

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 4: Congenital Heart Disease



A Scientific Statement From the American Heart Association and American College of Cardiology

George F. Van Hare, MD, FACC,
*Chair**

Michael J. Ackerman, MD, PhD,
FACC*

Juli-anne K. Evangelista, DNP,
APRN, CPNP-AC, FACC*

Richard J. Kovacs, MD, FAHA, FACC*
Robert J. Myerburg, MD, FACC*

Keri M. Shafer, MD*

Carole A. Warnes, MD, FACC*

Reginald L. Washington, MD, FAHA*

Congenital heart disease (CHD) is the most common form of serious birth defect, occurring in 8 per 1,000 live births (1). The past several decades have seen dramatic improvements in survival with palliative or corrective heart surgery, such that there are now more adult patients than pediatric patients alive with CHD. Although restriction from competitive athletics may well be indicated for some, the great majority of patients can and should engage in some form of physical activity and should avoid a sedentary lifestyle. Clinicians should encourage their patients

to engage in healthy physical activities, bearing in mind specific features in some patients, such as residual obstruction, pulmonary vascular disease, low systemic ventricular function, and preexisting arrhythmias in the presence of implanted cardiac rhythm devices such as pacemakers and implantable cardioverter-defibrillators. In addition, the physiological effects of athletic activities at high altitude should be considered for patients with elevated pulmonary vascular resistance. These issues are covered elsewhere in this document. Fortunately,

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although repaired CHD is clearly associated with the development of arrhythmias such as atrial flutter and ventricular tachycardia, exercise does not appear to contribute to the risk.

The level of sports participation recommended includes consideration of both the training and the competitive aspects of the activity but must be individualized to the particular patient, taking into account the patient's functional status and history of surgery. Noninvasive testing, such as formal exercise testing, Holter monitoring, echocardiography, and cardiac magnetic resonance imaging studies, is also often useful.

TYPES OF CONGENITAL DEFECTS

Simple Shunting Lesions (Atrial Septal Defect, Ventricular Septal Defect, Patent Ductus Arteriosus), Treated and Untreated

Of the 8 most common subtypes of CHD, ventricular septal defect (VSD; 34%), atrial septal defect (ASD; 13%), and patent ductus arteriosus (PDA; 10%), respectively, are the most common (2). With rare exceptions, patients with hemodynamically insignificant CHD such as VSD, ASD, and PDA may participate competitively in all sports. There are no demonstrative data that children with hemodynamically insignificant VSD (open or after closure), ASD (open or after closure), or PDA (open or after closure) require exercise limitations or that these lesions are related to acknowledged episodes of sudden cardiac death (SCD) (3,4). Patients with associated pulmonary hypertension secondary to the above-mentioned lesions that is hemodynamically significant can develop acute symptoms, including reduced exercise capacity or, more importantly, arrhythmias, syncope, chest pain, or sudden death (5,6). For the purposes of this document, pulmonary hypertension is defined as a mean pulmonary artery pressure >25 mm Hg or a pulmonary vascular resistance index of >3 Wood units.

Patients with right-to-left shunting may become more cyanotic during exercise, at least in part because of changes in the ratio of systemic vascular resistance to pulmonary vascular resistance, which can result in increased hypoxemia. Therefore, full clinical assessment, including laboratory and exercise testing, should be considered before any physical activity, because this population represents a very high risk of sudden death (6). Additional precautions should be taken when these patients are exercising at altitude, because the pulmonary vascular resistance generally rises, thus increasing the degree of hypoxemia and cardiac workload.

Children with open or surgically closed VSDs have a normal exercise capacity despite a mild chronotropic

limitation in the latter. Some data suggest that aerobic capacity is reduced in patients with open or closed VSDs, as well as in patients with closed ASDs. Abnormal right ventricular (RV) and pulmonary pressure can also occur in those with isolated VSDs; however, these findings did not impact these exercise recommendations or identify any episodes of SCD (7).

ASD: Untreated

Recommendations

1. It is recommended that athletes with small defects (<6 mm), normal right-sided heart volume, and no pulmonary hypertension should be allowed to participate in all sports (*Class I; Level of Evidence C*).
2. It is recommended that athletes with a large ASD and no pulmonary hypertension should be allowed to participate in all sports (*Class I; Level of Evidence C*).
3. Athletes with an ASD and pulmonary hypertension may be considered for participation in low-intensity class IA sports (*Class I; Level of Evidence C*).
4. Athletes with associated pulmonary vascular obstructive disease who have cyanosis and a large right-to-left shunt should be restricted from participation in all competitive sports, with the possible exception of class IA sports (*Class III; Level of Evidence C*).

ASD: After Surgical Repair or Closure by Interventional Catheterization

Recommendations

1. Three to 6 months after operation or intervention, athletes without pulmonary hypertension, myocardial dysfunction, or arrhythmias may participate in all sports (*Class I; Level of Evidence C*).
2. After operation or intervention, patients with pulmonary hypertension, arrhythmias, or myocardial dysfunction may be considered for participation in low-intensity class IA sports (*Class IIb; Level of Evidence C*).

VSD: Untreated

Recommendations

1. An athlete with a small or restrictive VSD with normal heart size and no pulmonary hypertension can participate in all sports (*Class I; Level of Evidence C*).
2. An athlete with a large, hemodynamically significant VSD and pulmonary hypertension may consider participation in only low-intensity class IA sports (*Class IIb; Level of Evidence C*).

VSD: After Surgical Repair or Closure by
Interventional Catheterization**Recommendations**

1. At 3 to 6 months after repair, asymptomatic athletes with no or a small residual defect and no evidence of pulmonary hypertension, ventricular or atrial tachyarrhythmia, or myocardial dysfunction can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with persistent pulmonary hypertension should be allowed to participate in class IA sports only (*Class I; Level of Evidence B*).
3. Athletes with symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree atrioventricular block should not participate in competitive sports until further evaluation by an electrophysiologist (*Class III; Level of Evidence C*).
4. Athletes with mild to moderate pulmonary hypertension or ventricular dysfunction should not participate in competitive sports, with the possible exception of low-intensity class IA sports (*Class III; Level of Evidence C*).

PDA: Untreated

Recommendations

1. Athletes with a small PDA, normal pulmonary artery pressure, and normal left-sided heart chamber dimension can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with a moderate or large PDA and persistent pulmonary hypertension should be allowed to participate in class IA sports only (*Class I; Level of Evidence B*).
3. Athletes with a moderate or large PDA that causes left ventricular (LV) enlargement should not participate in competitive sports until surgical or interventional catheterization closure (*Class III; Level of Evidence C*).

PDA: Treated (After Surgical Repair or Closure by
Interventional Catheterization)**Recommendations**

1. After recovery from catheter or surgical PDA closure, athletes with no evidence of pulmonary hypertension can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with residual pulmonary artery hypertension should be restricted from participation in all competitive sports, with the possible exception of class IA sports (*Class I; Level of Evidence B*).

Pulmonary Valve Stenosis: Treated and Untreated

Mild valvar pulmonary stenosis (PS) is characterized by a systolic ejection murmur, a systolic ejection click that

varies with respiration, and a normal ECG. Decisions are based on estimated severity by use of Doppler-derived peak instantaneous gradients. A gradient <40 mm Hg indicates mild PS, 40 to 60 mm Hg indicates moderate PS, and >60 mm Hg indicates severe PS. Treatment can be by surgery or more commonly by balloon valvuloplasty. Adequate relief means a resolution of symptoms or a reduction in gradient to <40 mm Hg.

Recommendations

1. Athletes with mild PS and normal RV function can participate in all competitive sports. Annual reevaluation is also recommended (*Class I; Level of Evidence B*).
2. Athletes treated by operation or balloon valvuloplasty who have achieved adequate relief of PS (gradient <40 mm Hg by Doppler) can participate in all competitive sports (*Class I; Level of Evidence B*).
3. Athletes with moderate or severe PS can consider participation only in low-intensity class IA and IB sports (*Class IIb; Level of Evidence B*).
4. Athletes with severe pulmonary insufficiency as demonstrated by marked RV enlargement can consider participation in low-intensity class IA and IB sports (*Class IIb; Level of Evidence B*).

Aortic Valve Stenosis: Treated and Untreated

Assessment of fully grown athletes with aortic stenosis (AS) is discussed in the Task Force 5 report on valvular heart disease (8). The following discussion pertains to recommendations in children and adolescents. Patients with AS are differentiated between those with mild, moderate, and severe AS by physical examination, ECG, and Doppler echocardiography. In all cases, regardless of the degree of stenosis, patients with a history of fatigue, light-headedness, dizziness, syncope, chest pain, or pallor on exercise deserve a full evaluation. Annual reevaluation is required for all patients with AS, because the disease can progress. Patients with severe AS are at risk of sudden death, particularly with exercise (9).

Mild AS is defined as a mean Doppler gradient of <25 mm Hg or a peak instantaneous Doppler gradient <40 mm Hg. On evaluation, patients should have a normal ECG, normal exercise tolerance, and no history of exercise-related chest pain, syncope, or atrial or ventricular tachyarrhythmia. Moderate AS is defined as a mean Doppler gradient of 25 to 40 mm Hg or a peak instantaneous Doppler gradient of 40 to 70 mm Hg. Patients should have only mild or no LV hypertrophy by echocardiogram and an absence of LV strain pattern on ECG, as well as a normal maximum exercise stress test without

evidence of ischemia or tachyarrhythmia, with normal exercise duration and blood pressure response. Severe AS is defined as a mean Doppler gradient >40 mm Hg or a peak instantaneous Doppler gradient >70 mm Hg. Such patients may have symptoms such as exercise intolerance, chest pain, near-syncope, or syncope and likely will have LV hypertrophy with strain on ECG, as well as an abnormal blood pressure response to exercise. For cases in which symptoms or findings on ECG or exercise test appear more severe than expected for the estimated severity by Doppler, cardiac catheterization may be indicated.

Treatment may be by surgery or balloon aortic valvuloplasty. After treatment, patients may be left with residual valve gradient, aortic insufficiency, or both and may experience recurrence or progression, and thus, continued clinical follow-up is needed.

Recommendations

- 1. Athletes with mild AS can participate in all competitive sports (Class I; Level of Evidence B).**
- 2. Athletes with severe AS can participate only in low-intensity class IA sports (Class I; Level of Evidence B).**
- 3. Athletes with moderate AS may be considered for participation in low static or low to moderate dynamic sports (class IA, IB, and IIA) (Class IIB; Level of Evidence B).**
- 4. Athletes with severe AS should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).**

AS After Surgery or Balloon Dilation

Recommendations

- 1. Athletes with residual AS may be considered for participation in sports according to the above recommendations based on severity (Class IIB; Level of Evidence C).**
- 2. Athletes with significant (moderate or severe) aortic valve insufficiency may participate in sports according to the recommendation of Task Force 5 in this document (8).**

Coarctation of the Aorta: Treated and Untreated

Coarctation may be discrete or in the form of a long segment and causes hypertension in the upper limbs and hypotension in the lower limbs. The severity is determined by a clinical examination that includes the arm/leg pressure gradient, exercise testing, echocardiographic studies, and magnetic resonance imaging. Coarctation is often considered part of a more general aortopathy with a medial abnormality, particularly when associated with

a bicuspid aortic valve. This renders the aorta more vulnerable to dilation, aneurysm formation, and dissection and rupture. There is a recognized association with cerebral aneurysms. Virtually all patients, except those with mild coarctation, will undergo intervention, in the form of either surgical repair or percutaneous balloon angioplasty and stenting.

Even after successful surgical repair or stent placement, residual abnormalities may persist. These include residual coarctation and aneurysm formation at the site of repair or stent. Because of the aortopathy, the ascending aorta may also dilate and even dissect and rupture. Systemic hypertension may persist and if not present at rest may also occur on exercise. Some patients may have residual LV hypertrophy, and many may have residual aortic valve disease when a concomitant bicuspid aortic valve is present. Lifetime follow-up is mandatory, and the potential for premature coronary artery disease has been reported.

Before a decision is made regarding exercise participation, a detailed evaluation should be conducted, which should include a physical examination, ECG, chest radiograph, exercise testing transthoracic echocardiographic evaluation of the aortic valve and aorta, and either magnetic resonance imaging or computed tomography angiography. Normal standards exist for peak systolic blood pressure on exercise testing, by age and sex (10,11). Magnetic resonance imaging or computed tomography imaging should be performed to evaluate the thoracic aorta in its entirety, because transthoracic echocardiographic imaging alone will not visualize the entire aorta, and both residual coarctation and aneurysm may be missed.

Coarctation of the Aorta: Untreated

Recommendations

- 1. Athletes with coarctation and without significant ascending aortic dilation (z score ≤ 3.0 ; a score of 3.0 equals 3 standard deviations from the mean for patient size) with a normal exercise test and a resting systolic blood pressure gradient <20 mm Hg between the upper and lower limbs and a peak systolic blood pressure not exceeding the 95th percentile of predicted with exercise can participate in all competitive sports (Class I; Level of Evidence C).**
- 2. Athletes with a systolic blood pressure arm/leg gradient >20 mm Hg or exercise-induced hypertension (a peak systolic blood pressure exceeding the 95th percentile of predicted with exercise) or with significant ascending aortic dilation (z score >3.0) may be considered for participation only in low-intensity class IA sports (Class IIB; Level of Evidence C).**

Coarctation of the Aorta: Treated by Surgery or Balloon and Stent**Recommendations**

- 1. Athletes who are >3 months past surgical repair or stent placement with <20 mm Hg arm/leg blood pressure gradient at rest, as well as (1) a normal exercise test with no significant dilation of the ascending aorta (z score <3.0), (2) no aneurysm at the site of coarctation intervention, and (3) no significant concomitant aortic valve disease, may be considered for participation in competitive sports, but with the exception of high-intensity static exercise (classes IIIA, IIIB, and IIIC), as well as sports that pose a danger of bodily collision (Class IIb; Level of Evidence C).**
- 2. Athletes with evidence of significant aortic dilation or aneurysm formation (not yet at a size to need surgical repair) may be considered for participation only in low-intensity (classes IA and IB) sports (Class IIb; Level of Evidence C).**

Elevated Pulmonary Vascular Resistance in CHD

Patients with pulmonary vascular disease and CHD are at risk of sudden death during sports activity. In those with shunts (commonly septal defects or complex CHD), cyanosis is usually present at rest (Eisenmenger syndrome) and worsens with exercise. Most of these patients self-limit their activity, and they should not participate in competitive sports, with the exception of low-intensity (class IA) sports. The benefits of a regular exercise program, however, including improved walk distance, peak oxygen consumption, quality of life, and functional class, have been demonstrated, and thus, physical activity that does not require maximal effort should be encouraged. This usually comprises physical activity that allows the patient to speak a sentence comfortably (the “talk test”), and 6-minute walk tests will facilitate guidance in this regard.

Patients with suspected residual pulmonary hypertension who have undergone prior surgical repair or catheter intervention for shunt lesions should have a complete hemodynamic evaluation by cardiac catheterization before engaging in competitive athletics. Pulmonary arterial hypertension is usually defined as a mean pulmonary artery pressure of >25 mm Hg and a pulmonary arteriolar resistance >3 Wood units. Decisions proscribing exercise for patients with mild degrees of pulmonary hypertension are quite arbitrary, and no evidence-based scientific data exist. Similarly, no data exist with regard to appropriate exercise prescriptions for patients with mild and moderate pulmonary hypertension, which emphasizes the need to collect prospective data.

Patients and families should be cautioned, however, concerning the potential effect of high altitude on the existing abnormal cardiopulmonary physiology, because this may lead to important further elevations in pulmonary vascular resistance in such patients, with adverse effects.

Recommendations

- 1. Patients with mean pulmonary artery pressure of <25 mm Hg can participate in all competitive sports (Class I; Level of Evidence B).**
- 2. Patients with moderate or severe pulmonary hypertension, with a mean pulmonary artery pressure >25 mm Hg, should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports. Complete evaluation and exercise prescription (physician guidance on exercise training) should be obtained before athletic participation (Class III; Level of Evidence B).**

Ventricular Dysfunction After CHD Surgery

It is not unusual for a patient to present with significant ventricular dysfunction early or late after surgery for CHD, and this dysfunction, of course, affects exercise performance. Assessment of ventricular function is more straightforward for patients with systemic LVs than for those with systemic RVs, but the use of cardiac magnetic resonance imaging has improved the assessment of RV function (12). In general, throughout this document, severe ventricular dysfunction is defined as an ejection fraction (EF) <40%, moderate dysfunction as EF 40% to 50%, and normal as EF >50%. It should be recognized that these definitions are somewhat arbitrary. Of course, the other characteristics of the patient's heart disease and repair should be considered as well, such as valvar stenosis and insufficiency.

Recommendations

- 1. Before participation in competitive sports, all athletes with ventricular dysfunction after CHD surgery should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Athletes with normal or near-normal systemic ventricular function (EF ≥50%) can participate in all sports (Class I; Level of Evidence B).**
- 3. It is reasonable for athletes with mildly diminished ventricular function (EF 40%-50%) to participate in low- and medium-intensity static and dynamic sports (classes IA, IB, and IIA and IIB) (Class IIb; Level of Evidence B).**

4. Athletes with moderately to severely diminished ventricular function (EF <40%) should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).

Cyanotic CHD, Including Tetralogy of Fallot

Cyanotic Heart Disease: Unoperated or With Palliative Shunts

Patients with congenital defects resulting in chronic cyanosis can reach adolescence and adulthood but have significantly diminished exercise tolerance, which correlates with clinical outcomes (13-15). Iron deficiency further exacerbates exercise intolerance, whereas select treatments may improve exercise capacity in this population (16,17). Cardiopulmonary exercise testing shows that significant desaturation occurs in these patients with exercise, with performance and symptoms related to underlying anatomy (14,18), including those with palliative shunts, because of changes in the balance between pulmonary and systemic vascular resistance. Full clinical assessment, including laboratory and exercise testing, should be considered before any physical activity, because this population represents a very high risk of sudden death (19). Additional caution should be taken when these patients are exercising at altitude ("Elevated Pulmonary Vascular Resistance in CHD"). Unfortunately, data to address the safety of participation in competitive sports in this population are lacking.

Recommendations

- 1. In athletes with unrepaired cyanotic heart disease, a complete evaluation is recommended, which should involve exercise testing. An exercise prescription based on clinical status and underlying anatomy should be obtained before athletic participation (Class I; Level of Evidence C).**
- 2. Athletes with unrepaired cyanotic heart disease who are clinically stable and without clinical symptoms of heart failure may be considered for participation in only low-intensity class IA sports (Class IIb; Level of Evidence C).**

Postoperative Tetralogy of Fallot

Most patients with tetralogy of Fallot currently undergo initial repair in the first 2 years of life but often develop clinically significant pulmonary valve dysfunction in adolescence or adulthood. Clinical evaluation of patients before participation in competitive sports should include assessment of pulmonary valve function and assessment of factors associated with increased risk of sudden death in this population (15,19-21). In particular, attention should be paid to

careful assessment of LV function (13,22). Exercise testing is recommended to evaluate ability to augment cardiovascular function during increasing exercise intensity and for evidence of exercise-related ECG changes suggestive of arrhythmia or ischemia. Given its prognostic utility, cardiopulmonary exercise testing should be considered to fully evaluate patients before sports participation, particularly those with evidence of residual lesions on physical examination or imaging assessment (13,15). We strongly caution against participation in high-intensity competitive sports for those with severe biventricular dysfunction, atrial or ventricular arrhythmias, and significant abnormalities on exercise testing or abnormal hemodynamic assessment. Evaluation of lung function with pulmonary function tests may also be useful to assess for evidence of underlying disease and optimization before sports participation (23). For participation in moderate- and high-intensity sports, the patient should be asymptomatic at rest and with exercise, as well as free (or relatively free) of risk factors associated with sudden death, although individualized assessment is key for assessment of additional anatomic anomalies such as anomalous coronary arteries or residual outflow tract obstruction. Specific data regarding safety of long-term high-intensity exercise are needed in tetralogy of Fallot patients with preserved ventricular function with moderate to severe regurgitation, because a blunted stroke-volume response with high-intensity exercise has been reported in this population. One could extrapolate from these data that exercise performance in class III sports would be limited, although there is insufficient evidence to understand cardiovascular risk for these athletes (24). Given this, we recommend serial clinical evaluation with assessment of ventricular function during the period of sports participation.

Recommendations

- 1. Before participation in competitive sports, it is recommended that all athletes with repaired tetralogy of Fallot should undergo evaluation, including clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Athletes without significant ventricular dysfunction (EF >50%), arrhythmias, or outflow tract obstruction may be considered for participation in moderate- to high-intensity sports (class II to III). To meet these criteria, the athlete must be able to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms (Class IIb; Level of Evidence B).**

3. Athletes with severe ventricular dysfunction (EF <40%), severe outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).

Transposition of the Great Arteries: After Atrial Switch (Mustard or Senning Operation)

The atrial switch procedure was reported in 1959 and was performed frequently for transposition of the great arteries (TGA) from approximately the 1960s to the 1990s. Thus, the significant majority of patients with this anatomy are adults, because survival into the third and fourth decades occurs in most patients. Exercise tolerance is diminished in this population and correlates with clinical outcomes (13,15). Recent studies show this population may be at higher risk of sudden death than other CHD populations (19,20). The strongest predictors of sudden death are the presence of prior arrhythmia and severe systemic ventricular dysfunction, although prior VSD, age at repair, QRS duration, and heart failure symptoms may also be associated with an increased risk (19,25-29). The population with TGA with atrial switch likely has a unique response to exercise given reports that a high proportion of sudden death events occur during exertion (28). This adds complexity to the evaluation before sports participation, because the pathophysiology and prevention strategies for SCD in this population are not well understood. Unfortunately, evidence of exercise-induced arrhythmias on routine clinical testing has not been shown to reliably predict exercise-induced SCD events (28). Thus, careful evaluation of clinical status with special attention to clinical history of arrhythmias, patency and structure of the venous baffles, systemic ventricular function, coronary artery anatomy, and presence of additional obstructive lesions (e.g., PS) is recommended. Severe systemic ventricular function is defined as an EF <40%. Clinical evaluation should include cardiopulmonary exercise testing with continuous oximetry before sports participation. Restriction from high-intensity activities should be considered in the presence of severe systemic ventricular dysfunction, persistent arrhythmias, hypoxia, or inability to increase cardiac output, blood pressure, or heart rate with exertion. In the absence of these findings, moderate-intensity sports participation may be safe (30). However, the effect of long-term exercise training on the systemic RV is not known. Therefore, we recommend serial clinical evaluation during the period of sports participation, with assessment of ventricular function to evaluate the medium- and long-term effects of exercise participation.

Evaluation for and optimization of pulmonary dysfunction are recommended (31).

Recommendations

1. It is recommended that before participation in competitive sports, all athletes who have undergone the Senning and Mustard procedure should undergo an evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).
2. Participation in competitive sports in those athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability (Class IIb; Level of Evidence C).
3. Athletes without clinically significant arrhythmias, ventricular dysfunction, exercise intolerance, or exercise-induced ischemia may be considered for participation in low- and moderate-intensity competitive sports (classes IA, IB, IIA, and IIB) (Class IIb; Level of Evidence C).
4. Athletes with severe clinical systemic RV dysfunction, severe RV outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports. (Class III; Level of Evidence C).

Congenitally Corrected TGA

Patients with congenitally corrected TGA (CCTGA) are often diagnosed in childhood, usually in the presence of additional defects, including PS, VSD, or systemic atrioventricular valve abnormalities (see appropriate sections for additional recommendations). In CCTGA, exercise tolerance is limited, and both exercise tolerance and ventricular function are predictive of adverse outcomes (13,15,32,33). Systemic atrioventricular valve dysfunction is not uncommon in this population and correlates with exercise performance (33). In a recent study, patients with CCTGA and additional defects were found to have a particularly high rate of sudden death (19). However, because of the small number of patients with this anatomy, it is difficult to determine the risk factors for this outcome, although systemic ventricular dysfunction and arrhythmias may correlate with these events. When evaluating patients before competitive sports participation, we recommend assessment of clinical stability with noninvasive imaging and cardiopulmonary exercise testing. Clinical assessment should include evaluation of systemic ventricular and atrioventricular valve function and coronary artery anatomy, as well as exclusion of outflow tract obstruction. One small study found that participation of patients with CCTGA in a 3-month

exercise training program of moderate to high intensity was not associated with clinical decline (30); however, the effect of long-term exercise training on the systemic RV is not known. Therefore, we recommend serial clinical evaluation during the period of sports participation, with assessment of ventricular function to evaluate the medium- and long-term effects of exercise participation.

Limited data are available to assess the risks associated with sports participation in those who have had a double-switch procedure that resulted in the redirection of pulmonary venous blood to the LV and aorta. However, assessment of the venous baffle and Rastelli or arterial switch integrity is required before consideration of sports participation.

Recommendations

- 1. It is recommended that before participation in competitive sports, all CCTGA athletes should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Participation in competitive sports in those CCTGA athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability (Class IIb; Level of Evidence C).**
- 3. Athletes with CCTGA and without clinically significant arrhythmias, ventricular dysfunction, exercise intolerance, or exercise-induced ischemia may be considered for participation in low- and moderate-intensity competitive sports (class IA and IB) (Class IIb; Level of Evidence C).**
- 4. Asymptomatic athletes with CCTGA and without abnormalities on clinical evaluation may be considered for participation in moderate- to high-intensity competitive sports (classes II and IIIB or IIIC) (Class IIb; Level of Evidence C).**
- 5. Athletes with severe clinical systemic RV dysfunction, severe RV outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence C).**

TGA, After Arterial Switch Procedure

Significant numbers of patients have now undergone the arterial switch procedure over the past 3 decades, and thus, many are at an age when sports participation is desired. Coronary stenosis or obstruction is fortunately rare, and concerns are mainly focused on the possibility of supraventricular PS at the site of anastomosis, which is rarely significant. Patients with symptoms such as

syncope or exertional chest pain should have a careful assessment of their coronary artery status, because sudden death has been reported late after arterial switch repair (34). Exercise studies are not particularly sensitive in this group of patients, and coronary angiography or other modalities such as computed tomography angiography may be necessary in those with significant symptoms (35). The issue of surveillance of asymptomatic patients after the arterial switch procedure is controversial.

Recommendations

- 1. It is recommended that before participation in competitive sports, athletes who have undergone the arterial switch procedure for TGA should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. It is reasonable for athletes with no cardiac symptoms, normal ventricular function, and no tachyarrhythmias after the arterial switch procedure for TGA to participate in all competitive sports (Class IIb; Level of Evidence C).**
- 3. After the arterial switch procedure for TGA, athletes with more than mild hemodynamic abnormalities or ventricular dysfunction may be considered for participation in low and moderate static/low dynamic competitive sports (classes IA, IB, IC, and IIA), provided that exercise testing is normal (Class IIb; Level of Evidence C).**
- 4. After the arterial switch procedure for TGA, athletes with evidence of coronary ischemia should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).**

Fontan Procedure

The Fontan operation, a complete redirection of systemic venous blood to the pulmonary arteries, is performed to palliate single-ventricle physiology. Patients with this circulation have significantly decreased exercise performance, and they are able to increase cardiac output during exercise through unique mechanisms (13,36,37). Limitation to exercise performance is multifactorial and correlates with morbidity and mortality (36,38). When patients are evaluated before sports participation, it is imperative to recognize that both the Fontan circulation and the underlying cardiac anatomy can be extremely variable among patients. As a result, thorough clinical assessment is recommended before sports participation. This clinical assessment should include evaluation for risk factors associated with sudden death (20,38,39). Additionally, comprehensive

cardiac imaging is recommended, as well as cardiopulmonary exercise testing with continuous oximetry. If there is significant exercise intolerance during a maximal effort test, as evidenced by such things as an inability to increase blood pressure or heart rate, systemic desaturation, or development of arrhythmias or other symptomatic limitations, the healthcare provider should strongly consider restriction from participation in moderate- and high-intensity competitive sports and training. If the recommended evaluation is unremarkable, participation in moderate-intensity and moderate-duration exercise can be considered. This recommendation is based on small studies that have shown evidence of improvement in some measures of fitness without evidence of clinical deterioration in those participating in moderate-intensity exercise training and resistance training (40,41). However, the safety of participation in high-intensity or high-duration sports is unknown. Additionally, evaluation and optimization of lung function before sports participation is recommended (42). All Fontan patients requiring chronic anticoagulation should be restricted from participation in contact sports.

Recommendations

- 1. It is recommended that before participation in competitive sports, all athletes who have undergone the Fontan procedure should undergo an evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Athletes who have undergone the Fontan procedure and who have no symptomatic heart failure or significantly abnormal intravascular hemodynamics can participate only in low-intensity class IA sports (Class I; Level of Evidence C).**
- 3. Participation in other sports may be considered on an individual basis with regard for the athlete's ability to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms (Class IIb; Level of Evidence C).**

Ebstein Anomaly of the Tricuspid Valve

The phenotypic spectrum of this malformation is extreme, ranging from minimal to profound tricuspid regurgitation and right-sided heart enlargement. If there is an atrial shunt, cyanosis may be present. A minority of patients with Ebstein anomaly will have preexcitation that could precipitate clinically important and symptomatic arrhythmias. Physical disability and increased risk for sudden death with exercise have been reported with severe cases. Risk stratification for exercise-related

arrhythmias remains imprecise for this anomaly. In patients for whom there is also evidence of Wolff-Parkinson-White syndrome or in whom a defibrillator has been implanted, the recommendations found in Task Force 9 (43) should be respected as well. Note that the recommendations below apply both before and after surgical plication and are based on the degree of valve regurgitation and existence of arrhythmias.

Recommendations

- 1. Patients with mild to moderate Ebstein anomaly (i.e., no cyanosis, normal RV size, tricuspid regurgitation that is moderate or less, and no evidence of atrial or ventricular arrhythmias) can be considered for participation in all sports (Class IIb; Level of Evidence C).**
- 2. Patients with Ebstein anomaly with severe tricuspid regurgitation but without evidence of arrhythmias on ambulatory electrocardiographic monitoring (except isolated premature contractions) may be considered for participation only in low-intensity class IA sports (Class IIb; Level of Evidence C).**

Congenital Coronary Anomalies

Anomalies of coronary arteries are second in frequency among identified structural causes of SCD in competitive athletes, accounting for $\approx 17\%$ of such deaths in the United States (44). Anomalous origins of coronary arteries from the wrong sinus of Valsalva or from the pulmonary artery are estimated to be present in $\approx 1\%$ of the overall population (45) but are proportionately far more common in athletes who die suddenly, as cited above. Although the vast majority of sudden deaths associated with coronary anomalies occur during or shortly after exercise (46), sudden death has been reported in the sedentary state (47).

The most common anomalous origin is the right coronary artery originating from the left sinus of Valsalva, but among athletes who have died suddenly, anomalous origin of the left main or left anterior descending coronary artery from the right sinus of Valsalva is far more prevalent. Furthermore, SCDs are most strongly associated with the pattern in which the anomalous left coronary artery passes between the aorta and main pulmonary artery. An anomalous origin of a coronary artery from the pulmonary artery is far less commonly observed in athletes who die suddenly and in fact often presents with myocardial infarction in infancy or early childhood. Nonetheless, some cases are not recognized until adolescence or adulthood and may be associated with sudden death in athletes, albeit rarely. Nonspecific electrocardiographic findings may be observed in adolescents with otherwise unrecognized anomalous coronary arteries arising from the pulmonary artery.

The ECG is an unreliable screening tool for suspecting or recognizing anomalous origin of coronary arteries before an event, and even stress tests are not uniformly positive among people with these anomalies (48). Clinical symptoms, such as exertional chest discomfort or dyspnea, may be helpful, but 2 reports suggest that 50% of SCDs associated with coronary artery anomalies were first events without prior symptoms (46,49). The best methods for identifying the anomaly include coronary angiography, computed tomography angiography, and magnetic resonance angiography. Although not uniformly successful, athletes undergoing echocardiographic studies for any reason should have careful attempts to identify the origins of the coronary arteries.

Surgical procedures are the only therapies available for correcting these anomalies (50), with return to intense athletic activities permitted after 3 months after the procedure with demonstration of the absence of ischemia on postoperative stress testing (51).

Recommendations

1. Athletes with anomalous origin of a coronary artery from the pulmonary artery can participate only in low-intensity class IA sports, whether or not they have had a prior myocardial infarction, and pending repair of the anomaly (*Class I; Level of Evidence C*).
2. Athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva should be evaluated by an exercise stress test. For those without either symptoms or a positive exercise stress test, permission to compete can be considered after adequate counseling of the athlete and/or the athlete's parents (in the case of a minor) as to risk and benefit, taking into consideration the uncertainty of accuracy of a negative stress test (*Class IIa; Level of Evidence C*).
3. After successful surgical repair of an anomalous origin from the wrong sinus, athletes may consider participation in all sports 3 months after surgery if the patient remains free of symptoms and an exercise stress test shows no evidence of ischemia or cardiac arrhythmias (*Class IIb; Level of Evidence C*).
4. After repair of anomalous origin of a coronary artery from the pulmonary artery, decisions regarding exercise restriction may be based on presence of sequelae such as myocardial infarction or ventricular dysfunction (*Class IIb; Level of Evidence C*).
5. Athletes with an anomalous origin of a left coronary artery from the right sinus of Valsalva, especially when the artery passes between the pulmonary artery and aorta, should be restricted from participation in all competitive sports, with the possible exception of class IA sports, before surgical repair. This recommendation applies whether the anomaly is identified as a consequence of symptoms or discovered incidentally (*Class III; Level of Evidence B*).
6. Nonoperated athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva who exhibit symptoms, arrhythmias, or signs of ischemia on exercise stress test should be restricted from participation in all competitive sports, with the possible exception of class IA sports, before a surgical repair (*Class III; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
George F. Van Hare	Washington University	None	None	None	None	None	None	None
Michael J. Ackerman	Mayo Clinic	NIH (R01 grants) [†]	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic [†]
Juli-anne K. Evangelista	Boston Children's Hospital	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None
Keri M. Shafer	Boston Children's Hospital	None	None	None	None	None	None	None
Carole A. Warnes	Mayo Clinic-Rochester	None	None	None	None	None	None	None
Reginald L. Washington	Rocky Mountain Hospital for Children	None	None	None	None	None	None	None

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*Modest.

[†]Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Emile Bacha	New York Presbyterian Hospital-Columbia University College of Physicians and Surgeons	None	None	None	None	None	None	None
Julian I.E. Hoffman	University of California	None	None	None	None	None	None	None
Robert D.B. Jaquiss	Duke University School of Medicine	None	None	None	None	None	None	None
Silvana M. Lawrence	Baylor College of Medicine	None	None	None	None	None	None	Vice President, Science and Research, Championship Hearts Foundation [†]
John Moore	UCSD	None	None	None	None	None	None	None
Karen K. Stout	University of Washington	None	None	None	None	None	None	None

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[†]Significant.

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