitudinal studies were granted from the Joint UCL/UCLH Ethics Committee and NRES Committee London – Bentham in Ethics of Human Research (REC reference number: 08/H0715/105). Relevant approvals were also gained from the Research & Development (R&D) department at UCLH.

5.4. Procedure

5.4.1. Setting

Both the cross-sectional and longitudinal studies were conducted at The Heart Hospital, part of the University College London Hospitals (UCLH) NHS Foundation Trust, that specialises in cardiac diagnosis and treatment.

5.4.2. Participants

Between March 2009 and June 2011 all patients who had routine follow-up appointments at the Grown-Up Congenital Heart (GUCH) unit of The Heart Hospital, London were selected for the cross-sectional study. All participants of the cross-sectional study were subsequently invited for the longitudinal study. Participants were included in the study if they met the following inclusion criteria:

- 16 years of age and older
- Diagnosed with either of the following: ToF, TGA, CoA, VSD, ASD, PS, SAS, and SV physiology
- Fluency in English to ensure the completion of questionnaires

Participants were excluded if they met the following criteria:

- Chromosomal conditions such as trisomy 21 (Down syndrome) and 22q11 deletion (DiGeorge syndrome)
- Severe learning difficulties/mental retardation as indicated in clinical notes
- Diagnosis of patent foramen ovale (PFO)
- Those with TGA who had arterial switch operation, as this treatment approach was adopted in the late 1980's
- Immediately prior to or post (6-months) a surgical intervention or pregnancy
- Poor hearing or eyesight, as recorded in patients' records
- Previous stroke
- Physically unable to attempt the exercise test (e.g. wheelchair-bound)

5.4.3. Identification of eligible participants

At the time of the present study, not many patients with ACHD were recorded into the newly established electronic record system at The Heart Hospital, London;

therefore, eligible participants were identified using two methods, which are described below.

5.4.3.1. The paper records

Eligible participants were identified in alphabetical order (surname, name) via hospital paper records, which documented all the *active* patients in the UCLH GUCH service. Once identified, a specialist nurse used the most recent clinical letter to assess eligibility against the inclusion/exclusion criteria. If the criteria were met, the participant's name and contact details were added into an electronic spreadsheet. If the potential participant did not meet the criteria, they were excluded from the study and their details were entered into a separate electronic spreadsheet.

Eligible participants were categorised into four equally sized diagnostic groups: Simple, ToF, TGA, and SV. People who had more than one diagnosis were categorised into one of the four diagnostic groups based on their more complex diagnosis. For example, a person with ASD and SV would have been categorised in the SV group. The final categorisations were based on the agreement between two consultant cardiologists, who were involved in the participants' regular care and were familiar with their clinical history. The diagnostic groups are described in section 5.5.2 (page 137). The clinical details of these diagnostic groups have been described in the clinical background (section 1.4, page 24).

The target maximum recruitment number was 90 participants from each of the four diagnostic groups (see section 5.6.1, page 154). All eligible participants with TGA and SV were invited to take part in the study; this was due to the small numbers of individuals with these diagnoses. For the Simple and ToF diagnoses, 90 participants were selected using a random number generator and were invited to take part in the

study. Whenever a negative response was received, another participant was randomly selected until the required number (n= 90) was achieved.

5.4.3.2. The clinic lists

The outpatients' clinic lists were reviewed every month during the recruitment period, in order to identify eligible participants that were not recorded in the hospital paper records[5]. In addition to meeting the recruitment target, this method was followed in order to ensure that newly referred individuals were not missed. This method was therefore complementary to the method described above. Eligible participants with upcoming appointments were invited to take part in the study.

5.4.4. Invitation

5.4.4.1. Cross-sectional study

Participants were invited for the study by post, which contained a formal invitation letter, an information sheet (see Appendix I) and an interest form, which participants were asked to complete and return in a freepost envelope. The information sheet included an explanation about the purpose of the study and the reason for which the participant was selected. Invited participants were informed about their role in the study, were explained about confidentiality issues, and their right to withdraw at any time. Finally, invited participants were provided with contact numbers in case they had any questions and comments about the study. If the participant had not responded within two weeks, a second invitation letter was sent with the same documents. If the participant had not responded after an additional two weeks, a telephone call was made.

[5] These participants were typically patients that were newly referred to the UCLH GUCH clinic from other clinics around the UK; hence, no paper records of them were available at the time.

All the enrolled participants were assigned a unique study identification number (e.g. 001, 002,…314). Appointment letters were sent to the participants for the same day of their upcoming outpatient appointment in order to minimise travelling burden and costs.

5.4.4.2. Longitudinal study

Participants were invited for the study by post, which contained a formal invitation letter, an information sheet (see Appendix J), and an interest form, which participants were asked to complete and return in a freepost envelope. Participants were informed that they were invited because they had participated in the cross-sectional study and that there was no obligation for participating in the longitudinal study. Telephone reminders were carried out, when participants did not respond to a second postal invitation. Appointment letters were sent to interested participants for the same day of their upcoming outpatient appointment. Postal questionnaire packs were mailed to participants whose next appointment was after the study end date in order to maximise recruitment.

5.4.5. Administration of questionnaires

5.4.5.1. Cross-sectional study

On the day of the assessment, participants were informed about the purpose of the study using the information sheet as a guide and were then asked to sign the consent form (see Appendix K). The latter indicated that the participant had enough time to consider their participation and that they agree to take part. Following the informed consent process, participants completed the questionnaires in a dedicated room at The Heart Hospital, London, with the researcher present available to answer potential questions. Participants received a £5 voucher to use for lunch.

Occasionally, participants had a long day of testing at the hospital; these participants were given the choice of completing the questionnaires at home. In these instances, participants were provided with a freepost envelope and telephone calls were conducted to answer possible questions and to ensure that the completed questionnaires were returned.

5.4.5.2. Longitudinal study

Follow-up administrations took place soon after the cross-sectional study was completed over a period of one year. For face-to-face administrations, the same informed consent process (see Appendix L) was followed as in the cross-sectional study (see previous section). For postal administrations, freepost envelopes were provided along with telephone numbers, on which participants could reach the researcher for any questions regarding the questionnaires. The same reminder protocol (described in section 5.4.5.1) was used for any participants who did not return the follow-up questionnaires. Participants did not receive a £5 voucher for lunch in the follow-up study.

5.5. Measures

The selection of measures was guided by the SRM and previous research. The study measures included demographic and clinical information as well as self-report questionnaires. Due to the large number of variables studied, where available, shorter versions of the questionnaires were used in order to minimise participant burden.

5.5.1. Demographic characteristics

Demographic data were collected during the cross-sectional study only using a self-report form. Demographic details included:

- Age

- Gender
- Ethnicity
- Marital status: married/in civil partnership, in relationship, single, divorced/separated, widowed
- Educational level: Primary, secondary, tertiary, undergraduate degree, master's degree, doctorate degree
- Employment status: Employed, self-employed, seeking job, housewife/husband, student, retired, long-term sick leave, unable to work

5.5.2. Clinical information

A consultant cardiologist examined the participants' paper and electronic hospital records (EPR, CDR) that included clinical reports, pre-assessment forms, and clinical testing reports and recorded detailed and comprehensive clinical information using a standard form during the cross-sectional study. Clinical information was also collected through electronic hospital records during the longitudinal study regarding further interventions, hospitalization days, and medication. The details of all clinical information obtained for the studies and the measurement details are presented in Table 5.1.

Participants were categorised into four diagnostic groups. The "Simple" group included acyanotic people with ASD (treated surgically or percutaneously), VSD (treated surgically), PS, SAS, and CoA (treated both in childhood and adulthood and re-coarctations). People with CoA who had aortic valve replacement were excluded, as this condition does not meet the merit of a simple defect. The "ToF" group included people with a diagnosis of ToF, including those with pulmonary atresia,

Major Aortopulmonary Collaterals (MAPCAs[6]), and those who had pulmonary valve replacement. The "TGA" group included people with a diagnosis of TGA who had Mustard or Senning operations (i.e. atrial switch), including those with implantable cardioverter defibrillators and pacemakers. People who had arterial switch operation were not included as this treatment approach was adopted in the late 1980's. Finally, the "SV" group included people with a single ventricle physiology, those who had the Fontan or TCPC operations, including those who are chronically cyanotic.

Cyanosis information included the number of days that participants were cyanosed before they had undergone their first intervention that restored their saturation levels (i.e. duration of cyanosis). For chronically cyanosed participants this represented the days cyanosed up until the study participation date.

Intervention history included the total number of cardiac-related interventions, including surgical operations and catheterization lab procedures since birth. Hospitalization days reflected the total number of days spend in hospital either for surgical/catheterization lab procedures or emergency hospitalizations since birth.

The tests for current O_2 saturation, VO_2 max, and ventricular function data outlined in Table 5.1 were performed as part of the routine monitoring of ACHD for all study participants. The most recent pulse oximetry result was recorded to provide information about current O_2 saturation.

[6] Arteries that develop naturally in embryonic life to supply blood to the lungs when pulmonary circulation is underdeveloped.

Table 5.1. Clinical information obtained from hospital records

Variable Group	*Measurement Details*
Disease characteristics	
Diagnostic groups	1) Simple 2) Tetralogy of Fallot 3) Transposition of the great arteries 4) Single ventricle
Co-morbidities total	Count
Arrhythmias	Yes/No
Cyanosis days (until study participation date)	Count
Intervention history	
Interventions total (includes cardiac-related surgical and catheterisation lab procedures)*	Count
Hospitalization days (since birth)*	Count
Current status	
Medication total*	Count
Current O_2 saturation	%
Functional status (NYHA)	Groups: 1) Class I 2) Classes II, III, IV
VO_2 max	mL/(kg·min)
Left & right ventricular ejection fraction	%

*Clinical variables for which information was collected at both cross-sectional and longitudinal studies.

Details of a recent exercise test were recorded in order to collect information on exercise capacity as measured by maximal oxygen uptake (VO_2 max). The gold standard in measuring VO_2 max is thought to be cardiopulmonary exercise testing (CPEX), where the assessment is conducted via a mouthpiece/mask fitted while the individual is exercising on a bike. However, not all people with ACHD are required to undergo this test as part of their treatment and most are prescribed a standard test of exercise capacity based on the (modified) Bruce protocol performed on a treadmill. In these cases, although not directly measured, VO_2 max may be predicted (calculated) using a formula that takes into account the duration of the test. In the

present study the standard formula[7] proposed by Bruce, Kusumi, and Hosmer (1973) for cardiac populations was used for people that had undergone a (modified) Bruce protocol test instead of CPEX. The correlation between measured VO_2 max and predicted (using the formula) is .93 for women and .87 for men with cardiac conditions (Bruce et al., 1973). For this reason, it was deemed a reliable way of obtaining comparable exercise capacity measurements for participants having different tests.

Ventricular function was documented by recording left and right ventricular ejection fraction based on the results from the most recent MRI (where available) and ECHO tests. For people with SV ventricular function was based on the function of the "single ventricle".

5.5.3. Psychosocial variables

Participants completed the same psychosocial measures in the cross-sectional and longitudinal studies. The psychosocial questionnaires are displayed in Appendix M. Participants were asked to respond to the questionnaires in relation to their ACHD.

5.5.3.1. Primary outcome – health-related quality of life

A disease-specific and a generic measure of HRQoL were utilised in this study. The generic measure was used to allow comparisons between people with ACHD and normative data. The disease-specific measure was used to study ACHD-specific HRQoL issues. Both measures were selected on the basis that together they address the multi-dimensionality of HRQoL (physical, occupational, psychological, interpersonal, and somatic sensation elements) highlighted in section 2.2.2 (page 47).

[7] VO2 max= 2.327*time(min) + 9.48 (Bruce et al., 1973)

Generic HRQoL – Short-Form 36 Health Survey Version 1®[8]

The SF-36v1® (Ware & Sherbourne, 1992) is the single most commonly used generic measure of HRQoL in healthy and ill populations. This measure was also identified in the systematic review (see Chapter 2) as the most frequently used generic HRQoL measure in ACHD. A benefit of the SF-36v1® is the use of norm-based scores (NBS), which allows for easy comparison with general population norms and facilitates interpretability and guards against floor and ceiling effects. A study of a large general population sample indicated that the SF-36v1® appeared to measure HRQoL more accurately, compared with the WHOQOL-Bref which tended to measure more global QoL (Huang, Wu, & Frangakis, 2006). Furthermore, a review of common HRQoL measures in ischemic heart disease found that the SF-36v1® offered more reliable, valid, and sensitive assessment of HRQoL compared with the Nottingham Health Profile and SIP (Dempster & Donnelly, 2000).

This 36-item instrument has eight subscales, which may be subsequently combined to form two components (Ware & Kosinski, 2001b). The subscales include physical functioning (10 items), role physical (role limitations due to physical health; 4 items), bodily pain (2 items), general health (5 items), which form the physical component summary, and vitality (4 items), social functioning (2 items), role emotional (role limitations due to mental/emotional health; 3 items), and mental health (5 items), which form the mental component summary. The measure includes an additional item (health transition), which is not used in the subscale scoring and thus was not included in the present study. The definitions of the SF-36v1® subscales are presented in Table 5.2.

[8] License was acquired for the use of SF-36v1©. The license agreement can be seen in Appendix N.

Table 5.2. Definition of the SF-36v1® subscales

	Subscale	*Definition*[*]
Physical component summary	Physical functioning	Performance of physical activities such as self-care, walking, and vigorous physical activities
	Role physical	The degree to which physical status interferes with a person's typical role activities
	Bodily pain	Intensity, duration, and frequency of bodily pain and limitations in usual activities due to pain
	General health	The beliefs and evaluations of a person's overall health
Mental component summary	Vitality	Feelings of energy; the absence of fatigue
	Role emotional	The degree to which emotional status interferes with a person's typical role activities
	Social functioning	The degree to which a person develops and maintains social relationships
	Mental health	A person's emotional, cognitive and intellectual status

[*]As defined by Saris-Baglama et al. (2010) in the QualityMetric Health Outcomes™ Scoring Software 4.0, User's Guide.

Participants are asked to rate their health status during the past 4 weeks. Response formats include 3- and 5-point Likert scales. The official software (QualityMetric Health Outcomes™ Scoring Software 4.0) was used for scoring the SF-36v1® subscales (Saris-Baglama et al., 2010). The eight subscales are linearly transformed into *T*-scores (i.e. NBS), with a mean of 50 and a standard deviation of 10. The software uses the 1998 general U.S. population norms. A NBS ranging between 0 and 100 is obtained for each subscale, with higher scores representing better HRQoL. The scoring software also produces the aggregated physical and mental component summaries (Maruish & DeRosa, 2009).

The measure has good content, concurrent, criterion, and construct validity and good internal consistency (α≥ .78 for subscales) (McHorney, Ware, Lu, & Sherbourne, 1994), which has been replicated in a significant amount of studies with different patient populations (Turner-Bowker, Bartley, & Ware, 2002). In addition to validity and reliability considerations, McHorney et al. (1994) noted ceiling (maximum scores) and floor (minimum scores) effects in the role physical and role emotional subscales as these represent the more coarse subscales in terms of response categories. However, empirical studies have shown that the SF-36 rarely misses significant differences in group level comparisons (Katz, Larson, Phillips, Fossel, & Liang, 1992) and reviews critically examining HRQoL measures in cardiac disease reported that, of all the generic HRQoL measures, the SF-36 appears to have fewer floor and ceiling effects and performs best (Thompson & Yu, 2003).

Disease-specific HRQoL – Congenital Heart Disease-TNO/AZL Adult Quality of Life

The CHD-TAAQOL is a disease-specific HRQoL measure for people with ACHD (Kamphuis et al., 2004). Its development is based on the pre-existing generic core measure, the TAAQOL by the Netherlands Organisation for Applied Scientific Research Academic Medical Centre. At the time of the present study, the CHD-TAAQOL was the only available measure specifically designed to assess HRQoL in ACHD, thus the selection of a disease-specific instrument was limited to this measure. The 26-item scale has three subscales: symptoms (9 items), worries (10 items), and impact cardiac surveillance (7 items). The definitions of the CHD-TAAQOL subscales are presented in Table 5.3.

Table 5.3. Definition of the CHD-TAAQOL subscales

Subscale	***Definition****
Symptoms	Specific cardiac symptoms (e.g. shortness of breath) weighted by emotional response to symptoms
Worries	ACHD-specific worries (e.g. having children) weighted by emotional response to worries
Impact cardiac surveillance	Cardiac surveillance weighted by emotional response to cardiac surveillance

*As defined by Kamphuis et al. (2004).

Each item consists of two questions. The first question refers to health and the second question refers to the emotional impact in case of a health status problem. Participants are asked about the frequency of specific problems during the last month except for the impact cardiac surveillance scale where participants are questioned about the last twelve months. For example:

[Symptom frequency] "How often in the last month did you look pale?"

[Emotional impact] "How much did that bother you?"

The response format for problem frequency consists of a three-point Likert scale: 1= *Never*, 2= *Occasionally*, 3= *Often*. The response format for the impact cardiac surveillance scale is: 1= *No*, 2= *Yes*. The response format for emotional impact is: 1= *Not at all*, 2= *A little*, 3= *Quite a lot*, 4= *Very much*. One single score is given for each pair of items (functional item and the corresponding emotional item). For example, if a person indicates that the occurrence of a particular problem was *often* but was not causing any emotional impact, then a score of 4 is assigned. The subscale scores are then linearly transformed, ranging from 0 to 100, with higher

scores indicating better HRQoL. The scoring procedures were conducted based on the official algorithms provided by Kamphuis et al. (2004).

The CHD-TAAQOL has good reliability (α= .77 for symptoms, α=.78 for impact cardiac surveillance, α= .82 for worries) and construct, convergent, and discriminant validity and appears to be free from floor and ceiling effects (Kamphuis et al., 2004).

5.5.3.2. Psychosocial variables

Illness perceptions – Brief Illness Perceptions Questionnaire

The Brief Illness Perceptions Questionnaire (Brief IPQ; Broadbent, Petrie, Main, & Weinman, 2006) is the brief version of the original Illness Perceptions Questionnaire (IPQ; Weinman, Petrie, Moss-Morris, & Horne, 1996) and its subsequent revised version (IPQ-R; Moss-Morris, Weinman, Petrie, Horne, Cameron, & Buick, 2002). The original IPQ measure was developed to measure the five illness representations of the SRM and is therefore theoretically derived. The IPQ and its revised and brief versions are the most commonly used measures of illness perceptions in cardiac disease as well as other chronic illnesses (Broadbent, Wilkes, Koschwanez, Weinman, Norton, & Petrie, 2015; Hagger & Orbell, 2003).

The Brief IPQ is a 9-item scale that provides rapid assessment of cognitive and emotional illness perceptions including consequences, timeline, controllability, curability, identity, concern, coherence, emotional representation and causal dimensions. Because of the congenital nature of ACHD, the causal dimension was not included in the present study. Definitions of the Brief IPQ subscales are presented in Table 5.4.

Table 5.4. Definition of the Brief IPQ subscales

Subscale	*Definition*[*]
Consequences	The expected effects of the illness
Timeline	How long the individual believes the illness will last
Controllability	The extent to which the individual believes that they can control the illness
Curability	The extent to which the individual believes that they can recover from the illness
Identity	The degree of experience of severe symptoms from the illness
Concern	The degree of concern about the illness
Coherence	The degree of understanding about the illness
Emotional representation	The extent to which the individual has negative reactions such as fear, anger, and distress from their illness

[*]As defined by Weinman et al. (1996) and Broadbent et al. (2006).

Each subscale is measured by a single item, which is rated using a continuous linear scale ranging from 0 to 10. Higher scores represent stronger perceptions in the subscale measured.

The brief version was selected over the IPQ-R (>80 items) to minimise participant burden. It is considered an appropriate alternative to the fuller version when a large number of variables is studied as it has good concurrent validity with the IPQ-R (Broadbent et al., 2006). Furthermore, Leventhal and Nerenz (1985) have argued that since illness perceptions are now well-defined, single items are more suited to their assessment rather than multiple items assessing latent factors. The Brief IPQ is also recommended for use in longitudinal research to study changes in illness perceptions over time (Petrie et al., 2007). Compared with the original IPQ, the Brief IPQ covers a wider array of illness perceptions including illness coherence and emotional

representations which are not addressed in the original measure. The Brief IPQ has acceptable test-retest reliability (α= .70 for consequences, α= .67 for timeline, α= .63 for controllability, α= .55 for curability, α= .65 for identity, α= .66 for concern, α= .48 for coherence, α= .65 for emotional representation) and good predictive validity in people recovering from myocardial infarction (Broadbent et al., 2006). The measure also has good discriminant validity, which is evidenced by its ability to distinguish between different illnesses (Broadbent et al., 2006). Data on floor and ceiling effects in the Brief IPQ are lacking but one study in low back pain indicates that ceiling effects are evident only in the timeline subscale (Løchting, Garratt, Storheim, Werner, & Grotle, 2013). More recently, a meta-analysis further demonstrated that the measure has good concurrent validity as the associations are consistent with theory and previous research, discriminant validity, predictive validity through longitudinal studies, and sensitivity to change through longitudinal studies and randomised controlled trials in various chronic illnesses including cardiovascular disease (Broadbent et al., 2015).

Coping strategies – Brief COPE

The Brief COPE was developed by Carver (1997) and it is an abbreviated version of the COPE Inventory (Carver, Scheier, & Weintraub, 1989). The Brief COPE assesses individuals' coping styles and strategies. The measure considers coping as motivated action and is focused on the self-regulatory processes underlying people's coping efforts. The COPE and Brief COPE measures are multidimensional and encompass a wide range of coping strategies. A study that compared the COPE with other well-established measures including the Ways of Coping Questionnaire (WCQ; Folkman & Lazarus, 1988) and the Coping Strategy Indicator (CSI; Amirkham,

1990) found that the COPE explained the most variance in psychosocial outcomes such as mood and QoL (Clark, Bormann, Cropanzano, & James, 1995). Furthermore, a large number of studies guided by the SRM have used the COPE measure to assess coping strategies in various chronic illnesses in the past (Hagger & Orbell, 2003) and more recently the Brief COPE has been the preferred method of assessing coping strategies in multiple studies in diabetes, HIV, chronic heart failure, and cancer (Bose, Bjorling, Elfstrom, Persson, & Saboonchi, 2015). For these reasons, the Brief COPE was also utilised in the present study.

The instrument consists of 28 items, which measure 14 distinct coping reactions (2 items each), from adaptive to problematic. The fourteen subscales and their definitions are presented in Table 5.5.

Participants are asked to indicate the degree to which they usually use each strategy when they experience stress related to their condition. The response format consists of a four-point Likert scale: 0= *I don't do this at all*, 1= *I do this a little bit*, 2= *I do this a moderate amount*, 3= *I do this a lot*. Mean scores for each subscale are obtained from the two items and they can range from 0 to 3, where higher scores indicate more use of the specific coping strategy. The author suggests that each subscale should be treated separately in the analysis.

This brief measure was created in response to criticism that the full version was too burdensome for respondents in terms of length of time to complete the questionnaire and because of the redundancy of items (n= 60). Carver (1997) demonstrated the soundness of the internal structure of the Brief COPE through exploratory factor analysis on the item set to obtain factor loadings, which indicated that the measure's factor structure was very similar to that of the full version.

Table 5.5. Definition of the Brief COPE subscales

Subscale	*Definition*[*]
Active coping	Taking active steps to try to remove or circumvent the stressor or to ameliorate its effects
Planning	Thinking about how to cope with a stressor (thinking about steps and action plans)
Positive reframing	Construing the stressor in positive terms
Acceptance	Accepting the reality of a stressful situation
Humour	Using humour to reduce the stressor
Religion	Tendency to turn to religion in times of stress
Using emotional support	Getting moral support, sympathy, or understanding
Using instrumental support	Seeking advice, assistance, or information
Self-distraction	Engaging in activities that serve to distract the person from thinking about the stressor
Denial	Trying to push the reality of the stressor away
Venting	Venting whatever distress or upset one is experiencing
Substance use	Using alcohol or other drugs to reduce the stressor
Behavioural disengagement	Reducing one's effort to deal with the stressor or giving-up
Self-blame	Criticising oneself for responsibility in the situation

Note. "Stressor" in the present study refers to stress relating to ACHD.
[*]As defined by Carver et al. (1989) and Carver (1997).

Considering that reliability is influenced by the number of items in a scale, the Brief COPE has minimally acceptable reliability with the values in each scale exceeding .50, and good construct validity (Carver, 1997). The theoretically-based 14-factor structure is also supported by a recent study of 606 adults through confirmatory factor analysis (Monzani, Steca, Greco, D'Addario, Cappelletti, & Pancani, 2015). The same study also reports good and excellent reliability across the 14 coping

dimensions and recommends the use of the Brief COPE as opposed to the full version and other lengthy coping instruments (e.g. WCQ) when a battery of measures is administered. Floor and ceiling effects in the Brief COPE have not been reported in the literature.

Anxiety – Six-item Short-Form of Spielberger State-Trait Anxiety Inventory

Anxiety was assessed using the STAI-6 developed by Marteau and Bekker (1992). This instrument is the abbreviated version of the 20-item state scale of the Spielberger State-Trait Anxiety Inventory (STAI) (Spielberger, Gorsuch, Lushene, Vagg, & Jacobs, 1983), which is the most frequently used measure of anxiety in cardiac populations (De Jong & Hall, 2006) and has also been used in ACHD (Müller et al., 2013a). The STAI-6 measures how a person feels at a given moment. It consists of six items, three indicating the absence of anxiety and three indicating the presence of anxiety. Participants are asked to give answers on statements on a four-point Likert scale using the following response format: 1= *Not at all*, 2= *Somewhat*, 3= *Moderately*, 4= *Very much*. The three items indicating the absence of anxiety are reverse-scored. The mean score of the scale is obtained from the six items and can range between 1 and 4, where higher scores reflect higher anxiety levels.

The short form was specifically designed for use in settings where time constraints or pragmatic considerations preclude the use of the full 20-item and it has been shown to be more acceptable by study participants compared with the full version (Marteau & Bekker, 1992). The STAI-6 has good reliability (α= .82) and concurrent validity when applying the six item questionnaire and the full-form of the state anxiety scale to four study groups (n=261) (Marteau & Bekker, 1992). The STAI-6

was also found to be as sensitive as the full-form scale to different degrees of anxiety. More recent evidence supports the finding that the STAI-6 is a reliable and valid instrument (Court, Greenland, & Margrain, 2010; Tluczek, Henriques, & Brown, 2009). Data on floor and ceiling effects in the STAI-6 are generally lacking but a study in cancer patients indicated the absence of such effects (van den Bergh, Korfage, Borsboom, Steyerberg, & Essink-Bot, 2009).

Depression – Center for Epidemiologic Studies Depression Scale

Depression was assessed with the abbreviated version of the CES-D (CES-D 10) which was developed by Andersen, Malmgren, Carter, and Patrick (1994). The original 20-item scale was developed by Radloff (1977). There are several measures of depression which have been utilized within the cardiac and ACHD literature including the BDI, HADS, and CES-D, all of which have similarly good psychometric properties. However, CES-D has been shown to be cognitively simpler (assessed by length, readability, and linguistic problems) compared with the BDI and HADS (Shumway, Sentell, Unick, & Bamberg, 2004). Both the brief and full versions, have been applied in a range of health settings since their development and have also been used in research involving spinal cord injury, diabetes and cardiovascular disease. CES-D 10 measures depressive symptomatology and consists of 10 items, two of them worded in the positive direction.

Participants rate how often each depressive symptom occurred during the past week. Responses are organised on a 4-point Likert scale: 0= *Rarely or none of the time (less than 1 day)*, 1= *Some or a little of the time (1-2 days)*, 2= *Occasionally or a moderate amount of the time (3-4 days)*, 3= *All of the time (5-7 days)*. The two positively worded items are reverse-scored. The summed score of the scale is

obtained from the 10 items and can range between 0 and 30, where higher scores indicate higher levels of depressive symptoms. There is considerable variation in the literature regarding a cut-off score to indicate clinical levels of depression. The cut-off score of 10 suggested by Andersen and colleagues (1994) was used in the present study; a score of 0 to 10 was classified as "without depressive symptoms" and a score of more than 10 was classified as "with depressive symptoms". At the cut-off point of 10, Boey (1999) found high sensitivity (.85) and specificity (.80), with a reduced misclassification rate at 17.5% compared with 23.5% when a cut-off point of 8 was used. In an outpatient sample of 1024 people with coronary heart disease the CES-D 10 had 76% sensitivity and 79% specificity compared with the Diagnostic Interview Schedule (McManus, Pipkin, & Whooley, 2005).

The brief version was developed to minimise participant response burden in multi-item surveys and research with physically ill populations as it takes just two minutes to complete. The CES-D 10 has acceptable convergent validity, internal consistency (α= .78 to .79), test-retest reliability (α=.44) over a period of three years, and predictive accuracy when compared to the original 20-item measure (Boey, 1999). Test-retest reliability over a period of 22 days was found to be .59 by authors (Andersen et al., 1994). Studies in chronic illnesses have reported that the CES-D 10 does not exhibit floor and ceiling effects (Amtmann et al., 2014; Miller, Anton, & Townson, 2008).

Perceived social support – Multidimensional Scale of Perceived Social Support

The Multidimensional Scale of Perceived Social Support (MSPSS) is a measure of subjective assessment of social support adequacy (Zimet, Dahlem, Zimet, & Farley, 1988). The MSPSS was selected over traditional network measures in order to

determine perceived social support, as opposed to the quantity of social support available, which is often assessed by the size and density of the network. Other multidimensional measures of perceived social support like the Medical Outcomes Study Social Support Survey (MOS-SSS; Sherbourne & Stewart, 1991) and the Interpersonal Support Evaluation List (ISEL; Cohen & Hoberman, 1983) are lengthy and do not distinguish between various sources of support. The Perceived Social Support Scale (PSSS; Procidano & Heller, 1983) is an alternative measure that assesses perceived support from family and friends but it has limited response options (yes/no), is lengthy, and does not assess support from a significant other. Therefore, the MSPSS was selected because a) it assesses multiple aspects of perceived social support from three sources, b) it is brief and concise compared with other measures, and c) it has been used extensively in cardiac populations (Stafford et al., 2009) and more recently in ACHD (Pike et al., 2012).

MSPSS assesses perceived social support from family, friends, and significant other using 12 items (4 items in each subscale). Support is assessed using a 5-point[9] Likert scale using the following response format: 1= *Strongly disagree*, 2= *Disagree*, 3= *Neither disagree nor agree*, 4= *Agree*, 5= *Strongly agree*. The mean score is calculated for each subscale as well as a mean total score of perceived support and can range between 1 and 5. Higher scores indicate higher levels perceived social support.

The instrument has construct validity, good internal consistency both at scale and subscale level ($\alpha \geq .85$) across different populations, and good test-retest reliability

[9] The revised version of the MSPSS utilises a 7-point scale. However, both scales were used by the authors of the measure (Zimet et al., 1990; Dahlem, Zimet, & Walker, 1991) and others (Cheng & Chan, 2004; Wongpakaran & Wongpakaran, 2012).

(.85) (Canty-Mitchell & Zimet, 2000; Zimet et al.,1988; Zimet, Powell, Farley, Werkman, & Berkoff, 1990). Ceiling effects have been observed in the MSPSS in some studies (Sawyer, Ayers, Young, Bradley, & Smith, 2012), while others did not find such effects (Zhou et al., 2015).

5.6. Statistical analysis

Statistical analysis was performed using IBM SPSS versions 20.0 and 21.0.

5.6.1. *A priori* power analysis

To estimate a statistically sufficient sample size for the cross-sectional study, G-Power (Faul, Erdfelder, Lang, & Buchner, 2007) was used to calculate the required number of participants for each diagnostic group. Power calculations were based on a four-group, cross-sectional study utilising one-way ANOVA to detect significant group differences in HRQoL. A systematic review of a large number of studies has indicated that a 0.5 S.D. is the minimally important difference in HRQoL in chronic illnesses, irrespective of whether the instrument is generic or disease-specific (Norman, Sloan, Wyrwich, 2003). Therefore, an approximately medium effect size (Cohen, 1988) was adopted in the power calculations to detect differences in the generic and disease-specific HRQoL measures. Sample size calculations indicated that a total sample size of 360 participants should be sought, with 90 participants per diagnostic group. These calculations were based upon equal group sizes, 90% power to detect differences at the .05 significance level, and an effect size of $f = .20$. The power calculations were based on high power (90%) to allow for possible loss of power ≤ 10%, in case the required sample size was not achieved.

For the longitudinal study, literature was consulted for guidance on the appropriate sample size for measuring change over time using Hierarchical Linear Models

(HLM). At present, there is no gold standard for calculating the sample size required for HLM, because such calculations are mathematically complicated and require parameters unknown to the researcher prior to data collection (Hayes, 2006). Tabachnick and Fidell (2007), however, suggest that at least 60 participants are required for each level of the model. As the data was structured into two levels (section 5.6.8.4, page 165), a sample of minimum 120 participants was required for the study to have sufficient power.

5.6.2. Data screening

Data were screened to ensure that all variables fell within possible ranges. All values that were out of range were crosschecked using the raw questionnaire data and corrected where necessary.

5.6.3. Missing value analysis

The same approach to missing value analysis and imputation was performed for the cross-sectional and longitudinal data. Mean item replacement was undertaken when calculating scale scores for cases that had ≤ 50% of items missing before conducting scale imputation, as suggested by Graham (2009). This method and the subsequent scale imputation excluded the SF-36v1®, for which missing data estimations are conducted within the official scoring software (see section 5.6.3.1, page 156 for description). Missing value analysis was performed at scale level except for demographic and clinical variables. Little's Missing Completely At Random (MCAR) test was conducted to check if differences existed between the missing values and the observed values (Little, 1988). An insignificant result ($p > .01$) suggests that data are MCAR and that imputation is appropriate.

Traditional methods of imputation include mean imputation, last observation carried forward, hot-deck imputation, regression imputation, and multiple imputations among others. Given that there was less than 5% missing data in the dataset (see Appendix O & Appendix P), using almost any of the above methods would yield similar results (Tabachnick & Fidell, 2007). Missing data was imputed at scale or subscale level using Bayesian stochastic regression (using chained equations, Markov Chain Monte Carlo-MCMC). Data were not imputed for the 11 cases missing the whole questionnaire packs of the cross-sectional study and for cases that were missing the whole questionnaire pack of the longitudinal study.

The imputation process was conducted in sets of variables (i.e. clinical, demographic, psychosocial questionnaires) due to logistical reasons (i.e. processing power and time). However, the predictive model for all imputations had the same predictors; specifically, all other variables within the dataset.

5.6.3.1. Missing data on the SF-36v1®

The SF-36v1® licence purchase is supplemented by a scoring software (QualityMetric Health Outcomes™ Scoring Software 4.0) (Saris-Baglama et al., 2010), which has built-in algorithms that deal with missing data prior to scoring the scales. The program offers two ways of estimating missing data:

i) Half-scale rule, where missing values are estimated based on the available data for the scales that have at least half of the items completed. The average score of the completed items replaces the missing values and scales are rendered missing when more than half of the items are missing. In turn, the component summary scores are calculated only when there are available data (either actual or estimated) on all eight scales.

ii) Maximum data recovery, where missing values are estimated when at least one item in the scale is completed. A scale is rendered missing when all items in that scale are missing. The component summary scores are calculated when at least seven of the eight scales have completed data (either actual or estimated). One of the seven completed scales that are required is physical functioning for the physical component summary and mental health for the mental component summary, in order for these summaries to be calculated.

For the present study, the official scoring software was used as per the scoring manual. The SF-36v1® data were exported into an excel spreadsheet and imported into the scoring software, according to the manual. In order to make best use of the data and owing to the low percentages of missing data, the maximum data recovery method was selected for missing data estimations. Scales that were still missing after the scoring process were recorded as such in order to avoid invalidating the software's missing data estimations. After the appropriate missing data estimations and scoring procedures were completed using the software, the SF-36v1® dataset was imported back to the main imputed SPSS dataset.

5.6.4. Outliers

Outliers are defined as observations that *"deviate so much from other observations as to arouse suspicions that it was generated by a different mechanism"* (Hawkins, 1980) and can be influential in statistical tests. Outliers were identified by graphical inspection and by standardised *z* scores greater than ±3.29 (Tabachnick & Fidell, 2007). There is disagreement amongst researchers on whether outliers should be removed from the analysis. Provided they are not erroneous data entries, outliers may contain valuable information and can represent the inherent variability of the

variable in question (Orr, Sackett, & DuBois, 1991). In addition, removing cases that have random outlying scores would result in significant reduction of the sample size. Therefore, outliers were retained in order to maximise sample size and because it was felt that they would provide valuable information regarding the relationships studied. Multivariate outliers are discussed in section 5.6.8.3 (page 162).

5.6.5. Normality

Some multivariate statistical tests require that the data be normally distributed. Because multivariate normality is not easily tested without conducting multiple variable combinations (Tabachnick & Fidell, 2007), univariate normality was studied. Tabachnick and Fidell (2007) recommend that normality tests are based on the grouped data (in this case diagnostic group) when the analysis consists of group comparisons. Statistically, the Shapiro-Wilk test can be used to test whether skewness and kurtosis are statistically significant. A significant result indicates non-normal distribution. However, in large samples (n> 200) the impact of skewness and kurtosis deviations diminishes making this test over-sensitive to violations of normality (Field, 2013). In the present study the distribution of the variables was also examined visually by exploring the data visually using Q-Q plots and histograms for skewness and kurtosis.

Floor and ceiling effects in the measures were also assessed. A floor effect occurs when a large proportion of people score at the lowest possible score, whereas a ceiling effect occurs when a large proportion of people score at the highest possible score. These effects may make it difficult to distinguish between groups and reduce measure responsiveness in longitudinal studies examining change (Fayers & Manchin, 2009). More specifically, a questionnaire may be unable to detect further

deterioration in a group of people if a high proportion of them scored at the scale minimum or further improvement if a high proportion scored at the scale maximum. In the present study, the distribution of the variables and the proportion of people scoring at the scale minimum/maximum were examined in order to assess floor and ceiling effects. When the median values were the same as the minimum or maximum values of a scale and a proportion of >25% of people scored at the scale minimum or maximum then floor and ceiling effects were considered substantial (McHorney et al., 1994). When ceiling and floor effects are present, data may be transformed but transformations may result in problems regarding interpretation of the data (Osborne, 2002).

A majority of the analyses presented in this thesis consisted of regressions, where normal distribution refers to the residuals rather than the variables. Section 5.6.8.3 (page 162) describes the method used for exploring the distribution of the residuals in the regression-based analyses.

Both parametric and non-parametric analyses were initially conducted where necessary; however, parametric analyses were presented when both approaches yielded similar results. This approach was followed because non-parametric tests may sometimes lead to loss of power for near-normal data (Pallant, 2013).

5.6.6. Internal reliability of scales and subscales

To assess the internal reliability of the measures, Cronbach's α was calculated and reported for scales or subscales containing three or more items. Because Cronbach's α is sensitive to the number of items in a scale, the inter-item correlation was also calculated and reported for two-item scales or subscales (Pallant, 2013). Cronbach's

α of >.70 (Pallant, 2013) and inter-item correlations between .20 and .50 are generally considered optimal (Clark & Watson, 1995).

5.6.7. Preliminary analyses

For all preliminary and main analyses, the level of statistical significance was set at $p < .01$ in order to minimise family-wise error due to multiple testing.

Means and standard deviations for continuous variables and frequencies with percentages for categorical variables were presented. One-way ANOVAs were used to examine differences in continuous demographic and clinical characteristics and questionnaire data between the four diagnostic groups. Chi-square (χ^2) tests were employed to test associations between categorical demographic/clinical variables and the four diagnostic groups. One-way ANOVAs and chi-square tests were also employed to examine differences in continuous demographic and clinical characteristics and questionnaire data between responders and non-responders in the longitudinal study. The Fisher's exact probability test was reported where cell counts were lower than five. Cramer's V (φ_c) (.10= small, .30= medium, .50= large) and eta squared (η^2) (.01= small, .06= medium, .14= large) were reported for χ^2 tests and one-way ANOVAs respectively.

5.6.8. Main analyses

5.6.8.1. Research question #1a: HRQoL comparisons in people with ACHD with normative data from the general population

The use of *T*-scores (NBS) in the SF-36v1® enables direct comparisons between the participants' mean scores and the population average. The use of US population norms in the present study may pose some limitations as these are not culture-specific. However, there are several reasons that informed the decision to use US

normative data. Firstly, there are currently no official UK norms recommended by the licensing company (QualityMetric Incorporated, personal communication, April 5, 2013). Secondly, published UK normative data from four studies are either outdated (pre-1996), geographically restricted, age-restricted, and overall non-representative of the UK population resulting in substantial differences between them (Bowling, Bond, Jenkinson, & Lamping, 1999). Thirdly, the SF-36v1® licensing company encourages researchers to use the official scoring software which at present only uses US population norms. Fourthly, US norms have been used as a point of comparison in other international studies in ACHD (e.g. Bruto et al., 2007). In light of the above limitations the present study used the official scoring software, which utilises more recent healthy population norms albeit from a different country, a wide age range, and a more representative sample of a healthy population.

A cut-off point of ≥ 1 standard deviation was selected to assess the percentage of participants scoring below or above the general population mean, based on the authors' recommendation (Ware & Kosinski, 2001a). To test whether the mean generic physical and psychosocial HRQoL (SF-36v1®) scores of the total sample as well as each of the diagnostic groups differ statistically from the norm mean, one-sample *t*-tests were employed. An alternative way of analysis is to conduct comparisons with age- and gender-matched controls or normative data. However, the SF-36v1® was not administered to matched controls, owing to time limitations and lack of resources. In addition, age- and gender- normative data are currently only available for the latest instrument version (i.e. SF-36v2®) (QualityMetric Incorporated, personal communication, April 5, 2013). Furthermore, since the question was how much of a normal life people with ACHD lead, no comparisons

with other chronic illnesses were made. Effect sizes for mean differences were calculated using Cohen's *d* (.20= small, .50= medium, .80= large) (Cohen, 1988) along with 99% Confidence Intervals (CI).

5.6.8.2. Research question #1b: HRQoL comparisons between four diagnostic groups

To examine differences between the four diagnostic groups in physical, psychosocial, and disease-specific HRQoL, and due to the slightly unequal sample sizes in the four diagnostic groups, one-way ANOVA with Gabriel (for equal variances) or Games-Howell (for unequal variances) post-hoc tests were employed, based on 99% CI. The Welch test was implemented where necessary (for unequal variances). Effect sizes for the proportion of variance explained were calculated using η^2 (.01= small, .06= medium, .14= large).

5.6.8.3. Research question #2: factors associated with HRQoL in ACHD

In order to examine the demographic, clinical, and psychosocial factors associated with HRQoL hierarchical multiple regressions were performed. Outcome variables included generic physical HRQoL (physical component summary and its subscales), generic psychosocial (mental component summary and its subscales) and all three scales of the disease-specific (CHD-TAAQOL) measure. Categorical predictor variables with more than two groups were dummy coded where appropriate.

Bivariate linear regressions were employed first, in order to reduce the number of predictor variables entered into the regressions and therefore preserve power (Field, 2013). The general rules regarding adequate sample size in multiple regression are $n \geq 50+8m$ (m= number of independent variables) when testing a model and $n \geq 104+m$ for testing individual predictors (Tabachnick & Fidell, 2007). Both equations

were calculated for each regression and the highest *n* required was taken as a general guideline. All regression models had sufficient sample size as a result of using the bivariate regressions to reduce the number of predictor variables.

The entry method was used instead of stepwise as the latter does not necessarily produce the best model if there are redundant predictors, but rather produces a statistical model which may be biased (Harrell, 2001; Tabachnick & Fidell, 2007). Demographic and clinical factors were entered into the first blocks followed by psychosocial variables. Mood variables were entered in the last block as these were found to be highly correlated with HRQoL outcomes in previous research. The order of entry of the predictors in the regressions is displayed in Figure 5.1. NYHA functional status was not included in the hierarchical multiple regressions because it covered clinical function measured by other clinical variables in the models.

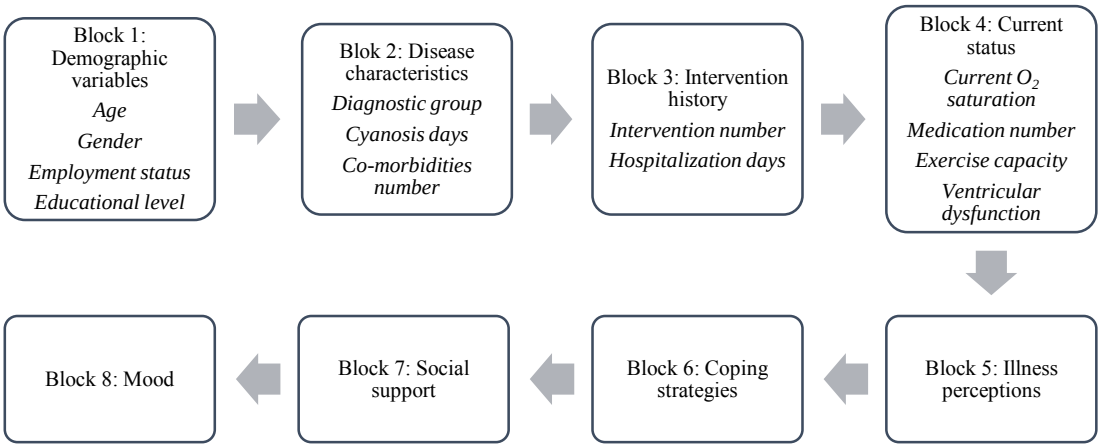


Figure 5.1. Order of entry of predictor variables in hierarchical multiple regressions

In order to assess whether the assumptions for regression analyses were met, evaluations were conducted for singularity, multicollinearity, independence of errors, outliers, normality, linearity, and homoscedasticity.

Singularity: To avoid singularity, either the subscale scores or the total scale scores were entered in the regressions. Specifically, for the perceived social support measure (MSPSS) the family, friends, and significant other support subscales were entered into the regressions, omitting the total scale score.

Multicollinearity: Prior to the regression analysis, correlations between the independent variables were investigated in order to check for strong correlations (≥ .80). This would indicate correlations that could potentially be problematic for the regression analysis. Collinearity diagnostics were also examined including Variance Inflation Factor (VIF), tolerance statistic, variance proportions, and eigenvalues. VIF indicates strong linear relationships between the independent variables and a general rule is that values ≥ 4 are indicative of possible problems, while values ≥ 10 are a definite cause of concern (Belsley, Kuh, & Welsch, 1980). Acceptable tolerance values were set at ≥ 0.20 with lower values indicating a problem (Field, 2013).

Independence of errors: The Durbin-Watson test was employed to test for independence of errors in the regressions. It tests whether there are correlations between adjacent residuals. Generally, a value close to 2 indicates no correlation and values less than 1 or greater than 3 may be problematic (Field, 2013).

Outliers: In the regressions, multivariate outliers were examined in detail and their influence on the overall model was assessed using Cook's distance (values > 1 were deemed problematic). In addition, leverage values greater than twice the average were deemed problematic. For the hierarchical linear models it was not appropriate to check for multivariate outliers given the independent variables were time, diagnostic group, and Diagnostic Group x Time which were categorical.

Normality, linearity, homoscedasticity: Frequency histograms and normal probability plots were examined to determine the distribution of the residuals. Scatter plots of standardized predicted scores against standardized residuals were examined to check for unusual patterns. The above assumptions are met provided that: the residuals are normally distributed about the predicted dependent variable scores, the residuals have a straight-line relationship with predicted dependent variable scores, and the residuals variance about predicted dependent variable scores is the same for all predicted scores (Tabachnick & Fidell, 2007). Non-linearity of the relationship between the independent variables and the dependent variable may lead to biased estimates of the true population values and heteroscedasticity may lead to type I errors (Osborne & Waters, 2002).

5.6.8.4. Research question #3: Changes over time in clinical, HRQoL, and other psychosocial factors

Hierarchical Linear Models (HLM) were used to examine changes over time in clinical, HRQoL, and psychosocial factors[10]. HLM are complex statistical models used to analyse variance in continuous outcome variables when the predictor variables have hierarchical structure. HLM involve both fixed and random effects (West, Welch, & Galecki, 2007).

HLM is a statistical analysis that has had an increasing use in health psychology research in recent years. HLM may be used to study complex relationships by including a number of covariates. The HLM method has a number of advantages over traditional methods such as repeated measures ANOVA [see Quené & van den Bergh (2004) or West (2009) for a more detailed description].

[10] Mixed within-between ANOVAs were used where HLM analyses did not converge.

In the present study, HLM was used over repeated measures ANOVA for two main reasons. Firstly, HLM account for the hierarchical structure of data evident in longitudinal designs where assessments at different time points are "nested" within individuals and are correlated (Figure 5.2). Traditional methods assume that the observations and residuals are independent. This may result in inflated standard errors of the regression coefficients. HLM allow specification of the within-individual error covariance structure that is most appropriate for the data. This is important because in longitudinal designs the assessment waves may be unequally spaced (Shek & Ma, 2011). Various covariance structures may be assessed by comparing the information criteria of each model, the commonest being -2 log likelihood. Lower -2 log likelihood values indicate better model fit.

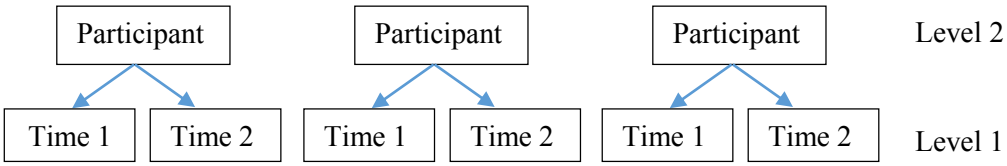


Figure 5.2. Hierarchical structure of data in the present study

Secondly, HLM are quite robust against missing data (West et al., 2007). Traditional methods exclude cases when they have missing data from an administration point. With HLM, the data from completed assessments are still used in the analysis even if an individual did not complete both time points thus preserving statistical power.

Before conducting HLM, the Intra-Class Correlation (ICC) was calculated in order to confirm the hierarchical structure of the data. The ICC estimates the amount of variance in the outcome explained by the grouping structure (Hox, 2002). High values suggest that the assumption of independence of errors is violated and that errors are correlated, hence the hierarchical structure is supported (Tabachnick &

Fidell, 2007). The ICC values confirmed the hierarchical structure of the data in this study.

HLM was used to examine changes in clinical and psychosocial factors over time and across diagnostic groups. Diagnostic group, time (entered as a categorical variable), and the interaction Diagnostic Group x Time were entered as fixed effects into the model and "Participant ID" was entered as a random effect. Due to the variability in the timing of the longitudinal assessments, differences were tested between the diagnostic groups to establish whether there was a need for it to be adjusted during HLM.

The selected method of estimation was the restricted maximum likelihood (REML) method. As opposed to maximum likelihood (ML), REML provides unbiased estimates of covariance parameters by considering the loss of degrees of freedom in the estimation of fixed effects (West et al., 2007). A "step-up" approach to modelling was adopted, whereby models were built from simple to more complex (West et al., 2007). The following steps were followed: 1. Unconditional or "null" model, 2. Added the fixed effect of time, 3. Added a repeated statement where the covariance structure for the residuals was defined (diagonal), 4. Added the fixed effect of control variables (follow-up years), 5. Added the fixed effect of the predictor of interest (diagnostic group) as well as the Diagnostic Group x Time interaction. The diagonal covariance structure for the residuals appeared to be the most appropriate for the data, which achieved convergence. It assumes a heterogeneous variance at each time point and an independent covariance between measurement times.

At each modelling step, the model fit may be evaluated using the information criteria (e.g. -2 restricted log likelihood). When the REML estimation method is used,

significance tests may be conducted when comparing models that share the same fixed effects and only differ in random effects (Snijders & Bosker, 2012). For this reason, significance tests were not appropriate in the present study. However, smaller values in the information criteria generally suggest a better model fit.

A significant Diagnostic Group x Time interaction was interpreted as an indication that changes were different by diagnostic group. In order to examine significant mean differences pairwise comparisons were requested from SPSS using Sidak's adjustment for multiple comparisons. Standardized adjusted effect sizes for significant mean differences were calculated using Cohen's *d* (.20= small, .50= medium, .80= large) (Cohen, 1988).

CHAPTER 6 – SAMPLE CHARACTERISTICS

6.1. Prologue

This chapter presents the baseline characteristics of the sample. It begins with a description of participation rates in the cross-sectional study using a flow diagram and continues with a description of missing value analysis. Examination of data assumptions is presented before the description of participant demographic, clinical, and psychosocial characteristics along with differences between the four diagnostic groups. The final section includes a discussion of the sample characteristics with comparisons to other available literature.

6.2. Participation rates

Current estimations indicate that approximately 5000 people are followed-up by specialist cardiologists at The Heart Hospital, London. However, not all 5000 were active (and therefore recorded in paper records) at the time of the study (see section 5.4.3, page 132). A total of 1199 active patients were identified as potential participants and were assessed for eligibility for the present study (Figure 6.1). One hundred and twenty-three (10.3%) were excluded based on the inclusion/exclusion criteria. All eligible participants with TGA and SV diagnoses were invited to take part in the study due to the limited number of people with these diagnoses, while a random sample of eligible participants with ToF and Simple diagnoses were selected for invitation (n= 236 and n= 197 respectively). Out of those invited, 273 declined (38.6%) and 81 (11.4%) did not respond to the invitation and telephone calls. Out of the 354 (50%) participants who agreed to take part, twenty-nine (11.3%) withdrew from the study without providing a reason and 11 (3.1%) participants did not

complete the psychosocial questionnaire pack. The final sample used for the main analyses consisted of 303 participants. Demographic and clinical information was reported for all participants with available data in those variables (n= 314; response rate: 44%).

Due to ethical reasons in relation to accessing the patient records of individuals who did not consent, examining differences in key demographic and clinical characteristics between participants and non-participants was not possible, except for diagnostic group. Chi-square analysis indicated an association between diagnostic group and response to the invitation to participate in the study, $\chi^2(3)= 19.29$, $p< .001$; $\varphi_c= .17$. The TGA group was more likely and the Simple group less likely to respond and complete the study.

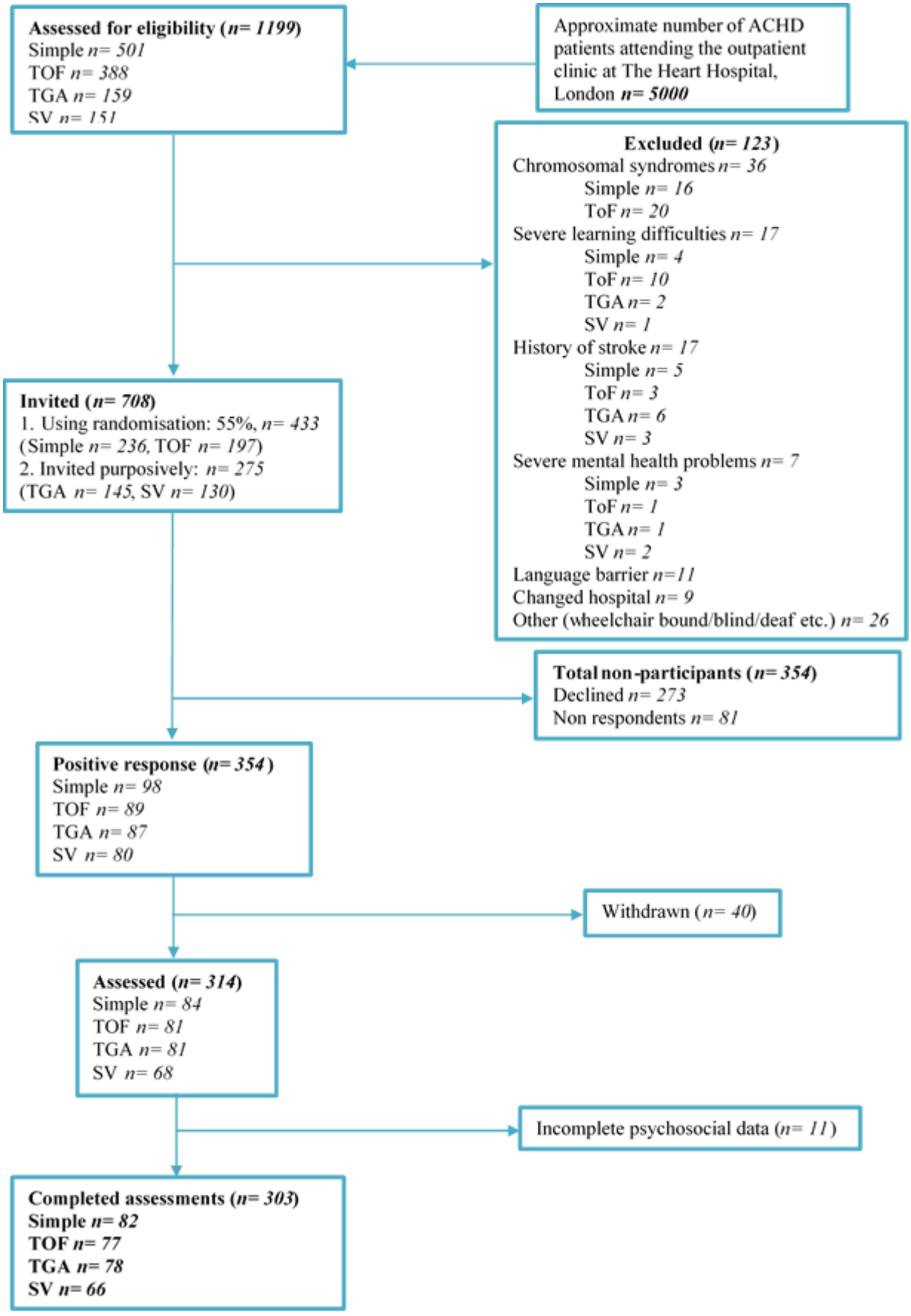


Figure 6.1. Flow diagram of participation rates for the cross-sectional study

6.3. Missing value analysis

Missing value analysis was performed at scale level except for demographic and clinical variables and is presented in Appendix O. In demographic and clinical characteristics, 61 (1.2%) items were missing out of 5024. In the psychosocial questionnaires 52 (0.4%) values were missing out of 12120 values. Little's MCAR test was conducted to check if differences existed between the missing values and the observed values (Little, 1988). The result was $p = .219$, suggesting that data were MCAR and that imputation is appropriate. Imputation was not performed for the eleven participants who did not complete the psychosocial questionnaires but for which clinical and demographic characteristics were available.

6.4. Data assumptions

6.4.1. Scale reliability

All scales and subscales of the psychosocial questionnaires used in the study showed satisfactory internal reliability based on Cronbach's α scores and/or inter-item correlations (Table 6.1) and were thus retained for the analyses.

Table 6.1. Internal reliability of scales/subscales used in the cross-sectional study

Scales & Subscales	*No. of Items*	*Cronbach α/ Inter-Item Correlations*[§]
SF-36		
Physical functioning	10	.91
Role physical	4	.88
Bodily pain	2	.88/ .82
General health	5	.80
Vitality	4	.87
Social functioning	2	.68/ .51
Role emotional	3	.85
Mental health	5	.82
CHD-TAAQOL		
Symptoms	9	.84
Worries	10	.82
Impact cardiac surveillance	7	.74
Brief COPE		
Planning	2	.69/ .53
Active coping	2	.70/ .53
Positive reframing	2	.73/ .57
Acceptance	2	.63/ .46
Humour	2	.90/ .81
Religion	2	.90/ .81
Seeking emotional support	2	.78/ .64
Seeking instrumental support	2	.82/ .69
Self-distraction	2	.58/ .41
Denial	2	.50/ .35
Venting	2	.62/ .45
Substance use	2	.93/ .87
Behavioural disengagement	2	.64/ .48
Self-blame	2	.62/ .48
STAI-6	6	.81
CES-D 10	10	.85
MSPSS		
Family support	4	.88
Friends support	4	.93
Special person support	4	.95

[§]Inter-item correlations are also reported for scales and subscales consisting of two items.

6.4.2. Normality

Normality was tested statistically using the Shapiro-Wilk test and visually by examining Q-Q plots and histograms for skewness and kurtosis. The Shapiro-Wilk test was significant ($p < .001$) for most variables. However, as described in section 5.6.5 (page 158) the test may be over-sensitive to violations of normality in large samples. Visual exploration of plots indicated a more normal distribution in some variables than the Shapiro-Wilk test suggested.

Due to the non-normality in some of the variables, floor and ceiling effects were examined by considering the median value of a scale and calculating the proportion of participants with the lowest or highest possible score in all measures. When the median values were the same as the minimum or maximum values of a scale and a proportion of >25% scored at the scale minimum or maximum then floor/ceiling effects were considered substantial (McHorney et al., 1994). Ceiling effects were observed in the role physical and role emotional subscales of the SF-36. Ceiling effects were observed only in the timeline subscales of the Brief IPQ. Floor effects were observed in the religion, denial, substance use, behavioural disengagement and self-blame subscales of the Brief COPE. No ceiling/floor effects were observed in the disease-specific HRQoL, anxiety, depression, and MSPSS measures.

Data were not transformed because preliminary transformations in some variables using square root and logarithmic base 10 failed to improve non-normality. In addition, transformations may result in problems regarding interpretation of the data (Osborne, 2002). It has also been argued that with large sample sizes the sampling distributions of means are normally distributed irrespective of univariate distributions according to the central limit theorem (Tabachnick & Fidell, 2007).

6.5. Demographic characteristics of the sample

The demographic characteristics of the total sample and each diagnostic group are displayed in Table 6.2.

Due to the small number of participant within some variables' categories, these were merged for some demographic variables. For ethnicity, all other groups other than "British" were categorised as "other". For marital status, the groups "married/civil partnership" and "in a relationship" were merged together and the groups "divorced", "separated", and "widowed" were merged into single. For educational level, the groups "primary education", "secondary education", "tertiary education" were merged into school level and the groups "undergraduate degree", "master's degree", and "doctorate degree" were merged into university level. For employment status, the groups "employed" and "self-employed" were merged together and the groups "student", "seeking job", "housewife/husband", "retired", "unable to work", and "long-term sick leave" were merged into unemployed.

Mean age was 33.2 (*S.D.* 10.7) years (range: 19-76) with 177 (56.4%) males and 137 (43.6%) females in the sample. Approximately half were married or in a relationship (50.6%). A total of 66.9% completed school level education, while 33.1% completed a higher degree. The majority were employed (70.4%), while 29.6% were unemployed.

There were age differences between diagnostic groups ($p < .001$). Games-Howell post-hoc comparisons indicated that the Simple group was older than the TGA (mean difference= 5.49, $p = .007$, 99% CI [0.22, 10.77]; $d = .51$) and SV (mean difference= 8.62, $p < .001$, 99% CI [3.05, 14.18]; $d = .76$). The ToF group was older than the SV

(mean difference= 6.18, *p*= .001, 99% CI [1.32, 11.03]). No other differences were observed between the diagnostic groups in demographic characteristics.

Table 6.2. Demographic characteristics of the sample by diagnostic group

Demographic Variable	*Total Sample* *n= 314*	*Simple* *n= 84*	*ToF* *n= 81*	*TGA* *n= 81*	*SV* *n= 68*	*Test Statistic*	*Sig.*	*Effect Size*
Age (*mean, S.D., range*)	33.2 (10.7)	37.1 (13.7)	34.6 (11)	31.6 (6.5)	28.4 (7.6)	$f(3,167.07)=$	**<.001**	$\eta^2= .09$
	18-76	19-76	19-66	19-50	18-58	10.29[§]		
Gender (*n*, %)						$\chi^2(3)= 10.22$	.017	$\varphi_c= .18$
Male	177 (56.4)	37 (44)	43 (53.1)	52 (64.2)	45 (66.2)			
Female	137 (43.6)	47 (56)	38 (46.9)	29 (35.8)	23 (33.8)			
Ethnicity[*] (*n,* **%**)								
British	266 (84.7)	68 (81)	72 (88.9)	72 (88.9)	54 (79.4)	$\chi^2(3)= 4.58$	.206	$\varphi_c= .12$
Other	48 (11.3)	16 (19)	9 (11.1)	9 (11.1)	14 (20.6)			
Marital status[*] (*n*, %)						$\chi^2(3)= 1.98$	.576	$\varphi_c= .08$
Married/relationship	159 (50.6)	45 (53.6)	44 (54.3)	36 (44.4)	34 (50)			
Single	155 (49.4)	39 (46.4)	37 (45.7)	45 (55.6)	34 (50)			
Educational level[*] (*n*, %)						$\chi^2(3)= 3.28$	.351	$\varphi_c= .10$
School level	210 (66.9)	53 (63.1)	55 (67.9)	60 (74.1)	42 (61.8)			
University level	104 (33.1)	31 (36.9)	26 (32.1)	21 (25.9)	26 (38.2)			
Employment status[*] (*n*, %)						$\chi^2(3)= 2.49$	.478	$\varphi_c= .09$
Employed	221 (70.4)	57 (67.9)	59 (72.8)	61 (75.3)	44 (64.7)			
Unemployed	93 (29.6)	27 (32.1)	22 (27.2)	20 (24.7)	24 (35.3)			

[*] Variables where categories were merged, [§] Welch Anova, φ_c: .10= small, .30= medium, .50= large. η^2: .01= small, .06= medium, .14= large.

6.6. Clinical characteristics of the sample

Details of the primary diagnosis by diagnostic group are presented in Table 6.3 and the remaining clinical characteristics are presented in Table 6.4.

Table 6.3. Primary diagnosis of the participants by diagnostic group

Diagnosis	***Total Sample n= 314 (n, %)***	***Simple n= 84***	***ToF n= 81***	***TGA n= 81***	***SV n= 68***
Transposition of the great arteries	83 (26.4)	0	0	81	2
Tetralogy of Fallot	81 (25.8)	0	81	0	0
Coarctation of the aorta	39 (12.4)	39	0	0	0
Ventricular septal defect	9 (2.9)	9	0	0	0
Secundum atrial septal defect	6 (1.9)	6	0	0	0
Atrial septal defect	18 (5.7)	18	0	0	0
Pulmonary valve stenosis	10 (3.2)	10	0	0	0
Sub-aortic stenosis	2 (0.6)	2	0	0	0
Tricuspid atresia	23 (7.3)	0	0	0	23
Double inlet left ventricle	14 (4.5)	0	0	0	14
Pulmonary atresia/intact ventricular septum	11 (3.5)	0	0	0	11
Pulmonary atresia/ ventricular septal defect	5 (1.6)	0	0	0	5
Double outlet right ventricle	5 (1.6)	0	0	0	5
Hypoplastic right ventricle	3 (1)	0	0	0	3
Isomerism	3 (1)	0	0	0	3
Hypoplastic left ventricle	1 (0.3)	0	0	0	1
Mitral atresia	1 (0.3)	0	0	0	1

Table 6.4. Clinical characteristics of the sample by diagnostic group

Clinical Variable	*Total Sample n= 314*	*Simple n= 84*	*ToF n= 81*	*TGA n= 81*	*SV n= 68*	*Test Statistic*	*Sig.*	*Effect Size*
Disease characteristics								
Co-morbidities no. (*mean, S.D., range*)	1 (1.1) 0-6	0.8 (1) 0-4	0.9 (1) 0-4	1.1 (1.2) 0-5	1.2 (1.1) 0-6	$f(3,310)= 1.40$	.243	$\eta^2= .01$
Arrhythmias (*n*, %)						$\chi^2(3)= 32.98$	**<.001**	$\varphi_c= .32$
Yes	83 (26.4)	7	15	34	27			
No	231 (73.6)	77	66	47	41			
Cyanosis days (*mean, S.D.*)	1317.4 (2712)	9.5 (87.3)	1510.6 (2251.9)	592.8 (1593.7)	3566 (4160.1)	$f(3,125.61)= 31.75^{\S}$	**<.001**	$\eta^2= .23$
Intervention history (*mean, S.D.*)								
Interventions no.	2.4 (1.4)	1.5 (.8)	2.4 (1.2)	2.6 (1.3)	3.2 (1.7)	$f(3,160.86)= 33.17^{\S}$	**<.001**	$\eta^2= .20$
Hospitalization days	40.9 (52.9)	17 (16.7)	46.5 (33.5)	35.4 (24.8)	70.2 (94.9)	$f(3,152.99)= 26.72^{\S}$	**<.001**	$\eta^2= .13$
Current status								
Medication no. (*mean, S.D.*)	1 (1.3)	0.8 (1.3)	0.5 (1.1)	1.1 (1.4)	1.5 (1.2)	$f(3,310)= 8.05$	**<.001**	$\eta^2= .08$
Current O_2 saturation (*mean, S.D.*)	95.9 (4.7)	98.2 (1.7)	97.3 (1.4)	96.2 (2.3)	91.1 (7.6)	$f(3,157.73)= 28^{\S}$	**<.001**	$\eta^2= .32$
NYHA class (*n*, %)						$\chi^2(3)= 6.14$	.105	$\varphi_c= .14$
Class I	275 (87.6)	79	70	71	55			
Class II, III, IV	39 (12.4)	5	11	10	13			
VO_2 Max (*mean, S.D.*)	28 (8.2)	30.3 (8)	27.2 (8.2)	27.2 (8.3)	27 (7.8)	$f(3,310)= 3.12$	.026	$\eta^2= .03$
Right ventricular function (*mean, S.D.*)	57.4 (8.7)	63.4 (5.1)	55.8 (8.9)	54.3 (7.6)	55.6 (9.5)	$f(3,161.22)= 36.76^{\S}$	**<.001**	$\eta^2= .18$
Left ventricular function (*mean, S.D.*)	60.9 (8.6)	64.7 (6.9)	59.9 (7.5)	62.8 (8.1)	55 (9.2)	$f(3,310)= 21.50$	**<.001**	$\eta^2= .17$

§Welch ANOVA.
φ_c: .10= small, .30= medium, .50= large. η^2: .01= small, .06= medium, .14= large.

6.6.1. Disease characteristics

There were no differences in the number of co-morbidities ($p > .01$) between diagnostic groups. A total of 24.6% of the sample experienced arrhythmias. Chi-square analysis indicated an association between diagnostic group and arrhythmias ($p < .001$). The TGA and SV were more likely to experience arrhythmias whereas the Simple group were less likely to.

There were differences in cyanosis days ($p < .001$) between diagnostic groups (Figure 6.2). The SV group experienced longer duration of cyanosis than the ToF (mean difference= 2055.41, $p = .002$, 99% CI [256.61, 3854.21]), TGA (mean difference= 2973.20, $p < .001$, 99% CI [1257.07, 4689.33]), and Simple (mean difference= 3556.49, $p < .001$, 99% CI [1924.65, 5188.33]). The ToF and TGA groups experienced longer duration of cyanosis than the Simple group (mean difference= 1501.08, $p < .001$, 99% CI [696.37, 2305.79] & mean difference= 583.29, $p = .008$, 99% CI [13.43, 1153.16] respectively).

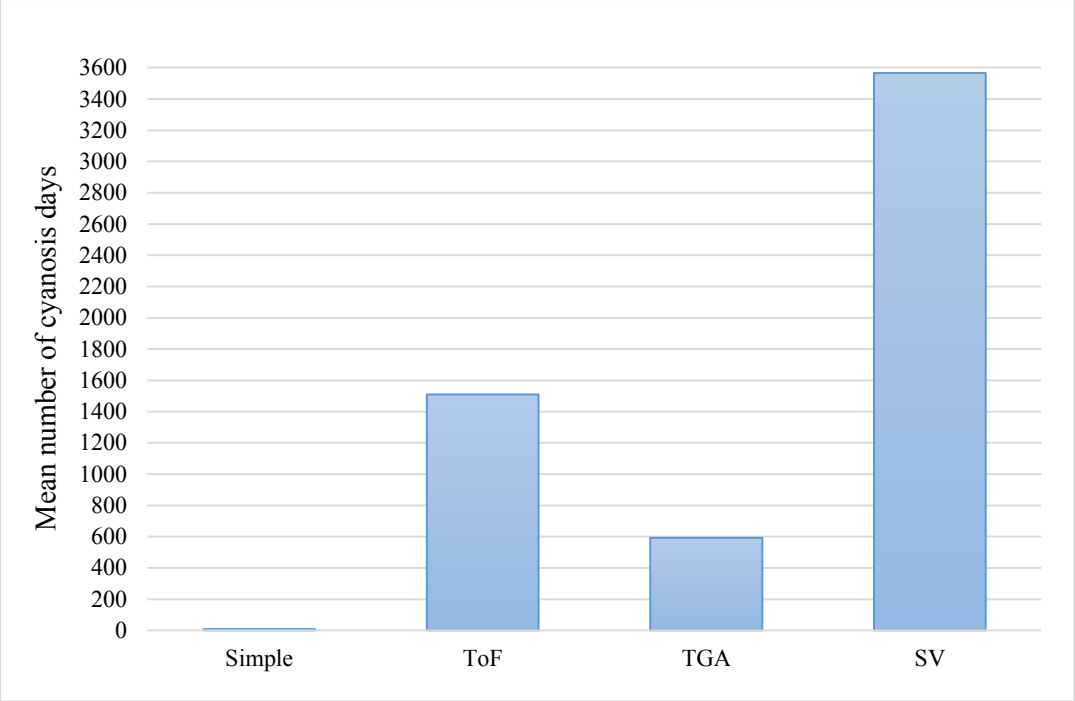


Figure 6.2. Mean number of cyanosis days by diagnostic group

6.6.2. Intervention history

There were differences in the number of interventions that patients had undergone ($p < .001$) between diagnostic groups (Figure 6.3). The SV group had undergone more interventions than the ToF (mean difference= 0.78, $p = .009$, 99% CI [0.01, 1.55]) and Simple (mean difference= 1.72, $p < .001$, 99% CI [1.02, 2.43]). In addition, the ToF and TGA groups had undergone more interventions than the Simple group (mean difference= 0.94, $p < .001$, 99% CI [0.43, 1.45] & mean difference= 1.18, $p < .001$, 99% CI [0.65, 1.71] respectively).

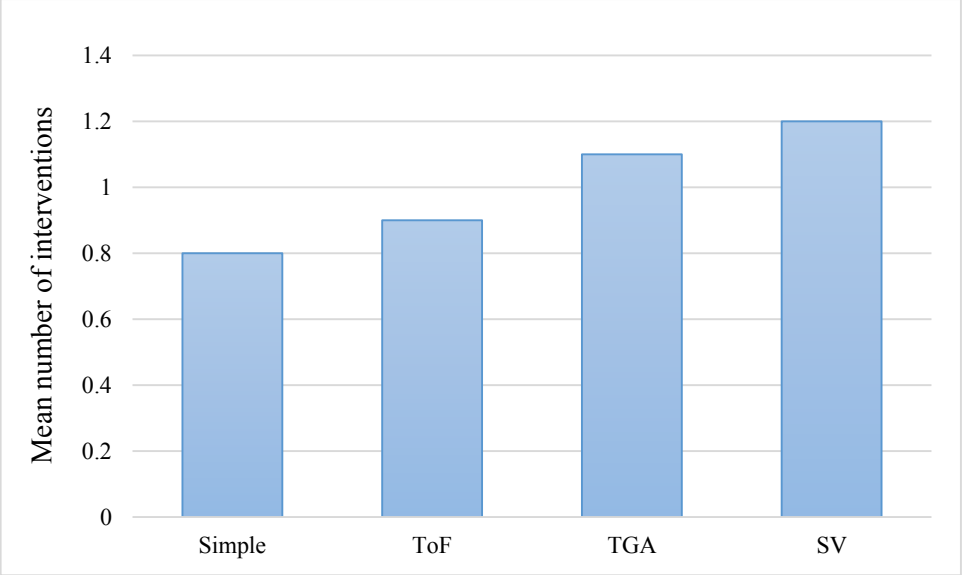


Figure 6.3. Mean number of interventions by diagnostic group

There were also differences in hospitalization days ($p < .001$) between diagnostic groups (Figure 6.4). The ToF, TGA, and SV groups each had more hospitalization days than the Simple group (mean difference= 29.50, $p < .001$, 99% CI [16.30, 42.69], mean difference= 18.40, $p < .001$, 99% CI [7.93, 28.86] & mean difference= 53.11, $p < .001$, 99% CI [15.49, 90.73] respectively).

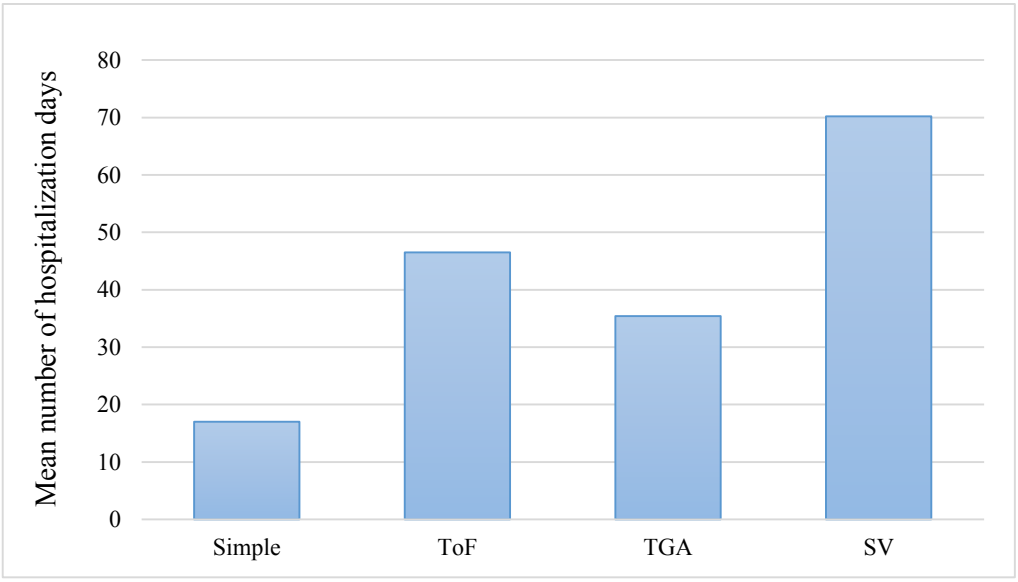


Figure 6.4. Mean number of hospitalization days by diagnostic group

6.6.3. Current status

There were differences in the number of medications people were taking ($p < .001$) between diagnostic groups (Figure 6.5). The SV group was taking more medications than the ToF (mean difference= 0.98, $p < .001$, 99% CI [0.33, 1.64]) and Simple (mean difference= 0.69, $p = .005$, 99% CI [0.04, 1.34]).

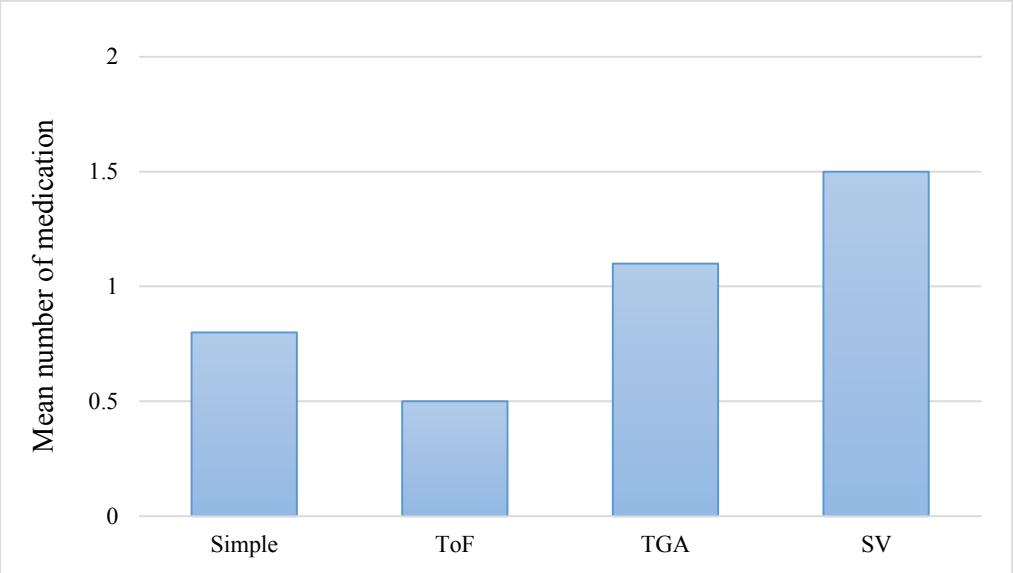


Figure 6.5. Mean number of medication by diagnostic group

There were differences in current O_2 saturation ($p < .001$) between diagnostic groups (Figure 6.6). The Simple group had significantly higher saturation levels than the ToF (mean difference= 0.92, $p = .002$, 99% CI [0.14, 1.70]), TGA (mean difference= 1.93, $p < .001$, 99% CI [0.93, 2.93]), and SV (mean difference= 7.06, $p < .001$, 99% CI [4.05, 10.08]). The ToF group had higher saturation levels than the TGA (mean difference= 1.01, $p = .006$, 99% CI [0.06, 1.97]) and the SV (mean difference= 6.14, $p < .001$, 99% CI [3.14, 9.14]). The TGA group had higher saturation levels than the SV (mean difference= 5.13, $p < .001$, 99% CI [2.07, 8.19]).

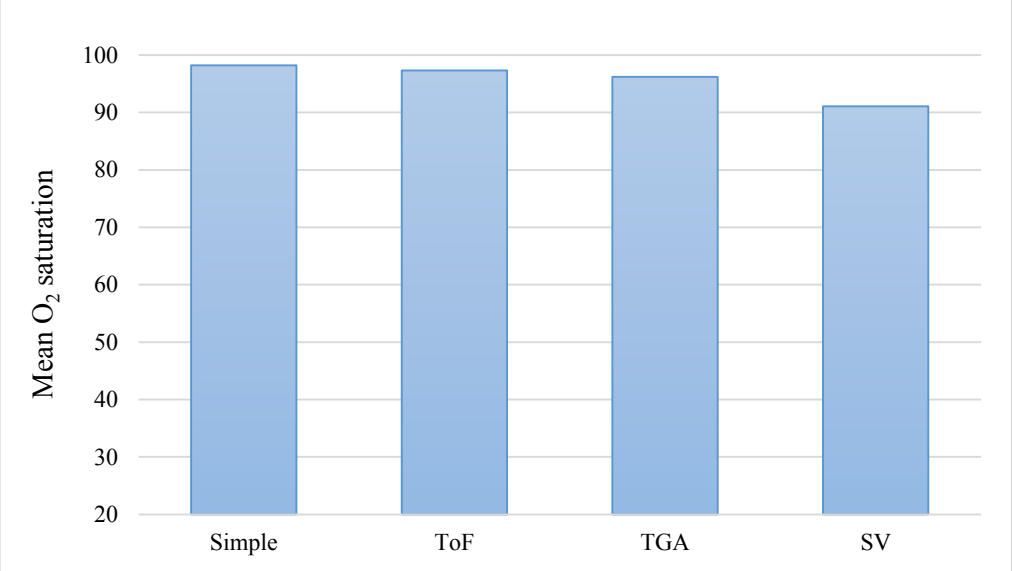


Figure 6.6. Mean O_2 saturation by diagnostic group

NYHA functional classes II, III, and IV were merged due to the small number of individuals in these groups. The majority were classified at NYHA functional class I, while 39% were at II, III, and IV. There was no association between functional status as measured by the NYHA classification and diagnostic group ($p > .01$). No significant effect of diagnostic group on the degree of exercise capacity as measured by VO_2 max was observed ($p > .01$).

There were differences in both right and left ventricular function ($p < .001$) between diagnostic groups (Figure 6.7 and Figure 6.8). The Simple group had better right ventricular function than the ToF (mean difference= 7.59, $p < .001$, 99% CI [3.98, 11.20]), TGA (mean difference= 9.11, $p < .001$, 99% CI [5.89, 12.33]), and SV (mean difference= 7.78, $p < .001$, 99% CI [3.69, 11.87]). In addition, the Simple group had better left ventricular function than the ToF (mean difference= 4.86, $p = .001$, 99% CI [0.96, 8.76]) and SV (mean difference= 9.78, $p < .001$, 99% CI [5.70, 13.86]) groups. The ToF and TGA groups had better left ventricular function than

the SV group (mean difference= 4.92, p= .001, 99% CI [0.81, 9.04] & mean difference= 7.87, p< .001, 99% CI [3.76, 11.99] respectively).

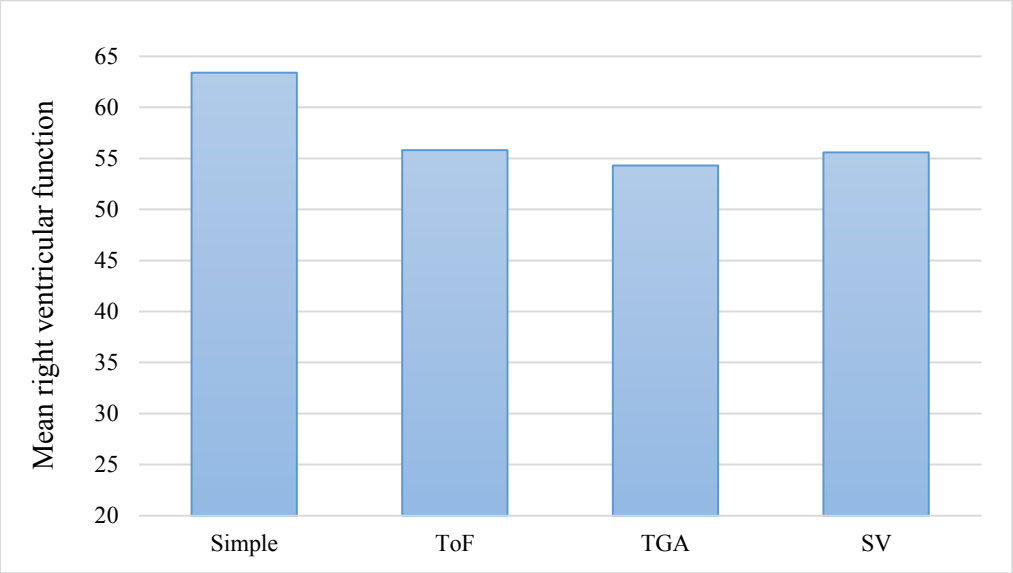


Figure 6.7. Mean right ventricular function by diagnostic group

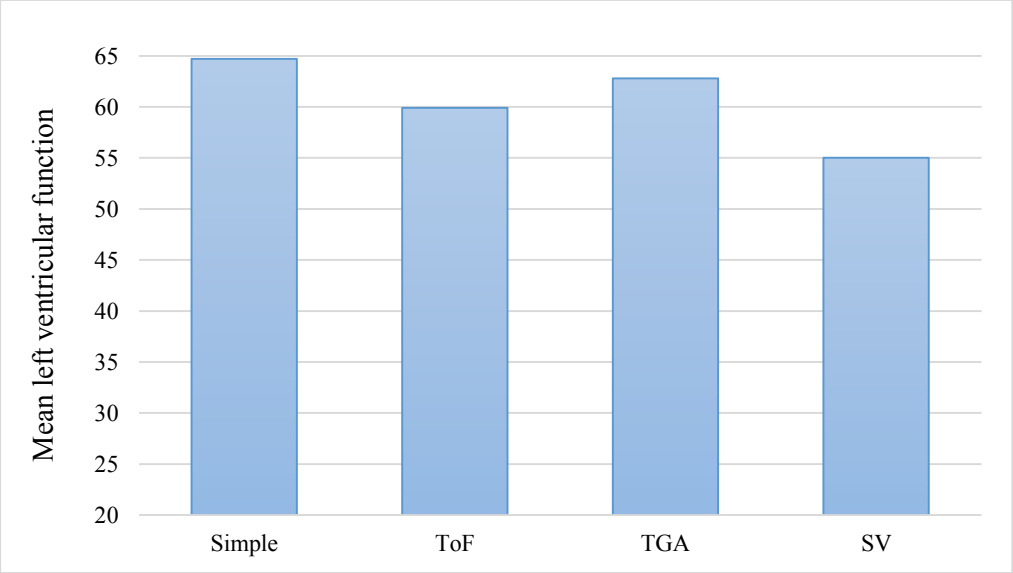


Figure 6.8. Mean left ventricular function by diagnostic group

6.7. Psychosocial predictor variables

6.7.1. Illness perceptions

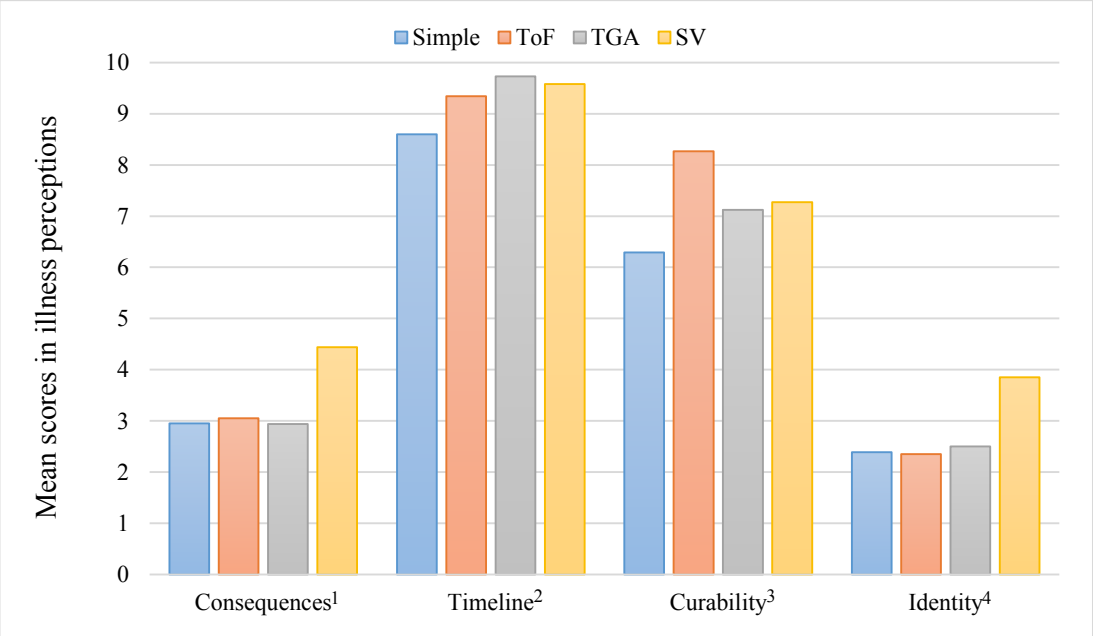
The mean scores and results from group comparisons on illness perceptions are reported in Table 6.5.

Table 6.5. Mean scores and group comparisons on illness perceptions

Illness Perceptions		*Mean (S.D.)*	*Model Parameters*			
			f	*df*	*Sig.*	η^2
Consequences			4.39§	162.59	**.005**	.05
	Simple	2.95 (2.78)				
	ToF	3.05 (2.50)				
	TGA	2.94 (2.37)				
	SV	4.44 (2.98)				
	Total	3.30 (2.71)				
Timeline			4.17§	154.63	**.007**	.05
	Simple	8.60 (2.82)				
	ToF	9.34 (2.04)				
	TGA	9.73 (1.03)				
	SV	9.58 (1.64)				
	Total	9.29 (2.06)				
Controllability			0.80§	163.13	.496	.01
	Simple	4.99 (3.34)				
	ToF	5.62 (2.70)				
	TGA	5.46 (2.58)				
	SV	5.08 (3.11)				
	Total	5.30 (2.94)				
Curability			7.02§	164.03	**<.001**	.06
	Simple	6.29 (3.43)				
	ToF	8.27 (2.23)				
	TGA	7.12 (2.79)				
	SV	7.27 (2.53)				
	Total	7.22 (2.88)				
Identity			6.89	299	**<.001**	.06
	Simple	2.39 (2.42)				
	ToF	2.35 (2.23)				
	TGA	2.50 (2.01)				
	SV	3.85 (2.45)				
	Total	2.73 (2.34)				
Concern			0.94	299	.423	.01
	Simple	3.96 (2.91)				
	ToF	4.36 (2.81)				
	TGA	3.83 (2.66)				
	SV	4.50 (2.88)				
	Total	4.15 (2.82)				
Coherence			1.62	299	.186	.02
	Simple	6.59 (2.65)				
	ToF	6.21 (2.32)				
	TGA	6.31 (2.40)				
	SV	5.68 (2.70)				
	Total	6.22 (2.52)				
Emotional representation			2.69§	162.76	.048	.03
	Simple	3.13 (3.22)				
	ToF	3.55 (3.01)				
	TGA	3.09 (2.69)				
	SV	4.45 (3.37)				
	Total	3.51 (3.10)				

§Welch ANOVA. η^2: .01= small, .06= medium, .14= large.
Scale 0 – 10, higher score = stronger perceptions.

ANOVA indicated a significant effect of diagnostic group on illness consequences ($p < .01$), timeline ($p < .01$), curability ($p < .001$), and identity ($p < .001$). Post-hoc analysis indicated that the SV group reported stronger perception of negative consequences as a result of their ACHD than those with TGA (mean difference= 1.50, $p = .007$, 99% CI [0.06, 2.95]) (Figure 6.9). The TGA group reported stronger beliefs about illness chronicity than the Simple group (mean difference= 1.13, $p = .005$, 99% CI [0.07, 2.19]). The ToF group reported a stronger belief about treatment effectiveness than the Simple (mean difference= 1.98, $p < .001$, 99% CI [0.57, 3.39]). The SV group attributed more severe symptoms to their condition than all the other groups (Simple: mean difference= 1.46, $p = .001$, 99% CI [0.27, 2.65], ToF: mean difference= 1.50, $p = .001$, 99% CI [0.29, 2.71], TGA: mean difference=1.35, $p = .003$, 99% CI [0.14, 2.56]).



All scales 0 – 10, higher scores = [1] severe illness consequences, [2] illness is chronic, [3] treatment extremely helpful, [4]many severe symptoms

Figure 6.9. Mean scores in illness perceptions by diagnostic group

6.7.2. Coping strategies

Table 6.6 presents the mean scores and results from group comparisons on coping strategies.

Table 6.6. Mean scores and group comparisons on coping strategies

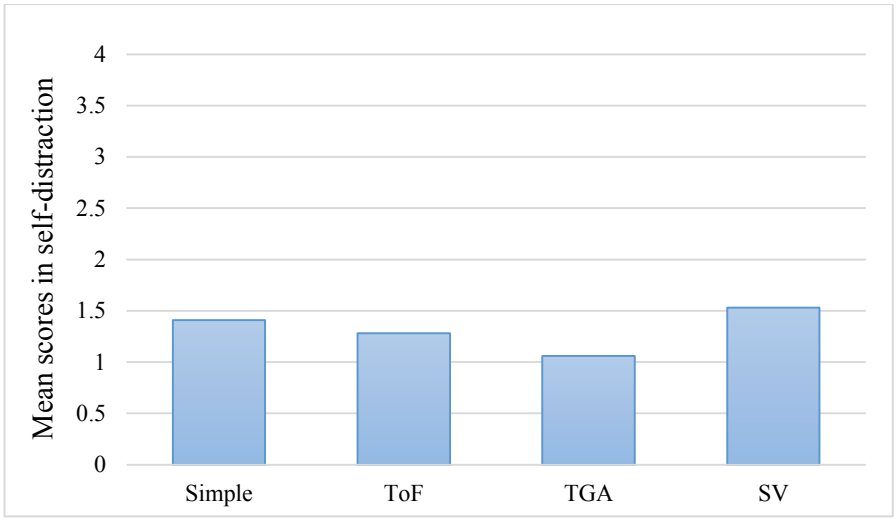
Coping Strategies		*Mean (S.D.)*	*Model Parameters*			
			f	*df*	*Sig.*	η^2
Planning			3.55	299	.015	.03
	Simple	1.45 (0.92)				
	ToF	1.47 (0.80)				
	TGA	1.13 (0.87)				
	SV	1.58 (0.90)				
	Total	1.40 (0.88)				
Active coping			2.03	299	.109	.02
	Simple	1.63 (0.87)				
	ToF	1.80 (0.80)				
	TGA	1.47 (0.83)				
	SV	1.70 (0.85)				
	Total	1.65 (0.84)				
Positive reframing			0.59	299	.625	.01
	Simple	1.58 (0.98)				
	ToF	1.51 (0.91)				
	TGA	1.42 (0.92)				
	SV	1.40 (0.90)				
	Total	1.48 (0.93)				
Acceptance			2.38	299	.070	.02
	Simple	2.24 (0.76)				
	ToF	2.32 (0.73)				
	TGA	2.13 (0.89)				
	SV	2.46 (0.70)				
	Total	2.28 (0.78)				
Humour			1.38	299	.251	.01
	Simple	1.21 (1.05)				
	ToF	1.27 (1.16)				
	TGA	1.13 (1.13)				
	SV	1.49 (1.04)				
	Total	1.27 (1.10)				
Religion			1.97*	160.08	.121	.01
	Simple	0.52 (0.83)				
	ToF	0.60 (1.01)				
	TGA	0.33 (0.64)				
	SV	0.58 (0.95)				
	Total	0.51 (0.87)				
Emotional support			1.52	299	.211	.01
	Simple	1.18 (0.85)				
	ToF	1.35 (0.93)				
	TGA	1.06 (0.93)				
	SV	1.30 (1.00)				
	Total	1.22 (0.92)				

Table 6.6 ***Continued***

Coping Strategies		*Mean (S.D.)*	*Model Parameters*			
			f	***df***	***Sig.***	**η^2**
Instrumental support			0.94	299	.420	.01
	Simple	1.00 (0.87)				
	ToF	1.24 (0.91)				
	TGA	1.11 (0.90)				
	SV	1.13 (0.94)				
	Total	1.11 (0.90)				
Self-distraction			4.20	299	**.006**	.04
	Simple	1.41 (0.91)				
	ToF	1.28 (0.85)				
	TGA	1.06 (0.89)				
	SV	1.53 (0.72)				
	Total	1.31 (0.86)				
Denial			0.14	299	.935	.00
	Simple	0.19 (0.42)				
	ToF	0.17 (0.40)				
	TGA	0.17 (0.47)				
	SV	0.21 (0.46)				
	Total	0.18 (0.44)				
Venting			2.73	299	.044	.03
	Simple	0.74 (0.65)				
	ToF	0.75 (0.81)				
	TGA	0.59 (0.71)				
	SV	0.94 (0.75)				
	Total	0.75 (0.74)				
Substance use			1.89*	160.31	.133	.02
	Simple	0.29 (0.60)				
	ToF	0.18 (0.40)				
	TGA	0.13 (0.39)				
	SV	0.29 (0.59)				
	Total	0.22 (0.51)				
Behavioural disengagement			1.22*	160.41	.304	.01
	Simple	0.26 (0.55)				
	ToF	0.16 (0.44)				
	TGA	0.19 (0.38)				
	SV	0.31 (0.59)				
	Total	0.23 (0.50)				
Self-blame			0.79	299	.503	.01
	Simple	0.55 (0.66)				
	ToF	0.40 (0.63)				
	TGA	0.47 (0.65)				
	SV	0.52 (0.73)				
	Total	0.49 (0.66)				

* Welch ANOVA. η^2: .01= small, .06= medium, .14= large.
Scale 0 – 3, higher score = greater use of coping strategy.

ANOVAs showed no differences in the use of coping strategies between the diagnostic groups ($p > .01$) except for self-distraction ($p < .01$) (Figure 6.10). The SV group used more self-distraction than the TGA group (mean difference= 0.47, $p = .006$, 99% [0.02, 0.92]).



Scale 0 – 4, higher score = greater use of self-distraction

Figure 6.10. Mean scores in self-distraction by diagnostic group

6.7.3. Social support

Table 6.7 presents the mean scores and group comparisons on social support. ANOVAs did not indicate statistically significant differences between the diagnostic groups ($p > .01$).

Table 6.7. Mean scores and group comparisons on social support

Social Support		*Mean (S.D.)*	*Model Parameters*			
			f	*df*	*Sig.*	η^2
Family support			1.68*	163.12	.174	.01
	Simple	4.17 (0.93)				
	ToF	4.35 (0.69)				
	TGA	4.19 (0.71)				
	SV	4.07 (0.83)				
	Total	4.20 (0.80)				
Friends support			1.02	299	.382	.01
	Simple	4.12 (0.88)				
	ToF	4.01 (0.83)				
	TGA	4.01 (0.81)				
	SV	3.86 (1.02)				
	Total	4.01 (0.88)				
Significant other support			2.63*	161.29	.052	.03
	Simple	4.32 (0.87)				
	ToF	4.43 (0.83)				
	TGA	4.17 (1.10)				
	SV	3.98 (1.11)				
	Total	4.24 (0.99)				

* Welch ANOVA. η^2: .01= small, .06= medium, .14= large.
Scale 1 – 5, higher score = greater support

6.7.4. Mood

Mean scores for anxiety are displayed in Table 6.8 and the percentage of the sample with depressive symptoms is displayed in Table 6.9. ANOVA and chi-square analysis did not suggest statistically significant differences between the diagnostic groups in anxiety levels and depressive symptoms (p> .01).

Table 6.8. Mean scores and group comparisons on anxiety levels

	Mean (S.D.)	*Model Parameters*			
		f	*df*	*Sig.*	η^2
Anxiety		1.62	299	.163	.02
Simple	1.68 (0.61)				
ToF	1.62 (0.56)				
TGA	1.60 (0.51)				
SV	1.80 (0.60)				
Total	1.67 (0.57)				

η^2: .01= small, .06= medium, .14= large.
Scale score 1 – 4, higher score = higher anxiety levels.

Table 6.9. Percentages of people with depressive symptoms and group comparisons

Depression n (%)	*Total*	*Simple*	*ToF*	*TGA*	*SV*	*Model Parameters*			
						χ^2	*df*	*Sig.*	φ_c
No depressive symptoms	238 (78.5)	59 (72)	63 (81.8)	68 (87.2)	48 (72.7)	7.38	3	.061	.16
With depressive symptoms	65 (21.5)	23 (28)	14 (18.2)	10 (12.8)	18 (27.3)				

φ_c: .10= small, .30= medium, .50= large.

6.8. Discussion

The following discussion focuses on comparisons between the current sample and other studies in ACHD.

6.8.1. Participation rates

Due to lack of reports regarding participation rates in previous studies, comparisons can only be made to one study. The participation rate in the current study was 44% compared with 59% in a previous study (Lane, Lip, & Millane, 2002). The lower participation rate might have been due to practical reasons. The administration of questionnaires was conducted in person and participants were asked to arrive at the hospital prior to their outpatient appointment, whereas in the study by Lane et al. (2002) questionnaires were sent by post.

The TGA group was more likely and the Simple group less likely to respond and complete the study. It is possible that individuals in the Simple group did not perceive themselves as having problems with their HRQoL and therefore had no interest in participating in the study. Because invited participants were not required to give a reason for not participating, this assumption and the reason for the participation rates for the Simple and TGA groups could not be explored further.

6.8.2. Demographic characteristics

This study was similar to other ACHD studies regarding the demographic characteristics of the sample. The higher percentage of male (56.4%) compared to female participants was also evident in the systematic review of 31 studies (average: 56.3% male) (Fteropoulli, Stygall, Cullen, Deanfield, & Newman, 2013). Prevalence rates of ACHD are typically higher for females than in males (Marelli, Mackie, Ionescu-Ittu, Rahme, & Pilote, 2007). Simpler diagnoses, including ASD and PS, are generally more common in females than males, while ToF and TGA are more common in males (Mulder, 2012; Perloff, 1991). The larger (combined) proportion of people in the ToF and TGA groups over people in the Simple group is likely to be the explanation for the higher percentage of males in the present study.

The proportion of people who were married or in a relationship was 50.6%. This is at the higher end but is consistent with previous studies, where the proportion of married participants ranged between 30% and 55% (Chen et al., 2011; Lane et al., 2002; Moons et al., 2006; Rose et al., 2005; Simko & McGinnis, 2003).

Previous reports regarding the percentage of people with ACHD completing university level education varied between 25-70% (Kamphuis et al., 2002c; Simko & McGinnis, 2003; Zomer et al., 2012b). The percentage in the present study was 33.1%, therefore within the reported range. This percentage was only slightly lower than the percentage of people who have a graduate degree (38%) in the UK in 2013 (Office for National Statistics, 2013). The proportion of employed participants in the study (70.4%) was within the range of 35-79% reported in the literature (Chen et al., 2011; Jefferies et al., 2004; Lane et al., 2002). This percentage was slightly lower than the employment rate (73%) in the UK in 2014 (Office for National Statistics,

2014). These findings potentially have implications regarding the provision of career advice to people with ACHD (Crossland, Jackson, Lyall, Burn, & O'Sullivan, 2005).

6.8.3. Clinical characteristics

As anticipated, there were differences between the four diagnostic groups. There was increased morbidity in the SV group in the form of chronic cyanosis, number of interventions, hospitalization days, and medication, and ventricular function compared with the ToF, TGA, and Simple groups. It was difficult to compare the current sample with previous studies in clinical characteristics due to the variability in diagnostic groups and the limited reports of participants' clinical characteristics in previous studies.

Comparisons can be made regarding the exclusion criteria. The current study excluded individuals with chromosomal conditions and/or severe learning difficulties or mental retardation, and history of stroke because the self-report measures required normal intellectual abilities and people who were able to complete the questionnaires unassisted. Similar exclusion criteria have been used in previous studies (Callus, et al., 2014; Chen et al., 2011; Irtel et al., 2005; Kamphuis et al., 2002b; Loup et al., 2009; Moons, et al., 2006; Moons et al., 2009; Saliba et al., 2001; Simko & McGinnis, 2005).

6.8.4. Illness perceptions

Only two studies have examined illness perceptions in ACHD, both of which have used the revised version of the IPQ to measure illness perceptions as opposed to the brief IPQ used in the present study. Therefore, the differences between the two measures limited direct comparisons between this and previous studies.

Overall, the low mean scores in the consequences, emotional representation, and concern subscales suggest that people believe that their condition has a relatively small impact on their lives. Similar findings have been reported by Schoormans et al. (2014). The findings of the present study also indicate that people do not believe that their condition causes many severe symptoms (identity subscale). Data from previous studies were not directly comparable to the present study. Riley et al. (2012) reported a mean of four symptoms experienced by people with ACHD in their study, the most common being palpitations, breathlessness, and fatigue. Although the findings of the present study may reflect the absence of severe symptoms in the sample, it is also possible that the treatment regimen helps ameliorate some of these symptoms. In fact, mean scores in the curability subscale were high, suggesting that people believe that they benefit from their treatment regimen. Mean scores in the controllability subscale were in the scale average, indicating neither strong nor weak beliefs regarding personal control. These findings are consistent with the findings reported by Riley et al. (2012) and Schoormans et al. (2014).

High mean scores in the timeline subscale suggest that people acknowledge the chronicity of their condition. Similar findings were reported in the study by Riley et al. (2012). This finding is not surprising considering the congenital nature of ACHD. The mean scores in the illness coherence subscale suggest that individuals in the present study have average understanding of their condition. Schoormans et al. (2014) reported higher levels, whereas Riley et al. (2012) reported similar levels of illness coherence compared with the present study. A possible explanation for the higher coherence level in the study by Schoormans et al. is their inclusion of a large

proportion (60%) of people with simple defects, which may be easier to understand as opposed to more complex defects.

The comparisons between the four diagnostic groups on illness perceptions indicated that the SV group perceived their condition and its impact more negatively than the other groups, as also reported by Schoormans et al. (2014). Negative illness perceptions in the SV group may be valid reflections of their poorer physical status. However, some negative perceptions in the Simple group warrant attention. These individuals reported weaker perceptions of illness chronicity than those with TGA and weaker perceptions of curability than those with ToF. One possible explanation for why people in the Simple group do not recognise the chronicity of their condition to the same degree as people with TGA is the nature of their condition, which is less complex and has fewer consequences. In addition, people in the Simple group are more likely to be medically managed as opposed to those with ToF who usually have reparative surgery early in their lives; hence, their weaker perceptions of treatment benefit.

6.8.5. Coping strategies

Participants in the present study engaged moderately in certain coping strategies, including acceptance and active coping, and to a lesser degree in others, including positive reframing, planning, self-distraction, humour, emotional support, instrumental support, denial, venting, and religion. In addition, mean scores in behavioural disengagement, substance use, and self-blame suggest minimal engagement in these coping strategies.

Similar findings have been reported in previous studies in ACHD that used the Utrecht Coping List and reported that the most used coping strategy is active

problem solving, while the least used is expression of emotions (Eslami et al., 2013; van Rijen et al., 2004). The findings of the present study are consistent with previous qualitative reports of normalization coping in people with ACHD (Berghammer et al., 2006; Claessens et al., 2005). Normalization includes a process of accepting and adjusting to one's limitations. This appears to be the case for participants in the present study who accept their condition and take active steps to cope with it.

It is important to note that participants engaged less in what is generally considered maladaptive coping, including self-distraction, denial, venting of negative emotions, behavioural disengagement, substance use, and self-blame. Such coping strategies have been commonly reported in qualitative studies of adolescents with CHD. Zahmacioglu et al. (2011), using qualitative interviews, found that denial is used by adolescents to cope with stressful interventions but is shortly followed by more active approaches. Similar findings have been reported by McMurray et al. (2001). It appears that as people get older they accept their condition and begin to adjust their lives based on their limitations (Chiang et al., 2011).

Comparisons between the four diagnostic groups did not reveal differences in the use of coping strategies with the exception of self-distraction. The findings indicated that the SV group uses self-distraction more than the TGA group. Previous studies showed that the adoption of coping strategies was independent of disease complexity (Enomoto et al., 2013; Eslami et al., 2014; van Rijen et al., 2004). It is likely that people with SV use more self-distraction than those with TGA because they experience greater emotional burden from their condition. Self-distraction may serve as an adaptive rather than maladaptive coping strategy, because it helps people with

SV to ameliorate stress associated with their condition (Tuncay, Musabak, Gok, & Kutlu, 2008).

6.8.6. Social support

The mean scores in perceived social support ranged between "agree" and "strongly agree", suggesting that participants perceive high levels of support from family, friends, and their significant others. Similar high levels of social support have been reported in a study by Pike et al. (2012) who used the same measure (MSPSS).

There were no differences between the four diagnostic groups in levels of perceived social support. No previous studies have examined levels of perceived social support across different diagnostic groups. However, high levels of social support have been reported in studies that included people with SV (Pike et al., 2012), ToF (Knowles et al., 2012), and studies with mixed samples (Rose et al., 2005).

6.8.7. Mood

Responses in anxiety ranged from "not at all" to "somewhat", suggesting that participants in this study experience low levels of anxiety. Similar levels of anxiety have been reported by Müller et al. (2013a) who also used the STAI. Previous studies that applied cut-off points to categorise people on the degree of anxiety and indicated that up to 63% of people with ACHD may experience elevated anxiety levels (Cohen et al., 2010; Eslami et al., 2013; Riley et al., 2012).

A total of 21.5% of the total sample exhibited depressive symptoms. Variable prevalence rates up to 41% have been reported in previous studies using self-report measures (Bang et al., 2013; Cohen et al., 2010; Eslami et al., 2013; Kourkoveli, et

al., 2014; Kovacs, et al., 2009b; Bromberg et al., 2003; Müller et al., 2012; Pike et al., 2012; Popelová et al., 2001; Riley et al., 2012).

No differences were observed between the four diagnostic groups in anxiety levels and the presence of depressive symptoms. Previous studies in ACHD failed to find any association between disease complexity and mood (Eslami et al., 2013; Kovacs et al., 2009b; Loup et al., 2009; Müller et al., 2012, Müller et al., 2013a; Popelová et al., 2001).

6.9. Summary

This chapter described the demographic, clinical, and psychosocial characteristics of the study sample. Overall, the findings suggest that the present sample of people with ACHD is comparable to previous studies in the literature. The next chapter will address the first aim of the study by describing the HRQoL of people with ACHD and differences between the four diagnostic groups.

CHAPTER 7 – IMPACT OF ADULT CONGENITAL HEART DISEASE ON HEALTH-RELATED QUALITY OF LIFE

7.1. Prologue

The previous chapter presented a description of the sample characteristics. The current chapter describes the analyses from the cross-sectional study to examine the impact of ACHD on HRQoL by comparing the HRQoL of people with ACHD with normative data from the general population and to examine differences in HRQoL between four diagnostic groups. It was hypothesized that people with ACHD will report significantly poorer HRQoL compared with the general population norm, predominantly in physical domains of HRQoL and that there will be differences in HRQoL between the diagnostic groups, predominantly between the Simple and SV groups (section 4.4, page 127). The following sections present the results of the *t*-test and ANOVA analyses and discussion of the findings.

7.2. HRQoL comparisons between people with ACHD and general population norms

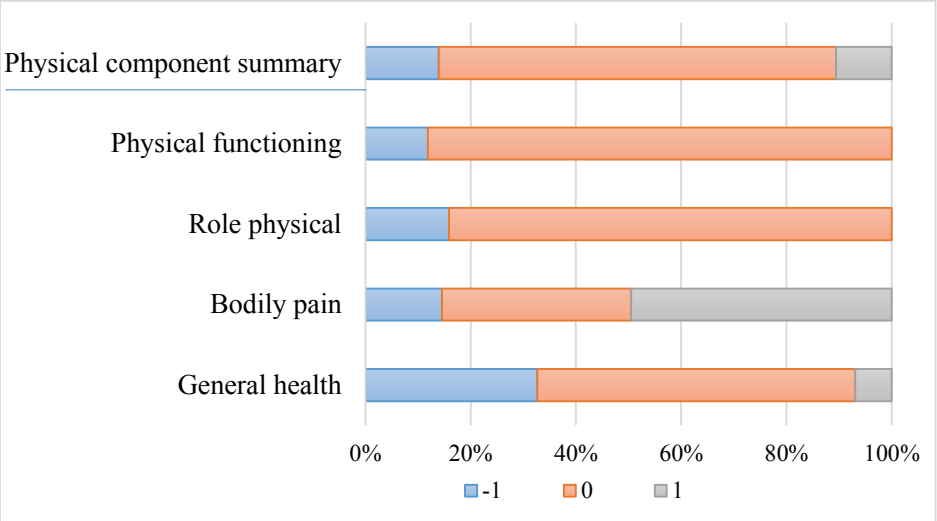
Comparisons between people with ACHD and normative data were conducted for generic HRQoL (SF-36v1®).

7.2.1. Physical HRQoL

The distribution of participants' scores in the physical component summary and its subscales are presented in the form of percentage of people that scored within the norm mean (0), 1 *S.D.* below (< -1), and 1 *S.D.* above (> 1) the norm mean.

In the total sample, the majority (range: 60.4- 88.1%) scored within the average range in the physical component summary and all of the subscales, followed by those

who scored below the norm mean (range: 11.9- 32.7%) (Figure 7.1). A smaller number (range: 0- 10.6%) scored above the norm mean. This excludes the bodily pain subscale, in which more people scored above the norm mean (49.5%) than within the average (36%) or below the nom mean (14.5%). None scored above the norm mean in the physical functioning and role physical subscales.

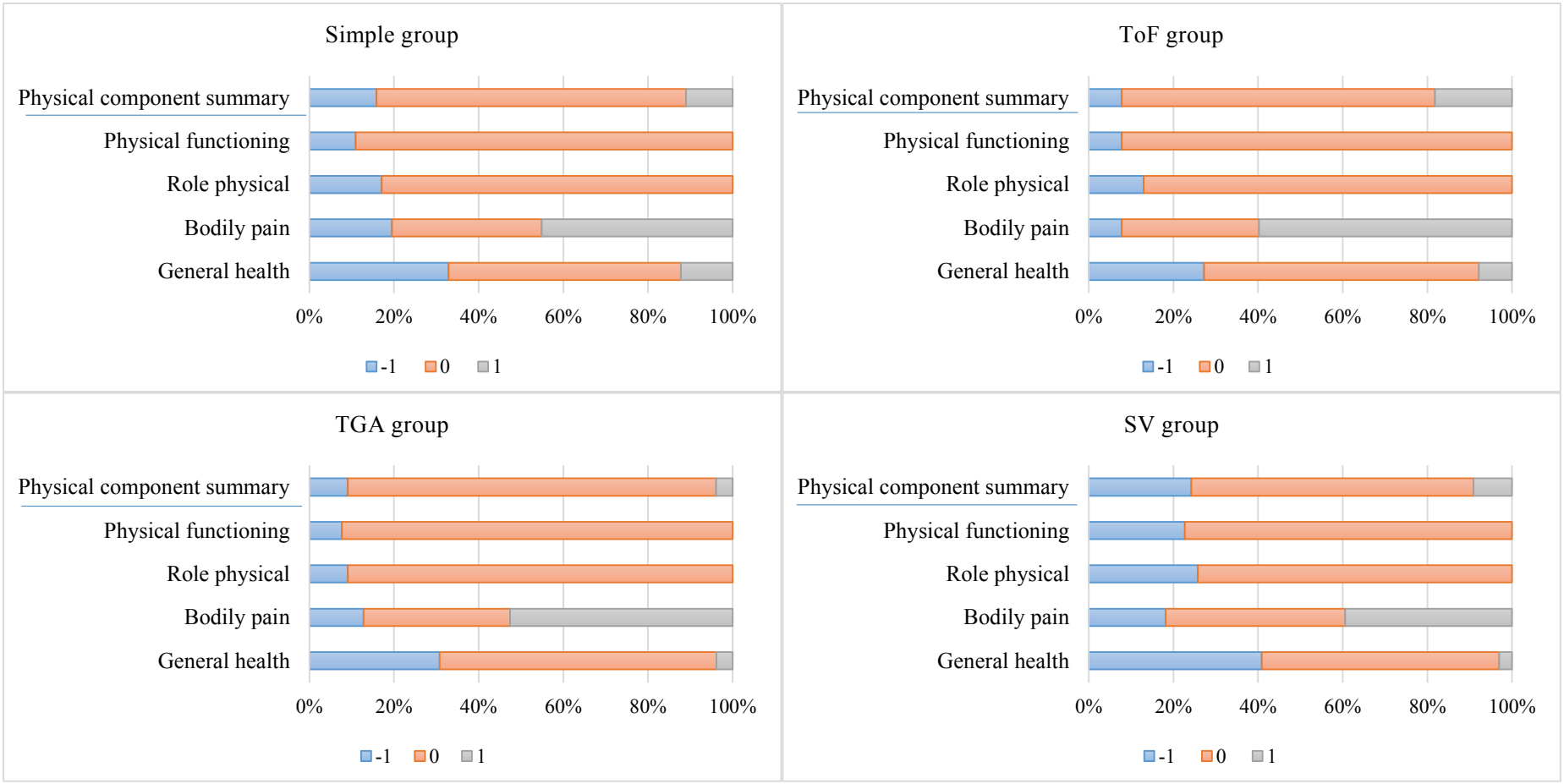


Note. -1= 1 *S.D.* or more below the norm mean, 0= within the norm mean, 1= 1 *S.D.* or more above the norm mean

Figure 7.1. Distribution of physical HRQoL scores for total sample

The same pattern of results was observed within each of the diagnostic groups, with two exceptions (Figure 7.2). Firstly, in the ToF group, more people scored above (18.2%) than below the norm mean (7.8%) in the physical component summary. Secondly, in the SV group, slightly more people scored within the average (42.4%) than above the norm mean (39.4%) in the bodily pain subscale.

More people scored below the norm mean in general health than any other physical HRQoL subscale. On the other hand, more people scored above the norm mean in bodily pain than any other subscale. This pattern was evident in the total sample and across all diagnostic groups.



Note. -1= 1 *S.D.* or more below the norm mean, 0= within the norm mean, 1= 1 *S.D.* or more above the norm mean

Figure 7.2. Distribution of physical HRQoL scores for each diagnostic group

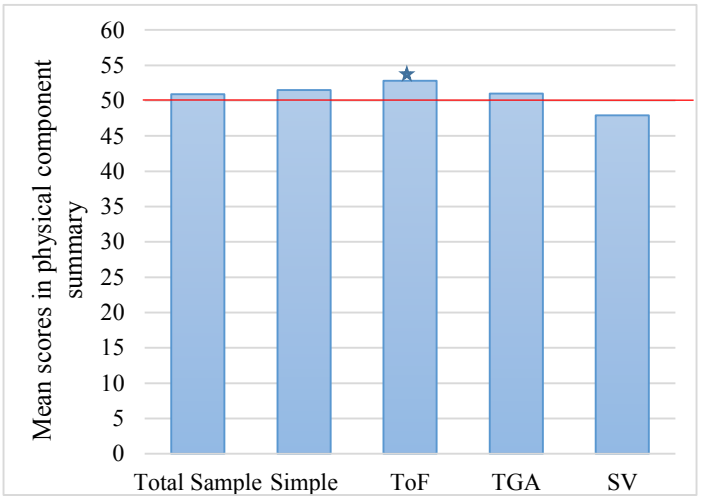
The standardised mean scores of the total sample and diagnostic groups for physical HRQoL compared with the norm mean and results from *t*-tests are presented in Table 7.1.

Table 7.1. *T*-tests: physical HRQoL comparisons with norms

Scale/Subscale		*Mean (S.D.)*	*T-test*	*Sig.*	*99% CI of the Difference*	*d*
Physical component summary						
	Total	50.9 (9.4)	*t*(301)= 1.74	.083	-0.46, 2.34	.09
	Simple	51.5 (10)	*t*(81)= 1.38	.170	-1.38, 4.42	.15
	ToF	52.8 (8.7)	*t*(76)= 2.84	**.006**	0.20, 5.44	.30
	TGA	51 (7.5)	*t*(76)= 1.18	.241	-1.25, 3.27	.11
	SV	47.9 (10.8)	*t*(65)= -1.56	.123	-5.59, 1.45	.20
Physical functioning						
	Total	49.8 (8.9)	*t*(302)= -0.35	.728	-1.50, 1.15	.02
	Simple	50.9 (9.2)	*t*(81)= 0.88	.381	-1.79, 3.57	.09
	ToF	51.4 (7.2)	*t*(76)= 1.70	.094	-0.78, 3.57	.16
	TGA	50.5 (7.5)	*t*(77)= 0.62	.538	-1.73, 2.78	.06
	SV	45.8 (10.6)	*t*(65)= -3.20	**.002**	-7.65, -0.71	.41
Role physical						
	Total	50.5 (9.7)	*t*(301)= 0.94	.346	-0.92, 1.97	.05
	Simple	50.4 (9.9)	*t*(81)= 0.34	.733	-2.52, 3.27	.04
	ToF	52.2 (8.5)	*t*(76)= 2.28	.025	-0.35, 4.75	.24
	TGA	51.3 (8.4)	*t*(76)= 1.33	.187	-1.26, 3.82	.14
	SV	47.9 (11.6)	*t*(65)= -1.48	.144	-5.92, 1.68	.20
Bodily pain						
	Total	54.5 (10.4)	*t*(302)= 7.46	**<.001**	2.91, 6.00	.44
	Simple	52.7 (11.8)	*t*(81)= 2.07	.042	-0.74, 6.16	.25
	ToF	56.7 (8.7)	*t*(76)= 6.79	**<.001**	4.12, 9.37	.72
	TGA	55.4 (10)	*t*(77)= 4.80	**<.001**	2.44, 8.39	.54
	SV	52.8 (10.3)	*t*(65)= 2.22	.030	-0.55, 6.20	.28
General health						
	Total	45.1 (10.6)	*t*(302)= -8.08	**<.001**	-6.47, -3.33	.48
	Simple	46 (11.5)	*t*(81)= -3.15	**.002**	-7.33, -0.65	.37
	ToF	46.8 (9.8)	*t*(76)= -2.83	**.006**	-6.14, -0.21	.33
	TGA	44.7 (9)	*t*(77)= -5.24	**<.001**	-7.99, -2.64	.56
	SV	42.4 (11.5)	*t*(65)= -5.32	**<.001**	-11.32, -3.78	.71

Note. Norm mean 50 (10). Positive and negative t values indicate better and poorer HRQoL respectively. Cohen's *d*: .20= small, .50= medium, .80= large.

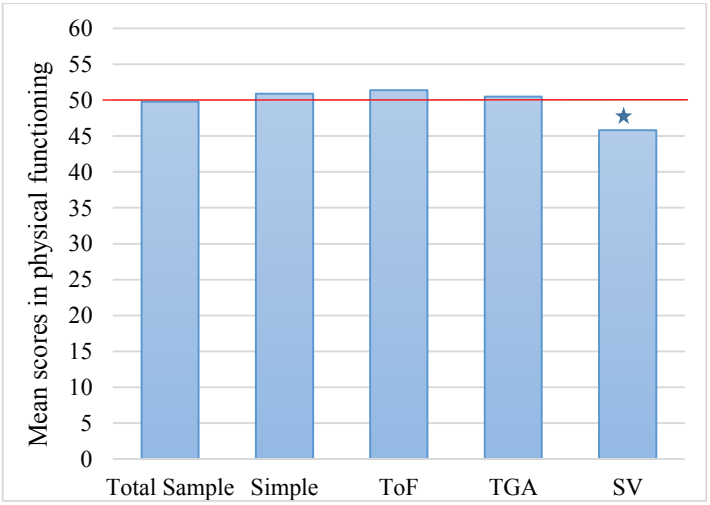
In the physical component summary, people with ToF reported significantly better HRQoL than the norm ($p < .01$) (Figure 7.3).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.3. Comparisons with norm in physical component summary

In the physical functioning subscale, the SV group reported poorer HRQoL than the norm ($p < .01$) (Figure 7.4).

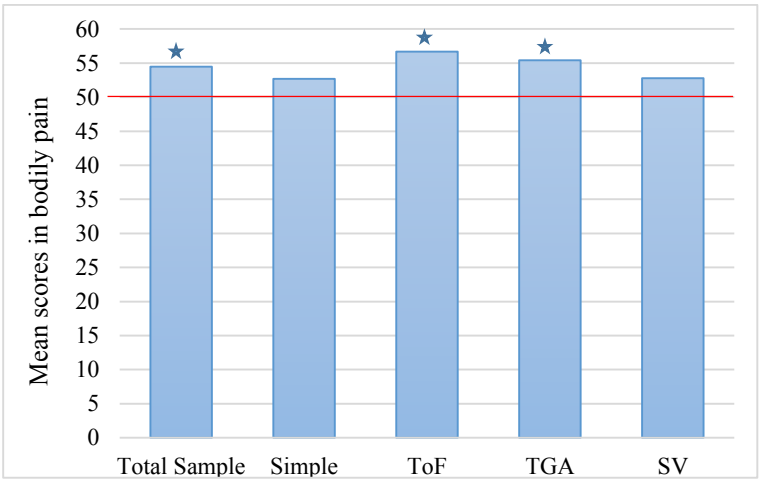


Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.4. Comparisons with norm in physical functioning

No differences were observed between the total sample and diagnostic groups and the norm in the role physical subscale. In the bodily pain subscale, the total sample

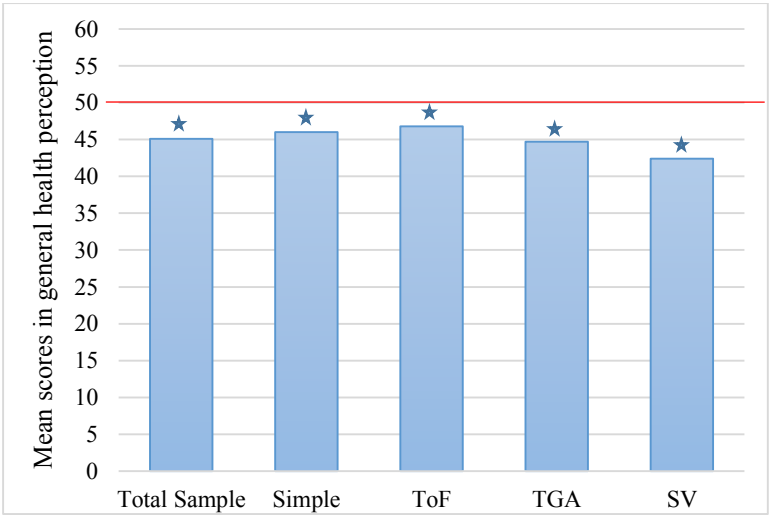
and ToF and TGA groups reported better HRQoL than the norm ($p < .001$) (Figure 7.5).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.5. Comparisons with norm in bodily pain

In the general health subscale, the total sample and all diagnostic groups reported poorer HRQoL than the norm ($p < .01$) (Figure 7.6).

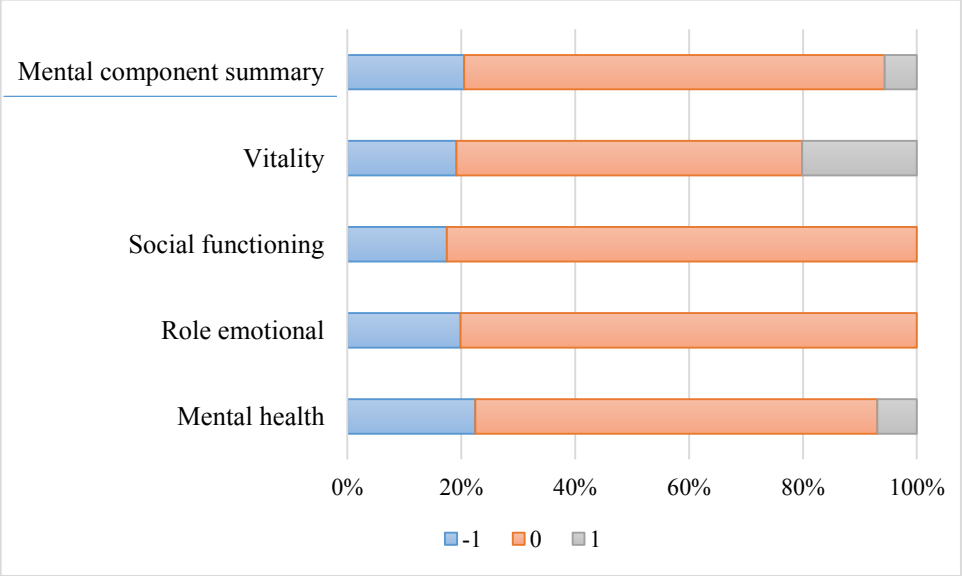


Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.6. Comparisons with norm in general health

7.2.2. Psychosocial HRQoL

In the total sample, the majority (range: 60.8- 82.5%) scored within the average in the mental component summary and all of the subscales, followed by those who scored below the norm mean (range: 17.5- 22.4%) (Figure 7.7). A smaller number (range: 0- 6.9%) scored above the norm mean. However, in the vitality subscale, slightly more people scored above the norm mean (20.1%) than below the nom mean (19.1%). None scored above the norm mean in the social functioning and role emotional subscales.



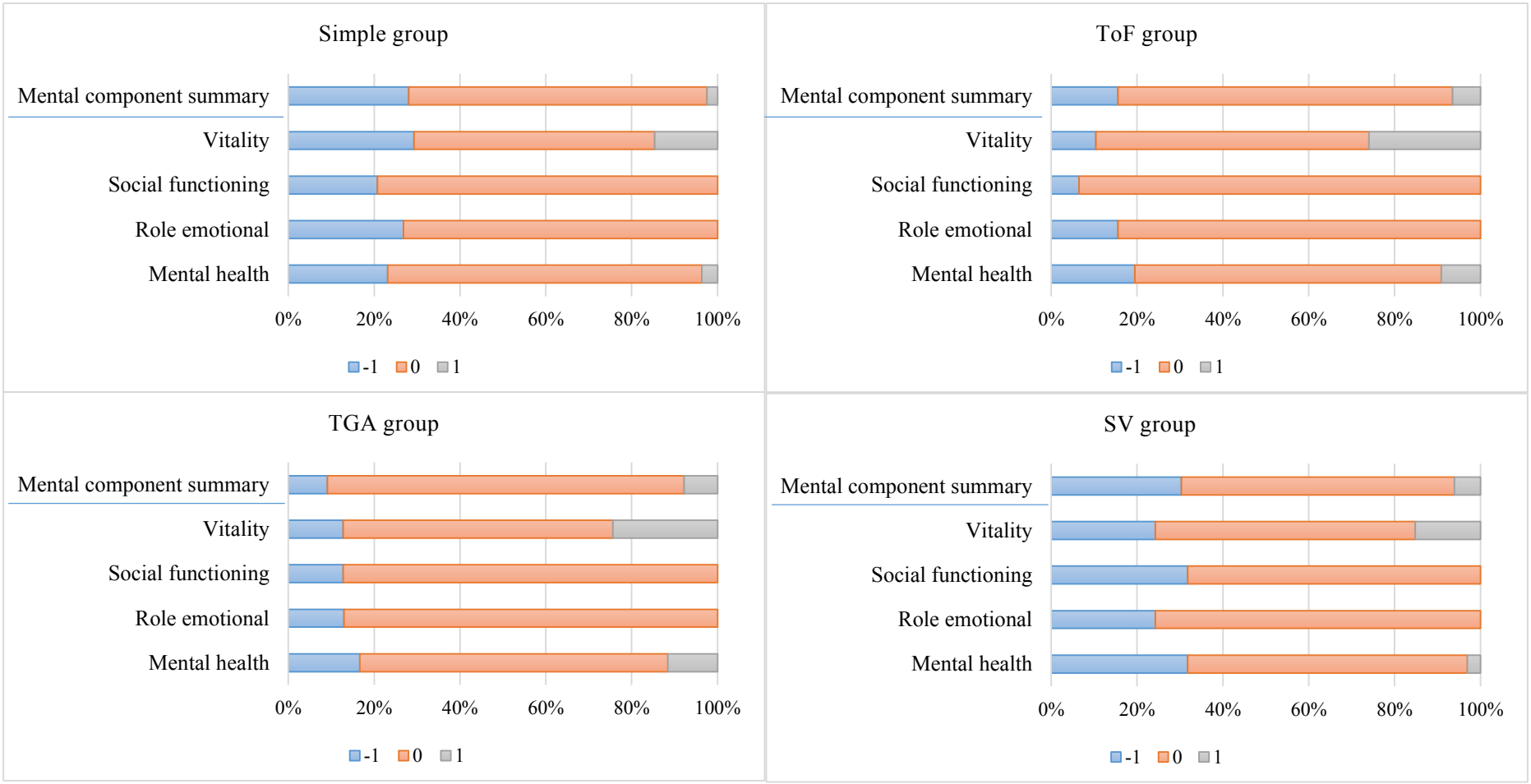
Note. -1= 1 *S.D.* or more below the norm mean, 0= within the norm mean, 1= 1 *S.D.* or more above the norm mean

Figure 7.7. Distribution of psychosocial HRQoL scores for total sample

The same pattern of results was observed within each of the diagnostic groups, with the exception of ToF and TGA groups (Figure 7.8). In these groups, more people scored above (ToF: 26%, TGA: 24.4%) than below the norm mean (ToF: 10.4%, TGA: 12.8%) in the vitality subscale.

More people scored below the norm mean in mental health than any other psychosocial HRQoL subscale. This pattern was evident in the total sample and in

the ToF, TGA, and SV groups. In the Simple group, more people scored below the norm in vitality than any other subscale. On the other hand, more people scored above the norm mean in vitality than any other subscale. This pattern was evident in the total sample and across all diagnostic groups.



Note. -1= 1 *S.D.* or more below the norm mean, 0= within the norm mean, 1= 1 *S.D.* or more above the norm mean

Figure 7.8. Distribution of psychosocial HRQoL scores for each diagnostic group

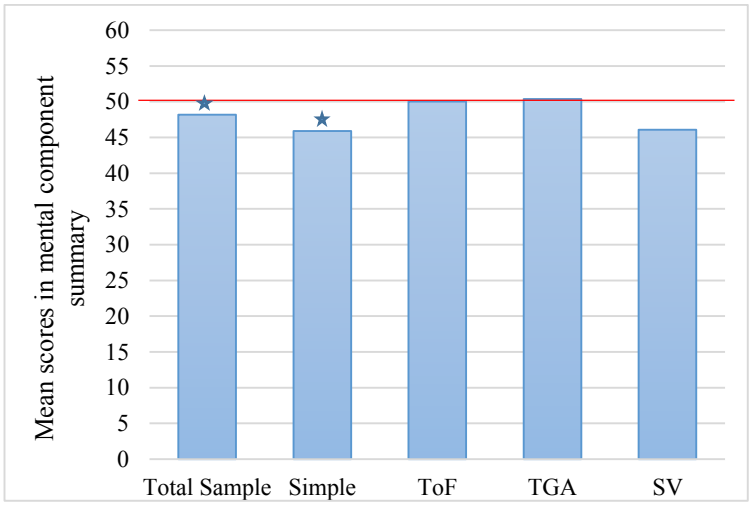
The standardised mean scores of the total sample and diagnostic groups for psychosocial HRQoL compared with the norm mean and results from *t*-tests are presented in Table 7.2.

Table 7.2. *T*-tests: psychosocial HRQoL comparisons with norms

Scale/Subscale		*Mean (S.D.)*	*T-test*	*Sig.*	*99% CI of the Difference*	*d*
Mental component summary						
	Total	48.2 (10.9)	*t*(301)= -2.95	**.003**	-3.47, -0.22	.17
	Simple	45.9 (12)	*t*(81)= -3.06	**.003**	-7.57, -0.56	.37
	ToF	50 (9.4)	*t*(76)= -0.00	.997	-2.83, 2.82	.00
	TGA	50.4 (9.1)	*t*(76)= 0.38	.703	-2.33, 3.12	.04
	SV	46.1 (12.2)	*t*(65)= -2.56	.013	-7.84, 0.14	.35
Vitality						
	Total	50.8 (10.6)	*t*(302)= 1.36	.176	-.075, 2.40	.08
	Simple	47.8 (11.1)	*t*(81)= -1.82	.073	-5.44, 1.00	.21
	ToF	53.6 (9.1)	*t*(76)= 3.43	**.001**	0.82, 6.32	.38
	TGA	53 (9.6)	*t*(77)= 2.72	**.008**	0.08, 5.85	.31
	SV	48.9 (11.5)	*t*(65)= -0.80	.429	-4.88, 2.63	.10
Social functioning						
	Total	48.7 (10.5)	*t*(302)= -2.13	.034	-2.84, 0.28	.13
	Simple	48.4 (10.6)	*t*(81)= -1.37	.175	-4.68, 1.48	.16
	ToF	51.4 (8.4)	*t*(76)= 1.48	.142	-1.11, 3.97	.15
	TGA	50 (8.9)	*t*(77)= -0.03	.976	-2.68, 2.62	.00
	SV	44.5 (12.9)	*t*(65)= -3.47	**.001**	-9.75, -1.30	.48
Role emotional						
	Total	49.2 (11)	*t*(301)= -1.21	.227	-2.40, 0.87	.08
	Simple	47.4 (12.6)	*t*(81)= -1.89	.062	-6.28, 1.03	.23
	ToF	50.3 (9.9)	*t*(76)= 0.25	.806	-2.70, 3.26	.03
	TGA	51.5 (8.9)	*t*(76)= 1.49	.139	-1.16, 4.18	.16
	SV	47.7 (11.8)	*t*(65)= -1.60	.114	-6.16, 1.52	.21
Mental health						
	Total	47.9 (10.7)	*t*(302)= -3.41	**.001**	-3.69, -0.50	.20
	Simple	46.9 (11.2)	*t*(81)= -2.53	.014	-6.40, 0.14	.29
	ToF	49.7 (9.6)	*t*(76)= -0.28	.778	-3.20, 2.58	.03
	TGA	49.2 (10)	*t*(77)= -0.69	.495	-3.78, 2.22	.08
	SV	45.6 (11.6)	*t*(65)= -3.12	**.003**	-8.22, -0.67	.41

Note. Norm mean 50 (10). Positive and negative t values indicate better and poorer HRQoL respectively. Cohen's *d*: .20= small, .50= medium, .80= large.

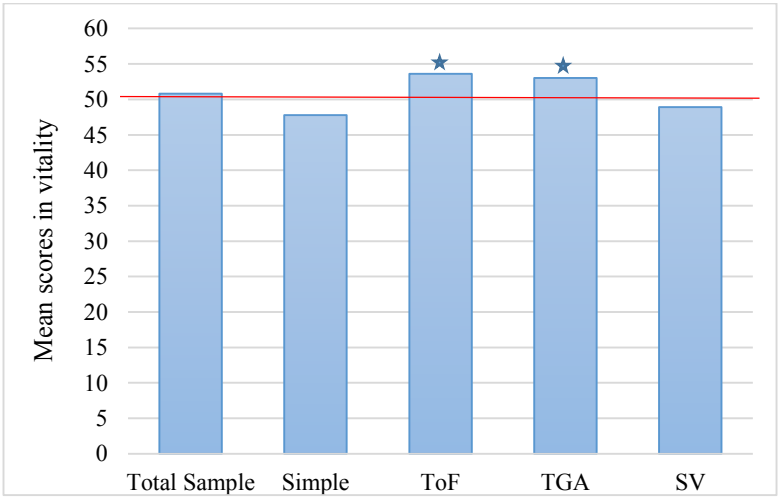
In the mental component summary, the total sample and Simple group reported poorer HRQoL than the norm ($p < .01$) (Figure 7.9).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.9. Comparisons with norm in mental component summary

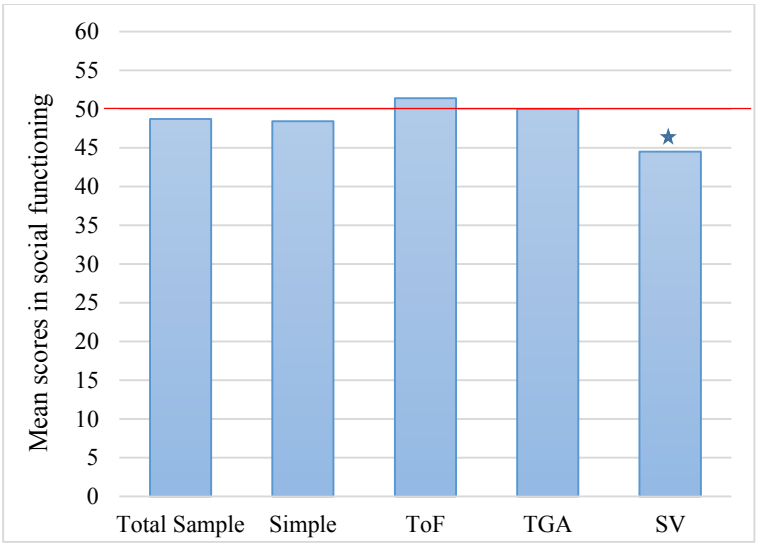
In the vitality subscale, the ToF and TGA groups reported better HRQoL than the norm ($p < .01$) (Figure 7.10).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.10. Comparisons with norm in vitality

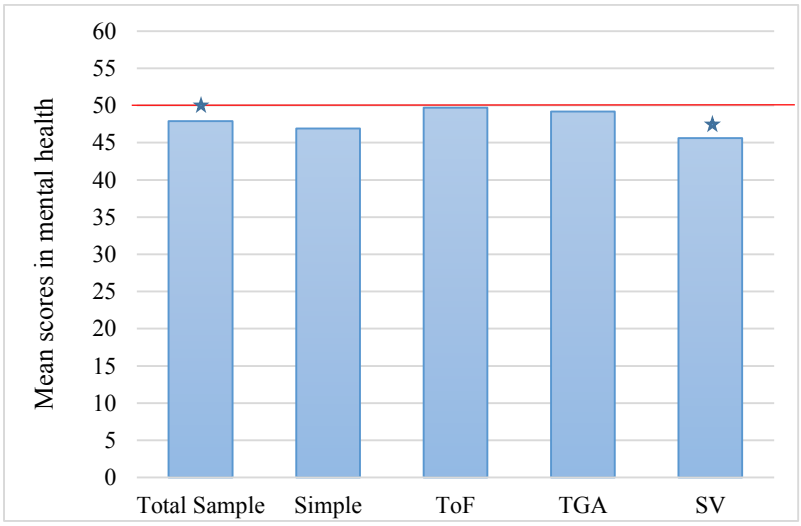
In social functioning, the SV group reported poorer HRQoL than the norm ($p < .01$) (Figure 7.11).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.11. Comparisons with norm in social functioning

No differences were observed between the total sample and diagnostic groups and the norm in the role emotional subscale. In the mental health subscale, the total sample and SV group reported poorer HRQoL than the norm ($p < .01$) (Figure 7.12).



Scale 0 – 100, higher score= better HRQoL.
Norm mean= 50. Star indicates significant difference between the group and the norm.

Figure 7.12. Comparisons with norm in mental health

7.3. HRQoL comparisons between diagnostic groups

Comparisons between the four diagnostic groups were conducted for the generic (physical and psychosocial) and disease-specific HRQoL.

7.3.1. Generic physical HRQoL

Results from the comparisons between the diagnostic groups for generic *physical* HRQoL are presented in Table 7.3.

Table 7.3. ANOVA: group comparisons in generic physical HRQoL

Scale/Subscale		***Mean (S.D.)***	***Model Parameters***			
			f	***df***	***Sig.***	η^2
Physical component summary			2.93*	161.15	.035	.03
	Simple	51.5 (10)				
	ToF	52.8 (8.7)				
	TGA	51 (7.5)				
	SV	47.9 (10.8)				
Physical functioning			4.60*	161.06	**.004**	.06
	Simple	50.9 (9.2)				
	ToF	51.4 (7.2)				
	TGA	50.5 (7.5)				
	SV	45.8 (10.6)				
Role physical			2.20*	161.10	.091	.03
	Simple	50.4 (9.9)				
	ToF	52.2 (8.5)				
	TGA	51.3 (8.4)				
	SV	47.9 (11.6)				
Bodily pain			3.01*	163.63	.032	.03
	Simple	52.7 (11.8)				
	ToF	56.7 (8.7)				
	TGA	55.4 (10)				
	SV	52.8 (10.3)				
General health			2.16*	162.54	.095	.02
	Simple	46 (11.5)				
	ToF	46.8 (9.8)				
	TGA	44.7 (9)				
	SV	42.4 (11.5)				

*Welch ANOVA.
Note. η^2: .01= small, .06= medium, .14= large.

There was no significant effect of diagnostic group on the physical component summary and the following subscales: role physical, bodily pain, general health (p> .01). There was a significant effect of diagnostic group on physical functioning (p< .01). Post-hoc comparisons indicated that the SV reported poorer physical functioning than the ToF group (mean difference= -5.58, p= .003, 99% CI [-10.50, -0.66]; d= .63) (see Figure 7.4, page 204).

7.3.2. Generic psychosocial HRQoL

The comparisons between the diagnostic groups for generic *psychosocial* HRQoL are presented in Table 7.4.

Table 7.4. ANOVA: group comparisons in generic psychosocial HRQoL

Scale/Subscale		*Mean (S.D.)*	*Model Parameters*			
			f	*df*	*Sig.*	η^2
Mental component summary			3.76*	161.55	.012	.04
	Simple	45.9 (12)				
	ToF	50 (9.4)				
	TGA	50.4 (9.1)				
	SV	46.1 (12.2)				
Vitality			6.07	299	**.001**	.06
	Simple	47.8 (11.1)				
	ToF	53.6 (9.1)				
	TGA	53 (9.6)				
	SV	48.9 (11.5)				
Social functioning			4.99*	160.68	**.002**	.06
	Simple	48.4 (10.6)				
	ToF	51.4 (8.4)				
	TGA	50 (8.9)				
	SV	44.5 (12.9)				
Role emotional			2.72*	161.90	.046	.03
	Simple	47.4 (12.6)				
	ToF	50.3 (9.9)				
	TGA	51.5 (8.9)				
	SV	47.7 (11.8)				
Mental health			2.47	299	.062	.02
	Simple	46.9 (11.2)				
	ToF	49.7 (9.6)				
	TGA	49.2 (10)				
	SV	45.6 (11.6)				

*Welch ANOVA.
Note. η^2: .01= small, .06= medium, .14= large.

There was no significant effect of diagnostic group on the mental component summary and the following subscales: role emotional, mental health (p> .01). There was a significant effect of diagnostic group on vitality, and social functioning (p< .01). Post-hoc analysis indicated that the Simple group reported less vitality than the ToF (mean difference= -5.79, p= .003, 99% CI [-10.99, -0.58]; d= .57) (see Figure 7.10, page 210). The SV group reported poorer social functioning than the ToF

(mean difference= -6.96, *p*= .002, 99% CI [-12.88, -1.03]; *d*= .65) group (see Figure 7.11, page 211).

7.3.3. Disease-specific HRQoL

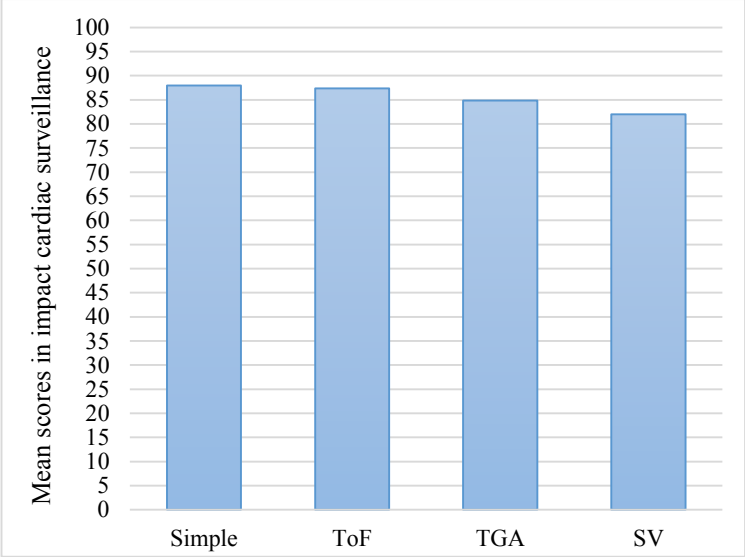
Results from the comparisons between the diagnostic groups for disease-specific HRQoL are presented in Table 7.5.

Table 7.5. ANOVA: group comparisons in disease-specific HRQoL

Subscale		***Mean (S.D.)***	***Model Parameters***			
			f	***df***	***Sig.***	η^2
Symptoms			4.02	299	**.008**	.04
	Simple	87.41 (14.22)				
	ToF	87.96 (12.28)				
	TGA	87.70 (13.04)				
	SV	80.92 (16.63)				
Impact cardiac surveillance			5.04	299	**.002**	.05
	Simple	87.96 (10.49)				
	ToF	87.38 (9.36)				
	TGA	84.88 (10.04)				
	SV	81.97 (11.36)				
Worries			4.03	299	**.008**	.04
	Simple	82.10 (14)				
	ToF	84.69 (13.60)				
	TGA	84.17 (15.32)				
	SV	76.79 (17.14)				

Note. η^2: .01= small, .06= medium, .14= large.

There was a significant effect of diagnostic group on all subscales, including symptoms, impact cardiac surveillance, and worries (p< .01). Post-hoc comparisons did not reveal statistically significant differences between the diagnostic groups in the symptoms and worries subscales. In the impact cardiac surveillance subscale, the SV group reported greater impact and therefore poorer HRQoL than the Simple (mean difference= -5.99, p= .003, 99% CI [-11.38, -0.60]; *d*= .55) (Figure 7.13).



Scale 0 – 100, higher score= better HRQoL.

Figure 7.13. Mean scores in impact cardiac surveillance by diagnostic group

7.4. Discussion

This chapter presented the analyses conducted to examine the impact of ACHD on HRQoL and to investigate differences in HRQoL between four diagnostic groups. It was postulated that people with ACHD would report significantly poorer HRQoL compared with the general population norm, predominantly in physical HRQoL. In addition, it was hypothesized that impairments in HRQoL would be predominant in the SV group who would report poorer HRQoL compared with the general population. Differences between diagnostic groups were expected to be prominent between the SV and Simple group, with the SV reporting poorer HRQoL. The next sections will discuss the findings of the present study in relation to previous literature and provide explanations for what these findings might mean.

7.4.1. Comparisons with general population norms in generic HRQoL

7.4.1.1. Physical HRQoL

The study showed that there were no differences between the total sample or the diagnostic groups and the general population norms in the physical component

summary of the SF-36. This finding supports those observed in a previous study that used the physical component summary in a mixed ACHD sample (Müller et al., 2013b). In another study however, Müller et al. (2014) found poorer HRQoL in the physical component summary in a mixed sample. This discrepancy may be attributed to the inclusion of a larger proportion of complex ACHD in their recent study, as this group (i.e. SV) tended towards lower scores in the physical component summary compared with the general population in the present study. A closer examination of the differences between the current sample (total and diagnostic groups) and the general population in the subscales that comprise the summary scale provides a more detailed account of HRQoL in various diagnostic groups.

The lack of impairment in most physical subscales of HRQoL across the total sample and diagnostic groups can help explain the normal physical component summary score. With the exception of poorer general health in the total sample and diagnostic groups and poorer physical functioning in the SV group, physical HRQoL in this sample was comparable and sometimes better than the general population in role limitations due to physical problems and bodily pain respectively.

General health was perceived to be poorer than the general population, irrespective of diagnostic group. There was some variability in the general health scores but a large proportion (30-40%) in each diagnostic group reported poorer general health, especially in the SV group. In accordance with these results, previous studies in ACHD have found poor general health in mixed samples (e.g. Enomoto et al., 2013; Hager & Hess, 2005; Jefferies et al, 2004; Riley et al., 2012). These findings suggest that people with ACHD seem to acknowledge that their general health is compromised due to their condition, hence their general health perception appears to

hold true. This may be the result of long-term management of ACHD. These individuals attend follow-up appointments in a specialist clinic and experience repeat interventions and long-term medication uptake, which may act as a continuous reminder that their health is compromised.

Despite poor general health, the total sample and the Simple, ToF, and TGA groups reported normal *physical functioning*, with only the SV group reporting poorer physical functioning than the general population. A small number of people with Simple, ToF, and TGA (<10%) also scored below the norm in this subscale but there was a larger proportion of lower scores in people with SV (>20%). These results differ from the majority of previous studies in ACHD that have found poorer physical functioning in mixed samples. However, very few of these studies have distinguished between diagnostic groups. Bruto et al. (2007) found poorer physical functioning in a mixed sample, yet when comparing the various diagnostic groups with norms, poorer physical functioning was only evident in the most complex. Consistent with the present study, previous studies that included a single diagnostic group reported normal physical functioning in Simple (Buys et al., 2013; Hanninen et al., 2011; Kahya Eren et al., 2013; Kamphuis et al., 2002a) and poorer physical functioning in SV samples (Idorn et al., 2013; Pike et al., 2012; van den Bosch et al., 2004; Winter et al., 2008). The findings of studies in ToF and TGA samples have been less consistent; while some studies reported no differences between people with ToF/TGA and the general population (e.g. Loup et al., 2009; Opić et al., 2013), others found poorer physical functioning in these individuals (e.g. Irtel et al., 2005; Hickey et al., 2012). The small sample sizes in some of these studies make it difficult to draw conclusions regarding the ToF and TGA groups.

The finding of lack of *role limitations due to physical problems* in the total sample and all diagnostic groups supports the findings of previous studies (Fteropoulli et al., 2013). Although not statistically significant, there was a tendency for lower scores in the SV group compared with the norms, which may help explain the small number studies in the systematic review that have found greater role limitations due to physical problems in people with SV than the general population (e.g. Hickey et al., 2012; Idorn et al., 2013; Pike et al., 2012; Winter et al., 2008). The findings of the present study suggest that people with ACHD are able to carry on with fulfilling their usual role activities without feeling that their condition interferes with them. It is likely that by accepting their limitations they are able to adjust to ACHD by either engaging in less physically demanding activities or even avoiding such activities altogether (Claessens et al., 2005).

It is also important to highlight that the total sample and especially the ToF and TGA groups experienced less *bodily pain*, whilst the Simple and SV groups had pain levels comparable with the general population. The majority of previous studies have found normal pain levels in people with ACHD, which may be explained by the inclusion of either a large proportion of people from the Simple and SV groups in mixed sample studies (e.g. Kamphuis et al., 2002b; Müller et al., 2011) or the inclusion of exclusively either of these two groups in others (e.g. Angeli et al., 2012; Buys et al., 2013; Hanninen et al., 2011; van den Bosch et al, 2004). Similar to the present study, a smaller number of studies also reported less pain in mixed (e.g. Bruto et al., 2007; Mokhles et al., 2011) and ToF (Hickey et al., 2012; Knowles et al., 2012) samples. One possible explanation for the normal or better scores in bodily pain may be that people with ACHD experience symptom relief following

interventions (Mokhles et al., 2011). This notion was further supported by Lane et al. (2002), who found that people with inoperable ACHD experienced greater pain than those who had surgery. Another plausible explanation is that the lifelong nature of ACHD may have resulted in a higher pain threshold, hence a different point of reference for judging bodily pain in people with ACHD than the general population.

Taken together, the findings regarding physical HRQoL indicate that people with ACHD as a whole experience good HRQoL. Examining each of the diagnostic groups in comparison to normative data demonstrated that physical limitations are prominent in the SV group. One implication of these findings is that it is potentially more informative to distinguish between diagnostic groups in order to establish what is causing the effect in studies with mixed samples.

7.4.1.2. Psychosocial HRQoL

The study showed differences in the mental component summary scores of the SF-36 with the total sample reporting poorer HRQoL compared with the norm. All three previous studies that used the mental component summary reported HRQoL comparable to the general population (Enomoto et al., 2013; Müller et al., 2013b; Müller et al., 2014). It is difficult to explain this discrepancy but it might be related to cultural differences as the abovementioned studies have been conducted in Japanese and German samples. Although the low scores evident in the present study can be mainly attributed to the Simple and SV groups. The SV group tended towards lower scores in the mental component summary but this did not reach statistical significance. The Simple group did, however, score significantly lower in the mental component summary compared with the general population. This finding corroborates those of a recent study with exclusively people with CoA (Buys, Budts,

Delecluse, Vanhees, 2013). The authors found that people with CoA experienced significant impairment in psychosocial domains of their HRQoL and argued that CoA may not be as simple as it was considered to be in the past. In the present study a large proportion (46%) of people comprising the Simple group had a CoA diagnosis. This rather surprising finding has important implications about the expected impact of ACHD on the HRQoL of people in the Simple group. This group is structurally simpler compared with the other diagnostic groups in the study and despite possible long-term complications, they are generally expected to be free from disease burden, evident from the minimal impact on their physical HRQoL (see section 7.4.1.1, page 216). However, the Simple group consisted of people that still require health monitoring at a specialist clinic as opposed to those who are physically well, free from long-term complications, and more likely to have been discharged and followed-up by their local general practitioners. Another possible explanation may be related to expectations. Being told that their condition is "simple" in nature, that their long-term prognosis is excellent, and that they are expected to live "normal" lives may lead to unrealistic expectations (Warnes, 2005). People in the Simple group may expect that their functioning levels will get back to their normal standards after what they may consider curative surgery, especially if they had operations during adulthood (Lane et al., 2002). If people's expectations are not fulfilled and they still experience minor limitations, this may result in greater emotional impact.

The findings in the specific subscales that comprise the mental component summary indicated poorer mental health in the total sample and SV group and poorer social functioning in the SV group compared with the general population. Participants had

comparable and sometimes better HRQoL than the general population in the subscales of role limitations due to emotional problems and vitality respectively.

The finding of poorer *mental health* in the total sample compared with the general population suggests significant emotional impact of ACHD in these individuals. This finding is contrary to the majority of studies identified in the systematic review (Fteropoulli et al., 2013) and can be attributed to the lower scores of the Simple and SV groups, which would have negatively influenced the mental health score of the total sample. Indeed, the scores of the ToF and TGA groups in this subscale were not different from the norms, whereas the scores of the SV group were significantly lower (and near significant for the Simple group). Furthermore, more than 30% of those with SV and 20-25% of those in the Simple group scored lower than the norm, while less than 20% of those with ToF and TGA scored lower than the norm. A detailed examination of the sample composition in previous studies revealed that normal mental health was mostly evident in samples that consisted of either people with ToF (e.g. Daliento et al., 2005), TGA (e.g. Müller et al. 2013c) or a large proportion of these diagnostic groups (e.g. Bruto et al., 2007; Loup et al., 2009). Some studies that have found normal mental health in samples with SV as opposed to the present study were small in sample size ($n < 60$), which might have resulted in lack of power to detect an effect. Two previous studies that included people with SV shared the findings of the present study regarding impaired mental health in this diagnostic group (e.g. Angeli et al., 2012; van den Bosch et al., 2004). These findings suggest that increased survival into adulthood in people with SV brings about emotional challenges that warrant attention. This was expected as people with

SV represent the most complex group which can pose significant physical and emotional burden.

In addition to poorer mental health, people with SV also reported poorer *social functioning,* while all other diagnostic groups and the total sample were comparable to the general population. More than 30% of those with SV scored lower than the norm in this subscale, while a smaller proportion did so in all other diagnostic groups. The majority of the studies in the systematic review reported normal social functioning in mixed ACHD samples, which is in agreement with the present study. However, the present study contradicts the findings of previous studies in SV samples, which have found normal social functioning (e.g. Angeli et al., 2012; Idorn et al., 2013; Pike et al., 2012; van den Bosch et al., 2004). This discrepancy may be attributed to rather smaller samples included in previous studies compared with the current study, which might have resulted in their lack of power to detect an effect.

Despite some limitations in the Simple and SV groups, participants did not differ significantly from the general population in *role limitations due to emotional problems* irrespective of diagnostic group. This finding is in line with other studies as evidenced in the systematic review (Fteropoulli et al., 2013). This finding complements the finding of lack of role limitations due to *physical* problems (section 7.4.1.1, page 216) and suggests that irrespective of whether people experience significant emotional burden from their condition their usual role activities remain unaffected. As mentioned earlier, this may be indicative of successful adaptation to ACHD and in this case its emotional burden. Some authors have suggested that people with ACHD develop a strong sense of coherence, which is characterised by a

feeling of understanding, manageability, and meaningfulness that helps them find meaning in their experiences and adapt to their condition (Moons et al., 2006).

This study also showed that the sample experienced similar and in the case of ToF and TGA better HRQoL in *vitality* compared with the general population. A large proportion of people with ToF and TGA (>20%) reported greater vitality than the norm as opposed to the smaller proportion (<15%) who reported lower vitality. The majority of previous studies reported normal vitality levels (e.g. Gratz et al., 2009), while a smaller number studies reported lower vitality levels (e.g. Müller et al., 2011) in people with ACHD compared with the general population. Only one recent study of people with TGA who had arterial switch (as opposed to atrial switch as in the current TGA sample) reported greater vitality in these individuals than the general population (Ruys et al., 2013). Although these findings are not directly comparable to the present study both studies suggest that people with TGA, regardless of the type of operation, have excellent outcomes in this particular domain. An explanation for the normal or high vitality levels may be that vitality was rated based on the degree of functioning, as people with ACHD may not engage in the same energy demanding activities as average healthy individuals generally do. If this is the case it is possible that people with ACHD have successfully adjusted to their limitations by engaging in less energy demanding activities more suited to their capabilities (Claessens et al., 2005). In this study, however, it was not possible to establish whether the higher than average vitality levels in people with ToF and TGA and their generally high ratings in most HRQoL subscales are due to them adjusting their expectations and activities to their capacity.

Overall, the findings regarding psychosocial HRQoL indicate that people may experience significant emotional burden as a result of living with ACHD, particularly people in the SV and Simple groups. On the other hand, people in the ToF and TGA groups appear to have normal psychosocial HRQoL. These findings raise interesting questions regarding patient expectations and the degree of adjustment of expectations based on people's capacity.

7.4.2. Comparisons between diagnostic groups in HRQoL

The impact of ACHD on HRQoL was explored further with comparisons between the four diagnostic groups on generic and disease-specific HRQoL.

7.4.2.1. Generic HRQoL

Comparisons between the diagnostic groups in physical HRQoL did not show differences in the physical component summary and the subscales of role limitations due to physical problems, bodily pain, and general health. There were, however, differences in physical functioning with the SV group experiencing poorer HRQoL than the ToF in this subscale. This finding is in line with previous research which has found impaired physical functioning in people with SV compared with other groups (e.g. Gratz et al., 2009). This was not surprising as people with SV experienced significant physical burden, including arrhythmias, long-term cyanosis, repeat interventions, multiple medication, and compromised ventricular function compared with the other diagnostic groups (see section 6.6, page 178). The multiple long-term complications associated with SV may lead to reduced physical functioning including an impact on walking, running, and other daily activities as evidenced in the comparisons with the general population (section 7.4.1.1, page 216). Notably, the SV group differed from the ToF but not the Simple group as

originally hypothesized. Generally, the ToF group appeared to have good HRQoL with a tendency towards higher scores than the other groups in physical domains, including physical functioning.

Further comparisons between the diagnostic groups in psychosocial HRQoL did not indicate differences in the mental component summary and the subscales of role limitations due to emotional problems and mental health. The SV group did report poorer social functioning compared with the ToF group. The physical manifestation of SV in the form of symptoms, repeat interventions and hospitalizations may hinder people's ability to participate in social activities and increase the likelihood of social isolation (McMurray et al., 2001). Similar to the group comparisons in physical functioning, the SV group differed from the ToF but not the Simple group as originally hypothesized. This can also be attributed to the tendency of the ToF group towards higher scores than the other diagnostic groups in psychosocial domains, including social functioning.

The generally good HRQoL in people with ToF was also evident in relation to vitality, where they reported better HRQoL compared with those with a Simple condition. This finding corroborates the findings of Ternestedt et al. (2001), who also found greater vitality in people with ToF than those with ASD. The authors proposed that people with ToF are generally more purposeful and less willing to compromise compared to those with ASD but did not provide detailed explanation for this assumption. It is difficult to establish the specific pathways that lead to the overall good HRQoL (including high vitality levels) evident in people with ToF compared with other diagnostic groups. To speculate, there may be a difference in their standards of what constitutes good HRQoL in this particular and other domains.

People in the Simple group may generally expect their vitality levels to be similar to healthy individuals, whereas those with ToF may have adjusted their expectations to their capacity. Further studies with more focus on the expectations of various diagnostic groups are therefore suggested.

7.4.2.2. Disease-specific HRQoL

Disease-specific HRQoL measures are believed to be more sensitive to subtle differences between people with various diagnoses. In the present study, however, the ACHD-specific measure failed for the most part to distinguish between the four diagnostic groups. A significant effect was observed for all three subscales, however, significant group differences were only observed in the *impact of cardiac surveillance* but not in *symptoms* and *worries*.

The study showed that people with SV experienced greater impact of cardiac surveillance compared with those in the Simple group. In the original CHD-TAAQOL validation study, Kamphuis and colleagues (2004) examined differences between two groups (mild and complex) and found poorer HRQoL in the complex group compared with the mild group on all three scales. The difference in findings with the current study may be attributed to differences in the composition of the samples. In the Kamphuis et al. study the mild group consisted of people not requiring treatment and specialist follow-up while the complex group consisted of all others who had undergone operations and were followed-up in a specialist clinic. In the current study the sample consisted of four diagnostic groups varied in structural complexity and were all followed-up in a specialist clinic. Thus, it may be that the CHD-TAAQOL is able to discriminate between relatively healthy and ill patients but not across the entire spectrum of ACHD complexity.

The findings of the present study suggest that people with SV find cardiac surveillance more burdening than those in the Simple group. The subscale of impact cardiac surveillance measures the impact (frequency weighted by emotional impact) of diagnostic tests and hospitalizations. It may be expected that the SV group would report greater impact of cardiac surveillance as due to the nature and complexity of this diagnostic group people need regular monitoring with blood, MRI, ECHO, ECG, and exercise testing annually or biannually. In addition, they are generally more likely to be hospitalized due to complications compared with the other diagnostic groups (see section 1.8, page 40 & section 6.6, page 178). Regardless of the frequency, cardiac surveillance may have a greater impact on people with SV due to its specific purpose. Generally, cardiac testing in these individuals is not only performed to ensure their continuous functioning but to also identify and manage long-term complications in a timely manner (Warnes et al., 2008).

This finding was supportive of the findings from group comparisons on generic HRQoL in that both identified HRQoL impairment in people with SV. The disease-specific measure, however, showed differences between the SV and the Simple group as it was originally hypothesized, whereas the generic measure showed differences between the SV and the ToF group. This may indicate that the CHD-TAAQOL has good discriminant validity in the impact of cardiac surveillance but not in the symptoms and worries scales where there was lack of group differences. Considering the observed group differences in the physical functioning subscale of the SF-36, it may be expected that diagnostic groups would differ in the symptoms scale of the CHD-TAAQOL. Validation of the measure was beyond the scope of the

study and it may be informative if future studies used the CHD-TAAQOL to validate further its use in people with ACHD.

7.5. Summary

The present study is one of the largest studies that examined HRQoL in people with ACHD and differences in HRQoL between four diagnostic groups. Overall, there were few differences between the total sample and the general population. The impact of ACHD on HRQoL was not predominant in physical HRQoL domains as hypothesized, rather some differences were observed between people with ACHD and the general population in both physical and psychosocial HRQoL domains. The comparisons between each diagnostic group and the general population further indicated that physical and psychosocial HRQoL impairments were as hypothesized predominant in the SV group. Notably, people in the Simple group also reported some impairment in psychosocial HRQoL. On the other hand, those with ToF and TGA generally reported good or excellent HRQoL in most domains compared with the general population. Comparisons between the diagnostic groups on generic and disease-specific HRQoL supported the above findings regarding HRQoL impairment in the SV group and the overall good HRQoL in the ToF group. Differences between diagnostic groups were mostly evident between the SV and ToF group, with the latter reporting poorer HRQoL. The categorisation of people with ACHD into four diagnostic groups helped identify specific HRQoL issues for each group and built on previous studies with ill-defined samples. The findings highlight the importance of distinguishing between diagnostic groups against generalising across the whole ACHD population.

As discussed in Chapter 3 underlying psychosocial processes may also help explain some of the evident variation in HRQoL. The next chapter presents the findings of regression analyses conducted to study the demographic, clinical, and psychosocial factors associated with HRQoL.

CHAPTER 8 – FACTORS ASSOCIATED WITH HEALTH-RELATED QUALITY OF LIFE IN ADULT CONGENITAL HEART DISEASE

8.1. Prologue

The previous chapter described the impact of ACHD on HRQoL and found an impact on both physical and psychosocial HRQoL domains. The SV group reported poorer physical and psychosocial HRQoL compared with the general population and other diagnostic groups. However, the Simple group also experienced some impairment in psychosocial HRQoL compared with the general population. Some of the evident variation in HRQoL can be further explained by psychosocial factors. At present, limited research has examined the role of psychosocial factors in the HRQoL of people with ACHD. This chapter investigates the factors associated with HRQoL in ACHD, this includes comprehensive demographic and clinical measures along with psychosocial constructs drawn from the SRM and previous research using hierarchical multiple regressions. It was hypothesized that psychosocial factors, including illness perceptions, coping strategies, mood, and social support will account for a significant amount of variance in HRQoL over and above that accounted for by demographic and clinical characteristics.

8.2. Factors associated with HRQoL in ACHD

Before multivariate analyses were performed correlations between the predictors were examined for multicollinearity (see Appendix Q). As none of the relationships exceeded .80, multicollinearity was not considered to be of concern.

Bivariate regressions were conducted to examine the relationships between the predictor variables and HRQoL outcomes, which included generic physical HRQoL

(physical component summary and the subscales of physical functioning, role physical, bodily pain, general health of the SF-36), generic psychosocial HRQoL (mental component summary and the subscales of vitality, social functioning, role emotional, mental health of the SF-36), and disease-specific HRQoL (symptoms, impact of cardiac surveillance, worries of the CHD-TAAQOL) (see Appendix R). Some consistent and statistically significant associations were observed.

Of all of the demographic factors employment status exhibited the most consistent associations with both physical and psychosocial HRQoL. No consistent associations were observed between demographic factors and disease-specific HRQoL. Clinical factors that were consistently associated with physical HRQoL included number of co-morbidities, number of medication, and current O_2 saturation. No consistent associations were observed between clinical factors and psychosocial HRQoL. Clinical factors that were consistently associated with disease-specific HRQoL included being in the ToF group (vs Simple), number of co-morbidities, and current O_2 saturation. Illness perceptions except for illness coherence and curability were consistently associated with physical, psychosocial, and disease-specific HRQoL. Of all the coping strategies, venting, behavioural disengagement, and self-blame exhibited consistent associations with physical, psychosocial, and disease-specific HRQoL. None of the social support variables were associated with physical HRQoL, whilst only friend support was consistently associated with psychosocial HRQoL. No consistent associations were observed between social support and disease-specific HRQoL. Both anxiety and the presence of depressive symptoms were consistently associated with physical, psychosocial, and disease-specific HRQoL.

Only significant predictors from the bivariate regressions were entered into the hierarchical multiple regression models. This resulted in a sufficient subject to predictor variable ratio as discussed in section 5.6.8.3 (page 162). The order in which the predictor variables were entered into the hierarchical multiple regressions and a detailed description of the method used is presented in Figure 5.1 (page 163). The results of the hierarchical multiple regressions are reported in the next sections for generic (physical and psychosocial of the SF-36) and disease-specific (CHD-TAAQOL) HRQoL. The final model of each regression is presented and the full hierarchical multiple regressions that include all the steps are displayed in Appendix S. A table with a summary of the significant predictors is presented at the end of the results section (page 251).

8.2.1. Factors associated with generic physical HRQoL

The final model for physical component summary is displayed in Table 8.1. Demographic variables, including education level, employment status, and age were entered in Step 1 and explained 7.9% of the variance in physical component summary. The addition of diagnostic group, cyanosis days, and number of co-morbidities in Step 2 explained an additional 11.6% of the variance in the physical component summary. Hospitalization days, entered in Step 3 did not explain additional variance in the outcome. Current O_2 saturation, number of medication, and VO_2 max, were entered in Step 4 and explained a further 3.3% of the variance. Illness perceptions were entered in Step 5 and explained an additional 27.5% of the variance. Neither coping strategies entered in Step 6, nor mood entered in Step 7 explained any additional variance. The final model was significant, $f(22,278)= 13.48$; $p< .001$ and explained 47.8% of the variance in physical component

summary. Illness identity was the only unique predictor in this final model indicating that a perception of more severe symptoms due to ACHD was associated with poorer physical HRQoL.

Table 8.1. Final regression model for physical component summary (*n*= 301)

Predictor Variables	**β**	***sr*²**	
Education level	.066	.061	
Employment status	.045	.041	
Age	-.010	-.008	
ToF	.099	.074	
TGA	.020	.015	
SV	.114	.069	
Cyanosis days	-.066	-.046	
Co-morbidities no.	-.032	-.023	
Hospitalization days	-.069	-.058	
Current O_2 saturation	.075	.049	
Medication no.	.006	.004	
VO_2 max	.015	.013	
Brief IPQ consequences	-.126	-.076	
Brief IPQ controllability	.038	.035	
Brief IPQ identity	**-.517****	**-.332**	
Brief IPQ concern	-.057	-.040	
Brief IPQ emotional representation	.081	.047	
Brief COPE religion	-.064	-.060	
Brief COPE venting	.042	.035	
Brief COPE behavioural disengagement	-.010	-.008	
STAI-6 anxiety	-.037	-.030	
CES-D 10 depression	-.003	-.003	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.079**	**.069**	**.079****
Step 2	**.195**	**.172**	**.116****
Step 3	.202	.178	.008
Step 4	**.235**	**.203**	**.033***
Step 5	**.510**	**.481**	**.275****
Step 6	.515	.481	.005
Step 7	.516	.478	.001

Note. Educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

Analyses were also conducted for the subscales comprising the physical component summary including physical functioning, role physical, bodily pain, and general

health. A fuller description of the hierarchical multiple regressions including all the steps is displayed in Appendix S.

8.2.1.1. Physical functioning

The final model for the physical functioning subscale is displayed in Table 8.2. The final model was significant, $f(26,275)= 17.08$; $p< .001$ and explained 58.1% of the total variance in the physical functioning subscale. The unique predictors in the final model were illness identity, consequences, and emotional representation. Perceptions of more severe symptoms and consequences due to ACHD and a perception of less emotional impact were associated with poorer physical functioning.

Table 8.2. Final regression model for physical functioning (*n*= 302)

Predictor Variables	**β**	***sr*²**	
Gender	-.096	-.081	
Employment status	.083	.074	
ToF	.010	.007	
TGA	-.037	-.026	
SV	-.013	-.008	
Cyanosis days	.016	.011	
Co-morbidities no.	-.116	-.078	
Arrhythmias	.056	.041	
Intervention no.	.019	.015	
Hospitalization days	-.066	-.053	
Current O_2 saturation	.132	.084	
Medication no.	-.014	-.010	
VO_2 max	.038	.031	
LVEF	.018	.015	
Brief IPQ consequences	**-.216***	**-.130**	
Brief IPQ identity	**-.381****	**-.246**	
Brief IPQ concern	-.096	-.066	
Brief IPQ emotional representation	**.172***	**.098**	
Brief COPE religion	-.049	-.045	
Brief COPE denial	-.102	-.093	
Brief COPE venting	.102	.082	
Brief COPE substance use	-.099	-.089	
Brief COPE behavioural disengagement	-.020	-.015	
Brief COPE self-blame	-.004	-.003	
STAI-6 anxiety	-.103	-.082	
CES-D 10 depression	-.064	-.047	
Step	***R*²**	***Adjusted R*²**	***ΔR*²**
Step 1	**.099**	**.093**	**.099****
Step 2	**.273**	**.253**	**.174****
Step 3	.283	.258	.009
Step 4	**.346**	**.314**	**.064****
Step 5	**.573**	**.546**	**.227****
Step 6	**.605**	**.571**	**.032***
Step 7	.618	.581	.012

Note. Gender: 0= male, 1= female, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.1.2. Role physical

The final model for the role physical subscale is displayed in Table 8.3. The final model was significant, $f(14,286)= 13.52$; $p< .001$ and explained 36.9% of the total variance in the role physical sub-domain. The unique predictors in the final model were employment status, illness identity, and presence of depressive symptoms.

Being unemployed, a perception of more severe symptoms due to ACHD and the presence of depressive symptoms were associated with greater role limitations due to physical problems.

Table 8.3. Final regression model for role physical (*n*= 301)

Predictor Variables	**β**	***sr*²**	
Employment status	**.140***	**.130**	
Cyanosis days	-.113	-.086	
Co-morbidities no.	.048	.044	
Current O_2 saturation	.001	.001	
Brief IPQ consequences	-.176	-.108	
Brief IPQ controllability	.009	.009	
Brief IPQ identity	**-.304****	**-.201**	
Brief IPQ concern	.027	.020	
Brief IPQ emotional representation	.089	.052	
Brief COPE venting	-.022	-.018	
Brief COPE behavioural disengagement	.001	.000	
Brief COPE self-blame	-.013	-.011	
STAI-6 anxiety	-.058	-.046	
CES-D 10 depression	**-.249****	**-.190**	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.085**	**.082**	**.085****
Step 2	**.140**	**.131**	**.055****
Step 3	.148	.137	.009
Step 4	**.342**	**.322**	**.194****
Step 5	.351	.324	.009
Step 6	**.398**	**.369**	**.047****

Note. Employment status: 0= unemployed, 1= employed, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.1.3. Bodily pain

The final model for the bodily pain subscale is presented in Table 8.4. The final model was significant, $f(15,287)= 10.99$; $p< .001$ and explained 33.2% of the total variance in the bodily pain sub-domain. The only unique predictor in the final model was illness identity, indicating that a perception of more severe symptoms due to ACHD was associated with greater bodily pain.

Table 8.4. Final regression model for bodily pain (*n*= 303)

Predictor Variables	**β**	***sr*²**	
Co-morbidities no.	-.019	-.015	
Current O_2 saturation	-.045	-.041	
Medication no.	.048	.038	
Brief IPQ consequences	.006	.004	
Brief IPQ controllability	.067	.063	
Brief IPQ identity	**-.455****	**-.300**	
Brief IPQ concern	-.061	-.043	
Brief IPQ emotional representation	.040	.024	
Brief COPE religion	-.090	-.088	
Brief COPE denial	-.110	-.102	
Brief COPE venting	-.038	-.031	
Brief COPE behavioural disengagement	.003	.002	
Brief COPE self-blame	.030	.025	
STAI-6 anxiety	-.138	-.110	
CES-D 10 depression	-.077	-.059	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.032**	**.029**	**.032***
Step 2	.051	.041	.019
Step 3	**.317**	**.299**	**.266****
Step 4	.344	.314	.027
Step 5	**.365**	**.332**	**.021***

Note. CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

8.2.1.4. General health

The final model for the general health subscale is displayed in Table 8.5. The final model was significant, *f*(21,281)= 12.73; *p*< .001 and explained 44.9% of the total variance in the general health sub-domain. The unique predictors in this final model were illness identity and presence of depressive symptoms. A perception of more severe symptoms due to ACHD and presence of depressive symptoms were associated with poorer general health.

Table 8.5. Final regression model for general health (*n*= 303)

Predictor Variables	**β**	***sr*²**	
Education level	.112	.106	
Employment status	.017	.016	
Co-morbidities no.	.004	.003	
Intervention no.	.004	.003	
Hospitalization days	-.036	-.030	
Current O_2 saturation	-.001	-.001	
Medication no.	-.095	-.073	
VO_2 max	.017	.015	
RVEF	.073	.065	
Brief IPQ consequences	-.049	-.030	
Brief IPQ controllability	.075	.068	
Brief IPQ curability	.111	.101	
Brief IPQ identity	**-.313****	**-.202**	
Brief IPQ concern	-.105	-.073	
Brief IPQ emotional representation	-.093	-.054	
Brief COPE venting	.020	.016	
Brief COPE substance use	-.038	-.035	
Brief COPE behavioural disengagement	-.002	-.001	
Brief COPE self-blame	.005	.004	
STAI-6 anxiety	.037	.029	
CES-D 10 depression	**-.186***	**-.140**	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.071**	**.064**	**.071****
Step 2	**.125**	**.116**	**.054****
Step 3	.133	.118	.008
Step 4	**.191**	**.167**	**.059****
Step 5	**.465**	**.437**	**.273****
Step 6	.468	.432	.003
Step 7	**.488**	**.449**	**.020***

Note. Educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

8.2.2. Factors associated with generic psychosocial HRQoL

The final model for the mental component summary is displayed in Table 8.6. Demographic variables were entered in Step 1, explaining 9.5% of the variance in mental component summary. Illness perceptions were entered in Step 2 and explained an additional 25% of the variance. Coping strategies, entered in Step 3, explained a further 12% of the variance. Social support, entered in Step 4, did not explain additional variance. Mood was entered in Step 5 and explained an additional

13.3% of the variance. The final model was significant, $f(20,281)= 21.54$; $p< .001$ and explained 57.7% of the variance in the mental component summary. The unique predictors in the final model were age, self-blame, anxiety, and presence of depressive symptoms. Younger age, greater use of self-blame as a coping strategy, greater anxiety, and the presence of depressive symptoms were associated with poorer psychosocial HRQoL.

Table 8.6. Final regression model for mental component summary (*n*= 302)

Predictor Variables	**β**	***sr*²**	
Age	**.164****	**.151**	
Employment status	.074	.068	
Brief IPQ consequences	-.040	-.024	
Brief IPQ controllability	-.008	-.007	
Brief IPQ curability	.034	.031	
Brief IPQ identity	-.036	-.024	
Brief IPQ concern	-.036	-.026	
Brief IPQ emotional representation	-.065	-.038	
Brief COPE positive reframing	.070	.060	
Brief COPE acceptance	.044	.038	
Brief COPE self-distraction	-.059	-.050	
Brief COPE denial	-.027	-.024	
Brief COPE venting	-.019	-.015	
Brief COPE substance use	.000	.000	
Brief COPE behavioural disengagement	.029	.023	
Brief COPE self-blame	**-.172****	**-.142**	
MSPSS friends support	.056	.050	
MSPSS significant other support	-.006	-.005	
STAI-6 anxiety	**-.131***	**-.099**	
CES-D 10 depression	**-.421****	**-.319**	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.095**	**.089**	**.095****
Step 2	**.345**	**.327**	**.249****
Step 3	**.465**	**.435**	**.121****
Step 4	.472	.438	.006
Step 5	**.605**	**.577**	**.134****

Note. Employment status: 0= unemployed, 1= employed, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

Analyses were conducted for the subscales comprising the mental component summary including vitality, social functioning, role emotional, and mental health. A

fuller description of the hierarchical multiple regressions including all the steps is displayed in Appendix S.

8.2.2.1. Vitality

The final model for the vitality subscale is displayed in Table 8.7. The final model was significant, $f(23,279) = 18.91$; $p < .001$ and explained 57.7% of the total variance in the vitality sub-domain. The unique predictors were diagnostic group, illness identity and present of depressive symptoms. Being in the ToF, TGA, and SV vs. being in the Simple group was associated with greater vitality. In addition, a perception of more severe symptoms due to ACHD and the presence of depressive symptoms were associated with less vitality.

Table 8.7. Final regression model for vitality (*n*= 303)

Predictor Variables	**β**	***sr*²**	
Employment status	.079	.071	
Gender	-.036	-.032	
ToF	**.175****	**.135**	
TGA	**.147***	**.114**	
SV	**.157***	**.102**	
Co-morbidities no.	-.003	-.002	
Current O_2 saturation	.030	.023	
Medication no.	.022	.017	
Brief IPQ consequences	-.078	-.047	
Brief IPQ controllability	.046	.042	
Brief IPQ curability	.012	.011	
Brief IPQ identity	**-.376****	**-.245**	
Brief IPQ concern	-.009	-.006	
Brief IPQ emotional representation	.008	.005	
Brief COPE acceptance	.091	.082	
Brief COPE self-distraction	-.062	-.051	
Brief COPE denial	-.047	-.042	
Brief COPE venting	.018	.014	
Brief COPE substance use	.005	.004	
Brief COPE behavioural disengagement	.026	.021	
Brief COPE self-blame	-.099	-.082	
STAI-6 anxiety	-.076	-.057	
CES-D 10 depression	**-.302****	**-.226**	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.095**	**.089**	**.095****
Step 2	**.177**	**.160**	**.082****
Step 3	.201	.179	.024
Step 4	**.491**	**.466**	**.290****
Step 5	**.544**	**.510**	**.053****
Step 6	**.609**	**.577**	**.065****

Note. Gender: 0= male, 1= female, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

8.2.2.2. Social functioning

The final model for the social functioning subscale is displayed in Table 8.8. The final model was significant, *f*(22,280)= 12.67; *p*< .001 and explained 45.9% of the total variance in the social functioning sub-domain. The unique predictors in this final model were illness identity, consequences, and presence of depressive symptoms. Perceptions of more severe symptoms and consequences due to ACHD

and the presence of depressive symptoms were associated with poorer social functioning.

Table 8.8. Final regression model for social functioning (*n*= 303)

Predictor Variables	**β**	***sr*²**	
Employment status	.055	.051	
ToF	.116	.091	
TGA	.022	.017	
SV	.002	.001	
Co-morbidities no.	.015	.011	
Hospitalization days	-.030	-.025	
Current O_2 saturation	.048	.037	
Medication no.	.070	.052	
Brief IPQ consequences	**-.224***	**-.134**	
Brief IPQ controllability	-.012	-.011	
Brief IPQ identity	**-.242****	**-.159**	
Brief IPQ concern	-.049	-.034	
Brief IPQ emotional representation	.006	.003	
Brief COPE self-distraction	-.030	-.026	
Brief COPE denial	-.030	-.027	
Brief COPE venting	.017	.013	
Brief COPE substance use	-.014	-.013	
Brief COPE behavioural disengagement	.012	.009	
Brief COPE self-blame	.019	.016	
MSPSS friends support	.034	.031	
STAI-6 anxiety	-.119	-.093	
CES-D 10 depression	**-.249****	**-.187**	
Step	***R*²**	***Adjusted R*²**	***ΔR*²**
Step 1	**.065**	**.062**	**.065****
Step 2	**.138**	**.124**	**.073****
Step 3	.142	.124	.004
Step 4	.162	.139	.020
Step 5	**.430**	**.405**	**.269****
Step 6	.439	.401	.009
Step 7	.442	.403	.004
Step 8	**.499**	**.459**	**.056****

Note. Employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.2.3. Role emotional

The final model for the role emotional subscale is displayed in Table 8.9. The final model was significant, $f(16,285)= 12.88$; $p< .001$ and explained 38.7% of the total variance in the role emotional sub-domain. The unique predictors in the final model

were self-blame and presence of depressive symptoms. Greater use of self-blame as a coping strategy and the presence of depressive symptoms were associated with greater role limitations due to emotional problems.

Table 8.9. Final regression model for role emotional (*n*= 302)

Predictor Variables	**β**	***sr*²**	
Employment status	.087	.081	
Brief IPQ consequences	-.085	-.051	
Brief IPQ controllability	.004	.003	
Brief IPQ identity	-.014	-.010	
Brief IPQ concern	.007	.005	
Brief IPQ emotional representation	-.011	-.006	
Brief COPE positive reframing	.033	.029	
Brief COPE acceptance	.030	.027	
Brief COPE self-distraction	-.052	-.045	
Brief COPE venting	-.021	-.017	
Brief COPE substance use	-.013	-.012	
Brief COPE behavioural disengagement	.025	.020	
Brief COPE self-blame	**-.146***	**-.123**	
MSPSS friends support	.080	.075	
STAI-6 anxiety	-.098	-.076	
CES-D 10 depression	**-.384****	**-.291**	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.063**	**.060**	**.063****
Step 2	**.218**	**.202**	**.155****
Step 3	**.301**	**.270**	**.084****
Step 4	.312	.278	.010
Step 5	**.420**	**.387**	**.108****

Note. Employment status: 0= unemployed, 1= employed, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.2.4. Mental health

The final model for the mental health subscale is displayed in Table 8.10. The final model was significant, $f(20,282)= 20.04$; $p< .001$ and explained 55.8% of the total variance in the mental health sub-domain. The unique predictors in this final model were age, self-blame, anxiety, and presence of depressive symptoms. Younger age, greater use of self-blame as a coping strategy, greater anxiety, and presence of depressive symptoms were associated with poorer mental health.

Table 8.10. Final regression model for mental health (*n*= 303)

Predictor Variables	β	sr^2	
Age	**.186****	**.172**	
Employment status	.082	.076	
Brief IPQ consequences	-.019	-.011	
Brief IPQ controllability	.020	.018	
Brief IPQ identity	-.050	-.033	
Brief IPQ concern	-.076	-.054	
Brief IPQ emotional representation	-.070	-.041	
Brief COPE positive reframing	.047	.040	
Brief COPE acceptance	.024	.021	
Brief COPE self-distraction	.001	.001	
Brief COPE denial	-.075	-.067	
Brief COPE venting	-.015	-.012	
Brief COPE substance use	.021	.019	
Brief COPE behavioural disengagement	-.004	-.004	
Brief COPE self-blame	**-.156***	**-.129**	
MSPSS friends support	.039	.033	
MSPSS family support	-.018	-.014	
MSPSS significant other support	-.019	-.016	
STAI-6 anxiety	**-.192****	**-.148**	
CES-D 10 depression	**-.356****	**-.270**	
Step	R^2	*Adjusted* R^2	ΔR^2
Step 1	**.096**	**.090**	**.096****
Step 2	**.351**	**.336**	**.255****
Step 3	**.463**	**.435**	**.112****
Step 4	.467	.433	.004
Step 5	**.587**	**.558**	**.120****

Note. Employment status: 0= unemployed, 1= employed, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.3. Factors associated with disease-specific HRQoL (CHD-TAAQOL)

8.2.3.1. Symptoms

The final model for symptoms is displayed in Table 8.11. Demographic variables were entered in Step 1, explaining 9.4% of the variance in symptoms. Addition of diagnostic group, cyanosis days, number of co-morbidities, and arrhythmias in Step 2 explained an additional 11.3% of the variance in the outcome. Hospitalization days, entered in Step 3, did not explain additional variance. Current O_2 saturation, number of medication, VO_2 max, and LVEF were entered in Step 4 and explained a

further 7.1% of the variance. Illness perceptions were entered in Step 5 and explained an additional 30.3% of the variance. Coping strategies, entered in Step 6, did not explain additional variance. Mood, entered in Step 7, explained an additional 4.1% of the variance in symptoms. The final model was significant, $f(26,275)= 18.29$; $p< .001$ and explained 59.9% of the total variance in symptoms. The unique predictors in the final model were illness identity and presence of depressive symptoms. A perception of more severe symptoms due to ACHD and the presence of depressive symptoms were associated with greater symptom impact.

Table 8.11. Final regression model for symptoms (*n*= 302)

Predictor Variables	**β**	***sr*²**	
Gender	-.051	-.043	
Employment status	.075	.067	
ToF	-.010	-.007	
TGA	-.022	-.016	
SV	-.012	-.007	
Cyanosis days	.042	.030	
Co-morbidities no.	-.047	-.032	
Arrhythmias	.031	.023	
Hospitalization days	.042	.035	
Current O_2 saturation	.096	.062	
Medication no.	-.007	-.005	
VO_2 max	.078	.066	
LVEF	.038	.033	
Brief IPQ consequences	-.060	-.035	
Brief IPQ controllability	-.018	-.017	
Brief IPQ identity	**-.431****	**-.279**	
Brief IPQ concern	-.107	-.074	
Brief IPQ emotional representation	.001	.001	
Brief COPE self-distraction	-.015	-.013	
Brief COPE denial	-.039	-.035	
Brief COPE venting	.038	.030	
Brief COPE substance use	-.008	-.007	
Brief COPE behavioural disengagement	.013	.010	
Brief COPE self-blame	-.029	-.024	
STAI-6 anxiety	-.097	-.075	
CES-D 10 depression	**-.215****	**-.160**	
Step	***R*²**	***Adjusted R*²**	***ΔR*²**
Step 1	**.094**	**.088**	**.094****
Step 2	**.207**	**.185**	**.113****
Step 3	.207	.182	.000
Step 4	**.278**	**.246**	**.071****
Step 5	**.581**	**.555**	**.303****
Step 6	.593	.558	.011
Step 7	**.634**	**.599**	**.041****

Note. Gender: 0= male, 1= female, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
**p*< .01, ***p*< .001

8.2.3.2. Impact of cardiac surveillance

The final model for impact of cardiac surveillance is displayed in Table 8.12. Diagnostic group, number of co-morbidities, and arrhythmias were entered in Step 1, explaining 12.6% of the variance in impact of cardiac surveillance. Addition of number of interventions in Step 2 and current O_2 saturation, number of medication,

and VO_2 max in Step 3 did not explain any additional variance. Illness perceptions were entered in Step 4 and explained further 16.8% of the variance in the outcome. Coping strategies, entered in Step 5, did not explain additional variance. Mood, entered in Step 6, did not explain additional variance. The final model was significant, $f(19,283)= 8.57$; $p< .001$ and explained 32.3% of the total variance in impact of cardiac surveillance. The unique predictors in the final model were planning and anxiety. Greater use of planning as a coping strategy and greater anxiety were associated with greater impact of cardiac surveillance.

Table 8.12. Final regression model for impact cardiac surveillance (*n*= 303)

Predictor Variables	**β**	***sr*²**	
ToF	.016	.012	
TGA	-.079	-.057	
SV	-.034	-.021	
Co-morbidities no.	.014	.010	
Arrhythmias	-.141	-.105	
Intervention no.	-.070	-.056	
Current O_2 saturation	.037	.028	
Medication no.	-.029	-.021	
VO_2 max	.011	.010	
Brief IPQ consequences	-.011	-.006	
Brief IPQ identity	-.142	-.093	
Brief IPQ concern	-.135	-.093	
Brief IPQ emotional representation	-.132	-.077	
Brief COPE planning	**-.138***	**-.130**	
Brief COPE venting	.019	.016	
Brief COPE substance use	.003	.002	
Brief COPE behavioural disengagement	-.088	-.074	
STAI-6 anxiety	**-.159***	**-.127**	
CES-D 10 depression	-.005	-.004	
Step	***R*²**	***Adjusted R*²**	**Δ*R*²**
Step 1	**.126**	**.111**	**.126****
Step 2	.129	.112	.003
Step 3	.154	.128	.024
Step 4	**.322**	**.292**	**.168****
Step 5	.347	.309	.025
Step 6	.365	.323	.018

Note. ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*$p< .01$, **$p< .001$

8.2.3.3. Worries

The final model for worries is displayed in Table 8.13. Only age was entered in Step 1, explaining 2.3% of the variance in worries. The addition of the diagnostic group and number of co-morbidities in Step 2 explained an additional 8.1% of the variance. Current O_2 saturation, entered in in Step 3, did not explain additional variance in the outcome. Illness perceptions were entered in Step 4 and explained an additional 32.1% of the variance. Coping strategies, entered in Step 5, explained a further 4.3% of the variance in worries. Social support, entered in Step 6, did not explain additional variance. Mood was entered in Step 7 and significantly explained an additional 3.7% of the variance. The final model was significant, $f(22,280) = 13.97$; $p < .001$ and explained 48.6% of the total variance in worries. The unique predictors in this final step were age, illness consequences, and presence of depressive symptoms. Younger age, a perception of more consequences due to ACHD, and the presence of depressive symptoms were associated with greater worries.

Table 8.13. Final regression model for worries (*n*= 303)

Predictor Variables	β	sr^2	
Age	**.218****	**.183**	
ToF	.061	.048	
TGA	.048	.036	
SV	.060	.038	
Co-morbidities no.	-.014	-.012	
Current O_2 saturation	.049	.037	
Brief IPQ consequences	**-.179***	**-.109**	
Brief IPQ controllability	-.023	-.021	
Brief IPQ curability	.067	.060	
Brief IPQ identity	-.137	-.089	
Brief IPQ concern	-.146	-.103	
Brief IPQ emotional representation	-.106	-.061	
Brief COPE planning	-.117	-.103	
Brief COPE self-distraction	-.024	-.020	
Brief COPE venting	.090	.071	
Brief COPE substance use	-.066	-.060	
Brief COPE behavioural disengagement	.084	.069	
Self-blame	-.126	-.103	
MSPSS friend support	.047	.040	
MSPSS family support	.021	.018	
STAI-6 anxiety	-.061	-.047	
CES-D 10 depression	**-.224****	**-.168**	
Step	***R^2***	***Adjusted R^2***	***ΔR^2***
Step 1	**.023**	**.020**	**.023***
Step 2	**.104**	**.089**	**.081****
Step 3	.116	.098	.012
Step 4	**.437**	**.414**	**.321****
Step 5	**.480**	**.447**	**.043***
Step 6	.487	.450	.006
Step 7	**.523**	**.486**	**.037****

Note. ToF, TGA, SV: dummy-coded- reference group was Simple, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
*p< .01, **p< .001

8.2.4. Summary of factors associated with HRQoL from hierarchical multiple regressions

Table 8.14 presents a summary of the factors associated with generic (physical & psychosocial) and disease-specific HRQoL in the final models.

Table 8.14. Summary of the factors associated with generic (physical & psychosocial) and disease-specific HRQoL from hierarchical multiple regressions

	Generic Physical HRQoL (Physical Component Summary and Subscales)					*Generic Psychosocial HRQoL (Mental Component Summary and Subscales)*					*Disease-specific HRQoL*		
	Physical component summary	***Physical functioning***	***Role physical***	***Bodily pain***	***General health***	***Mental component summary***	***Vitality***	***Social functioning***	***Role emotional***	***Mental health***	***Symptoms***	***Impact of cardiac surveillance***	***Worries***
Employment status			▲										
Age						▲				▲			▲
TOF							▲						
TGA							▲						
SV							▲						
Brief IPQ consequences		▼						▼					▼
Brief IPQ identity	▼	▼	▼	▼	▼		▼	▼			▼		
Brief IPQ emotional representation		▲											
Brief COPE self-blame						▼			▼	▼			
Brief COPE planning												▼	
STAI anxiety						▼				▼		▼	
CES-D 10 depression			▼		▼	▼	▼	▼	▼	▼	▼		▼

▲Significant positive relationship, ▼Significant negative relationship

Note. Employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms. HRQoL scale 0 – 100, higher scores= better HRQoL.

8.3. Examining the assumptions of hierarchical multiple regression analyses

Across all regression models VIF values were below 4 and tolerance values were greater than 0.20 indicating that there was no multicollinearity issues in the hierarchical multiple regressions. Multivariate outliers can be found in Appendix T. Cook's distance indicated that these multivariate outliers did not influence the results of the hierarchical multiple regressions and were therefore retained in the analyses. The Durbin-Watson values for all regressions were within the acceptable limit of 2, indicating that the assumption of independence of errors was met. Examination of the plots indicated that the assumption of normality of residuals was met for most regressions. Some deviation from normality was observed in role emotional. The assumptions of linearity and homoscedasticity were met for most of the analyses. There appeared to be an unequal spread of values above or below the zero line for physical functioning, role physical, bodily pain, social functioning, and role emotional sub-domains of the SF-36v1®. Although these patterns were observed in the data, they were not sufficiently pronounced as to be a cause for concern. This may weaken the analyses, but it does not invalidate them (Tabachnick & Fidell, 2007).

8.4. Discussion

This chapter presented the analyses conducted to investigate the demographic, clinical, and psychosocial factors associated with both generic and disease-specific HRQoL in ACHD. It was hypothesized that psychosocial factors, including illness perceptions, coping strategies, mood, and social support would account for a significant amount of variance in HRQoL, over and above that accounted for by demographic and clinical characteristics. Some, but not all, of these psychosocial

factors explained more variance and were significantly associated with HRQoL and over and above demographic and clinical factors as predicted.

8.4.1. Factors associated with generic physical HRQoL

8.4.1.1. Demographic factors

Demographic factors explained only a small proportion of the variance in physical HRQoL, including the physical component summary and the subscales of physical functioning, role physical, and general health (6-9%). Of note, employment status was found to predict the subscale of role limitations due to physical problems in the final model but not the physical component summary score which encompasses this subscale. This finding is in agreement with a study of 676 people with ACHD by Vigl et al. (2011) who did not find an association between unemployment and the physical component summary. Employment status was a consistent predictor of the physical component summary and the subscales of physical functioning and general health; however, this association was supplanted when clinical variables were entered into the models. This suggests that this relationship may be related to disease complexity as unemployment has been previously found to be associated with more complex ACHD (Kamphuis et al., 2002c).

Employment status was, however, associated with the subscale of role physical in the final model, suggesting that unemployed individuals experienced greater role limitations due to physical problems compared with employed individuals. It is likely that people with ACHD are unemployed because they experience more physical problems and greater role limitations which prevent them from gaining and maintaining employment. However, the cross-sectional nature of this study limits the ability to establish the direction of this relationship.

8.4.1.2. Clinical factors

Diagnostic group, cyanosis days, and co-morbidities number together and current O_2 saturation, exercise capacity, medication number, and ventricular function together explained some of the variance (3-17% and 3-6% respectively) in the physical component summary and all of the subscales. However, clinical factors were not found to predict physical HRQoL and were supplanted by illness perceptions. This finding is in agreement with a longitudinal study of 845 people with ACHD by Schoormans et al. (2014), who found that disease complexity was not associated with the physical component summary of the SF-36 in a model that included illness perceptions. It appears that the effects of clinical factors are encompassed in the relationship between illness perceptions and physical HRQoL. Psychosocial factors, including illness perceptions explained more of the variance in physical HRQoL and were significant unique predictors of physical HRQoL over and above clinical factors, which is in agreement with previous studies in ACHD (Chen et al., 2011; Pike et al., 2012; Riley et al., 2012; Schoormans et al., 2014).

8.4.1.3. Psychosocial factors

After accounting for demographic and clinical characteristics, illness perceptions consistently explained the largest proportion of variance (19-28%) in the physical component summary and all of the subscales. Negative illness perceptions, specifically perceiving more severe symptoms related to ACHD was associated with poorer physical HRQoL in the physical component summary and all physical subscales of HRQoL, including poorer physical functioning, greater role limitations due to physical problems, greater bodily pain, and poorer general health. Two previous studies in ACHD used the IPQ-R as opposed to the Brief IPQ used in the

present study but did not examine the relationship between illness identity and physical HRQoL (Riley et al., 2012; Schoormans et al., 2014). The findings support previous studies in other chronic conditions, in both a meta-analysis and systematic review (Hagger & Orbell, 2003; Petrie et al., 2007), including studies in coronary artery disease (Stafford et al., 2009) and myocardial infarction (Weinman et al., 1996). Hagger and Orbell (2003) found strong relationships between illness identity and physical and role functioning over and above clinical factors across a range of long-term conditions. The results of the current study also corroborate those of Scharloo et al. (1998), who found that the largest proportion of variance in SF-36 outcomes was explained by illness identity in people with rheumatoid arthritis, chronic obstructive pulmonary disease, and psoriasis. Taken together, these findings suggest the importance of subjective perceptions of symptoms over and above clinical factors for physical HRQoL across chronic conditions, including people with ACHD. As mentioned earlier, illness perceptions are likely to mediate the relationship between clinical factors and physical HRQoL as their effect became insignificant after the inclusion of illness perceptions in the models.

Other illness perceptions measured in the Brief IPQ, including illness coherence, controllability, and curability, were not found to be associated with the physical component summary confirming the findings of Riley et al. (2012) and Schoormans et al. (2014). Schoormans et al. (2014) found that illness controllability and coherence were not predictive of the physical component summary in a longitudinal study of 845 people with ACHD. This is likely attributed to the congenital nature of ACHD as recent data in coronary heart disease suggests that illness coherence and controllability are associated with HRQoL but only in people who experience an

unexpected medical event rather than outpatients (Foxwell, Morley, & Frizelle, 2013). The authors argued that an unexpected rather than a familiar situation would require people to have an increased illness understanding and sense of control in order to successfully adapt.

This study did not corroborate some the findings of Schoormans et al. (2014) who reported that perceiving the illness to have more serious consequences and less emotional consequences at baseline were predictive of poorer physical HRQoL in the physical component summary in the two-year follow-up. This is possibly due to the inclusion of the illness identity subscale in the present study, which shared some variance with these illness perceptions and was the strongest predictor of physical HRQoL. Illness consequences and emotional representation were, however, associated with the subscale of physical functioning, which is encompassed in the physical component summary. Firstly, perceiving the illness to have more serious consequences was associated with poorer physical functioning, a finding that is consistent with previous studies in coronary heart disease (e.g. Stafford et al., 2009) and myocardial infarction (Alsén, Brink, Persson, Brändström, & Karlson, 2010; French et al., 2005). Secondly, surprisingly a perception of fewer emotional consequences as a result of ACHD was associated with poorer instead of better physical functioning. Results from bivariate analyses showed an inverse rather than a positive relationship between the two variables. This association was limited to only physical functioning and no other domains. For these reasons it was assumed that this may be a spurious finding possibly the result of suppression effects, whereby the change in the nature of the association is due to the inclusion of other variables controlled for in the model (Tu, Gunnell, & Gilthorpe, 2008). More research will

likely help determine the nature of the relationship between illness emotional representation and physical HRQoL.

In addition to illness perceptions, anxiety and the presence of depressive symptoms also explained a small proportion of the variance in some domains of physical HRQoL, including role limitations due to physical problems (5%) and general health (2%) but not the physical component summary.

Contrasting the present findings regarding the physical component summary, a smaller study of 99 people with ACHD found that higher depression levels (measured with the HADS) were associated with poorer HRQoL in the physical component summary (Riley et al., 2012). Other studies in ACHD that utilised different measures of HRQoL (WHOQOL-Bref) and depression also found a significant association between depression and overall physical HRQoL (Chen et al., 2011; Rose et al., 2005). These conflicting findings may be a result of methodological differences but also reflect the findings observed in the wider cardiac literature, where depression has been more consistently associated with psychosocial as opposed to physical HRQoL (Stafford et al., 2007) (see section 8.4.2.3, page 260).

The findings in relation to the subscales, however, indicated that people with depressive symptoms reported more role limitations due to physical problems and poorer general health compared with those with no depressive symptoms, two subscales that are encompassed in the physical component summary. It may be that depressive symptoms have a greater role in relation to certain domains of physical HRQoL than other, which may be masked when solely examining aggregated scales, also evidenced by research in pulmonary hypertension and diabetes (Ali et al., 2010; Vanhoof et al., 2014). A possible explanation for these findings may be that

depression "amplifies" the effects of a medical condition like ACHD on physical HRQoL (Gaynes et al., 2002). Nearly a quarter of participants in this study had depressive symptoms and all participants irrespective of diagnostic group reported diminished general health (see section 7.4.1.1, page 216). These findings further highlight the importance of assessing depressive symptoms in people with ACHD as these symptoms can pose additional physical burden even on those that are clinically stable (Stafford et al., 2007).

8.4.2. Factors associated with generic psychosocial HRQoL

8.4.2.1. Demographic factors

Demographic factors explained the same small proportion of variance in psychosocial HRQoL, including the mental component summary and the subscales of vitality, social functioning, role emotional, and mental health (6-9%), as they did in physical HRQoL. Age was found to be associated with the mental component summary and the subscale of mental health in the final models. In particular, younger individuals experienced poorer overall psychosocial HRQoL and poorer mental health compared with older ones. Previous research has reported variable findings regarding the association between age and psychosocial HRQoL in ACHD. The findings of the present study are in agreement with the longitudinal study by Schoormans et al. (2014), where older age was a significant unique predictor of better scores on the mental component summary of the SF-36 at the two-year follow-up. Silva et al. (2011) also found that younger individuals reported poorer psychological HRQoL (as measured by the WHOQOL-Bref) than older individuals with ACHD. However, other studies in ACHD have not observed a relationship between people's age and their psychosocial HRQoL (Hickey et al., 2012; Kahya

Eren et al., 2013; Loup et al., 2009; van Rijen et al. 2005a) possibly due to the exclusion of people with SV. This group was younger than the other diagnostic groups included in the present study (see section 6.5, page 175) and were found to experience diminished psychosocial HRQoL (see section 7.4.1.2, page 220). A possible explanation for the association between age and psychosocial HRQoL is that people adapt to living with ACHD and cope better as they age (Bruto et al., 2007). The findings may have implications for the assessment of people's psychological needs throughout their lives, especially with the growing number of individuals with SV surviving well into adulthood resulting in an older population (DoH, 2006).

8.4.2.2. Clinical factors

Clinical factors were not part of the multivariate analysis for the mental component summary and the subscales of role emotional and mental health as they were not associated with these outcomes in the bivariate analyses. Diagnostic group and co-morbidities number did, however, explain a small proportion of the variance (7-8%) in the subscales of vitality and social functioning. Overall, clinical factors were found to be poor predictors of psychosocial HRQoL, in a model that included psychosocial factors as these explained more variance in psychosocial HRQoL. This finding is consistent with previous studies in ACHD (Chen et al., 2011; Riley et al., 2012; Rose et al., 2005).

Diagnostic group was only found to be associated with vitality in the final model. Being in the ToF, TGA, and SV groups as opposed to the Simple (reference group) was associated with greater vitality. This finding, in part, gives further support to the differences between the diagnostic groups found in earlier analyses (section 7.4.1.1,

page 216), whereby people in the Simple group had significantly lower vitality levels than other groups and especially people with ToF. Ternestedt et al. (2001) also reported greater vitality in people with ToF compared with ASD when the converse may be expected. In Chapter 7, it was speculated that people in different diagnostic groups may have differing expectations about their vitality. People in the Simple group may expect their functioning to be similar to healthy individuals, while the other diagnostic groups may have adjusted their expectations according to their capacity. This is an area worthy of investigation in future studies.

8.4.2.3. Psychosocial factors

After accounting for demographic and clinical characteristics, illness perceptions consistently explained the largest proportion of variance (16-29%) in psychosocial HRQoL, including the mental component summary and all of the subscales. However, illness perceptions were not found to be associated with the mental component summary in the final model. Schoormans et al. (2014) found that the mental component summary was only predicted by negative emotional representation. In the present study, emotional representation was associated with the mental component summary before being superseded by mood in the final model. This can be attributed to the similarity between constructs or the effects of emotional representation may have been encompassed in the relationship between mood and psychosocial HRQoL. Riley et al. (2012) also failed to find an association between illness perceptions and the mental component summary in a model that included anxiety and depression.

The findings in relation to the subscales, however, indicated that perceiving more severe symptoms due to ACHD was associated with less vitality and poorer social

functioning. A perception of more severe consequences as a result of ACHD was also associated with poorer social functioning. These findings support the earlier findings in relation to physical HRQoL and further suggest that negative perceptions about their symptoms and the consequences of ACHD can potentially be detrimental for only certain domains of people's psychosocial HRQoL. These findings are also in agreement with a meta-analysis of studies across long-term conditions, which found that illness identity and consequences were negatively associated with social functioning and vitality (Hagger & Orbell, 2003). Such negative perceptions may be accurate in people with SV, who reported poorer social functioning compared with the general population (section 7.4.1.2, page 220). However, in people in the Simple group, where there was variation in vitality (section 6.7.1, page 185), negative perceptions about symptoms may not represent their actual clinical state, leading to misconceptions about the condition and its impact (Schoormans et al., 2014).

In addition to illness perceptions, coping explained 5-12% of the variance in psychosocial HRQoL, including the mental component summary and the subscales of vitality, role limitations due to emotional problems, and mental health. Only the use of self-blame was associated with poorer psychosocial HRQoL in the mental component summary as well as greater role limitations due to emotional problems and poorer mental health in the final models. This finding supports previous studies in people with heart failure (Graven et al., 2014; Klein et al., 2007), cancer (Shapiro, McCue, Heyman, Dey, & Haller, 2010), and across other chronic conditions (Bombardier, D'Amico, & Jordan, 1990). Bombardier et al. (1990) found that the use of self-blame was associated with poorer psychosocial HRQoL after controlling for disease complexity in a sample of 101 individuals with various chronic conditions. It

is believed that long-term use of maladaptive coping strategies, such as self-blame, can be problematic as they aggravate negative emotions and distract people from actively improving their situation (Carver, 1989). This has important implications for people with ACHD. Because of the congenital nature of their illness, these individuals have more time to accept the reality of their condition and take steps towards improving and sustaining their HRQoL. However, such efforts can be impaired when they blame themselves or are being overly critical of themselves (Klein et al., 2007). It may therefore be beneficial to identify and address maladaptive self-blame in people with ACHD in order to help them cope successfully.

Conversely, the lack of a relationship between a majority of the coping strategies and particularly adaptive coping (planning, active coping, emotional support, and instrumental support) and psychosocial HRQoL in both bivariate and multivariate analyses is in line with previous studies in heart failure (Klein et al., 2007), end-stage renal disease and congestive heart failure (Kristofferzon, Lindqvist, & Nilsson, 2011), and asthma and chronic obstructive pulmonary disease (Hesselink et al., 2004). This appears to be a common occurrence in the chronic illnesses literature and has been attributed to their incurable nature, as the adoption of emotion-focused coping may be more effective in ameliorating the emotional impact and therefore maintaining psychosocial HRQoL (Petrie & Reynolds, 2007). However, people with ACHD in present study reported using predominantly adaptive coping strategies, including acceptance and active coping (section 6.7.2, page 188). It is also possible that the lack of an association between these coping strategies and psychosocial HRQoL is due to the operationalization of coping in this study. Previous studies in

other populations that have shared similar findings have concluded that generic coping checklists may be problematic if coping is successful or when people use habitual coping behaviours which they may not be able to identify (Steed, Newman, & Hardman, 1999). Some authors have argued in favour of disease-specific coping measures (Hagger & Orbell, 2003), which are not currently available in ACHD. More research on this topic needs to be undertaken before the association between coping and psychosocial HRQoL is more clearly understood.

Mood, including anxiety and depressive symptoms also explained a proportion of the variance (6-13%) in the mental component summary and all of the subscales. The presence of depressive symptoms was associated with poorer HRQoL in the mental component summary and all of the subscales, including lower vitality, poorer social functioning, more role limitations due to emotional problems, and poorer mental health in the final models. Higher anxiety levels were only associated with poorer HRQoL in the mental component summary and poorer mental health. These findings confirm previous studies in ACHD (Chen et al., 2011; Müller et al., 2012; Müller et al., 2013a; Riley et al., 2012). Chen et al. (2011) found that after accounting for demographic and clinical variables, psychological distress (anxiety, depression) was the strongest predictor of both psychological and social HRQoL as measured by the WHOQOL-Bref. These findings have also been replicated across various ACHD samples including people with SV (Pike et al., 2012) and people with ASD (Cohen et al., 2010). The present study extends these findings by demonstrating that mood can potentially influence all aspects of psychosocial HRQoL including vitality, social functioning, role limitations, and mental health. The present and previous studies in ACHD are also in agreement with the wider cardiac literature, in which

mood disorders are considered amongst the strongest predictors of poor psychosocial HRQoL (Ruo et al., 2003). These findings coupled with the considerable incidence of depressive symptoms in 22% of the sample in this study, may help explain the impairment in psychosocial HRQoL observed across the total sample in Chapter 7 (section 7.4.1.2, page 220). It may also help explain the impaired psychosocial HRQoL in the SV and Simple groups, which had the highest proportion of people with depressive symptomatology (27% and 28% respectively).

A possible explanation for this relationship is that depressive symptomatology poses an additional burden on people with ACHD, making them more vulnerable to experiencing poorer psychosocial HRQoL and limitations in their social activities and their social role, while also encouraging a more negative perception in these outcomes (Stafford et al., 2007). Alternatively, for some subscales it could be an issue with definition. According to the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV), depressive symptoms include reduced energy levels and decreased interest in activities (American Psychiatric Association, 2000). Assuming the CESD-10's validity as a measure of symptoms relating to clinical depression, it may reflect the vitality, impaired social functioning, and mental health aspects of HRQoL, posing possible overlap between the concepts. Therefore, it would not be surprising that the presence of depressive symptomatology was associated with poorer HRQoL in these subscales. However, the measure did not tackle these concepts directly and depressive symptomatology was also associated with certain aspects of physical HRQoL. Overall, these findings highlight the importance of measuring and addressing anxiety and depressive symptoms in people with ACHD, as these can potentially be detrimental for their HRQoL.

It is important to note that social support did not explain significant variance and was not associated with either the mental component summary or any of the subscales in the multivariate analyses. This finding suggests that despite the high social support levels experienced by people in the present study (section 6.7.3, page 190), other factors are more important in explaining variation in psychosocial HRQoL. In contrast to these findings, Pike et al. (2012) found that social support was a significant unique predictor of overall HRQoL rather than the mental component summary and its subscales in a smaller sample of people with SV, after accounting for the effects of depression and functional status. Rose et al. (2005) also found that social support was a significant determinant of psychological and social HRQoL as measured by the WHOQOL-Bref in people with ACHD. Both these studies did not account for other confounding variables including illness perceptions, which were found to be amongst the strongest predictors of psychosocial HRQoL in the present study.

There are several possible explanations for the lack of a relationship between social support and psychosocial HRQoL. Firstly, this lack of relationship may be partially explained by the lack of variability in social support. It may also be that social support had an indirect impact on HRQoL through other factors included in the multivariate analyses. Recent studies in other populations found that depression may serve as a mediator between social support and psychosocial HRQoL (Wicke et al., 2014). Evidence also suggests that in addition to a direct effect on HRQoL, social support may have an indirect effect by serving as a "buffer" to reduce the emotional impact associated with an illness (Cohen & Wills, 1985). While testing the abovementioned assumptions was beyond the scope of the present study, their

examination in future studies may provide important insight into the relationship between social support and HRQoL in ACHD.

8.4.3. Factors associated with disease-specific HRQoL

8.4.3.1. Demographic factors

Demographic factors explained a small proportion of the variance in the symptoms (9%) and worries (2%) scales of the CHD-TAAQOL. Age remained a significant unique predictor of the worries subscale in the final model. Younger people reported greater ACHD-specific worries as opposed to older individuals. This finding is consistent with the findings in generic psychosocial HRQoL, as discussed in section 8.4.2.1 (page 258). Schoormans et al. (2014) also found that younger age was a significant predictor of greater worries (also measured using the CHD-TAAQOL) at the two year follow-up. Inspection of the items within the worries subscale provides further insight into this finding. It may be expected that as people with ACHD age they worry less about educational attainment, employment opportunities, and being able to have children, all of which may be important issues for younger people. The finding may therefore be an artefact of the type of worries specified within the scale rather than worries in general. A broader measure or inclusion of older age specific items may have made the measure more relevant to this population. This finding does however have important implications for the provision of information for younger adults, especially during key periods such as their transition from child to adult care (Sable et al., 2011).

8.4.3.2. Clinical factors

Diagnostic group, co-morbidities, cyanosis days, and presence of arrhythmias together explained a proportion of variance in all subscales of the disease-specific

HRQoL including symptoms, impact of cardiac surveillance, and worries (8-13%). Current O_2 saturation, medication number, exercise capacity, and ventricular function together also explained significant variance but only in symptoms (7%). However, none of the clinical variables were associated with disease-specific HRQoL in the final models and became insignificant after the inclusion of illness perceptions. This is consistent with the findings for physical and psychosocial HRQoL, whereby clinical variables were found to be poor unique predictors of HRQoL when psychosocial factors are accounted for (see section 8.4.1.2, page 254 and section 8.4.2.2, page 259) and may signify that illness perceptions mediate the relationship between clinical factors and disease-specific HRQoL.

8.4.3.3. Psychosocial factors

After adjusting for demographic and clinical characteristics, illness perceptions explained a substantial amount of variance (17-32%) in all three subscales of the CHD-TAAQOL. Perceiving ACHD to have more serious consequences was associated with greater worries, a finding that is in agreement with the longitudinal study by Schoormans et al. (2014). Furthermore, a perception of more severe symptoms due to ACHD was found to be associated with greater symptom impact. It may be argued that this relationship is an artefact of overlapping definitions, yet their distinction is subtle but important. The symptoms subscale of the CHD-TAAQOL measures the frequency of symptoms, but weighted by their emotional impact on the person. Whilst the illness identity subscale of the Brief IPQ quantifies the degree to which people experience symptoms attributable to ACHD and is a cognitive rather than emotional evaluation. Although it may not be possible to draw clear conclusions regarding this particular association, the strong relationships between illness identity

and all other physical and psychosocial HRQoL domains discussed in sections 8.4.1.3 (page 254) and 8.4.2.3 (page 260), provide further support for its importance in both generic and disease-specific HRQoL.

Coping explained 4% of the variance but only in the worries scale. The findings indicated that greater use of planning as a coping strategy was found to be associated with greater impact of cardiac surveillance in the final model. Planning is a problem-focused coping strategy, which involves the formation of plans towards dealing with a problem and it is thought to be one of the most important coping strategies in chronic illness that facilitates adjustment (Felton et al., 1984). Owing to the cross-sectional design of the study it is difficult to draw conclusions on causality for this particular association. It may be expected that people who reported greater impact of cardiac surveillance would need to engage in more planning in order to accommodate cardiac testing and hospital appointments.

Anxiety and depressive symptoms also explained some variance (4%) in symptoms and worries. The findings indicated that individuals with depressive symptoms reported greater symptom impact and greater worries than those with no depressive symptoms, whilst higher levels of anxiety were associated with greater impact of cardiac surveillance. The findings are consistent with those in relation to generic HRQoL, further supporting the wider notion that anxiety and depression can potentially be detrimental for HRQoL in cardiac populations (Ruo et al., 2003). Ruo et al. (2003) also found that depressive symptoms were associated with greater symptom burden in people with coronary artery disease and this has been attributed to the additional functional limitations people with depression suffer from (Stafford et al., 2007). Individuals with depressive symptoms in the present study were also

found to experience greater worries in relation to their education, employment, their ability to have children, and independent living. The finding that higher levels of anxiety were associated with greater impact of cardiac surveillance was not surprising; the impact of cardiac surveillance subscale assesses the frequency of cardiac surveillance but weighted by its emotional impact on the person, hence it is not independent of mood. In addition, anxiety was measured in its state form, where participants were asked about their feelings at the particular time of the assessment and these assessments generally took place in the hospital. It is likely that the hospital environment itself might have been a confounding factor as it may be connected to negative experiences, such as previous hospitalizations and may also be a reminder that their health is not ideal. However, it is difficult to determine the direction of the relationship between anxiety and the impact of cardiac surveillance based on cross-sectional data. It is also likely that greater impact of cardiac surveillance might have resulted in higher levels of anxiety in patients.

8.5. Summary

This chapter presented the results of hierarchical multiple regressions conducted to examine the factors associated with HRQoL in people with ACHD. Overall, the findings indicated that psychosocial factors and specifically illness perceptions significantly explained a large proportion of variance in HRQoL after adjusting for demographic, clinical, and other psychosocial factors. Coping and mood significantly explained a smaller proportion of variance in HRQoL but predominantly in psychosocial domains. Psychosocial factors, including illness perceptions and specifically illness identity and consequences, the use of self-blame as a coping strategy, and mood were found to be strong unique predictors of

HRQoL. The findings provided further support to the wealth of literature in other chronic populations, which highlighted the importance of psychosocial factors for understanding the factors associated with people's HRQoL. The limitations of the cross-sectional study and the theoretical, clinical, and research implications of the findings will be addressed in the general discussion in Chapter 11.

CHAPTER 9 – LONGITUDINAL CHANGES IN HEALTH-RELATED QUALITY OF LIFE AND PSYCHOSOCIAL FUNCTIONING

9.1. Prologue

The previous chapters described and discussed the findings of the cross-sectional study. Chapter 2 (section 2.6.1.1, page 96) highlighted that very few studies have examined changes in HRQoL over time in people with ACHD. In addition, no studies have examined changes in psychosocial functioning over time in ACHD, including the perceptions people hold about their condition, the coping strategies they utilise, their perceptions of social support, and their mood (section 3.8, page 119). The longitudinal study in this thesis was designed to address both these issues. This chapter describes the analyses conducted using hierarchical linear models to examine changes in psychosocial factors and HRQoL over time and concludes with a discussion of the findings.

9.2. Response rate and sample attrition

Figure 9.1 displays the flow diagram for the participant recruitment process.

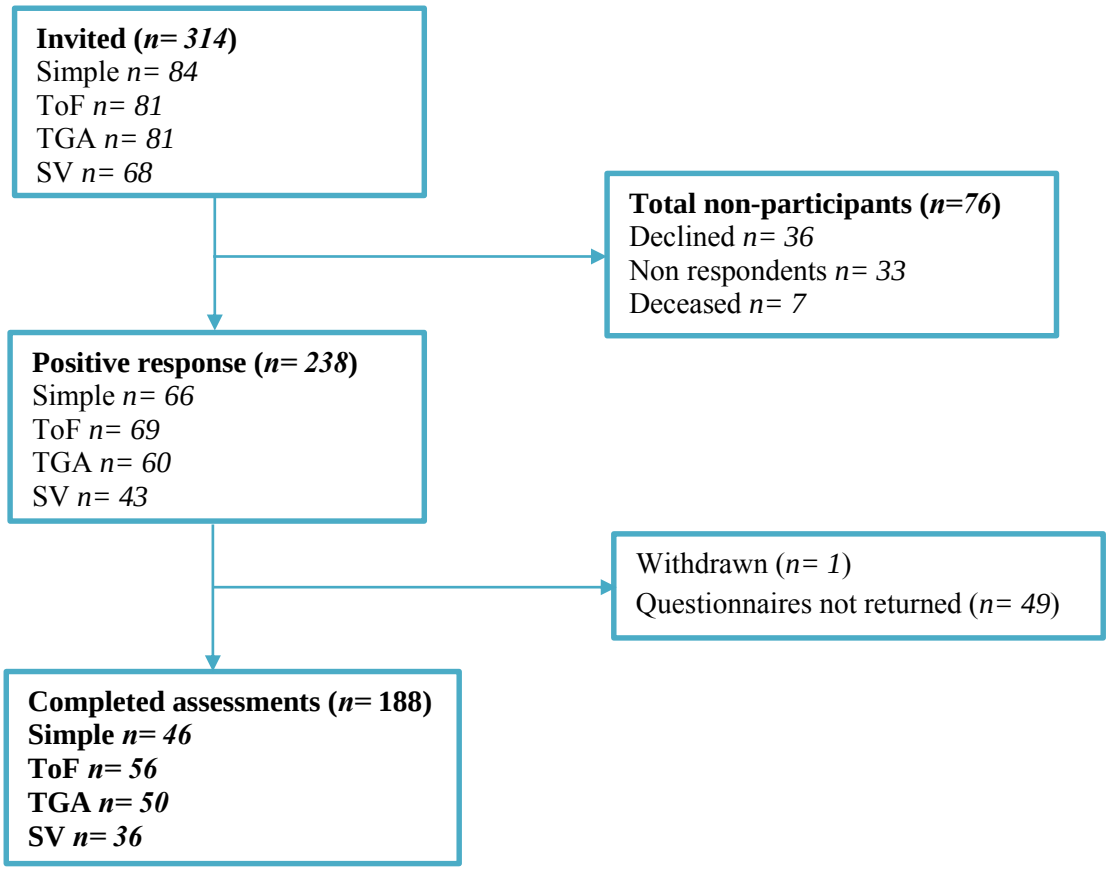


Figure 9.1. Flow diagram of the participant recruitment process for the longitudinal study

A total of 314 prospective participants, all in the initial cross-sectional study were invited to participate in the follow-up study. Of the 238 (75.8%) who consented to take part, one (0.4%) participant withdrew and 49 (20.6%) failed to return the postal questionnaire packs. The final sample for the longitudinal study consisted of 188 participants, resulting in a 60% response rate.

9.3. Missing value analysis

Missing value analysis was performed at scale level and is presented in Appendix P. A total of 14 (0.2%) values were missing of 7520. Little's MCAR suggested that the data were MCAR (p= .283) and that imputation is appropriate. Imputation was not performed for participants who did not complete the follow-up assessments as

hierarchical linear models still use all available data without deleting cases that have completed only one of the assessments.

9.4. Scale reliability

All scales and subscales of the questionnaires demonstrated satisfactory internal reliability based on Cronbach's α scores and/or inter-item correlations (Table 9.1).

Table 9.1. Internal reliability of scales/subscales at follow-up

Scales & Subscales	*No. of Items*	*Cronbach α/ Inter-Item Correlations*[§]
SF-36		
Physical functioning	10	.93
Role physical	4	.93
Bodily pain	2	.80/ .68
General health	5	.86
Vitality	4	.87
Social functioning	2	.85/ .75
Role emotional	3	.80
Mental health	5	.84
CHD-TAAQOL		
Symptoms	9	.82
Worries	10	.85
Impact cardiac surveillance	7	.77
Brief COPE		
Planning	2	.72/ .56
Active coping	2	.84/ .72
Positive reframing	2	.76/ .62
Acceptance	2	.64/ .47
Humour	2	.91/ .84
Religion	2	.94/ .88
Seeking emotional support	2	.83/ .70
Seeking instrumental support	2	.84/ .72
Self-distraction	2	.62/ .45
Denial	2	.81/ .68
Venting	2	.64/ .48
Substance use	2	.96/ .93
Behavioural disengagement	2	.47/ .38
Self-blame	2	.57/ .42
STAI-6	6	.84
CES-D 10	10	.82
MSPSS		
Family support	4	.95
Friends support	4	.94
Special person support	4	.97

[§]Inter-item correlations are also reported for scales and subscales consisting of two items.

9.5. Differences between responders and non-responders

There were no significant differences in demographic and clinical characteristics between people who completed the follow-up assessment and those that did not (Table 9.2).

Table 9.2. Comparisons between responders and non-responders in demographic and clinical characteristics

Characteristic	***Responders (n= 188)***	***Non-Responders (n= 126)***	***Test Statistic***	***Sig.***
Age (*mean, S.D.)*	34 (11.5)	31.9 (9.3)	$f(1,312)= 2.94$	.088
Gender (*n, %*)			$\chi^2(1)= 0.48$	.490
Male	103 (58.2)	74 (41.8)		
Female	85 (62)	52 (38)		
Marital status (*n, %*)			$\chi^2(1)= 2.04$	.154
Married	89 (56)	70 (44)		
Single	99 (63.9)	56 (36.1)		
Educational level (*n, %*)			$\chi^2(1)= 1.97$	.161
School	120 (57.1)	90 (42.9)		
University	68 (65.4)	36 (34.6)		
Employment status (*n, %*)			$\chi^2(1)= 1.39$	.238
Employed	137 (62)	84 (38)		
Unemployed	51 (54.8)	42 (45.2)		
Diagnostic group (*n, %*)			$\chi^2(3)= 5.28$	.152
Simple	46 (54.8)	38 (45.2)		
ToF	56 (69.1)	25 (30.9)		
TGA	50 (61.7)	31 (38.3)		
SV	36 (52.9)	32 (47.1)		
Co-morbidities no. (*mean, S.D.*)	1 (1.09)	.92 (1.09)	$f(1,312)= 0.40$	.526
Cyanosis days (*mean, S.D.*)	1311.16 (2819.48)	1326.73 (2554.28)	$f(1,312)= 0.00$	.960
Interventions no. (*mean, S.D.*)	2.35 (1.36)	2.4 (1.44)	$f(1,312)= 0.11$	.738
Medication no. (*mean, S.D.*)	0.97 (1.32)	0.94 (1.27)	$f(1,312)= 0.05$	.833

There were also no significant differences on any of the psychosocial measures between people who completed the follow-up assessment and those that did not, except for depression (Table 9.3). The chi-square test was significant ($\chi^2(1)= 8.10$, $p< .004$, $\varphi_c= .16$) suggesting a significant relationship between depressive symptoms and completion of the follow-up assessment. Individuals with depressive symptoms were more likely to have not completed the follow-up assessment. The effect size was however small.

Table 9.3. Comparisons between responders and non-responders in psychosocial measures

Variable (mean, S.D.)	*Responders*	*Non-responders*	*Test Statistic*	*Sig.*
Physical component summary	50.7 (9.3)	51.3 (9.6)	$f(1,300)= 0.27$	.601
Physical functioning	50.2 (8.5)	49.3 (9.5)	$f(1,301)= 0.77$	.380
Role physical	50.5 (9.4)	50.5 (10.3)	$f(1,300)= 0.00$	.981
Bodily pain	53.8 (10.7)	55.6 (9.9)	$f(1,301)= 2.15$	.143
General health	45.3 (10.5)	44.8 (10.7)	$f(1,301)= 0.21$	.649
Mental component summary	48.7 (47.3)	47.3 (11.3)	$f(1,300)= 1.21$	.272
Vitality	51 (10.5)	50.5 (10.8)	$f(1,301)= 0.15$	.700
Social functioning	48.9 (10.6)	48.4 (10.3)	$f(1,301)= 0.21$	.647
Role emotional	49.5 (10.8)	48.9 (11.3)	$f(1,300)= 0.19$	.668
Mental health	48.7 (10.4)	46.6 (11.1)	$f(1,301)= 2.91$	.089
Symptoms	85.9 (14.6)	86.7 (13.7)	$f(1,301)= 0.20$	.659
Worries	82.4 (15.7)	81.7 (14.4)	$f(1,301)= 0.16$	.686
Impact cardiac surveillance	85.6 (10.4)	85.8 (10.8)	$f(1,301)= 0.02$	.884
Brief IPQ consequences	3.2 (2.7)	3.4 (2.8)	$f(1,301)= 0.24$	.625
Brief IPQ timeline	9.3 (2.1)	9.3 (2)	$f(1,301)= 0.03$	.865
Brief IPQ controllability	5.4 (2.8)	5.1 (3.2)	$f(1,301)= 0.64$	.424
Brief IPQ curability	7.4 (2.7)	6.9 (3.2)	$f(1,301)= 2.05$	.153
Brief IPQ identity	2.8 (2.4)	2.6 (2.3)	$f(1,301)= 0.30$	.582
Brief IPQ concern	4.2 (2.7)	4.1 (3)	$f(1,301)= 0.05$	.822
Brief IPQ coherence	6.4 (2.4)	5.9 (2.7)	$f(1,301)= 2.99$	.085
Brief IPQ emotional representation	3.3 (2.9)	3.8 (3.4)	$f(1,301)= 2.07$	.151
Brief COPE planning	1.5 (0.8)	1.3 (0.9)	$f(1,301)= 4.86$	.028
Brief COPE active coping	1.7 (0.8)	1.5 (0.9)	$f(1,301)= 6.23$	.013
Brief COPE positive reframing	1.5 (0.9)	1.4 (0.9)	$f(1,301)= 1.42$	.234
Brief COPE acceptance	2.3 (0.7)	2.2 (0.8)	$f(1,301)= 2.00$	.159
Brief COPE humour	1.3 (1.1)	1.3 (1.1)	$f(1,301)= 0.03$	.870
Brief COPE religion	0.5 (0.9)	0.6 (0.9)	$f(1,301)= 1.31$	.254
Brief COPE emotional support	1.2 (0.9)	1.2 (1)	$f(1,301)= 0.06$	.800
Brief COPE instrumental support	1.2 (0.9)	1 (0.9)	$f(1,301)= 2.35$	.126
Brief COPE self-distraction	1.3 (0.8)	1.3 (0.9)	$f(1,301)= 0.00$	.980
Brief COPE denial	0.2 (0.4)	0.2 (0.4)	$f(1,301)= 0.33$	.565
Brief COPE venting	0.7 (0.7)	0.8 (0.8)	$f(1,301)= 0.42$	.519
Brief COPE substance use	0.2 (0.4)	0.3 (0.6)	$f(1,301)= 0.84$	.359
Brief COPE behavioural disengagement	0.2 (0.5)	0.3 (0.6)	$f(1,301)= 0.93$	.336
Brief COPE self-blame	0.5 (0.7)	0.5 (0.7)	$f(1,301)= 0.03$	.865
MSPSS family support	4.2 (0.7)	4.2 (0.9)	$f(1,301)= 0.00$	.969
MSPSS friend support	4 (0.8)	4 (0.9)	$f(1,301)= 0.11$	.742
MSPSS significant other support	4.3 (0.9)	4.2 (1.1)	$f(1,301)= 1.13$	.288
Anxiety	1.6 (0.6)	1.7 (0.6)	$f(1,301)= 1.92$	.167
Depression (*n, %*)			$\chi^2(1)= 8.10$	**.004**
No depressive symptoms	156 (65.5)	82 (34.5)		
With depressive symptoms	30 (46.2)	35 (53.8)		

9.6. Changes over time in clinical and psychosocial factors

The mean follow-up was 3.3 years (*S.D.*= 0.68, range: 1.7-4.5). The analysis indicated that the interval between baseline and follow-up assessments was significantly different between diagnostic groups [f (3,184)= 3.37, p= .020]; hence, follow-up years was included as a covariate (fixed effect) in the HLMs or mixed within-between ANOVAs.

F-test results are presented for the analyses conducted to determine the effect of time, diagnostic group, and Diagnostic Group x Time interaction. The main effects of diagnostic group are presented but are not interpreted as these analyses replicate those discussed in Chapter 7. The main effects of time are reported for each outcome, however their interpretation is not recommended when interaction effects are significant (Pallant, 2013). Therefore, the main effects of time were interpreted only when interaction effects were not significant. A significant time effect was interpreted as an indication of significant change over time. A significant Diagnostic Group x Time interaction was interpreted as an indication that changes over time were different by diagnostic group. Where within-between ANOVAs were conducted this is indicated with the results in the associated tables. Effect sizes (Cohen's *d*) are reported where differences between diagnostic groups are significant. Statistical tests for which results were significant (p< .01) are presented in bold.

9.6.1. Changes over time in clinical factors

Table 9.4 presents the results of the mixed within-between ANOVAs. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on clinical factors, indicating no change over time and across

diagnostic groups. See Appendix U for a graphical representation of clinical factors over time.

Table 9.4. Adjusted means (*S.D.*) and effects for clinical factors (*n*= 188)

Clinical Factors	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Intervention no.*			*f*(1,183)= 1.59, *p*= .208	*f*(3,183)= 10.54, *p*< .001	*f*(3,183)= 3.35, *p*= .020
Simple	1.46 (1.28)	1.67 (1.73)			
ToF	2.43 (1.27)	2.58 (1.73)			
TGA	2.76 (1.27)	3.12 (1.73)			
SV	2.81 (1.27)	3.52 (1.72)			
Total	2.36 (1.28)	2.72 (1.74)			
Hospitalization days*			*f*(1,183)= 3.00, *p*= .085	*f*(3,183)= 15.42, *p*< .001	*f*(3,183)= 1.99, *p*= .118
Simple	18.59 (28.70)	19.80 (29.69)			
ToF	48.78 (28.59)	49.22 (29.57)			
TGA	36.09 (28.58)	37.06 (29.56)			
SV	58.07 (28.57)	60.76 (29.54)			
Total	40.38 (28.81)	41.71 (29.79)			
Medication no.*			*f*(1,183)= 0.54, *p*= .464	*f*(3,183)= 3.85, *p*= .011	*f*(3,183)= 0.43, *p*= .733
Simple	0.88 (1.29)	1.48 (2.27)			
ToF	0.57 (1.29)	1.39 (2.25)			
TGA	0.99 (1.29)	1.62 (2.26)			
SV	1.67 (1.28)	2.61 (2.25)			
Total	1.03 (1.29)	1.78 (2.28)			

*Based on mixed within-between ANOVA analyses.

9.6.2. Changes over time in HRQoL

Table 9.5 presents the results of the HLM for generic physical HRQoL, including the physical component summary and all of the subscales. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on physical HRQoL, indicating no change over time and across diagnostic groups. Figure 9.2 presents a graphical representation of the physical component summary over time. Figures 9.3 to 9.6 present a graphical representation of the physical subscales over time.

Table 9.5. HLM: Adjusted means (*S.D.*) and effects for generic physical HRQoL (*n*= 188)

HRQoL Subscale	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Physical component summary			*f*(1,181.61)= 0.46, *p*= .499	*f*(3,183.19)= 3.71, *p*= .013	*f*(3,181.61)= 1.18, *p*= .319
Simple	52.85 (9.12)	53.44 (9.87)			
ToF	51.82 (9.13)	49.79 (9.79)			
TGA	50.35 (9.09)	50.80 (9.83)			
SV	47.13 (9.13)	46.49 (9.77)			
Total	50.54 (9.18)	50.13 (9.90)			
Physical functioning			*f*(1,183.84)= 3.57, *p*= .060	*f*(3,183.66)= 5.77, *p*< .001	*f*(3,183.87)= 1.28, *p*= .283
Simple	52.70 (8.26)	52.40 (9.93)			
ToF	51.25 (8.28)	48.33 (9.89)			
TGA	50.22 (8.25)	49.78 (9.89)			
SV	45.42 (8.27)	44.66 (9.89)			
Total	49.90 (8.32)	48.79 (10)			
Role physical			*f*(1,183.79)= 0.11, *p*= .739	*f*(3,183.49)= 3.20, *p*= .025	*f*(3,183.81)= 0.32, *p*= .810
Simple	51.94 (9.35)	52.75 (9.90)			
ToF	51.36 (9.35)	51.49 (9.87)			
TGA	50.55 (9.33)	51.61 (9.86)			
SV	47.58 (9.35)	46.60 (9.87)			
Total	50.36 (9.42)	50.61 (9.97)			
Bodily pain			*f*(1,182.81)= 0.02, *p*= .899	*f*(3,183.34)= 1.14, *p*= .336	*f*(3,182.88)= 1.95, *p*= .124
Simple	52.49 (10.60)	55.21 (10.53)			
ToF	56.04 (10.60)	53.79 (10.46)			
TGA	54.16 (10.58)	53.40 (10.49)			
SV	51.37 (10.60)	51.26 (10.49)			
Total	53.51 (10.68)	53.42 (10.60)			
General health			*f*(1,183.47)= 0.21, *p*= .647	*f*(3,183.45)= 4.12, *p*= .007	*f*(3,183.50)= 0.39, *p*= .757
Simple	48.16 (10.34)	48.37 (11.56)			
ToF	45.91 (10.36)	46.19 (11.52)			
TGA	44.93 (10.31)	44.87 (11.52)			
SV	41.52 (10.35)	39.85 (11.51)			
Total	45.13 (10.42)	44.82 (11.63)			

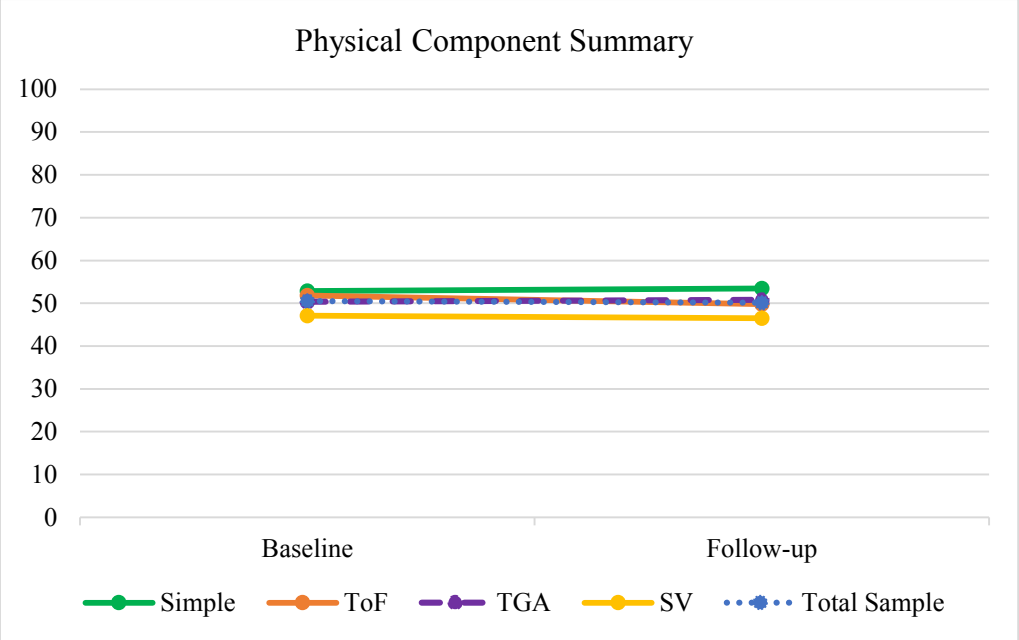


Figure 9.2. Graphical representation of the physical component summary over time

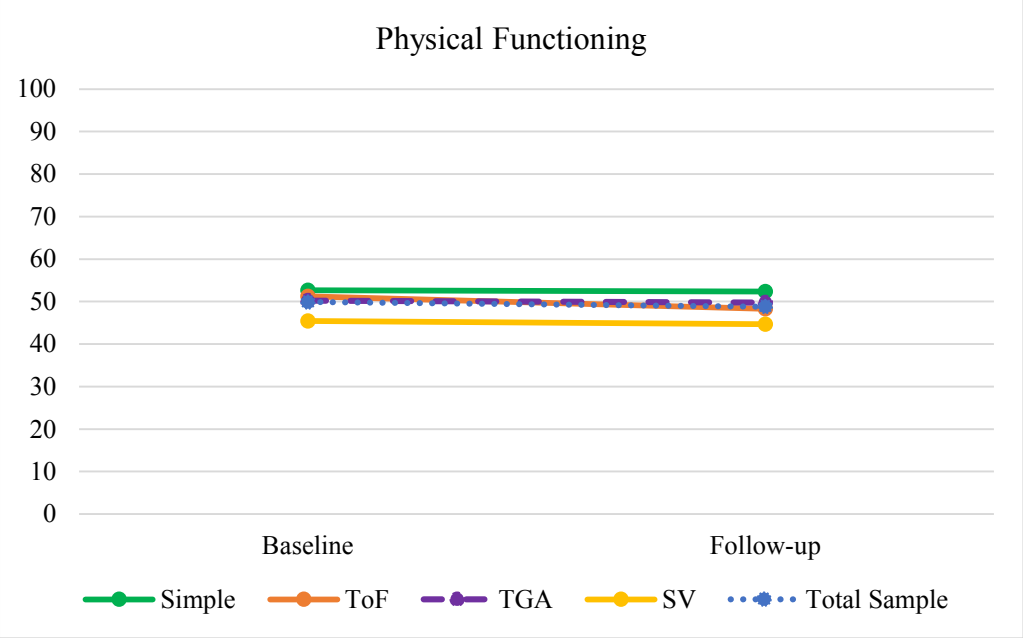


Figure 9.3. Graphical representation of the physical functioning subscale over time

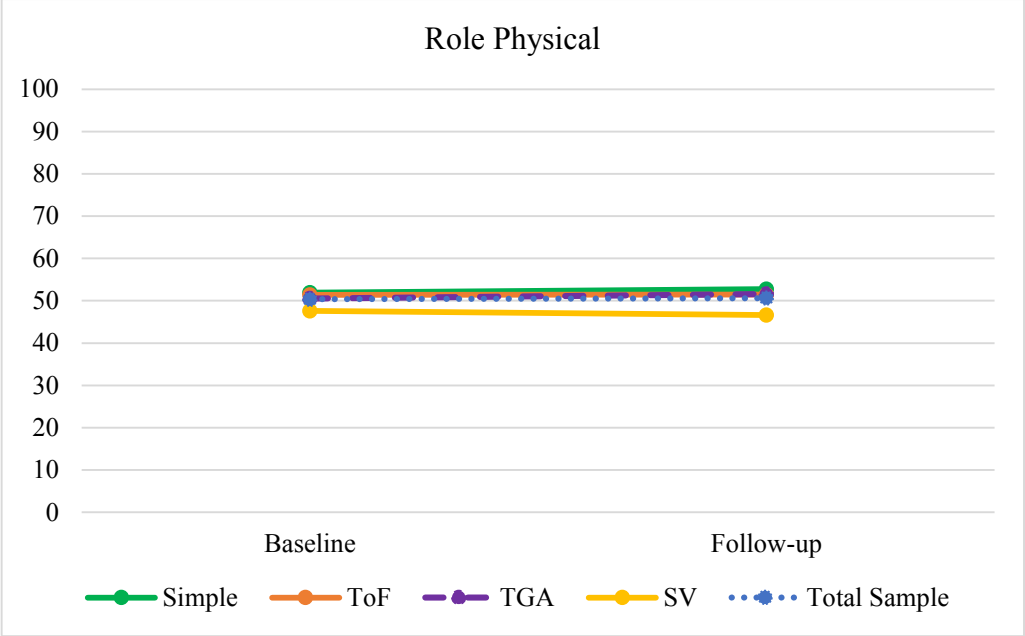


Figure 9.4. Graphical representation of the role physical subscale over time

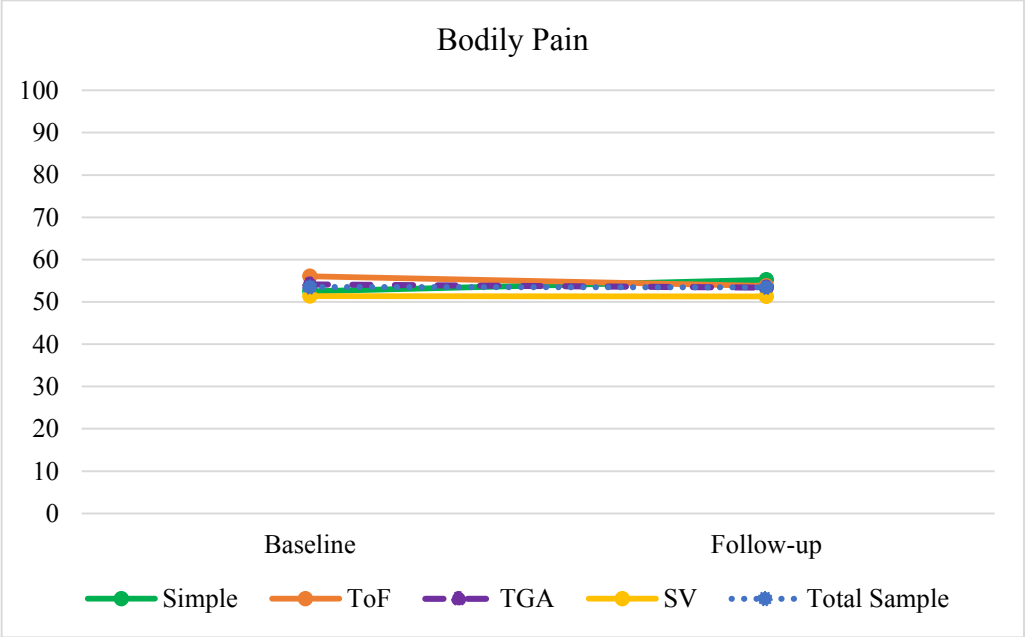


Figure 9.5. Graphical representation of the bodily pain subscale over time

Figure 9.6. Graphical representation of the general health subscale over time

Table 9.6 presents the results of the HLM for generic psychosocial HRQoL, including the mental component summary and all of the subscales. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on psychosocial HRQoL, indicating no change over time and across diagnostic groups. Figure 9.7 presents a graphical representation of the mental component summary over time. Figures 9.8 to 9.11 present a graphical representation of the psychosocial subscales over time.

Table 9.6. HLM: Adjusted means (*S.D.*) and effects for generic psychosocial HRQoL (*n*= 188)

HRQoL Subscale	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Mental component summary			*f*(1,182.11)= 0.07, *p*= .793	*f*(3,182.99)= 3.60, *p*= .015	*f*(3,182.09)= 0.90, *p*= .442
Simple	46.72 (10.54)	48.27 (10.55)			
ToF	50.80 (10.54)	51.23 (10.49)			
TGA	50.36 (10.52)	48.62 (10.51)			
SV	45.40 (10.54)	44.34 (10.63)			
Total	48.32 (10.62)	48.12 (10.61)			
Vitality			*f*(1,182.30)= 0.05, *p*= .820	*f*(3,183.52)= 3.50, *p*= .017	*f*(3,182.29)= 2.39, *p*= .071
Simple	48.14 (10.19)	51.04 (10.93)			
ToF	54.23 (10.20)	52.18 (10.86)			
TGA	52.71 (10.16)	51.38 (10.89)			
SV	47.25 (9.11)	47.05 (10.99)			
Total	50.58 (10.27)	50.41 (10.98)			
Social functioning			*f*(1,183.08)= 0.13, *p*= .717	*f*(3,183.42)= 4.28, *p*= .006	*f*(3,183.14)= 0.06, *p*= .980
Simple	49.51 (10.44)	49.23 (11.16)			
ToF	51.20 (10.45)	50.64 (11.10)			
TGA	49.70 (10.42)	49.06 (11.13)			
SV	43.82 (10.45)	44.13 (11.13)			
Total	48.56 (10.53)	48.26 (11.23)			
Role emotional			*f*(1,182.81)= 0.70, *p*= .406	*f*(3,182.69)= 2.54, *p*= .058	*f*(3,182.88)= 2.09, *p*= .103
Simple	49.17 (10.88)	50.87 (10.74)			
ToF	49.59 (10.87)	51.04 (10.70)			
TGA	51.05 (10.86)	48.27 (10.71)			
SV	47.34 (10.87)	43.83 (10.71)			
Total	49.29 (10.97)	48.50 (10.80)			
Mental health			*f*(1,181.49)= 0.01, *p*= .931	*f*(3,182.76)= 3.01, *p*= .032	*f*(3,181.47)= 0.42, *p*= .741
Simple	47.35 (10.28)	48.42 (10.51)			
ToF	51.32 (10.28)	50.33 (10.44)			
TGA	49.61 (10.26)	49.38 (10.47)			
SV	44.87 (10.28)	45.27 (10.57)			
Total	48.29 (10.35)	48.35 (10.56)			

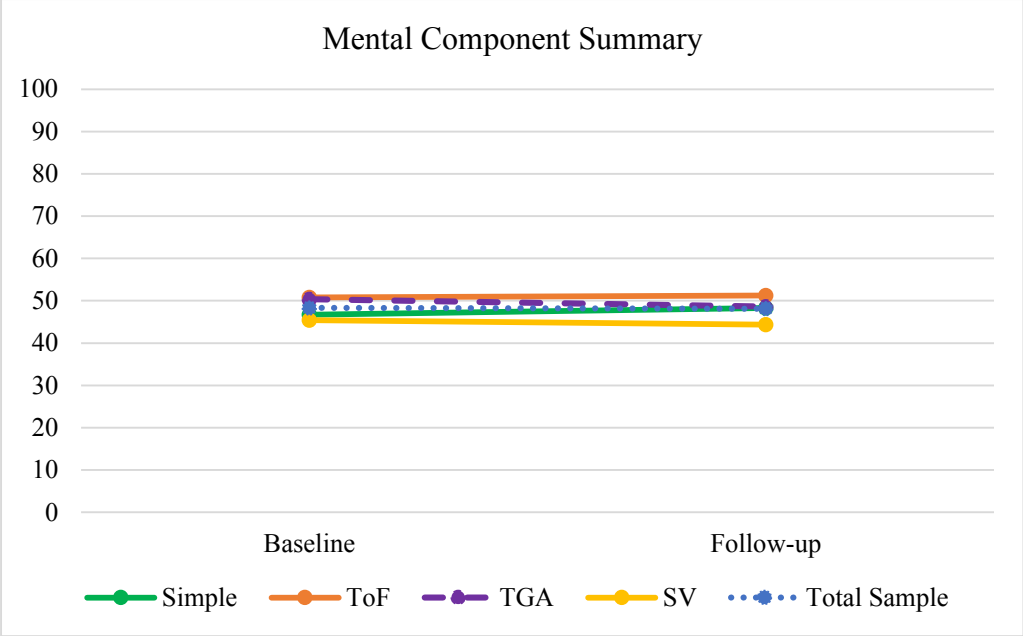


Figure 9.7. Graphical representation of the mental component summary over time

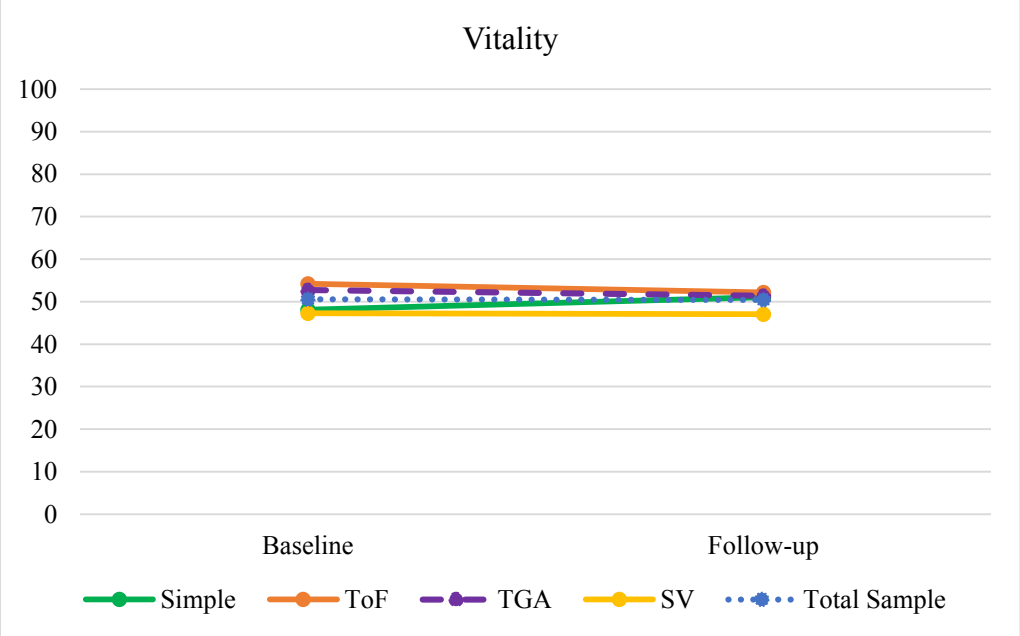


Figure 9.8. Graphical representation of the vitality subscale over time

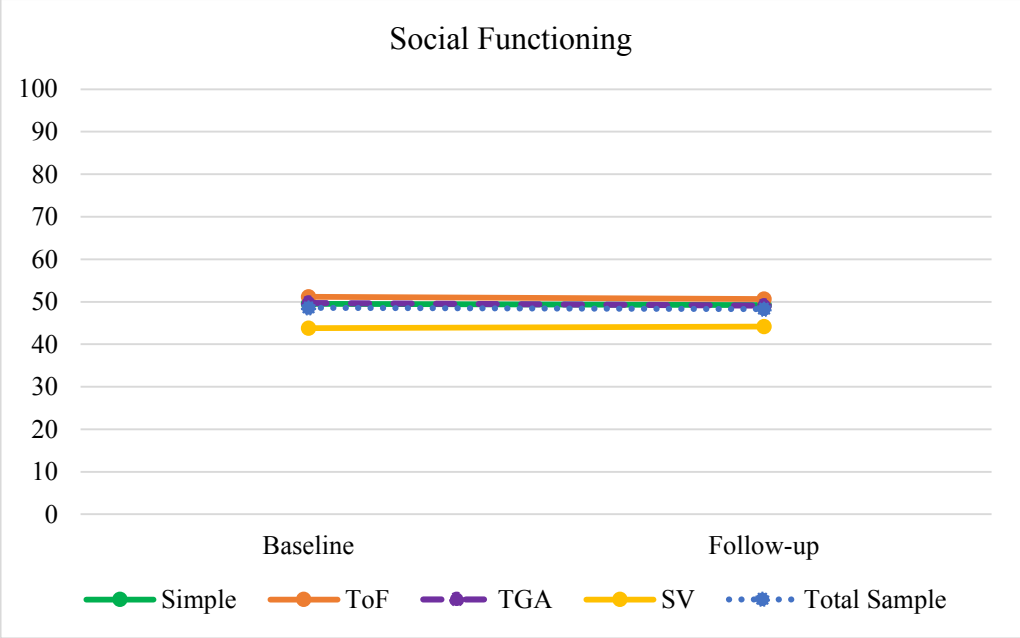


Figure 9.9. Graphical representation of the social functioning subscale over time

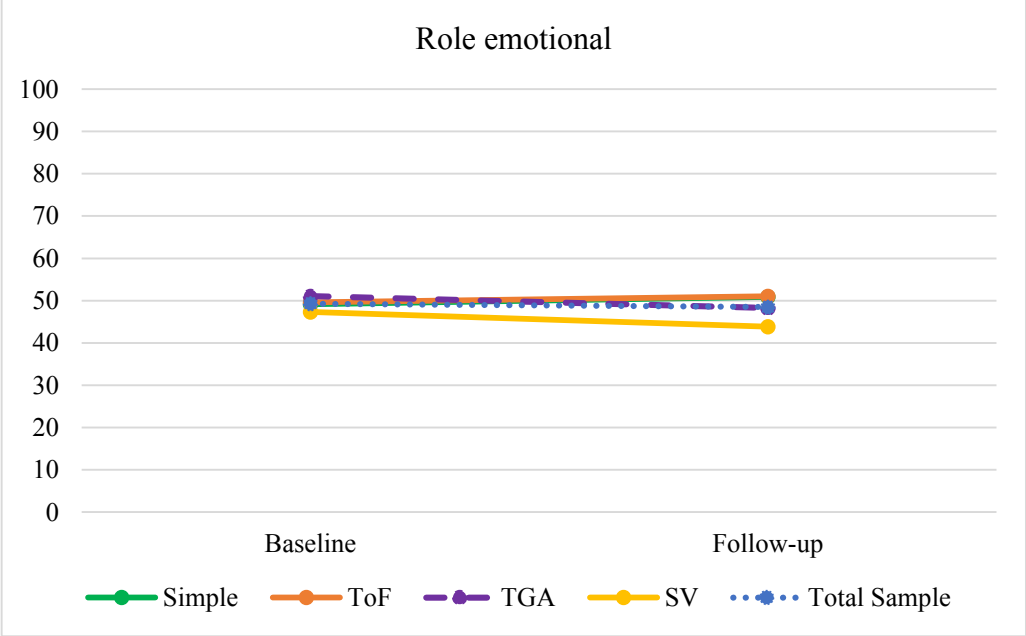


Figure 9.10. Graphical representation of the role emotional subscale over time

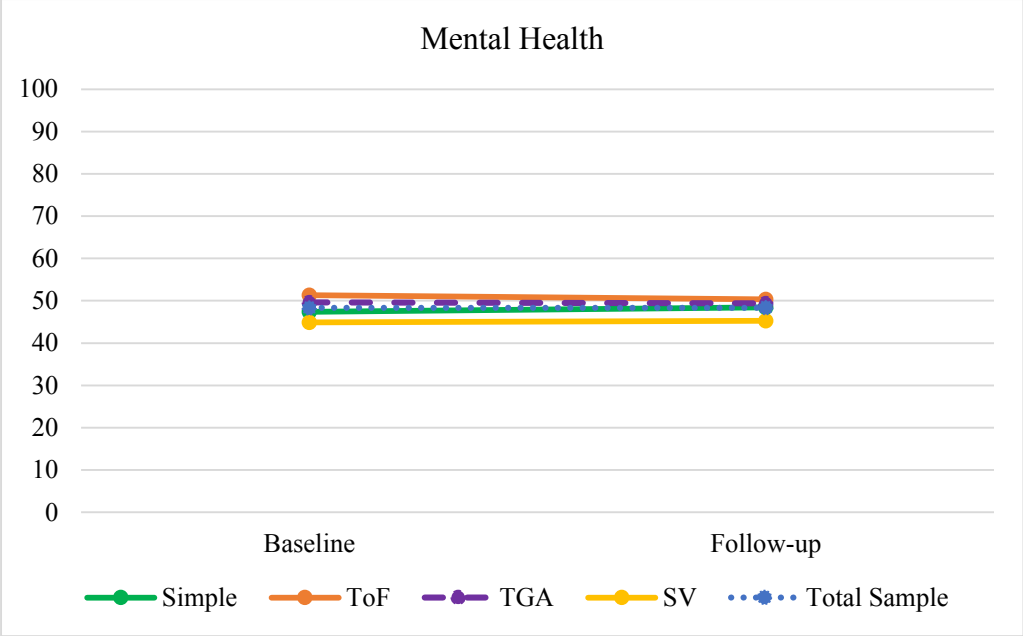


Figure 9.11. Graphical representation of the mental health subscale over time

Table 9.7 presents the results of the HLM for disease-specific HRQoL. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on disease-specific HRQoL, indicating no change over time and across diagnostic groups. Figures 9.12 to 9.14 present a graphical representation of disease-specific HRQoL over time.

Table 9.7. HLM: Adjusted means (*S.D.*) effects for disease-specific HRQoL (*n*= 188)

HRQoL Subscale	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Symptoms			*f*(1,182.53)= 0.20, *p*= .655	*f*(3,183.40)= 3.93, *p*= .010	*f*(3,182.53)= 1.21, *p*= .308
Simple	88.77 (14.30)	88.43 (13.35)			
ToF	87.72 (14.34)	85.42 (13.30)			
TGA	86.95 (14.26)	86.34 (13.29)			
SV	78.41 (14.33)	80.30 (13.30)			
Total	85.46 (14.43)	85.12 (13.41)			
Impact cardiac surveillance			*f*(1,183.11)= 2.54, *p*= .113	*f*(3,182.75)= 14.81, *p*< .001	*f*(3,183.13)= 1.22, *p*= .304
Simple	89.30 (10.06)	91.96 (9.59)			
ToF	86.90 (10.05)	89.46 (9.56)			
TGA	84.78 (10.04)	86.71 (9.55)			
SV	80.29 (10.04)	78.63 (9.56)			
Total	85.32 (10.13)	86.69 (9.65)			
Worries			*f*(1,183.14)= 0.13, *p*= .718	*f*(3,183.42)= 5.41, *p*= .001	*f*(3,183.16)= 0.03, *p*= .993
Simple	83.55 (15.33)	83.02 (15.69)			
ToF	85.91 (15.36)	85.64 (15.63)			
TGA	83.39 (15.29)	83.49 (15.63)			
SV	74.24 (15.36)	73.63 (15.63)			
Total	81.77 (15.45)	81.44 (15.77)			

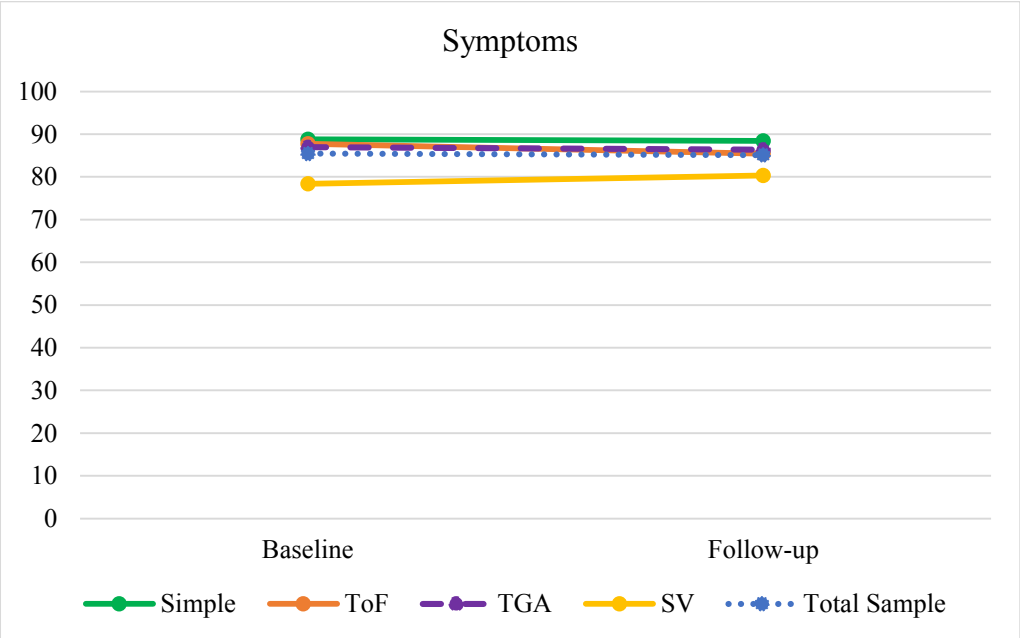


Figure 9.12. Graphical representation of symptoms over time

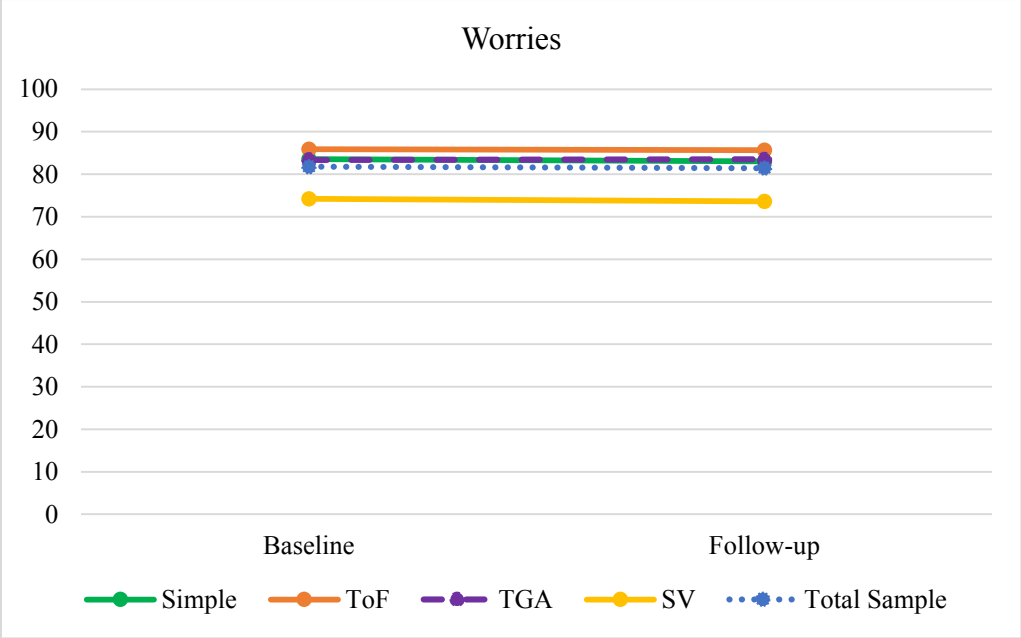


Figure 9.13. Graphical representation of worries over time

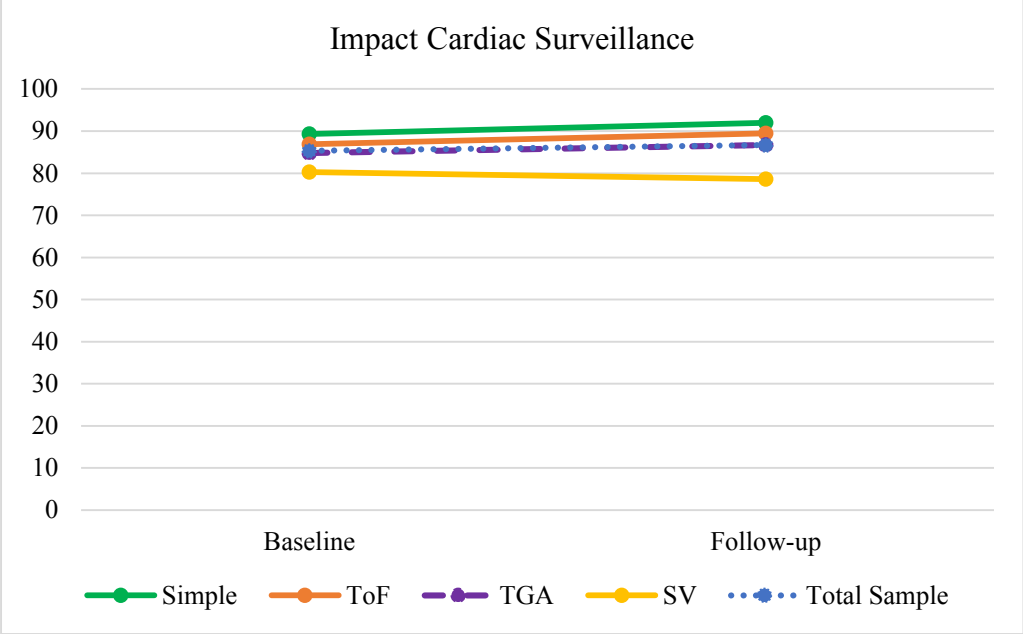


Figure 9.14. Graphical representation of impact of cardiac surveillance over time

9.6.3. Changes over time in illness perceptions

Table 9.8 presents the results of the HLM for illness perceptions. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on the Brief IPQ subscales of consequences, timeline, identity,

concern, and emotional representation, indicating no change over time and across diagnostic groups. See Appendix V for a graphical representation of these results.

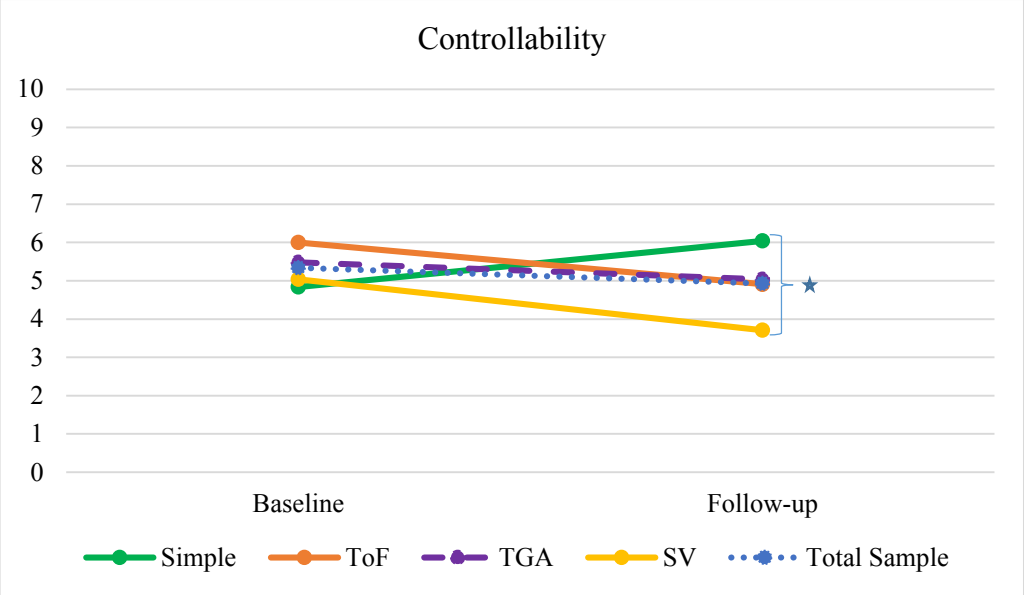
Table 9.8. HLM: Adjusted means (*S.D.*) and effects for illness perceptions (*n*= 188)

Illness Perceptions	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Consequences			*f*(1,183.20)= 0.01, *p*= .920	*f*(3,183.36)= 10.02, *p*< .001	*f*(3,183.22)= 1.22, *p*= .305
Simple	2.54 (2.56)	1.99 (2.62)			
ToF	2.91 (2.56)	3.04 (2.61)			
TGA	3.04 (2.55)	3.06 (2.61)			
SV	4.84 (2.56)	5.18 (2.61)			
Total	3.34 (2.58)	3.32 (2.63)			
Timeline			*f*(1,182.32)= 0.21, *p*= .645	*f*(3,182.50)= 5.04, *p*= .002	*f*(3,182.34)= 1.87, *p*= .137
Simple	8.38 (2.05)	8.37 (2.30)			
ToF	9.40 (2.05)	8.67 (2.29)			
TGA	9.71 (2.04)	9.71 (2.29)			
SV	9.70 (2.05)	10 (2.29)			
Total	9.30 (2.06)	9.24 (2.32)			
Controllability			*f*(1,182.17)= 2.66, *p*= .104	*f*(3,181.87)= 1.81, *p*= .147	*f*(3,182.20)= 5.00, ***p*= .002**
Simple	4.84 (2.79)	6.04 (3.11)			
ToF	6 (2.78)	4.91 (3.10)			
TGA	5.48 (2.79)	5.04 (3.10)			
SV	5.04 (2.78)	3.71 (3.10)			
Total	5.34 (2.81)	4.93 (3.13)			
Curability			*f*(1,183.69)= 0.04, *p*= .833	*f*(3,183.21)= 0.82, *p*= .483	*f*(3,183.72)= 4.45, ***p*= .005**
Simple	6.62 (2.64)	7.94 (2.83)			
ToF	8.15 (2.64)	7.43 (2.82)			
TGA	7.26 (2.64)	7.60 (2.82)			
SV	7.46 (2.64)	6.71 (2.82)			
Total	7.37 (2.66)	7.42 (2.85)			
Identity			*f*(1,183.37)= 2.23, *p*= .138	*f*(3,183.36)= 8.69, *p*< .001	*f*(3,183.39)= 2.05, *p*= .109
Simple	2.33 (2.27)	2.03 (2.46)			
ToF	2.24 (2.28)	2.88 (2.45)			
TGA	2.75 (2.27)	2.83 (2.45)			
SV	4.20 (2.28)	4.70 (2.45)			
Total	2.88 (2.29)	3.11 (2.47)			
Concern			*f*(1,181.89)= 0.54, *p*= .464	*f*(3,182.09)= 2.12, *p*= .100	*f*(3,181.91)= 0.77, *p*= .511
Simple	3.95 (2.73)	3.46 (2.96)			
ToF	4.10 (2.73)	3.78 (2.95)			
TGA	4.05 (2.72)	3.93 (2.95)			
SV	4.84 (2.73)	5.20 (2.95)			
Total	4.24 (2.75)	4.09 (2.98)			

Table 9.8 ***Continued***

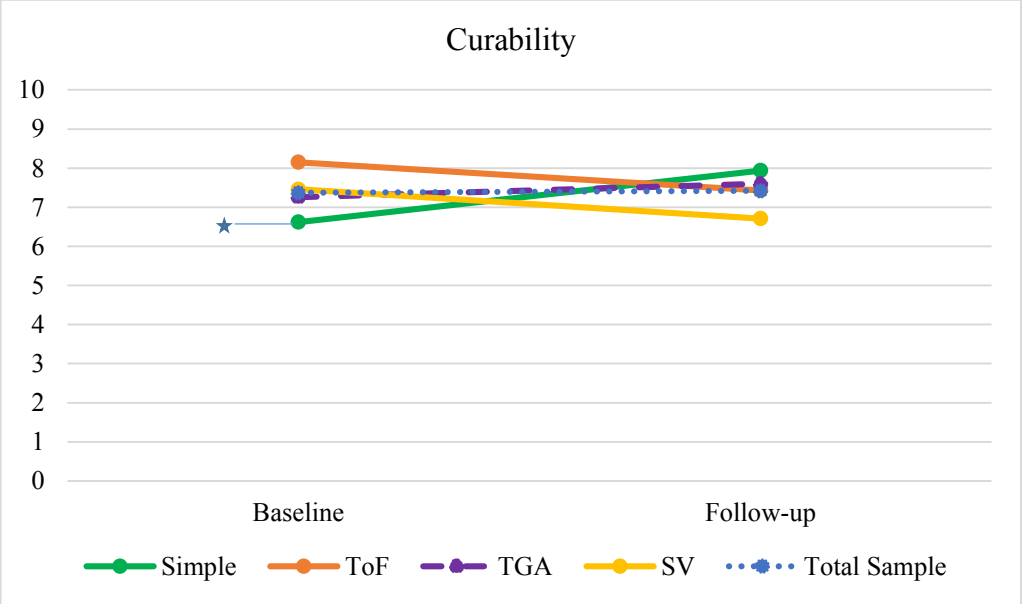
Illness Perceptions	***Baseline (Mean, S.D.)***	***Follow-up (Mean, S.D.)***	***Effect of Time***	***Effect of Diagnostic Group***	***Effect of Diagnostic Group x Time***
Coherence			$f(1,182.41)=55.07$, ***p*< .001**	$f(3,182.80)=0.83$, $p=.479$	$f(3,182.43)=0.03$, $p=.994$
Simple	6.75 (2.42)	7.93 (2.27)			
ToF	6.36 (2.42)	7.45 (2.26)			
TGA	6.39 (2.42)	7.46 (2.26)			
SV	6.05 (2.42)	7.19 (2.26)			
Total	6.39 (2.44)	7.51 (2.28)			
Emotional representation			$f(1,181.41)=0.71$, $p=.400$	$f(3,182.80)=0.83$, $p=.479$	$f(3,182.43)=0.03$, $p=.994$
Simple	2.77 (2.86)	2.07 (3.04)			
ToF	3.14 (2.87)	2.90 (3.03)			
TGA	3.14 (2.86)	3.20 (3.03)			
SV	4.58 (2.87)	4.80 (3.03)			
Total	3.41 (2.89)	3.24 (3.06)			

The interaction effects for diagnostic group and time were significant for controllability and curability. Pairwise comparisons indicated that the change in illness controllability was different for the SV group than for the Simple group; the SV group exhibited a significant reduction in perceptions about illness controllability over time compared with the Simple group which exhibited a significant increase in illness controllability (mean difference= -2.34, $p=.005$, 99% CI [-4.53, -0.15]; $d=.78$) (Figure 9.2). Pairwise comparisons for interactions effects on curability did not reach statistical significance. However, there was an increase in perceptions of curability within the Simple group over time (mean difference= -1.32, $p=.005$, 99% CI [-2.54, -0.10]; $d=.44$) (Figure 9.3).



Note. Star indicates significant interaction effect between diagnostic group (Simple vs. SV) and time

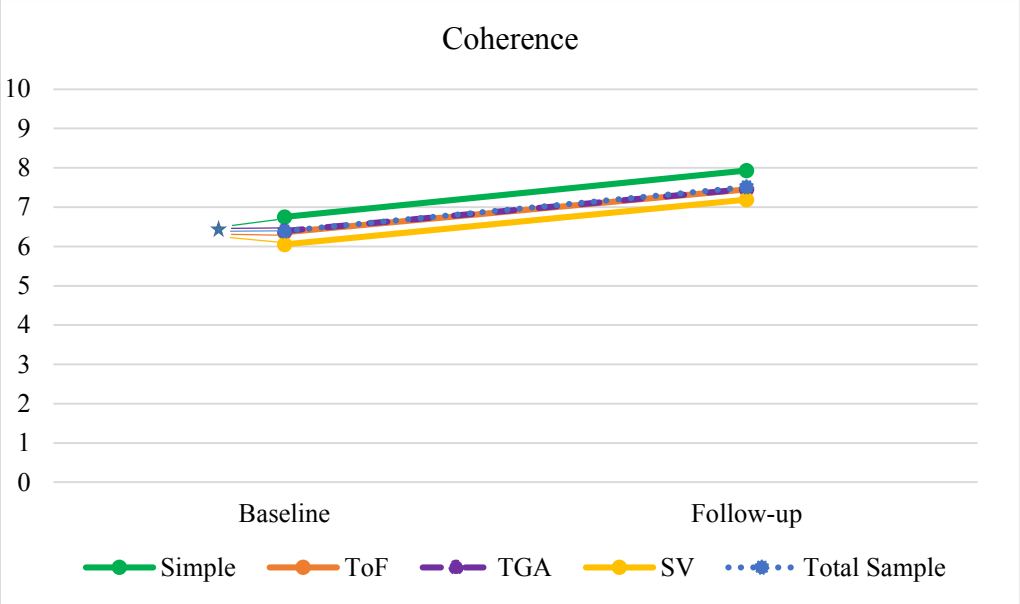
Figure 9.15. Graphical representation of change over time in illness controllability



Note. Star indicates significant main effect for time in the Simple group

Figure 9.16. Graphical representation of change over time in illness curability

There was a significant effect for time on the illness coherence subscale, with a significant increase in scores over time for the whole sample and all diagnostic groups (mean difference= -1.12, $p< .001$, 99% CI [-1.51, -0.73]; $d= .48$) (Figure 9.4).



Note. Star indicates significant main effect for time in the total sample and all diagnostic groups

Figure 9.17. Graphical representation of change over time in illness coherence

9.6.4. Changes over time in coping

Table 9.9 presents the HLM for each of the coping strategies. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on any of the coping strategies, indicating no change over time and across diagnostic groups. See Appendix W for a graphical representation of coping over time.

Table 9.9. HLM: Adjusted means (*S.D.*) and effects for coping strategies (*n*= 188)

Coping Strategies	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Planning			*f*(1,183.62)= 0.85, *p*= .358	*f*(3,183.32)= 3.00, *p*= .032	*f*(3,183.64)= 1.03, *p*= .381
Simple	1.42 (0.85)	1.19 (0.90)			
ToF	1.55 (0.85)	1.42 (0.90)			
TGA	1.30 (0.85)	1.37 (0.90)			
SV	1.73 (0.85)	1.76 (0.89)			
Total	1.50 (0.85)	1.44 (0.90)			
Active coping			*f*(1,183.75)= 0.84, *p*= .359	*f*(3,183.35)= 5.47, *p*= .001	*f*(3,183.78)= 0.30, *p*= .827
Simple	1.53 (0.80)	1.45 (0.92)			
ToF	1.93 (0.80)	1.82 (0.92)			
TGA	1.57 (0.80)	1.44 (0.92)			
SV	1.94 (0.80)	2 (0.92)			
Total	1.74 (0.80)	1.68 (0.93)			
Positive reframing			*f*(1,183.14)= 0.38, *p*= .540	*f*(3,183.14)= 1.62, *p*= .186	*f*(3,183.16)= 3.25, *p*= .023
Simple	1.53 (0.95)	1.23 (0.94)			
ToF	1.65 (0.94)	1.83 (0.94)			
TGA	1.53 (0.95)	1.53 (0.94)			
SV	1.37 (0.95)	1.67 (0.94)			
Total	1.52 (0.95)	1.56 (0.95)			
Acceptance			*f*(1,183.76)= 0.25, *p*= .621	*f*(3,183.32)= 3.34, *p*= .021	*f*(3,183.78)= 0.79, *p*= .503
Simple	2.20 (0.73)	2.08 (0.73)			
ToF	2.41 (0.73)	2.43 (0.73)			
TGA	2.25 (0.73)	2.39 (0.74)			
SV	2.48 (0.73)	2.57 (0.73)			
Total	2.33 (0.74)	2.37 (0.74)			
Humour			*f*(1,182.65)= 0.56, *p*= .454	*f*(3,182.72)= 0.80, *p*= .497	*f*(3,182.68)= 1.17, *p*= .323
Simple	1.28 (1.09)	1.12 (1.13)			
ToF	1.19 (1.09)	1.28 (1.12)			
TGA	1.18 (1.08)	1.41 (1.12)			
SV	1.47 (1.09)	1.54 (1.12)			
Total	1.28 (1.09)	1.34 (1.14)			
Religion			*f*(1,182.65)= 0.03, *p*= .875	*f*(3,183.24)= 0.40, *p*= .753	*f*(3,182.66)= 1.00, *p*= .393
Simple	0.47 (0.85)	0.55 (0.85)			
ToF	0.56 (0.85)	0.48 (0.85)			
TGA	0.40 (0.85)	0.36 (0.85)			
SV	0.37 (0.85)	0.44 (0.85)			
Total	0.45 (0.86)	0.46 (0.86)			
Emotional support			*f*(1,182.28)= 0.00, *p*= .962	*f*(3,182.27)= 1.48, *p*= .221	*f*(3,182.31)= 1.05, *p*= .372
Simple	1.12 (0.91)	0.97 (0.98)			
ToF	1.38 (0.91)	1.31 (0.97)			
TGA	1.11 (0.91)	1.26 (0.97)			
SV	1.31 (0.91)	1.39 (0.97)			
Total	1.23 (0.91)	1.23 (0.99)			

Table 9.9 ***Continued***

Coping Strategies	***Baseline (Mean, S.D.)***	***Follow-up (Mean, S.D.)***	***Effect of Time***	***Effect of Diagnostic Group***	***Effect of Diagnostic Group x Time***
Instrumental support			$f(1,182.84)=0.62$, $p=.433$	$f(3,182.75)=1.43$, $p=.237$	$f(3,182.87)=2.08$, $p=.105$
Simple	1.05 (0.92)	0.84 (0.92)			
ToF	1.26 (0.92)	1.17 (0.92)			
TGA	1.25 (0.92)	1.07 (0.92)			
SV	1.13 (0.92)	1.39 (0.92)			
Total	1.17 (0.93)	1.12 (0.93)			
Self-distraction			$f(1,182.38)=0.23$, $p=.634$	$f(3,182.11)=4.23$, $p=.006$	$f(3,182.41)=1.64$, $p=.182$
Simple	1.37 (0.80)	1.16 (0.88)			
ToF	1.24 (0.80)	1.13 (0.88)			
TGA	1.09 (0.80)	1.28 (0.88)			
SV	1.67 (0.80)	1.67 (0.88)			
Total	1.34 (0.80)	1.31 (0.89)			
Denial			$f(1,183.85)=0.53$, $p=.469$	$f(3,183.52)=1.71$, $p=.166$	$f(3,183.88)=1.94$, $p=.124$
Simple	0.20 (0.45)	0.05 (0.47)			
ToF	0.15 (0.45)	0.15 (0.47)			
TGA	0.22 (0.45)	0.14 (0.47)			
SV	0.25 (0.45)	0.36 (0.47)			
Total	0.20 (0.45)	0.17 (0.47)			
Venting			$f(1,182.44)=0.42$, $p=.516$	$f(3,182.14)=4.49$, $p=.005$	$f(3,182.47)=0.70$, $p=.554$
Simple	0.67 (0.68)	0.72 (0.69)			
ToF	0.74 (0.68)	0.63 (0.69)			
TGA	0.59 (0.68)	0.47 (0.69)			
SV	0.96 (0.68)	1 (0.68)			
Total	0.74 (0.68)	0.70 (0.69)			
Substance use			$f(1,182.60)=0.10$, $p=.751$	$f(3,182.11)=0.97$, $p=.406$	$f(3,182.63)=0.98$, $p=.403$
Simple	0.29 (0.44)	0.19 (0.48)			
ToF	0.16 (0.45)	0.11 (0.48)			
TGA	0.18 (0.45)	0.16 (0.48)			
SV	0.19 (0.44)	0.30 (0.48)			
Total	0.21 (0.45)	0.19 (0.48)			
Behavioural disengagement			$f(1,183.78)=0.07$, $p=.786$	$f(3,183.46)=2.13$, $p=.097$	$f(3,183.80)=1.24$, $p=.298$
Simple	0.20 (0.45)	0.16 (0.47)			
ToF	0.10 (0.45)	0.15 (0.46)			
TGA	0.26 (0.45)	0.14 (0.47)			
SV	0.30 (0.45)	0.36 (0.46)			
Total	0.21 (0.45)	0.20 (0.47)			
Self-blame			$f(1,182.16)=1.99$, $p=.160$	$f(3,181.80)=1.34$, $p=.264$	$f(3,182.19)=0.68$, $p=.565$
Simple	0.55 (0.66)	0.38 (0.61)			
ToF	0.36 (0.66)	0.36 (0.61)			
TGA	0.55 (0.67)	0.40 (0.61)			
SV	0.57 (0.66)	0.57 (0.62)			
Total	0.51 (0.67)	0.43 (0.62)			

9.6.5. Changes over time in social support

Table 9.10 presents the HLM for social support. There were no statistically significant interaction effects between diagnostic group and time or main effects for time on levels of social support, indicating no change over time and across diagnostic groups. See Appendix X for a graphical representation of social support over time.

Table 9.10. HLM: Adjusted means (*S.D.*) and effects for social support (*n*= 188)

Social Support	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Family support			*f*(1,184.03)= 2.79, *p*= .096	*f*(3,184.18)= 2.32, *p*= .077	*f*(3,184.06)= 1.34, *p*= .262
Simple	4.05 (0.74)	3.73 (1.04)			
ToF	4.37 (0.73)	4.21 (1.04)			
TGA	4.22 (0.74)	4.18 (1.04)			
SV	4.12 (0.74)	4.18 (1.04)			
Total	4.19 (0.74)	4.08 (1.06)			
Friends support			*f*(1,183.51)= 3.19, *p*= .076	*f*(3,183.23)= 0.62, *p*= .602	*f*(3,183.54)= 0.74, *p*= .531
Simple	4.16 (0.85)	3.93 (0.98)			
ToF	4 (0.85)	3.79 (0.98)			
TGA	4.06 (0.85)	3.98 (0.98)			
SV	3.85 (0.85)	3.87 (0.98)			
Total	4.02 (0.85)	3.89 (0.99)			
Significant other support			*f*(1,183.61)= 1.69, *p*= .195	*f*(3,183.28= 0.88, *p*= .454	*f*(3,183.64)= 0.13, *p*= .940
Simple	4.27 (0.91)	4.08 (1.14)			
ToF	4.45 (0.91)	4.30 (1.14)			
TGA	4.20 (0.90)	4.13 (1.13)			
SV	4.16 (0.91)	4.11 (1.13)			
Total	4.27 (0.91)	4.16 (1.15)			

9.6.6. Changes over time in mood

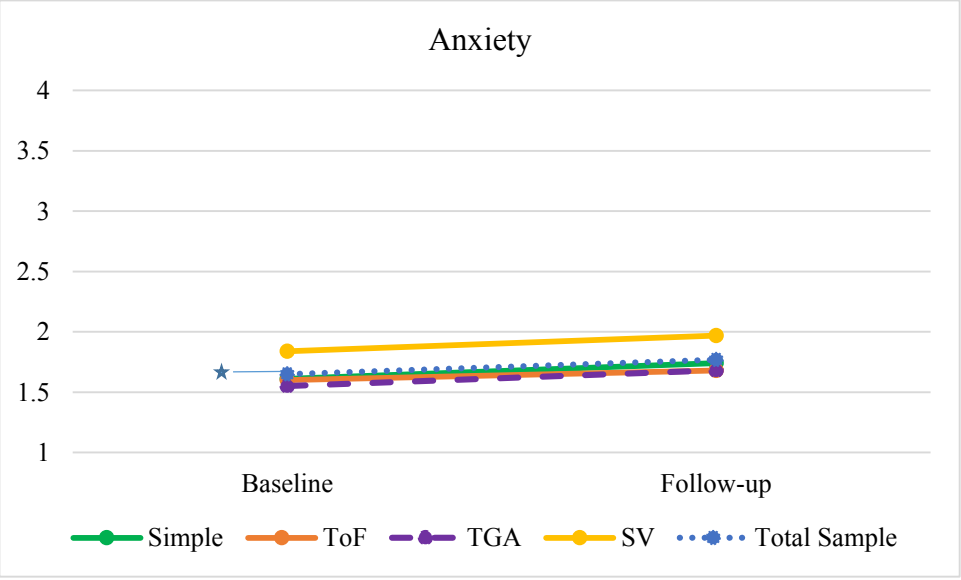
Table 9.11 presents the HLM for anxiety and depression. There was a significant time effect on levels of anxiety, with anxiety increasing over time in the whole sample (mean difference= -0.12, p= .007, 99% CI [-0.23, -0.01]; *d*= .21) (Figure 9.5). There were no statistically significant interaction effects between diagnostic

group and time or main effects for time on levels of depression, indicating no change over time and across diagnostic groups. See Appendix Y for a graphical representation of depression over time.

Table 9.11. HLM: Adjusted means (*S.D.*) and effects for mood (*n*= 188)

Mood	*Baseline (Mean, S.D.)*	*Follow-up (Mean, S.D.)*	*Effect of Time*	*Effect of Diagnostic Group*	*Effect of Diagnostic Group x Time*
Anxiety			*f*(1,183.63)= 7.41, ***p*= .007**	*f*(3,183.32)= 2.72, *p*= .046	*f*(3,183.66)= 0.12, *p*= .951
Simple	1.61 (0.56)	1.74 (0.62)			
ToF	1.60 (0.55)	1.68 (0.62)			
TGA	1.55 (0.55)	1.68 (0.62)			
SV	1.84 (0.55)	1.97 (0.62)			
Total	1.65 (0.56)	1.77 (0.63)			
Depression*			*f*(1,183.30)= 4.30, *p*= .039	*f*(3,183.37)= 3.71, *p*= .013	*f*(3,183.33)= 1.29, *p*= .280
Simple	7.46 (4.95)	7.42 (5.26)			
ToF	5.57 (4.95)	5.80 (5.25)			
TGA	6 (4.94)	7.04 (5.24)			
SV	8.17 (4.96)	9.69 (5.24)			
Total	6.83 (4.99)	7.49 (5.29)			

*HLM analysis was conducted on the continuous scale due to convergence issues.



Note. Star indicates significant main effect for time in the total sample

Figure 9.18. Graphical representation of change over time in anxiety

9.7. Discussion

9.7.1. Changes over time in clinical factors

No changes were observed over time in the clinical factors measured, including the number of interventions, number of days spent in hospital, and number of medication. These findings suggest that this sample of people with ACHD was clinically stable over the assessment period. This would have been expected as participants in the longitudinal study were not specifically assessed following a major clinical event, including changes in the treatment regimen, further interventions, or hospitalizations, especially as disease progression and reoperation timing varies even between people with the same diagnosis (Warnes et al., 2008). In addition, the length of follow-up may have been too short to allow for significant disease progression to take place. This enables any changes in HRQoL to be assessed against a background of clinical stability.

9.7.2. Changes over time in HRQoL

Overall, no changes in generic or disease-specific HRQoL were observed suggesting stability in HRQoL in this population over the time period of the follow-up. Three previous longitudinal studies in ACHD have also reported minimal changes in HRQoL. A small study in one diagnostic group examined changes in HRQoL as measured by the SF-36 over nine years in 35 individuals with TGA who had undergone a Mustard operation (Ebenroth & Hurwitz, 2007). They found no changes in HRQoL domains with the exception of a decrease in vitality. In the present study vitality scores in the TGA group were lower at follow-up but this did not reach statistical significance. Skoglund et al. (2014) examined changes in HRQoL over three years in 103 individuals with previous RVOTO surgery (see page 29), a sample

that primarily consisted of people with ToF. With the exception of a decrease in general health, no changes in HRQoL were observed. In the present study the ToF group exhibited slightly better general health at follow-up, which was not statistically significant. Skoglund et al. (2014) did use the EQ-VAS and EQ-5D, rather than the SF-36 as in this study. van Rijen and colleagues (2005a) followed-up 243 individuals with ACHD over 10 years using the Heart Patients Psychological Questionnaire. They found increased negative emotions and decreased social inhibition (feeling unpleasant among other people) over time. These findings are not directly comparable to the present study.

The above studies examined HRQoL at a different stage to the current study. They assessed participants between their 20s to middle adulthood (30s) whereas the present study examined participants on average from mid to late 30s. The challenges transitioning from young to middle adulthood are likely to be different to those in the older group in the present study. In addition, the follow-up in the studies differed markedly from the present study. Ebenroth & Hurwitz (2007) and van Rijen et al. (2005a) had an interval of almost 10 years between the assessments with a variation in follow-up years among participants, not controlled for in the analysis as opposed to the present study. It is thus unclear whether the minimal changes in HRQoL reported in previous studies were true changes or a result of measurement limitations.

It is possible that the relatively short interval between the two time points did not allow significant changes to take place in HRQoL. This stability of the SF-36 over moderate time periods has also been demonstrated in other chronic yet clinically stable populations. A study of a clinically stable sample of people with systemic

lupus erythematosus from Canada, U.S., and the UK indicated that the SF-36 physical and mental component summary scores remained stable over 4 years (Panopalis et al., 2005). The authors argued that stability in HRQoL was due to the long-term nature of the condition, which allowed people to adapt and therefore maintain their HRQoL. Considering that the responsiveness of the SF-36 has been demonstrated in the literature (Garratt, Ruta, Abdalla, & Russell, 1994), the findings of the present study more likely suggest that no true change has taken place in HRQoL over the assessment period. This is further supported by the lack of changes in the disease-specific CHD-TAAQOL, which is believed to be more responsive to changes in HRQoL over time (Garratt et al., 1994).

Some authors argue that measuring HRQoL longitudinally is particularly challenging as a response shift may take place in the interval between the assessment points. Therefore it could be argued that the lack of change in HRQoL was a result of redefinition of the concept of HRQoL and/or change in people's internal standards and values (Schwartz & Sprangers, 1999). However, considering the relatively short follow up time period and the clinical stability observed in the current sample it is unlikely that substantial response shift effects have taken place in the study (Ahmed, Sawatzky, Levesque, Ehrmann-Feldman, & Schwartz, 2014).

9.7.3. Changes over time in psychosocial factors

The SRM postulates a dynamic nature of illness perceptions, whereby changes in the illness experience, such as change in illness manifestation (e.g. new symptoms) or the introduction of a new treatment can result in people reformulating their initial perceptions of their illness (Leventhal et al., 1998). The findings indicated that the whole sample experienced an increase in illness coherence, which suggests that

people with ACHD increased their illness understanding over the period of the follow-up. Increase in illness coherence has also been reported in people with diabetes over a period of 2 years post-diagnosis (Lawson et al., 2008). Bijsterbosch et al. (2009) also found an increase in illness coherence in people with chronic osteoarthritis over a period of 6 years, which was greater in people who did not experience progression of disability than those who did. The lifelong management of people with ACHD in a specialist clinic and the continual provision of information from healthcare professionals during outpatient appointments may have enhanced people's understanding of their condition. Specifically, individuals attending the GUCH outpatient clinic at The Heart Hospital, London typically meet with specialist nurses prior to seeing their consultant, where they are reminded about the nature of their condition, implications for their functioning, their treatment regimen, and potential future complications. However, without quantifying the amount and nature of the information given to or sought by these individuals, it is not possible to attribute these improvements directly to these consultations. Although the present sample remained clinically stable over the assessment period, the change in illness coherence may be related to other factors not measured in the study.

Some changes in illness perceptions were also observed in certain diagnostic groups. People in the Simple group showed an increase in perceptions of illness curability over time. This group also had an increase in perceptions of illness controllability compared with those in the SV group who reported a decline over the assessment period. This finding suggests that people with SV felt less in control compared with people in the Simple group who felt more in control of their condition and experienced the benefits of their treatment over the period of the follow-up. Limited

longitudinal research in other chronic populations, including people with osteoarthritis and myocardial infarction, used the IPQ-R and has also showed that perceptions about personal control decrease over time (Bijsterbosch et al., 2009; Petrie & Weinman, 1997; Sheldrick, Tarrier, Berry, & Kincey, 2006). Bijsterbosch et al. (2009) found that people with osteoarthritis who experienced progression of disability reported decreased perceptions of control in the two-year follow-up compared with people who did not experience progression of disability. Foster et al. (2008) also found that increase in perceptions of personal control was associated with good clinical outcome in people with low back pain.

The sample in the current study was clinically stable as measured by clinical events such as further interventions, hospitalization, and medication. In light of the clinical stability it is possible that some items of the Brief IPQ do not offer reliable measurement of illness perceptions over time. There are limited studies that have reported findings on the psychometric properties of the Brief IPQ. These studies reported satisfactory test-retest reliability over a short period of time (1-3 weeks) in most dimensions of the Brief IPQ, yet test-retest agreement in illness controllability and coherence was low (de Raaij, Schröder, Maissan, Pool, & Wittink, 2012; Hallegraef, van der Schans, Krijnen, & de Greef, 2013; Løchting et al., 2013). The Brief IPQ and especially the items measuring illness controllability and coherence may be further evaluated in future studies in ACHD to establish their test-retest reliability.

Perceptions about the symptoms and consequences experienced as a result of ACHD remained unchanged over the time of the follow-up. Petrie and Weinman (1997) also reported stability in these illness perceptions in people with myocardiac infarction

over time. They argued that the initial perceptions of the impact of cardiac disease are readily available to people, because they are well defined to the degree that they become lay beliefs. The beliefs people hold about the impact of ACHD appear to be similarly stable over time, which may be explained by the similarity of some symptoms and consequences between congenital and acquired cardiac disease.

It is evident that the pattern of change in illness perceptions is similar to other chronic conditions. It should be noted, however, that other chronic conditions that have their onset during adulthood (e.g. myocardial infarction) may have different trajectories in relation to illness perceptions. Initially, myocardial infarction may be perceived in acute terms and with passing time people may form more chronic illness perceptions attached to other associated cardiac conditions such as hypertension (Petrie & Weinman, 1997). On the other hand, ACHD is congenital and therefore trajectories of illness perceptions may follow a different pattern. Future studies with multiple assessments may provide further insight on trajectories of change in the illness perceptions of people with ACHD.

There were increased levels of anxiety over the assessment period, yet the effect size was rather small and the overall level of anxiety was low. The reason for this increase is unclear but it may be attributable to situational factors as state anxiety is naturally transitory (Marteau & Bekker, 1992). Notably, participants did not alter their use of their coping strategies over the assessment period or perceived a change in their social support. This implies that for people with ACHD family, friends, and significant others provided the same levels of support over the period of the follow-up. These findings may have important implications for identifying and addressing persistent engagement in maladaptive coping strategies, including self-blame which

was found to be significantly associated with HRQoL in the cross-sectional study (section 8.4.2.3, page 260).

9.8. Summary

This chapter examined changes in HRQoL and other psychosocial factors over time. The results indicated that clinical factors, HRQoL, and a majority of the psychosocial factors remained stable over time. Over the assessment period there was an increase in illness coherence and anxiety across the whole sample, increase in perceptions of curability within the Simple group and increase in perceptions of illness controllability in the Simple group compared with the SV group. These findings suggest an overall stability in the HRQoL and psychosocial functioning of people with ACHD. The limitations of the longitudinal study and the implications of the findings will be addressed in the general discussion in Chapter 11.

CHAPTER 10 – THE EXPERIENCES OF PEOPLE WITH ACHD: A QUALITATIVE STUDY

10.1. Prologue

The previous chapters described the quantitative component of the thesis. Chapters 7 and 8 described the results of the cross-sectional study examining the impact of ACHD on HRQoL and the demographic, clinical, and psychosocial factors associated with HRQoL in people with ACHD. Chapter 9 described the findings from the longitudinal study examining changes in HRQoL and psychosocial functioning over time. Chapter 4 highlighted that very few qualitative studies focused on adults with ACHD (Berghammer et al., 2006; Claessens et al., 2005; Cornett & Simms, 2014; Horner et al., 2000; Overgaard et al., 2013). These studies were not focused on HRQoL and included predominantly younger adults with more complex conditions. Living with and adapting to a chronic and progressive condition like ACHD is a complex experience and can benefit from in-depth exploration. The overall purpose of the qualitative study was to further explore the experiences of people with ACHD in relation to their HRQoL and how people adjust to living with the condition. The rationale and purpose of this mixed methods approach was discussed in Chapter 4 (section 4.5, page 128).

The present study aims to:

- Aim 4a: Explore the experiences of people on how ACHD had influenced their HRQoL in physical, social, occupational, and psychological domains over the years.
- Aim 4b: Explore how people adjust to living with ACHD.

10.2. Methodology

10.2.1. Ethical approval

Ethical approval was granted by the Joint UCL/UCLH Ethics Committee and NRES Committee London – Bentham in Ethics of Human Research (REC reference number: 08/H0715/105).

10.2.2. Design

This study utilised semi-structured interviews with participants who completed both the cross-sectional and longitudinal study. Individual interviews were selected over focus groups because the purpose of the study was to gain in-depth accounts of the participants' experiences.

10.2.3. Participants and consent

Participants who had completed the quantitative studies were eligible for participation in this study. A purposive sampling approach was followed to recruit participants from all four diagnostic groups and to make sure that gender was adequately represented.

After the completion of the quantitative longitudinal assessments, participants were informed about the qualitative study in person and their interest was recorded. Interested participants received a formal invitation letter and an information sheet (Appendix Z). Interviews were arranged for a time convenient to the participants. Written consent was obtained before the interview (Appendix AA). Participants were reminded the interviews were to be audio-recorded, confidential, and that the transcripts would not include personally identifiable information.

10.2.4. Location of the interviews and safety protocol

Participants were offered a choice of location either at home, at the hospital or university at a time convenient for them and all participants indicated their preference of conducting the interview at the hospital. Interviews were conducted in designated quiet rooms at The Heart Hospital, London. It was ensured that all rooms were supplied with telephones and that the researcher could easily contact the research team in case of safety concerns. It was also ensured that members of the clinical team at The Heart Hospital, London were aware of the time and location of the interview.

10.2.5. Topic guide

The interview schedule (Figure 10.1) was developed by the researcher in collaboration with the supervisory team. The interview schedule was designed to cover topics relating to the impact of ACHD, the ways people cope with living with ACHD, priorities, future expectations, and social support. The process of developing the schedule began with establishing the subjects to be covered by reviewing the relevant literature (e.g. Berghammer et al., 2006; Claessens et al., 2005) and further issues raised by participants in informal conversations during the quantitative studies. The subjects were discussed with the supervisory team, in terms of how they group together into broad topic areas, how questions may be phrased, prompts, and subject coverage. Two independent researchers with extensive experience in qualitative research then reviewed the interview schedule and made recommendations with regards to how it will work in practice, wording, and length.

The semi-structured approach allowed for key topics of interest to be explored, while also permitting flexibility in the order those questions were asked. This approach also allowed for new themes to emerge (Ritchie & Lewis, 2003).

All interviews were conducted and transcribed verbatim by the researcher. Brief notes were taken after each interview summarizing the researcher's reflections, impressions about the interview, the general attitude of the participant, moments of discomfort or emotional charge, and emergent themes. These notes were taken into consideration during the analysis. A detailed reflective account can be found in Appendix BB.

Introduction: Purpose of study, confidentiality, consent, demographic information

Example questions (to be used as a general guideline):

General

1. How is it to live with congenital heart disease?

Impact of ACHD

2. How is ACHD influencing or not influencing your life?
Prompts: impact on current physical health, symptoms, physical limitations, health changes over the years.

3. How much does your condition affect your social life?
Prompts: social activities, disclosure, changes over the years.

4. How much does/did your condition affect/ed your working life?
Prompts: disclosure, adjustments at work, changes over the years.

5. How does it feel living with congenital heart disease?
Prompts: ways your condition affects you emotionally, emotions experienced, changes over the years.

Coping

6. How would you deal with problems arising from your health?
Prompts: coping with symptoms, physical limitations, social limitations, and work limitations.

7. How would you deal with negative emotions arising from your condition?
Prompts: steps or approach taken to cope with negative emotions.

Future expectations

8. What are your future expectations regarding your quality of life?
Prompts: extend of health influencing quality of life.

Social support

9. Who is your main source of support?

10. How would you describe the current function of your support system?

11. What sort of support do you mainly need?

Domain importance

12. What does having a good quality of life mean to you?

13. Which area(s) of your life are most important to you at the moment and why?

14. What do you think you can do to make your quality of life better?

Closing: Additional comments, participant impressions, thanking, confidentiality.

Figure 10.1. Interview schedule for the qualitative study

10.2.6. Sample size

The approach to sample size in quantitative research, where larger numbers are generally more desirable, is not applicable to qualitative research, where the sample size reflects the depth and richness of information that describes a phenomenon (O'Reilly & Parker, 2013). Data saturation was defined as the emergence of no new themes relating to the *"pre-established conceptual categories"* or study research question and it was ensured by setting an initial recruitment target and estimating how many further interviews would be needed before no new themes emerged, and applying a 'stopping criterion' of the number of consecutive interviews that elicited no new themes or ideas (Francis et al., 2010). The present study aimed to include an initial sample of 10 participants, with a "stopping criterion" of two consecutive interviews that did not elicit any new themes before recruitment ended, ensuring an equal number of female and male participants and at least two participants from each diagnostic group.

10.2.7. Data analysis

The systematic guide to Framework Analysis (FA) suggested by Ritchie and Lewis (2003) was used to analyse the data. Framework analysis is a type of thematic analysis that allows the development of a thematic framework, consisting of main themes and related sub-themes (Ritchie & Spencer, 1994). This method was selected because it involves distinct steps and analysis may take a theme-based, case-based approach or a combination of the two (Ward, Furber, Tierney, & Swallow, 2013).

Alternative approaches to data analysis were reviewed and compared. Grounded theory (Glaser & Strauss, 2009) is concerned with generating theory for a particular phenomenon and generally requires larger sample sizes to reach data saturation and

is therefore more time consuming (O'Reilly & Parker, 2013). Since the purpose of the present study was not to generate theory, grounded theory was deemed inappropriate. Interpretative Phenomenological Analysis (IPA) was not selected because it focuses both on an individual's unique experiences and on patterns of meaning across participants (Smith, Flowers, & Larkin, 2009). The current study was not concerned with the experience of individuals per se but the experiences across the sample. IPA also generally requires a small homogeneous sample (3-6), which within the purpose of the present study was not deemed appropriate for exploring the range of participants' experiences (Smith et al., 2009). The present study was informed by previous literature and the quantitative component of the thesis and it has been argued that IPA may not be appropriate within pre-existing theoretical frameworks (Brocki & Wearden, 2006). Moreover, there is no consistency in the methodology used for IPA or a detailed procedure for conducting data analysis. Thematic analysis is a method of identifying, analysing, and reporting patterns (themes) within data (Braun & Clarke, 2006). This method was considered an appropriate alternative to FA. However, FA was selected rather than thematic analysis for three main reasons:

1) FA is considered as an explicit and straightforward approach to thematic analysis, which allows flexibility and provides a transparent path of the analytical procedures with the use of charted data summaries (Ward et al., 2013). This method was deemed more appropriate as it would enable the experienced supervisory team to oversee the analysis using an audit trail and would facilitate the discussion of ideas using charted summaries within the team.

2) FA allows for the combination of case-based and theme-based approaches to thematic analysis; this would facilitate the identifications of patterns both across different groups of cases and within a single case and allow for comparisons to be made (Ritchie & Lewis, 2003).

3) FA is shaped by existing ideas and can address specific questions. However, it enables a combination of inductive and deductive approaches; this was significant for the present study, which had specific issues to explore but also allowed new themes to emerge.

The analysis involved a number of stages and was conducted using paper-based methods and the computer software NVivo, version 10 for Windows by QSR International. Specifically, analysis involved the following stages:

1. Familiarisation: According to Spencer, Ritchie, and O' Connor (2003), the purpose of this stage is to *"gain an overview of the data and become thoroughly familiar with the data"* (p. 221). Familiarisation with the data was achieved by listening to the audiotapes, reading the interview transcripts, and studying field notes made following each interview. The familiarisation process was facilitated by the researcher's involvement in all stages of the research, including conducting and transcribing the interviews. During this step, recurring themes were identified and recorded in a spreadsheet. Generally, it is not necessary to review all the material but rather a selection of the data set (Srivastava & Thompson, 2009); however, because of the small number of interviews conducted for the present study, it was decided that all data should be included at this stage. This ensured that no data were overlooked and that less frequent themes were detected.

2. Developing a conceptual framework: During this stage the recurrent themes identified in the previous stage along with the topics introduced through the interview schedule are used to form a conceptual framework or "index" (Spencer et al., 2003). Recurrent themes were grouped under broader main themes to form an overall framework. The development of the framework was conducted using paper-based methods, where post-it notes were used to record the recurrent themes. This method facilitated the sorting and re-sorting of sub-themes under the main themes. The domains explored during the interviews were taken into consideration when developing the conceptual framework. However, a flexible approach was maintained, whereby new emerging themes were also recorded and integrated into the framework. A worked example from this analysis stage is presented in Appendix CC. Ritchie and Spencer (1994) highlight that this framework is tentative and refinement is possible during further stages.

3. Indexing the data: At this point, the interview transcripts were uploaded into NVivo. During this analysis stage, the conceptual framework developed in the previous stage was applied systematically to all of the interviews. Interview transcripts were re-read and sections of the data were indexed to corresponding themes. Emerging interconnections between themes were noted for subsequent associative analyses. At this stage, a refinement of the initial conceptual framework is common. For example, themes may be added, removed, merged, or subdivided in order to describe the data in a more suitable way (Ritchie, Spencer, & O' Connor, 2003). In the present study further immersion in the data resulted in the refinement, combination, and development of themes and sub-themes. It became apparent that

some sub-themes belonged to multiple main themes. Hence, the framework was developed to represent the data more accurately (see example in Appendix DD).

4. Sorting, charting, and summarising the data by theme: During this step, the themes are sorted so that those with similar content are located together (Ritchie et al., 2003). This involved the creation of case by theme charts, with summaries of what was said by each participant using the framework matrix function in NVivo. Charts retained the "feel" of participants' experiences by using exact quotations, where possible (Gale, Heath, Cameron, Rashid, & Redwood, 2013). In addition, references to illustrative quotations were noted within charts (see example in Appendix EE).

5. Mapping and interpretation: The final step of the analysis involves a close examination of the themes and sub-themes, whereby differences in the representation of a phenomenon are identified, patterns are detected, associations are mapped, and explanations are explored (Gale et al., 2013) (see example in Appendix EE).

10.2.7.1. Rigor and Trustworthiness

The following actions were taken to ensure rigor and trustworthiness:

1. An audit trail of the data analysis process was recorded with descriptions and explanations of decisions about conceptualization, combination, and omission of themes, the development of the initial framework, and the framework refinement. This was used to discuss the process of data analysis with the supervisory team.
2. Reflective accounts were recorded at the end of each interview, which included the researcher's feelings and impressions about the interview, the

general attitude of the participant, and emotionally charged or uncomfortable moments. These accounts were taken into consideration during data analysis. A detailed reflective account can be found in Appendix BB.

3. External validation of 25% (*n*= 3) of the interviews was conducted by an independent examiner who coded the transcripts without prior knowledge of the conceptual framework. Differences in coding were discussed until an agreement was achieved (see section 10.3.2, page 316).
4. A "thick description" approach was adopted for the presentation of the findings. Descriptive interpretations were accompanied by excerpts from the participants' transcripts, which allow the reader to verify the validity of the interpretations and potentially to consider their "transferability" to other contexts (Ritchie & Lewis, 2003).

10.3. Results

10.3.1. Sample characteristics

Seventy-eight participants were eligible to take part in the study and were approached by the researcher between January 2013 and January 2014. Twenty-four participants agreed to take part in the study. Twelve could not attend an interview prior to the completion of the study due to other commitments. A total of 12 semi-structured interviews were therefore conducted. This is considered an appropriate sample size for mixed methods studies (Francis et al., 2010). Importantly, at this point no new themes were emerging, hence data saturation was achieved. Interviews lasted between 30 minutes and 1 hour 42 minute, allowing participants to elaborate on their experiences as much as they wished. Basic demographic and clinical characteristics of the participants are presented in Table 10.1. Pseudonyms were assigned to each participant to protect their identity.

Table 10.1. Characteristics of participants interviewed

Participant Pseudonym	*Age*	*Gender*	*Diagnostic Group*	*Marital Status*	*Employment Status*
Anastasia	45	Female	TGA	Married	Unemployed
Maria	25	Female	SV	In a relationship	Employed
Doreen	37	Female	SV	Married	Unemployed
Chloe	36	Female	TGA	Married	Employed
Janis	35	Female	SV	Married	Employed
Sophia	27	Female	Simple	In a relationship	Employed
Ron	54	Male	TGA	Married	Employed
Phil	37	Male	ToF	Married	Employed
Robert	40	Male	TGA	Married	Employed
Leo	32	Male	SV	Single	Unemployed
Jim	36	Male	ToF	Married	Employed
Peter	39	Male	Simple	In a relationship	Employed

10.3.2. External validation of coding

The coding by the independent researcher was compared to the coding conducted by the current researcher and no major inconsistencies were observed. The only discrepancies were the codes “impact of treatment” and “information from healthcare professionals” assigned by the independent researcher. After a discussion it was agreed that “impact of treatment” had been captured in the original conceptual framework as “physical and social consequences” and “information from healthcare professionals” had been captured in the original framework as either “seeking social support” or “receipt of social support”.

10.3.3. Themes

A thematic map was developed during the final stage of the analysis (Figure 10.2). Three overarching processes were identified within the data; the first reflected the

narratives regarding the impact of living with ACHD on people's HRQoL and the second reflected the narratives regarding the processes through which people coped with ACHD. Embedded into these two processes was the overarching theme of change over time which reflected the narratives about the impact of changes in health and treatment over the illness trajectory.

Four main themes were identified within the coping process: 1) priorities and goals, 2) acceptance and adjustment, 3) gaining perspective, and 4) social influence. Four main themes were identified within the impact of ACHD: 1) physical and social consequences, 2) occupational consequences, 3) illness disclosure, and 4) psychological consequences. Within some of these themes, sub-themes were identified (Figure 10.3).

Change over time was an over-arching theme that reflected the dynamic facet of living with ACHD and ran through many of the participants' narratives. People appeared to be in a dynamic state, where periods of health deterioration and change in the treatment regimen influenced both their experiences about their HRQoL and the ways they coped with ACHD. These changes in health over time made the physical, social, occupational, and emotional consequences of ACHD more salient for some participants and challenged their coping efforts, specifically their attempts to accept and re-adjust and gain perspective. This dynamic facet of living and coping with ACHD was also evident during major changes across people's adult lives, including establishing, maintaining, and leaving employment and family planning. Adjustment was therefore not an endpoint but an ongoing process across the illness trajectory. The overarching theme of change over time was inherent in the processes of coping and the impact of ACHD on HRQoL and not independent of the themes

and sub-themes identified in Figure 10.3. Therefore, it is described in terms of the impact of changes in health and treatment within the relevant themes or sub-themes, which are presented in the following sections. Illustrative quotes are provided along with the themes and sub-themes to support the interpretations.

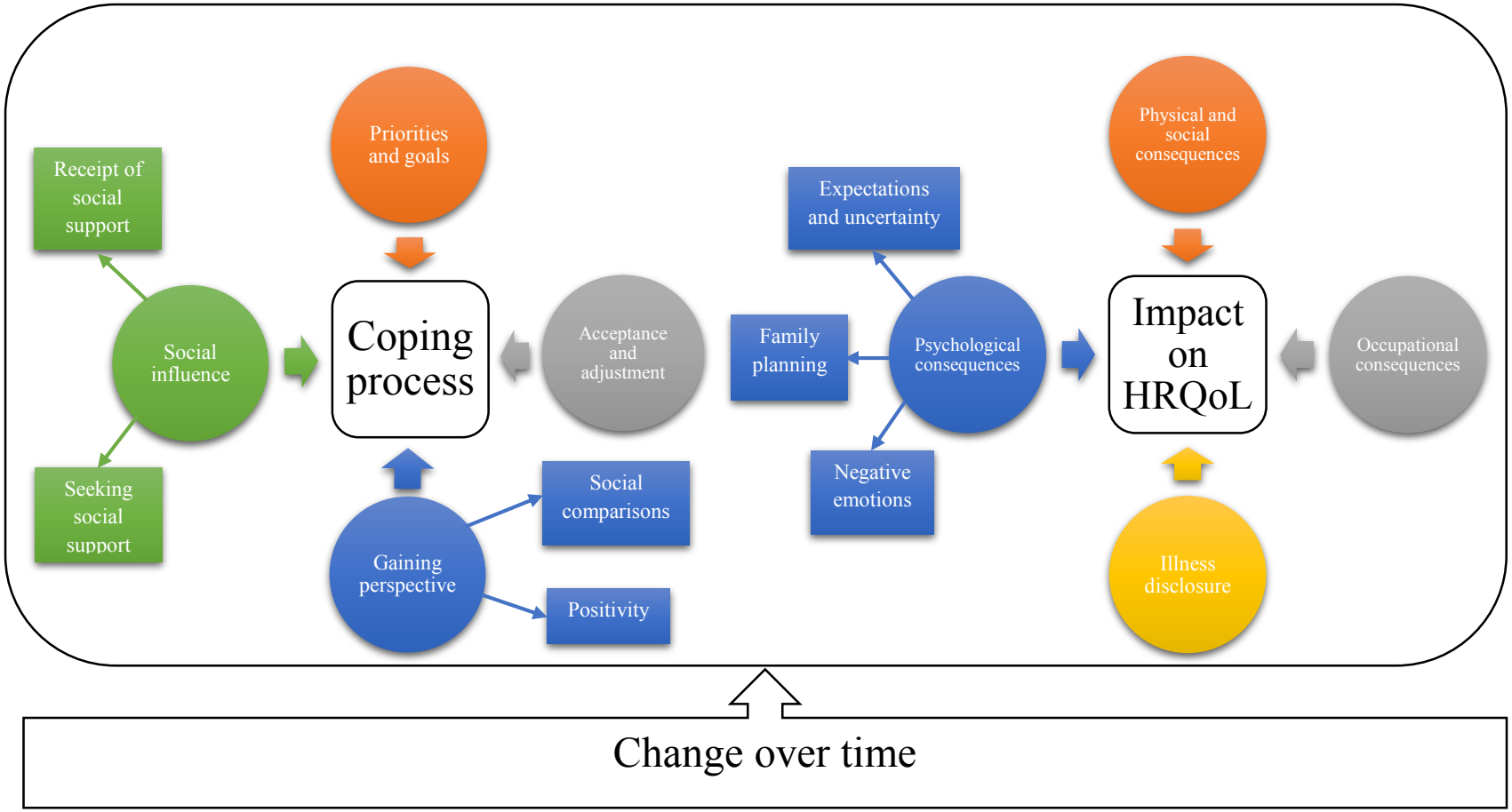


Figure 10.2. Thematic map of the three overarching processes and the eight main themes within these processes

Theme 1: Priorities and goals
Theme 2: Acceptance and adjustment
Theme 3: Gaining perspective
Sub-theme: Social comparisons
Sub-theme: Positivity
Theme 4: Social influence
Sub-theme: Receipt of social support
Sub-theme: Seeking social support
Theme 5: Physical and social consequences of ACHD
Theme 6: Occupational consequences of ACHD
Theme 7: Illness disclosure
Theme 8: Psychological consequences of ACHD
Sub-theme: Negative emotions
Sub-theme: Family planning
Sub-theme: Expectations and uncertainty

Figure 10.3. The final themes and sub-themes identified during framework analysis

10.3.3.1. Priorities and goals

Most participants talked about their health being a priority to maintaining their QoL, in terms of *"being able"* to do the things they wanted to do within their limitations and without being *"inhibited"* by their heart condition. Health and physical ability was a recurrent theme across many of the interviews, followed by having a job that provided financial stability, *"being happy"*, and maintaining social relationships.

> *"Just your health. It's got to be your health comes first. And then just enjoying your family, enjoying your social life, and again work. You got to enjoy, to me, if you don't enjoy work don't do it." – Ron, 54, TGA*

> *"Being healthy. Being able to sort of go out do whatever you want sort of thing. Not being worried about sort of money stuff. Just yeah just sort of have the freedom to do whatever you want without any sort of restrictions." – Phil, 37, ToF*

Some individuals, for whom health issues appeared to be less of a concern, talked about prioritising their work and personal lives and keeping a *"balance"* between the two. One person, who prioritised his personal life, appreciated that health may become the focal point when it deteriorates.

> *"…I've diverted far more personal time towards my relationship rather than friends and other areas of my life […] It [health] isn't an issue 'till it becomes an issue. You don't think about it until it goes wrong. You take it for granted until then and then when it does have an impact then everything has to be built around it." – Peter, 39, Simple*

Most participants felt unsure about whether they could improve or control their QoL. Some felt that at times, their health was *"in control"* of them rather than the opposite.

> *"I feel like the only control I have is making sure that…my body is as good as it can be. So I do keep walking and I do keep my weight off. The other parts of my life, I have total control over as much as my heart condition lets me, if you like." – Janis, 35, SV*

These perceptions were accompanied by descriptions of how they prioritised and directed more attention and efforts towards their health, by engaging in healthy lifestyle behaviours, including controlling their eating habits and exercising. Some participants emphasized that keeping healthy was important to them because they wished to *"be there"* for their families.

> *"But I think right now it's probably a physical point of view because I wanna be there for my family, I wanna be there for my son, I wanna be able to do things with him." – Jim, 36, ToF*

"I'm putting a lot of input into my health at the moment. I mentioned before that over in the last three years I've lost a lot of weight and I got to the point where I thought that logically a smaller body mass means your heart has to work less hard. So I did lose three stone and made myself better that way." – Janis, 35, SV

10.3.3.2. Acceptance and adjustment

Across many of the interviews there was an underlying narrative that participants had accepted their condition and achieved some normalcy in their lives, being able to carry on with *"day-to-day living"* despite limitations resulting from ACHD. They talked about how they have *"always lived with it"* and *"got used to not feeling 100%"* and how they had accepted their condition as being part of their lives and their identity (*"who they are"*).

"Accept it. It's there, that's the way it is. It isn't going to change so it's no use getting upset over it. I suppose that's how I emotionally deal with it I just say "Well it's not gonna change so."" – Leo, 32, SV

"…you can make a big deal out of it if you want to or you just accept it as being there and you deal with it if issues come up. And I think that's what I've tried to do with my life…" – Jim, 36, ToF

They described how they *"get on"* with their condition and their lives by *"knowing their limits"* and being *"realistic"* about what they were able to do. Participants described that they found ways to cope with their limitations by *"planning"* ahead, *"pacing"* themselves when they did something physically demanding, and *"avoiding"* situations, which may be challenging for them.

"I know my limits if you know what I mean. I don't run around, I can't play football or I don't go to the gym or anything like that but yeah I get by. If there's something I

feel I can't do, I just won't do it like just, I'll just sit out of something […] there are certain things you sort of limit yourself, you know what yourself you can do" – Ron, 54, TGA

"I just focus on what I need to do at that right moment in time, just take a step back and just think about it a little bit further whether I can do or not […] just focusing on certain house chores I would do, but I will do it and I'll have a rest afterwards so I just pace myself to do it…" – Robert, 40, TGA

"There's always gonna be limits. That's just parts of living with a heart condition. And it's just finding ways of them not being a problem. So we don't go to India for holiday. We go somewhere closer. That's, to me that's just common sense." – Janis, 35, SV

"I probably just think of strategies around it, sort of saying, just find a way of not having to face a particular issue if it was going to be a problem for me. I guess if you take going to a holiday of altitude, rather than sort of pushing on through it and seeing how I go, I would just not really go on that holiday, cause it might be a problem and I don't wanna get into a problem situation." – Peter, 39, Simple

Participants also characterised their treatment regimen, including follow-up appointments and diagnostic tests as *"routine"* and *"second nature"*. Those who had interventions during adulthood also talked about how these had helped them get *"back to normality"*.

"So used to them [hospital visits] now, it's no problem at all. You're sort of used to the people, what, the various things that you have to do. Sort of, it's like a routine, you just get used to it." – Phil, 37, ToF

Most people talked about how they had learned to successfully manage their limitations and integrate their treatment regimen into their lives. Some described how they attempted to adjust to changes in their health and treatment regimen. A younger person, who was experiencing recent health deterioration, talked about how she thought of her condition as a *"barrier"* to achieving a previous state of functioning. She described her experience of how she struggled with the limitations caused by her health changing and the process of gradually accepting and re-adjusting her plan to work around her limitations.

> *"I'm always striving to get back to that point where I could part-time work, be at Uni, and be physically active. That's what I aspire to be able to do again and I find it hard that I can't […] So I'm trying to come to terms with the fact that there are certain jobs I can't do, there are certain things that I'm not gonna be able to do but that's not the end of the world. I'm just gonna have to change my plan. I struggle a bit with a change of my plan…" – Maria, 25, SV*

Similarly, another person described the process of adjusting to a recent change in her treatment regimen and specifically the introduction of Warfarin medication and the installation of a pacemaker as part of her daily life.

> *"About three weeks ago I had a pacemaker fitted so I'm kind of being a bit careful with that. So just kind of sort of getting used to things […] When I first started my Warfarin I was like "Oh, be careful of what I eat". I was just like stop. They tailor it to you, you do whatever you like, if it's different one week then it's different. Don't try and make it the numbers. They'll make the medication match what you're doing and it took me about two months to actually get that kind of balance. But I'm still kind of like thinking about what I'm drinking […] "I've got a pacemaker, does that*

mean I can't drive? Or I can drive? Do I not drive? Do I tell them?" That sort of thing so yeah it's the admin bit." – Chloe, 36, TGA

10.3.3.3. Gaining perspective

Participants described how they often tried to find meaning in their experiences of living with ACHD and gain perspective. One individual described her experience of putting things into perspective despite the significant impact that her heart condition had on her life.

"There are times when it [heart condition] feels like the only thing in my life and it's really overwhelming and I can't deal with it. But if I'm looking from the outside […] if my heart was fine then I would have a sickly perfect life. So I suppose, in some ways you can't have everything. And I've got nice things. So there, that's a bit sickly [laughs] but it's true." – Janis, 35, SV

<u>Social comparisons</u>

Some people engaged in "downward" comparisons, describing how their situation could have been *"worse"* and that comparing themselves with other people had helped them gain perspective and adjust to their condition.

"I've met up with people who have heart conditions as well and they were born with it, it affects them differently because it's a lot worse so. It depends on severity but I just think that you need to kind of realise what you've got and understand what you've got and just try to have the best life you can." – Jim, 36, ToF

"I just feel like I can still get around and I know there's worse people off than me that got the same condition as me so I'm just happy and glad that I can do what I can do basically." – Ron, 54, TGA

Others engaged in "upward" comparisons, describing how they gained perspective by comparing themselves with people who coped really well despite being in a more difficult situation than them.

> *"I met a man, when I went in for my pacemaker and he'd had, he was on like his 14th pacemaker or something. He had his 1st when he was 6 months. And I spoke to him and he was the cheeriest person ever and I was like "Wow, you've been through loads". Me I only had one lead, that's not really anything, is it? It's like I'm gonna have to put things into perspective […] And I'm like "Oh ok. So this isn't the big cloud."" – Chloe, 36, TGA*

Positivity

Gaining perspective was also linked to feelings of luck and gratefulness. Some people believed that they were in a fortunate position and appreciated that they were *"lucky"* to be alive and to have reached this far in their lives having being born with congenital heart disease. Some female patients felt lucky and *"privileged"* to have got married and to have had families of their own.

> *"…I do feel very lucky […] I think time-wise from when I was born I was very lucky that I was, quite at the early days of babies being cured with this condition […] I suppose just emotionally, just that the fact that, yeah, the realisation that I am here and that I was a lucky one." – Anastasia, 45, TGA*

Others described how they tried to reframe their situation in more positive terms by *"adjusting negative motions into positive ones"* or how they generally tried to keep a positive outlook.

> *"…in everyday life I'm really positive about it. Tried my best, well I do, I make the best out of my situation as much as I can, I'm trying to think outside the box." – Maria, 25, SV*

Positive outlook and seeing the positive aspects of their situation helped some people cope with their treatment demands, for example operations and the need to attend regular outpatient appointments.

> *"Well I think I'm quite a positive person... I always had a positive outlook so I don't know, but I imagine it probably just helps you make better recovery and more speedy, cause you're like "Oh it'll be fine, it'll be fine"" – Sophia, 27, Simple*

> *I'm a bit of a positive person really [chuckles]. I don't like to really dwell on negative sides... I sort of twist it to make it more enjoyable, having to come down to London to visit the hospital every once a year or every couple, twice a year. I sort of, I don't mind it a great deal… It's a good excuse to, to venture down London and shop and see the sights and go round the clubs and that sort of things. So I try and treat it as positively as I can. – Leo, 32, SV*

For some participants, who experienced a change in their health, maintaining a positive outlook and reframing the negative into positive was difficult due to uncertainty about the future. They talked about how they had actively sought counselling in order to cope with uncertainty, which subsequently had helped them achieve a positive outlook, see the big picture, and appreciate other aspects of their life.

10.3.3.4. Social influence

Although social support was intended as a pre-determined topic to be tackled during the interviews, the role of people's social environment was a recurrent and

unprompted theme in all of the participants' narratives that was often linked to other themes pertaining to the coping process. All participants identified their social environment, including their partner, family, and friends, as a beneficial resource that helped them cope with their changing condition and its treatment (seeking social support). The nature of social support was further elaborated under the sub-theme of receipt of social support.

Receipt of social support

All participants talked about having a supportive environment, which helped them cope with their condition. They also described how they were receiving the right type and amount of support they needed. Those who were married or in a relationship talked about how their partner provided them with both instrumental and emotional support. They referred to their partner as being *"always there"* and willing to talk about anything that worried them about their condition. Some mentioned that the experiences they had shared with their partner had brought them closer together, to the point where their support was *"unsaid"*. Others commented on how their partner was *"practical"*, while some people used the terms *"confidence-building"* and *"encouraging"*.

> *"…emotional support and physical support and that he'll just do stuff so he knows I don't have to do it. And that's real support. Just get on and do stuff and it's, and I don't have to say to him "I'm tired". He will say "You look tired" or "I'm gonna take our son out so you can have rest". So I don't have to go to him all the time and say "I really need some time out". He'll just get on and do it. And I think that support is just unsaid but brilliant. And that helps me." – Doreen, 37, SV*

> *"…he'll be suggesting like sort of to go out and encourage me to go out and ask me how it was like at work, and encourage me to go back to work. But equally if I can't do stuff he took, he worked from home the week after I had my pacemaker put in. So he was sort of preparing the food, more going out and taking the take-aways." – Chloe, 36, TGA*

Some commented on how their partner helps them maintain a positive outlook.

> *"I think he's [husband] the positive to my negative. So it works quite well cause actually he'll say, you know "It was a positive appointment. You know, they said this, and this, and this and to me that is really positive"…I suppose he balances me out." – Janis, 35, SV*

Most people also talked about how their parents and especially their mother were concerned about them and provided them with emotional and practical support when they needed it. They commented on how their parents had been there throughout their lives and how they were *"still involved"* in their healthcare. A few individuals described their mother as *"fussy"* but mentioned that they preferred that approach and that they *"understood"* their concern.

> *"I know it worries them [parents] and they're always "Oh how did you get on today at the hospital?" So they're always concerned about, or want to know what's going on during the day and how I'm feeling at the end of the day." – Robert, 40, TGA*

> *"Well I think my mum is quite, she had always been quite involved anyway, but she's like, she's not like more overly involved in with me than she is with my sister […] my mum does worry about us but I think that was probably something she had anyway without, maybe the medical things have made it more pronounced than it would have been, I don't know, but yeah, I guess I'd rather have somebody who was too fussy than not fussy enough I think." – Sophia, 27, Simple*

> *"…my mum's approach is just been built up over the years. She needs to constantly check and, so that's built upon the years because of the worrying and that but she tries to tame it back a bit now." – Leo, 32, SV*

Many people described how their friends were understanding and accommodating of their physical limitations by adjusting the group activities based on the individual's abilities. They felt able to talk to their friends and be emotionally supported.

> *"…they [friends] accept it and they do things to me, really, you know "And myself are gonna do this. Will you be able to do that?" and I'll say "Yeah". If I say no they'd go "Oh we're not gonna do that, we'll do something else."" – Ron, 54, TGA*

One person, whose friends also lived with other chronic conditions, commented on how they all supported and *"kept an eye out"* for each other.

> *"I've got other friends who've got conditions where they can't go out at times because they feel unwell so they understand when I don't feel, when I say "Well I'm not coming out, I can't go cause I'm feeling". They will all be understanding cause they're all on the same boat at times […] they all are relatively sympathetic." – Leo, 32, SV*

<u>Seeking social support</u>

The availability of social support encouraged people to seek help when needed. Many of them commented on seeking support from their environment as a way of coping with their condition. Some also talked about contacting their healthcare professionals first in order to obtain information and reassurance about their condition and treatment.

> *"I phone the nurses first. So usually I sit down and I'm like "Right. Is this different, is this normal?" And then if I'm worried I'm like "Well I must be worried about*

something" […] You know, there is always somebody at the end of the phone during the week […] very factual but they are quite good at sorting out how you feeling and sort of just calming you down." – Chloe, 36, TGA

Others described how they turned to friends and family for emotional support.

"…my family and I are really close and I've got lots of really good friends […] I think usually if you have something you worry about, you can usually talk about it with somebody so, and then find an answer through talking." – Sophia, 27, Simple

"I just cope, I don't know how. Well, I talk a lot with my wife about vivid things related to my health or and well I always talk about things like that yeah. I'll let my emotions come out to my wife more than anybody else I suppose." – Robert, 40, TGA

Although all participants acknowledged that having support was very important to them, some pointed out that they sometimes prevented themselves from seeking support from their family because they did not want them to *"worry"*.

"I'm probably less willing to let mum know about any problems now because she worries and I suppose as I got older I've appreciated what that worry means so. But she is always there to pick up the pieces." – Anastasia, 45, TGA

"You hide things, you hide your feelings as much and sometimes from your partner, you know, you don't want them to worry. You're worrying but, you know. I guess there is two ways you can deal with that. Sharing it helps you but sometimes actually "Do I want her to worry as much as I'm worrying about it?"" – Jim, 36, ToF

10.3.3.5. Physical and social consequences of ACHD

Most participants described their general health as *"good"*, *"pretty good"*, *"quite good"*, or *"alright"*. Some mentioned that their health was *"not too bad"* or *"fairly good"* stating weight issues and fitness levels as the main reasons for dissatisfaction.

> *"Fairly good I think. I'd like to be fitter…I'd like to lose some weight and be fitter which I think it would probably be general mid-forties mom thing." – Anastasia, 45, TGA*

Most people described a range of symptoms associated with their condition. The most common symptoms were shortness of breath when exerting themselves, an overall feeling of tiredness, headaches, light-headedness, and dizziness. One person characterised her condition as *"my heart poorly"*.

> *"I noticed when I try to do a lot of running or something like that, I do find I get out of breath really quickly." – Jim, 36, ToF*

Some people cited *"feeling hearty"* and having a *"racing heart"* which resulted in hospitalization. One person described a recent occasion when she had to be taken into hospital because of an acute arrhythmic episode.

> *"When I had my suspected TIA I was literally, I was just walking down the street….I had really bad double vision, like literally somebody had taken my eyes and stuck inside razors….And they phoned me an ambulance" – Chloe, 36, TGA*

Some participants described how their symptoms had progressed through the years and had made them realise that their health *"becomes a big deal"* as they aged.

> *"I suppose really it's been in the last 10 years that things have started to, obviously I'm getting older, and my heart's getting older and things have started to happen perhaps that weren't really thought about before then." – Janis, 35, SV*

The experience of symptoms, particularly tiredness, shortness of breath, and arrhythmias were consistently linked with physical limitations. The majority of participants described being physically limited by their condition in the type and amount of activities they could do and their engagement in sports and activities such as swimming and running.

> *"I get short of breath, I can't run too far. I've done my exercise last year on the running machine. So I can't run too far. I like to do a little bit of swimming if I go out with my family….I'll do a couple of whips or a couple of them swimming and that would be enough for me then but I find that…I get out of breath easier." – Ron, 54, TGA*

Some people compared themselves to their peers and described that since their childhood, they *"struggle to keep up with others"* due to their limited exercise capacity.

> *"It's been hard all my life cause I was watching children who were physically able to and I wasn't […] I've caught up in my own time I should say so. On the physical way I could never compete with anybody anyway I mean my brothers' been, are always physical and they wanted to do a lot of things… I wasn't able to do what they could do. I still can't now, you know. Growing up with, still growing up with it, I think there's "Oh I can't do that"" – Robert, 40, TGA*

A few people mentioned that there was variation in what they could do day-to-day depending on their health and how ill they were feeling. A few people indicated that the weather could have a *"massive effect"* on their ability to do things.

"…my health just dips and peaks and I find that difficult because one day I'll be able to do something, the next I won't so….So one day I'll be able to do something, I'll be able to walk the dog, the next day I won't be able to walk the dog." – Maria, 25, SV

"Some days I don't feel like I've done everything I've wanted to do. But I think is also cause things just takes so long to do but also cause I know I don't want to run around like a lunatic cause I'm tired as well…I feel it more definitely, those limitations in the winter when it's just harder to do stuff." – Doreen, 37, SV

It appeared that for some people the degree to which symptoms and physical limitations had influenced their lives was linked to the experience of health decline which resulted in a significant impact on their lives.

"I'm noticing exercising, walking, going up the stairs gets me a lot way out of breath. Now that could well be the effect of my, my valve is leaking more now." – Jim, 36, ToF

Most people described that their physical limitations had, at times, interfered with their social activities. They described occasions when they felt *"exhausted"* and *"not physically able"* to go out, or occasions when they had to leave a social gathering because they were having *"a particularly bad day"*. There were some people, who talked about being unable to engage in social sporting activities that they enjoyed, for example mountaineering, climbing, and swimming.

"I used to really enjoy climbing, swimming, I can't really do them anymore. I think socially it's changed a lot. It's been a lot less active my social life." – Maria, 25, SV

The treatment regimen and particularly medication appeared to influence people's social lives as they needed to regulate their alcohol consumption and their eating

habits. One person said that taking Warfarin had *"ruined [his] drinking sessions"*. Some talked about their condition and treatment interfering with holiday planning.

> *"I want to go to Italy […] and just having all my admin in order. Whereas before I didn't have any pills and I didn't have a pacemaker and stuff like that. So that's all kind of new for me and I was like "Ok. So don't forget of what I eat. When do I get my blood tests before I go?"" – Chloe, 36, TGA*

Others reported that their condition had not influenced their social life and their activities. However, these persons described their hobbies and activities as *"light"* and not physically demanding.

> *"I haven't noticed it to affect my ability to tap dance so I'm still as bad or as good as I was before […] I mean playing the violin isn't that, quite so athletic, I suppose" – Sophia, 27, Simple*

10.3.3.6. Occupational consequences of ACHD

There was a common narrative across some people who were in employment at the time or who were employed in the past, that their condition influenced their work life. They talked about reducing their working hours because they were "tiring" themselves and it was *"extremely difficult"* to remain in full-time employment.

> *"I've reduced my hours purely as well as on that basis as well as being the area of work where I was working was becoming more. So it was no way I could do that full-on anyway. Cause the warehouse where I was in, was, some of the products were removed and other products within our warehouse which were too physical, manhandling things so it was. I'd be doing that for a 9-hour day, as early as late, as early as late, so too much for me to be honest." – Robert, 40, TGA*

These people also described having to adjust the nature of work that they were doing, for example reducing manual or more physically demanding work because of their condition and symptoms. One person described her experience of how she needed to adjust her work responsibilities because of a sudden health deterioration.

> *"…when I was going they were saying my heart rate was quite low with my heart monitor stuff. They were like "Have you passed out?" And because I work in a laboratory that's not great. So there was a period of time when they were like "Oh, you don't go in the lab". So then I was just sitting on my desk, "Oh yeah this is fun", just doing sort of menial desk-based stuff. Not actually the job that, part of my job, but not the job that I wanted to do." – Chloe, 36, TGA*

Some female participants who were employed in the past talked about their decision to retire after having their baby because they felt it was too difficult to *"manage"* motherhood and working.

> *"…then six months down the line I put her [baby] into nursery and went back to work part-time and I think actually I should have given myself more time. So as the years went on I think that sort of built up 'till in the end I just thought "I can't, can't do this anymore, I need a proper break" in terms of a year, or years rather than just months, you know. So I suppose in that way my heart condition yes, did affect the decision but indirectly really." – Anastasia, 45, TGA*

Some people also described that their condition limited their choice of work in that they needed to find a *"flexible"* job that would be accommodating to their condition and their limitations.

> *"I probably, in today's world, they wouldn't accommodate someone like me that has to have medical appointments and go sick. I tend to get flues and colds probably a bit*

quicker than anyone else […] so I just went into jobs that I could do easy which is driving." – Ron, 54, TGA

Another younger individual, who was starting her first job after finishing university, described her job responsibilities and how these were adapted for her condition and treatment demands.

"It's [job] part time, it's twenty hours a week and I can choose my hours, which is really handy, because obviously with hospital appointments and some days I'm not very well and things like that. It'll just be going in and doing group activities, like quizzes, bingo, little bit of sort of exercise that oldies can do. So nothing [chuckles] strenuous, sitting on a chair, which is my kind of exercise and I can do that." – Maria, 25, SV

Some people mentioned that their condition did not influence their working life and that the only occasion that it did interfere with their work was when they had to attend their outpatient appointments.

"The only time it [condition] affects it [working life], it's when I have to take time to come to hospital for appointments. Obviously that's not very often so minimal impact I would say." – Phil, 37, ToF

10.3.3.7. Disclosure of illness

There was an underlying narrative across all interviews that disclosure of their condition to close friends had not been an issue. Participants highlighted the importance of disclosure as opposed to leaving people to *"guess"* why they could not do a particular task. Others mentioned that their thoracotomy scar was evidence that they had undergone some sort of surgery and therefore could not hide it from

their social circle. One person emphasized the safety aspects of disclosure especially in emergency situations, where people would need to know.

> *"I've always been really honest. It's like "This is me". And, you know, I think it's really important, you know, it's not fair on them, to go out with people and them not know […] it's not fair to put them in a position where something might happen to me." – Janis, 35, SV*

One person acknowledged that disclosing his condition had become easier in adulthood because adults were more interested and understanding.

> *"It's a lot easier to tell people as you, when you're older. People understand more, wanna listen more. I think it's difficult when you're younger. You know, you're walking around with scars on your body […] when you get older you're more mature, you're more concerned and stuff like that. So it's just easier to disclose it. It's nothing, there's nothing there that I need to hide and I don't feel that I need to hide it." – Jim, 36, ToF*

A few people found it *"hard"* to disclose their condition to new friends or acquaintances. A younger person described that she had difficulty in finding a *"balance"* of when or how to disclose her condition and engaged in cost-benefit decisions about disclosure. She described some social situations, where she *"struggled"* and worried about how other people would perceive her.

> *"When you first meet people, obviously, I don't wanna be like "Oh by the way I've got a heart condition". That's not appropriate. At the same time when they're like "Ok, let's just go walk to this place" and there's a big hill, and I think "Oh I can't walk there I need to get a taxi, or a lift". And then I'd start think "Oh but they're just gonna think I'm lazy. I don't want people to think I'm lazy". Or when I have to sit down, if*

there's music on, cause I can't and they're like "Oh no, come on, come on dance" and I can't dance but I don't want them to just think I'm boring." – Maria, 25, SV

The majority of participants described their experience of disclosing their condition to their employer as *"fine"* owing to the fact that they had to fill in forms or undergo medical tests. Most of them appreciated that they had to be *"honest"* and *"open"* with their employers. They also referred to the importance of disclosing changes in their health that could potentially affect their work.

"…when I know that things are possibly gonna get worse then you tell them that "I'm gonna have to go for a lot more tests and it's gonna involve obviously having days off. And there's a possibility at some point where I'd need to have quite a bit of time off if I have an operation."" – Jim, 36, ToF

Some people mentioned that they were unsure that they would secure the job after disclosing their condition due to fear of discrimination.

"I'm not sure I've disclosed it as much as I should have, like on reflection and in all honesty. I've told them of my heart condition, I told them I need, I will have a lot of hospital appointments, but I've said that I don't think it will affect the job, which I'm not sure is entirely truthful. But I think if you go in there and lay it all down, I don't know. I wouldn't give me a job." – Maria, 25, SV

Another individual wanted her condition to be *"private"* and was particularly careful not to get herself into situations where she experienced symptoms, especially shortness of breath, in front of other people. She described her experience of not disclosing her condition to her work colleagues until she actually had to.

"I remember at my old job, old, old job we went skiing. And I didn't say anything cause like I know what is skiing and I didn't think it would be that bad but basically

it was a nightmare when I went to a height-like ski up the hill. And I was like "Actually dudes I can't, I can't do this". And they said "Oh well why didn't you say anything?" I was like "Well, cause it's not what I actually need to talk about it."" – Doreen, 37, SV

10.3.3.8. Psychological consequences of ACHD

Negative emotions

Many people talked about how their physical limitations affected them emotionally. They mentioned that not being able to do what they wanted, made them feel *"down"* and *"frustrated"*. Many expressed a wish that they could do more and *"achieve"* what they wanted regarding their family role, social participation, and work.

"So you feel a bit, the thing that you can't join in as much sort of thing. You feel a bit down with that side of thing. You can't run around so much with your grandchildren. And my kids as they were growing up as well. […] I can honestly say I've never been depressed over my heart condition. I've been a bit, like sometimes when you can't join in on certain activities, you're a bit "Oh I wish I could do that, I wish I could do this."" – Ron, 54, TGA

"…you're out with friends and you, just it's time to go home because you're absolutely shattered, everyone else will be moving on somewhere else, it's "I can't, I need to go home, I'm really tired". And so it can be frustrating when you're enjoying yourself out and you just feel too tired and you go home so. Emotionally it can be frustrating any time." – Leo, 32, SV

One person, who did not experience significant physical limitations except for extreme sporting activities, felt *"resentment"* when he was unable to engage in these activities.

> *"I think if you are physically limited and unable to take part in sporting activities and social activities, things that you want to […] there's always a bit of resentment that you are unable to do what other people take for granted" – Peter, 39, Simple*

Some individuals, who were experiencing a change in their health, described feeling *"depressed"* at times and worried.

> *"I think where I've had an issue in the last couple of months...where I had all the problems with, possibly with the heart getting worse […] it'd become a bit too much for me around November time and I'd been depressed and a bit down. But things are better now. And that's probably the only time I've ever let health issues affect me." – Jim, 36, ToF*

Some described that being told by healthcare professionals that *"they are not fine"* had made them more *"aware"* of their condition and hence more anxious and *"obsessive"* with their heart and symptoms.

> *"When you're fine, it's fine. When you're not fine or people tell you you're not fine it's kind of a bit all-consuming. It's like everything you worry about. Every time you wake up you're like "Oh is it that?" Every time that you can feel pain it's like "Oh do I need to go to the hospital?" Whereas before I felt a bit of a fool going for check-ups and they were "Oh yeah you're all fine". […] I was sitting there like wearing my heart rate monitor last year for days, plotting the graphs, seeing how fast it was." – Chloe, 36, TGA*

For one person, this increased heart-related anxiety had made her more hesitant about engaging in activities that could potentially cause further symptoms; this led to loss of confidence in her ability to engage in activities that required physical effort.

> *"So slightly nervous about pushing myself physically in case that [atrial flutter] happens because when it does happen I'm just sort of completely floored, obviously if I end up in hospital so […] the atrial flutter did knock my confidence a lot as far as pushing myself physically. So I sort of try to overcome that now as well really and get going again before it's too late." – Anastasia, 45, TGA*

Some participants who did not generally experience significant physical burden from their heart condition reported that they were not particularly worried about it. One person talked about not noticing the *"heart stuff"* because of other more serious co-morbid conditions.

> *"I don't really notice it […] I think just because I was so used to having a lot of things, when I was a child, I don't really. This is kind of like an extra thing that I don't really, it doesn't really bother me, that much." – Sophia, 27, Simple*

Some others talked about previous interventions and how these were *"rough"*, *"stressful"*, and *"dramatic"*. One person refused to talk about his latest heart operation because it made him feel *"uncomfortable"*. Another individual felt that the emotional impact of her heart operation was more apparent compared with other operations, which were unrelated to her heart.

> *"…when I had the heart surgery, that was, it was more, it felt more traumatic than the other operations I'd had. But I don't know if that was because I was older and I could understand the risks a bit more, or if it is because it was the heart so it was a bit more I don't know feels more serious, I don't know […] I think when I went home I was so relieved I was like, I did cry a lot when I got home." – Sophia, 27, Simple*

Family planning

Female participants talked about emotional issues surrounding pregnancy and family planning. Those who did not have children, talked about how a future pregnancy might be *"overwhelming"* and that it is something they were concerned about. Those that did have children talked about the *"emotional"* impact of being advised that they could not have (any more) children. One person, who went on to adopt her son, described how she needed more clear and timely information from healthcare professionals regarding her capacity to withstand the challenges of pregnancy.

> *"I really wish I understand and I understood more clearly that I couldn't have children, a lot earlier, earlier stage of my life...rather than when I was already in my thirties. Cause I think I would have done things [cries] slightly differently […] just felt like "Ok I'm 34 now and I'm now been told I can't, it just wouldn't be good to have kids". I should have been 24." – Doreen, 37, SV*

Another person felt some *"pressure"* as she was advised to have her children before the age of 30. Although she went on to have two children, she described that her experience was emotional.

> *"Probably later when I wanted children that was a bit of an emotional thing I suppose that hang over me. That I was, I think when I was 18, I was told "Yes you will be able to have children but you must get on and do it before you get 30". So that was always this sort of, a bit of a pressure I suppose." – Anastasia, 45, TGA*

Expectations and uncertainty

Uncertainty was a common narrative across many interviews and was closely linked to expectations about the future. The majority of people acknowledged that health deterioration was inevitable in the future saying that *"it will get worse before it gets*

better", that *"the only way is down"* and that they would probably need to have further interventions.

> *"Eventually I will start to get symptoms, eventually I will need to have some kind of procedure." – Peter, 39, Simple*

Others felt unsure about the course of their condition indicating that *"it's a bit of an unknown"*.

> *"I don't know. No, I really don't know and I don't think I've ever been given an answer by the doctors on that either." – Anastasia, 45, TGA*

For some, health deterioration was already their reality and faced long-term complications and changes in their treatment regimen. In these individuals the emotional consequences of living with ACHD (described in the previous sub-theme) were enhanced by a feeling of uncertainty about the future and their ability to follow-through on future plans. Uncertainty was heightened by health care professionals not being able to give them *"definitive answers"*. One person mentioned that she had to seek professional help because of how this uncertainty had affected her emotionally.

> *"...we are at an age where people before us didn't survive. So we are the test group of people. So nobody can give you any answers. So that does take a lot of coming to terms with really […] Psychologically, I have had periods, especially over the last few years, where it has really affected me and I've been offered stress [therapy] […] just purely, you don't know what's gonna happen so it's not like you can ever, you don't ever have a real plan "This is gonna happen and then you gonna get to this stage and this is gonna happen". So psychologically you have to come to terms with the fact that actually you just have to live in the here and now, you can't think too much about what's gonna happen because actually nobody knows." – Janis, 35, SV*

Furthermore, the possibility of future interventions was described by some people as *"always in the back of [their] mind"*. They felt *"worried"* and often wondered about future interventions; feelings which appeared to be mostly prominent around the time of their follow-up appointments. In addition to their follow-up appointments being a reminder of possible interventions, people talked about the *"uncertainty"* about the timing of an upcoming intervention.

> *"I guess you've always got it there, at the back of your mind that you've had a problem and that something could happen in the future. It's always there, you know. I've always sort of come in back to the hospital every couple of years or whenever it is. I know they've mentioned that I'm gonna have to have another procedure at some point later on so. Obviously a bit worried about it" – Phil, 37, ToF*

> *"I guess it's more the anticipation of what might happen in the future as you get older and the fact that you probably will need an operation at some point and what that means for your quality of life […] rather than any impact now." – Peter, 39, Simple*

> *"I'd like to get a good, if the valve's leaking more, do we, are talking about operation in the next year or two years. I'd rather kind of get it firmed up, because it does play in the mind." – Jim, 36, ToF*

10.4. Discussion

The aim of the present study was to explore the experiences of people with ACHD and to gain a deeper understanding of how they adjust to living with their condition. There were three overarching processes identified across the narratives; the impact of ACHD, the coping process, and change over time. People with ACHD appeared to have developed effective ways of coping with their condition and its treatment. Coping with ACHD appeared to be a result of interplay of factors including setting

goals and priorities, acceptance and adjustment, gaining perspective, and social influences. These factors influenced the degree to which people experienced an impact on their HRQoL as a result of ACHD. There was a range of consequences as a result of living with ACHD, including, physical, social, occupational, and psychological, as well as issues relating to disclosure (Figure 10.2, page 319). As mentioned earlier, the process of change over time was embedded in the participants' narratives, which suggested the dynamic aspect of living with ACHD and its progressive nature. Changes in health and treatment over time influenced people's HRQoL experiences and their coping efforts. They described a number of symptoms and limitations caused by their health deteriorating and by the introduction of new treatment regimens, which had an evident impact on their lives. Such changes in the illness trajectory over time appeared to initiate a new cycle of adjustment to different limitations and treatment routines that needed to be accepted and incorporated into their lives. During these periods people also experienced increased uncertainty and negative emotions and tried to gain perspective and find meaning in these new experiences. In addition, the life situation appeared to be significant for how people experienced the consequences of ACHD and how they coped with them. For example life events such as finding work, retirement, and starting a family brought new insights on the nature of what it is like to live with and adjust to ACHD. The impact of change over time is addressed further within the themes, which will now be discussed in relation to previous research. Synthesis of the findings of this study in relation to the quantitative studies described in the previous chapters and the general implications for theory, research and clinical practice will be addressed in Chapter 11.

10.4.1. Priorities and goals

Most people placed significant importance on health and physical ability along with financial stability, happiness, and maintaining social relationships. These findings provide further support to the assumption that HRQoL is a multidimensional concept that may contain physical, occupational, psychological, interpersonal, and somatic sensation aspects (Schipper et al., 1996).

On the whole people prioritised their health above any other domain of their life. Although a few people tended to place more importance on their personal and work lives. Most people expressed doubt about the degree of control they have over their QoL more broadly and especially their health. Lack of control in people with ACHD has also been reported in a study of 12 individuals with moderate and complex ACHD by Claessens et al. (2005), who reported that people felt they were being controlled and "dominated" by their condition. In the present study people described how they took active steps in an attempt to maintain their health, including eating healthy and exercising. This finding suggests that despite feeling that their illness is uncontrollable, people still strive to gain control over certain aspects of their health. Rönning et al. (2008) found that receiving information about their condition and support resources helped people feel more in control of their ACHD. This approach to patient education may therefore be useful in enhancing people's sense of control.

10.4.2. Acceptance and adjustment

The narratives of many participants revolved around achieving normalcy in their lives by accepting their condition and integrating it into their personal identity and their lives. People acknowledged their condition, were aware of their limitations, and attempted to adjust their lives based on their abilities. Deatrick and colleagues (1999)

have argued that this is a process of normalization, which occurs early in people's lives and is key to the way families cope with a chronically ill child. They identified several elements of normalization evident within families with chronically ill children: a) acknowledging of the impairment, b) defining life as normal, c) minimising the social consequences of the illness, d) engaging in behaviour that are consistent with normality, e) incorporating a treatment regimen consistent with normality. The family of chronically ill children attempt to form their own normality by integrating the child's illness and treatment into their daily lives. Multiple qualitative studies with children and adolescents with CHD have found normalization to be integral in adjusting to their condition (Andresen, Andersen, Lindberg, Døhlen, & Fosse, 2014; Chiang et al., 2011; Gantt, 2002; Horner et al., 2000; Kools, Gilliss, & Tong, 1999; Shearer et al., 2013; Tong et al., 1998; Zahmacioglu et al., 2011).

The findings of present study suggest that this process of normalisation and specifically acceptance and adjustment is part of people's coping attempts throughout their life course and not just in childhood. This notion has been supported in previous qualitative studies of people with ACHD (Berghammer et al., 2006; Claessens et al., 2005). Acceptance of the presence of an underlying health problem and the limitations associated with it was the first step towards achieving normality for individuals in the present study. These people defined normality based on their capabilities and had knowledge about what they were able to do. For example, by planning ahead, pacing themselves, and avoiding social activities that were beyond their limitations. In addition, they constructed their normality to include their treatment regimen.

The degree to which people adjusted to living with ACHD was largely influenced by the stability of their health. Those that had experienced changes in their health and treatment over time often struggled to incorporate these into their lives. This dynamic nature of adjustment has also been identified in previous studies in ACHD (Claessens et al., 2005; Gantt, 2002) and other chronic illnesses (Deatrick et al., 1999). Claessens et al. (2005) found that illness progression and the impact of ACHD influenced people's attempts to achieve normalcy. Gantt (2002) conducted interviews with mother-daughter dyads, which included people with ACHD and found evidence that adjustment was influenced by worsening in symptoms, further surgeries, and hospitalizations. These events made normalization a difficult goal to achieve (Gantt, 2002). The findings of the present study indicated that experience of health changes resulted in people trying to readjust to the increasing demands of their condition and treatment. This is an important finding because as people with ACHD move through life, they are progressively faced with inevitable long-term complications and changes to their treatment. Some authors suggest that, although the course of ACHD may not be predictable for many individuals, providing timely information about possible changes may help people accept them as part of their life and aid better adjustment when they do occur in the future (Moons et al., 2001).

10.4.3. Gaining perspective

People's coping efforts also consisted of trying to find meaning in their experiences of living with ACHD and gain perspective over their condition. Claessens et al. (2005) also found that people's efforts to adjust with ACHD involved "putting things in perspective". In the present study people with ACHD attempted to gain

perspective by comparing themselves to other people in similar situations, appreciating their survival and achievements, and maintaining a positive attitude.

Participants engaged in both upwards and downwards social comparisons with other patients. Upwards comparisons were mainly associated with attempts to cope with a new treatment regimen, where individuals compared themselves to people who managed to adjust successfully. Downwards comparisons were associated with participants' need to adjust to their condition and health changes by comparing themselves with other people in *"worse"* situations. Previous studies have found that during adolescence people compared themselves and their functioning with healthy individuals, which enhanced a feeling of being different (Berghammer et al., 2006; Claessens et al., 2005). This study extends the findings of previous studies by demonstrating that people with ACHD also engage in positive social comparisons during their adult lives. For example, an individual who had her first pacemaker fitted at the time of the interview, described how comparing herself with another person who had multiple pacemaker fittings throughout the years, made her gain perspective and subsequently minimised the emotional burden of this change in her treatment regimen. The findings in relation to the benefits of social comparisons support the findings of a recent review on chronic illnesses which reported that comparisons made by individuals with others who were *coping* better as opposed to coping poorly and with others who were experiencing a *more severe illness* as opposed to less severe were beneficial for their adjustment (Arigo, Suls, & Smyth, 2014). The findings of the present study are important in light of evidence suggesting that people with ACHD need mentorship programmes in order to better cope with their condition (Pagé, Kovacs, Irvine, 2012).

Participants also felt *"lucky"* and *"privileged"* to have survived and to have achieved their life plans, such as having a family. This stemmed from the recognition of the treatment advances in ACHD during the period of their birth and that patients before that did not have the same chances of survival. This phenomenon was expressed as "beating the odds" in a study by Horner et al. (2000). Thinking in these terms helped participants in the present study gain perspective over their condition and realise that they had achieved their goals despite being born with a life-threatening condition. Adopting a positive outlook and reframing negative emotions and situations were key processes that helped people cope with their condition, enabling them to view the illness and treatment disruptions including operations and regular follow-up appointments, as manageable and generally in more positive terms. This made people shift focus from the negative aspects of living with ACHD to more positive life experiences such as their social life, family, and work and therefore gain perspective. Similar positive attitudes have been reported in children and adolescents (Birks et al., 2007; Tong et al., 1998) as well as in young people with ACHD (Overgaard et al., 2013). Positive reframing is a cognitive coping strategy where a person appraises a difficult situation more positively and is believed to encourage positive affect (Folkman & Moskowitz, 2000). Studies in chronic illnesses have also demonstrated that positive reframing may buffer against the stress associated with a chronic condition and contribute towards gaining perspective and finding meaning (Folkman & Moskowitz, 2000). Moreover, a study in congestive heart failure found that people who coped through seeing their illness in a positive light experienced higher levels of meaning (Park, Malone, Suresh, Bliss, & Rosen, 2008). In the present study, individuals who experienced changes in their health and were facing the possibility of further interventions, gaining perspective through positive

reframing was more difficult due to uncertainty and it involved a more active approach like seeking psychological counselling. This was helping them place their heart condition within the bigger picture and appreciate other positive aspects of their lives. These findings provide further support to studies indicating the efficacy of providing psychological support for people with ACHD particularly during periods of health decline and introduction of changes in their treatment regimen as it may help people cope more effectively with their condition and the uncertainty (see section 10.4.8, page, 359) associated with illness progression (Rönning et al., 2008).

10.4.4. Social influence

All participants in the study highlighted the importance of having a supportive environment that helped them cope with the demands of their condition and treatment regimen. They expressed high levels of support from their friends and family. These findings are in line with quantitative studies that have reported equal to or higher levels of support in people with ACHD compared with the general population (Knowles et al., 2012; Pike et al., 2012). People also described how they receive the right type and amount of support they need, thus indicating no inconsistencies between their preferred and received support. This finding has been reported in a previous quantitative study, which indicated that people with ACHD reported fewer inconsistencies between their preferred and received support compared with healthy individuals (van Rijen et al., 2004). Congruence between preferred support and received support has been highlighted in the literature as potentially important for people's well-being (Thoits, 1995).

Partners provided *practical support* by sharing everyday responsibilities that required physical effort, whilst friends were accommodating by engaging in

activities suited to the individual's limitations, minimising in this way the physical and social consequences of ACHD. These findings have important implications about helping people with ACHD build and maintain supportive social relationships, considering that children and adolescents with CHD may experience impaired social functioning, including engagement in social activities and establishing social relationships (e.g. McMurray et al., 2001).

People also described how they received *emotional support* from their family and friends. It has been suggested that social support can enhance people's efforts to cope with the stress associated with their illness, thus aiding as a "buffer" (Heaney & Israel, 2008). People in the present study sought to reduce the emotional burden of living with ACHD through sharing their worries and negative emotions and seeking reassurance from their family and friends. It is important to note that most people also reported seeking and receiving information and reassurance about their condition and treatment from their healthcare professionals and more specifically nurse specialists. This is reassuring given that research in the U.S. (Kools et al., 2002) found that people with ACHD felt that they lacked support from nurses, who did not have adequate experience and sensitivity towards their challenges. Notably, this study was conducted in the U.S. where such problems in the care and provision of information to people with ACHD have been found (Webb, 2010). Promoting supportive relationships between people with ACHD and clinical staff, where the former feel comfortable seeking information and communicating their needs, may aid their efforts to adjust to their condition (Cornett & Simms, 2014).

For some people, parental involvement in their healthcare and in ensuring their well-being had been ongoing since their childhood. There were few reports of parental

over-protection in this study, which is a common theme across qualitative studies with children with CHD but also in people with ACHD (Claessens et al., 2005; Kools et al., 2002). Some authors have argued that parental over-protection may hinder independence in children with CHD and the development of social skills later in adulthood (McMurray et al., 2001; Tong et al., 1998). Research also highlighted that gaining autonomy from parents might take longer in people with ACHD as opposed to healthy individuals (Overgaard et al., 2013). In the present study it was evident that individuals managed to gain their autonomy, even those who were experiencing some parental over-protection. They also expressed an understanding and even a preference towards parental involvement because for them, it was indicative of genuine concern.

Most importantly, parental over-protection did not appear to have an impact on the people's social functioning. One explanation may be older age, as most were well into their thirties and therefore had years of establishing social relationships outside of the family environment. A previous longitudinal study suggested that social inhibition in people with ACHD diminishes over time resulting in them feeling more secure in social situations (van Rijen et al., 2005a). This finding coupled with the findings of the present study suggest that as people get older they manage to gain their independence, develop their social skills, and maintain their social functioning without feeling restricted by parental over-protection.

It is important to note that a few individuals reported that they were, at times, hesitant about reaching out to close people because they did not want to worry them. Similar findings have been reported in another qualitative study with people with ACHD (Cornett & Simms, 2014), which found that people's help-seeking behaviour

was complicated by them wishing to "protect" their family members. The authors argued that not seeking support may hinder the positive effects of the social environment on people's coping efforts. Encouraging people with ACHD to effectively communicate their needs may help them receive the support that is most suited to their needs but may also help them adjust to their condition.

10.4.5. Physical and social consequences of ACHD

Participants perceived their health to be generally good, yet the physical manifestation of ACHD was evident in most people. Severe symptoms such as arrhythmias and cyanosis were cited by some people. For most people the experience of symptoms resulted in some physical and social limitations, including reduced exercise capacity, engagements in sporting activities, and travelling. These limitations appeared to vary from day-to-day in some people. A previous qualitative study about the experiences of people with SV noted significant symptoms and physical limitations in relation to daily and social life (Overgaard et al., 2013). The authors also noted that most of these individuals managed to find their own pace and adjust to their limitations. The findings of the present study also suggest that the degree to which symptoms and subsequent physical and social limitations interfered with people's lives depended on whether they managed to successfully incorporate them into their daily lives (see section 10.4.2, page 347). As mentioned earlier, adjustment to ACHD is a lifelong and dynamic process, dependent on changes in health and treatment. It may be important for people to have knowledge about potential changes in their physical functioning as lack of knowledge or even misconceptions may leave people with ACHD struggling to achieve normalcy although that may not be possible as they age and face further complications

(Rönning et al., 2008). Many participants in the study mentioned that changes in their health were unexpected and *"weren't really thought about before then"*. In addition, although most people acknowledged that their health might deteriorate (further) in the future, they still felt uncertain about how it would impact their lives (see section 10.4.8, page 359).

People with ACHD have previously expressed a desire for structured and timely information about potential changes in their health and treatment and the possible consequences to their physical and social functioning that may help them cope with the demands of their condition and treatment (Rönning et al., 2008). They have also described how receiving continuous information from healthcare professionals had previously helped their knowledge of what to expect and had provided them with tools to manage their symptoms and limitations (Rönning et al., 2008). Taken together, the findings of the present and previous studies suggest that continuous provision of timely information across people's lifespan may help them form realistic expectations about their condition and illness progression.

10.4.6. Occupational consequences of ACHD

The physical impact of ACHD in the form of symptoms and physical limitations had a direct effect on the people's ability to fulfil their occupational role. Most people appeared to be well educated and had skilled or professional jobs. However, their physical status seemed to have an increasing impact on their ability to work full-time, their engagement in their usual job responsibilities, and early retirement due to family planning or reduced capacity. For a few people, their condition had influenced their choice of career or limited their ability to work in the first place. Claessens et al. (2005) also found that people with ACHD experienced employment

issues. Another large study of employment in people with ACHD found that although they did receive career advice, they found it less helpful than healthy individuals. This was attributed to inappropriate advice that was not specific to ACHD (Crossland et al., 2005). The authors suggested that educating employers about the physical challenges of people with ACHD might help these individuals' chances of finding a job specific to their capabilities with an understanding employer. Together, these findings suggest that employment issues are persistent challenges even for people currently in employment, especially as they age and their physical limitations become more prominent.

In addition, female participants appear to have particular needs in relation to balancing work and motherhood, which reflects the employment challenges experienced by women in general (Perrons, 2009). Some authors have suggested that adjustments such as working at home may be a practical way of helping people with ACHD that wish to stay in employment (Kamphuis et al., 2002c). However, it is also important that people with ACHD themselves are educated about how illness progression might influence their work life as it may help them take proactive steps towards disclosing their condition (section 10.4.7) and making arrangements with their employer or even early retirement.

10.4.7. Illness disclosure

The findings of this study indicated that people with ACHD did not generally experience problems with disclosing their condition to their close social circle. Disclosure was more difficult with new friends or acquaintances and people appeared to engage in cost-benefit decisions about whether to disclose their condition. This was also evident in a recent study, which suggested that people with

ACHD were unsure about when and how to disclose their condition (Cornett & Simms, 2014). Children and adolescents with CHD have also reported difficulties in disclosing their condition because of fear of discrimination and social exclusion (Horner et al., 2000; Tong et al., 1998). Disclosure challenges have been reported in other visible and invisible chronic conditions (Joachim & Acorn, 2000) and it appears that ACHD is no exception. The findings of the present study also suggest that disclosure within the social circle becomes less challenging for adults as they had managed to develop supportive social relationships.

Most people currently working were comfortable with disclosing their condition to employers, particularly when there was a need for adjustments in work responsibilities. However, some of them expressed apprehension about fully disclosing their condition during job interviews because of fear of discrimination. A previous qualitative study with adolescents with CHD reported that they were concerned over discrimination from future employers mainly because they had already experienced discrimination in their lives (McMurray et al., 2001). In the present study it was apparent that some people carried these concerns through to their adult lives supporting findings from various chronic conditions (Joachim & Acorn, 2000). With the *Disability Discrimination Act* (DDA) 1995 in place in the UK, it is generally expected that people should not be concerned about discrimination and in turn employers should not discriminate against people with long-term disabilities. Some people with ACHD may not be fully aware of their rights for equal opportunities in employment and may still face discrimination. Hence, combining provision of information about employment laws for disabled

people with patient education about career and employment might help people with ACHD to pursue employment and encourage them to disclose their condition.

10.4.8. Psychological consequences of ACHD

Most people expressed feelings of *"frustration"* in relation to their physical limitations and not being able to fulfil their typical role at home, work, and within their social circle. The emotional burden of ACHD was most prominent during periods of health deterioration and interventions, where people described feeling depressed, anxious, and less confident in their capabilities. Furthermore, previous interventions were characterised as *"dramatic"*. These findings suggest that illness progression and changes in health status are accompanied by increased psychological consequences. Similar findings have been reported in a recent qualitative study about the psychological impact of ACHD (Cornett & Simms, 2014). The authors found that people with ACHD felt depressed over their condition and unexpected cardiac episodes and further operations left patients shocked and traumatised. The findings from both studies suggest that long-term complications and interventions during adulthood can have a significant emotional impact on people with ACHD.

It is also important to note that female participants found pregnancy and family planning to be distressing. Claessens et al. (2005) also found that pregnancy was a particular concern for women with ACHD. Horner et al. (2000) reported that being informed that they were not able to have children was devastating for women with ACHD. In the present study women who did not have children were concerned about potential pregnancy issues in the future, whilst women who did have children described these experiences as being particularly emotional. One person, who did not understand doctors' advice regarding pregnancy, emphasized the importance of

receiving clearer and timely information about her ability to withstand pregnancy. These findings suggest the need for more structured education about pregnancy and family planning for women with ACHD and potentially the provision of psychological support for women informed that they cannot bear children (Moons, De Geest, & Budts, 2002).

The findings of the present study also indicated that the emotional consequences of ACHD were enhanced by a general feeling of uncertainty. Although most people acknowledged the possibility of health deterioration and further interventions and even though some of them had already experienced these changes, they still felt uncertain. This uncertainty left them feeling even more worried and anxious. Feelings of uncertainty have been reported multiple times in children and adolescents with CHD (Lee & Kim, 2012; Tong et al., 1998) and in the adult population (Claessens et al., 2005; Horner et al., 2000) suggesting that this is an ongoing issue for these individuals throughout their lifetime. Uncertainty has also been reported in other chronic conditions and evidence suggests it may be detrimental for people's HRQoL and overall adjustment (Cleanthous, Newman, Shipley, Isenberg, & Cano, 2013). Feelings of uncertainty were enhanced by a "wait and see" attitude from healthcare professionals in the present study. These findings support the Uncertainty in Illness Theory (UIT; Mishel, 1988), which posits that the primary source of uncertainty is the underlying illness, whilst healthcare professionals are considered as a secondary source of uncertainty.

As mentioned earlier, advising people about potential long-term health changes might help ameliorate these feelings of uncertainty and prepare them for the possibility that their health will deteriorate and that they will need to undergo further

interventions. However, a qualitative study with people with systemic lupus erythematosus and rheumatoid arthritis found that uncertainty may persist even when people are well informed about their condition, suggesting the distinction between the subjective perception of uncertainty and the objective lack of information (Cleanthous et al., 2013). Furthermore, some people in the present study sought professional psychological support to help them cope with uncertainty-related stress. Taken together, these findings suggest that provision of information needs to be accompanied by psychological support readily available for individuals who require it, as it might help them cope with the increasing emotional impact resulting from uncertainty. The need for psychological support is further supported by data which indicate that up to 40% of people with ACHD may seek mental health treatment at some point in their lives and generally favour the provision of psychological treatment (Kovacs et al., 2009a). However, a qualitative exploration of people's educational needs highlighted that the provision of psychological support was an "unfulfilled" need for people with ACHD as they lacked knowledge about the availability of such resources (Rönning et al., 2008). Identification of individuals in need of psychological support by their clinicians might not be straightforward as people with ACHD do not always share their negative emotions with healthcare professionals (Cornett & Simms, 2014). In the present study most people reported seeking emotional support from family and friends and some reported contacting their healthcare professionals for reassurance. On the other hand, some appeared to be apprehensive about sharing their emotions in order to protect family and loved ones (section 10.4.4, page 352). This finding highlights the importance of not just the provision of psychological support but also the need to encourage people with ACHD to actively seek support.

10.5. Summary

The present study explored the experiences of people with ACHD and the ways they adjust to living with ACHD. The overall narrative indicated that people with ACHD utilised coping strategies that helped them minimise the impact of ACHD on their HRQoL in terms of physical, social, occupational, and psychological functioning. People with ACHD had clear goals in relation to their health and managed to adjust to living with ACHD by accepting their condition and integrating their limitations and treatment into their daily lives. They sought to gain perspective over their experiences by comparing themselves to others in similar situations and maintaining a positive outlook. People's social environment appeared to have an important role in helping them cope with their condition and it was these coping efforts that influenced the degree to which they experienced an impact on their physical, social, occupational, and psychological HRQoL. People with ACHD experienced limitations in their activities, impact on their work lives, and a number of negative emotions. Uncertainty about the future was a common concern amongst all people and for women family planning and pregnancy was particularly emotional. Health deterioration and changes in the treatment regimen over time appeared to be influencing both the coping attempts and the impact of living with the condition. The findings of this study could help inform patient education and psychological support especially in relation to illness and treatment progression.

The following chapter will now bring together the findings from the quantitative and qualitative enquiries in order to discuss the contribution of the thesis to existing knowledge and the implications for research, theory, and practice.

CHAPTER 11 – GENERAL DISCUSSION

11.1. Prologue

The findings of each study have been discussed in their corresponding chapters in the thesis. This final chapter offers a broad discussion of the key findings of the quantitative and qualitative studies reported in Chapters 7 to 10 and how these fit with previous research reported in Chapters 2 and 3 of the thesis. The discussion begins with the main aims of the thesis followed by a synthesis of the findings from the quantitative and qualitative studies. The overall aims are discussed in relation to how these studies make a contribution to the literature in the area. The chapter concludes with the strengths and weaknesses of the studies in the thesis, a discussion of the implications for practice, and recommendations for future research.

11.2. Thesis aims

The overall aim of the study was to investigate HRQoL and the factors associated with HRQoL in ACHD. It also sought to examine changes in HRQoL and psychosocial functioning over time. The qualitative study aimed to examine the experiences of people with ACHD and how they adjust to living with ACHD. The specific aims and hypotheses are described in more detail in section 4.3 (page 127).

11.3. Synthesis of the findings from the quantitative and qualitative studies

The following sections seek to integrate the findings from the quantitative and qualitative studies in this thesis and discuss the overarching conclusions within the existing literature and the main implications of the study as a whole. The integration of the findings from mixed methods at the interpretation stage of a research is considered important for achieving the full potential of this approach (O'Cathain,

Murphy, & Nicholl, 2010). The assimilated findings provided various insights with regards to the role of structural complexity in relation to HRQoL, the dynamic aspects of HRQoL, general methodological considerations, and the wider theoretical implications within the context of the SRM.

11.3.1. The role of structural complexity in relation to HRQoL in ACHD

As indicated in Chapter 1, ACHD consists of a heterogeneous group with differing structural changes in the heart. This heterogeneity coupled with the lack of a gold standard in the categorisation of ACHD in general and structural complexity in particular, has resulted in much research that has incorporated heterogeneous groups when examining HRQoL. In the systematic review of the literature in Chapter 2 it became evident that the lack of definition of these mixed samples produced inconsistent findings across studies that examined HRQoL in ACHD and limited the ability to draw clear inferences about the impact of ACHD on the HRQoL of various groups that fall under the umbrella ACHD. The present study sought to examine HRQoL using four diagnostic groups defined by the structural changes of their heart defect. Comparisons were made between these groups and to the general healthy population.

The main findings from the quantitative study indicated that the SV group experienced reduced HRQoL in physical and psychosocial domains compared with both the general population and all the other groups of ACHD in the study. Impaired psychosocial HRQoL was also evident in the Simple group compared with the general population. These findings have important implications regarding the expected psychosocial impact of ACHD, especially in certain diagnostic groups. The significant physical and psychosocial morbidity may be expected in people with SV

due to the complexity of their condition. Both previous quantitative and qualitative studies in ACHD have supported this finding (e.g. Angeli et al., 2012; van den Bosch et al., 2004, Overgaard et al., 2013). The individuals with SV have increased illness and treatment demands with a higher risk of complications, need for frequent health monitoring, and multiple interventions throughout their lifetime which would be expected to have an impact on their HRQoL.

In contrast, people in the Simple group require less frequent outpatient appointments and are generally expected to be free from disease burden and to have good HRQoL. Some previous studies that focused exclusively on people with ASD (Ternestedt et al., 2001) and CoA (Buys et al., 2014), both of which were included in the Simple group in the present study, also reported diminished psychosocial HRQoL in this group compared with the general population supporting the findings of the present study. These findings indicate that the relationship between structural complexity and HRQoL is not a simple linear relationship where decreasing structural complexity leads to better HRQoL.

The issue for the Simple group may be more related to their perception driven by illness and treatment experiences, which may play a role in the evaluation of their HRQoL. Earlier experiences including surgery may shape people's expectations about their future functioning. A study by Lane and colleagues (2002) found that people who were deemed surgically cured (a sample that consisted of people with ASD, VSD, PS, AS) reported poorer HRQoL compared with the general population and with other groups not considered to be cured. In light of this evidence, it is possible that people in the Simple group in this study tend to perceive that they have been "cured" early in their lives, thereby entering adulthood with unrealistic

expectations and misconceptions about their functioning and the extent to which they will remain as patients and their medical needs later in life. The use of surgical terms such as "total correction" by medical teams may further enhance these perceptions of cure (Warnes, 2005). The realisation later in life that their health will likely decline more rapidly than anticipated and that they may require further medical or surgical intervention may come as a shock (Saidi et al., 2007). They may have built up expectations that they will be like the general population and this may be their reference point when completing a HRQoL questionnaire.

It is also the case that the Simple group are likely to have long periods of stability in functioning which may reinforce their belief that they are cured. Their emotional response to any deterioration and limitations and changes in their health may be more extreme (Saidi et al., 2007). It was evident from the qualitative data of the present study that people's experiences of sudden health deterioration or receiving information from healthcare professionals about the possibility of further health decline and re-operation during their adulthood were not always congruent with their expectations resulting in uncertainty and increased negative emotional consequences. These findings suggest that it is possible that the mismatch between expectations about the illness course and experience may lead to significant emotional impact. It may be that the emotional consequences of ACHD are more salient for people in the Simple group because they may find it harder to adjust their expectations, especially since they are more likely to experience longer periods of health stability without symptoms and re-interventions.

If this interpretation is correct, these findings are important considering the evidence that suggest that people may form misconceptions and lack clear understanding

about their condition during their childhood which they may carry into adulthood (Veldtman et al., 2000). A study of people with ACHD further identified gaps in their knowledge especially with regards to the physical restrictions and the purpose of the follow-up, suggesting that they may lack understanding about illness progression and may experience unnecessary self-imposed restrictions which may subsequently impact on their social integration (Moons et al., 2001). This was apparent in the interviews of the present study, where people noted that lack of clear information from doctors heightened uncertainty about their condition, its treatment, and its future impact on their lives.

The pattern of quantitative and qualitative findings in the present study also indicated a lack of a coherent understanding of the illness as a whole as people did not attribute many severe symptoms and consequences to their ACHD, had an average understanding of their condition, felt unsure about the degree of personal control over their ACHD but believed that their treatment was beneficial. These perceptions along with negative emotions were found to be influential for people's HRQoL in the quantitative study. More specifically, people who perceived more severe symptoms and greater consequences due to their ACHD, those with higher anxiety levels and those with depressive symptoms had poorer HRQoL. Furthermore, perceived uncertainty appeared to be related to increased negative emotions including frustration, stress, and anxiety in the qualitative study. These findings are in agreement with the wider chronic illness literature that has demonstrated the impact of negative illness perceptions and mood problems on HRQoL in heart failure, coronary heart disease, myocardial infarction, cancer, arthritis, diabetes, asthma, HIV, and epilepsy (Gaynes et al., 2002; Hagger & Orbell, 2003; Hallas et

al., 2011; Petrie et al., 2007, Ruo et al., 2003; Stafford et al., 2007). The findings also support the conclusions of Schoormans and colleagues (2014) who have reported that negative emotional and cognitive perceptions were significant determinants of poorer HRQoL outcomes two years later. Thus it appears that although there is some evidence of structural complexity driving HRQoL (e.g. SV group) people's perceptions about their ACHD and their emotions also appear to have a significant influence on HRQoL.

It may be expected that people with more complex ACHD hold more negative illness perceptions and mood problems. This was evident in the present study, where the SV group perceived their condition and its impact more negatively than the other groups and had a high percentage (27.3%) of people with depressive symptoms. This may help explain their poorer HRQoL compared with the general healthy population and the other groups in this study. For this group, such negative illness perceptions may therefore be accurate reflections of their poorer clinical state. However, the study by Schoormans and colleagues (2014) indicated that there were people with less complex conditions who also had unwarranted negative illness perceptions about their condition and its emotional impact which negatively influenced their HRQoL. In the present study, the Simple group had the highest percentage (28%) of people with depressive symptoms. People in the Simple group also had a more negative perception about illness curability and a more positive perception about illness chronicity (shorter duration) than other groups suggesting that they did not view their current treatment as helpful and did not believe that their illness would last as long as other groups, which appear to be contradictory. As discussed earlier, the expectations and perceptions people form following their treatment experiences may

explain the impaired psychosocial HRQoL in this group. It is possible that the lower scores in the curability subscale in these adults reflect the realisation that past treatments were not curative and that further problems and re-operations are likely after a long period of clinical stability. Alternatively, there may be people who perceive their current treatment as less helpful and potentially intrusive because they are physically well but they are still required to attend regular appointments. Although the study did not set out to examine the role of treatment perceptions in detail, this is an area of investigation that may be fruitful in accounting for the differences observed in this thesis. This will be discussed in the following section.

11.3.1.1. Implications for the use of structural complexity in HRQoL research

The findings discussed in this section raise the important question of the extent to which categorisation of ACHD based on structural complexity is of value when studying HRQoL. The ACHD population is highly diverse consisting of various diagnoses and structural changes. Guideline reports from various organisations around the world argue for their formulation for the classification of ACHD, each proposing their own in order to guide clinical practice and no gold standard exists. These categorisations are often broad and while they differ in the way they categorise specific diagnoses, all commonly attach labels like simple/mild, moderate, and complex/severe in order to describe the groups by disease complexity. Perhaps the most widely known is the Task Force 1 classification system of the American Heart Association, which is based on the underlying diagnosis and the need for regular monitoring (Warnes et al., 2001).

The way in which such broad systems categorise various diagnoses has received criticism in recent publications for their inability to capture the true impact of illness

on people's experiences (Jackson et al., 2015). According to the Task Force 1, TGA and SV are both categorised as complex. However, SV is associated with increased morbidity, mortality, and long-term complications (e.g. hemodynamic issues) that may not be typical in other diagnoses including TGA (Jackson et al., 2015). It is questionable the extent to which this and other similar broad categorisations sufficiently differentiate between the most complex and comparatively less complex diagnoses. Another way used to categorise ACHD is the use of the NYHA classification. However, this classification represents various levels of functional status and is not specific to ACHD, thus it does not take into account the underlying structural change in the heart and by extension the heterogeneity of ACHD.

In view of the limitations of previous categorisation efforts, the present study followed an approach that allowed for clear distinction between various diagnostic groups defined by their structural changes in the heart. There were evident differences between the diagnostic groups and the general population providing some support for a categorisation based on structural complexity. However, the unexpected finding with regards to the impaired psychosocial HRQoL in the Simple group may suggest that this categorisation could be potentially expanded to include factors relating to expectations and illness and treatment experience. A more comprehensive evaluation of the treatment history (e.g. nature of treatment – curative operation, corrective operation, palliative operation, inoperable, medication) along with an assessment of expectations should be considered to supplement any use of the classification based on structural complexity. This will enable an examination of the classification based on structural complexity to be investigated in relation to expectations and perceptions about treatment and subsequently HRQoL.

Considering the importance of illness perceptions in relation to HRQoL in this study, the assessment of treatment perceptions may be useful in establishing whether these vary between the diagnostic groups. Evidence from cardiac disease have indicated that treatment perceptions are more complex than the single-item approach of the present study and qualitatively different than illness perceptions (Hirani, Patterson, & Newman, 2008).

Measures have been developed to examine in more detail the various aspects of treatment perceptions (e.g. value, concerns, decision satisfaction, and cure). The assessment of treatment perceptions is particularly relevant in ACHD due to the fact that treatment is a major aspect of people's experience of their condition. A variety of different treatment approaches may be followed even within a diagnostic group. A person in the Simple group who has received surgery deemed "total correction" and who experiences long periods of clinical stability and minimal limitations would likely perceive their current treatment differently to that of someone with SV who has received palliative surgery and continues to experience significant limitations. It would be expected that these differing perceptions are likely have an impact on their reports of HRQoL. In light of the evidence discussed above that suggested differences in HRQoL between treatment groups (cured, palliative, medical etc.) in ACHD (Lane et al., 2002) coupled with the differences observed between diagnostic groups in illness perceptions in the present study and their role in relation to HRQoL point to the need for further research in order to establish whether treatment characteristics interact with structural complexity to influence illness and treatment perceptions and subsequently HRQoL.

In summary, categorisation by structural complexity in the present study has provided some value in differentiation of ACHD and HRQoL along with illness perceptions and mood. This approach addressed some of the limitations of previous approaches in studying ACHD as a homogeneous group and has helped laid the road for future research. In terms of outcomes research the consideration of factors relating to treatment experience need to be considered in combination or alongside structural complexity in order to examine how their relationship with illness and treatment perceptions and mood may influence HRQoL in ACHD.

11.3.2. The dynamic aspects of HRQoL in ACHD

11.3.2.1. The life stage

ACHD is a lifelong condition that for most people (>60%) is diagnosed early in their lives (Knowles & Hunter, 2014). People with ACHD are expected to go through the same life stages and developmental tasks as healthy individuals but they have to do so by adapting to illness-related, treatment-related, and life challenges at each stage. Guidelines have been published highlighting the need to consider the various life stages along with their relevant developmental tasks when helping people adjust to ACHD (Foster et al., 2001).

The findings of the present study indicated that younger people have poorer psychosocial HRQoL and greater ACHD-specific worries (e.g. employment, family planning) compared with older individuals. This finding supports previous studies in ACHD that have found positive relationship between age and psychosocial HRQoL (Silva et al., 2011; Schoormans et al., 2014). In particular, people in the present qualitative study reflected on how earlier experiences during their young adulthood such as previous operations, challenges in family planning and pregnancies had been

particularly distressing. This is not surprising since young adulthood is a transitional life stage where establishing independence and social relationships, pursuing employment, and starting a family are pertinent issues as opposed to the relative life stability that characterises older adulthood (Sable et al., 2011).

Comparisons between the longitudinal findings of the present study and previous studies in ACHD can provide further insight with regards to life stage and HRQoL. In the present study HRQoL remained stable over the period of the follow-up, whereas previous studies found changes in certain HRQoL domains. Ebenroth and Hurwitz (2007) reported decrease in vitality in people with TGA over a period of 9 years, while van Rijen and colleagues (2005a) reported increase in negative emotions and decrease in social inhibition in a mixed sample over a period of 10 years. A potential explanation for the discrepancy is that previous studies captured changes in HRQoL over a much wider time span and during a transitional period in the people's lives (from young to middle adulthood, 18 to 30 years of age) when their condition may be particularly salient due to the challenges associated with young adulthood.

Whereas the present study examined participants on average from mid to late 30s which may be a relatively stable period of life for most of the participants. During their younger adulthood people with ACHD have to come to terms with their career choices, the timing issues and risks associated with pregnancy while also establishing independence and autonomy from their parents. This period may be particularly distressing because people with ACHD transitioned from adolescence where they may have experienced delay in developmental milestones because of their illness, including completing their education and choosing a career, increasing

their knowledge of their illness and responsibility for their medical care, and establishing independence from overprotective parents (Warnes et al., 2008).

This is further supported by evidence from a recent meta-analysis of young adults with various paediatric chronic illnesses including CHD and healthy individuals. Young adulthood (18-30 years) appeared to be a critical period where young adults with chronic illness had difficulties finding employment, leaving the parental home, marrying, and becoming parents compared with their healthy counterparts (Pinquart, 2014). However the fundamental limitation of the present study was the short follow-up time that was possible thereby limiting any longer term changes over time in HRQoL.

The impact of ACHD on HRQoL was prominent in younger individuals but it appeared to subside as people grew older. This is interesting considering that as people get older and enter their third and fourth decade in life they may face other developmental challenges (e.g. maintaining rather than gaining employment, revising career goals, facing the prospect of premature death and its impact on the family) that may influence their HRQoL (Sable, 2011).

These findings need to also be considered within the wider context and in relation to aging and HRQoL in the general population. A large cross-cultural study with over 10,000 participants from the US and UK general population has found that age had a negative effect on physical HRQoL but a positive effect on psychosocial HRQoL (Franco et al., 2012). The authors attributed these findings to the deterioration of body functions and capabilities and better coping abilities and adaptation in people as they grow older respectively. Older age was not associated with a reduction in physical HRQoL in the present study possibly because people with ACHD would

have not have experienced “normal” health like an average healthy individual. It is also likely that as people with ACHD age they interpret emergent impairments in their functioning as part of the aging process (Bruto et al., 2001).

On the other hand, the increase in psychosocial HRQoL with increasing age may be attributed to well-developed coping strategies and maturation processes. For example, research in healthy populations has indicated that HRQoL can increase with age due to adjustment in goals and personal growth (Netuveli & Blane, 2008). Studies in other chronic illnesses provided similar explanations. In a mixed methods study examining the impact of age on HRQoL in people with heart failure Moser and colleagues (2013) found that as people aged they gained perspective by appreciating their accomplishments (e.g. having a family), seeing the positive, and comparing themselves to other people in similar situations. The pattern of findings in the quantitative and qualitative studies in the present thesis indicated that people with ACHD engaged in adaptive coping strategies including acceptance, normalization, social comparisons, and maintaining a positive outlook. This is consistent with previous studies in ACHD (e.g. Claessens et al., 2005; van Rijen et al., 2004).

Although the studies in this thesis cannot definitively answer the question whether the coping strategies people used had evolved since their young adulthood, comparisons with studies in younger populations do provide some suggestive evidence. In contrast to the more adaptive coping strategies used by people in the present study, denial and avoidance emerged as the dominant coping strategies in studies with children, adolescents and young adults with CHD (e.g. Zahmacioglu et al., 2011). This may suggest how maturing and growing up with ACHD can encourage the use of more adaptive coping strategies in people with ACHD.

Furthermore, the present study also revealed that the illness trajectory can significantly influence how people cope with their condition and adjust to living with ACHD. The next section will discuss the findings in relation to how illness progression can influence people's experiences with living and adjusting to ACHD.

11.3.2.2. The illness trajectory

Despite the usual developmental challenges, people with ACHD also have to adapt and cope with the challenges caused by their underlying condition. According to the SRM changes in health over the course of a chronic illness can lead to a redefinition of the illness representation and the coping strategies people adopt to adjust to their illness (Leventhal et al., 2012). Periods of health deterioration or a change in the treatment may therefore redefine the illness experience.

Changes in health and treatment over the course of the study were prominent in the narratives of some participants in the qualitative study and they influenced their perceptions about the impact of ACHD on their HRQoL as well as their coping efforts. People who reported a change in their health and treatment over time struggled to cope with this change and during these periods people strived to adjust to the changing demands of their illness by enhancing the use of coping efforts. On the other hand, there was evidence of clinical stability in the sample as a whole in the quantitative follow-up study, as measured by recorded clinical data on the number of interventions, hospitalizations, and medication during the follow-up study. While these are key indicators of illness progression, there are other clinical factors not measured in the follow-up study, which may signify a change in people's clinical status over the period of the follow-up. For example, even more subtle changes such as a new symptom, new information from healthcare professionals, or further

diagnostic tests to detect health deterioration can often evoke a process of redefinition of the illness experience (Leventhal et al., 2012). These subtle changes may not be reflected in the clinical records. It is also possible that the stability in health and treatment observed in the follow-up study is due to the fact that illness progression as measured by clinical records is variable in people with ACHD. The observation of clinical stability as well as stability of HRQoL and psychosocial factors at the group level may be accounted for by the short length of follow-up and that people were not assessed in relation to any clinical event.

This is in contrast to the qualitative study where the interviews focused on individual experiences and revealed the variability in illness experience amongst individuals. The interviews allowed people to reflect on their experiences, take and express a longer term perspective, and therefore captured the individual and dynamic processes in the illness experience. While HRQoL appeared to be relatively stable over time in the questionnaires, there was considerable ongoing readjustment taking place for people who experienced illness progression in the qualitative study. This may help explain why certain illness perceptions changed (e.g. increase in illness coherence) while all other factors remained stable during the quantitative follow-up study.

Previous studies in chronic illnesses, including osteoarthritis and diabetes have also found that changes in illness perceptions over time were associated with illness progression (Bijsterbosch et al., 2009; Lawson et al., 2008). These changes may be part of people's continuous efforts to make sense of new information and gain sense of coherence, establish a stable self-image, cope with the increasing demands of their condition and treatment, and maintain or restore their HRQoL.

Although coping strategies and social support had little or no contribution in explaining HRQoL in the quantitative study, the impact of the illness trajectory was evident in the use of coping strategies by people in the qualitative study. People's coping efforts were influenced by illness progression, as the changing demands of their condition and treatment caused people to intensify their coping efforts. It may be that coping and social support become more influential for HRQoL when illness conditions show a marked change, when they encounter a situation where they need to re-evaluate their coping strategies and social support in order to maintain or restore their HRQoL. In the qualitative study, coping efforts, including social comparisons, positivity, and social support varied, even day-to-day, dependent on situational factors revealing the dynamic aspects of these processes in people with ACHD. In line with previous studies in ACHD (Berghammer et al., 2006; Claessens et al., 2005), the degree to which symptoms, limitations, and treatment demands interfered with people's lives was dependent on whether they were able to accept and incorporate them into their daily lives. The qualitative study indicated that this was intensified during periods of illness progression and symptomatic flares. In an effort to cope with their illness and treatment as well as the increased uncertainty that accompanied changes in these aspects, people also attempted to gain perspective by comparing themselves to others in similar positions and viewing their illness and treatment in more positive terms. Furthermore, in order to cope with the loss of control caused by their illness some people re-prioritised and focused on the aspects they could control, for example their diet and exercise.

Qualitative research in chronic illness have previously suggested that "letting go" of control was linked to sense of coherence, which included a search for meaning and

accepting that certain aspects of the illness are not controllable (Aujoulat, Marcolongo, Bonadiman, & Deccache, 2008). Similarly, embracing uncertainty and viewing it as opportunity rather than threat is believed to aid adjustment in chronic illness (Mishel & Clayton, 2008). Considerable research is now focused on examining the role of hope, benefit finding, sense making, personal growth, and other related concepts pertaining to the positive aspects of living with a chronic illness and how these may aid adjustment (de Ridder, Geenen, Kuijer, & van Middendorp, 2008). In view of this evidence, the present findings suggest that perceived uncertainty and lack of personal control may be viewed as a potential opportunity for personal growth and with the use adaptive coping strategies (e.g. social comparisons, positive re-interpretation, re-prioritisation) people can experience positive outcomes including improved HRQoL.

In the qualitative study, social support was also an important coping resource that served various functions, ranging from instrumental (helping with responsibilities, adjusting social activities), emotional (encouragement, confidence-building, understanding), to informational (getting information from healthcare professionals) and played a significant role in people's adjustment. In line with previous studies (Pike et al., 2012), the quantitative and qualitative findings of the study indicated that people perceived high levels of social support from their environment and support previous studies reporting that people with ACHD experience few inconsistencies between their preferred and received support (van Rijen et al., 2004). This may be due to the shared experiences of the illness between people with ACHD and their partner, family, and friends that resulted in social support practises becoming habitual or as one interviewee characterised it "just unsaid but brilliant".

Especially during stressful periods of health deterioration people reported seeking informational support from healthcare professionals and turned to their social environment for emotional support in order to cope with their condition. This may suggest that the benefits of social support for HRQoL are situation-specific (DeLongis & Holtzman, 2005) and may explain why social support was not found to be associated with HRQoL in the quantitative analyses of the present study or previous studies in other chronic illnesses including heart failure (Luttik, Jaarsma, Moser, Sanderman, & van Veldhuisen, 2005) and stroke (Kruithof, van Mierlo, Visser-Meily, van Heugten, & Post, 2013).

In summary, the findings from the qualitative study indicated that the way people coped with ACHD was influenced by illness progression as the changing demands of their condition and treatment caused them to intensify their coping efforts. This suggests that coping is an ongoing process that can vary in intensity depending on the illness trajectory perhaps making coping strategies and social support more influential for HRQoL during stressful periods of health deterioration and change in treatment. This is common in chronic illnesses where people continually need to readjust to the demands of their illness, cope with different facets of the illness, and adapt their coping strategies according to the stressor at hand and at particular timeframes (DeLongis & Holtzman, 2005).

11.3.2.3. Implications for the study of change

The discussion in this section indicated that HRQoL in ACHD needs to be viewed within people's life stage as each is associated with various different developmental tasks which can pose additional challenges that people need to cope with in addition to illness-related issues. Much of the existing literature included mixed samples of

adolescents and adults which complicates the interpretation of the findings with regards to HRQoL in ACHD (Apers, Luyckx, & Moons, 2013a). Viewing the findings of the present study within the existing literature indicated that there are critical periods (i.e. transitional periods) in people's lives when they may be vulnerable to experiencing reduced HRQoL. Furthermore, changes in HRQoL need to also be viewed within the broader life context of the individual as there are key transitional periods where these changes are more likely to occur (e.g. adolescence to adulthood, young to middle adulthood). These ongoing processes across people's lives are important in the context of a long-term condition like ACHD as flexibility may be required in order for people to adjust.

ACHD is a progressive illness and like other chronic illnesses is characterised by stressful periods of health deterioration, flare-ups, and changes in the treatment regimen which can make adjustment complex, with considerable variability across persons, time, and contexts (Hoyt & Stanton, 2012). More research is needed to study how HRQoL may change following such periods and what factors may predict HRQoL. Studies with targeted follow-ups (e.g. before or after interventions) may help determine the best timing for interventions targeting modifiable factors including illness perceptions and mood that aim to improve HRQoL. As the qualitative findings suggest the ways people mobilise their coping efforts can also evolve not just over people's life course but also over the illness trajectory making their effects on HRQoL situation- and time-dependent. Compared with quantitative studies, qualitative studies as in the case of this thesis are important in capturing the dynamic aspects of coping and social support.

The period of follow-up may be extended in future longitudinal studies with multiple targeted follow-ups in order to capture trajectories of change (both individual and group) in these processes and how they may influence HRQoL. Another method that allows for the examination of the fluctuations in both an individual and group level are daily process methods which involve day-to-day monitoring of the study variables (DeLongis & Holtzman, 2005). This method may not only reduce recall error but may accurately capture daily variations in the use of coping strategies and social support and their influence in HRQoL and may detect more subtle changes that correspond to those observed in the present qualitative study. This may contribute to a better understanding of the role of coping and social support in relation to HRQoL in ACHD especially during critical periods in the illness trajectory. Furthermore, a different approach to studying coping, such as clustering people with ACHD on the basis of coping profiles may provide further insight about the relationship between coping profiles and HRQoL across the illness trajectory.

11.3.3. General methodological considerations

Some methodological points have been discussed in the previous sections in particular the categorisation of ACHD, sample definition, and methods to assess the dynamic aspects of the factors under study. The present study also raised some general methodological considerations with regards to the development of disease-specific measures and the use of mixed methods.

The present study utilised both generic and disease-specific measures to examine HRQoL in ACHD as their combination is believed to offer information on general HRQoL and disease-specific issues in the population under study (Fletcher et al., 1992). The selection of a disease-specific HRQoL measure was limited as the CHD-

TAAQOL is the only available measure for use in ACHD. However, as discussed in Chapter 7 of the thesis the CHD-TAAQOL had limited capacity in discriminating between the four diagnostic groups of the study. Significant group differences were only observed in the impact of cardiac surveillance but not in symptoms and worries subscales. Thus the measure was only able to discriminate between the groups in the more objective subscale (i.e. impact of cardiac surveillance).

Taking into account the group differences observed in certain subscales of the generic SF-36 measure (e.g. physical functioning, vitality, social functioning), it was expected that group differences would be even more pronounced in the disease-specific measure since it is concerned with issues more relevant to ACHD. Examining the mean scores in the subscales of the CHD-TAAQOL across the four diagnostic groups indicated that the measure may be able to discriminate between the most complex (SV) and the least complex (Simple) group but not between the intermediate groups (e.g. ToF, TGA). This is not surprising since in the initial validation study by Kamphuis et al. (2004) the measure was used to distinguish between mild (not requiring specialist follow-up) and complex (requiring specialist follow-up) ACHD.

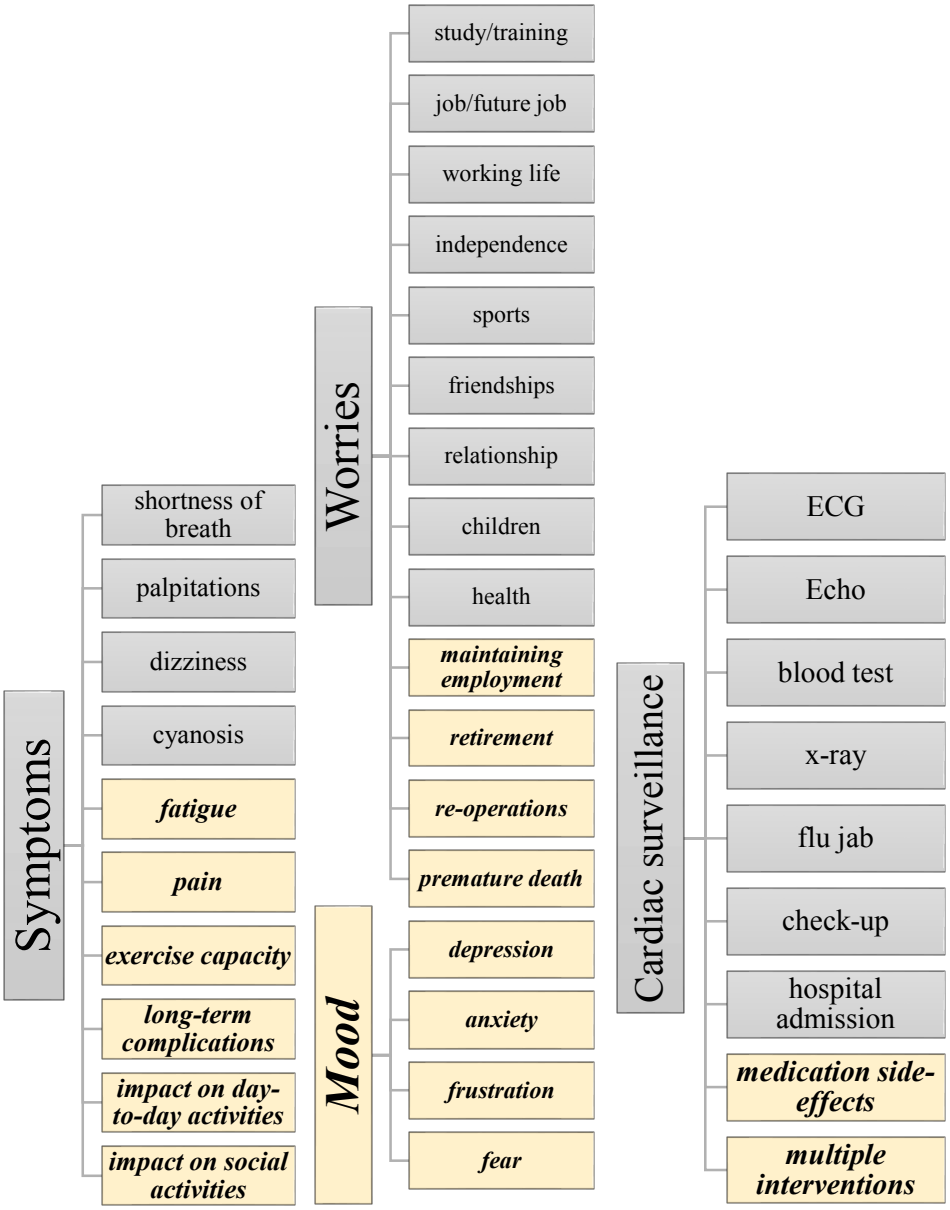
Considering the heterogeneity of ACHD and the fact that SV is substantially different from TGA and ToF (different symptomatology, residual issues, and long-term complications) a disease-specific measure with more sensitivity is needed that has the ability to discriminate across the full spectrum of structural complexity in ACHD and not just between the relatively healthy and ill populations.

Two main problems may have contributed to the limitations of this measure including the specific issues it tackles and the scoring of the subscales. The CHD-

TAAQOL includes various symptoms like shortness of breath, palpitations, dizziness, and cyanosis but not the full range of symptoms in ACHD, thus excluding others like fatigue, pain and exercise capacity or long-term complications of having ACHD. Furthermore, it does not tackle the impact of ACHD on physical and social activities in much detail and although there is a whole subscale on cardiac surveillance, the impact of medication and side effects from treatment are not addressed. The emotional impact of ACHD is addressed by the worries subscale which focuses on the degree of worrying about various issues but not directly assessing negative emotions as a result of ACHD. Overall, the issues covered by the CHD-TAAQOL may be more relevant to younger adults as issues relevant to the older population are not included (e.g. maintaining employment, retirement, re-operations, premature death) (Foster et al., 2001). Figure 11.1 depicts the domains currently tackled by the CHD-TAAQOL and the suggested additions.

The response format (frequency and emotional impact) of the measure may also be considered problematic especially for the items in the worries subscale. For example, asking participants how much they are worried about a specific aspect of their lives and then asking them how much this worrying bothered them may be redundant and unnecessarily complicated. Furthermore, the scoring of the responses assumes that when a problem is absent then the person is not bothered about it. This may not always apply in the case of the cardiac surveillance scale where a person might be bothered even if they have not had a specific test because they may feel that they needed to. These are some of the limitations of the CHD-TAAQOL that need to be addressed when developing new disease-specific HRQoL measures.

There has been significant progress in the children and adolescent literature with the development of disease-specific measures for the paediatric population including the Congenital Heart Disease Quality of Life Questionnaire (ConQoL; Macran et al., 2006) and the Pediatric Cardiac Quality of Life Inventory (PCQLI; Marino et al., 2008). However, there is lack of development of disease-specific measures for the adult population and is being increasingly recognised by researchers calling for disease-specific PROMs in ACHD (Cedars & Spertus, 2014). A disease-specific HRQoL measure in ACHD would need to be comprehensive and cover issues relevant to a wide age range across people's adult life and the ability to discriminate between ACHD groups. The development of psychometrically sound, disease-specific HRQoL measures is necessary to evaluate the effects of randomized controlled trials of new treatment approaches and changes in HRQoL following interventions and over time (Marino et al., 2008).



Note. Highlighted boxes indicate the suggested additional domains

Figure 11.1. Domains currently tackled by the CHD-TAAQOL and suggested additions

In addition to HRQoL, research in other chronic illnesses has previously highlighted the various limitations of generic coping measures and the need for disease-specific measures of coping. Generic measures like the Brief COPE do not distinguish whether certain coping strategies are effortful or habitual for the individual (Steed et al., 1999), or if they are effective and how effective they are. If coping is successful or when people use habitual coping behaviours, they may not be able to identify

what strategies they use to cope with ACHD in general. This is particularly relevant in ACHD because people may have already developed effective (or ineffective) and habitual coping strategies once they reach adulthood (having lived with the condition for most of their lives).

Furthermore, asking people to consider how they usually cope with the stress caused by ACHD may be simplistic because ACHD as a stressor envelops several other sub-stressors. It is hard to discern which stressor (i.e. the context) influences a person's responses on the measure as a whole or even individual items. The limitations of generic coping measures are believed to be addressed to some extent by disease-specific coping measures. Some authors argue that due to the dynamic nature of coping it may be more important to assess whether a person copes in a flexible and complete way rather than the type of coping they use as assessed by generic coping measures (Maes, Leventhal, & de Ridder, 1996). For example, people with chronic illness may need a variety of strategies to cope with various different stressors related to their illness and thus situation-specific coping behaviour may need to be assessed. In addition to disease-specific measures, daily diary methods discussed in the previous section can provide more information about the process and evolution of coping in the everyday lives of people with ACHD.

Qualitative methods are also useful in studying the way people cope with ACHD and the use of mixed methods is considered an appropriate methodology to study the complexity of coping (Tashakkori, Brown, & Borghese, 2010). Coping and social support were two areas where the use of a mixed methods approach in the present study contributed to a better understanding of these processes in ACHD. The quantitative component dealt with the generic aspects of coping and social support,

whereas the qualitative component provided depth with specific contextual examples and the processes underlying these constructs.

While there are inherent challenges when attempting to compare inconsistent findings from two different methodologies, they are best viewed as complementary that provide various plausible explanations for a relationship and open new areas of investigation (Tashakkori et al., 2010). The fact that the qualitative rather than the quantitative data provided more detail about the importance of coping and social support in relation to HRQoL supports the relevance of mixed methods approach in ACHD. As opposed to explaining variance in HRQoL which was the purpose of the quantitative component, the qualitative study revealed the meaning behind people's scale scores. The qualitative study also revealed the disease-specific coping strategies that people used, some of which were not captured in the generic measure used in the quantitative study. The use of social comparisons and re-prioritisation as coping strategies are examples of the qualitative findings complementing the quantitative. The interviews allowed people to provide the context of coping and the nature of the stressor. For example symptoms, limitations, treatment regimen, uncertainty, and lack of control were all ACHD-related stressors that guided people's coping efforts to restore and maintain their HRQoL.

Finally, the interviews captured the dynamic elements of coping and social support indicating that in addition to the day-to-day challenges that people had to cope with, changes in the illness and treatment demands over the illness trajectory also influenced people's coping efforts making them more relevant for HRQoL as people needed to re-evaluate and enhance their coping efforts in order to restore or maintain their HRQoL. The mixed methods approach was valuable in studying these

processes thereby overcoming the logistical limitations of including multiple follow-ups or daily diary assessments.

The concurrent embedded design used in the present study allowed for key aspects of the quantitative component to be explored further with the qualitative component. Alternatively, a sequential design where the qualitative study is conducted first would have revealed the pertinent issues for people with ACHD (e.g. uncertainty) which could have been evaluated in the subsequent quantitative study to determine their influence on HRQoL at population level. However, the concurrent embedded design was deemed appropriate for the present study which used the well-established SRM as a framework for testable research questions. The findings from the qualitative study complemented the findings of the quantitative studies and revealed constructs including uncertainty which may be used to expand the SRM. The theoretical implications of the study findings within the context of the SRM are discussed in the following section.

11.3.4. Theoretical implications

The aim of this section is to discuss the findings within the context of the SRM and the extent to which other factors fit within the SRM to enhance understanding about HRQoL in ACHD. According to the SRM, people's emotional and cognitive representations of their illness play a significant role for outcomes including HRQoL (Leventhal et al., 1992). The findings of the quantitative study in the present thesis also found that people's illness perceptions were associated with HRQoL, supporting the key role of cognitive processing in relation to HRQoL.

Further evidence for the importance of cognitive factors can be seen from the findings of the qualitative study relating to uncertainty. Uncertainty has been the

focus of the UIT by Mishel (1990) who conceptualized uncertainty as a cognitive state, with set antecedents (i.e. stimuli, healthcare professionals/social environment) and an appraisal process (i.e. appraised as danger vs opportunity) which influences coping and subsequently adaptation. Based on previous findings in chronic illness, uncertainty can negatively influence HRQoL and overall adjustment (Cleanthous et al., 2013). The findings of the present qualitative study further indicated that uncertainty in people with ACHD was associated with increased negative emotions especially during periods of health deterioration. This could be interpreted within the SRM in terms of parallel cognitive (uncertainty) and emotional (worry) representation of a health threat (health deterioration). Elements of the UIT and in particular uncertainty could be therefore incorporated into the cognitive representation component of the SRM and future research may establish whether it influences or is influenced by illness perceptions or interacts with them to influence outcomes using validated measures of uncertainty (e.g. Mishel uncertainty in illness scale).

Another expansion for the SRM concerns the incorporation of treatment perceptions or beliefs. As mentioned earlier in the discussion the importance of treatment perceptions has been demonstrated in other chronic illnesses and evidence has suggested that when considered together with illness perceptions in coronary heart disease they explain greater variance in outcomes than when each is considered alone (Newman et al., 2012). Leventhal and colleagues (1997) suggested that because treatments are designed to treat the illness, treatment perceptions may be viewed along the same dimensions of illness perceptions (e.g. time for effectiveness, ability to cure/control, consequences, symptoms etc.). Measures of treatment

perceptions examine in more detail various dimensions including value (treatment benefit), concerns (anxiety and worry of undergoing treatment), decision satisfaction (informational needs, decision-making process, and suitability of treatment), and cure (Hirani et al., 2008).

The findings of the present study indicated differences between diagnostic groups in illness perceptions and demonstrated their importance for HRQoL. These findings along with evidence of the impact of type of treatment on HRQoL in previous studies (e.g. Lane et al., 2002) suggest that treatment perceptions may be a useful addition to the study of HRQoL in ACHD. Irrespective of their label (e.g. correction, palliation, medication etc.), most treatments in ACHD are directed at controlling rather than curing the underlying defect (with various success levels and need for re-intervention in the long-term). Yet treatments may be perceived as cures rather than controllers of the condition by people with ACHD (Saidi et al., 2007).

Examining both illness and treatment perceptions may offer greater insight into people's perspectives and expectations especially prior to re-intervention during their adult lives. Rather than the sequential examination approach followed in the present study (see figure 5.1, page 163), it would be valuable to study the interaction between clinical and cognitive factors; whether people in the four diagnostic groups perceive their treatment differently, particularly in the cure dimension, and how these perceptions relate to their illness perceptions and their HRQoL.

The practical implications of the findings discussed in the previous sections are addressed in section 11.6 (page 406).

11.4. Strengths and weaknesses of the thesis

As with all research, there are methodological factors that may have affected the results or areas that require further exploration. The strengths and weaknesses of the quantitative and qualitative components of the thesis are addressed next.

11.4.1. Quantitative studies

11.4.1.1. Sample size

Firstly, with 314 participants included, the cross-sectional study is one of the largest to have examined the HRQoL of people with ACHD to date. This was an adequately powered sample to find an effect in the ANOVA and regression analyses presented in Chapters 7 and 8 respectively. Although power calculations for HLM analyses are complex, the sample size for the longitudinal study presented in Chapter 9 satisfied the criteria recommended by Tabachnick and Fidell (2007), whereby a minimum of 120 participants is required for two-level models. Therefore, both studies had enough power to test the research questions and hypotheses stated in Chapter 4.

11.4.1.2. Sample composition

It should be acknowledged that certain diagnoses might have been under-represented, for example some diagnoses in the Simple group, which consisted of a large percentage of people with CoA. This could potentially limit generalizability across the entire population of people with ACHD, especially people who are followed-up by their local GPs as opposed to those who attend specialist outpatient clinics. However, including a representative sample of the several different diagnoses would require a very large sample size, a problem acknowledged across the entire ACHD literature. Although the time frame and resources in the present

study did not allow for the inclusion of all possible ACHD diagnoses, the most common were well represented across the diagnostic groups.

A strength of the current study compared with previous literature is the categorisation of participants in diagnostic groups which were clearly defined in their inclusion and exclusion criteria. This facilitated comparisons between four distinct diagnostic groups in Chapter 7.

11.4.1.3. Selection and non-response bias

The lack of an electronic records system of all individuals attending the GUCH clinic at The Heart Hospital, London influenced the recruitment strategies used in the study to identify potential participants, which might have inadvertently resulted in selection bias. Active patients were identified through paper forms, however not all of them were recorded in these forms. As discussed in Chapter 1 (section 1.9, page 44), a common issue in ACHD clinics around the UK is the large number of people lost to follow-up, especially during the transition period from paediatric to adult clinics (Wray et al., 2013). It is likely that people who were lost to follow-up are in better health and have better HRQoL than those who are still followed-up at the GUCH clinic. Although not a limitation of the study itself, this has influenced the recruitment strategies used in the current study to identify potential participants and might have resulted in some selection bias. However, efforts have been made to include newly referred individuals identified from outpatients' clinic lists, who would be otherwise missed if the recruitment was limited to paper forms (section 5.4.3.2, page 134).

As with many longitudinal studies, there is a question of whether sample attrition resulted in non-response bias. The response rate in the study was 60%. Participants

who completed the follow-up study were found to be similar to those who did not in all variables measured but depressive symptoms. As participants who did not complete both assessments were more likely to have depressive symptoms, it is possible that they represent a group who experienced significant change in either their HRQoL or psychosocial functioning. However, it should be noted that the effect size was rather small which is reassuring.

11.4.1.4. Measures

All studies described in this thesis relied on the use of self-report measures, which are believed to be subject to self-representation and recall bias. In order to minimise these bias, it was ensured that participants were aware that they were no right or wrong answers and that their responses were anonymous and confidential.

The study did not include a composite measure of socioeconomic status. The common indicators of educational level and employment status were recorded, however income was not assessed as it can be volatile and fluctuate throughout the year/lifetime. A comprehensive and accurate assessment of socioeconomic status would require a large number of questions and coupled with the sensitivity of the topic (especially income), may result in a low response rate (Grundy & Holt, 2001). Since health inequalities was not the focus of the study, a comprehensive measure of SES was not deemed necessary, especially as education and employment were recorded.

The selection of a disease-specific measure of HRQoL used in the present study was limited to the CHD-TAAQOL, which compared with the generic measure did not appear to be sensitive enough to detect differences between the four diagnostic groups (section 7.4.2.2, page 227 and section 11.3.3, page 382). This suggests that

the measure may be lacking discriminant validity. The CHD-TAAQOL is relatively new and the present study is the first to have used this ACHD-specific measure to study differences in HRQoL between diagnostic groups. Therefore, the validity of the measure needs to be further examined in future studies of people with ACHD.

The use of brief versions of some measures may be considered a limitation. For example, in the Brief IPQ each of the illness perceptions dimensions is measured using a single item, thus the nature of some constructs may not have been fully captured within the items. The revised version of the IPQ (i.e. IPQ-R) might have provided more detailed evaluation of people's illness perceptions. However, owing to the large number of constructs being examined, a pragmatic decision was made to include brief versions of measures, where available, in order to minimise participant burden.

Although floor/ceiling effects are most commonly discussed within the context of experimental research, they can also influence non-experimental research. As reported in Chapter 5, a few subscales of the measures exhibited floor/ceiling effects, which may impact on the ability of a test to detect significant group differences (caused by heterogeneity of variance across groups) and may reduce measure responsiveness in longitudinal studies examining change. In the present study, heterogeneity of variance was taken into account in the ANOVA for group differences by employing the Welch test where appropriate, which is more robust against this violation. In addition, comparisons of the findings from parametric and non-parametric tests revealed the same differences, increasing the confidence in the findings reported in the thesis. Transformations may be conducted to deal with non-

normality and ceiling/floor effects, however in the present study preliminary transformations failed to improve the distribution of the variables affected.

It is possible that lack of change in the variables affected by floor/ceiling effects reflected the limited potential for improvement or deterioration during the follow-up. It is unclear what systematic impact these effects may have had on the results of the longitudinal study but their presence means that although there appeared to be no change in the affected variables over during the follow-up, there might have been little room for improvement/deterioration (Hessling, Traxel, & Schmidt, 2004).

This may have greater implications for future intervention studies that aim to evaluate change in HRQoL and other psychosocial variables as the responsiveness of the measures may be compromised by floor and ceiling effects. However, it should be noted that generic HRQoL measures are often affected by these effects, as a result of using population-based measures in chronic illness samples. Substantial ceiling effects were observed only in two subscales of the SF-36 in the present study. As Bindman, Keane, and Lurie (1990) argue, ceiling effects are less of a concern because at a practical level researchers are less likely to search for improvements in HRQoL among those who already have excellent HRQoL. Thus, the SF-36 may still be an appropriate measure for use in future intervention studies, especially if it is accompanied by disease-specific HRQoL assessments.

One strength of the thesis is the inclusion of a wide range of psychosocial measures, including illness perceptions, coping strategies, mood, and social support. This enabled a comprehensive evaluation of the relative contribution of psychosocial factors to HRQoL in ACHD. As with all studies, however, some measures were not included in the study. Some authors have suggested that, in addition to the factors

included in the SRM, dispositional factors such as personality traits may moderate people's illness perceptions and coping strategies (e.g. dispositional optimism, Carver et al., 1989).

11.4.1.5. Cross-sectional analysis

The cross-sectional study found a range of demographic, clinical, and psychosocial factors that were associated with HRQoL in this sample of people with ACHD. However, there are inherent limitations in causality and directionality inferences based on cross-sectional data. Replication of the relationships between the factors studied is suggested in future longitudinal studies.

11.4.1.6. Longitudinal analysis

By adopting more advanced statistical analyses (HLM), the present study has overcome some of the limitations associated with traditional repeated measures approaches (ANOVA, paired *t*-tests, Wilcoxon signed rank test) used in previous studies. This statistical method takes into account the within-participant variation and the fact that assessments taken from the same participant at different time points are not independent (Quené & van den Bergh, 2004; West, 2009). The strengths of HLM over traditional methods have been discussed in more detail in section 5.6.8.4 (page 165).

It is well known that longitudinal studies require time and resources. In the present study the data collection for the follow-up study lasted a year compared with the cross-sectional study for which data collection lasted longer; this was done to maximise recruitment and thus achieve an adequate sample size for the analyses. This resulted in a variation in the length of follow-up, which could have influenced

the degree of change. However, efforts were made to minimise these effects by controlling for the variation in the analyses.

Furthermore, with only two time-points, the longitudinal study was limited in examining trajectories of change in HRQoL and identification of crucial time points where changes in clinical and psychosocial status had taken place that might have influenced HRQoL. Although multiple assessments would have potentially provided valuable information, it was beyond the scope and the resources available for the present thesis.

11.4.2. Qualitative study

11.4.2.1. Bias

Rapport with the participants was facilitated by the researcher's involvement in conducting the quantitative component of the study. Participants may have been more inclined to participate in the study because of familiarity between them and the researcher. In addition, due to the researcher's involvement in the questionnaire assessments, there might have been pre-conceptions regarding the interviewees' HRQoL. However, efforts were made to disregard these pre-conceptions and preserve an open and inquisitive approach during the interviews. To some extent, pre-conceptions would have been overcome by the supervisory team reviewing the interview topic guide before the interviews and by keeping a reflective diary, which helped understand the researcher's perspective.

11.4.2.2. Interview setting

Guidelines regarding interview settings are scarce but it is generally suggested that the setting is convenient and easily accessible by participants (Mack, Woodsong, MacQueen, Guest, & Namey, 2005). All participants in this study preferred the

interview to be conducted at the hospital as it was convenient to them. It was also ensured that interviews took place in a quiet room away from distractions. The choice of the clinical setting may have inadvertently influenced the participant's responses, for example people who were awaiting test results and decisions regarding upcoming interventions.

11.4.2.3. Generalisability

Twelve people were interviewed for the present study, which may be criticised as insufficient for generalisability to the wider population of people with ACHD. However, generalisability in qualitative research refers to inferential and representational generalisation (Ritchie & Lewis, 2003). Representational generalisation was sought by including a diverse sample in terms of age, gender, and diagnostic group. Inferential generalisation, which involves presentation of the findings that allow the reader to assess whether the findings could be transferred to other settings, was ensured by providing a "thick" or rich description of the findings.

The number of participants reflected the average typically included in qualitative studies in ACHD and it was enough to achieve data saturation. However, it should be acknowledged that some diagnostic groups were under-represented, in particular the Simple group which consisted of two cases. Time limitations did not allow for the inclusion of at least three cases per group, as it is generally recommended when comparing sub-groups in qualitative studies (Onwuegbuzie & Leech, 2007).

11.4.2.4. Rigor and trustworthiness

Rigor and trustworthiness was actively sought by the adoption of various strategies including the use of audit trail for the data analysis, the use of reflective accounts during data analysis, the process of external validation for the coding process, and

the inclusion of "thick" description for the presentation of findings. These approaches contributed towards the validity and reliability of the present study.

11.4.2.5. Mixed methods

The use of mixed method approach in this thesis provided an enriched understanding of HRQoL in ACHD with a synthesis of the findings from both approaches (section 11.3, page 363). This facilitated triangulation of the study findings by highlighting both the similarities and differences from the two approaches. The mixed methods approach also painted a more complete picture of the impact of ACHD on HRQoL and how people adjust to living with ACHD.

A challenge in the integration and interpretation of the findings from the quantitative and qualitative studies is making appropriate meta-inferences or generalisations, considering that the sample size of the qualitative component is less than 10% of the sample included in the quantitative component. The inclusion of a smaller sample size (n= 12) in the qualitative study may be justified by the concurrent embedded design, whereby the qualitative component is supplemental to the dominant quantitative component. Collins, Onwuegbuzie, & Jiao (2007) have argued that statistical generalisations that are *interpretive consistent* (consistency between the inferences made and the sampling design) may be justified if the dominant component is the one with the large sample, which is the case of the present thesis. Including large samples in both quantitative and qualitative studies would provide a stronger basis for interpretive consistency; however, the time constraints of the present study did not allow for the recruitment of a large number of participants in the qualitative study.

The concurrent embedded design of the study, whereby the quantitative component was dominant and the qualitative was supplemental is one of the many methods of mixed methods research. The synthesis of quantitative and qualitative methods is rapidly growing and new techniques are likely to arise in the future (Pope, Mays, and Popay, 2007).

11.4.3. Public and patient involvement in research

The present study did not involve the public and patients during the design and conduct. It is being increasingly recognised that public and patient involvement (PPI) in research can have multiple benefits including acquiring a different perspective on the research topic by people who live with ACHD and making the research more relevant by guiding it towards issues that interest and concern them. In addition PPI can improve the quality of the research through the choice of appropriate methods and materials and increase participation (INVOLVE, 2012).

Consulting patients and their carers in the present study would perhaps help clarify the research questions and affirm their importance. It could also have improved participation rates through advice on recruitment strategies and data collection methods and provide patients' perspective on the materials and information sheets. This is especially relevant with respect to the development of the topic guide of the qualitative study and the subsequent development of the themes. Although the time and financial constraints of the study did not allow for PPI during the design and conduct, there is potential for involving patients and carers during the dissemination stage. Specifically, the study findings will be discussed with patients and carers in order to get advice on producing coherent reports and lay summaries for the study participants and identify other relevant audiences such as ACHD patient networks.

Future studies, including interventions, may benefit from involving patients and carers early in the research process. Patients may offer advice on the ethical considerations, the focus of the intervention and its relevance, feasible recruitment strategies and help with the development of patient information materials. Furthermore, patients may help with the interpretation and dissemination of the findings. PPI can be especially beneficial for intervention studies in order to ensure their feasibility and acceptability by patients (Brett, Staniszewska, Mockford, Seers, Herron-Marx, & Bayliss, 2010).

11.5. Directions for future research

As the systematic review in Chapter 2 and the introduction in Chapter 3 highlighted, few studies have moved beyond the biomedical model in studying HRQoL in ACHD. This could be attributed to the fact that ACHD has been considered a novel area of study for years. However, there has been increasing interest towards studying the psychosocial factors that may influence HRQoL.

The analyses conducted in Chapter 7 revealed that ACHD may result in significant emotional burden in the Simple group compared with the relatively more complex groups (i.e. ToF and TGA) who reported good HRQoL. An interesting avenue of future research is the in depth exploration of expectations in relation to HRQoL in ACHD, particularly across people in various diagnostic groups in order to test whether people form differing expectations especially following their treatment.

The study discussed in Chapter 8 represents the first attempt to study the factors associated with HRQoL in ACHD by including a wide range of demographic, clinical, and psychosocial factors. Future studies may adopt similar approaches in order to examine whether the novel findings from the present study can be replicated

in other samples. Overall, illness perceptions were useful in explaining a large percentage of variance in HRQoL. As the Brief IPQ was identified as a potential a limitation in the present thesis, future studies could use more elaborated versions such as the IPQ-R to measure illness perceptions. In addition, measurement of sense of coherence could be extended to include the aspect of meaningfulness, which was identified as an important part of people's experiences in the qualitative study. Antonovsky's Sense of Coherence scale (SOC) (Antonovsky, 1993) includes a meaningfulness subscale, which could be potentially used in conjunction with illness perceptions measures in future studies. In addition, further studies need to be conducted in order to fully understand the role of coping and social support in relation to HRQoL in ACHD.

Considering that the findings from regression analyses were based on cross-sectional data, future studies may adopt longitudinal designs to examine the temporal stability of the associations identified within the thesis. Furthermore, it is important that future studies use multiple follow-ups in order to explore trajectories of change in HRQoL and the psychosocial factors that influence such changes.

Future research in ACHD investigating people's illness perceptions might focus on developing an intervention to change negative illness perceptions and correct misconceptions. Petrie, Cameron, Ellis, Buick, and Weinman (2002) examined the effectiveness of a brief in-hospital intervention to alter strong negative illness perceptions in people with myocardial infarction. The intervention involved provision of information, challenge of negative illness perceptions, development of personalized action plans, and revision of action plans. Compared with the control group (standard care), the intervention group showed positive changes in their illness

perceptions, earlier return to work, and fewer angina symptoms at a 3-month follow-up. Broadbent, Ellis, Thomas, Gamble, and Petrie (2009) designed a similar randomised controlled trial, which replicated the findings of Petrie and colleagues. The effectiveness of illness perceptions-based interventions has been demonstrated in other chronic populations, including haemodialysis (Karamanidou, Weinman, & Horne, 2008) and type 2 diabetes (Davies et al., 2008; Keogh et al., 2011). These interventions appeared to be successful in improving psychological and HRQoL outcomes and are a promising area for future research in ACHD.

In addition, future research might focus on developing psychological interventions to treat depression in people with ACHD. Research has provided some support for the effectiveness of psychological interventions in minimising mood problems in cardiac populations (Newman, Hirani, Stygall, & Fteropoulli, 2012). In the ENRICHD study, the intervention group of people with myocardial infarction received cognitive behavioural therapy, which included active problem solving, behavioural activation, and challenge of depressive thoughts with antidepressant medication when required. This study indicated significant improvement in HRQoL (Carney et al., 2004).

Intervention studies and cardiac rehabilitation programs in ACHD and the wider CHD population have predominantly focused on improving exercise capacity (Tikkanen, Oyaga, Riaño, Álvaro, & Rhodes, 2012). In addition, a recent review commissioned by Cochrane failed to identify any psychological interventions for depression in people with ACHD (Lane, Millane, & Lip, 2013). It has been argued that mood and HRQoL should also serve as endpoints in future interventions rather than clinical outcomes alone (Linden, 2000). Currently, one study is underway, which aims to assess the feasibility of a randomized controlled trial of a group

intervention aimed at improving psychosocial functioning, QoL, and resilience in people with ACHD (Kovacs & Irvine, 2013). This study could provide valuable insights for future intervention studies.

Many of the interventions addressing illness perceptions took place during hospitalization, while those addressing mood generally took place after an acute episode or following diagnosis; these indicated the importance of timing in the delivery of interventions. Timing may be especially important for people with ACHD, many of whom will face health deterioration that may require further intervention during their adult lives. Interventions aimed at changing negative perceptions may be delivered at such key time points or "windows of opportunity" in order to improve the people's HRQoL after hospital discharge. Delivering interventions during an acute illness phase may be more effective as individuals are more amenable to interventions (Petrie et al., 2002).

In contrast to interventions targeting illness perceptions, interventions aimed at modifying mood may be more successful when delivered after rather than during an acute episode, due to emotional effects that may be situational (Whooley, 2006). Future intervention studies in ACHD need to assess crucial timings at which they may prove more beneficial for improving the people's HRQoL.

Another avenue for research is the possibility of addressing both negative illness perceptions and mood problems concurrently. A systematic review of randomised controlled trials, which aimed at changing negative illness perceptions in coronary heart disease, indicated that there is lack of high quality research that hinders the ability to draw conclusions (Goulding, Furze, & Birks, 2010). The authors highlighted that interventions that include a combination of cognitive behavioural

therapy and counselling are generally more successful in improving behavioural, functional, and psychological outcomes.

11.6. Implications for practice

The findings of the studies have a number of implications for clinical practice. A good starting point could be the routine assessment of people's perceptions about their condition as negative illness perceptions can potentially have a detrimental impact on their HRQoL. The Brief IPQ, although not as comprehensive as the IPQ-R, is a short and useful measure that can be used as part of routine care for a quick assessment of illness perceptions. This may help draw attention to misconceptions that individuals hold about their condition that could be tackled by clinicians or specialist nurses during routine outpatient appointments.

A more challenging issue in patient education is the provision of clear and personalised information regarding health deterioration, changes in treatment, and further interventions. Due to the generally unpredictable nature of many chronic conditions including ACHD, clinicians often provide vague or insufficient information (Rönning et al., 2008). Indeed, the findings of the qualitative study in the present thesis indicated that people were struggling with uncertainty, which was enhanced by the "wait and see" attitude of their clinicians. Clinicians and health care professionals could routinely assess the people's needs during their outpatient appointments and provide timely information regarding the illness and treatment course, education, employment, and family planning issues. This could help people with ACHD accept potential health changes, form realistic expectations especially after treatment, ameliorate uncertainty to some degree, and increase their sense of control (Rönning et al., 2008).

Furthermore, it is important to identify people who are in need of psychological support. The findings from the study indicated the importance of mood problems as a risk factor for poor HRQoL. Short, self-report measures such as the CES-D 10 used in the present study, or other measures like the HADS and BDI are useful for a quick assessment of depressive symptomatology and could therefore be incorporated into the people's routine care. This could be an effective way of identifying individuals at risk in a timely manner and referring them for professional psychological support. It is also important that psychological support is readily available as previous reports have highlighted that the provision of psychological support was an "unfulfilled" need for people with ACHD as they were unaware about the availability of such resources (Rönning et al., 2008). Another possibility for the provision of psychological support may be through peer support groups. As the qualitative study suggested, social comparisons helped people with ACHD cope with their condition and treatment. In another qualitative study, people with ACHD talked about the need of support services such as group therapy, mentorship programs, and patient conferences (Pagé et al., 2012). The Internet may also be a potential way to provide psychological support to these people (Kovacs et al., 2009a).

It is important to note that the findings of the longitudinal and qualitative studies in the present thesis indicated that the people's educational and psychological needs can vary throughout their lives. Therefore, it may be beneficial for health care professionals to re-assess people's perceptions and emotional status regularly, especially during crucial periods such as transition from child to adult clinics, health changes, introduction of new treatment regimens, and before or after re-

interventions. This can help address people's needs more effectively and minimise consultation times by providing only relevant information tailored to people's individual needs during a particular stage in their lives and their journey with chronic illness.

Despite the potential benefits of regular assessment of illness perceptions and emotional status there are still a number of barriers that need to be overcome before such PROMs are widely adopted in healthcare practice. A recent systematic review of professionals' experiences identified several barriers in using PROMs including increase in workload, lack of clear guidelines about the data collection process, and lack of training in analysis and interpretation (Boyce, Browne, & Greenhalgh, 2014).

Although there are also organizational challenges with regards to adopting PROMs in practice that need to be addressed, a number of facilitators have been identified for their implementation by healthcare professionals. Education of healthcare professionals about the use of PROMs and their interpretation is considered paramount (Antunes, Harding, & Higginson, 2014). Engaging healthcare professionals and patients in PROMs-related assessment strategies may enhance acceptability and understanding about their usefulness in clinical practice (Wray, Brown, Marino, & Franklin, 2011).

Furthermore, it has been argued that workloads can be reduced if PROMs feedback is integrated naturally into the consultation process, especially with the use of data collection technology (Boyce et al., 2014). Specifically, the American Heart Association recommends the use of electronic systems for data collection (e.g. tablet computers or computer stations) in outpatient clinic waiting rooms, with built-in scoring algorithms which make the results immediately available to healthcare

professionals (Rumsfeld et al., 2013). Other electronic methods include web-based data collection that can take place in people's homes (Black, 2013). These data may then be incorporated into electronic patient records. PROMs assessment within the NHS is currently being applied only with regards to some surgical procedures, however they are likely to be introduced across a wider range of NHS services, including routine clinical practice along with the introduction of financial incentives (Devlin & Appleby, 2010). Studies are therefore needed to assess the feasibility and acceptability of all the above PROMs data collection methods in ACHD clinics as well as their potential to improve HRQoL outcomes in ACHD.

11.7. Overall conclusions

The present thesis has made a number of contributions to the literature, specifically to the understanding of HRQoL in four distinct diagnostic groups and the factors associated with HRQoL in ACHD using a mixed methods approach. It demonstrated that people with ACHD experience HRQoL impairments, with the SV group experiencing reduced physical and psychosocial HRQoL and the Simple group experiencing reduced psychosocial HRQoL compared with the general healthy population. This has important implications about distinguishing between diagnostic groups when studying HRQoL in ACHD. With the inclusion of a wide range of factors in studying HRQoL in ACHD, the study demonstrated the relative importance of psychosocial factors, in particular negative illness perceptions about the symptoms and consequences of ACHD and mood problems in explaining (unique) variance in HRQoL. The qualitative study has added to the understanding of people's experiences about the impact of ACHD and how they adjust to living with ACHD. Although HRQoL and other psychosocial factors remained stable over

a period of 3.3 years, the qualitative interviews revealed the dynamic day-to-day status of people living with ACHD. The impact of ACHD was evident by the various physical, psychological, social, and occupational consequences of ACHD. The coping process involved people accepting and adjusting to their condition, setting goals and priorities, and gaining perspective with the help of their social environment and was influenced by changes in health and treatment. The thesis offered important implications for clinical practice with regards to the provision of clear and timely information and the assessment of illness perceptions and mood problems during key periods in people's lives and provided a basis for future longitudinal research in studying the role of psychosocial factors for HRQoL in ACHD across the illness trajectory and people's life span. It also highlighted factors amenable to change that can potentially inform the development of intervention studies aiming to improve HRQoL in people with ACHD.

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APPENDICES

Appendix A. E-mail correspondence for image usage rights from Texas Heart Institute

From: Ken Hoge [mailto[redacted]
Sent: 24 July 2014 00:50
To: Fteropoulli, Theodora
Subject: RE: Use of website images for research

Dear Theodora,

The Texas Heart Institute is pleased to grant you conditional permission to use the following 2 copyrighted graphics from our website at http://texasheart.org in your academic work as described in your request below.

We require:
1. The copyright must be noted and the Institute's website address must be listed accompanying all use of the images as follows: Image copyright Texas Heart Institute, www.texasheart.org
2. No commercial gain may be derived from the sale or distribution of this publication by the author or the publisher.
3. Texas Heart Institute reserves all rights and does not grant permission for any subsequent republication, commercial or otherwise, of these images in any media (print, web, video, etc.).

Permission to reproduce this image is contingent upon acceptance of these terms. No rights to any further use beyond this one time are hereby granted.

The authors are: Department of Scientific Publications, Texas Heart Institute:
http://www.texasheart.org/AboutUs/Depart/scipub.cfm

Thank you for your request.

Ken

Ken Hoge
Manager, Visual Communication Services
[redacted] | [redacted] | www.texasheart.org
6770 Bertner Ave., MC 1-134, Houston, TX 77030
Fax: [redacted] | Pager: [redacted]

Appendix B. E-mail correspondence for image usage rights from National Heart, Lung, and Blood Institute (NHLBI)

NHLBI Web Manager (NIH/NHLBI) <[redacted]>
Mon 14/07/2014 16:53
To: Fteropoulli, Theodora <[redacted]>; NHLBI Web Manager (NIH/NHLBI) <[redacted]>;

Theodora,

Thank you for your courtesy in requesting permission to use the information published by the National Heart, Lung, and Blood Institute (NHLBI). No copyright exists on documents generated by the U.S. government, so please feel free to use any information that we provide. We simply request that you cite the NHLBI (as a part of the NIH and the U.S. Department of Health and Human Services), as well as the source document, and if you make any implications or draw any conclusions that are not in the original NHLBI document, that you make a clear statement to that effect in your publication. In addition, the material should not be used in any direct or indirect product endorsement.

NHLBI Web Manager
Center for Biomedical Informatics (CBI)
National Heart, Lung, and Blood Institute (NHLBI)
6705 Rockledge Drive, Room 6086
Bethesda, MD 20892
Phone[redacted]

Appendix C. E-mail correspondence for image usage rights from British Heart Foundation

From: Resources <resources@bhf.org.uk>
Sent: 01 September 2014 14:51
To: Fteropoulli, Theodora
Subject: RE: Permission to use BHF Images for academic purposes

Dear Ms Fteropoulli,

Here is your copyright permission. We are happy to grant it on this occasion. Please let me know if you require the images in another format.

Permission to Use illustrations: Hypoplastic left heart, Tricuspid atresia, Cavopulmonary shunt, Total cavopulmonary connection.

I write to confirm that the British Heart Foundation ("BHF") is willing to consent to the use of the copyright works listed below (the "Works") by Theodora Fteropoulli ("You") in your PhD publication and subsequently within your PhD publication on City Research Online digital archive, on the following conditions:
1. No other use of the Works will be made by You without the prior consent of BHF.
2. You may use the Works for the period of 1 year, commencing on today's date and only in the UK, excepting the accepted use in the City Research Online digital archive.
3. All use of the Works by You shall be in accordance with honest business practices and shall not in any way adversely affect or damage the Works or BHF.
4. All goodwill or other rights accruing from the use of the Works by You shall belong to BHF.
5. All future use of the Works by You shall cease within 7 days of receipt of notice of termination of the permission granted in this email from BHF by You.
6. Any further use of the Works or any other material owned by BHF in any other publication or other manner shall require and be subject to the express written consent of BHF.
7. BHF gives no warranties, indemnity or representation in respect of your reproduction or use of the Works and their use by you.
8. All use of the Work by you shall be accompanied by the following statement: "© British Heart Foundation, 2014. Reproduced with kind permission of the British Heart Foundation."

Yours sincerely
Dr Matthew McArdle

Appendix E. PRISMA 2009 checklist

Section/topic	#	Checklist item	Reported on page #	Notes
TITLE				
Title	1	Identify the report as a systematic review, meta-analysis, or both.	46	
ABSTRACT				
Structured summary	2	Provide a structured summary including, as applicable: background; objectives; data sources; study eligibility criteria, participants, and interventions; study appraisal and synthesis methods; results; limitations; conclusions and implications of key findings; systematic review registration number.	No	
INTRODUCTION				
Rationale	3	Describe the rationale for the review in the context of what is already known.	46-51	
Objectives	4	Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS).	51	
METHODS				
Protocol and registration	5	Indicate if a review protocol exists, if and where it can be accessed (e.g., Web address), and, if available, provide registration information including registration number.	No	
Eligibility criteria	6	Specify study characteristics (e.g., PICOS, length of follow-up) and report characteristics (e.g., years considered, language, publication status) used as criteria for eligibility, giving rationale.	53-55	
Information sources	7	Describe all information sources (e.g., databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched.	52-53	
Search	8	Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated.	Appendix F	
Study selection	9	State the process for selecting studies (i.e., screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis).	55	

Section/topic	#	Checklist item	Reported on page #	Notes
Data collection process	10	Describe method of data extraction from reports (e.g., piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators.	56-57	
Data items	11	List and define all variables for which data were sought (e.g., PICOS, funding sources) and any assumptions and simplifications made.	55-57	
Risk of bias in individual studies	12	Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis.	No	Quality was assessed not risk of bias
Summary measures	13	State the principal summary measures (e.g., risk ratio, difference in means).	No	Not a meta-analysis
Synthesis of results	14	Describe the methods of handling data and combining results of studies, if done, including measures of consistency (e.g., I^2) for each meta-analysis.	55-57	
Risk of bias across studies	15	Specify any assessment of risk of bias that may affect the cumulative evidence (e.g., publication bias, selective reporting within studies).	No	Quality was assessed not risk of bias
Additional analyses	16	Describe methods of additional analyses (e.g., sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified.	n/a	
RESULTS				
Study selection	17	Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram.	58-59, Figure 2.1	
Study characteristics	18	For each study, present characteristics for which data were extracted (e.g., study size, PICOS, follow-up period) and provide the citations.	60-67, Table 2.2 80-86, Table 2.6	

Section/topic	#	Checklist item	Reported on page #	Notes
Risk of bias within studies	19	Present data on risk of bias of each study and, if available, any outcome level assessment (see item 12).	No	Quality was assessed not risk of bias
Results of individual studies	20	For all outcomes considered (benefits or harms), present, for each study: (a) simple summary data for each intervention group (b) effect estimates and confidence intervals, ideally with a forest plot.	67-80, Table 2.3 Table 2.4 Table 2.5 87-91 Table 2.7	
Synthesis of results	21	Present results of each meta-analysis done, including confidence intervals and measures of consistency.	n/a	
Risk of bias across studies	22	Present results of any assessment of risk of bias across studies (see Item 15).	No	Quality was assessed not risk of bias
Additional analysis	23	Give results of additional analyses, if done (e.g., sensitivity or subgroup analyses, meta-regression [see Item 16]).	n/a	
DISCUSSION				
Summary of evidence	24	Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (e.g., healthcare providers, users, and policy makers).	91-99	
Limitations	25	Discuss limitations at study and outcome level (e.g., risk of bias), and at review-level (e.g., incomplete retrieval of identified research, reporting bias).	100-102	
Conclusions	26	Provide a general interpretation of the results in the context of other evidence, and implications for future research.	102	
FUNDING				
Funding	27	Describe sources of funding for the systematic review and other support (e.g., supply of data); role of funders for the systematic review.	No	Provided in publication

Appendix F. Systematic review search strategy: ScienceDirect example

Science Direct [all fields]

{quality of life} OR

{life satisfaction} OR

{well being}

AND

GUCH OR

ACHD OR

{congenital heart disease} OR

{congenital cardiac disease} OR

{congenital heart defect} OR

{congenital cardiac defect} AND adult

Limit to journal articles

Hits: 931

Appendix G. Data extraction form used for the systematic review

General Information
Article Title: **Author(s):** **Source:**
Sample Characteristics
Target Population: **Inclusion Criteria:** **Exclusion Criteria:** **Sample Characteristics: CHD Group:** **Norms/Control Group (if applicable):**
Study Characteristics
Aim(s): **Design:** **QoL/ HRQoL Definition:** **QoL/ HRQoL Measures:** **Other Measures:** **Statistical Analysis:**
Main Findings

Appendix H. Quality index used for the systematic review (adapted from Downs & Black, 1998)

	YES	Partially	NO	Unable to determine
REPORTING				
1. The hypothesis/ aim/ objective(s) of the study were clearly described	2	1	0	0
2. The main outcomes to be measured were clearly described in the Introduction or Methods section	2	1	0	0
3. The characteristics of the patients included in the study were clearly described	2	1	0	0
4. The principal confounders in the sample were clearly described	2	1	0	0
5. The main findings of the study were clearly described	2	1	0	0
6. Does the study provide estimates of the random variability in the data for the main outcomes?	2	1	0	0
7. Actual probability values have been reported (e.g. .035 rather than <.05) for the main outcomes except where the probability value was less than .001	2	1	0	0
EXTERNAL VALIDITY				
8. The persons asked to participate in the study were representative of the entire population from which they were recruited	2	1	0	0
9. The persons who were prepared to participate were representative of the entire population from which they recruited	2	1	0	0
10. The inclusion/exclusion criteria were made explicit	2	1	0	0
INTERNAL VALIDITY- (Bias)				
11. The study design was appropriate for answering the research question(s)	2	1	0	0
12. The statistical tests used to assess the main outcomes were appropriate	2	1	0	0
13. The variable measures used were accurate (valid and reliable)	2	1	0	0
INTERNAL VALIDITY- (Selection Bias)				
14. The participants were similar in terms of prognostic factors	2	1	0	0
15. There was a control group (2 for case-control, 1 for normative data, 0 for no control group)	2	1	0	0
16. There were adequate adjustment for confounding in the analyses from which the main findings were drawn	2	1	0	0
17. Potential differences between respondents and non-respondents were reported	2	1	0	0
POWER				
18. The study had sufficient power to detect a clinically important effect where the probability value for a difference being due to chance was less than 5%	2	1	0	0

Appendix I. Information sheet used in the cross-sectional study

University College London Hospitals NHS
NHS Foundation Trust

Consultants:
Dr Shay Cullen
Dr Fiona Walker
Prof Philipp Bonhoeffer
Prof John Deanfield

Surgeons:
Mr Victor Tsang
Ms Carin van Doorn
Mr Martin Kostolny

Clinical Nurse Specialists:
Ruth Brooks
Marie Francis
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Website: www.uclh.org

UCLH Project ID number : 08/0326

CONFIDENTIAL
INFORMATION SHEET (Version 2-12/08)

Quality of life in GUCH patients.

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Jan Stygall, Fiona Kennedy, Marie Francis, Nathalie Picaut, Katie Austin, Theodora Fteropoulli, Manavi Tyagi, Anna Davies.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

You are being invited to take part in a research study. Before you decide it is important for you to understand why the research is being conducted and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part.

What is the purpose of the study?

Although various studies have been conducted into the quality of life of children with congenital heart disease very little has been carried out with adults with congenital heart disease. In order to better plan our services to support long term care we would find it extremely valuable to have an understanding of how congenital heart disease affects quality of life. Therefore, we in the GUCH Unit at the Heart Hospital, UCLH, and the Unit of Behavioural Medicine at UCL would like to find out how your heart condition affects your life and your functioning. The information you give us will then be used to improve the long-term care of adults with congenital heart disease.

Why have I been chosen?

You are being asked to take part in this study because you have a diagnosis of congenital heart disease. Approximately, 360 patients will take part in this study over the next 3 years.

Do I have to take part?

It is up to you to decide whether or not to take part. If you do decide to take part you will be given this information sheet to keep and be asked to sign a consent form. If you decide to take part you are still free to withdraw at any time and without giving a reason. A decision to withdraw at any time, or a decision not to take part, will not affect the standard of care you receive.

What is involved in the study?

If you agree to take part in this study you will be asked to attend the Heart Hospital on the same day of your next outpatient appointment. During the research appointment a researcher will take you through some questionnaires aimed to find out how your heart condition has impacted upon your life. The appointment will take approximately 45 mins.

What are the possible benefits of taking part?

Whilst there are no immediate benefits for people participating in this study, it is hoped that this work will contribute to improving the long-term care of adults with congenital heart disease.

Confidentiality of records

We also need permission to access your medical records, which relate directly to this study. All the information we obtain will be strictly confidential. UCL will overview the collection, storage and handling of the data and Professor Newman, in his capacity of chief investigator, will be responsible for security and access to the data. Only study investigators (named above) will have access to the data. The information collected during the study, with exception of your name, will be stored and analysed confidentially in a computer. No identifiers on the data held by computer will enable a third party to link the data to you. A study ID number will be assigned to you and this will appear on all data including medical information and questionnaires. All data will be kept strictly confidential and secured under lock and key in UCL. The data will be stored for 5 years after the study has been completed. The results of this study may be published within the medical literature, however, no personal details will be revealed. Copies of the publications will be available to you from the researchers. A report of the findings of the research will be sent to all interested participants in approximately 3½ years from the start of the study.

Comments or concerns during the study

If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on 020 7040 0878. If you remain unhappy and wish to complain formally you can do this through the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this information sheet.

Ethics Committee Review

All proposals for research using human subjects are reviewed by an ethics committee before they can proceed. This proposal was reviewed by the Joint UCL/UCLH Ethics Committee in Ethics of Human Research.

Thank you for taking the time to read this. If you decide to take part you will be given a copy of this information sheet and a signed consent form to keep.

Appendix J. Information sheet used in the longitudinal study

University College London Hospitals NHS
NHS Foundation Trust

Consultants:
Dr Shay Cullen
Dr Fiona Walker
Prof Philipp Bonhoeffer
Prof John Deanfield

Surgeons:
Mr Victor Tsang
Ms Carin van Doorn
Mr Martin Kostolny

Clinical Nurse Specialists:
Ruth Brooks
Marie Francis
Fiona Kennedy
Kerry Romer

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Website: www.uclh.org

UCLH Project ID number : 08/0326

CONFIDENTIAL
INFORMATION SHEET (Version 3-11/11)

Quality of life in GUCH patients, follow-up

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Jan Stygall, Fiona Kennedy, Marie Francis, Nathalie Picaut, Theodora Fteropoulli, Manavi Tyagi.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

You are being invited to take part in a follow-up research study. Before you decide it is important for you to understand why the research is being conducted and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part.

What is the purpose of the study?

Although various studies have been conducted into the quality of life of children with congenital heart disease very little has been carried out with adults with congenital heart disease. Moreover, there is little research examining how further treatment and disease progression affects the quality of life of GUCH patients. In order to better plan our services to support long term care we would find it extremely valuable to have an understanding of how congenital heart disease affects quality of life. Therefore, we in the GUCH Unit at the Heart Hospital, UCLH, and the Health Services Research Group at City University would like to find out how your heart condition affects your life and your functioning in the longer term, after your initial assessment.

The information you give us will then be used to improve the long-term care of adults with congenital heart disease.

Why have I been chosen?

You are being asked to take part in this follow-up study because you have a diagnosis of congenital heart disease and you have participated in the first phase of the study, about 2 years ago. Approximately, 220 patients will take part in this study over the next year.

Do I have to take part?

It is up to you to decide whether or not to take part. If you do decide to take part you will be given this information sheet to keep and be asked to sign a consent form. If you decide to take part you are still free to withdraw at any time and without giving a reason. A decision to withdraw at any time, or a decision not to take part, will not affect the standard of care you receive. Your contribution to the first phase of this study will also remain unaffected.

What is involved in the study?

If you agree to take part in this study you will be asked to attend the Heart Hospital on the same day of your next outpatient appointment. The same procedure applies as in the first phase of the study. During the research appointment a researcher will take you through some questionnaires aimed to find out how your heart condition has impacted upon your life. The full appointment will take approximately 45mins.

What are the possible benefits of taking part?

Whilst there are no immediate benefits for people participating in this study, it is hoped that this work will contribute to improving the long-term care of adults with congenital heart disease.

Confidentiality of records

We also need permission to access your medical records, which relate directly to this study. All the information we obtain will be strictly confidential. City University will overview the collection, storage and handling of the data and Professor Newman, in his capacity of chief investigator, will be responsible for security and access to the data. Only study investigators (named above) will have access to the data. The information collected during the study, with exception of your name, will be stored and analysed confidentially in a computer. No identifiers on the data held by computer will enable a third party to link the data to you. A study ID number, assigned to you during the first phase of the study will appear on all data including medical information and questionnaires. All data will be kept strictly confidential and secured under lock and key in City University. The data will be stored for 5 years after the study has been completed. The results of this study may be published within the medical literature, however, no personal details will be revealed. Copies of the publications will be available to you from the researchers. A report of the findings of the follow-up research will be sent to all interested participants in approximately 3½ years from the start of the study.

Comments or concerns during the study

If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on 020 7040 0871/0878. If you remain unhappy and wish to complain formally you can do this through

the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this information sheet.

Ethics Committee Review

All proposals for research using human subjects are reviewed by an ethics committee before they can proceed. This proposal was reviewed by the Joint UCL/UCLH Ethics Committee and NRES Committee London – Bentham in Ethics of Human Research.

Thank you for taking the time to read this. If you decide to take part you will be given a copy of this information sheet and a signed consent form to keep.

Appendix K. Consent form used in the cross-sectional study

University College London Hospitals NHS
NHS Foundation Trust

GUCH Office
The Heart Hospital
16-18 Westmoreland Street
London W1G 8PH
Tel: 020 7573 8808
Nurses: 020 7573 8872
Fax: 020 7573 8807
Email: guch.dept@uclh.org
Website: www.uclh.org

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 2-12/08)

Quality of life in GUCH patients.

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Jan Stygall, Fiona Kennedy, Marie Francis, Nathalie Picaut, Katie Austin, Theodora Fteropoulli, Manavi Tyagi.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

Please read the following statements and initial box

1. I confirm that I have read and understand the information sheet (version2-12/08) for the above study and have had the opportunity to ask questions. ☐

2. I confirm that I have had sufficient time to consider whether or not I want to be included in the study. ☐

3. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my medical care or legal rights being affected. ☐

4. I understand that all data will be kept strictly confidential to this research and give permission for the above investigators to have access to my medical records. ☐

5. I agree to take part in the above study. ☐

Continued on next page/

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 2-12/08)

Signed: _____ Date: _____

Full name in block letters: _____

Signed (investigator): _____ Date: _____

Full name in block letters: _____

Comments or concerns during the study.

If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on (telephone number yet to be provided). If you remain unhappy and wish to complain formally you can do this through the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this consent form.

1 form for patient
1 to be kept as part of the study documentation
1 to be kept with hospital notes

Appendix L. Consent form used in the longitudinal study

University College London Hospitals NHS
NHS Foundation Trust

GUCH Office
The Heart Hospital
16-18 Westmoreland Street
London W1G 8PH
Tel: 020 7573 8808
Nurses: 020 7573 8872
Fax: 020 7573 8807
Email: guch.dept@uclh.org
Website: www.uclh.org

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 3-11/11)

Quality of life in GUCH patients, follow-up

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Fiona Kennedy, Nathalie Picaut, Theodora Fteropoulli, Manavi Tyagi.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

Please read the following statements and initial box

1. I confirm that I have read and understand the information sheet (version 3-11/2011) for the above study and have had the opportunity to ask questions. ☐

2. I confirm that I have had sufficient time to consider whether or not I want to be included in the study. ☐

3. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my medical care or legal rights being affected. ☐

4. I understand that data collected during the study, may be looked at by individuals from UCLH NHS Foundation Trust, from regulatory authorities or from the NHS Trust, Where it is relevant to my part in this research, I give permission for these individuals to have access to my records. ☐

5. I agree to take part in the above study. ☐

Continued on next page/

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 3-11/11)

Signed: _____ Date: _____

Full name in block letters: _____

Signed (investigator): _____ Date: _____

Full name in block letters: _____

Comments or concerns during the study.

If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on (telephone number yet to be provided). If you remain unhappy and wish to complain formally you can do this through the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this consent form.

1 form for patient
1 to be kept as part of the study documentation
1 to be kept with hospital notes

Appendix M. Psychosocial questionnaire pack

Quality of Life in GUCH patients

Participant Questionnaire Booklet

Researcher Notes

Participant ID

Thank you for agreeing to take part in this study looking at quality of life in adults with congenital heart disease. The information you give us today will contribute to improving the long term care of adults with congenital heart disease.

This booklet contains a number of questionnaires which you are kindly asked to complete. Please take time to read and understand the individual instructions for each questionnaire.

There are no right or wrong answers; we are just interested in your opinions. If you do not understand any of the questions or the instructions provided, please do not hesitate to ask the researcher for assistance.

Please mark the boxes with a **cross** [X]. Should you make a mistake, please colour the box in completely and mark your correct selection.

Thank you again for taking the time to complete this study. We assure you that your responses will be kept completely confidential.

Many Thanks,

The GUCH Team

SF-36 HEALTH SURVEY

INSTRUCTIONS: This survey asks for your views about your health. This information will help keep track of how you feel and how well you are able to do your usual activities.

Answer every question by marking the answer as indicated. If you are unsure about how to answer a question, please give the best answer you can.

1. In general, would you say your health is:

(circle one)

Excellent 1

Very good 2

Good 3

Fair 4

Poor 5

2. Compared to one year ago, how would you rate your health in general now?

(circle one)

Much better now than one year ago 1

Somewhat better now than one year ago 2

About the same as one year ago 3

Somewhat worse now than one year ago 4

Much worse now than one year ago 5

3. The following questions are about activities you might do during a typical day. Does your health now limit you in these activities? If so, how much?

(circle one number on each line)

ACTIVITIES	**Yes, Limited A Lot**	**Yes, Limited A Little**	**No, Not Limited At All**
a. **Vigorous activities**, such as running, lifting heavy objects, participating in strenuous sports	1	2	3
b. **Moderate activities**, such as moving a table, pushing a vacuum cleaner, bowling, or playing golf	1	2	3
c. Lifting or carrying groceries	1	2	3
d. Climbing **several** flights of stairs	1	2	3
e. Climbing **one** flight of stairs	1	2	3
f. Bending, kneeling, or stooping	1	2	3
g. Walking **more than a mile**	1	2	3
h. Walking **half a mile**	1	2	3
i. Walking **one hundred yards**	1	2	3
j. Bathing or dressing yourself	1	2	3

4. During the past 4 weeks, have you had any of the following problems with your work or other regular daily activities as a result of your physical health?

(circle one number on each line)

	YES	**NO**
a. Cut down on the **amount of time** you spent on work or other activities	1	2
b. **Accomplished less** than you would like	1	2
c. Were limited in the **kind** of work or other activities	1	2
d. Had **difficulty** performing the work or other activities (for example, it took extra effort)	1	2

5. During the past 4 weeks, have you had any of the following problems with your work or other regular daily activities as a result of any emotional problems (such as feeling depressed or anxious)?

(circle one number on each line)

	YES	NO
a. Cut down on the **amount of time** you spent on work or other activities	1	2
b. **Accomplished less** than you would like	1	2
c. Didn't do work or other activities as **carefully** as usual	1	2

6. During the past 4 weeks, to what extent has your physical health or emotional problems interfered with your normal social activities with family, friends, neighbours, or groups?

(circle one)

Not at all 1

Slightly 2

Moderately 3

Quite a bit 4

Extremely 5

7. How much bodily pain have you had during the past 4 weeks?

(circle one)

None 1

Very mild 2

Mild 3

Moderate 4

Severe 5

Very severe 6

8. During the past 4 weeks, how much did pain interfere with your normal work (including both work outside the home and housework)?

(circle one)

Not at all 1

A little bit.....2

Moderately 3

Quite a bit.....4

Extremely 5

9. These questions are about how you feel and how things have been with you during the past 4 weeks. For each question, please give the one answer that comes closest to the way you have been feeling. How much of the time during the past 4 weeks –

(circle one number on each line)

	All of the Time	**Most of the Time**	**A Good Bit of the Time**	**Some of the Time**	**A Little of the Time**	**None of the Time**
a. Did you feel full of life?	1	2	3	4	5	6
b. Have you been a very nervous person?	1	2	3	4	5	6
c. Have you felt so down in the dumps that nothing could cheer you up?	1	2	3	4	5	6
d. Have you felt calm and peaceful?	1	2	3	4	5	6
e. Did you have a lot of energy?	1	2	3	4	5	6
f. Have you felt downhearted and low?	1	2	3	4	5	6
g. Did you feel worn out?	1	2	3	4	5	6
h. Have you been a happy person?	1	2	3	4	5	6
i. Did you feel tired?	1	2	3	4	5	6

10. During the past 4 weeks, how much of the time has your physical health or emotional problems interfered with your social activities (like visiting with friends, relatives, etc.)?

(circle one)

All of the time 1

Most of the time 2

Some of the time 3

A little of the time 4

None of the time 5

11. How TRUE or FALSE is each of the following statements for you?

(circle one number on each line)

	Definitely True	**Mostly True**	**Don't Know**	**Mostly False**	**Definitely False**
a. I seem to get ill more easily than other people	1	2	3	4	5
b. I am as healthy as anybody I know	1	2	3	4	5
c. I expect my health to get worse	1	2	3	4	5
d. My health is excellent	1	2	3	4	5

The experience of symptoms, treatment, and emotions may be different for each person with congenital heart disease. In the following questionnaire we are interested to learn how your heart condition affects your quality of life. The questions relate to different aspects of your heart condition.

Please answer the questions by putting a cross in the box which is most applicable to **you**.

1a. How often in the **last month** were you short of breath after strolling **100 yards (91m)**?

☐ Never ☐ Occasionally ☐ Often

1b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

2a. How often in the **last month** were you short of breath after strolling **between half a mile and 3 miles (0.8-4.8km)**?

☐ Never ☐ Occasionally ☐ Often

2b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

3a. How often in the **last month** did you have difficulty breathing while lying down?

☐ Never ☐ Occasionally ☐ Often

3b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

4a. How often in the **last month** did you have to get up a lot at night to go to the toilet?

☐ Never ☐ Occasionally ☐ Often

4b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

5a. How often in the **last month** did you have excess fluid in your ankles, legs, and/or stomach?

☐ Never ☐ Occasionally ☐ Often

5b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

6a. How often in the **last month** did you have palpitations?

☐ Never ☐ Occasionally ☐ Often

6b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

7a. How often in the **last month** did you feel dizzy?

☐ Never ☐ Occasionally ☐ Often

7b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

8a. How often in the **last month** did your lips turn blue when you exerted yourself?

☐ Never ☐ Occasionally ☐ Often

8b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

9a. How often in the **last month** did you look pale?

☐ Never ☐ Occasionally ☐ Often

9b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

10a. How often in the **last month** were you worried about your study/training?

☐ Never ☐ Occasionally ☐ Often

10b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

11a. How often in the **last month** were you worried about your job/future job?

☐ Never ☐ Occasionally ☐ Often

11b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

12a. How often in the **last month** were you worried about your working life?

☐ Never ☐ Occasionally ☐ Often

12b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

13a. How often in the **last month** were you worried about being able to live independently?

☐ Never ☐ Occasionally ☐ Often

13b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

14a. How often in the **last month** were you worried about being home alone?

☐ Never ☐ Occasionally ☐ Often

14b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

15a. How often in the **last month** were you worried about exertion or sports?

☐ Never ☐ Occasionally ☐ Often

15b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

16a. How often in the **last month** were you worried about having good friends?

☐ Never ☐ Occasionally ☐ Often

16b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

17a. How often in the **last month** were you worried about a steady relationship?

☐ Never ☐ Occasionally ☐ Often

17b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

18a. How often in the **last month** were you worried about being able to have children?

☐ Never ☐ Occasionally ☐ Often

18b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

19a. How often in the **last month** were you worried about your health?

☐ Never ☐ Occasionally ☐ Often

19b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

20a. In the **last twelve months** have you had an ECG (heart trace)?

☐ No ☐ Yes

20b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

21a. In the **last twelve months** have you had an ultrasound heart test (echocardiograph)?

☐ No ☐ Yes

21b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

22a. In the **last twelve months** have you had a blood sample taken because of your heart condition?

☐ No ☐ Yes

22b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

23a. In the **last twelve months** have you had an x-ray of your heart and lungs because of your heart condition?

☐ No ☐ Yes

23b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

24a. In the **last twelve months** have you had a flu jab because of your heart condition?

☐ No ☐ Yes

24b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

25a. In the **last twelve months** have you seen a consultant for a check-up because of your heart condition?

☐ No ☐ Yes

25b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

26a. In the **last twelve months** have you been admitted to hospital because of your heart condition?

☐ No ☐ Yes

26b. How much did that bother you?

☐ Not at all ☐ A little ☐ Quite a lot ☐ Very much

When people experience a condition such as congenital heart disease, they form specific beliefs in order to make sense of that condition. In the following questionnaire we are interested to find out **your own** personal views of your heart condition **NOT** what you think other people believe.

For the following questions, please put a cross in the box number that best corresponds to **your views**.

1. How much does your illness affect your life?

No affect at all — Severely affects my life

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

2. How long do you think your illness will continue?

A very short time — Forever

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

3. How much control do you feel you have over your illness?

Absolutely no control — Extreme amount of control

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

4. How much do you think your treatment can help your illness?

Not at all — Extremely helpful

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

5. How much do you experience symptoms from your illness?

No symptoms at all — Many severe symptoms

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

6. How concerned are you about your illness?

No at all concerned — Extremely concerned

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

7. How well do you understand your illness?

Don't understand at all — Understand very clearly

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

8. How does your illness affect you emotionally? (e.g. does it make you angry, scared, upset, or depressed?)

Not at all affected emotionally — Extremely affected emotionally

0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10 ☐

We are interested in how people respond to having congenital heart disease. There are lots of ways to try to deal with problems which may arise due to your condition. This questionnaire asks you to indicate what you generally do and feel when you have problems related to your heart condition. Obviously, different events bring out somewhat different responses, but think about **what you usually do** when you are **experiencing stress** relating to your heart condition.

Please respond to each of the following items by putting a cross in the box that best describes what you have been doing. There are no "right" or "wrong" answers, so choose the most accurate answer for **YOU**, not what you think "most people" would say or do.

	I don't do this at all	I do this a little bit	I do this a moderate amount	I do this a lot
1. I try to come up with a strategy about what to do.	☐	☐	☐	☐
2. I concentrate my efforts on doing something about the situation I am in.	☐	☐	☐	☐
3. I try to see it in a different light, to make it seem more positive.	☐	☐	☐	☐
4. I accept the reality of the fact that it has happened.	☐	☐	☐	☐
5. I make jokes about it.	☐	☐	☐	☐
6. I pray or meditate.	☐	☐	☐	☐
7. I get comfort and understanding from someone.	☐	☐	☐	☐
8. I try to get advice or help from other people about what to do.	☐	☐	☐	☐
9. I turn to work or other activities to take my mind off things.	☐	☐	☐	☐
10. I say to myself "this isn't real".	☐	☐	☐	☐
11. I say things to let my unpleasant feelings escape.	☐	☐	☐	☐
12. I use alcohol or other drugs to help me get through it.	☐	☐	☐	☐
13. I give up trying to deal with it.	☐	☐	☐	☐
14. I blame myself for the things that happened.	☐	☐	☐	☐

	I don't do this at all	I do this a little bit	I do this a moderate amount	I do this a lot
15. I take action to try and make the situation better.	☐	☐	☐	☐
16. I think hard about what steps to take.	☐	☐	☐	☐
17. I look for something good in what is happening.	☐	☐	☐	☐
18. I learn to live with it.	☐	☐	☐	☐
19. I try to find comfort in my religion or spiritual beliefs.	☐	☐	☐	☐
20. I make fun of the situation.	☐	☐	☐	☐
21. I do something to think about it less, such as going to movies, watching TV, reading, daydreaming, sleeping, or shopping.	☐	☐	☐	☐
22. I get emotional support from others.	☐	☐	☐	☐
23. I express my negative feelings.	☐	☐	☐	☐
24. I refuse to believe that it is happening.	☐	☐	☐	☐
25. I get help and advice from other people.	☐	☐	☐	☐
26. I use alcohol or other drugs to make myself feel better.	☐	☐	☐	☐
27. I give up the attempt to cope.	☐	☐	☐	☐
28. I criticise myself.	☐	☐	☐	☐

A number of statements which people have used to describe themselves are given below. Read each statement and then put a cross in the appropriate box to indicate how you feel ***right now, at this moment***. There are no right or wrong answers. Do not spend too much time on any one statement but give the answer that seems to describe your **present feelings** best.

	Not at all	Somewhat	Moderately	Very much
1. I feel calm.	☐	☐	☐	☐
2. I am tense.	☐	☐	☐	☐
3. I feel upset.	☐	☐	☐	☐
4. I am relaxed.	☐	☐	☐	☐
5. I feel content.	☐	☐	☐	☐
6. I am worried	☐	☐	☐	☐

Below is a list of some of the ways you may have felt or behaved. Please indicate how often you have felt this way during the **past week** (please put a cross in one box on each row).

	Rarely or none of the time (less than 1 day)	Some or a little of the time (1-2 days)	Occasionally or a moderate amount of time (3-4 days)	All of the time (5-7 days)
1. I was bothered by things that don't usually bother me.	☐	☐	☐	☐
2. I had trouble keeping my mind on what I was doing.	☐	☐	☐	☐
3. I felt depressed.	☐	☐	☐	☐
4. I felt that everything I did was an effort.	☐	☐	☐	☐
5. I felt hopeful about the future.	☐	☐	☐	☐
6. I felt tearful.	☐	☐	☐	☐
7. My sleep was restless.	☐	☐	☐	☐
8. I was happy.	☐	☐	☐	☐
9. I felt lonely.	☐	☐	☐	☐
10. I could not "get going".	☐	☐	☐	☐

We are interested in how much you feel you are supported by your friends, family, and a special person in your life. Please put a cross in the box of one of the five responses to indicate how much you agree with each statement.

	Strongly disagree	Disagree	Neither disagree nor agree	Agree	Strongly agree
1. There is a special person who is around when I am in need.	☐	☐	☐	☐	☐
2. There is a special person with whom I share my joys and sorrows.	☐	☐	☐	☐	☐
3. My family really tries to help me.	☐	☐	☐	☐	☐
4. I get the emotional help and support I need from my family.	☐	☐	☐	☐	☐
5. I have a special person who is a real source of comfort to me.	☐	☐	☐	☐	☐
6. My friends really try to help me.	☐	☐	☐	☐	☐
7. I can count on my friends when things go wrong.	☐	☐	☐	☐	☐
8. I can talk about my problems with my family.	☐	☐	☐	☐	☐
9. I have friends with whom I can share my joys and sorrows.	☐	☐	☐	☐	☐
10. There is a special person in my life who cares about my feelings.	☐	☐	☐	☐	☐
11. My family is willing to help me make decisions.	☐	☐	☐	☐	☐
12. I can talk about my problems with my friends.	☐	☐	☐	☐	☐

Any other comments you would like to add regarding your quality of life:

There are no more questions.

Thank you very much for completing this booklet.

Please make sure you have answered all relevant questions and hand the booklet back to the researcher.

Thank you once again for your time.
The GUCH team

Appendix N. License agreement for SF-36v1©

QualityMetric

NON-COMMERCIAL LICENSE AGREEMENT

Office of Grants and Scholarly Research (OGSR)

License Number: QM012947

Effective Date: March 8, 2012

Licensee Name: **City University London**

Licensee Address: Room A224, College Building, Northampton Square London, EC1V 0HB, UK

Approved Purpose: Non-commercial academic research – Grant funded by government agency or non-profit organization

The extent and impact of cognitive functioning on quality of life, mood, and social integration of grown-up congenital heart (GUCH) patients
Study Name: BACKDATED LICENSE / SURVEY REVIEWED BY SCIENCE & APPROVED FOR THIS STUDY ONLY
Study Type: GRANT FUNDED

Therapeutic Area: Heart and Circulation

Other Definitions: As indicated on Appendix B "License Agreement – Details", including without limitation: Licensed Surveys, Modes, Fees, Administrations, Services, Approved Languages and (if applicable) License Term

Licensee accepts and agrees to the terms of this Non-Commercial License Agreement (the "Agreement") from the Office of Scholarly Grants and Research (OGSR) of QualityMetric Incorporated ("QM") as of the Effective Date.

Subject to the terms of this Agreement, including the QualityMetric Non-Commercial License Terms and Conditions attached as Appendix A: (a) QM grants to Licensee, and Licensee accepts, a non-exclusive, non-transferable, non-assignable, non-sublicensable worldwide license to use, solely for the Approved Use and during the License Term, the Licensed Surveys in the authorized Modes and Approved Languages indicated on Appendix B and to administer the Licensed Surveys only up to the Approved Administrations (and to make up to such number of exact reproductions of the Licensed Surveys necessary to support such administrations) in any combination of the specific Licensed Surveys and Approved Languages and Modes and to use any related software provided by QM and (b) Licensee agrees to pay the Fee and other applicable charges in accordance with the attached invoice.

Capitalized terms used in this Agreement and not otherwise defined herein shall have the meanings assigned to them in Appendix A. The appendices attached hereto are incorporated into and made a part of this Agreement for all purposes.

EXECUTED, as of the Effective Date, by the duly authorized representatives as set forth below.

QualityMetric Incorporated	**City University London**
[QM] Signature: [redacted]	[Licensee] Signature: [redacted]
Name: Michelle White	Name: PROFESSOR STANTON NEWMAN
Title: Director of Consulting Science	Title: DEAN OF HEALTH SCIENCES.
Date: 12 MAR 2012	Date: 9TH MARCH 2012.

[redacted]

Appendix O. Missing value analysis for cross-sectional study

Variables	*Missing Subscales (%§)*
DEMOGRAPHIC	
Age	0
Gender	0
Marital status	0
Educational level	11 (3.5)
Employment status	11 (3.5)
CLINICAL	
Co-morbidities no.	3 (1)
Arrhythmias	3 (1)
Cyanosis days	12 (3.8)
Interventions no.	3 (1)
Hospitalization days	8 (2.5)
Medication no.	3 (1)
Current saturation	4 (1.3)
VO_2 Max	0
Right ventricular function	0
Left ventricular function	0
NYHA class	3 (1)
Total values: *n*= 5024	**Missing values: *n*= 61 (1.2%)**
PSYCHOSOCIAL	
SF-36	
Physical functioning	0
Role physical	1 (0.3)
Bodily pain	0
General health	0
Vitality	0
Social functioning	0
Role emotional	1 (0.3)
Mental health	0
Physical component summary	1 (0.3)
Mental component summary	1 (0.3)
CHD-TAAQOL	
Symptoms	2 (0.7)
Worries	0
Impact cardiac surveillance	1 (0.3)
Brief IPQ	
Consequences	1 (0.3)
Timeline	2 (0.7)
Controllability	1 (0.3)
Curability	9 (3)
Identity	0
Concern	0

Variables	*Missing Subscales (%)*
Coherence	1 (0.3)
Emotional representation	1
Brief COPE	
Planning	2 (0.7)
Active coping	2 (0.7)
Positive reframing	2 (0.7)
Acceptance	2 (0.7)
Humour	2 (0.7)
Religion	2 (0.7)
Seeking emotional support	2 (0.7)
Seeking instrumental support	2 (0.7)
Self-distraction	2 (0.7)
Denial	2 (0.7)
Venting	2 (0.7)
Substance use	2 (0.7)
Behavioural disengagement	2 (0.7)
Self-blame	2 (0.7)
STAI-6	0
CES-D 10	2 (0.7)
MSPSS	
Family support	0
Friends support	0
Special person support	0
Total values: *n*= 12120	**Missing values: *n*= 52 (0.4%)**

[§]Percentage of values missing *within* the particular scale/subscale.

Note. Missing value analysis for psychosocial questionnaires did not include the cases with incomplete psychosocial assessments (n=11) as these were not to be imputed.

Appendix P. Missing value analysis for longitudinal study

Variables	*Missing Subscales (%§)*
CLINICAL	
Interventions no.	0
Hospitalization days	0
Medication no.	0
Total values: *n*= 564	**Missing values: *n*= 0**
SF-36	
Physical functioning	0
Role physical	0
Bodily pain	1 (0.5)
General health	0
Vitality	2 (1.1)
Social functioning	1 (0.5)
Role emotional	1 (0.5)
Mental health	2 (1.1)
Physical component summary	2 (1.1)
Mental component summary	2 (1.1)
CHD-TAAQOL	
Symptoms	0
Worries	0
Impact cardiac surveillance	0
Brief IPQ	
Consequences	0
Timeline	0
Controllability	0
Curability	2 (1.1)
Identity	1 (0.5)
Concern	0
Coherence	0
Emotional representation	0
Brief COPE	
Planning	0
Active coping	0
Positive reframing	0
Acceptance	0
Humour	0
Religion	0
Seeking emotional support	0
Seeking instrumental support	0
Self-distraction	0
Denial	0
Venting	0
Substance use	0

Variables	*Missing Subscales (%)*
Behavioural disengagement	0
Self-blame	0
STAI-6	0
CES-D 10	0
Family support	0
Friends support	0
Special person support	0
Total values: *n*= 7520	**Missing values: *n*= 14 (0.2%)**

[§]Percentage of values missing *within* the particular scale/subscale.

Appendix Q. Correlation matrix of predictor variables

	1	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*	*11*	*12*	*13*	*14*
1. Age	-													
2. Gender§	**.158****	-												
3. Marital status§	**-.373****	**-.137***	-											
4. Education level§	**-.135***	-.005	.090	-										
5. Employment status§	-.006	**-.189****	-.099	.101	-									
6. TOF§	.081	.039	-.043	-.013	.032	-								
7. TGA§	-.087	-.093	.073	-.090	.064	**-.348****	-							
8. SV§	**-.232****	-.104	.007	.057	-.065	**-.310****	**-.310****	-						
9. Co-morbidities no.	**.315****	.014	-.077	-.042	**-.161****	-.050	.044	.087	-					
10. Arrhythmias§	**.152****	-.047	-.086	.008	-.102	-.106	**.208****	**.158****	**.552****	-				
11. Cyanosis days	.017	-.037	-.046	.102	-.033	.061	**-.158****	**.433****	**.241****	**.163***	-			
12. Interventions no.	-.096	**-.139***	-.008	**-.125***	**-.152****	.010	.109	**.304****	**.294****	**.401****	**.235****	-		
13. Hospitalisation days	.062	-.060	-.037	.016	**-.158****	.063	-.061	**.291****	**.404****	**.256****	**.228****	**.445****	-	
14. Medication no.	**.274****	-.054	-.099	-.096	**-.114***	**-.193****	.043	**.227****	**.544****	**.433****	**.250****	**.328****	**.310****	-

Continued

	1	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*	*11*	*12*	*13*	*14*
15. Current saturation	.032	.002	.024	.005	**.139***	**.170****	.042	**-.542****	**-.276****	**-.139***	**-.637****	**-.227****	**-.263****	**-.181****
16. VO_2 max	**-.283****	**-.264****	**.158****	**.158****	**.149****	-.056	-.058	-.063	**-.320****	**-.220****	**-.196****	**-.261****	**-.187****	**-.292****
17. LVEF	.031	.102	-.028	.019	.047	-.068	**.134***	**-.361****	**-.172****	**-.205****	**-.288****	**-.271****	**-.237****	**-.210****
18. RVEF	.062	**.125***	-.021	.104	.053	-.109	**-.213****	**-.109**	**-.133***	**-.297****	**-.168****	**-.384****	**-.220****	**-.169****
19. NYHA class§	**.190****	.097	-.082	-.101	**-.263****	.021	-.001	.107	**.457****	**.344****	**.246****	**.295****	**.365****	**.453****
20. Consequences	.055	.032	-.016	-.085	**-.279****	-.053	-.079	**.223****	**.310****	**.194****	**.183****	**.220****	**.245****	**.350****
21. Timeline	**-.118***	-.085	.056	.013	.042	.013	**.126***	.073	.013	.069	-.066	**.158****	.089	-.003
22. Controllability	.071	-.045	-.053	-.032	**.129***	.066	.034	-.039	-.001	-.045	-.011	.078	.017	-.026
23. Curability	.066	**-.123***	-.038	-.045	.005	**.214****	-.022	.009	.024	.101	.065	.138	.096	.100
24. Identity	**.135***	.082	-.044	**-.139***	**-.180****	-.094	-.057	**.253****	**.330****	**.243****	**.228****	**.213****	**.232****	**.361****
25. Concern	.031	-.015	.043	-.060	**-.184****	.045	-.066	.066	**.227****	**.150****	.105	.075	**.143***	**.323****
26. Coherence	**.169****	.106	-.080	**.144***	**-.114***	-.003	.020	**-.113***	**.147***	.023	-.008	-.028	.066	**.113***
27. Emotional representation	-.046	.109	.020	-.009	**-.233****	.006	-.081	**.160****	**.247****	**.143***	.091	.076	**.126***	**.224****
28. Planning	.102	**.117***	**-.116***	**.210****	.096	.048	**-.178****	.104	.013	.005	.065	.042	.028	.071

Continued

	1	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*	*11*	*12*	*13*	*14*
29. Active coping	.093	.101	-.099	**.189****	.061	.105	**-.121***	.031	.039	.013	.050	.037	.030	.014
30. Positive reframing	**.124***	**.132***	-.034	.021	.035	.016	-.037	-.046	-.048	-.048	.031	-.062	-.110	-.073
31. Acceptance	.106	.028	-.100	.069	.074	.033	**-.115***	**.123***	-.040	.091	.044	.054	-.010	-.006
32. Humour	.010	-.011	-.036	-.046	-.044	.002	-.072	.108	.010	-.004	.059	.060	-.036	.034
33. Religion	**.126***	**.214****	-.051	.085	-.047	.067	**-.116***	.043	.061	.048	**.219****	-.018	-.016	.021
34. Emotional support	-.053	**.195****	-.040	.061	-.032	.084	-.102	.044	.059	-.003	.027	.033	-.031	.093
35. Instrumental support	-.074	.105	-.005	.052	.008	.080	-.005	.007	.053	.016	.006	-.012	-.001	.057
36. Self-distraction	-.045	**.165****	.034	.068	.027	-.023	**-.175****	**.133***	.022	-.028	**.119***	-.043	.017	.089
37. Denial	**.114***	.104	.005	-.075	.004	-.021	-.016	.033	.025	.023	.095	-.029	.009	**.113***
38. Venting	-.019	.064	-.012	-.029	-.094	-.001	**-.126***	**.138***	**.116***	.063	.049	.064	.068	.069
39. Substance use	.014	-.095	**.121***	.021	**-.146***	-.047	-.103	.068	.086	.099	-.011	.082	.081	.024
40. Behavioural disengagement	-.031	-.022	.077	-.090	-.086	-.075	-.048	.090	.082	.007	.051	.052	-.024	.093
41. Self-blame	**.124***	.093	-.026	-.023	-.085	-.074	-.017	.029	**.130***	.057	-.036	-.031	-.032	.087
42. Anxiety	.010	**.157****	.072	-.070	**-.161****	-.050	-.074	**.118***	**.174****	.061	.045	-.016	**.114***	**.139***

Continued

	1	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*	*11*	*12*	*13*	*14*
43. Depression[§]	-.022	.079	.040	-.028	**-.246****	-.047	**-.124***	.075	**.165****	-.011	.098	-.052	.101	**.117***
44. Family support	.059	**.131***	-.101	.041	.006	.109	-.010	-.084	-.009	.025	-.028	.038	**-.137***	.052
45. Friends support	.036	**.185****	-.028	-.011	.089	.003	.001	-.087	-.088	-.033	-.023	-.004	**-.176****	-.019
46. Special person support	**.176****	**.224****	**-.390****	-.062	**.120***	.112	-.038	**-.134***	-.005	.029	-.076	-.024	-.055	.022

Continued

	15	*16*	*17*	*18*	*19*	*20*	*21*	*22*	*23*	*24*	*25*	*26*	*27*	*28*
15. Current saturation	-													
16. VO_2 max	**.204***	-												
17. LVEF	**.291****	**.151****	-											
18. RVEF	**.169****	**.225****	**.566****	-										
19. NYHA class§	**-.299****	**-.291****	**-.149****	**-.191****	-									
20. Consequences	**-.263****	**-.256****	**-.224****	**-.160****	**.331****	-								
21. Timeline	-.028	-.065	.056	-.089	.026	.092	-							
22. Controllability	.040	-.038	.057	-.003	-.071	**-.211****	-.083	-						
23. Curability	.027	-.045	-.078	**-.168****	.027	-.066	.036	**.264****	-					
24. Identity	**-.325****	**-.290****	**-.219****	-.112	**.375****	**.709****	.064	**-.159****	-.075	-				
25. Concern	**-.147***	**-.149****	**-.149****	**-.128***	**.346****	**.582****	.062	**-.176****	.019	**.536****	-			
26. Coherence	.051	-.037	-.001	-.001	.011	-.005	-.030	**.155****	**.141***	-.068	-.026	-		
27. Emotional representation	**-.159****	**-.158****	**-.138***	**-.116***	**.247****	**.643****	.002	**-.221****	-.077	**.545****	**.646****	-.020	-	
28. Planning	-.021	-.010	-.109	-.025	.008	**.115***	-.077	.076	.111	.104	.073	**.128***	.105	-

Continued

	15	*16*	*17*	*18*	*19*	*20*	*21*	*22*	*23*	*24*	*25*	*26*	*27*	*28*
29. Active coping	.028	.006	-.006	-.024	.037	.104	.020	**.185****	**.199****	.081	.088	.105	.007	**.750****
30. Positive reframing	.056	-.051	.015	-.012	.024	-.042	-.113	**.219****	.081	-.022	-.036	.041	-.056	**.435****
31. Acceptance	-.008	-.067	-.074	-.101	-.037	.017	-.005	.038	.051	.028	-.095	**.113***	**-.115***	**.198****
32. Humour	.003	-.010	-.042	-.002	.079	.092	-.107	.108	**.141***	.094	-.088	-.054	.057	**.201****
33. Religion	-.089	**-.166****	-.087	-.018	**.120***	.099	**-.141***	-.004	.062	**.129***	.078	-.003	**.137***	**.121***
34. Emotional support	-.035	**-.115***	-.091	-.101	.043	**.120***	-.047	.087	**.116***	**.166****	**.119***	.061	**.153****	**.305****
35. Instrumental support	.002	-.023	-.089	-.078	-.015	.084	.002	.087	**.119***	.060	**.165****	.085	.106	**.409****
36. Self-distraction	**-.120***	-.089	**-.124***	-.059	.057	**.271****	-.019	.010	.017	**.209****	**.201****	.006	**.287****	**.381****
37. Denial	-.064	-.093	-.086	.006	.073	**.118***	-.082	.001	-.013	**.128***	**.126***	-.045	**.182****	**.142***
38. Venting	**-.113***	-.089	-.019	-.016	.094	**.345****	.032	-.100	.017	**.299****	**.316****	.003	**.511****	**.219****
39. Substance use	-.021	-.054	-.008	.034	**.115***	**.187****	-.029	-.079	-.065	**.134***	**.169****	**-.161****	**.215****	.077
40. Behavioural disengagement	**-.142***	-.070	-.054	.018	.043	**.291****	.031	**-.177****	**-.148***	**.249****	**.259****	**-.219****	**.400****	.085
41. Self-blame	-.019	-.087	-.002	.051	.091	**.237****	.039	-.108	-.053	**.163****	**.162****	-.008	**.306****	**.208****
42. Anxiety	**-.130***	**-.136***	-.051	.013	**.200****	**.365****	-.036	**-.283****	**-.256****	**.272****	**.316****	**-.129***	**.476****	-.046

Continued

	15	*16*	*17*	*18*	*19*	*20*	*21*	*22*	*23*	*24*	*25*	*26*	*27*	*28*
43. Depression[§]	**-.160**[**]	-.092	-.016	.000	**.173**[**]	**.400**[**]	-.007	**-.205**[**]	**-149**[**]	**.333**[**]	**.313**[**]	-.027	**.518**[**]	.013
44. Family support	.043	-.093	.076	.015	-.023	**-.133**[*]	-.032	**.196**[**]	**.163**[**]	-.075	-.068	.086	-.082	**.131**[*]
45. Friends support	.053	-.087	.041	.024	-.087	**-.170**[**]	-.023	**.186**[**]	**.119**[*]	-.057	-.088	.030	**-.132**[*]	.075
46. Special person support	.072	**-.127**[*]	.093	.036	.071	-.107	-.048	.083	**.145**[*]	-.035	-.071	.021	**-.118**[*]	**.198**[**]

Continued

	29	*30*	*31*	*32*	*33*	*34*	*35*	*36*	*37*	*38*	*39*	*40*	*41*	*42*
29. Active coping	-													
30. Positive reframing	**.516***	-												
31. Acceptance	**.277****	**.382****	-											
32. Humour	**.269****	**.358****	**.216****	-										
33. Religion	.067	**.172****	**.133***	.056	-									
34. Emotional support	**.259****	**.256****	**.150****	**.217****	**.238****	-								
35. Instrumental support	**.379****	**.217****	.054	**.130***	**.150****	**.637****	-							
36. Self-distraction	**.319****	**.216****	**.185****	**.216****	.087	**.390****	**.359****	-						
37. Denial	.066	.008	**-.131***	**.119***	.090	.075	.105	**.242****	-					
38. Venting	**.156****	.037	.017	**.122***	.074	**.251****	**.306****	**.339****	**.190****	-				
39. Substance use	.005	.007	-.033	.072	-.100	-.017	.001	**.169****	.012	**.207****	-			
40. Behavioural disengagement	-.070	-.074	**-.162****	.040	.105	.015	.046	**.174****	**.339****	**.320****	**.277****	-		
41. Self-blame	.057	-.035	-.108	.098	.020	.109	**.189****	**.254****	**.226****	**.396****	**.154****	**.402****	-	
42. Anxiety	**-.178****	**-.244****	**-.245****	**-.133***	.047	-.036	-.020	.073	**.182****	**.271****	**.208****	**.357****	**.307****	-

Continued

	29	*30*	*31*	*32*	*33*	*34*	*35*	*36*	*37*	*38*	*39*	*40*	*41*	*42*
43. Depression[§]	-.106	**-.181****	**-.173****	-.022	.071	.042	.035	**.169****	**.185****	**.354****	**.215****	**.411****	**.356****	**.508****
44. Family support	**.135***	**.151****	**.145***	.081	.090	**.351****	**.196****	.064	-.022	-.011	**-.210****	**-.133***	-.093	**-.259****
45. Friends support	.079	**.185****	**.144***	**.126***	**.164****	**.277****	**.158****	.078	-.014	.017	**-.172****	-.104	-.091	**-.235****
46. Special person support	**.231****	**.197****	**.127***	.094	.049	**.267****	**.133***	.065	.054	-.009	**-.154****	-.062	-.062	**-.166****

Continued

	43	*44*	*45*	*46*
43. Depression[§]	-			
44. Family support	**-.176****	-		
45. Friends support	**-.169****	**.454****	-	
46. Special person support	**-.158****	**.413****	**.322****	-

*p<.05, **p<.01.

[§] Gender: 0= male, 1= female, Marital status: 0= married/in relationship, 1= single, Educational level: 0= school level, 1= university level, Employment status: 0= unemployed, 1= employed, TOF, TGA, SV: dummy-coded diagnostic group, Arrhythmias: 0= no arrhythmias, 1= arrhythmias, NYHA class: 0= Class I, 1= Classes II, III, IV, Depression: 0= no depressive symptoms, 1= with depressive symptoms.

Appendix R. Bivariate regressions: factors associated with generic physical and psychosocial and disease-specific HRQoL

Predictor Variable	*Physical HRQOL*					*Psychosocial HRQOL*					*Disease-specific HRQoL*		
	Physical Component Summary	***Physical Functioning***	***Role Physical***	***Bodily Pain***	***General Health***	***Mental Component Summary***	***Vitality***	***Social Functioning***	***Role Emotional***	***Mental Health***	***Symptoms***	***Impact Cardiac Surveillance***	***Worries***
Age	**-.153***	-.125	-.060	-.056	-.013	**.160***	-.033	.101	.065	**.164***	-.108	-.008	**.153***
Gender§	-.112	**-.163***	-.049	-.138	-.019	-.056	**-.170***	-.037	-.056	-.075	**-.155***	.004	-.030
Educational level§	**.156***	.124	.054	.135	**.181***	-.001	.032	.053	-.010	.061	.112	.033	-.069
Employment status§	**.212****	**.290****	**.287****	.126	**.211****	**.261****	**.284****	**.256****	**.251****	**.261****	**.289****	.111	.147
ToF§	.118	.103	.101	.129	.096	.099	**.151***	**.151***	.056	.098	.072	.092	.099
SV§	**-.170***	**-.238****	-.144	-.083	-.133	-.098	-.097	**-.214****	-.075	-.116	**-.196***	**-.188***	**-.186***
Co-morbidities no.	**-.312****	**-.381****	**-.182***	**-.178***	**-.269****	-.095	**-.227****	**-.205****	-.138	-.083	**-.318****	**-.268****	**-.171***
Arrhythmias§	-.135	**-.198***	-.015	-.008	-.138	.043	-.029	-.022	.018	.016	**-.166***	**-.290****	-.057
Cyanosis days	**-.261****	**-.289****	**-.242****	-.093	-.113	.021	-.068	-.143	.005	-.039	**-.180***	-.102	-.101
Intervention no.	-.131	**-.182***	-.050	.059	**-.152***	.059	.019	-.069	.060	.022	-.124	**-.222****	-.034
Hospitalization days	**-.235****	**-.294****	-.149	-.111	**-.195***	-.048	-.094	**-.182***	-.063	-.075	**-.183***	-.146	-.145
Medication no.	**-.305****	**-.340****	-.143	**-.171***	**-.322****	-.076	**-.209****	**-.194***	-.073	-.093	**-.305****	**-.295****	-.133

Continued

Predictor Variable	***Physical HRQOL***					***Psychosocial HRQOL***					***Disease-specific HRQoL***		
	Physical Component Summary	***Physical Functioning***	***Role Physical***	***Bodily Pain***	***General Health***	***Mental Component Summary***	***Vitality***	***Social Functioning***	***Role Emotional***	***Mental Health***	***Symptoms***	***Impact Cardiac Surveillance***	***Worries***
Current O_2 saturation	**.334****	**.411****	**.279****	**.152***	**.214****	.087	**.198***	**.270****	.072	.137	**.323****	**.215****	**.205****
VO_2 max	**.266****	**.331****	.130	.124	**.224****	.022	.136	.139	.034	.073	**.320****	**.193***	.114
RVEF	.090	.098	.042	-.071	**.157***	-.061	-.056	.021	-.070	-.010	.120	.125	.134
LVEF	.137	**.209****	.085	.029	.114	.028	.016	.124	.025	.056	**.192***	.084	**.142**
Brief IPQ consequences	**-.553****	**-.614****	**-.499****	**-.426****	**-.545****	**-.426****	**-.544****	**-.581****	**-.378****	**-.425****	**-.615****	**-.432****	**-.557****
Brief IPQ controllability	**.155***	.145	**.158***	**.192***	**.232****	**.227****	**.238****	**.183***	**.183***	**.243****	**.166***	.138	**.186***
Brief IPQ curability	.073	.085	.062	.103	**.154***	**.165***	**.171***	.113	.141	.140	.072	.016	**.161***
Brief IPQ identity	**-.677****	**-.667****	**-.512****	**-.541****	**-.597****	**-.332****	**-.587****	**-.555****	**-.288****	**-.344****	**-.698****	**-.444****	**-.480****
Brief IPQ concern	**-.406****	**-.441****	**-.316****	**-.365****	**-.474****	**-.352****	**-.411****	**-.415****	**-.272****	**-.386****	**-.508****	**-.416****	**-.476****
Brief IPQ emotional representation	**-.362****	**-.405****	**-.374****	**-.392****	**-.504****	**-.527****	**-.512****	**-.493****	**-.415****	**-.539****	**-.550****	**-.455****	**-.550****

Continued

Predictor Variable	***Physical HRQOL***					***Psychosocial HRQOL***					***Disease-specific HRQoL***		
	Physical Component Summary	***Physical Functioning***	***Role Physical***	***Bodily Pain***	***General Health***	***Mental Component Summary***	***Vitality***	***Social Functioning***	***Role Emotional***	***Mental Health***	***Symptoms***	***Impact Cardiac Surveillance***	***Worries***
Brief COPE planning	-.061	-.041	-.052	-.109	-.011	-.037	-.064	-.046	-.062	-.013	-.056	**-.168***	**-.154***
Brief COPE positive reframing	-.056	-.011	.027	-.044	.137	**.224****	.146	.062	**.152***	**.212****	.007	.069	.103
Brief COPE acceptance	-.015	.050	.000	.057	.107	**.222****	**.166***	.101	**.152***	**.218****	.047	.061	.068
Brief COPE religion	**-.156***	**-.153***	-.101	**-.168***	-.049	-.007	-.044	-.077	-.047	-.022	-.133	-.108	-.110
Brief COPE self-distraction	-.118	-.147	-.124	-.147	-.140	**-.212****	**-.240****	**-.199****	**-.172***	**-.175***	**-.214****	-.094	**-.258****
Brief COPE denial	-.133	**-.205****	-.110	**-.207****	-.072	**-.188***	**-.214****	**-.154***	-.136	**-.229****	**-.190***	-.145	-.117
Brief COPE venting	**-.170***	**-.174***	**-.244****	**-.251****	**-.255****	**-.361****	**-.319****	**-.280****	**-.297****	**-.357****	**-.294****	**-.253****	**-.285****
Brief COPE substance use	-.108	**-.197***	-.139	-.039	**-.171***	**-.202****	**-.185***	**-.191***	**-.192***	**-.175***	**-.165***	**-.150***	**-.234****

Continued

Predictor Variable	***Physical HRQOL***					***Psychosocial HRQOL***					***Disease-specific HRQoL***		
	Physical Component Summary	***Physical Functioning***	***Role Physical***	***Bodily Pain***	***General Health***	***Mental Component Summary***	***Vitality***	***Social Functioning***	***Role Emotional***	***Mental Health***	***Symptoms***	***Impact Cardiac Surveillance***	***Worries***
Brief COPE behavioural disengagement	**-.192***	**-.264****	**-.232****	**-.239****	**-.272****	**-.374****	**-.339****	**-.289****	**-.301****	**-.396****	**-.307****	**-.290****	**-.280****
Brief COPE self-blame	-.072	**-.178***	**-.194***	**-.155***	**-.179***	**-.419****	**-.352****	**-.217****	**-.372****	**-.401****	**-.253****	-.125	**-.303****
MSPSS family support	.084	.103	.074	.143	.082	.136	.105	.146	.085	**.160***	.091	.040	**.170***
MSPSS friend support	.033	.106	.107	.051	.097	**.220****	.127	**.165***	**.212****	**.194***	.075	.072	**.189***
MSPSS significant other support	-.031	.014	.052	-.026	.079	**.180***	.087	.117	.142	**.151***	.043	.013	.146
STAI-6 anxiety	**-.243****	**-.364****	**-.313****	**-.333****	**-.317****	**-.525****	**-.447****	**-.420****	**-.429****	**-.550****	**-.422****	**-.342****	**-.402****
CES-D 10 depression§	**-.261****	**-.361****	**-.444****	**-.327****	**-.427****	**-.686****	**-.598****	**-.513****	**-.585****	**-.652****	**-.510****	**-.274****	**-.507****

Note. Values represent standardised beta coefficients.
*p< .01, **p< .001. §Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.
Note. Beta coefficients are standardized. Marital status, TGA, brief IPQ timeline, brief IPQ coherence, brief COPE active coping, brief COPE humour, brief COPE emotional support, and brief COPE instrumental support were excluded from the table because they did not have a significant relationship with any of the outcome variables.

Appendix S. Full hierarchical multiple regressions

Physical component summary of the SF-36 (*n*= 301)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.079	.069	.079			**<.001**
	Education level				.117	.115	.040
	Employment status				.203	.202	**<.001**
	Age				-.119	-.118	.036
2		.195	.172	.116			**<.001**
	Education level				.146	.141	**.008**
	Employment status				.153	.149	**.005**
	Age				-.065	-.057	.283
	ToF				.109	.087	.098
	TGA				.010	.008	.884
	SV				-.046	-.032	.537
	Cyanosis days				-.185	-.156	**.003**
	Co-morbidities no.				-.213	-.191	**<.001**
3		.202	.178	.008			.094
	Education level				.150	.144	**.006**
	Employment status				.143	.139	**.008**
	Age				-.061	-.053	.316
	ToF				.135	.105	.047
	TGA				.024	.019	.721
	SV				-.007	-.005	.926
	Cyanosis days				-.185	-.157	**.003**
	Co-morbidities no.				-.177	-.149	**.005**
	Hospitalization days				-.103	-.088	.094
4		.235	.203	.033			**.007**
	Education level				.121	.115	.026
	Employment status				.113	.107	.038
	Age				-.017	-.014	.783
	ToF				.125	.095	.065
	TGA				.071	.054	.294
	SV				.091	.057	.273
	Cyanosis days				-.076	-.054	.295
	Co-morbidities no.				-.084	-.062	.227
	Hospitalization days				-.092	-.078	.130
	Current O_2 saturation				.183	.122	.019
	Medication no.				-.125	-.095	.066
	VO_2 max				.104	.091	.078
5		.510	.481	.275			**<.001**
	Education level				.062	.059	.158
	Employment status				.050	.046	.272
	Age				-.016	-.013	.761
	ToF				.104	.078	.061
	TGA				.025	.019	.651
	SV				.119	.073	.080
	Cyanosis days				-.084	-.060	.153
	Co-morbidities no.				-.032	-.024	.570
	Hospitalization days				-.065	-.055	.187
	Current O_2 saturation				.069	.046	.272
	Medication no.				.009	.006	.880
	VO_2 max				.026	.022	.589
	Brief IPQ consequences				-.126	-.077	.067
	Brief IPQ controllability				.046	.044	.296

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief IPQ identity				-.517	-.334	**<.001**
	Brief IPQ concern				-.055	-.038	.356
	Brief IPQ emotional representation				.073	.048	.246
6		.515	.481	.005			.418
	Education level				.068	.063	.129
	Employment status				.046	.042	.312
	Age				-.011	-.009	.835
	ToF				.102	.077	.066
	TGA				.023	.017	.676
	SV				.115	.070	.092
	Cyanosis days				-.065	-.045	.276
	Co-morbidities no.				-.033	-.025	.555
	Hospitalization days				-.071	-.060	.150
	Current O_2 saturation				.076	.050	.235
	Medication no.				.007	.005	.905
	VO_2 max				.018	.015	.711
	Brief IPQ consequences				-.129	-.078	.061
	Brief IPQ controllability				.044	.042	.311
	Brief IPQ identity				-.513	-.331	**<.001**
	Brief IPQ concern				-.057	-.040	.341
	Brief IPQ emotional representation				.067	.041	.330
	Brief COPE religion				-.063	-.059	.160
	Brief COPE venting				.042	.035	.401
	Brief COPE behavioural disengagement				-.017	-.015	.719
7		.516	.478	.001			.747
	Education level				.066	.061	.143
	Employment status				.045	.041	.323
	Age				-.010	-.008	.840
	ToF				.099	.074	.077
	TGA				.020	.015	.720
	SV				.114	.069	.101
	Cyanosis days				-.066	-.046	.274
	Co-morbidities no.				-.032	-.023	.578
	Hospitalization days				-.069	-.058	.167
	Current O_2 saturation				.075	.049	.242
	Medication no.				.006	.004	.921
	VO_2 max				.015	.013	.753
	Brief IPQ consequences				-.126	-.076	.068
	Brief IPQ controllability				.038	.035	.403
	Brief IPQ identity				-.517	-.332	**<.001**
	Brief IPQ concern				-.057	-.040	.340
	Brief IPQ emotional representation				.081	.047	.264
	Brief COPE religion				-.064	-.060	.154
	Brief COPE venting				.042	.035	.400
	Brief COPE behavioural disengagement				-.010	-.008	.841
	STAI-6 anxiety				-.037	-.030	.479
	CES-D 10 depression				-.003	-.003	.951

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Physical functioning subscale of the SF-36 (*n*= 301)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.099	.093	.099			**<.001**
	Gender				-.104	-.102	.064
	Employment status				.279	.274	**<.001**
2		.273	.253	.174			**<.001**
	Gender				-.139	-.134	**.008**
	Employment status				.210	.202	**<.001**
	ToF				.043	.034	.490
	TGA				-.028	-.022	.663
	SV				-.151	-.108	.030
	Cyanosis days				-.134	-.115	.022
	Co-morbidities no.				-.329	-.267	**<.001**
	Arrhythmias				.044	.035	.483
3		.283	.258	.009			.151
	Gender				-.145	-.139	**.006**
	Employment status				.198	.189	**<.001**
	ToF				.072	.055	.269
	TGA				-.011	-.008	.865
	SV				-.107	-.070	.158
	Cyanosis days				-.134	-.114	.022
	Co-morbidities no.				-.286	-.218	**<.001**
	Arrhythmias				.044	.034	.493
	Intervention no.				-.012	-.009	.854
	Hospitalization days				-.110	-.090	.070
4		.346	.314	.064			**<.001**
	Gender				-.115	-.103	.032
	Employment status				.167	.156	**.001**
	ToF				.063	.047	.330
	TGA				.036	.026	.588
	SV				.016	.010	.841
	Cyanosis days				.016	.010	.829
	Co-morbidities no.				-.145	-.100	.038
	Arrhythmias				.037	.028	.558
	Intervention no.				.038	.029	.541
	Hospitalization days				-.103	-.084	.079
	Current O_2 saturation				.241	.159	**.001**
	Medication no.				-.157	-.120	.012
	VO_2 max				.125	.106	.027
	LVEF				.080	.071	.140
5		.573	.546	.227			**<.001**
	Gender				-.102	-.090	.021
	Employment status				.099	.090	.021
	ToF				.044	.032	.415
	TGA				-.009	-.007	.860
	SV				.014	.008	.832
	Cyanosis days				-.004	-.003	.940

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Co-morbidities no.				-.112	-.076	.050
	Arrhythmias				.050	.038	.333
	Intervention no.				.044	.034	.387
	Hospitalization days				-.078	-.063	.105
	Current O_2 saturation				.137	.089	.022
	Medication no.				-.017	-.013	.745
	VO_2 max				.059	.049	.209
	LVEF				.026	.023	.549
	Brief IPQ consequences				-.234	-.142	**<.001**
	Brief IPQ identity				-.374	-.244	**<.001**
	Brief IPQ concern				-.094	-.065	.095
	Brief IPQ emotional representation				.102	.068	.083
6		.605	.571	.032			**.001**
	Gender				-.106	-.091	.017
	Employment status				.091	.082	.030
	ToF				.016	.011	.766
	TGA				-.032	-.022	.554
	SV				-.013	-.008	.837
	Cyanosis days				.015	.011	.778
	Co-morbidities no.				-.123	-.082	.030
	Arrhythmias				.061	.046	.229
	Intervention no.				.039	.030	.433
	Hospitalization days				-.081	-.065	.085
	Current O_2 saturation				.140	.089	.018
	Medication no.				-.015	-.011	.771
	VO_2 max				.041	.034	.365
	LVEF				.013	.011	.767
	Brief IPQ consequences				-.231	-.139	**<.001**
	Brief IPQ identity				-.375	-.244	**<.001**
	Brief IPQ concern				-.093	-.064	.092
	Brief IPQ emotional representation				.119	.071	.061
	Brief COPE religion				-.045	-.041	.282
	Brief COPE denial				-.107	-.097	.011
	Brief COPE venting				.104	.083	.028
	Brief COPE substance use				-.109	-.099	**.009**
	Brief COPE behavioural disengagement				-.046	-.037	.333
	Brief COPE self-blame				-.025	-.021	.583
7		.618	.581	.012			.014
	Gender				-.096	-.081	.030
	Employment status				.083	.074	.048
	ToF				.010	.007	.842
	TGA				-.037	-.026	.484
	SV				-.013	-.008	.833
	Cyanosis days				.016	.011	.768
	Co-morbidities no.				-.116	-.078	.037
	Arrhythmias				.056	.041	.270

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Intervention no.				.019	.015	.696
	Hospitalization days				-.066	-.053	.159
	Current O_2 saturation				.132	.084	.025
	Medication no.				-.014	-.010	.786
	VO_2 max				.038	.031	.400
	LVEF				.018	.015	.680
	Brief IPQ consequences				-.216	-.130	**.001**
	Brief IPQ identity				-.381	-.246	**<.001**
	Brief IPQ concern				-.096	-.066	.079
	Brief IPQ emotional representation				.172	.098	**.009**
	Brief COPE religion				-.049	-.045	.231
	Brief COPE denial				-.102	-.093	.013
	Brief COPE venting				.102	.082	.029
	Brief COPE substance use				-.099	-.089	.018
	Brief COPE behavioural disengagement				-.020	-.015	.681
	Brief COPE self-blame				-.004	-.003	.929
	STAI-6 anxiety				-.103	-.082	.029
	CES-D 10 depression				-.064	-.047	.204

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Role physical subscale of the SF-36 (*n*= 301)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.085	.082	.085			**<.001**
	Employment status				.291	.291	**<.001**
2		.140	.131	.055			**<.001**
	Employment status				.270	.266	**<.001**
	Cyanosis days				-.196	-.192	**<.001**
	Co-morbidities no.				-.093	-.089	.098
3		.148	.137	.009			.082
	Employment status				.253	.246	**<.001**
	Cyanosis days				-.122	-.094	.081
	Co-morbidities no.				-.079	-.075	.165
	Current O_2 saturation				.124	.093	.082
4		.342	.322	.194			**<.001**
	Employment status				.168	.157	**.001**
	Cyanosis days				-.111	-.085	.075
	Co-morbidities no.				.043	.039	.412
	Current O_2 saturation				.019	.014	.774
	Brief IPQ consequences				-.203	-.125	**.009**
	Brief IPQ controllability				.041	.040	.403
	Brief IPQ identity				-.305	-.203	**<.001**
	Brief IPQ concern				.051	.037	.443
	Brief IPQ emotional representation				-.062	-.042	.375
5		.351	.324	.009			.275
	Employment status				.171	.160	**.001**
	Cyanosis days				-.118	-.091	.057
	Co-morbidities no.				.045	.041	.392
	Current O_2 saturation				.012	.009	.850
	Brief IPQ consequences				-.194	-.119	.013
	Brief IPQ controllability				.035	.033	.484
	Brief IPQ identity				-.306	-.203	**<.001**
	Brief IPQ concern				.044	.031	.509
	Brief IPQ emotional representation				-.010	-.006	.898
	Brief COPE venting				-.033	-.027	.574
	Brief COPE behavioural disengagement				-.051	-.044	.355
	Brief COPE self-blame				-.057	-.049	.302
6		.398	.369	.047			**<.001**
	Employment status				.140	.130	**.005**
	Cyanosis days				-.113	-.086	.061
	Co-morbidities no.				.048	.044	.342
	Current O_2 saturation				.001	.001	.984
	Brief IPQ consequences				-.176	-.108	.020
	Brief IPQ controllability				.009	.009	.850
	Brief IPQ identity				-.304	-.201	**<.001**
	Brief IPQ concern				.027	.020	.669
	Brief IPQ emotional representation				.089	.052	.254
	Brief COPE venting				-.022	-.018	.694

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief COPE behavioural disengagement				.001	.000	.999
	Brief COPE self-blame				-.013	-.011	.811
	STAI-6 anxiety				-.058	-.046	.313
	CES-D 10 depression				-.249	-.190	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Bodily pain subscale of the SF-36 (*n*= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.032	.029	.032			**.002**
	Co-morbidities no.				-.178	-.178	**.002**
2		.051	.041	.019			.052
	Co-morbidities no.				-.095	-.078	.169
	Current O_2 saturation				.110	.106	.061
	Medication no.				-.100	-.084	.138
3		.317	.299	.266			**<.001**
	Co-morbidities no.				-.019	-.016	.745
	Current O_2 saturation				-.023	-.021	.660
	Medication no.				.044	.035	.469
	Brief IPQ consequences				-.005	-.003	.948
	Brief IPQ controllability				.090	.087	.072
	Brief IPQ identity				-.462	-.307	**<.001**
	Brief IPQ concern				-.051	-.036	.449
	Brief IPQ emotional representation				-.093	-.063	.194
4		.344	.314	.027			.041
	Co-morbidities no.				-.030	-.024	.614
	Current O_2 saturation				-.035	-.033	.494
	Medication no.				.053	.042	.383
	Brief IPQ consequences				-.011	-.007	.883
	Brief IPQ controllability				.096	.092	.054
	Brief IPQ identity				-.448	-.296	**<.001**
	Brief IPQ concern				-.057	-.040	.400
	Brief IPQ emotional representation				-.030	-.019	.696
	Brief COPE religion				-.086	-.084	.077
	Brief COPE denial				-.120	-.111	.021
	Brief COPE venting				-.037	-.030	.529
	Brief COPE behavioural disengagement				-.027	-.022	.638
	Brief COPE self-blame				.002	.002	.970
5		.365	.332	.021			**.009**
	Co-morbidities no.				-.019	-.015	.749
	Current O_2 saturation				-.045	-.041	.382
	Medication no.				.048	.038	.417
	Brief IPQ consequences				.006	.004	.936
	Brief IPQ controllability				.067	.063	.182
	Brief IPQ identity				-.455	-.300	**<.001**
	Brief IPQ concern				-.061	-.043	.358
	Brief IPQ emotional representation				.040	.024	.617
	Brief COPE religion				-.090	-.088	.061
	Brief COPE denial				-.110	-.102	.031
	Brief COPE venting				-.038	-.031	.515
	Brief COPE behavioural disengagement				.003	.002	.961
	Brief COPE self-blame				.030	.025	.590
	STAI-6 anxiety				-.138	-.110	.020
	CES-D 10 depression				-.077	-.059	.207

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

General health subscale of the SF-36 (*n*= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.071	.064	.071			**<.001**
	Education level				.162	.161	**.004**
	Employment status				.195	.194	**.001**
2		.125	.116	.054			**<.001**
	Education level				.156	.155	**.004**
	Employment status				.156	.154	**.005**
	Co-morbidities no.				-.237	-.233	**<.001**
3		.133	.118	.008			.271
	Education level				.159	.157	**.004**
	Employment status				.147	.143	**.009**
	Co-morbidities no.				-.198	-.178	**.001**
	Intervention no.				-.013	-.012	.827
	Hospitalization days				-.090	-.076	.160
4		.191	.167	.059			**<.001**
	Education level				.134	.130	.014
	Employment status				.138	.134	.011
	Co-morbidities no.				-.059	-.046	.383
	Intervention no.				.069	.057	.281
	Hospitalization days				-.061	-.051	.333
	Current O_2 saturation				.115	.108	.041
	Medication no.				-.216	-.176	**.001**
	VO_2 max				.063	.056	.286
	RVEF				.079	.072	.169
5		.465	.437	.273			**<.001**
	Education level				.113	.108	.013
	Employment status				.043	.040	.359
	Co-morbidities no.				.001	.001	.991
	Intervention no.				.027	.022	.616
	Hospitalization days				-.047	-.039	.368
	Current O_2 saturation				.007	.006	.881
	Medication no.				-.093	-.072	.095
	VO_2 max				.014	.013	.772
	RVEF				.069	.062	.152
	Brief IPQ consequences				-.064	-.039	.367
	Brief IPQ controllability				.080	.074	.089
	Brief IPQ curability				.122	.113	**.009**
	Brief IPQ identity				-.319	-.208	**<.001**
	Brief IPQ concern				-.093	-.065	.131
	Brief IPQ emotional representation				-.155	-.104	.017
6		.468	.432	.003			.769
	Education level				.111	.106	.015
	Employment status				.040	.037	.398
	Co-morbidities no.				.003	.002	.956
	Intervention no.				.033	.026	.544
	Hospitalization days				-.050	-.041	.344
	Current O_2 saturation				.007	.006	.885
	Medication no.				-.096	-.074	.089
	VO_2 max				.013	.011	.799
	RVEF				.077	.068	.119

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief IPQ consequences				-.058	-.035	.418
	Brief IPQ controllability				.076	.070	.106
	Brief IPQ curability				.116	.107	.014
	Brief IPQ identity				-.322	-.209	**<.001**
	Brief IPQ concern				-.091	-.064	.143
	Brief IPQ emotional representation				-.138	-.084	.053
	Brief COPE venting				.009	.007	.868
	Brief COPE substance use				-.044	-.041	.341
	Brief COPE behavioural disengagement				-.030	-.025	.569
	Brief COPE self-blame				-.015	-.013	.766
7		.488	.449	.020			**.005**
	Education level				.112	.106	.013
	Employment status				.017	.016	.712
	Co-morbidities no.				.004	.003	.939
	Intervention no.				.004	.003	.935
	Hospitalization days				-.036	-.030	.485
	Current O_2 saturation				-.001	-.001	.986
	Medication no.				-.095	-.073	.088
	VO_2 max				.017	.015	.732
	RVEF				.073	.065	.131
	Brief IPQ consequences				-.049	-.030	.486
	Brief IPQ controllability				.075	.068	.110
	Brief IPQ curability				.111	.101	.019
	Brief IPQ identity				-.313	-.202	**<.001**
	Brief IPQ concern				-.105	-.073	.087
	Brief IPQ emotional representation				-.093	-.054	.205
	Brief COPE venting				.020	.016	.713
	Brief COPE substance use				-.038	-.035	.409
	Brief COPE behavioural disengagement				-.002	-.001	.975
	Brief COPE self-blame				.005	.004	.919
	STAI-6 anxiety				.037	.029	.498
	CES-D 10 depression				-.186	-.140	**.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Mental component summary of the SF-36 (*n*= 302)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.095	.089	.095			**<.001**
	Age				.165	.165	**.003**
	Employment status				.264	.264	**<.001**
2		.345	.327	.249			**<.001**
	Age				.143	.139	**.004**
	Employment status				.126	.121	.011
	Brief IPQ consequences				-.123	-.076	.111
	Brief IPQ controllability				.058	.054	.251
	Brief IPQ curability				.101	.097	.042
	Brief IPQ identity				-.017	-.012	.807
	Brief IPQ concern				.005	.004	.939
	Brief IPQ emotional representation				-.385	-.260	**<.001**
3		.465	.435	.121			**<.001**
	Age				.160	.149	**.001**
	Employment status				.125	.117	**.007**
	Brief IPQ consequences				-.090	-.055	.209
	Brief IPQ controllability				.019	.017	.699
	Brief IPQ curability				.088	.083	.056
	Brief IPQ identity				-.041	-.028	.521
	Brief IPQ concern				-.010	-.007	.872
	Brief IPQ emotional representation				-.232	-.142	**.001**
	Brief COPE positive reframing				.143	.126	**.004**
	Brief COPE acceptance				.083	.072	.098
	Brief COPE self-distraction				-.055	-.046	.286
	Brief COPE denial				-.047	-.042	.332
	Brief COPE venting				-.043	-.034	.429
	Brief COPE substance use				-.037	-.034	.431
	Brief COPE behavioural disengagement				-.039	-.031	.469
	Brief COPE self-blame				-.244	-.205	**<.001**
4		.472	.438	.006			.190
	Age				.156	.144	**.001**
	Employment status				.124	.116	**.008**
	Brief IPQ consequences				-.071	-.042	.327
	Brief IPQ controllability				.011	.010	.825
	Brief IPQ curability				.083	.077	.074
	Brief IPQ identity				-.051	-.034	.426
	Brief IPQ concern				-.012	-.009	.843
	Brief IPQ emotional representation				-.226	-.138	**.002**
	Brief COPE positive reframing				.134	.116	**.008**
	Brief COPE acceptance				.077	.067	.121
	Brief COPE self-distraction				-.064	-.054	.213
	Brief COPE denial				-.046	-.041	.348
	Brief COPE venting				-.053	-.042	.334
	Brief COPE substance use				-.022	-.020	.641
	Brief COPE behavioural disengagement				-.041	-.033	.443
	Brief COPE self-blame				-.237	-.198	**<.001**

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	MSPSS friends support				.081	.073	.093
	MSPSS significant other support				.012	.011	.794
5		.605	.577	.134			**<.001**
	Age				.164	.151	**<.001**
	Employment status				.074	.068	.070
	Brief IPQ consequences				-.040	-.024	.524
	Brief IPQ controllability				-.008	-.007	.858
	Brief IPQ curability				.034	.031	.407
	Brief IPQ identity				-.036	-.024	.521
	Brief IPQ concern				-.036	-.026	.493
	Brief IPQ emotional representation				-.065	-.038	.311
	Brief COPE positive reframing				.070	.060	.111
	Brief COPE acceptance				.044	.038	.309
	Brief COPE self-distraction				-.059	-.050	.186
	Brief COPE denial				-.027	-.024	.527
	Brief COPE venting				-.019	-.015	.693
	Brief COPE substance use				.000	.000	.998
	Brief COPE behavioural disengagement				.029	.023	.545
	Brief COPE self-blame				-.172	-.142	**<.001**
	MSPSS friends support				.056	.050	.179
	MSPSS significant other support				-.006	-.005	.888
	STAI-6 anxiety				-.131	-.099	**.008**
	CES-D 10 depression				-.421	-.319	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Vitality subscale of the SF-36 (n= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.095	.089	.095			**<.001**
	Employment status				.261	.257	**<.001**
	Gender				-.123	-.121	.029
2		.177	.160	.082			**<.001**
	Employment status				.215	.207	**<.001**
	Gender				-.113	-.109	.040
	ToF				.223	.185	**.001**
	TGA				.197	.161	**.002**
	SV				.047	.039	.458
	Co-morbidities no.				-.195	-.191	**<.001**
3		.201	.179	.024			.013
	Employment status				.201	.193	**<.001**
	Gender				-.112	-.108	.039
	ToF				.225	.184	**<.001**
	TGA				.228	.183	**.001**
	SV				.165	.113	.031
	Co-morbidities no.				-.098	-.078	.135
	Current O_2 saturation				.165	.130	.013
	Medication no.				-.119	-.096	.067
4		.491	.466	.290			**<.001**
	Employment status				.104	.097	.022
	Gender				-.049	-.046	.271
	ToF				.228	.179	**<.001**
	TGA				.208	.166	**<.001**
	SV				.232	.155	**<.001**
	Co-morbidities no.				-.009	-.007	.862
	Current O_2 saturation				.078	.061	.151
	Medication no.				.011	.009	.839
	Brief IPQ consequences				-.117	-.071	.091
	Brief IPQ controllability				.068	.064	.130
	Brief IPQ curability				.045	.041	.326
	Brief IPQ identity				-.361	-.237	**<.001**
	Brief IPQ concern				.014	.010	.813
	Brief IPQ emotional representation				-.210	-.140	**.001**
5		.544	.510	.053			**<.001**
	Employment status				.107	.097	.017
	Gender				-.040	-.036	.371
	ToF				.195	.151	**<.001**
	TGA				.194	.152	**<.001**
	SV				.195	.129	**.001**
	Co-morbidities no.				-.005	-.004	.920
	Current O_2 saturation				.064	.049	.228
	Medication no.				.018	.014	.737
	Brief IPQ consequences				-.108	-.065	.108
	Brief IPQ controllability				.067	.062	.125
	Brief IPQ curability				.042	.038	.347
	Brief IPQ identity				-.381	-.248	**<.001**
	Brief IPQ concern				.014	.010	.807
	Brief IPQ emotional representation				-.111	-.066	.101
	Brief COPE acceptance				.129	.119	**.003**
	Brief COPE self-distraction				-.047	-.039	.332

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief COPE denial				-.058	-.052	.195
	Brief COPE venting				.000	.000	1.000
	Brief COPE substance use				-.014	-.013	.749
	Brief COPE behavioural disengagement				-.019	-.015	.711
	Brief COPE self-blame				-.150	-.126	**.002**
6		.609	.577	.065			**<.001**
	Employment status				.079	.071	.059
	Gender				-.036	-.032	.391
	ToF				.175	.135	**<.001**
	TGA				.147	.114	**.003**
	SV				.157	.102	**.007**
	Co-morbidities no.				-.003	-.002	.949
	Current O_2 saturation				.030	.023	.542
	Medication no.				.022	.017	.659
	Brief IPQ consequences				-.078	-.047	.211
	Brief IPQ controllability				.046	.042	.265
	Brief IPQ curability				.012	.011	.773
	Brief IPQ identity				-.376	-.245	**<.001**
	Brief IPQ concern				-.009	-.006	.864
	Brief IPQ emotional representation				.008	.005	.898
	Brief COPE acceptance				.091	.082	.029
	Brief COPE self-distraction				-.062	-.051	.172
	Brief COPE denial				-.047	-.042	.267
	Brief COPE venting				.018	.014	.700
	Brief COPE substance use				.005	.004	.913
	Brief COPE behavioural disengagement				.026	.021	.583
	Brief COPE self-blame				-.099	-.082	.028
	STAI-6 anxiety				-.076	-.057	.128
	CES-D 10 depression				-.302	-.226	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Social functioning subscale of the SF-36 (*n*= 303)

Block	*Predictor Variables*	*R^2*	*Adjusted R^2*	*ΔR^2*	β	*sr^2*	*Sig.*
1		.065	.062	.065			**<.001**
	Employment status				.256	.256	**<.001**
2		.138	.124	.073			**<.001**
	Employment status				.213	.209	**<.001**
	ToF				.121	.100	.064
	TGA				.065	.054	.318
	SV				-.132	-.111	.040
	Co-morbidities no.				-.159	-.156	**.004**
3		.142	.124	.004			.265
	Employment status				.207	.202	**<.001**
	ToF				.137	.111	.040
	TGA				.074	.060	.262
	SV				-.107	-.084	.118
	Co-morbidities no.				-.133	-.121	.026
	Hospitalization days				-.070	-.060	.265
4		.162	.139	.020			.032
	Employment status				.192	.187	**.001**
	ToF				.143	.115	.033
	TGA				.104	.084	.118
	SV				.004	.003	.962
	Co-morbidities no.				-.064	-.049	.362
	Hospitalization days				-.065	-.056	.297
	Current O_2 saturation				.169	.133	.013
	Medication no.				-.071	-.057	.286
5		.430	.405	.269			**<.001**
	Employment status				.076	.072	.108
	ToF				.161	.128	**.004**
	TGA				.078	.063	.158
	SV				.057	.037	.400
	Co-morbidities no.				.015	.011	.805
	Hospitalization days				-.055	-.046	.298
	Current O_2 saturation				.086	.067	.134
	Medication no.				.073	.055	.213
	Brief IPQ consequences				-.261	-.159	**<.001**
	Brief IPQ controllability				.029	.028	.535
	Brief IPQ identity				-.235	-.155	**.001**
	Brief IPQ concern				-.027	-.019	.675
	Brief IPQ emotional representation				-.159	-.106	.017
6		.439	.401	.009			.626
	Employment status				.076	.070	.117
	ToF				.149	.117	**.009**
	TGA				.069	.055	.222
	SV				.049	.032	.472
	Co-morbidities no.				.016	.012	.794
	Hospitalization days				-.060	-.050	.262
	Current O_2 saturation				.082	.063	.161
	Medication no.				.076	.057	.200
	Brief IPQ consequences				-.254	-.153	**.001**
	Brief IPQ controllability				.026	.024	.585
	Brief IPQ identity				-.236	-.156	**.001**
	Brief IPQ concern				-.029	-.020	.654
	Brief IPQ emotional representation				-.126	-.077	.086
	Brief COPE self-distraction				.001	.001	.978

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief COPE denial				-.049	-.045	.316
	Brief COPE venting				.013	.011	.810
	Brief COPE substance use				-.038	-.035	.428
	Brief COPE behavioural disengagement				-.038	-.030	.499
	Brief COPE self-blame				-.038	-.032	.470
7		.442	.403	.004			.184
	Employment status				.077	.071	.111
	ToF				.151	.119	**.008**
	TGA				.072	.057	.204
	SV				.054	.036	.422
	Co-morbidities no.				.018	.013	.765
	Hospitalization days				-.050	-.041	.352
	Current O_2 saturation				.083	.064	.154
	Medication no.				.070	.053	.236
	Brief IPQ consequences				-.242	-.145	**.001**
	Brief IPQ controllability				.016	.015	.732
	Brief IPQ identity				-.246	-.161	**<.001**
	Brief IPQ concern				-.029	-.020	.651
	Brief IPQ emotional representation				-.121	-.073	.100
	Brief COPE self-distraction				-.007	-.006	.894
	Brief COPE denial				-.047	-.043	.338
	Brief COPE venting				.004	.004	.937
	Brief COPE substance use				-.029	-.026	.555
	Brief COPE behavioural disengagement				-.037	-.030	.507
	Brief COPE self-blame				-.032	-.027	.542
	MSPSS friends support				.064	.059	.184
8		.499	.459	.056			**<.001**
	Employment status				.055	.051	.231
	ToF				.116	.091	.033
	TGA				.022	.017	.685
	SV				.002	.001	.977
	Co-morbidities no.				.015	.011	.795
	Hospitalization days				-.030	-.025	.555
	Current O_2 saturation				.048	.037	.389
	Medication no.				.070	.052	.217
	Brief IPQ consequences				-.224	-.134	**.002**
	Brief IPQ controllability				-.012	-.011	.800
	Brief IPQ identity				-.242	-.159	**<.001**
	Brief IPQ concern				-.049	-.034	.422
	Brief IPQ emotional representation				.006	.003	.939
	Brief COPE self-distraction				-.030	-.026	.542
	Brief COPE denial				-.030	-.027	.525
	Brief COPE venting				.017	.013	.755
	Brief COPE substance use				-.014	-.013	.762
	Brief COPE behavioural disengagement				.012	.009	.825
	Brief COPE self-blame				.019	.016	.708
	MSPSS friends support				.034	.031	.463
	STAI-6 anxiety				-.119	-.093	.029
	CES-D 10 depression				-.249	-.187	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Role emotional subscale of the SF-36 (*n*= 302)

Block	*Predictor Variables*	*R^2*	*Adjusted R^2*	*ΔR^2*	β	*sr^2*	*Sig.*
1		.063	.060	.063			**<.001**
	Employment status				.251	.251	**<.001**
2		.218	.202	.155			**<.001**
	Employment status				.135	.129	.013
	Brief IPQ consequences				-.167	-.103	.047
	Brief IPQ controllability				.074	.072	.166
	Brief IPQ identity				-.003	-.002	.973
	Brief IPQ concern				.054	.039	.448
	Brief IPQ emotional representation				-.292	-.199	**<.001**
3		.301	.270	.084			**<.001**
	Employment status				.131	.124	.013
	Brief IPQ consequences				-.135	-.082	.096
	Brief IPQ controllability				.040	.037	.448
	Brief IPQ identity				-.021	-.014	.770
	Brief IPQ concern				.040	.029	.561
	Brief IPQ emotional representation				-.164	-.100	.043
	Brief COPE positive reframing				.099	.088	.076
	Brief COPE acceptance				.069	.060	.221
	Brief COPE self-distraction				-.048	-.042	.394
	Brief COPE venting				-.036	-.028	.565
	Brief COPE substance use				-.050	-.047	.338
	Brief COPE behavioural disengagement				-.045	-.038	.440
	Brief COPE self-blame				-.214	-.183	**<.001**
4		.312	.278	.010			.041
	Employment status				.132	.124	.012
	Brief IPQ consequences				-.111	-.067	.174
	Brief IPQ controllability				.027	.025	.603
	Brief IPQ identity				-.034	-.023	.641
	Brief IPQ concern				.036	.026	.599
	Brief IPQ emotional representation				-.155	-.095	.053
	Brief COPE positive reframing				.089	.078	.112
	Brief COPE acceptance				.061	.054	.271
	Brief COPE self-distraction				-.059	-.051	.299
	Brief COPE venting				-.048	-.038	.437
	Brief COPE substance use				-.033	-.031	.529
	Brief COPE behavioural disengagement				-.046	-.039	.429
	Brief COPE self-blame				-.207	-.177	**<.001**
	MSPSS friends support				.108	.101	.041
5		.420	.387	.108			**<.001**
	Employment status				.087	.081	.075
	Brief IPQ consequences				-.085	-.051	.259
	Brief IPQ controllability				.004	.003	.939
	Brief IPQ identity				-.014	-.010	.832
	Brief IPQ concern				.007	.005	.909
	Brief IPQ emotional representation				-.011	-.006	.888
	Brief COPE positive reframing				.033	.029	.527

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief COPE acceptance				.030	.027	.556
	Brief COPE self-distraction				-.052	-.045	.317
	Brief COPE venting				-.021	-.017	.710
	Brief COPE substance use				-.013	-.012	.786
	Brief COPE behavioural disengagement				.025	.020	.654
	Brief COPE self-blame				-.146	-.123	**.007**
	MSPSS friends support				.080	.075	.100
	STAI-6 anxiety				-.098	-.076	.093
	CES-D 10 depression				-.384	-.291	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Mental health subscale of the SF-36 (*n*= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.096	.090	.096			**<.001**
	Age				.168	.168	**.002**
	Employment status				.264	.264	**<.001**
2		.351	.336	.255			**<.001**
	Age				.152	.148	**.002**
	Employment status				.125	.119	.012
	Brief IPQ consequences				-.082	-.051	.280
	Brief IPQ controllability				.102	.098	.037
	Brief IPQ identity				-.041	-.028	.557
	Brief IPQ concern				-.030	-.022	.642
	Brief IPQ emotional representation				-.386	-.262	**<.001**
3		.463	.435	.112			**<.001**
	Age				.178	.167	**<.001**
	Employment status				.125	.118	**.007**
	Brief IPQ consequences				-.064	-.039	.367
	Brief IPQ controllability				.061	.057	.186
	Brief IPQ identity				-.057	-.038	.376
	Brief IPQ concern				-.046	-.033	.449
	Brief IPQ emotional representation				-.239	-.146	**.001**
	Brief COPE positive reframing				.116	.102	.019
	Brief COPE acceptance				.064	.056	.198
	Brief COPE self-distraction				.006	.005	.908
	Brief COPE denial				-.097	-.086	.047
	Brief COPE venting				-.034	-.027	.528
	Brief COPE substance use				-.012	-.011	.800
	Brief COPE behavioural disengagement				-.074	-.060	.169
	Brief COPE self-blame				-.224	-.188	**<.001**
4		.467	.433	.004			.567
	Age				.177	.163	**<.001**
	Employment status				.128	.120	**.006**
	Brief IPQ consequences				-.049	-.029	.501
	Brief IPQ controllability				.051	.047	.279
	Brief IPQ identity				-.063	-.042	.332
	Brief IPQ concern				-.048	-.035	.426
	Brief IPQ emotional representation				-.237	-.144	**.001**
	Brief COPE positive reframing				.111	.097	.026
	Brief COPE acceptance				.059	.051	.242
	Brief COPE self-distraction				-.002	-.001	.973
	Brief COPE denial				-.095	-.084	.052
	Brief COPE venting				-.042	-.033	.447
	Brief COPE substance use				.001	.001	.980
	Brief COPE behavioural disengagement				-.074	-.060	.169
	Brief COPE self-blame				-.219	-.183	**<.001**
	MSPSS friends support				.053	.045	.295
	MSPSS family support				.028	.023	.603
	MSPSS significant other support				-.009	-.008	.855

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
5		.587	.558	.120			**<.001**
	Age				.186	.172	**<.001**
	Employment status				.082	.076	.048
	Brief IPQ consequences				-.019	-.011	.768
	Brief IPQ controllability				.020	.018	.629
	Brief IPQ identity				-.050	-.033	.384
	Brief IPQ concern				-.076	-.054	.157
	Brief IPQ emotional representation				-.070	-.041	.288
	Brief COPE positive reframing				.047	.040	.295
	Brief COPE acceptance				.024	.021	.592
	Brief COPE self-distraction				.001	.001	.984
	Brief COPE denial				-.075	-.067	.081
	Brief COPE venting				-.015	-.012	.763
	Brief COPE substance use				.021	.019	.626
	Brief COPE behavioural disengagement				-.004	-.004	.926
	Brief COPE self-blame				-.156	-.129	**.001**
	MSPSS friends support				.039	.033	.385
	MSPSS family support				-.018	-.014	.709
	MSPSS significant other support				-.019	-.016	.667
	STAI-6 anxiety				-.192	-.148	**<.001**
	CES-D 10 depression				-.356	-.270	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Symptoms scale of the CHD-TAAQOL (*n*= 302)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.094	.088	.094			**<.001**
	Gender				-.104	-.102	.064
	Employment status				.270	.266	**<.001**
2		.207	.185	.113			**<.001**
	Gender				-.132	-.128	.015
	Employment status				.212	.204	**<.001**
	ToF				.020	.016	.761
	TGA				-.008	-.007	.900
	SV				-.147	-.106	.043
	Cyanosis days				-.078	-.067	.199
	Co-morbidities no.				-.272	-.221	**<.001**
	Arrhythmias				.036	.029	.581
3		.207	.182	.000			.996
	Gender				-.132	-.127	.015
	Employment status				.212	.203	**<.001**
	ToF				.020	.016	.766
	TGA				-.008	-.006	.901
	SV				-.147	-.101	.053
	Cyanosis days				-.078	-.067	.200
	Co-morbidities no.				-.272	-.208	**<.001**
	Arrhythmias				.036	.029	.582
	Hospitalization days				.000	.000	.996
4		.278	.246	.071			**<.001**
	Gender				-.102	-.092	.066
	Employment status				.176	.166	**.001**
	ToF				.025	.019	.711
	TGA				.053	.040	.425
	SV				-.006	-.004	.943
	Cyanosis days				.069	.050	.317
	Co-morbidities no.				-.124	-.085	.091
	Arrhythmias				.044	.033	.506
	Hospitalization days				.021	.018	.719
	Current O_2 saturation				.234	.154	**.002**
	Medication no.				-.162	-.124	.014
	VO_2 max				.145	.125	.013
	LVEF				.086	.076	.131
5		.581	.555	.303			**<.001**
	Gender				-.061	-.054	.160
	Employment status				.093	.085	.028
	ToF				.027	.020	.599
	TGA				.015	.011	.773
	SV				.026	.016	.678
	Cyanosis days				.032	.023	.545
	Co-morbidities no.				-.062	-.042	.279
	Arrhythmias				.058	.044	.251
	Hospitalization days				.030	.025	.513
	Current O_2 saturation				.118	.077	.046
	Medication no.				-.012	-.009	.819
	VO_2 max				.085	.072	.063
	LVEF				.029	.025	.509
	Brief IPQ consequences				-.088	-.053	.166
	Brief IPQ controllability				.014	.014	.725
	Brief IPQ identity				-.428	-.278	**<.001**
	Brief IPQ concern				-.088	-.061	.115

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief IPQ emotional representation				-.134	-.088	.023
6		.593	.558	.011			.257
	Gender				-.062	-.053	.169
	Employment status				.092	.083	.032
	ToF				.014	.010	.794
	TGA				.008	.006	.876
	SV				.017	.010	.788
	Cyanosis days				.032	.023	.547
	Co-morbidities no.				-.058	-.039	.309
	Arrhythmias				.058	.044	.251
	Hospitalization days				.022	.018	.636
	Current O_2 saturation				.118	.076	.048
	Medication no.				-.008	-.006	.878
	VO_2 max				.078	.065	.090
	LVEF				.026	.023	.557
	Brief IPQ consequences				-.081	-.049	.207
	Brief IPQ controllability				.010	.009	.815
	Brief IPQ identity				-.431	-.280	**<.001**
	Brief IPQ concern				-.093	-.064	.097
	Brief IPQ emotional representation				-.104	-.063	.104
	Brief COPE self-distraction				.009	.008	.833
	Brief COPE denial				-.054	-.049	.205
	Brief COPE venting				.031	.025	.523
	Brief COPE substance use				-.028	-.025	.516
	Brief COPE behavioural disengagement				-.028	-.022	.564
	Brief COPE self-blame				-.075	-.063	.103
7		.634	.599	.041			**<.001**
	Gender				-.051	-.043	.239
	Employment status				.075	.067	.069
	ToF				-.010	-.007	.847
	TGA				-.022	-.016	.669
	SV				-.012	-.007	.841
	Cyanosis days				.042	.030	.414
	Co-morbidities no.				-.047	-.032	.385
	Arrhythmias				.031	.023	.528
	Hospitalization days				.042	.035	.339
	Current O_2 saturation				.096	.062	.092
	Medication no.				-.007	-.005	.896
	VO_2 max				.078	.066	.072
	LVEF				.038	.033	.370
	Brief IPQ consequences				-.060	-.035	.332
	Brief IPQ controllability				-.018	-.017	.640
	Brief IPQ identity				-.431	-.279	**<.001**
	Brief IPQ concern				-.107	-.074	.045
	Brief IPQ emotional representation				.001	.001	.987
	Brief COPE self-distraction				-.015	-.013	.724
	Brief COPE denial				-.039	-.035	.339
	Brief COPE venting				.038	.030	.409
	Brief COPE substance use				-.008	-.007	.854
	Brief COPE behavioural disengagement				.013	.010	.781

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief COPE self-blame				-.029	-.024	.518
	STAI-6 anxiety				-.097	-.075	.042
	CES-D 10 depression				-.215	-.160	**<.001**

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Impact of cardiac surveillance scale of the CHD-TAAQOL (n= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.126	.111	.126			**<.001**
	ToF				-.002	-.002	.973
	TGA				-.056	-.045	.412
	SV				-.171	-.140	.011
	Co-morbidities no.				-.164	-.137	.012
	Arrhythmias				-.163	-.130	.017
2		.129	.112	.003			.292
	ToF				.016	.013	.811
	TGA				-.036	-.027	.613
	SV				-.141	-.106	.052
	Co-morbidities no.				-.155	-.128	.019
	Arrhythmias				-.148	-.114	.036
	Intervention no.				-.069	-.057	.292
3		.154	.128	.024			.040
	ToF				.011	.009	.870
	TGA				-.017	-.013	.813
	SV				-.064	-.041	.450
	Co-morbidities no.				-.046	-.033	.540
	Arrhythmias				-.145	-.110	.041
	Intervention no.				-.043	-.035	.515
	Current O_2 saturation				.102	.079	.143
	Medication no.				-.139	-.108	.045
	VO_2 max				.066	.060	.266
4		.322	.292	.168			**<.001**
	ToF				.033	.026	.594
	TGA				-.045	-.034	.489
	SV				-.031	-.019	.694
	Co-morbidities no.				.013	.009	.850
	Arrhythmias				-.133	-.100	.039
	Intervention no.				-.064	-.052	.285
	Current O_2 saturation				.047	.035	.465
	Medication no.				-.039	-.029	.550
	VO_2 max				.020	.018	.716
	Brief IPQ consequences				-.040	-.024	.616
	Brief IPQ identity				-.136	-.089	.067
	Brief IPQ concern				-.129	-.090	.064
	Brief IPQ emotional representation				-.232	-.156	**.001**
5		.347	.309	.025			.027
	ToF				.019	.015	.759
	TGA				-.075	-.054	.258
	SV				-.042	-.026	.584
	Co-morbidities no.				.002	.002	.971
	Arrhythmias				-.138	-.103	.032
	Intervention no.				-.051	-.041	.390
	Current O_2 saturation				.038	.028	.552
	Medication no.				-.031	-.023	.638
	VO_2 max				.022	.020	.681

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	Brief IPQ consequences				-.030	-.018	.701
	Brief IPQ identity				-.131	-.086	.075
	Brief IPQ concern				-.134	-.093	.053
	Brief IPQ emotional representation				-.185	-.113	.019
	Brief COPE planning				-.119	-.114	.018
	Brief COPE venting				.015	.013	.792
	Brief COPE substance use				-.009	-.009	.855
	Brief COPE behavioural disengagement				-.117	-.103	.033
6		.365	.323	.018			.020
	ToF				.016	.012	.802
	TGA				-.079	-.057	.228
	SV				-.034	-.021	.661
	Co-morbidities no.				.014	.010	.834
	Arrhythmias				-.141	-.105	.027
	Intervention no.				-.070	-.056	.241
	Current O_2 saturation				.037	.028	.558
	Medication no.				-.029	-.021	.651
	VO_2 max				.011	.010	.832
	Brief IPQ consequences				-.011	-.006	.892
	Brief IPQ identity				-.142	-.093	.051
	Brief IPQ concern				-.135	-.093	.050
	Brief IPQ emotional representation				-.132	-.077	.106
	Brief COPE planning				-.138	-.130	**.006**
	Brief COPE venting				.019	.016	.739
	Brief COPE substance use				.003	.002	.959
	Brief COPE behavioural disengagement				-.088	-.074	.117
	STAI-6 anxiety				-.159	-.127	**.008**
	CES-D 10 depression				-.005	-.004	.939

Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

Worries scale of the CHD-TAAQOL (n= 303)

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
1		.023	.020	.023			**.008**
	Age				.153	.153	**.008**
2		.104	.089	.081			**<.001**
	Age				.221	.196	**<.001**
	ToF				.102	.084	.126
	TGA				.132	.106	.054
	SV				-.045	-.036	.517
	Co-morbidities no.				-.241	-.223	**<.001**
3		.116	.098	.012			.044
	Age				.229	.203	**<.001**
	ToF				.114	.094	.085
	TGA				.157	.124	.024
	SV				.042	.028	.609
	Co-morbidities no.				-.214	-.194	**<.001**
	Current O_2 saturation				.138	.110	.044
4		.437	.414	.321			**<.001**
	Age				.197	.170	**<.001**
	ToF				.096	.076	.084
	TGA				.104	.082	.065
	SV				.083	.054	.222
	Co-morbidities no.				-.031	-.027	.545
	Current O_2 saturation				.057	.044	.314
	Brief IPQ consequences				-.246	-.153	**.001**
	Brief IPQ controllability				-.002	-.002	.966
	Brief IPQ curability				.089	.082	.064
	Brief IPQ identity				-.116	-.076	.087
	Brief IPQ concern				-.122	-.087	.050
	Brief IPQ emotional representation				-.224	-.150	**.001**
5		.480	.447	.043			**.001**
	Age				.235	.198	**<.001**
	ToF				.077	.061	.157
	TGA				.089	.068	.112
	SV				.093	.060	.160
	Co-morbidities no.				-.025	-.022	.610
	Current O_2 saturation				.079	.061	.157
	Brief IPQ consequences				-.215	-.132	**.002**
	Brief IPQ controllability				-.001	-.001	.990
	Brief IPQ curability				.091	.083	.053
	Brief IPQ identity				-.135	-.088	.042
	Brief IPQ concern				-.130	-.092	.032
	Brief IPQ emotional representation				-.203	-.124	**.004**
	Brief COPE planning				-.090	-.080	.061
	Brief COPE self-distraction				-.004	-.003	.940
	Brief COPE venting				.085	.067	.117
	Brief COPE substance use				-.092	-.087	.044
	Brief COPE behavioural disengagement				.046	.038	.375
	Self-blame				-.180	-.151	**.001**
6		.487	.450	.006			.173
	Age				.233	.196	**<.001**
	ToF				.082	.064	.136
	TGA				.093	.071	.097

Block	*Predictor Variables*	R^2	*Adjusted* R^2	ΔR^2	β	sr^2	*Sig.*
	SV				.104	.067	.116
	Co-morbidities no.				-.023	-.020	.641
	Current O_2 saturation				.080	.062	.149
	Brief IPQ consequences				-.197	-.120	**.005**
	Brief IPQ controllability				-.013	-.012	.782
	Brief IPQ curability				.083	.075	.079
	Brief IPQ identity				-.145	-.094	.029
	Brief IPQ concern				-.130	-.092	.032
	Brief IPQ emotional representation				-.201	-.122	**.004**
	Brief COPE planning				-.097	-.086	.044
	Brief COPE self-distraction				-.014	-.012	.775
	Brief COPE venting				.075	.060	.164
	Brief COPE substance use				-.075	-.069	.108
	Brief COPE behavioural disengagement				.047	.039	.359
	Self-blame				-.171	-.142	**.001**
	MSPSS friend support				.061	.052	.221
	MSPSS family support				.041	.035	.410
7		.523	.486	.037			**<.001**
	Age				.218	.183	**<.001**
	ToF				.061	.048	.250
	TGA				.048	.036	.384
	SV				.060	.038	.361
	Co-morbidities no.				-.014	-.012	.765
	Current O_2 saturation				.049	.037	.365
	Brief IPQ consequences				-.179	-.109	**.009**
	Brief IPQ controllability				-.023	-.021	.606
	Brief IPQ curability				.067	.060	.150
	Brief IPQ identity				-.137	-.089	.033
	Brief IPQ concern				-.146	-.103	.013
	Brief IPQ emotional representation				-.106	-.061	.138
	Brief COPE planning				-.117	-.103	.013
	Brief COPE self-distraction				-.024	-.020	.626
	Brief COPE venting				.090	.071	.087
	Brief COPE substance use				-.066	-.060	.147
	Brief COPE behavioural disengagement				.084	.069	.094
	Self-blame				-.126	-.103	.013
	MSPSS friend support				.047	.040	.330
	MSPSS family support				.021	.018	.659
	STAI-6 anxiety				-.061	-.047	.261
	CES-D 10 depression				-.224	-.168	**<.001**

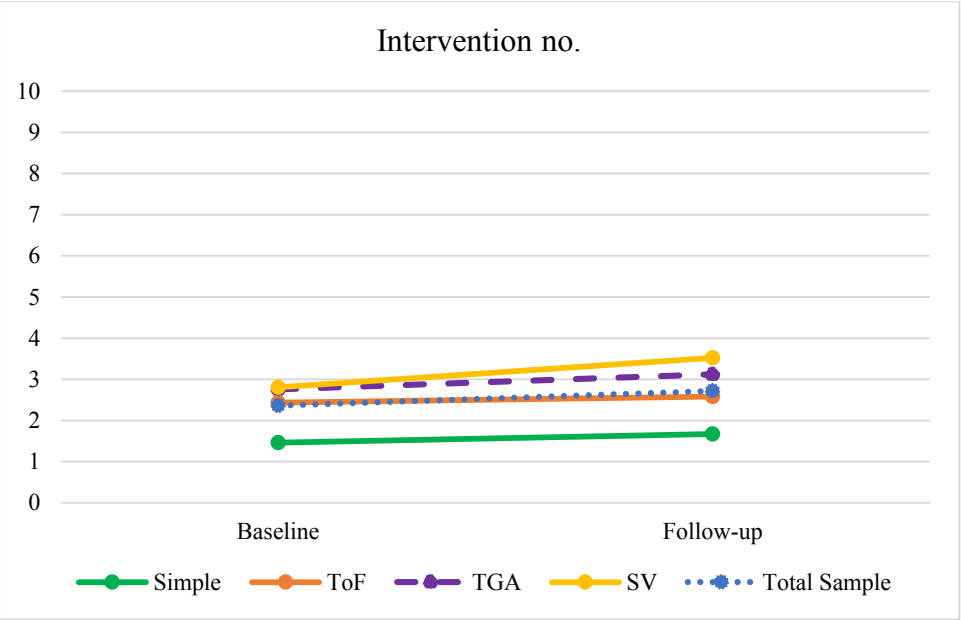
Note. Gender: 0= male, 1= female, educational level: 0= school, 1= university, employment status: 0= unemployed, 1= employed, ToF, TGA, SV: dummy-coded- reference group was Simple, arrhythmias: 0= no arrhythmias, 1= arrhythmias, CES-D 10: 0= no depressive symptoms, 1= with depressive symptoms.

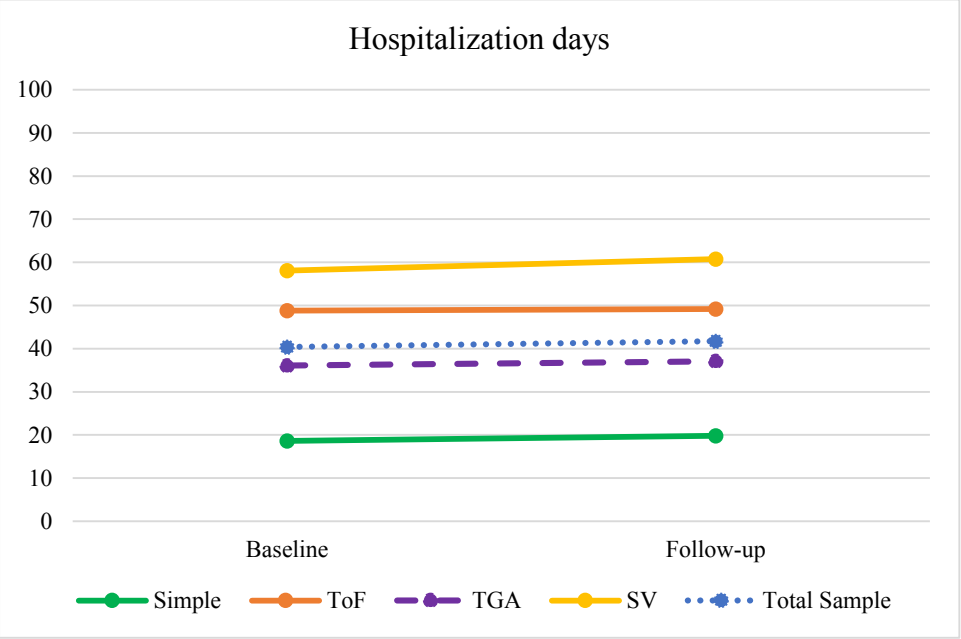
Appendix T. Multivariate outliers in regression analyses

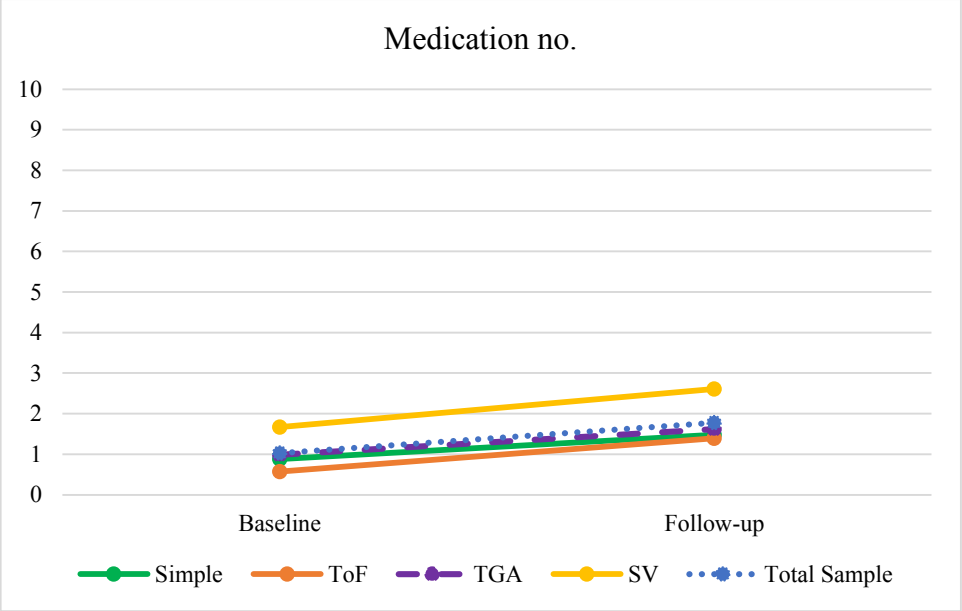
Regression Model (DV)	*Case ID*	*Standardized Residual*	*Cook's Distance*
Physical component summary	Case 272	-4.62	0.03
Physical functioning	Case 272	-4.90	0.04
	Case 179	-4.22	0.03
	Case 112	-4.10	0.12
Role physical	Case 24	-3.73	0.02
	Case 252	-3.70	0.03
	Case 272	-3.51	0.02
Bodily pain	n/a	n/a	n/a
General health	n/a	n/a	n/a
Mental component summary	n/a	n/a	n/a
Vitality	Case 92	-3.43	0.03
	Case 123	-3.51	0.06
Social functioning	Case 30	-3.71	0.07
	Case 179	-3.80	0.03
Role emotional	Case 29	-3.32	0.02
	Case 131	-3.41	0.06
Mental health	Case 136	3.68	0.09
	Case 305	-3.60	0.03
Symptoms	Case 135	-3.62	0.03
Worries	Case 10	-3.61	0.09
	Case 12	-3.48	0.11
Impact cardiac surveillance	n/a	n/a	n/a

Note. ±3.29 threshold

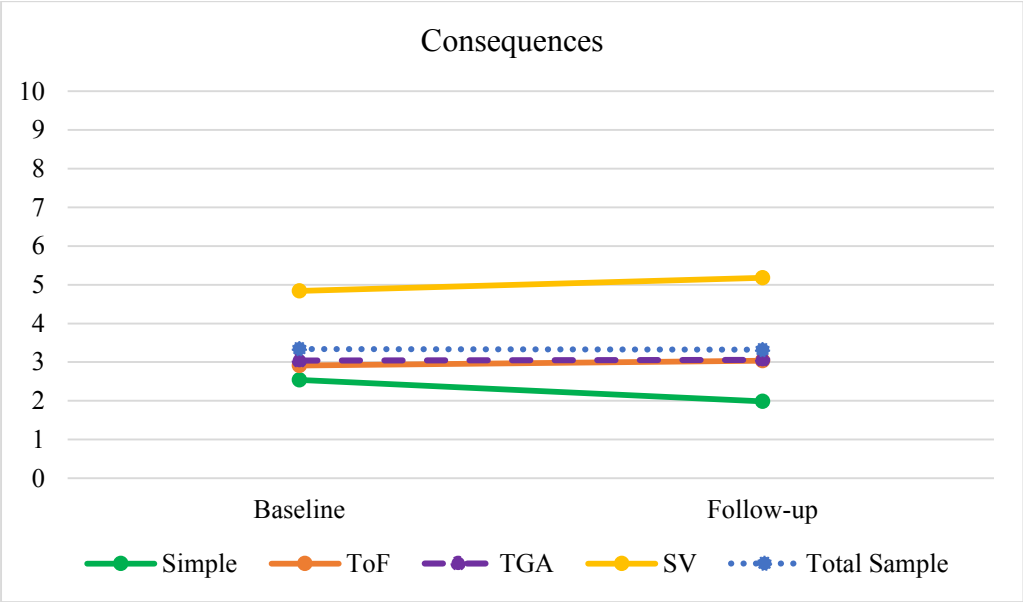
Appendix U. Graphical representation of clinical factors over time

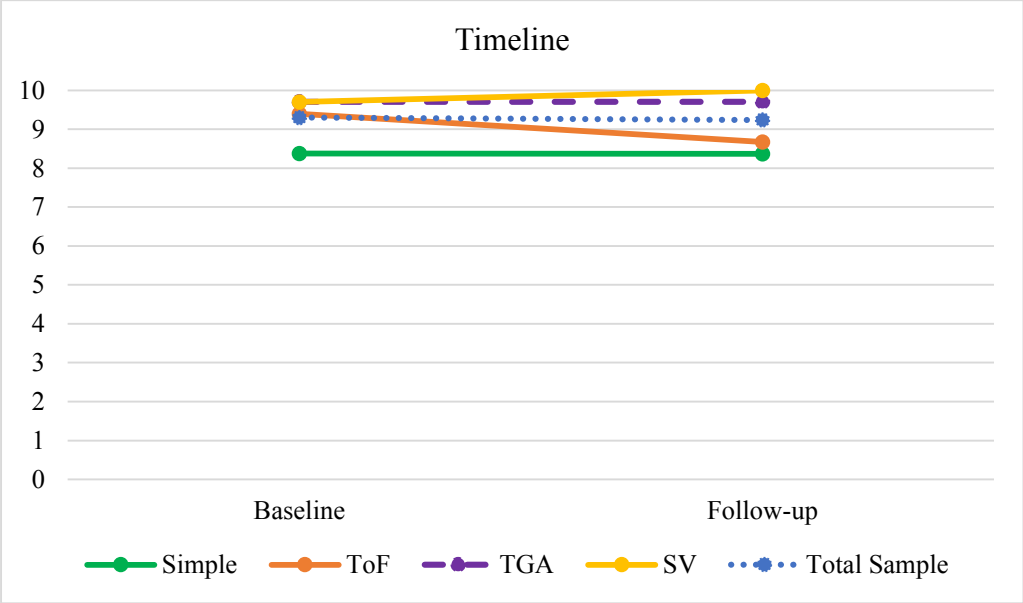


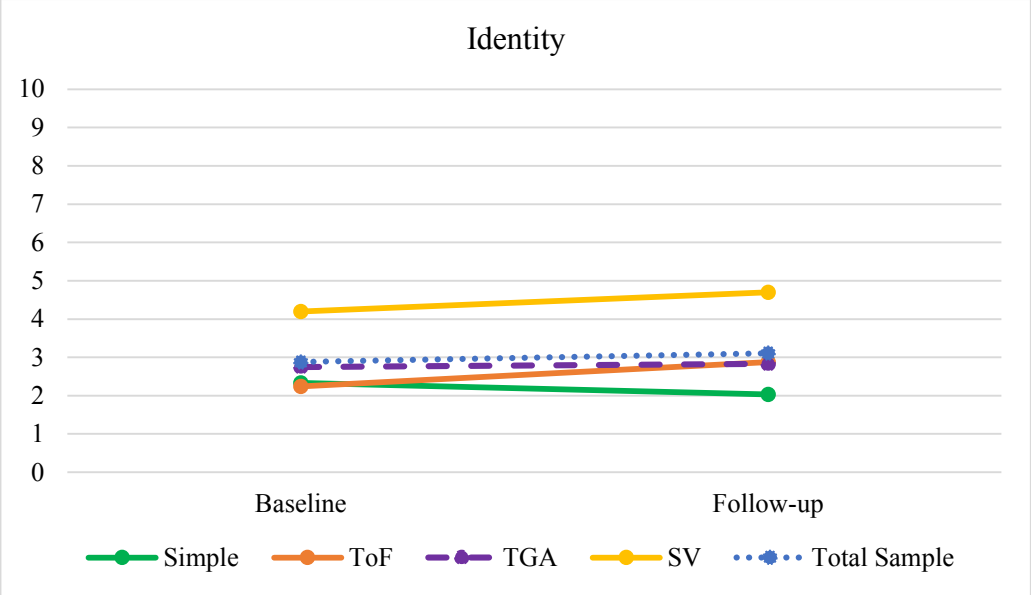


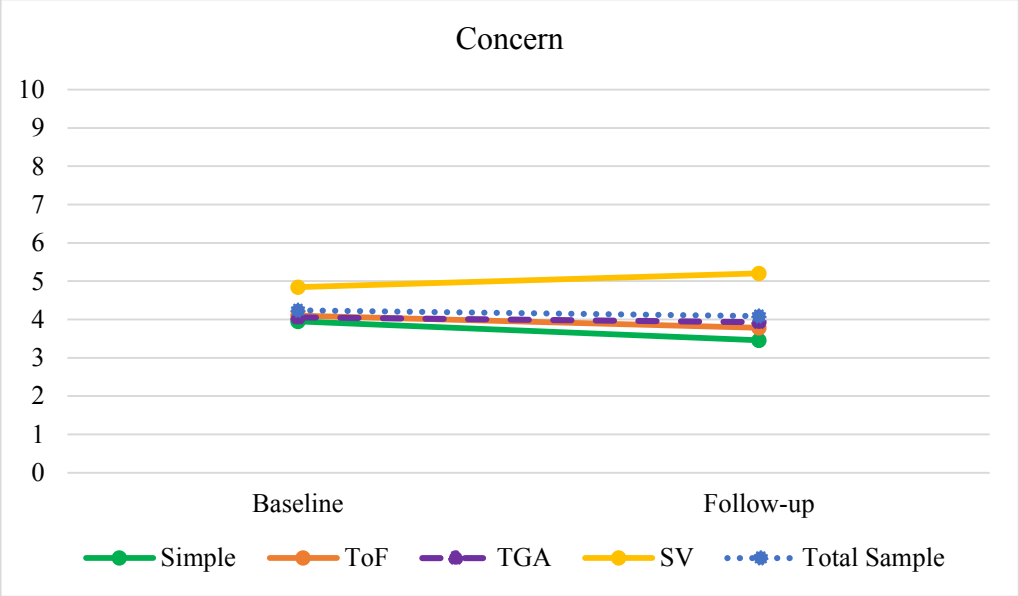

Medication no.
10
9
8
7
6
5
4
3
2
1
0
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample

Appendix V. Graphical representation of illness perceptions over time






Identity
10
9
8
7
6
5
4
3
2
1
0
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample


Concern
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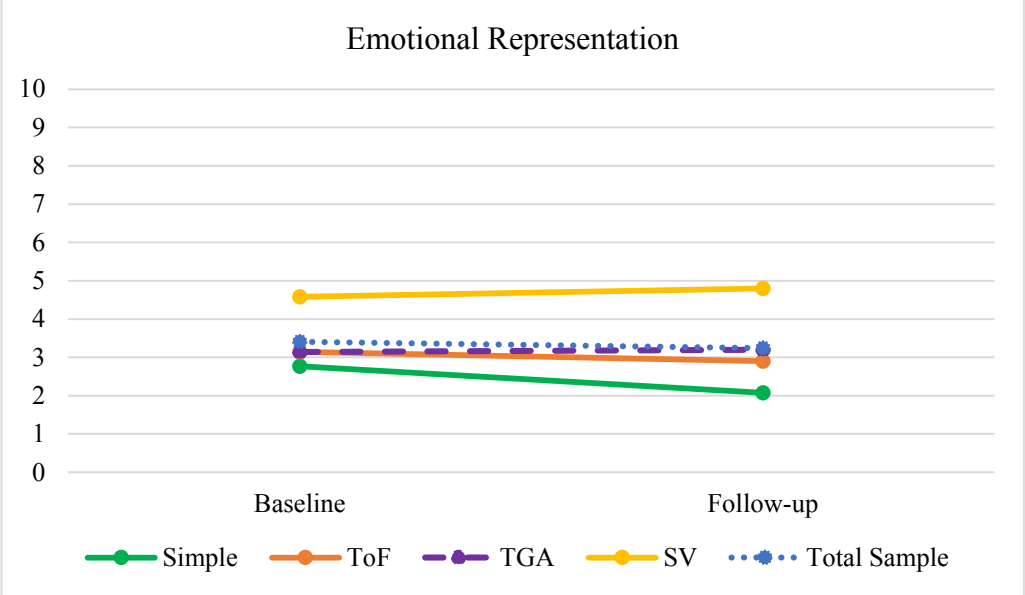
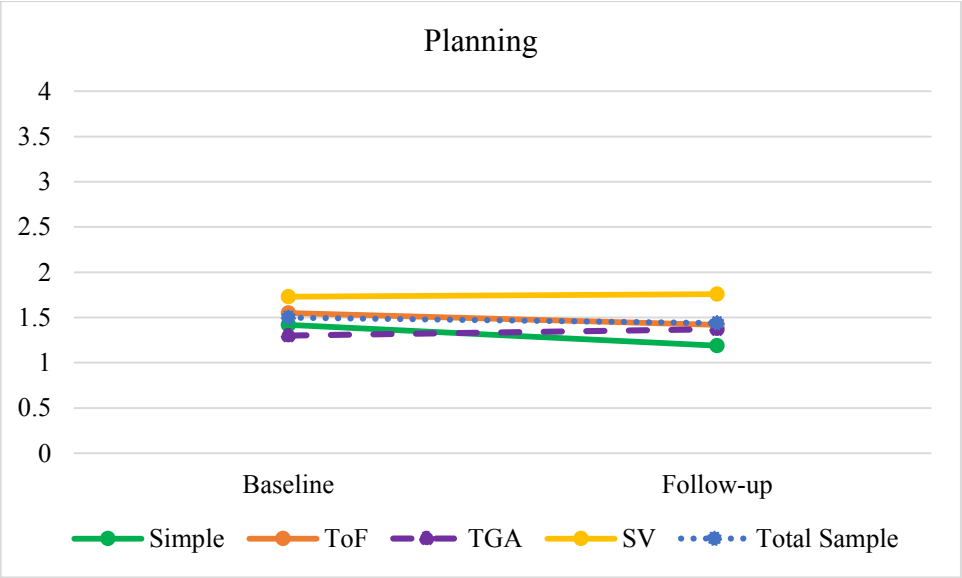
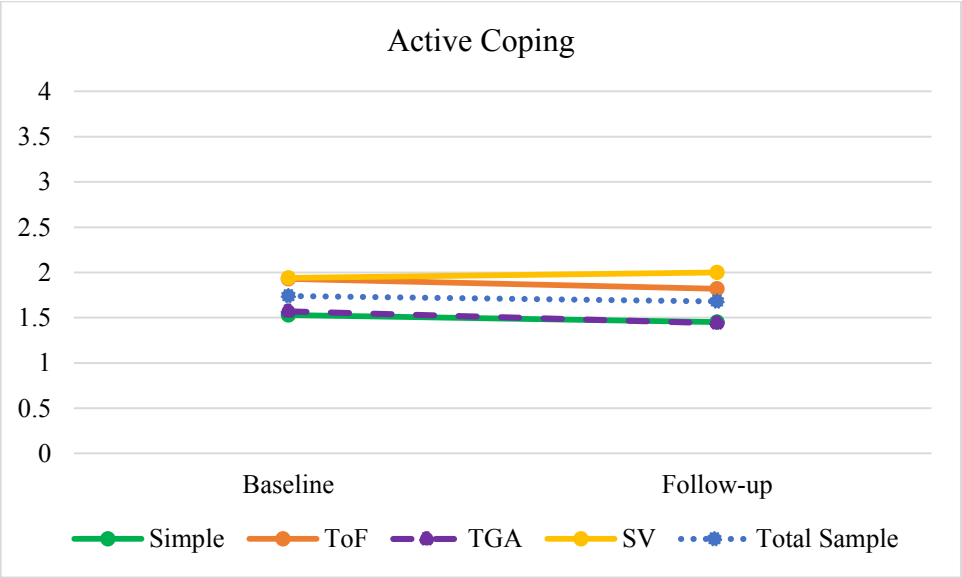
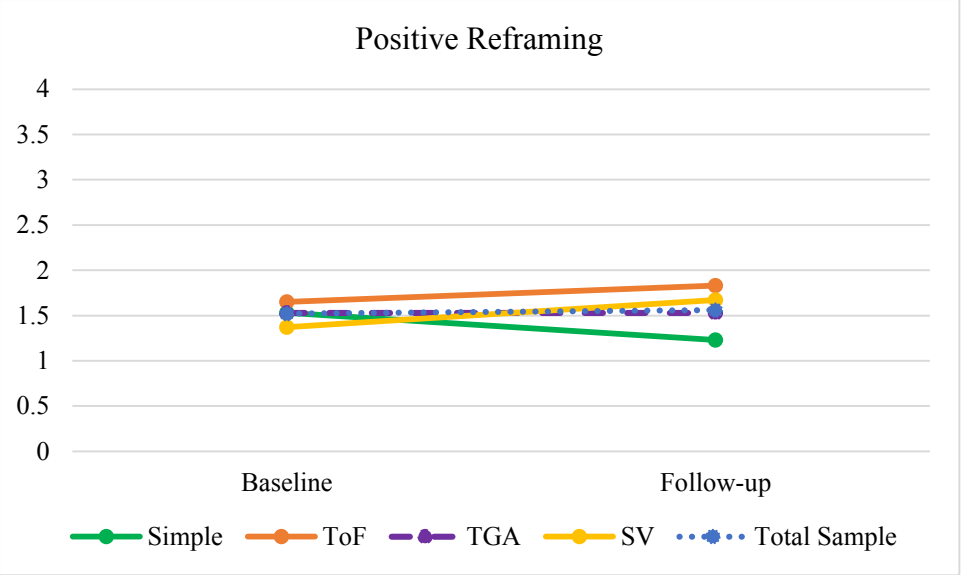
9
8
7
6
5
4
3
2
1
0
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample


Emotional Representation
10
9
8
7
6
5
4
3
2
1
0
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample

Appendix W. Graphical representation of coping strategies over time






Positive Reframing
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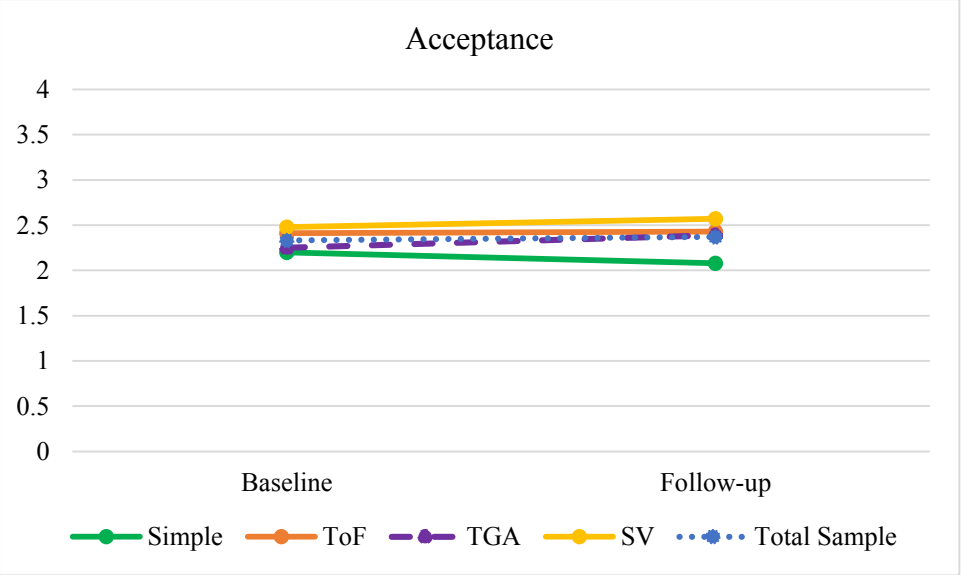
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample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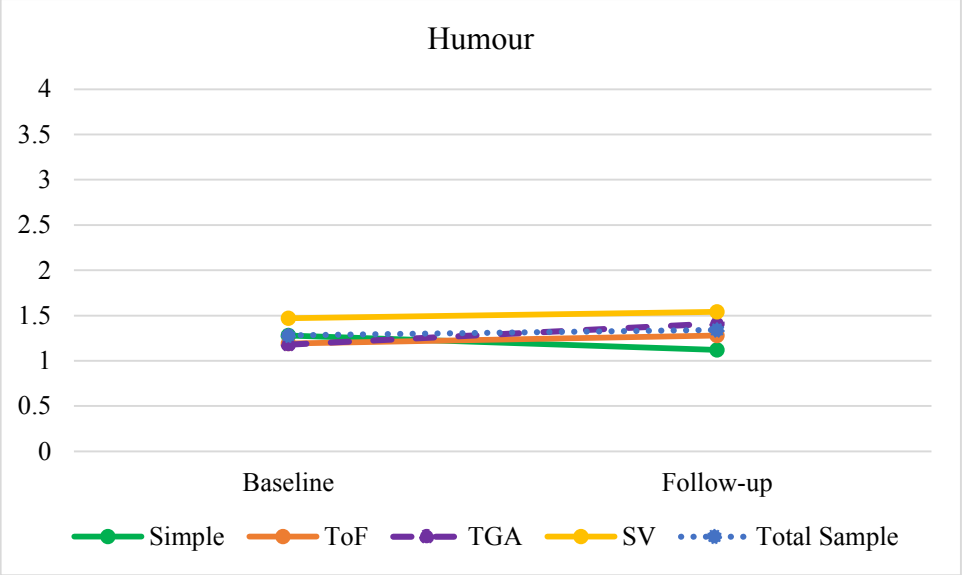
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Follow-up
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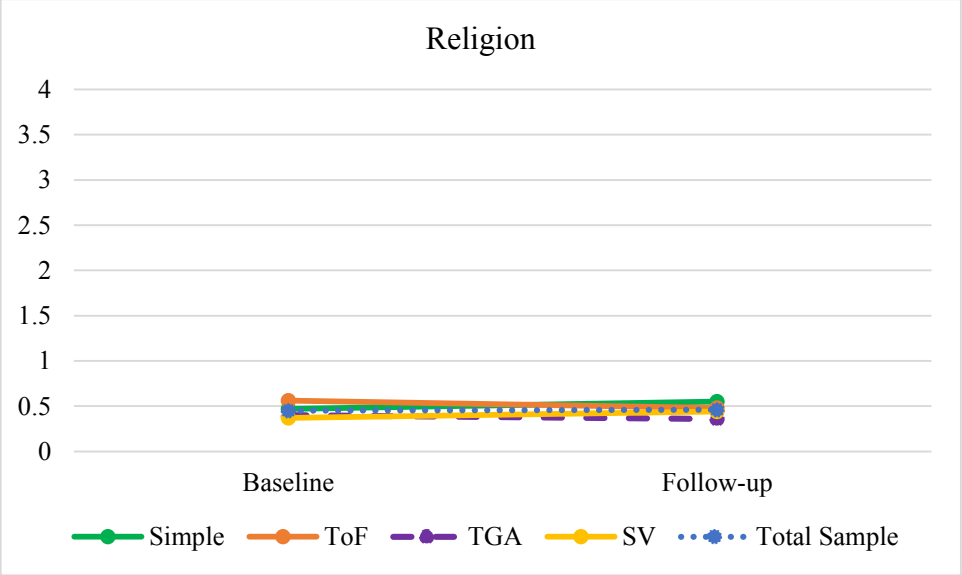
TGA
SV
Total Sample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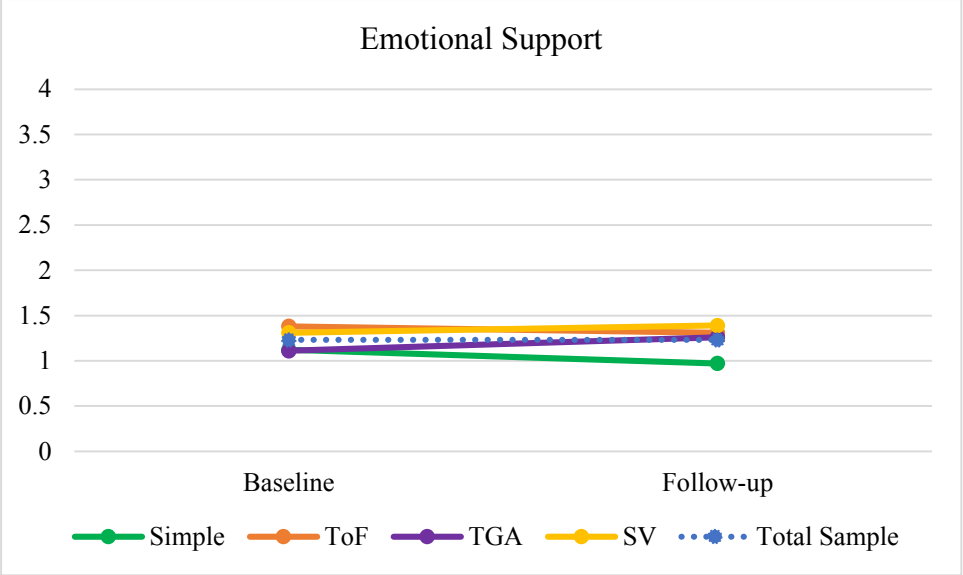
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Follow-up
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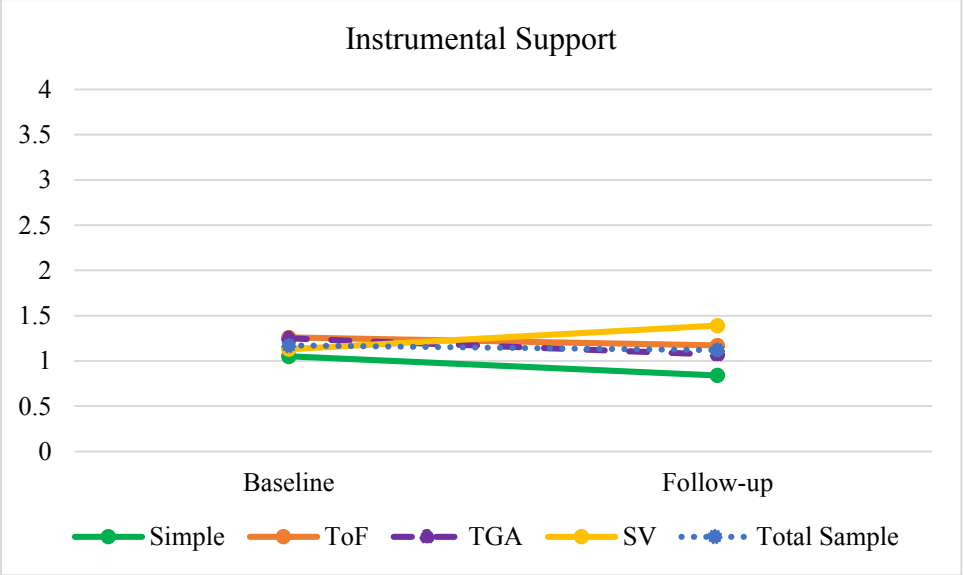
ToF
TGA
SV
Total Sample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Follow-up
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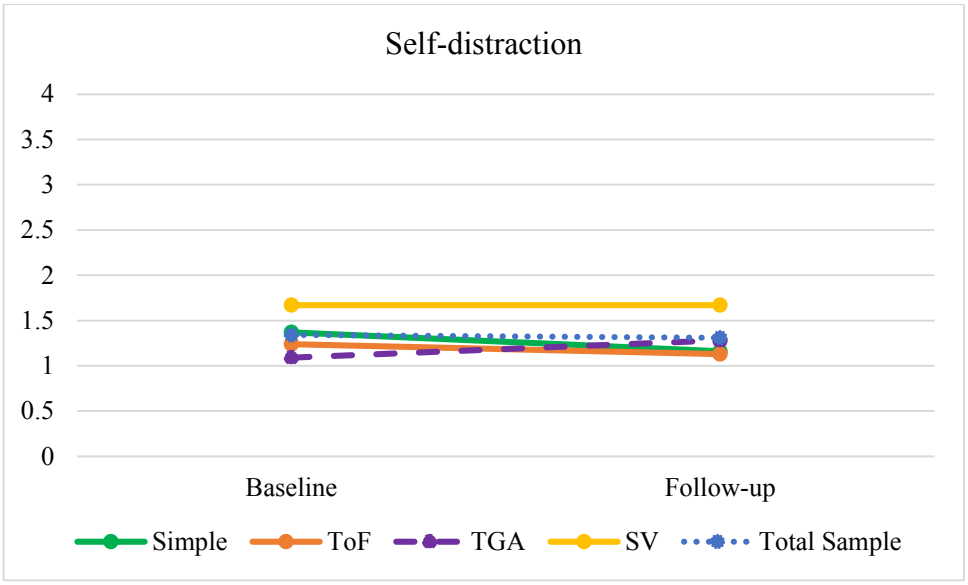
SV
Total Sample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Follow-up
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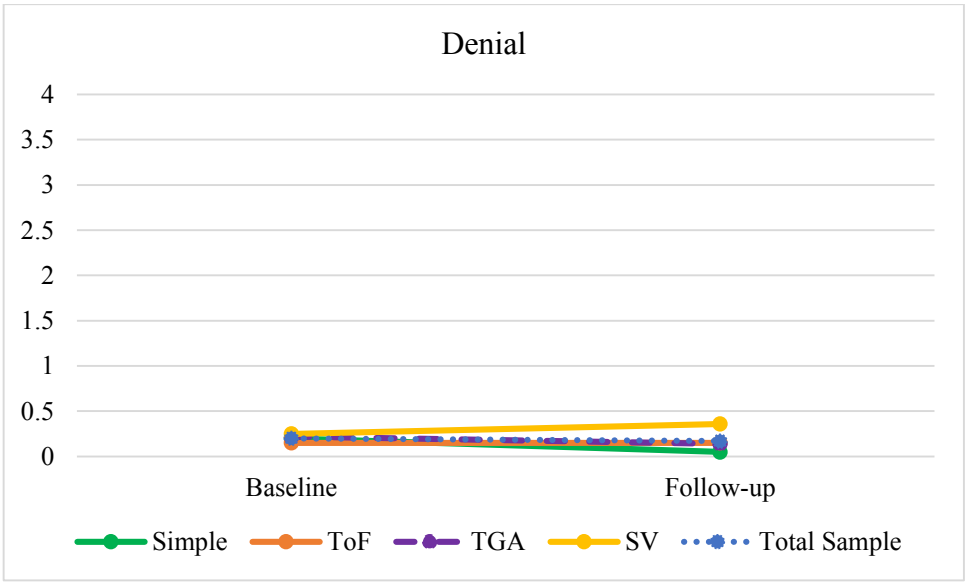
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Follow-up
Simple
ToF
TGA
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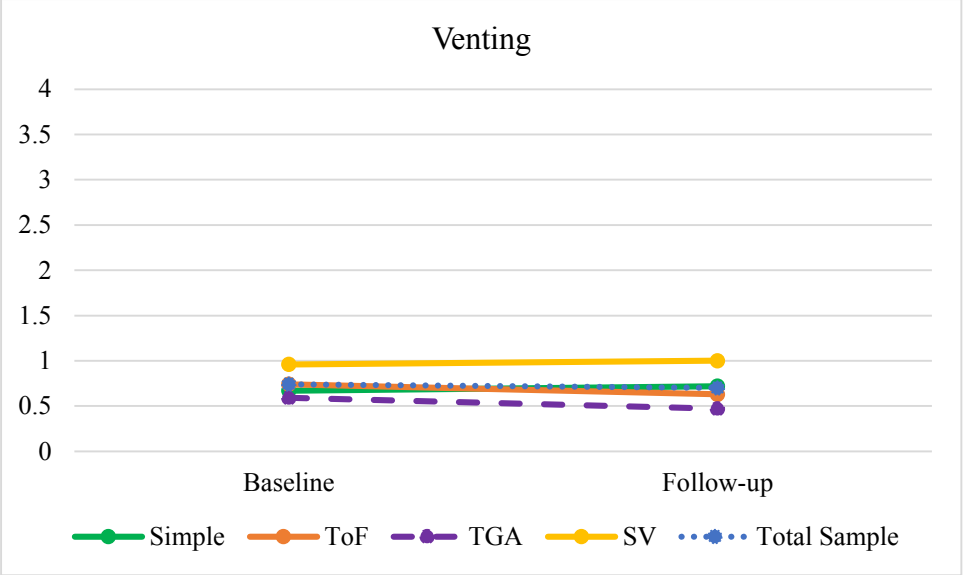
Total Sample


Venting
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Baseline
Follow-up
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ToF
TGA
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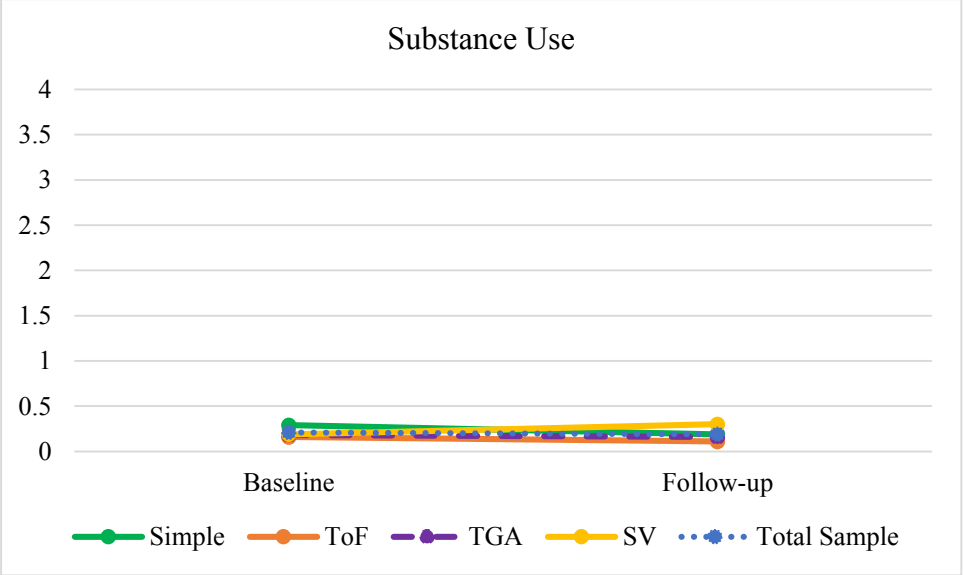
Total Sample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Follow-up
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TGA
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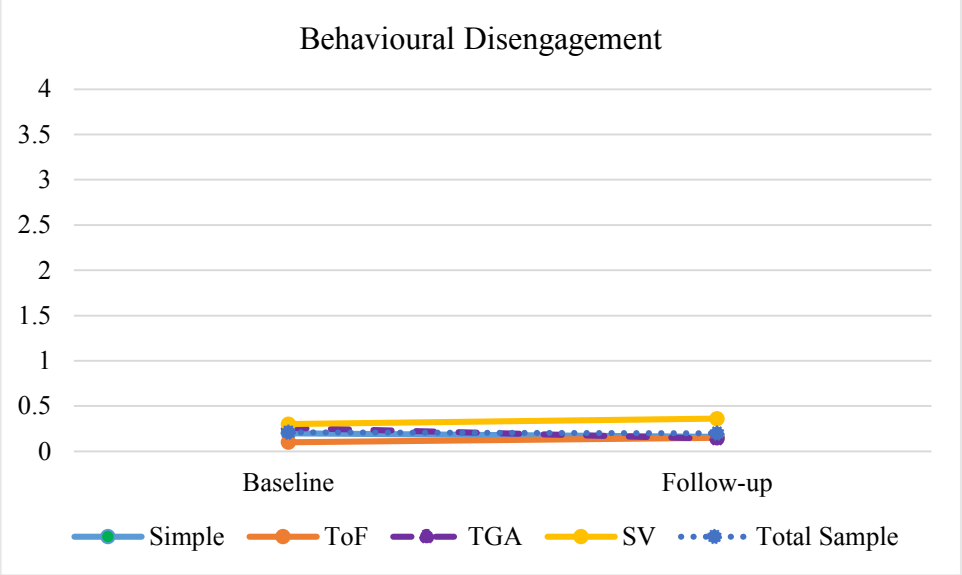
Total Sample


Behavioural Disengagement
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2
1.5
1
0.5
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Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample


Self-blame
4
3.5
3
2.5
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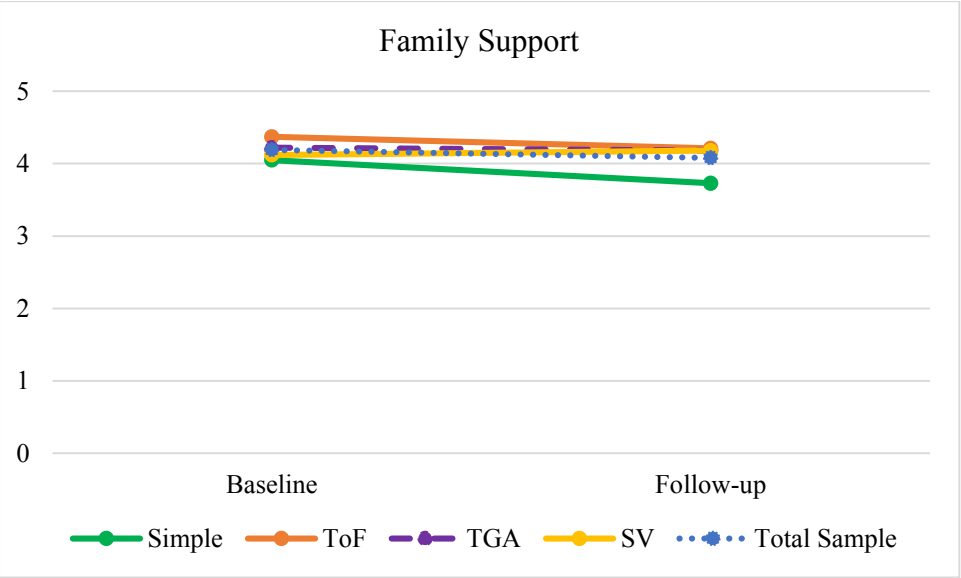
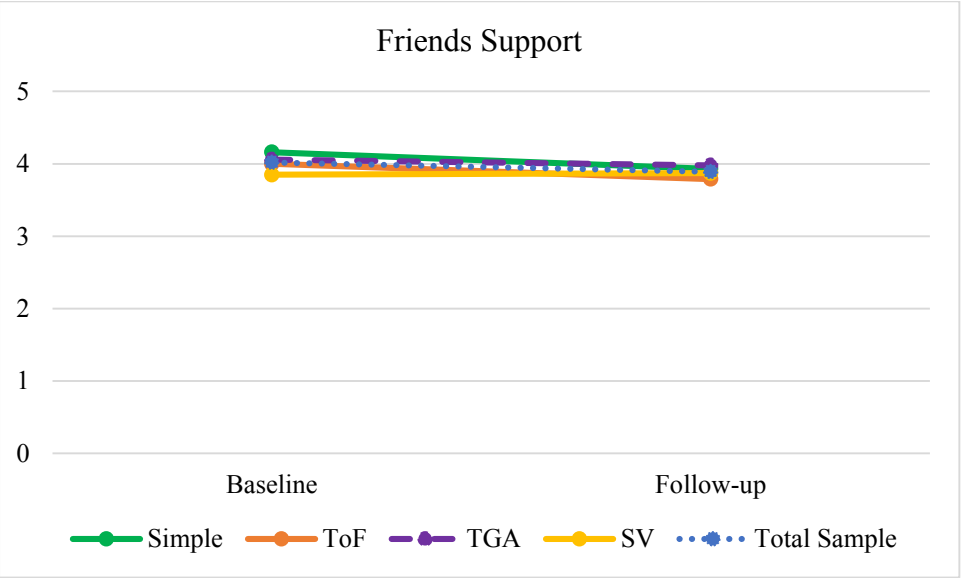
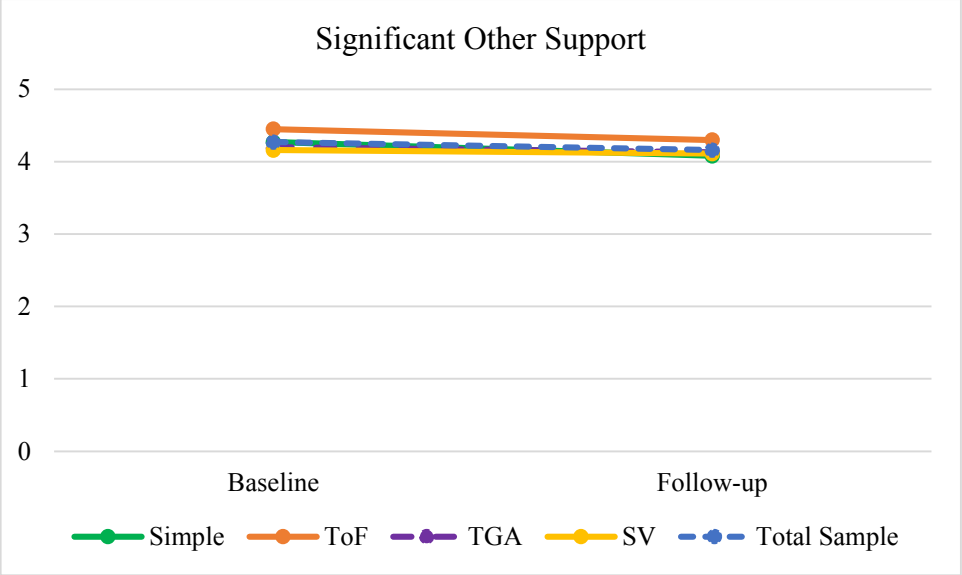
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample

Appendix X. Graphical representation of social support over time






Significant Other Support
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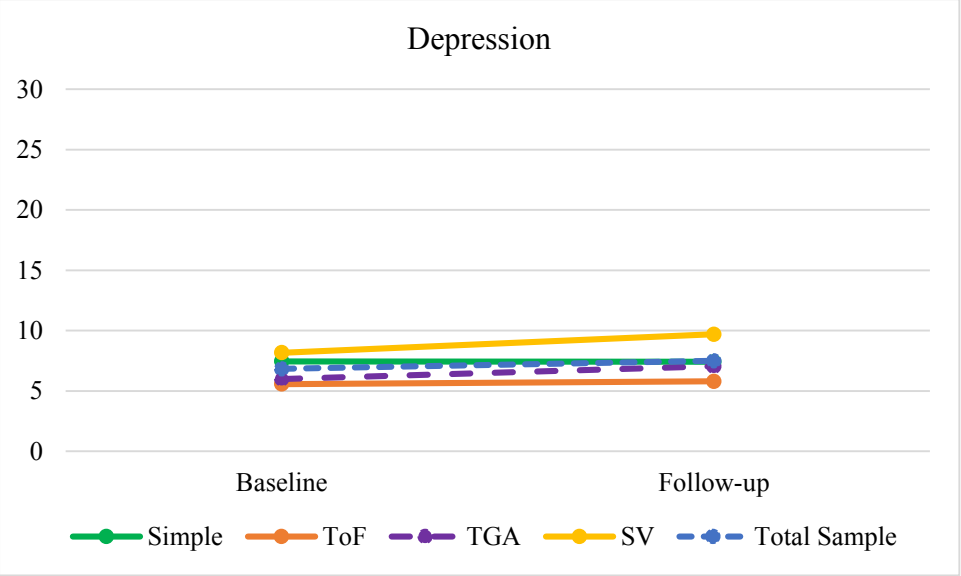
Baseline
Follow-up
Simple
ToF
TGA
SV
Total Sample

Appendix Y. Graphical representation of depression over time



Appendix Z. Information sheet used in the qualitative study

University College London Hospitals NHS
NHS Foundation Trust

Consultants:
Dr Shay Cullen
Dr Fiona Walker
Prof Philipp Bonhoeffer
Prof John Deanfield

Surgeons:
Mr Victor Tsang
Ms Carin van Doorn
Mr Martin Kostolny

Clinical Nurse Specialists:
Ruth Brooks
Marie Francis
Fiona Kennedy
Kerry Romer

GUCH Office
The Heart Hospital
16-18 Westmoreland Street
London W1G 8PH
Tel: 020 7573 8808
Nurses: 020 7573 8872
Fax: 020 7573 8807
Email: guch.dept@uclh.org
Website: www.uclh.org

UCLH Project ID number : 08/0326

CONFIDENTIAL
INFORMATION SHEET (Version 4-11/11)

Quality of life in GUCH patients, qualitative study

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Fiona Kennedy, Nathalie Picaut, Theodora Fteropoulli, Manavi Tyagi.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

You are being invited to take part in a qualitative study. Before you decide it is important for you to understand why the research is being conducted and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part.

What is the purpose of the study?
Although various studies have been conducted into the quality of life of children with congenital heart disease very little has been carried out with adults with congenital heart disease. Moreover, there is little research examining the patients' personal experiences about their quality of life. In order to better plan our services to support long term care we would find it extremely valuable to have an understanding of how congenital heart disease affects quality of life. Therefore, we in the GUCH Unit at the Heart Hospital, UCLH, and the Health Services Research Group at City University would like to find out how you feel your heart condition affects your quality of life. The information you give us will then be used to improve the long-term care of adults with congenital heart disease.

Why have I been chosen?
You are being asked to take part in this study because you have a diagnosis of congenital heart disease and you have participated in the first/second phase of the study.

Do I have to take part?
It is up to you to decide whether or not to take part. If you do decide to take part you will be given this information sheet to keep and be asked to sign a consent form. If you decide to take part you are still free to withdraw at any time and without giving a reason. A decision to withdraw at any time, or a decision not to take part, will not affect the standard of care you receive. Your contribution to the other phases of this study will also remain unaffected.

What is involved in the study?
If you agree to take part in this study you will be asked to attend the Heart Hospital on a day and at a time that suit your schedule. The meeting will involve an interview with the researcher which will be audio taped. You will be asked a few questions about your experiences with living with congenital heart disease. The conversation will largely be guided by what and how much information you want to share with us. Please note that you will be free to share only as much information as you wish. It is anticipated that the process will take approximately 60 minutes.

What are the possible benefits of taking part?
Whilst there are no immediate benefits for people participating in this study, it is hoped that this work will contribute to improving the long-term care of adults with congenital heart disease.

Confidentiality of records
We also need permission to access your medical records, which relate directly to this study. All the information we obtain will be strictly confidential. City University will overview the collection, storage and handling of the data and Professor Newman, in his capacity of chief investigator, will be responsible for security and access to the data. Only study investigators (named above) will have access to the data. The information collected during the study, with exception of your name, will be stored and analysed confidentially in a computer. No identifiers on the data held by computer will enable a third party to link the data to you. A study ID number, assigned to you during the first phase of the study will appear on all data including medical information and questionnaires. All data will be kept strictly confidential and secured under lock and key in City University. The data will be stored for 5 years after the study has been completed. The results of this study may be published within the medical literature, however, no personal details will be revealed. Copies of the publications will be available to you from the researchers. A report of the findings of the research will be sent to all interested participants in approximately 2 years from the start of the study.

Comments or concerns during the study
If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on 020 7040 0871/0878. If you remain unhappy and wish to complain formally you can do this through the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this information sheet.

Ethics Committee Review
All proposals for research using human subjects are reviewed by an ethics committee before they can proceed. This proposal was reviewed by the Joint UCL/UCLH Ethics Committee (Bentham) in Ethics of Human Research.

Thank you for taking the time to read this. If you decide to take part you will be given a copy of this information sheet and a signed consent form to keep.

Appendix AA. Consent form used in the qualitative study

University College London Hospitals NHS
NHS Foundation Trust

GUCH Office
The Heart Hospital
16-18 Westmoreland Street
London W1G 8PH
Tel: 020 7573 8808
Nurses: 020 7573 8872
Fax: 020 7573 8807
Email: guch.dept@uclh.org
Website: www.uclh.org

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 4-11/11)

Quality of life in GUCH patients, qualitative study

Investigators:	Professor Stanton Newman, Professor John Deanfield, Dr Shay Cullen, Fiona Kennedy, Nathalie Picaut, Theodora Fteropoulli, Manavi Tyagi.
Contact details:	GUCH Unit, Heart Hospital, 16-18 Westmoreland St., London W1G 8PH Tel: 020 7573 8889

Please read the following statements and initial box

1. I confirm that I have read and understand the information sheet (Version 4, 11/11) for the above study and have had the opportunity to ask questions. ☐

2. I confirm that I have had sufficient time to consider whether or not I want to be included in the study. ☐

3. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my medical care or legal rights being affected. ☐

4. I understand that all data will be kept strictly confidential to this research and give permission for the above investigators to have access to my medical records. ☐

5. I agree for the interview to be audio recorded and the recordings will be destroyed and only an anonymous written copy of my interview will be kept for research purposes. ☐

6. I agree to take part in the above study. ☐

Continued on next page/

UCLH Project ID number: 08/0326

CONFIDENTIAL
CONSENT FORM (Version 4, 11/11)

Signed: _____ Date: _____

Full name in block letters: _____

Signed (investigator): _____ Date: _____

Full name in block letters: _____

Comments or concerns during the study.

If you have a concern about any aspect of this study you should speak to the researchers who will do their best to answer your questions either at your appointment or on 020 7040 0871/0878. If you remain unhappy and wish to complain formally you can do this through the Complaints Manager, UCLH, 2nd Floor West, 250, Euston Road, London NW1 2PQ. Please quote the UCLH project number at the top of this consent form.

1 form for patient
1 to be kept as part of the study documentation
1 to be kept with hospital notes

Appendix BB. Reflective account for qualitative study

My PhD rests on a pragmatic paradigm, guided by *"what"* the research problem is and *"how"* it can be researched (Creswell & Plano Clark, 2011). It is a paradigm that embraces and values the contribution of both qualitative and quantitative approaches to research. During my qualitative enquiry, I was concerned about how I can objectively interpret and present people's experiences. I understand that the subjectivity of the researcher is recognised in qualitative research because it not only guides the selection of topic and formulation of questions but also the methodology and interpretation of the data. Therefore, the researcher needs to reflect on their values and assumptions they bring to the study in order to recognise their subjectivity and facilitate objective comprehension of the world (Ratner, 2002). What follows is a reflection on my own background, the methods and data collection, and the data analysis and interpretation.

Researcher background

I am female, educated to postgraduate level, and from a Greek cultural background. This study was not just an academic endeavour but also reflects my personal and academic interests. My research interests have always revolved around the impact and adjustment to chronic illness, with particular focus on cardiovascular disease. My father's diagnosis of coronary heart disease and his subsequent coronary artery bypass surgery played a significant role in my choice of a career in health psychology and informed the subject of both my undergraduate and postgraduate theses. My undergraduate thesis focused on the role of self-regulation in Cypriot cardiac patients. My postgraduate thesis examined the relationship between dispositional optimism, illness perceptions, and coping in patients undergoing treatment for cardiovascular disease. After completing my MSc in health psychology I wished to keep my research focus on cardiovascular disease. I worked on a large study of cognitive and psychosocial functioning in ACHD for three years. This work enhanced my interest in this understudied population and influenced my decision to pursue a

PhD on the subject. I was hoping that the study would contribute to our understanding of HRQoL in ACHD and how we can help people adjust to this chronic condition.

I had little prior experience interviewing participants for qualitative research in health psychology. However, during my PhD I attended a two-day workshop at the NatCen which involved a "hands-on" approach to conducting face-to-face interviews. This workshop had proved very helpful and made me feel confident in my ability to conduct the qualitative interviews. Further workshops in qualitative data analysis using framework analysis (developed by the NatCen) provided me with sufficient skills to conduct the research, including practical exercises in the analysis of actual data using framework analysis.

My experience during recruitment and data collection for the quantitative studies provided me with invaluable experience on how to approach and talk to people about their HRQoL. After completing the questionnaires, many people were very keen to discuss their experiences with living with ACHD beyond the questions in the psychosocial measures. This experience of informal conversations with participants taught me how to maintain an objective yet friendly position and be reflective on my thoughts and feelings about the experiences that they shared with me.

Reflection on the methods and data collection

I appreciate that my pre-conceptions about people's experiences regarding their HRQoL derived from the informal chats might have influenced my interview guide. However, I made every effort in setting these pre-conceptions aside when finalising the interview guide. I believe some of these pre-conceived notions were overcome by having the interview guide reviewed and revised by the supervisory team and two independent experts in qualitative research. In addition, none of the participants in the interviews were part of the earlier informal chats I had during the quantitative studies which minimised the effects of my pre-conceptions.

During the interviews I introduced myself as PhD student at City University London. I explained the aims of the study to participants and ensured that they understood that they could terminate the interview if they felt uncomfortable. I was aware of the emotional burden that ACHD can cause and I tried to approach topics with sensitivity and compassion. I generally approached this study with the understanding that qualitative data is rich in detail and felt it was inappropriate to interrupt participants even if they were going slightly off-topic. I did not push participants to give more detail where they did not wish to or where they found it difficult to articulate their views. This explains the short or long length of some interviews.

During the interviews, two female participants became upset when they described particular experiences relating to pregnancy and uncertainty. I paused the tape recorder and gave them the opportunity to have a break. I asked them whether they wanted to terminate the interview but on both occasions participants wished to continue. I did not feel uncomfortable at all and remained calm and empathetic, knowing that pregnancy is a very sensitive topic for women with ACHD. I provided water and tissues for the participants and the interviews continued. On reflection, me being a female researcher, may have influenced what participants were willing to share with me. Specifically, the challenges of family planning and pregnancy discussed by female participants may not have come up with a male researcher. On the other hand, male participants discussed in much detail how their ACHD had influenced (or not influenced) their occupational life. This resonated with my experiences of growing up in a primarily patriarchal culture, where the father is the main bread-winner in the family. Furthermore, it is possible that my gender made some male participants more hesitant in sharing negative emotional experiences because they may have thought I would view them as vulnerable.

One particular interview with a male participant made me feel uncomfortable; he provided short yes/no answers despite my various attempts to prompt him. This interview was

therefore very short in duration. On occasion, I found myself asking one or two questions in a closed-ended way. As I became aware of that, I rectified it quickly. This participant had a Simple defect which did not have a significant impact on his HRQoL. This experience initially led me to think carefully about my interview topic guide in relation to people who generally did not experience impact on their HRQoL as a result of their ACHD. However, subsequent interviews with people with similar experiences had proved to be quite insightful in the ways they coped with their condition. In addition, studying my reflective notes from that interview suggested to me that the particular participant found it difficult to articulate his views and experiences and was generally not open about sharing details.

Overall, I was able to establish rapport with most of the participants quite quickly. This was not surprising since the participants were already familiar with the study and its purpose and had already met me during the quantitative studies. I got the impression that the participants felt that they were part of the research team instead of being researched and I was moved by their commitment to the study, and their praises about its significance. From my part I felt very lucky to be able to get such an insight of their experiences.

At the end of each interview I ensured that participants were not left in an emotional state and assured them about confidentiality. Overall, participants were very keen to sharing their experiences. The variety of experiences acquired from the interviews provided some reassurance in relation to potential bias in participant selection.

On reflection, I now realise that during the interview process I was perhaps viewed not as an "insider" but rather a "bridge" or a "medium" between the participants and the healthcare system. For many of them, there weren't many opportunities to discuss their experiences and feelings about living with ACHD in such depth. As a result, I felt a responsibility towards the participants to be able to translate their experiences in an accurate and objective manner.

Reflection on data analysis and interpretation

The overall approach I took for the data analysis was to search for common themes or patterns across the participants' narratives. Since there are no rules regarding what constitutes a theme, within the present study a theme needed to *"capture something important about the data in relation to the research question, and represent some level of patterned response or meaning within the data set"* (Braun & Clarke, 2006). This meant that themes were not considered only in terms of occurrence but also in terms of their importance to the research questions. Since framework analysis enables both inductive and deductive approaches, my data analysis was based on specific questions and drew on previous research but also allowed for new themes to emerge.

There are two points which I believe are particularly relevant in my analysis and interpretation. Coming into this study and as a health psychology student, I was already familiar with the literature in ACHD before data collection and analysis. I was aware that people with ACHD face various challenges and I was prepared for themes to emerge about physical limitations, emotional impact, uncertainty, and social relationships; these themes were apparent in the participants' narratives. Moreover, I share the view of most researchers in this field that HRQoL is a multidimensional concept. This has likely influenced my analysis and structure of the findings with regards to the impact of ACHD on the various areas of people's lives. Participants who described a negative impact of ACHD on their lives were perhaps given particular weight during data analysis partly because their experiences resonated with the theoretical literature I was familiar with. However, I made sure to present positive or opposing narratives where it was relevant. My background in psychology may have also influenced my focus on narratives highlighting psychosocial processes that helped people cope and adjust to living with ACHD, in particular acceptance and the role of social support. It is possible that participants might have focused on other issues (e.g. satisfaction with healthcare) had I not somehow guided the conversation towards this direction.

Furthermore, the qualitative interviews were supplemental and designed to complement the dominant quantitative component of the thesis, thus the analysis and interpretation of the interview data were influenced by the quantitative study. Although it was not the aim of the qualitative study to look at the experiences of people within various diagnostic groups separately from each other, there were some data that suggested the differing ways ACHD had an impact on these groups of people. I discussed these differing experiences but I have not explored them in much depth due to the limitations of the study in terms of time and resources to allow for data saturation within groups. It is possible that these observations were influenced by my assumptions that the four diagnostic groups would experience challenges to differing degrees.

Throughout the analysis and interpretation I was influenced by my knowledge of the field. I drew from existing literature and concepts from theories (normalization and uncertainty are two examples) to make sense of the data. During the final stages, I realised that there was a need to consciously choose the data that I was going to present, which were relevant to the research questions of the study. I understand that these decisions were influenced by various factors such as writing an interesting and coherent story to be presented to the wider academic community that remained true to the participants' experiences. Thus, I selected quotes that were more illustrative of the point I wanted to get across, ensuring that all participants were represented. Overall, I recognise that my research background may have had an impact on the interpretation and the presentation of the participants' experiences.

Appendix CC. Stage 2 of framework analysis: grouping categories thematically and developing an initial conceptual framework *(example of initial themes & sub-themes that were later developed into the overarching theme of coping)*

Key theme	Sub-themes	Description	Example of indexed data
Coping	Adjusting to physical limitations	Process of accepting the condition and their limitations and *how* they incorporate them into their lives.	*"just focusing on certain house chores I would do, but I will do it and I'll have a rest afterwards so I just... pace myself to do it and the jobs really in the house"*
	Adjusting to treatment	Accepting and adjusting to their treatment regimen.	*"So used to them [hospital visits] now, it's no, no problem at all. You're sort of used to the people, what, you know, the various things that you have to do. Sort of, sort of it's like a routine"*
	Being positive	Positive feelings of luck, positive outlook/optimism, and gaining perspective.	*"I suppose just emotionally...just that the fact that, yeah, that the, the realisation that "I am here" and that I was a lucky one"*
	Seeking support	Actively seeking practical support, advice, and emotional support from family, friends, and HCPs. Also includes deliberately hiding facts and feelings from others.	*"Sharing it helps you eem but sometimes, you know, actually "Do I want her to worry as much as I'm worrying about it?" Probably not so, you know, you keep that bit inside you"*
	Social comparisons	Comparisons with other people including those in worse and better condition.	*"There's people in the world with much worse heart conditions than me but travel the world…It's like I'm gonna have to put things into perspective"*
Personal control	n/a	Whether (or not) they feel that they have control over their QoL and what they think they can do to change it.	*"I don't think, I'm not in control. Your health's in control isn't it? You can't control your health, well you can...but obviously I can't control it. If my heart wants to go...funny or...what, I can't control that"*

Key theme	Sub-themes	Description	Example of indexed data
Social support	Support from partner	Ways their partner provides support.	*"he [husband] really pushes me to do all of those things. So he might remind me to take my tablets"*
	Support from parents	Ways their parents provide support.	*"I think my mum is more physical support for when I'm too ill to do things. She will come over and, like I said previously help me cook dinner"*
	Support from friends	Ways their friends provide support.	*"I was, got really upset, my friend was straight round after work, saying "Right if we can't do this, we can't do it. There's no point you making yourself all over it or having your plan" and I needed that then"*
	Support from employer and colleagues	Ways their employer and colleagues provide support.	*"we've got a good team at work, there's a good team of us so...there's always some support there"*
	Support from healthcare professionals	Ways HCPs provide support.	*"N [nurse] gave me one of these...sort of relaxation CDs, you know, with the breathing and the visualisation and stuff. And that actually helped quite a lot"*
	Preferred support	Type of support they want/need.	*"I like to think I'm quite tough emotionally. So when I physically can't do things it's the physical things that I need help with"*
QoL meaning	Good QoL meaning	What good QoL means for them.	*"It's got to be your health comes first. Eeh and then just enjoying your family, enjoying your social life, and again work"*
	Priorities	Life domains they prioritise and/or give particular attention to.	*"I think my health is more important. Eem...looking after myself and looking after....my partner is more important"*

Appendix DD. Stage 3 of framework analysis: developing and refining themes and sub-themes from stage 2 *(example of coping)*

Key theme	***Sub-themes***	***Description***
Gaining perspective	Social comparisons Positivity	Gaining perspective was developed as a main theme under which *positivity* and *social comparisons* were coded as sub-themes. Both were directed towards gaining perspective based on the coded data.
Acceptance & adjustment	n/a	Formed by merging *adjustment to physical limitations* and *adjustment to treatment*. Treatment was not independent of disease impact.
Social influence	Receipt of social support Seeking social support	Social influence was developed as a main theme under which *receipt of social support* and *seeking social support* were coded as sub-themes. Seeking social support as a coping mechanism was not independent of the availability of social support.
Priorities & goals	n/a	QoL meaning and personal control themes were merged into this theme as coded data reflected how people prioritise and how they try to control and improve aspects of their lives.

Appendix EE. Stages 4 & 5 of framework analysis: creating case by theme charts and interpretation *(example of acceptance & adjustment)*

Theme: Acceptance & adjustment

Case*	Summary	Illustrative quotations	Notes/comments on patterns, relationships, explanations
Ron male, 54, TGA	Knowledge and acceptance of his limitations, getting on with it, avoiding activities he can't do, adjusting speed/time for chores, leading normal life, hospital appointments part of his life, planning.	*"I know my limits if you know what I mean. I don't run around, I can't play football or I don't go to the gym or anything like that but yeah I get by[…] there are certain things you sort of limit yourself, you know what yourself you can do"*	❖ Acceptance of ACHD as part of life and identity, acknowledging limitations, adjusting to limitations, and achieving normalcy. ❖ Similar to normalization concept in previous studies – ongoing process? ❖ Evident impact of health deterioration and change in treatment regimen on attempts to adjust – dynamic adjustment. ❖ Degree of successful adjustment related to degree of consequences (see theme physical consequences)
Janis female, 35, SV	Avoiding activities she can't do, concentrates on what she can do, acknowledging limits.	*"There's always gonna be limits. That's just parts of living with a heart condition. And it's just finding ways of them not being a problem. So we don't go to India for holiday. We go somewhere closer. That's, to me that's just common sense."*	
Peter male, 39, Simple	Thinking strategies around his condition, avoiding activities that may cause problems, planning ahead.	*"[…] I guess if you take going to a holiday of altitude, rather than sort of pushing on through it and seeing how I go, I would just not really go on that holiday, cause it might be a problem and I don't wanna get into a problem situation."*	
Robert male, 40, TGA	Thinking ahead about activities, pacing himself for chores, plan the day, knowledge of his limits, not pushing himself.	*"just focusing on certain house chores I would do, but I will do it and I'll have a rest afterwards so I just pace myself to do it…"*	
Jim male, 36, ToF	Always lived with it, accept it, dealing with issues as they come, acceptance of possible interventions, hospital appointments and tests as *second nature*, part of his life, normal life.	*"…you can make a big deal out of it if you want to or you just accept it as being there and you deal with it if issues come up. And I think that's what I've tried to do with my life…"*	

*Example chart includes only a subset of cases.