



Congenital heart disease and exercise

John M. Dent, MD

*Adult Echocardiography and Exercise Stress Laboratories, University of Virginia Health System,
Box 800662, Charlottesville, VA 22908, USA*

Successful treatment of patients with congenital heart disease (CHD), combined with population growth, has increased the likelihood that physicians will encounter these patients during the course of daily practice. Because of the known health and psychologic benefits of exercise, determining the safety of exercise and prescribing a rational exercise program are important in the evaluation and management of CHD patients. There are a number of challenges involved in this task. First, there is very little prospective data available about the safety of vigorous exercise in patients with CHD; whereas we know from autopsy studies that some forms of CHD can precipitate sudden death during exercise, particularly when undetected, applying this information to the patient in your office with known disease can be difficult. The protean manifestations of CHD are also a challenge for the noncardiologist, who may be confused by ambiguous terminology and the absence of complete medical records. Surgical treatment for CHD modifies both anatomy and the hemodynamic impact of CHD, requiring an ongoing assessment of changes in hemodynamics and recognition of emerging complications as these patients reach adulthood. Finally, the well-publicized deaths of athletes attributed to CHD and the pressures of competitive athletics can complicate the physician's task by adding social and even political pressures to the care of these special patients.

This article will address these issues by providing a practical approach to assessing the safety of exercise in patients with known or suspected CHD.

General approach

The evaluation of patients with CHD begins with the retrieval of medical records, particularly surgical reports and diagnostic testing. Nowhere in practice

E-mail address: jmd5k@hscmail.mcc.virginia.edu

is it more important to have these records, because the patients' anatomic and hemodynamic status will determine their ability to exercise, and it can be very difficult to identify cardiac structure and function appropriately in a patient with one or more corrective surgeries. Using these records and data obtained from a thorough history and physical examination, one can usually assess the patient's general condition and functional status, but more accurate assessment may require diagnostic testing.

Echocardiography and MR imaging can provide extensive information about both anatomy and function. Because of the accuracy of these noninvasive tests, cardiac catheterization is used less commonly today than in the past. Information available from echocardiography includes general anatomy, chamber sizes and ventricular function, valvular function, and presence of shunt flows. A useful measurement obtainable for most patients is the estimation of pulmonary artery pressure from Doppler of the tricuspid regurgitation (TR) signal (Fig. 1). Using the modified Bernoulli equation and an estimate of right atrial pressure, the peak velocity of the TR jet can be converted to a highly accurate estimate of peak pulmonary artery pressure [1]. Because decisions about exercise prescription for patients with CHD often depend on the presence or absence of pulmonary hypertension, this single measurement may be the most important information obtained from an echocardiogram.

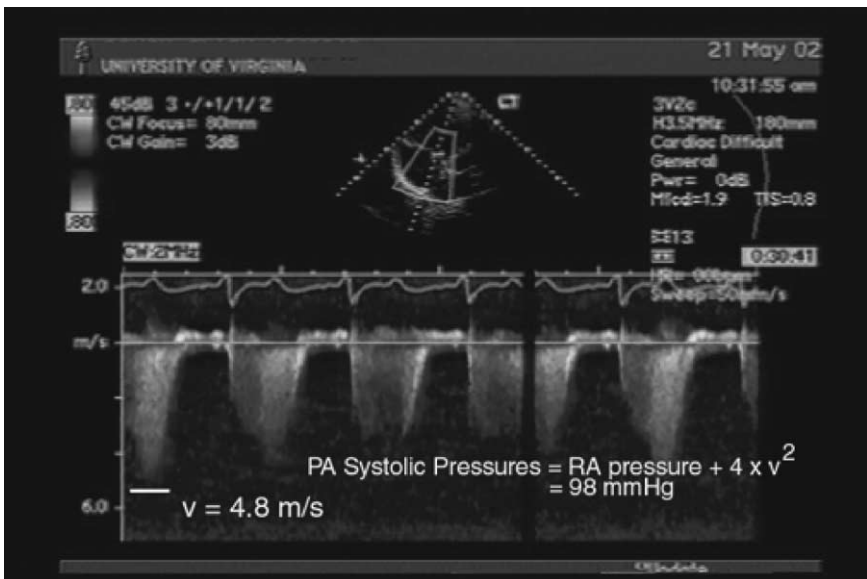


Fig. 1. Doppler echocardiogram of a patient with pulmonary hypertension. Using the tricuspid regurgitation jet velocity and the modified Bernoulli equation, estimated pulmonary artery peak pressure is 98 mm Hg. This patient with CHD would not be eligible for competitive sports.

Though guidelines for exercise participation often depend on whether the patient is symptomatic or not, it can be quite difficult to be certain about the presence or lack of symptoms, particularly in sedentary patients who wish to start an exercise regimen. The sedentary patient may report dyspnea on exertion, which may be caused by deconditioning or underlying cardiac decompensation. In selected cases, exercise testing can provide data about exercise tolerance and, when combined with analysis of expired gases, can give objective information about the patient's functional status [2–6].

Quite often, patients are referred to physicians after another health care provider detects a heart murmur, which raises a question of possible CHD and requires evaluation prior to clearance for sports. Because echocardiography is expensive, it should not be used routinely for all patients with heart murmurs. A thorough exam including auscultation with provocative maneuvers will allow you to select the subset with murmurs that may indicate underlying heart disease and who do require echocardiography [7].

Classification of CHD

The nonspecialist faces the challenge of understanding CHD terminology. The classification of patients with known CHD begins with the determination of whether the patient is cyanotic or acyanotic, which can often be determined from physical examination and oximetry. Within each of these categories, lesions can be divided based on anatomic location, as well as the presence or absence of shunt.

To understand more complex anatomy such as tetralogy of Fallot or transpositions of chambers and vessels, one tries to follow the flow of blood from the systemic circulation as it enters the heart; by following the vena cavae with echocardiography, one can determine which chamber accepts this blood, then follow it through the ventricle and out the large vessel attached to this ventricle. There are a number of clues that are used to determine which chamber attaches to which; for example, the right ventricle can be identified by a prominent band of muscle called the moderator band. The same process is used on the left side of the heart, beginning with the identification of the pulmonary veins and the chamber into which they empty. Because some forms of CHD closely resemble others, with minor variations, two specialists may label a patient's anatomy differently.

Surgery for CHD may be either corrective or palliative. An example of corrective surgery is the anatomic switch procedure for transposition of the aorta and pulmonary artery, where the surgeon attaches the anatomically correct arteries to their functional ventricles, and a normal or near-normal life expectancy can be achieved. Palliative surgery occurs when complete correction is impossible; for example, if the patient has only a single ventricle, the surgeon may constrict the pulmonary artery with a band to reduce pulmonary blood flow and optimize the balance of flow between the pulmonary and systemic circulations. This patient will be highly likely to develop later complications that would jeopardize sports participation.

Classification of sport activities

Assessing the potential impact of competitive athletics on a patient with CHD requires knowledge of the patient's particular condition and an understanding of the demands of the proposed physical activity. Exercise is typically classified by the type of exercise, either predominantly dynamic or predominantly static, because the cardiac impact differs significantly. Dynamic exercise such as running requires changes in muscle length and joint movement with low intramuscular force, whereas static exercise such as weight lifting imposes a different burden of large intramuscular force and less change in muscle length and joint movement [8]. Dynamic exercise imposes a volume load on the ventricles, whereas static exercise produces a pressure load on the heart and arterial circulation.

Echocardiography of trained athletes can reveal evidence of these loads on the left ventricle [9,10]. Athletes who participate in predominantly dynamic exercise training develop left ventricular (LV) diastolic enlargement and increased wall mass, but the wall mass may not be obvious on echocardiography because it is distributed over a larger ventricle. On the other hand, athletes who perform static training can develop LV hypertrophy (LVH) that may be difficult to distinguish from that associated with either hypertension or some forms of hypertrophic cardiomyopathy.

Most types of athletic activity can be classified easily by the degree of dynamic and static exercise. For example, bowling is a low-static/low-dynamic activity, whereas, at the other end of the spectrum, decathlon is a high-static/high-dynamic activity. Physicians can use this classification combined with knowledge of the patient's anatomic abnormality and physiology to decide whether a particular sport should be attempted.

A second consideration in athletics is the potential for collisions during sports. Some patients with CHD may be cleared for sports as long as the potential for collisions is low. For example, patients with Marfan syndrome who lack a family history of sudden death and have no evidence of aortic root dilation can participate in low and moderate sports activity, but they are at increased risk of injury with collisions. Thus, archery and fencing would be acceptable, but football would not be allowed. Similarly, patients recovering from cardiac surgery should not engage in sports with the potential for collision until their incisions have healed, sternal stability is reestablished, and postoperative complications such as pericarditis and atrial arrhythmias have resolved.

Sudden cardiac death in CHD

Patients with CHD can be divided into two distinct groups: those with known characterized syndromes, and those in the population with unrecognized CHD who are at risk for sudden cardiac death during exercise. Though it is impossible

to measure the risk of competitive athletic training for patients with CHD, the majority of patients who die suddenly do so during intense physical activity—either during training or competition. In younger patients, hypertrophic cardiomyopathy (HCM) is the most common finding at autopsy, accounting for approximately one third of deaths. Coronary artery anomalies, typically a single coronary artery or unusual origin of these vessels, account for an additional 20% of deaths. The only other form of CHD with a significant association is congenital aortic stenosis, which is associated with approximately 4% of sudden deaths in younger patients [11,12].

Screening for undetected CHD in order to prevent sudden cardiac death is expensive and difficult. First, though the sudden deaths of athletes appear frequently in the news media, the actual incidence of sudden death during athletics is extraordinarily low—less than 1 death per 100,000 athletes [11]. Second, routine screening with a noninvasive test such as echocardiography would be prohibitively expensive, and would probably identify only the patients with aortic stenosis and HCM. As is well established for screening tests in a low-pretest probability group, the positive predictive value of an abnormal finding on echo would be very low, and many athletes would likely be disqualified based on findings of unknown significance. For example, it can be difficult to differentiate some forms of HCM from normal, compensatory hypertrophy seen in athletes. For these reasons, the current standard of care for athletes who wish to perform in competitive athletics is simply a thorough family history and physical examination by a licensed physician. The widespread use of routine screening tests will need to be validated prospectively from a cost-effectiveness perspective before this can be recommended.

Guidelines

The paradox of exercise-induced sudden cardiac death is that the only way to prevent it is to prohibit intense physical activity, but, if physicians were to prohibit such activity in all patients with CHD, they would deny the great majority of patients the well-known health benefits of exercise in order to prevent the very rare occurrence of sudden death. Though the rarity of both CHD and sudden death has resulted in a dearth of prospective studies, it is possible to formulate a rational exercise prescription for most patients. The most recent guidelines for exercise participation were published in a “Bethesda Conference Report,” a comprehensive document that should be reviewed by any physicians caring for patients with CHD [13].

The importance of basing medical decision making on these guidelines was demonstrated in the high-profile case of a college athlete who sued Northwestern University after being denied participation on the basketball team. The case was ultimately decided in favor of Northwestern, and the court decision emphasized the utility of expert guidelines because medical opinion may vary widely in individual cases. The precedent set by this decision reinforced the

critical role of the sports physician in deciding about eligibility for individual athletes with CHD [14].

Specific conditions

Shunts

Most shunt lesions are initially acyanotic because there is a predominance of left to right flow, which means that oxygenated blood returns through the pulmonary circulation. Typical shunts include atrial septal defects (ASD) (Fig. 2), ventricular septal defects (VSD), and a patent ductus arteriosus (PDA). The presence of symptoms is usually determined by the amount of shunt because an increasing shunt reduces the mechanical efficiency of the heart. Children with large shunts usually present in childhood with congestive heart failure, but, if the shunt is small, they may remain asymptomatic throughout life. Patients with VSDs are usually diagnosed in childhood because of the intensity of the heart murmur produced by the left ventricle as it propels blood into the lower-pressure right ventricle, and, similarly, the loud continuous murmur of a PDA is apparent on even the most cursory of auscultatory exams. Because the physical findings in patients with ASDs may be subtle, including fixed splitting of the second heart

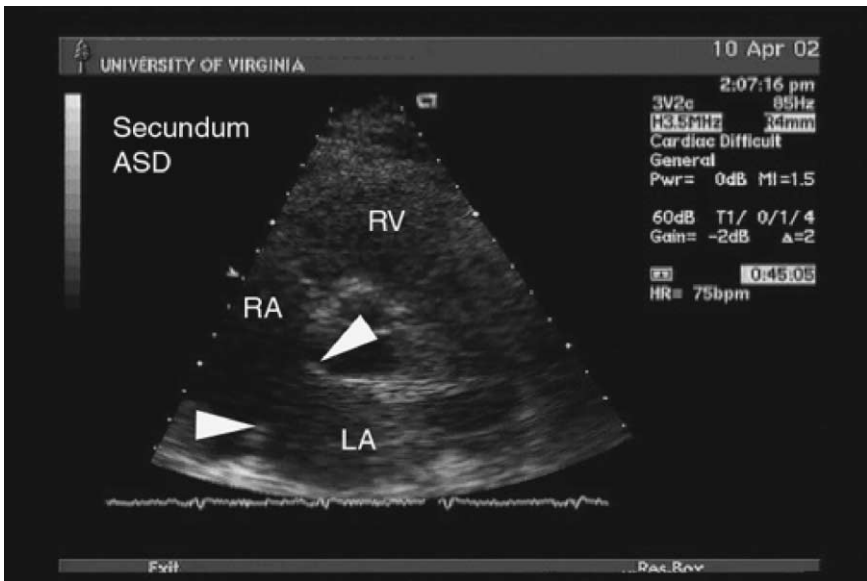


Fig. 2. Two-dimensional parasternal echocardiogram of a patient with a large secundum ASD. The image shows a large defect in the interatrial septum, along with an enlarged right ventricle. Color Doppler imaging can also be used to demonstrate the direction of flow across the ASD. LA, left atrium; RA, right atrium.

sound and a pulmonary flow murmur, patients with small to moderate ASDs may escape detection until later in life, when the atrial shunt increases because of a subtle increase in left atrial pressure associated with the aging process.

The most common symptoms in patients with left-to-right shunts are dyspnea and easy fatigue. Patients with ASDs with normal pulmonary artery pressure can participate in all competitive sports, although it is of course possible that their exertion will be limited by fatigue from the shunt. Patients with small to moderate VSDs and normal pulmonary artery pressure may participate in all sports as well, but patients with large VSDs should be limited to low-level sports until they undergo repair of the VSD [15].

Patients with ASDs and VSDs can develop cyanotic shunts if there is an increase in pulmonary artery resistance in response to increased flow. Normally, the pulmonary circulation is a low-resistance circuit, even in the presence of markedly increased blood flow. For largely unknown reasons, some patients do not tolerate chronic high pulmonary flow states, and there is a progressive increase in pulmonary vascular resistance, with obliteration of capillaries. When pulmonary artery pressure approaches systemic blood pressure, the shunt direction can reverse, leading to right-to-left shunting and cyanosis, which is called Eisenmenger syndrome. Patients who have progressed to Eisenmenger physiology are unlikely to attempt significant exercise because of limiting symptoms, and they should not participate in competitive sports [15,16].

It is important to understand the significance of pulmonary hypertension in patients with CHD because many of the guidelines for exercise participation differentiate patients according to this measurement. Pulmonary artery (PA) pressure may increase because of increased pulmonary vascular resistance from obliteration of the capillary bed, as occurs in Eisenmenger syndrome. PA pressure will also rise, however, whenever there is elevated left atrial pressure, even in the absence of high pulmonary vascular resistance, because such an increase is mandated to achieve the forward flow of blood into the left atrium and ventricle. This type of compensatory pulmonary hypertension is often reversible if the underlying lesion can be corrected, and left atrial pressure returns closer to normal, whereas the fixed pulmonary hypertension of Eisenmenger syndrome portends a poor prognosis. Finally, it is critical to recognize that pulmonary artery pressure is the product of flow (cardiac output) and resistance. If cardiac output falls, for example because of right ventricular (RV) dysfunction, then pulmonary artery pressure will fall even in the presence of high pulmonary resistance (Fig. 3). For this reason, it is important to understand that low pulmonary artery pressure may camouflage the presence of severely increased pulmonary vascular resistance in a patient with RV failure, and such a patient may be more compromised than a patient with a much higher PA pressure.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is a common genetic disorder, occurring in approximately 1 in 500 of the population, which is characterized by thickened,

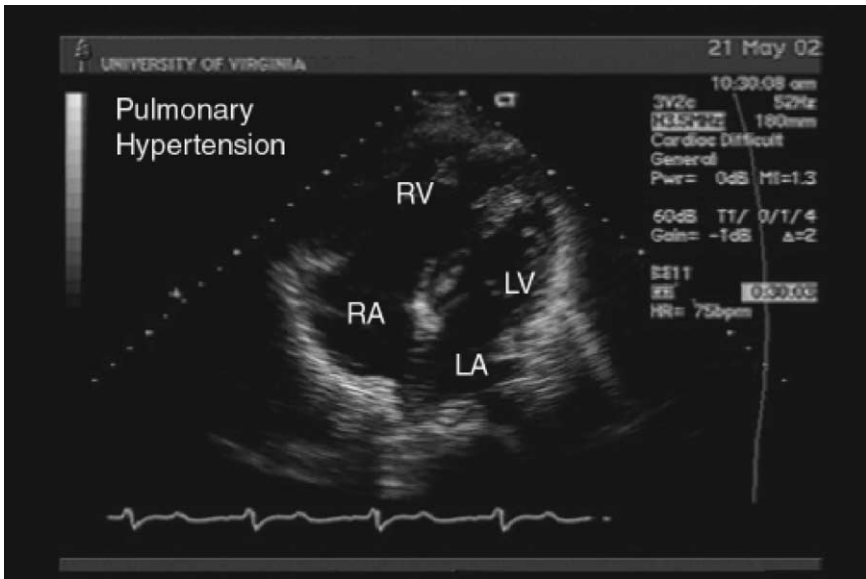


Fig. 3. Two-dimensional apical echocardiogram of a patient with severe pulmonary hypertension and RV enlargement from an ASD. The heart is displayed upside-down by convention, and the massively enlarged RV dwarfs the smaller left ventricle (LV). This patient's RV dysfunction could lead to a reduction in pulmonary artery pressure caused by reduced cardiac output, despite severely elevated pulmonary vascular resistance.

abnormal myocardium. Rather than a single disease, it should be considered a family of disorders with variable manifestations. The most common syndrome is characterized by excessive thickness of the upper ventricular septum, which narrows the LV outflow tract because of abnormal systolic anterior motion of the mitral valve (SAM), and produces a dynamic outflow obstruction [17–19]. Other patients may have localized thickening of the LV apex or diffusely increased wall thickness. Intraventricular pressure gradients may be absent at rest and only appear during exercise or when LV filling is reduced.

The thickened myocardium of HCM requires increased filling pressures in order to maintain cardiac output, so left atrial pressure rises, and dyspnea is the most common symptom. Patients with obstruction caused by SAM also develop mitral regurgitation, with a consequent increase in left atrial pressure. Patients may develop arrhythmias including atrial fibrillation and ventricular ectopy, which may lead to syncope or sudden death. Though sudden death tends to occur in families with the most lethal mutations, which can be predicted from a thorough family history, it is impossible to predict which patients are at lesser risk of this complication until they reach later life. Complicating the screening for HCM is the fact that children with the disease may have normal myocardial thickness until they approach puberty, when myocardial thickness can increase dramatically. Population-based studies suggest that there is a substantial pool of

undiagnosed patients in the community, whose disease would not be predicted based on familial inheritance [19].

After family history, the most useful information is obtained from echocardiography, which demonstrates thickened myocardium (Fig. 4A,B), and Doppler can show the characteristic dynamic “saber-shaped” pattern of LV outflow obstruction (Fig. 4C). Though the extent of obstruction and the resulting LV pressure gradient tend to correlate with symptoms, some patients without obstruction may be severely symptomatic.

Treatment of HCM remains controversial. The standard treatment consists of high-dose beta blockers and verapamil, which suppress LV contractility and lower the pressure gradient, along with diuretics to lower left atrial pressure and reduce dyspnea. Patients treated with these medications may experience exercise limitation because of a reduced cardioacceleratory response. They may also suffer from orthostatic dizziness caused by the combined effects of the outflow tract obstruction and the pharmacologic blockade.

More recently, dual chamber pacing has been used in patients with HCM to reduce the outflow gradient by reducing the force of systole through asynchro-

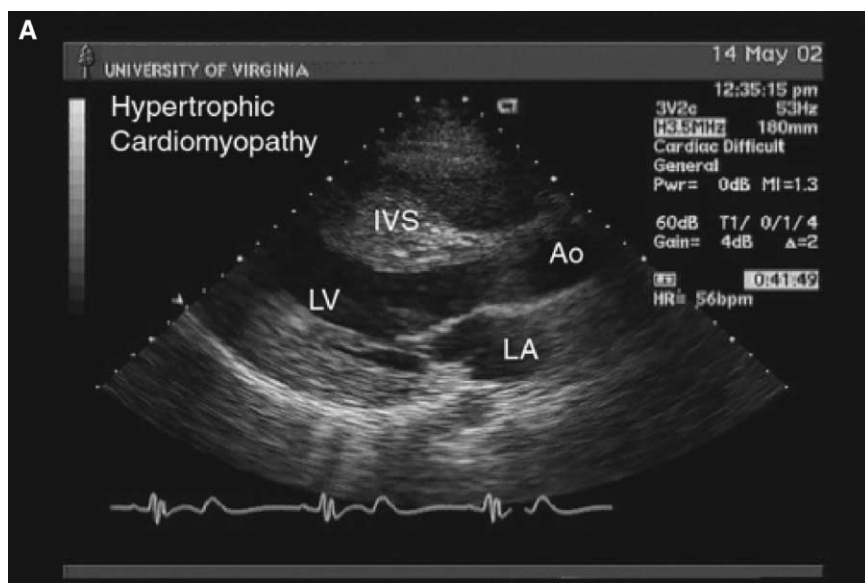


Fig. 4. (A) Two-dimensional parasternal echocardiogram of a patient with hypertrophic cardiomyopathy. Note the markedly thickened interventricular septum (IVS), which measured over 2 cm (normal septal thickness is 0.5–1.0 cm). (B) Two-dimensional apical echocardiogram of the same patient with HCM. The entire left ventricle is thickened, with a small cavity; in some patients; the thickening is most pronounced in the septum, whereas in others, the wall thickening is diffuse. (C) Doppler echocardiogram in a patient with HCM and dynamic outflow tract obstruction; the characteristic saber-shaped signal is indicative of a gradient that increases during systole. Using the Bernoulli equation, this patient’s outflow gradient was measured at a peak of 64 mm Hg, indicative of severe obstruction. This patient would be ineligible for competitive sports.

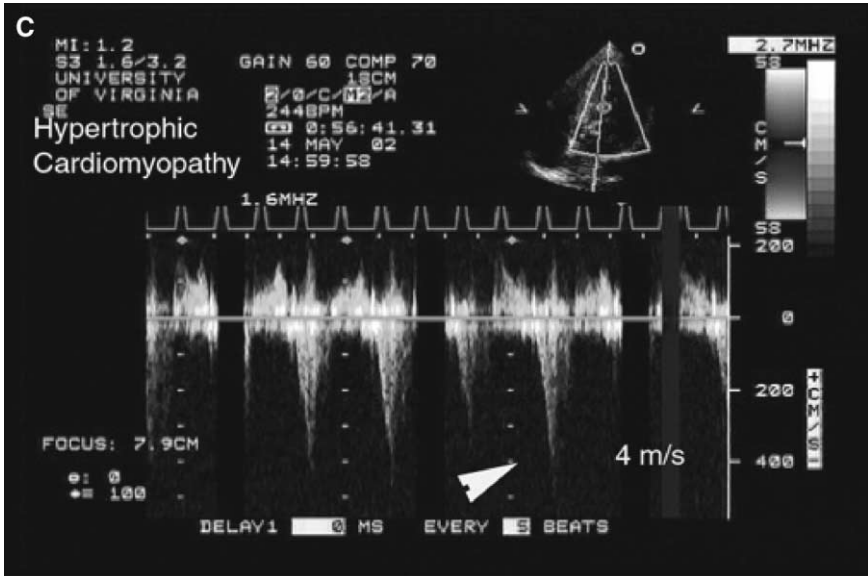
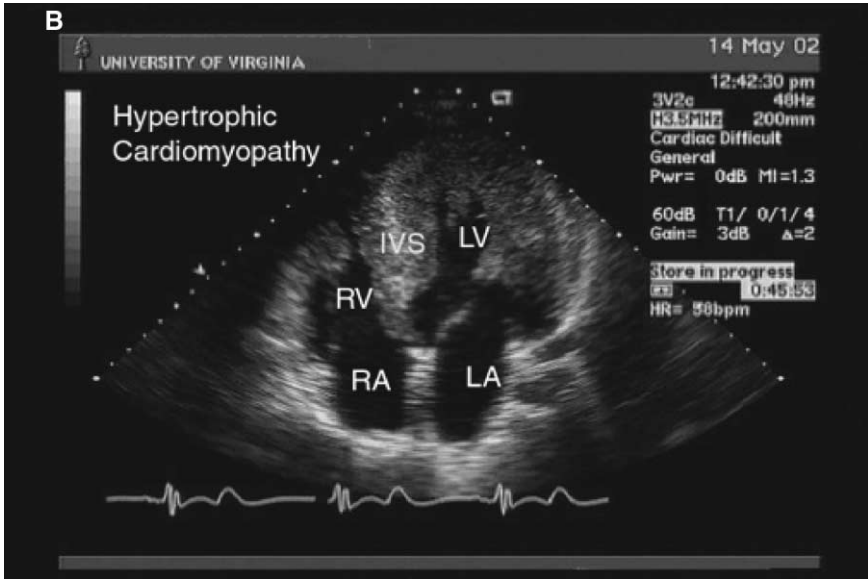


Fig. 4 (continued).

nous activation of the left ventricle; this approach remains controversial. One limitation of the pacemaker treatment is that cardiac output may drop at the higher heart rates encountered during exercise. Surgical removal of a portion of the upper septum, called septal myectomy, can significantly reduce the pressure gradient as well, but excellent results critically depend on patient selection and

surgical experience. An intriguing nonsurgical technique is percutaneous septal ablation in which alcohol injected into a selectively catheterized septal branch of the left anterior descending coronary artery produces a controlled septal infarction, with a resulting reduction in the pressure gradient. All of these mechanical techniques carry the potential for complications, and long-term follow-up data are unavailable [19].

The use of prophylactic internal cardiac defibrillators (ICDs) may extend the life of patients with HCM at high risk of sudden death, but implanting them in all patients with the disease would be costly because of both the expense and morbidity of the devices. If a patient with an ICD wishes to exercise, it is mandatory to ensure that sinus tachycardia does not trigger an unneeded shock, so the target heart rate of exercise must be below the rate threshold of the ICD. This is most often required in younger patients, where beta blockers can be used to reduce the cardioacceleratory response to exercise. Additionally, patients should be informed of the possibility of lead fractures, which can lead to either device failure or inappropriate shocks. An athlete with a malfunctioning ICD may present a risk of physical injury to other athletes, so some forms of competition may be inappropriate. For these reasons, patients with ICDs placed for aborted sudden death should not participate in high-level competitive sports.

Current guidelines specify that patients with unequivocal HCM should not be allowed to participate in competitive athletics because sudden death may be precipitated [17]. It is unclear whether placement of an ICD should modify this rule. Athletes older than 30 may have a lower risk of sudden death, assuming that many patients with the most lethal variants experience death at a younger age. For these patients, it may be possible to select an asymptomatic, lower-risk subset who lack a family history of sudden death, do not demonstrate ventricular arrhythmias on ambulatory monitoring, and have relatively moderate findings on echocardiography and myocardial perfusion imaging. These patients may be allowed to exercise at moderate intensity as long as they understand the limitations of our risk prediction ability.

It is important to recognize that not all LVH is a sign of disease because the ventricle can hypertrophy in response to vigorous training [10]. Characterized by symmetric LVH, extremes of the so-called “athletic heart” can be difficult to differentiate from mild forms of HCM. Additionally, patients with some forms of HCM do not develop either resting or provokable outflow tract obstruction, so the absence of such a pressure gradient cannot be used to exclude the diagnosis of HCM.

Prohibiting competitive athletics for a promising athlete because of HCM may not just remove the pleasure of competition but may also jeopardize future income from professional sports. Often, the physician-patient relationship is therefore threatened by the intrusion of third parties, including parents and coaches [14]. The physician may be challenged further by the lack of a definitive diagnosis in borderline cases because LVH can be a part of the normal response to exercise. Finally, newer treatments for HCM, including use of implantable cardiac defibrillators to prevent sudden death, may allow previously disqualified

athletes to participate with the understanding that it is impossible to remove the risk of death completely from competitive athletics. The final decision to allow or prohibit sports for the patient with HCM will therefore always be variable and individualized.

Bicuspid aortic valve

Bicuspid aortic valves are the most common congenital cardiac anomaly, occurring in about 1% of the population. These valves are susceptible to the development of either stenosis or regurgitation, which can occur at any point of an individual's life. Though aortic regurgitation may compromise athletic performance, aortic stenosis also carries the potential to precipitate sudden death during athletics and accounts for about 4% of deaths during training or competition [11,12]. In addition, because the aorta is abnormal in patients with bicuspid valves, the incidence of aortic dissection is increased; it is possible that exercise might precipitate dissection caused by increased shear forces.

Detecting significant aortic stenosis relies on the auscultatory skill of the examining physician. The presence of the characteristic "crescendo-decrescendo" murmur combined with a frequent apical aortic click can suggest the presence of a bicuspid valve that echocardiography can confirm through typical two-dimensional (Fig. 5A) and Doppler (Fig. 5B) findings. Though cardiac catheterization previously played a major role in measuring the abnormal pressure gradient, the accuracy of high-quality echocardiography has diminished the utility of the test for these patients. Echocardiography can be used to estimate the severity of disease, based either on the resting pressure gradient (higher means more severe stenosis), or on reduction in aortic valve area.

Asymptomatic patients with mild aortic stenosis can be allowed to participate in any form of competitive athletics. When the disease progresses to moderate obstruction, then asymptomatic athletes may still be allowed to participate in low- to moderate- static/dynamic exercise (examples include baseball, archery, and golf) as long as echocardiography does not demonstrate more than mild LVH, and they can complete a normal graded exercise treadmill test. Patients with severe aortic stenosis or moderate stenosis and symptoms should be prohibited from competitive athletics [15]. This final recommendation imposes a burden on the patient with asymptomatic aortic stenosis who is not yet a surgical candidate but who must sometimes discontinue activities such as running and tennis. These patients may be allowed to maintain fitness through walking, which will help the physician to detect onset of symptoms and the need for surgery.

Treatment options for symptomatic patients with congenital aortic stenosis include balloon or surgical valvuloplasty, performed in younger patients in order to postpone the need for prosthetic valve replacement, and valve replacement. For younger patients who wish to avoid anticoagulation, the aortic valve can be replaced with a homograft (preserved cadaveric aortic valve), with a porcine prosthesis, or in an innovative procedure, with the patient's own pulmonary valve in the Ross procedure. Each of these treatments will allow participation in

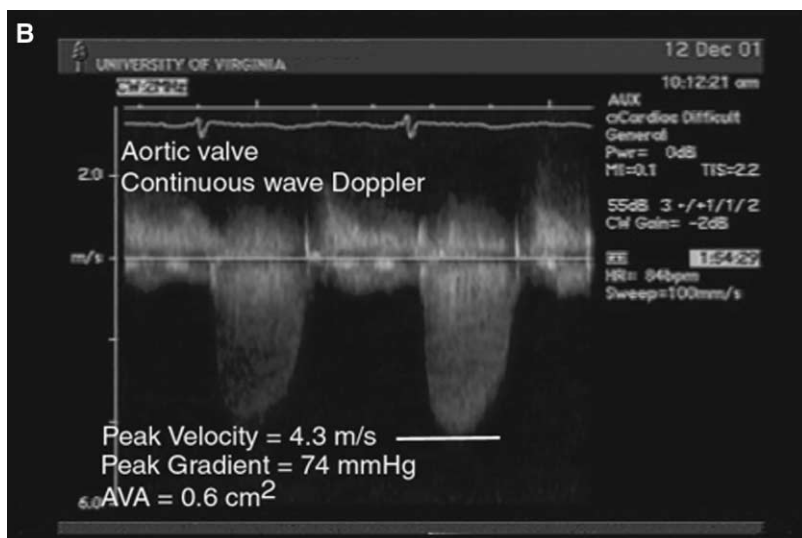
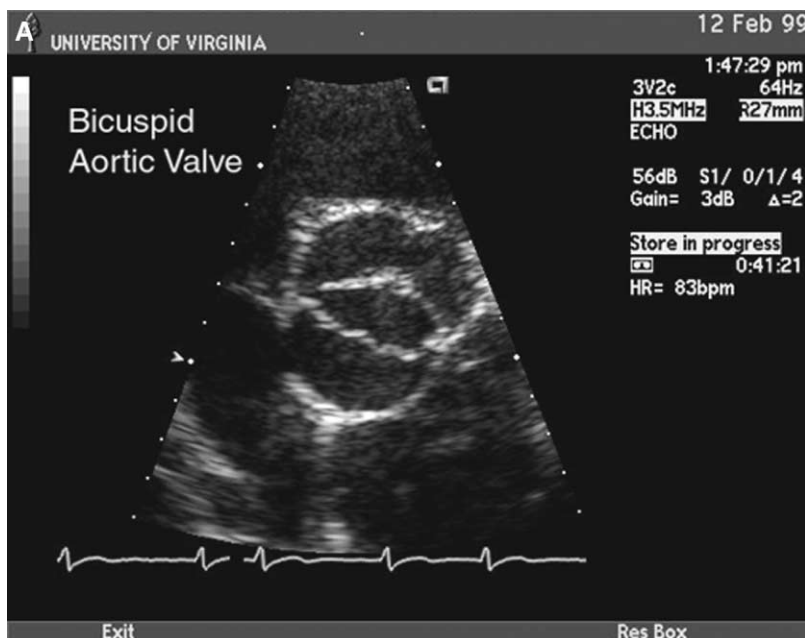


Fig. 5. (A) Two-dimensional echocardiogram of a bicuspid aortic valve, demonstrating fusion of the left and right coronary cusps. This patient has adequate valve opening to exclude significant stenosis, and high level exercise would be permissible. (B) Doppler echocardiogram of a patient with severe aortic stenosis; the peak transvalvular gradient was 74 mm Hg. Using the continuity principle, the patient's estimated aortic valve area is 0.6 cm². This patient would not be allowed to participate in high-level sports whether symptomatic or asymptomatic.

competitive athletics that involves the potential for physical impact during exercise, although the sacrifice may include the need for future reoperation because of valve failure.

Coronary artery anomalies

Coronary artery anomalies are uncommon findings in the general population but may account for up to 20% of deaths during sports [11,12]. Unfortunately, the diagnosis is rarely made prior to death in younger patients, where sudden death may be the first symptom. Anomalous origin of the left main coronary artery from the pulmonary artery can lead to congestive heart failure or sudden death in childhood or adulthood; the diagnosis is suspected in cases of LV dysfunction. Occasionally, these anomalies are detected in older patients referred for cardiac catheterization, and a decision must be made as to need for treatment and permissibility of exercise. Anomalous origin of the left coronary artery has been associated with sudden death, particularly when it passes between the aorta and pulmonary artery. Patients with these anomalies should not engage in strenuous exercise unless the condition is corrected surgically and adequate myocardial perfusion is assessed with stress testing.

Tetralogy of Fallot

The tetralogy of Fallot consists of VSD, RV hypertrophy, an abnormal and over-riding aorta, and pulmonic stenosis. Originally, palliative surgery was performed with a systemic-to-pulmonary shunt to increase pulmonary blood flow, called the Blalock-Taussig shunt. Because most patients with tetralogy now undergo surgical correction of these lesions as a child, the majority of patients encountered will be much more functional than the previous generation. In order to reach a decision about sports participation, it is vital to know the extent of the correction and the presence of residual defects because not all patients will enjoy the same surgical results. Patients with successful repairs and near-normal hemodynamics can be cleared for all levels of sports participation. A significant problem in a subset of postoperative patients, however, is the presence of severe pulmonary regurgitation that results from repair of the stenotic valve; in these patients and those with residual pulmonary hypertension or arrhythmias, exertion should be restricted to only low-level activities [15].

Marfan syndrome

The Marfan syndrome is characterized by an inherited defect in the production of fibrillin, a component of connective tissue. Affected individuals typically develop dilation of the ascending aorta and a predisposition to aortic dissection, along with mitral valve prolapse and systemic manifestations. As with HCM, genetic analysis has demonstrated a significant heterogeneity [20], so patients with Marfan syndrome experience a wide variation in the extent of cardiac abnormalities and the risk of dissection. Echocardiography can show the dilated

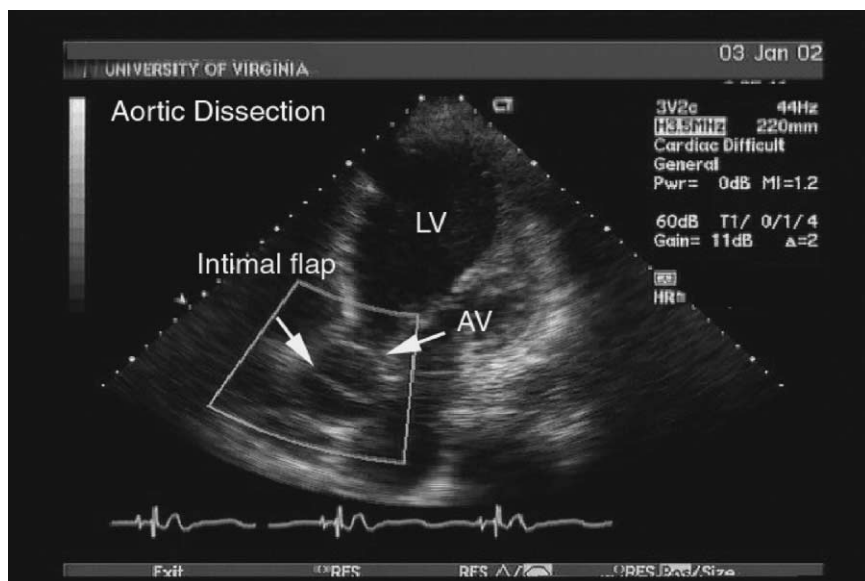


Fig. 6. Two-dimensional echocardiogram of a patient with Marfan syndrome who experienced an acute aortic dissection. The intimal flap can be seen just above the aortic valve in this inverted image. Sudden death can be precipitated by pericardial tamponade, although this patient had no evidence of pericardial effusion.

ascending aorta and, in cases of dissection, can demonstrate the dissection flap (Fig. 6).

The risk of aortic dissection can be partially predicted by measuring the ascending aorta with echocardiography, and by determining the family history of dissection. Because cases may occur sporadically, however, in some patients there is an unknown risk of dissection. Patients with borderline enlargement of the ascending aorta are treated with beta blocking medications to reduce the risk of dissection. Once the aortic root exceeds 5.0 cm, most authorities recommend prophylactic replacement of the ascending aorta. There is controversy about whether the aortic valve and sinuses of Valsalva should be preserved or replaced.

Patients with Marfan syndrome and aortic root enlargement should not participate in competitive athletics unless they first undergo root replacement [15,21]. If the aortic valve is replaced, then avoidance of a mechanical prosthesis with required warfarin anticoagulation is desirable for the young athlete, although later reoperation for valve failure is a consequence of this strategy. Additionally, patients with Marfan syndrome should avoid sports having a potential for collision injuries caused by their underlying connective tissue disorder.

Tricuspid atresia

Patients with tricuspid atresia are born with an incomplete or absent tricuspid valve, typically in combination with an ASD. Because they lack a functioning

right ventricle, surgery is palliative and consists of connecting the right atrium to the right pulmonary artery; the most commonly performed operation is called the Fontan procedure. Because blood flow to the lungs is largely passive, exercise performance in these patients will be reduced, although they may still be highly functional [2–5,15,20,22,23]. Exercise performance will be particularly compromised if they develop atrial arrhythmias or LV dysfunction.

Clearance for patients with the Fontan operation to participate in greater than low-level athletics requires exercise stress testing to determine functional status and echocardiography to measure LV function. Oximetry may be combined with exercise stress testing to obtain further objective evidence of functional status [15].

Ebstein's anomaly

Ebstein's anomaly is an unusual syndrome characterized by apical displacement of the tricuspid valve. This defect effectively reduces the functional size of the right ventricle because a portion of the right atrium is "ventricularized." There is substantial variability in the impact of this lesion, which is determined primarily by the extent of apical displacement. Exercise performance can be further jeopardized by the presence of a secundum ASD, present in up to 85% of patients. Atrial arrhythmias are also common because of the abnormal right atrium and the frequent presence of conductive bypass tracts in the atrium and ventricle.

Echocardiography is very useful in determining the extent of the anomaly. If the tricuspid valve displacement is mild, there is no cyanosis, and there is no history of arrhythmia, then sports participation is unlimited. If the anomaly is severe, sports participation is prohibited. Selected patients can undergo tricuspid valve repair; following successful surgery, they can be reevaluated with echocardiography, exercise stress testing, and ambulatory ECG monitoring. If the results of these tests are favorable, then more vigorous athletics is a possibility [15].

Patent ductus arteriosus

The ductus arteriosus connects the descending aorta to the left pulmonary artery during fetal life and usually closes shortly after birth. In a subset of patients, the ductus remains open, sometimes as an isolated anomaly, and sometimes as a part of a more complex anomaly. Patients with persistently patent ductus experience a left to right shunt as long as pulmonary artery pressure is lower than systemic pressure. The hemodynamic consequences of this lesion are somewhat analogous to mitral regurgitation because blood that leaves the left ventricle returns to the left atrium and ventricle (by way of the pulmonary circulation), requiring ventricular enlargement to accommodate the shunt volume. Echocardiography demonstrates LV and left atrial enlargement and can provide an estimate of the degree of shunt, along with pulmonary artery pressure.

Most patients with a patent ductus arteriosus undergo corrective surgery in childhood. Selective catheter-based ablation of the ductus is also possible, using coils or an occlusion device. Patients undergoing successful correction can

participate in all athletics as long as pulmonary artery pressure (measured by echocardiography) is not significantly elevated [15].

Pulmonary valve stenosis

Pulmonary valve stenosis (PS) leads to reduced cardiac output and easy fatigue as a result. The degree of stenosis is easily quantitated by echocardiography, using the modified Bernoulli equation. Patients with moderate to severe PS are candidates for balloon dilatation of the valve using a catheter-based approach. Following successful balloon valvuloplasty, patients should be followed with echocardiography as the gradient occasionally returns. Patients who undergo successful dilatation can generally be cleared to participate in sports with no limitation [15].

Aortic coarctation

Coarctation of the aorta is a congenital narrowing of the descending thoracic aorta with resultant upper extremity hypertension. The diagnosis is suspected in younger patients with hypertension who have a significant differential in arm and leg blood pressure, and it can be confirmed by either echocardiography or MR imaging. It is important to recognize the common association of this disorder with bicuspid aortic valves as well as aortic dissection. Surgical correction of this disorder consists of enlarging the aorta with either patch material or adjacent arterial tissue. Some patients remain significantly hypertensive even after corrective surgery. Additionally, patients who have undergone some types of surgery or balloon dilatation may be at risk for focal aneurysms at the repair site, so follow-up imaging with either CT or magnetic resonance should be conducted, particularly in patients who underwent procedures more than a decade ago.

Patients undergoing successful surgery who have at most a low residual pressure gradient in the aorta and who have normal blood pressure may participate in competitive athletics after recovering from surgery. These patients should not participate, however, in high-intensity static exercise for 1 year after surgery, and they should avoid powerlifting, which leads to unacceptable aortic wall stress. Patients with suboptimal surgical results or who develop aneurysms and aortic wall thinning can participate only in low-level exercise unless their condition is amenable to reoperation and repair. [15].

Summary

Though initially challenging, the process of determining appropriate levels of exercise for patients with congenital heart disease can be broken down into several practical steps:

List 1: Summary of approach to CHD patients and exercise

- Get the records:
Surgical reports, diagnostic test results, office visits, admissions

Table 1
Examples of specific conditions and exercise prescription

No exercise restriction	Low-intensity competitive sports	No competitive sports
ASD, normal pulmonary artery pressure	ASD, pulmonary hypertension	ASD, Eisenmenger physiology
VSD, small to moderate, asymptomatic	VSD, large	VSD, Eisenmenger physiology
PDA, small		PDA, large, unrepaired
PS, low gradient, asymptomatic	PS, high gradient, asymptomatic	PS, high gradient, symptomatic
AS, mild	AS, moderate, asymptomatic, nl exercise test, no LVH	Severe AS
Tetralogy of Fallot, corrected, near-normal pulmonary artery pressure		Tetralogy of Fallot, uncorrected or residual pulmonary hypertension

AS, aortic stenosis; ASD, atrial septal defect; LVH, left ventricular hypertrophy; PDA, patent ductus arteriosus; PS, pulmonic stenosis; VSD, ventricular septal defect.

- Obtain family history:
Family members with sudden death increase risk.
- Thorough physical exam:
Special attention to auscultation of murmurs
- Appropriate diagnostic testing:
Noninvasive testing is usually adequate.
- Review guidelines:
26th Bethesda Conference, 1994 [13]
- Make recommendation:
Be specific about types of exercise allowed.
- Reassess at least annually:
Patients' status may change over time [24].

The use of this algorithm and review of available guidelines, in combination with selected consultation with other specialists, should allow the sports physician to prescribe exercise for the majority of these patients confidently. Examples of several congenital defects of varying severity, and the appropriate exercise prescription, are listed in Table 1.

References

[1] Yock PG, Popp RL. Noninvasive estimation of right ventricular systolic pressure by Doppler ultrasound in patients with tricuspid regurgitation. *Circulation* 1984;70:657–62.

[2] Miyairi T, Kawauchi M, Takamoto S, Morizuki O, Furuse A. Oxygen utilization and hemodynamic response during exercise in children after Fontan procedure. *Jpn Heart J* 1998;39(5):659–69.

[3] Mocellin R, Gildein P. Velocity of oxygen uptake response at the onset of exercise: A comparison between children after cardiac surgery and healthy boys. *Pediatric Cardiology* 1999; 20(1):17–20; discussion 21.

- [4] Ohuchi H, Arakaki Y, Hiraumi Y, et al. Cardiorespiratory response during exercise in patients with cyanotic congenital heart disease with and without a Fontan operation and in patients with congestive heart failure. *Int J Cardiol* 1998;66(3):241–51.
- [5] Ohuchi H, Katou Y, Arakaki Y, Kamiya T. Alveolar-arterial gas tension differences during progressive exercise in patients after the Fontan operation. *Jpn Circ J* 1997;61(5):402–12.
- [6] Washington RL. Cardiorespiratory testing: anaerobic threshold/respiratory threshold. *Pediatric Cardiology* 1999;20(1):12–5; discussion 16.
- [7] Grewe K, Crawford MH, O'Rourke RA. Differentiation of cardiac murmurs by dynamic auscultation. *Curr Probl Cardiol* 1988;13(10):669–721.
- [8] Mitchell JH, Haskell WL, Raven PB. Classification of sports. *J Am Coll Cardiol* 1994;24: 864–6.
- [9] Douglas PS, O'Toole ML, Hiller WDB, Reichel N. Different effects of prolonged exercise on the right and left ventricles. *J Am Coll Cardiol* 1990;15:64–9.
- [10] Turpeinen AK, Kuikka JT, Vanninen E, et al. Athletic heart: a metabolic, anatomical, and functional study. *Med Sci Sports Exerc* 1996;28(1):33–40.
- [11] Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic and pathologic profiles. *JAMA* 1996;276:199–204.
- [12] Van Camp SP, Bloor CM, Mueller FO, et al. Nontraumatic sports death in high school and college athletes. *Med Sci Sports Exerc* 1995;27:641–7.
- [13] Maron BJ, Mitchell JH, Isner JM, McKenna WJ. 26th Bethesda conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. *J Am Coll Cardiol* 1994;24:845–99.
- [14] Maron BJ, Mitten MJ, Quandt EF, Zipes DP. Competitive athletes with cardiovascular disease—The case of Nicholas Knapp. *N Engl J Med* 1998;339:1632–5.
- [15] Graham Jr TP, Bricker JT, James FW, Strong WB. Task force 1: congenital heart disease. *J Am Coll Cardiol* 1994;24:867–73.
- [16] Sietsema KE. Cyanotic congenital heart disease: dynamics of oxygen uptake and ventilation during exercise. *J Am Coll Cardiol* 1991;18(2):322–3.
- [17] Maron BJ, Isner JM, McKenna WJ. Task Force 3: hypertrophic cardiomyopathy, myocarditis, and other myopericardial diseases and mitral valve prolapse. *J Am Coll Cardiol* 1994;24: 880–5.
- [18] Maron BJ, Moller JH, Seidman CE, Vincent GM, Dietz HC, Moss AJ, et al. Impact of laboratory molecular diagnosis on contemporary diagnostic criteria for genetically transmitted cardiovascular diseases: hypertrophic cardiomyopathy, long-QT syndrome, and Marfan syndrome. A statement for healthcare professionals from the Councils on Clinical Cardiology, Cardiovascular Disease in the Young, and Basic Science. American Heart Association. *Circulation* 1998;98(14): 1460–71.
- [19] Maron BJ. Hypertrophic cardiomyopathy: a systematic review. *JAMA* 2002;287(10):1308–20.
- [20] Nijbroek G, Sood S, McIntosh I, et al. Fifteen novel FBNI mutations causing Marfan syndrome detected by heteroduplex analysis of genomic amplicons. *Am J Hum Genet* 1995; 57:8–21.
- [21] Braverman AC. Exercise and the Marfan syndrome. *Medicine & Science in Sports & Exercise* 1998;30(10):S387–95.
- [22] Driscoll DJ. Exercise responses in functional single ventricle before and after Fontan operation. *Prog Ped Cardiol* 1993;2:44–9.
- [23] Magosso E, Cavalcanti S, Ursino M. Theoretical analysis of rest and exercise hemodynamics in patients with total cavopulmonary connection. *Am J Physiol Heart Circ Physiol* 2002;282(3): H1018–34.
- [24] Maron BJ, Thompson PD, Puffer JC, McGrew CA, Strong WB, Douglas PS, et al. Cardiovascular preparticipation screening of competitive athletes: a statement for health professionals from the sudden death committee (clinical cardiology) and congenital heart defects committee (Cardiovascular disease in the young). American Heart Association *Circulation* 1996;94:850–6.