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The effects of a low-intensity exercise program on the fitness level of a 22 year-old female with congenital univentricular heart repair

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THE EFFECTS OF A LOW-INTENSITY EXERCISE PROGRAM ON THE FITNESS LEVEL OF A 22 YEAR-OLD FEMALE WITH CONGENITAL UNIVENTRICULAR HEART REPAIR

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Table of Contents

THE EFFECTS OF A LOW-INTENSITY EXERCISE PROGRAM ON THE FITNESS LEVEL OF A 22 YEAR-OLD FEMALE WITH CONGENITAL UNIVENTRICULAR HEART REPAIR

Purpose: This case study examined the effects of exercise on a 22 year-old female with a repaired univentricular heart through early, modified Fontan technique. Improvements in oxygen consumption, blood pressure, heart rate and body composition were observed after a 12-week supervised fitness program. This program considers reversing hypo-activity and the subsequent deconditioning and obesity associated with this patient.

INTRODUCTION

Congenital heart disease (CHD) is a rare occurrence in the general population affecting approximately 8 out of every 1,000 live births (Park, 1984). Incidences increase with the number of the family members who have a defect. Early detection is crucial for effective treatment and appropriate management of symptoms and limitations that accompany the disease. CHD encompasses many classifications of defects based on the symptoms and testing demonstrated at birth. CHDs can be linked to the mother's maternal history. Physicans who suspect CHD's in newborns must determine the status of the mother's general health with specific questioning concerning diabetes or other metabolic disorders. During the first trimester infections such as rubella, herpes and AIDS may be associated with a variety of congenital defects (Veasy, 1995). The mother's age and lifestyle are important factors in the normal development of the fetus. Women at the beginning and end of childbearing years have higher incidences of congenital

anomalies. Usage of alcohol and drug abuse has also been linked to cardiac defects from fetal alcohol syndrome (American Heart Association, 1998).

A child or infant with potential cardiac problems should be examined for obvious physical symptoms of distress. Cyanosis may not be readily apparent in a newborn child with suspected CHD. This is associated with decreased arterial saturation, which in mild cases is usually 85% or lower before it is detected in patients with normal hemoglobin (Park, 1984). Long-standing arterial desaturation of more than a few months usually results in clubbing of the fingernails and toenails. This widening of the fingertips and thickening of nail beds can also be associated with other diseases as well as with normal persons (genetic trait). Such factors as respiratory rate and dyspnea should be noted especially if altered after eating. A child's general state of well being should be observed for any abnormal behavioral problems, along with signs of possible chromosomal abnormalities. After physical inspection aggressive testing should take place to determine the cause of symptoms. Abnormal blood pressures and auscultation of the heart sounds detecting murmurs require further investigations. Electrocardiography, chest roentgenography, echocardiography, or cardiac catheterization should be used to diagnosis the pathology. Most newborns diagnosed with CHD require palliative procedures in later childhood to keep the demands of the growing body matched with their blood pumping capabilities. Some may need temporary corrections as infants until a later repair can take place with less risk to the child's survival.

The lifetime success of living with congenital heart disease requires routine supervision by a cardiologist, possible future corrective surgery, and proper attention/education to lifestyle by the patient. The purpose for this study to examine the

role of exercise on a sedentary, overweight female with a rare univentricular heart defect (less than 1% of all congenital disease). The current literature states that patients with congenital defects have limited exercise capacity, as reflected in reduced cardiac output at rest and during exercise (Graham, Bricker, James, & Strong, 1994). Reduced exercise ability combined with a commonly observed psychological phenomenon of those with chronic disease to "withdraw, become depressed, and be disabled by inactivity" or what can be observed as The Disability Syndrome (Marley, 1997), may contribute to progressive weight gain and early mortality of CHD patients.

THE PATIENT

This 22 year-old female was born without cyanosis but with a detected heart murmur. Her mother developed diabetes during pregnancy and no family history of CHD was determined. Eight months later the infant underwent cardiac catheterization because of cardiomegaly and increased pulmonary blood flow on a chest roentgenogram. The cardiac catheterization disclosed the presence of a single ventricle with unrestricted pulmonary blood flow. Shortly thereafter, she underwent banding of the pulmonary artery and closure of the patent ductus arteriosis to allow survival, alleviate symptoms and preserve the myocardium for later repair. Although this palliative procedure is not as popular as it once was, it is still the most feasible for children with unrestricted pulmonary flow and preventing pulmonary vascular disease until corrective repair can be done (Danilowicz, Presti, & Colvin, 1990). Years later a gradual increase in her hemoglobin and hematocrit concentrations indicated progressive restriction of pulmonary flow as noted by the attending pediatric cardiologist. Symptoms of breathlessness,

cyanosis, and fatigue accompanied the increased blood concentrations making surgery immanent.

At five years of age a Fontan-type procedure was performed at the Mayo Clinic to separate arterial and venous return so that adequate oxygen would be available to the systemic circulation. This patient's current physiology is not thoroughly known according to her supervising pediatric cardiologist; however, commonalties for all single-ventricle exist in the literature. Prior to surgery her undeveloped right ventricle gave rise to one chamber that presented as the systemic pumping mechanism for oxygenated and deoxygenated blood. The deoxygenated blood from the vena cavae to the right atrium was unable to pass through the ventricle due to tricuspid atresia. An atrial septal defect allowed passage of blood to the left atrium where it mixed with oxygenated blood pumped in through the lungs where it was then pumped into the single ventricle. Both pulmonary and aortic valves returned the partially oxygenated blood to the lungs and to the systemic circulation (Figure 1).

Post-surgery she underwent regular testing to evaluate the repair and associated hematologic changes. Transesophageal echocardiographs taken by her cardiologist demonstrated moderate but stable ventricular dysfunction, recording a stasis artifact (stagnation of fluid) in the venous chamber and part of the Fontan anastomosis. No aortic regurgitation was found but some mild artioventricular valve regurgitation was noted. Electrocardiogram revealed sinus tachycardia with exercise with the P-wave becoming buried in the preceding T-wave giving the appearance of junctional tachycardia. First degree AV block, right axis deviation, strain pattern, left atrial enlargement and right ventricular hypertrophy were also observed. Blood profiles were normal except for

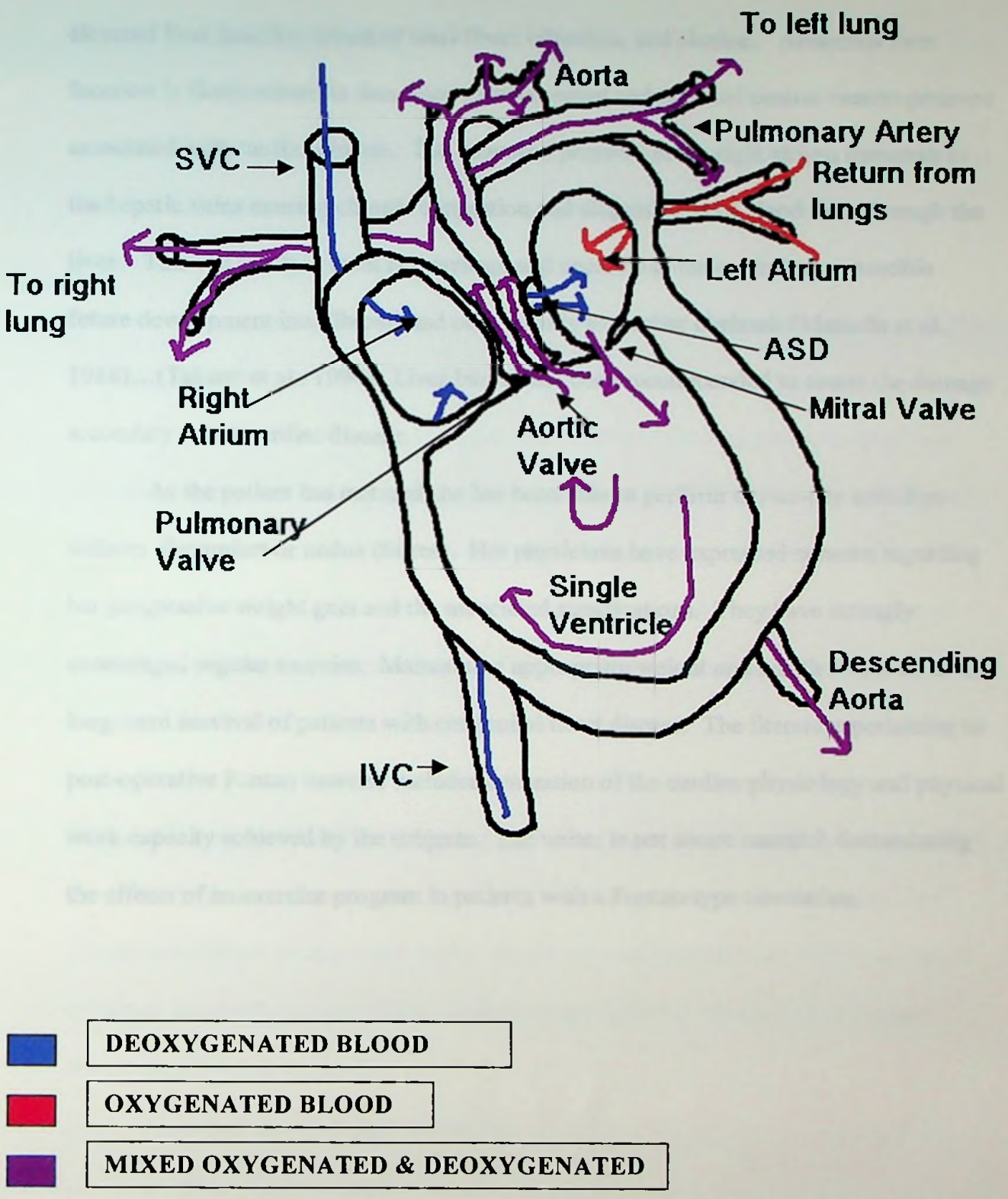

Figure 1. Univentricular Heart With Tricuspid Atresia.

elevated liver function values of total/direct billirubin, and alanine. Abnormal liver function is likely related to decreased cardiac output and elevated central venous pressure associated with cardiac disease. The increased pressure in the right atrium transmits to the hepatic veins causing chronic congestion and sluggishness of blood flow through the liver. This can result in focal hemorrhage and necrosis of the central area; possible future development into fibrosis and occasionally to cardiac cirrhosis (Matsuda et al., 1988)...(Takano et al., 1994). Liver biopsy has been recommended to assess the damage secondary to her cardiac disease.

As the patient has matured she has been able to perform day-to-day activities without discomfort or undue distress. Her physicians have expressed concern regarding her progressive weight gain and the associated ramifications. They have strongly encouraged regular exercise. Maintaining appropriate weight and health is critical to the long-term survival of patients with congenital heart disease. The literature pertaining to post-operative Fontan exercise includes evaluation of the cardiac physiology and physical work capacity achieved by the subjects. The writer is not aware research documenting the effects of an exercise program in patients with a Fontan-type circulation.

REVIEW OF LITERATURE

To understand the Fontan-type circulation, one must abandon conventional cardiac anatomy and physiology. The Fontan design is based on pulmonary flow directly from the right atrium to the pulmonary artery without an interposed ventricle to create systemic blood flow (Figure 2). The late surgical anastomosis consisted of valved conduits with recent (including this patient) atriopulmonary connections constructed from generous pieces of pericardium (Colvin, 1998).

Blood circulation flow to the right atrium through the surgical connection and pulmonary artery to the lungs is dependent on the pressure differentials in the body and pulmonary resistance. Surgical success is reliant on the integrity of the pulmonary circulation. Thick walled and narrow vessels offering excessive resistance to passive blood flow will negate Fontan surgery. Pulmonary resistance may present after the Fontan. The elevated venous pressure required to overcome this resistance and maintain blood flow may cause systemic diffusion of blood from venous walls. Facial puffiness, pleural effusion, abdominal swelling and poor absorption of nutrients from the intestines are common signs and symptoms. Low lung blood vessel resistance is just one vital component patients must possess before the Fontan can be performed with a successful outcome. Research has established the following eligibility criteria for the Fontan procedure (Fisher et al., 1995):

- Possess normal right atrium size
- Over 4 years of age
- Normal heart rhythm
- No atrio-ventricular valve leak

- Ejection fraction over 60%
- Adequate pulmonary artery size
- Successful previous shunt surgery
- No abnormalities of systemic venous drainage
- Pulmonary lung artery pressure below 15 mmHG

These selection criteria permit patients to achieve survival rates of up to 70% over 5 to 15 year post Fontan. The procedure, however, is not a cure for congenital defects and brings possible complications and risks of early mortality (Driscoll et al., 1992). Patients unsuited for the Fontan may benefit from alternative options such as cardiac transplantation.

Post Operative Risks

Fontan patients with subsequent increased atria pressure commonly develop *protein losing enteropathy* (PLE). Elevated venous pressure reduces intestinal protein. As protein levels in the stomach decrease, so do the concentration levels in the blood. When the levels decrease the fluid seeps out of the blood vessels into the abdomen, lung cavity, ankles and around the heart giving the patient edematons. This protein deficiency can cause diarrhea and nausea. Protein loss can be detected through blood protein measurements of albumin. The function of albumin is colloid osmotic pressure, holding fluid in the blood. Levels below 3.5 g/dl should be treated with medication, higher protein dietary modifications, or surgery to correct the altered heart function and lung blood vessel distortion (Feldt et al., 1996).

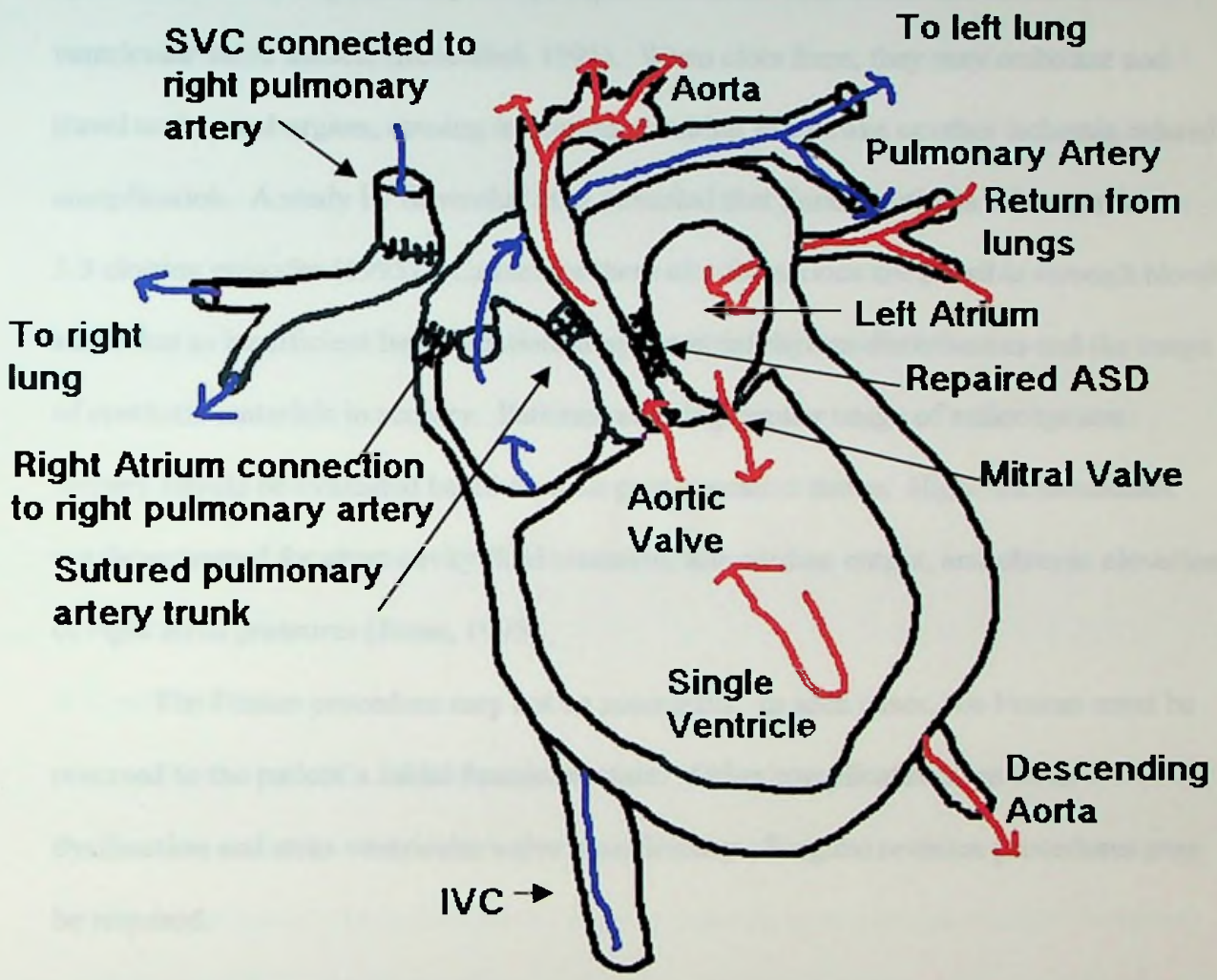

Figure 2. Univentricular Heart Repair Through Fontan Procedure.

Other risks for some Fontan patients are the development of thrombotic events making the usage of anticoagulants part of the routine clinical care. Such thrombi may form on the surface of a patch used to construct the atrial connection for a Fontan operation, inside the pulmonary artery, superior/inferior vena cavas and on the atrio-ventricular valve leaflets (Rosenthal, 1995). When clots form, they may embolize and travel to the vital organs, causing infarction, resulting in a stroke or other ischemia related complication. A study by Rosenthal et al. revealed that Fontan patients will experience 3.9 clotting episodes (1995). Causes for these clot formations are possible through blood stasis due to insufficient heart function, frequent atrial rhythm disturbances and the usage of synthetic materials in surgery. Patients requiring regular usage of anticoagulant therapy should be evaluated based on their post-operative status. High-risk candidates are those treated for chest cavity fluid retention, low cardiac output, and chronic elevation of right atrial pressures (Jonas, 1995).

The Fontan procedure may not be successful. In such cases, the Fontan must be reversed to the patient's initial functional state. Other complications are liver dysfunction and atrio-ventricular valve insufficiency. Surgical revision procedures may be required.

Quality of Life after the Fontan

The Mayo clinic conducted a fifteen-year follow-up of Fontan patients. Of the 352 patients 10.5% viewed their health as excellent five years post Fontan (i.e., had no heart related symptoms or no cardiac medications except for digitalis, and were able to perform as much or more physical activity as their peers) and 50.6% felt they had an

intermediate outcome. Patients questioned about their health ten to fifteen years post surgery had significantly lower outcomes (Driscoll, 1992). This suggests a slow decline in functional status in some patients after the Fontan operation. The Fontan procedure does not guarantee clinical success for univentricular heart patients but with continued improvements in medical technology it can provide a better quality of life. Most Fontan patients lead normal lives holding down jobs, engage in moderate exercise, go to school and although rare have children. In the past, female patients post Fontan were discouraged from becoming pregnant due to small sample studies that showed many early miscarriages and spontaneous abortions compared to healthy females. More recent studies, however, present a more positive outlook showing pregnancy and birth being well tolerated after a Fontan operation, although the incidence of miscarriage is still slightly higher than normal and should be monitored closely (Cannobio, 1996).

Exercise Guidelines

Physical exercise has become a mainstay for achieving and maintaining optimal health and preventing disease. Many active adults receive their healthy start in early childhood when activity is nurtured by parents, schools, peers, etc. as a vital component of their physiological growth. These adults may become more likely to continue this activity pattern later in life and prevent the "creeping obesity" associated with maturity. Many children, however, without normal physical activity patterns have impaired fitness levels and consequently more difficulty in maintaining an appropriate body weight and an associated deterioration of their functional work capacity later in life. Constraints against these children may also encompass a host of psychological factors related to their

disease that negatively affect their motivation for physical activity. Such factors as parental/teacher overprotection, social embarrassment, personal fear of death, and ignorance of proper exercise prescription should be addressed when evaluating the activity history of a diseased individual (Saris, 1985).

Although, Fontan patients have lowered exercise tolerance, they can still benefit from many of the activities normal subjects enjoy. The American College of Cardiology classifies the types of sport activities recommended for Fontan procedure recipients. According to Mitchel et al., categories of sport are recognized as having static (involving development of a relatively large intramuscular force with little or no change in muscle length or movement) and dynamic components (involving changes in muscle length and joint movement with rhythmic contractions that develop a relatively small intramuscular force) (1994). Fontan patients can participate in low-intensity competitive sports (class IA) of either moderate or low static demand if they exhibit normal or near-normal ventricular function and oxygen saturation and near-normal tolerance on formal exercise testing (Graham, Bricker, James, & Strong, 1994). These activities include: golf, bowling, billiards, archery, diving, equestrian and motorcycling.

Aerobic Capacity and Heart Rate Response

Long-term hypo-activity among patients with congenital heart disease is likely attributed to the marked decrease in physical work capacity and fatigue associated with the heart pathology. More specifically, those with single ventricle who have undergone the Fontan procedure demonstrate impaired aerobic capacity during exercise compared with control groups as measured by maximal oxygen consumption (VO_2 max) (Mahle,

Wernovsky, & Bridges, 1999). Studies of adults after Fontan operations demonstrate on the average significantly lower maximum heart rates (mean 149 b/min) (Nir et al., 1993)...(Grant et al., 1988) and maximum oxygen consumptions (mean 25.27 ml/kg/min) (Nir et al., 1993)...(Grant et al., 1988)...(Zellers et al., 1989). Blunted heart rate responses in post-operative Fontan patients could be from sinus node or sinus node artery damage during surgical procedures or progressive scarring along suture lines with late occurrences of sinus node dysfunction. (Kurer, Tanner, & Vetter, 1991). Different investigators have reported a variety of heart rate patterns during progressive exercise for patients after the Fontan operation, suggesting variation among individuals and the modifications used in correction of the defect.

Cardiac Output

Reduced exercise capacity in Fontan patients is linked to abnormal cardiac output. Grant et al. reported that seven out of thirteen subjects with normal cardiac output at rest had low peak oxygen consumption with a maximum heart rate similar to the control group (Grant et al.,1988). This implies a decrease in stroke volume during exercise with respect to a VO_2 to heart rate slope compared with healthy subjects (Troutman, Barstow, Galindo, & Cooper, 1998). Studies comparing Fontan-type patients with hypoplastic left heart syndrome to total cavopulmonary connections demonstrated similar findings in regard to reduced maximum oxygen consumption implying similar aerobic capacity and cardiac output responses (Joshi, Carey, Simpson, & Paridon, 1997). Rosenthal et al. also compared similar atriopulmonary and total cavopulmonary connection Fontan procedures and determined the same maximum exercise performance although there were findings of

variances in physiological adaptations and slightly different cardiac outputs at rest (1995).

Harrison et al. measured decreased exercise ejection fractions in adult patients 6.7 years after Fontan repair as 40 ± 15% compared with controls of 70 ± 8% (1995). The relationship between ejection fraction and ventricular contractility is still unclear in Fontan physiology because preload to the ventricle is reduced in the unusual pulmonary circulation lacking the impact of a pulmonary ventricle (Uemura et al., 1995). Findings of abnormal ventricular contraction during exercise compared with similar studies indicate this as very common in clinically well patients after the Fontan operation, but can not be predicted from exercise capacity or measurements of ventricular contraction at rest (Del Torso, Kelly, Kalff, & Venables, 1985). The implications for this phenomenon remain unclear.

Oxygen Saturation

Subjects with Fontan physiology have slightly lower than normal blood oxygen saturation levels at rest, which decrease further with exercise. Mayo Clinic Fontan patients 2 months to 13.4 years post operation reported SaO_2 values from 92.49 ± 2.89 to 89.51 ± 4.17 at rest and at peak exercise (Durongpisitkul et al., 1997). According to Grant et al. this is due to ventilation perfusion imbalances where abnormal distribution of blood flow increases to the upper lobes of the lung and increasing physiological dead space (1988). This increased upper lobe blood flow creates pulmonary vascular resistance making oxygen loading difficult into the blood of a ventilated lung. Durongpisitkul et al. mentions that preoperative left pulmonary artery stenosis and those with classic Glenn

anastomosis (connection of the superior vena cava to pulmonary artery bypassing the right ventricle), known to be associated with abnormal pulmonary arteriovenous openings, may play roles in the decreased oxygen saturation after the Fontan operation (1997).

Ventilatory Response

Studies performed on the role of the right ventricle in the Fontan circulation during exercise by Rhodes et al. reveals low minute ventilation compared to controls but excessive ventilation for their VO_2 level, possible through the maldistribution of pulmonary blood flow (1998). In contrast, Troutman et al. found excessive ventilation in Fontan subjects as determined by V_E/Vco_2 measures, but attributed the values more to increased ratio of deadspace to tidal volume and changes in chemoreceptor setpoint for arterial carbon dioxide tension (1998). This high physiological dead space can be caused by abnormal distribution of ventilation and low pulmonary blood flow due to areas of the lungs being ventilated but not perfused with reduced movement of carbon dioxide into alveolar gas. Thus, a decrease in the concentration of carbon dioxide in mixed expired and end-tidal gas measurements (Grant et al., 1988).

Arrhythmias

Most Fontan patients experience cardiac arrhythmias during exercise. A study by Harrison et al. tested 30 patients with 24-hour ambulatory EKG monitoring revealed predominant sinus rhythm, atrial flutter, atrial tachycardia, accelerated junctional tachycardia, paced rhythm, and premature ventricular contractions (1995). Reported

abnormalities include sinus node dysfunction, atrial fibrillation and flutter. These potentially fatal arrhythmias may be caused by sinus node or sinus node artery damage during surgical procedures or progressive scarring along suture lines, increased atrial pressures, stretching of the atrium, decreased function of the ventricles, and AV-valve leaks. Antiarrhythmic medications have been successful in controlling the disturbances at rest. Some patients, however, may require pacemaker insertion if they present with complete heart blocks, tachy-brady syndrome, or frequent arrhythmias (Kurer, Tanner, & Vetter, 1991). In the event of resuscitation, basic cardiac life support should be supplemented with manual external abdominal compressions (four upper abdominal compressions for ten seconds each followed by a pause of five seconds given each minute). This will increase the flow into the lungs during relaxation, resulting in maximal filling of the main ventricle and adequate cardiac output (Prabhat & Suresh, 1994).

METHODOLOGY

The patient was required to read and sign an informed consent form explaining the purpose, benefits, confidentiality, risks and discomforts associated with this study. The form includes information for the patient to contact the Human Subjects Research Review Committee and all investigators with any questions and concerns she may have while participating in the study. The description of discomforts and assumed risks involved with testing and exercise training included: fatigue during and after exercise, dizziness, delayed onset muscle soreness (DOMS), nausea, shortness of breath, muscle injury, syncope, elevated heart rates and blood pressures, cardiac arrhythmias, and possible death. The subject was instructed in proper exercise technique, exercise prescription strategies and methods to develop personal exercise programs. The patient was given a Medical Profile Test that included a detailed history, physical examination by her cardiologist, multi-stage exercise test (MSET), pulmonary function test, resting metabolism and body composition analysis prior to initiating the training program.

Exercise Testing

The MSET VO_2 test was conducted with a Marquette Case - 16 Exercise Testing System using a modified Bruce treadmill protocol at the beginning and at the end of the 12-week study. A twelve-lead EKG with exercise leads monitored heart rate. This protocol began with a constant walking speed of 1.7 m/hr at 0% grade with increased intensity at each three-minute stage. Signs, symptoms and subjective ratings of perceived exertion were measured by the Borg's scale to correlate with heart rate at the end of each stage. Oxygen saturation levels were monitored during the test and throughout the study

by Masimo SET Radical finger oximeter. Blood pressures were monitored manually before and after exercise testing and at the beginning of each intensity stage. Respiratory gas measurements were calculated every 20-seconds using a Sensormedics 2900 Energy Expenditure Unit through indirect calorimetry. The expired air was collected through a non-rebreathing valve attached to a facemask covering the nose and mouth. The patient was encouraged to exercise to exhaustion. Criteria for stopping the test were oxygen saturation levels below 90%, subject perceived exertion, respiratory exchange ratio of at least 1.1 or any abnormal rhythm disturbances as evaluated by the attending physician. A recovery period occurred for twelve minutes at 1.7 mph and 0% grade.

Resting pulmonary mechanics were testing prior to testing consisting of forced expiratory volume at one second (FEV_1) and functional vital capacity (FVC). Breathing reserve (BR) at maximal exercise was calculated and expressed as a percentage (BR%=[MVV-VE/MVV]X100). The results of the spirometry were compared with data for healthy adults (Adams, 1998) and research for univentricular heart patients.

The patient's body composition was tested in a bod pod using plethysmography (Life Measurement Instruments) before and after the exercise program. Resting metabolism was checked using indirect calorimetry for initial evaluation of abnormal metabolism (possible thyroid dysfunction as indicated by enlarged thyroid).

Exercise Training

The exercise prescription was designed from the metabolic exercise testing data using the ventilatory equivalent method (Figure 3). American College of Sports Medicine (ACSM) Guidelines for Exercise Testing and Prescription for moderate levels of intensity (40-60% of maximal oxygen uptake) were applied (1996). Exercise workloads were determined by ACSM metabolic equations set at approximately 60% of the patient's oxygen consumption in order to maintain exercise for one hour without undue fatigue (Swain, 1997). All cardiovascular workouts were based on a target heart rate of 133 beats per minute and caloric expenditure of 100 kilocalories progressing to 300 kilocalories near completion of the study. Progression of exercise intensity was dependent on the subject's monitored vitals and perceived exertion. Treadmill workouts were conducted in the Cardiac Rehabilitation Laboratory three days per week for 12-weeks. The duration of study was comparable to other endurance studies where fitness changes occurred (Melanson, Freedson, & Jungbluth, 1996)...(Acevedo & Goldfarb, 1989)...(Taunton et al., 1996)...(Albrecht et al., 1998)...(Block et al., 1990). Each session consisted of a 10 minute warm-up and cool-down with 20-30 minutes of exercise duration. Light progressive resistance exercise was introduced the last month of the study per request of the patient's physical therapist. She was instructed on proper lifting technique, progression of intensity and breathing techniques. Exercise was supervised by exercise physiologists with a physician available during all scheduled sessions. Telemetry was used to monitor cardiac changes during the exercise with defibrillator and crash cart on-hand.

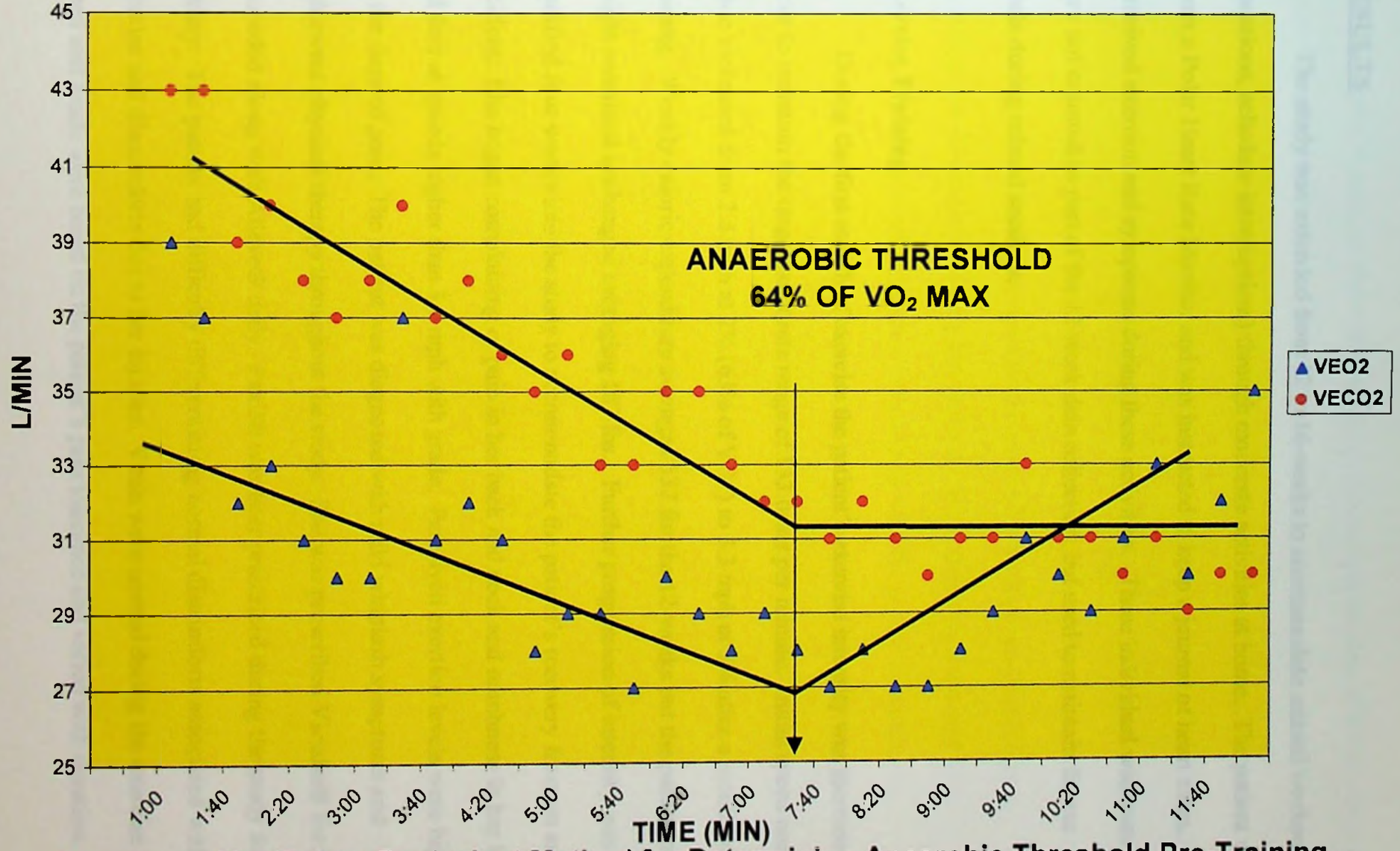

Figure 3. Ventilatory Equivalent Method for Determining Anaerobic Threshold Pre-Training.

RESULTS

The study was extended from 12 to 16-weeks to accommodate missed workouts (vacations, schedule interruptions) through moderate activities at home. The patient was given a Polar Heart Rate Monitor and was instructed to keep a journal of heart rates, perceived exertion and symptoms during these activities. These individual workouts were not counted as part of the 12-week data collection, but used to maintain fitness levels during missed sessions.

Exercise Training

During the first month of exercise the patient's exercise intensity was increased in order to maintain the target heart rate range of 133 beats per minute. Initial speed and grade increased from 2.5 mph at 2% (63% of VO_2) to 3.3 mph at 3% after a month of training. Weekly caloric expenditure averaged 537 for the 12-weeks but the patient's weight remained unchanged averaging 187 lbs. Further progression of intensity was modified four weeks into the study to accommodate the patient's recovery from an auto accident. She began complaining of pain in her back and neck and numbness in her legs and feet at speeds higher than 3.0 mph with grade. Perceived exertion levels were higher on the days of pain. The patient was diagnosed with mild whiplash symptoms and underwent physical therapy throughout the study. She was prescribed Vicadin® for pain as needed along with Alleve® daily. Paxil® was later prescribed during the study for anxiety. The patient had difficulty differentiating normal discomforts associated with exercise and discomforts due to her injuries. Vitals were normal during the exercise but speed and grade were based on the patient's pain tolerance and varied with duration. She

walked longer (30 minutes plus warm-up and cool-down) on the days intensity was less. Heart rates generally stayed below 133 beats per minute on those days. Table 1 demonstrates heart rate, blood pressure, oxygen saturation and RPE ranges noted throughout the exercise program. Resting data was measured an average of 3.5 minutes before and after exercise in a seated position.

There were no significant ECG changes during the study except for one session where an arrhythmia of frequent premature ventricular contractions (PVC) (Lown Classification 2) developed during the warm-up and increased in frequency with intensity. The session was terminated prematurely with the patient exhibiting no symptoms. The PVC's subsided with cessation of exercise and she was closely monitored. The physician attributed the arrhythmia to the patient having eaten prior to exercise and increased myocardial oxygen demand. Exercise was also terminated when the patient had an episode of syncope. All vitals were normal except for a slightly lowered blood pressure at rest. Upon examination, the physician could find no significant reason for the event.

Exercise Testing

Maximal aerobic capacity. The patient exercised to exhaustion during both tests and neither were terminated by the examiners. She achieved an R-value of 1.15 on the first test and 1.23 on the second depicting maximal effort. Maximal oxygen consumption was subnormal in both tests but improved slightly for this patient.

Table 1. Exercise Training Data.

	PRE-EXERCISE RESTING VALUES	EXERCISE VALUES	POST-EXERCISE RESTING VALUES
HR (b/min)	77-98	133	86-108
BP (mmHg)	100-125 70-80	125-165 70-82	98-130 68-90
SaO_2 (%)	94-98	92-95	93-96
RPE		10 TO 13	

Oxygen saturation at maximal exercise. SaO_2 levels for the first test were 92% and 93% for the second test. Various factors contribute to low peak exercise SaO_2 levels including values at rest (higher resting values have higher peak exercise values), age of Fontan procedure (younger ages have higher values), and type of corrections involved with the Fontan (Nir et al., 1993).

Maximal Heart rate response and RPE. The maximal heart rate during the first test was 168 beats per minute and 154 beats per minute for the second test. The patient was able to achieve a MET level of 6.3 instead of 5.7 with a lower maximal heart rate. The patient achieved 90% of normal predicted values for test one and 83% in test two. Although, a higher workload was achieved with a lower heat rate, anaerobic threshold remained unchanged at 61% of the VO_2 max (Figure 4). Perceived exertion ratings of 18-19 were noted at maximal exercise.

Blood Pressure. Blood pressure response to exercise was normal. Systolic pressure rose significantly during the first test. Diastolic rose less during the second test.

Ventilation. MVV values measured prior to treadmill testing were in the 90^{th} percentile of predicted. Breathing reserve at maximal exercise for both tests were 30% of predicted values, normal values being 20% - 50% for healthy adults (Wasserman et al., 1999). Minute ventilation increased with exercise in both tests. The ventilatory equivalent values for oxygen and carbon dioxide remained unchanged for both tests as shown in Tables 2 and 3.

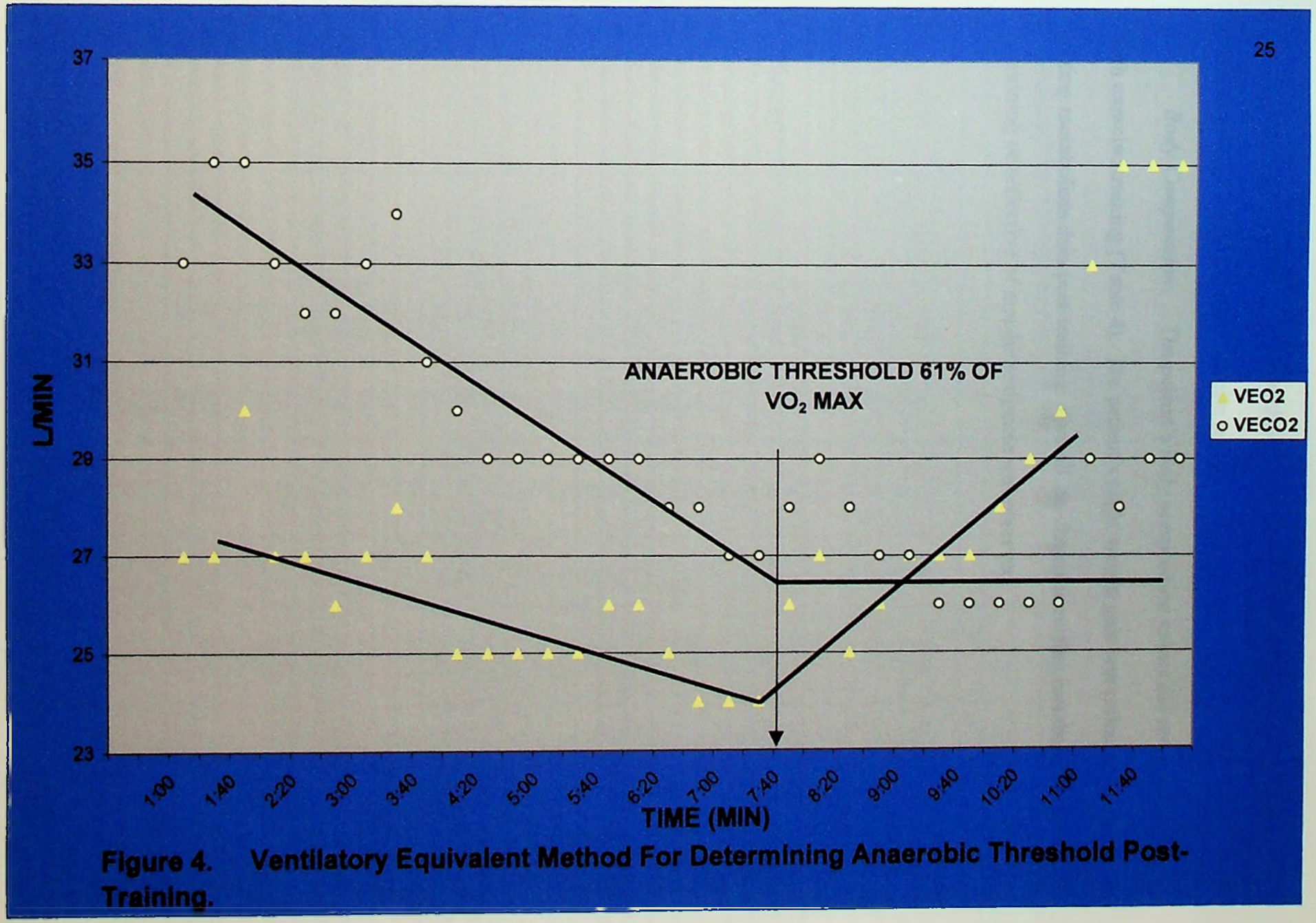

Figure 4. Ventilatory Equivalent Method For Determining Anaerobic Threshold Post-Training.

Electocardiogram. Contour changes consistent with those reported in the literature were observed during exercise testing.

Body Composition. The patient's body composition values did not improve with exercise training (Table 4). The patient's slight weight gain was reflected in the resting metabolism data post-training. Overall, no change in resting metabolism specific to training or reflective of metabolic disease was observed.

Table 2. Metabolic Testing Data Prior to Exercise Training.

				MVV	92					
				FVC	71					
				FEV1	79					
				BR	37%					
	BLOOD								VO2	
	PRESSURE	SAO2%	TIME	VEO2	VECO2	HR	VO2	VCO2	ML/KG/MIN	R
XERCISE										
TAGE 0	128/88	96	1:00	39	43	124	638	566	7.51	0.89
			1:20	37	43	126	646	559	7.6	0.87
..7 MPH			1:40	32	39	122	829	692	9.75	0.83
			2:00	33	40	122	803	657	9.45	0.82
0%			2:20	31	38		720	592	8.47	0.82
			2:40	30	37	123	773	632	9.1	0.82
AGE 1/2	128/86	94	3:00	30	38	119	969	773	11.4	0.8
			3:20	37	40	122	698	634	8.21	0.91
..7 MPH			3:40	31	37	121	797	678	9.38	0.85
			4:00	32	38	124	887	758	10.44	0.85
, GRADE			4:20	31	36	125	860	727	10.12	0.84
			4:40	28	35	127	947	773	11.15	0.82
			5:00	29	36	127	1019	830	11.98	0.82
			5:20	29	33	127	922	797	10.85	0.86
			5:40	27	33	127	1008	826	11.86	0.82
TAGE 1	160/90	93	6:00	30	35	127	951	830	11.1	0.87
			6:20	29	35	128	1071	891	12.6	0.83
..7 MPH			6:40	28	33	129	1067	923	12.55	0.87
			7:00	29	32		1054	941	12.4	0.89
% GRADE			7:20	28	32		1086	949	12.77	0.87
			7:40	27	31		1291	1100	15.1	0.85
			8:00	28	32		1217	1081	14.32	0.89
			8:20	27	31	138	1153	1019	13.56	0.88
			8:40	27	30	138	1256	1110	14.77	0.88
TAGE 2	172/88	92	9:00	28	31	141	1196	1090	14.07	0.91
			9:20	29	31	142	1437	1350	16.91	0.94
5 MPH			9:40	31	33		1268	1169	14.92	0.92
			10:00	30	31	150	1383	1305	16.27	0.94
, GRADE			10:20	29	31		1445	1381	17	0.96
			10:40	31	30	159	1519	1559	17.87	1.03
			11:00	33	31	163	1437	1551	16.91	1.08
			11:20	30	29	166	1527	1582	17.96	1.04
			11:40	32	30	166	1478	1577	17.38	1.07
			12:00	35	30	168	1688	1945	19.86	1.15

Table 3. Metabolic Testing Data Post-Exercise Training.

MVV	96
FVC	76
FEV1	86
BR	31%

	BLOOD PRESSURE	SAO2%	TIME	VEO2	VECO2	HR	VO2	VCO2	VO2 ML/KG/MIN	R
XERCISE										
STAGE 0	122/78	95	1:00	27	33		724	597	8.42	0.82
			1:20	27	35	94	1007	771	11.71	0.77
..7 MPH			1:40	30	35		701	598	8.15	0.85
			2:00	27	33	94	821	661	9.55	0.81
0%			2:20	27	32	97	909	753	10.57	0.83
			2:40	26	32		885	739	10.29	0.83
			3:00	27	33	100	1111	911	12.92	0.82
TAGE 1/2	124/76	94	3:20	28	34		968	802	11.26	0.83
			3:40	27	31		861	740	10.01	0.86
..7 MPH			4:00	25	30		1091	913	12.69	0.84
			4:20	25	29	101	1081	916	12.57	0.85
%, GRADE			4:40	25	29		1038	897	12.07	0.86
			5:00	25	29		986	859	11.47	0.87
			5:20	25	29		1183	1029	13.76	0.87
			5:40	26	29	106	1106	960	12.86	0.87
TAGE 1	128/78	93	6:00	26	29		1145	1024	13.31	0.89
			6:20	25	28		1278	1109	14.86	0.87
..7 MPH			6:40	24	28	111	1161	1021	13.5	0.88
			7:00	24	27		1300	1173	15.12	0.9
% GRADE			7:20	24	27	113	1400	1246	16.28	0.89
			7:40	26	28	119	1209	1126	14.06	0.93
			8:00	27	29		1300	1198	15.12	0.92
TAGE 2	132/78	94	8:20	25	28		1506	1361	17.51	0.9
			8:40	26	27	125	1540	1458	17.91	0.95
..5 MPH			9:00	27	27	138	1505	1525	17.5	1.01
			9:20	27	26		1624	1679	18.88	1.03
% GRADE			9:40	27	26	136	1739	1835	20.22	1.06
			10:00	28	26		1705	1877	19.83	1.1
			10:20	29	26	141	1744	1963	20.28	1.15
TAGE 3	134/80	93	10:40	30	26		1635	1885	19.01	1.15
			11:00	33	29	144	1968	2252	22.88	1.14
..4 MPH			11:20	35	28	147	1725	2125	20.06	1.23
			11:40	35	29	150	1905	2288	22.15	1.2
% GRADE			12:00	35	29	154	1737	2101	20.2	1.21

Table 4. Body Composition Changes With Exercise Training.

	TOTAL WEIGHT (lbs)	LBW (lbs)	FAT WEIGHT (lbs)	% FAT
PRE-EXERCISE	185	100.7	84.7	45.8
POST-EXERCISE	188	97.5	90.5	48.1

DISCUSSION

This study has examined the fitness effects of exercise on a patient after a Fontan operation. The purpose was to demonstrate that a Fontan patient who follows an individually prescribed exercise program should be able to increase their exercise tolerance and aerobic capacity. Exercise intervention can serve to increase quality of living and prevent further cardiac complications associated with a sedentary lifestyle and obesity attributed to the low work capacity and muscular fitness common among these patients. Designing an exercise program for the Fontan patient means special consideration must be given to their unique physiological responses to overload.

Oxygen Saturation

Pulse Oximetry should be used to monitor oxygen saturation levels during exercise in the Fontan patient. Desaturation is common with exercise and is attributed to ventilation perfusion mismatches and poor oxygenation in the lungs from arteriovenous shunts (Gewillig et al., 1990). Although, this patient demonstrated mild desaturation during the low intensity exercise, it became greater with increased intensity. Factors such as the age of repair and the type of Fontan used influences the amount of desaturation in these patients. The American College of Sports Medicine recommends maintaining SaO_2 levels of >88% during exercise training for pulmonary patients (1996). Supplemental oxygen has been useful for patients having difficulty breathing while SaO_2 levels are adequate. According to the literature some Fontan patients may desaturate below 88% with no dyspnea. These patients should be monitored closely on the basis of their particular condition.

Ventilation

VE/VO_2 and VE/CO_2 in Fontan patients tends to be higher during exercise testing than that of control groups. This patient was within normal ranges found in similar testing of Fontan patients (Zeller et al., 1989)...(Nir et al., 1993). VE/VO_2 values prior to the Fontan surgery are elevated among these patients but tend to decrease to normal post surgery. An increase in oxygen consumption with exercise seems to explain near normal values due to better ventilation perfusion matching for oxygen. Abnormal ventilation may be induced by the patients increased need to dissipate CO_2 in maintaining normal pH. According to Chua et al. univentricular patients have a lowered hypoxic chemosensitivity than of Fontan patients. More importantly, there seems to be increased ventilation for a given level of arterial CO_2 tension (VE/VCO_2) which also shifts toward normal values post surgery (1997).

Pulmonary Function

The patient's FEV_1 values were within normal range and FVC values were considered low. According to healthy lung norms, the FEV_1/FVC ratio of 90% may be indicative of some lung restriction (Ruppel, Gregg, 1994). Fontan patients have alterations in pulmonary mechanics possibly due to scar tissue resulting from thoractomies and lung perfusion abnormalities (Mahle et al., 1999). The values for this test could have been affected by patient effort. Difficulties wearing the nose clip while simultaneous mouth breathing and obesity were factors to consider. In studies where pulmonary function values were reported, Fontan patients measured lower than normal

values. More specific clinical testing would need to be performed to exclusively determine restrictive lung impairment. MVV values were measured to determine the patient's lung power. The ability to breathe was not limiting to the patient during exercise as determined by the breathing reserve.

Oxygen Consumption, Heart Rate and Blood Pressure

Oxygen consumption values improved 3.0 ml/kg/min after 12-weeks of exercise. Although, the VO_2 was below normal values the patient demonstrated an ability to improve. The literature has discussed the limited oxygen consumption levels for Fontan patients as related to their abnormal stroke volume response (Zellers et al., 1989). This patient's increased oxygen consumption was likely attributed to better peripheral extraction and increased a-$\overline{V}$ O_2 difference. Myocardial improvements may be more evident with exercise programs of higher intensity. More studies are needed to determine if and how much Fontan patients can improve cardiac output with long-term exercise.

Maximal heart rate and blood pressure was lower with an increased workload. Heart rate reserve (HRR) was higher during the second test by 13 beats/min. Although, higher oxygen consumption was obtained, lowered maximal heart rate with training could not be explained. This decreased HR-VO_2 slope with training may be indicative of physiological adaptations unique to the Fontan patient (Figures 5 and 6). Nir et al., reported that Fontan patients tested the second time post operatively (mean 3.5 years) had a decrease in maximal heart rate of 1.8 beats (1993). Possible factors responsible for this decline were an increase in age, age of Fontan procedure, and time between testings.

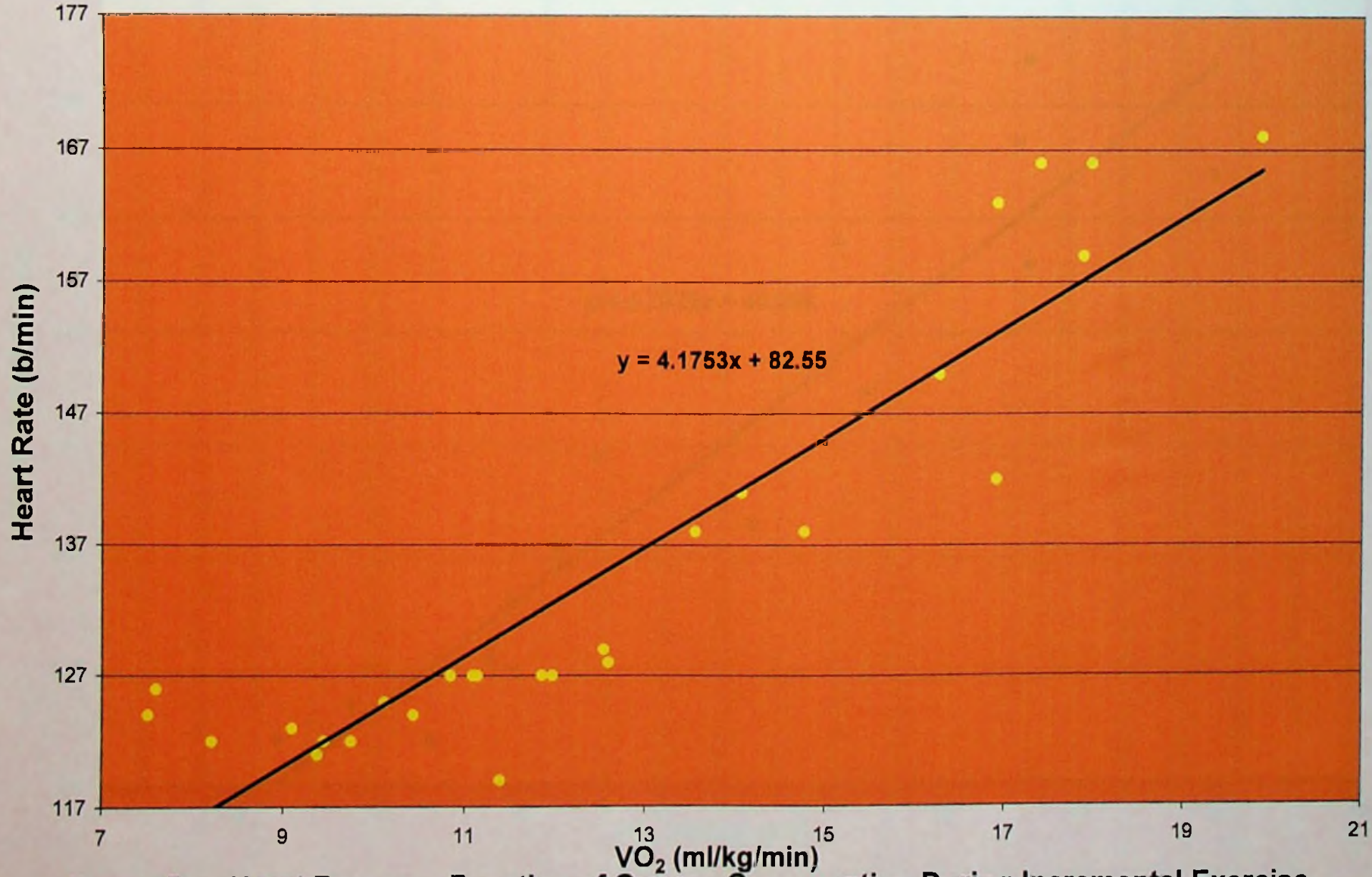

Figure 5. Heart Rate as a Function of Oxygen Consumption During Incremental Exercise Pre-Training.

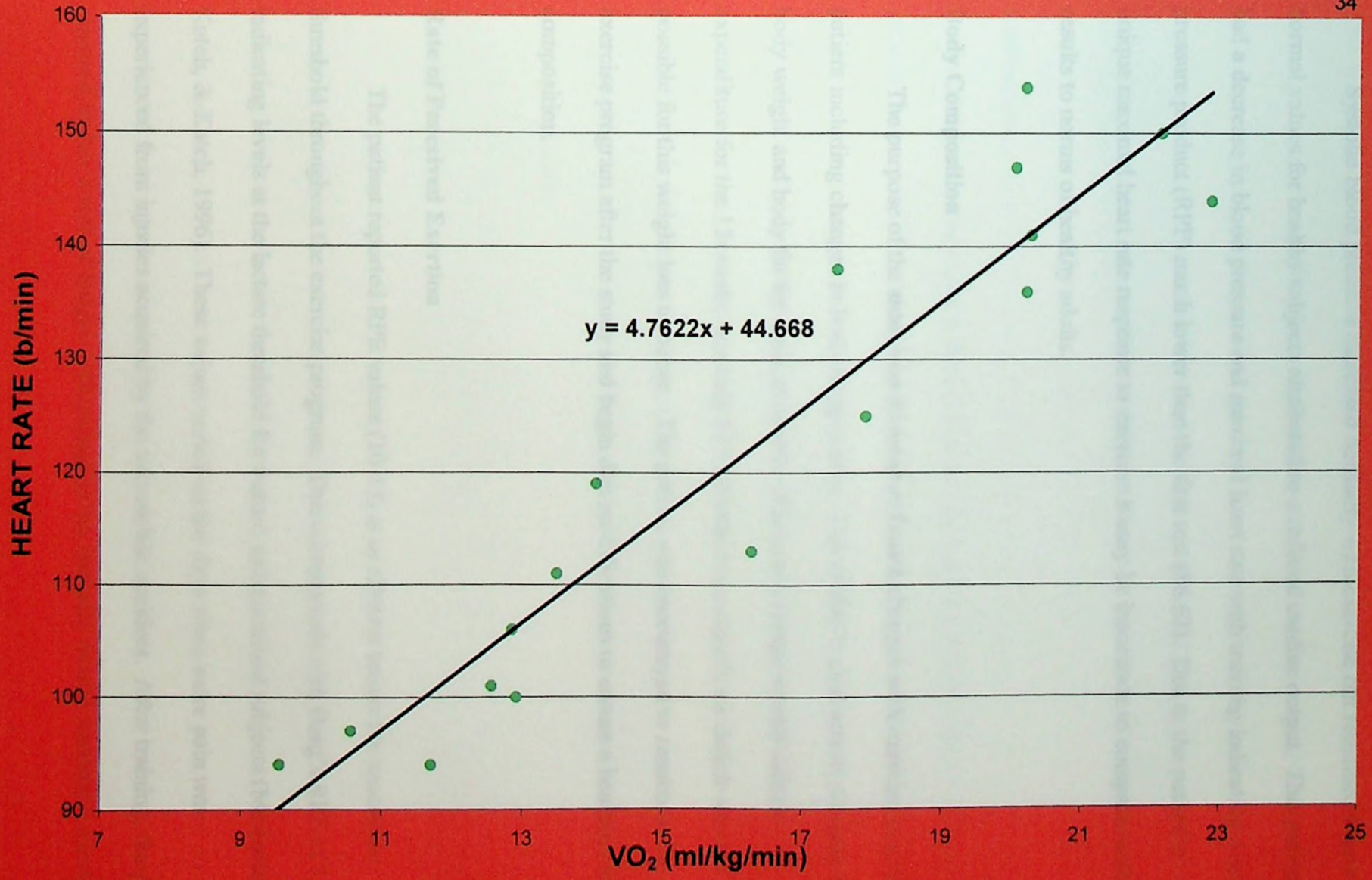

Figure 6. Heart Rate as a Function of Oxygen Consumption During Incremental Exercise Post-Training.

Systolic blood pressure increased normally with exercise but remained below normal values for healthy subjects attributed to a reduced cardiac output. This patient had a decrease in blood pressure and maximal heart rate with training indicating a rate pressure product (RPP) much lower than the first test (86.62). Due to the patient's unique maximal heart rate response to exercise it may be inaccurate to compare such results to norms of healthy adults.

Body Composition

The purpose of the study was to monitor fitness changes with exercise in a Fontan patient including changes in body composition. The subject's goal was to decrease her body weight and body fat for optimal health. Since the average weekly caloric expenditure for the 12-weeks was only 537 kilocalories a significant deficit was not possible for this weight loss to occur. The patient was encouraged to remain in the exercise program after the study and begin diet modifications to ensure a healthier body composition.

Rate of Perceived Exertion

The patient reported RPE values (10-13) at or slightly below the anaerobic threshold throughout the exercise program. This corresponds with Borg's RPE of 11.0 indicating levels at the lactate threshold for trained and untrained subjects (McArdle, Katch, & Katch, 1996). These values varied on the days when more pain was experienced from injuries acquired by the automobile accident. After training, the patient reported higher levels of fatigue with testing as the intensity neared maximum exertion.

This was possibly related to familiarity with the test and the ability to exert more effort (although lactate threshold levels were unchanged). The patient revealed having difficulty comparing her levels of discomfort to the appropriate RPE values because of her inexperience with exercise. RPE was a useful measure for gauging exercise intensity particularly when compared with changing vital signs.

Limitations

Comparing the exercise data of one Fontan patient to studies using multiple subjects was an obvious limitation. Although, the values were true of this individual the results can not be inferred on other Fontan patients exclusively. The methodology had to be modified during the study after the patient's car accident altering the fitness improvements made during testing. The patient also began light resistance training during the later part of the study per request of her physical therapist. Anti-anxiety and pain relief medication was prescribed during this time altering the patient's psychological and physical state.

Conclusion

According to this study, a Fontan patient was able to increase the amount of external work through training with less evidence of cardiac stress. This was corroborated by the patient's reporting of having less fatigue and shortness of breath during her daily activities. Although, this patient's aerobic capacity was below normal, the capacity to improve was evident.

The propensity for Fontan patients to abstain from regular physical activity may be related to psychological and sociological influences rather than from the disability itself. Interestingly enough, Troutman et al., reported that "Fontan patients probably do not participate in physical activity of the same duration and intensity as do healthy children and therefore report abnormal responses to exercise due to deconditioning rather than from real physiologic impairment" (1998). A study by Grundy et al. on physical activity and morbidity in overweight obese individuals, indicated that being physically active reduced obesity-related chronic diseases and decreased risk for early death. Also, active and fit persons who were overweight or obese had lower morbidity and mortality risk than normal weight persons who were sedentary (1999). This information would suggest that Fontan patients who are overweight should engage in regular exercise to prevent future cardiac complications associated with obesity.

Informed Consent to Participate in the Case Study of a Low-Intensity Exercise Program on a Congenital Univentricular Heart Repair

I understand that I am giving my consent to participate in this case study. I also understand that the purpose of this study is to determine the effects of a low-intensity exercise program on the fitness level of a 22 year-old female with congenital univentricular heart repair.

1. Procedures:

I realize that in order to determine the effects of the exercise prescription, it is necessary to participate in testing sessions before and after the twenty-week program in the Marshall University Human Performance Laboratory. More specifically I will participate in the following procedures:

To determine my fitness level I will perform a multi-stage exercise test (MSET) with a twelve lead EKG to maximal effort. The actual test will last as long as it takes for me to reach a perceived exertional level. I understand that I can stop the test at anytime, and for any reason. I also understand that the test may be stopped by the physician if I exhibit dangerous arrhythmias detected by EKG monitoring or any other obvious symptoms that may be of risk.

To determine body composition I will be required to sit in a body box for several minutes. Pulmonary function values will be examined by a spirometer that will require me to breathe maximally.

To examine the effects of exercise, I am required to follow a cardiovascular training program. This will be done primarily through treadmill walking at various speeds and inclines. I understand that I must wear heart-monitoring telemetry during all training sessions. All testing and training sessions will be supervised by a medical physician.

Beth Phillips
Subject's Signature

October 3 2000
Date

APPROVED

MU

2. Risks and Discomforts:

I may experience a sensation of breathlessness, nausea, dizziness and fatigue during the MSET. I understand that these sensations may also occur during the training sessions.

I may experience delayed onset of muscle soreness. This may occur up to 48 hours after training, and can last up to four days after training.

I understand that I may exhibit elevated heart rates, blood pressure, cardiac arrhythmias and possible death.

I understand that I may experience muscular injury from the exercise and falling or stumbling off of the treadmill.

I also understand that in the event of illness or injury as a result of participation in this study, no compensation, financial or otherwise will be provided by Marshall University, or the investigators.

3. Benefits:

By participating in this study, I will learn proper exercise technique, understand exercise prescription strategies and receive instruction that will permit me to develop personal exercise programs.

4. Alternatives:

I understand that I am not required to participate in this study and may withdraw my consent and discontinue participation at any time without penalty or loss of benefit to myself.

IRB
SEP 13 2000
APPROVED
MU

Subject's Signature

October 3, 2000
Date

5. Confidentiality:

I agree that the information gathered from this study may be used in medical and scientific settings. This includes publication, presentation and access to my clinical records by the Institutional Review Board. I understand that my identity will remain confidential to the extent permitted by law and Marshall University.

I have been advised that if I have any questions concerning the study I may contact Dr. William P. Marley at 696-2936 or Alice Elkins at (304) 757-7834. If I have any questions regarding my rights as a participant in a research study I may call Henry K. Driscoll M.D., Chairperson, Marshall University Institutional Review Board at 696-7320.

Prior to participation in this study I will be given a copy of this consent form. I understand the procedures and hereby agree to participate in the study previously described.

Bob Phillips — Subject's Signature

October 3 2000 — Date

Alice A. Elkins — Witness

10/5/00 — Date

IRB APPROVAL EXPIRES 2001

William P. Marley — Principal Investigator

10.5.2000 — Date

IRB SEP 13 2000

APPROVED

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