

OCCUPATIONAL PERFORMANCE CHARACTERISTICS OF CHILDREN AND  
ADOLESCENTS WITH CONGENITAL HEART DISEASE.

By

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B.App.Sc.OT., The University of South Australia, 1983

A THESIS SUBMITTED IN PARTIAL FULFILMENT OF  
THE REQUIREMENTS FOR THE DEGREE OF

MASTER OF SCIENCE

in

THE FACULTY OF GRADUATE STUDIES

(School of Rehabilitation Sciences)

We accept this thesis as conforming

to the required standard

THE UNIVERSITY OF BRITISH COLUMBIA

July 1999

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Date August 9<sup>th</sup> 1999

## Abstract

This study explored the occupational performance characteristics of children and adolescents who have congenital heart disease. Children with congenital heart disease have been found to be at risk for neurological deficits and to experience an increase in problem behaviors, learning and academic difficulties and an increase in emotional or mental health disorders. Recently, pediatric cardiologists have advocated the establishment of rehabilitation programs for these children focusing almost exclusively on exercise. This somewhat narrow focus does not encompass the multitude of deficits that often accompany congenital heart disease. Occupational therapy theory provides a broader framework for exploring the range of difficulties these children are experiencing as the literature suggests they may be under-performing in occupational performance areas of self-care, productivity and leisure.

This prospective descriptive study assessed children and adolescents with congenital heart disease to determine their ability to participate in self-care, productivity and leisure tasks. The sample of 35 participants aged 7.0 – 16.5 years underwent assessment using standardized measures. The data were analyzed descriptively and inferentially. The t-test was used to determine whether a statistically significant difference existed between the group means and the test means.

Participants had complex congenital heart disease and had experienced at least one open-heart surgery with cardiopulmonary bypass and hypothermia, a median of 9.3 years prior to participation. In comparison to test means, participants were found to have significantly reduced skills in self-care, productivity and leisure as reported by parents. They also had significantly reduced skills in visual motor integration and motor control

as measured directly. There was no significant difference found in performance on either visual perception or fine motor skills and participants reported a moderately high sense of self-worth.

Contrary to anecdotal opinion, this study found that children with congenital heart disease do not grow out of early difficulties during middle childhood. Further investigation is required into the efficacy of interventions aimed at increasing their ability to undertake meaningful occupations of childhood. Rehabilitation programs must address more than exercise. In addition, intra-rater reliability analysis suggested that a review of the process used to establish inter-rater reliability in commonly used tests and measures is warranted.

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## Acknowledgements

The completion of this masters' thesis represents a milestone on a voyage of adventure undertaken since mid 1995 when our small family decided to 'leave home'. Our Canadian experience has been supported by many people, not least of whom, are those who we left behind.

My admiration and gratitude go to my supervisor Dr. Susan Harris, who continued to offer positive support and encouragement throughout her own most difficult year. As well, I sincerely appreciate the assistance provided by my committee members, Dr. Anne Carswell, and especially Dr. George Sandor, whose initial receptiveness to the idea of including occupational therapy outcomes to his study and his ongoing encouragement, enabled this project to occur. The Demonstration Project to Evaluate the Benefits of Exercise Rehabilitation in Children with Congenital or Acquired Heart Disease, of which this study is a component, received generous financial support from The Heart and Stroke Foundation of British Columbia and Yukon and British Columbia's Children's Hospital Foundation.

I am grateful for the many opportunities I have had in Canada, and the support received from colleagues at the British Columbia Children's Hospital. I especially valued Dr. Jim Potts ongoing assistance in the development and implementation of the demonstration project and Laurie Ainsworth's statistical assistance.

Finally, my deepest appreciation goes to Wes, Tyson and Phoebe, who have 'lived the experience' with me, and without me when I spent hours at the computer. Wes' enthusiasm, reflective thinking, lively discussions and sense of humor greatly increased my enjoyment of the process.

## Chapter I: Rationale and Significance of the Study

### Rationale

Occupational therapists provide therapy for children with disease or disability in the belief that by enabling successful engagement in childhood occupations, quality of life will be improved (CAOT, 1997). The role of the occupational therapist in intervening with children with known neurological or significant developmental disorders has been well developed and documented (Pratt & Allen, 1989). Less well documented, and perhaps less well developed, is the role of therapists with children and families who experience congenital heart disease. A literature search utilizing Medline (1966-1999), CINAHL (1982-1999) and Current Contents (1996-1999) databases found only two references to research conducted by occupational therapists, specific to children with congenital heart disease. This suggests that children with congenital heart disease have not been clearly identified by therapists, as a group who may experience difficulties in participating in developmentally appropriate childhood occupations. However, research in other fields has demonstrated that some children with congenital heart disease are experiencing difficulties in many areas of their lives and a number will experience either transitory or permanent neurological deficits (du Plessis et al., 1995; Fallon, Aparicio, Elliott, & Kirkham, 1995; Ferry, 1987; Miller, Egli, Contant, Baylen, & Myers, 1995; Mravinac, 1991; Smith Wical & Tomasi, 1990).

Congenital heart disease occurs with an incidence of about 0.7% of live births with 85% of these infants surviving to adulthood (Sparacino, 1994). Most defects are detected in infancy or early childhood although some may remain undetected until adulthood. Improved surgical and medical management of congenital heart disease has

provided many children with either a definitive repair, resulting in anatomically or physiologically normal circulation, or palliative procedures designed to manage the pulmonary blood flow (Higgins, 1994). Regardless of the type or success of the surgical intervention, most will require ongoing monitoring throughout their lives (Higgins, 1994; Higgins & Reid, 1994).

While the cause of congenital heart disease is not fully understood, it is known to be associated with maternal conditions such as diabetes, lupus, and some prescription and non-prescription drugs. Chromosomal anomalies such as trisomy 21, Turner's syndrome, Noonan's syndrome and the DiGeorge or CHARGE configuration of anomalies are also associated with an increased incidence of congenital heart disease. These children may come to the attention of therapists because of the neuro-developmental consequences of their syndrome or disorder, rather than the presence of a heart defect. However, those children with congenital heart disease who do not have an associated congenital anomaly have also been shown to be at risk of neuro-developmental consequences from either the defect itself or from complications of the surgical procedures they experience (for example, Ferry, 1987; Jonas, 1998; Limperopoulos et al., 1999).

In addition to the literature related to neurological outcomes for children with congenital heart disease, psychosocial and quality of life studies have focused on a wide range of issues. This literature described children with congenital heart disease as having an increase in problem behaviors (Casey, Sykes, Craig, Power, & Mulholland, 1996; Janus & Goldberg, 1995; O'Dougherty, Wright, Garmezzy, Loewenson, & Torres, 1983; Utens et al., 1993), decreased global psychological functioning (DeMaso, Beardsley,

Silbert, & Fyler, 1990; Morris, Krawiecki, Wright, & Walter, 1993; O'Dougherty et al., 1983), lower self esteem (Kitchen, 1978), poorer psycho-social functioning (Casey et al., 1996; Spurkland, Bjornstad, Lindberg, & Seem, 1993; Wright, Jarvis, Wannamaker, & Cook, 1985), learning and academic difficulties (Morris et al., 1993; O'Dougherty, Wright, Loewenson, & Torres, 1985; Wright & Nolan, 1994) and emotional (Utens et al., 1993) or mental health disorders (Wright & Nolan, 1994). This population has been presented as being at risk for ongoing developmental and educational needs, perhaps requiring additional resources (Morris et al., 1993; Spurkland et al., 1993; Wright & Nolan, 1994).

Our ability to generalize the findings of these studies is somewhat limited. With the passage of time, surgical and medical techniques change, with subsequent altered experiences for the children and their families. Are the findings reported in the past decade relevant to today's children when the children studied underwent surgery 10-20 years ago? Certainly they may be relevant to today's older teenagers and young adults, however, we have a continually changing population of younger children with needs that may be quite different.

Adults with cardiovascular disease have benefited from well-developed and well-researched cardiac rehabilitation programs encompassing exercise, psychosocial and occupational components. These programs have included education, counseling, vocational retraining, and interventions to reduce stress and increase coping (Cronin, 1992; Tooth & McKenna, 1996; Vaccaro, Galioto, Bradley, Hansen, & Vaccaro, 1984; Wilde & Hall, 1995). Unlike their adult counterparts with cardiovascular disease (Tooth & McKenna, 1996), children with congenital heart disease do not consistently receive

well-developed and comprehensive rehabilitation programs (Vaccaro et al., 1984; Washington, 1992). The growing body of literature and research in the pediatric population has to date focused on the introduction of exercise rehabilitation programs aimed at improving oxygen consumption and aerobic fitness. While some studies on the effect of exercise rehabilitation commented on improvements in independence and self confidence (Calzolari et al., 1990), adaptations to activities of daily living (Galioto, 1990), and increases in socialization (Ruttenberg, Adams, Orsmond, Conlee, & Fisher, 1983), they did not present data related to these suggested outcomes, or develop programs to address those specific areas of functioning.

The Canadian Model of Occupational Performance (CAOT, 1997) provides occupational therapists with a theoretical framework from which to consider the impact of disability or illness upon occupational performance tasks, and how to approach the challenges for the individual within their environment. Occupational performance, or the ability of an individual to perform an activity in a satisfactory way, is influenced by factors in three domains, the person, his or her environment and the occupation itself. At the person level of the model, consideration is given to the physical, affective and cognitive components of the individual. These three components describe the skills and attributes utilized by the individual to engage in occupational performance tasks. The research reviewed suggests that children with congenital heart disease may experience deficits at the person level of the model, related to their heart defect or the repair process. Deficits of performance in these personal skills may reduce participation and success in childhood occupations related to self-care, productivity or leisure.

Exercise rehabilitation has been shown to improve exercise tolerance and performance (Koch, Galioto, Vaccaro, Vaccaro, & Buckenmeyer, 1988; Sklansky, Pivarnik, Smith, Morris, & Bricker, 1994; Thomassoni, Galioto, Vaccaro, & Vaccaro, 1990) and exercise rehabilitation is now being advocated (Galioto & Tomassoni, 1993; Thomassoni et al., 1990). However, whether exercise rehabilitation is sufficient to address the total needs of these children is not clear. Studies have found that children with congenital heart disease have poor psychosocial functioning (Casey et al., 1996; Spurland et al., 1993; Wright et al., 1985), and may have neurological insults as a result of surgery or hypoxia (DeMaso et al., 1990; Ferry, 1987). It is logical to assume that these difficulties will impact the physical, cognitive or affective skills and abilities of the child. Indeed the previous studies cited support this rationale (Casey et al., 1996; Koch et al., 1988; O'Dougherty et al., 1985; Sklansky et al., 1994; Utens et al., 1993; Wright & Nolan, 1994). The findings of these studies suggest that children with congenital heart disease are at risk of reduced physical, cognitive or affective skills which in turn may limit their ability to participate in the childhood occupations of self-care, productivity or leisure.

There is a need for systematic measurement of the skills and abilities of children with congenital heart disease, beyond their exercise tolerance and aerobic capacity, to enable appropriate and comprehensive rehabilitation programs to be developed. Assessment of the child's ability to participate in childhood occupations would enlarge the lens through which we view these children today, and would provide an opportunity to ascertain, whether or not, children with congenital heart disease have deficits of performance in self-care, productivity or leisure.

### Significance

This project gathered empirical data on the self-care, productivity and leisure abilities of children with congenital heart disease. This descriptive study gathered data about children, ages 6-16 years, the analysis of which, contributes to the body of knowledge regarding their ability to participate in childhood occupations, after undergoing medical and surgical treatment in the 1980's to early 1990's.

### Objectives

The study represented a component of a grant-funded project entitled Demonstration Project to Evaluate the Benefits of Exercise Rehabilitation in Children with Congenital or Acquired Heart Disease (Sandor, G. Principal Investigator, 1998 - 1999). The demonstration project was designed to implement and evaluate an exercise rehabilitation program. It compared a treatment group who participated in hospital-based and home-based rehabilitation programs with a control group who were assessed only. The primary outcome measures of the demonstration project were aerobic capacity and exercise tolerance in children with congenital heart disease. Additionally, gross motor skills, strength and flexibility were assessed as part of the demonstration project.

As children with congenital heart disease had also been shown to have neurological deficits and negative psychosocial outcomes (for example, Limperopoulos et al., 1999; Wright & Nolan, 1994), a more comprehensive evaluation of the sample was warranted. In order to determine how these children were doing in their daily lives, the study aimed to assess components of the children's occupational performance in self-care, productivity and leisure. Standardized assessment tools were utilized and outcomes

compared with normative data. This study utilized the pre-treatment data of both the treatment and control groups from the demonstration project.

Data were gathered on 1) the child's current level of participation in tasks of self-care, productivity and leisure, as reported by their parents; 2) specific performance components that contributed to success in childhood occupations, in fine motor (physical) and visual-perceptual (cognitive) areas; and 3) the child or adolescent's perception of him or herself as an academic or sports-person. In addition, the child or adolescent's perception of themselves in areas of physical appearance, behavior, social skills and general self-worth and self-esteem were measured. These measures tapped affective performance components.

#### Research Questions and Hypotheses

1. Is there a difference in the development of occupational performance abilities in self-care, productivity and leisure, as measured by standardized assessment tools, between children 6 – 16 years of age, with repaired or palliated congenital heart defects and their typical peers?

The following null hypotheses were tested;

##### Self care

- 1.1. The self-care skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the daily living skills domain of the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984).

## Productivity

1.2 The communication skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the communication domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984).

## Performance components.

1.3 The fine motor skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the Bruininks Oseretsky Test of Motor Proficiency (Bruininks, 1978).

1.4 The visual motor integration skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery, 1997).

1.5 The visual perceptual skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery, 1997).

1.6 The motor control skills of children and adolescents with congenital heart disease are equivalent to those of the population in the standardized sample of the Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery, 1997).

## Leisure

1.7. The socialization skills of children and adolescents with congenital heart disease are equivalent to the population in the standardized sample of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984).

2. Is there a difference in the self-perception profile and self-esteem of children and adolescents, 6 - 16 years of age with repaired or palliated congenital heart defect, as measured by the Self-Perception Profile for Children (Harter, 1985), and the Self-Perception Profile for Adolescents (Harter, 1988).

The subjects will complete the profile appropriate to their age and the following null hypotheses were tested;

2.1. That children or adolescents with congenital heart disease will demonstrate a self-perception profile of scholastic competence, social acceptance, athletic competence, physical appearance, behavioral conduct, job competence, romantic appeal, close friendship and global self worth as measured by the Self-Perception Profile for Children (or Adolescents), (Harter, 1985; 1988) within the normative pattern.

2.2 That children or adolescents with congenital heart disease will demonstrate self-esteem, as calculated by determining the discrepancy between the self-perception domains and the importance domains in the Self-Perception Profile for Children (or Adolescents), (Harter, 1985; 1988) within the normative pattern.

## Definition of Terms and Acronyms

### List of Terms

Arterial Switch Procedure. Infants with isolated transposition of the great arteries may undergo this procedure in the first two weeks of life. The pulmonary artery is anastomosed to the right ventricle and the aorta anastomosed to the left ventricle, above the respective valves. This procedure also involves transfer and reattachment of the coronary arteries (Daberkow & Washington, 1989).

Atrial Septal Defect. A communication, or hole in the septum between the two atrial chambers of the heart, typically resulting in a left to right shunt of blood which increases the blood flow through the pulmonary arteries (Le Blanc & Williams, 1993).

Eisenmenger's Syndrome. Where a primary heart defect is associated with pulmonary hypertension (Le Blanc & Williams, 1993).

Fontan Procedure. Complex surgical procedure to enable a single functional ventricle to act as the systemic pump, with venous return being shunted to flow to the pulmonary artery and lungs without passing through a ventricle. Repair is usually completed in stages with one or more palliative procedures occurring before the Fontan. Palliative procedures may include Blalock Taussig (BT) shunt or Glenn anastomosis. The BT shunt or Glenn procedures are designed to divert systemic blood directly to the pulmonary arteries. (Driscoll et al., 1991; Greely, 1996; Leatham, Bull, & Braimbridge, 1991).

Hypertrophic Obstructive Cardiomyopathy. A form of left ventricular outflow tract obstruction (Le Blanc & Williams, 1993).

Mustard Procedure. A corrective procedure for transposition of the great arteries prior to the advent of the arterial switch. Mustard procedure is still used where there is late presentation of transposition of the great arteries or associated lesions. The procedure involves intra-atrial redirection of blood flow to allow oxygenated blood to be directed to the body rather than lungs. The Mustard procedure was first used in the early 1960's. Prior to this, the Senning procedure was used which also involved atrial redirection of blood (Daberkow & Washington, 1989; Le Blanc & Williams, 1993).

Patent Ductus Arteriosus. The ductus arteriosus is a small vessel communicating between the pulmonary artery and the aorta allowing blood to bypass the non-aerated lungs in the fetus. The ductus usually closes in the first hours or days of life, however in some infants remains open (patent) allowing re-circulation of systemic blood through the lungs (Le Blanc & Williams, 1993).

Tetralogy of Fallot. A heart defect involving ventricular septal defect, overriding aorta, right ventricular hypertrophy and obstruction to the right ventricular outflow tract of varying degrees, ranging from pulmonary stenosis to atresia (Le Blanc & Williams, 1993; Runton, 1988).

Transposition of the Great Arteries. A heart defect where the pulmonary artery arises from the left ventricle and the aorta from the right, resulting in deoxygenated blood being re-circulated around the body and oxygenated blood returning to the lungs. This defect is frequently associated with other lesions, which may result in mixing of oxygenated and deoxygenated blood (Daberkow & Washington, 1989; Le Blanc & Williams, 1993).

Ventricular Septal Defect. A communication in the septum between the two ventricles, which may occur as an isolated lesion or in combination with other defects (Daberkow & Washington, 1989).

List of Acronyms

ASD	– Atrial septal defect
BT shunt	– Blalock Taussig shunt
CAOT	– Canadian Association of Occupational Therapists
CHD	– Congenital heart defect
CMOP	– Canadian Model of Occupational Performance
PDA	– Patent ductus arteriosus
SEM	– Standard error of measurement
TGA	– Transposition of the great arteries
TET	– Tetralogy of Fallot
VSD	– Ventricular septal defect

## Chapter II: Review of the Literature

This chapter will provide a review of the literature. The first section will review occupational therapy theory, providing a framework and supportive rationale for identifying the complex needs surrounding this population. Literature will then be presented on the incidence of congenital heart disease and the known neurological complications of heart disease and its management. Following this, an in-depth review of studies of children with congenital heart disease that included outcome measures most closely related to occupational therapy, will be provided. Finally, the chapter will conclude with a brief summary of the current status of pediatric rehabilitation programs and a summary and critique of the literature presented.

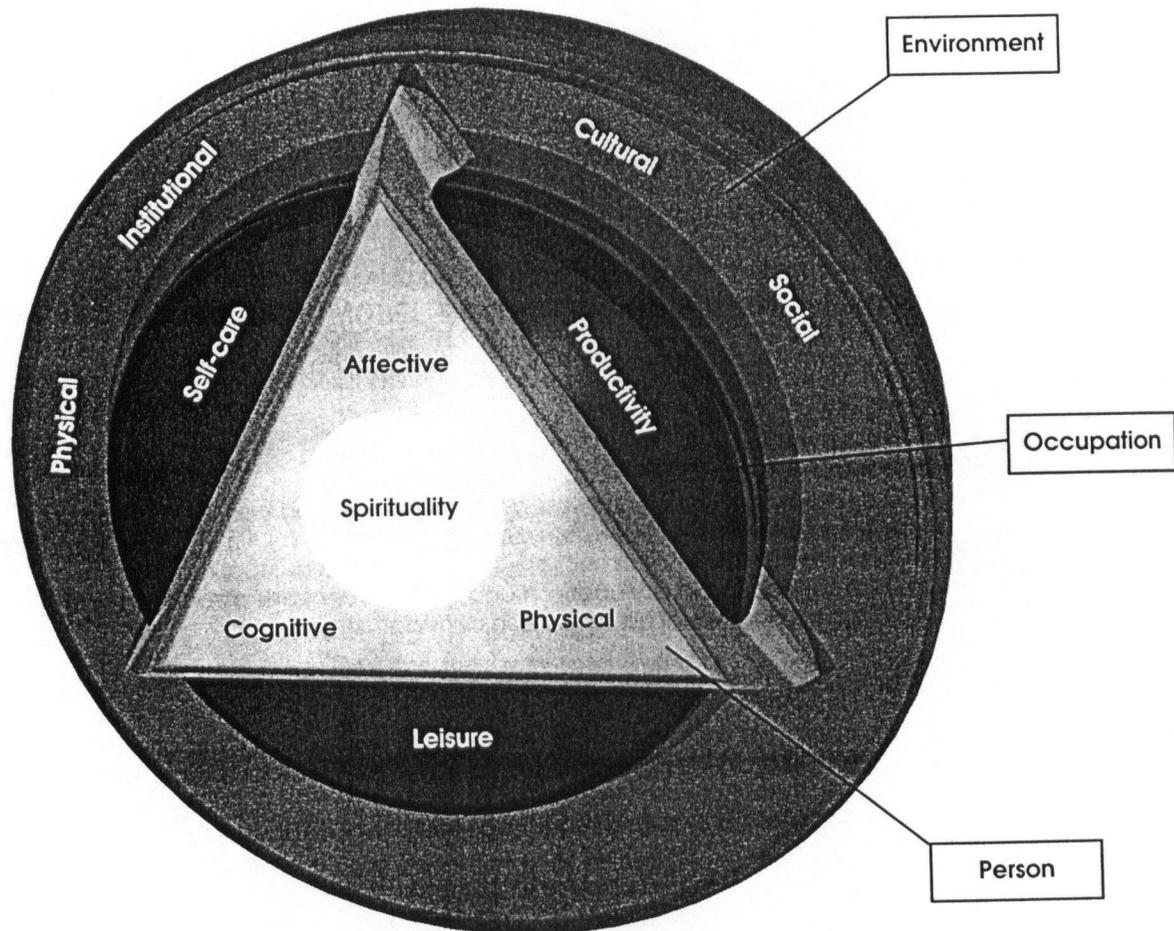
### Theoretical Framework

The Canadian Model of Occupational Performance (CMOP), (CAOT, 1997) was chosen as the theoretical model for this study. The CMOP provides a way of looking at human occupation over the life span. It embodies a client-centered approach to understanding the complexity of what individuals need to do to participate in “life activities” (Baum & Law, 1996, p. 279), using their personal strengths and the resources of their respective environments. The model also provides a way for occupational therapists to consider the impact of disability or illness upon specific occupational performance tasks and of approaching the challenges of adaptation for the individual, and of the environment. This client-centered, multi-layered framework is used to tease out the complex issues and concerns that characterize the experiences of children and adolescents with congenital heart disease.

Occupational performance, or the ability of an individual to perform an activity in a satisfactory way, is influenced by factors in three domains, the person, his or her environment and the occupation itself. The inner spiritual person is the core of the individual and of the model. The three dimensional figure (see Figure 1, CAOT, 1997), represents the dynamic interaction of each level of the model. At the person level, consideration is given to the physical, or doing part of the person, the affective or feeling part, and the cognitive or thinking component of the person. These three performance components describe the skills and attributes utilized by the individual to engage in occupational performance tasks. Physical skills include gross and fine motor skills as well as underlying postural and equilibrium responses. Cognitive skills include communication and development of perceptual and academic skills. Interactions and experiences of the child or adolescent with caregivers and people within his or her extended community provides opportunity for development of affective and social skills.

The environment is considered by the model to be composed of cultural, institutional, physical and social resources that can be used to develop strategies to facilitate the resolution of occupational performance issues. The cultural environment will encompass beliefs about health and disability that influence a person. The institutional environment includes resources in the healthcare community. The physical dimensions of the environment include technology and equipment resources, and family or caregiver supports are a part of the social environment. Environmental factors may also contribute to, or be solely responsible for, occupational performance difficulties.

Figure 1

Canadian Model of Occupational Performance (CMOP)

Canadian Model of Occupational Performance (CMOP), from Enabling Occupation: An Occupational Therapy Perspective (p. 32), by Canadian Association of Occupational Therapists, 1997, Ottawa: CAOT Publications ACE. Copyright 1997 Canadian Association of Occupational Therapists. Reprinted with permission.

The model also facilitates an understanding of the complexities of occupation itself, of the intricacies of the activities of self-care or looking after personal needs, of productivity whether as a child or adult, and of leisure or those activities which restore and refresh the individual. Occupation is a basic human need encompassing those activities people carry out to look after themselves, to participate in the community and to interact with the environment. Occupation is culturally defined and age-related. In childhood and adolescence, self-care tasks are those done routinely to maintain health and well-being in the environment. This includes tasks of personal care and contributions to domestic activities as the child gets older. The productive role of childhood is to learn through movement, play, attending school and, in adolescence, also encompasses the introduction to paid work. Leisure activities include preferences for, and skills in, social interactions and activities undertaken for enjoyment (CAOT, 1997).

Spirituality, as the core of the model, is seen as “the experience of meaning in everyday life” (Urbanowski & Vargo, 1994, p. 89). The model’s idea of spirituality includes concepts of will, drive, motivation and self-determination. Spirituality includes the “innate essence of self” (CAOT, 1997, p. 43) and leads to an appreciation of the uniqueness of every individual within the therapeutic context. Egan and DeLaat (1994) described the individual’s spirit as being expressed through engagement in occupation. The model is situated within a developmental framework and spirituality can also be viewed developmentally; however it is beyond the scope of this review to explore the developmental stages of expressed spirituality.

The CMOP (CAOT, 1997), encourages a holistic approach to occupational performance, encompassing each level of the model. It focuses attention on the skills and

attributes of the children and adolescents and their ability to successfully participate in appropriate occupations within their environmental contexts. The dynamic relationship between the multiple layers of the model highlights the interactions between the person, the environment and the occupation. Changes at any level of the model will have an impact across the layers. Optimal occupational performance will occur when there is congruity between the various levels of the model, that is, the person, his or her spirit, the occupations he or she undertakes within his or her environmental context.

The purpose of this study was to explore the experiences of children and adolescents who have congenital heart disease at the person and occupation levels of the model. Occupational therapy programs and treatment strategies for children have been described in the literature and there are a variety of assessment tools and behavior checklists available to assess pediatric occupational performance components (Asher, 1989). These can be utilized to determine the status of children prior to the development and implementation of treatment programs. However, a literature search utilizing Medline (1966-1999), CINAHL (1982-1999) and Current Contents (1996- March 1999) databases, found no journal references of occupational therapy programs specific to children with congenital heart disease, and therefore no reference to their ability to participate in childhood occupations.

#### Congenital Heart Disease: Incidence and Configurations

Congenital heart disease is a “structural or functional heart disease that is present at birth” (Hoffman, 1990, p. 25). It occurs with a reported incidence of between 0.7 and 1% of live births (Daberkow & Washington, 1989; Hoffman, 1990; Sparacino, 1994).

This conservative estimate of the overall incidence excludes those fetuses who were still-born or aborted early in the pregnancy, and those children with minor lesions that were not detected (Hoffman, 1990). An understanding of the various factors involved in determining a cardiac defect is evolving slowly, as the overall incidence is relatively low and specific defects and combinations of defects even lower. Pathogenesis is thought to be multi-factorial, including maternal and environmental factors, a genetic predisposition and the timing of the insult in relation to embryological development (Daberkow & Washington, 1989; Ferencz & Villasenor, 1991). Ferencz and Villasenor (1991) concluded that there was evidence of genetic predisposition, chromosomal abnormalities or inheritable disorders in 25% of children presenting with congenital heart disease. The authors recommended further research into the effect of environmental toxins such as lead, pesticides and solvents. More recently, studies have reported increasing identification of genes responsible for congenital heart defects (Belmont, 1998).

The heart is the central pumping system of the body responsible for circulating the blood efficiently. Disruption to the pumping mechanism or to the major vessels leading to and from the heart will impact the efficiency and effectiveness of the circulatory system. Clinical signs and symptoms of infants with severe congenital heart disease may include cyanosis, respiratory distress, or congestive heart failure. Cyanosis is a blue discoloration of the nails, lips, or skin resulting from excessive concentration of deoxygenated hemoglobin. Respiratory distress may be seen as an increase in respiratory rate or in the effort to breathe. Congestive heart failure can occur, as the heart becomes less able to meet all of the body's circulatory requirements. This may result in an increase in heart rate, cardiac enlargement, changes in cardiac rhythm, poor cardiac

output, tachypnea, decreased urine output and edema, diaphoresis (sweating), reduced activity or exercise tolerance and early fatigue (Daberkow & Washington, 1989; Le Blanc & Williams, 1993).

Each of these symptoms may effect the child's immediate ability to participate in developmentally appropriate activity as well as having long-term implications. Minor anomalies or defects such as small atrial or ventricular septal defects which involve a communicating hole within the atrial or ventricular septa (Runton, 1988), may not be detected in the infant, or may undergo spontaneous closure as the infant grows. However, many defects will require medical management of the symptoms or surgical repair at some point (Daberkow & Washington, 1989; Limperopoulos et al., 1999; Neveux, 1996; Sparacino, 1994).

Surgical repair may involve procedures that occur during cardiac catheterization, closed or open-heart surgery. Cardiac catheterization occurs under general anaesthetic and involves the passing of a catheter through a major vein or artery, up into the heart. An angiogram is taken to visualize structures. Some manipulation of vessels or valves, such as dilatation, or palliative procedures to increase the supply of oxygenated blood to the body by mixing venous and arterial blood at the atrial level, can occur during catheterization (Le Blanc & Williams, 1993). Closed heart surgery is used to repair structures external to the heart, such as the great vessels. Open-heart surgery is required when repairing the heart itself, and involves the use of cardiopulmonary bypass. This includes septal repairs, valve repair or replacement, or reconstruction of complex lesions such as transposition of the great arteries, tetralogy of Fallot, or univentricular heart (Daberkow & Washington, 1989; Le Blanc & Williams, 1993; Runton, 1988).

As techniques for repair and palliation of congenital heart disease have continued to improve, and more than 85% of these children are now surviving (Sparacino, 1994), attention is being given to the quality of that survival. Research and literature can be found related to medical or surgical management and survival (Higgins, 1994; Higgins & Reid, 1994; Sparacino, 1994), exercise tolerance and cardiac function (Galioto & Tomassoni, 1993; Koster, 1994; Thomassoni, 1996; Washington, 1992), quality of life (Casey, Craig, & Mulholland, 1994; Laane et al., 1997), and to psychosocial and behavioral outcomes (Casey et al., 1996; DeMaso et al., 1990; Kong, Tay, Yip, & Chay, 1986; Kramer, Awiszus, Sterzel, van Halteren, & Classen, 1989; Champaine, Nadelman, Rosenthal, Behrendt, & Sloan, 1990; Silbert, Wolff, Mayer, Rosenthal, & Nadas, 1969; Spurkland et al., 1993; Utens et al., 1993; Wright & Nolan, 1994). This population has been presented as at risk for ongoing developmental and educational needs, perhaps requiring additional resources. Indeed, Samango-Sprouse and Sudderby (1997) described a “behavioral phenotype of children with cardiac disease with a low normal IQ and perceptual-motor delays who are temperamentally more difficult.” (p.91). This clearly suggests that these children and their families would benefit from an in-depth review by therapists who are concerned with the developmental needs of children.

#### Neurological Sequaele to Congenital Heart Disease

Adverse neuro-developmental effects of congenital heart disease have been linked to chronic hypoxia, abnormal circulation leading to an increased risk of thrombosis, bacterial endocarditis, and brain abscess. A number of children with congenital heart disease will also have co-existing or associated congenital syndromes. Chronic hypoxia

has been postulated to have long term developmental consequences. O'Dougherty et al. (1985) studied 31 elementary students with repaired transposition of the great arteries. These children had a mean age at surgery of 2.03 years, and experienced chronic hypoxia until repaired. They were matched with a control group of children with no known neurological or learning difficulties. The authors found that 77% of the children with congenital heart disease had mild to moderately abnormal neurological examination, obtained similar mean IQ scores to the control group, but were under performing in tests of academic achievement. The authors postulated that chronic hypoxia may lead to an "attention deficit disorder with hypo-activity" (O'Dougherty et al., 1985, p.45).

Currently, transposition of the great arteries is usually repaired in the first weeks of life, thus reducing the length of time when children are hypoxic. However, there are children with other heart defects that will result in prolonged hypoxia, or who were born prior to the availability of this repair procedure, for whom these findings may be significant. Children with tetralogy of Fallot may also experience prolonged hypoxia and thus also have associated abnormalities of blood viscosity and be at risk of cerebro-vascular accidents (Cottrill & Kaplan, 1973; Park & Neches, 1993).

Coarctation of the aorta has been associated with an increased incidence of intracranial aneurysm and with spinal cord damage if collateral vessels within the spinal canal dilate and compress the cord. Spinal cord damage can also occur as a complication of surgery during aortic arch repairs (Kirkham, 1998; Park & Neches, 1993). These are uncommon findings however, and as diagnostic and management techniques improve, the incidence of these complications is further reduced.

Infective endocarditis, originating on a prosthetic implant such as a valve, or where there is abnormal shunting of blood within the heart, may result in dislodged emboli causing a cerebro-vascular accident (Park & Neches, 1993). Brain abscess can occur and has been found in up to 2% of children with cyanotic heart defects.

Pathogenesis of central nervous system infection was thought to be multi-factorial including hypoxia with ischemic insult and multiple systemic emboli (Park & Neches, 1993).

Co-existing congenital anomalies or syndromes that occur with congenital heart disease such as Down syndrome will also have neuro-developmental consequences. Children with Down syndrome are typically hypotonic, and experience delayed or altered patterns of development of motor and cognitive skills (Carr, 1995). Between 23 –56% of children with Down syndrome have been reported to have congenital heart disease. Children with Down syndrome thus account for nearly 10% of this population (Hoffman, 1990). Ferencz and Villasenor (Ferencz & Villasenor, 1991) report that 12% of children with congenital heart disease have a chromosomal anomaly and 8% will have an associated syndrome. Certain heart defects, for example hypoplastic left heart, are also associated with a higher incidence of congenital brain abnormality as shown on autopsy (Park & Neches, 1993).

In a recently published paper by Limperopoulos et al. (1999), the neurological status of infants with congenital heart disease was investigated prior to surgery. This prospective study of 56 consecutive referrals to an acute care pediatric facility in Montreal, Canada, found neurobehavioral and neurological deficits in 56 – 58% of neonates. Infants were assessed independently by a neurologist, and an occupational

therapist who assessed the infants' neurobehavioral responses using the Einstein Neonatal Neurobehavioral Assessment Scale (Duam et al. 1977, as cited in Limperopoulos et al., 1999). Infants were excluded from the study if their gestational age was less than 36 weeks, had hypoplastic left heart, or known extra-cardiac defects with a central nervous system component or a known central nervous system insult not related to their heart defect. Neurological deficits found included diffuse hypotonia, jitteriness, no suck, motor asymmetry, decreased muscle power in limbs, cranial nerve abnormalities, poor state regulation, and reduced visual or auditory orienting responses.

In summary, the literature related to the incidence and spectrum of congenital heart disease indicates that the presence of a heart defect places children at increased risk of an adverse neurological outcome. It appears that there is an association between having a heart defect and neurological deficits. Co-existing congenital anomalies or syndromes, prolonged hypoxia, infection or circulatory changes resulting in ischemic cerebral events, may all contribute to the incidence of neurological insult.

#### Neurological complications of cardiac surgery.

As management of congenital heart disease has improved, the incidence of neurological complications resulting from prolonged hypoxia, bacterial endocarditis or brain abscess has decreased. The focus of research has therefore moved to examining surgical complications that may result, as more complex repairs have become possible. The range of post-surgical neurological problems reported includes peripheral nerve damage (Mravinac, 1991), cerebro-vascular accidents resulting in motor deficits (Cottrill & Kaplan, 1973; du Plessis et al., 1995; Mravinac, 1991), seizures, choreoathetosis (Smith Wical & Tomasi, 1990), neuro-ophthalmic defects (Mravinac, 1991), cognitive

deficits, coma or altered levels of consciousness and death (Bellinger et al., 1995; du Plessis, Newburger, & Jonas, 1994; Fallon et al., 1995; Ferry, 1987; Jonas, 1998; Kirkham, 1998; Mravinac, 1991; Newburger et al., 1993; O'Dougherty et al., 1985; Swain, 1993). The main cause of the central nervous system damage that can occur is cerebral ischemia resulting in either focal lesions such as a stroke, or in generalized lesions including periventricular leukomalacia, parasagittal cerebral injury or selective neuronal necrosis. As surgery is undertaken on younger infants, the incidence of complications related to living with an un-repaired heart, are being replaced with the impact of altered conditions during surgery on the very immature brain. Deep hypothermia, lowered or arrested circulation, and relative blood pH levels, all impact the metabolic environment of the developing brain leaving it vulnerable to damage (Jonas, 1998; Kirkham, 1998).

The components of cardiac surgery that are thought to be correlated with adverse neurological outcome are those to do with managing circulation and cell metabolism during surgery, that is, duration of deep hypothermia, cardiac bypass, circulatory arrest or low flow and reperfusion (Jonas, Newburger, & Volpe, 1996; Jonas, 1998; Jonas et al., 1993; Kirkham, 1998; Park & Neches, 1993).

Protective strategies within surgery have developed as knowledge regarding mechanisms of central nervous system injury have grown. Additionally, as techniques in managing blood flow have improved, so has the survival of those infants with complex congenital heart disease for whom there was no possible repair in the past. Low flow cardiopulmonary bypass or circulatory arrest, in combination with deep hypothermia, is used during cardiac surgery to obtain relatively blood-free and motionless access to the

heart. However, low blood flow and circulatory arrest play a role in causing neurological injury in some of these infants (du Plessis et al., 1995; Jonas, 1998; Kirkham, 1998; Newburger et al., 1993). To control blood flow and bleeding within the area of surgery, extracorporeal circulation is established, with blood diverted to a heart lung machine. The heart-lung machine controls gas exchange and blood temperature. During cardiac repair both the pulmonary and coronary circulations are made hypothermic and ischemic. If total circulatory arrest is required, the entire body and brain are included in this process (Jonas et al., 1996).

Both the ischemia and the process of re-perfusion are believed to play a role in the tissue damage that can occur during cardiac surgery. Cerebral ischemia has been shown to precipitate a range of biochemical alterations that begin with the injury and continue into the recovery phase. As the blood flow to the brain is reduced, oxygenation is decreased, and metabolic functions shift from aerobic to anaerobic. However, anaerobic metabolism is not sufficient to meet the energy needs of cerebral tissue. Other metabolic changes, which occur concurrently with this shift, increase the disturbances within the cells. Disturbance of both cerebral perfusion and metabolism may continue into the recovery period, increasing the period of destruction of central nervous system tissue that results in ischemic brain damage (Jonas et al., 1996). During low flow or circulatory arrest there is also a change in the level of activation of leukocytes and platelets that may result in adherence and accumulation of leukocytes in the microcirculatory system. This process, which can result in plugged capillaries and tissue damage, can occur in any organ system, skeletal muscle or in the brain (Jonas et al., 1996).

Deep hypothermia has been found to have a protective effect on cerebral tissue during cardiopulmonary bypass that may be related to the reduced energy requirements of cooled tissues, as well as to interactions between hypothermia and the metabolic changes that typically occur during ischemic events. Even very slight cooling of 3 - 6 degrees Celsius has been shown to have positive effect. During total circulatory arrest, infants may be cooled to as low as 10 - 15 degrees Celsius. Mild hypothermia post surgery has also been shown to reduce the delayed neuronal death that occurs after an ischemic injury (Jonas, 1998). Complex management of the metabolic environment during surgery, including control of blood pH and calcium levels, all contribute to improved neurological outcome in these infants.

The relative incidence of adverse neurological outcome is very difficult to discern from the literature due to varying research methods. In a summary statement of the magnitude of neurological complications in congenital heart disease, Jonas et al. (1996) reported the incidence to be between 10% and 30%. More specific studies have reported that 9% of survivors of congenital heart disease have experienced seizures (Ferry, 1987), 16.6 % of children undergoing repair of arch anomalies have adverse neurological events (Fallon et al., 1995), 6% of children with congenital heart disease undergoing routine surgery have acute neurological disorders (Kirkham, 1998) and 3% have mild cerebral palsy (Jonas, 1998). Miller et al.'s (1995) study into post-operative complications of heart surgery provides an example of findings in this field. These authors conducted a prospective study of 91 full-term infants, who underwent open-heart surgery with hypothermia and either circulatory arrest or low flow bypass. The infants underwent neurological examination before and after surgery to evaluate short-term

neurological outcome. All infants had complex congenital heart disease, including hypoplastic left heart, interrupted aortic arch, transposition of the great arteries, tetralogy of Fallot, septal defects, valve atresia or anomalous vascular connections. Significant findings included 18% overall mortality, seizures in 15% of the infants, transitory movement disorders in 11%, hypotonia in 43% before surgery and 34% after, hypertonia (generalized or asymmetrical) in 12%, and abnormal cranial ultrasound in 20%.

Relationships between outcome and surgical procedures were also analyzed. These authors believe that adverse neurological outcome is multi-factorial. They found that only the duration of deep hypothermia showed a direct relationship to adverse neurological outcome. In contrast to previous studies, such as that by Newburger et al. (1993), there was no difference in outcome when comparing low-flow bypass with circulatory arrest during the surgery. Miller et al.'s (1995) study did not address the more subtle cognitive deficits that may be associated with mild neurological damage, nor did it examine long-term outcome.

Surgical processes may also cause peripheral nerve damage resulting in sensory and motor dysfunction. The most common areas of damage include stretch injury to the brachial plexus during sternal retraction (up to 23% in open-heart surgeries) (Mravinac, 1991), damage to the facial nerve during intubation, or the common peroneal nerve after femoral bypass (Mravinac, 1991; Park & Neches, 1993). When the left recurrent laryngeal nerve is isolated during closed heart surgery, particularly during aortic arch repair, it may be compressed and damaged. The incidence of damage to this nerve during arch repairs has been reported at 4-5% (Park & Neches, 1993).

The literature reviewed provides a baseline of information that suggests that children with congenital heart disease also have a significant risk of neurological complications. One study found that 56 – 58% of infants with congenital heart disease had an abnormal neurological examination prior to surgery (Limperopoulos et al., 1999). In addition, up to 20% of children have been found to have an associated chromosomal anomaly or syndrome (Ferencz & Villasenor, 1991), and between 10 and 30% will experience post surgical complications (Jonas et al., 1996). The purpose of much of the research reviewed was to explore surgical methodology so that continuous improvement of surgical technique will result in a reduction of adverse outcome.

#### Psychosocial Outcomes in Children with Congenital Heart Disease

Children with congenital heart disease have also been the focus of studies examining psychosocial and functional outcomes. Topics addressed have included self-perception (Wray & Sensky, 1998), self-esteem (Kitchen, 1978), quality of life (Casey et al., 1994; Laane et al., 1997), behavior (Casey et al., 1996; Janus & Goldberg, 1995; Utens et al., 1993), psychological functioning (DeMaso et al., 1990; Morris et al., 1993), central nervous system function (DeMaso et al., 1990; O'Dougherty et al., 1983), general abilities and school performance (Morris et al., 1993; O'Dougherty et al., 1985; Wright & Nolan, 1994). These studies have assessed the child's functioning in areas that can be related to occupational therapy theory. School performance may be considered a component of productivity in children; and quality of life studies have measured the child's ability to participate in normal childhood activities, including personal and leisure tasks. Psychological functioning, central nervous system function and behavioral

measures tap the performance components, that is, the cognitive, physical and affective skills of the child.

In a review of the literature on children with congenital heart disease, Kitchen (1978) concluded that there was lowered self-esteem in this population that was not related to the severity of the disease process. She also found inconsistent results in the literature investigating relationships between cognitive functioning and congenital heart disease. That is, in some studies, poorer cognitive functioning was associated with lower self-esteem, and in others, this relationship was not demonstrated.

In 1997, Wray and Sensky developed their own assessment tool to measure the self-perception of children with congenital heart disease, using a semantic differential of strength, mood, self-liking, loneliness, safety, anger, goodness and health. Scores for 31 children with congenital heart disease were compared with those of 41 children undergoing bone marrow transplant and those of 44 healthy children. Measures were taken both before and after treatment. Significant pre-treatment differences were reported in self-perception in both the children with congenital heart disease before surgery, and the children undergoing bone marrow transplant, prior to transplant, compared with the healthy group. Children with congenital heart disease improved so much after surgical treatment that there was no difference between them and their healthy peers upon re-assessment, one year after treatment.

Laane and colleagues (1997) studied quality of life in 200 children with congenital heart disease using a standardized instrument, which measured the "essential life spheres" (p. 975) of external living conditions, interpersonal and personal psychological conditions. There were no significant differences between children with

congenital heart disease and control children ( $n = 301$ ), for overall quality of life. There was however a trend toward higher perceived quality of life for the children with congenital heart disease, raising many questions about the concept of quality of life, how it is measured and the role of coping mechanisms.

Casey and colleagues (1994) compared quality of life for 26 children who had complex cyanotic heart disease, with an age and sex-matched group of children with innocent murmurs. Assessments of ongoing symptoms, exercise tolerance and ability to participate in normal childhood activities were the outcome measures. The complex group demonstrated ongoing somatic symptoms, poorer exercise tolerance and some reduction in school attendance. Interestingly, parents were found to have underestimated their child's exercise tolerance in 80% of the children with complex cyanotic heart disease. However it was not always the parents who restricted the children's activity but sometimes the children themselves.

A number of studies have reported poor behavioral adjustment in children with congenital heart disease. As a component of the 1994 study by the same authors, Casey and colleagues (1996) reported on the children's behavioral adjustment and found that children with complex heart lesions showed greater withdrawal, more social problems and less participation in social activities. Janus and Goldberg (1995), in their study of sibling empathy and behavioral adjustment of 28 dyads of children with congenital heart disease and a sibling, also found support for the belief that children with congenital heart disease exhibit greater behavioral concerns than those without. In 1993, Utens and colleagues studied behavioral and emotional problems in 323 children and adolescents with congenital heart disease who underwent surgery between 1968 and 1980. Parent

and self-reported emotional or behavioral problems were compared with those reported for typical peers utilizing standardized assessments. It was found that children and adolescents with congenital heart disease experienced higher problem scores than their peers. Unlike the findings by DeMaso and colleagues (1990), even if the sample was limited to those children with intelligence quotients within the normal range, there were still significantly more problem behaviors noted.

Interestingly, Utens and colleagues (1994) completed a similar study on 288 young adults with congenital heart disease and found favorable outcomes in emotional, intellectual and social functioning. The authors discussed possible mechanisms, such as denial, that may play a role in promoting more positive psychosocial outcomes, as these children become young adults.

In 1993, Spurkland and colleagues reported on the mental health and psychosocial functioning of 26 adolescents with complex congenital heart disease. This group was matched for sex and age with a comparison group of 26 adolescents who had repaired atrial septal defects. Psychiatric assessment, a parent-reported behavior checklist and a somatic assessment were completed. One-third of the children in the complex group were found to be seriously dysfunctional on the Children's Global Assessment Scale (Schaffer et al. 1985, as cited in Spurkland et al., 1993), compared with 4% (one adolescent) in the comparison group. The authors concluded that adolescents with complex congenital heart disease are at high risk for psychiatric disorder. However, those adolescents with higher physical functioning performed better, suggesting that good physical functioning may be a protective factor.

Adaptive functioning has been measured in a number of different ways. Wright and colleagues (1985) studied 188 young adults with congenital heart disease utilizing muscular endurance, motor ability tests, and a self-administered questionnaire about play, leisure, restrictions imposed on them as children, and their current school and work experiences, social patterns and problems. They concluded that physical functioning scores were below average for this group and that the young adults described feeling different or frustrated as children in 30% of cases, and left out socially in 17% of cases.

O'Dougherty and colleagues (1983) constructed a risk model to determine later competence in 31 children with transposition of the great arteries (hereafter TGA) who had survived the Mustard procedure between 1967 and 1977. The model addressed multiple medical and socioeconomic risk factors in this population. Neurological examination, intelligence testing, perceptual-motor and behavioral testing, electroencephalogram, Pattern Visual Evoked Potential, socioeconomic status, and current life stress indicators were utilized. While 80% of the children demonstrated average or above average intelligence, 42% of the total group exhibited learning disabilities or behavior problems requiring specialized class placement or individualized programs. Seventy-seven percent of the sample had an abnormal neurological examination. They found that children who sustained one negative medical or socioeconomic event were more resilient than those who went on to experience multiple events. The role of prolonged hypoxia in overall development was addressed, with the implication that the earlier the heart repair, the better the outcome. Children undergoing the Mustard procedure in those years were typically older than 2 years of age. In 1985, from analyses of data gathered in the 1983 study by the same authors,

O'Dougherty and colleagues reported that prolonged hypoxia negatively impacted neurological motor functioning, cognitive functioning and school achievement.

DeMaso and colleagues (1990) completed a retrospective study of children with congenital heart defects using New England Regional Infant Cardiac Program data. They compared psychological functioning and global central nervous system functioning among 63 children with TGA, 77 children with tetralogy of Fallot (hereafter TET), and 36 children who spontaneously recovered from congenital heart defects. A large battery of psychometric tests was completed during the New England follow up study when the children were between 5.5 years and 6.3 years. All of these children had been diagnosed between 1968 and 1972. A global psychological functioning scale and global central nervous system dysfunction scale were constructed to amalgamate the scores from testing. Both the children with TGA and those with TET had poorer global psychological function and greater central nervous system dysfunction than the group that spontaneously recovered. When controlling for low intelligence quotient and central nervous system dysfunction, however, these children did not demonstrate significantly more emotional disorders than the recovered group.

Morris and colleagues (1993) assessed the neurological, academic and adaptive functioning of children who survived cardiac arrest in hospital. They utilized a battery of standardized assessments to study 25 children, 2 to 15 years of age, 20 of whom had congenital heart disease. While more than 80% of the children were found to have a normal neurological examination, greater than 50% of the children performed at or below the first standard deviation below the normative mean on variables related to intelligence, visual perceptual functioning, fine and gross motor control, achievement and adaptive

behavior. The authors concluded that, despite a normal neurological exam, many of these children would require special education services. This study addressed a particular subgroup of children with congenital heart disease, those that suffer cardiac arrest. However, the authors questioned whether it was the presence of congenital heart disease that caused the poor outcome as opposed to the arrest.

In a follow up to the Morris et al. (1993) study, Bloom et al (1997) studied the effect of cardiac arrest on 32 children with congenital heart disease. They compared 16 children who experienced cardiac arrest with 16 children who did not. The groups were assessed for general cognitive functioning, gross and fine motor skills, memory and adaptive behavior. The group who experienced cardiac arrest performed significantly poorer on each assessment, in comparison to those who did not have a cardiac arrest. The mean performance for the group who did not sustain cardiac arrest was close to the test mean for all assessments with the exception of adaptive behavior, which was lower. The authors concluded that while those with cardiac arrest potentially experienced reduced neuro-psychological or adaptive development, the relationship between cardiac arrest and overall disease severity was more predictive of outcome than cardiac arrest on its own.

Wright and Nolan (1994) studied 29 children between 7 and 12 years of age with TGA or TET who underwent surgical repair between 1979 and 1984. This group was compared to 36 children who were diagnosed with minor defects, such as small atrial septal defects or innocent murmurs. Comprehensive assessment of school performance in reading, writing, arithmetic, self-perception, Wechsler Intelligence Scale for Children - Revised (Wechsler, 1974, as cited in Wright & Nolan, 1994), parent rated behavior checklist, and motor and neurological examinations were conducted. Children with

repaired TGA or TET performed significantly worse in academic tests, had lower intelligence quotients, greater parent-reported behavioral problems, and poorer balance responses than the comparison group. However, self-perception ratings, short-term memory and teacher-reported maladjustment were not significantly different between the two groups. The authors concluded that children with TGA or TET were at great risk of having school-related difficulties.

#### Exercise Rehabilitation for Children with Congenital Heart Disease

In the 1980's and 1990's, research included not only survival data but also outcomes such as exercise tolerance and the effects of physical training on these children's performance (Calzolari et al., 1990; Goldberg et al., 1981; Ruttenberg et al., 1983; Thomassoni et al., 1990). The findings of these studies, that exercise tolerance and aerobic capacity can be improved in children after cardiac surgery, have resulted in recommendations for the development of pediatric cardiac rehabilitation programs (Galioto & Tomassoni, 1992; Koster, 1994; Thomassoni, 1996). The focus of these programs to date has been to provide exercise training. Indeed, Galioto and Tomassoni (1993) define cardiac rehabilitation as "supervised, progressive exercise training to improve aerobic fitness in patients with impaired cardiovascular responses to exercise." (p. 50).

Many exercise rehabilitation research papers have also indicated concurrent influence on development of psychosocial factors. Findings have included increases in independence, initiative, and self-confidence (Calzolari et al., 1990), adaptations to daily living (Galioto, 1990), increases in socialization (Ruttenberg et al., 1983), and a "more

normal lifestyle" (Goldberg et al., 1981; Koch et al., 1988). While these papers presented largely anecdotal evidence, Rowland (1990) supported their findings in his belief that the beneficial effect of exercise on factors such as stress, development of self-concept and maintenance of mental well-being in children was well recognized.

### Conclusion

Differential history limits the generalizability of the cited studies. The time delay between the diagnosis and management of the defect and the study outcomes presents difficulties generalizing these findings to today's children who experience different medical and surgical interventions. TGA is now typically treated by the more successful arterial switch operation in the first few days of life, rather than the Mustard procedure (Wright & Nolan, 1994). When many of the children in these studies were assessed and treated, children with multiple problems or syndromes such as Down syndrome were not being treated for their heart defect (Carr, 1995; Utens et al., 1993; Utens et al., 1994). Today in British Columbia, children with Down syndrome account for 10% of the cardiac population (Cender, March, 1998; personal communication), significantly altering population characteristics. A number of these studies assessed young adults as a means of recalling the child's experience (Utens et al., 1994; Wright et al., 1985). A number of studies were retrospective (DeMaso et al., 1990), or utilized non-standardized assessments (Kitchen, 1978; Wright et al., 1985). Overall, vast arrays of different assessment tools have been utilized to measure many aspects of functioning in children with congenital heart disease.

The research cited suggests that children with surgically repaired congenital heart disease are at risk for a wide range of difficulties. They experience poor self-esteem or self-perception (Kitchen, 1978; Wray & Sensky, 1998), behavioral difficulties (Casey et al., 1996; Janus & Goldberg, 1995; Morris et al., 1993; O'Dougherty et al., 1985; Utens et al., 1993), and poor psychological or psychosocial functioning (DeMaso et al., 1990; Spurkland et al., 1993; Wright et al., 1985). This group is at risk for central nervous system damage (DeMaso et al., 1990; Limperopoulos et al., 1999; O'Dougherty et al., 1983), and compromised school performance (O'Dougherty et al., 1985; Wright & Nolan, 1994). However, some contradictory results are also present. Wright and Nolan (1994) found no difference in self-perception, Morris and colleagues (1993) reported normal neurological exams, and Laane and colleagues (1997) showed a trend for higher quality of life in the children with cardiac problems.

The difficulties these children have been shown to experience in exercise tolerance, behavior, self-esteem, psychosocial functioning, and central nervous system functioning suggest that they do not bring the same level of physical, affective and cognitive skills to their childhood tasks and occupations as their typical peers. With poorer development of these performance components, these children are at risk for reduced ability to participate in age-appropriate childhood occupations of self-care, productivity and leisure. Wright and Nolan's (1994) finding that these children are under-performing at school supports this conclusion.

Despite findings that children with congenital heart disease are under-performing in many areas, only cardio-vascular exercise programs are being advocated as the means of rehabilitation of this population (Galioto & Tomassoni, 1992; Thomassoni, 1996).

This trend apparently relies on anecdotal reports that exercise will positively impact the overall quality of life and psychosocial functioning of these children. However it appears from the literature reviewed that congenital heart disease can have a wide-ranging effect on the adaptive behavior or occupational performance of children.

Occupational therapists are rehabilitation specialists and, as such, play a role in the identification of needs and advocacy for appropriate rehabilitation programs for children with congenital heart disease. Occupational therapy theory provides a rationale to support a structured look at how these children are doing in their daily lives. Exercise rehabilitation is a logical and well-supported treatment option for children with heart disease. However, these children have also been shown to have significant difficulties both in the development of skills at the person level of the CMOP (CAOT, 1997) and in adapting to daily living as seen in their ability to successfully participate in meaningful occupations of childhood.

As cardiac rehabilitation program planning becomes more commonplace for children, a holistic approach to needs identification is warranted. Some studies into exercise rehabilitation report anecdotal evidence of improvements in psychosocial areas. None of them, however, provided empirical evidence of these findings. For comprehensive rehabilitation programs to be developed and implemented, occupational performance outcomes can and should be assessed using well-developed and standardized assessment tools.

### Chapter III: Study Design and Methods

The following section outlines the study's methodology, including subject selection, recruitment and characteristics. Selection criteria, reliability and validity of the assessment tools are presented along with methods used to establish intra-rater reliability. Finally, procedures used to complete the project and analyze the data are provided.

#### Design

The project used a prospective descriptive research design to describe the occupational performance characteristics of children and adolescents between 6 and 16 years of age who had congenital heart disease. The independent variable was therefore congenital heart disease, and the dependent variables were the occupational performance characteristics. Retrospective chart audits were used to identify children and adolescents who met the inclusion criteria for the grant funded project Demonstration Project to Evaluate the Benefits of Exercise Rehabilitation in Children with Congenital or Acquired Heart Disease (Sandor 1998-1999). The children who participated in the overall project constituted the sample of interest for this study.

#### Sample

##### Subject Recruitment

The children and adolescents recruited had received surgical and medical management for TGA, TET or had undergone the Fontan procedure for complex or univentricular heart. Because the primary grant-funded project had an experimental

component designed to stress the exercise potential of these children, children exhibiting the following conditions were excluded, 1. surgery within the previous six weeks; 2. uncontrolled arrhythmias; 3. severe aortic or pulmonary stenosis; 4. Eisenmenger's complex; 5. obstructive hypertrophic cardiomyopathy; 6. acute inflammatory myocardial disease; 7. echocardiographic windows that were not suitable for obtaining the measurements required; 8. special needs requiring one-to-one supervision; or 9. children who were unwilling or unable to cooperate. Potential candidates were screened by their primary cardiologist to ensure physician permission prior to family contact. Families of those children who satisfied the inclusion and exclusion criteria were contacted by letter and invited to participate (see Appendix A).

Informed consent was obtained by having the parent, or legal guardian of the child, sign a consent form (see Appendix B). All children recruited were assessed for cardiac, exercise, development and functional abilities. Occupational performance components were assessed by the occupational therapist on the second visit of each child, prior to any other testing that day.

There were 82 children and adolescents between 6 and 16 years who had TGA, TET or undergone the Fontan procedure who lived in the Lower Mainland of British Columbia, Canada, and were deemed eligible by their cardiologist for inclusion in the project. On invitation, 35 families agreed to participate. This sample of 35 children and adolescents aged between 7.0 and 16.5 years constituted 42.7% of the sampling frame of those eligible. The relative frequencies of each diagnosis are presented in Table 1, and demonstrate comparable frequencies between the sampling frame and the final sample. Using DeMasó et al.'s (1991) Cardiologist's Perception of Medical Severity rating scale

(see Appendix C), each subject's cardiac diagnosis was rated at either level 4 or 5, indicating that they had marked or severe disorders.

Previous studies have found the ratio of boys to girls to differ according to the cardiac diagnosis. The ratio of boys to girls with TGA was found to be 4:1, for those with TET the ratio of boys to girls was 1:1, and in those who undergo the Fontan procedure the ratio of boys to girls was just under 2:1 (Hoffman, 1990; Silber, 1987). In this sample, the gender balance was not reflective of the epidemiological findings. The numbers of participants with TGA or who had the Fontan procedure were quite small, however, boys with TGA out-numbered girls 2:1 and girls who had the Fontan procedure out-numbered boys 2:1. In the largest group, those with TET, where the literature suggests equal incidence, boys out-numbered girls 3:1. It is unclear whether girls were more likely to have increased cardiac severity and therefore be disqualified by their cardiologists, or whether some other gender bias, for example that it was deemed more important for boys to participate in an exercise rehabilitation project than girls, was operating in this group. As those with TET constituted 60% of the sampling frame, this gender bias was also reflected in the final sample.

Table 1.

Frequency table of diagnoses and gender in the sampling frame and sample

Group	N (%)	n (%)
Total	82 (100%)	35 (42.7%)
TGA	21 (25.6%)	10 [28.6%]
TET	49 (59.8%)	22 [62.9%]
Fontan	12 (14.6%)	3 [8.6%]
Girls	26 (31.7%)	12 [34.3%]
Boys	56 (68.3%)	23 [65.7%]

Note. N = number of children in sampling frame; n = number of children in sample;

Values in squared brackets represent percentages of the sample.

Power analysis.

Initial sample size analysis indicated that for a desired power of 0.8 and a moderate effect size, a sample of 54 subjects would be required. However, a moderate to large effect size was found for five of the seven hypotheses and power analyses conducted indicated powers of 0.91 – 0.99% probability of rejecting a false null hypothesis in these cases (see Table 2). There was insufficient power to detect a significant difference in fine motor and visual perceptual skills. For these two hypotheses, the power of the study to detect a moderate effect size was calculated and will be presented with the results.

Table 2

Table of the study's power to detect a false null hypothesis

Hypothesis	n	Effect size	Power
1.1 Self-care skills	35	0.84	>0.99
1.2 Communication skills	35	1.03	>0.99
1.3 Fine motor skills*	31	0.19	0.01
1.4 Visual motor integration skills	35	0.66	0.91
1.5 Visual perceptual skills	35	0.17	0.06
1.6 Motor control skills	35	0.95	>0.99
1.7 Socialization skills	35	1.17	>0.99

Note. \* n = 31 as 4 participants exceeded the test age limit.

Subject Characteristics

The medical records of the participants were reviewed to obtain demographic and descriptive data. Sample characteristics related to age of child, gender of child, parent interviewed and co-existing diagnoses have been provided in Table 3.1.

Table 3.1

Summary table of sample characteristics

Variable	Mean (SD)	n (%)
Age (years)		
Total	11.23 (2.6)	35 (100%)
Girls	12.17 (3.2)	12 (34.3%)
Boys	10.74 (2.3)	23 (65.7%)
Parent interviewed		
Mother		27 (77.1%)
Father		8 (22.9%)
Co-existing diagnosis		
Di George		2 (5.7%)
Hypospadias		2 (5.7%)
Developmental delay		3 (8.6%)
Cleft lip/palate		1 (2.9%)
Asthma		2 (5.7%)
Enuresis		2 (5.7%)
Otitis media		1 (2.9%)
Other		3 (8.6%)
Prematurity ( $\leq 37$ weeks) [n = 32]		3 [9.4%]

Note. Values enclosed in parentheses represent standard deviations (SD) or percentages (%). Values enclosed in square brackets represent number for whom data were available.

Co-existing diagnosis = those participants with non-cardiac co-morbidity.

Additional data relating to medical severity were collected, such as cardiac surgical parameters and complications. All participants underwent definitive repair between 1983 and 1995 (median year = 1989). A cross-tabulation of the type and frequency of surgical procedures experienced by each diagnostic group are presented in Table 3.2, along with data describing the median age of participants at first, definitive, and last surgical procedures. Balloon septostomy and Blalock Taussig shunts are both considered interim or palliative procedures that improve flow or oxygenation of blood but do not repair the defect. The TET repair, atrial or arterial switch, Rastelli and Fontan are all considered definitive repairs (Le Blanc & Williams, 1993), although residual cardiovascular dysfunction is common. A number of participants also required subsequent surgeries, such as valve repair or replacement, or conduit replacement as they outgrew implants or due to ongoing dysfunction.

Table 3.2

Type and frequency of cardiac surgical procedures experienced by participants

Surgical procedure	TGA	TET	Fontan
	C (o)	C (o)	C (o)
Balloon septostomy	8 (8)	0	1 (1)
Blalock Taussig shunt	5 (6)	11 (14)	3 (5)
TET repair	-	22 (22)	-
Atrial switch – Mustard/Senning	5 (5)	-	-
Arterial switch	2 (2)	-	-
Rastelli procedure for TGA	3 (3)	-	-
Coarctation repair	1 (1)	0	0
Glenn anastomosis	-	-	0
Fontan	-	-	3 (3)
Other procedure, not at time of full repair			
Valve repair/replacement	0	1 (1)	0
Conduit repair	0	1 (1)	0
Pacemaker insertion	0	1 (1)	0
Median age in months			
First procedure	[3.33]	[6.9]	[1.0]
Definitive repair	[9.8]	[19.3]	[45.5]
Last procedure	[9.8]	[20.1]	[45.5]

Note. C = number of children who had procedure; (o) = total number of occurrences of the procedure; values in square brackets represent median number of procedures.

Cardiac surgical parameters experienced by participants are presented in Table 3.3 and relate to the whole sample. Oxygen saturation obtained prior to definitive repair and hemoglobin on day of definitive repair, were used as indicators of the degree of hypoxia and the potential incidence of reduced blood flow due to increases in hemoglobin. Both of these parameters have been implicated in studies addressing neurological outcome in children with congenital heart disease (Kirkham, 1998; O'Dougherty et al., 1985). Severe hypoxia and polycythemia result in increased blood viscosity and subsequent "peripheral sludging" (Park & Neches, 1993, p. 442), which may result in an increased incidence of cerebro-vascular accidents due to clotting abnormalities. Normal oxygen saturation is 100%, although values in the mid to high 90's are acceptable. Hemoglobin levels vary according to age, with values as high as 19 grams per deci-liter (g/dL) in the newborn. Hemoglobin levels then fall, with 10 to 13 g/dL considered normal in children over three months of age (Walters & Abelson, 1996). The high median hemoglobin level and low median oxygen saturation experienced by this group provided some indication that participants may have been at risk of neurological insult due to these factors. However, the interactions and relationships between these parameters and potential neurological insults such as cerebro-vascular accidents, is complex involving other factors that are beyond the scope of this review (Cottrill & Kaplan, 1973; Greely, 1996; Park & Neches, 1993).

Numbers of hospitalizations, length of critical care required and the number of surgeries participants experienced, all provided indicators of disease severity for this group. Days of inotropic support provided an indication of the degree of cardiac support required immediately following cardiac surgery. Low cardiac output after

cardiopulmonary bypass may be related to poor ventricular function or pulmonary hypertension (Greely, 1996). Inotropic support included infusions of dopamine, dobutamine or adrenalin that were provided to assist with cardiac output. There was only one participant who received vancomycin and gentomycin antibiotic therapy for a short period after cardiac surgery.

Table 3.3

Disease severity indicators

Variable	Median	Min/max	Percent	C (o)
O <sub>2</sub> sat. before definitive repair (%) [n = 34]	81.5	51 / 99		
Hb. before definitive repair (g/dL) [n = 32]	15.0	10.1 / 21.7		
<b>Hospitalizations</b>				
Number of admissions	3	1 / 15		35 (129)
Number of days admitted	23	7 / 33		35 (1138)
Number ICU days	11	2 / 32		35 (436)
Number of days ventilated	6	2 / 25		35
Number of days inotropic support	4	0 / 16		35
<b>Surgeries</b>				
Cardiac	2	1 / 4	100%	35 (68)
Open heart	1	1 / 2	100%	35 (38)
Closed heart	1	0 / 3	60%	21 (29)
Sternal closure post open-heart	0	0 / 2	28.6%	10 (12)
Cardiac catheterization	2	1 / 4	100%	35 (72)
Non-cardiac	0	0 / 6	40%	14 (27)

Note. Percent = percentage of participants experiencing procedure; C = number of children; (o) = occurrences; O<sub>2</sub> Sat. = oxygen saturation; Hb. = hemoglobin; ICU = intensive care unit. Values in square brackets indicate number for whom data were available.

Greater detail regarding the type and parameters of cardiopulmonary bypass are also provided in Table 3.4 as many authors have explored the relationships between these procedures and the incidence of gross or subtle neurological complications. The components of cardiac surgery that have been correlated with adverse neurological outcomes are those to do with managing circulation and cell metabolism during surgery, that is, duration of deep hypothermia, cardiac bypass, circulatory arrest or low flow and reperfusion (Jonas, 1998; Jonas et al., 1993; Kirkham, 1998; Park & Neches, 1993).

Unfortunately, recording of surgical parameters had changed significantly over the period that the participants underwent their open-heart surgeries, and some parameters that are now considered important were not recorded for a number of participants. Those participants who experienced cardiopulmonary bypass between 1983 and 1987 had a few bypass parameters recorded on the surgeon's written report. Participants who underwent cardiopulmonary bypass between 1989 and the early to mid 1990's had further details recorded on a form entitled 'Perfusion Operative Report'. From 1995, comprehensive data were clearly and consistently recorded on the 'Perfusion Services Cardiopulmonary Bypass Information Record'. Data not available from either of the perfusion records were obtained from the anesthetist's log, or the surgeon's report. Consequently Table 3.4 provides limited data for the length of time participants were cooled, the length of time kept at coldest temperature and the coldest temperature reached. These are parameters that are known to place children at risk for neurological damage, however, due to data recovery difficulties, the data that have been presented should be interpreted with caution.

Kirkham (1998) has proposed safety limits, suggesting that at 28° C, circulatory arrest can be safely managed for 11-19 minutes, and at 18°C it can be managed for 39-65 minutes. In addition to the data presented in Table 3.4, there were 26 participants for whom data related to the length of time at coldest temperature were available. Of these, 12 experienced deep hypothermia (16 –20°C), and 14 experienced moderate hypothermia (22 - 30°C). While many of these children were cooled for long periods, none were in circulatory arrest for longer periods than those recommended by Kirkham (1998). Recent literature now also highlights the importance of the rate of cooling and re-warming; (Jonas et al., 1996; Jonas, 1998; Kirkham, 1998), however, investigation of this was beyond the scope of this study.

Table 3.4

Cardiopulmonary bypass parameters

Parameter	Median	Min / Max	Percent	C (o)	
<b>Type of bypass</b>					
Partial			5.7%	2 (2)	
Total, moderate hypothermia ( $\geq 22^\circ$ )			45.7%	16 (16)	
Total, deep hypothermia ( $<22^\circ$ )			20%	7 (7)	
With circulatory arrest			40%	14 (15)	
<b>Bypass parameters<sup>f</sup></b>					
Bypass (minutes)	n = 35,5	106	15 / 181	100%	35 (40)
Cross clamp (minutes)	n = 35,5	56.5	0 / 141	100%	35 (40)
Circulatory arrest (minutes)	n = 35,5	0	0 / 64	37.1%	13 (15)
Total hypothermia (minutes)	n = 28,5	140	34 / 280		28 (33)
Time at coldest (minutes)	n = 23,5	52.5	15 / 205		26 (28)
Temperature (Celsius)	n = 34,5	24	16 / 32		34 (39)

Note. C = number of children who had procedure; (o) = number of occurrences;  $f = 5$

children had 2 bypass surgeries; all bypass values were obtained after combining 1<sup>st</sup> & 2<sup>nd</sup>

bypass; n = number of children for whom data were available for 1<sup>st</sup>, 2<sup>nd</sup> bypass.

A significant proportion of participants had complications related to their cardiac surgery as can be viewed in Table 3.5, although only one participant experienced a significant neurological event, hypoxic ischemic encephalopathy, as an intra-surgical event. This participant underwent surgery in another Canadian province thus limiting the data available to us; however, it appears that the most significant impact was that of cortical visual impairment that resolved over time. This participant and two others had seizures post-operatively; none however remained on seizure medication after discharge from the time of their definitive repair.

Table 3.5

Frequency and type of complication experienced due to cardiac condition or surgery

Complication	n	Percent
Pre-operative, cardiac arrest	1	2.9%
Post surgical		
Sepsis	2	5.7%
Prolonged chest tube drainage	8	22.9%
Post pericardial syndrome (PPS)	8	22.9%
Peripheral nerve injury	4	11.4%
Seizures	3	8.6%
Cerebro-vascular accident	0	0
Cardiac or respiratory arrest	0	0
CNS infection	0	0
Myocardial infarct	0	0
Wound infection	4	11.4%
Encephalopathy	1	2.9%
Other	5	14.3%
None	10	28.6%

Note. Some children had more than one complication. Peripheral nerve injury = Horner's syndrome (3), laryngeal nerve injury (1); Other = Renal or peripheral dialysis (2), deep vein thrombosis (1), significant bleeding (2).

Most participants had some form of relatively minor residual cardiovascular dysfunction as seen in Table 3.6. On entry to the study, however, all but one individual had normal oxygen saturation, and only six were taking medications, either Coumadin (2), or Aspirin (3) to reduce the risk of blood clotting, or Enalapril (1) for hypertension. In Table 3.6, flow obstruction or regurgitation included pulmonary stenosis, pulmonary regurgitation, tricuspid regurgitation or outflow obstruction. Hypertrophy included atrial hypertrophy (1), right ventricular hypertrophy (10), left ventricular hypertrophy (1) and cardiac enlargement (1). Dysrhythmia was classified as second or third degree heart block requiring pacing or medication for management. One of these individuals had a pacemaker inserted after the definitive repair, and one was scheduled to have a pacemaker inserted two months after this assessment. Conduction disorders were relatively minor and included first-degree heart block, right bundle branch block, or intraventricular conduction delay, which are expected to occur after the myocardium is cut. None of the conduction disorders required medical or surgical intervention. Two participants were excluded from the demonstration project after the initial assessment as they were found to require further surgery, one for pacemaker insertion and the other for conduit replacement.

At time of entry to the study, the participants were between 0.4 and 15.2 years (median = 8.2 years) past their last procedure, and between 2.4 and 15.2 years (median = 9.3 years) past their definitive repair.

Table 3.6

Residual cardiac dysfunction or other disorder

Residual difficulties	n	Percent
<u>Post discharge/on follow-up</u>		
Urinary tract infections	2	5.9%
Psychiatric assessment	1	2.9%
Migraine	1	2.9%
<u>Residual cardiac dysfunction</u>		
Minor flow obstruction or regurgitation	13	37.1%
Mod. or Sig. flow obstruction or regurgitation	3	8.6%
Minor residual VSD or patch leak	7	20%
Hypertrophy	13	37.1%
Dysrhythmias	2	5.9%
Conduction disorders	25	71.4%
Pacemaker inserted	1	2.9%
<u>On entry to study</u>		
Oxygen saturation's at rest < 96%	1	2.9%
Medications	6	17.1%
Known to require further surgery	2	5.9%

Note. Many participants had more than one dysfunction. CNS = central nervous system;

VSD = ventricular septal defect; Mod. = moderate; Sig. = significant.

In summary, the relative frequency of both gender and diagnosis within the sample of 35 suggested that this group could be considered demographically representative of the sampling frame. There is, however, a gender bias towards boys in comparison to the literature. The disease severity data indicated that the participants experienced significant surgical and medical interventions in the first few years of life, placing them at risk for gross and/or subtle neurological deficits that may have impacted their performance on the assessments utilized within this study. There were, however, only two individuals with known congenital syndromes, one diagnosed early in life, and the other, 6 months after the study began, and one other participant who experienced a known, significant neurological event.

### Measures Utilized

#### Instrument Selection

Appropriate measurement tools were selected using the following criteria: 1) the measurement tool assessed occupational performance areas pertinent to children between the ages of 6-16 years, including self care, productivity and leisure tasks; 2) the tools were standardized to enable comparison with a normative sample; and 3) the instruments had been used in previous cardiac studies so that information obtained built on the developing knowledge of this population. However, child fatigue and the need to conserve the power of the study limited the final selection of measurement tools.

Occupational performance assessment involved measurement of the child's participation in self-care, productivity and leisure tasks. These areas encompass all those things that people need to do to look after themselves and others, to enjoy life and

ultimately to contribute to their community (CAOT, 1997). As previously discussed, a child's ability to participate in childhood occupations depends, in part, on the skills and abilities they possess in physical, cognitive and affective areas. Some aspects of these performance components were also assessed.

#### Self-care.

Self-care tasks were defined as the development of personal independence in personal hygiene, eating, dressing, and personal safety. In the older child and adolescent, self-care also encompassed household and domestic responsibilities, access and utilization of community resources and development of basic work habits. The daily living skills domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) provided a standardized, semi-structured interview format for evaluating the development of these skills, as perceived by the parent. The daily living skills domain had three sub-domains: personal, domestic and community living skills. The personal domain assessed personal hygiene, independence in feeding, dressing and toileting, and the individual's ability to look after their own health and wellbeing. The domestic sub-domain assessed the individual's ability to contribute to, or complete household tasks including cooking, cleaning and basic home-maintenance. The community skills sub-domain assessed personal safety in the community, the ability to use resources such as the telephone, go shopping, develop a budget and develop work skills.

The Vineland Scales have been used in previous research. DeMaso and colleagues utilized an earlier Vineland Social Maturity Scale (1965, as cited in DeMaso et al., 1990) in their study of children with TGA, TET and spontaneously recovered peers. In this study, the Vineland results were incorporated into a global psychological

functioning scale. Seventy percent of children with TGA, 75% of children with TET and 55% of the spontaneously recovered group demonstrated mild to severe difficulties on the global psychological functioning scale. Morris and colleagues (1993) also used the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) to study children surviving cardiac arrest and Bloom et al (1997) used the Vineland to compare outcomes of children with congenital heart disease who did or did not sustain cardiac arrest.

### Productivity.

Productivity in childhood was defined as the child's ability to engage in learning through movement, play and school or academic tasks. Productivity was measured using the communication domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984), which tapped the parent's perception of how their child or adolescent was succeeding at school. The communication domain provided a standardized format for evaluating the parent's perception of the development of writing, reading and expressive language as components of communication. These skills provided a measure of the child's ability to participate in productivity tasks as related to school. The use of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) in cardiac studies has been previously outlined.

In addition to the Vineland assessment, the fine motor sub-tests of the Bruininks Oseretsky Test of Motor Proficiency (Bruininks, 1978), and the Beery-Buktenica Test of Visual-Motor Integration (Beery, 1997) were administered. These three assessments did not measure occupational performance. They were used to evaluate some of the underlying performance components that may contribute to success in tasks of productivity. The Beery-Buktenica Test of Visual-Motor Integration (Beery, 1997) was

designed to assess the child's ability to copy increasingly complex geometrical designs. Visual motor integration was defined as "the degree to which visual perception and finger hand movements are well coordinated" (Beery, 1997, p. 19). Since the development of the original version of this test by Beery in 1967, it has been extensively used in educational, psychological and medical fields (Beery, 1997). The child's ability to copy geometric designs has been shown to correlate significantly with academic achievement (Wright & DeMers, 1982, as cited in Beery, 1997) and with neurological problems (Williams & Ashcroft, 1993, as cited in Beery, 1997). This suggested that scores gained on the Beery test would provide additional information on the underlying skills these children bring to the tasks of productivity. In addition to assessing visual motor integration, the 1997 version has two supplemental components, designed to measure visual perception and motor control skills. These components assist in isolating the skills required to complete the visual motor integration component. They were administered to provide an indication of the participants' ability to develop cognitive and physical performance components at the person level of the model.

The Beery Test of Visual Motor Integration (Beery, 1967, as cited in Morris et al., 1993) has been used in previous outcome studies in children with congenital heart disease. Morris and colleagues (1993) assessed the neuro-psychological status and achievement of children who survived in-hospital cardiac arrest. Sixty-four percent of the children assessed, scored at or below one standard deviation below the mean on the test of Visual Motor Integration.

The upper limb coordination and fine motor composite of the Bruininks-Oseretsky Test of Motor Proficiency (Bruininks, 1978) were used to assess the child's

ability to engage in childhood tasks requiring fine motor skills. These sub-tests provided information about the child's ability to use their arms and hands quickly, in coordination with their eyes, in order to participate in higher level motor tasks. The fine motor composite had three sub-tests, response speed, which measured how quickly the hand stopped a moving visual stimulus; visual motor control, which measured the ability of the child to complete a number of paper and pencil tasks; and upper limb speed and dexterity, which measured the ability of the child to move the arms and hands with speed and precision.

Bruininks (1978) cited literature to support the importance of speed of response, integration of visual-perceptual and motor responses and precision of fine motor responses in the development of psycho-motor abilities (Cratty, 1967; Fleishman, 1964; and Guilford 1958 all cited in Bruininks, 1978), for success in reading and handwriting (Wedell, 1973, as cited in Bruininks, 1978) and for skilled motor performance in activities of work, play and sport. Thus the development of this tool supported its use as a measure of the physical performance skills of participants which may contribute to their ability to participate in tasks of productivity as well as other aspects of occupational performance.

Selected sub-tests of the Bruininks Test of Visual Motor Integration (Bruininks, 1978) were utilized by Wright and Nolan (1994) in their study on the impact of heart disease on school performance. Children with heart disease performed worse than controls on the balance sub-test of the assessment, but not on the visual motor or fine motor measures. Wright and Nolan (1994) also found a positive correlation between performance on the balance sub-test of the Bruininks-Oseretsky Test of Motor

Proficiency (Bruininks, 1978) and overall outcome, regardless of whether the children were in the control or the case group.

#### Leisure.

Leisure in childhood was defined as those activities done for enjoyment, social interaction and personal reward. The socialization domain of the Vineland Adaptive Behavior Scale (Sparrow et al., 1984), was used as a measure of interpersonal relationships, play and leisure participation, and coping skills.

#### Additional Measures.

The Maladaptive Behavior Domain of the Vineland Adaptive Behavior Scale (Sparrow et al., 1984), measured undesirable behaviors that can interfere with a child's adaptive functioning or ability to participate in childhood occupations across all three domains: self-care, productivity and leisure. Behavior problems were identified by a number of authors investigating children with congenital heart disease (Casey et al., 1996; Janus & Goldberg, 1995; Morris et al., 1993; O'Dougherty et al., 1983; Utens et al., 1993), although these authors typically used more comprehensive measures than the maladaptive behavior domain of the Vineland. Use of this domain completed the Vineland battery and provided some information on parental perception of behavior in these children.

To balance the parent-perceived behavioral profile from the Vineland (Sparrow et al., 1984), the Self-Perception Profile for Children (and Adolescents) (Harter, 1985; 1988) was administered to tap the children's perception of themselves as academics and as sports-people. It also assessed their perceived popularity with peers, their happiness with their physical appearance, their behavior and their global self worth. The Harter

(1985) profile has been utilized in a study of school performance of children with TGA, (Wright & Nolan, 1994). No differences were seen between the control group (those with cardiac murmur but requiring no treatment) and the case group (those with TGA).

The Self-Perception Profile for Children (Harter, 1985) contained the following domains: 1. Scholastic competence - the child's perception of their ability in scholastic areas. 2. Social acceptance - the degree that the child felt accepted by peers and their sense of popularity. 3. Athletic competence - the child's perception of their ability to participate in sports and games. 4. Physical appearance - how the child felt about their height, weight, looks and general body structure. 5. Behavioral conduct - how the child felt about how they behave, and the extent to which they act the way they are expected. 6. Global self-worth - a general category looking at how much the child liked herself and whether she was happy with the way she lived her life. The global self-worth scale was not intended as a measure of general competence.

Harter later (1988) developed a Self-Perception Profile for Adolescents from grade 8 to grade 11. This profile was divided into nine domains, and expanded the Children's Profile with the addition of the following domains: 1. Job competence - how the adolescent felt about their ability to do work and how well they did at work they had done. 2. Romantic appeal - how attractive the adolescent felt to those in whom they were interested and whether they felt they were fun and interesting when on a date. 3. Close friendship- the ability to make close friends.

In summary, the assessment tools chosen, that is, the Vineland Adaptive Behavior Scales, Beery-Buktenica Test of Visual Motor Integration, and Bruininks Oseretsky Test of Motor Proficiency, provided information on the child's performance across the three

occupational performance domains of self care, productivity and leisure and a limited measure of skill development at the person level of the model. Additionally, the Self-Perception Profiles for Children, and Adolescents (Harter, 1985,1988), provided information on the children's perception of themselves in these three areas. All of the assessments were validated on normative samples and reliability information provided within the manuals was within acceptable limits for the analyses selected for this study. The final selection criterion, that the tools were used in previous cardiac studies, was also met for each assessment, albeit in a limited manner.

### Reliability

#### Test validity and reliability.

Comprehensive data regarding the standardization processes were available in the respective manuals of the assessments chosen. Reliability and validity testing was undertaken with each tool, as outlined in the test manuals, demonstrating that the tools were valid and reliable. This information has been summarized in Appendix D.

#### Intra-rater reliability.

Intra-rater reliability of the examiner was established by re-scoring 10-20% of the sample data, one to three months after the initial scoring. Ten individual workbooks of the Beery-Buktenica test of visual motor integration, the motor control sub-test (Beery, 1997) and five workbooks of the visual motor control sub-test of the Bruininks Oseretsky Test of Motor Proficiency (Bruininks, 1978), were re-scored. Test booklets were selected by chance by a colleague, had identifying features removed, and were re-scored by the examiner. Two other sub-tests of the Bruininks Oseretsky test, that is, the upper limb

coordination and fine motor speed and dexterity sub-test for five subjects were video taped and re-scored, and five semi-structured interviews of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) were audio-taped and re-scored.

Reliability was established using the limits of agreement method as outlined by Altman and Bland (1993; 1986). As 10% of the sample resulted in a very small number of subjects, test-retest scores were combined in the following manner. Both the visual motor integration and motor control components of the Beery were analyzed together ( $n = 20$ ), all three sub-tests of the Bruininks were analyzed together ( $n = 15$ ), and the three domains and the composite score of the Vineland ( $n = 20$ ) were analyzed together. Scores were combined in this manner as visual analysis of the scatter-plots that were constructed demonstrated that an individual's test-retest scores for each of the components within an assessment battery were independent of each other.

Scatter-plots were constructed of the 'difference between the first and second scores' and the 'mean of the first and second scores'. A 95% limit of agreement was established by plotting 2 standard deviations of the difference between the scores. Only one score fell outside the limits on the Beery and Vineland sets, indicating that there was 95% agreement between the scores. In the Bruininks set, 93.3% of the scores fell within the 95% limit of agreement, again with only one score falling outside the 95% limit. The difference between these two results lies in the sample size. In addition, the differences appeared to be normally distributed, suggesting that there was no consistent bias in the test-retest scores. It was therefore determined that the intra-rater reliability was within acceptable limits. Table 4 presents the median, minimum and maximum difference in scores for the three assessment batteries.

Table 4

Table of differences between 1<sup>st</sup> and 2<sup>nd</sup> standard scores obtained in each assessment battery

Test battery	Test $\mu$ ( $\sigma$ )	Median	Minimum	Maximum
Beery n = 20	100 (15)	0	-7.0	5.0
Bruininks* n = 15	50 (10)	0	-1.0	2.0
Vineland n = 20	100 (15)	0	-4.0	6.0

Note.  $\mu$  = test mean; ( $\sigma$ ) = test standard deviation. \* = Raw scores used.

### Procedures

Ethical approval was obtained from both the University of British Columbia and British Columbia's Children's Hospital ethical review committees, prior to initiation of the grant-funded project. Following recruitment and gaining of written consent, each participant and one parent were scheduled for assessment at the British Columbia's Children's Hospital. The child or adolescent attended a 1-hour session to complete the three assessments.

Each participant underwent assessment of occupational performance characteristics of self-care, productivity and leisure, and completed a self-perception profile. Standardized assessment tools were utilized in the following order; 1) Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery, 1997); 2) Fine Motor composite of the Bruininks-Oseretsky Test of Motor Proficiency (Bruininks, 1978); 3) Self-Perception Profile for Children, or Adolescents (Harter, 1985; 1988). These three

assessments took one hour to complete. At the completion of the child or adolescent's assessment, the Vineland Adaptive Behavior Scales interview was conducted privately with one parent, taking from 45 – 75 minutes to complete (Sparrow et al., 1984).

All assessments of the children and adolescents occurred in the same room and utilized standardized equipment and furniture as appropriate to the size of the participant. Parents were interviewed in one of two rooms depending on scheduling. All instructions were provided in the standardized manner defined by each assessment tool. The children then went on to complete the physiotherapy and cardiology assessments related to the grant-funded demonstration project. All occupational therapy assessments were conducted by the author, an occupational therapist, with knowledge and experience in administration of each of the assessment tools utilized. Pre-test training occurred prior to testing the first subject, on five children from an unrelated sample of convenience. Each participant was assessed prior to the parent interview and retrospective chart review. Therefore, the examiner was blind to the specific cardiac diagnosis, any complicating medical conditions and developmental or schooling difficulties experienced by the child or adolescent at the time of the assessment.

#### Data Management and Analysis

Data were managed using the Statistical Software Package for the Social Sciences (SPSS). All data were kept secure and confidential. Child and parent responses to each assessment were collected on paper during the assessment process. Raw scores were then converted to standard scores and results entered onto a statistical spreadsheet designed for the purpose.

### Hypotheses 1.1 – 1.7

Standard scores were obtained for the components of interest in each assessment. Each domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) had a mean of 100 and standard deviation of 15. The fine motor composite for the Bruininks-Oseretsky Test of Motor Proficiency (Bruininks, 1978), had a mean of 50 and standard deviation of 10. The Beery Test of Visual-Motor Integration (Beery, 1997), had a mean of 100 and standard deviation of 15 for each component.

Statistical reasoning was used to describe the distribution and variability of each data set and visual analysis of histograms and stem and leaf plots used to determine that data were essentially normally distributed. This concurred with Glenberg's assertion that "the t-test is robust and can be utilized if the data are not 'grossly skewed' (Glenberg, 1988, p. 262). Therefore, statistically significant differences between children with congenital heart disease and the standardization data of each assessment were tested using a two-tailed independent t-test with experiment-wise alpha set at 0.05.

Alpha level inflation was controlled using the Bonferoni adjustment, as there were multiple t-tests conducted. Alpha was set at 0.0125 ( $0.05 \div 4$ ) for the data collected from the children, with 34 ( $n - 1$ ) degrees of freedom, and a decision to reject the null hypothesis if  $t \leq -2.75$  or  $t \geq 2.75$  (Glenberg, 1988, Table C, p. 510). For data collected from the parents, alpha was set at 0.017 ( $0.05 \div 3$ ), with 34 ( $n - 1$ ) degrees of freedom and a decision to reject the null hypothesis if  $t \leq -2.46$  or  $t \geq 2.46$  (Glenberg, 1988, Table C, p. 510).

### Hypotheses 2.1 – 2.2

Group means for each sub-scale measuring perceived competency in the Self-Perception Profiles for Children (or Adolescents) (Harter, 1985; 1988), were calculated and compared with the mean and standard deviation data provided within the manual. For children, the overall mean provided in the manual was 3.0; however, the test standard deviations fluctuated between .5 and .85 depending on sub-scale and gender. For adolescents, the overall mean provided in the manual was 2.9, and the test standard deviations fluctuated between .5 and .75 also depending on sub-scale and gender. The sub-scale's large variance in comparison to the unit of measurement, suggested that individuals were likely to vary considerably in their responses.

A measure of self-esteem was obtained by calculating the discrepancy between the individuals' mean perceived competence in each sub-scale and the mean importance for each sub-scale. Harter (1985; 1988) suggested that in only those sub-scales where the importance rating was greater than 3.0 would self-esteem be influenced. When the discrepancy between the importance rating and the competence rating was calculated, a positive score indicated success in areas deemed important and therefore high self-esteem. If the result was negative and large, a lack of success in areas deemed important was indicated, and self-esteem was considered to be low. An association between the global self-worth score and this discrepancy score was expected.

## Chapter IV: Results

Occupational Performance Characteristics of Children and Adolescents with  
Congenital Heart Disease

Self-Care Abilities

1.1 The self-care abilities of children and adolescents with congenital heart disease were determined by comparison of their daily living standard scores with the population parameter provided by the daily living skills domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984). Table 5 provides summary statistics of the participant's daily living scores.

Table 5

Table of descriptive statistics related to daily living standard scores

Descriptive	Statistic
Mean	87.40
Median	88.00
Standard deviation	12.51
Minimum	61
Maximum	112
Percent of scores $\leq$ 5 <sup>th</sup> percentile	20.0%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was rejected,  $t = -5.96$  ( $p < .0001$ ). Children and adolescents with congenital heart disease had statistically significantly lower self-care abilities.

### Productivity

1.2. The productivity of children and adolescents with congenital heart disease was determined by comparison of their communication standard scores with the population parameter provided by the communication domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984). Table 6 provides summary statistics of the participant's communication scores.

Table 6

Table of descriptive statistics related to communication standard scores

Descriptive	Statistic
Mean	85.0
Median	87.0
Standard deviation	15.0
Minimum	53
Maximum	118
Percent of scores $\leq$ 5 <sup>th</sup> percentile	28.6%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was rejected,  $t = -5.92$  ( $p < .0001$ ). Children and adolescents with congenital heart disease had statistically significantly lower communication skills.

In addition, it was found that 31.4% ( $n = 11$ ), of participants received learning assistance (LAC) at school, while 11.4% were reported by parents to be "straight A" students.

Performance components.

1.3. The fine motor abilities of children and adolescents with congenital heart disease were determined by comparison of their fine motor composite standard scores with the population parameter provided by the fine motor composite of the Bruininks Oseretsky Test of Motor Proficiency (Bruininks, 1978). Table 7 provides summary statistics of the participant's fine motor scores.

Table 7

Table of descriptive statistics related to fine motor composite standard scores

Descriptive	Statistic
Mean*	50.06
Median	54.00
Standard deviation	13.59
Minimum	23
Maximum	68
Percent of scores $\leq$ 5 <sup>th</sup> percentile	16.1%

Note. Test mean = 50; standard deviation = 10;

\*n = 31, as 4 participants exceeded the test age limit.

The null hypothesis,  $H_0: \mu_1 = 50$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was retained,  $t = 0.026$  ( $p = .98$ ). There is insufficient evidence to suggest that children and adolescents with congenital heart disease had significantly different fine motor abilities.

As there was no clinically or statistically significant difference between the sample and the test mean, the power of this study to detect a moderate effect size was determined and found to be .51 ( $n = 31$ ,  $\alpha = .01$ ,  $\delta = .65$ ; where  $\delta$  is predicted effect size).

Despite there being no difference between the test and sample mean, there was increased variability in the sample data with a standard deviation of 13.59 compared with the test standard deviation of 10, and just over 16% of participants scored at or below the fifth percentile. There are three sub-tests, which are combined to compute the fine motor composite score: response speed, visual motor control and upper limb speed and dexterity. In order to explore the fine motor skills of this group further, descriptive statistics related to the sub-tests of the fine motor composite are presented in Tables 8, 9 and 10.

Table 8

Table of descriptive statistics related to the response speed standard scores

Descriptive	Statistic
Mean*	14.8
Median	15.0
Standard deviation	5.5
Minimum	4
Maximum	27
Percent of scores $\leq 2 \sigma$ below test $\mu$	3.2%

Note. Test mean ( $\mu$ ) = 15; standard deviation ( $\sigma$ ) = 5; \*  $n = 31$ .

There was no clinically significant difference between the participants' scores and the test parameters for response speed.

Table 9

Table of descriptive statistics related to the visual motor control standard scores

Descriptive	Statistic
Mean*	16.4
Median	19.0
Standard deviation	5.7
Minimum	1
Maximum	23
Percent of scores $\leq 2 \sigma$ below test $\mu$	3.2%

Note. Test mean ( $\mu$ ) = 15; standard deviation ( $\sigma$ ) = 5; \* n= 31.

The group mean and standard deviations for visual motor control were not clinically different from the test parameters, however this group achieved a very high median score suggesting negatively skewed data.

Table 10

Table of descriptive statistics related to the upper limb speed and dexterity standard scores

Descriptive	Statistic
Mean*	12.6
Median	14.0
Standard deviation	6.5
Minimum	1
Maximum	23
Percent of scores $\leq 2 \sigma$ below test $\mu$	19.4%

Note. Test mean ( $\mu$ ) = 15; standard deviation ( $\sigma$ ) = 5; \* n= 31.

There was a slightly lower group mean for upper limb speed and dexterity, although this would not necessarily be considered clinically significant. However, over 19% of the group achieved scores that were less than or equal to, two standard deviations below the test mean, which suggested that a sub-group of participants experienced significant difficulty with this component of the fine motor composite.

In addition to the fine motor composite, descriptive data were collected on the upper limb coordination sub-test of the Bruininks Oseretsky Test of Motor Proficiency and participants were found to achieve lower standard scores in this sub-test than the population parameter provided by the test manual. While the difference between the means was not subjected to statistical analysis, the groups' mean of 9.58 was just over one standard deviation below the sub-test population mean and was considered clinically significantly different. Table 11 provides summary statistics of the participant's upper limb coordination scores.

Table 11

Table of descriptive statistics related to upper limb coordination standard scores

Descriptive	Statistic
Mean*	9.4
Median	9.0
Standard deviation	4.6
Minimum	1
Maximum	19
Percent of scores $\leq$ 5 <sup>th</sup> percentile	22.6%

Note. Sub-test mean = 15, standard deviation = 5; \* n = 31.

As there was a similarity between the percentage of participants with poor performance in the upper limb coordination and fine motor composite standard scores, the relationship between these two data sets was explored. There was a moderately low, positive relationship between the fine motor composite standard score and the upper limb coordination standard score ( $r = .45$ ).

1.4 The visual motor integration abilities of children and adolescents with congenital heart disease were determined by comparison of their visual motor integration standard scores with the population parameter of the visual motor integration component of the Beery-Buktenica Test of Visual Motor Integration (Beery, 1997). Table 12 provides summary statistics of the participant's visual motor integration scores.

Table 12

Table of descriptive statistics related to visual motor integration standard scores

Descriptive	Statistic
Mean	89.11
Median	88.00
Standard deviation	19.8
Minimum	49
Maximum	122
Percent of scores $\leq$ 5 <sup>th</sup> percentile	34.3%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was rejected,  $t = -3.26$  ( $p < .003$ ). Children and adolescents with congenital heart disease had statistically significantly lower visual-motor integration abilities.

1.5 The visual perceptual abilities of children and adolescents with congenital heart disease were determined by comparison of their visual perceptual standard scores with the population parameter of the visual perceptual component of the Beery-Buktenica Test of Visual Motor Integration (Beery, 1997). Table 13 provides summary statistics of the participant's visual perceptual scores.

Table 13

Table of descriptive statistics related to visual perceptual standard scores

Descriptive	Statistic
Mean	97.20
Median	98.00
Standard deviation	17.3
Minimum	57
Maximum	124
Percent of scores $\leq 5^{\text{th}}$ percentile	17.1%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was retained,  $t = -0.96$  ( $p = .35$ ). There is insufficient evidence

to suggest that children and adolescents with congenital heart disease had statistically significantly different visual perceptual abilities.

As there was no clinically or statistically significant difference between the sample and the test mean, the power of this study to detect a moderate effect size was determined and found to be .65 ( $n = 35$ ,  $\alpha = .01$ ,  $\delta = .65$ ; where  $\delta$  is predicted effect size).

The variability of the sample data was similar to the test data, however, 17.1% of participants scored at or below the 5th percentile.

1.6 The motor control abilities of children and adolescents with congenital heart disease were determined by comparison of their motor control standard scores with the population parameter of the motor control component of the Beery-Buktenica Test of Visual Motor Integration (Beery, 1997). Table 14 provides summary statistics of the participant's motor control scores.

Table 14

Table of descriptive statistics related to motor control standard scores

Descriptive	Statistic
Mean	85.83
Median	88.00
Standard deviation	13.2
Minimum	56
Maximum	106
Percent of scores $\leq$ 5 <sup>th</sup> percentile	25.7%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was rejected,  $t = -6.35$  ( $p < .0001$ ). Children and adolescents with congenital heart disease had statistically significantly lower motor control abilities.

### Leisure

1.7 The leisure abilities of children and adolescents with congenital heart disease were determined by comparison of their socialization standard scores with the population parameter of the socialization domain of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984). Table 15 provides summary statistics of the participant's socialization scores.

Table 15

#### Table of descriptive statistics related to socialization standard scores

Descriptive	Statistic
Mean	84.97
Median	87.00
Standard deviation	11.9
Minimum	62
Maximum	107
Percent of scores $\leq$ 5 <sup>th</sup> percentile	20.0%

Note. Test mean = 100; standard deviation = 15.

The null hypothesis,  $H_0: \mu_1 = 100$ , (where  $\mu_1$  = sample mean of children with congenital heart disease), was rejected,  $t = -7.45$  ( $p < .0001$ ). Children and adolescents with congenital heart disease had statistically significantly lower socialization skills.

## Additional Measures

### Self-perception profiles.

2.1 The self-perception profiles of children and adolescents with congenital heart disease were determined by comparison with the profiles provided by Harter (1985; 1988), and found to fit the normative pattern. Group means for each competence sub-domain were within one standard deviation above or below the mean established by Harter (1985; 1988). Table 16 provides descriptive statistics related to how competent the participants felt in each of the domains. There were only 34 subjects for this component as one participant did not understand the concept well enough to complete the task.

Table 16

Table of descriptive statistics related to group scores for the 9 competence sub-domains of the Self-Perception Profiles for Children (and Adolescents)

Competence Sub-domain	n	Minimum	Maximum	Mean (SD)
Scholastic competence	34	1.00	3.83	2.70 (.70)
Social acceptance	34	1.17	4.00	3.12 (.77)
Athletic competence	34	1.40	4.00	2.92 (.72)
Physical appearance	34	1.67	4.00	3.30 (.68)
Behavioral competence	34	1.20	4.00	2.88 (.67)
Job competence	7	2.00	3.80	3.29 (.62)
Romantic appeal	7	1.80	4.00	2.69 (.85)
Close friendship	7	2.80	4.00	3.60 (.48)
Global self-worth	34	1.50	4.00	3.33 (.73)

Note. Values in parentheses are standard deviations.

Overall test mean = 3.00, standard deviation = .68.

Where n = 7, these are the adolescent profiles.

In completing sub-domain items, 1 = really not competent; 2 = sort of not competent; 3 = sort of competent; 4 = really competent.

Table 17 provides descriptive statistics on how important it was to participants to be competent in each of the domains. An overall test mean and standard deviation for the importance ratings was not provided as these scores were utilized to calculate discrepancies rather than for comparative purposes.

Table 17

Table of descriptive statistics related to group scores for the 9 importance sub-domains of the Self-Perception Profiles for Children (and Adolescents)

Importance Sub-domain	n	Minimum	Maximum	Mean (SD)
Scholastic competence	34	2.50	4.00	3.26 (.60)
Social acceptance	34	1.00	4.00	2.50 (.90)
Athletic competence	34	1.00	4.00	2.74 (.89)
Physical appearance	34	1.00	4.00	2.34 (.92)
Behavioral competence	34	1.50	4.00	3.54 (.59)
Job competence	7	2.00	4.00	3.71 (.76)
Romantic appeal	7	2.00	4.00	3.29 (.70)
Close friendship	7	3.00	4.00	3.71 (.39)

Note. Values in parentheses are standard deviations. Where  $n = 7$ , these are the adolescent profiles.

In completing sub-domain items, 1 = really not important; 2 = sort of unimportant; 3 = sort of important; 4 = really important.

The self-esteem profiles were obtained by subtracting the importance score from the competence score for each participant and constructing a group mean of the difference. When the importance score was higher than the competence score, a negative impact on self-esteem in that sub-domain was implied. Harter (1985; 1988) suggested that only those domains where importance was rated 3 or higher would impact self-esteem. The global self-worth component was not subject to this process, as it was assumed that it would be important to all individuals to have high self-worth.

As a group, all domains except social, athletic and physical appearance had a mean importance of three or more indicating that they were deemed to be important. The three domains that scored less than three had high variability. Self-esteem scores were positive, indicating high self-esteem for social acceptance, athletic competence and physical appearance. The self-esteem scores for scholastic, behavioral, and job competency, romantic appeal and ability to make close friends were all negative, however, none was greater than  $-0.68$ . This suggested that none of these would significantly impact self-esteem in this group. Constructing a group self-esteem profile in this manner and comparing it to the graph of norms on the relationship between self-worth and the competence/importance discrepancy score, provided by Harter (1985, p. 25; 1988, p. 26), supported the overall finding that this group of participants had a high sense of their self-worth. Table 18 provides descriptive statistics related to the group self-esteem profiles. Visual comparison between Table 18 and Table 16 confirms the presence of either low negative or positive scores for self-esteem and a moderately high score for global self-worth.

Table 18

Table of descriptive statistics related to group self-esteem scores for the 9 sub-domains of the Self-Perception Profiles for Children (and Adolescents)

Self-esteem Sub-domain	n	Minimum	Maximum	Mean (SD)
Scholastic competence	34	-2.33	1.33	-0.56 (.81)
Social acceptance	34	-1.50	2.60	0.66 (.95)
Athletic competence	34	-1.50	1.83	0.18 (.81)
Physical appearance	34	-1.00	2.50	0.96 (1.0)
Behavioral competence	34	-2.50	0.30	-0.68 (.66)
Job competence	7	-0.80	0.00	-0.43 (.31)
Romantic appeal	7	-2.20	0.60	-0.60 (.98)
Close friendship	7	-1.20	0.50	-0.11 (.53)

Note. Values in parentheses are standard deviations. Where n = 7, these are the adolescent profiles.

#### The Vineland Adaptive Behavior Composite Score.

Descriptive statistics related to the Vineland Adaptive Behavior Composite standard scores are provided in Table 19. The composite score is obtained by combining the standard scores of the three domains, communication, daily living and socialization. There was a highly clinically significant difference between the group mean and the test mean in the adaptive behavior composite. Forty percent of participants scored at or below the fifth percentile for adaptive behavior.

Table 19

Table of descriptive statistics related to adaptive behavior composite standard scores

Descriptive	Statistic
Mean	81.8
Median	82.0
Standard deviation	13.0
Minimum	55
Maximum	109
Percent of scores $\leq$ 5 <sup>th</sup> percentile	40%

Note. Test mean = 100; standard deviation = 15.

Maladaptive behavior.

The final component of the Vineland Adaptive Behavior Scales was the completion of the Maladaptive Behavior Domain, which explored behaviors that participants exhibited that were undesirable. There were 27 items, parents indicated whether the child would never, sometimes or often display the behavior. Examples from this domain included being overly dependent, sucking thumb or fingers, becoming withdrawn, wetting the bed, being anxious, or excessively unhappy, having poor concentration, displaying aggressive, negative or dishonest behaviors. Raw scores were tallied, and the sum compared to age group ratings and described as 'non-significant', 'intermediate' or 'significant'. Ratings varied according to age, however, a score of  $\leq$  4 or 5 negative behaviors was generally considered non-significant, while scores over 10 – 12 were considered significant. Ratings were categorized using the percentile ranks of

the national standardization sample. A ranking of 'significant' indicated that the individual scored in the "extreme 16% of individuals the same age" (Sparrow et al., 1984, p. 119). Table 20 provides the frequencies of responses to this domain with 71.4% of parents describing their children or adolescents as displaying intermediate or significant behavior problems.

Table 20

Frequency table of maladaptive behavior rating

Rating	n	Percent
Non significant	10	28.6%
Intermediate	16	45.7%
Significant	9	25.7%

Parent perception of development.

On completion of the three sub-domains and maladaptive behavior domain of the Vineland Adaptive Behavior Scales, respondents were asked to provide a global statement about the overall development of their child. The following question was asked; "Do you think your child/adolescent is typical for his or her age, or younger or older in comparison to others the same age?" Table 21 provides frequency of responses to this question. In contrast to the responses given to each of the preceding domains, most parents (74.2%) believed their child or adolescent was developing at a rate that was typical for, or older than, their age.

Table 21

Frequency table of parent rating of development

Rating	n	Percent
Younger	9	25.7%
Typical	20	57.1%
Older	6	17.1%

Supplemental Results

In order to explore the relationships between findings, a number of correlational analyses were conducted. No probability (p) values or statistical conclusions will be presented, as the risk for type 1 errors became unacceptable. Table 22 provides Pearson Product Moment Correlations for the relationship between the varying assessment components.

Table 22

Correlation (r) of standard scores between assessment sub-tests

Variable	MC	VP	FMC	Com	DL	Soc
Visual motor integration	.63	.52	.60	.59	.42	.39
Motor control		.40	.55	.38	.24	.24
Visual perception			.52	.27	.24	.05
Fine motor composite				.30	.37	.34
Communication					.59	.53

Note. MC = motor control; VP = visual perception; FMC = fine motor composite; Com = communication; DL = daily living; Soc = socialization.

Visual motor integration demonstrated a moderate and positive relationship with a number of assessment scores other than those within the Beery assessment battery.

Performance on socialization domain did not appear to be related to performance on any other component except for communication, as would be expected, as it is a component of the same assessment battery.

Table 23 presents Pearson Product Moment Correlations for the relationship between the varying assessment components and a few demographic variables.

Table 23

Correlation (r) of assessment results and demographic characteristics

Variable	Age	Ao. Sat.	Temp.	Bypass	Hosp.
Visual motor integration	.04	.12	-.05	.24	-.01
Motor control	.29	.01	.03	.41	.14
Visual perception	-.09	.03	.07	-.01	-.21
Fine motor composite	.20	.10	.16	.22	-.16
Communication	.24	.20	.08	.22	.10
Daily living	.05	.14	-.09	.25	-.26
Socialization	.15	.13	-.02	.47	-.04

Note. Age = age at definitive repair; Ao. Sat. = aortic saturation level prior to definitive repair; Temp. = temperature cooled to during bypass; Bypass = minutes of bypass during open-heart surgery; Hosp. = number of hospitalizations.

Performance on the motor control component of the Beery, and socialization skills of the Vineland appeared to have a moderate and positive relationship with length of time on bypass during open-heart surgery. Age at time of definitive repair appeared to have little relationship with any test variable, except perhaps motor control.

As the results of a number of the sub-tests indicated significantly poorer performances for up to 40% of the group, there was an attempt to explore whether specific demographic variables would identify a subgroup of participants who experienced greater difficulty. A look at the raw data suggested that only a very few individuals experienced global deficits, most others demonstrated varying areas of weakness and strength. Visual analysis of scatter-plots of the data related to hypotheses 1.1 – 1.7, which were marked to identify either boys versus girls, or the three diagnostic groups, did not reveal any consistent pattern. In addition there were no significant correlations between age and performance.

The data were also analyzed to determine whether a selection bias existed. The demonstration project was initially designed as a randomized control trial. However, it was somewhat difficult to obtain agreement to participate from families due to the time commitment involved in full participation. Ultimately, families were given the ability to choose the level of their involvement by choosing to be in the treatment versus control group. This led to concerns that only parents whose children were experiencing difficulties ultimately participated. To partially address this concern, the data were analyzed to compare results between those children who chose to attend treatment sessions and those who only agreed to participate if they could be in the control group.

The treatment and control groups were comparable in numbers, ( $n = 17$ ,  $n = 18$ ), mean age (10.6, 11.9), number of boys (10, 13) and diagnoses, with one exception; that all participants who underwent the Fontan procedure ( $n = 3$ ), were in the treatment group.

The data were analyzed for differences between the means achieved by the treatment and control groups for each hypothesis using non-directional t-tests. As this was an exploratory look at the differences, probability ( $p$ ) values will not be presented. The control group's means for each hypothesis was higher than the treatment group (see Table 24). However, there were no statistically significant differences between the means of the treatment or control groups for any of the hypotheses. The difference between the treatment and control group means for the daily living domain of the Vineland approached significance; however, both control and treatment group means were significantly below the test mean. While the difference between the treatment and control groups in visual motor integration was not significant, in contrast to the whole group, the control group mean was not significantly different to the test mean of 100. The variability of the data in both the control and treatment groups for visual motor integration was quite high.

Table 24

Mean score and t value for each hypothesis for treatment and control groups

Hypothesis	Treatment (n = 17) Mean (SD)	Control (n = 18) Mean (SD)	t value
1.1 Daily living	83.3 (15.3)	91.3 (7.8)	-1.93
1.2 Communication	83.0 (17.5)	86.9 (12.5)	-0.76
1.3 Fine motor composite*	49.2 (15.0)	50.9 (12.5)	-0.33
1.4 Visual motor integration	85.2 (20.8)	92.8 (18.5)	-1.13
1.5 Visual perceptual	94.7 (19.0)	99.6 (15.8)	-0.82
1.6 Motor control	85.8 (16.2)	85.9 (10.1)	-0.03
1.7 Socialization	82.6 (15.0)	87.2 (7.8)	-1.14

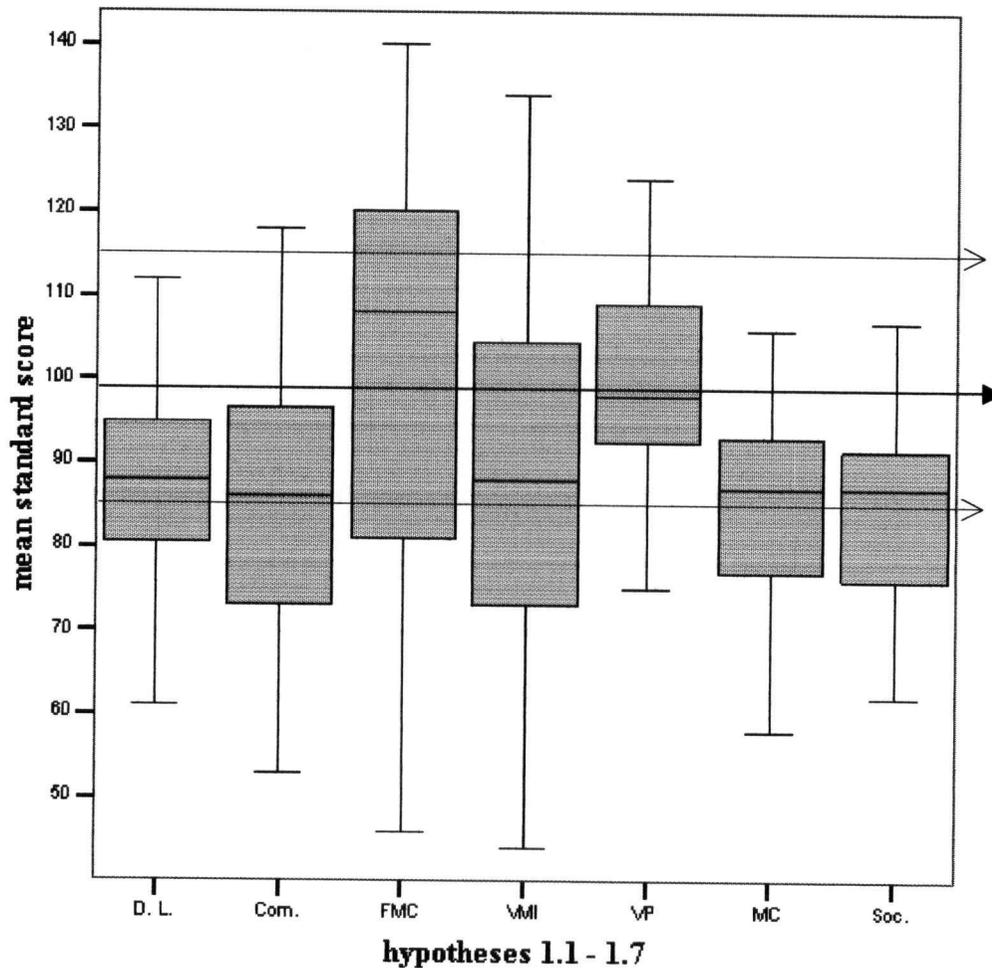
Note. All test means = 100, standard deviations (SD) = 15, except \* where test mean = 50 and standard deviation = 10. t values computed for difference between the treatment and control group means.

Summary

Figure 2 provides a graphic demonstration of the occupational performance of children and adolescents with congenital heart disease. Note that on this figure, the fine motor composite score has been transformed to a mean of 100 and standard deviation of 15. This sample was found to be experiencing difficulties in each occupational performance domain, that is, self-care, productivity and leisure. When performance components were examined, the participants exhibited strengths in visual perceptual and

fine motor abilities, however were under-performing in motor control and visual motor integration tasks. Overall, the participants had moderately high self-worth and self-esteem. The parents of the participants tended to believe that the children and adolescents were developing typically for their age, though they were exhibiting moderate to significant numbers of negative behaviors.

Figure 2

Interactive box-plot of standard scores for hypotheses 1.1 – 1.7

Note. Fine motor composite data have been transformed. D.L. = daily living; Com. = communication; FMC = fine motor control; VMI = visual motor integration; VP = visual perception; MC = motor control; Soc. = socialization;  $\rightarrow$  = approximately 1 standard deviation above or below test mean;  $\rightarrow$  = approximately the test mean. Box-plot is constructed around median, 75% of scores within the box, minimum & maximum scores represented by whiskers.

## CHAPTER V: Discussion

This chapter will provide a summary and explanation of the results of the study. The results will then be compared and contrasted with the findings reported in the literature. The implications of the findings from theoretical, research and practice perspectives will be discussed. Following this, the limitations of the study will be presented and suggestions for future research and practice made.

### Summary of Results

The participants of this study were children and adolescents aged 7.0 to 16.5 years who had TGA, TET or undergone the Fontan procedure for congenital heart disease. The median length of time from definitive repair to assessment within this study was just over nine years. All participants had complex congenital heart disease necessitating between one and four cardiac surgeries including at least one open-heart surgery, between 1983 and 1995, with cardiopulmonary bypass and hypothermia. Many participants also experienced circulatory arrest during their open-heart surgeries, and all for whom data were available, experienced bypass parameters within proposed safety limits (Kirkham, 1998). Only two participants had diagnosed associated congenital anomalies, namely Di George sequence, although a number had co-existing non-cardiac diagnoses such as asthma, hypospadias, or developmental delay. While a number of participants experienced post-surgical complications, only one had a significant neurological event during surgery resulting in seizures and temporary cortical visual impairment.

Participants were found to have reduced ability to participate in age-appropriate occupations of childhood, including tasks of self-care, productivity and leisure.

Participants' self-care abilities, as measured by the Vineland Adaptive Behavior Scales

(Sparrow et al., 1984), were significantly reduced. There was less variability in the sample data in comparison to the test variance, and 20% of the sample performed at or below the fifth percentile, suggesting that the parents of this group consistently reported that participants were under-performing in daily living skills. Activities reported on included personal independence in hygiene, dressing, and an awareness of one's own health. Participation in domestic activities, for example, assisting in meal preparation, cooking, tidying or cleaning were measured, as was growing independence in community tasks such as exhibiting safe behaviors in the community, using the telephone, and developing skills in managing money. In older adolescents, participation and success at paid employment was also explored.

The ability of participants to engage successfully in tasks of productivity was also reduced. Parents consistently reported that participants were under-performing in communication skills, as assessed by the Vineland, and that 31.4% of the participants were receiving learning assistance at school. The communication domain of the Vineland in these age groups focuses on development of reading, writing and verbal skills.

Direct assessment of the participants focused on their skills in physical and cognitive performance components. While the assessments used did not measure occupational performance, participant responses provided a limited measure of their underlying skills. There was insufficient evidence to suggest that participants had significantly different fine motor skills from their age peers. However, there was increased variability in the data in comparison to the test variance, and just over 16% of the group performed at or below the fifth percentile. This suggested that perhaps a

subgroup of participants were experiencing difficulties in this area, or that the power of the study to detect a difference was limited by the sample size.

Participants were found to perform clinically significantly lower than the test mean in the upper-limb coordination sub-test of the Bruininks (1978). The sample mean was just over one standard deviation below the test mean, and 22.6% of participants performed at or below the fifth percentile. The correlation between fine motor composite scores and the upper limb coordination scores was positive and moderately low. This suggested that it was not necessarily the same individuals who experienced the most difficulty with these two components of the Bruininks test. There may have been a stronger correlation between the upper limb coordination sub-test and the gross motor components of the Bruininks, with the poor performance on the upper limb coordination component reflecting the groups' deficits in gross motor skills rather than fine. However, this requires further investigation.

Elements of the participants' cognitive performance components were assessed using the visual motor integration and visual perceptual components of the Beery test (1997). Participants were found to have significantly lower visual motor integration skills than the test mean. In contrast, there was insufficient evidence to suggest that the participants had lower visual perceptual skills. Despite the lack of statistical significance between the means, there was evidence to suggest that a subgroup of participants was experiencing clinically significant difficulties in visual perceptual skills, with just over 17% performing at or below the fifth percentile. There was a low-moderate relationship between poor performance in the visual perceptual component of the Beery test, and poor performance in the fine motor composite of the Bruininks, suggesting that a subgroup of

participants experienced a range of difficulties. However, visual perceptual performance was not strongly related to school performance as measured by the communication skills' component of the Vineland test (Sparrow et al., 1984). This study demonstrated limited power to detect a moderate difference in visual perceptual skills.

The motor control component of the Beery (1997) was administered to assess pencil control, as an element of the individual's physical performance abilities. The participants demonstrated statistically significantly lower motor control skills than the test mean. This is in contrast to their Bruininks fine motor composite scores, although there was a moderate relationship between the scores of these two assessments. Poor performances in the motor control and visual motor integration sub-tests of the Beery suggested that the key concern in this group was one of motor output as opposed to a visual perceptual or global deficit.

The group was found to have a lower ability to participate in age-appropriate leisure pursuits and demonstrate social skill development, as measured by the socialization domain of the Vineland. This domain addressed those activities undertaken for fun and enjoyment, the participants' ability to make friends and establish relationships, their coping abilities and social skills required to engage in interactions with others.

The self-perception profiles indicated that as a group, the participants felt competent scholastically, socially, athletically, and behaviorally, and that they were happy with their physical appearance. The adolescents in the group also indicated that they felt competent at work they had done, in forming close friendships and that they had

romantic appeal. Overall, the groups' sense of self-worth and self-esteem was moderately high.

Response to the maladaptive behavior domain of the Vineland (Sparrow et al., 1984) found that a large proportion of participants exhibited moderate to significant behavioral difficulties as reported by parents. In contrast to both this scale and the below average scores obtained on the communication, daily living and socialization domains of the Vineland, most parents reported that they felt their child to be developing at a rate and level that was typical for, or older than, his or her age. This suggested that these parents had lower expectations of their child's need to be independent in the activities and tasks measured by the Vineland. However, one quarter of the parents interviewed indicated that they felt their child was developing at a rate that was 'younger' than their age.

As a number of the findings suggested that a subgroup of participants were experiencing difficulties, visual analysis of scatter-plots marked by different variables was undertaken to attempt to uncover identifying characteristics of this group. There did not appear to be any consistent bias towards age, diagnostic group or gender; however, this analysis was entirely preliminary and limited by the sample size and the distribution of those demographics across the sample.

Further analysis was undertaken to explore potential relationships between measurement tools and demographic or disease severity data. Performance on the visual motor integration component of the Beery, was found to have a moderate and positive relationship with performance on the visual perceptual and motor control sub-tests of the Beery, the fine motor composite of the Bruininks and the communication, daily living and socialization domains of the Vineland. The motor control component of the Beery

also had a low-moderate correlation with the length of time participants experienced bypass during open-heart surgery. These preliminary findings suggest that the Beery Test of Visual Motor Integration could be an important screening tool for identification of children at risk. In contrast to the expectations reported by past literature (Ferry, 1987; O'Dougherty et al., 1985), age at definitive repair did not appear to be related to the outcomes measured in this study.

In summary, the results of this study found that children and adolescents with congenital heart disease had a reduced ability to participate in childhood occupations of self-care, productivity and leisure, as measured by parent report. They were also found to have statistically significantly poorer skills on direct assessment, in visual motor integration and motor control and to have clinically significantly lower skills in upper limb coordination. In contrast, visual perceptual and fine motor skills were a relative strength of this group. In addition, this group of children and adolescents with congenital heart disease reported that they felt moderately competent at a range of childhood activities, and had a moderately high sense of self-worth and self-esteem.

#### Integration of Findings with Past Literature

The findings of this study supported many of those reported in the literature. DeMaso et al. (1990), Morris et al. (1993) and Bloom et al. (1997) found that children with congenital heart disease demonstrated mild to moderate difficulties in the Vineland Adaptive Behavior Scales. DeMaso et al (1990) studied a similar population; that is, children who had TGA or TET and compared them to other children who had minor heart defects requiring no surgery. However a global psychological functioning scale was

constructed, and so direct comparisons of performance between their study and this, were not possible. In contrast, Morris et al.'s (1993) sample was somewhat different, consisting of a group of children who had survived in-hospital cardiac arrest, most of whom had congenital heart disease. Morris et al.'s reported means for the standard scores of each domain of the Vineland, were very similar to those found in the present study, ranging from 83.0 to 87.2. In fact the battery composite standard score for the group in the present study was 81.8, while Morris et al. reported a battery composite standard score of 81.1. The sample demographics were certainly different from the present study, though Morris et al. postulated that the presence of the heart defect may have had more influence on participants' scores than the cardiac arrest.

In a follow up to the Morris et al. (1993) study, Bloom et al. (1997) utilized the Vineland in a study of the impact of cardiac arrest on children with congenital heart disease. These authors compared children with complex congenital heart disease who either did, or did not have a cardiac arrest. Those children who sustained a cardiac arrest had significantly poorer scores on the Vineland (mean 81.5), a very similar mean to the group in the present study. The group mean (91.6), for those who did not sustain a cardiac arrest was also lower than the test mean of 100. Bloom et al. (1997) concluded that cardiac arrest significantly increased risk of functional difficulties. The present study found a group of children with congenital heart disease, only one of whom sustained cardiac arrest, who performed at a similarly low level on the Vineland. This suggested that cardiac arrest is not the only contributor to lowered adaptive behavior.

The Beery test of visual motor integration (1997) was utilized by Morris et al. (1993) who found that 64% of the children scored at or below one standard deviation

below the mean with an overall group standard score mean of 82.7. Morris et al.'s sample performed more poorly than the sample in this study, perhaps reflective of the significance of a cardiac arrest in conjunction with congenital heart disease on visual motor integration.

This study's findings, that children and adolescents with congenital heart disease did not perform worse than the test mean on fine motor and visual motor tasks as measured by the Bruininks, supported Wright and Nolan's findings (1994). Wright and Nolan utilized the Bruininks when they studied children who had TGA or TET, who had undergone open-heart surgery between 1979 and 1984, that is a little earlier than the group in this study. They also found that there was no difference in performance between their group and the test mean on the fine motor components. In contrast to the report by Wright and Nolan, the present study found some evidence to suggest that a subgroup of children with congenital heart disease experienced deficits of fine motor and visual perceptual skills. It was not however possible to identify specific diagnostic or other demographic characteristics of this subgroup. In addition, the sample size of both the present study and Wright and Nolan's (1994) study ( $n = 29$ ), may have been insufficient to detect a difference in these skills.

The Harter Self-Perception Profiles (1985; 1988) were also utilized by Wright and Nolan (1994). Wright and Nolan compared their case (those with TGA and TET) and control groups (those with congenital heart disease who required no surgery) and found no differences between them on the self-perception profiles. Unfortunately Wright and Nolan did not provide enough information to determine whether either group differed significantly from the sample provided by Harter (1985; 1988). The findings of the

present study are in contrast to Kitchen's (1978) earlier literature review. Kitchen found that the literature she reviewed suggested that children with congenital heart disease had a lower self-esteem, regardless of disease severity. Kitchen's review is now dated however, as it was set in a period where many children with complex congenital heart disease were either not surviving, or whose definitive repairs were conducted at later ages, with less positive outcome.

In general, the findings of this study, that children and adolescents with congenital heart disease experienced difficulties that limited specific skill development and participation in childhood activities, supported the literature presented previously. Other authors have utilized different assessment tools and found poor psychosocial functioning (DeMaso et al., 1990; Spurkland et al., 1993; Wright & Nolan, 1994), compromised school performance (O'Dougherty et al., 1985; Wright & Nolan, 1994) and behavioral difficulties (Casey et al., 1996; Janus & Goldberg, 1995; O'Dougherty et al., 1983; Utens et al., 1993). The present study also found an increase in problem behaviors as reported by parents in the maladaptive behavior domain of the Vineland.

Previous studies have measured quality of life, with contradictory findings. Casey et al. (1994) studied 26 children with uni-ventricular heart, similar to the Fontan group of this study, and found reduced quality of life. They defined quality of life as it related to the child's ability to participate in normal childhood activities. Laane et al. (1997) studied three groups of children with heart disease ( $n = 164$  in total), those not requiring surgery, those requiring surgery, and those who also had an associated congenital syndrome, and compared them with children without heart disease ( $n = 301$ ). Laane et al. found a trend for higher quality of life in the group with treated heart disease.

Interestingly they defined quality of life as it related to the child's external living conditions, interpersonal conditions such as social supports and family interactions and personal conditions including self-esteem, basic mood and activity satisfaction.

While this study did not directly measure quality of life, the results provided some information about a few of the domains that influence quality of life as determined by the literature previously discussed. If self-esteem and satisfaction with how activity is performed influence quality of life, then the self-perception profiles completed in this study and the parent perception that the participants were developing 'typically', suggested that this group's quality of life would have been positively impacted. However, if like Casey et al. (1994), quality of life is considered to be a function of successful participation in childhood activities, the measurement tools utilized in this study might suggest a lowered quality of life.

This study did not purport to measure quality of life, however the theoretical model, CMOP (CAOT, 1997), proposes that optimal functioning in developmentally appropriate occupations will enhance quality of life. The seemingly contradictory result of this study, and between Laane et al. (1997) and Casey et al.'s (1994) studies can be related to the underlying difficulty of an inadequately developed conceptual framework related to quality of life. Casey et al. (1994) presented a very narrow definition of quality of life; that related only to the children's ability to participate in physical activity or attend school. Laane et al. (1997), however, had a more fully developed framework that addressed three spheres of life, external, internal and interactive. Quality of life is a burgeoning subject in the medical literature with outcomes increasingly being defined by the individual's sense of well-being. A recent study reported a 180% increase in Medline

citations of 'quality of life' or 'health status' between 1985 and 1993, in comparison to a 77% increase in the same period for citations in Medline on 'health' (Hornberger & Lenert, 1996). However, to accurately assess quality of life, a thoroughly formulated definition and conceptual framework is required, one that accommodates the dynamic nature of quality of life as it influences and is influenced by, many domains of life.

The CMOP (CAOT, 1997) provides a method of viewing multiple life domains, however, specific conclusions about quality of life can not necessarily be drawn from measures of those domains. The difficulty lies in the adjustments or "trade-offs" (Schipper, Clinch, & Olweny, 1996, p. 13) that are made as health status or competence changes. For example, many participants in this study indicated a lack of competence in one or two domains of the Harter (1985; 1988). It was then common for these individuals to rate those domains as less important to their sense of self, in comparison to the domains in which they felt competent, thus maintaining a high sense of self-worth and self-esteem. There were, of course, a number of individuals for whom this was not true and who rated their self-worth as low. While self-worth and self-esteem are not synonymous with quality of life, they do provide a measure of one's sense of well-being. This however, remains only one aspect of quality of life. Casey et al.'s (1994) study entitled "Quality of Life in Surgically Palliated Congenital Heart Disease" which only explored physical functioning, does not provide a convincing argument that children with congenital heart disease have a lowered quality of life. Laane et al.'s (1997) study found a trend for higher quality of life in the children with the most complex congenital heart disease. These findings and the present study's analysis of the self-perception profiles

support the concept of trade-off's or adjustments that are made to accommodate changing circumstances to maintain a positive outlook.

There is very little in the occupational therapy literature with which to compare the findings of this study. There were two studies found that had first authors who were identified as occupational therapists. Limperopoulos et al. (1999), addressed the neurological status of infants with congenital heart disease prior to surgery, and Wright et al. (1985) reported on the adaptive functioning of adults with congenital heart disease. These two and the presently reported study examined individuals with congenital heart disease during three different periods across the life span. Limperopoulos et al.'s (1999) findings were that over half the infants they studied were at risk for neuro-developmental difficulties even prior to cardiac surgery. The present study suggested that children and adolescents with congenital heart disease were experiencing difficulties in successfully engaging in childhood occupations. Wright et al. (1985) found lower scores for muscular endurance and physical activity levels. The remainder of Wright et al.'s study focused on recall of childhood experiences and found that while the young adults recalled feeling different or frustrated as children in 30% of cases, the group as a whole demonstrated normal social behavior profiles in adulthood. Uten's et al. (1993) study of adolescents and then young adults (1994) with congenital heart disease explored the changes that they found between these age groups. Utens et al. found adolescents to have greater emotional and behavioral problems than same-aged peers. In contrast, young adults with congenital heart disease were no longer found to experience these difficulties. These authors also discussed the adjustments people make to accommodate health changes, and suggested that denial might be one of the mechanisms at work.

The limited occupational therapy evidence that is available supports the need for therapeutic involvement with children who have congenital heart disease. These children have been shown to be at significant risk of neuro-developmental deficits, to be under-performing in childhood occupations by parent report, and to have reduced physical activity levels in adulthood. It might be tempting to suggest that as reports show young adults with congenital heart disease to be vital and contributing members of the community, therapeutic interventions in childhood are unnecessary. In addition, it seems that even if these individuals do not feel competent, they are able to make internal adjustments that allow them to maintain high self-esteem and positive quality of life despite health concerns. However, the ultimate goal of occupational therapy is to promote the ability of the individual and their family to achieve the things they want and need to do. Therapeutic interventions that address the deficits that these children have demonstrated may effectively increase the congruence between occupation, person and environment that is essential for optimal occupational performance.

### Implications of the Findings

#### Theoretical Implications

The CMOP (CAOT, 1997) proved to be an effective theoretical platform from which to conduct this study. The findings of the study suggested that children and adolescents, who had congenital heart disease, experienced disruption at the person, occupation and environmental levels of the model. Although the measurement tools primarily focused on the occupation and person level of the model, the retrospective chart

audit also provided evidence of altered environmental experiences within the participants' lives.

Because this was a descriptive study, it was not possible to elucidate the specific mechanisms that ultimately resulted in lowered occupational performance. However, there appears to be a strong relationship between congenital heart disease and lowered occupational performance in self-care, productivity and leisure as reported by parents. There was also a relationship between participants' performance on two of the assessments at the person level of the model, and the parent perception at the occupation level of the model. Due to design limitations, it was not possible to evaluate fully the physical, cognitive and affective skills of the participants. There is evidence to suggest that children with congenital heart disease have motor control deficits that impact performance in high level fine motor tasks, such as the Beery Test of Visual Motor Integration (1997). Exploratory analysis suggested that the participants' performances on the visual motor integration and motor control components were related to performance at the occupation level of the model. Cognitive skills related to visual perception were relatively well developed, as were affective skills as measured by self-worth and self-esteem ratings.

The study did not evaluate the impact of environmental factors on occupational performance. The retrospective chart audit provided evidence that participants experienced significantly altered environments in comparison to healthy peers. These participants had between one and 15 hospitalizations each, encompassing a total of 1138 days, experienced open heart surgery and had at least one stay in the intensive care unit. This, in itself, constituted an altered environment. However, the long-term implications

of this experience, either directly on the participants or on the parenting they subsequently received, were not explored in this study.

The CMOP (CAOT, 1997) embodies a client-centered approach in which the goal of therapy is to address the components of occupational performance by exploration of what is wanted by, or needed by clients, in order for them to successfully participate in occupations that are meaningful to them. It is interesting to note that while there were a number of parents who were actively seeking support for the difficulties they perceived their child to be experiencing, most parents indicated that despite lowered performances, they felt their child to be typical. This has implications for theory-driven research. Ideally, the theory behind the CMOP would demand that research be driven by client demand, in partnership with, and consultation and collaboration between clients and therapists. However, the profession also has a responsibility to explore outcomes related to occupational performance as advances in medicine continue to push the boundaries of survival for those with complex congenital issues. Anecdotal evidence suggests that while families and physicians are focused on survival, there is less room for thought regarding the various components of developmental outcome. As medical outcomes become more positive, a dialogue must be entered into regarding continued improvement of the quality of these outcomes. This study aimed to explore aspects of occupational performance, as a step toward entering a meaningful interaction with families who have children with congenital heart disease, to ensure that occupational therapy interventions that promote occupational performance are provided.

### Research Implications

Use of the limits of agreement procedure (Altman & Bland, 1993) to determine intra-rater reliability highlighted some interesting factors. For example, in the Beery it was found that a difference of one raw score, out of a potential 27, could make up to a 7-point difference in standard scores. Likewise, in the Vineland, a difference of 2 or 3 raw scores out of a potential 100 – 150, could also make a difference of up to a 6 standard score points. A similar difference occurred in comparing raw to standard scores in the Bruininks, however the difference was scaled down as would be expected due to the lower test mean and standard deviation. Each of the differences in standard scores obtained were within the SEM's provided by the test manuals. The variability demonstrated by the limits of agreement method strongly reinforces the need to utilize confidence intervals when making clinical interpretations of individual performances. In addition, unless very high consistency exists within the rater, the chance of making clinically inappropriate decisions is quite high, that is small errors in raw scores, rapidly make large differences in standard scores.

Bland and Altman's (Altman & Bland, 1993; Bland & Altman, 1986), method for determining the agreement between two measurements was logically very sound and clinically rigorous. However, research in the health field has often involved small samples, and consequently the probability of having one or two scores constitute more than 5% of the test-retest sample would be quite high. Bland and Altman (1993; 1986) recommended the use of a 95% limit of agreement. They also suggested, however, that a clinical decision is ultimately required to determine whether the difference in scores would have impacted treatment choice or recommendations made, and therefore, whether

the extent of agreement was adequate. The clinical reasoning that is required for this, is quite different to that traditionally used when determining reliability.

Rehabilitation and medical research has in the past, relied on statistical analyses such as the Pearson Product Moment Correlation to determine the extent of test-retest or inter-rater reliability. Indeed, each of the standardized tests utilized in this study, which are in common usage in clinical practice, also used correlational analyses to establish reliability. Achieving a reliability coefficient of 0.95 is not the same as establishing 95% agreement between scores. Thus the process of establishing the reliability of many tools utilized in rehabilitation for repeat measures is called into question.

### Implications for Clinical Practice

#### Therapeutic interventions for children with congenital heart disease.

The results of this study highlighted the need for occupational therapy involvement with children who have congenital heart disease. The median length of time since the participants had their corrective surgeries was 9.3 years, and yet they were still found to be experiencing occupational performance deficits. While there are a group of children whose participation may be inhibited by cardio-vascular limitations, there also appears to be a significant group who are limited by co-existing neurological and developmental deficits. It is likely that those individuals who experience significant neurological events, such as a cerebro-vascular accident, or who have a co-existing congenital syndrome will receive occupational therapy input. However there is a significant proportion of children with congenital heart disease who are experiencing

more subtle deficits that impact their ability to participate successfully in childhood occupations, who are not identified and who do not receive therapeutic interventions.

This study focused on outcomes in middle childhood, supporting the notion that difficulties experienced in the immediate post operative period do not necessarily resolve. It is likely that children could be identified at a much younger age and interventions negotiated with families to maximize the child and family's capacity to undertake the tasks they need, or want to do, that is to promote optimal occupational performance. This in turn, may provide timely building blocks that support development of personal independence, school entry and participation, and increase the child's options for leisure pursuits including integration of physical activity and sport.

#### Use of standardized tests.

Standardized tests and measures are in common usage in occupational therapy. The process of establishing intra-rater reliability in this study strongly reinforces the need for careful administration and the use of confidence intervals when reporting findings. Quality assurance practices within therapy departments might also include establishment of assessor reliability as a routine component of professional development.

### Limitations

#### Generalization of Results

Generalization of these results was limited to those children and adolescents with complex congenital heart disease, including TGA, TET and those defects requiring surgical repair through the Fontan procedure, from the early 1980's to the early 1990's. These limitations were applied because the rapidly changing medical and surgical

management of children with congenital heart disease may influence the period of time children experience hypoxia, the complications that arise from open-heart surgery, the length of hospitalization and the attitude of parents and community to participation of these children in daily activities.

Any statistically significant results could not be assumed to be indicative of the whole population of children with complex congenital heart disease, principally because the data collection did not fulfill the required assumptions for randomized sampling to allow such generalization. The sampling frame included 82 children who fulfilled the inclusion criteria of the demonstration project, all of whom were invited to participate. However, it was somewhat difficult to obtain agreement, principally because involvement in the demonstration project necessitated considerable time and effort on behalf of the families. Like any research involving human subjects, there is the potential that there is an important difference between those who volunteer and those who do not. The key concern in this design was that the 35 families who agreed to participate perhaps did so because those parents believed their children were experiencing difficulties.

In order to partially address this concern, the data were analyzed for differences between the treatment and control groups of the demonstration project. Those participants who were in the control group committed themselves to attending for three assessments, while the treatment group was assessed three times, and also attended twice-weekly treatment sessions for four months. The participants were not randomly assigned to groups as agreement to participate was ultimately linked to the parents' ability to choose the level of their involvement.

The comparison between the treatment and control groups suggested that there were some differences in achievement and skill between them, with the control group apparently not experiencing the same degree of difficulty as the treatment group. However, with the exception of performances on the visual motor integration sub-test, these differences did not alter the findings of the study.

As the overall purpose of this study was to explore and describe the occupational performance characteristics of children with congenital heart disease, a study sample that consisted of over 42% of the potential sample, lent credibility to the findings. In addition, the similarity of distribution of gender and diagnosis also suggested that a reasonable cross-section of the sampling frame was obtained. Results from this study were viewed as indicative of important trends in the development of children and adolescents with congenital heart disease.

### Causation

The research design and statistical analysis utilized in this study could not be used to attribute causation between congenital heart disease and the identified trends. This study identified a situation, related to the occupational performance abilities of children and adolescents with congenital heart disease, in comparison to their peers who do not have congenital heart disease. Further specific quantitative or qualitative studies are needed to investigate why any trends identified by this study exist.

### Analysis and Statistical Power

It was determined that the study had power of .91 to .99 to reject five of the seven null hypotheses. The power to detect a moderate effect size in the remaining two hypotheses, that is those related to fine motor control and visual perceptual skills, was insufficient. There was a 49% chance that a moderate effect size in the fine motor component would not be detected in this sample, and a 35% chance that a moderate effect size would not be detected in the visual perceptual component. This lack of power is of concern, particularly as past research which supported the findings in these areas was conducted on a similar sample size (Wright & Nolan, 1994). In order to partially address this concern, the standard scores for the two sub-tests that were found by Wilson, Polatajko, Kaplan and Faris, (1994) to provide the “greatest degree of discrimination between children with and without motor problems” (p. 15), were also presented. The descriptive statistics of these two sub-tests also suggested that these participants were not clinically different from the test mean.

### Measurement

There are a number of limitations related to measurement. The reliability, validity and standardization processes of each of the tools utilized have been provided in Appendix D. Concerns regarding the establishment of reliability have already been discussed. It should also be noted that each tool was standardized on pediatric populations in the United States. The assumption was made that children on the North American continent would be similar in their responses to each of these assessments. This may not be true. In particular, the Lower Mainland of British Columbia is likely to

differ from the USA in distribution of ethnicity. It is unclear what impact this may have had on participant responses to the assessments.

It should also be noted that the Bruininks motor test and the Vineland Adaptive Behavior Scales were standardized in the mid 1970's and early 1980's respectively, indicating that the normative data are 10-20 years old. It is possible that the standardization data for both these tools are outdated and no longer valid. However, they remain in common usage across Canada, and Wilson et al. (1994) have made recommendations regarding the clinical applicability of the data obtained.

Wilson et al. (1994) suggested that the visual motor control sub-test of the Bruininks was one of four sub-tests most likely to discriminate between those children with motor problems and those without. During the administration of the Bruininks in this study, it was observed that many participants rapidly reached the raw score test limit of this sub-test. This impression was confirmed when comparisons were made between the Bruininks fine motor scores and the Beery motor control scores. While there was a positive relationship between the scores obtained on the Beery and the Bruininks, the Beery motor control component was more likely to identify participants who were having motor difficulties than the Bruininks visual motor control sub-test. The value of the Beery as a research and clinical assessment tool was well supported by the findings of this study. It was sensitive to moderate deficits, performance on the visual motor integration sub-test was found to be related to performance on other assessments and the motor control component related to length of time on bypass during open-heart surgery.

Use of the Vineland Adaptive Behavior Scales (Sparrow et al., 1984) to determine childhood occupational performance entailed reliance on parent report of performance.

In pediatrics it is common to utilize parent report as a component of the assessment process, and studies have demonstrated consistency between parent-report and child-report of concepts such as health status assessment (Glaser, Davies, Walker, & Brazier, 1997). In contrast, the study conducted by Casey et al. (1994) found that 80% of parents underestimated their children's exercise tolerance by up to a third, suggesting a lack of consistency in direct versus surrogate measures. While the Vineland has been well-validated and utilized in a number of other studies, it remains an indirect assessment. The supplemental use of direct observation and assessment of participants, and the positive relationship between direct and indirect assessments, for example, visual-motor integration and communication sub-tests, lent credibility to the findings.

The Harter self-perception profiles provided a valuable opportunity to explore the participants' own sense of achievement, competence and self-worth. Participant responses to completing this varied. For example, one adolescent boy commented that it gave him pleasure to reflect on his abilities and acknowledge that he was okay. Those children whose sense of self-worth was not high found the task somewhat daunting. As this tool has only been validated on a small sample, it was not appropriate to make statistical inferences from the data obtained. It would, however, be a valuable tool to use with children in clinical practice when exploring occupational performance needs from a client-centered perspective.

#### Future Directions for Research

Future research must focus on a number of issues related to the needs of children with congenital heart disease, the role of occupational therapy in meeting these needs and

the reliability of measurement tools utilized in therapy. The CMOP can assist in focusing research questions that relate to the person, occupation and environmental components of the model. At the person level, the past literature suggests that over 50% of infants with congenital heart disease will experience neurological deficits (Limperopoulos et al., 1999), and the present study found that they do not 'grow out of' difficulties experienced in early childhood. Therefore, perhaps all children with congenital heart disease should undergo developmental assessment. The results of this study suggest that some skill deficits relate to a particular subset of children. If so, determining both how this subset is to be identified, when assessment should be provided and what intervention would be effective, is imperative. The relationship between length of time on cardio-pulmonary bypass during open-heart surgery and outcomes on this study may provide one starting point for exploring the identifying characteristics of those most at risk.

At the occupation level of the model, many children were reported by parents to be under-performing in childhood occupations. The specific factors that cause this deficit have not been identified. Can deficits be solely attributed to the neurological and physical implications of heart disease or are deficits also related to the impact of congenital heart disease on parents and their parenting? In addition, an exploration of the relationship between reported behavior and direct measures in the field of occupational therapy is warranted.

If the question of the impact of congenital heart disease on parenting is raised, then the environmental component of the model comes into focus. How powerful would information versus treatment be, that is, would assessment of the young child, followed

by education and information-sharing with the parent be effective in promoting increased occupational performance? The other key environment for children is that of the school. While school achievement has been measured in this population in the past, research exploring the children's school experiences more directly has not occurred. It is not clear how the school system perceives and supports children with congenital heart disease.

Continued exploration into the efficacy of therapeutic interventions is also required. What form should therapeutic interventions take, are any of them effective and if so, how can they be delivered in a manner that is both cost-effective and equitable regardless of geographic location? If therapists in local communities are to undertake treatment of this group, what training or education will they require?

As therapists contribute to the building of evidence-based health care, the integration of new ways of measuring outcome is critical. If improved quality of life is a desired outcome, how will it be effectively measured? In addition, if standardized measurement tools are to provide valid information, reliability is key. The reliance on generation of reliability coefficients is no longer supported. The application of Bland and Altman's (1993; 1986) limits of agreement methodology is required and rehabilitation practitioners should move to incorporate this process into both their research and clinical practice.

### Summary and Conclusions

This study found that the occupational performance abilities, as reported by the parents of this sample of 35 children and adolescents with congenital heart disease, were lower than those of their typical peers. These participants were also found to have

deficits in performance components, particularly in visual motor integration and motor control. Their ability to complete tasks requiring visual perceptual or fine motor skills was not significantly different from their age peers. These results support those presented in the past literature; children with congenital heart disease experience a wide range of difficulties.

Analysis of the measures utilized suggested that the visual motor integration sub-test of the Beery might be a good screening tool for this population as it was moderately correlated with a number of the other assessment tools utilized. Analysis of intra-rater reliability using the limits of agreement methods led the author to question the reliability data provided by many of the standardized tests and measures in common usage in therapy.

Analysis of the relationship between the assessments undertaken by participants and their disease severity data found only length of bypass time during open-heart surgery to be of predictive interest. Contrary to what was suggested by the literature, age at definitive repair did not appear to be related to outcome on the measures utilized in this study.

Contrary to anecdotal or popular opinion, these children, who were a median of 9 years post corrective surgery, had not outgrown their difficulties. The findings suggest that children with congenital heart disease may require comprehensive rehabilitation programs as opposed to programs that focus exclusively on exercise. Recommendations from the study are that young children with congenital heart disease should receive routine occupational therapy assessment and then, intervention as appropriate. Further occupational therapy research is required into: 1. reliability of frequently used tests and

measures; 2. age at which assessment of children with congenital heart disease should occur; and 3. efficacy of any interventions applied to this population.

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Appendix A

Initial Letter of Contact

Appendix B

Informed Consent

# THE UNIVERSITY OF BRITISH COLUMBIA



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## INFORMED CONSENT

### Project Title:

"A demonstration project to evaluate the benefits of exercise rehabilitation in children with congenital or acquired heart disease."

### Principal Investigator:

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Dr. Donald C. McKenzie  
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The University of British Columbia  
Vancouver, B.C.

### Background:

Congenital heart disease occurs in 6 to 8 per 1000 live births, resulting in an incidence of approximately 0.7%. Advances in the medical and surgical management of these children has greatly improved their overall health in the past decade. However, many children have a low level of physical fitness and poor exercise tolerance because their congenital lesion either limits blood flow, creates a volume overload in the heart, or limits oxygen delivery to the working muscles. In addition, apprehension on the part of the child or his/her parents, friends, teachers, or family physician may prevent the child from participating in regular physical activity.

The purpose of this study is to initiate a 12-week supervised exercise rehabilitation program for children with congenital heart disease, reintroduce home-based physical activity to the child, and to prescribe a 24-week program of exercise for all children who complete the supervised rehabilitation program. We believe that the implementation of an exercise rehabilitation program will be successful in improving exercise tolerance, cardiac function, and the functional independence and quality of life of children with congenital heart disease.

Study Procedures:

**Developmental and Functional Abilities Assessment**

All subjects will undergo a developmental and functional abilities assessment by a physiotherapist and an occupational therapist prior to participating in the exercise rehabilitation program, and at the end of both the 12-week supervised exercise program and the 6-month home-based exercise program. The physiotherapist will evaluate your child's gross motor skills (running speed and agility, balance, bilateral coordination, strength and flexibility). The occupational therapist will assess fine motor skills, visual perception, visual motor skills, personal and social skills, and several quality of life measures.

**Exercise Testing**

Your child's height and weight will be measured. He/she will then exercise on a stationary bicycle while measurements of heart and lung function are taken. Every 3 minutes the resistance on the bicycle ergometer will be increased and your child will exercise until they become fatigued (volitional fatigue) or want to stop for some other reason. During the exercise test your child's heart rate and rhythm, blood pressure, and oxygen saturation will be monitored. Your child will be given an exercise diary to take home to record any physical activity that they do during the study.

Risks:

There are minimal risks associated with exercise. Careful monitoring of your child's electrocardiogram, blood pressure, and oxygen saturation during exercise will help us to ensure that your child can safely exercise. If your child develops abnormal symptoms or complaints, he/she may stop exercising immediately. Your child may experience mild, temporary fatigue during exercise and in the immediate post-exercise period. This is not an uncommon finding in people who are exercising.

Withdrawal from Study:

I understand that if I decide not to participate in this study, or withdraw my child from this study at any time, it will not in any way jeopardize my child's medical care.

Confidentiality:

I understand that all personal information about my child will be confidential. All documents will be identified only by a numeric code and kept in a locked filing cabinet. My child will not be identified by name in any written reports from this study.

Contact:

I understand that if I have any questions or desire further information with respect to this study, I should contact Dr. George Sandor at 875-2295. If I have any concerns about my child's treatment rights as a research subject, I should contact the Director of Research Services at the University of British Columbia, Dr. Richard Spratley, at 822-8598.

Subject's Consent:

I understand that participation in this study is entirely voluntary and that I may refuse to have my child participate, or may withdraw my child from the study at any time, without any consequences to his/her continuing medical care.

I understand that my child's commitment to this study will be limited to completing the three exercise studies (3 hours), and three developmental and functional abilities assessments by a physiotherapist and occupational therapist (9 hours).

I acknowledge having received a copy of this consent form for my own records.

I consent to having my child participate in this study.

\_\_\_\_\_  
Subject's Name

\_\_\_\_\_  
Parent/Guardian's Signature

\_\_\_\_\_  
Date

\_\_\_\_\_  
Witness' Signature

\_\_\_\_\_  
Date

\_\_\_\_\_  
Investigator's Signature

\_\_\_\_\_  
Date

## Appendix C

Cardiologist's Perception of Medical Severity Rating Scale (DeMaso et al. 1991, p.141):

1. No or insignificant disorder - disorder has no impact on child's health.
2. Mild disorder – lesion requires no operative intervention, only long-term follow-up (e.g. small ventricular septal defect).
3. Moderate disorder – child is asymptomatic, but has had or will require operation, easy repair (e.g. atrial septal defect).
4. Marked disorder – child quite symptomatic, has had or will require major difficult repair ( e.g. tetralogy of Fallot, transposition of the great arteries)
5. Severe disorder – uncorrectable cardiac lesions or only complex palliative repair possible (e.g. pulmonary vascular obstruction, Fontan repair, valve replacement).

## Appendix D

### Test Validity and Reliability

#### Vineland Adaptive Behavior Scale

The Vineland Adaptive Behavior Scale (Sparrow et al. 1984) was developed and standardized in the United States of America (hereafter U.S.A.) using 3000 individuals, roughly equal boys and girls, with approximately 100 children in each age group from birth to eighteen years. The population used to create the standardization was stratified according to the 1980 U.S.A. census for parental education, ethnicity, community size, child's educational placement and preschool attendance and distribution across four regions. Standardized norms have been developed for each age group's performance in each domain and as a total score.

Comprehensive reliability and validity data are presented in the manual for the Vineland Adaptive Behavior Scales (Sparrow et al. 1984) and will be briefly summarized here. Reliability coefficients are provided for split-half or internal consistency reliability, test-retest reliability and inter-rater reliability using Pearson coefficients. The median split-half reliability coefficients for the survey form are reported on the total national sample and range from 0.69 for the personal subdomain to 0.84 for the written subdomain. Test-retest reliability coefficients for the four domains of the survey form are reported for 484 individuals from the national sample and range from 0.81 to 0.88. Inter-rater reliability was assessed for 160 individuals from the national sample and ranges from 0.62 for the socialization domain to 0.78 for the motor skills domain. Intra-class coefficients were also calculated for test-retest and inter-rater reliability and are reported to be 0.99 and 0.98 respectively.

Standard errors of measurement (hereafter SEM) were computed using the split-half reliability coefficients, and a standard deviation of 15. The SEMs are reported for

each age group within the 11 sub-domains and range from 2.2 to 8.2 standard score units, at the 68% confidence level.

Tests for construct validity indicated that the Vineland Adaptive Behavior Scales exhibited higher correlations with other adaptive behavior scores, such as the Vineland Social Maturity Scale (Doll, 1965, as cited in Sparrow et al. 1984), Adaptive Inventory for Children (Mercer & Lewis, 1978, as cited in Sparrow et al. 1984) than with intelligence measures such as Kaufman Assessment Battery for Children (Kaufman & Kaufman, 1983, as cited in Sparrow et al. 1984) and Peabody Picture Vocabulary Test - Revised (Dunn & Dunn, 1981, as cited in Sparrow et al. 1984). There were only moderate correlations with the adaptive behavior tests and thus the Vineland was assumed not to be an unnecessary duplication. The comprehensive development of this scale as a diagnostic, program planning and research tool, makes it a valuable measure of adaptive functioning in children with congenital heart disease.

#### Test of Visual-Motor Integration

The original Test of Visual-Motor Integration (Beery, 1967) was normed in 1964 on 1030 children and then cross-validated in 1981 and 1989, with 2060 and 2734 children respectively. In 1996, the test was normed again in conjunction with two supplemental tests of visual perception and motor control. The 1996 sample included 2614 children from the U.S.A., stratified according to the 1990 census for gender, ethnicity, residence, geographic area, socioeconomic level and across ages 3 years 0 months to 17 years 11 months.

Comprehensive reliability and validity data are presented in the test manual (Beery, 1997) and will be briefly summarized here. Internal consistency was calculated using 50 children from each age group in the national sample. Internal consistency was

calculated by the Rasch-Wright analysis as an odd-even split-half correlation resulting in a coefficient of 0.88, and also by coefficient alpha with an overall coefficient of 0.82.

SEM's were calculated for standard scores with a mean of 100 and standard deviation of 15, using the split-half coefficient and are provided for each age group, ranging from 4 to 6 standard units of measurement, at the 68% confidence level. Test-retest reliability was calculated on data from 122 children between 6 and 10 years, with overall test-retest raw score coefficients of 0.87 for the visual motor integration test, 0.84 for the visual perception supplement and 0.83 for the motor control supplement. Interrater reliability was calculated using a randomly selected sample of 100 children from the national sample and found to be 0.94 for the Visual Motor Integration Test, 0.98 for the Visual Perception supplement and 0.95 for the Motor Control supplement.

The Beery (1997) manual provides details as to the comprehensive development of this tool which support the content validity of the test. Concurrent validity was tested during the 1996 standardization process by comparison to two other copying tests and found to be acceptable. Construct validity was tested against a number of hypotheses and the test was found to correlate strongly with age, with coefficients of 0.83 for visual motor integration, 0.75 for the visual perceptual supplement and 0.74 for the motor control supplement. Correlations with academic achievement were found to be 0.63 for visual motor integration, 0.29 for visual perceptual supplement, and 0.40 for the motor control supplement. The test was also found to be sensitive to varying disabling conditions. These correlations suggest that the Beery Test of Visual Motor Integration (Beery, 1997), can be used to gain insight into the ability of children with congenital heart disease to participate and be successful at school and thus be an indicator of productivity in childhood.

The Beery Test of Visual Motor Integration (1997) was developed and validated for three purposes: 1) to identify difficulties in a child's development of visual motor

integration; 2) to assess the effectiveness of treatment strategies and 3) to serve as a research tool. The raw scores for each age group, calculated at 2 month intervals, can be converted to standard scores for each component of the test. The normative data will be utilized to compare the performance of the children with congenital or acquired heart disease with their age-related peers.

### Bruininks-Oseretsky Test of Motor Proficiency

The Bruininks-Oseretsky Test of Motor Proficiency (Bruininks, 1978) was developed in the U.S.A. and standardized on 800 children from the age range of 4.5 years to 14.5 years. The children of the national sample were randomly selected from across the country and stratified according to the 1970 U.S.A. census, for age, sex, race, community size and geographic region. Norms were developed for each age range for both boys and girls for standard scores, with a mean of 50 and standard deviation of 10 on the whole test, and for gross motor and fine motor composites separately. The fine motor composite will be utilized to compare the performance of the children with congenital heart disease with their age and sex matched peers.

Literature and research support for the validity of the Bruininks-Oseretsky Test of Motor Proficiency (1978) as a developmental assessment of motor skills is presented in the manual. Evidence of construct validity is cited through comparison with other motor assessments through correlation of performance with increasing age (with a median correlation coefficient for the total sample of 0.78), and by analysis of internal consistency (0.77 for visual motor control and 0.81 for upper limb speed and dexterity).

Factor analysis was also completed to support the grouping of items within the subtests. While the factor analysis gave some support for the grouping of items within the gross motor and upper limb subtests, the fine motor subtest items did not cluster

together as separate factors but rather were found to load significantly on the general motor ability factor.

In examining reliability of this test, only test-retest reliability was studied. Because most of the items are dependent on speed of response, internal consistency estimates that use coefficient alpha or split-half reliability were not deemed appropriate. Test-retest reliability was calculated during a special study of 63 second-grade children and 63 sixth-grade children representing a range of abilities. The reliability coefficient for the Fine Motor Composite was 0.88 for grade two children, and 0.68 for grade six children.

SEM was calculated using the test-retest coefficients from the two grades, and found to be 4.7 standard units of measurement, at the 68% confidence level, for the Fine Motor Composite which has a mean of 50 and standard deviation of 10. Inter-rater reliability was calculated using data from two special studies using subtest 7 only (visual motor control), because the scoring of these items was deemed to require more judgment than the other items. Inter-rater reliability from the two studies found coefficients of 0.98 and 0.90, suggesting strong consistency can be achieved.

### Self-Perception Profiles for Children

The Self-Perception Profiles for Children (Harter, 1985) were developed in the U.S.A. using children from Colorado, 90% of whom were Caucasian. Harter (1985) administered the scale to four samples of children from 3rd to 8th grade with between 91 and 612 children in each grade, and roughly equal numbers of boys and girls. Internal consistency based on Cronbach's alpha, is reported and was consistent across the four samples within each domain. The lowest internal consistency was for the behavioral conduct subscale in sample D at 0.71, and the highest internal consistency was for the athletic competence subscale in sample B at 0.86. Factor analysis using oblique rotation

was undertaken to determine whether the subscales measured different components of self-perception. Factor loadings for the five subscales (excluding global self-worth) were found to be substantial with no cross-loadings of greater than 0.18, indicating that the subscales were measuring differing components of self-perception.

Means and standard deviations are provided for boys and girls in each sample across the six grades for each subscale and will be utilized as the normative data against which the children with congenital heart disease will be compared.

Harter (1988) administered the revised questionnaire to four samples of adolescents in Colorado, two samples for each of the four grades. There were 100 to 205 adolescents in each grade with roughly equal numbers of boys and girls. The sample was 90% Caucasian representing households from lower middle to upper middle class. Internal consistency was calculated using Cronbach's alpha and results presented show that the lowest subscale internal consistency on the revised scale was for sample D in the job competency domain at 0.74, and the highest was for sample A in the job competency domain at 0.91. There was greater variability across samples for job competency, scholastic competency, and social acceptance in comparison to the reasonably consistent reliability values reported across samples within domains in the children's profile. Factor analysis was also completed for eight of the subdomains (excluding global self-worth) demonstrating substantial factor loadings with no cross-loadings of greater than 0.3, indicating that the factors define different components of self-perception. Means and standard deviations for males and females in the four samples across four grades are provided and will be utilized as the normative profile against which the adolescents with congenital heart disease will be compared.

Reliability data and normalization procedures of both the Harter Scales (1985, 1988) are more limited than for those of the other three tests selected. The population sizes are much smaller, and were not stratified as the others were according to U.S.A.

census data. While comparisons between the children with congenital heart disease and these norms will be made, conclusions will necessarily be more guarded.