Clinical Profile of the Adolescent/Adult Fontan Survivor

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ABSTRACT

Objectives. The study aims to describe the clinical profile of the adult Fontan survivor and identify the worries, symptoms, and the impact of cardiac surveillance most commonly experienced.
Design. A descriptive, cross-sectional design was used.
Setting. The study was performed in outpatient adult and pediatric cardiology clinics in university-affiliated and private practice offices.
Patients. Fifty-four adolescent and adult patients with single ventricle congenital heart disease who have undergone the Fontan procedure participated in the study. The mean age was 26 ± 9 years with 52% female and 63% Caucasian.
Outcome Measures. Demographic and clinical data were obtained by a standard intake form and retrospective chart reviews. The Congenital Heart Disease TNO/AZL Adult Quality Of Life questionnaire was completed to assess worries, symptoms, and the impact of cardiac surveillance.
Results. The majority were single (73%), employed or full-time students (93%), with health insurance (94%), had a single left ventricle (78%), the diagnosis of tricuspid atresia or double inlet left ventricle (59%), lateral tunnel Fontan type (44%), history of arrhythmias (76%), left ventricle ejection fraction percentage >50 (66%), oxygen saturations >90% (70%), frequent headaches (50%), scoliosis (22%), varicose veins, ascites, and liver cirrhosis (46%), normal body mass index (59%), and New York Heart Association class I (48%) and II–III (52%). Primary worries related to current health (83%), job/employment (69%), ability to work, (61%) and living independently (54%). The most bothersome symptoms were shortness of breath with ambulation (69%), dizziness (61%), and palpitations (61%).
Conclusions. Fontan survivors experience residua and sequelae from multiple surgical procedures and the natural disease course. Our results support the need for ongoing assessment of both physical symptoms and psychosocial concerns, and suggest the need for multiple risk factor intervention strategies that improve physical and emotional health in Fontan survivors

Key Words. Single Ventricle; Congenital Heart Disease; Fontan; Health-related Quality of Life

Introduction

Congenital heart disease (CHD) not only is a disease of pediatrics but also represents a chronic adult disease population. The clinical profile of the general CHD population is changing related to advanced age, improved surgical techniques, and the survival of more complex congenital heart defects. Approximately 1.3 million children in the United States with significant heart defects have survived into adulthood.[1] At the present time, there are as many adults with CHD as there are children with this disease.[2] Furthermore, this population is estimated to grow at a rate of 5% per year.[3] The current and future resources required to provide optimal care for this growing population reflect the increased size and number of adult CHD programs across the United States.[4]

Over the past two decades, life expectancy for children with single ventricle CHD has increased significantly. This is related to

advancements in surgical technique and perioperative care. Single ventricle CHD is a subgroup of complex CHD and is estimated to represent only 9–12% of all congenital heart anomalies.[3] Current surgical management is tailored to the specific type of single ventricle defect. However, the majority require multiple surgical procedures. Thus, the completion of the Fontan while providing satisfactory palliation, still leaves the patient with a single right or left ventricle at risk for late cardiac failure, exercise intolerance, and arrhythmias.[5]

Despite improved life expectancy, many older patients with a single ventricle have residua and sequelae from multiple surgical palliations or the natural disease course. Physiologically, the Fontan population is prone to disease complications related to right atrial enlargement and residual atrial septal defect (i.e., arrhythmias, thrombus, hypoxemia, stroke), chronic passive venous congestion (i.e., ascites, liver cirrhosis), and single ventricle failure as described in long-term outcome studies after the Fontan procedure.[5] A portion of patients who have undergone the Fontan procedure may ultimately require heart transplantation related to long-term postoperative morbidities associated with single ventricle dysfunction.[6] Furthermore, the individual with single ventricle heart disease is subject to frequent cardiac surveillance, hospitalizations, diminished exercise tolerance, and need for multiple daily medications.

Specific complaints or symptoms such as fatigue or shortness of breath with exertion, cyanosis (lips and nail beds), dizziness, and palpitations have been previously described in the literature.[7–10] The symptoms can be physically debilitating and emotionally stressful in the Fontan patient. However, the clinical profile of the adolescent/adult Fontan survivor and their worries have not been described in the literature.

The purpose of this study is to describe the clinical profile of adolescent/adult Fontan survivors; and identify the worries, symptoms, and the impact of cardiac surveillance most commonly experienced.

Methods

In this study, the clinical profile, worries, symptom,s and impact of cardiac surveillance concerns were identified in a group of adult Fontan survivors. This study is a descriptive, cross-sectional design.

Sample and Setting

The appropriate Institutional Review Boards reviewed and approved the study. A convenience sample of 54 adolescents and adults, age 15–50, participated in the study between July 2006 and February 2007. The inclusion criteria were as follows: (1) age 15 years and older; (2) English literacy; (3) single ventricle diagnosis; and (4) prior Fontan completion. Exclusion criteria were as follows: (1) severe visual, cognitive, or psychiatric problems precluding informed consent and self-administered questionnaire completion; and (2) recent hospitalization or surgery (<3 months). Most patients should return to baseline physical activity or better within 3 months 'postsurgical procedure or hospitalization.

In the Ahmanson-UCLA Adult Congenital Heart Disease clinic database, 76 Fontan patients were identified, 15 were deceased, 21 had moved or were lost to follow-up, and 40 were identified as eligible. Out of the 40 eligible, 27 responded to the physician's referral letter and were approached in person or via the telephone for study participation. At Children's Hospital Los Angeles, 18 late adolescents were eligible, one was excluded secondary to severe developmental delay, with a total of 17 who agreed to participate. Additional subjects were referred to an outside cardiologist informed of the study and were also recruited from local adult CHD support group meetings. Thirteen subjects were eligible and agreed to participate. However, three subjects from the support group meeting did not return their completed demographic data form, questionnaire, and cardiologist contact information and were excluded. A total of 57 adolescents and adult Fontan patients were consented, with 54 completing study participation.

Measures

Sociodemographic and Clinical Data

Demographic data were obtained from the participants on a standardized intake form included with the study questionnaire. Clinical data were obtained from a detailed medical record review with the use of a standardized form and included information related to cardiac anatomy, single ventricle type, Fontan characteristics (type, fenestration, revisions), date and age of pre- and post-Fontan cardiac surgical procedures, current echocardiogram results (atrioventricular valve regurgitation, quantitative assessment of single left ventricle ejection fraction), New York Heart

Association (NYHA) class, oxygen saturation level, history of arrhythmias and treatment, need for pacemaker, current medications, body mass index (BMI), symptoms and other noncardiac medical conditions and surgical procedures.

Worries, Symptoms, and Impact Cardiac Surveillance

The Congenital Heart Disease—TNO/AZL Adult Quality Of Life (CHD-TAAQOL) is a developed disease-specific health-related quality of life (HRQOL) instrument for young adults with CHD.[11] However, this tool was used to identify worries, symptoms, and the impact of cardiac surveillance as determinants of HRQOL. The CHD-TAAQOL consists of 26 items and three scales: Worries (10 items), Symptoms (9 items), and Impact Cardiac Surveillance (e.g., routine cardiac testing and medical follow-up) (7 items). For each item, the perceived frequency is assessed using a 3-point rating scale indicating "never," "occasionally," and "often." If a problem occurred occasionally or often, then the level of distress associated with the problem was determined through the use of four response categories: "not at all," "a little," "quite a lot," and "very much." These two scales are transformed to an ordinal 5-point rating scale for coding purposes, with score 1 corresponding to "problem does not occur" and scores 2–5 corresponding to the respective emotional response.[12] For example, if a patient indicated that the occurrence of a particular problem was "often," but was not causing distress, then a score of 2 was assigned.[12] In order to identify the problems or concerns associated with distress (ranging from "a little" to "very much"), the subscale items with transformed scores ≥ 3 were totaled and ranked according to the highest percentage. The ranking of individual items is not consistent with the tools' original description. However, other authors have used similar methods to identify the most distressing worries and symptoms.[12,13]

The transformed scores for each scale were summed and calculated into a mean score ranging from 0 to 100, with higher scores indicating better HRQOL. Cronbach's alpha for the three scales were Worries .82, Symptoms .77, and Impact Cardiac Surveillance .78.[11] The CHD-TAAQOL is the only available disease-specific self-report instrument for the desired age range and has been found to be a valid and reliable measure of HRQOL in CHD patients.[11,13] The instrument was modified for use in the United States by conversion from the metric system to imperial units (e.g., how often in the last month were you short of breath strolling less than 100 meters [yards]?). The internal reliability (Cronbach's α) for the three scales in this study were .79, .68, and .71 for Worries, Symptoms, and the Impact of Cardiac Surveillance, respectively.

Procedure

Eligible adolescents/adults who have undergone the Fontan procedure were identified by the patient's cardiologist, nurse practitioner, or directly responded to the investigator by telephone to study advertisements or mailed physician referral letters. Additional recruitment was sought through local CHD support group meetings secondary to decreased response from mailed physician referral letters. Eligibility was assessed by the investigator either over the telephone or in person, and informed consent/assent was obtained. For patients less than 18 years of age, parent(s) and adolescent were approached together for participation with parental consent and adolescent assent required for participation. Once written informed consent/assent was obtained, the participant's clinical information was collected via chart review by the investigator.

The CHD-TAAQOL and a demographic intake form were completed by participants. Most participants either completed the questionnaire during a clinic visit or were given a self-addressed, stamped envelop to return the questionnaire once completed. A telephone number was provided so participants could call with any questions. Upon receipt of the completed questionnaire, the instrument was checked to make sure participants did not inadvertently leave any items unanswered. Participants were offered the chance to complete missing items, but were allowed to leave items blank if desired. A monetary compensation of $20 was provided upon completion of the questionnaire in appreciation for participation.

Data Analysis

Descriptive statistics of demographic, clinical variables and CHD-TAAQOL scales (worries, symptoms, and cardiac surveillance) were expressed in frequencies, percentages, means, and standard deviations. All analyses were performed using SPSS for Windows (version 13.0, SPSS Inc., Chicago, IL, USA).

Table 1. Demographic Characteristics of Adolescent/Adult Fontan Survivors (n = 54)

Demographic	n (%) or Mean ± SD
Age, (15–50), mean ± SD	26 ± 9
Gender	
Male	26 (48)
Female	28 (52)
Ethnicity	
Caucasian	34 (63)
Hispanic	11 (20)
Other	9 (17)
Marital status	
Single	40 (70)
Married	10 (19)
Living with partner	4 (8)
Biological children	
None	49 (91)
1–2	5 (9)
Education	
≤High school	21 (39)
≤College	28 (52)
Beyond college	5 (9)
Employment status	
Student (full-time)	14 (26)
Employed	35 (67)
Unemployed/Disabled	5 (7)
Insurance	
PPO	20 (37)
HMO	14 (26)
MediCal/SSI/Disability	17 (32)
Self-pay/Uninsured	3 (5)

SD, standard deviation; HMO, health maintenance organization; PPO, prepaid organization; SSI, social security insurance.

Results

Patient Characteristics

Demographic and clinical characteristics of the study sample are listed in Tables 1 and 2, respectively. The demographic characteristics identified an even distribution of males and females, the majority were Caucasian (63%), single (73%), employed or full-time student (93%), and with health insurance (94%). The clinical characteristics identified that the majority were single left ventricle (78%), the diagnosis of tricuspid atresia or double inlet left ventricle (59%), lateral tunnel Fontan type (44%), history of arrhythmias (76%), left ventricle ejection fraction percentage >50 (66%), oxygen saturations >90% (70%), frequent headaches (50%), scoliosis (22%), varicose veins, ascites and liver cirrhosis (46%), normal BMI (59%), and NYHA class I (48%) and II–III (52%). The symptoms and other medical conditions listed in Table 2 represent a wide range of symptoms or conditions extracted from the medical chart review.

Worries, Symptoms, and Impact Cardiac Surveillance

The CHD-TAAQOL mean scale scores and items that were associated with any distress (transformed

Table 2. Clinical Characteristics of Adolescent/Adult Fontan Survivors (N = 54)

Clinical Variables	n (%)
Ventricle type	
Left	42 (78)
Right	9 (17)
Undetermined	3 (5)
Diagnosis	
Tricuspid atresia	19 (35)
Double inlet left ventricle	13 (24)
Hypoplastic RV	8 (15)
HLHS / Variants	4 (7)
Double outlet right ventricle	3 (6)
Atrioventricular canal	2 (4)
Ebsteins	1 (2)
Other complex anatomy	4 (7)
Fontan type	
Classic RA to PA	15 (28)
Bjork RA to RV	2 (4)
Lateral tunnel	24 (44)
Extracardiac	13 (24)
Fontan fenestration	
Yes	20 (33)
Fontan revision	
Yes	18 (33)
Stage 1 palliation	
BT shunt	15 (28)
PA band	7 (13)
Norwood / BT shunt	4 (7)
Waterston	4 (7)
Central shunt	3 (6)
NYHA	
I	26 (48)
II	17 (32)
III	11 (20)
LVEF%	
≤50	14 (33)
>50	28 (67)
AVV regurgitation	
Trace	18 (44)
Mild	20 (49)
Moderate–severe	3 (7)
Number of daily medications	
≥3	33 (61)
<3	21 (39)
Cardiac medications	
Aspirin	23 (43)
Coumadin	25 (46)
Diuretics	23 (43)
Ace inhibitor	31 (57)
Beta-blocker	15 (28)
Antiarrhythmic	18 (33)
History of arrhythmias	
Yes	41 (76)
Type of arrhythmia	
Atrial fibrillation/flutter/atrial tachycardia	42 (78)
Sinus node dysfunction	10 (19)
Ventricular tachycardia	7 (13)
Junctional rhythm	5 (9)
Pacemaker	
Yes	22 (41)
Oxygen saturation	
≤90	17 (31)
>90	37 (69)
Body mass index	
Underweight (<18.5)	5 (10)
Normal (18.5–24.9)	31 (59)
Overweight (25–29.9)	9 (17)
Obesity (>30)	2 (4)
Symptoms and other medical conditions	
Fatigue	28 (52)
Palpitations	27 (50)
Headaches	27 (50)
Anxiety symptoms	17 (32)
Scoliosis	12 (22)
Varicose veins	11 (20)
Ascites	10 (19)
Liver cirrhosis	8 (15)
Hypothyroidism	7 (13)
Cyanosis	7 (13)
Listed for heart transplant	4 (8)
Protein-losing enteropathy	2 (4)

RV, right ventricle; RA, right atrium; HLHS, hypoplastic left heart syndrome; PA, pulmonary artery; BT, blalock taussig; NYHA, New York Heart Association; LVEF%, left ventricle ejection fraction; AVV, atrioventricular valve.

Table 3. CHD-TAAQOL—Worries, Symptoms, and Impact of Cardiac Surveillance for Whom Patients Reported Distress

Variable	n (%)*	Mean ± SD
Worries		70.4 ± 18.6
Current health	45 (83)	
Job/future job	37 (69)	
Working life	33 (61)	
Living independently	29 (54)	
Exertion or sports	26 (48)	
Education	25 (46)	
Relationships	25 (46)	
Having children	25 (46)	
Having friends	24 (44)	
Being home alone	18 (33)	
Symptoms		77.4 ± 13.8
Shortness of breath walking ½ to 3 miles	37 (69)	
Palpitations	33 (61)	
Dizziness	33 (61)	
Nocturia	26 (48)	
Shortness of breath walking < 100 yards	23 (43)	
Edema (ankle, legs, abdomen)	15 (28)	
Looking pale	15 (28)	
Cyanosis during exercising	14 (26)	
Orthopnea	12 (22)	
Impact of cardiac surveillance		80 ± 10.3
Cardiac hospital admission	19 (35)	
Cardiology visit	16 (30)	
Echocardiogram	13 (24)	
Taking blood	13 (24)	
Electrocardiogram	12 (22)	
Chest x-ray	11 (20)	
Flu vaccine	6 (11)	

*Percentages totaled and ranked according to transformed scores ≥3 on a scale of 1–5.
Scale range 0–100; higher scores indicating less symptoms, worries and impact of cardiac surveillance, or better HRQOL.
SD, standard deviation; CHD-TAAQOL, Congenital Heart Disease TNO/AZL Adult Quality Of Life; HRQOL, health-related quality of life.

scores ≥3 on a scale of 1–5) ranked by percentage under worries, symptoms, and impact of cardiac surveillance are shown in Table 3. The mean scores (worries 70.4 ± 18.6; symptoms 77.4 ± 13.8; impact of cardiac surveillance 80 ± 10.3) indicate perceived symptoms and worries related to their HRQOL (scale range 0–100, higher scores indicating less symptoms and worries or better HRQOL). However, the identified worries, symptoms, and impact of cardiac surveillance are viewed as determinants of HRQOL and not a direct indicator. The primary worries were related to current health (83%), job/employment (69%), ability to work (61%), and living independently (54%). The most bothersome symptoms were shortness of breath with ambulation (69%), dizziness (61%), and palpitations (61%). Furthermore, cardiac hospital admissions (35%) and cardiology visits (30%) were of most concern under impact of cardiac surveillance.

Discussion

Patient Characteristics

In this study, the Fontan survivors had a mean age of 26 ± 9, were Caucasian, single, attended college or trade school, and are employed with some form of health insurance. Conversely, previous studies have reported patients with complex CHD to have difficulty maintaining employment and health insurance related to physical limitations or a "pre-existing condition," with 10–22% of adults uninsured.[14,15] In addition, only five participants had biological children (four male and one female)

Fontan survivors in this study had single left ventricles (tricuspid atresia, double inlet left ventricle), lateral tunnel Fontan type, and NYHA classification I or II. These findings are consistent with other studies examining adolescent and adult Fontan survivors.[6–8,16–18] As survival improves for patient with single right ventricle defects, the clinical profile will change to reflect this single ventricle subgroup and its acquired medical conditions.

As early mortality in the Fontan population decreases, late morbidity is becoming of great interest. The following symptoms or medical conditions identified in the chart review could potentially be explained from long-term Fontan physiology, chronic antiarrhythmic therapy, surgical sequelae from a previous era, or occur independently from their heart disease.

Arrhythmias/Hypothyroidism

Previous studies have identified atrial arrhythmias, exercise intolerance, ventricular failure, cyanosis, protein-losing-enteropathy, and thrombotic events as a concern for long-term outcomes.[7,8] The incidence of arrhythmias in this study was 76%, with the most common type being atrial fibrillation, flutter, or atrial tachycardia. These arrhythmias are notoriously resistant to antiarrhythmic pharmacological therapy and can be associated with rapid hemodynamic deterioration.[10,16] Nonpharmacologic management options include catheter or surgical ablation (e.g., Maze procedure), automatic implantable internal defibrillators, and new generation pacemakers designed to prevent and treat atrial tachyarrhythmias. Furthermore, the risk of thromboembolic events from frequent atrial tachyarrhythmias may warrant antiplatelet or anticoagulation therapy. In this study, 43% of the participants were on antiplatelet therapy, 46% anticoagulation therapy, and 41% required pacemaker placement. These

findings confirm the extensive literature on anticoagulation, arrhythmias, and pacemaker use in this population.[10,19]

Most patients in this study were treated with amiodarone or sotolol for atrial arrhythmias. However, thyroid dysfunction was a medical condition identified as a long-term complication of antiarrhythmic therapy, especially with the use of amiodarone. If amiodarone use is unavoidable, vigilant monitoring of thyroid function should be performed on a routine basis or with any change in clinical condition.

Varicose Veins, Abdominal Ascites, Liver Cirrhosis

The medical findings of varicose veins, abdominal ascites, and liver cirrhosis are likely the result of chronic passive venous congestion related to Fontan physiology and reduced ventricular function. In this study, liver cirrhosis/fibrosis was an incidental finding on chest magnetic resonance imaging scans to evaluate heart function or Fontan baffle/conduit obstruction. In addition, none of the eight participants with cirrhosis had a documented history of alcohol abuse and they were classified as having "cardiac cirrhosis." Hepatic changes have been documented related to Fontan duration and hepatic vein pressure with the development of liver fibrosis.[20,21] In addition, the risk of gastro-esophageal varices and regenerative liver nodules, a precursor to hepatocellular carcinoma, can develop in this population. However, the question still remains as to when liver dysfunction will develop and compromise patient survival.

Scoliosis/Orthopedic Conditions

Another interesting medical condition identified was the presence of scoliosis with some patients requiring Harrington rod placement. In this study, the majority of patients underwent thoracotomy incisions for systemic to pulmonary artery shunts or pulmonary artery banding procedures as their first palliative procedure in the late 1950s to early 1980s. The most common approach for the systemic to pulmonary artery shunts was the posterior lateral thoracotomy incision. Early surgical approaches often required the division of the latissimus dorsi and serratus anterior muscles with rib removal to obtain better visualization and access to the heart structures. Consequently, this leaves the patient at risk for musculoskeletal deformities. These findings confirm previous CHD studies that identified an increase prevalence of scoliosis in patients with thoracotomy incisions operated at an early age.[22,23] Furthermore, scoliosis was primarily a thoracic curvature, averaging 10–20 degrees and directed to the side of the surgical approach. The current muscle-sparing approach to thoracotomy procedures may help minimize the development of scoliosis. However, scoliosis should be routinely monitored in all thoracotomy patients until skeletal maturation is complete.

Headaches

Another medical condition of interest is the complaint of headaches in Fontan survivors. Headaches have obtained recent attention as a frequently identified subjective complaint.[24,25] In this study, 50% of the participants complained of frequent headaches or migraines. There are many potential triggers or causes for headaches in the general population. However, some Fontan patients have potential physiologic explanations for headaches such as elevated central venous or Glenn shunt pressures, reduced oxygen saturation levels (e.g., <90%) secondary to residual cyanosis and subsequent development of hyperviscosity symptoms.[10] Conversely, only 30% of study participants had oxygen saturation levels less than 90% and 13% with visible cyanosis on clinical examination. However, the question remains as to cause of headaches in cyanotic and noncyanotic Fontan patients. These findings require further investigation into the cause of headaches and potential treatment in Fontan survivors.

Functional Status/BMI

The NYHA class is a frequently used objective measure of functional or physical status. Previous studies identified correlations between the physical functioning domain of HRQOL and NYHA class in adult survivors with CHD.[9,16,26] Furthermore, one would assume that deficits in physical functioning would contribute to increased weight or BMI. In this study, almost 50% of participants were NYHA classes II and III. However, the BMI of participants demonstrated normal weight in 59%, with 10% underweight and 20% overweight. Most children with complex heart disease have abnormal growth parameters in height and weight.[1] The abnormal growth is thought to be related to low energy intakes and high resting energy expenditures as children. However, gastrointestinal malabsorption syndrome can develop over time and is thought to be related to chronically elevated central venous pressures and low cardiac output (e.g., protein-losing enteropathy). The physical findings of cardiac cachexia, as described in the adult heart failure literature,[27] require further investigating into the applicability of this term in the failing Fontan population.

Worries, Symptoms, and Impact Cardiac Surveillance

For many patients with a single ventricle, their heart defect impacts their life on a daily basis. The study finding showed mean scale scores ranging from 70 to 80 (scale 0–100, with higher scores indicating better HRQOL). However, the CHD-TAAQOL was used in this study to identify worries, symptoms, and impact of cardiac surveillance which are viewed as determinants of HRQOL and not a direct measurement. Kamphuis and colleagues[11] reported scale scores for worries (84), symptoms (86), and cardiac surveillance (85) in a group of patients with complex heart disease. However, the study population was younger (17–32 years of age), with only a small portion diagnosed with complex single ventricle defects. This could explain their findings of scale scores in the mid to high 80s. Furthermore, there are very few nonproxy-reported studies in the literature specifically addressing the determinants of HRQOL or quality of life in adolescent/adult Fontan survivors in the United States.[28] The majority of HRQOL research in CHD has been conducted at Canadian and European centers.[7,8,11,13,16,29] However, the generalizability of the results to the single ventricle population in the United States is questionable because of cultural, social, and economic difference. In addition, most studies examined all types of CHD and only a small portion of the study participants had single ventricle CHD.[7,11,13,16,29]

Worries of the adolescent/adult Fontan survivor were related to their general health, job/future job, ability to work, and live independently. Furthermore, over 50% of the study participants are single and living with their parents into adulthood. Moons and colleagues[13] reported similar findings of 55% unmarried and living with their parents in 626 patients with CHD. Living independently is a concern that has emerged as a common theme in qualitative studies in CHD.[29–31] Several investigators report a "feeling of being different," along with negative psychosocial outcomes such as diminished self-esteem, anxiety, depression, and poor emotional or social adjustment.[26,29–35] Others have identified concerns for the illness burden on the family, ability to work, self-management of illness, fear of sudden death and pregnancy.[30–32] The need for dialog with parents and patients during routine healthcare visits to address these concerns can help to facilitate the responsibilities of self-care in preparation to live independently and transition to adult care providers.

Symptoms of shortness of breath with walking, dizziness, and palpitations were perceived as more distressing in the Fontan survivors. These symptoms can be debilitating and provoke the fear of a life-threatening arrhythmia and sudden death. Saliba and colleagues[7] identified worse health status with age in adults with univentricular heart disease. This was thought to be related to uncertainties associated with the natural disease course. However, worse symptoms may have a more physiologic basis related to decrease ventricular function over time. Furthermore, routine assessment and symptom management may help alleviate emotional stress and improve HRQOL in Fontan survivors.

The impact of routine cardiac surveillance was not perceived as emotionally distressing. Most patients with single ventricle CHD have grown up with frequent cardiology and hospital visits. The familiarity with routine cardiac testing and required medical follow-up may explain the higher mean scores in this scale.

Limitations

One of the primary limitations to this study is a small sample size from a numeric value but is comparable with previous studies.[7,8,16,18] However, single ventricle heart disease only represents a small subgroup of CHD and recruiting only adolescent/adult survivors further reduces the sample. Another limitation in this study is that the sample is primarily representative of single left ventricle Fontan patients. The majority of single right ventricle patients are starting to reach young adulthood and beginning the transition to adult CHD clinics. The sample also represents the surgical progression of the Fontan operation and reduces the generalizability of the findings to a specific Fontan type. In addition, this population may be healthier than normal secondary to selection bias (e.g., patients currently receiving medical follow-up, agree to participate, speak English, met inclusion/exclusion criteria) and may not be generalizable to the entire population of Fontan survivors. In this descriptive, cross-sectional design, medical history data were collected from retrospective chart review and questionnaire data on worries and symptoms from one point in time. Future longitudinal studies will be required to assess the changing clinical profile of Fontan survivors over time. Nevertheless, despite these limitations, this study contributes to the adolescent/adult CHD literature by describing the clinical

profile of the Fontan survivor and their psychosocial concerns.

Conclusion

The clinical profile of the Fontan survivor identifies residua and sequelae from multiple surgical procedures, medications, and the natural disease course. This includes arrhythmias, varicose veins, liver cirrhosis, ascites, scoliosis, chronic headaches, and thyroid dysfunction. The clinical symptoms of shortness of breath, palpitations, dizziness and worries of general health, employment, ability to work, and live independently were identified as being most stressful to this population. The clinical profile will continue to change with advanced age, improved medical and surgical management, and survival of more complex single right ventricle Fontan patients. Our results support the need for ongoing assessment of both physical symptoms and psychosocial concerns, and suggest the need for multiple risk factor intervention strategies that improve physical and emotional health in Fontan survivors.

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Conflict of interest: None.

Accepted in final form: November 14, 2010.

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