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**Appendix 1. Author Relationships With Industry and Others**

Name	Consultant	Research Grant	Speaker's Bureau	Stock Holder	Expert Witness Testimony
Dr. Barry J. Maron	None	• Medtronic	None	None	• 1996, Defense, Northwestern vs. Knapp
Dr. Pamela S. Douglas	None	None	None	None	None
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## Task Force 2: Congenital Heart Disease

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### GENERAL CONSIDERATIONS

The most common congenital heart lesions that have been associated with sudden death during sports participation are hypertrophic cardiomyopathy, coronary artery anomalies, Marfan syndrome, and aortic valve disease (1–3). Less common lesions include complex defects, such as transposition and single ventricle, and those with associated pulmonary vascular disease.

The recommendations presented are intended to provide broad guidelines for patients with congenital heart defects (4). When questions about the safety of sports participation arise, there is no substitute for a comprehensive evaluation by a knowledgeable and experienced physician. Exercise testing can be useful, particularly if symptoms, the electrocardiogram (ECG), and blood pressure are monitored during conditions that simulate the sport in question. Arrhythmias discussed in this Task Force are usually identified by exercise testing or some form of long-term monitoring (including ambulatory Holter and event recording). Serial evaluations may be required because of changing hemodynamic status with time.

### TYPES OF CONGENITAL DEFECTS

**Atrial septal defect (ASD)—untreated.** Most children with ASD are asymptomatic, and closure is usually carried out before they are active in competitive sports. An ECG and echocardiogram are required for evaluation. Small atrial defects are characterized by minimal or no right ventricular volume overload, moderate or large defects have significant volume overload but pulmonary hypertension is unusual.

*Recommendations:*

1. **Athletes with small defects, normal right heart volume, and no pulmonary hypertension can participate in all sports.**
2. **Athletes with a large ASD and normal pulmonary artery pressure can participate in all competitive sports.**
3. **Athletes with an ASD and mild pulmonary hypertension can participate in low-intensity competitive sports (class IA). Patients with associated pulmonary vascular obstructive disease who have cyanosis and a large right-to-left shunt cannot participate in competitive sports.**

4. **Athletes with ASD and symptomatic atrial or ventricular tachyarrhythmia or moderate-to-severe mitral regurgitation should follow the recommendations in Task Force 1: Preparticipation Screening and Diagnosis of Cardiovascular Disease in Athletes, and Task Force 6: Coronary Artery Disease, respectively.**

**ASD—closed at operation or by interventional catheterization.** The ASDs usually are completely closed by operation or device insertion. When closure is performed in childhood there is little or no residual right ventricular enlargement. Supraventricular arrhythmias can occur after closure and are more common when the defect is repaired later in life (5–7). Evaluation should include an estimate of cardiac performance, pulmonary vascular resistance and right ventricular size, and a search for conduction or rhythm disturbances. A chest radiograph, ECG, and echocardiogram usually are needed. Patients with preoperative pulmonary hypertension, a right-to-left shunt, or both, before operation require postoperative assessment of pulmonary artery pressure with Doppler echocardiography or cardiac catheterization.

*Recommendations:*

1. **Three to six months after the operation or intervention, patients can participate in all sports unless the following are present: 1) evidence of pulmonary hypertension; 2) symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree heart block; and 3) evidence of myocardial dysfunction.**
2. **Patients with any of these abnormalities should have an exercise evaluation and an individualized exercise prescription with respect to competitive sports. For related recommendations on competitive sports participation, see the section entitled Elevated Pulmonary Resistance and Ventricular Dysfunction After Cardiac Surgery in this Task Force report, as well as Task Force 7: Arrhythmias.**

**Ventricular septal defect (VSD)—untreated.** The VSDs are categorized as small, moderate, or large. If physical examination and echocardiography indicate normal heart size and normal pulmonary artery pressure in patients with a suspected small VSD, further evaluation is not required.

A patient with a VSD that does not fit into the small defect category may require further investigation. Cardiac catheterization might be required.

A moderate-sized defect with low pulmonary vascular resistance will have a pulmonary/systemic flow ratio of about 1.5 to 1.9. A large defect with low or mildly increased pulmonary vascular resistance is defined as a pulmonary/systemic flow ratio greater than or equal to 2 and pulmonary resistance less than 3 U/m<sup>2</sup>. Large right-to-left shunts are discussed in the section entitled Elevated Pulmonary Resistance.

*Recommendations:*

1. **Athletes with a VSD and normal pulmonary artery pressure can participate in all sports.**
2. **Athletes with a large VSD who do not have marked elevation of pulmonary resistance are candidates for repair, and full participation in all sports would normally occur after a successful VSD closure.**

**VSD—closed at operation or by interventional catheterization.** Successful repair is characterized by the absence of symptoms, absence of significant shunt, cardiomegaly, or arrhythmias, and the presence of normal pulmonary artery pressure. Minimal diagnostic evaluation after surgery before participation in sports includes a chest radiograph, ECG, and echocardiogram. Patients with residual left or right ventricular enlargement, myocardial dysfunction, or pulmonary hypertension may require an exercise test or cardiac catheterization before a decision for participation in sports can be made.

*Recommendations:*

1. **At three to six months after repair, asymptomatic athletes with no defect or only a small residual defect can participate in all competitive sports if they have no evidence of pulmonary artery hypertension, ventricular or atrial tachyarrhythmia, or myocardial dysfunction.**
2. **Athletes with symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree atrioventricular (AV) block should follow the recommendations in Task Force 7: Arrhythmias. Athletes with mild-to-moderate pulmonary hypertension or ventricular dysfunction should follow the recommendations in the section entitled Elevated Pulmonary Resistance or Ventricular Dysfunction After Cardiac Surgery.**
3. **Athletes with persistent, severe pulmonary hypertension cannot participate in competitive sports (see section entitled Elevated Pulmonary Resistance).**

**Patent ductus arteriosus (PDA)—untreated.** The patient with a small PDA has a characteristic murmur, absence of symptoms, and normal left heart chamber dimensions. Those with a larger patent ductus have cardiomegaly and widened pulse pressure. There may be evidence of pulmonary hypertension. Minimal diagnostic studies generally include echocardiography.

*Recommendations:*

1. **Athletes with a small PDA and normal left heart chamber dimension can participate in all competitive sports.**
2. **Athletes with a moderate or large PDA, causing left ventricular (LV) enlargement, should undergo surgical or interventional catheterization closure before unrestricted competition.**

**Table 1.** Definitions of Mild, Moderate, and Severe Aortic Stenosis

	Peak-to-Peak Systolic Gradient (Catherization)	Mean Echo Doppler Gradient (CW)	Peak Instantaneous Echo Doppler Gradient (CW)*
Mild AS	less than 30 mm Hg	less than 25 mm Hg	less than 40 mm Hg
Moderate AS	30 to 50 mm Hg	25 to 40 mm Hg	40 to 70 mm Hg
Severe AS	greater than 50 mm Hg	greater than 40 mm Hg	greater than 70 mm Hg

\*Gradients obtained from apical window usually most predictive of catheter gradient.

3. For athletes with a moderate or large PDA, severe pulmonary hypertension, and cyanosis, see the section entitled Elevated Pulmonary Resistance.

**PDA, closed at operation or by interventional catheterization.** A successful result is characterized by the absence of symptoms, a normal cardiac examination, and normal echo/Doppler study.

**Recommendations:**

1. Three months after PDA closure, patients with no symptoms, with normal cardiac examination, and with no evidence of pulmonary hypertension or LV enlargement can participate in all competitive sports.
2. For athletes with residual pulmonary artery hypertension see the section entitled Elevated Pulmonary Resistance.

**Pulmonary valve stenosis (PS)—untreated.** Mild stenosis is characterized by a systolic ejection murmur, variable ejection click, and a normal ECG. A Doppler peak instantaneous gradient less than 40 mm Hg usually indicates mild stenosis, 40 to 60 mm Hg moderate stenosis, and greater than 60 mm Hg severe stenosis. Most patients with a gradient 50 mm Hg or greater undergo balloon valvuloplasty.

**Recommendations:**

1. Athletes with a peak systolic gradient less than 40 mm Hg and normal right ventricular function can participate in all competitive sports if no symptoms are present. Annual re-evaluation is recommended.
2. Athletes with a peak systolic gradient greater than 40 mm Hg can participate in low-intensity competitive sports (classes IA and IB). Patients in this category usually are referred for balloon valvuloplasty or operative valvotomy before sports participation.

**PS—treated by operation or balloon valvuloplasty.** Adequate relief of PS is characterized by the absence or improvement in symptoms, an improved physical examination, and Doppler study showing mild or no residual gradient and mild or no pulmonary valvular regurgitation.

**Recommendations:**

1. Athletes with no or only residual mild PS and normal ventricular function without symptoms can participate in all competitive sports. Participation in sports can begin two to four weeks after balloon valvulo-

plasty. After operation, an interval of approximately three months is suggested before resuming sports participation.

2. Athletes with a persistent peak systolic gradient greater than 40 mm Hg should follow the same recommendations as those for patients before treatment.
3. Athletes with severe pulmonary incompetence characterized by a marked right ventricular enlargement can participate in class IA and IB competitive sports.

**Aortic valve stenosis (AS)—untreated.** This section discusses congenital valvular AS in young patients; AS in adults is addressed in Task Force 3. Differentiation between mild and either moderate or severe AS is accomplished by physical examination, ECG, and Doppler echocardiography. The distinction between moderate or severe stenosis is more difficult and may require cardiac catheterization in rare situations when clinical examination, ECG, echocardiography, and/or other data are discrepant. Patients with a history of fatigue, light-headedness, dizziness, syncope, chest pain, or pallor on exercise deserve a full evaluation, which may include cardiac catheterization and exercise testing. Because AS may progress, periodic re-evaluation is needed. Sudden death is more likely to occur in patients with severe LV hypertrophy, exertional syncope, chest pain or dyspnea and a LV strain pattern on the ECG. Between approximately 20% and 80% of sudden deaths in patients with severe AS have been found to occur on physical exertion (2,8).

For the purpose of these recommendations, Table 1 summarizes the definitions of mild moderate and severe AS. It should be emphasized that virtually all patients classified as moderate or severe AS would be expected to have left ventricular hypertrophy by echocardiography.

**Recommendations:**

1. Athletes with mild AS can participate in all competitive sports if they have a normal ECG, normal exercise tolerance, and no history of exercise-related chest pain, syncope, or atrial or ventricular tachyarrhythmia associated with symptoms.
2. Athletes with moderate AS can participate in low static/low-to-moderate dynamic, and moderate static/low-to-moderate dynamic (classes IA, IB, and IIA) competitive sports if the following conditions are met:
  - Mild or no LV hypertrophy by echocardiography and the absence of LV strain pattern on the ECG.

- Normal exercise test without evidence of myocardial ischemia or atrial or ventricular tachyarrhythmia and with normal exercise duration and blood pressure response. Those athletes with supraventricular tachycardia or multiple or complex ventricular tachyarrhythmias at rest or with exercise can participate only in low-intensity competitive sports, classes IA and IB.
  - Absence of symptoms, as defined in the preceding text.
3. Athletes with severe AS should not participate 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 operation or balloon valvuloplasty.** After operation, a variable degree of residual stenosis or regurgitation, or both, can be present. Re-evaluation by physical examination, ECG, and echocardiography is necessary for reassessment. In addition, exercise stress testing or catheterization, or both, can be required for patients whose physiologic and anatomic severity cannot otherwise be determined.

**Recommendations:**

1. Athletes with residual mild, moderate, or severe stenosis should follow the same recommendations as previously defined for untreated patients.
2. Athletes with moderate to severe aortic regurgitation should follow the recommendations in Task Force 3: Valvular Heart Disease.
3. Because of the propensity for recurrence of LV outflow obstruction in discrete subaortic stenosis postoperatively, these patients require continued follow-up and re-evaluation annually for exercise recommendations. This recommendation also applies to all other forms of fixed AS.

**Coarctation of the aorta—untreated.** This abnormality is characterized by obstruction usually present in the juxtaductal or juxtaligamentary area. Severity is assessed by the arm and leg pressure gradient, physical examination, exercise testing, echocardiographic/Doppler studies, and cardiac magnetic resonance (CMR) studies. Virtually all patients, except those with mild coarctation, will undergo either surgical repair or balloon dilation/stenting.

**Recommendations:**

1. Athletes with mild coarctation and the absence of large collateral vessels or significant aortic root dilation ( $z$ -score 3.0 or less) (score 3.0 = 3 standard deviations from the mean for patient size), with a normal exercise test and a small pressure gradient at rest (usually 20 mm Hg or less between upper and lower limbs), and a peak systolic blood pressure 230

mm Hg or less with exercise can engage in all competitive sports.

2. Athletes with a systolic arm/leg gradient more than 20 mm Hg or exercise-induced hypertension with a systolic blood pressure more than 230 mm Hg can engage in only low-intensity competitive sports (class IA) until treated.

**Coarctation of the aorta—treated by surgery or balloon angioplasty.** The majority of patients will have coarctation repair or balloon arterioplasty performed during childhood. After repair, abnormalities can persist, such as a mild residual gradient, ventricular hypertrophy, systemic hypertension, and residual obstruction evident on exercise (9,10). Before a decision for participation, minimal diagnostic studies, including a chest radiograph, ECG, exercise testing, and echocardiographic evaluation of LV function and aortic anatomy, should be done. Magnetic resonance imaging can be useful to document residual anatomical abnormalities, aortic dilation, or aneurysm formation.

**Recommendations:**

1. Participation in sports, three or more months after surgical or balloon angioplasty for coarctation of the aorta, is permitted for athletes with a 20 mm Hg or less arm/leg blood pressure gradient at rest and a normal peak systolic blood pressure during rest and exercise.
2. During the first postoperative year, athletes should refrain from high-intensity static exercise (classes IIIA, IIIB, and IIIC) and sports that pose the danger of bodily collision.
3. After three months, if patients continue to be asymptomatic, with normal blood pressure at rest and exercise, all sports are permissible except those with a large static component (particularly, classes IIIA, IIIB, and IIIC).
4. For athletes with evidence of significant aortic dilation, wall thinning, or aneurysm formation, participation should be restricted to low-intensity competitive sports (classes IA and IB).

**Elevated pulmonary resistance with congenital heart disease.** Patients who have pulmonary vascular disease and congenital heart disease are at risk for sudden death during sports activity. As pulmonary vascular obstruction progresses, these patients develop cyanosis at rest and intense cyanosis with exercise. Although most of these patients self-limit their activity, they should not participate in competitive sports.

Patients who have suspected elevated pulmonary artery pressure after operation or interventional catheterization for shunt lesions should be evaluated by echocardiography and/or cardiac catheterization before engaging in competitive athletics.

*Recommendations:*

1. If pulmonary artery peak systolic pressure is 30 mm Hg or less, athletes can participate in all sports.
2. If pulmonary artery pressure is more than 30 mm Hg, a full evaluation and individual exercise prescription are required for athletic participation.

**Ventricular dysfunction after cardiac surgery.** Left and/or right ventricular dysfunction can occur after surgical treatment of both simple and complex congenital heart diseases and affect exercise performance. Periodic assessment of ventricular function is required for participation in sports because ventricular function may deteriorate over time.

*Recommendations:*

1. For full participation, normal or near-normal ventricular function is required (ejection fraction 50% or more).
2. Athletes with mildly depressed ventricular function (ejection fraction 40% to 50%) should participate in low-intensity static competitive sports only (classes IA, IB, and IC).
3. Athletes with moderately to severely depressed ventricular function (ejection fraction less than 40%) should not participate in competitive sports.

**Cyanotic congenital cardiac disease—unoperated.** Cyanotic congenital heart disease is associated with exercise intolerance and progressive hypoxemia with increasing effort. Patients are unlikely to engage in competitive sports because of their own self-limiting activity. There are rare patients with cyanotic congenital heart disease (such as pulmonary stenosis or mildly elevated pulmonary vascular resistance plus atrial or ventricular defects) who reach adolescence or even adult life with mild cyanosis at rest and shortness of breath only with exercise. These patients may experience a profound increase in cyanosis during sports participation.

*Recommendation:*

1. Patients with untreated cyanotic heart disease can usually participate in low-intensity competitive sports of only class IA.

**Postoperative palliated cyanotic congenital heart disease.** Palliative surgical intervention 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:*

1. Patients can participate in low-intensity competitive sports (class IA), provided that the following criteria are met:

- Arterial saturation remains above approximately 80%
- Tachyarrhythmias associated with symptoms of impaired consciousness not present
- There is not moderate/severe ventricular dysfunction.

**Postoperative tetralogy of Fallot (T/F).** The current treatment for T/F is early repair, and patients have varying degrees of pulmonary stenosis and mild/moderate pulmonary insufficiency. Diagnostic evaluation usually includes physical examination, chest radiograph, echocardiography, CMR, ambulatory ECG monitoring, and exercise testing. Cardiac catheterization and/or exercise testing may be required for complete evaluation in selected patients, particularly those with significant cardiomegaly and/or symptoms. Patients with important residual abnormalities, such as a significant left-to-right shunt, right ventricular hypertension, moderate-to-severe pulmonary regurgitation, or right ventricular dysfunction, who also have a history of syncope and/or arrhythmia, may be at risk for sudden death (11).

*Recommendations:*

1. Athletes with an excellent repair should be allowed to participate in all sports, providing that the following criteria are met:
  - Normal or near-normal right heart pressure
  - No or only mild right ventricular volume overload
  - No evidence of a significant residual shunt
  - No atrial or ventricular tachyarrhythmia abnormality on ambulatory ECG monitoring or exercise testing
2. Patients with marked pulmonary regurgitation and right ventricular volume overload, residual right ventricular hypertension (peak systolic right ventricular pressure greater than or equal to 50% systemic pressure), or atrial or ventricular tachyarrhythmias, should participate in low-intensity competitive sports only (class IA).

**Transposition of the great arteries (TGA)—postoperative Mustard or Senning operation.** Patients who have had atrial repair of TGA can have significant hemodynamic abnormalities including impaired systemic venous return, abnormal right ventricular (systemic ventricular) function, pulmonary stenosis or pulmonary hypertension, abnormalities of pulmonary venous return, tricuspid insufficiency, and significant atrial or ventricular arrhythmias (12). After atrial repair, the right ventricle is subjected to systemic pressure, and because of the intrinsic properties of the right ventricle its reserve is believed to be less than that of the left ventricle. The consequences of hypertrophy and dilation in a trained athlete after an otherwise excellent atrial repair of transposition are unknown. Evaluation before training and competition in moderate- and low-intensity sports should include history and examination, chest radiograph, ECG, echocardiogram, CMR, ambulatory ECG

monitoring, and exercise testing. For patients in whom data are unclear with regard to hemodynamic abnormalities or ventricular function, cardiac catheterization may be necessary.

**Recommendations:**

1. **Selected patients can engage in low and moderate static/low dynamic competitive sports (classes IA and IIA) provided there is:**
  - Mild or no cardiac chamber enlargement on chest radiograph, echocardiography, or CMR
  - No history of atrial flutter, supraventricular tachycardia, or ventricular tachyarrhythmia
  - No history of syncope or other cardiac symptoms
  - A normal exercise test defined as normal duration, workload, heart rate, ECG, and blood pressure response for age and gender.
2. **Patients not in this category require an individualized exercise prescription.**

**Postoperative arterial switch for TGA.** A significant cohort of patients have had arterial switch repair of TGA 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 and neo-aortic root dilation). Only limited exercise data are available.

**Recommendations:**

1. **Athletes with normal ventricular function, normal exercise test, and no atrial or ventricular tachyarrhythmias can participate in all sports.**
2. **Athletes with more than mild hemodynamic abnormalities or ventricular dysfunction can participate in low and moderate static/low dynamic competitive sports (classes IA, IB, IC, and IIA), provided that their exercise test is normal.**

**Congenitally corrected transposition of the great arteries (CCTGA).** Usually, CCTGA is associated with other congenital malformations of the heart, such as ventricular septal defect, pulmonary stenosis, and systemic AV valve abnormalities, which may dictate the level of participation in competitive sports. These patients are at risk for development of supraventricular tachycardia and spontaneous AV block.

**Recommendations:**

1. **Asymptomatic patients with CCTGA without other cardiac abnormalities may be eligible for participation in class IA and IIA sports if there is no systemic ventricle enlargement, no evidence of atrial or ventricular tachyarrhythmia on ambulatory ECG monitoring or exercise testing or normal exercise tests (including normal maximum oxygen consumption for age and gender).**

2. **Periodic re-evaluation is important to detect development of arrhythmias and deterioration of systemic (right) ventricular function and systemic (tricuspid) AV valve regurgitation. In particular, sports with a large static component (classes IIIA, IIIB, IIIC) such as power weightlifting are not recommended.**

**Postoperative Fontan operation.** The Fontan operation is characterized by systemic venous return bypassing the right ventricle. The operation is used for the long-term palliation of patients with tricuspid atresia or other complex types of single ventricle. Although many patients improve clinically after the Fontan operation, they usually have limited exercise capacity and reduced cardiac output at rest and during exercise (13). Postoperative arrhythmias have been associated with significant morbidity and mortality. Diagnostic evaluation before sports participation should include a chest radiograph, ECG, echocardiography or CMR, and exercise testing with oxygen saturations.

**Recommendations:**

1. **Athletes can participate in low-intensity competitive sports (class IA).**
2. **Athletes can engage in class IB sports if they have normal ventricular function and oxygen saturation.**

**Ebstein's anomaly.** A great deal of variability exists in the severity of this malformation, which is characterized by variable degrees of tricuspid regurgitation and right-heart enlargement caused by a malformed and displaced tricuspid valve. Cyanosis may be present due to atrial right-to-left shunting. Even mild cases may be associated with important arrhythmias. Severe cases can be associated with physical disability and increased risk for sudden death with exercise.

**Recommendations:**

1. **Athletes with a mild expression of Ebstein's anomaly without cyanosis, with normal right ventricular size, and with no evidence of atrial or ventricular tachyarrhythmias can participate in all sports.**
2. **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 Holter monitoring other than isolated premature contractions.**
3. **Athletes with severe Ebstein's anomaly are precluded from all sports participation. However, after surgical repair, low-intensity competitive sports (class IA) can be permitted if tricuspid regurgitation is absent or mild, cardiac chamber size on chest radiograph or by echocardiography is not substantially increased, and symptomatic atrial or ventricular tachyarrhythmias are not present on ambulatory ECG monitoring and exercise test. Selected athletes with an excellent hemodynamic result after repair may be permitted additional participation on an individual basis.**

**Congenital coronary artery anomalies.** Congenital coronary anomalies of wrong sinus origin are the second most common cardiovascular cause of sudden death in young athletes (1). The most common of these malformations is anomalous origin of the left main coronary artery from the anterior (right) sinus of Valsalva, with an acute angled bend coursing between the pulmonary trunk and the anterior aspect of the aorta (14,15). Rare cases of anomalous origin of the right coronary artery from the left coronary sinus, congenitally hypoplastic coronary arteries, and anomalous origin of the left main coronary artery from the pulmonary trunk have also been associated with sudden cardiac death during exercise (1). Identification of these anomalies during life can be difficult because patients often do not experience warning symptoms, and rest and exercise ECGs are usually normal. Coronary anomalies should be considered in athletes with exertional syncope or symptomatic ventricular arrhythmia and should be investigated using appropriate studies such as echocardiography, CMR, or ultrafast computed tomography imaging. Coronary arteriography is indicated if other studies are not diagnostic. Surgery is usually performed when the diagnosis is made (16).

**Recommendations:**

1. **Detection of coronary anomalies of wrong sinus origin in which a coronary artery passes between great arteries should result in exclusion from all participation in competitive sports.**
2. **Participation in all sports three months after successful operation would be permitted for an athlete without ischemia, ventricular or tachyarrhythmia, or dysfunction during maximal exercise testing.**
3. **Athletes with previous myocardial infarction (MI) should follow the appropriate recommendations in Task Force 6: Coronary Artery Disease.**

A discussion of coronary artery myocardial bridging appears in Task Force 6: Coronary Artery Disease.

**Kawasaki disease.** Kawasaki disease, an acute, self-limited vasculitis of unknown etiology, is now the most common cause of acquired heart disease in children in the U.S. (17). Coronary artery aneurysms develop in approximately 20% of untreated children and in 4% of those treated with high-dose intravenous gammaglobulin in the acute phase (18). Coronary aneurysms, together with progressive coronary artery stenosis, can lead to ischemic heart disease, MI, or sudden death (19). Because coronary artery morphology evolves over time, the risk of exercise for the individual patient may change. Patients without coronary artery changes on echocardiography at any stage of the illness appear to have risk for ventricular tachyarrhythmias and sudden death similar to that of the normal population in the first 20 years after illness onset (19). When aneurysms regress to normal lumen diameter, structural and functional coronary abnormalities persist (20). Arteries with persistent

aneurysmal morphology may develop stenoses or occlusion, increasing the risk of myocardial ischemia (21). The risk associated with competitive sports in individuals who have had Kawasaki disease depends upon the degree of coronary artery involvement. Of note, because of the general cardiovascular benefits of exercise, all patients with Kawasaki disease should avoid a sedentary lifestyle.

**Recommendations:**

1. **Patients with no coronary artery abnormalities or transient coronary artery ectasia resolving during the convalescent phase of the disease are encouraged to participate in all sports after six to eight weeks.**
2. **Patients with regressed aneurysms can participate in all competitive sports if they have no evidence of exercise-induced ischemia by stress testing with myocardial perfusion imaging.**
3. **Patients with isolated small- to medium-sized aneurysms in one or more coronary arteries and judged to be at low risk for ischemic complications (normal left ventricular function, absence of exercise-induced ischemia or arrhythmia) may participate in low to moderate static and dynamic competitive sports (classes IA, IB, IIA, and IIB). Stress testing with evaluation of myocardial perfusion should be repeated at one- to two-year intervals to monitor ischemia and guide further recommendations about sports competition.**
4. **Patients with one or more large coronary aneurysms or multiple (segmented) or complex aneurysms with or without obstruction to coronary flow may participate in class IA and IIA sports if they have no evidence of reversible ischemia on stress testing, normal LV function, and absence of exercise-induced arrhythmia. Stress testing with evaluation of myocardial perfusion should be repeated at one-year intervals to monitor ischemia and guide further recommendations about sports competition.**
5. **Athletes with recent MI or revascularization should avoid competitive sports until their recovery is complete—usually six to eight weeks. Those with normal LV ejection fraction, exercise tolerance, absence of reversible ischemia or myocardial perfusion testing, and absence of exercise-induced arrhythmias can participate in class IA and IB sports. Those with left ventricular ejection fraction less than 40%, exercise intolerance, or exercise-induced ventricular tachyarrhythmias should not participate in competitive sports.**
6. **Patients with coronary lesions who are taking anti-coagulants and/or antiplatelet drugs (aspirin, clopidogrel) should not participate in sports that pose a danger of high speed collision.**

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## Appendix 1. Author Relationships With Industry and Others

Name	Research Grant	Expert Witness Testimony
Dr. David Driscoll	None	None
Dr. Welton M. Gersony	None	None
Dr. Thomas P. Graham	None	None
Dr. Jane W. Newburger	<ul style="list-style-type: none"> <li>• Pfizer</li> <li>• Philips</li> </ul>	<ul style="list-style-type: none"> <li>None</li> <li>None</li> </ul>
Dr. Albert Rocchini	None	None
Dr. Jeffrey Towbin	None	<ul style="list-style-type: none"> <li>None</li> <li>• 2005, Defense, Myocarditis, sudden death in child</li> <li>• 2004, Defense, Myocarditis, sudden death in child</li> <li>• 2004, Defense, Myocarditis, sudden death in child</li> <li>• 2002, Defense, Myocarditis, sudden death in child</li> <li>• 2000, Defense, Myocarditis, sudden death in child</li> <li>• 1996, Plaintiff, Myocarditis, sudden death in professional athlete</li> <li>• 1995, Plaintiff, Myocarditis, sudden death in professional athlete</li> </ul>