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RESEARCH ARTICLE

CONGENITAL HEART DISEASE

***Dr. Adesola Adefemi Muritala**

Department of Physiology, Faculty of Basic Medical Sciences, Ladoke Akintola University of Technology, P.M.B 4000, Ogbomoso, Oyo State, Nigeria

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**Corresponding author:*
Dr. Adesola Adefemi Muritala

ABSTRACT

The focus of this paper is the children with identified Cardiovascular Abnormality, and its goal is to develop prudent consensus recommendations regarding the eligibility of such children for competition in a sport. To accomplish this objective. I have attempted to ascertain by way of consensus which Cardiovascular Abnormalities and with what degree of severity would place the competitive athlete at increased risk for sudden death or disease progression, thereby justifying a medical recommendation against participation in certain sports. Both parents and physicians are involved with efforts to maximize recreational and competitive sports participation in Children with congenital heart disease while at the same time minimizing any potential risks. In this regard, it is fortunate that significant morbidity or mortality precipitated by exercise in patients with congenital heart disease is rare. Indeed, there are relatively few congenital heart lesions that have been associated with sudden death during sports participation. The most common are hypertrophic cardiomyopathy, congenital coronary artery anomalities, Marfan's syndrome, and myocarditis with less common causes including Valvular aortic stenosis, and complex defects tetrology of fallot, complete transposition and single ventricle, and those associated with Pulmonary Vascular Disease. The recommendations for the permitted level of activity are offered only as guidelines.

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INTRODUCTION

Both parents and physicians are involved with efforts to maximize recreational and competitivesports participation in children with congenital heart disease while at the same time minimizing any potential risks. In this regard, it is fortunate that significant morbidity or mortality precipitated by exercise in patients with congenital heart disease is rare. Indeed, there are relatively few congenital heart lesions that have been associated with sudden death during sports participation (Driscoll, 1985; Freed, 1984; Garson, 1985; Lambert, 1974). The most common are hypertrophic cardiomyopathy, congenital coronary artery anomalies, Marfan's syndrome, and myocarditis, with less common causes including valvular aortic stenosis, and complex defects such as tetralogy of Fallot, complete transposition and single ventricle, and those associated with pulmonary vascular disease. This list encompasses a small percentage of the total number of patients with congenital heart disease and even some of these patients, with appropriate testing, may engage in low intensity (Class IA [see Table 1 in Classification of Sports]) forms of competitive sports under certain conditions.

Although the risk of sudden cardiac death is the most compelling concern for the, health professional evaluating the athlete with congenital heart disease, progressive hemodynamic deterioration with time is also an additional potential consequence of intense, chronic exercise in some circumstances. These recommendations for the permitted level of activity are offered only as guidelines. The physician with knowledge of the particular patient, including the severity of the lesion and that individual's physiologic and psychologic response to training and competition, may choose to modify these recommendations in selected instances. Indeed, the excessive restriction of physical activity in children with cardiac malformations has the potential for causing considerable physical and psychological harm (Bar-or, 1983; Garson, 1974; Linde, 1982; Mocellin, 1976; Reybrouck, 1991). Eligibility recommendations for sports participation for children with congenital cardiac malformations have become somewhat more permissive over the past several years, without evidence for detrimental effects (Sklansky, 1993). For each patient the history, physical exam, and knowledge of the individual child's course become extremely important. Patients with a prior history of symptomatic arrhythmia, syncope, pulmonary hypertension, or myocardial dysfunction deserve special emphasis since such

historical data can put these patients in a higher risk category. Patients with significant heart disease who have a history of any of these abnormalities deserve full evaluation. Important hemodynamic information can be obtained by echocardiographic/Doppler assessment. Other evaluations may include exercise testing, ambulatory ECG monitoring, cardiac catheterizatiori, and invasive electrophysiologic studies under certain conditions. The congenital heart defects are presented to provide broad guidelines for patients with shunt lesions, obstructive lesions, and the more common complex cyanotic malformations (Maron, 1982). In the mild form of these common anomalies, most sports will be permissible. In the moderate form of these anomalies, immediate levels of sports participation may be safe but evaluation is required. In the severe form of most of these abnormalities, strenuous exercise could be detrimental to certain patients (Lambert, 1974). It is important to stress that there is no substitute for a comprehensive history and physical examination by a physician with interest and expertise in patients with congenital heart disease to make recommendations for patients in whom there are questions about the severity of the abnormality and the optimal exercise prescription. Exercise testing can be useful, particularly with monitoring of symptoms, ECG, and blood pressure during conditions simulating both the dynamic and static demands of the sport in question. In general, serial yearly evaluations are required because of changing hemodynamics with growth and with changing severity of the congenital cardiac defect.

TYPES OF CONGENITAL DEFECTS

Atrial Septal Defect, Untreated: Atrial septal defect indicates any persistent communication between the atria. Most patients with atrial septal defect in childhood are without symptoms and operation is generally carried out before- the age at which childrenare normally active in sports. Current practice usually mandates a history, physical exam, ECG, and echocardiogram for complete evaluation and categorization of patients with atrial septal defect. Small defects are characterized by minimal or no right ventricular volume overload; moderate or large defects have significant volume overload and can have variable degrees of pulmonary hypertension.

Recommendations

- Athletes with small defects and without pulmonary hypertension can participate in all competitive sports.
- Athletes with significant pulmonary hypertension and/or right-to-left shunts can participate only in low intensity competitive sports (Class IA). Also, athletes in the severe Eisenmenger class with marked cyanosis and large right-to-left shunting cannot participate in competitive sports. See section, Elevated Pulmonary Resistance.
- Athletes with symptomatic supraventricular or ventricular arrhythmia or significant mitral regurgitation should follow recommendations for athletes with these abnormalities in Task Force 6 and Task Force 2, respectively.

Atrial Septal Defect, Closed at Operation or by Interventional Catheterization: Atrial septal defect usually is completely corrected by operation. Generally, when operation is performed in childhood there is little or no residual right ventricular enlargement. Supraventricular arrhythmias can occur after repair of the defect and are generally more common when the defect is repaired later in life (Bink-Boelkens, 1983; Sealy, 1969; Vetter, 1982). The evaluation for competitive sports should include an estimate of cardiac performance, pulmonary vascular resistance, right ventricular size, and a search for conduction or rhythm disturbances. Minimal diagnostic studies are usually needed before a decision can be made regarding competitive sports, including a chest x-ray and ECG. If cardiomegaly is present by physical exam or x-ray, echocardiography may be useful. Patients with preoperative pulmonary hypertension and/or right-to-left shunting prior to operation require echocardiographic assessment of pulmonary artery pressure with Doppler or cardiac catheterization.

Recommendation

Six months after the operation or intervention, athletes can participate in all competitive sports unless the following are present:

- Evidence of pulmonary hypertension
- Arrhythmias associated with symptoms
- Evidence for myocardial dysfunction

Athletes with any of these residual abnormalities should have exercise evaluations and individualized exercise prescriptions. For further guidance in this regard, refer to Elevated Pulmonary Resistance and Ventricular Dysfunction in this Task Force report as well as Task Force 6.

Ventricular Septal Defect, Untreated: For purposes of sports recommendations, ventricular septal defects can be categorized as small, moderate, or large. The physician experienced in the evaluation of patients with congenital heart disease usually can make the diagnosis of small ventricular septal defect by clinical features alone. If physical examination and ancillary studies indicate normal heart size and normal pulmonary artery pressure in patients suspected clinically of having a small ventricular septal defect, then further evaluation is not required. An individual with a ventricular septal defect who does not readily fit into the category of small defect requires further investigation, usually echocardiography. If this modality is not diagnostic for a moderate-sized defect with normal pulmonary artery pressure then cardiac catheterization may be required. Large right-to-left shunts are discussed under Elevated Pulmonary Resistance. A moderate defect with low pulmonary vascular resistance will have a pulmonary-to-systemic flow ratio of approximately 1.5-1.9. A large defect with low or mildly increased pulmonary vascular resistance is defined as a pulmonary systemic flow ratio of ≥ 2 and pulmonary resistance <3 units-M^2.

Recommendations

- Athletes with a small or moderate ventricular septal defect can participate in all competitive sports.
- Selected athletes with a large ventricular septal defect can participate in some low intensity competitive sports (Class IA). Those athletes who do not have marked elevation of pulmonary resistance are candidates for repair and high intensity sportsparticipation can await successful operation.

Ventricular Septal Defect, Closed at Operation or by Interventional Catheterization: Successful repair is characterized by the absence of symptoms, absence of significant cardiomegaly or arrhythmias, and normal

Table 1. Classification of Sports: Based on peak components during competition

	A. Low Dynamic	B. Moderate Dynamic	C. High Dynamic
I. Low static	Billiards	Baseball	Badminton
	Bowling	Softball	Cross-country skiing (classic technique)
	Cricket	Table tennis	Field hockey*
	Curling	Tennis (doubles)	Orienteering
	Golf	Volleyball	Race walking
	Riflery		Racquetball
			Running (long-distance)
			Soccer*
			Squash
			Tennis (singles)
II. Moderate static	Archery	Fencing	Basketball*
	Auto racing*+	Field events (jumping)	Ice hockey*
	Diving*+	Figure skating*	Cross-country skiing (skating technique)
	Equestrian*+	Football (American)	Football (Australian rules)*
	Motorcycling*+	Rodeoing*+	Lacrosse*
		Rugby*	Running (middle-distance)
		Running (sprint)	Swimming
		Surfing*+ Synchronized swimming+	Team handball
III. High static	Bobsledding*+	Body building*+	Boxing*
	Field events (throwing)	Downhill skiing*+	Canoeing/kayaking
	Gymnastics*+	Wresting*	Cycling*+
	Karate/judo*		Decathlon
	Luge*+		Rowing
	Sailing		Speed skating
	Rock climbing*+		
	Water skiing*+		
	Weight lifting*+		
	Wind surfing*+		

* Danger of bodily collision. + Increased risk if syncope occurs.

pulmonary artery pressure. A small residual defect with the characteristic murmur can be present. Minimal diagnostic evaluation before sports participation following surgery includes chest radiograph, and ECG. Echocardiographic/Doppler studies are useful for patients in whom the examination, chest radiograph, and ECG are not diagnostic. Patients with residual cardiomegaly, myocardial dysfunction, or questionable pulmonary hypertension may require an exercise test or catheterization before a decision for sports participation can be made.

Recommendations

- Athletes ≥6 months following repair, with no residual defect and no symptoms, can participate in all competitive sports if they have no evidence for pulmonary artery hypertension, arrhythmia, or myocardial dysfunction.
- Athletes with residual small defects and no pulmonary hypertension and no symptoms can also participate in all competitive sports.
- Athletes with residual moderate or large defects can participate in low intensity sports (Class IA), but should have repair performed.
- Athletes with persistent severe pulmonary hypertension cannot participate in any competitive sports (see section, Elevated Pulmonary Resistance).
- Athletes with significant arrhythmias (see Task Force 6).
- Athletes with mild to moderate pulmonary hypertension or ventricular dysfunction (also see sections, Elevated Pulmonary Resistance and Ventricular Dysfunction after Cardiac Surgery).

Patent DuctusArteriosus, Untreated: The individual with a small patent ductusarteriosushas a characteristic murmur, the absence of symptoms, and normal heart size. Those with larger patent ductushave cardiomegaly and collapsing pulses with widened pulse pressure on physical examination. There may or may not be evidence of pulmonary hypertension. Minimum diagnostic studies generally include echocardiography, but in patients with atypical findings, cardiac catheterization may be required.

Recommendations

- Athletes with a small patent ductusarteriosus can participate in all competitive sports.
- Athletes with a moderate or large patent ductus, causing left ventricular enlargement, must have it repaired or closed by interventional catheterization before unrestricted competition.
- Athletes with large patent ductus and severe pulmonary hypertension should be characterized under Eisenmenger Syndrome, see below.

Patent DuctusArteriosus, Closed at Operation or by Interventional Catheterization: A successful result is characterized by the absence of symptoms and normal cardiac examination. Recommendations:

- Athletes after operative or interventional closure of a patent ductus who have no symptoms, a normal cardiac examination with no evidence of pulmonary hypertension or cardiac enlargement can participate in all competitive sports 3 months after repair.
- Athletes with residual pulmonary artery hypertension must be restricted if pulmonary hypertension persists by noninvasive or invasive testing (also, see section, Elevated Pulmonary Resistance).

Pulmonary Valve Stenosis, Untreated: The severity of pulmonary stenosis generally can bedefined by clinical features. Mild stenosis is characterized by a systolic ejection

murmur and variable ejection click, and a normal electrocardiogram or mild right ventricular hypertrophy. Echocardiographic/Doppler studies of peak instantaneous systolic pressure gradients are now widely used to evaluate severity. Mild stenosis is usually characterized by a transvalvular gradient < 40 mm Hg; moderate stenosis, 40-70 mm Hg; and severe stenosis > 70 mm Hg. Most patients with gradient a ≥50 mm Hg have balloon valvuloplasty performed.

Recommendations

- Athletes with peak systolic gradient of <50 mm Hg and normal right ventricular function can participate inall competitive sports if no symptoms are present. Annual reevaluation is recommended.
- Athletes with a peak systolic gradient of >50 mmHg can participate in low intensity competitive sports (Class IA). Patients in this category are likely to be referred for intervention with balloon valvuloplasty oroperative valvotomy.

Pulmonary Valve Stenosis, Treated by Surgery or Balloon Valvuloplasty: Adequate relief of pulmonary stenosis is characterized by absence of symptoms as well as physical examination and echocardiographic/Doppler study indicating a gradient in the mild range.

Recommendations

- Athletes with adequate relief of obstruction and normal ventricular function without symptoms can participate in all competitive sports. If balloon valvuloplastyis performed, participation in competitive sports can begin approximately 1 month after intervention. If operation is required, an interval of approximately 3 monthswould be reasonable before beginning to participate in competitive sports.
- Athletes with persistent peak systolic gradients >50 mm Hg should follow the same recommendations as forpatients before treatment.
- Athletes with severe pulmonary incompetence characterized by a marked rightventricular enlargement require individual assessment.

Aortic Valve Stenosis, Untreated: This section deals with congenital valvular aortic stenosis in young patients. Aortic stenosis in adults is discussed in Task Force 2. Congenital aortic stenosis is usually easily identifiable by a constant apical ejection click and a systolic ejection murmur heard maximally at the upper right sternal border. Differentiation between mild and either moderate or severe aortic stenosis is readily accomplished with ECG and Doppler echocardiography, as well as the physical examination. The distinction, however, between moderate or severe stenosis is more difficult and may require cardiac catheterization to clarify. Patients with a history of fatigue, light-headedness, dizziness, syncope, chest pain, or pallor with exercise deserve a full evaluation, usually including catheterization and exercise testing. The lesion may progress in severity with time, and consequently periodic reevaluation is necessary. Sudden death is more likely to occur in patients with severe disease, severe left ventricular hypertrophy, exertional syncope, chest pain or dyspnea, and left ventricular strain pattern on ECG. Between 20% and 80% of deaths in patients with severe aortic stenosis have been found to occur with physical exertion (4,6,14). For purposes of these sports recommendations, *mild* aortic stenosis is, in general, characterized by a resting peak instantaneous systolic pressure gradient (in the presence of normal cardiac output) of ≤20 mm Hg by catheterization or Doppler echocardiography, *moderate* stenosis 21-49 mm Hg, and *severe* stenosis ≥ 50 mm Hg. This classification is much different from that used for making clinical decisions regarding balloon valvuloplastyor surgery and represents a conservative approach tailored to strenuous sports participation in this anomaly because of the known problems of sudden death with severe aortic stenosis and the difficulty in achieving an up-to-date appraisal of severity for a lesion that can progress with time. It should be noted that, in contrast to the present discussion of *congenital* aortic valve stenosis severity that relies on peak instantaneous pressure gradient estimated with continuous wave Doppler, the section on *acquired* aortic stenosis (which appears in Task Force 2) assesses severity with respect to the *mean* aortic valve gradient.

Recommendations

- Athletes with mild aortic stenosis can participate in all competitive sports if they have a normal ECG and exercise tolerance, and no history of exercise-related chest pain, syncope, or arrhythmia associated with symptoms.
- Athletes with moderate aortic stenosis can participate in low static/low moderate dynamic and moderatestatic/low dynamic (Classes IA and IB and IIA) competitive sports if the followingconditions are met:
- Mild or no left ventricular hypertrophy by echocar-diography and the absence of left ventricular strain onECG.
- Normal exercise test with no evidence for ischemia or arrhythmia, normal exercise duration, and normalblood pressure response.
- An absence of symptoms as defined above.
- 3.Athletes with severe aortic stenosis should not par-ticipate in competitive sports.

The criteria in this section also apply to athletes with *discrete (membranous) subaortic stenosis* and *supravalvular aortic stenosis.*

Aortic Stenosis, Treated by Surgery or Balloon Valvuloplasty: After operation, a variable degree of residual stenosis and/or regurgitation can be present. Reevaluation byphysical exam, ECG, and echocardiogram is necessary for reassessment. In addition, .exercise stress testing and/or catheterization can be required for patients whose physiological and anatomical severity cannot otherwise be determined. Recommendations:

- Athletes with residual mild, moderate, or severe stenosis should follow the same recommendations as defined above.
- Athletes with moderate to severe aortic regurgitation should follow the guidelines in Task Force 2.

Coarctation of the Aorta, Untreated: This abnormality is characterized by an obstruction usually in the juxtaductal or juxtaligamental area with elevated blood pressure in the upper

limbs and relatively normal or decreased pressure in the lower limbs. Severity is assessed by the arm and leg pressure gradient, by physical exam, by exercise testing, and by echocardiographic/Doppler studies. Virtually all patients, except those with mild coarctation, will have either surgical repair or balloon dilatation.

Recommendations

- Athletes with mildcoarctation and the absence of large collateral vessels or severe aortic root dilation andwith normal exercise test and a small pressure gradient at rest (usually ≤20 mm Hg between upper and lower extremities; these pressure gradients usually are measured best by Doppler determination of systolic pressure inarms and legs using cuffs of appropriate size) and peaksystolic blood pressure with exercise ≤ 230 mm Hg can engage in all competitive sports.
- Athletes with resting systolic arm to leg gradient > 20 mm Hg or exercise induced hypertension with asystolic blood pressure> 230 mm Hg can engage only in low intensity competitive sports (Class IA) until treated.

Coarctation of the Aorta, Treated by Surgery or Balloon Arterioplasty: The majority of patients will have coarctation repair or balloon arterioplasty performed during childhood. After repair, abnormalities can persist, such as mild residual gradients, ventricular hypertrophy, systemic hypertension, and residual obstruction evident on exercise (8,13,20) prior to a decision regarding eligibility for sports participation, minimal diagnostic studies which include chest radiograph, exercise testing, and evaluation of left ventricular function should be done. In patients with balloon angioplasty, magnetic resonance imaging can be useful to determine if an aneurysm is present.

Recommendation:

Sports participation ≥ 6 months after coafetationtreatment is permitted for athletes in whom there is 20 mm Hg arm to leg blood pressure gradient at and peak systolic blood pressure during rest andexeccise is normal. (These pressure gradients usually are measured best by Doppler determination of systolic pressure in arms and legs using cuffs of appropriate size.) Athletes should refrain from high intensity static exercise (Classes IIIA, IIIB, and IIIC) and no sports with the danger of bodily collision during the first postoperative year. After 1 year, if patients continue to be asymptomatic with normal blood pressure at rest and exercise, all sports are permissible except power-lifting. For athletes with evidence of significant aortic dilatation, wall thinning, or aneurysm formation, participation should be restricted to low intensity competitive sports (Class IA).

Elevated Pulmonary Resistance: Patients with pulmonary vascular disease due to Eisenmenger syndrome are at risk for sudden death during intense sports activity. As pulmonary vascular obstruction progresses, these patients develop cyanosis at rest and intense cyanosis with exercise. Most of these patients have self-limited activity. These patients should not participate in competitive sports. Patients with suspected elevated pulmonary artery pressure after surgery or interventional catheterization for shunt lesions should have an estimation of this pressure by echocardiography or cardiac catheterization before engaging in competitive athletics.

Recommendations

- If pulmonary artery peak systolic pressure is ≤40 mm Hg, athletes can participate in all competitive sports.
- If pulmonary artery pressure is >40 mm Hg, an individual evaluation and exercise prescription are required for participation in competitive athletics.

Ventricular Dysfunction after Cardiac Surgery: Ventricular dysfunction can occur after treatment of both simple and complex congenital heart disease and can affect exercise performance, and therefore the recommendations for sports participation. Evaluation of left ventricular function can be performed noninvasively in most patients and is most useful when ejection parameters are measured simultaneously with an estimate of afterload. Evaluation of right ventricular function can be performed with radionuclide angiography, magnetic resonance imaging, angiocardiography, and less precisely with echocardiography. Load independent assessment of right ventricular function is not possible at present. Periodic assessment of ventricular function is required for continued recommendations for participation because ventricular function may deteriorate with time in some patients.

Recommendations

- For full participation in all competitive sports, normal or near normal ventricular function is required.
- Athletes with mildly depressed ventricular functionshould participate only in low intensity static competitive sports (Classes IA, IB, and IC).
- Athletes with moderately depressed ventricular function should in general participate only in low intensity competitive sports (Class IA).

Cyanotic Congenital Cardiac Disease, Unoperated: In most patients, cyanotic congenital heart disease produces exercise intolerance and progressive hypoxemia with increasing effort. Patients are unlikely to want to engage in competitive sports because of their own self-limiting activity. There are rare patients with cyanotic congenital heart disease who reach adolescence or even adult life with mild resting cyanosis and shortness of breath only with exercise. These patients may experience a rapid and profound decrease in arterial saturation during sports participation.

Recommendation: Athletes with unoperated cyanotic heart disease usually can participate only in low intensity competitive sports (Class IA), but individualized exercise prescriptions are recommended.

Postoperative Palliated Cyanotic Congenital Heart Disease: Palliative surgery can be performed to increase pulmonary blood flow in patients with decreased flow or to limit blood flow in those with excessive flow. Often these patients have significant relief of symptoms at rest but arterial desaturation during exercise frequently persists.

Recommendation: Athletes can usually participate in low intensity competitive sports (Class IA) provided the following criteria are met:

- Arterial saturation remains above approximately 80%.
- Symptomatic arrhythmias are not present.
- There is no symptomatic ventricular dysfunction.
- Athletes have near-normal physical working capacity on exercise testing.

Postoperative Tetralogy of Fallot: The current standard of treatment for tetralogy of Fallot is reparative operation in early childhood. Currently, most patients have good relief of stenosis with minimal residual pulmonary stenosis and only mild to moderate pulmonary insufficiency. Diagnostic evaluation for a decision regarding sports participation usually includes physical examination, chest radiograph, and ambulatory ECG monitoring and exercise testing. Echocardiography and/or cardiac catheterization may be required for complete evaluation in selected patients, particularly those with significant cardiomegaly and/or symptoms. Patients with important residual abnormalities such as significant left-to-right shunting, moderate to severe pulmonary regurgitation, or right ventricular dysfunction who also have a history of syncope or ventricular arrhythmia are at risk for sudden death (9).

Recommendations

- Athletes with an excellent result after repair of tetralogy of Fallot should be allowed to participate in all competitive sports providing there is:
  - Normal or near normal right heart pressure.
  - Only mild right ventricular volume overload.
  - No evidence for significant residual left-to-right shunting.
  - No rhythm abnormality on ambulatory ECG monitoring or exercise testing.
- Athletes with marked pulmonary regurgitation, residual right ventricular hypertension (peak systolicright ventricular pressure $\geq$0.5 systemic pressure), and/or rhythm abnormalities as defined in Task Force 6 should participate in only low intensity competitive sports (Class IA).

Transposition of the Great Arteries, Postoperative Mustard, or Senning Operation: Patients who have had atrial repair of transposition of the great arteries can have significant hemodynamic abnormalities despite clinical well-being. Such abnormalities include those of systemic venous return, abnormal right ventricular or systemic ventricular function, pulmonary stenosis or pulmonary hypertension, abnormalities of pulmonary venous return, tricuspid insufficiency, and significant atrial or ventricular arrhythmias (12). Left ventricular dilatation and hypertrophy develop in a normal highly fit young athlete. In transposition patients following atrial repair, it is the right ventricle that would be subjected to stress in the young athlete. Because of the anatomy of the right ventricle, its reserve is felt to be intrinsically less than that of the left ventricle and the consequences of hypertrophy and dilatation of the right ventricle of a highly trained individual after an otherwise excellent atrial repair of transposition are unknown. For these reasons, high physical intensity isometric or dynamic sports are not recommended for this group. Evaluation before competition in moderate and low intensity sports should include history and examination, chest radiograph, ECG, echocardiogram, ambulatory ECG monitoring, and exercise testing. For patients in whom data are unclear regarding hemodynamic abnormalities or ventricular function, cardiac catheterization may be necessary.

Recommendations

- Selected athletes can engage in low and moderate static, and low dynamic competitive sports (Classes IA and IIA) if there is:
- No significant cardiac enlargement on chest radiograph.
- No history of atrial flutter or ventricular arrhythmia (see Task Force 6).
- No history of syncope.
- A normal exercise test.
- Athletes who do not fit in this category require individualized exercise prescriptions.

Congenially Corrected Transposition of the Great Arteries
Congenitally corrected transposition of the great arteries is usually associated with other congenital malformations of the heart (such as ventricular septal defect and/or pulmonary stenosis and systemic atrioventricular valve abnormalities). These associated defects can preclude sports participation in many circumstances, depending on severity. The individual with congenitally corrected transposition is at risk for development of supraventricular tachycardia and for the late spontaneous development of atrioventricular block.

Recommendations: Some patients who have congenitally corrected transposition without other cardiac abnormalities may be eligible for full sports participation if there is no cardio-megaly and no evidence of arrhythmia by ambulatory ECG or exercise testing. Periodic reevaluation is important, however, to monitor the development of arrhythmias as well as for deterioration of systemic (right) ventricular function and systemic (tricuspid) atrioventricular valve regurgitation.

Postoperative Arterial Switch for Transposition of the Great Arteries: There is a significant cohort of young individuals who have had successful arterial switch repair of transposition and are now old enough to engage in competitive sports. These patients have a low prevalence of ventricular dysfunction, arrhythmia with symptoms, and hemodynamic sequelae (with the possible exception of pulmonary artery or anastomotic stenosis). Only limited exercise data are available for this group.

Recommendations:

- Six months after surgery there are no restrictions from sports competition if there is normal heart size, no residual defects, normal ventricular function and exercise test, and absence of arrhythmias associated with symptoms. However, because of the theoretical possibility of producing or increasing aortic regurgitation, high static sports associated with severe static exercise (Classes IIIA, IIIB, and IIIC) which could increase blood pressure to high levels should be discouraged.
- Athletes with more than mild hemodynamic abnormalities or ventricular dysfunction can participate in lowstatic and moderate static/low dynamic competitive sports (Classes IA, IB, 1C, and IIA), provided their exercise test is normal.

Postoperative Fontan Operation: The Fontan operation is characterized by a communication from right atrium to pulmonary artery without an effective right-sided pumping chamber. The operation is used for the long-term palliation of patients with tricuspid atresia or other complex type of single

ventricle. Although many patients are improved clinically after the Fontan operation, they usually have limited exercise capacity, as reflected in reduced cardiac output at rest and with exercise (5). Postoperative arrhythmias have been associated with significant morbidity and mortality. Diagnostic evaluation before sports participation should include chest radiograph, ECG, and exercise testing. If there is a possibility of ventricular dysfunction, echocardiography and/or other modalities to assess ventricular function should be used.

Recommendation: Athletes can participate in low intensity competitive sports (Class IA). Selected individuals can engage in sports of either moderate demand or low static demand if they have normal or near-normal ventricular function, normal or near-normal oxygen saturation, and near-normal exercise tolerance on formal exercise testing.

Ebstein's Anomaly: There is a great deal of variability in the severity of this malformation. Even mild cases may be associated with important arrhythmias. Severe cases can have physical disability and are at increased risk for sudden death with exercise.

Recommendations

- Mild expressions of Ebstein's anomaly without cyanosis, with nearly normal heart size, and with no evidence of arrhythmia can participate in all sports.
- Athletes with tricuspid regurgitation of moderate severity can participate in low intensity competitive sports (Class IA) if there is no evidence of arrhythmia on ambulatory ECG.
- Athletes with severe Ebstein's anomaly are precluded from all competitive sports participation. However, following surgical repair, low intensity competitive sports (Class IA) can be permitted if tricuspid regurgitation is absent or mild, heart size on chest radiograph is not substantially increased, and arrhythmia is not identified on ambulatory ECG and exercise testing. Selected athletes with an excellent hemodynamic result of repairmay be permitted additional participation on an individually assessed basis.

Congenital Coronary Anomalies: Sudden death in young athletes has been associated with the rare but important anomaly in which the left main coronary artery originates anomalously from the anterior right sinus of Valsalva, makes an acute angled bend, and courses between the pulmonary trunk and theanterior aspect of the aorta (3,18). Rare cases of anomalous right coronary from the left coronary sinus, congenitally hypoplastic coronary arteries, and anomalous origin of the left coronary artery from the pulmonary trunk have also been associated with sudden cardiac death during exercise. Identification of these anomalies during life is unreliable because patients usually do not experience warning symptoms, and noninvasive diagnosis is difficult. These abnormalities should be considered in athletes with exertional syncope or near-fatal arrhythmia, and investigated with echocardiography, exercise testing, and ultimately coronary arteriography if indicated.

Recommendations:

- Detection of these abnormalities should result in exclusion from all competitive sports participation. Surgical treatment (when feasible) probably decreases the risk for sudden death.
- Sports participation ≥6 months after surgery wouldbe permitted for an individual without ischemia during maximal exercise testing.
- Athletes with prior myocardial infarction should follow appropriate recommendations in Task Force 5,Coronary Artery Disease.

The discussion of coronary arterial myocardial bridging appears in Task Force 5, Coronary Artery Disease.

Marfan's Syndrome: Marfan's syndrome is characterized by arachnodactyly, tall stature, pectusexcavatum, kyphoscoliosis, and lenticular dislocation. The cardiovascular manifestations are mitral valve prolapse and aortic dilatation. The risk of aortic regurgitation and aortic dissection is greatest with severe aortic dilatation, but dissection (especially of the distal aorta) can occur in patients with only mild aortic dilatation. Aortic dissection is a known cause of sudden death in competitive athletes (18). Diagnosis is suspected by skeletal features and confirmed by eye examination and echocardiography (to evaluate the magnitude of aortic dilatation).

Recommendations:

- Athletes without a family history of premature sudden death and without evidence of aortic root dilatation or mitral regurgitation can participate in moderate low static and low dynamic competitive sports (Classes IAand IIA). Echocardiographic measurement of aortic root dimension should be repeated every 6 months for continued sports participation.
- Athletes *with* aortic root dilatation can participate only in low intensity competitive sports (Class IA).
- Athletes with Marfan's syndrome should not participate in sports with the risk of bodily collision.
- See Task Force 2, Acquired Valvular Heart Disease,for criteria related to valvular aortic regurgitation in Marfan's syndrome.

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