CASE STUDY

Exercise Testing and Training in a High School Athlete Following Atrial Septal Defect Repair

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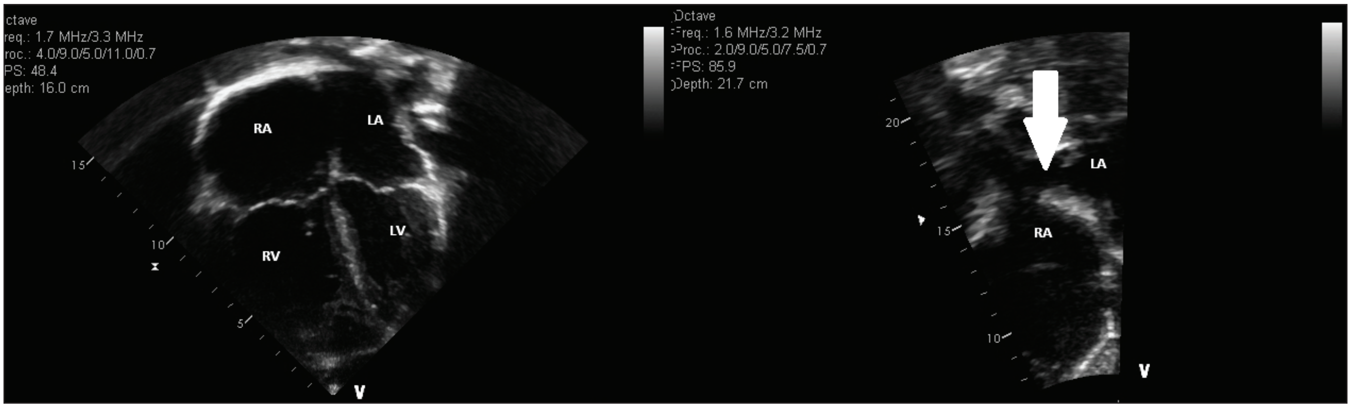
BACKGROUND

In autumn 2013 during a routine check-up, JT, an apparently healthy 16-year-old, presented to his primary care physician with a heart murmur. Despite being an active teenager who played tennis for his high school team and with no overt symptoms, a pediatric cardiology referral was made. An ECG was performed showing normal sinus rhythm with a right bundle branch block. An echocardiogram revealed sinus venosus atrial septal defect (ASD), partially anomalous pulmonary veins, and enlargement in both the right and left atria (Figure 1). However, no functional abnormalities were noted for the ventricles (e.g., left ventricular ejection fraction was >55%), and the remainder of the echocardiogram assessment was normal. Percutaneous transcatheter closure (*see Management section*) was not possible; therefore, JT required an open heart surgical correction.

The repair of a sinus venosus ASD in the pediatric population is an elective procedure as it is rarely acutely hemodynamically significant. However, if left untreated, over time an ASD can cause progressive enlargement of the right atrium and ventricle increasing the likelihood of arrhythmias and decreased cardiac function. ASDs can also cause excessive blood flow to the lungs, which can lead to pulmonary hypertension.

JT and his family elected to schedule his surgery within the first week of June 2014, allowing him greater recovery and training time prior to the start of the high school tennis season in the fall. Closure of the sinus venosus ASD and repair of the partially anomalous pulmonary veins was performed without surgical or post-operative complication. A post-surgery echocardiogram (Figure 2) shows a reduced right atrium and ventricle size. Following an uneventful 3 d hospitalization, JT was discharged to home.

FIGURE 1. Echocardiogram study demonstrating a sinus venosus ASD.



Left pane: Apical four-chamber view showing right atrial and ventricular enlargement. Right pane: Two-dimensional image of a sinus venosus ASD (arrow) as seen in a subcostal sagittal view.

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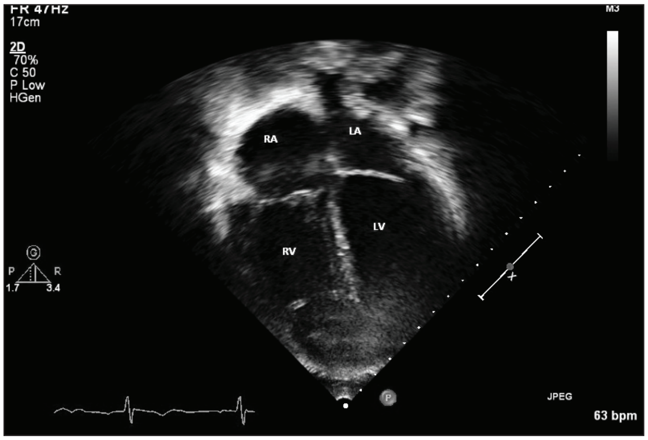
FIGURE 2. Post-surgery echocardiogram demonstrating decreased right atrial and ventricular size.



Prior to discharge, a clinical exercise physiologist met with JT and developed a 2 wk home-based exercise program prior to enrollment in the outpatient cardiac rehabilitation (CR) program at Nationwide Children's Hospital in Columbus, Ohio. The home-based program consisted of daily walking, beginning with 10 min at a light intensity. Over the next 2 wk, this regimen progressed to 25 min daily with intensity remaining light. The upper intensity limit was defined by JT's inability to continuously speak comfortably as he walked (11). Additionally, JT was asked to perform daily upper extremity range-of-motion exercises. The primary aims of the program were to increase exercise tolerance, counteract the effects of bed rest, and to promote basic movement patterns that would encourage sternal and surgical incision healing.

As an athlete, JT had experience with exercise training. He also had a strong desire to return to competition for his senior year of high school. JT began playing tennis in his early childhood. In high school, he began to play year round for both his school and a traveling team. Prior to his surgery, he performed resistance training three times per week at a community recreation center. He also was active with regular bike rides with friends. Adherence, motivation, work ethic, and socioeconomic constructs were deemed to be minimal barriers for JT and his family.

JT returned with his mother for a cardiology clinic follow-up appointment 2.5 wk following discharge. JT reported being adherent with his home exercise program and commented that he wished to do more. Per JT's cardiologist, there were no concerns with his incision site, and JT could begin outpatient cardiac rehabilitation with the following restrictions: 1) no competitive tennis, 2) no driving, 3) no full range of motion trunk rotation as this movement caused him pain, and 4) no heavy lifting.

Prior to beginning cardiac rehabilitation, a graded exercise stress test with measured gas exchange and continuous ECG monitoring was performed. JT tolerated the test without symptoms, and the ECG was negative for arrhythmia and ST segment change. JT's cardiorespiratory fitness was excellent with a peak $\dot{V}O_2$ of 52.1 $mL \cdot kg^{-1} \cdot min^{-1}$, which was 104% of his age predicted value (3). The peak respiratory exchange ratio (RER) was 1.15 and indicated a maximal effort. Total exercise time was 10 min 17 s on the Bruce treadmill protocol, and he achieved a peak heart rate of 173 $beats \cdot min^{-1}$. Despite having an average functional capacity, the rationale for cardiac rehabilitation was threefold: 1) provide additional surveillance during exercise, 2) continue to improve fitness, and 3) increase JTs confidence and self-efficacy, with the goal of safe high school sports participation in the fall.

JT participated in CR at Nationwide Children's Hospital 2 $d \cdot wk^{-1}$ for 6 wk. There was an expectation that JT would adhere to additional exercise at home. The CR sessions were 1.5 hr in duration with 1 hr devoted to exercise training and the remaining 0.5 hr rotated between behavioral and occupational therapy. This schedule was in accordance with CR at Nationwide Children's Hospital, which is a multidisciplinary program staffed by a clinical exercise physiologist, a physical therapist, a registered dietician, an occupational therapist, a psychologist, a recreational therapist, and a massage therapist. Services provided to each participant depend on the needs of that patient as well as insurance coverage. After three CR sessions, the behavioral and occupational therapy services were discontinued as JT and the therapists agreed these were no longer indicated.

JT's exercise sessions consisted of approximately 30 min devoted to the treadmill and/or cycling, 20 min of resistance training, and 10 min of range-of-motion training with the primary aim of returning his ability to play competitive tennis. The exercise intensity was assessed using the heart rate response with a range of 120 ± 5 $beats \cdot min^{-1}$, which corresponded to his anaerobic threshold (AT) as determined during the cardiopulmonary exercise test. Initially, the treadmill workload was set between 4.8 and 5.6 km/h (3.0 and 3.5 mph) at a 0% incline (i.e., 6 METs). The cycle workloads were set between 110 and 130 W. One to two resistance exercises were performed each for the upper and lower body for two to three sets and for 10 to 15 repetitions at a weight that created fatigue near the last several repetitions. Free weights and ankle weights were also used. Partial curl-ups were performed for abdominal strengthening.

JT progressed through the CR program without symptoms and expressed that the initial exercise was "easy." Following session three, continuous electrocardiogram (ECG) monitoring was no longer warranted as a normal heart rate response and no arrhythmias were observed. By his third week, JT was allowed to exercise at heart rates 10 to 20 $beats \cdot min^{-1}$ above AT. By the fourth week, a dynamic warm-up and 5 x 1 min run intervals performed on the treadmill (~10 METs) were incorporated. Resistance training progressed from single- to multi-joint exercises, and JT was no longer limited by incision pain. At this point, JT began playing recreational tennis with his mother. During JT's final two weeks of CR, 5 x 1 min high-intensity directional intervals were incorporated to provide a training stimulus similar to tennis. These intervals were performed on the treadmill and involved 1 x 20 s of side shuffles to the left, 1 x 20 s backward running, and 1 x 20 s side shuffles to the right. Though familiarization was required, JT reported enjoying this workout. His final exercise session was an outdoor active game of Frisbee. Throughout the CR program, his attendance was perfect, and he reported adherence with his home exercise.

Upon completion of the CR program, his cardiorespiratory fitness was reassessed. There was a 20% improvement in both peak $\dot{V}O_2$ and the $\dot{V}O_2$ value at AT. The RER was similar between both tests (i.e., >1.1). His quality of life score, according to the PedsQL™ 4.0 teen questionnaire, also showed improvement (from 13 to 18). In November 2014, he competed in a USTA tennis tournament. At his 1 yr follow-up from initial presentation, he and his parents reported he was back to all his usual activities and pleased with his clinical progress.

DISCUSSION

Epidemiology

In the United States, congenital heart disease (CHD) affects four to eight of every 1,000 live births (12). While atrial septal defects (ASD) are one of the most common types of congenital heart defects, accounting for 8 to 10% of congenital heart disorders, sinus venosus ASDs only account for 5% of all ASDs (2). Most ASDs occur as an idiopathic heart defect, although there are genetic abnormalities that are associated with ASDs (16).

Etiology

A sinus venosus ASD results secondary to the malposition of the insertion of the superior vena cava, which is caused by incomplete reabsorption of the sinus venosus. This incomplete reabsorption results in a defect in the wall of the septum between the left and right atria and is located posterior and superior to the fossa ovalis. A sinus venosus defect is commonly associated with partially abnormal pulmonary venous return, as was the case with JT. An ASD will result in shunting between the right and left atrium as determined by the compliance of the ventricles. In most cases, the right ventricle is more compliant than the left ventricle and, therefore, will result in a left-to-right shunt and subsequent increased pulmonary blood flow.

Diagnosis and Clinical Manifestations

The left-to-right shunt resulting from a large ASD leads to volume overload on the right heart causing dilation of the right atrium and ventricle. The increased pulmonary blood flow can lead to hypertensive pulmonary vascular disease; however, this effect is generally not seen until later in life. Infants and younger children with ASDs are generally asymptomatic and diagnosed secondary to auscultation. Older children are also often asymptomatic, but may complain of fatigue and dyspnea. Again, the ASD is usually diagnosed on the basis of an ejection murmur on exam as well as a wide and fixed splitting of the second heart sound or after an abnormal chest x-ray. A large ASD can cause cardiomegaly, which can be seen on chest x-ray, in addition to right atrial prominence and increased pulmonary vascular markings. Typical ECG findings include an RSR' pattern in the right precordial leads, right axis deviation, and tall P waves in lead II corresponding to right ventricular volume overload and right atrial enlargement. With small ASDs, the ECG is usually normal.

Management

Small ASDs that are hemodynamically insignificant may not require closure. Larger defects are electively closed in asymptomatic patients to prevent long-term complications. Secundum (a portion of the septum) ASDs can potentially be closed with percutaneous devices in the cardiac catheterization laboratory, or may require surgical closure if a percutaneous device cannot be safely placed. Sinus venosus ASDs require surgical closure secondary to their position in the right atrium as well as their association with anomalous pulmonary veins. This was the case for JT. An autologous pericardial patch can be used to close a sinus venosus ASD without anomalous pulmonary venous return. However, in most cases, anomalous pulmonary veins are associated with the defect, and then surgical closure must redirect the veins into the left atrium as well as close the ASD.

PEDIATRIC EXERCISE STRESS TESTING

Clinical pediatric exercise testing has many applications in children with and without heart disease. Comprehensive reviews have been published for pediatric exercise testing (10) as well as exercise testing and training specifically in CHD (13). While there are many similarities between pediatric and adult graded exercise testing, important differences do exist in regards to age, test indication and termination, equipment, and physiological responses. Pediatric exercise testing is most common in the teenage years. However, patients as young as 6 years old can be tested when an appropriate test indication exists.

Indications for testing are dependent upon the unique diagnoses of the patient and include, but are not limited to, evaluations for ECG abnormalities (e.g., loss of pre-excitation in Wolff-Parkinson-White or QTc elongation), assessment of functional capacity in structurally abnormal hearts (e.g., single ventricle or tetralogy of Fallot), and evaluation of exercise-induced bronchospasm. Because exercise tests are effort-dependent, criteria such as an RER exceeding 1.1 or a peak heart rate greater than 200 beats·min^{-1} are often cited as measures of good volitional effort (5). However, for reasons not entirely clear, children's peak RER values tend to be lower than adults (13). Therefore an RER >1.05 may be a more acceptable indicator of a true maximal effort in those under 19 yr (13).

Children are routinely tested with equipment analogous to what would be seen in an adult lab. An electrocardiogram, pulse oximetry, blood pressure, and metabolic parameters must be properly fitted to each patient for tests conducted on either a treadmill or cycle ergometer. Neurological, neurodevelopmental, orthopedic, and other comorbidities are often associated with CHD and must be considered when selecting test mode. With walking the gait should be carefully monitored as ambulating on a moving surface or maintaining pedaling cadence can be challenging for a child or patient with comorbidities, particularly in the setting of a diagnostic test with unfamiliar equipment.

Compared with adults, ongoing physical maturation alters the physiological responses to acute exercise in children. These changes are summarized in Table 1 (1). Of note, while absolute $\dot{V}O_2$ values in children are lower, relative $\dot{V}O_2$ values are higher. It has been reported that scaling $\dot{V}O_2$ to total body mass is an inferior method for quantifying growth-related changes in oxygen consumption. Total lean mass or total lower leg lean mass may be superior methods for normalizing $\dot{V}O_2$ values in children. However, the application of assessment of lean mass presents a variety of practical challenges for many clinical sites (7,13). Analyzing changes in the percent of predicted peak $\dot{V}O_2$ is an alternative method for evaluating $\dot{V}O_2$ changes in children over time as predictive equations by Cooper and Weiler-Ravell are generated from height and gender, not weight; albeit the predictive equations are less reliable with children who are <130 cm in height (3,12,13).

PEDIATRIC CARDIAC REHABILITATION

In a systematic review of pediatric CR programs, Tikkanen analyzed 18 programs with structured exercise training (14). The safety and efficacy of exercise in children with CHD was supported in this review. Most studies reported acute benefits of CR with one study showing the benefits were sustained 1 yr post CR (14). Rhodes and colleagues reported in a cohort of children aged 8 to 18 yr with serious CHD that peak $\dot{V}O_2$, percent predicted peak $\dot{V}O_2$ (12), and AT each significantly improved following participation in CR.

Current exercise recommendations exist for healthy children and children with CHD who are competing in sports (1,8,9). Tikkanen recommends the structure of pediatric CR programs should include aerobic, strength, and flexibility training 2 to 3 $d \cdot wk^{-1}$ for 12 wk with a minimum duration of 40 min of moderate intensity exercise set at an individual's AT (14). CHD patients often exhibit chronotropic incompetence or are prescribed a beta blocker, thereby limiting the usefulness of peak heart rate predictive methods (e.g., 220-age) in the determination of an exercise training intensity range (6,14). The chosen mode for aerobic exercise can be any exercise that is dynamic, rhythmic, and utilizes large muscle groups. The exercise mode may be selected based on available equipment, patient preference, and patient's comorbidities. Incorporation of games, sport balls, music, and age-appropriate incentives add enthusiasm and motivation to the exercise session (12).

Although a specific exercise prescription for resistance training in the pediatric CHD has not been established, the importance of resistance training is evidenced by the strong correlation of muscle strength and exercise tolerance as well as the finding that adults with only a single ventricle have sarcopenia (4,14). It is reasonable to apply American College of Sports Medicine resistance training guidelines used in adult CR with the pediatric CHD population (1).

TABLE 1. Comparison of cardiopulmonary measures between healthy adults and children (1).

	Greater in Adults	Greater in Children
Peak oxygen uptake (absolute) $L \cdot min^{-1}$	X	
Peak oxygen uptake (relative) $mL \cdot kg^{-1} \cdot min^{-1}$		X
Peak heart rate $beats \cdot min^{-1}$		X
Peak cardiac output $L \cdot min^{-1}$	X	
Stroke volume $L \cdot beat^{-1}$	X	
Systolic blood pressure mm Hg	X	
Diastolic blood pressure mm Hg	X	
Respiratory rate $breaths \cdot min^{-1}$		X
Tidal volume $L \cdot breath^{-1}$	X	
Minute ventilation $L \cdot min^{-1}$	X	
Respiratory exchange ratio	X	

SUMMARY

While adult CR programs are numerous and widely studied, pediatric CR programs are largely underutilized. Cardiac rehabilitation in the pediatric population is often overlooked as a first-line approach to secondary intervention (14). Additionally, in a population where exercise capacity is reduced overprotection by physicians, parents, teachers, coaches, or the children themselves can lead to further deconditioning (8,13,14). Such barriers warrant further exploration.

The clinical management of pediatric patients with CHD has made tremendous advancements. The use of CR in this population has the potential to provide invaluable adjuvant treatment and, as in the case of JT, can provide a method to help achieve excellent functional capacity, patient confidence, and clinical outcomes.

Keywords: atrial septal defect, cardiac rehabilitation, pediatric

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