Physical activity and exercise among patients with congenital heart disease: towards a model of pediatric cardiac rehabilitation

by

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A thesis submitted in conformity with the requirements
for the degree of Doctor of Philosophy

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2017

Abstract

Patients with congenital heart disease (CHD) have reduced physical activity compared to healthy peers. Past interventions have focused on young children with CHD and improvements in exercise capacity achieved with exercise training. Regular physical activity is beneficial for CHD patients to achieve optimal long-term physical and psychosocial outcomes. This research sought to understand the physical activity behaviour of adolescents (13-17 years old) and emerging adults (18-25 years old) with CHD. In the first study, longitudinal trends in a cohort of patients with Fontan circulation (n=33; 21±3years) indicated a 31% reduction in moderate-to-vigorous physical activity (MVPA) over a seven-year period from childhood to adulthood. More vigorous physical activity as a child was associated with greater physical activity as an adult. A cross-sectional study (Study 2) including qualitative interviews in a separate cohort of emerging adults with CHD (n=15; 21±3years) identified that emerging adults perceived family as supportive, appreciated parental involvement, accepted their CHD as normal, contextualized activity in terms of school and sports, felt that activity accumulated at work was sufficient, and participated in physical activity to be healthy. This cohort of emerging adults reported moderate self-efficacy

for exercise, high quality of life, and low heart-focused anxiety. Daily MVPA measured by accelerometer was 26 minutes/day. In a third study, adolescents with CHD (n=31; 15±3years) participated in a randomized control trial of an adapted Motivational Interviewing (MI) behavioural intervention compared to a control condition. Participants achieved 21 minutes/day of MVPA and reported high self-efficacy and quality of life. Participants (n=15) who received the behavioural intervention did not improve physical activity, fitness, or psychosocial outcomes compared to control patients. No participants met the current physical activity recommendations. Physical activity promotion should be a priority for the pediatric CHD population and should include age-appropriate interventions. Behavioural interventions require more information derived from additional research to determine the most feasible and effective approach to improve physical activity among patients with CHD across the lifespan. Family-based interventions that include vigorous activities in childhood may be one approach to help patients achieve higher levels of activity in adulthood.

Acknowledgments

I would like to gratefully acknowledge and thank all of the patients who contributed to this thesis research. I would also like to acknowledge the support I received from the Hospital for Sick Children (Labatt Family Heart Centre), University Health Network (Toronto Congenital Cardiac Centre for Adults), and the Heart and Stroke Foundation of Canada.

Thank you to my primary supervisor, Dr. Brian McCrindle, for your tremendous guidance and support throughout this program. Your focus and drive towards success was instrumental and your generosity is greatly appreciated.

To my committee members, Dr. Sherry Grace and Dr. Ken Allison, thank you for providing insight into the development and production of this thesis research. You provided superb mentorship that helped me develop as a researcher.

Thank you to my collaborators and co-investigators, particularly Dr. Adrienne Kovacs, Dr. Gina Dimitropoulos, Dr. Jane Schneiderman, and Dr. Laura Banks. I'd like to also thank all the staff and fellow students in the Cardiovascular Clinical Research Unit, and the staff at the Hospital for Sick Children for your support and encouragement over the years.

A special thank you to my family and friends for supporting me over the years and learning to stop asking me "when will you be done?".

Last but not least, thank you to my wife, Frances. I couldn't have done any of this without your love, support, patience, and willingness to listen anytime I needed to talk. It's been a long haul and you've been there every step (and tumble) along the way! And to my children, Audrey and Everett, thank you for being the best possible distraction these last few years. You continue to teach me more than school ever could - and thank you for learning to sleep through the night so daddy could work…

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List of Abbreviations

ACHD	Adult Congenital Heart Disease
AMI	Adapted Motivational Interviewing
AS	Aortic Stenosis
ASD	Atrial Septal Defect
AT	Anaerobic Threshold
AV	Atrioventricular
AVSD	Atrioventricular Septal Defect
BAV	Bicuspid Aortic Valve
BIA	Bio-electrical Impedance Analysis
BMI	Body Mass Index
CACPR	Canadian Association of Cardiovascular Prevention and Rehabilitation
CAQ	Cardiac Anxiety Questionnaire
ccTGA	Congenitally-corrected Transposition of the Great Arteries
CEP	Certified Exercise Physiologist
CFS	Canadian Fitness Survey
CHD	Congenital Heart Disease
CHMS	Canadian Health Measures Survey
CHQ	Child Health Questionnaire
CI	Confidence Interval
CoA	Coarctation of the Aorta
CONSORT	Consolidated Standard of Reporting Trials
CPT	Cardiopulmonary-Exercise Testing
CSA	Computer Science Application
DORV	Double-outlet Right Ventricle
FITT	Frequency, Intensity, Type, Time
HAES	Habitual Activity Estimation Scale
HBM	Health Belief Model
HLHS	Hypoplastic Left Heart Syndrome
HR	Heart Rate
HRHS	Hypoplastic Right Heart Syndrome
HRQL	Health-related Quality of Life
IAA	Interrupted Aortic Arch
LV	Left Ventricle
mCAFT	Modified Canadian Aerobic Fitness Test
mHealth	Mobile Health
MI	Motivational Interviewing
MINT	Motivational Interviewing Network of Trainers
MITI	Motivational Interviewing Treatment Integrity
MS	Mitral Atresia
MS	Mitral Stenosis
MVPA	Moderate-to-Vigorous Physical Activity
NHANES	National Health and Nutrition Examination Survey
NYHA	New York Heart Association

OR	Odds Ratio
PAQ-A	Physical Activity Questionnaire for Adolescents
PAQ-C	Physical Activity Questionnaire for Children
PDA	Patent Ductus Arteriosus
PedsQL	Pediatric Quality of Life
PFO	Patent Foramen Ovale
PHN	Pediatric Heart Network
PS	Pulmonary Stenosis
PTA	Patent Truncus Arteriosus
PVR	Pulmonary Vascular Resistance
QoL	Quality of Life
RCT	Randomized Controlled Trial
REACT	Rehabilitative Exercise and Activity Clinical Trial
REAL	Rehabilitative Exercise and Activity for Life
RQ	Respiratory Quotient
RV	Right Ventricle
SES	Self Efficacy Scale
SMS	Short Messaging Services
SV	Single Ventricle
TAPVR	Total Anomalous Pulmonary Venus Return
TCCCA	Toronto Congenital Cardiac Centre for Adults
TGA	Transposition of the Great Arteries
TOF	Tetralogy of Fallot
TTM	Transtheoretical Model
VAS	Visual Analogue Scale
VSD	Ventricular Septal Defect
WHO	World Health Organization
YRBS	Youth Risk Behaviour Survey

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Chapter 1
Introduction

1 Research Overview

1.1 Background and Rationale

Congenital heart disease (CHD) is defined as "the presence of a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" [1]. CHD occurs in approximately 1-4% of all live births [2]. Thanks to advances in medical and surgical practice, it is estimated that there are now more adults living with CHD than children. Due to increased survival of CHD patients, long-term clinical care and management of patients with CHD is increasingly important. Patients with CHD are not "cured" of their heart disease and should be followed by a specialized CHD care team. In addition, there is a growing need to address the long-term care required for patients with CHD to maintain health and prevent illness. Strategies to adopt a healthy lifestyle that includes regular physical activity should be introduced early in the patient's care.

Research has shown that children and adults with CHD are less physically active than others without CHD. This inactivity often occurs despite successful surgical correction of the underlying structural defect and subsequent adequate cardiac function. Reduced physical activity participation may be related to parental overprotection and/or self-limitation due to a lack of clarity around safe and appropriate activity recommendations [3-5]. Furthermore, recommendations are commonly focused on restriction rather than promotion of physical activity and provided with limited detail (i.e., no contact sports, no competitive sports) [6, 7].

Physical activity and exercise recommendations for the CHD population have traditionally taken a prescriptive approach [8, 9]. Given the chronic nature of CHD, interventions that focus on exercise prescription alone may not provide the necessary behavioural aspect to adopt and maintain an active lifestyle. Integrating behavioural interventions into the clinical care of patients may serve as an effective long-term strategy to achieve and maintain a healthy lifestyle.

The goal of this research was to understand the physical activity behaviour among adolescents and emerging adults with CHD. The results of this research could be used as a starting point for the development of future clinical exercise and physical activity interventions and/or programs.

1.2 Interventions: Past and Present

Physical activity and exercise training interventions have traditionally aimed to improve physiological outcomes such as exercise capacity, blood pressure, and cardiorespiratory function [10, 11]. The delivery of physical activity and exercise training interventions has often taken a prescriptive approach, addressing the "what, when, and where" aspects of an individualized intervention. Deviation from a prescription is often discouraged and there is an underlying expectation that patients adhere to the prescription provided [12]. Prescriptive interventions often require additional resources (i.e., space, equipment) and trained personnel (i.e., certified exercise physiologists, medical supervision) to execute an effective regimen. Prescriptive exercise interventions demonstrate short-term benefits and often lack long-term effectiveness to maintain desirable health outcomes. Behavioural interventions on the other hand, may be viewed as interventions designed to affect an individual's actions towards their health with an underlying

goal to help change their behavior and may be applied at the individual, community, and population levels [12].

Vaccaro et al. described the basic components of a pediatric cardiac rehabilitation program that would ideally include exercise testing and prescription that focused strictly on the aerobic capacity of the patient [13]. The authors recommended a program based on 12 weeks of hospital-based exercise sessions performed or offered twice per week. Tikkanen et al. identified sixteen pediatric CHD rehabilitation programs between 1981-2010 that included an exercise-training component (Table 1) [8]. These programs included a mix of aerobic and resistance training, with few including flexibility, coordination, and education components. The duration of these programs ranged from 2 weeks to 10 months with 2-3 sessions/week for approximately 30-60 minutes/session. The primary outcome of these programs was mainly focused on aerobic capacity. None of the programs explicitly described physical activity behaviour as a primary outcome, nor included a behavioural component aimed to improve physical activity.

The daily physical activity recommendation for children is to accumulate 60-minutes of moderate-to-vigorous intensity physical activity (MVPA) every day [14]. A gap remains in the literature regarding optimal rehabilitation strategies for patients with CHD. The focus of previous research has been physiological improvements with exercise training in children with CHD, yet research also supports the role of regular physical activity to improve additional health-related outcomes in the lives of patients with CHD [15]. Additional research is necessary to expand our understanding of physical activity behaviour among adolescents and adults with CHD. Therefore, this research will inform novel exercise and physical activity interventions with the ultimate goal to conceptualize a pediatric cardiac rehabilitation program that will bring evidence into practice.

Table 1. Summary of studies including cardiac rehabilitation programs for young (4-25 years old) CHD patients (updated/adapted from Tikkanen et al. 2012).

References	Location	Patient Population	Ages	N	Patients Treated	Type of Exercise	Duration	Frequency	Intensity	Sessions
Morrison [16]	Academic fitness centre, home	Heterogeneous	12-20	143	72	Education, Motivational Interviewing	6 months	1 day, monthly follow-up	Not specified	Not specified
Duppen [17]	Supervised fitness centre	TOF/Fontan	10-25	93	56	Aerobic	12 weeks	3/week	60-70% of HR reserve	60 min
McBride [18]	Hospital	Heterogeneous	10-16	20	20	Aerobic, resistance	2-10 months	3/week	HR at AT, 60% of MVC	30-60min
Rhodes [19]	Hospital	Heterogeneous	8-18	33	15	Aerobic, resistance	3 months	2/week	HR at AT, light resistance	60 min
Moalla [20]	Home	Heterogeneous	12-15	18	10	Aerobic	12 weeks	3/week	HR at AT	45 min
Brassard [21]	Home (2), Hospital (3)	Fontan	11-26	5	5	Aerobic, resistance	2 months	3/week	50-80% peak VO_2	20-30 min
Opocher [22]	Hospital + Home	Fontan	7-12	10	10	Aerobic	8 months	2/week	50-70% peak VO_2	30-45 min
Rhodes [23]	Hospital	Heterogeneous	8-17	16	16	Aerobic, resistance	3 months	2/week	HR at AT, light resistance	60 min
Minamisawa [24]	Home	TOF	11-25	11	11	Aerobic	2-3 months	2-3/week	60-80% peak HR	40 min
Fredriksen [25]	Hospital or supervised gym	Heterogeneous	10-16	93	55	Aerobic, resistance, flexibility, education	2 weeks or 5 months	Daily or 2/week	65-80% peak HR	Not specified
Sklansky [26]	Hospital	TOF	6-16	11	11	Aerobic	2 months	3/week	60-70% peak HR	30 min
Balfour [27]	Hospital	Heterogeneous	13.5-19.8	6	6	Aerobic, education	3 months	3/week	>70% peak HR	30-40 min
Calzolari [28]	Hospital	TOF	6-16.5	18	9	Aerobic, respiratory	3 months	3/week	60-70% peak HR	60 min
Longmuir [29]	Home	Heterogeneous	4.7-14.3	40	17	Aerobic, resistance, flexibility, coordination	6 weeks	2/week	Capability of talking during exercise	Not specified
Longmuir [30]	Home	Heterogeneous	4.7-14.3	60	30	Aerobic, resistance, flexibility, coordination	6 weeks	2/week	Capability of talking during exercise	Not specified
Bradley [31]	Hospital	TOF/TGA	4-13	9	9	Aerobic, resistance, flexibility, coordination	12 weeks	2/week	60-80% peak VO_2	60 min
Ruttenburg [32]	Hospital	Heterogeneous	7-18	21	21	Aerobic	9 weeks	3/week	65-75% peak HR	30 min
Goldberg [33]	Home	TOF/VSD	7-18	26	26	Aerobic	6 weeks	3-4/week	50-70% peak VO_2	45 min

TOF: Tetralogy of Fallot; AT: anaerobic threshold; HR; heart rate; VSD; ventricular septal defect; TGA: transposition of the great arteries; MVC; maximal voluntary contraction.

1.3 Research Goals

Previous physical activity and exercise research among the CHD population has primarily focused on young children and prescriptive exercise training interventions [8, 34]. This research aims to advance the field by studying physical activity behaviours among adolescents and emerging adults with CHD. The goals for this research were to:

1. Determine long-term physical activity trends among a population of patients with complex CHD between childhood and early adulthood.

2. Identify physical activity behaviours and perceptions towards physical activity among emerging adults with CHD.

3. Determine the feasibility of an adapted Motivational Interviewing (MI) behavioural intervention to improve physical activity among adolescents with CHD.

2 Literature Review

2.1 Congenital Heart Disease

2.1.1 Prevalence of CHD

Among congenital defects, CHD is the most common congenital defect anomaly, accounting for 28% of all major congenital anomalies [35]. It is estimated that CHD occurs in approximately 1-4% of all live births, resulting in approximately 1 million newborns with CHD each year world-wide [2]. Between 1930 and 2010, the reported birth prevalence of CHD increased from 0.6 per 1000 to 9.1 per 1000 live births. It is estimated that the prevalence of CHD increases at a rate of 5% per year [36]. The highest birth prevalence of CHD occurs in Asia (9.3 per 1000 live births) while the lowest reported birth prevalence was found in Africa (1.9 per 1000 live births) [37]. The difference between regions may be attributed to a higher parental consanguinity present in the Asian population [38, 39]. The birth prevalence of CHD in Canada has declined from 107.1 per 10,000 births to 85.1 per 10,000 births between 1998-2009, respectively (Figure 1) [40]. Increased prenatal screening and diagnoses that result in termination may account for a 21% reduction in the birth prevalence of CHD, particularly in complex CHD associated with other genetic anomalies.

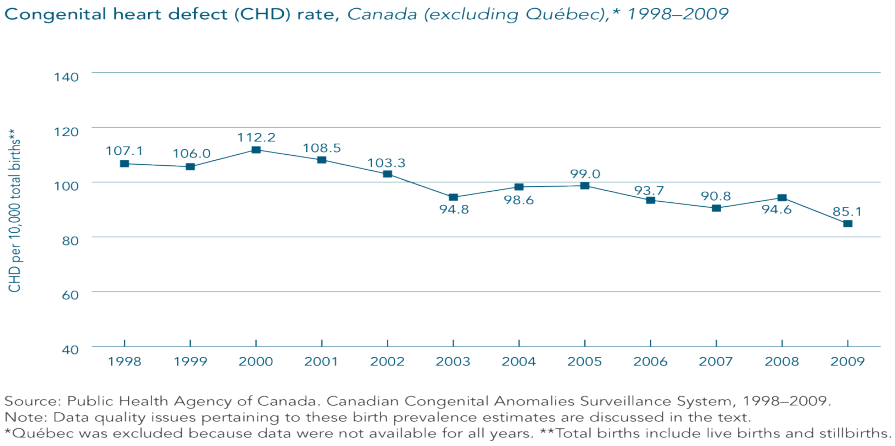

Figure 1. Birth prevalence of CHD in Canada (1998-2009).

The clinical presentation of CHD and time of diagnosis makes it difficult to determine an accurate prevalence, as the prevalence is "the number of living subjects with a particular disease during a specific time frame" [2]. For example, individuals living with undiagnosed CHD and those who died prior to assessment are excluded from prevalence estimates. Improvements in screening and diagnostic technology may account for a reduction of undiagnosed CHD and a more accurate prevalence estimate over time [2]. Differences in the availability, use, and accuracy of pre-natal diagnostic services may further complicate prevalence reports [41].

Overall trends indicate that there is a significant and growing population of individuals living with CHD that require long-term cardiac care. Infant mortality has decreased among the CHD population and now over two-thirds of the CHD population is adults living with some form of CHD [40]. A complete understanding of the long-term consequences faced by the aging CHD population is lacking. It is also important to understand adult-onset morbidities including obesity, hypertension, sleep apnea, and diabetes that may compound the effects from CHD [42]. Long-term surveillance and longitudinal research are necessary to improve our knowledge regarding the physical, neurodevelopmental, psychosocial, and reproductive outcomes for CHD patients across the lifespan.

2.1.2 Pathophysiology of CHD

Risk Factors: The precise etiology of CHD is unknown. Multiple risk factors associated with CHD development have been identified, including both genetic and non-genetic factors. (i.e., environmental teratogens, maternal exposures, lifestyle risk factors) [43]. Evidence supporting the non-genetic risk factors is inconsistent but has identified important considerations for further

research, including the association between CHD and vitamin supplementation, maternal illness, drug exposure (anti-retroviral medications, thalidomide, retinoic acid, valproate, phenytoin, and opioid analgesics), environmental exposure, and paternal factors [44]. Additional emerging non-genetic risk factors include obesity with associated diabetes mellitus and hypercholesterolemia and certain illnesses/conditions (phenylketonuria, influenza, lupus, epilepsy, and rubella infections) [38, 42, 44, 45]. The emerging evidence of non-inherited risk factors associated with CHD has led to recommendations for prospective parents to reduce the risk of having a child with CHD [44].

Genetic Contribution to CHD Development: Genetic research in CHD aims to discover the patho-biological basis of disease and definition of disease risk [46]. This is particularly difficult provided that CHD affects almost all heart structures and the development of the heart structures are extremely complex (Figure 2) [47]. The development of CHD has a well-established genetic contribution, with 15-20% of CHDs associated with known genetic disorders [48]. Patients with CHD exhibit associated syndromes or conditions in 25-40% of cases [43]. Common genetic syndromes associated with CHD include: Down syndrome (40-50%, percentages indicate rate of CHD occurrence in each genetic syndrome), Turner syndrome (25-45%), DiGeorge Syndrome or 22q11.2 deletion syndrome (70-75%), Williams syndrome (75-80%), Noonan syndrome (70-80%), Kabuki syndrome (31-55%), and Alagille syndrome (90%) [49].

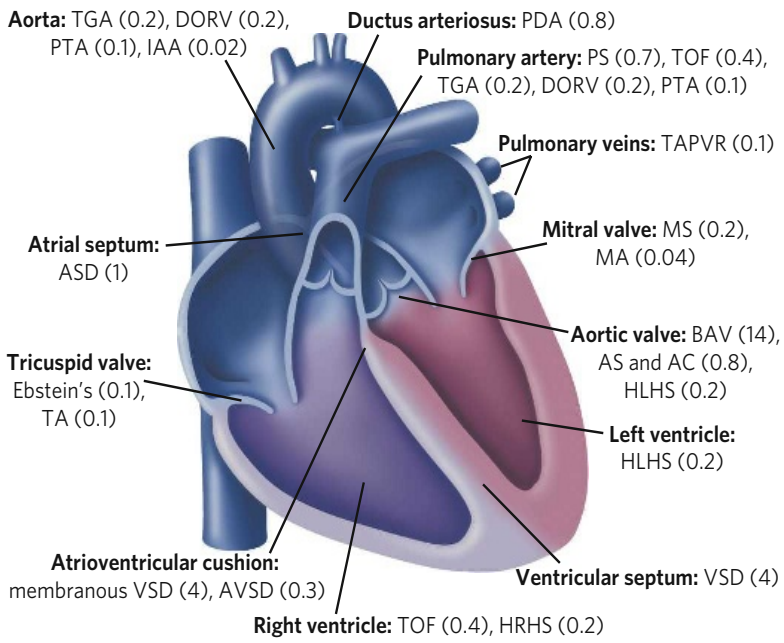

Figure 2. This diagram of the adult heart illustrates the structures that are affected by CHD, with the estimated incidence of each disease per 1,000 live births indicated in parentheses. (TGA: Transposition of the Great Arteries; DORV: double-outlet right ventricle; PTA: persistent truncus arteriosus; IAA: interrupted aortic arch; PDA: patent ductus arteriosous; PS: pulmonary artery stenosis; TOF: Tetralogy of Fallot; TAPVR: total anomalous pulmonary venous return; MS: mitral stenosis; MA: mitral atresia; BAV: bicuspid aortic valve; AS: aortic stenosis; AC: aortic coarctation; HLHS: hypoplastic left heart syndrome; VSD: ventricular septal defect; HRHS: hypoplatic right heart syndrome; AVSD: atrioventricular septal defect; Estein's: Ebstein's anomaly of the tricuspid valve; ASD: atrial septal defect). (Image courtesy of F. Yeung, University of Toronto, Canada) (from Bruneau et al. 2008).

Fahed et al. provided a comprehensive summary of genetic underpinnings of CHD [38]. Genetic CHD mutations can be autosomal dominant, recessive, or X-linked, with great heterogeneity in terms of expression and clinical presentation. While an extensive description of the genetic contribution of CHD goes beyond the scope of this research, it is worth mentioning that the genetic basis for CHD is a rapidly changing field. The complexity of CHD development and its intrinsic strong association with additional genetic conditions supports genome sequencing that may direct personalized therapies for patients [50]. Further studies are needed in order to elucidate the genetics behind CHD and to develop new strategies to prevent CHD.

2.1.3 Classification of CHD

A full spectrum of cardiac lesions exists in the broader CHD diagnosis. Some defects remain undiagnosed and asymptomatic while others are life threatening and require multiple surgeries shortly after birth. The eight most common subtypes of CHD include aortic stenosis, atrial septal defect, coarctation of the aorta, patent ductus arteriosus, pulmonary stenosis, transposition of the great arteries, tetralogy of Fallot, and ventricular septal defect (Figure 3) [37].

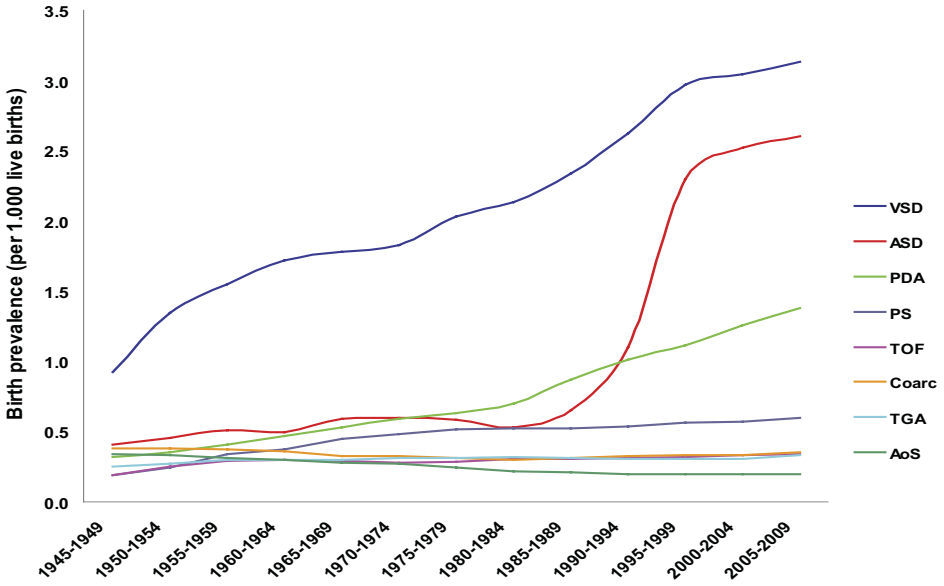

Figure 3. Worldwide birth prevalence of the eight most common CHD from 1945 until 2010. (VSD: ventricular septal defect; ASD: atrial septal defect; PDA: patent ductus arteriosus; PS: pulmonary stenosis; TOF: Tetralogy of Fallot; Coarc: Coarctation of the aorta; TGA: Transposition of the Great Arteries; Aos: aortic stenosis (from van der Linde et al. 2011).

A CHD diagnosis may include mild to severe physiological consequences. With a wide variety of CHD diagnoses that affect different structures, specific surgical or therapeutic interventions are available to help maintain or improve cardiac function (i.e., cardiac output). Therefore, the following section describes CHD with regards to the different hemodynamic consequences between shunt, obstructive, and complex CHD lesions [51-53].

Shunt Lesions: In normal cardiac development, complete septation occurs that separates the heart into right (pulmonary) and left (systemic) systems, with each system consisting of one atria and one ventricle (four chambers) (Figure 4). These separated systems maintain de-oxygenated (pulmonary) and oxygenated (systemic) blood in the circulation. The pulmonary and systemic circulatory systems run in parallel with a 1:1 volume relationship [52]. In shunt lesions, malformation can occur as left-to-right (oxygenated blood returning to the lungs and thus reducing the total cardiac output to the systemic circulation) or right-to-left shunts (de-oxygenated blood enters the systemic circulation resulting in reduced blood oxygen concentration). The two most common septal defects include atrial septal defects (ASD) and ventricular septal defects (VSD). The severity of an ASD and VSD depend on the size (opening) of the shunt and the location along the septum. The size and location of the shunt ultimately dictates the volume differential between the pulmonary and systemic systems and symptom severity [52]. Less severe shunt lesions include a patent foramen ovale (PFO) and a patent ductus arteriosus (PDA). These lesions are the most common remnants of the fetal circulatory system

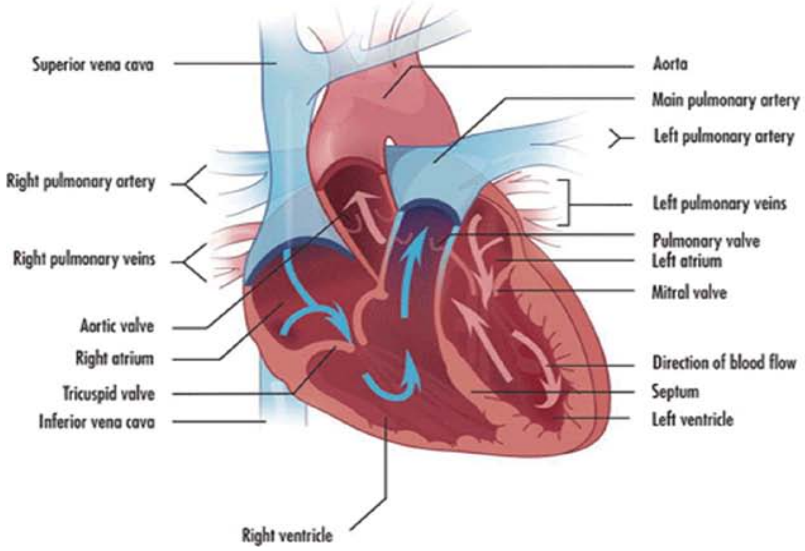

Figure 4. Diagram of a normal heart and structures.

that may persist after birth. A PFO or PDA defect may remain undiagnosed due to the insignificant impact on the circulation and overall physiology (especially compared to ASD and VSD morphology).

Total anomalous pulmonary venous return (TAPVR) is a left-to-right shunt that occurs due to the pulmonary veins returning oxygenated blood from the lungs into the right atrium via the superior vena cava [54]. In normal circulation, the pulmonary veins return oxygenated blood from the lungs to the left atrium to be pumped to the systemic circulation. In TAPVR, the oxygen rich blood is not pumped to the systemic circulation and is unnecessarily sent to the pulmonary circulation. Some level of mixing must occur, usually from right-to-left shunting across an atrial communication or a PDA, which allows blood from the right atrium to pass into the left atrium, and thus brings some of the oxygenated blood into the systemic circulation [55]. This results in reduced blood-oxygen saturation in the systemic circulation and increased pulmonary over-circulation as a result of increased blood volume in the pulmonary circulation. Following surgical repair of the venous connection, patients may experience pulmonary hypertension or post-operative pulmonary venous obstruction, both of which result in poor outcomes and increased risk of death in the first few years of life [54].

Obstructive Lesions: The narrowing or complete occlusion of outflow tracts, valves, or blood vessels introduce downstream cardiovascular morphological consequences. An important consequence is increased afterload resulting in ventricular hypertrophy, characterized by thickening of the ventricular wall, reduced compliance, and high atrial filling pressure [51]. Increased afterload is also a well-known contributor of reduced stroke volume and ultimately reduced cardiac output (Cardiac Output = Stroke Volume x Heart Rate). The severity of the physiological challenges depends on the location of the obstruction.

Right-sided obstructions occur in the outlet portion of the right ventricle (RV) proximal to the pulmonary valve (subvalvar stenosis), at the valve itself (valvar pulmonary stenosis), distal to the valve in the main pulmonary artery (MPA; supravalvar stenosis), or more peripherally in the branch pulmonary arteries [51]. Isolated pulmonary stenosis occurs in approximately 10% of all CHD cases. Obstructive lesions may also be associated with other cardiac malformations or syndromes (i.e., tetralogy of Fallot (TOF), Williams syndrome) or as a result of surgical intervention (i.e., Blalock-Taussig procedure, arterial switch) [56].

Left-sided obstructive lesions occur at the subvalvar, valvar, or supravalvar level, and at distal locations. Increased afterload contributes to left-ventricular hypertrophy and may also affect pulmonary venous pressure and lead to pulmonary edema [51]. Aortic valve malformation is relatively common in the general population, with a bicuspid aortic valve (BAV) being present in approximately 1% of cases, and aortic valve stenosis in 3-8% of the CHD population [51]. Valvar lesions may be progressive in nature and symptoms may not occur in childhood and tend to appear later in adulthood [57]. Similar to right-sided obstructions, left-sided obstructions are also associated with numerous genetic syndromes, including Noonan's syndrome (subaortic stenosis), William's syndrome (supravalvar aortic stenosis), and Turner's syndrome (coarctation of the aorta).

Coarctation of the aorta (CoA) is a narrowing of the aorta and occurs in 5-10% of all CHD cases, and is associated with BAV in up to 50-85% of patients [51, 58]. Patients with CoA and BAV experience greater complications compared to patients with CoA without BAV, including ascending and descending aortic aneurysms, aortic dissection, and aortic rupture [59]. The presence of CoA with BAV may also place the left ventricle (LV) under additional stress and potentially result in heart failure [59]. Surgical procedures may help improve blood flow in the

aorta and reduce the aforementioned complications; however, even following surgery, patients may continue to experience complications and develop morbidities explained by poor hemodynamic and vascular biomechanics [59]. Patients with CoA may experience exercise-induced hypertension in the upper limb, but the exact cause of this remains unknown [60, 61]. Correria et al. reported that exercise-induced hypertension and abnormal blood pressure at rest and after exercise remain prevalent among adults with repaired CoA and should, therefore, not be considered a simple lesion that is fixed following repair [61].

Complex Congenital Heart Disease: Structural malformations of the heart may result in complex CHD that involve a combination of shunt lesions, obstructions, atrial or ventricular hypoplasia, and abnormal vessel connections [53]. The most common forms of complex CHD include transposition of the great arteries (TGA), TOF, single-ventricle physiology and hypolastic left heart syndrome.

Development of TGA is complex and may result in different atrioventricular connections between the morphological atria, ventricles, pulmonary artery, and aorta [62]. Complete TGA results from the inverted outflow tracts arising from the otherwise normally developed heart. In this form of TGA, the aorta arises from the RV and the pulmonary artery arises from the LV. The aorta is positioned at the RV and serves as the RV outflow tract while the pulmonary artery is positioned at the LV and becomes the LV outflow tract. This arrangement is unfavourable as deoxygenated blood is returned to the systemic circulation and the oxygenated blood remains in the pulmonary circulation and is not delivered to the body tissues (i.e., there are two systems, functioning in parallel to each other). Patients with complete TGA are cyanotic at birth and require intervention (i.e., balloon septostomy) to improve mixing and to help alleviate cyanosis by increasing the size of an ASD to allow systemic blood to enter the pulmonary circulation for

oxygenation and allow oxygenated blood from the pulmonary circuit to enter the systemic circulation [62]. Additionally, medication is given to maintain patency of the ductus arteriosus, providing another site for mixing. Another form of TGA is “congenitally corrected” (ccTGA) such that improper ventricular looping in development resulted in a reversed right and left ventricle that have the concordant inversion of the atrioventricular connections (pulmonary artery arises from the morphological left ventricle and the aorta arises from the morphological right ventricle). As a result, the systemic and pulmonary circulatory systems remain functional and in parallel. Patients with ccTGA may remain asymptomatic for much of their life, or develop failure of the morphologic, systemic right ventricle or the atrioventricular valve.

TOF consists of four distinct characteristics: over-riding aorta, right pulmonary valve obstruction, RV hypertrophy, and a VSD [63]. An infant with TOF has reduced blood oxygenation saturation due to the morphology that impeded pulmonary blood flow and left-to-right shunt across the VSD. Surgical intervention after birth redirects blood flow to the lungs with a complete TOF repair occurring between 3 to 6 months of age. Most children with repaired TOF continue to live normal lives but complications may arise in adulthood, primarily from pulmonary valve regurgitation resulting in right ventricular dilation. Therefore, it is important for patients with TOF to be followed closely at a specialized centre that can identify pulmonary valve regurgitation early and plan the necessary interventions to prevent RV dilation [53, 63].

Single-ventricle physiology includes functional single ventricle, absent or hypoplastic pumping chambers, absent or hypoplastic atrioventricular (AV) valves, or multiple complex VSDs that do not allow for a biventricular repair [53, 64]. The circulatory system runs in parallel (as opposed to series in the normal heart) and relies on one ventricle to pump blood out of the heart to the pulmonary and systemic circulation. The physiological challenge with univentricular

morphology is dependent on a number of factors, including outflow, inflow, and/or flow across the atrial septum, systemic and pulmonary venous return, pulmonary vascular resistance, and AV valve regurgitation [65]. Surgical advancements, namely the Fontan procedure, has allowed for increased survival of patients with single-ventricle physiology.

The Fontan procedure is the final palliative procedure in a series of multiple surgeries that allow for progressive adaptation of the heart and lungs to the changing cardiac anatomy. The ultimate goal of each stage is to place the systemic and pulmonary circulations in series, with a remaining single pumping (ventricle) chamber. The first stage aims to relieve systemic obstruction and provide sufficient pulmonary blood flow to permit oxygen delivery (and thus reduce cyanosis) and pulmonary arterial growth [66]. This shunt, commonly referred to as the Blalock-Taussig shunt, is a temporary solution until the patient displays sufficient pulmonary arterial growth with low pulmonary vascular resistance (PVR). The second stage usually occurs at 2-6 months and involves the connection of the superior vena cava to the proximal pulmonary artery. This procedure, known as the Glenn procedure, reduces the volume load on the ventricle and directs deoxygenated blood to the lungs and low-pressure pulmonary blood flow. The inferior vena cava carrying deoxygenated blood and the pulmonary veins carrying oxygenated blood both empty into the single ventricle, resulting in continued desaturation (80-85%). As the final stage palliative surgery, the Fontan procedure directs blood flow from the inferior vena cava to the pulmonary circulation. In some cases, a small fenestration (opening) is needed to maintain low PVR that serves to limit caval pressure and congestion, as well as increase systemic ventricle pre-load and cardiac output [66].

Regardless of the CHD complexity, it is important to emphasize the need for patients with CHD to receive specialized care. Patients with CHD may be unaware of their disease progression and

are also at increased risk of cardiovascular complications or developing other secondary co-morbidities, which necessitates long-term follow-up [67]. Therefore, it is important to consider cardiovascular disease risk factor management to help reduce the risk or severity of acquired heart disease secondary to CHD.

2.1.4 Prognosis

Prior to the 1950s, the highest mortality rate of children born with CHD occurred in the first week of life. Between 1979-1997, death due to an underlying CHD in the Unites States declined at approximately 2% per year [68]. The rate of death associated with or due to a CHD was greatest among infants and the greatest improvement in childhood mortality included those with complex CHD [68, 69]. While there has been a reduction in mortality over the years, CHD remains the leading cause of death in the first year of life excluding infectious disease [70]. Patients living with CHD are at increased risk of developing physical and developmental disabilities compared to otherwise healthy populations. Lummert et al. reported that adults with CHD (n=830; 18-79 years old) representing a variety of acyanotic and cyanotic CHD diagnoses also had non-cardiac co-morbidities from the fields of respiratory medicine, gastroenterology/hepatology, nephrology, endocrinology, gynecology/obstetrics, neurology/psychiatry, hematology, otorhinolaryngology, orthopaedics, and dermatology [71]. Developmental delays are commonly documented among the CHD population, including general growth, gross motor and coordination, feeding and swallowing, and speech and language problems [72-74]. Presence and progression of these delays should be identified, monitored, and treated in early childhood when possible to avoid more severe delays later in life. Establishing adequate eating and swallowing practices is particularly important to meet the metabolic demands that accompany CHD [72, 73].

Beyond early childhood, patients with CHD may face additional challenges and co-morbidities that become apparent later in life. Razzaghi et al. reported increased rates of asthma and ear infections in children with CHD compared to controls [75]. When considering specific neurodevelopmental outcomes, Razzaghi et al. reported that children with CHD in the age range of 2 to 17 years old had higher odds for autism spectrum disorder (odds ratio (OR)=4.6), attention deficit hyperactivity disorder/attention deficit disorder (OR=1.6), and intellectual disability (OR=9.1). Massin et al. documented that approximately 20% of patients with CHD had a significant co-morbidity (Table 2) and a co-morbidity was acquired in the first year of life in approximately 8% of cases [76]. Furthermore, there is an overall understanding that neurodevelopmental and psychosocial outcomes worsen with increasing CHD severity [77].

Table 2. Prevalence of important non-cardiac co-morbidities among 224 patients with CHD over a 10-year period (from Massin et al. 2007).

Subspecialty	**Co-morbidity**
Neurology (n=139; 13.1%)	Mental retardation (8.7%) Epilepsy (1.6%) Cerebral Palsy (1.2%) Acquired hemipeligia (0.5%) Ataxia (0.3%) Other conditions (1.8%)
Pulmonology (n=36; 3.4%)	Asthmas (2.4%) Bronchomalacia (0.2%) Bronchectasy (0.2%) Other conditions (0.7%)
Orthopaedics (n=26; 2.5%)	Scoliosis (1.1%) Hip dysplasia (0.3%) Pied bot (0.3%) Other conditions (0.9%)
Nephro-urology (n=19; 1.8%)	Enuresis (0.7%) Hydronephosis (0.2%) Renal dysplasia (0.2%) Vesicoureteral reflux (0.2%) Other conditions (0.6%)
Gastroenerology (n=14; 1.3%)	Esophageal atresia (5.7%) Anal atresia (0.3%) Tracho-esophogeal fistula (0.2%) Other conditions (0.3%)
Endocrinology (n=13; 1.2%)	Intrauterine growth restriction (0.5%) Diabetes Mellitus (0.4%) Hypothydroidia (0.2%) Other conditions (0.2%)
Other subspecialties (n=26; 2.5%)	Visual deficit (0.6%) Surdity (0.4%) Cancer (0.3%) Immunodeficiency (0.2%) Other conditions (1.5%)

The presence of significant co-morbidities further complicates the physical, psychosocial, and emotional well-being of patients with CHD [76, 77]. Work by Uzark et al. identified that children with cardiovascular disease, including CHD, report reduced quality of life compared to

healthy peers, and 20% of patients reported significantly impaired psychosocial quality of life [77]. Patients with more complex CHD and with residual lesions generally experience a lower quality of life and lower life expectancy [8].While there remain inconsistencies in the literature with respect to the quality of life reported by CHD patients, patients with multiple co-morbidities face even more challenges and require highly specialized and integrated care for optimal outcomes [76]. For a more detailed review of the psychosocial outcomes in CHD patients, see section 2.4.

A shift from post-operative survival towards long term physical and psychosocial functioning is warranted to match an aging CHD population that requires additional surveillance. The full scope of the necessary CHD services and interventions across the lifespan remains unknown. The long-term health outcomes of patients with CHD continue to evolve due to continuous improvements in diagnosis, surgery, and medical care.

Adult congenital heart disease (ACHD): The emerging ACHD population is relatively understudied with fewer resources focused on ACHD care compared to the larger acquired heart disease population. It is estimated that 1 million adults in the Unites States are living with some form of CHD and there are now more adults living with CHD than children [40, 78]. The chronic course of CHD also presents numerous social, educational, and occupational challenges for emerging adults [79, 80]. ACHD patients experience unique needs compared to adults with acquired heart disease (Table 3).

The ACHD population also experiences medical complications at a younger age, including arrhythmias, haematological problems, secondary pulmonary hypertension, heart failure, and death [36, 81]. Lui et al. described the management of cardiovascular risk factors among the

ACHD population and multifactorial progression from childhood to adulthood with respect to cardiovascular disease risk in the CHD population (Figure 5) [67].

Table 3. Comparison of CHD and coronary heart disease (from Ferguson et al. 2016).

	Moderate to Complex CHD	Coronary Artery Disease
Medical Differences		
Typical age at diagnosis	Fetus or newborn	Older adulthood
Timing of surgeries/interventions	Newborn, childhood, adolescence, adulthood	Adulthood
Average age of patients	20s and 30s	50s, 60s, and 70s
Average age at death	40s	70s
Recommended follow-up at specialized tertiary centre	Always	Occasionally
Psychological Differences		
Coping focus	Living with lifelong chronic condition	Recovery
Impact on psychosocial development	Low to high	n/a
Risk of parental over-protection	Increased	n/a
Transition from pediatric to adult care	Yes	No
Impact on education and career planning	Low to high	Low
Sex differences in mood/anxiety	Minimal	Women > men
General public's familiarity with disease	Lower	Higher

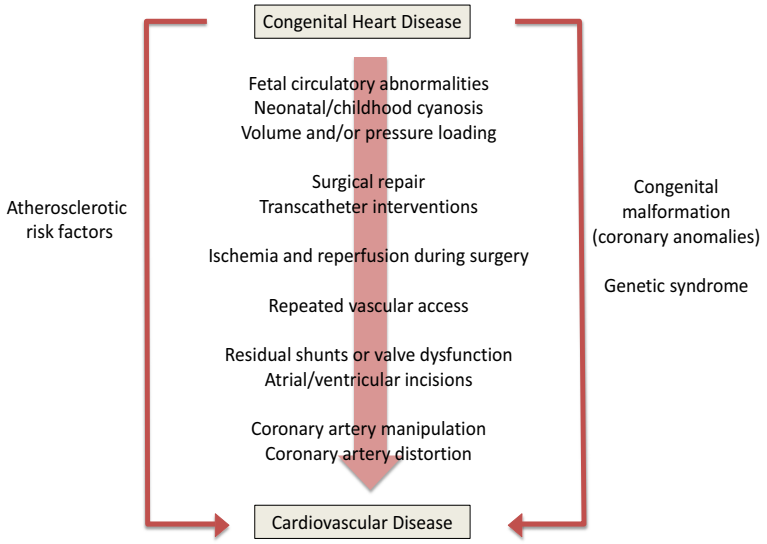

Figure 5. Overview of potential mechanism of cardiovascular disease progression in adults with CHD (from Lui et al. 2014).

Transition to ACHD care: The concept of transition to adult care should be viewed as a process rather than a single event such as transfer of care. Transition refers to the "purposeful and planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems" [82]. The transition process involves a change in the approach to medical care including learning new strategies for managing health, coping with medical decisions, and facing morbidities and the possibility of early mortality [83]. A number of key stakeholders should be involved in the transition process to contribute to a successful transition process from pediatric to adult cardiac care, including the patient, parent/guardian, pediatric-provider, and adult-provider [84]. The lack of appropriate follow-up for CHD patients and unsuccessful transition to adult care may result in delayed recognition of new or progressing cardiac problems [85, 86]. Young adults with CHD may experience greater cardiac complications later in adulthood [87-89]. Therefore, the continuity of medical care is the primary aim of transition programs [90].

While specialized ACHD programs are available to support this growing patient population, CHD patients are often lost to follow-up. Mackie et al. studied a population of CHD patients (n=643; 52% male) from 1983 to 2005 in Quebec, Canada, to determine the proportion of CHD patients who received cardiac care throughout childhood and early adulthood and identify risk factors for failure of follow-up [91]. Only 39% of ACHD patients (18-22 years old) were seen by a cardiologist, and 87% of ACHD patients were evaluated by a primary care physician. Furthermore, approximately 20% of adults with severe CHD failed to be seen by a cardiologist despite having access to and being in contact with the healthcare system [91]. Reid et al. reported that only 47% of 19-21 year old patients with CHD saw a cardiologist in a congenital cardiac clinic [92]. These authors identified several factors independently associated with higher risk of loss of follow-up during adolescence and emerging adulthood including male sex, having a

simple (shunt) CHD, no cardiac-related hospitalization before age six, fewer cardiologist visits, fewer non-cardiology physician visits, and last cardiology visit outside a university hospital setting. Gurvitz et al. reported that 42% of ACHD patients experience a 3-year gap in cardiac care, with patients 19-20 years old being the most common age group to experience this gap [83]. Despite existing transition programs, additional research is needed to evaluate interventions to improve transition and how these interventions impact the long-term management of ACHD patients.

2.2 Physical Activity and CHD

2.2.1 Population Physical Activity Trends

Consistent and strong evidence demonstrates the benefits of physical activity on physical (i.e., cardiovascular health) [93, 94] and psychosocial (i.e., self-esteem, anxiety, and depression) [95] health. Lifestyle risk factors including poor diet, smoking, stress, sleep disturbances, and sedentary lifestyles contribute to the progression of cardiovascular disease [96]. Physical activity is also an important component of the management of patients with chronic disease. Research has shown benefits of regular physical activity for cardiovascular, musculoskeletal, and psychological health for patients with chronic disease [97-99].

Evidence-based Canadian physical activity guidelines recommend a minimum of 60 minutes of daily MVPA for adolescents (≤17 years of age) and 150 minutes of weekly MVPA for adults (≥18 years of age) for optimal health [100]. The Canadian Society for Exercise Physiology describes moderate-intensity activities as those that make a person sweat a little and breathe harder, while vigorous-intensity activities make a person sweat and be ‘out of breath’ [100]. The

current physical activity guidelines were informed by a body of research that determined the optimal amount of physical activity to improve cardiorespiratory fitness as well as additional health outcomes [14]. These guidelines were based on evidence from a healthy population to accrue health benefits and may serve as a general guide for patients with a chronic disease. However, more detailed clinical exercise and physical activity recommendations are warranted to meet the unique needs of patients living with a chronic disease, including CHD.

Craig et al. reported 20-year trends in adult physical activity using data collected through the Canadian Fitness Survey (CFS) between 1980-2000 [101]. The CFS used adaptations from the Minnesota Leisure-Time Physical Activity Questionnaire, a self-report instrument that tracks physical activity in terms of time spent doing different activities. In this study, physical activity was calculated as a multiple of basal resting energy (1 metabolic equivalent, or 1 MET) and sufficient activity was defined as a minimum of 3MET-hours/day or 1260MET-minutes/week, which was equivalent to 60-minutes of walking every day for 12 months. Results from the CFS indicated an active population in Canada, particularly between 1980-1990 [102]. Bryan and Katzmarzyk reported that the proportion of adults (18-55 years) accumulating 30-60minutes of MVPA/day increased from 54% to 65% between 1994-1998 and 2001-2007, respectively [103]. Data for this study were derived from the National Population Health Surveys (1994-1998) and the Canadian Community Health Surveys (2001-2007), and were based primarily on a self-report instrument with a limited set of activities for patients to base their report of their level of activity over the last three months.

The U.S. National Health and Nutrition Examination Survey (NHANES) implemented a population physical activity evaluation with objective (accelerometer-based) assessments of physical activity [104]. Accelerometer data from NHANES between 2003-2004 indicated that

adults achieved the recommended 30 minutes of MVPA most days of the week. In contrast, children and youth did not meet the recommended daily level of activity, and the lowest prevalence of recommendation adherence was among adolescents [104]. Riddoch et al. also reported poor level of physical activity assessed by accelerometers in 11-year old children as part of the Avon Longitudinal Study of Parents and Children [105]. This study reported that children achieved 20 min/day of MVPA and only 2.5% of children met the activity recommendations (≥60min of MVPA/day). Allison et al. compared the decline in physical activity among adolescents between US and Ontario based on data collected by the 2001 Youth Risk Behavior Survey and the 2001 Ontario Student Drug Use Survey, respectively. Results indicated a steady decline in physical activity between ages 14 and 18 years [106].

The Canadian Health Measures Survey (CHMS) is a population-based study that included accelerometer-measured physical activity levels of Canadian children, adolescents, and adults between 2007-2009 [107, 108]. Results from the CHMS indicated that only 7% of Canadian children and youth and 15% of adults [107] met the current national recommendations. Canadian youth spent the majority of their time (62%) in sedentary pursuits. Adult physical activity as measured by the CHMS indicated that only 5% of adults engaged in MVPA on a regular basis (defined as 30min/day on 5 days/week) and 47% of adults accumulated 30 minutes of MVPA less than 1 day/week [107].

Despite the well-known benefits of regular physical activity, recent reports indicate that the majority of the population fails to accumulate sufficient physical activity. This is particularly evident in the above studies that employed objective, accelerometer-based physical activity assessments. Moreover, physical activity is a complex and multi-faceted behaviour with a number of barriers that may limit one`s ability to engage in physically activity pursuits.

Barriers to physical activity: There are significant barriers that limit physical activity participation. Barriers to physical activity participation may include environmental, biological, and psychological factors [109, 110]. An "ecological model" of behaviour change represents a dynamic interaction between these factors that changes over time (Figure 6).

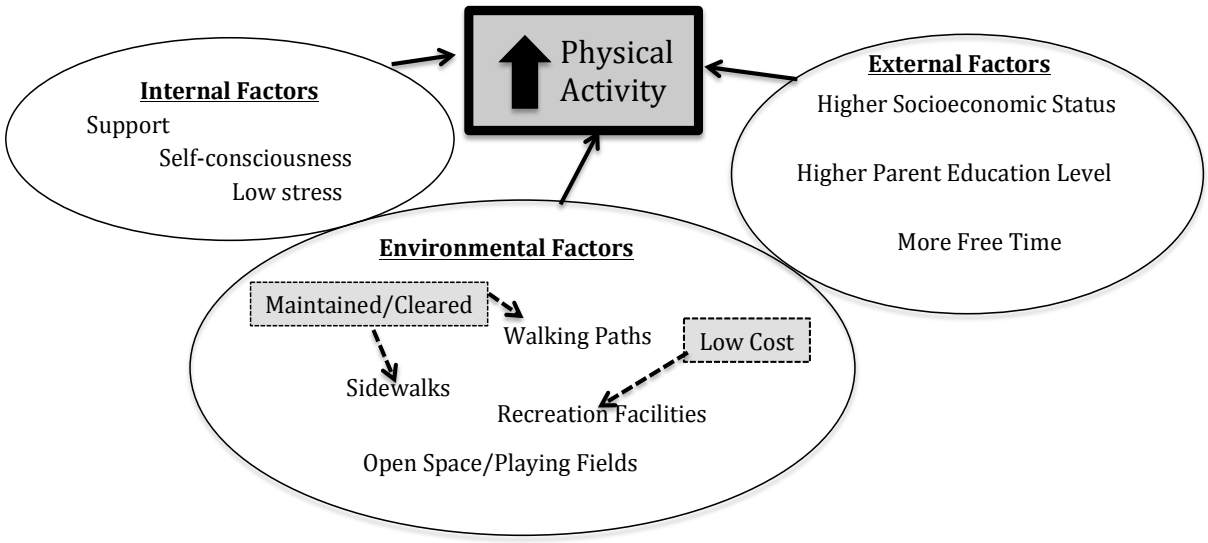

Figure 6. Ecological model including environmental, internal, and external factors that influence physical activity levels.

Environmental factors known to influence physical activity participation may include access to recreational facilities, parks, open space/playing fields, walking paths, sidewalk [111-113]. Tucker et al. evaluated the environmental factors associated with physical activity behaviour among youth (grade 7 and 8) [113]. Children with recreational facilities located in their neighbourhood were more active than children without recreational facilities (>180 min/day vs. <60 min/day, respectively). The Public Health Agency of Canada surveyed Canadians to understand the relationship between neighbourhoods and physical activity participation [114]. Survey results indicated that people participated in more leisure activities and active transport if the neighbourhood included low-cost recreational facilities and well-maintained sidewalks.

Individual perceptions regarding physical activity behaviour may also influence one's participation in physical activity. Allison et al. studied the perceived barriers to physical activity among adolescents [115]. A lack of time was identified as the main barrier, while psychosocial barriers (i.e., lack of support, feeling stressed, self-consciousness) were not perceived as major barriers. These results are consistent within the literature, such that external factors tend to be more influential on activity participation as perceived by the individual compared to internal factors [116, 117]. Likewise, Pouliou et al. reported that household socioeconomic status and maternal education were strongly associated with physical activity of children rather than environmental factors like accessibility to leisure space or safety perceptions [111].

2.2.2 Physical Activity in Patients with CHD

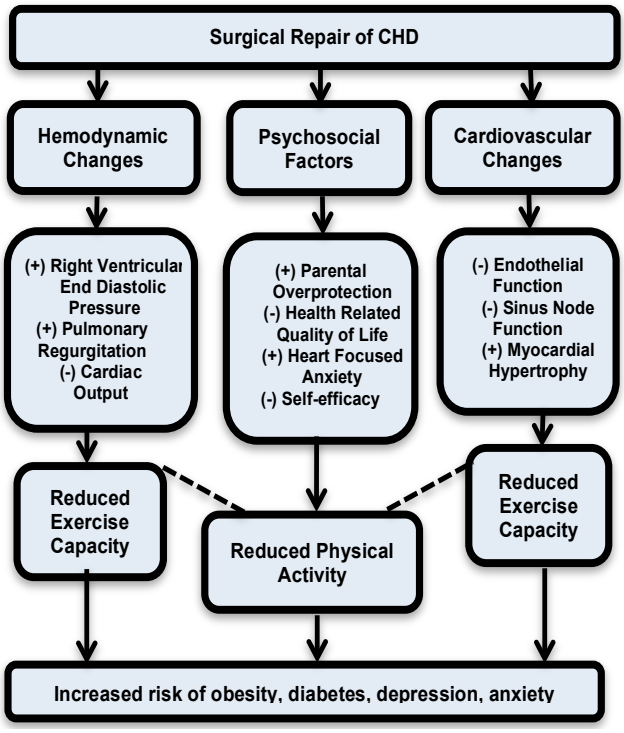

Figure 7. Long-term implications following CHD repair that challenge physical activity participation.

The hemodynamic, psychosocial, and cardiovascular changes that occur following surgical repair of CHD ultimately contribute to an increased risk of reduced activity participation and development of co-morbidities (Figure 7). Regular physical activity is important for the long-term health of children and adults with CHD and is beneficial for the CHD population to offset co-morbidities and improve both physical and psychosocial health outcomes.

Patients with CHD may experience substantial cardiovascular and cardiopulmonary limitations, particularly patients with moderate to severe CHD, including reduced cardiac output, impaired contractility, and disrupted blood flow due to endothelial dysfunction. Patients with CHD may also experience other non-cardiac physical limitations including neurologic deficits and orthopedic problems, which may prevent full participation in physical pursuits. Patients with CHD may also experience psychosocial barriers resulting in reduced activity participation, including reduced self-efficacy, health-related quality of life, and increased parental overprotection. Despite these limiting factors, most patients with CHD are recommended to participate in some form of physical activity [118].

Evidence supports regular physical activity to improve health outcomes in patients with CHD [7, 119, 120]. Despite these recommendations, patients with CHD display lower physical activity levels compared to healthy peers. Studies that included a heterogeneous sample in terms of CHD diagnoses and ages also reported insufficient activity (i.e., <60min of MVPA per day) [16, 25, 121, 122]. An inactive lifestyle contributes to the increased likelihood of patients to become obese [6]. Pinto et al. reported the prevalence of obesity and overweight among children 6-19 years old with heart disease, including acquired and congenital heart disease [6]. This study also investigated the impact of obesity on systolic blood pressure and documentation of weight counseling in referral notes to the primary care physician. This study found that patients with CHD have a high prevalence of overweight and obesity, with almost 25% of patients with moderate CHD and over 15% of patients with complex CHD considered obese or overweight (according to BMI percentile for age and sex). In addition, systolic blood pressure was higher among overweight and obese patients with acquired or congenital heart disease (median blood pressure: 75.7mmHg and 79.9mmHg, respectively) compared to patients of normal weight (62.1mmHg) ($p<0.001$). Documentation of weight counseling was present for only 13% of obese

patients and 9% of overweight patients with acquired or congenital heart disease (data were not presented as CHD compared to other heart disease). Therefore, the authors recommended that patients with CHD should be encouraged to participate in physical activity as a means to reduce the risk of overweight and obesity.

The persistency of sedentary behaviour into adulthood places these patients at increased risk of compounding their CHD with additional cardiovascular diseases [120, 123]. On the other hand, many studies report that children with CHD who are more physically active demonstrate improved physical functioning and reduced psychological problems including depression and anxiety [15, 124, 125]. Dulfer et al. conducted a systematic review to identify the association between exercise capacity, physical activity, and psychosocial functioning (including quality of life, emotional or behavioural problems, self-efficacy, and depressive symptoms) among patients with CHD [15]. From this review, two studies measured self-reported physical activity and self-efficacy among patients with mild to great CHD (10-18 years old), and found that increased physical activity was associated with increased self-efficacy [7, 126]. Another study by Muller et al. measured physical activity objectively with a tri-axial accelerometer among patients with single ventricle after total-cavopulmonary connection repair (n=57; 8-52 years old) and found that higher activity level was associated with higher self-reported mental health scale [127]. This study also reported that 72% (n=41) of patients with total cavopulmonary connection met the physical activity recommendations (≥60 minutes of activity, ≥5 days per week), and demonstrated a normal activity pattern. McCrindle et al. studied a separate cohort of patients with Fontan circulation (n=147; 7-18 years old), and found that higher objectively measured physical activity (uni-axial accelerometer) was associated with higher parent-reported general health [128]. Duppen et al. reported that patients with TOF or Fontan circulation (n=54; median age (IQR): 15.2 (12.6-17.6)) who participated in an exercise training intervention (12-week

program with 3 training sessions per week focused on aerobic exercise) reported improved cognitive functioning and motor functioning compared to control [17, 129].

Surveillance of physical activity among the CHD population has not been rigorously investigated and longitudinal data measuring physical activity are particularly scarce. Research on this topic is primarily based on cross-sectional studies with mixed methodologies in terms of measurement tools (i.e., subjective vs. objective assessments). McCrindle et al. reported that children with complex CHD (Fontan circulation) accumulated physical activity levels that were below the 50th percentile compared to healthy controls (Figure 8) [130]. The observed reduced physical activity levels of Fontan patients occurred despite adequate cardiac function and was associated with an increased number of surgeries prior to the Fontan procedure, hypoplastic left heart syndrome diagnosis, and dyspnea during exercise [130].

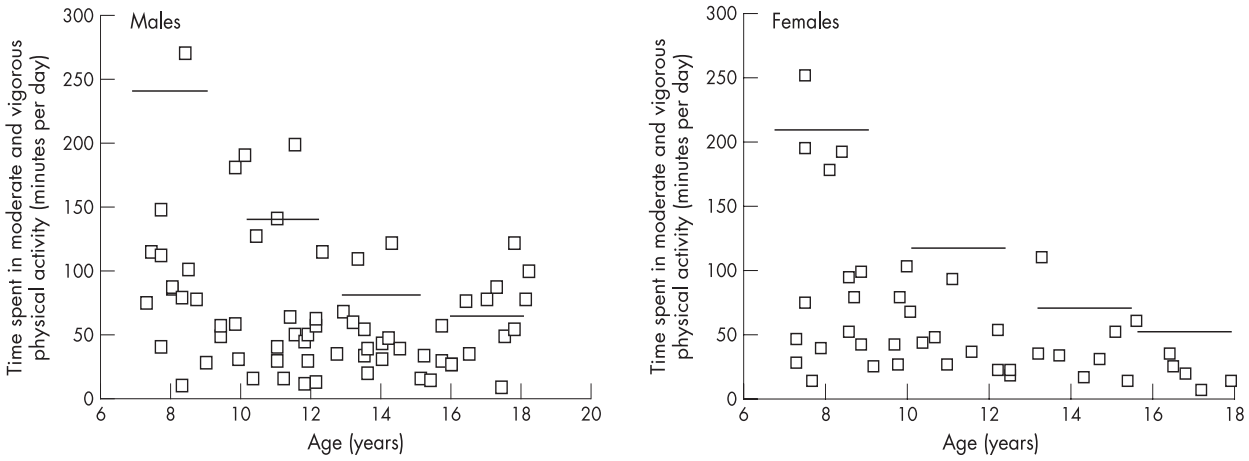

Figure 8. Physical activity (daily minutes of MVPA) for male (left) and female (right) Fontan patients by age. Reference lines represent the age-group 50th percentile for normal, healty children (from McCrindle et al. 2007).

Lunt et al. compared physical activity levels of adolescents with mild or severe CHD and healthy peers, and reported that 70% of adolescents with CHD participated in adequate or vigorous levels of physical activity [131]. While the overall aerobic activity recommendations were met in

both the CHD and healthy reference populations, patients with CHD performed less vigorous intensity activity compared to the healthy reference group. Patients with CHD also reported comparable self-efficacy towards physical activity and family support as the reference population. The difference between physical activity intensities could not be explained by differences in self-efficacy between CHD and healthy groups.

In another study, Duppen et al. reported physical activity levels and cardiopulmonary fitness of patients with TOF or Fontan circulation following exercise training intervention using a randomized controlled trial design [17]. Patients with Fontan and TOF spent comparable percentage of time in moderate-to-vigorous physical activities per day compared to healthy control subjects both at baseline (14.8±9.5% vs. 11.8±6.8%, respectively) and following an exercise training intervention (12.7±8.1 vs. 11.8±6.2%, respectively). This study contrasts other work that reported reduced physical activity levels compared to healthy reference populations [121, 122, 130].

Morrison et al. described similar activity levels among patients with CHD at baseline randomized to intervention or control groups (28.4±20.1 vs. 32.7±28.7 min MVPA/day, respectively) [16]. Patients who participated in a group session to explore motivation towards activity using motivational interviewing and visualization techniques improved their activity level post-intervention (28.4±20.1 vs. 57.2±32.2 min MVPA/day, respectively) while the activity level among control patients did not change (32.7±28.7 vs. 29.2±27.3, respectively). A high reported standard deviation in both groups indicated a large range in the data and potentially skewed data; however, the range or median values with confidence intervals rather than mean and standard deviation were not reported. This study supported the use of a behavioural change approach (MI) to improve activity levels among the CHD population.

Tikkanen et al. completed a systematic review of pediatric cardiac rehabilitation/exercise training programs for patients with CHD and identified only two studies (out of 15) that reported physical activity level as an outcome variable [8, 19, 25]. Rhodes et al. implemented a supervised exercise-training program (2 sessions/week, 60 minutes/session, including aerobic and light resistance training) with a heterogenous CHD patient population (n=33; 8-18 years old) [19]. Physical activity was assessed using a self-report instrument (8-item questionnaire asking about the nature and frequency of their exercise activities and limitations). Patients who completed the rehabilitation intervention reported more frequent exercise participation and perceived an improvement in their exercise capacity one year post-intervention [19]. Fredriksen et al. conducted a non-randomized control trial of exercise training whereby patients completed exercise training at a rehabilitation centre (2-week sport camp) or at home (5-months) [25]. Objectively measured physical activity (accelerometer) data was combined for analysis between the two groups, and results indicated an improvement in physical activity following the exercise training [25]. Similarly, in a more recent systematic review, Duppen et al. identified two studies out of 29 that looked at physical activity as an outcome measure, one in adults (Dua et al. 2010) and one in children (Fredriksen et al., included in the review by Tikkanen et al.) [25, 34, 132]. The study by Dua et al. reported an improvement in physical activity level among young adults with CHD following an exercise training intervention [132]

There is a lack of intervention studies that assess physical activity as an outcome variable, particularly as a primary outcome variable. Given the evidence that supports physical activity participation among the CHD population and contrasting findings with respect to intervention modality (i.e., prescriptive training vs. behavioural approaches), future exercise training and physical activity studies should include physical activity as an outcome measurement to identify treatment effects on health outcomes.

Barriers to physical activity participation in CHD patients: In addition to the barriers present in the general population, patients with CHD face unique challenges that may further limit their physical activity participation. For example, Longmuir and McCrindle reported that perceptions of physical activity restrictions vary significantly between cardiologists, parents, and health records, which negatively influence physical activity participation [133]. In addition, the same study identified large variation in the physical activity restrictions for patients with Fontan circulation as reported by the responsible cardiologist, emphasizing the need for clinical guidelines (beyond competitive sport) in this population [133]. Limited patient knowledge about physical activity participation may also be a result of lack of information, as identified by Lunt et al. who reported that less than 20% of patients with CHD received formal guidance around physical activity from their care provider [131]. Kendall et al. investigated the effectiveness of a questionnaire to help identify patients and families who required (or requested) additional advice about physical activity and exercise [4]. The screening tool was administered to patients and parents and included a series of five questions about activity, including one question that asked patients to identify the level of activity most appropriate for them given their heart condition (taken from the American Heart Association guidelines for recreational activity)[4]. Follow-up was completed in person in the cardiac clinic or by telephone with a cardiac physiotherapist. The main concern (65%; 50/77) reported was lack of knowledge about appropriate activities and those they should avoid [4]. Questionnaire administration and follow-up was easily implemented and provided a new method to address important exercise questions from patients and families.

Moola et al. reported physical activity perceptions of children and adolescents with CHD and identified social barriers to activity participation [134]. Young patients with CHD face challenges with disclosing their condition to others and the fear of marginalization from social activities. However, the dilemma in not disclosing the condition was met with potentially higher

expectations to perform to higher standards. Physical education class was one particular area that caused important barriers to physical activity participation, including the demanding physical education curriculum, insufficient time for rest, inappropriate responses from physical education teachers, and bullying [134].

In summary, patients with CHD experience a number of physical and psychosocial challenges that may influence physical activity participation. While some studies report lower physical activity levels in CHD patients (mostly complex CHD), others report comparable activity levels to healthy controls. Evidence suggests that physical activity is beneficial for patients with CHD and should be encouraged throughout their lives, including clinical, school, and home environments. Additional longitudinal research is needed to assess physical activity trends among the CHD population, identify factors associated with changes in physical activity, and assess long-term physical and psychosocial outcomes that may be related to physical activity.

2.2.3 Physical Activity Recommendations for CHD Patients

Published physical activity and exercise guidelines for patients with CHD are available from the American Heart Association and European Society of Cardiology [118, 135, 136]. Published recommendations have focused primarily on competitive sport participation and highlight specific sport restrictions [118, 137]. Takken et al. outlined physical activity recommendations for pediatric CHD patients, including important clinical recommendations for activity promotion [135]. Despite precautions for some patients, particularly those with complex CHD, the majority of patients are able to be physically active to the same level recommended for the healthy population. Physical activity guidelines support the long-standing evidence that regular physical

activity participation is important to accrue health benefits. The past and current paradigm for delivering physical activity recommendations has focused on restriction of certain activities with limited detail provided about activities that are safe and appropriate for the patient. The recommendations reported by Takken et al. may help shift this paradigm towards a model focused on the promotion of activity in the CHD population rather than restriction.

Longmuir et al. further emphasized the need to develop physical activity promotion strategies and programs that are integrated into the clinical care of patients with CHD [118]. These recommendations outlined counseling practices to guide health practitioners in promoting physical activity among their patients. The healthcare practitioner is encouraged to engage the patient and family in a conversation about physical activity to learn about three important aspects that may dictate referral to a physical activity/exercise specialist:

1. Identify the patient's readiness and/or ability to change their physical activity behaviour.
2. Identify the specific physical activity dimensions important to the patient, including mobility, object manipulation, cognitive function, behaviour or social skills, communication and perception, and fitness.
3. Identify important clinical considerations that may influence the patient's physical activity.

The above recommendations are meant to guide the clinical management of pediatric patients with CHD. Improved survival of CHD patients has also lead to recommendations for the clinical management of ACHD patients with regards to physical activity and exercise recommendations [136]. There is a general lack of research on exercise and physical activity for ACHD patients due to the relatively new era of adults living with CHD. Overall, physical activity counseling should continue in the care of adults with CHD that is specific to patient's clinical presentation.

The European Society of Cardiology provided high-level recommendations for the most common forms of CHD in the adult population [136]. These ACHD physical activity guidelines are focused on exercise and sport participation recommendations. In most cases, patients with normal hemodynamics who are free from significant arrhythmia should be unrestricted regarding activity.

The effectiveness of physical activity and exercise interventions to improve health outcomes has been demonstrated for the CHD population [8, 34], with necessary precautions for some patients as described above. Despite this evidence, CHD-specific programs are typically limited to research settings, provide interventions over a short duration, and focus on changes in exercise capacity (refer to section 2.3.3 for more details regarding exercise interventions). The CHD population might benefit from a more structured program that encourages early childhood activity participation and fosters healthy, active lifestyle behaviours that continue into adolescence and adulthood.

2.3 Fitness of Patients with CHD

2.3.1 Cardiopulmonary exercise testing

Standardized exercise testing provides objective insights about one's exercise capacity and abnormal responses to exercise for both symptomatic and asymptomatic patients [138-140]. Poor cardiopulmonary fitness is also a risk factor for hospitalization and death in patients with CHD [141]. The main variables assessed during cardiopulmonary exercise testing (CPT), interpretation of findings and common indications in CHD patients have been previously reported [138]. The gold standard for assessing cardiac function and aerobic capacity is

ascertained by measuring the peak oxygen uptake (VO_2peak). Overall, patients with CHD typically have reduced exercise capacity. Paridon et al. reported that only 28% of patients with Fontan circulation achieved a VO_2peak value within normal range [142]. However, this study also highlighted that the range of values included some patients who achieved a VO_2peak above predicted (112% of predicted). Other studies showed a difference in aerobic capacity between patients with complete and incomplete repair of CHD [143]. Rosenblum et al. reported that patients with incomplete repair (i.e., presence of residual lesions identified by echocardiography) possessed significantly lower mean peak work rate, age-adjusted oxygen (O_2) pulse, and a higher ratio of the increase in ventilation to the increase in carbon dioxide output during exercise (VE/VCO_2 ratio), but similar VO_2peak compared to healthy controls [143]. Therefore, despite evidence from the literature that indicates a reduced exercise capacity among patients with CHD, current data should be interpreted cautiously when providing individualized recommendations.

2.3.2 Functional Fitness Testing

Physical fitness extends beyond CPT and variables collected from such testing. Physical fitness also includes body composition, muscle strength, muscle endurance, balance and flexibility [9]. Inclusion of fitness outcomes beyond maximal aerobic capacity provides a more comprehensive assessment of a patient's fitness level.

The physical fitness of Canadians was measured as part of the CHMS [144]. Results from this work indicated that children and adults display reduced fitness compared to earlier population-based fitness assessments. Klausen et al. described health related fitness of patients with complex CHD including cardiorespiratory fitness, muscle strength, body composition, and questions

regarding health behaviour [145]. Cluster analysis identified strongly interrelated variables that resulted in three clusters based on the characteristics of the health-related fitness outcomes. The three clusters (Robust, Moderately Robust, and Less Robust) included 158 adolescents (41% female) with various CHD diagnoses (33% CoA, 22% TGA, 13% TOF, and 32% other complex diagnosis). VO_2peak and muscle strength differed significantly between clusters, while BMI differed between clusters for girls only. Lifestyle behaviours differed between clusters for girls such that girls in the Robust cluster demonstrated active lifestyles while girls in the Less Robust cluster demonstrated more sedentary lifestyles. There were no differences in lifestyle behaviours between clusters for boys, except for their enjoyment of physical activity that was lower in the Less Robust cluster compared to the Robust cluster. Klausen et al. also highlighted that health-related fitness outcomes are attributed to challenges faced in adolescence, including physical growth, biological maturation, and behavioural development. In addition, this study addressed the importance of including physical activity recommendations in health promotion practices among adolescents, particularly girls who tend to show more significant declines in physical activity during adolescence. Allison et al. did not find a difference between male and female adolescents (14-18 years), and attributed the decline in physical activity observed in this group to the development of new interests and increased pressures that reduce the time available to be physically active [106]. Furthermore, some patients with CHD displayed strong outcomes in the Robust group that may in fact encourage extreme and potentially high-risk activities if their fitness is perceived as supra-optimal. Therefore, physical activity and exercise counseling should also focus on these patients with respect to symptoms arising from extreme sport participation [145].

Recent work by Longmuir et al. reported that children with Fontan circulation displayed body composition and strength similar to healthy peers [146]. This study highlighted the importance of

assessing multiple fitness parameters to identify potential areas for increased improvement and training. Longmuir et al. used fitness assessment protocols from the CHMS to measure muscular endurance, muscular strength, flexibility, and coordination [144]. This approach allowed for comparison to population fitness results with CHD patients. A more comprehensive and holistic assessment of the patients' physical fitness compared to aerobic capacity (CPT) alone or single-item assessment s (i.e., muscular strength) may provide useful information to understand various aspects of the patient's functional fitness. However, the systematic review by Duppen et al. of exercise training programs in CHD patients reported that only 5/29 exercise training programs assessed muscle strength and one study assessed body composition [34]. Therefore, future research should include additional functional fitness assessments to better characterize the CHD population.

Overall, the ability to provide specific exercise recommendations for patients relies on a comprehensive fitness assessment. However, few studies including CHD patients include a range of fitness assessments and focus primarily on the maximal aerobic capacity through a standard CPT. As a result, many exercise-training programs for patients rely on the maximal exercise measures to base exercise recommendations and neglect other aspects of the patient's fitness.

2.3.3 Exercise Training for Patients with CHD

Exercise training recommendations for patients with CHD generally focus on the aerobic component of exercise and competitive sport participation [9, 10, 135]. Exercise-training programs and interventions typically follow an adult-oriented model, likely due in part to the available literature on the topic. However, more age-appropriate interventions should be

explored to improve acceptability and adherence to meet patients' needs and expectations [11]. Many patients with CHD are unrestricted and able to participate in any form of exercise or physical activity and, therefore, should strive to meet the physical activity recommendations for the general population [11, 118]. However, some patients do have exercise precautions and are unable to rely on the general population recommendations. Therefore, personalized exercise recommendations should be included in the clinical management of patients with CHD.

Structured exercise training is safe and feasible for both child and adult patients with CHD [23, 25, 132, 147]. Fredriksen et al. applied an exercise training intervention among patients with CHD (10-16 years old) that included either a 2-week home-based training program (control group) or 5-month facility-based program (training group) [25]. Each program included various sports and activities aimed to improve balance, strength, stamina, coordination, and flexibility. Exercise training improved VO_2peak for patients in the training group (134.9 vs. 140.2 ml/kg-0.67/min; p=0.014) but not the control group (154.8 vs. 157.4 ml/kg-0.67/min; p=0.249). Exercise training also improved physical activity levels (15.0 vs. 15.1 log activity counts; p=0.028). Control patients also improved their physical activity levels but this was not statistically significant (14.8 vs. 15.0 log activity counts; p=0.053). This study was the first to objectively assess physical activity using an activity monitor before and after a training intervention among young patients with CHD. Study limitations included the difference between patients in each group at baseline, high dropout rate, small sample size, and heterogeneity of CHD diagnoses. Despite these limitations, Fredriksen et al. identified important considerations with respect to training program compliance and preference for rehabilitation centres over home-based activity programs.

Rhodes et al. demonstrated that a structured rehabilitation program for patients with serious CHD improved aerobic capacity [19]. Patients (n=16; 8-16 years old) participated in a supervised exercise session twice per week (one hour/session) for 12 weeks. Activities included games and light resistance exercise. Patients were recommended to engage in activity twice per week at home, but adherence was not monitored. Control patients did not receive the exercise training intervention, but did have similar CHD diagnoses as the intervention group. VO_2peak improved after the rehabilitation program in the intervention group (26.4±9.1 vs. 30.7±9.2 ml/kg/min; $p<0.05$). This improvement was sustained by patients six months post-intervention [19]. Furthermore, patients reported exercising more often and perceived their exercise capacity was improved compared to the previous year, whereas control patients did not. Intervention patients also scored higher on the Child Health Questionnaire (measure of health status) in the emotional, physical, and behavioural domains (albeit not statistically significant), while the control patients declined in these domains. This study supported the work by Fredriksen et al., such that an institutional, supervised exercise program can improve the fitness of patients with CHD. However, Rhodes et al. did not monitor the physical activity of patients post-rehabilitation or during the six-month follow-up.

Morrison et al. held an activity day that explored the patient's motivation to exercise and discussed ways to increase activity over a 6-month period follow-up period [16]. This self-implemented, unsupervised program provided patients with a written plan to follow over 6-months at their own volition. Control patients did not participate in the group activity day and received their usual care. Intervention patients improved their predicted VO_2max between baseline and post-intervention (35.0±7.4 vs. 37.4±8.8 ml/kg/min; $p=0.02$) while predicted VO_2max in control patients was unchanged (37.8±8.6 vs. 37.5±8.6 ml/kg/min; p-value not reported). Physical activity level (average MVPA per day) assessed with accelerometer increased

in the intervention group between baseline and post-intervention (28.4±20.1 vs. 57.2±32.2 min/day; $p<0.001$) but decreased slightly in control patients (32.7±28.7 vs. 29.2±27.3 min/day; p-value not reported). Although the authors discussed the benefits of exercise training to improve physical activity among the CHD population, the intervention activity days differed greatly from other exercise training studies that involved structured, supervised sessions at a central facility [23, 25, 148]. Despite the methodological differences, the work by Morrison et al. did employ a new approach using behavioural therapy to address exercise behaviours among patients.

Work by Longmuir et al. described home-based exercise and activity training among young children with CHD [29, 149]. This research investigated play-based, parent-led, and patient-oriented programs to improve physical and neurodevelopmental outcomes. Qualitative interviews with parents of children with TGA or Fontan circulation indicated that having a variety of activities that the child enjoyed and that could fit into the family's schedule were key factors for program adherence. Parents also appreciated the program structure due to increased time spent as a family in learning the activities, including siblings. Although play-based programs differ significantly compared to supervised, exercise-training sessions, this approach may be more appropriate for the pediatric CHD population while offering similar overall effects. Home-based programs may help improve access to exercise programs when attending centrally located and/or facility-based locations to participate in an exercise-training program may be challenging [149].

Duppen et al. conducted a systematic review of physical exercise training programs in children and young adults with CHD [34]. The authors described 29 articles (including those described above), in which the mean training intervention duration was 12 weeks (range: 6-52 weeks) with an average frequency of three sessions/week (range: one session/week – daily). There was a mix

of home-based (7/29), institution-based (10/29), and combined (12/29) intervention models. The underlying CHD diagnosis varied between study groups such that 10 studies included a single diagnosis (Fontan, TOF, or TGA), while the remaining 19 studies included multiple diagnoses. Cardiopulmonary exercise testing was conducted in 24 studies with a reported mean increase in VO_2peak of 2.6 ml/kg/min. Improvements in VO_2peak at ventilatory threshold without an improvement in VO_2peak was reported in two studies [20, 150]. A decrease in VO_2peak was reported in two studies [26, 151], which attributed this decrease to improvements in exercise economy (improved efficiency to transfer raw energy into mechanical energy). No change in VO_2peak or related measures was reported in three studies [152-154]. Overall, the studies included in this review showed a positive effect for exercise capacity, fitness, and physical activity. While some serious adverse events were reported (e.g., seizure, transient ischemic attack, ventricular arrhythmia), these events did not occur as a direct result of exercise training, and the authors highlighted the safety of exercise training for CHD patients. Thus, in this study, exercise training was deemed safe and effective for CHD patients, which is supported across the literature [34, 132, 147].

There remain substantial limitations with regards to established outcome measures to compare studies and provide benchmarking [11, 34]. A standardized approach for exercise-training research, including consistent outcomes, will help identify best practices for clinical practice. Duppen et al. included the following recommendations for future exercise-training interventions:

1. 9-weeks of supervised (in-person or with adequate monitoring tools) training.
2. Sufficient power to demonstrate effect of training.
3. Include well-defined, homogenous sample.
4. Randomization including a control group.

5. Study design should allow for measurements to determine changes in the following: cardiorespiratory fitness (i.e., cardiopulmonary exercise test or exercise capacity), physical activity levels, motor function, and changes in cardiac function (i.e., cardiac output, ejection fraction, stroke volume).

Duppen et al. also outlined that future studies should aim to address evidence gaps in the field, including proper outcome measures for CHD patients and the optimal frequency, intensity, type, and time/duration of each session and the overall exercise training program. Analysis should also aim to investigate diagnosis-specific training effects to better understand the acceptance and trainability of patients with different CHD diagnoses [34].

2.3.4 Physical Activity and Exercise Precautions

While full participation in physical activity should be encouraged in the majority of patients with CHD, some precautions are required in specific cases [118, 135, 136]. The focus of guidelines has been on sport participation, particularly competitive sports [137, 155], but more recent published guidelines include information regarding physical activity, recreational sport, and exercise training [118, 135]. Information about the dynamic (aerobic and anaerobic) and static (strength) components involved in different physical activities is important to better identify the type of activities appropriate for each specific CHD (Figure 9) [156]. Takken et al. published physical activity recommendations for the most common CHD conditions including specific clinical recommendations for various single lesions [135]. Their work also addressed the paucity of high level evidence based guidelines and recommendations, as the basis of the current data available in the literature are derived from small studies, expert opinion, and case studies.

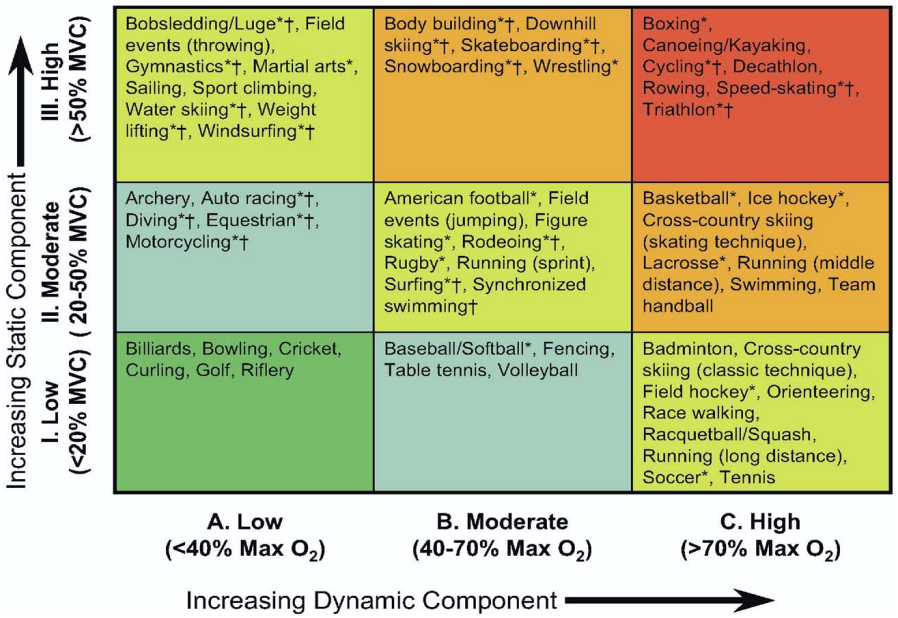

Figure 9. Classification of sports based on the relative static (percentage of maximal voluntary contraction; MVC) and dynamic (percentage of maximal oxygen uptake; $MaxO_2$) contributions to each sport (from Mitchell et al. 2005).

Longmuir et al. provided best practices with regards to physical activity promotion in the presence of activity precautions [118]. In this study, they suggest that the delivery of exercise and physical activity recommendations should provide as many details as possible to limit confusion and improve the confidence of patients and parents to participate in approved activities. Recommendations should be tailored to the individual clinical situation and take into account the unique needs of the patient, parents, and family-members. The conversation about physical activity and exercise should be bi-directional, allowing for questions from the patient and parents. Longmuir et al. also described the use of MI techniques that have proved to be a useful approach to understand the patient's motivation to change their behaviour, explore ambivalence towards behaviour change, and create action plans with the patient to assist adoption of recommendations and instill change.

2.4 Psychosocial Outcomes and CHD

Patients with CHD have less favourable psychosocial outcomes, including lower global quality of life (QoL), social adjustment, and self-efficacy [157, 158]. QoL is a multidimensional construct that includes a person's subjective perception of physical, emotional, satisfaction and cognitive functioning [159]. This construct can provide a crude assessment of an individual's perception of his/her current status that incorporates global interactions and experiences throughout life. According to the World Health Organization (WHO), QoL should be contextualized with respect to the individual's culture and value systems and in relation to their goals, expectations, standards and concerns [160]. QoL is a complex concept influenced by the person's physical health, psychological state, level of independence, social relationships, personal beliefs, and relationship to salient features of their environment [160]. Multiple definitions of QoL exist and a variety of ways to assess QoL are available, but no optimal strategy for its evaluation is recognized with a general consensus [161-163]. Regardless of the specific approach utilized, QoL assessments typically involve an operational definition including the individual's behaviour or level of functioning and/or the individual's perceived health status or wellbeing [162].

Systematic reviews on the topic of QoL and CHD have been completed, and the results are quite conflicting, presenting a wide range of outcomes and factors affecting QoL in this population [163]. Due to the discrepancies observed in published results and increased survival of CHD patients, investigations into underlying factors thought to influence QoL, such as parenting styles, social support and coping mechanisms, may help to more accurately describe the QoL of patients with CHD. For example, parenting a child with a chronic illness can pose challenges that

alter the effectiveness of parenting. Parents of children with CHD have reported heightened distress during the first years of the child's life compared to parents of healthy children [164]. Rassart et al. found that different parenting styles impact the perceived health of children with CHD [165]. More favourable health outcomes were reported by patients whose parents were more democratic, whereas children with authoritarian, overprotective, and psychologically controlling parents displayed an increased risk of poor perceived health over time. These results support the need for discussion regarding parenting skills and perceived patient health to be integrated into clinical prevention and intervention efforts. Healthcare professionals should aim to assess parenting behaviour, as this may influence the patient's perceived health and predict future health outcomes, including QoL [166].

Although QoL assessment is common within chronic disease populations, additional psychosocial assessments may provide important insights into the overall health of patients at the individual level. Self-efficacy is defined as one's confidence in being able to execute a specific action [167], and is another important dimension in the CHD population that may inform physical activity behaviour [126]. As described by Bandura, one's perceived self-efficacy influences choice of behaviour and activities and represents a main tenet of motivation and behaviour change [167]. In the CHD population, self-efficacy is generally reported as low and commonly investigated in the context of physical activity self-efficacy (i.e., the belief in one's ability to perform physical activity) [126]. Physical activity participation has been more strongly associated with self-efficacy rather than the severity of the disease among adolescents with CHD [7]. Therefore, self-efficacy should be assessed broadly across the CHD population, as patients with less-severe CHD may still have reduced confidence to be physically active and benefit from interventions to improve their self-efficacy.

Patients with CHD also face psychosocial challenges that may influence physical functioning. Kovacs et al. reported specific challenges experience by ACHD patients, including emotional, psychological, and social challenges [158]. Improving the psychosocial factors influencing the lives of CHD patients early in life may serve to optimize their care across the lifespan.

2.5 Physical Activity Assessment Techniques

There are many objective and subjective data collection techniques to assess physical activity levels. At the core of these assessments is the intention to collect data related to physical activity, which is defined as "any bodily movement produced by skeletal muscles that results in energy expenditure" [168]. Physical activity and exercise are terms often used interchangeably but a substantial difference exists between the two. Exercise is defined as a more structured, planned, and repetitive type of physical activity to improve or maintain physical fitness (i.e., cardiopulmonary, muscle strength, muscle endurance, flexibility) [168]. Regular physical activity assessment is important due to its well-established inverse relationship with chronic disease [169, 170]. Different techniques are used to assess physical activity across the scientific community, and the lack of a standard approach makes it difficult to compare outcomes between studies. Much of the current debate has focused on subjective versus objective techniques; however, each method has advantages and disadvantages and may be more suited in different types of research or among different populations. For example, the higher-level of precision of objective methods is often weighed against the low cost and easy implementation of most subjective methods [169, 171]. The following section will provide an overview of the different types of physical activity assessments commonly used in the literature.

2.5.1 Subjective physical activity assessments

Subjective physical activity assessments are the most common type, given the low-cost and limited burden on the patient [172-174]. Self-report is the most common method, and requires the patient to recall their activity and sometimes provide details regarding frequency, duration, type and intensity of executed activities [172]. Self-report instruments may include self-administered or interviewer-administered recall questionnaires, activity logs or diaries, and proxy reports [173]. Self-report instruments may also collect qualitative and/or quantitative information with respect to physical activity behaviour. Self-reports can be administered among diverse populations, be modified to meet the needs of specific populations or research studies, and assess multiple dimension of the behaviour, without affecting the behaviour in question [173]. Despite these benefits, data collected by self-report instruments may present an over-estimation of activity participation and low completion rates (particularly for daily logs/diaries) [175, 176]. Individual interpretation of questions (unless assistance is provided to help patients understand the question) may affect the quality of responses [169]. In addition to the aforementioned limitations, the use of subjective physical activity assessments in the pediatric populations raises other challenges, mainly related to the young age of the subjects. Difficulties in accurately recalling activity, reliably perception of activity intensity, and ability of properly documenting unplanned or sporadic activities are the most common amongst young children [169, 170, 177]. It is recommended children below 10 years of age not use self-report instruments, primarily due to low reliability at young ages [169]. Proxy reports (i.e., parent reports) of the child's activity participation are one method to collect the same information. However, proxy reports collect information from the adult's perception of the child's activity and may not represent the child's true activity behaviour [170].

A number of reviews have been completed to measure the reliability and validity of subjective, self-report tools to accurately assess physical activity. Focusing specifically on young people less than 19 years of age, Biddle et al. evaluated 20 assessments and received detailed evaluation by leading experts in the field of physical activity measurement [178]. From these 20 assessments, three were subsequently supported by experts for surveillance of activity among youth: the Physical Activity Questionnaire for Older Children and Adolescents (PAQ-C and PAC-A, respectively) [179], Youth Risk Behaviour Survey (YRBS) [180], and Teen Health Survey [181].

The PAQ-C (8-13 years old) and PAC-A (14-20 years old) are 9-item self-administered assessments that ask patients to recall the frequency (scale from "no", 1-2, 3-4, 5-6, or 7 times or more per week) of their participation for various activities during the last seven days. Patients are also asked about activity participation during physical education class, recess, lunchtime, after school, evenings, and the most recent weekend. The PAQ-C and PAQ-A assess activity based a minimum 10-minute bout of MVPA. The YRBS (10-21 years old) is a 5-item assessment that measures both moderate and vigorous physical activity by asking patients to recall activity participation over one year or seven days. This short assessment also includes a measure of sedentary behaviour. The Teen Health Survey (14-17 years old) is only 2-items and was adapted from the YBRS for possible use in the primary care setting. Application of this assessment among pre-teens has not been evaluated.

Each of the above assessments received expert support for use in surveillance of physical activity based on validity and reliability and ease-of-use while still providing a robust measure of activity. While self-report assessments may be preferred for large-scale use for the reasons previously described, a more detailed evaluation of physical activity may be necessary for non-

surveillance work. Therefore, alternative methods to collect physical activity data may be warranted that include objective assessment of activity participation.

2.5.2 Objective physical activity assessments

Technological advancements have improved objective assessments of physical activity, including increased precision, accuracy, and versatility, and use in different populations [176]. Double-labeled water is considered the "gold standard" to measure energy expenditure and is considered valid and reliable criterion [182]. The overarching theory behind double-labeled water is that "oxygen turnover in a body is dominated by the flow of water through the body as well as inspired oxygen and expired carbon dioxide" [183]. This technique requires an oral dose of radio-labeled isotope ($^{2}H_2{}^{18}O$) that results in body water being labeled with ^{2}H or ^{18}O. Energy expenditure is measured by the rate of elimination between ^{2}H (representing the elimination of water) and ^{18}O (representing elimination of water and CO_2). The difference between the rate of elimination is proportional to CO_2 production (i.e., [$^{18}O - {}^{2}H$] or [(water+CO_2) – (water)] = CO_2 production). This method, though highly precise, is impractical to administer on a large scale because it requires many resources to execute the necessary protocol, and is expensive. In addition, this technique estimates total energy expenditure over a minimum 3-days period and fails to deliver information about activity behaviour frequency, duration, or intensity with further resolution (daily or hourly) [182].

Objective measurement devices primarily include pedometers and accelerometers [169, 170, 176]. Pedometers measure movement using a mechanical spring that registers movement in the vertical direction. This measurement represents movement at the hip level during ambulation (walking) movements. The pedometer device sums each movement to provide a total number of steps taken. However, a pedometer device does not measure intensity, duration or frequency of

the activity, or differentiate types of activities [171, 184]. This lack of detailed information with respect to a given activity represents the main limitation for the use of pedometers for studies that aim to characterize the physical activity profile of a specific population. Compared to the doubly-labeled water technique, pedometers do offer a relatively inexpensive, easy-to-use, objective method to collect physical activity information on a larger scale [171].

The use of accelerometers in studies to measure physical activity has increased greatly and they are now the most common method among studies in youth [185]. Their use helps to objectively assess physical activity in populations where subjective measures had been previously relied upon. Accelerometers collect data regarding frequency, intensity, and duration of a given activity, unlike the pedometer or doubly-labeled water. A substantial reduction in the cost associated with accelerometers over the last decade has also led to its increased use in research, particularly in large-scale studies. These devices use a piezoelectric transducer to measure changes in acceleration (velocity/second, or m/s^2) in one or multiple planes of movement (i.e., x-, y-, z-axis) [171]. Movement is registered in all three planes and integrated as a summed "activity count". The activity counts are converted to pre-determined "cut-points" using a computer software program. The cut-points have been developed under controlled conditions in a laboratory setting that test the accelerometer against a range of activities and movement intensities [186, 187]. A number of accelerometer devices are available, including the Caltrac®, Computer and Science Application (CSA) monitor, and Actigraph devices. The Caltrac was one of the first accelerometer devices available and the most frequently studied [176]. This device is approximately the size of a pocket calculator and collects acceleration in a single (vertical) axis [188]. Data are presented as activity counts and estimates of energy expenditure can be derived if other data (height, weight, age, gender) are available. The Caltrac lacks an in-board storage computer but includes easy access to device controls by users; however, this access also

increases the risk of device tampering [171]. The CSA monitor is smaller and has internal computer memory storage that allow for future analysis. This device collects acceleration from a single-axis like the Caltrac, and is therefore unable to account for activities including motion in the other two dimensions. The ability to download activity data collected by the CSA device allows for data analysis using different cut-points, whereas the Caltrac cut-point thresholds must be determined prior to data collection. More recent tri-axial accelerometers measure acceleration in all three dimensions, include in-board memory storage, and remain small and light-weight. The Actigraph GT3X+ model is one example of a tri-axial accelerometer that stores activity data at a sampling rate between 30 to 100Hz [189]. These raw data can be downloaded to the corresponding ActiLife software for analysis using a specified epoch (e.g., activity counts per 10 seconds). The ActiLife software can be used to apply established accelerometer cut-points to the raw data to calculate activity intensity and estimates of energy expenditure. However, the analysis of data and use of accelerometer cut-points is highly debated due to substantial variation in data with different cut-points [171, 186, 189, 190]. Controversy remains regarding cut-points variability, and future research should address the significant differences between currently used cut-points in order to standardized the approach and allow for wide-spread comparison between studies. Corder et al. described the large variation between the use of cut-point values from four different conventions applied to a sample of 12- and 13-year olds to measure MVPA (Figure 10) [171]. As a result, uncertainty remains as to preferred and definitive cut-point values to measure physical activity. Furthermore, many of the cut-points defined for accelerometer-based studies were developed with healthy populations free of chronic disease. Stephens et al. reported new energy expenditure equations and accelerometer cut-points derived from a validation study among patients with CHD, cystic fibrosis, dermatomyositis, juvenile arthritis, inherited muscle

disease, and haemophilia [191]. These new disease-specific cut-points may represent the reference point for future research involving accelerometer-based physical activity assessments.

2.5.3 Future considerations for physical activity assessments

The use of subjective and objective methods to measure physical activity varies across research studies, each methodology with its own respective benefits and shortcomings. Population-based activity assessments may move towards an objective measure as the cost to implement new technologies is low and provides additional details. However, reliability and validity studies

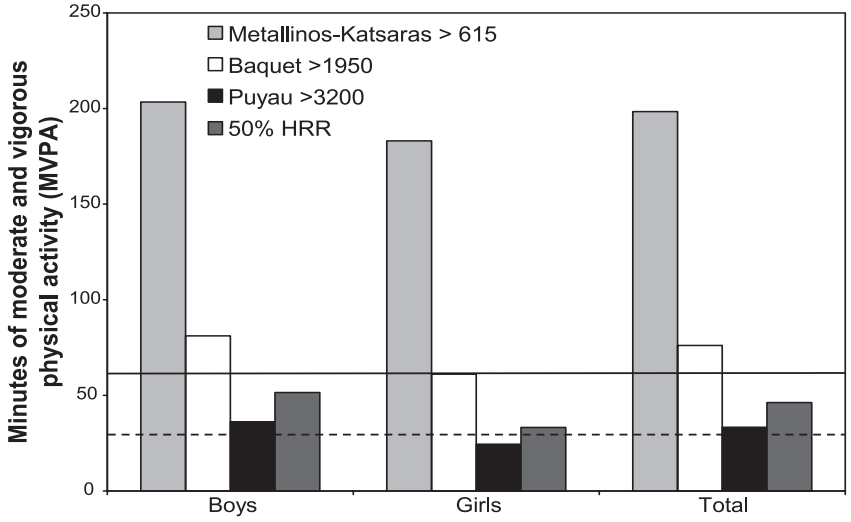

Figure 10. Variation in MVPA results based on different accelerometer cut-points applied to a sample of 12- and 13-year old children. Reference lines indicate activity recommendations for adults (30 min MVPA/day) and children (60 min MVPA/day (from Corder et al. 2008).

should be conducted in new populations where objective assessments are novel. Future projects should also aim to provide additional information about physical activity behaviour outcomes with respect to gender and ethnicity (i.e., subgroup analysis) for each methodology.

2.6 Physical Activity Interventions

Physical inactivity has reached epidemic proportions worldwide and is the fourth leading risk factor for non-communicable disease [192]. In order to address the acute and chronic health impacts associated with physical inactivity, health professionals have developed interventions aimed to increase activity participation. Physical activity interventions often involve the delivery of information (prescriptive), behavioural change strategies alone, or in combination [193, 194]. The following section provides an overview of each approach and, where applicable, the use of the intervention to help improve the physical activity of the CHD population.

2.6.1 Prescriptive Interventions

Physical activity interventions typically focus on providing information in the form of an exercise prescription. This approach often uses the FITT (frequency, intensity, type, and time) principle to guide activity recommendations [135, 195]. The FITT principle is widely recognized and used among the exercise physiology governing bodies including the American College of Sports Medicine and the Canadian Society of Exercise Physiology [196, 197] and supported by other organizations, researchers, and practicing clinicians [195, 198, 199]. The underlying aim is to provide a tailored exercise prescription that is relative to the individual based on their needs [200, 201]. The practitioner is also encouraged to document the exercise prescription on a pad of paper to resemble a typical prescription for medication [201]. The use of a written exercise prescription is thought to “elevate” the seriousness of the exercise prescription, looked as an order rather than simple recommendation [195]. Although this approach provides detailed documentation of the exercise prescription, it may also result in the medicalization of physical inactivity. Medicalization can be defined as “a process by which human problems come to be defined and treated as medical problems” [202]. The medicalization of physical inactivity may

hold negative consequences like pathologizing normal behaviour, disempowering individuals when subject to control by medical professionals or models of care, de-contextualizing the patient's experience, and depoliticising social problems [202]. This process may negatively frame conversations intended to promote activity, and further deter the patient from engaging in physical activity.

Prescriptive physical activity interventions are common among children and adolescents, particularly with regards to weight management and the prevention of obesity and type 2 diabetes [203-205]. van Sluijs et al. reviewed interventions among healthy children and adolescents that aimed to increase physical activity [204]. Interventions were categorized as educational, environmental, or combined. The majority of interventions (82%) for children were school-based interventions, as well as almost all of the interventions for adolescents. Approximately 50% of the interventions for children and 70% of interventions for adolescents were education-focused. Results indicated that educational interventions alone did not have an overall effect on physical activity participation. Results from combined interventions were inconclusive to report any effect on physical activity among children, while showing an overall positive effect for adolescents.

2.6.2 Behavioural Interventions

Many programs that aim to increase physical activity include a behaviour change component that is informed by social-cognitive theories [193]. Numerous behaviour change theories exist that have been applied in healthcare, including the Theory of Planned Behaviour, Protection Motivation Theory, Self-Determination Theory, Social Cognitive Theory, Health Belief Model, and Transtheoretical Model [206]. Physical activity interventions that are based on an identified theory are more efficacious than intervention that are otherwise atheoretical in their application

[193]. The use of a specific behaviour change theory may depend on factors like characteristics of the study populations (cognitive ability), delivery model (in-person, group, population, mail), and resources (counselor time, ability to change the environment). Central to many health behaviour change theories is a single construct known as self-efficacy.

Self-efficacy refers to one's confidence in their ability to perform a certain activity or task despite challenges or obstacles [167]. The use of self-efficacy as an outcome variable is appealing given that it is a modifiable factor drawing on personal experiences, persuasion, and vicarious experiences (learned from observing others), or modeling [207]. Self-efficacy is primarily an individual-based construct (rather than population based) and may be influenced by a number of both internal (personal mastery, physiological feedback) and external (vicarious experience, verbal persuasion) factors [208]. Intrinsic motivation is one factor that may influence one's ability to accept and change a given behaviour. This concept is heavily embedded in the Transtheoretical Model of Change (TTM) and can be assessed by asking questions that indicate one's readiness to change.

2.6.2.1 Transtheoretical Model of Change

The TTM was first described by Prochaska in 1997 for changing addiction behaviour but has since been used for a number of behaviours, including physical activity [209]. The TTM posits that individuals move through and between the following five stages (Figure 11): pre-contemplation (no intention to change), contemplation (intention to change in the next 6 months), preparation (making small changes), action (making large changes within the last 6 months), and maintenance (adopting change for the last 6 months or more) [210]. Rather than a linear progression, individuals often experience a cyclical pattern of progression-regression throughout these stages [211]. Progression through the five stages of change is mediated by three

main factors: self-efficacy for change, balancing advantages and disadvantages, and use of strategies/techniques to modify thoughts, feelings, and behaviour (processes of change) [210].

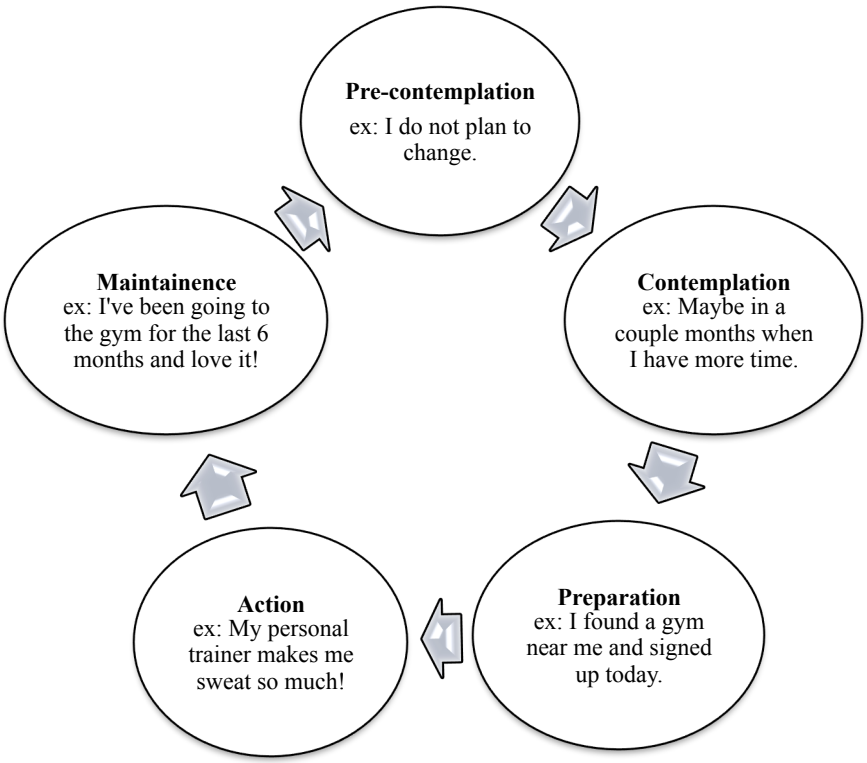

Figure 11. Diagram of the Stages of Change described in the Transtheoretical Model.

Marshall and Biddle conducted a meta-analysis of 71 published TTM-based interventions for physical activity and exercise, including cross-sectional (n=54), longitudinal (n=6), quasi-experimental (n=10), and randomized control trial (n=1) studies [210]. Results indicated that physical activity levels increased as patients moved to the next highest stage of change, with the greatest effect size between Preparation to Action stages (d=0.85; CI: 0.64-1.07) [210]. However, the authors also discussed some limitations in the use of stage-based assessments for physical activity, as the criteria for adequate physical activity differed across studies (i.e., 15 minute bouts x 3 days/week vs. 30 minutes x 5 days/week). Furthermore, when stages were used

to categorize activity levels (i.e., inactive=pre-contemplation and contemplation vs. active=action and maintenance), patients over-estimated their activity level. Results from the meta-analysis also indicated that patients experienced an increase in activity level when they progressed from pre-contemplation to contemplation, despite these two stages being considered "inactive". Therefore, the stage-based approach to classify activity levels may not be sensitive enough to represent the real changes in activity level between stages. It is recommended that for future research, criteria to describe physical activity levels using the stages of change should be standardized to help patients understand and conceptualize different activity intensities [210].

The TTM is a stand-alone behaviour change theory that can be applied across different interventions. Some studies and researchers have linked the TTM with Motivational Interviewing (MI); however, the two should not be used interchangeably nor should it be thought that MI is based on the theoretical components of the TTM [212]. Miller and Rollnick stated that "the TTM is intended to provide a comprehensive conceptual model of how and why changes occur, whereas MI is a specific clinical method to enhance personal motivation for change" [212]. In addition, MI is not a theory on its own and theoretical assumptions from MI are just now being identified and explored [212]. Nonetheless, MI continues to be used in the healthcare setting to change individual's behaviours.

2.6.2.2 Motivational Interviewing

First described by Rollnick and Miller in the early 1980s, MI was created to help people resolve ambivalence and make a positive change [213]. A more recent definition from Rollnick and Miller in 2009 defined MI as, "a collaborative, person-centred form of guiding to elicit and strengthen motivation for change." [212]. MI was developed in the substance abuse and addiction field and has since expanded to a variety of health behaviours, including weight loss,

physical activity, smoking cessation, and treatment/medication adherence [214]. The genesis of MI occurred as a means to change the substance abuse treatment paradigm by including a treatment intervention that enhanced one's motivation to change rather than blaming one for being unmotivated [212]. Therefore, the changes started to focus on the client's current level of readiness, which is likely where most of the connection between TTM and MI had been derived [215].

As a client-centered method, the interviewer guides the conversation towards a target behaviour while eliciting the client's desire, abilities, reasons, and need for changing that behaviour [213]. These areas form the basis of "change-talk" on which the MI counselor will try to identify and build upon throughout the conversation. Change talk is considered the client's movement towards change and can be identified with phrases like "I want to…", "I can…", "I will…", or "I plan to…" [216]. The counselor may reflect such change talk statements, which will let the client hear their own explanations and motivations for change and help them become more committed to changing their behaviour [213]. Focusing on "change talk" and directing the conversation towards change will help the patient to hear their own arguments for change (Figure 12) [213].

Figure 12. Change talk to facilitate behaviour change.

The general principles of MI include: expressing empathy (understanding and accepting the issues from the client's perspective), developing discrepancy (identifying differences between current behaviour and their goals), avoiding argumentation and confrontation (assume position that the client is responsible for their decision to change), rolling with resistance (engage the client in problem solving), and supporting self-efficacy and optimism (focus on past successes

and highlight the client's abilities with supportive statements) [215]. Throughout the conversation, the interviewer uses reflective listening and summarizes the client's thoughts while being empathetic and supportive. Reflective listening shows the client that the MI provider is listening and provides an opportunity for the client to elaborate, amplify, confirm, or correct points reflected by the provider [216]. This approach allows the practitioner to demonstrate the collaborative nature of the conversation and the client can be presented with a summary of their own self-motivational statements [217]. Miller and Rollnick emphasize that practitioners must possess the "spirit of MI" [212]. The "MI spirit" is based on three main elements, including collaboration between the practitioner and client, evocation of the client's reasons for change, and emphasizing the autonomy of the client to decide whether, when, and how to change [213, 217]. The presence of the MI spirit in sessions has been evaluated and adherence to this spirit is reliably measurable and is predictive of treatment outcomes [218-222].

Hettema et al. completed a meta-analysis of MI interventions, primarily from clinical trials, to assess MI efficacy to change a variety of health behaviours, including alcohol use, smoking, HIV/AIDs, drug abuse, treatment compliance, gambling, intimate relationships, water purification/safety, eating disorders, and diet and exercise [213]. Among the 72 studies included, 31 studies investigated the efficacy of MI combined with another type of intervention (adaptation of MI; AMI), while the remaining 41 studies investigated the efficacy of MI alone or MI combined with personal feedback (motivational enhancement therapy, the most common type of AMI). The duration of the MI intervention ranged from 15 minutes to 12 hours (mean=2.24 hours; SD=2.15) between studies. One of these studies by Rubak et al. reported that MI can be administered in brief encounters (15 minutes), while more than one session increases the likelihood of effect [214]. The majority of studies (74%) used a manual or specific training to administer the MI intervention; however, the use of a manual for MI interventions was found to

be unnecessary and, in fact, studies that did not use a manual demonstrated an effect size that was double than the one observed in studies with a manual [212, 213]. Manual-guided MI may instruct providers to meet certain milestones (i.e., creating a change plan) before the client has reached that stage. This mismatch between adhering to manual requirements with clients that are less ready for change opposes good MI practice [213].

A specific meta-analysis from Hettema et al. included a small number of studies (n=4) that looked at the use of MI to improve diet and exercise behaviour [223-226]. These studies included randomized controlled trials to evaluate the efficacy of MI to *(n=sample size)*: deliver education to patients with hyperlipidemia (n=85), improve fruit and vegetable consumption among African Americans (n=574), improve adherence to an obesity management program and glucose monitoring in older obese women (n=26), and deliver a lifestyle modification program across multiple behaviour domains (e.g., reduce alcohol consumption, dietary fat and salt intake, smoking cessation, and increasing physical activity) (n=166). The combined effect size of MI to improve diet and exercise behaviour across these four studies was 0.78 (95% CI: 0.41-1.16). Only one study completed a follow-up assessment following MI intervention and found an effect size of 0.14 (-0.16, 0.44). The effect of MI to improve diet and exercise long-term remains largely unknown and requires additional research. Significant variability in effect sizes within the same study population exists between studies, despite efforts to standardize training and treatment procedures [213, 214]. For example, the Project MATCH study on the treatment of alcohol use disorders included nine study sites, with significant variation of effect noted between sites and between therapists from the same site [227]. Furthermore, in their meta-analysis, Hettema et al. reported that the effect size of MI interventions is most pronounced early in the intervention period, while follow-up assessments showed a significant decrease in the effect size over time [213].

Gruhl and Van Leuven described studies that used MI with obese or overweight adolescents with the goal to improve activity and diet behaviours [216]. Overall, evidence suggests that MI is an effective approach to help adolescents manage and prevent obesity by changing their physical activity and eating behaviours. Despite the promising use of MI to change behaviour, Gruhl and Van Leuven highlight a shortage of evidence-based research on the efficacy of MI in children and adolescents with some encouraging results from limited studies in this cohort.

Schwartz et al. evaluated the feasibility of MI delivered by pediatricians and dietitians in the primary care setting [228]. Patients (n=91; 3 to 7 years) with a BMI between the 85th and 95th percentile, or of normal weight with a parent with a BMI >30 kg/m^2, were randomized to receive the control condition (usual care), minimal intervention (pediatrician only), or intensive intervention (pediatrician and dietitian). Results indicated that MI reduced the BMI percentile of children (n=91; 3 to 7 years old) in each group by 0.9 (control), 1.9 (minimal), and 2.6 (intensive) (non-significant differences between groups). Changes in specific behaviour patterns included a statistically significant decrease in snack intake in the minimal intervention group compared to control ($p=0.01$), and decrease in dining out in the intensive intervention group compared to the minimal intervention group ($p=0.04$). There was no significant difference in changes in behaviour for consumption of sweetened drinks, intake of fruits and vegetables, or television viewing within or between groups ($p>0.05$).

Saelens et al. administered a 20-week intervention and found no difference between the control group (provider-prescribed direct approach throughout the 20-weeks) versus the intervention group (self-directed, MI-approach after the 5th week) [229]. A reduction in BMI was reported in both groups, indicating that MI was equally effective compared to the direct-approach. Neumark-Sztainer et al. studied teenage girls who enrolled in an all-girls physical education class and

randomized them to receive 16 weeks of standard programming or a new program that included individual MI sessions, nutrition and social support, weekly lunch get-togethers, and parent outreach activities [230]. The intervention groups showed improvements in their stage of change for physical activity, physical activity goal setting behaviours, and self-efficacy to overcome barriers to physical activity.

Borrello et al. conducted a recent systematic review regarding MI in childhood obesity treatment for children 2-11 years old [231]. The authors concluded that MI is an acceptable method for treating obesity among children, but the efficacy of MI to improve BMI could not be made based on a lack of empirical evidence. Christison et al. described a nutrition and physical activity counseling tool paired with a MI intervention in the primary care during well-child visits to identify and address "obesigenic" behaviours [232]. This intervention was accepted by practitioners and families/patients, and showed positive outcomes related to meeting goals and sustained behaviour at 6-months follow-up.

A systematic review by Morton et al. included 22 MI studies with physical activity as an outcome measure of MI intervention [215]. Half of the studies reported a positive effect from the MI intervention. However, the majority of studies (n=17) used additional components combined with MI (i.e., exercise prescription, advice, exercise vouchers, walking groups, pedometer). Only five studies reported no or minimal use of additional components as part of the MI intervention, and only one of these studies showed a positive between-group effect [233]. In addition, two other studies that included MI without additional components demonstrated positive within-group changes in physical activity [234, 235].

All studies presented above raised methodological issues among the data on MI currently available in the literature, including non-randomization of patients, small sample size, absence of

MI training details, and lack of practitioner supervision or feedback [216, 231]. In addition, Morton et al. highlight that MI can mean different things to different researchers or practitioners, thus acting as a major barrier to the replication of MI-based studies within primary care contexts [215]. Future work should look to diversify the application of MI among the pediatric population to focus on physical activity promotion. In addition, more rigorous and standardized methodologies should be investigated to help inform MI interventions. Follow-up is also needed to determine the long-term effects of MI to improve behaviour change.

2.6.2.3 Motivational Interviewing for CHD patients

MI may be an effective approach to help patients with CHD to achieve a number of health behaviours associated with improved long-term health. A scientific statement from the American Heart Association described the role of behaviour change counseling in the primary care setting for patients with CHD and recommended assessing the patient's readiness to change and using MI to help build intrinsic motivation [118]. Despite these recommendations to use behaviour change counseling methods like MI among the CHD population to improve health behaviours, only one study referring to MI as part of an exercise training intervention is available [16].

Morrison et al. investigated the use of an exercise-based motivational session followed by a structured exercise-training program to increase physical activity and improve psychosocial well-being among adolescents (n=143; 60% male; mean age 15.6±2.27 years old) with CHD [16]. Patients enrolled in this study had major CHD (n=104; 73%) or minor CHD (n=39; 27%). Patients were randomized to the intervention (n=72) or control group (n=71), with no differences in CHD diagnosis between groups. Patients in the intervention group were invited to one of six activity days, which included a MI-based group session that explored motivation towards exercise using key MI elements (e.g., including rating the importance of exercise, confidence and

readiness to increase their activity level, and discussing the pros and cons of exercise) and visualization techniques. Control-group patients received their usual level of cardiac care. Following the one-day activity session, patients in the intervention group reported higher importance of exercise (7.8±2.3; $p<0.01$), confidence to change behaviour (7.6±2.1; $p<0.01$), and readiness to change behaviour (7.7±2.0; $p<0.01$). Patients in the intervention group also displayed improvements in physical activity (MVPA) at reassessment compared to their baseline assessment (28.4±20.1 vs. 57.2±32.2 minutes of MVPA per day, respectively; $p<0.001$) compared to the control group (32.7±28.7 vs. 29.2±27.3 minutes of MVPA per day; no p-value reported). The work by Morrison et al. showed encouraging results; however, replication of the study methodology among CHD patients from a different institution that may possess different resources or patient characteristics should be considered.

Despite the lack of research on MI-based interventions in children with CHD, MI has been used among adult cardiac patients to help improve outcomes in a cardiac rehabilitation setting [236-240]. Given that adult cardiac rehabilitation programs help reduce mortality by improving risk factors and lifestyle behaviour, integrating MI into a typical program structure may empower patients to make a positive change [239]. Adult cardiac rehabilitation programs face challenges with program attendance and adherence, often stemming from a lack of motivation from patients [238]. MI may be used to help improve attendance and retention of patients in cardiac rehabilitation [237].

McGrady et al. studied the effect of a MI and stress management intervention to improve cardiac rehabilitation program adherence among adults following myocardial infarction, coronary artery bypass graft surgery, stable angina, chronic heart failure, or other diagnosis (i.e., stent placement, valve replacement, aortic aneurism repair, atrial fibrillation, and heart transplant) [237]. The MI

intervention included four sessions that focused on the following topics: 1) identifying personal strengths to support self-efficacy; 2) elicit change talk and promote optimism for healthy lifestyles; 3) managing negative thoughts and recognizing past successes; and 4) create a strategic plan to accomplish rehab goals. This study demonstrated the MI helped patients attend 30 out of 36 sessions, while those who did not receive MI and dropped out of the program only completed 6 out of 36 sessions ($p<0.001$).

Patients of cardiac rehabilitation may also benefit from MI to improve program outcomes, namely exercise capacity, physical activity, and quality of life. Beckie and Beckstead conducted a randomized controlled trial among women (n=252; mean age 63±12 years old) in a cardiac rehabilitation program who received traditional programming or tailored programming that included MI [236]. Patients who received MI demonstrated a greater increase in QoL compared to those who receive traditional programming. This increased QoL was sustained 6-months post-intervention for patients in the MI group but not among patients in the traditional programming group. Brodie et al. investigated the use of MI to improve physical activity among patients (n=60; mean age 78±6 years) with chronic heart failure [239]. Patients received standard programming (provision of information and recommendations to increase physical activity), MI (home-based sessions on exploring ambivalence to physical activity), or a combined intervention (standard programming + MI). Patients who received MI alone or in combination with standard programming improved their energy expenditure (2.4kcal/kg/day and 2.3 kcal/kg/day, respectively). Patients who received standard programming were less active, as demonstrated by reduced energy expenditure post-intervention (0.1kcal/kg/day). In this study, the use of MI to improved physical activity levels, while standard programming alone did not facilitate physical activity participation. Interestingly, exercise capacity as measured by a 6-minute walk test

increased in all three groups (Group 1: Standard care + MI; Group 2: Standard care; Group 3: MI), with no differences between groups (Table 4).

Table 4. Six minute walk test in each group at baseline and post-intervention (from Brodie et al. 2005).

Group	Baseline	5 months	Significance p<
1	89.5 (73.4)	109.5 (71.4)	0.0001
2	157 (100)	181 (116.2)	0.0001
3	97.2 (85.4)	119.3 (95.7)	0.0001

With the exception of the work by Morrison et al. (2013), all studies presented above were conducted in adults with heart disease. Despite promising results, there is still a lack of data in the literature on the efficacy of MI to help patients with CHD improve their exercise behaviour. Additional research is needed to substantiate the evidence that MI is an effective method to help patients with CHD improve their exercise behaviour.

2.6.3 Alternative Interventions

The traditional and most common delivery method for physical activity interventions is in-person consultation with health professionals. A new era of healthcare service delivery based on new technology is emerging that includes mobile and wearable devices. This new approach, called mHealth, is defined by the National Institutes of Health as "the use of mobile and wireless devices to improve health outcomes, health care services, and health research" [241]. In 2014, there were over 6 billion mobile subscribers, accounting for 93% of the world's population [242]. Mobile devices, primarily smartphones, provide a means to access Internet services as well as mobile applications relied upon for mHealth interventions. Almost 20% of users reported downloading a mobile app that helps track or manage their health, with the most popular being those that help track physical activity [242].

The American Heart Association released a scientific statement that reviewed evidence in 69 mHealth studies to help reduce cardiovascular disease risk factors including weight management, physical activity, diabetes mellitus, smoking cessation, hypertension, and dyslipidemia [243]. Among these 69 mHealth studies, 14 were focused on physical activity, with nine of them being Internet-based and the others included short messaging services (SMS) text messaging (n=2), personal digital assistant or email (n=2) reminders, and pedometer tracking (n=1). In nine of the 14 studies, a significant increase in physical activity was reported in patients who received an mHealth intervention compared to a control group (n=5 internet interventions, n=2 SMS interventions, n=2 personal digital assistant/email interventions)[243]. The mean age of patients across the studies was 49.9±7.8years old and all studies occurred between 2005-2014. Each of these studies evaluated physical activity differently, including increases in step counts (pedometers), and moderate intensity activity, moderate-to-vigorous intensity activity, or weekly physical activity (accelerometry or self-report questionnaire). The overall message derived from these studies is encouraging and supports the use of mHealth technology to help patients improve cardiovascular risk. However, additional research is needed that includes a standardized definition of physical activity outcome measures that are investigated in randomized controlled trials. Additionally, as outlined in the American Heart Association Scientific Statement, there remains a gap in this line of research with respect to pediatric-focused mHealth interventions compared to research conducted among adults [243].

Niksch highlighted that pediatric mHealth solutions are not much different than pharmaceuticals and medical device innovations, such that children are at risk of being underserved as seen in other fields of research [241]. The advances made in mHealth may help to serve the pediatric population with new interventions that were previously fragmented due to geographical challenges, limited institutional resources, family structure, and financial limitations [241].

mHealth with respect to cardiac patient care is a growing area as both preventative (primary and secondary) and treatment options. Martinez-Perez et al. reviewed cardiology-focused mobile apps and identified 710 relevant apps [244]. From these 710 apps, approximately 40% were intended for use by the general population while less than 10% were intended for specific patient populations. The results of this review also highlighted that there are no mobile applications for patients with CHD or a heart transplant.

Cardiac rehabilitation programs include a large exercise and physical activity component, which has recently been delivered with mHealth technology. Several barriers limiting access to hospital/institution-based cardiac rehabilitation programs have been reported, including transportation challenges, lack of interest, dislike of classes and hospitals, work or domestic commitments, rural residence, location and accessibility and parking [245, 246]. Fortunately, home-based models of program administration have been shown to be as effective as facility-based programs [43]. mHealth technology may provide a means to further support home-based programs and to help increase patient participation by reducing some of the identified barriers to program access [246].

Maddison et al. described the rationale and methods of the “heart exercise and remote technologies” (HEART) trial [247]. The HEART trial included an SMS text message intervention that provided behavioural strategies (goal setting, exercise scheduling, and overcoming barriers) to increase adherence [247]. Internet support was also available for patients to document and monitor their progress, access motivational and peer-support (role-modeling) videos, and other cardiac rehabilitation information. This study intervention was developed out of a formal research process informed by Pfaeffi et al., including the conceptualization of the intervention, formative research to inform the development, pre-testing the intervention content,

pilot study, pragmatic RCT, and qualitative research [248]. This process engaged patients who were referred to or currently participating in the cardiac rehabilitation program. The behavioural strategies were grounded in the self-efficacy theory of behaviour change. In a separate report of the HEART trial by Maddison et al., results indicated that patients who received the mHealth intervention increased their self-reported leisure time physical activity (320 vs. 383 minutes/week) and walking (450 vs. 512 minutes/week) [249]. Intervention patients also improved their self-efficacy to be active following the intervention period compared to control (78% vs. 72%; (0.2-12.2); p=0.04).

Walters et al. developed a cardiac rehabilitation care model that utilized a mobile phone as the primary communication medium to support patients through the program [250]. This six-week, home-based program aligned with the Australian cardiac rehabilitation program and included a target physical activity time of 30 minutes of moderate intensity activity most days of the week. The program underwent a separate investigation to determine the effect of the smartphone-based home service delivery of cardiac rehab [245]. Patients who received the mHealth intervention showed greater adherence to the program components (completion of the Active Australia Survey, pedometer-measured walking activity over seven days, and 6-minute walk test) compared to patients who received traditional programming (94% vs. 68% adherence, respectively; $p<0.05$). Completion of the cardiac rehabilitation was 33% higher in the mHealth group compared to traditional community-based cardiac rehab programming (80% vs. 47% completed, respectively; $p<0.05$). Additionally, both groups demonstrated improvements in dietary profiles, triglycerides, mental health, and exercise capacity. The mHealth group also showed slight improvements in waist circumference and weight, and significant improvements in health-related quality of life compared to traditional programming patients. In this study, the mHealth intervention not only demonstrated improvements in uptake and completion of the

home-based program but also provided a means to collect objective exercise measurements rather than previously relied upon self-report methods [245].

Wearable devices represent a new method of monitoring physical activity that could change health care programs by empowering patients to take control of their health [251]. Wearable devices typically have an accelerometer to measure the duration and intensity of activity, and some devices are paired with third-party software programs to provide real-time data analysis indicating the user's progress. Others may include more advanced technical capabilities, including heart rate monitoring, heart rhythm monitoring, and glucose monitoring. There is increasing interest in the type of data collected by third party applications to enable interoperability with existing health information systems [251]. However, empirical research in the area of wearable devices and the efficacy of these devices to improve health outcomes or change behaviour is limited [252].

Many clinicians and researchers are hesitant to support mHealth, and a number of published reports recommend caution for the clinical use of mHealth and wearable devices based on limited data available [253]. Burke et al. state that clinicians should not take the lack of research on the topic as a reason to ignore the potential utility and effectiveness of mHealth technology, but rather accept the challenge of producing evidence to guide the future development and implementation of mHealth tools in clinical practice [243]. Given the rapid changes in technology over a short timeframe, there is an urgent need to develop viable research methodologies to evaluate mHealth usability and efficacy outcomes. mHealth research requires an interdisciplinary approach that integrates psychological, clinical, technical, design, and regulatory perspectives [252, 253]. In addition, the development of privacy, confidentiality, and data management/security policies and processes should be investigated. Future mHealth

interventions should at least demonstrate adequate privacy policies integrated into their design, include a behaviour-change theoretical underpinnings, and content should be referenced to a credible source [253].

2.7 Cardiac Rehabilitation

2.7.1 History of Cardiac Rehabilitation

Cardiac rehabilitation (cardiac rehab) is defined by the World Health Organization as: *the sum of activities required to influence favourably the underlying cause of the disease, as well as to ensure the patient the best possible physical, mental and social conditions, so that they may, by their own efforts, preserve or resume when lost, as normal a place as possible in the life of the community."*[254]

The historical course of cardiac rehab was first documented in 1772 as a case report by Heberden who described the improvements of a patient's condition attributed to 30 minutes of manual labour in 1772 [255]. In this report, early cardiac care included strict mobility restriction and minimum six-months of bed rest, which resulted in severe deconditioning of patients following a cardiac event. This approach contributed to reduced functional capacity, increased length of stay, and increased morbidity and mortality. The first formal form of rehabilitation for cardiac patients included chair-based exercises in the 1940's [256]. Exercise as a rehabilitation strategy challenged the status quo of cardiac care in the 1950's while additional supporting evidence became available. Evidence showed that those with more active occupations (i.e., ticket sellers or post-men) compared to less active occupations (i.e., bus drivers or clerks) displayed reduced risk of coronary artery disease and experiencing fatal cardiac infarction identified the potential role for exercise as an intervention [257]. Reports that highlighted the negative impacts associated

with prolonged mobility restriction and immobilization began to emerge that further supported physical exercise and mobilization of patients to combat deconditioning [258].

Cardiac rehab was originally designed to help the growing population of adults who experienced a cardiac event to regain autonomy and participate in regular physical activity. The most common diagnoses referred to cardiac rehab programs include: acute myocardial infarction, stable angina pectoris, coronary artery bypass graft surgery, heart valve repair or replacement, percutaneous transluminal coronary angioplasty and heart transplantation or heart-lung transplantation [255]. The early focus of cardiac rehab programs was to improve the functional capacity of patients using exercise-training principles. This remains a primary component of cardiac rehab to inform the development of tailored exercise and physical activity prescriptions [259, 260]. Cardiac rehab programs now include well-established core components that aim to improve primary clinical outcomes (reduced all-cause and cardiac mortality, nonfatal re-infarction and reduced hospitalization rates) and changes in modifiable risk factors (total cholesterol, triglycerides and systolic blood pressure) [120, 260, 261]. Cardiac rehab has evolved to include multifaceted program components to improve the physical and psychosocial outcomes of cardiac patients, as supported by the American Heart Association and American Association of Cardiovascular and Pulmonary Rehabilitation [262].

2.7.2 Adult Cardiac Rehabilitation Model

Cardiac rehab has traditionally been hospital- or facility-based and the benefits of this program model have been widely reported and summarized in systematic reviews and meta-analyses [263-266]. Some of the reported benefits of cardiac rehabilitation include reduction in total mortality, cardiovascular mortality, reduced hospital admission, lower risk of re-infarction, improvements in functional capacity, and improvements in cardiovascular disease risk factors

(i.e., lipid levels, systolic blood pressure, self-reported smoking) [266-269]. Psychosocial benefits of cardiac rehab have also been reported, and include improved health-related quality of life (HRQL) and reduced depression symptoms [270, 271].

Cardiac rehab was first described in Canada in the 1970's as an exercise-based program for post-myocardial infarction patients [272]. The cardiac rehab program model has evolved to include the following core components: 1) systematic patient referral processes; 2) patient assessment; 3) health behaviour interventions and risk factor modification; 4) adaptations of program models to improve accessibility (especially for under-served populations); 5) development of self-management techniques based around individualized assessment, problem-solving, goal-setting and follow-up; 6) exercise training; 7) leisure-time activities; 8) outcomes assessment; 9) continuous quality improvement programs; and 10) professional development [264]. These core components are derived from the Canadian Guidelines for Cardiac Rehabilitation and Cardiovascular Disease Prevention, 3rd Edition, and align with other reports from the American Heart Association and American Association of Cardiovascular and Pulmonary Rehabilitation, and European Association for Cardiovascular Prevention and Rehabilitation (Table 5).

While the core components of cardiac rehabilitation programs are well established, various delivery models including hospital- and home-based programs exist. It was reported that 70% of cardiac rehabilitation programs in Canada are delivered in a hospital as a supervised, site-based program [273]. Cardiac rehab is typically administered in three phases: inpatient, ambulatory outpatient, and maintenance phases [274]. The first phase is often short, given the limited time the patient remains in hospital, thereby limiting the amount of time to provide patient education [255]. Within Phase II, typical programs consist of 2-3 sessions/week (while some report 1-7

Table 5. Basic program components and multi-disciplinary team to facilitate cardiac rehabilitation programs (adapted from King et al. 2012 and Dalal 2015).

PROGRAM COMPONENTS	
Patient assessment	Long-term management
Lifestyle and risk factor management: - physical activity counseling - exercise training - diet/nutritional counseling - smoking cessation	Cardioprotective therapies Psychosocial health Audit and evaluation
Medical risk factor management: - weight - blood pressure - lipid - diabetes	Health behaviour change and education
TEAM MEMBERS	
Physician (cardiologist, community cardiologist, physician, general practitioner with special interest) Physiotherapist Occupational Therapist	Psychologist Nurse Specialist Dietitian Exercise Specialist Clerical Administrator

sessions/week) with a median duration of 5-6 months [273, 275]. Phase II is when patients may receive structured exercise programs in addition to other complementary components, including consultations with physicians, nurses, dietitians, pharmacists, psychologists, social workers, or other allied health professionals. The administration of Phase III is generally a continuation of Phase II components aimed to facilitate life-long maintenance of physical fitness and risk factor reduction strategies [255].

Home-based delivery models may occur in combination with facility-based programs or be an independent program model. In Ontario, approximately 70% of cardiac rehab programs are home-based and 11% of cardiac rehab patients attend a home-based program [273]. Home-based models have been shown to be equally effective to improve clinical and health related quality of life [276]. Dalal et al. conducted a systematic review and meta-analysis of home- versus centre-

based cardiac rehab that included 12 studies [276]. Centre-based programs were defined as "a supervised group based program undertaken in a hospital or community setting, such as a sports centre", whereas home-based programming was defined as "a structured program, with clear objectives for the patients, including monitoring, follow-up visits, letters, telephone calls from staff, or at least self-monitoring diaries". Pooled results indicated there was no difference in short-term (3-12 months) improvements of exercise capacity between facility-based and home-based programs. Long-term (14-24 months) improvements in exercise capacity indicated a favourable outcome among the home-based exercise program compared to the facility-based programs (although non-significant). Reported improvements in diastolic blood pressure were seen in centre-based programs at 3-12 months follow-up, while there was no different between centre-based compared to home-based programs for changes in systolic blood pressure at 3-12 or 12-24 month follow-up. Changes in blood lipid profiles (total cholesterol, low density lipoprotein cholesterol, and triglycerides) showed no difference between program types, whereas an increased concentration of high-density lipoprotein cholesterol was reported in centre-based compared to home-based programs. Change in self-reported smoking behaviour and health-related quality of life showed no difference between centre- and home-based programs.

Cardiac Rehab Referral Limitations: Despite the reported benefits of cardiac rehab, referral to established cardiac rehab programs present a major limitation to uptake and utilization [255]. Among cardiac rehab programs in Canada, the United States, and United Kingdom, referral to cardiac rehab is approximately 30% [277]. Other reports indicate that almost 80% of eligible patients are not referred to cardiac rehab in the United States [278]. Low referral rates are partly due to a lack of knowledge about cardiac rehab program locations, lack of standardized referral processes, inconvenient referral systems, perception that programs are low quality, lack of discharge communication from previously-referred patients, long distance to cardiac rehab

program for the patient to travel, perceptions of low patient motivation, and lack of clarity regarding who in the health care team is responsible for referral [264].

Following referral, only 14-35% of patients after a myocardial infarction and 31% of patients after bypass surgery participated in a cardiac rehab program [41, 279]. In Ontario, the most recent report of cardiac rehab participation from 2004 indicated that 22% of eligible patients attended a cardiac rehabilitation program [280]. Furthermore, referral and uptake is even lower among some populations, including women, the elderly, and minorities [281-287].

The evidence to support facility-based programs continues to grow, and emerging evidence from home-based cardiac rehab programs is encouraging. Improving patient referral and patient uptake/completion rates is a necessary area of research to fill the existing gaps in the current cardiac rehab paradigm. Additional research is also needed to understand optimal home-based program models to help increase the reach of cardiac rehab programs and improve accessibility. The use of technology and mobile devices may also provide a new means to administer programs, as described in section 2.6.3.

2.7.3 Pediatric Cardiac Rehabilitation

Vaccaro et al. first recommended a pediatric cardiac rehab program aimed to improve the functional capacity of pediatric cardiac patients [13]. The main components were derived from existing adult-based recommendations, including clinical examination and exercise testing, and exercise training and prescription. The program structure was focused exclusively on exercise training and recommended a 12-week hospital-based program, including two sessions per week (one hour per session) that would progressively increase in exercise intensity and duration. It was

recommended that a physical education specialist (i.e., elementary physical education teacher) with the oversight of a pediatric cardiologist and expertise of an exercise physiologist work to develop and administer the intervention. This multi-disciplinary team was recommended to provide age-appropriate and enjoyable activities for patients that produced adequate cardiovascular stress.

Two recent reviews summarized pediatric cardiac rehab and pediatric physical exercise training studies in CHD patients [8, 34]. Tikkanen et al. completed a systematic review of pediatric cardiac rehab programs that included a structured exercise training component [8]. There were 16 studies included in this review, including individual randomized control trials or observational study with dramatic effect (n=8), case series, case-control, or historically controlled studies (n=7), and non-randomized controlled cohort and follow-up study (n=1). None of the studies included a randomized control trial design. Results supported short-term benefits of cardiac rehab programs with structured exercise training; however, long-term benefits are understudied and require additional research to fully understand any sustained effects of cardiac rehab in the CHD population. Tikkanen et al. recommended a pediatric cardiac rehab program model informed by the articles in the review and using information from existing programs developed for other pediatric chronic disease (cystic fibrosis, renal transplantation, obesity). The authors recommended a 12-week program with 2-3 sessions per week, each session lasting at least 40 minutes, and including aerobic, resistance, and flexibility training, and education and psychological interventions.

A systematic review by Duppen et al. included studies with any type of exercise intervention, not limited to a structured exercise training program (unlike the review by Tikkanen et al.) among both children and young adults with CHD [34]. From the 29 studies included, there were 31

articles reviewed (including two studies that reported on acute and long-term outcomes in separate articles). Studies in children contributed 19 articles (11 of these were included in the review by Tikkanen et al.), four studies in children and young adults (two of these included in the review by Tikkanen et al.), and six studies including only adults. The majority of studies (n=18) were cohort study design, followed by a cohort with control group design (n=7), RCT (n=3), and a case report (n=1). Interventions lasted 12-weeks on average, with 3 sessions/week, and ranged from 5-60 minutes per session. Program models included facility-based/supervised training sessions (n=10), home-based programs (n=7), or a combination of supervised and home-based models (n=12). Results across the studies indicated an overall positive effect of physical exercise training to improve VO_2peak, muscle strength, and physical activity. Duppen et al. supported physical exercise training for the CHD population and encouraged future research to evaluate a core set of outcome measures to aid in benchmarking between studies and present firm conclusions.

The current adult cardiac rehab model recommends many different components as described above (see section 2.7.2), including education and psychological services. Children with CHD experience psychosocial challenges, including reduced QoL and self-efficacy. Furthermore, childhood onset of mental health problems occur in 70% of cases and almost 30% of adults with CHD possess a mental health issue [288, 289]. Therefore, psychological and psychosocial outcomes among the pediatric CHD population should be included in programs to identify problems early in childhood and manage these conditions into adulthood. Only four of the studies reported by Tikkanen et al. incorporated education into the program, whereas psychological assessment and counseling were not reported in any studies [8]. Duppen et al. focused on exercise-training programs, where six studies that included QoL or other

psychological assessments showed significant improvements, while one study showed no improvement in QoL [34].

Exercise and physical activity have been shown to attenuate psychological distress, but requires further research among the CHD population. Amedro et al. reported a significant correlation between patient reported HRQL and VO_2peak ($r=0.27$, $p<0.001$), patient reported HRQL and anaerobic threshold ($r=0.22$; $p<0.01$), parent proxy-reports of patient HRQL and VO_2peak ($r=0.43$; $p<0.0001$), and parent proxy-reports of patient HRQL and anaerobic threshold ($r=0.31$; $p<0.0001$) [125]. This study also identified an inverse relationship between lower anaerobic threshold values and lower VO_2peak with increasing CHD severity. The authors suggested the need to explore HRQL in the context of a pediatric cardiac rehab program, as only four programs reviewed by Duppen et al. included a HRQL outcome measure.

A major limitation of reported pediatric cardiac rehab programs was the lack of long-term follow-up of patients, despite well-documented acute benefits of the programs. Only two studies showed that physical fitness can be sustained up to 5-years after the intervention [19, 29]. In addition, the lack of randomization, heterogeneity of patient diagnoses, and lack of agreement on outcome measures used between studies, prevents definitive conclusion regarding the efficacy of cardiac rehab programs for the CHD population [8].

The inclusion of CHD patients in cardiac rehab programs remains limited, and even the most recent reviews of cardiac rehab do not acknowledge CHD as a potential patient group that could benefit from cardiac rehab services [261, 275]. Physical activity interventions are safe, feasible, and beneficial in adolescents and adults with CHD [23, 118, 132, 290]. Existing evidence regarding physical activity interventions from the general population and chronic disease populations may inform the development of a CHD-specific program. Compelling evidence

supports cardiac rehab as a secondary prevention strategy to combat modifiable risk factors, many of which are prevalent in the CHD population. Furthermore, a shift towards understanding and promoting physical activity behaviour to improve psychosocial aspects of CHD patients has emerged.

Cardiac rehab may provide a viable model to achieve current CHD exercise recommendations in a structured, supervised program. The current literature including pediatric cardiac rehab includes adaptations of general exercise training and adult-based cardiac rehab program components. The ability to leverage existing cardiac rehab models for use with ACHD patients requires additional research to assess the system-level implications (i.e., referral, utilization, adherence, cost-effectiveness) as well as individual-level outcomes (i.e., patient preferences, risk reduction and clinical outcomes). However, the direct application of an adult-based model with pediatric patients with CHD, especially children and adolescents, may be inappropriate. The pediatric CHD population presents specific healthcare needs that require novel methods to administer short- and long-term care. Home-based models and the use of technology to facilitate the program may be a more suitable approach to meet the needs of pediatric CHD patients. Exercise interventions continue to highlight the importance of adequate physical functioning and remain a cornerstone of CHD care. Preliminary studies of cardiac rehab interventions in children and adolescents with CHD have shown improvements in maximal exercise capacity to near-normal levels and improvements in physical activity [29]. However, these programs focus on the exercise prescription aspect of rehab and often neglect behaviour change interventions.

2.8 Overall Findings and Gap

The management of CHD patients has shifted from a focus on post-operative survival towards improving the functional health status of patients. The role of exercise and physical activity in the clinical care of patients with CHD has evolved since the first reports by Vaccaro et al. in the 1970s, with a large body of literature to support exercise-training to improve exercise capacity [15, 34]. Meanwhile, exercise-training interventions have shown mixed effects on physical activity outcomes. This may be due to differences in how physical activity was measured between studies (subjective vs. objective assessments of physical activity), or the prescriptive nature of these interventions. There remains a lack of behaviour change strategies or methods integrated in exercise-training interventions to help CHD patients move towards positive change and improve their physical activity behaviour. Additional research is needed to explore how physical activity interventions can be enhanced in the CHD population.

Interventions focused on improvements in physical activity should be investigated across the lifespan of CHD patients to identify optimal, age-appropriate strategies. The following gaps were identified from the existing literature in order to describe the current state of physical activity research among the CHD population, in which this thesis research was conducted.

Firstly, there is a lack of longitudinal research among CHD patients that focuses on changes in physical activity behaviours over time. Most research has been cross-sectional with inconsistent methods that limit comparison between studies (i.e., subjective, self-report instruments versus objective, accelerometer-based assessments). Given that CHD is a chronic condition, and survival beyond childhood has increased significantly, it is important to describe long-term physical activity trends to help identify key areas or modifiable factors that could help improve physical activity participation as patients continue to grow and develop.

It is recognized that CHD is a complex condition that includes physical and psychosocial sequelae. The impact of the impaired cardiovascular physiology in CHD on numerous physical outcomes (e.g., exercise capacity, muscular strength, gross-motor development, physical activity) is widely reported in the literature and a primary outcome for many exercise-training interventions. The psychosocial consequences of CHD have also been reported, with mixed reports regarding reduced quality of life. These objective measures have been used to explain potential contributing factors to reduced physical activity participation in the CHD cohort. However, it is also important to explore the patient's perspective towards physical activity and how growing up with CHD could influence physical activity behaviours. Qualitative research may provide rich data to help contextualize how patients view physical activity and identify new areas for future research that build on patient experiences.

Last but not least, few interventions target improvements in physical activity in CHD patients, despite the well-known physical and psychological benefits that accompany regular physical activity participation. There is longstanding evidence that exercise capacity is an important prognostic indicator among CHD patients. Exercise-based interventions that aim to improve exercise capacity are most common to demonstrate physiological adaptations to exercise training. These interventions are typically conducted in a controlled, supervised environment using a prescriptive approach. Overall, exercise-training studies have shown modest improvements (+2.6ml/kg/min increase in peak oxygen consumption) in exercise capacity among the CHD patients [34]. Some studies have investigated physical activity as an outcome following exercise training; however, interventions aimed at improving physical activity behaviours among the CHD patient are lacking. The use of behavioural interventions in

particular may help facilitate physical activity participation and establish important strategies to sustain regular physical activity.

The following research aims to address these gaps and potentially identify new areas that will ultimately inform a program of research focused on investigating a comprehensive, pediatric cardiac rehabilitation program.

3 Summary of Thesis Research

3.1 Rationale for Thesis

The CHD population continues to grow at a rate of 5% annually and more children are surviving into adulthood. The focus of clinical care has shifted to include functional outcomes that will allow patients to live productive lives. Functional health status includes both physical and psychosocial components. The ability to be physically active is a main indicator of one's physical health. As a group, CHD patients are generally less active than healthy peers and demonstrate reduced physical functional health status. Previous research has mainly focused on children with CHD or adults with acquired heart disease. There is a need to understand the physical activity behaviour of adolescents and young adults to inform the best approach to improve the clinical management for long-term health of these patients.

3.2 Purpose of Thesis

Physical activity and exercise participation should be discussed, advised, and promoted throughout the clinical care of CHD patients. This thesis research identified the physical activity levels, behaviours and perceptions towards activity of adolescents and emerging adults with CHD. A behavioural intervention to improve physical activity was also developed and assessed. Ultimately, the results of this thesis may be used to inform the development of future programs and/or interventions that serve to improve the physical activity and health of CHD patients.

3.3 Aims of Thesis

1. To determine the longitudinal trends in physical activity levels among a cohort of Fontan patients and to explore medical and psychosocial factors associated with physical activity levels.

2. To describe the physical activity perceptions and behaviours of emerging adults with CHD.

3. To evaluate the feasibility and efficacy of an adapted Motivational Interviewing behavioural intervention to improve the physical activity levels of adolescents with CHD.

4 Physical activity among young adults with Fontan circulation

4.1 Study Contributions

Dr. Brian McCrindle and Adam McKillop conceptualized the design of this study as part of the Pediatric Heart Network Fontan Follow-up Study. Adam McKillop, with assistance from Elizabeth Niedra, Elizabeth Radojewski, and Patrica Walter, completed participant follow-up and recruitment. Adam McKillop, with assistance by Elizabeth Radojewski and Patrica Walter, completed study data collection. Adam McKillop, with assistance from Dr. Cedric Manlhiot, completed data analysis. Adam McKillop, with assistance from Dr. Cedric Manlhiot and Dr. Brian McCrindle, completed data interpretation. The primary supervisor for this study was Dr. Brian McCrindle.

4.2 Introduction

Normal cardiac development results in four functional chambers (two atria and two ventricles) separated into the pulmonary (right-heart) and systemic (left-heart) systems that work in series [291]. Single-ventricle physiology occurs when one ventricle (right or left) is under-developed or absent (Figure 13). This results in the pulmonary and systemic circulation functioning in parallel rather than series and mixing of oxygenated and de-oxygenated blood. The single-ventricle experiences chronic volume over-load, oxygen desaturation, and lacks the functional capacity to fully support systemic circulation [291, 292]. Although rare (2-3% of all CHD diagnoses), single-ventricle physiology is a life-threatening condition that requires multiple surgeries throughout infancy and early childhood.

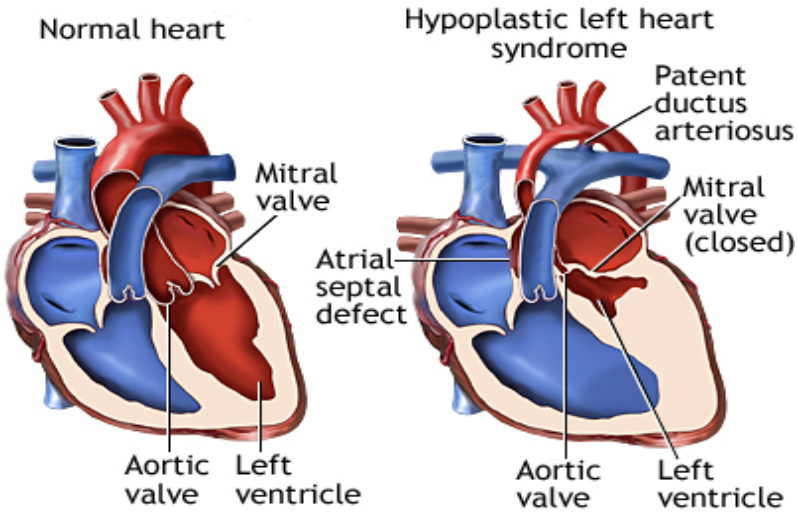

Figure 13. Normal heart compared to hypoplastic left heart syndrome.

Patients with single-ventricle physiology undergo a series of staged surgical palliation procedures throughout infancy including the Norwood, Sano, or Hybrid procedure (<3 months old), followed by the Glenn Procedure (3-9 months of age), with the final Fontan procedure completed around age four years. The Fontan procedure ultimately places the pulmonary and systemic circulatory systems in series with a single pumping (systemic) ventricle (Figure 14).

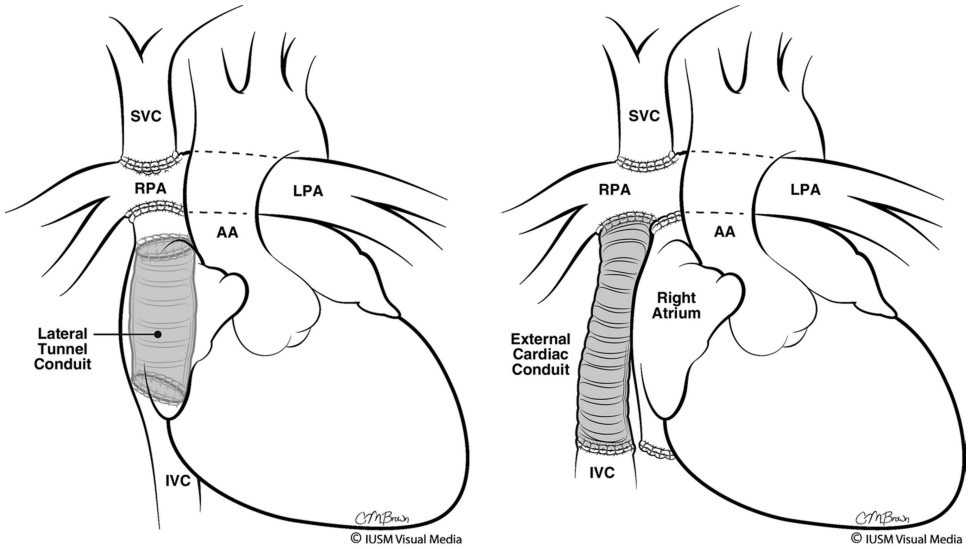

Figure 14. Diagram showing two different types of Fontan circulation: a heart with lateral tunnel conduit (left) and a heart with an external cardiac conduit (right). SVC, superior vena cava; RPA, right pulmonary artery; IVC, inferior vena cava; AA, aorta; LPA, left pulmonary artery (from Fredenburg 2011).

Patients with Fontan circulation experience a number of different physiological and hemodynamic constraints. Fontan patients experience chronic low cardiac output, largely as a result of simultaneous decreased ventricular preload and increased afterload [293]. Patients demonstrate a blunted heart rate response to exercise and inability to increase stroke volume, thus contributing to reduced cardiac output [66]. This hemodynamic limitation is further compounded by poor pulmonary vascular capacitance. The pulmonary vascular bed of Fontan patients is unable to recruit or distend pulmonary vasculature in response to increased cardiac demand, resulting in increased pulmonary vascular resistance with exercise [293].

Advancements in surgical and medical care have contributed to an improved survival for Fontan patients such that 65% of patients survive to 5 years of age and 55% of patients survive to 10 years of age [292]. The clinical management of the Fontan population has shifted from achieving post-operative survival towards improving the patient's functional status, including both physical and psychosocial well-being [130]. Physical activity is a contributor to physical health that may also indicate one's exercise capacity and self-efficacy. Children with Fontan circulation are less active than their healthy peers, and this reduced physical activity participation is a function of modifiable factors unrelated to cardiac status, such as perceived general health and parent/child perspectives on the child's physical abilities [130, 294]. Fontan patients also demonstrate reduced exercise capacity compared to healthy children of similar ages, but this appears to be unrelated to reduced physical activity [135, 295].

Previous cross-sectional research described the physical activity of children with Fontan circulation [130, 142, 294]. It is important to expand the current evidence with longitudinal research to yield more precise estimates of physical activity levels in Fontan patients and their relationship to various associated factors over time. Due to the positive effects on exercise

capacity and other aspects of physical health, achievement of recommended physical activity levels is important for continued health in Fontan survivors [135, 296, 297].

This study sought to determine long-term trends in physical activity levels after the Fontan procedure through longitudinal data collection, and to determine the relationship between physical activity levels and associated patient and clinical factors.

Study aims:

1. To determine changes in the physical activity level of Fontan patients between childhood and emerging adulthood.

2. To determine factors associated with physical activity level in emerging adulthood and factors associated with physical activity in emerging adulthood.

Hypothesis: The physical activity level in a cohort of Fontan patients would be reduced between childhood and emerging adulthood.

4.3 Methods

Pediatric Heart Network: The Pediatric Heart Network (PHN) consists of 7 pediatric cardiac centres in the United States and Canada, a data-coordinating centre at the New England Research Institutes, and the PHN Chair. The PHN performs clinical studies with funding from the National Heart, Lung, and Blood Institute of the National Institutes of Health. The PHN studied Fontan patients longitudinally throughout the Fontan 1, Fontan 2, and Fontan 3 studies, along with associated ancillary studies (Figure 15).

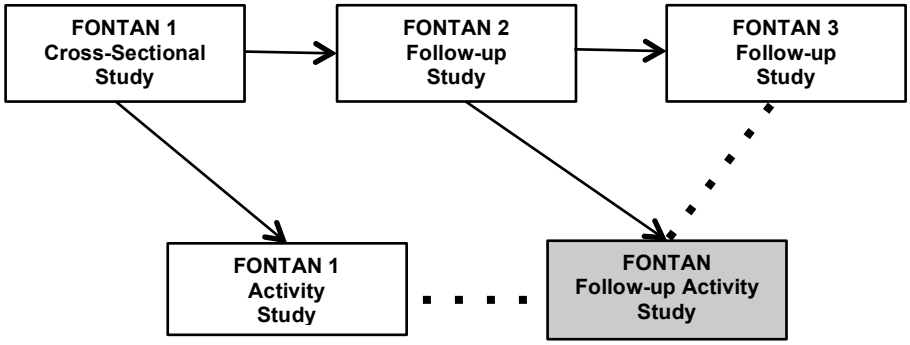

Figure 15. Overview of previous PHN studies and the Fontan Follow-up Activity Study. Data collected from Fontan 1, Fontan 2, and Fontan 3 were used in the Fontan Follow-up Activity Study.

Previous PHN Fontan Studies: The **Fontan 1 Cross Sectional Study** assessed the association between measures of functional status, ventricular function and exercise performance in pediatric Fontan patients [298]. Ancillary to this study was the **Fontan 1 Activity Study** that measured the activity levels of children and adolescents and explored relationships between physical activity, patient and medical characteristics, attitudes towards exercise, and exercise capacity and functional status [128]. The **Fontan 2 Follow-up Study** aimed to re-assess patients who participated in the Fontan 1 Cross-Sectional Study to measure vital status, functional health status, interim medical events, access to health care and self-reported availability and willingness to participate in future studies. The Fontan 2 Follow-up Study enrolled 85% of the Fontan 1 survivors and found 87% of these willing to participate in a future study. The **Fontan 3 Follow-up Study** collected data including health status questionnaires, maximal exercise testing, echocardiography and assessment of B-type natriuretic peptide (BNP) levels for comparison to data obtained during the previous Fontan 1 Study. Additional data included medical record review and questionnaires to assess socioeconomic status, family functioning, and access to health care. The Fontan 3 study also collected biological specimens for storage in a central repository for future genetic studies.

The Fontan Follow-up Activity Study: this study aimed to determine long-term trends in physical activity levels since the Fontan 1 Activity Study through longitudinal data collection using accelerometry, and to determine the relationship between physical activity levels and various associated factors. This study was a longitudinal observational cohort follow-up study that included data from the above PHN studies described above [130, 298]. This study included Fontan survivors who participated in the Fontan 2 Follow-up Study at the Hospital for Sick Children in Toronto, Ontario, Canada. The Fontan Follow-up Activity Study assessed long-term changes in physical activity levels after the Fontan procedure and examined associated factors with physical activity levels, including demographic information, medical history, and functional status.

Approach: Data in the Fontan Follow-up Activity Study included a combination of data collected from the Fontan Cross-Sectional Study, Fontan 1 Activity Study, Fontan 2 Follow-up Study, and Fontan 3 Follow-up Study. Longitudinal data were obtained by performing follow-up assessment of Fontan survivors followed at the Hospital for Sick Children in Toronto, Ontario, Canada. Clinical factors including patient characteristics, medical history, and functional status were evaluated to determine associations with physical activity levels.

Patients: Patients (n=104) from the Hospital for Sick Children who participated fully in the previous PHN cross-sectional research (Fontan 2 Follow-up Study) were approached for participation. From those approached for participation, 65 patients had completed the physical activity assessment during the Fontan 1 Activity Study. The inclusion and exclusion criteria were identical to those used to select subjects for the Fontan 2 Follow-up Study, as outlined below:

Inclusion criteria: All subjects enrolled were assessed for vital status and those currently alive with a Fontan circulation were approached for consent.

Exclusion criteria: Subjects who have died or had a cardiac transplant since Fontan 1.

Recruitment: Eligible patients were mailed an introductory letter explaining the premise of the study and research consent forms to review. An opt-out card with a prepaid envelope was also provided and patients were asked to return the card by mail to indicate their willingness to participate in the study or not, or to be contacted for additional information. If they did wish to participate, the consent form was reviewed over the telephone and verbal consent was obtained and documented. The patient was asked to sign and return the consent form that they had received in the return addressed envelope with postage.

Study Measures

Accelerometer Assessment of Physical Activity Levels: Accelerometry is a well-validated means of measuring habitual physical activity levels in children and young adults [299-301]. Accelerometry has been shown to be an objective and reliable measurement tool through comparisons with indirect calorimetry during free-living exercises, and is a valid means to obtain objective physical activity levels among adolescents and adults with CHD. Longitudinal assessment of physical activity levels included accelerometer data obtained previously from the Fontan 1 Activity Study and follow-up physical activity collected as part of this Fontan Follow-up Activity Study.

Physical activity level in the Fontan 1 Activity Study was measured using the ActiGraph GT1M uniaxial accelerometer, as described previously [128]. This model collected activity counts in the vertical direction (y-axis) only. The device was programmed to store physical activity using a

60-second epoch. Activity counts were converted to intensity level (METS) using age-appropriate cut-points [302]. Data had been previously analyzed as part of the Fontan 1 Activity Study and were made available as time (minutes per day) spent in moderate and vigorous intensity physical activity for each participant to include in the analysis for this Fontan Follow-up Activity Study.

Physical activity levels measured in the Fontan Follow-up Activity Study were measured using a more recent ActiGraph model, the tri-axial ActiGraph GT3X+ accelerometer (Manufacturing Technology, Fort Walton Beach, FL, USA). This model was not commercially available during the Fontan 1 Activity Study, but was a device upgrade used for the Fontan Follow-up Activity Study. This model collected activity counts in three directions (x, y, and z axes). The device was programmed for a 15-second sampling interval and stored the activity count per 60 seconds to memory at the end of each successive interval. Upon return of the accelerometers, stored activity counts for each 60-second epoch were downloaded for analysis (ActiLife 6.1). Daily activity counts were converted to intensity levels achieved per day **based on age-specific cut points [302], providing a final estimate of time spent doing physical activity at different intensity levels.**

At both time-points (Fontan 1 Activity Study and Fontan Follow-up Activity Study), participants were asked to wear the monitor during all waking hours securely at the mid-axial line with an adjustable elastic waist belt. Participants were asked to wear the device for seven days (including at least one weekend day), with three full days of wear-time (10 hours) defined as the acceptable minimum for inclusion in analysis. The accelerometers were to be removed during sleep, bathing, and any water activities. Logbooks were provided to track days the device was worn and reasons for device removal.

***Assessment of Patient and Clinical Factors*:** Factors included demographic (age, sex), medical history, exercise test data, and functional status (Child Health Questionnaire; CHQ) data collected in the Fontan 1 Cross-Sectional Study. The most recent cardiopulmonary exercise test results were abstracted from the participant's medical record to determine any changes in exercise capacity and exercise-related parameters over time. Exercise test data were also used to identify factors associated with physical activity over time. The CHQ offers a generic measure of functional health status and includes both child (CHQ-CF87) and parent (CHQ-PF50) versions. The CHQ included scaled domains of physical, behavioural, emotional, social, and family well-being, along with four categorical single item domains. A higher score on the CHQ indicated higher functional status.

Data Analysis: Descriptive analysis was used for data cleaning and description of the study participants and study results. Values were presented as frequencies and means with standard deviations where appropriate. Mean imputation was used to calculate for missing values. Univariable regression analysis was performed to determine factors associated with MVPA (dependent variable) in adulthood at follow-up, including patient characteristics, medical history, exercise test variables, and functional health status scores (independent variables). Factors associated with MVPA that had a significance of $p>0.20$ were included in a bootstrap-bagging algorithm (1000 resamples). Variables with high reliability (>50%) (defined as percent of resamples in which a given variable was selected) were then included in a multivariable linear regression model with backward selection of variables to obtain a final model. All statistical analyses were performed using SAS v9.3 (SAS statistical software, Cary NC).

4.4 Results

Study Participation (Figure 16): Eligible patients included 104 patients between 14-28 years who participated in the Fontan 2 Follow-up Study at the Hospital for Sick Children in Toronto, Ontario, Canada. Patients who declined participation for the activity study (n=13; 11%) returned an opt-out card by mail, while 51 patients (58%) did not return the opt-out card or were unable to be reached by phone to complete the consent process. A total of 40 patients consented to participate in the study but seven patients were lost to follow-up after consent was received and could not be contacted to coordinate delivery of the activity monitor. Therefore, 33 patients (31%) participated in this Fontan Activity Study.

Patient Characteristics: Patient characteristics and medical history are summarized in Table 6. The mean age of patients was 21±3 years (52% male). The mean age at Fontan surgery was 4.2±2.1 years (range 1.5-14 years) and the mean time since Fontan repair at the time of this study was 13.1±5.1 years.

Physical Activity Assessment: Physical activity data were available for 22 patients from the Fontan 1 Activity Study. Although accelerometers were provided to all 33 patients in the Fontan Follow-up Activity Study, activity data were excluded for analysis due to non-compliance (< 3 days total wear time) and device malfunction for 10 patients. Therefore, physical activity data was available for 23 patients in this Fontan Follow-up Activity Study. In addition, six patients with activity data from the Fontan 1 Activity Study did not have data available from the Fontan Follow-up Activity Study. Therefore, activity data from both the Fontan 1 Activity Study and this Fontan Follow-up Activity Study were available for 17 patients (Table 7). Patients had significant reductions in daily minutes of MVPA from the Fontan 1 Activity Study compared to

the Fontan Follow-up Activity Study (39±28 minutes of MVPA vs. 27±17 minutes of MVPA, respectively; $p<0.01$) (Figure 17).

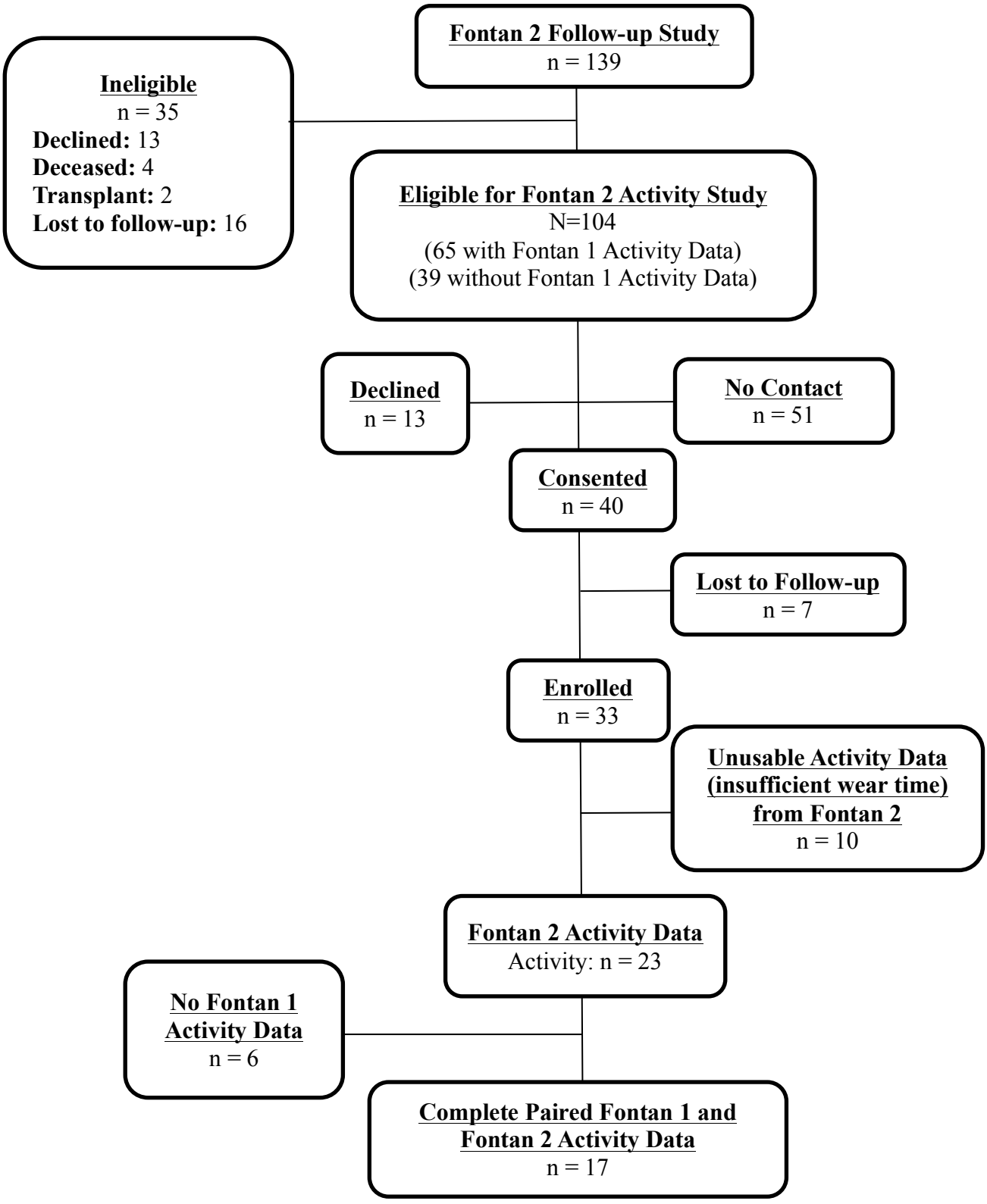

Figure 16. Fontan Activity Study patient recruitment diagram.

Table 6. Patient medical and surgical characteristics of the 33 patients enrolled in the Fontan Follow-up Activity Study from the PHN Fontan 1 Activity Study. (N=33)

Variable	Value
Sex (n; % male)	16 (48.5%)
Age at Fontan procedure (years as mean±SD)	4.2±2.9
Time since most recent Fontan (years as mean±SD)	16.9±3.3
# Cardiac surgical procedures pre-Fontan (mean±SD)	4.1±2.4
# Pre-Fontan single ventricle palliation surgeries (mean±SD)	2.8±1.8
# Pre-Fontan Single-ventricle surgeries (mean±SD)	0
# Pre-Fontan other cardiac surgeries (mean±SD)	1.3±1.1
# Cardiac catheterizations pre-Fontan (mean±SD)	0.9±1.9
Significant decreased systolic ventricular function (%)	8 (24)
History of arrhythmia (%)	10 (30)
History of stroke (%)	1 (3)
History of thrombosis (%)	8 (24)
Weight at Fontan (kg)	16.7±10
Fenestration performed (%)	22 (67)
Time on cardiopulmonary bypass (minutes as mean±SD)	126±55
Protein-losing enteropathy (%)	0

Table 7. Patient physical activity results completed during the PHN Fontan 1 Activity Study and the current Fontan 2 Ancillary Activity Study (mean±SD).	
Fontan 1 Activity Study (N=22*)	
Moderate Activity (min/day)	37.39±25.3
Vigorous Activity (min/day)	1.8±4.5
MVPA (min/day)	39.2±27.7
Fontan Follow-up Activity Study (N=23§)	
Valid wear days	6.2±1.1
Total Wear Time (hr/day)	11.8±0.7
Sedentary (min/day)	535.9±125.8
Light Activity (min/day)	101.3±45.5
Lifestyle Activity (min/day)	41.8±23.5
Moderate Activity (min/day)	24.8±15.3
Vigorous Activity (min/day)	2.7±3.9
MVPA (min/day)	27.5±19.2
*Accelerometer data from the Fontan 1 Activity assessment was available for 22 of the 33 patients enrolled in the current Fontan Follow-up Activity Study; §23 patients enrolled in the Fontan Follow-up Activity Study completed the physical activity assessment and had usable accelerometer data.	

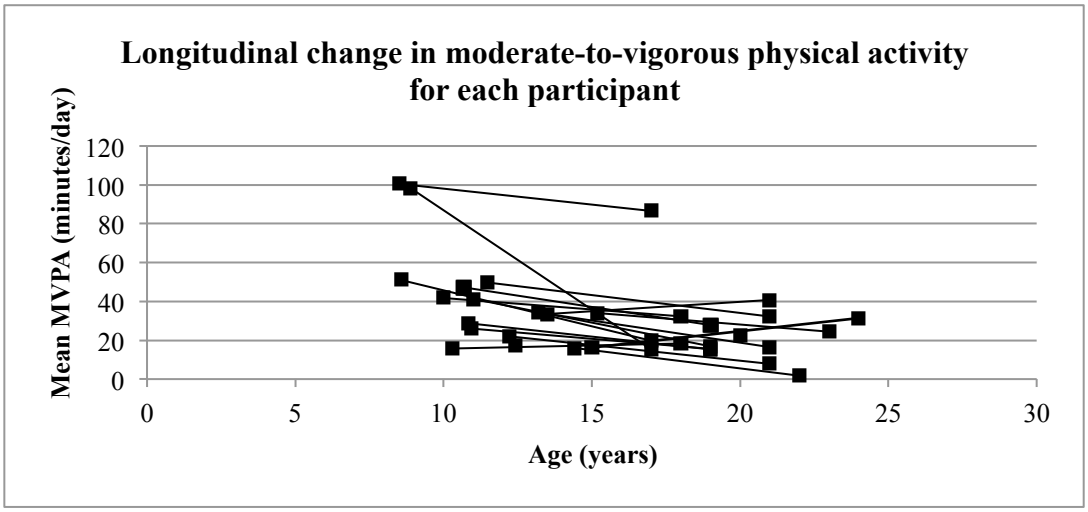

Figure 17. Change in physical activity (minutes MVPA per day) between childhood and adulthood for each patient (n=17 paired accelerometer data between Fontan 1 Activity Study and Fontan Follow-up Activity Study; n=7 males; 41%).

Cardiopulmonary exercise test: Peak oxygen consumption (VO_2peak) was unchanged for patients from Fontan 1 (n=30; 28.3±6.4 ml/kg/min; 62% predicted) to the most recent exercise test results (n=28; 27.8±8.5 ml/kg/min; 75% predicted). Respiratory quotient (RQ) results from the Fontan 1 Activity Study indicated that Fontan patients did not reach maximal effort (RQ ≤1.1) whereas more recent exercise test results from older Fontan patients indicated a maximal effort (RQ>1.1). Exercise test results from each study are summarized in Table 8.

Table 8. Patient exercise test results completed during the PHN Fontan 1 Study and the most recent cardiopulmonary exercise test obtained by medical record review with permission during the Fontan Follow-up Activity Study (mean±SD).

Variable	**Fontan 1 Activity Study** (N=30*)	**Fontan Follow-up Activity Study** (N=28§)
Age (years)	13.1±3.3	19.8±4.6
Height (cm)	150.0±17.4	169.2±9.0
Weight (kg)	47.4±19.7	71.9±16.7
Peak heart rate (beats/min)	165.3±25.5	165.2±22.0
Peak heart rate (% predicted)	89%	94%
VO_2peak (ml/kg/min)	28.3±6.4	27.8±8.5
VO_2peak (% predicted)	62%	75%
Respiratory Quotient	1.06±0.11	1.18±0.10

*Exercise test results from the Fontan 1 Cross-Sectional Study were available for 30 of the 33 patients enrolled in the current Fontan Follow-up Activity Study; § Recent exercise test results were available for 28 of the 33 patients enrolled in the Fontan Follow-up Activity Study.

Functional Status: Patients (n=33) and parents (n=33) completed the CHQ as part of the Fontan 1 Cross-Sectional Study to assess functional status. The distribution of scores for both parent- and patient-completed CHQ questionnaires is in Table 9.

Table 9. Functional health status (CHQ) score distribution for parents and patients collected from the Fontan Cross-sectional Study.		
CHQ Domain	**Parent-reported (n=33)**	**Child-reported (n=33)**
Scaled Domains		
Physical functioning	87.9±14.9	88.6±12.6
Role/Social Limits – Physical	88.9±23.8	91.4±14.5
Role/Social Limits – Emotional	83.8±29.4	89.3±23.2
Role/Social Limits – Behavioural	-	91.4±15.8
Bodily Pain	89.1±16.1	85.0±17.7
Behaviour	72.9±21.6	79.8±14.3
Mental Health	75.8±14.8	80.8±11.1
Self-esteem	73.8±21.1	79.7±15.4
General health perception	58.5±19.7	67.3±16.6
Family activities	79.0±24.2	83.0±19.3
Single-item domains		
Family Cohesion (100=excellent, 0=poor)	76.5±22.8	67.4±29.0
Change in Health (5=much better, 1=much worse than 1 year ago)	3.4±0.7	3.6±0.8
Global Health (100=excellent, 0=poor)	86.9±14.9	77.7±16.3
Global Behaviour (100=excellent, 0=poor)	83.5±22.8	84.0±15.7

Factors associated with physical activity at follow-up (n=23): Univariable regression analysis indicated that male sex was associated with a greater MVPA at follow-up (estimate[standard error];p-value) (+15.7[6.2] minutes MVPA/day; p=0.01). Lower physical activity at follow-up was associated with greater mean pulmonary arterial pressure at Fontan procedure (-3.0[1.5] minutes MVPA/day for every 1mmHg increase; p=0.04). A modest association was present between lower physical activity at follow-up and a higher number of surgeries prior to the Fontan procedure (-5.3[2.8] minutes MVPA/day; p=0.06). There was a significant positive association between MVPA at follow-up and higher childhood VO_2peak (+1.4[0.7] minutes MVPA/day; p=0.04). Higher MVPA at follow-up was also associated with higher childhood

moderate (+0.37[0.18]; 0.04), vigorous (+3.18[0.44]; <0.0001), and moderate-to-vigorous physical activity (+0.40[0.14]; p=0.004). MVPA at follow-up also showed a modest positive association with higher VO_2peak at follow-up (+0.88[0.46]; p=0.06).

Univariable regression analysis also showed that higher adult MVPA was positively associated with higher resting forced vital capacity (+7.9[3.5]; 0.02), higher resting force expiratory volume (+11.7[4.6]; 0.01), higher VO_2peak (L/min) (+18.7[5.7]; 0.001), higher VCO_2peak (+14.9[4.5]; 0.001), higher maximal Watts (+0.38(0.08); <0.001), higher O_2-pulse (+1.74(0.84); 0.04), higher VE BTPS (+0.28(0.11); 0.01), and higher VT max (+14.0[7.11]; 0.05).

Variable reduction following bootstrap identified the following variables to be included in a multivariable regression analysis with adult MVPA as the dependent variable: child vigorous activity, number of medications at discharge, gender, and adult VO_2peak (R^2-adj=0.90). Results indicated that higher MVPA as an adult was associated with greater vigorous physical activity as a child (+2.6[0.28]; <0.0001). Lower activity as an adult was associated with more medications at discharge following Fontan (-4.2[1.0]; <0.0001) and female gender (-8.1[3.3]; 0.01). Adult VO_2peak was modestly associated with adult MVPA (+5.1[1.0]; 0.07).

4.5 Discussion

This study sought to determine longitudinal changes in physical activity levels in a cohort of Fontan survivors over a seven-year period. Factors associated with physical activity levels were also identified. Results from this study demonstrated a mean 31% reduction in MVPA over a 7-year period. The Canadian Health Measures Survey (CHMS) is a population-based study that assessed the physical activity of healthy children and adults between 2007-2009 [108]. Patients

in the current study accumulated 25% less daily activity in childhood compared to healthy children of similar age (39 vs. 52 minutes/day, respectively)[108]. This finding is consistent with published work in the Fontan cohort that assessed physical activity, fitness, and body composition [146, 303]. Adults with Fontan circulation in the Fontan Follow-up Activity Study accumulated slightly more MVPA per day than healthy adults of similar age (27 min/day vs. 24 min/day, respectively)[107]. Due to limited studies that assessed physical activity in adults with Fontan circulation, it is difficult to comment regarding the representativeness of these findings. However, participation in a study that measures physical activity may include potentially biased patients that are more interested in physical activity and therefore demonstrate comparable activity levels to the general population.

Dua et al. assessed the physical activity of CHD patients (n=61; 31.7±10.9 years old) and reported similar daily MVPA as found in our study (26 minutes of MVPA per day) [5]. They noted that complex CHD patients accumulated significantly less MVPA compared to less complex patients (18.0±14.1min/day vs. 35.1±22.6, respectively). Sandberg et al. recently reported that adults with simple (n=40; median age (IQR): 30(20.7)) and complex (n=40; 33.7(29.4)) CHD exhibited similar physical activity patterns to healthy controls (n=42; 31.4(24.1)) [304]. Activity was considered moderate intensity if the heart rate of patients increased 1.75 times the resting heart rate as measured from the ActiHeart device, or if the heart rate reached and exceeded the heart rate measured at 3 minutes during submaximal step testing. There was no difference in the amount of time spent in MVPA between patients with CHD (simple or complex lesions) and control patients. Our study measured activity using a tri-axial accelerometer using pre-determined activity count cut-points that represent moderate-to-vigorous intensity independent of heart rate. The discrepancy between the results reported in our study compared to Sandberg et al. may be explained by the device and analysis method used to classify

the intensity of activity. This also emphasizes the need for additional research with more consistent methods between studies to provide valid comparisons. In addition, regional differences may contribute to this discrepancy, as published cohorts were derived from the United Kingdom [5], Sweden [304], and Canadian patients.

The overall reduction in physical activity levels between childhood and adulthood as observed in the Fontan Follow-up Activity Study has been previously reported among healthy populations [305, 306]. Previous longitudinal reports of physical activity trends have primarily relied on self-reported recall assessments (questionnaires), with few employing objective accelerometer-based measures of activity [307]. Telama et al. tracked physical activity between childhood and adulthood to investigate the stability of physical activity participation over a 21-year period [307]. Results showed high levels of activity accumulated from 9-18 years old, which predicted a high level of activity in adulthood.

Greater daily MVPA of adults with Fontan circulation was associated with greater vigorous physical activity levels in childhood. Previous cross-sectional research in this cohort showed that children with Fontan circulation have reduced physical activity levels compared to healthy peers and this reduced physical activity occurred independent of exercise capacity [130]. Lunt et al. reported that adolescents with CHD participated in less vigorous-intensity physical activity than healthy peers [131]. Therefore, interventions that help patients safely participate in vigorous-intensity activities may be warranted. Duppen and colleagues undertook a systematic review of the effects of physical exercise training programs in children and young adults with CHD [34]. This review identified one study by Dua et al. that assessed physical activity in adults with CHD (n=61; 31.7±10.9 years), which reported 21.7±16.9 minutes of MVPA per day at baseline prior

to exercise training, which increased to 31.9±27.0 minutes of MVPA at follow-up ($p<0.001$) [132].

The benefits of engaging in regular physical activity are well established, including both physical and psychosocial outcomes. Research completed to date emphasizes the importance of improvements in exercise capacity as a measure of improved health. Our results showed that exercise capacity was not associated with physical activity levels, and thus may not be an appropriate measure to predict physical activity behaviour as an adult. Despite the clinical value of determining aerobic capacity of patients to understand cardiac function and the patient's physiologic response to maximal exercise (i.e., arrhythmia, syncope, ventricular function, heart rate recovery), exercise capacity in relation to physical activity participation should remain as a complementary variable to describe the patient's functional status.

The reduced activity levels of CHD patients may be a result of parental overprotection experienced during childhood. Parental overprotection has been previously thought to negatively impact physical activity participation [308]. Reduced activity levels as an adult may have been impacted by misinterpreting exercise recommendations received during childhood, practicing independent decision making as adult, or learning to manage their health while experiencing substantial life transitions as an adult with CHD.

Improved physical activity promotion strategies should be developed for children with CHD in order to achieve increased physical activity levels, particularly regarding time spent in vigorous activity. Successful interventions in childhood may benefit patients as they enter adulthood and result in greater MVPA as an adult. Physical activity behaviours adopted in childhood should be made a priority in pediatric care in order to provide healthy behaviours that can continue into adulthood. This is particularly important for Fontan survivors, as patients with complex CHD are

at higher risk of developing conditions like atherosclerosis, coronary artery disease, or heart failure, and regular physical activity may serve to prevent additional complications associated co-morbidities like obesity, diabetes, or high cholesterol [309].

Patients in our study demonstrated 62%-predicted and 75%-predicted exercise capacity (VO_2peak) between adulthood and childhood, respectively. These values are similar to other reports [310], and therefore the 31% decline in physical activity observed in our patients may not be explained by changes in exercise capacity. Muller et al. reported that adults with Fontan circulation had a peak VO_2 of 63% predicted. In addition, they identified a slow decline in exercise capacity among CHD patients over time, represented a reduction by ~1% per year of the predicted VO_2peak [310]. Dulfer et al. showed that Fontan patients failed to achieve increases in exercise capacity following a 12-week exercise training intervention; however, this was attributed to a high baseline VO_2peak (33ml/kg/min), which was 5 ml/kg/min higher than our study group [15].

4.6 Limitations

The results of this study should be interpreted in light of the following limitations. The poor recruitment rate and final sample size highlight challenges faced in longitudinal research in this cohort. The small sample size derived from a single institution limits the generalizability of results to all patients with Fontan circulation and a more representative sample may produce different results. Furthermore, the small sample size may have contributed to an over-fit multi-variable regression model given the number of variables remaining in the model. In addition, the high R^2 may have been a result of collinear variables included in the final model. This requires further investigation to determine collinearity of variables and their respective contribution to physical activity. A different accelerometer model was used to measure physical activity at each

time point. The Fontan 1 Activity Study measured activity in the vertical direction only and thus captured movements like walking and running. In contrast, the Fontan Follow-up Activity Study physical activity measurement included data from all three axes and thus captured activity counts representing movement in all directions and those experienced in activities of daily living (i.e., jumping, bending, standing, laying down) [311]. This may have resulted in greater minutes of activity included in the follow-up study; however, a decline in physical activity was still demonstrated despite accelerometer data that included movement in all directions rather than a single axis. Vanhelst et al. compared physical activity measurement using uni-axial and tri-axial accelerometers [312]. Results indicated good agreement between data collected using the different accelerometers and the authors concluded that either monitor could be used to measure physical activity. Cut-points used for accelerometer analysis in this study were derived from healthy cohorts and may not account for disease-specific activity patterns and behaviours. CHD-specific cut-points have become recently reported and warrant further investigation [191]. Accelerometer assessments were not matched by season for each patient between activity assessment times (i.e., a patient may have worn the monitor in the spring in their first assessment and in the winter for their second assessment). Given the difficulties faced with patient recruitment and compliance, study measures were obtained when most convenient for the patient. Future longitudinal surveillance research should consider a season-matched approach to assess activity during the same season; however, this approach may not be appropriate for more short-term interventions. Furthermore, this study did not include longitudinal physical fitness assessments (i.e., muscular strength, muscular endurance, flexibility) that may have identified additional underlying factors to explain physical activity trends reported in this study.

4.7 Conclusions

This study identified a decline in physical activity levels into adulthood among a cohort of Fontan patients followed over a 7-year period. Results from this study indicate that childhood physical activities should include vigorous intensity activities when possible to help patients achieve higher physical activity levels as an adult. Individualized counseling to promote physical activity participation during childhood should be developed to identify appropriate activities for patients. Future research should include prospective studies with additional physiologic and psychosocial outcomes to identify potentially modifiable factors that contribute to physical activity behaviour.

This study described the physical activity trends among a cohort of Fontan patients as they entered adulthood and identified associated factors. The complex nature of CHD warrants a deeper understanding of factors that contribute to the decline in physical activity participation as observed in this study. In addition, potential facilitators should be explored that may help inform the development of interventions that target improvements in physical activity. A qualitative approach may help identify important information that is missed when using quantitative study designs. Collecting qualitative data from patients could provide the patient's perspective regarding physical activity and be used to develop interventions that meet the needs of the CHD population.

5 Physical activity perceptions and behaviours among emerging adults with CHD

5.1 Study Contributions

Adam McKillop, assisted by Dr. Brian McCrindle and Dr. Adrienne Kovacs, conceptualized this study. Adam McKillop, assisted by Dr. Gina Dimitropoulos, developed the qualitative component of this study (i.e., approach, interview questions, analysis plan). Adam McKillop completed participant recruitment and data collection. Adam McKillop completed interview transcriptions, with assistance by Mimi Bandyopadhyay. Adam McKillop, assisted by Dr. Gina Dimitropoulos and Dr. Adrienne Kovacs, completed qualitative interview analysis and interpretation. Adam McKillop, with assistance from Dr. Adrienne Kovacs, completed quantitative data analysis. The primary supervisor for this study was Dr. Adrienne Kovacs.

5.2 Introduction

Approximately 90% of children with CHD survive into adulthood [85]. The number of adults living with CHD continues to rise and there are now more adults than children living with CHD in North America [40]. The emerging clinical population of adults with CHD of moderate to great complexity requires life-long cardiac care at specialized centres to manage their chronic condition. Many adults with CHD may consider themselves “cured” or “fixed”; however, CHD is considered a chronic condition as these patients are at increased risk of arrhythmias, endocarditis, and congestive heart failure [313].

As a group, adults with CHD are less active than those without CHD and many report a lower quality of life [5]. Patients with CHD may be discouraged from physical activity due to an

increased fear of heart failure or sudden death [314]. Due to these physical or self-imposed limitations, many adults with CHD experience physical activity restrictions throughout childhood [158]. Research has shown that adults with CHD often possess greater heart-focused anxiety and reduced self-efficacy towards physical activity and exercise [308]. In addition, approximately one-third of adults with CHD meet the diagnostic criteria for mood and anxiety disorders [289]. A sedentary lifestyle is associated with obesity, high cholesterol, hypertension, reduced exercise capacity, depression and anxiety [123, 309]. Regular physical activity and exercise has been shown to attenuate mood and anxiety symptoms, improve quality of life, and is important to combat the development of co-morbidities present in the general population as a result of poor lifestyle management [93-95].

Previous research primarily focused on understanding patient and parent perceptions of physical activity for children and youth with CHD [315, 316]. Among children with complex CHD, studies indicated that physical activity levels were below the 50th percentile compared to healthy controls [130]. Most children and adolescents with CHD are willing to participate in physical activity [294]; however, significant barriers to physical activity exist. For example, conflicting physical activity restrictions often exist between cardiologists, parents, and health records, and these discrepancies may negatively influence physical activity participation [133]. Moola et al. identified social barriers for physical activity among youth with CHD, including challenges faced with disclosure of their heart condition, participating in the physical education setting, and understanding their body [134]. This qualitative study also highlighted the need for healthcare professionals to address the social and cultural environment in which the patient lives. Although research has shown that adults with CHD can benefit from organized exercise programs [5, 132, 290], qualitative research may help identify physical activity perceptions among the adult CHD population.

Despite evidence-based guidelines to promote physical activity among the CHD population [118], little is known about how adults with CHD perceive physical activity and exercise. Young adults with CHD may experience a burden of adult-onset complications or illnesses and reduced physical functioning while navigating independent decision making [317]. In particular, emerging adults (18-25 years old) represents a distinct population that possess unique experiences as they approach young adulthood [318]. Emerging adults encounter significant changes in life, including identity development, increased sense of autonomy, and coping with the altered parent–child relationships [319]. In addition, emerging adults with CHD are often faced with challenges in maintaining healthy behaviours [317]. Qualitative research may serve to fill the existing gap in the adult CHD literature to provide greater insight into the patient perceptions of activity participation among this emerging population. Therefore, it is important to understand the perceptions of physical activity as they relate to the experience of entering adulthood with CHD. This information may help identify key experiences to improve physical activity promotion among the ACHD population.

This study sought to explore exercise perceptions and behaviours among emerging adults with CHD. The main study aims were to:

1. Describe the perceptions of physical activity and exercise from childhood through emerging adult using semi-structured qualitative interviews.

2. Objectively measure the physical activity level of patients using accelerometers to characterize the study population.

3. Assess psychosocial outcomes to identify potential factors correlated with physical activity level.

5.3 Methods

This cross-sectional study received institutional ethics board approval from the University Health Network (Toronto, Canada). Study participation included an in-person interview to explore physical activity behaviours and perceptions of young adults with CHD, completion of psychosocial questionnaires (which they could take home to complete), accelerometer-based assessment of activity, and medical record review.

Patients: Patients ages 18-25 years old were recruited from the Toronto Congenital Cardiac Centre for Adults (TCCCA) within the Peter Munk Cardiac Centre at the University Health Network in Toronto, Ontario from November 2014 and June 2015. All patients had undergone prior repair of CHD of moderate to great complexity [78], and were at least one year after most recent open-heart surgery, and had the language and cognitive abilities to independently provide informed consent to participate in an interview and complete surveys. Enrolment continued until data saturation for the qualitative component of the study was achieved whereby no new themes were identified following analysis of the most recent transcript.

Approach: Clinic lists and medical charts were reviewed each week to identify eligible patients to be recruited. Patients provided verbal and written consent and were encouraged to ask questions at any time during the consent process. After informed consent was received, patients completed the following study procedures.

Qualitative Research Design: This study described the perceptions of patients by exploring their experience with physical activity throughout their life. An over-arching theoretical framework did not inform this study, nor were data collected to generate new theories. To this end, a qualitative descriptive approach was used, as it is not tied to a pre-existing theoretical or philosophical commitment [320]. Basic or fundamental qualitative description is a valuable

method that interprets the data with low-inference [320]. In line with qualitative description design, participants were selected using purposeful sampling and data were collected using semi-structured interviews [321]. Data were analyzed using thematic analysis that applied a low-level of interpretation beyond the experiences as described by participants and reported in similar language used by participants [322].

Qualitative Interview: Interviews collected a detailed narrative from emerging adults with CHD to understand their perceptions of physical activity as a child, adolescent, and as an emerging adult (Table 10). This qualitative assessment used semi-structured, one-on-one interviews followed by thematic analysis to identify emerging themes. Interviews aimed to address the following topics: (i) childhood physical activity, (ii) importance of physical activity, (iii) perceptions about physical activity, and (iv) outcome expectations regarding physical activity. The first question for all interviews was, "*Tell me what it was like growing up with congenital heart disease*." The interview progressed from questions about childhood towards the patients' outlook regarding activity participation and life as an emerging adult with CHD. All patients were asked, "*Do you have anything else to add with regards to your physical activity or exercise experience?*" as a closing question.

Qualitative Interview Setting: All interviews were completed on the same day as the patients' regularly scheduled clinical appointment. Following the clinical appointment, patients were invited to an exam room or small conference room to complete the interview. This room was in the same area as the clinical appointment. The patient was asked to sit at the table located in the room, and the interviewer sat across from the patient. The interviewer confirmed with the patient that it was okay to record the session for transcription and data analysis. The audio-recorder was placed on a table in between the patient and interviewer. Once the audio-recorder was set-up, the

patient and interviewer continued with the interview questions. The interviewer used the questions outlined in Table 10 as a general guide for the interview and asked probing questions to elicit more information from the patient on a particular topic. Each interview progressed through the different topics outlined in Table 10 and finished with any final comments by the participant that they felt would contribute to the understanding of their perceptions towards physical activity.

Table 10. Qualitative interview questions.

1. **Childhood physical activity (past):**
   - *a.* What was it like growing up with CHD?
   - *b.* How active were you growing up? What physical activities did you do as a child?
   - *c.* What physical activities were you unable to do as a child? What prevented you from doing some of these activities? What was that like for you?
   - *d.* What helped you be active? What were some thoughts and feelings you had that may have helped you be active?
   - *e.* What situations prevented you from being active? What were some thoughts or feelings you had that may have prevented you from being (more) active?
   - *f.* How involved were your parents with your physical activity? What did your parents tell you to do or not do growing up? How did you respond to their involvement? How did your parents' involvement change over time? How involved are your parents today?
2. **Importance of physical activity (present):**
   - *a.* How has your physical activity participation changed since childhood?
   - *b.* How important is physical activity in your life?
     - **i.** What are your thoughts about exercise now? What has contributed to the changes in your views about exercise?
     - **ii.** What is more important than exercise?
3. **Perceptions towards physical activity (present/future):**
   - *a.* Please describe what type of physical activity you engage in now. What are your reasons for engaging in physical activity?
   - *b.* What types of physical activity do you participate in currently?
   - *c.* What helps you participate in physical activity?
   - *d.* What stops you from participating in physical activity?
   - *e.* What might help you become more physically active?
   - *f.* What role has technology played in your activity participation? How could technology help you become more active?
   - *g.* What does it mean to you that you can (cannot) participate in physical activity? What are your feelings and thoughts about physical activity now?
4. **Outcome expectations regarding physical activity (future):**
   - *a.* What do you believe are the benefits of physical activity for people with CHD? What may the benefits be for you?
   - *b.* What do you believe are the short and longer-term physical activity goals for people with CHD? What about for you?

Questionnaires: Patients provided basic background information (e.g., marital status, if they have children, highest education level, student status, work situation, and current and/or past psychological treatments). The *Readiness to Change* questionnaire [323] was used to assess the patient's readiness to change their physical activity behaviour. This classified each patient into one of five stages of change: pre-contemplation, contemplation, preparation, action, or maintenance. The *Self-Efficacy for Exercise Scale* measured the patients' self-efficacy to be active [324, 325]. Global quality of life was assessed using a 10cm *Visual Analog Scale (VAS)*, which has been previously applied in the adult population and recommended as an assessment of global quality of life in clinical research [326-330]. Patients were asked to indicate their overall quality of life from zero (worst imaginable quality of life) to 100 (best imaginable quality of life). The *Cardiac Anxiety Questionnaire* was used to assess the patients' current level of heart-focused anxiety and feelings towards physical activity [308, 331]. Assessment of the patients' physical activity was measured using the self-report *Habitual Activity Estimation Scale (HAES)* [332]. A *Patient-based New York Heart Association (NYHA) Classification Assessment* was used to identify patient-reported NYHA classification, as described by [333].

Accelerometer-measured Physical Activity: Patients were provided with an accelerometer (ActiGraph, wGT3X+, Pensicola, CA) to measure their daily physical activity. Patients were asked to wear the monitor around their waist over their right hip at the mid-axillary line for seven days starting on the day after their clinic visit. The collection period was to include five weekdays and two weekend days. The device was programmed for a 15-second sampling interval and stored the activity count per 60 seconds to memory at the end of each successive interval. A logbook was provided to patients to record the days they chose to wear the monitor and any unusual circumstances that influenced activity participation (i.e., long bus trips, illnesses, etc.). Patients were asked to return the accelerometer using a pre-paid postage envelope

provided to them. Upon return of the accelerometers, stored activity counts for each 60-second interval were downloaded for analysis (ActiLife 6.1). Daily activity counts were converted to intensity levels achieved per day **based on age-specific cut points [302], providing a final estimate of time spent doing physical activity at different intensity levels.**

Medical Record Review: Patient medical records were used to abstract patients' clinical history including diagnosis, age at surgery(ies), years since surgical repair, surgical or medical complications related to their cardiac diagnosis, documented history of psychological difficulties and/or treatment, results of the most recent cardiopulmonary exercise test, documented exercise restrictions recommended by the cardiologist, and the number of hospitalizations for cardiac-related problems.

Data Analysis

Qualitative Data Analysis: All interviews were conducted, transcribed, and analyzed by a single investigator (A.M.). Thematic analysis was used to identify, analyze, and report specific patterns, or themes, in the data [334]. Though not bound to a single theory, such as Grounded Theory or Interpretative Phenomenological Analysis, thematic analysis is widely used in qualitative research and provides a level of flexibility and access to the data [334]. A six-phase approach to thematic analysis was used as described by Braun and Clarke and included the following phases: 1) familiarization with data; 2) generation of initial codes; 3) searching for themes; 4) reviewing themes; 5) defining and naming themes; 6) producing the report. Provisional results were reviewed with members of the study team to discuss the interview progress, preliminary themes, and reliability of the analysis. An iterative approach was relied upon to share and discuss interpretations of the results throughout the data collection and analysis period.

Quantitative Data Analysis: Data from questionnaire and accelerometer-based assessments were calculated as mean±standard deviation. Given the small sample size and heterogeneity of patient characteristics, results were analyzed for descriptive purposes only.

5.4 Results

Patients (Figure 18): Clinic lists were reviewed each week to identify eligible patients. A total of 170 patients were screened, including medical chart review to confirm inclusion and exclusion criteria. Patients were determined to be ineligible (n=142) if there was no ACHD clinic medical record, had a simple CHD diagnosis, documented cognitive impairment, or other complex medical conditions. Eligible patients (n=28) were approached to participate in the study. Patients declined to participate (n=13) in the study due to time limitations and unwillingness to extend their clinic visit or reschedule. Study enrolment included 15 patients who agreed to participate in the study and completed the consent process. One-on-one interviews were completed for each patient. Accelerometer data were available for 10 patients, as two devices showed insufficient wear time and three were not returned. Questionnaire data were available for 12 patients, as three patients completed the assessments at home but did not return the questionnaire package. Therefore, data were available from the interview, accelerometer, and questionnaire components of the study for 10 of the 15 enrolled patients.

Patient Demographics (Table 11): Fifteen patients (5 females and 10 males) participated in this study. The mean age of patients was 21±3 years and all patients were single and without children. Most patients completed some university/college (n=8; 53%) and were currently students (n=11; 73%), while other patients completed high school (n=2; 13%) or had a university or college degree (n=5; 33%). Patients reported working fulltime (n=3; 20%), part-time (n=7;

47%), or unemployed (n=5; 33%). One patient reported receiving previous psychological treatment (talk therapy). No patients reported taking medication for a psychological problem.

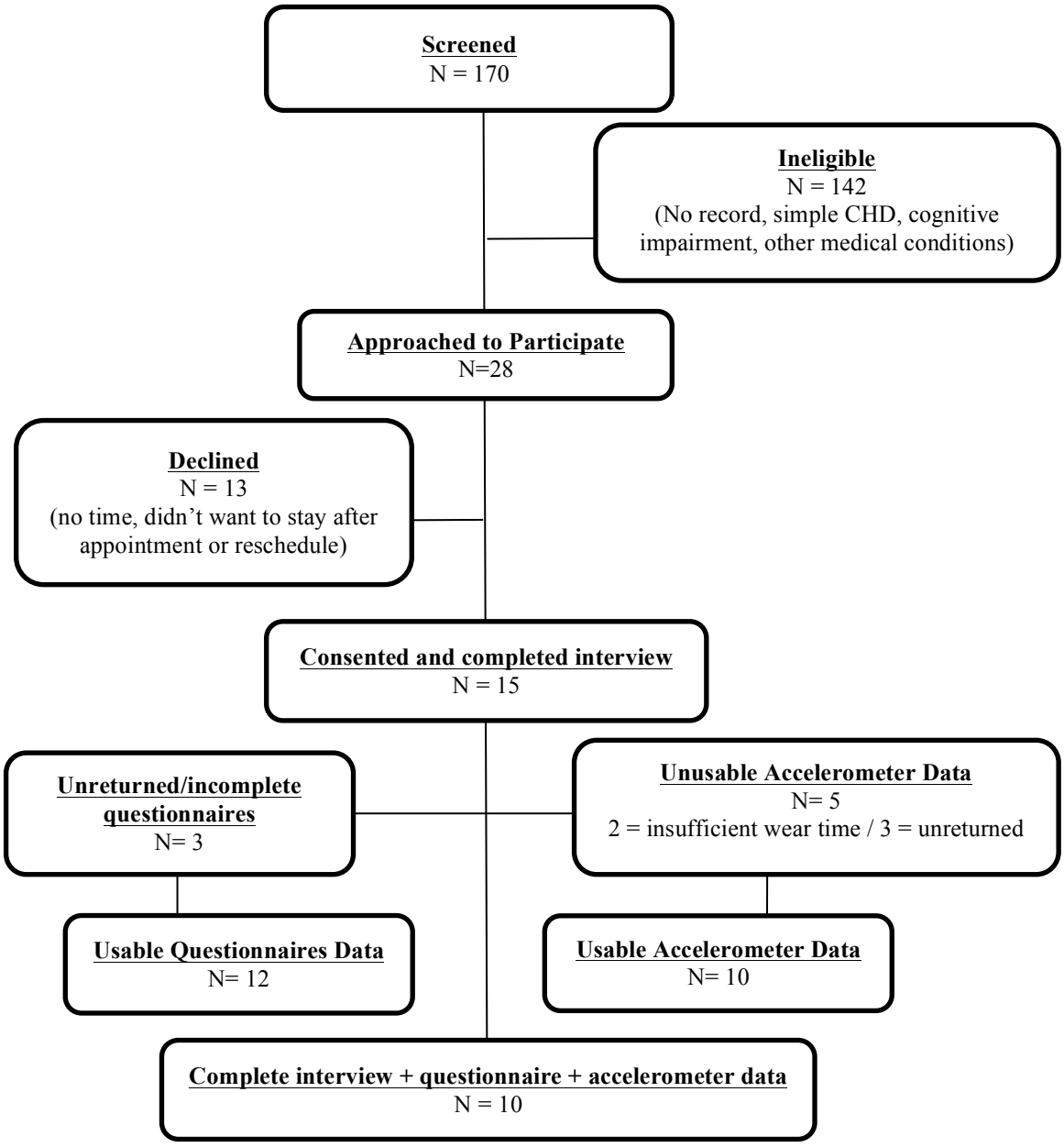

Figure 18. ACHD Exercise Study patient recruitment diagram.

Patient Medical History: Medical chart review indicated that patients had CHD of moderate (n=10, 67%) or great (n=5, 33%) complexity with a mean number of surgeries of 2±2 and 14±8 years since the most recent surgery. Cardiac diagnosis included transposition of the great arteries (TGA) (n=1), atrio-ventricular septal defect (AVSD)(n=2), biscuspid aortic valve with significant regurgitation (n=2), congenitally-corrected TGA (n=1), coarctation of the aorta (n=2), hypoplastic left heart syndrome (HLHS) (n=2), pulmonary atresia (n=1), pulmonary stenosis (n=1), and tetralogy of Fallot (TOF) (n=3). No patients had a documented history of arrhythmia or implanted cardiac device. No patient had a documented psychological diagnosis or treatment. Three patients had documented exercise restrictions in the medical chart advising to avoid isometric/strenuous weightlifting, while the other 12 patients had no documented exercise restrictions.

Table 11. Patient characteristics (N=15).

Variable	
Age (years)	21±3
Gender	
Male	10 (67%)
Female	5 (23%)
Single	15 (100%)
Education	
College/University	5 (20%)
Some college/university	8 (47%)
High School	2 (13%)
Employment Full-time Part-time Unemployed	 3 (20%) 7 (47%) 5 (33%)
Primary CHD Diagnosis	
Tetralogy of Fallot	3 (20%)
Atrioventricular Septal Defect	2 (13%)
Bicuspid aortic valve	2 (13%)
Coarctation of the Aorta	2 (13%)
Hypoplastic Left Heart Syndrome	2 (13%)
Transposition of the Great Arteries	1 (7%)
Congenitally-corrected TGA	1 (7%)
Pulmonary Atresia	1 (7 %)
Pulmonary Stenosis	1 (7%)
Documented Exercise Restriction	
Yes (isometric exercise)	3 (20%)
No	12 (80%)

Exercise Capacity (n=11): The most recent exercise test results were available for 11 patients who completed testing in the ACHD clinic. Exercise testing was not completed at the ACHD clinic at the time of this study for four patients. The mean peak oxygen consumption was 30±6 ml/kg/min, representing 72% of predicted. Resting heart rate increased from 77±9 beats per minute at rest to 160±27 beat per minute at maximal effort, representing 78% of predicted max heart rate. A respiratory exchange ratio of >1.1 was achieved in all tests, indicating a maximal effort. The average duration of the exercise test was 14±4 minutes.

Physical Activity (n=10): Complete accelerometer assessments were available for 10 patients, as three accelerometers were not received and two returned monitors displayed insufficient wear-times (minimum requirement was two weekdays and one weekend day). Accelerometer data from the ten patients with usable accelerometer data is summarized in Table 12.

Table 12. Descriptive statistics for physical activity assessment reported as minutes per day (n=10).

	Mean	**Range**
Total valid wear days	6±0.9	4 – 7
Wear time (hr/day)	11±0.8	10 – 15
Sedentary (min/day)	567±116	258 – 562
Light Activity (min/day)	67±21	38 – 101
Moderate Activity (min/day)	23±14	7 – 46
Vigorous Activity (min/day)	3±2	1 – 8
MVPA (min/day)	26±16	8 – 54

Intensity levels based on Freedson et al. 1998; MVPA: moderate-to-vigorous physical activity.

Habitual Activity Estimation Scale (n=10): The HAES questionnaire provided a subjective report of activity participation during the weekend and weekdays. Over half of the patients (55%) reported being "somewhat active or lower" and 45% reported being "active or very active" during the week. Patients reported being somewhat active for 326±198 minutes (~5 hours) per day and very active 191±197 minutes (~3 hrs) per day. Weekend activity was reported

as “somewhat active or lower” for 58% of patients and “active or very active” in 42% of patients. Patients reported being somewhat active for 299±160 minutes (~5hrs) and very active for 134±140 minutes (~2 hours) on weekends.

Psychosocial Outcomes (n=12): Some patients requested to complete the questionnaires at home to reduce the time spent in clinic but three patients did not return the questionnaires by mail as requested. Despite multiple follow-up attempts, these patients could not be contacted. The Readiness to Change questionnaire determined that the majority of patients (n=8) were in the *Maintenance* phase, while 1 patient was in the *Contemplation* phase, 1 patient in the *Action* phase, and 2 patients in the *Preparation* phase with regards to changing their physical activity participation. Patients classified themselves as being in NYHA Class I (n=6) or NYHA Class II (n=6). Results for quality of life, self-efficacy, and cardiac anxiety psychosocial questionnaires are summarized in Table 13.

Table 13. Descriptive statistics for psychosocial assessments (n=12).

Questionnaire	Mean	SD	Range
Quality of Life	84	11	60-100
Self-efficacy for Exercise	55	22	19-81
Heart-focused Anxiety			
CAQ total score	25	13	8-53
CAQ mean score	1.39	0.73	0-3
NYHA Functional Classification			
I	6	-	-
II	6	-	-
III	0	-	-
IV	0	-	-

Qualitative Interviews (n=15):

Thematic analysis of interviews identified the following six themes: 1) Importance of Family, 2) Parental Support-Not Overprotection, 3) Adaptation for continued activity participation, 4) Influence of School, 5) Occupational Activity, and 6) Activity for Health

Importance of Family: The majority of patients (n=12) acknowledged their family as being supportive of their participation in activity throughout childhood. When asked, "*What helped you be active as a child?"* patients described how parents and/or siblings played a role in their activity participation.

"Besides my parents, my sister – she is also really active. So even if it was a crumby day out, she would always get me off the couch or get me out of bed at 7 to go ride our bikes…it helps me get through it, when she keeps playing because I love watching her play too if I can't play the sport."

Patients generally described positive experiences throughout childhood and encouraging interactions with their parents and family members. Some patients (n=3) commented on the role of older siblings that encouraged the patient to be more active. Patients discussed the support drawn from siblings and parents as strong role models. Family members often participated in activity along with the patient and facilitated activity participation.

"*…I have an older brother, and he too is very active, so I think seeing him be that way growing up was normal to me – that people are active because my parents are…so just my family being active was normal to me*."

Overall, patients reported that family provided a positive support system that helped them to be confident in their activity participation growing up. Patients also commented that family support and involvement continued into adulthood but to a less extent compared to childhood. Parental support in adulthood took more of a passive role but patients indicated that their parents still checked-in and inquired about their activities and health.

Parental Support – Not Overprotection: Patients (n=12) described the involvement of their parents as being supportive and encouraging and did not identify over-protective parenting styles. Patients experienced a positive upbringing with respect to their parents' involvement and described opportunities to participate in physical activity and sports.

"…I think it made me feel more supported and like I was – I just felt like I had somebody that was saying, "you're allowed to do this" and they were supporting me, whereas if I didn't have that, I probably would have not been as excited to go and do all these things I did."

Some patients also recalled parents that facilitated activity participation, including purchasing equipment, program registration, and travelling with sports teams. Patients perceived this as the parents' way to help them stay healthy or to fulfill the parent's desire to have them play sports and fit in with other children.

"Like if I wanted to sign up for whatever it was, they would sign me up and even if they thought I wasn't going to be the best, they said at least give it a try. And if she doesn't like it or she can't do it, we will just pull her out. They always encouraged me to try whatever I wanted to do."

"I never really noticed until the last couple of years how supportive and encouraging they were. But over the years, you could obviously tell that they were, but you just never really notice until now. But you think back and you just owe them so much..."

Patients described physical activity restrictions or precautions in childhood as instructed by the cardiologist. The role of the parent in enforcing any exercise precautions was seen as compliance to the cardiologist's recommendations, and not necessarily coming directly from the parent. Therefore, the parent was not necessarily regarded as over-protective, but rather the messenger for the cardiologist and supportive towards their child's activity goals.

Adaptation for continued physical activity participation: The majority of patients (n=11) recalled a relatively normal childhood and rarely considered themselves as being different from their peers. Although the perception of a normal childhood was commonly discussed, patients also commented on differences between themselves and their peers in terms of endurance and coordination. The existence of some physical limitations did not deter patients from activity participation. Patients experienced some exercise intolerance growing up and were unable to participate fully in some activities. Patients managed these challenges by either working within their limitations or choosing an activity that met their expectations and in which they were able to participate fully.

"Growing up I learned to get used to it. Like sometimes I would have to take a break and my friends would understand…but I noticed I would take longer breaks than my friends."

Patients commented that they never experienced life without CHD and acknowledged that growing up with CHD was a normal part of their life. Some patients (n=3) said that they never noticed their CHD growing up, except missing school and coming to the hospital for appointments. Patients felt the CHD had no bearing on decisions they made with respect to physical activity choices. One patient compared herself to peers and described an appreciation for the ability to be physically active despite having complex CHD.

"If a kid with half a heart can do it, a kid with a full heart can do it…by me showing them [other children without CHD] I can be active with half a heart, and you with a full healthy body and whole heart, you will be able to do it too."

Some patients (n=4) recalled instances of being left out from opportunities, but seemed to manage this experience in childhood. Patients' perseverance and positive attitude towards activity helped them navigate the societal expectations, especially in school settings, in order to remain active.

" I lived a pretty normal life growing up. The only things were certain restrictions – like I couldn't play hockey, I couldn't play contact sports. But otherwise I grew up completely normal. So that was really the only limitation I had growing up…like I was still a really active kid - I could still run and bike, and whatever. I didn't have problems with that."

Influence of School: Patients discussed their activity as a child in the context of school, physical education, and sports participation. Daily physical activities in the context of activities of daily living (i.e., active transport, house work, leisure activities) were not recalled as frequently or with as much detail compared to the experiences from school. Furthermore, description of activity generally involved a comparison of peers who were regarded as healthier and a group with whom the patients aspired to be included.

"I had a bit of trouble keeping up with my peers…but it wasn't anything serious, it was just I couldn't really keep up that much…it was just being picked last for teams."

"I was actually able to do what the average kid was doing. And then near high school I was able to do more activity but I was not exactly on the sports team or anything like that because I was too scared to try out for anything."

Occupational Activity: Questions about physical activity in adulthood resulted in comments regarding their activity level in the context of their work environment. This was apparent across patients regardless of reported activity levels throughout the day. Occupational activity was regarded as high volume and patients described this as sufficient activity to meet daily recommendations. However, patients also acknowledged that it would likely be better if they did more activity outside of work, but they were too tired after a long workday.

"I'm still on my feet all day and I'm running around or whatever. Like I still have to do some manual labour, but when I was a kid it was just recreational. Like just skateboarding or whatever. Whereas now I have work and I'm up on my feet all day."

"As I've gotten older I do a lot less physical activity. I mean I do a lot of physical related tasks with my job, but I wouldn't say that I'm going out to go for a run or anything like that. I'm not a super active person."

One patient described the need to change occupations due to the high physical demands of the job. The patient described a conversation about physical activity during a recent clinical appointment and the reality of having CHD and working in a physically demanding job.

"… I should look at getting another job because, like right now, it's not bad…so he [cardiologist] is like "you may be able to do it for the next 15-years but based on other people with your condition…you won't be able to do it."

Activity for Health: Patients (n=11) commented that a main reason to participate in activity as an adult was primarily for health reasons. This was partly attributed to their increased independence and improved self-management of their condition. Patients also drew comparisons from other aspects in their life where they experienced an increased independence (i.e., living

independently, paying bills, establishing relationships) and sought to apply this to their CHD care. While patients described their reasons for participation in activity during childhood as primarily recreational, they commented that the reasons for participating in activity as an adult were primarily related to health.

"…because you should be happy and enjoy things in life as a child and that's why I did it as a kid. And now it's important because of health. I'm older and I understand health more, so I also do it [activity] for that factor."

Activity in adulthood was also described in the context of maintaining good health and prevention of disease to delay future intervention. Patients described an uncertainty regarding long-term outcomes but knew that regular physical activity might be beneficial to help combat co-morbidities. Patients described the many benefits of regular physical activity, including both physical (i.e., cardiovascular fitness and weight management) as well as psychosocial aspects (i.e., improved mood and decreased feelings of anxiety and depression).

"No matter what school work gets involved, like health wise, I know I am going to get sick again with my heart, so I just try to push it off as much as possible by staying active and eating healthy."

5.5 Discussion

The current literature regarding physical activity and exercise behaviour among the CHD population focuses on children, with limited reports of adult CHD patients. Qualitative research with emerging adults with CHD is particularly scarce. Therefore, this study examined the physical activity and exercise perceptions and behaviours of emerging adults with CHD.

Physical activity among emerging adults with CHD: Our study is one of few that objectively assessed the physical activity of emerging adults with CHD using accelerometers. Previous research by Dua et al. reported that adults with CHD accumulated 26 minutes of MVPA per day, consistent with the results from our study [5]. Work by Sandberg et al. and Buys et al. included self-reported activity measures of activity among ACHD patients that indicated suboptimal levels of physical activity [304, 335]. In contrast, Muller et al. reported that 76% of their ACHD study population reported achieving the daily activity recommendation for adults with a mean moderate activity level (>3 METS) of 59.2+39.7 min/day [336]. Jackson et al. also reported that emerging adults accumulated adequate MVPA using a self-report instrument [337]. These findings are in contrast to others, including our study, and may indicate that adults with CHD do not experience reduced activity levels. However, this requires further investigation using more rigorous and consistent activity assessments. In particular, the use of accelerometers to define activity participation in this population may provide an objective assessment and avoid pitfalls associated with self-reported instruments [338, 339].

Previous work in both healthy and CHD populations reported a decline in physical activity with age [104, 336]. Exercise training interventions are feasible and have shown some efficacy to increase exercise capacity and physical activity levels in ACHD patients [132, 290]. The European Society of Cardiology and the American Heart Association have published exercise and activity recommendations for common CHD diagnoses [118, 136]. While the adult model of cardiac rehabilitation programs may be an option for some adults (particularly for those ACHD patients with similar characteristics to adults with acquired heart disease), facility-based rehabilitation programs pose significant logistical challenges to young cardiac patients. Furthermore, the long-term benefits and safety of such exercise interventions for the ACHD

population are unknown. Further investigation is needed to better inform the development of programs designed to meet the short-term and long-term needs of ACHD patients.

Favourable psychosocial outcomes among emerging adults with CHD: Moons et al. measured the quality of life of ACHD patients using a VAS similar to the one used in our study, and indicated a median score of 80/100 among their patients [327]. This is comparable to the quality of life of patients in our current study (mean score of 84/100). The perceived ability to be physically active discussed during the interviews is concordant with this high quality of life score. Furthermore, our results agree with the body of literature indicating that exercise is a marker of one's quality of life [25, 340, 341]. The high quality of life reported in our study might have been expected as the study population was screened and tended to exclude those patients with identified risk factors for diminished quality of life, including social impediments, trait anxiety, lower educational level, orthopaedic problems, psychosocial problems, and age >23 years [158, 342, 343].

Results from the self-efficacy for exercise assessment indicated that patients with CHD were confident they could participate in physical activity. Patients in the current study reported a wide range of self-efficacy (19-81 out of a possible 90 points). Dua et al. reported that adults with CHD had some belief that they could participate in physical activity [5]. Self-efficacy is a key component in many exercise-training programs that aim to build self-efficacy using interventions based on the Social Cognitive Theory. Structured programs for the ACHD population that include elements derived from theoretical frameworks may lead to improvements in self-efficacy, especially those who reported low quality of life.

Results from the cardiac anxiety questionnaire are comparable to work completed by Ong et al. that reported a mean CAQ score of 1.29 among a cohort of ACHD patients [308]. Patients in the

current study reported a low heart-focused anxiety, indicating that concerns about their heart during activity are unlikely to contribute to avoidance to be physically active. Furthermore, results from the interviews identified that patients felt supported by parents and did not experience over-protective parenting as previously described in the CHD population [308, 344, 345]. Turgeon et al. described the negative impact that overprotective parenting has on the patient's feelings of dependency and self-efficacy [346]. Taken together, patients in our study reported growing up in an encouraging environment that fostered greater self-efficacy to be active. This likely contributed to the reported low cardiac anxiety and greater participation in a range of activities throughout childhood.

Qualitative Interview: Interviewer experience

During the interview, the first few questions helped to establish rapport between the patient and interviewer. These questions also provided information for the interviewer to learn more about the patient's life as they grew up with CHD. It was noted that patients were generally more forthcoming with information as the interview progressed. This was particularly evident when asked about their current perceptions towards physical activity compared to questions about their childhood. This may have been a result of rapport being established during the earlier questions about childhood physical activity. Patients seemed more willing to expand on thoughts that were more relevant as a young adult, while answers about their childhood experience did not provide as much detail about physical activity participation. Furthermore, questions that asked patients to consider their future goals and outlook as an older adult with CHD seemed difficult for patients to answer. In some cases, the request to consider their future was almost abstract and lacking relevant context to elicit substantial responses. Identifying long-term physical activity goals was

particularly challenging for patients. This may be related to a general sense of uncertainty about how their CHD may influence their future physical activity participation.

Qualitative Interview: main findings and next steps.

Following qualitative data analysis, patients viewed family involvement in the their physical activity pursuits as a positive factor. Family-based interventions applied throughout childhood may be the first step in fostering a positive attitude towards activity participation. Patients described positive and supportive experiences from childhood and recalled family-based activity participation. The physical activity behaviour and health (i.e., overweight status) of parents are well known correlates of child physical activity levels [347, 348]. Therefore, it is not surprising that patients described an overall active childhood and limited activity restrictions from parents in this study. Kendall et al. reported the views of parents of children with CHD, whereby parents described the importance of providing a "normal" upbringing for their child [349]. Parents described feeling stressed as their child with CHD reached secondary school due increased physical demands, but trusted their child to be aware of their own activity limitations. Parents also commented on the need to receive detailed and individualized physical activity recommendations for children with CHD. Moola et al. described physical activity perceptions of parents of children with CHD or cystic fibrosis [350]. Parents described the stress and challenges associated with raising a child with a chronic condition, the barriers to physical activity participation, and the importance of role modeling to help their children be active. Parents also described the benefits of activity for their children; however, parents of children with CHD had more trouble explaining the benefits of activity compared to parents of children with cystic fibrosis. Stieber et al. investigated the feasibility of a home-based, parent-delivered neurodevelopmental rehabilitation program for children ages 12-26 months with CHD [147].

This 10-week intervention included five two-week sessions that included play-based activity options to be completed 20 minutes per day. Qualitative interviews with parents indicated that increased family interaction and time spent doing the activities was an important outcome in this study. In addition, parents engaged older siblings in the activities to demonstrate the activity or task. This type of family interaction in the context of physical activity interventions aligns with the results from our study, whereby patients recalled positive experiences including parent and sibling involvement in their physical activities.

Patients in our study often considered activities performed during one's employment to be sufficient to meet physical activity recommendation levels, and reported limited time and/or energy to participate in activity outside of work. Examples of adequate activity included lifting heavy objects, stocking shelves, or walking a lot. The ability to perform physical tasks at work may be expected for some patients; however, occupational challenges have been identified for adults with CHD [79, 313]. Sluman et al. conducted qualitative interviews with a cohort of young adults (ages 22-35 years) with CHD and identified barriers (increased physical load, lack of time to recover from work, and poor employee-employer relationship) and facilitators (low physical demands at work, autonomy to choose tasks and increased recovery time from work, and supportive employers and colleagues) [79]. Patients in our study did not discuss occupational outcomes or challenges in the context of barriers/facilitators, but viewed success in terms of doing well in school, obtaining work, and establishing a career. They may not have experienced the occupational challenges as described my Sluman et al. due to the younger age (18-25 years old) and occupational status (73% of patients were students) in our study. One patient did, however, comment on the need to change jobs in the future due to the physical demands as recommended by their cardiologist. This case was similar to one described in a case report of a patient with tetralogy of Fallot that experienced minimal cardiac problems throughout

adolescence and early adulthood but encountered serious cardiac issues in his mid-30s that impacted not only his career opportunities but also self-image and self-esteem [313]. Although a small sample size precluded in depth correlation analysis between psychosocial and physical measures in our current study, Kovacs et al. reported a positive association between lower annual income with increased heart-focused anxiety in a cohort of cardiac patients [351]. At the time of the interview in our study, almost 90% of patients were completing or have completed a university/college degree and two-thirds of patients reported working (part-time or full-time). In combination with low cardiac anxiety, these results may indicate a positive trajectory for patients in our study in terms of occupational success.

Future research and application of the Social Cognitive Theory: The qualitative interviews uncovered important factors that may be interpreted using a Social Cognitive Theory (SCT) perspective. The SCT, as described by Bandura in the 1970s, emphasizes reciprocal determinism and the interaction between people and their environments [352]. More specifically, human behaviour is seen as the product of personal, behavioural, and environmental factors [207].

Young adults with CHD recalled growing up in positive environments that may have contributed to positive attitudes towards physical activity. In addition, young adults described growing up with active siblings and parents who were viewed as active role-models. These examples draw upon the observational learning construct of the SCT and exemplify the environmental influence on behaviour within the SCT. Adults with CHD also reported a moderate self-efficacy for exercise and barriers to physical activities including work and school commitments, and lack of time and energy to be physically active. Furthermore, the reasons to be physically active changed from childhood to adulthood. This change may be attributed to new environments (e.g., moving away from home), personal factors (e.g., establishing new friendships), and increased self-

management of their CHD. These new experiences may require a new set of skills that had not been acquired during their childhood, which relate to the self-regulation construct of the SCT that is achieved through actions like self-monitoring, goal-setting, feedback on performance, self-reward, self-instruction, and enlistment of social supports [207].

Chen et al. applied a social-cognitive approach to understand the influences on personal beliefs about exercise and the effects of external environments on their exercise behaviour [353]. Patients with mild congenital heart disease (n=126; 12-18 years) across three sites in Taiwan completed questionnaires that collected data about personal beliefs about exercise, interpersonal influences on exercise, and the availability of environments for exercise. Patients were also asked to complete a 7-day exercise log. Results indicated that the most important factor in determining MVPA was personal exercise beliefs, namely perceived exercise self-efficacy. Personal exercise belief had a mediating effect on the relationship between interpersonal factors and MVPA, and peer influence on MVPA [353]. Therefore, this study supports future work that aims to improve personal beliefs (i.e., self-efficacy for exercise) and thus applying a SCT approach.

Our descriptive study provides preliminary insights into different areas that could inform the development of future research informed by the SCT. The application of the SCT to understand CHD physical activity requires additional research to further explore the personal, behavioural, and environmental factors that shape physical activity behaviour in this population. Investigating the relationship between these factors is necessary given the changes in survival of CHD patients and thus new challenges and opportunities that patients with CHD face today.

5.6 Limitations

The results of this study should be interpreted with the following limitations in mind. Firstly, we worked with a small sample size based on the qualitative methodology to end recruitment upon data saturation. This limited our ability to collect sufficient quantitative data for more in-depth analysis. Moreover, due to incomplete data from accelerometer and questionnaire assessments, the sample size for quantitative results was further reduced and may not be completely reflective of the emerging adult CHD population, limiting generalizability of findings. Patients were approached to participate in a study about physical activity perceptions and behaviours. This may have introduced recruitment bias towards patients that were already active and inclined to participate in a study regarding physical activity. While the cross-sectional study design successfully characterized the study population, the lack of a control group of healthy patients and longitudinal data collection limits the ability to comment on long-term trends in this population compared to otherwise healthy populations.

5.7 Conclusion

This study provides important insights into how emerging adults with CHD view physical activity. Patients reported an overall positive experience regarding the medical care they received as a child and recalled an upbringing similar to healthy peers in terms of physical activity. Family played a substantial role in helping patients feel supported to participate in physical activity, primarily through parental encouragement and sibling involvement in activity. Participation in activity as an adult with CHD shifted towards improvement in health rather than enjoyment as experienced in childhood. Patients also described increasing independence and associated challenges experienced as an adult including managing priorities like school, work, relationships, and self-care. These results suggest that childhood interventions that include

family-based interventions may improve physical activity participation in CHD patients that might be carried forward into adulthood.

Family-based physical activity interventions among the CHD population are safe, feasible, and demonstrate improvements in physical outcomes [146]. These interventions have been primarily investigated among young children with CHD, their siblings, and parents. The lifelong management of CHD patients should include age-appropriate interventions that encourage the patient to be physically active. It may be particularly important to develop interventions that help adolescents with CHD establish an active lifestyle early in life, as adolescence is an important time when key health behaviours, including physical activity, are established. One approach may be to help build intrinsic motivation to make changes in their life that that allow for more regular physical activity. This may be best accomplished using a behavioural intervention that is adapted to the uniqueness of the adolescent CHD population.

6 Rehabilitative exercise and activity clinical trial in congenital heart disease (REACT in CHD): a pilot study

6.1 Study Contributions

Adam McKillop, assisted by Dr. Brian McCrindle, conceptualized this study. Adam McKillop conducted participant recruitment and data collection. Jane Schneiderman, or a designated Clinical Exercise Physiologist identified by Jane Schneiderman, completed fitness assessments. Adam McKillop completed all MI sessions with participants. Alexander Di Biagio, a research volunteer, completed all MI session transcriptions for subsequent fidelity measure. Dr. Maya Obadia completed the MI fidelity assessment. Adam McKillop, with assistance by Sue Kanoatovas, a research volunteer, completed all data entry and cleaning. Adam McKillop, with assistance from Dr. Cedric Manlhiot, completed quantitative data analysis. Adam McKillop, with assistance from Dr. Brian McCrindle, completed data interpretation. The primary supervisor for this study was Dr. Brian McCrindle.

6.2 Introduction

Children with CHD may have reduced functional health status or impaired quality of life [354, 355] and experience a ‘burden of disease’ resulting in detrimental psychosocial effects that continue into adulthood [315]. It is important to encourage regular physical activity in CHD patients during their clinical care to help improve their physical and psychosocial health [93-95]. Physical activity and exercise recommendations are generally delivered using a prescriptive approach with limited patient engagement. Reduced physical activity in children with CHD may

be attributed to parental overprotection and lack of consistent activity advice provided by cardiologists and parents, resulting in uncertainties regarding safe and appropriate activities [9, 308]. Increasing the engagement of adolescent patients in conversations about physical activity participation is particularly important as they begin to develop increased self-management skills and independence as they enter adulthood.

Although measuring the exercise capacity of patients with CHD provides diagnostic and prognostic information to help inform exercise and physical activity recommendations, interventions to improve exercise capacity are resource intensive and with short-term benefits [34]. Evidence suggests that improvements in physical activity levels might be achieved with behavioural interventions that require fewer resources and could be easily incorporated into the clinical conversation. Motivational Interviewing (MI) is one approach that may be used to help adolescents change their physical activity behaviour. MI takes a non-judgmental, patient-centered approach to evoke, not impose, motivation aimed at resolving ambivalence [356-358]. MI began as a technique first described to help with substance abuse, but it has since expanded to address health behaviours, including diet and exercise, diabetes self-management, medication adherence, and oral hygiene. Despite the overall effectiveness of MI-based behavioural interventions to improve health behaviours in adults [239, 359], the application of MI in the pediatric patient population has been limited.

There is a need to address the reduced activity levels among young patients with CHD. MI is a promising behaviour counseling intervention to help adolescent CHD patients. This approach can be readily adopted into practice, administered by a variety of practitioners, and delivered individually, in a group, in person, or by telephone [358]. This study builds on existing evidence that supports the importance of regular physical activity among the CHD population.

This study sought to determine the feasibility and efficacy an adapted MI behavioural intervention to improve physical activity among adolescents with CHD.

Study Aims:

1. To determine the feasibility of an adapted MI behavioural intervention to improve physical activity behaviour in among adolescents with CHD.

2. To objectively measure physical activity pre- and post-intervention to determine the efficacy of the behavioural intervention to change physical activity levels (MVPA per day).

3. To determine changes in physical fitness and psychosocial outcomes following the intervention.

Hypothesis: Patients will agree to study randomization, complete all baseline assessments, accept the MI intervention (if randomized to the MI group), and complete the individualized exercise prescription independently.

Secondary hypothesis: Patients who receive the intervention will demonstrate improvements in daily physical activity (minutes of MVPA per day) and psychosocial measures (stage of change, self-efficacy for exercise, and quality of life) while fitness indicators will not change within or between groups.

6.3 Methods

Design and Setting

This study was a pilot randomized controlled trial of an adapted MI intervention for adolescents with CHD, devised to increase physical activity. It was 12-weeks with 2-arms (intervention and control). The Research Ethics Board at the Hospital for Sick Children approved this study.

Eligible adolescents with CHD followed at the Labatt Family Heart Centre at the Hospital for Sick Children (Toronto, Ontario) in the outpatient cardiology clinic were approached to participate. Information about this study was provided by mail or in person during the patient's outpatient follow-up appointment. Written informed consent was obtained.

Inclusion/Exclusion Criteria

Inclusion Criteria: Adolescents (13-17 years of age) with prior repair of CHD of any complexity and at least one year after their most recent open-heart surgery, were eligible to participate.

Exclusion Criteria: Patients with exercise contraindication/limitations as identified by the responsible cardiologist (i.e., history of arrhythmias, syncope, hypoxia, pulmonary hypertension), significant cognitive disorders that would limit the completion of questionnaires and full participation in the MI sessions, and other medical conditions that may influence physical activity participation were excluded.

Randomization

Included patients were randomly allocated to the Intervention (MI) or Control group. A random number generator was used to create blocks of 2 or 4 to complete 1:1 group randomization

within each block. Group assignment was concealed by sealing of tamperproof, consecutively numbered envelopes by a researcher remote from the investigators patients. Patients were assigned the next envelope in sequence.

Personalized Physical Activity and Exercise Prescription: A Certified Exercise Physiologist (Canadian Society of Exercise Physiology Certified Exercise Physiologist) prepared a 12-week, individualized exercise prescription for each patient based on their baseline physical fitness (see assessments below), self-reported current activity participation, and any information provided by the patients that may affect exercise prescription (i.e., sport participation, work schedule, religious commitments). The 12-week exercise prescription progressively increased in frequency, intensity and duration towards guideline-based physical activity recommendations for adolescents (60 minutes of daily MVPA)[100]. Consideration was given to the patient's built environment, previous exercise experience, exercise interests, and personal schedule. Patients received one telephone call after 4 weeks and one telephone call after 8 weeks by the exercise physiologist to evaluate the need for modification to the prescription. Patients allocated to the control group did not receive additional contact beyond these follow-up calls. The exercise physiologist was blinded to the group allocation of each patient.

MI Intervention: Patients randomly allocated to the MI group participated in adapted MI sessions to explore and resolve ambivalence regarding physical activity. The underlying conceptualization was to focus this intervention on aspects that were primarily behavioural and intrinsic; it was designed to be collaborative in delivery, and with counseling aimed at stages of change [323] and increasing exercise self-efficacy. Key MI principles were relied upon, including autonomy, empathy, reflective listening, summaries, and asking open-ended questions

[360]. The intervention also took into consideration the patient's life stage as an adolescent, and their growing independence and hence requirement for self-management skills.

The intervention consisted of bi-weekly sessions by telephone over the 12-week period (i.e., six sessions). Each session was an average of 15 minutes in duration. The first session aimed to build rapport with the patient, understand their current views about exercise and physical activity, their general self-reported activity level, and assess the importance of physical activity in their life. The confidence to change behaviour was assessed and decisional balance (i.e., advantages and disadvantages of making a change) was explored as appropriate. Subsequent sessions built on the previous session(s) with progression towards building a reasonable plan with the patient that would help improve their physical activity and exercise participation. Conversations also included a description of a "typical day" for patients to discuss potential times in the day to add or improve the quality (i.e., increased intensity) of their physical activity. Subsequent sessions aimed to check-in on the progress since the previous session, address any outstanding issues that were not fully addressed during the previous session, and generally explored new factors that may help the patient move towards change. Where appropriate and with patient permission, information was provided regarding exercise techniques or suggestions based on their progress with the exercise prescription. All telephone sessions were recorded with patient permission.

Intervention Delivery: The counsellor (A.M.) received training from the MI Network of Trainers (MINT) during a 2-weekend training course. Supervised practice sessions were completed following the training course, where individualized feedback was provided from MI trainers and peers. A trained MI instructor with the MINT reviewed 25% of the recordings to assess MI fidelity using the Motivational Interviewing Treatment Integrity (MITI) standards [323, 361].

Study Measures

All patients were invited to complete the following assessments at baseline and during their post-test visit.

Feasibility: First, to test the feasibility of delivering the intervention, the number of completed sessions was recorded. Moreover, to test whether the intervention had the desired conceptual impact, stage of change and self-efficacy were assessed. The *Readiness to Change* [323] questionnaire is a 4-item questionnaire validated in the pediatric and adult population, and was used to assess the patient's motivation to change physical activity behaviours. Scores range from 0-4 with scores categorized to stage. The *Self Efficacy Scale (SES) for Physical Activity* was administered, consisting of 8 items scored from 0-40 (maximum score 80, with higher scores denoting greater self-efficacy). It has been administered in children [362] and adolescents [7] with CHD with good psychometric performance.

Physical activity: Patients were provided an Actigraph wGT3X-Plus Triaxial Activity Monitor (Actilife, Pensacola, FL, USA). It has been shown to be valid and reliable using treadmill walking at known speed and a laboratory shaker [363]. The monitor was to be worn over the right hip during waking hours for 7 days (2-weekend days and 5 weekdays), except when bathing or swimming. The device was programmed for a 15-second sampling interval and stored the activity count per 60 seconds to memory at the end of each successive interval. Patients were provided with a log-book to record the dates and times they wore the monitor. Patients returned the accelerometer in a pre-paid envelop. Accelerometer data was downloaded to a computer using available software (ActiLife) to determine activity intensity levels using previously reported physical activity intensity cut-points and important data filtering considerations [107, 108, 364]. Accelerometer data was analyzed where a minimum of three days of data was

received (minimum wear time of 10 hours for a valid day). Physical activity intensity was categorized using previously reported cut-points (sedentary=0-100 counts per minute (cpm); light=101-2295 cpm; moderate=2296-4011 cpm; vigorous= ≥4012cpm). These cut-point conventions (Evenson et al. 2008) were selected based on the published recommendation by [190]. Minutes per day were averaged at each intensity and the primary endpoint was change in MVPA.

Physical Fitness Indicators: Anthropometric measures included standing height and body weight (to compute body mass index (BMI=kg/m^2), as well as waist circumference (average of two measurements taken at the narrowest point above the iliac crest) [365]. Aerobic fitness was assessed using the Modified Canadian Aerobic Fitness Test, whereby patients were asked to take alternating steps to a set cadence from audio cues to compute oxygen consumption; flexibility (sit-and-reach), muscular strength (grip strength), and muscular endurance (partial curl-up) were also assessed using standard protocols [365].

Quality of Life: A *Visual Analog Scale (VAS)* was administered to assess global quality of life. The anchors were 0 to represent 'worst possible quality of life' and continued to a value of 100 to represent 'best possible quality of life' [328]. Dimensions of quality of life were assessed using the *Pediatric Quality of Life Inventory (PedsQL™) Teen (13-18) Report* [366]. Scores for each of 4 domains (range 0-100) were used to calculate summary scores including physical, psychosocial, and total quality of life, with higher scores denoting greater quality of life.

Statistical Analysis

First, to test the first objective (feasibility of study intervention), the mean number of sessions in which the intervention group participated was computed. To compare pre-test characteristics between the MI and Control groups to test the randomization, the Fisher's exact test was used for

all categorical variables and Student's t-test assuming unequal variance between samples for continuous variables (Satterthwaite methods). Secondary outcomes (change in physical activity, physical fitness, and psychosocial factors) were analyzed using the change in physical activity as the dependent variables in individual linear regression models adjusted for group. All statistical analyses were performed using SAS v9.4 (SAS statistical software, Cary NC). A level of $p<0.05$ was considered statistically significant.

Sample Size: As per published recommendations for pilot studies, formal pre-study sample size and power calculations need not be completed [367, 368]. Rather, pilot study sample size should be adequate to estimate recruitment rates [368]. In order to estimate effect sizes from this study, a sample size of 15-20 patients per group was recommended [369]. Thus, this pilot study proceeded with the target subject numbers to recruit 20 adolescents to complete each intervention (n=40 total patients).

6.4 Results

Respondent Characteristics: Patient flow is shown in Figure 19, adapted from the Consolidated Standard of Reporting Trials (CONSORT) statement [370]. Of the 757 patients screened during the recruitment period (March 2013 to March 2015), 299 were eligible and subsequently contacted to participate. Of those 299 eligible patients contacted, 72/299 (9.5%) declined and 173/299 (22.8%) did not respond to phone calls and email communication, despite repeated follow-up attempts. Reasons for declining included time constraints, travel distance, perception that the patient was already active enough, self-reported restriction to physical activity, or a general lack of interest in participating in the study. Overall, 54 patients consented and were enrolled in the study. Of the 55 patients, seven (13.0%) patients did not respond to follow-up

phone calls to schedule their initial assessment, and three (5.6%) indicated they were unwilling to complete study assessments due to lack of time or they were no longer interested in participating. Therefore, a total of 44 (81.5%) patients were randomized: 20 to the MI group (45.4%) and 24 to the Control group (54.5%). Following randomization, three (15.0%) patients withdrew from the MI group due to lack of time to complete the study. At the baseline assessment, one patient (5%) in the MI group reported recent episodes of feeling light-headed, and therefore testing was not completed. This patient was not interested in continuing in the study and was subsequently withdrawn. There was also one (5%) patient from the MI group that could not be contacted to schedule the first assessment and was therefore lost to follow-up.

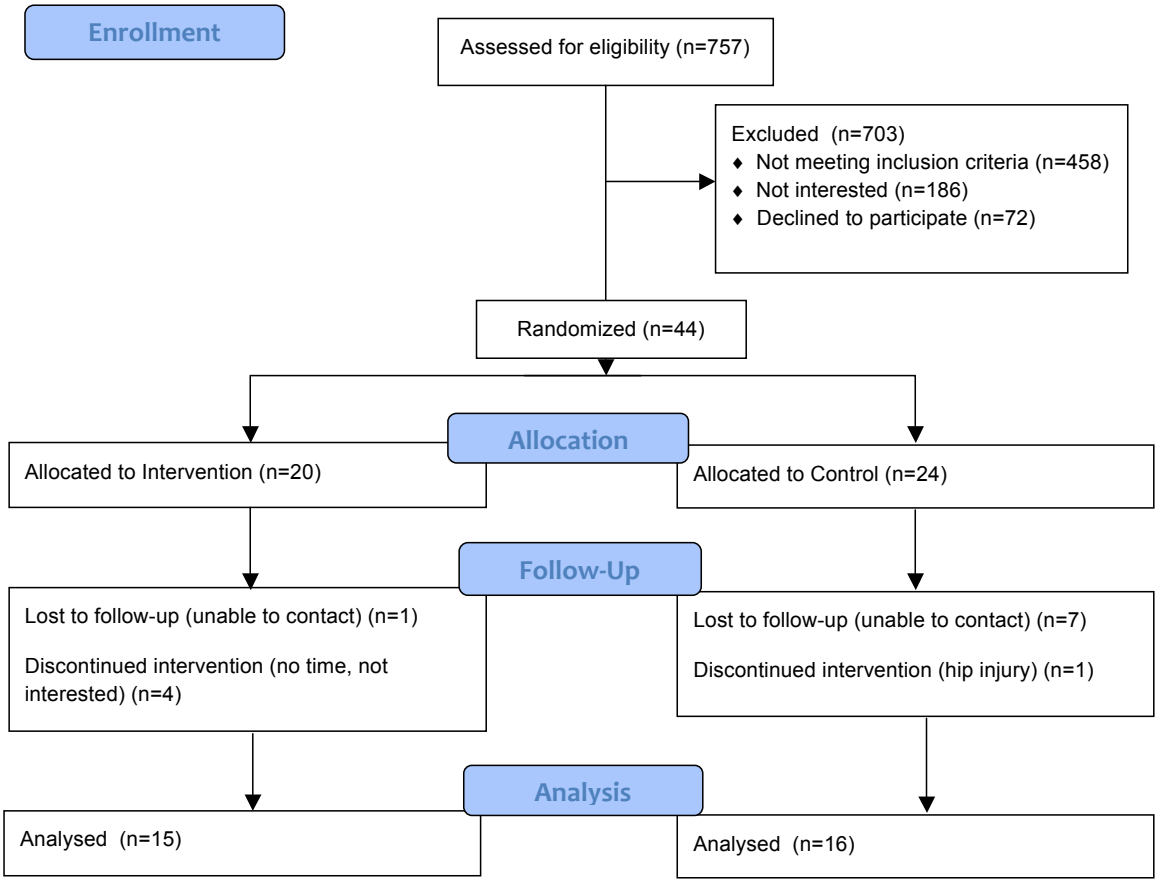

Figure 19. Patient flow diagram (adapted from CONSORT statement; Turner et al. 2012).

Table 14. Patient characteristics at baseline by group.			
	Control	**MI**	**p-value**
n	16	15	
Sex (male, n (%))	7 (44%)	10 (67%)	0.20
Age	15.0±1.5	15.1±1.5	0.81
Height (cm)	166.4±8.5	165.3±9.7	0.73
Weight (kg)	60.9±14.7	60.8±18.0	0.99

Intervention Feasibility: Patients in the MI group completed 4.2±1.2 sessions. Table 15 displays the self-efficacy and stage of change by time and group. There was no significant difference by group at pre-test (t=-1.43; p=0.16). The control group showed an increased stage at post-test (paired t=-2.55, p=0.02) whereas the MI group did not show an increase in stage (paired t=-0.56, p=0.58). There were no differences within group for self-efficacy for physical activity.

Table 15. Change in psychosocial indicators (self-efficacy and stage of change) by group and time.						
	Control			**MI**		
	PRE	POST	p	PRE	POST	p
SES for Physical Activity	29.7±4.6	30.3±3.5	0.67	31.1±4.7	30.3±5.7	0.65
Stage of Change						
Mean Stage of Change	4.0±1.1	4.8±0.5	0.02	4.5±0.9	4.7±1.0	0.58
Stage distribution						
1	0	0	-	0	0	-
2	2	0	-	1	1	-
3	4	1	-	1	0	-
4	2	1	-	2	0	-
5	8	14	-	11	14	-

Outcomes: Figure 20 displays mean MVPA by group and time, with mean scores for physical activity at each intensity reported in Table 16. At pre-test, there were no significant differences in physical activity between groups as expected, and no patients met the MVPA exercise guideline recommendations. Contrary to the secondary hypothesis, there were no significant

differences in the change in MVPA within the MI group (paired t=1.48; p=0.17) or Control Group (paired t=0.48, p=0.64). Change in physical activity for each participant in the Control and MI groups are shown in Figures 21 and 22, respectively.

Table 16. Patient physical activity results by group and time (mean±SD).

	CONTROL			MI		
Intensity	PRE	POST	p	PRE	POST	p
Sedentary	328.5±153.1	283.6±127.2	0.41	302.4±57.6	299.9±86	0.93
Light	73.1±39.0	70.5±37.0	0.86	74.6±23.4	65.7±31.9	0.42
Moderate	13.5±10.1	12.9±6.7	0.85	13.9±5.4	12.1±5.6	0.40
Vigorous	7.5±6.6	7.3±5.6	0.93	6.3±4.3	4.2±3.1	0.16
MVPA	21.1±15.4	20.2±13.5	0.87	20.1±8.9	16.2±8.3	0.25

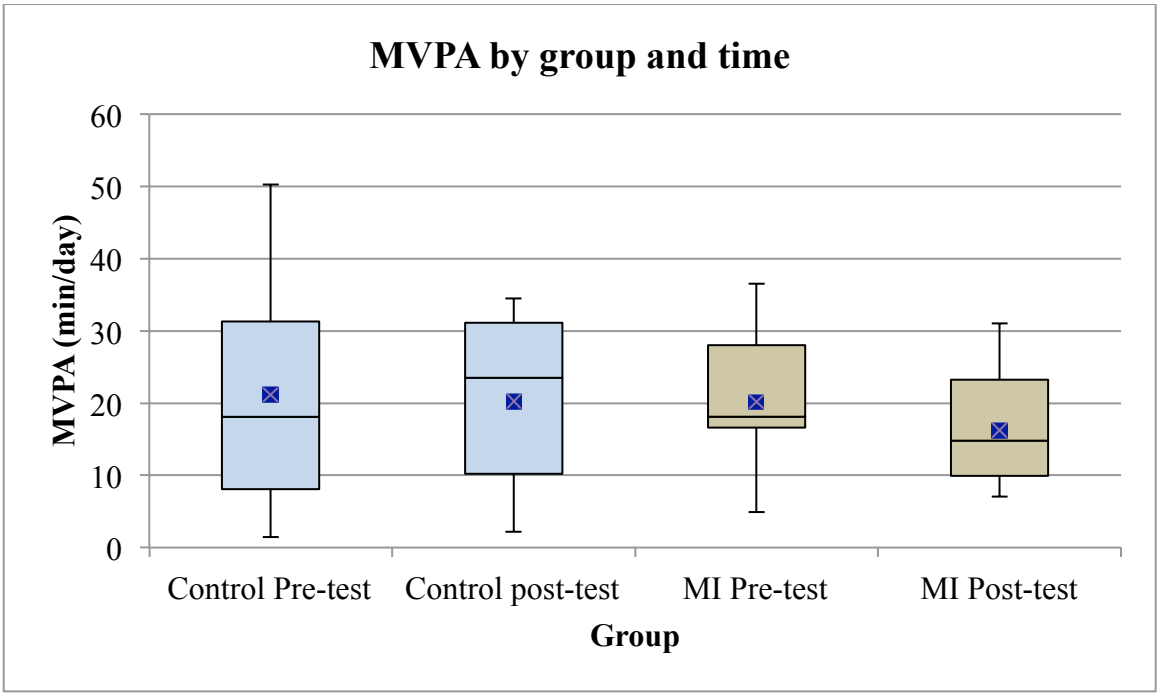

Figure 10. Change in moderate-to-vigorous physical activity (MVPA) between pre-test and post-test in Control and MI participants.

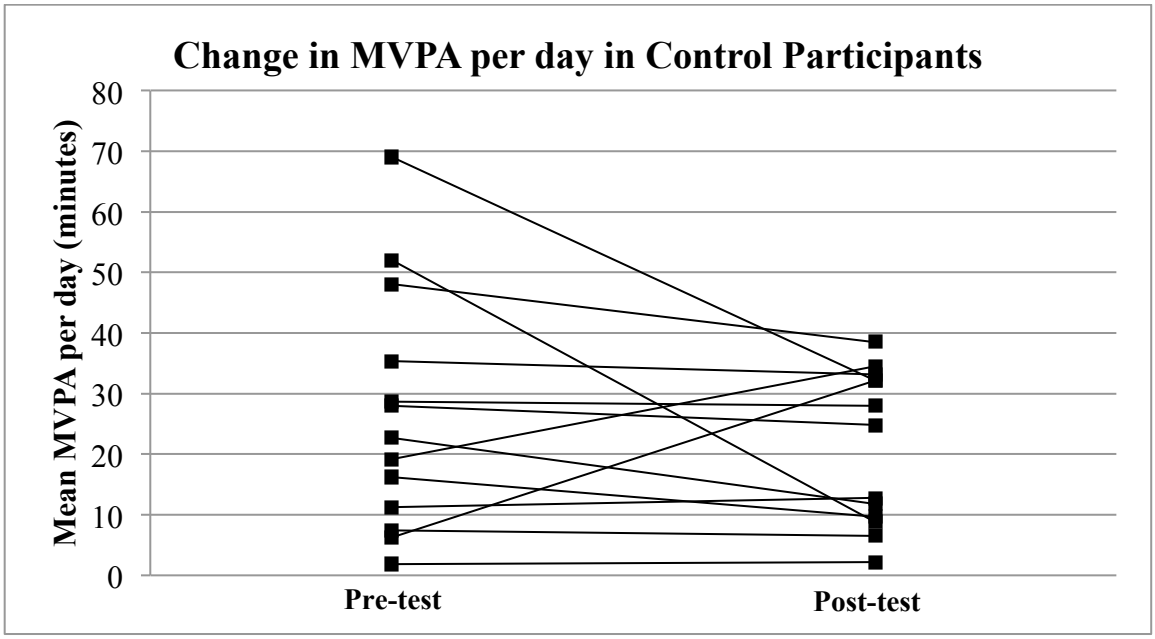

Figure 21. Change in physical activity between baseline to post-intervention in Control group patients. MVPA = moderate-to-vigorous physical activity (minutes per day).

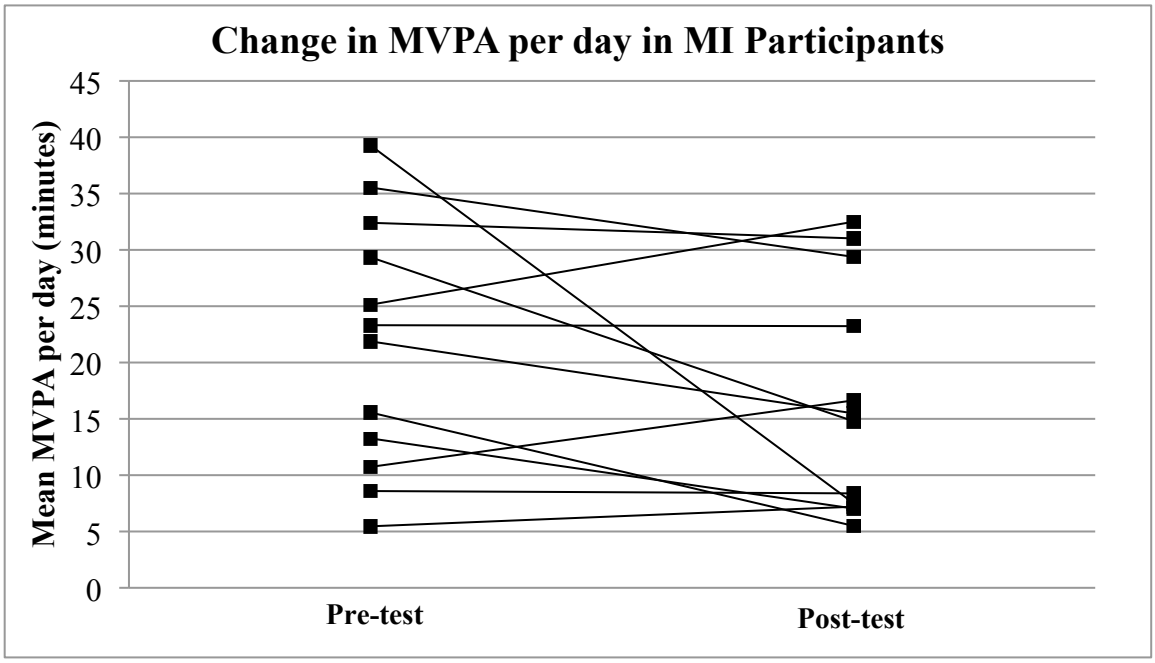

Figure 22. Change in physical activity between baseline to post-intervention in MI group patients. MVPA=moderate-to-vigorous physical activity (minutes per day).

Physical fitness indicators are reported in Table 17. Overall, six (20.0%) patients were considered overweight or obese (i.e., BMI >25 kg/m^2) at pre-test and 4 males (12.9%) were considered at increased or high health risk (i.e., men>94cm), while all females were considered at low health risk (i.e., women> 80cm) [365]. As shown, there were no significant differences in the change in any physical fitness indicator within groups. Compared to previously reported fitness indicators among other Canadian adolescents, patients demonstrated lower predicted O_2-consumption [144]. Measures of BMI, waist circumference, flexibility, muscular endurance, and grip strength were comparable between patients with CHD and healthy peers [144].

Table 17. Patient fitness results by group and time (mean±SD).

	CONTROL			**MI**		
Variable	PRE	POST	p	PRE	POST	p
BMI	21.9±5.2	21.9±5.4	0.99	22.1±5.1	22.0±4.8	0.98
Waist Circumference	74.8±13.1	74.2±13.3	0.91	74.9±12.7	74.7±11.6	0.96
O_2-consumption (ml/mg/min)	26.9±4.7	26.8±4.5	0.95	30.7±4.5	30.2±4.7	0.95
Flexibility (cm)	22.6±12.3	22.7±10.9	0.98	20.4±10.5	20.0±10.8	0.98
Muscular Endurance (#)	22.1±6.0	20.1±8.8	0.49	22.4±4.6	22.7±3.3	0.49
Grip Strength (lbs)	61.1±12.6	64.7±14.3	0.47	67.6±19.8	69.1±18.6	0.47

Patients with CHD reported favourable quality of life. There were no significant differences in the changes within or between groups in scores (Table 18).

Table 18. Patient psychosocial outcomes by group and time (mean±SD).

	CONTROL			**MI**		
Variable	PRE	POST	p	PRE	POST	p
Quality of Life	74.9±14.2	74.9±13.1	0.99	79.3±12.7	77.6±11.4	0.72
PedsQL						
Dimension Scores						
Emotional	67.8±22.1	72.2±16.1	0.52	82.3±11.9	81.7±16.3	0.90
Social	89.1±14.0	87.2±10.3	0.67	87.7±10.0	85.3±13.2	0.59
School	74.1±12.7	73.1±13.6	0.84	75.3±10.8	73.0±14.7	0.62
Physical	81.0±9.2	81.6±9.0	0.86	84.2±11.2	84.2±11.2	1.00
Summary Scores						
Psychological	77.0±13.1	77.5±5	0.90	81.8±8.0	80.0±11.3	0.62
Physical	81.0±9.2	81.6±9.0	0.86	84.2±11.2	84.2±11.2	1.00
Total	78.0±11.3	78.5±9.9	0.89	82.4±7.8	81.0±10.4	0.69

MI Fidelity: A review of 25% (n=28) of all available audio recordings of MI sessions identified that MI principles were adhered to 55% of the time throughout the reviewed sessions. According to establish MI fidelity guidelines, this score indicated a below-competency level of MI.

6.5 Discussion

It is well established that physical activity and exercise can improve psychosocial well-being for healthy individuals and those with chronic disease, including CHD [93-95]. Adolescents with CHD are known to have reduced physical activity compared to healthy peers and are at increased risk of developing modifiable co-morbidities. We sought to determine the feasibility of an adapted MI behavioural intervention to improve physical activity among adolescents with CHD.

Study Feasibility: This study highlighted feasibility challenges including recruitment and retention, as well as intervention administration and acceptance. The recruitment process included reviewing the medical records of patients scheduled for an out-patient follow-up appointment to determine eligibility. A total of 299 patients were eligible to participate (from 757 screened patients) and subsequently contacted, with only 54 patients consenting to participate (18.1% enrolment rate). Retention was also a challenge, such that 23 of the 54 patients either withdrew from the study (n=8) or were unable to be contacted for follow-up (n=15), representing a 42.6% attrition rate following study enrolment. Overall, 31 (10.4%) patients of the 299 eligible completed the study assessments. The study intervention was designed to limit the amount of in-person interaction that required patients to attend the hospital, including study assessments that corresponding with scheduled clinic visits (if possible), telephone-based MI sessions, and sending accelerometers by mail. However, patients in the MI group completed 25% less sessions than originally planned. Furthermore, the incentive for

participation in this study was a record of volunteer hours that could be applied to their secondary-school volunteer requirements, and reimbursement for parking. Increasing the incentive may have helped patients remain enrolled and engaged in the study (i.e., gift certificate for participating, entered in draw to win a prize).

Physical activity among adolescents with CHD: This study identified that adolescents with CHD did not achieve the national physical activity recommendation to achieve 60-minutes of moderate to vigorous physical activity every day. The average minutes of MVPA per day of patients with CHD in this study were approximately 30 minutes lower per day compared to published values of healthy children [108]. This may be attributed to the different accelerometer data analysis conventions (i.e., devices used, cut-points) applied to each sample. In addition, patients with CHD demonstrated reduced physical activity levels despite reporting favourable psychosocial outcomes, including self-efficacy to exercise and quality of life. Reports of patients with CHD having comparable psychosocial outcomes with healthy cohorts exist, and in some cases higher reports of quality of life and self-efficacy [15, 126, 163, 371]. Dulfer et al. investigated the association between exercise capacity, physical activity, and psychosocial outcomes among children and adolescents with CHD [15]. Higher exercise capacity was generally associated with greater physical quality of life but seldom associated with psychosocial domains of quality of life [15]. This review identified two studies that measured physical activity (self-reported) that indicated greater activity was associated with greater self-efficacy. Overall, the work by Dulfer et al. indicated that greater exercise capacity was associated with greater physical quality of life but not psychosocial quality of life.

Change in physical activity following an adapted MI intervention: Patients who received the adapted MI intervention accumulated less physical activity between pre- and post-test

(statistically insignificant) while control group patients appeared to maintain their physical activity over the 12-week test period. This finding contradicts the hypothesized increase in physical activity among patients who received the MI intervention.

The failure of the adapted MI intervention could be attributed to several factors, including insufficient dose, the adaptation of MI, intervention delivery via telephone rather than in-person, or characteristics of the counsellor. When considered in relation to other interventions in this area, this is the first reported study to use an adapted MI intervention among adolescents with CHD to improve physical activity. An MI-based intervention was used by Morrison et al. in a cohort of adolescents with CHD and found an improvement in daily physical activity and exercise capacity [16]. The intervention was in the form of a single exercise session informed by MI principles to educate patients about exercise and activity. Following this session, each patient received a personalized exercise plan over a 6-month period. The MI-based intervention described by Morrison et al. differed significantly from our intervention (group vs. individual, in person vs. telephone, single session vs. multiple sessions, respectively), which raises methodological differences that make it difficult to compare the MI treatment effect with our study [16, 372]. In addition, a lack of detail regarding the MI-specific components of the intervention by Morrison et al. makes it difficult to understand intervention similarities. Furthermore, the work by Morrison et al. did not report fidelity measures, and as indicated in our work, this is a crucial piece to MI interventions to evaluate the application of MI principles.

McCarthy et al. reported a process evaluation of an exercise intervention using MI among adults with heart failure [372, 373]. This included details regarding the recruitment and retention of patients, implementation of the MI intervention, and assessment of reach to the target population. The implementation measures included fidelity, dose, and context of the study intervention.

During the recruitment period, 20% (n=4) of the 20 audio-recorded tapes were evaluated by an expert in MI to determine the level of adherence to MI principles. Our study conducted the fidelity assessment upon completion of all MI sessions. The fidelity assessment for our study was based on 25% of the recordings, resulting in 28 recordings (compared to 4 records reviewed in the work by McCarthy et al.). From the four recordings reviewed for MI fidelity, the first two recordings received a score of 40 and 50% MI-adherent [372]. These scores are similar to the MI-adherence in our study (55%). The similar scores between studies may have been expected given that our study did not complete fidelity measures until the end of the study and therefore did not modify the administration of the MI intervention that may have resulted in improved MI-adherence. Feedback from the fidelity assessment in the study by McCarthy et al. resulted in revisions to the MI exercise-counselling guide to improve questioning techniques and communication [372, 373]. Therefore, earlier fidelity assessment and opportunity to modify the MI intervention through an iterative process may have improved the MI-adherence outcome in our study. However, the application of MI to the adolescent CHD population remains a novel approach to improve health behaviour and there remains a need for additional research to determine the clinical utility of MI among pediatric patients [374]. Furthermore, a more in-depth process evaluation of a MI intervention among the pediatric population may identify important aspects of the MI intervention to improve research study designs and potential clinical interventions.

Integrating prescriptive and behavioural components: Exercise training interventions administered in the CHD population have focused on improvements in exercise capacity with few reports on physical activity outcomes. Tikkanen et al. identified 18 studies with a structured exercise-training component for pediatric patients (up to age 18) with CHD [8]. Overall, exercise-training studies among patients with CHD presented mixed results in terms of

improvements in exercise capacity, physical activity, and psychosocial outcomes. None of the studies included in this review included behaviour change approach to improve physical activity. Physical activity was an outcome measure in two studies [23, 301]. Rhodes et al. assessed physical activity with a self-report questionnaire regarding time spent in different activities, while Fredriksen et al. used an accelerometer to objectively measure physical activity levels of patients. Following exercise training, two studies reported sustained benefits with respect to exercise capacity and physical fitness but did not report sustained benefits in physical activity participation [19, 29]. We did not conduct follow-up with patients beyond the final post-intervention assessment; however, a follow-up study may determine the long-term effects of our intervention.

Work completed by Longmuir et al. reported the benefits of a home-based rehabilitation program for children with CHD, including motor-skill development, improvements in physical activity, gross motor skill, exercise capacity, and fitness among children with Fontan circulation [149]. This work not only highlights the feasibility of a home-based pediatric cardiac rehabilitation program, but also speaks to the importance of considering a more comprehensive rehabilitation program for children that includes physical activity and play-based activities. Similar work should be investigated with home-based exercise and activity training programs among children with various CHD diagnoses (not only Fontan) and including adolescents with CHD. Furthermore, integrating components of the work by Longmuir et al. (family-based activity and education about activity), exercise prescription and exercise training, with behaviour-change strategies as used in this study, may determine any cumulative effect of such interventions. A multi-component intervention that also includes patients with a range of CHD diagnoses, physical abilities, and psychosocial outcomes may identify factors associated with success in the program (i.e., do some patients respond differently to exercise training/prescription, while others

benefit more from behaviour change). This may be used to inform the development of a pediatric cardiac rehabilitation program that tailors the intervention to best suit the patient based on each individual patient.

MI is a promising behaviour change technique that is supported by the American Heart Association in order to build intrinsic motivation among CHD patients [118, 375]. A review of existing studies that used MI to improve health behaviours (diet and exercise, diabetes, oral health) indicated an overall benefit of MI compared to usual care [373]. However, existing reviews on this topic did not include any studies including patients with CHD and more research is needed to identify the feasibility and efficacy of MI to improve health outcomes in CHD patients.

6.6 Limitations

The results of this study are reported in light of the following limitations. Our study was limited to a single institution with a heterogeneous cohort of patients with a range of CHD diagnoses from simple to great complexity. Therefore, we are unable to delineate disease-specific effects of the intervention. This study included an adapted MI behavioural intervention to improve physical activity. However, sessions did not adhere with MI principles and were below competency level to be considered MI. This may be a result of limited training working directly with the adolescent CHD population, and without additional audit-feedback during the study to adjust and improve the quality of MI delivered. This limits the ability to state that the intervention was a "true" MI intervention and speaks to the need for ongoing training, audits, and booster sessions in order to maintain a high level of MI fidelity throughout the study period [372]. Patients were not blinded to the study intervention and this may have contributed to differential treatment within each group or modified behaviour by the patient following randomization [376].

Furthermore, data analysis did not apply an intention-to-treat approach given the small sample size and high attrition in each group. Future studies with sufficiently large sample sizes should apply the intention-to-treat analysis to maintain the prognostic balance and sample size of each group following randomization [377]. In addition, retention in the study groups was low and thus limits the overall feasibility of this intervention and further limited the ability to ascertain the overall effect of the adapted MI intervention to improve physical activity between groups.

6.7 Conclusions

In conclusion, the adapted MI intervention was not sufficiently engaging for adolescents with CHD. The intervention did not have a measurable impact on the stage of change, self-efficacy for exercise. Moreover, the intervention had no effect on physical activity, physical fitness, or quality of life. Studies that investigate the use of behavioural interventions to improve physical activity among youth, especially those with CHD, are limited and more novel approaches to promote physical activity among the adolescent CHD population are needed.

7 Discussion

This thesis described the long-term physical activity trends of patients with complex CHD between childhood and adulthood, explored the physical activity behaviours and perceptions of emerging adults with CHD, and examined the use of a behavioural intervention to help improve physical activity behaviour among adolescents with CHD. The following discussion will describe how this research adds to the existing evidence in the area of physical activity and exercise in CHD patients, outline limitations in this research, identify areas for future research opportunities, and propose a pediatric cardiac rehab model for consideration.

7.1 Physical activity promotion in CHD: more work to be done

Given the chronic nature of CHD and risk of developing co-morbidities, especially in complex CHD patients, physical activity should be discussed during outpatient follow-up visits to facilitate a positive change. Early adoption of a healthy, active lifestyle during childhood may continue into adulthood and provide long-term health benefits. Therefore, it is important to identify optimal physical activity interventions to help children and adolescents establish positive health behaviours.

The delivery of information about physical activity in the clinical setting is largely provided in a didactic manner (i.e., patients are told what to do). In response, this prescriptive approach may be met with resistance from patients accompanied by a limited understanding and motivation to change their behaviour. In addition, clinical visits may already have a negative association for patients and be considered a burden, overwhelming, or intimidating. Furthermore, the healthcare provider (e.g., cardiologist, nurse) who delivers this information may be considered to be in a

position of authority or power, further limiting the acceptance of the information and increasing the likelihood of resistance to this information. A key consideration is patient engagement during clinical visits with respect to the uptake and comprehension of physical activity and exercise information. This is especially important provided that children view physical activity as being fun and do not necessarily associate physical activity with improved health until later in life. The physical activity counseling paradigm should be re-evaluated such that information is presented in a format that limits a prescriptive approach, where the patient feels more engaged with the information, and the necessary resources are known and made available to the health care team during physical activity counseling. The overarching tone should be one that encourages physical activity rather than a medical treatment or requirement.

Many CHD patients are able to perform various types of physical activity and are considered unrestricted from activity participation. Patients that are free from activity restrictions should be encouraged to participate in physical activity on a regular basis to prevent the development of co-morbidities. The current approach in the clinical setting continues to focus on restriction of activities and often falls short of adequate promotion of activities that are suitable for the patients. This approach is perpetuated by published physical activity and exercise recommendations for CHD patients that outline activity restrictions, with limited activity promotion strategies offered to help guide clinical practice [9]. Some CHD patients, particularly those with complex CHD, do require specific activity and exercise precautions in order to reduce the risk of a cardiac event or injury. While it is important to clearly outline physical activity limitations for patients in order to communicate important risk-reduction information, patients should also be provided information about safe and appropriate activities.

Physical activity promotion should be integral to the clinical care of CHD patients and be supported with details with respect to frequency, intensity, time, and type of activity. This approach, although embedded in an exercise-prescription approach, offers patients and families information to base decisions about activity participation. Unfortunately, clinicians remain uncertain about the details of physical activity and exercise recommendations, resulting in poor documentation and knowledge translation for patients and families [4, 349]. More detailed information for patients regarding specific activities or exercise regimens often fall outside the scope of treating clinicians and may require the expertise of an exercise specialist. For example, a Certified Clinical Exercise Physiologist brings a depth of understanding of exercise science and physiology that can be applied across many different clinical conditions, including CHD. This resource should be made available to patients requesting additional information about physical activity and exercise, even if considered unrestricted and able to participate at the level of the current physical activity recommendations. There is a need to bridge the gap between published physical activity guidelines and leverage available resources to help clinicians, patients, and families provide the best possible information and care with respect to physical activity and exercise recommendations.

7.2 Physical activity levels among CHD patients: could be worse but could be better

The reported physical activity level of patients with CHD varies throughout the literature. Some studies reported physical activity levels among CHD patients that are lower than healthy peers [121, 122, 128], while others reported physical activity levels above their peers and, in fact, met or exceeded current physical activity recommendations [17, 131]. Part of this variability could be

explained by the methods used to assess physical activity, specifically using self-reported measures that tend to over-estimate activity levels. Furthermore, reports of low physical activity among the CHD population may be derived from a cohort of complex CHD patients and inappropriately generalized to include all CHD cohorts. Duppen et al. reported that most exercise training studies include a wide range of CHD diagnoses, many with small sample sizes that limit sub-group analysis [34]. Therefore, more research is needed that includes larger sample size to identify differences in physical activity levels between CHD diagnoses.

The physical activity level of adolescents and adults with CHD in our work was below that of non-CHD and otherwise healthy children and adults reported in the CHMS. More specifically, the mean MVPA per day among Fontan survivors collected during the Fontan 1 Activity Study (childhood) was 39±28 minutes of MVPA per day. This is approximately 13 minutes of MVPA per day less than children without CHD as reported in the CHMS [372]. These results also indicate that adolescents and adults with CHD in our study did not meet current physical activity recommendations. This is in contrast to a recent study by Duppen et al., which reported that 70% of patients (10-25 years) met the current physical activity guidelines (60 minutes of MVPA per day) [17]. In our REACT in CHD study, one patient (control group) accumulated >60minutes of MVPA per day at baseline, which dropped to 32 minutes of MVPA per day at follow-up. Only three ACHD patients accumulated over 30 minutes of MVPA per day.

Recent reports have indicated sufficient physical activity levels among children with CHD may be due to improvements in the clinical management of CHD patients, thus providing CHD patients with improved functional outcomes. Despite some earlier reports of clinicians providing unnecessary restriction due to a lack of published recommendations, guidelines are now available to help clinicians deliver physical activity and exercise counseling. Physical activity

may be more recognized and accepted by clinicians as a means to improve health, as newer evidence has been made available to support this notion. However, the evaluation of the uptake and integration of published guidelines into clinical practice requires additional research.

The patients included in our studies accumulated sub-optimal physical activity levels and less physical activity than peers without CHD, despite reporting favourable psychosocial outcomes including quality of life, health-related quality of life, self-efficacy for exercise, and cardiac anxiety. This may be related to the exclusion criteria used in our studies, which excluded patients with significant cognitive or physical impairments that would preclude full participation in the study and completion of all study assessments. This may have resulted in recruiting a homogenous sample of patients with respect to favourable psychosocial factors. However, one may expect that the positive psychosocial outcomes as reported by our patients may contribute to more minutes of physical activity. Therefore, the lower physical activity levels displayed by patients with CHD in this thesis research cannot be explained by corresponding poor psychosocial outcomes, and there appear to be other contributing factors.

Potential contributing factors to explain poor physical activity levels of CHD patients:

There are likely multiple factors related to the reduced physical activity level demonstrated by our patients. Firstly, in the REACT in CHD study, telephone MI sessions with adolescents with CHD identified that school commitments and time management were perceived as barriers to physical activity participation. This was more apparent during the telephone calls that occurred during mid-term or exam periods (large work load and pressure to succeed) versus at the beginning of a school semester (less school work, projects, exams). Patients considered success in school very important and often placed school commitments as a top priority. These findings agree with previous work among adolescents who identified internal factors (individual

characteristics, low-priority for physical activity, and use of technology) and external factors (influence of peers/family, lack of time, and inaccessibility and cost of facilities) [378, 379].

Some patients also described challenges balancing extra-curricular activities with schoolwork and, thus, having even less time to devote to increasing their physical activity. It was clear through these conversations that patients still considered physical activity as a long-duration, planned, structured exercise session (i.e., playing a sport, going to the gym). Very few patients considered opportunities to improve their physical activity through other means of changing their activities of daily living. Patients stated that a rigid school schedule limited time for physical activity participation between classes or during lunch hour. The after-school period was identified as the best opportunity to increase activity. Patients described a greater likelihood to be active during the after-school period, including walking home from school, adding some additional time/distance to the walk home, or completing an exercise-session at home with bodyweight exercises or exercise equipment (free weights, aerobic machines, exercise videos). We did not specifically evaluate the amount of time patients spent in active pursuits during the after school period. This would be a worthwhile area to explore and would provide more detailed information about the opportunities to be active for patients.

Patients described challenges being active with their friends who were mostly inactive and preferred sedentary activities (i.e., video games, watching TV, going to the movies). Although patients described the benefits of being active and expressed a desire to be more active, they found it difficult to be active with friends that did not want to be active. Patients described most of their leisure time as spending time with friends and found it challenging to find independent leisure time to be physically active. This was partly due to the importance placed on spending time with friends, which seemed to outweigh the importance placed on being physically active.

In only a few patients was an active friend group described, where the leisure time spent with friends included some form of physical activity (i.e., riding bikes, playing sports).

The preference to socialize with friends is expected, as adolescence is an important time to establish peer acceptance and good peer relationships that help with identify formation [125]. Although peer socialization is important, adolescents with CHD should be provided with strategies to maintain a healthy active lifestyle while maintaining relationships with peers. This is particularly important in adolescents, as youth with CHD may not be comfortable disclosing their CHD diagnosis with peers and may require additional support to learn about CHD-specific activities [315].

Social Cognitive Theory as it applies to physical activity behaviour: The social context in which physical activity opportunities exist is an important consideration when trying to counsel patients about physical activity. Findings from this thesis work can be related to Bandura's Social Cognitive Theory, which outlines the behavioural, cognitive, and environmental factors that contribute to one's behaviour [352]. This thesis research can be represented in a social cognitive theory context given the various examples linked to each of the three components of this theory (Figure 23).

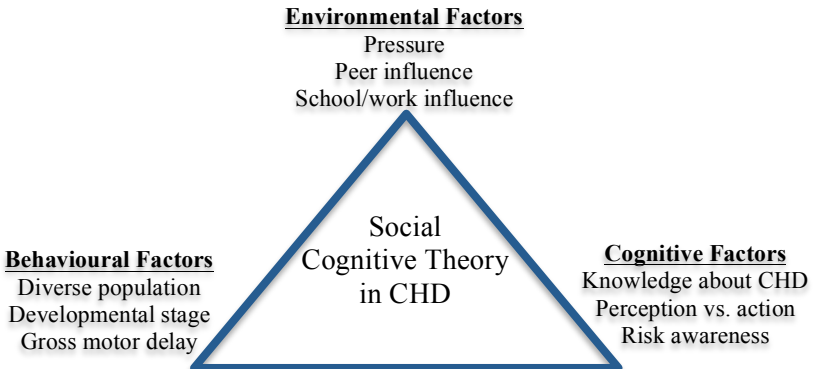

Figure 23. Application of the Social Cognitive Theory to describe the physical activity behaviour among the CHD population.

The social contributors that influence decision-making and behaviour change must be considered when applying physical activity interventions into the lives of patients with CHD. The role of friends and peers is important when counseling patients about physical activity, including the potential negative influences that drive patients towards a more sedentary lifestyle or positive scenarios that facilitate activity participation. It is also important to work with the individual to help them build the independence to be physically active in spite of challenging negative social situations and societal norms that may hinder activity participation (i.e., strategies to improve one's self-efficacy and building intrinsic motivation). Perhaps more relevant is the necessary coaching to identify opportunities to be physically active while differentiating between habitual physical activity and a structured exercise training session.

Patients included in the ACHD study described positive perceptions towards physical activity participation growing up. Childhood physical activity was described as an opportunity to spend time with friends and have fun rather than improve health. There were few perceived barriers to activity as a child, including illness, injury, or surgery. As the ACHD patients described their current perception of physical activity, barriers to be physically active included school (post-secondary) commitments, work schedules and demands, and a perception that the activity accumulated at work was sufficient to maintain a healthy lifestyle. Furthermore, some patients placed a low importance on physical activity given that they were of normal weight and did not have other risk factors that would indicate a need to improve their activity level (i.e., obesity, high cholesterol, high blood pressure). In contrast to being active for primarily recreational purposes in childhood, physical activity was primarily considered a means to improve their health in adulthood. This was attributed to becoming increasingly aware of their condition, the severity of the condition if not managed, and the importance of physical activity to prevent co-morbidities. Learning more about the underlying CHD diagnosis at a younger age was discussed

and perceived as beneficial, as patients indicated they may have been more aware of the future implications as they entered adulthood. While patients described positive experiences and recalled some form of physical activity counseling in the care they received during childhood and adolescence, it is during adulthood that patients begin to place an increased importance on the knowledge of their condition. This is likely a result of developing greater independence and associated independent decision making that accompanies emerging adulthood.

The physical activity levels of patients with CHD may be associated with factors present in their daily lives. A more comprehensive evaluation of factors should include school, peers/social, vocational, and family interactions that may influence activity participation. These factors are likely to have changed over time and present new challenges to patients' lives (i.e., patients with CHD from 10 years ago experienced a different life than CHD patients today), particularly during adolescence as mentioned in the REACT in CHD study. These factors remain understudied with respect to how they impact physical activity levels in the CHD population and may provide new insights to develop strategies to improve patient outcomes.

7.3 Psychosocial outcomes in CHD patients

Psychosocial outcomes, mainly quality of life, health-related quality of life, and self-efficacy, have been reported as lower in the CHD population compared to those without CHD. One of the limitations in this area of research is the lack of consensus regarding outcome measures, including psychosocial assessments. It is important to not only understand the psychosocial factors related to CHD but also explore the relationship between physical (i.e., exercise capacity, physical activity) and psychosocial outcomes [15]. Defining clinically relevant psychosocial assessments in future research will help guide the integration of psychosocial interventions into

clinical practice for a more comprehensive management of patients with CHD beyond the physical aspects of CHD care.

Overall, patients in this thesis research reported favourable psychosocial outcomes, including quality of life, self-efficacy for exercise, and cardiac anxiety. This may have been a result of limiting recruitment to patients that did not exhibit significant physical or psychological impairments. Patients may have also been more interested in participating in the studies simply by having a positive attitude towards physical activity and exercise. While there resulted indicated an overall positive psychosocial profile, there was a wide range of psychosocial responses across these thesis studies. This demonstrates the diversity of the CHD population and need to consider the unique needs of the individual patient. The wide range of physical activity levels and psychosocial outcomes also supports individualized physical activity and exercise counseling strategies rather than applying a single approach for all CHD patients. By considering the individual as unique rather than a standard set of recommendations, the patient can receive an appropriate physical activity and exercise plan to fit their lifestyle.

7.4 MI to improve physical activity levels in CHD patients

This thesis work included the use of an adapted MI behavioural intervention to help patients with CHD improve their daily physical activity level. Only one other study reported the use of an intervention that used MI principles among adolescents with CHD [16]. Therefore, the REACT in CHD study in this thesis is one of few studies that used a one-on-one adapted MI intervention among adolescents with CHD to improve their physical activity behaviour.

The adapted MI intervention was assessed to be 55% adherent with MI principles (90% adherent would be considered beginner proficiency in MI) [372]. The low MI adherence rate is testament to the necessary training and continuous audit of sessions to ensure the person administering the MI is meeting the standards. Although the MI provider (A.M.) received MI training and completed supervised MI sessions with feedback by MI trainers prior to study commencement, the sessions completed during the REACT in CHD intervention were unsupervised and did not undergo audit-feedback exercises. Oversight of the intervention by an MI trainer should have been implemented throughout the intervention to help ensure MI principles were met. In addition, regular audit and feedback sessions with an MI trainer may have prevented deviations from MI principles in subsequent sessions and resulted in a truly representative MI intervention.

Feasibility considerations of a behavioural intervention for adolescents with CHD:

The REACT in CHD study administered a phone-based, adapted MI behavioural intervention for adolescents with CHD aimed to help improve their physical activity behaviour. This research identified important feasibility considerations and raised questions about this approach as being accepted by youth with CHD. Telephone calls were chosen as a means to communicate with the patient for each session, proposed as bi-weekly sessions to occur over the 12-week intervention period (up to six sessions). All patients accepted a telephone-based intervention and no patients expressed concern or opposition to speaking with study personnel on the telephone. However, scheduling of telephone calls for this cohort was challenging during a 12-week period. Challenges included finding a time that was suitable for the patient that would not disrupt their school day and using a phone-number that was easily accessible during this time. The challenges faced in this study resulted in only 4/6 proposed sessions on average. Email communication was also relied upon for some patients to help schedule telephone sessions, as this was the preferred

medium of communication for patients. Despite best efforts to follow-up with patients and confirm previously scheduled calls, patients were often unavailable and the session was delayed. Reasons for missing a scheduled call included illness, vacation, or simply no answer. As a result, the time between sessions varied among the patients, including three to four weeks between sessions in some cases due to scheduling challenges. Therefore, a telephone-based intervention may not be appropriate in the adolescent CHD population and alternative communication techniques should be explored (e.g., text messaging, online interventions, or gamification) to improve the engagement of patients in the behaviour change process. Face-to-face MI sessions are commonly used but present additional feasibility issues related to travelling to a specified location, especially among adolescents [217, 380]; however, a face-to-face approach should be explored with trained community partners to limit long-distance travel and encourage session completion.

7.5 Perceptions of physical activity: family, health, and the future

Emerging adults with CHD described their perceptions of physical activity in the past, present, and future. Interviews were used to explore patient perceptions and identified the importance of family to help support and encourage activity participation during childhood. Adults with CHD were able to explicitly recall the positive role modeling of parents and siblings with respect to physical activity as a child. The positive role of family indicated that patients recalled supportive parents and did not describe an over-protective childhood. This finding is in contrast to previous reports that described over-protective parenting styles experienced by patients with CHD[158, 381]. This may be due to the limited observation of over-protecting parenting styles by patients as children. For example, patients may not be aware of conversations that parents have with teachers/coaches that may prevent their child's full participation. Similarly, patients may be

unaware of opportunities that their parents did not present to them during childhood, which may have limited the patient's knowledge of an over-protective upbringing. Patients also confirmed that physical activity limitations experienced during childhood were imposed by the cardiologist and stated that restrictions placed on activity by parents were simply being implemented by parents. Therefore, even if the parent did place limitations on the patient's activity, this was viewed more as an enforcement of limitations from the cardiologist rather than over-protective parents.

The positive influence of family on the patient's perceptions about physical activity should be highlighted during clinical appointments and included in conversations about physical activity. This may be particularly important for parents who are inactive and, thus, providing a potentially negative role-model for patients. Inactive family units should be considered for group physical activity counseling and provided family-based interventions to help parents adopt healthy, active lifestyles that may positively influence their child's activity. Families that are active should be acknowledged and encouraged to continue living an active lifestyle as a family, as this not only helps the patient but also beneficial for everyone in the family. Family members should be considered assets in providing recommendations and family-based interventions. This approach should be integrated into a family-based and patient-centric care model to help patients improve their physical activity.

ACHD patients perceived that physical activity was a means to maintain their health, whereas having fun and being with friends were the main reasons reported for being active in childhood. Adolescents with CHD also discussed the importance of spending time with friends during their leisure time, often in sedentary pursuits. As a result, patients found it difficult to find additional time to be physically active. Despite adolescents with CHD stating that physical activity is

important to be healthy, conversations with adolescents with CHD did not directly explore the importance of physical activity in the context of health in the same way it was discussed during interviews with ACHD patients. The importance of staying healthy as a more prominent reason to be active as an adult may be attributed to the greater understanding of the disease and potential risk factors of living an inactive, unhealthy life. Patients may have also been presented information differently by their ACHD healthcare team compared to their visits during their appointments in a pediatric setting when information is typically delivered to parents. The delivery of information from an ACHD clinician may be more matter-of-fact and directed towards the patient rather than parents who may otherwise shield or deflect mention of disease severity or poor outcomes. Clinical visits during adolescence should include conversations about the severity of the CHD and emphasize that building a strong foundation of healthy behaviours during childhood is important for life as an adult with CHD.

7.6 Health Belief Model: a theoretical framework to help CHD patients improve physical activity behaviour

Overall, more work is needed to understand how behavioural interventions may be applied to the CHD patient population to help improve physical activity levels. More specifically, investigations into the application of behaviour change theories (i.e., Health Belief Model, Social Cognitive Theory, Transtheoretical Model) that inform behavioural interventions are needed. Behavioural interventions have been used widely across clinical populations and can be tailored to meet the needs of diverse patient groups [382-384].

Structuring a physical activity intervention guided by an overarching health behaviour change model may facilitate the assessment of an individual's behaviour and the underlying

psychological processes that drive behaviour [385]. A health behaviour change model that informs a physical activity intervention depends on social, psychological, and environmental factors to [386]; however, definitive evidence that supports the use of behavioural interventions in the CHD population is lacking.

This thesis research identified important social, psychological, environmental factors among the CHD population that could be better described in the context of an existing health behaviour change mode. For example, the Health Belief Model (HBM) was originally described in the 1950's as a theoretical model of health behaviour change related to preventing, controlling, or screening for illness [207]. The HBM has been applied in cancer screening, HIV prevention and testing, dietary compliance, and physical activity participation to prevent cardiovascular disease [387]. There are six main constructs (perceived susceptibility, severity, benefits, barriers, and self-efficacy, and cues to action) that comprise the HBM, along with individual factors that may influence steps taken to make a change. The HBM has not been reported as a theoretical framework to guide physical activity interventions among the CHD population but has been applied to other chronic diseases. The use of the HBM to help CHD patients improve their lifestyle in the context of preventive cardiology may provide a framework to base interventions and clinical strategies to discuss physical activity and exercise with patients. Each construct from the HBM may be linked with physical activity and exercise counseling in CHD patients (Table 19).

Table 19. The application of the Health Belief Model (HBM) to physical activity counseling in CHD.

HBM Construct	Application for physical activity counseling for CHD patients
Perceived Susceptibility	Patients should be advised on current and future risks of sedentary lifestyle behaviour in the context of their CHD diagnosis. A description and/or comparison of populations may help contextualize the risk of getting disease but direct relationships should be drawn between perceived susceptibility and the individual's physical activity behaviour.
Perceived Severity	Patients should be made aware of potential complications and interventions. Physical activity should be encouraged to help patients maintain a healthy lifestyle that matches the future outlook for patients should future complications or interventions result in activity precautions.
Perceived Benefits	The benefits of physical activity should be highlighted for patients, including both mental and physical benefits. Clinicians should help patients explore benefits not only in the present state, but also with respect to long-term outcomes as the patient enters adulthood.
Perceived Barriers	Patients should be asked to describe potential barriers to physical activity. Clinicians should address any misperceptions regarding self-imposed activity precautions and counsel patients accordingly to help reduce or remove these barriers.
Self-efficacy	A patient-reported, validated self-efficacy assessment may be used to evaluate the patient's self-efficacy for physical activity. Clinicians should work with the patients to build confidence to overcome potential barriers that may prevent changing their physical activity behaviour.
Cues to Action	The clinical presentation of patients (i.e., increased weight, poor diet, trouble sleeping) may necessitate a change towards improving physical activity behaviour. Clinicians should identify such indications to help the patient recognize other cues in their life that may point towards making a change. A testimonial from a prominent public figure with CHD may also motivate others to take action.

It is important to note that perceptions outlined are those of the patients and that the focus is on prevention rather than treatment. Furthermore, the HBM posits that an individual's motivation to make a change can be deconstructed into three categories: individual perceptions, modifying behaviors, and likelihood of action [207]. Therefore, the intrinsic motivation that MI attempts to elicit may align with the intrinsic perceptions included in the HBM that contribute to behaviour change (perceived threat, susceptibility, benefits, and barriers). One technique used in MI is to explore decisional balance, including the pros (benefits) and cons (barriers) to change. This technique may be applied in a behaviour change intervention guided by the HBM to provide context to the conversation with patients. Another central MI technique is reflective listening, which may be useful to confirm the patient's perceptions during a conversation [372]. In addition, MI may complement the HBM by working towards changing a specific behaviour that may reduce the susceptibility of the disease.

The HBM may be more applicable for adults with CHD as these patients identified the susceptibility and severity of their disease and commented on the importance of physical activity to maintain their health. In addition, cues to action in the ACHD population tended to occur, as information from the cardiologist was more direct and made the patients more aware of the potential for re-intervention or future complications. However, one may argue that the HBM may in fact be more appropriately applied in the adolescent population to help patients recognize susceptibility and severity of the problem if they do not improve their physical activity behaviour. In addition, it is during adolescence when physical activity and exercise behaviours are developed and, thus, interventions to improve activity should focus on adolescents [306].

The delivery method of interventions guided by theory should also be examined to determine an approach accepted by patients. Artinian et al. provided an extensive overview of interventions to

improve physical activity and diet behaviour to prevent cardiovascular disease among adults [375]. Behaviour change was discussed in this scientific review and cognitive-behavioural counseling recommendations were provided to help counsel individuals. Strategies to facilitate behaviour change included goal setting, self-monitoring, frequency and duration of provider contact, feedback/reinforcement, self-efficacy enhancement, incentives, modeling, problem solving, relapse prevention, and MI. Artinian et al. also discussed intervention delivery models, including individual, group, computer/technology, and multi-component interventions.

Group interventions generally result in increased social interaction, support from other individual facing similar challenges, role modeling, and positive observational learning. The format of group interventions typically includes didactic education, counseling and behaivour change strategies (i.e., goal setting). Skill building and practice may also be included in group-based interventions [375].

Individual interventions provide an opportunity for more tailored or personalized counseling of recommendations in the context of the patient's life. Most successful individual interventions to improve physical activity behaviour include a health risk appraisal, activity counseling, and/or cognitive-behavioural strategies. Individual interventions may be delivered in-person, by telephone, electronically, or using combined methods.

Technology or internet-based interventions are relatively new and rapidly expanding methods to deliver behavioural interventions[388]. Advantages of using this method include increased reach of the intervention, easy storage of information, ease of updating information, provide personalized feedback, cost effectiveness and convenience for users, access to populations in isolation to help reduce stigma or feeling embarrassed, timeliness, user control and autonomy, supplier control of intervention content and delivery, and adapting information for specific

populations [375]. Although results of technology- or internet-based methods to improve behaviour are encouraging, there may be limitations to some populations (low-income, minority populations). In addition, internet- or computer-based interventions may not be appropriate to facilitate physical activity because the user may be stationary when receiving the intervention. Mobile devices may be more useful to deliver a behavioural intervention for users so as to encourage physical activity and exercise.

Patients with CHD may experience unique challenges and could benefit from behaviour change interventions across the lifespan. Longitudinal studies are also necessary to identify optimal strategies offered throughout childhood, adolescence, and adulthood to ensure age and development-appropriate interventions are provided. The above behavioural intervention methods should be applied and evaluated to identify the needs of patients as they progress towards adulthood.

7.7 Use of technology to deliver interventions for CHD patients

The use of new technology (i.e., mobile apps, wearable technology, and telemedicine) was not used in this thesis work. The telephone-based behavioural intervention among adolescents with CHD was a new concept that has not been reported previously; however, this approach may not be a suitable or appropriate form of communication for a relatively young patient population. Telephone calls may be an outdated mode to administer a behavioural intervention for youth, while short messaging service (texting) or video calls (Skype) may be preferred forms of communication. In addition, online interventions that include gamification or the use of an avatar to administer the intervention may improve acceptance of a behavioural intervention rather than the typical patient-provider model used in the REACT in CHD study [389, 390].

When asked, "*What role has technology played in your activity participation? How could technology help you become more active?",* ACHD patients did not recall using technology to support their physical activity or exercise growing up during adolescents or currently as a young adult. In fact, patients generally described technology as being a negative influence on physical activity and contributing to the sedentary lifestyles observed in today's society (i.e., watching television and playing video games contribute to poor lifestyle habits). Given the young ACHD population included 18-25 year olds, it may be unlikely that technology to help support physical activity and exercise was readily available during adolescence like it is today. In addition, technology to support CHD patients is lacking and integration into the clinical setting is even more limited. Patients did not completely discount the potential use of new technology to help patients with CHD be active; however, more work in this area is needed to help design an appropriate technology to meet the needs of CHD patients.

7.8 Physical activity and exercise counseling for CHD patients

This thesis research identified important considerations regarding physical activity counseling of patients during adolescence and emerging adulthood. The adapted MI intervention used in the REACT in CHD study included conversations that explored and identified potential barriers to change and reasons to be more active as perceived by the patients. Individual interviews with ACHD patients described experiences during childhood and provided retrospective insights into physical activity counseling preferences during childhood and as an adult.

The ability to keep-up with one's peers is commonly relied upon as a gross indication of the patient's functional status. An inability to keep-up with peers is regarded as a functional limitation for the patient. Given the data presented in recent reports on sub-optimal activity

levels of the general population and comparable activity levels between patients with CHD and those without CHD, this raises the question of the usefulness of peer comparison as a proxy of functional status. Furthermore, as indicated by qualitative research in this area, young patients self-report a comparison to normal peers and often describe a desire to "be normal" [134]. In essence, by asking patients questions like, "Are you able to keep up with your friends or the other kids?" asks the patient to examine their life from a non-normal, or potentially sub-optimal, perspective. Furthermore, this method also asks patients to consider their peers as a benchmark to strive towards, yet research has shown that the general population falls well below the national daily physical activity guidelines [108].

The overall approach to physical activity counseling should be re-evaluated to better align with the changing demographic of CHD patients and their peers. Conversations about physical activity and exercise should be an integral component of all follow-up appointments and physical activity should be encouraged for all members of the family. Resources to help facilitate the conversation should be readily available for the healthcare team and in turn provided to patients and families if requested and/or deemed appropriate and useful. The patient should be considered unique and be provided with detailed physical activity and exercise recommendations that are not only appropriate given their clinical presentation but also align with the patient's goals and expectations. Delivery of the physical activity information should be encouraging and focus on promotion rather than restriction. The option to generate a specific exercise prescription with a Certified Clinical Exercise Physiologist should be made available to patients.

7.9 Towards a model of pediatric cardiac rehabilitation: where do we go from here?

Children and young adults with CHD should participate in regular physical activity throughout their life. Research continues to support physical activity and exercise interventions to help facilitate the adoption of a healthy, active lifestyle for the CHD population. Despite published physical activity and exercise training guidelines for pediatric and adult CHD patients, few clinical programs exists. Formal pediatric cardiac rehab programs are lacking and often limited to research settings. Over the last few decades, emerging evidence has provided encouraging results to support pediatric cardiac rehab but remain inconclusive due to limited studies with small samples, non-randomized control trial designs, and inconsistent outcomes measures between studies.

Research has focused on young children with CHD with few studies evaluating outcomes in adolescent CHD patients and fewer still among adults with CHD. Furthermore, results from previous studies describe the short-term effect of an intervention while the long-term effects are unknown. These interventions have typically focused on physical exercise training using a prescriptive approach and reported changes in aerobic capacity, muscle strength, cardiac function, and blood pressure.

The psychosocial aspect of pediatric CHD care remains understudied. Regular physical activity and exercise has been shown to reduce psychological distress symptoms, improve self-efficacy, and improve quality of life. Research to explore the relationship between physical activity and psychosocial outcomes in the context of a pediatric cardiac rehab model is required to identify how physical activity and exercise facilitated effects psychosocial outcomes.

7.10 Conceptual model to inform intervention development

This thesis research, along with the evidence from existing literature, was used to conceptualize future interventions for patients with CHD (Figure 24). CHD is a chronic condition and necessitates a life-long approach to manage patients with respect to their physical activity behaviour. This diagram outlines considerations in childhood, adolescence, emerging adulthood, and adulthood, as indicated at the bottom of the diagram including current physical activity recommendation (minutes of MVPA/day). The main factors thought to influence the patient's physical activity participation, including physical, psychosocial, and environmental factors, are identified in middle of the diagram. The thin dotted line presents parental involvement in the care of CHD patients and how this changes between each stage (particularly from childhood to emerging adulthood). In addition, the thick dashed line shows the increased uptake and acceptance of technology (i.e., mHealth) throughout each stage. In general, physical activity interventions should aim to address factors listed in the middle section of the diagram.

The current physical activity and exercise-training paradigm may require changes to match the needs of patients, particularly where parental involvement and technology intersect during adolescence. This may be an important and perhaps optimal time to re-frame the delivery of information to improve patient knowledge about the condition, foster independence, and engage the patient in their care. Leveraging available technology that interests young patients with CHD may help facilitate more independent action towards improving their physical activity behaviour that can continue into adulthood. In addition, according to this thesis research, the role of parents may shift towards one of encouragement and support and not necessarily be viewed as over-protective, particularly with increased knowledge about their child's condition and clarity around activity precautions.

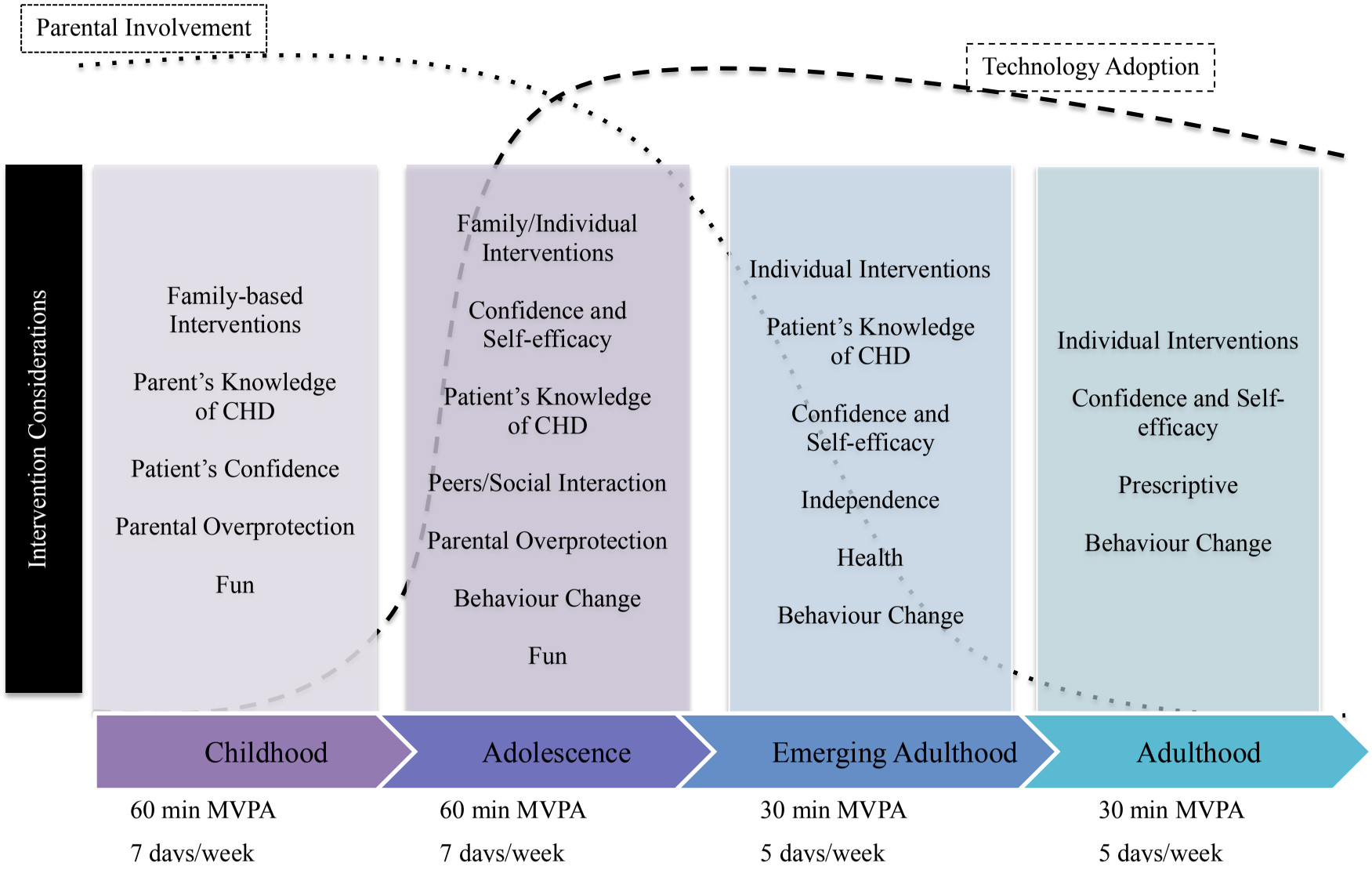

Figure 24. Conceptual diagram to demonstrate various factors that influence physical activity participation among patients with CHD between childhood and adulthood

8 Synthesis

8.1 Pediatric Cardiac Rehabilitation: steps in the right direction

Formal programs to help pediatric patients manage their CHD throughout the lifespan and meet the needs of patients surviving beyond adolescence and into adulthood are limited. It is important to address poor lifestyle behaviours in childhood to prevent detrimental patterns that may continue into adulthood. Furthermore, engaging patients during childhood may improve their knowledge about their CHD and emphasize the importance of maintaining a healthy, active lifestyle.

Structured pediatric exercise training programs have shown improvements in the physical fitness and psychosocial outcomes of patients with CHD [34]. Programs studied to date have generally been short-term (12-weeks) and limited to an exercise training (prescriptive) approach that asks patients to adhere to a set regimen. While cardiac rehab programs include an exercise-training element that remains central to the program model, these programs are now multi-disciplinary and include professionals to counsel nutrition, psychosocial issues, and medication. This comprehensive approach is needed to address the multiple dimensions involved in the patient's life and functional status. The following section proposes a pediatric cardiac rehabilitation program for patients with CHD and the various components as informed by this thesis research and the existing literature on the topic.

8.2 A Pediatric Cardiac Rehab Program for CHD Patients

The existing literature regarding pediatric cardiac rehab and CHD exercise training along with this thesis work can be used to inform the development of a formal pediatric cardiac rehab program. It is important to note that the following proposed program is focused on pediatric patients with CHD and should not be considered an adaptation of an adult-based model of cardiac rehab for children. This program acknowledges that pediatric CHD patients are unique and therefore require a corresponding cardiac rehab program developed to meet the evolving needs of this patient population.

8.2.1 Referral Guidelines:

Referral to a pediatric cardiac rehab program for patients with CHD should not mimic the work done in the adult-based model, as patients with CHD do not experience the same "event" that necessitates referral to a program. A pediatric cardiac rehab program should be considered a primary prevention strategy to reduce the impact of cardiovascular disease risk factors commonly associated with CHD. Furthermore, a pediatric cardiac rehab program could also serve to support patients and families with respect to physical activity and exercise training, along with other lifestyle modifications. The program could also support pediatric patients with acquired cardiovascular disease risk factors (i.e., obesity, diabetes, high cholesterol), those who experienced a cardiovascular or cerebrovascular event (i.e., pediatric stroke or heart attack), or underwent cardiac re-intervention (i.e., surgery, procedure, cardiac device implantation). The complexity and chronic course of CHD from birth and varying severity of the disease provides a strong rationale for a formal program to support patients and families with respect to physical activity and exercise training.

Children with CHD should be referred to a pediatric cardiac rehab program if the patient:

1. Describes their daily physical activity as being less than the recommended physical activity level of 60 minutes per day at moderate-to-vigorous intensity.
2. Demonstrates significantly impaired exercise capacity (<75% predicted) following a standardized clinical exercise test.
3. Reports self-limiting physical activity behaviour and/or parents report unnecessarily enforcing physical activity limitations.
4. Is classified as over-weight or obese based on pediatric BMI or has a waist to height ratio of greater than 50%.
5. Has other cardiovascular disease risk factors including high cholesterol, diabetes, high blood pressure that may be managed with lifestyle modification including physical activity and exercise training and nutrition counseling.

8.2.2 Intake Assessment:

Following referral to a pediatric cardiac rehab program, the patient should undergo a set of standardized assessments to document their current physical and psychosocial status. The main purpose of these assessments should be to identify priority areas to be addressed in the program. The assessments should include self-reported information provided by both the patient and parents. The parents should provide separate proxy reports if the patient is unable to respond due to cognitive limitations.

Physical Assessments: A number of physical fitness assessments have been applied among CHD patients as well as in healthy populations [34, 391]. The following includes a collection of highly relied upon assessments that provide sufficient information to base an exercise plan. The list is intentionally not exhaustive but allows for a comprehensive assessment of different physical parameters worth tracking in a program.

1. **Cardiopulmonary Exercise Test:** A standardized cardiopulmonary exercise test should be completed as a gold standard clinical assessment to measure the patient's response to maximal exercise [11]. This exercise test will also identify any important exercise-induced arrhythmias that may necessitate precautions in the program and subsequent activity and exercise recommendations. Most patients will undergo a clinical exercise test throughout their CHD care. An exercise test may be requested if the patient experienced a cardiac event or intervention, or experiences substantial changes in their physical functioning throughout the program.

2. **Functional Fitness Assessment:** A set of standardized fitness assessments will provide additional information about the overall fitness of patients with CHD. These tests should include assessments of muscular strength (hand grip), muscular endurance (partial curl-up), flexibility (sit-and-reach), and aerobic step test (modified Canadian Aerobic Fitness Test; mCAFT)[144] [134] [134] . Additional assessments that have been used in the CHD population include the 6-minute walk test that may serve as an alternative to the mCAFT and requires limited resources to administer [150]. Muscular power may also be assessed using a vertical jump test [392]; however, this has not been reported in the CHD population and requires further investigation as to the utility of this type of assessment. These fitness assessments are important to inform an individualized, structured exercise training program that aligns with the patient's individual fitness goals while accounting for their current fitness level.

3. **Body Composition:** Basic anthropometry should be assessed for all patients including height, weight, BMI, waist circumference, and waist-to-height ratio. The use of skin-fold

assessments may provide a low-cost option to measure percent body fat and fat-free (lean) muscle mass if experienced and trained personnel are available. If available, bioelectrical impedance analysis (BIA) or BodPod analysis may also be used to measure body composition. BIA offers an indirect method to determine body composition by measuring the body's resistance to an electrical current. This measure results in values for total body water, fat free mass, and fat mass [393]. The BodPod uses air displacement plethysmography to determine the volume of air displaced by the patients to determine total body volume. A series of equations that include variables like height, weight, and sex determine body composition including percent body fat and fat-free mass [394]. Collecting body composition data for patients provides additional insight to gauge program progress while accounting for growth and development. An accurate and non-judgmental explanation of body composition data should be provided to patients to avoid and/or address potential body image concerns.

4. **Physical Activity:** Accelerometers are widely used in the clinical population to collect objective measures of physical activity levels [172, 189, 395]. Accelerometers should be used to objectively measure physical activity levels among CHD patients. This approach will avoid overestimation of time spent physically active compared to self-report methods to measure physical activity. Accelerometer assessment of physical activity should be completed over a 7-day period during waking hours using a tri-axial accelerometer. A minimum of four wear-days (including at least one weekend day) and at least 10 hours of wear-time per day should be included for analysis. A logbook, or some form of tracking the wear-time of the device, should be included during the assessment period to verify days that the monitor was worn and to identify any discrepancies with wear-time

measured by the device. This will help identify any compliance issues and inform the decision for re-assessment should the requirements not be met. Detailed and clear instructions should be provided both verbally (with demonstration of use) and in writing to the patient to help reduce improper use [151]. Compliance to the accelerometer protocol may also be enhanced by providing monetary compensation, keeping a daily journal, frequent phone calls (three during monitoring period), and including a control condition [396].

Nutritional Assessment: Patients with CHD may experience malnutrition and sub-optimal nutritional status prior to and immediately following surgical repair [397, 398]. Long-term post-operative nutritional status should be assessed to understand baseline nutritional status and food consumption record that can guide nutritional counseling. A nutritional assessment is also important to ensure patients are receiving adequate and high-quality energy supply before beginning an exercise training intervention. Patients should complete a food frequency questionnaire to document food intake [399]. A Registered Dietitian should review this assessment and indicate if a more in-depth nutritional counseling or dietary intervention is warranted prior to starting the cardiac rehab program. If an intervention is advised, the dietitian should continue to work with the patient and family to improve the nutritional status of the patient throughout the cardiac rehab program.

Psychosocial Assessments: The psychosocial well-being of patients with CHD should be evaluated during the intake assessment to identify any immediate need for additional resources to help the patient/family. Secondly these assessments will inform the development of an individualized program that meets the current state of patients, as well as offer a baseline

comparison for patients to track progress and improvements throughout the program. The following assessments should be included in the intake of new patients:

1. **Readiness to Change:** Patients should be asked to report their current stage of change (pre-contemplation, contemplation, preparation, action, or maintenance) with respect to physical activity behaviour [400]. Classifying patients into one of these stages will help inform future counseling approaches that aim to move patient towards action to make a change. The individualized program should account for their current stage of change and behavioural change interventions should be used to help patients identify and resolve any ambivalence to change. Marcus et al. provide a 4-item assessment tool to measures readiness to change physical activity that indicated the current patient's current stage.

2. **Self-efficacy for Physical Activity and Exercise:** Self-efficacy is a core construct to many behavioural change theories and counseling approaches. Identifying one's self-efficacy to be physically active will help inform behaviour change techniques that may focus on methods to improve self-efficacy (i.e., vicarious learning, mastery, social interaction). Self-efficacy can be measured using the Self-efficacy for Exercise Scale developed by Dishman et al., and used previously in young patients with CHD (10-14 years old) by Ray and Henry [126]. This is a short 8-item assessment, anchored from 1 (disagree a lot) to 5 (agree a lot). Higher scores indicate a greater confidence the patient can be physically active.

3. **Knowledge of activity and risk assessment:** Each patient and family may demonstrate varying knowledge of the CHD and associated activity recommendations and potential risk of activity. An assessment to determine the current level of understanding about

activity recommendations will help inform the depth of information required by patients and families. A simple method similar to one used by Kendall et al. may be sufficient to provide an appropriate level of detail to inform recommendations while being easy to administer and follow-up with families [4].

Consultation with Certified Clinical Exercise Physiologist: Each patient referred to the cardiac rehab program should undergo a one-on-one consultation with a Certified Clinical Exercise Physiologist to review the results of the intake assessments. This consultation should occur separate from the clinical assessment and occur in an environment that encourages activity and limits exposure to the typical clinical settings. The consultation should occur both as a family and individually with the patient. A family-based consultation should include a discussion about parental and family activity behaviour and identify the potential to foster a family-based model that includes improvements in physical activity and exercise behaviour. This will allow the physiologist to learn more about the patient, family, and their environment to optimize future activity recommendations. Furthermore, community and online resources should be shared with the family when available and/or requested.

An individual consultation with the patient should be completed to discuss goals and identify potential barriers that may impede success. This approach will also help reduce the parental influence on patient responses. A one-on-one consultation with the patient may also identify new considerations independent from those discussed with parents. An individualized activity and exercise plan should be developed with the patient that aligns with their goals and accounts for their current physical and psychosocial status from the intake assessments.

8.2.3 Program Delivery Options

Reports of pediatric cardiac rehab programs have largely been facility-based programs that focus on supervised exercise training sessions. These programs have also been largely research-based programs and published reports do not describe a resulting clinical program integrated within the clinical management of patients with CHD. Evidence indicates that exercise training administered in a facility-based setting does improve physical activity and cardiorespiratory fitness of patients with CHD. This work has generally been short term with only few reports of long-term (1-5 years) follow-up. Changes in physical activity and psychosocial outcomes have also been reported but with large variability and inconsistency in terms of a training effect on these outcomes.

A recent Cochrane systematic review of adult cardiac rehab programs revealed that no differences exist between home-based and institution-based programs in terms of mortality, cardiac events, or exercise capacity [401]. Home-based programs foster a natural environment for patients to engage in exercise and activities of daily living [402]. In addition, patients in cardiac rehabilitation programs have indicated that travel time and distance to hospital-based programs were major drawbacks compared to home-based programs [263]. This limitation is further compounded by the inability of some adolescents and young adults to travel independently to institution-based settings. This finding supports the imperative to consider home-based programs for cardiac patients. Unfortunately, this review did not include pediatric cardiac CHD programs. Existing research using a home-based cardiac rehab model for pediatric patients with CHD is encouraging and should be pursued in the development of a pediatric cardiac rehab program. Given the available evidence to support both facility and home-based models from adult- and pediatric-exercise training programs, a pediatric cardiac rehab program

should be made available to patients throughout their care while being followed at a pediatric institution. This program should meet the unique needs of young patients and families by offering facility- and home-based components that complement each other rather than providing two independent delivery models.

Facility component: The intake assessment should be completed at a facility that is equipped and staffed with trained personnel to administer all necessary assessments. The facility should have adequate space to administer all physical assessments, particular the submaximal fitness assessment. A standardized cardiopulmonary exercise test should be administered if one is not available from a recent clinical assessment that may have prompted referral to the program. A cardiopulmonary exercise test should be repeated if the date of the last test was six months ago or more, or if the patient described a significant change in their fitness and/or physical activity behaviour.

The consultation with the CEP should also occur at the facility following the intake assessment. During this assessment period, the CEP and patient/family should discuss program delivery options that best suit their needs, availability, and ability. A facility-based program option should be made available for the patient/family if they require substantial oversight to learn new movements/exercises. Facility-based programming should focus on building the necessary skills, and refining techniques while improving self-efficacy. This delivery option should not be developed as a long-term option for patients and families but rather provide the fundamentals for patients and families to execute the recommendations independently.

Patients should be encouraged to return to the facility for a supervised training session within 1-2 weeks of the intake assessment. This first one-on-one training session with the CEP should introduce the patient to the proposed training plan. The patient should receive an overview of the

proposed exercises and modify the plan where necessary. Proper self-monitoring techniques and general safety and use of any exercise equipment should also be reviewed. All exercise equipment should be provided to the patient to continue with the training program at home (i.e., exercise bands, balls, free-weights, skipping ropes, etc). At least one additional one-on-one training session should be scheduled two weeks after the introductory session to review the plan and progress since their first session. This session may be done at the facility, or conducted remotely using available technology with video capabilities. Following this second training session, the CEP should determine if a third training session is needed to continue working on specific exercises or address any other deficiencies in the program. If a third session is not warranted, the CEP should ensure the patient is prepared with all necessary resources and equipment to continue with the training plan at home.

The CEP should help the patient incorporate exercise-training sessions into their routine. This is a particularly challenging task, especially for adolescents with additional academic commitments. Time management tools and strategies may be relied upon to facilitate a discussion with patients, including available technologies or behaviour change techniques. This discussion may also involve parents if some of the barriers to completing the program could be removed with increased parental support (i.e., drop-off/pick-up from gym).

Home component: The patient should begin a home-based training program following approval from a cardiologist who reviewed the intake assessments results and approved the exercise plan as outlined by the CEP. The facility-based training should provide the patients with the necessary skills to complete the training program independently at home and/or a facility of their choice (i.e., community centre, local gym, school). A home-based program should reduce the time and travel to a facility program while providing a familiar and perhaps preferred environment for the

patient to complete the training program. The training program should continue to be overseen by the CEP with bi-weekly to monthly progress check-ins. The CEP may recommend the patient return to the facility for a re-assessment or to observe the patient during a training session to correct techniques or modify the program.

Communication with the CEP should be available throughout the training program. This may include telephone, email communication, text-messaging, or video-calls. The use of video calls may help the CEP observe the patient at home during a training session to review a certain exercise or movement. Improvements in the available technology and remote monitoring capabilities may also allow the CEP to assess other parameters, including heart rate, respiratory rate, oxygen saturation, or rate of perceived exertion. A remote-monitoring and communication system should be available to all patients to reduce travel to the hospital. Monitored home-based sessions may occur during a more convenient time for the patient while maintaining contact with their care team when necessary [403].

Remote monitoring may also include a more pervasive approach through wearable technology worn by the patient. This approach may allow the patient to experience their day to day activities and complete exercise training sessions more naturally (i.e., without checking in with the CEP). Remote monitoring has been studied as an obesity intervention among adolescents and has emerged as a novel approach in adult-based cardiac rehabilitation programs [249, 404, 405]. Establishing a combined remote monitoring and communication system allows for the CEP to assess and track the patient's progress and compliance. The CEP may also communication directly with the patient to provide encouragement and help the patient maintain their program progress.

8.2.4 Program Elements

Although the delivery model may be different than an adult-oriented program, the elements included in a pediatric cardiac rehab program are much the same. A pediatric cardiac rehab program should include the following core elements.

1. **Exercise training:** Exercise training should remain a central component of a pediatric cardiac rehab program. The type of exercise training should depend on the age of the patient and align with the results of the intake assessment and goals discussed during the CEP consultation.

   Children (4-6 years old): exercise training should include a home-based, play-based model for young children. This training should be facilitated by parents and be closely monitored by a CEP to track progress, ensure a variety of activities are available to avoid boredom/lack of interest, and appropriateness for activity given changes in cardiac condition or fitness. The focus of exercise training should be gross-motor skill development, fine motor skill development, coordination, and aerobic fitness. An online or mHealth component may be considered to supplement information, provide a means to share credible resources, and communicate with the CEP.

   Children and Pre-adolescents (7-12 years old): exercise training should remain primarily home-based with parental facilitation. The training program should begin to introduce more independent forms of activity that do not rely on parental facilitation. School physical education may introduce new activities and associated challenges with learning new forms of activity. However, physical education class should not be considered part of an exercise-training program but rather supplementary to the training recommendations.

Sport participation may also be introduced at this age with scheduled training/practice sessions. The CEP should determine if and/or how sport training can be integrated into a training program.

Adolescents (13-17 years old): the foundation of exercise training developed during childhood should be maintained. An exercise-training program for adolescents may progressively move towards more structured or traditional exercise training modalities (i.e., running, cycling, swimming, resistance training) with guidance by a CEP to recommend details regarding frequency, intensity and duration parameters. A blend of home-based and facility-based (gym) programming options may be available for patients. These options remain unsupervised and training on self-regulation and monitoring should be introduced to patients (i.e., talk-test, measure heart rate by palpation). There may also be an increased competitive nature to certain activities and self-monitoring should also be encouraged to avoid over-exertion or injury.

The patient should also be encouraged to take increased responsibility for scheduling training sessions, tracking progress, identifying potential barriers to being active and devising strategies to overcome these barriers to remain adherent with their training program. This approach may help the patient prepare for emerging adulthood, marking a significant change in their routine and lifestyle with greater independence and less parental involvement and oversight.

Emerging Adult (18-25 years old): the transition from pediatric to adult care institutions presents a number of challenges, including continuity of care. The resources needed to address exercise-training requirements for ACHD patients are based largely on acquired

heart disease and those implemented in adult-based cardiac rehab programs. However, this approach may be inappropriate for ACHD patients with different demographic and clinical characteristics. Exercise training among the ACHD population should continue to be overseen by a CEP, and perhaps with the same CEP from the patient's pediatric care centre. This approach may help the patient maintain an existing program with a familiar care provider during the transition of care process. At a very least, similar to the rest of the patient's transition of care requirements, an exercise-training program established at a pediatric institution should be transitioned to the ACHD team.

The ACHD team may have access to an online or mHealth system that was used to help patients engage with their exercise training information during their pediatric care. Making the historical data available for the ACHD team may provide detailed information about the patient's exercise training regimen and identify areas for improvement and important successes in their program (i.e., meeting goals, substantial progress, and improved exercise test results).

2. **Physical Activity Counseling:** The current physical activity recommendations should be relied upon to facilitate a discussion about physical activity behaviour that includes an overview of associated health benefits. Patients should be asked about what types of activities constitutes "physical activity" in their day-to-day. One approach that may be useful is using an "elicit-provide-elicit" format. Using this approach, patients are asked to describe their current activity behaviour and perceptions, feedback is provided that may include a comparison to current recommendations with examples of physical activity not identified by patients/families, and finally asking for their thoughts about activity with this new information. Basic education about physical activity can be introduced if the

patient permits the sharing of this information, including any additional complementary resources (i.e., websites, pamphlets, community organization recommendations). This approach respects the patient's autonomy (which further aligns with an MI approach to physical activity counseling) and current knowledge on the topic.

Integrating MI into physical activity counseling offers a non-judgmental approach to help identify and resolve ambivalence towards changing behaviours that influence physical activity participation. The Readiness to Change assessment completed during the in-take process will help identify the current stage of change. This can be used to open the conversation and confirm the current stage with the patient through a discussion about their current physical activity participation.

Physical activity counseling should provide an opportunity for patients/families to explore their activity participation with guidance and input from an activity counselor. This counseling could help identify discrepancies and offer recommendations to help improve activity behaviour. At least one physical activity counseling session should be offered for each patient. This single session can also be used to establish a rapport with the patient and families for future consultations using MI. Each subsequent session should still include MI, and build on each subsequent session using key MI principles (i.e., identify change talk, reflective listening, summaries, decisional balance). During the activity counseling sessions, patients may be asked to indicate their importance, confidence, and readiness to make a change using a simple subjective feedback scale (i.e., "On a scale from 0 to 10, with 0 being not important/confident/ready to change my behaviour to 10 being very important/confident/ready to make a change, where do you

see yourself?). This will provide the counselor with subjective feedback from the user to help direct the conversation towards certain ambivalence towards making a change.

Accelerometer results from the intake assessment should also be reviewed in relation to the recommendations and current perceptions of their activity. The accelerometer results should be reviewed in the context of a "snapshot" of their activity levels representative of their lifestyle. The review of these results may initiate a discussion about challenges to activity during this assessment.

3. **Nutrition/Diet Counseling:** A Registered Dietitian should meet with patients and families to review current nutrition practices and facilitate a discussion about diet choices. The approach for nutrition counseling should promote a balanced diet based on recommendations from a Registered Dietitian. Goal setting should also be included in this consultation and recommendations should be based on these goals. A consultation with a dietitian will provide patients and families with the necessary information to improve their diet choices and focus on risk factor modification (i.e., lipid, cholesterol, and diabetes management)

4. **Psychology Services:** Access to psychology services should be made available to patients if concerns are identified by clinicians involved in their circle of care or brought up during patient visits. Support for patients is important to help improve psychosocial aspects including self-efficacy, mood and anxiety disorders, mental health, socialization with others, and navigating the complexity of adolescence and building skills to become more independent as an adults with CHD.

5. **Health Promotion:** General health promotion and education should be included in the program that focuses on smoking prevention, and safe drug and alcohol use. The aim should be to provide education about the deleterious health consequence of smoking and drug and alcohol use in the context of the CHD and associated risk factors. General public health promotion material and approaches may be sufficient but supplementary material should highlight the importance of maintaining a healthy lifestyle to prevent disease.

8.2.5 Program Maintenance and Transition

Patients enrolled in a pediatric cardiac rehab program should remain enrolled for 12-weeks. During this time, the patient should be provided support to help them adhere to the training plan and be continuously engaged in the other components offered by the program (i.e., physical activity counseling, nutrition/diet, psychology services, health promotion). The program should help the patient acquire the necessary knowledge and improved self-efficacy to continue the program independently. The CEP should provide a progress report that is reviewed with the patient for clarity. The patient and CEP should also work together to formulate new goals and identify the best approach to meet these goals moving forward in a more independent form of training. If the patient completes the 12-week program and continues to be followed at the referring pediatric institution, the patient should be offered re-assessment and counseling as needed. The patient may also be eligible to re-enroll during their time at the pediatric institution if referral to the program is received. Long-term enrollment (>12-weeks) may be an option for patients that undergo re-intervention, experience substantial functional limitations (i.e., heart failure), or complete the first session as a child and require re-assessment and new programming as an adolescent. The patient should repeat the intake assessments to document changes observed

since the change in status or since the first program. This re-assessment serves to inform a new set of goals as the patient moves into a new phase of training.

If the patient is enrolled in the pediatric cardiac rehab program, transfer from pediatric to adult care should include a detailed plan with regards to the transfer to exercise and physical activity knowledge between providers. The final progress report should be included in the patient's medical chart and key recommendations for the patient as an adult should be outlined for the new care providers. Coordination with an adult-based exercise program affiliated with the adult care centre should occur when possible.

8.2.6 Support and endorsement for pediatric cardiac rehab

The Canadian Association of Cardiovascular Prevention and Rehabilitation (CACPR) is a national organization that provides inter-disciplinary expertise related to cardiovascular disease prevention and rehabilitation. The focus of the CACPR has been on adult-based cardiovascular disease, namely acquired/ischemic heart disease and stroke. While cardiac rehab programs for adults have evolved over the last few decades, the CACPR has not integrated pediatric-based programs or research into their goal to enhance cardiovascular disease and rehabilitation knowledge and clinical care. Given the growing ACHD population and preliminary evidence that ACHD patients can participate in cardiac rehab, pediatric and adult cardiac rehab programs may overlap. This presents a cardiac rehab life-cycle for CHD patients that may allow for the coordination and continuity of cardiac rehab programs for patients and families. The CACPR may be the best-suited organization with the relevant cardiac rehab program knowledge and research expertise to support a pediatric cardiac rehab program that would ultimately prepare patients for adult care and thus adult cardiac rehab programs.

9 Limitations

This thesis work should be viewed in light of the following limitations. The recruitment of patients was challenging and resulted in a small sample size for the Fontan Activity Study and REACT in CHD study. This may have been due to the age of patients and difficulty contacting adolescents and young adults. This was particularly evident for patients from the Fontan Activity Study as the eligible patients were identified from a pre-existing enrolment list from a previous study and yet contacting patients was extremely challenging. Patients were attending post-secondary institutions during the recruitment period and were often living away from home. The contact information was updated but locating and messages may not have been communicated. Data collected from the earlier Fontan study (Fontan Cross-sectional Study) was incomplete for some patients that were enrolled in the Fontan Activity Study and limited the ability to complete a full repeated measures analysis between assessment periods. Furthermore, a small sample size in each study limited the ability to include multiple regression analysis to identify additional associated factors. The inclusion/exclusion criteria did not account for incomplete data sets collected during the earlier study. Only patients with a complete data set from the Fontan Cross-sectional Study, including accelerometer data, should have been eligible for the Fontan Activity Study.

The REACT in CHD study experienced poor enrolment rates given the substantial number of patients screened and approached to participate. This may be a result of the overall lack of appeal of a physical activity intervention among CHD patients (i.e., patients are not interested in participating due to the purpose of the study). In addition, patients also declined due to lack of time to attend the hospital for visits or complete the study intervention (telephone sessions). The approach used in the study may have prevented participation in the study. The use of a telephone

intervention may not have been as appealing for patients compared to more modern means of communication, including text-messaging or video calls (Skype). An online or mHealth approach may also have resulted in greater acceptance of the intervention.

Patients recruited in the REACT in CHD study reported favourable outcome measures at baseline and following the study intervention. This may be a result of patient bias towards physical activity, and a greater interest and positive perception of physical activity at baseline. Patients were not screened or excluded from the study if they demonstrated positive outcomes at baseline.

Accelerometer measurements provide an objective measure of physical activity that relies on the patient to adhere to specific procedures to ensure data quality. Although accelerometer data is not subject to over-estimation like self-report instruments, patients may also mishandle the accelerometer (i.e., move or throw the accelerometer) that misrepresents actual movement. Logbooks should be used to verify data collected by the accelerometer to avoid including non-wear movement data. Relying on courier services to send and receive devices cannot only result in lost devices, but also lost data that requires re-assessment (if possible). In addition, the loss of devices also results in the cost of replacing the device itself. Finally, relying on the patient to return the device immediately after use can sometimes be problematic and prevent the collection of accelerometer data for another patient if the device remains in the patient's possession after use. Therefore, although accelerometers provide high-quality, objective measures of activity data, there remain logistical challenges in coordinating the delivery and receipt of monitors with usable data for analysis.

10 Future Research Opportunities

This research focused on the physical activity and exercise behaviour of adolescents and emerging adults with CHD. A longitudinal assessment of physical activity among Fontan patients was the first study to report physical activity trends in this cohort between childhood and emerging adulthood. A behavioural approach was used to help adolescent CHD patients increase their physical activity. The use of a behavioural rather than prescriptive approach remains a novel strategy to help CHD patients become more physically active. Emerging adults with CHD shared their perceptions about physical activity and exercise using qualitative interviews. This was the first study to report perceptions from their past, in the present, and their outlook on the future. This body of research focused on adolescents and emerging adults given that the existing research in this area included children and there was a lack of information about this age group. While this research does contribute to the literature, there remain exciting opportunities to continue this line of inquiry to improve our understanding of physical activity and exercise in the lives of patients with CHD. The results of this thesis research have important considerations for future research that can be used to inform clinical practice and the development of a CHD-specific pediatric cardiac rehab program.

10.1 Physical activity and exercise research in CHD: standardize for success

There is a need to develop consistent and standardized approaches to physical activity and exercise research among the CHD population. The evidence to support more formal and structured programs is available across pediatric and adult CHD settings. However, the variety of methodologies and outcome measures used to report this evidence limits the ability to accurately

compare research and ultimately identify an optimal approach to help generate evidence to inform clinical practice. The work to date has focused on exercise-training outcomes with limited attention placed on understanding the physical activity and psychosocial outcomes of CHD patients. A comprehensive evaluation of the CHD patient across the lifespan is needed to inform the development of a clinical pediatric cardiac rehab program that can service this unique patient population.

A working group should be formed that includes individuals with expertise in pediatric cardiology (i.e., pediatric cardiologist), CHD patient management (i.e., nurses, nurse practitioners), exercise science (i.e., Certified Clinical Exercise Physiologist, physiotherapist), physical activity (i.e., kinesiologist, physical education teacher, sports instructor), and behavioural interventions/behaviour change (i.e., psychologist, social worker). Additional health professionals may also be considered, including pharmacists, surgeons, occupational therapists, genetic counselors, and dietitians. Parent and/or patient representation should be considered to gather their perspectives on the research process, proposed assessments, and data collection preferences. This approach aligns with a more patient-centric approach to care delivery, engages the patient in the research process, and may lead to greater acceptance of the proposed research approach in the future. This working group should identify a core set of assessments and outcomes that provide a comprehensive and clinically relevant representation of the patient. The assessments should be readily available for researchers and instructions about proper administration of the assessments should be clearly outlined for researchers to mitigate variation between researchers. The multi-disciplinary representation of the working group highlights the complexity of CHD care to identify best practices to optimize care. Coordination between the relevant clinical groups may serve to reduce redundancy in research data collection while offering an opportunity for inter-professional collaboration. Identifying a core set of assessments

and outcomes may also facilitate multi-site collaboration with consistent data collection practices. This coordinated and collaborative approach could help increase the number of patients included for more in-depth analysis to identify factors associated with physical activity and exercise outcomes.

10.2 Long-term outcomes for long-term survival: what are the sustained effects?

There remains a paucity of longitudinal research to identify long-term outcomes in this growing clinical population with respect to physical activity and exercise outcomes. Only two studies reported long-term follow-up outcomes following an exercise training intervention. Both studies reported sustained effects of the training intervention and provided encouraging results to support exercise training to improve both short-term and long-term outcomes. Long-term follow-up data are unavailable for other types of interventions, including behavioural interventions or combined interventions. There is a need to evaluate the long-term physical activity trends of CHD patients across childhood, adolescence, and adulthood that receive different types of interventions to identify important trends and associated factors that may influence physical activity participation and exercise performance. Future research should aim to collect long-term follow-up data, particularly for clinical trial or intervention studies to determine any sustained effects of the intervention to help patients improve their physical activity behaviour. One approach to collecting long-term activity data may be to integrate annual physical activity tracking into the clinical care of patient. Alternatively, mHealth tools may also provide a means to track physical activity between clinical visits. The use of the patient's mobile device with a suitable application to measure physical activity may be an attractive approach for patients.

However, the use of mHealth for this purpose requires additional investigation to ensure all regulatory (privacy and security) requirements are in place.

10.3 Subgroup CHD research to identify diagnosis-specific physical activity needs and preferences

This thesis research focused on the physical activity and exercise behaviour of adolescents and emerging adults with a full range of mild to complex CHD. The sample size in each study was inadequate to complete a sub-group analysis by diagnosis in the REACT in CHD group. In addition, regression analysis to identify factors associated with physical activity was not completed due to the small sample size. Future research should aim to identify differences by diagnosis or at least by CHD severity (i.e., mild, moderate, complex). This approach will help identify differences in the response to physical activity and exercise interventions by diagnosis group. Conducting research with a more homogenous sample of patients with the same CHD diagnosis may help focus the analysis to identify specific factors associated with activity and exercise outcomes. As a result, these findings may influence how the interventions are applied in the clinical setting to a specific patient population and provide useful details regarding clinical practice guidelines.

10.4 Including CHD patients with significant co-morbidities in future physical activity and exercise research

Patients with CHD may have significant co-morbidities that may influence their ability to participate fully in some physical activities or limit their ability to complete self-report instruments. These patients are often excluded from physical activity and exercise training

research studies, as was the case in this thesis research. Research on the physical activity and exercise behaviour of patients with CHD and other co-morbidities is scarce and limits the ability to provide specific clinical recommendations for these patients. Future research should include patients that have specific developmental, musculoskeletal or cognitive impairments to identify physical activity and exercise preferences and behaviours unique to an otherwise overlooked group of patients. This research will also help identify any safety and adverse event occurrences in this population to help clinicians and parents address concerns regarding injury or medical events resulting from physical activity participation. This is particularly important as regular physical activity and exercise may help maintain the health status with respect to other co-morbidities as well as the underlying CHD. Patients with multiple chronic diseases may also experience over-protective parenting and/or unnecessary self-restriction from activity if insufficient detail is provided to make informed decisions about physical activities that are safe and appropriate.

10.5 **R**ehabilitative **E**xercise and **A**ctivity for **L**ife (REAL): a proposed preventative cardiology research program for CHD patients

The next proposed series of studies would inform a larger program of research and build on the lessons learned to investigate alternative approaches to help CHD patients improve their physical activity behaviour across the lifespan. The Rehabilitative Exercise and Activity for Life (REAL) research program would aim to investigate the physical, medical, psychosocial, and environmental factors associated with physical activity and exercise investigations among CHD patients. The REAL program would primarily follow patients from childhood to emerging

adulthood and offer novel interventions aimed to educate, facilitate, and monitor physical activity and exercise outcomes.

Future research conducted through the REAL program would be guided by the Health Belief Model to help adolescents contextualize their condition with respect to physical activity and exercise performance. The Health Belief Model would provide an overarching theoretical framework to guide interventions relevant to each construct within the Health Belief Model (perceived susceptibility, perceived severity, perceived benefits, perceived barriers, cues to action, and self-efficacy) that aims to improve physical activity. While the individual interventions to be investigated within the REAL program may include other theoretical frameworks or models (e.g., Motivational Interviewing, Social Cognitive Theory), the basis of the REAL program would be one of health promotion and positive health behaviour change. The Health Belief Model may also help predict why some patients change their health behaviours or maintain an active lifestyle while other patients do not. The acceptance of the CHD diagnosis, personal perceptions of susceptibility and potential consequences of the CHD, and susceptibility to illness or disease as a result of the CHD could be explored and applied to future physical activity and exercise interventions.

The REAL program would also leverage emerging mHealth technology to improve the engagement of patients with their activity recommendations and provide a means to track activity participation. The use of mHealth for physical activity and exercise interventions has not been reported in the CHD population. This evolving field requires rapid attention to meet the ubiquitous nature of mobile technology among young patients who are true technology natives. There are many opportunities related to mHealth, including mobile apps and wearable

technology, that may revolutionize how physical activity and exercise research is conducted in this population.

The REAL research program will address the following questions:

1. **How can the Health Belief Model guide interventions to help patients with CHD understand their diagnosis in the context of physical activity and exercise participation and ultimately improve physical and psychosocial outcomes?**

The Health Belief Model would be used to conceptualize new interventions that introduce patients to perceptions of disease susceptibility and disease severity (perceived threat of disease). Education around the benefits and barriers to physical activity would be provided to patients. Self-efficacy would be central to the interventions and align with the exercise training and physical activity counseling interventions. Behavioural interventions should be explored that aim to increase self-efficacy to be physically active. Despite the results presented in this thesis work, MI should continue to be explored as a behaviour change intervention. Future research should evaluate the use of MI with increased training and frequent MI fidelity audits of sessions to ensure MI adherence. The delivery of the MI intervention (i.e., in-person, telephone, online) and frequency of sessions should also be explored to identify an optimal intervention approach that is accepted by patients and provides positive outcomes.

2. **How can mHealth technology be used to help patients become more engaged with the physical activity recommendations?**

The use of mHealth technology for physical activity and exercise research has not been evaluated in the CHD population and would be introduced to patients in future research. Initial feasibility

and pilot research would be conducted to assess overall usability and acceptance of this new technology. Integrating mHealth technology into the research environment could facilitate coordinated and widespread data collection using mobile device. For example, the use of open-source research platforms like Apple's ResearchKit may be a new approach to collect useful health data at the population level. mHealth may also be leveraged to deliver interventions (e.g., messages, videos) and serve as self-sufficient intervention that does not rely heavily on personnel to deliver the intervention.

3. **What are the long-term effects of physical activity and exercise interventions on physical and psychosocial outcomes among CHD patients and how do these change between childhood, adolescence, and adulthood?**

This research program would track the physical activity level of patients on a 6-18 month cycle (to account for seasonal variation) to identify trends in physical activity behaviours. Activity assessments would be assessed using an accelerometer. In addition, concurrent research would aim to identify factors associated with changes in physical activity, including medical, psychosocial, physical, and environmental factors. Ultimately, a registry of physical activity data may be created that would serve as a central data repository for physical activity data. This database could be available for other researchers to contribute to and access patient data for more robust statistical analysis.

4. **What resources are needed to support family-based physical activity interventions for children with CHD and how do these intervention influence future (adolescent and adult) physical activity behaviour?**

Family-based activity interventions should be encouraged and supported during the pediatric care of CHD patients. While the main focus of the clinical care and conversations about physical remains on the patient, improving the physical activity participation of parents may result in positive perceptions of activity from the patient. This research program would develop and evaluate the necessary resources to support family-based physical activity interventions.

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