Task Force 3: Valvular Heart Disease

Robert O. Bonow, MD, FACC, *Chair*
Melvin D. Cheitlin, MD, FACC, Michael H. Crawford, MD, FACC, Pamela S. Douglas, MD, FACC

GENERAL CONSIDERATIONS

The following valvular disorders are considered here in the context of competitive athletes: mitral stenosis (MS), mitral regurgitation (MR), aortic stenosis (AS), aortic regurgitation (AR), tricuspid regurgitation (TR), tricuspid stenosis, and multivalvular disease. Recommendations are also provided for patients who have undergone valve repair or replacement.

The *diagnosis* of valvular heart disease usually can be made on the basis of the characteristic murmurs and associated findings on physical examination. Estimation of the *severity* of valvular disease is more difficult. In athletes, where secondary gain in denying symptoms is likely, the history may be unreliable. In truly asymptomatic patients, severity can usually be determined from the physical examination and Doppler echocardiography. Additional testing is often not necessary, but the electrocardiogram (ECG) and chest radiograph can also aid in assessing severity, and radionuclide angiography or cardiac magnetic resonance are helpful in assessing left ventricular (LV) function when the echocardiogram is of suboptimal quality.

Doppler echocardiography is the most important diagnostic test for valvular heart disease and is generally reliable in distinguishing the severity of the lesion and assessing hemodynamics and ventricular compensation. Although very sensitive in identifying regurgitant jets, quantitation can be more difficult. Recent guidelines have provided recommendations for quantifying valvular regurgitation (1). Only rarely are cardiac catheterization and angiography necessary to determine the hemodynamic severity of valvular lesions.

Valvular regurgitation is frequently detected in normal subjects, including those without murmurs. With pulsed Doppler echocardiography, TR has been reported in 24% to 96% of normal subjects, MR in 10% to 40%, pulmonic regurgitation in 18% to 92%, and AR in 0% to 33% (2,3). In athletes, the prevalence of valvular regurgitation detected by Doppler is even higher, with at least one regurgitant jet found in greater than 90% of subjects and triple-valve regurgitation in 20% (4). The vast majority of these jets represents a trivial regurgitant volume and is of no clinical significance.

Although considerable information exists concerning the natural history, development of symptoms, importance of LV function, and indications for surgical or catheter-based intervention in patients with valvular heart disease (5), there are few data with regard to the rate of progression of the valvular disease (especially in those exercising vigorously) or factors that influence the rate of progression. The rate of progression of aortic stenosis is highly variable among individuals and difficult to predict (6). Chronic MR may increase in severity with time but is more likely to do so in patients who experience a new chordal rupture and flail mitral leaflet (7). Little is known about the influence of strenuous exertion on the progression of ventricular dysfunction, especially when that exertion is periodic in nature. When valvular disease coexists with another cardiovascular abnormality, such as arrhythmias or coronary artery disease (CAD), recommendations with regard to eligibility for sports should be based on the most restrictive of these guidelines.

The present recommendations with regard to the permitted level of athletic activity are offered only as guidelines. Physicians with knowledge of an individual athlete, including the severity of the lesion and the physiological and psychological response to competition, may liberalize these recommendations in selected instances. The recommendations are for those athletes who are asymptomatic, as symptoms will prevent vigorous exercise in most subjects and, in most circumstances, symptoms represent an indication for valve replacement or repair (5). In situations in which the history may be unreliable (e.g., when secondary gain in denying symptoms is likely), exercise tolerance testing is useful in confirming normal effort tolerance and suitability for the proposed athletic activity.

MITRAL STENOSIS

The etiology of MS is almost always rheumatic. Most patients with significant MS will be sufficiently symptomatic that participation in competitive sports is not an issue, but patients with mild-to-moderate MS may be asymptomatic even with strenuous exercise. Although MS rarely causes sudden death, exercise (with an increase in heart rate and cardiac output) can cause sudden marked increases in pulmonary capillary and pulmonary artery pressures, at times resulting in sudden acute pulmonary edema (8). Furthermore, the long-term effect of repeated exertion-related increases in pulmonary artery wedge and pulmonary artery pressures on the lungs or right ventricle is unknown. The effect of even periodic strenuous exercise on the likelihood of developing atrial fibrillation is also not known. When atrial fibrillation occurs, even patients with mild MS must be anticoagulated. The aforementioned considerations must be understood by the patient and the family in considering participation in strenuous competitive activity. Another problem associated with MS is systemic embolization, occurring most commonly in the presence of atrial fibrillation, but there is no evidence that this potential complication is provoked by strenuous exercise.

Evaluation. Clues regarding the hemodynamic severity of MS may often be obtained from the history, physical examination, ECG, and chest radiograph, but accurate noninvasive assessment of severity requires two-dimensional and Doppler echocardiography (9). In patients with MS and minimal or no symptoms who wish to engage in competitive sports, exercise stress testing should be performed to at least the level of activity approximating the exercise demands of the sport, particularly when there is a question as to the severity of the MS. In addition, pulmonary artery systolic pressure during exercise can be estimated noninvasively by Doppler echocardiography and may be helpful in making a decision as to how much activity is safe, even if the severity of MS in an individual patient is estimated to be only mild (5). If this is not practical or possible, then right heart catheterization with exercise can be performed to measure the pulmonary capillary wedge pressure and pulmonary artery pressure.

Hemodynamic severity of MS can be categorized as follows: *mild* = mitral valve area greater than 1.5 cm^2, exercise pulmonary artery wedge pressure less than or equal to 20 mm Hg, or rest pulmonary artery systolic pressure less than 35 mm Hg; *moderate* = mitral valve area 1.0 to 1.5 cm^2, exercise pulmonary artery wedge pressure less than or equal to 25 mm Hg, or rest pulmonary artery systolic pressure less than or equal to 50 mm Hg; *severe* = mitral valve area less than 1.0 cm^2, exercise pulmonary artery wedge pressure greater than 25 mm Hg, or rest pulmonary systolic pressure greater than 50 mm Hg.

Recommendations:

1. **Athletes with mild MS (as previously defined) in sinus rhythm with peak pulmonary artery systolic pressure during exercise less than 50 mm Hg can participate in all competitive sports. Athletes with mild MS in atrial fibrillation, or a history of atrial fibrillation, are discussed in recommendation 4 below.**
2. **Athletes with moderate MS (as previously defined), either in sinus rhythm or in atrial fibrillation, with peak pulmonary artery systolic pressure during exercise less than 50 mm Hg can participate in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB [see Table 1 in Task Force Report 8: Classification of Sports]).**
3. **Athletes with severe MS in either sinus rhythm or atrial fibrillation or those with peak pulmonary artery pressure greater than 50 mm Hg during exercise should not participate in any competitive sports.**
4. **Patients with MS of any severity who are in atrial fibrillation or have a history of atrial fibrillation, and who must receive anticoagulation therapy, should not engage in any competitive sports involving the risk for bodily contact (see Task Force Report 8: Classification of Sports) or possible trauma.**

MITRAL REGURGITATION

Mitral regurgitation, unlike MS, has a variety of etiologies, the most common of which is mitral valve prolapse (myxomatous mitral valve). Other common causes are rheumatic heart disease, infective endocarditis, CAD, connective tissue diseases (such as Marfan syndrome), and dilated cardiomyopathy. The recommendations outlined in this section are for patients with primary valvular MR rather than MR secondary to CAD or other conditions causing LV dilation (see Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome, and Task Force 6: Coronary Artery Disease).

Evaluation. Mitral regurgitation can be detected by the characteristic physical findings and confirmed by Doppler echocardiography. The severity of the MR is related to the magnitude of the regurgitant volume, which results in LV dilation and increased left atrial pressure. The increased LV diastolic volume enhances total LV stroke volume enough to accommodate the regurgitant volume and to maintain the forward stroke volume within normal limits. The low impedance presented by regurgitation into the left atrium unloads the left ventricle during ventricular systole, such that measures of LV pump function (e.g., ejection fraction) tend to overestimate true myocardial performance (10).

The severity of chronic MR can be adequately judged by noninvasive techniques, principally two-dimensional and Doppler echocardiography. The larger the jet area, and the wider the jet at its origin above the valve, the more severe the regurgitation. The entry of the jet into the atrial appendage or pulmonary vein or systolic reversal of the flow in pulmonary veins are all indicators of severity. Various measures of the severity of MR have been described (1,11). In some patients with eccentric jets or those impinging on the atrial wall, the assessment may be difficult (1). Generally, the LV diastolic volume reflects the severity of chronic MR. *However, it should be noted that the upper limit of normal LV size is increased in the healthy, highly trained athlete. In a series of elite athletes, echocardiographic LV end-diastolic dimensions as high as 66 mm were recorded in women (mean, 48 mm) and up to 70 mm in men (mean, 55 mm)* (12). *LV end-diastolic dimensions greater than or equal to 55 mm were observed in 45% of participants and greater than 60 mm in 14% of participants. Therefore, assessment of LV enlargement in a highly trained athlete with known or suspected valvular heart disease must take this issue into consideration.* Hence, for purposes of this discussion, an LV end-diastolic dimension greater than 60 mm is considered likely to represent the effects of LV volume overload due to valvular disease and not per se to physiologically based exercise training.

Patients with chronic MR should be followed longitudinally with serial echocardiograms (5). A decrease in ejection fraction and/or increase in end-systolic volume with time is a helpful marker of declining LV function and an indication of having reached the limits of cardiac compensation.

Effects of exercise. In general, exercise produces no significant change or a mild decrease in the regurgitant fraction because of reduced systemic vascular resistance. However, patients with elevation of heart rate (increased systolic ejection time per min) or blood pressure with exercise may manifest marked increases in regurgitant volume and pulmonary capillary pressures. Hence, static exercise that increases arterial pressure is potentially deleterious. Ejection fraction usually does not change or decreases slightly with exercise, although the ejection fraction response may be completely normal in young, asymptomatic subjects.

The etiology of MR may be important in making recommendations concerning heavy physical activity. In patients with MR secondary to previous infective endocarditis or ruptured chordae, the valve tissues theoretically could be further damaged or torn by marked sustained increases in LV systolic pressure.

Recommendations:

1. **Athletes with mild to moderate MR who are in sinus rhythm with normal LV size and function and with normal pulmonary artery pressures can participate in all competitive sports.**
2. **Athletes with mild to moderate MR in sinus rhythm with normal LV systolic function at rest and mild LV enlargement (compatible with that which may result solely from athletic training [less than 60 mm {12}]) can participate in some low and moderate static and low, moderate, and high dynamic competitive sports (classes IA, IB, 1C, IIA, IIB, and IIC).**
3. **Athletes with severe MR and definite LV enlargement (greater than or equal to 60 mm [12]), pulmonary hypertension, or any degree of LV systolic dysfunction at rest should not participate in any competitive sports.**
4. **Patients in atrial fibrillation or a history of atrial fibrillation who are receiving long-term anticoagulation should not engage in sports involving any risk for bodily contact (see Task Force 8: Classification of Sports) or danger of trauma.**

AORTIC STENOSIS

The diagnosis of AS is established by the characteristic physical findings and two-dimensional and Doppler echocardiography. The three most common etiologies are: 1) rheumatic, 2) congenital, and 3) calcific or degenerative. The majority of young adults with AS participating in competitive athletics have congenital lesions.

Evaluation. Continuous-wave Doppler echocardiography can reliably estimate the severity of AS, especially in the presence of normal cardiac output, which is the case in the great majority of those engaging in competitive sports (13).

Symptoms of dyspnea, syncope, or angina pectoris occur late in the course of AS (14), and the likelihood of sudden death increases significantly with the onset of symptoms. Because even transient symptoms are so important in marking the onset of increased risk of sudden death, the physician must be aware that dyspnea, near-syncope, and even syncope are likely to be unreported in competitive athletes. Although sudden death is more frequent in symptomatic patients with severe AS, it may also occur in completely asymptomatic patients (15). When doubt persists with regard to the severity of AS after Doppler study, or if a patient with mild-to-moderate AS has symptoms, cardiac catheterization should be performed. Sudden death is rare with mild AS.

Athletes with a history of syncope, even with mild AS, should be carefully evaluated by a cardiologist. This should include assessment of arrhythmias with exercise. Syncope should be regarded as a possible surrogate for spontaneously aborted sudden death and should be thoroughly investigated (see Task Force 7: Arrhythmias).

Severity of AS measured by continuous-wave Doppler echocardiography (or in those instances previously noted by cardiac catheterization) is categorized as follows with respect to the calculated aortic valve area: *mild* = greater than 1.5 cm^2; *moderate* = 1.0 to 1.5 cm^2; and *severe* = less than or equal to 1.0 cm^2 (5). This translates roughly (assuming that athletes have normal cardiac output) to the estimated mean aortic valve pressure gradient as follows: *mild* = less than 25 mm Hg; *moderate* = 25 to 40 mm Hg; and *severe* = greater than 40 mm Hg (5).

Because AS is often progressive, periodic re-evaluation at least yearly is necessary and should be performed by a physician with expertise in cardiology. This reassessment includes physical examination and Doppler echocardiography, but may require cardiac catheterization in selected patients as previously noted. In addition, Holter monitoring with intense exercise resembling competition is recommended to detect ventricular arrhythmias in patients with AS who wish to participate in competitive athletics.

In patients with AS, a markedly elevated cardiac output or peripheral vascular resistance for sustained periods of time could result in an exaggerated valvular gradient and a marked increase in LV systolic pressure. Given these precautions, the following recommendations can be made.

Recommendations:

1. **Athletes with mild AS can participate in all competitive sports, but should undergo serial evaluations of AS severity on at least an annual basis.**
2. **Athletes with moderate AS can engage in low-intensity competitive sports (class IA). Selected athletes may participate in low and moderate static or low and moderate dynamic competitive sports (classes IA, IB, and IIA) if exercise tolerance testing to at least the level of activity achieved in competition demonstrates satisfactory exercise capacity without symptoms, ST-segment depression or ventricular tachyarrhythmias, and with a normal 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 (class IA).

3. **Patients with severe AS or symptomatic patients with moderate AS should not engage in any competitive sports.**

AORTIC REGURGITATION

Aortic regurgitation has multiple etiologies, as any disease affecting the aortic valve, annulus, or proximal ascending aorta can result in AR. The common etiologies are: 1) congenital bicuspid aortic valve; 2) rheumatic heart disease; 3) infective endocarditis; and 4) aortic root diseases, including Marfan syndrome, ascending aortic aneurysm, aortic dissection, systemic hypertension, and rheumatoid spondylitis. Aortic regurgitation increases LV diastolic volume and stroke volume, which may ultimately lead to LV systolic dysfunction (10). In addition, myocardial oxygen supply/demand imbalance may develop because of the increased wall stress, LV hypertrophy, and reduced diastolic blood pressure (reduced coronary perfusion pressure).

Patients with severe AR may remain asymptomatic and athletic for many years, but angina pectoris, syncope, and ventricular arrhythmias ultimately may appear. Sudden death is rare among asymptomatic patients (less than 0.2% per year [5]) but can occur.

Evaluation. The hemodynamic severity of AR can be assessed noninvasively by physical examination (the severity being reflected by the degree of LV dilation and the peripheral signs of AR), chest radiography, and echocardiography. As noted previously, the upper limit of normal LV end-diastolic size is increased in the healthy, highly trained athlete (12), and this may well affect assessment of LV enlargement in the setting of AR.

Because of the importance of assessing LV function and the size of the aortic root and proximal ascending aorta in determining the etiology of AR, with resulting implications for athletic participation, evaluation by echocardiography is essential. Doppler echocardiography is very sensitive in detecting any degree of AR. Similar to MR, the greatest difficulty arises in differentiating moderate and severe AR. Qualitatively, the width of the regurgitant jet and the proportion of the LV outflow tract occupied by the jet are related to the severity of the AR, as is the slope velocity profile of the diastolic jet. The regurgitant volume can also be measured quantitatively by Doppler methods (1).

The LV function should be assessed serially by two-dimensional echocardiography (5). Radionuclide angiography or cardiac magnetic resonance may be helpful if echocardiograms are of suboptimal quality. Exercise testing can be useful in assessing exercise capacity, especially in those patients having nonspecific or mild symptoms, and it is recommended that testing be performed to at least the level of exertion required by the proposed competitive sport. Holter monitoring with intense exercise resembling competition is recommended to detect ventricular arrhythmias in patients who wish to participate in competitive athletics.

Effects of exercise. With exercise, regurgitant volume decreases because of the decrease in peripheral vascular resistance that reduces diastolic blood pressure and the decrease in diastolic filling period that accompanies the increase in heart rate (16). Because of these changes in preload and afterload, the failure of the ejection fraction to increase with exercise is of uncertain significance, and there are insufficient data with which to use this finding in formulating recommendations regarding participation in competitive athletics. There are also no data to define whether severe increases in physical activity permanently affect the function of the left ventricle.

For purposes of the following recommendations, hemodynamic severity of AR is graded as follows: *mild* = absent to slight peripheral signs of AR, normal LV size; *moderate* = peripheral signs of AR with mild-to-moderate increases in LV size with normal systolic function; and *severe* = peripheral signs of AR with severe LV enlargement and/or LV systolic dysfunction.

Recommendations:

1. **Athletes with mild or moderate AR, but with LV end-diastolic size that is normal or only mildly increased, consistent with that which may result solely from athletic training (12), can participate in all competitive sports. In selected instances, athletes with AR and moderate LV enlargement (60 to 65 mm) can engage in low and moderate static and low, moderate, and high dynamic competitive sports (classes IA, IB, 1C, IIA, IIB, and IIC) if exercise tolerance testing to at least the level of activity achieved in competition demonstrates no symptoms or ventricular arrhythmias. Those with asymptomatic nonsustained ventricular tachycardia at rest or with exertion should participate in low-intensity competitive sports only (class IA) (see also Task Force 7: Arrhythmias).**
2. **Athletes with severe AR and LV diastolic diameter greater than 65 mm as well as those with mild or moderate AR and symptoms (regardless of LV dimension) should not participate in any competitive sports.**
3. **Those with AR and significant dilation of the proximal ascending aorta (greater than 45 mm) can engage only in low-intensity competitive sports (class IA). These criteria do not apply to athletes with Marfan syndrome and AR, in whom the risks of aortic dissection and rupture are high, and any degree of aortic dilatation would be sufficient to prohibit competitive athletics, as discussed in Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome.**

JACC Vol. 45, No. 8, 2005
April 19, 2005:1334–40

BICUSPID AORTIC VALVES WITH AORTIC ROOT DILATATION

There is growing awareness that many patients with bicuspid aortic valves have disorders of vascular connective tissue, involving loss of elastic tissue, that may result in aortic root dilatation even in the absence of hemodynamically significant AS or AR (17,18). These patients have a risk of aortic dissection; surgery to repair the aorta has been recommended for those patients with greatly enlarged aortic roots (5,18). Recommendations for athletic participation in patients with bicuspid valve disease and associated aortic root dilatation are based on limited data, but with the understanding that aortic dissection can occur in some patients with aortic root diameters less than 50 mm. The recommendations that follow are for patients with bicuspid valves and associated aortic root enlargement. If such patients also have significant AS or AR, these recommendations should be considered in concert with those discussed in the present Task Force for patients with AS or AR. The following recommendations do not pertain to patients with Marfan syndrome, which are discussed in Task Force 4.

Recommendations:

1. **Patients with bicuspid aortic valves with no aortic root dilatation (less than 40 mm or the equivalent according to body surface area in children and adolescents) and no significant AS or AR may participate in all competitive sports.**
2. **Patients with bicuspid aortic valves and dilated aortic roots between 40 and 45 mm may participate in low and moderate static or low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB), but should avoid any sports in these categories that involve the potential for bodily collision or trauma.**
3. **Patients with bicuspid aortic valves and dilated aortic roots greater than 45 mm can participate in only low-intensity competitive sports (class IA).**

TRICUSPID REGURGITATION

Tricuspid regurgitation is most often secondary to right ventricular (RV) dilation and failure due to pulmonary or RV hypertension. Other etiologies that may affect athletes include rheumatic heart disease, tricuspid valve prolapse, infective endocarditis, congenital anomalies such as Ebstein's anomaly, and the sequelae of surgery for congenital heart disease leaving the patient with RV dilatation. Recommendations here with respect to athletes are for those with primary TR.

Primary TR leads to RV volume overload with the risk of subsequent RV failure as well as increased systemic venous pressure and its consequences. Severity of TR and estimation of right atrial and RV pressures can be determined noninvasively from physical examination, chest radiograph, and Doppler echocardiography. Occasionally, right-heart catheterization is necessary when these measures cannot be determined noninvasively. There is no evidence that the athlete with isolated primary TR is placed in jeopardy by engaging in heavy physical exertion.

Recommendation:

1. **Athletes with primary TR, regardless of severity, with normal RV function in the absence of right atrial pressure greater than 20 mm Hg or elevation of RV systolic pressure can engage in all competitive sports.**

TRICUSPID STENOSIS

Tricuspid stenosis is for the most part caused by rheumatic heart disease and is nearly always associated with MS. In that instance, the patient should be judged according to the severity of the MS. Isolated tricuspid stenosis is rare. Such patients should undergo exercise testing at least to the level anticipated in their sport. If asymptomatic, athletes may compete in all competitive activities.

MULTIVALVULAR DISEASE

Multivalvular disease occurs in the context of rheumatic heart disease, myxomatous valvular disease, and infective endocarditis. The lesions can be diagnosed by physical examination and Doppler echocardiography. The relative contribution of each of the lesions may be difficult to assess noninvasively, and cardiac catheterization and angiocardiography are frequently necessary to resolve these distinctions.

Recommendation:

1. **Multiple lesions of moderate severity may have additive physiologic effects. In general, athletes with significant multiple valvular disease should not participate in any competitive sports.**

PROSTHETIC HEART VALVES

Several general comments apply to all patients who have undergone valve replacement. First, although most patients improve after valve replacement and many become asymptomatic, the long-term mortality after operation is greater than that of a normal population of similar age. Second, a transvalvular gradient of varying severity is present in most patients after valve replacement (5,19). Third, although hemodynamic variables at rest may be essentially normal after valve replacement, many patients have an abnormal response to exercise (20). Finally, after valve replacement (with few exceptions) patients with mechanical prostheses require anticoagulant agents, and those with bioprosthetic valves in sinus rhythm usually do not. In assessing the athlete's capacity for physical activity, exercise stress testing to at least the level of activity performed in the competitive sport is valuable.

Effects of exercise. There are insufficient data to determine whether vigorous repetitive exercise after valve replacement has any long-lasting effect on ventricular or prosthetic valve function. The patient should be made aware of these deficiencies in our knowledge before deciding whether to participate in competitive athletics. Because mechanical and most tissue valves have reduced effective valve areas, they perform best at normal heart rates. Therefore, a sustained heart rate greater than 120 beats/min might result in elevated valve gradients and cardiac outputs that are less than normally expected.

Recommendations:

1. **Athletes with a bioprosthetic *mitral valve* not taking anticoagulant agents and who have normal valvular function and normal or near-normal LV function can participate in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB).**
2. **Athletes with a mechanical or bioprosthetic *aortic valve*, with normal valve function and with normal LV function, can engage in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, and IIA). Athletes participating in greater than low-intensity competitive sports (class IA) should undergo exercise testing to at least the level of activity achieved in competition to evaluate exercise tolerance and symptomatic and hemodynamic responses.**
3. **Independent of other considerations, athletes with a mechanical or bioprosthetic *mitral valve* or *aortic valve* who are taking anticoagulant agents should not engage in sports involving the risk of bodily contact (see Task Force 8: Classification of Sports) or the danger of trauma.**

VALVE REPAIR OR PERCUTANEOUS MITRAL BALLOON VALVOTOMY

Several general comments apply to athletes who have undergone valve repair or percutaneous mitral balloon mitral valvotomy (PMBV). The benefits of surgical mitral valve repair for MR appear to persist long term, but the effects of strenuous exercise on repaired mitral valves have not been studied systematically. After treatment for MS with PMBV or either closed or open surgical mitral commissurotomy, the patient should have no disability if there has been no injury to the left ventricle or development of significant MR. After aortic valvuloplasty in young patients with congenital AS, there is the risk of subsequent endocarditis, AR, or recurrent AS. Athletes should be evaluated for competitive sports on the basis of their residual hemodynamic state.

Recommendations:

1. **For patients with MS who have undergone successful PMBV or surgical commissurotomy, recommendations for participation in competitive sports should be based on the residual severity of the MS or MR, as in patients without operation. Capacity to engage in physical exercise should be evaluated with an exercise tolerance test at least to the level of anticipated activity. Patients with LV dysfunction should be restricted from athletic participation in the same context as those without operation.**
2. **Athletes who have undergone mitral valve repair for MR should not engage in sports involving the risk or likelihood of bodily contact (see Task Force 8: Classification of Sports) or possible trauma, which might disrupt the repair. They can participate in low-intensity competitive sports (class IA) and, in selected athletes, in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, and IIA).**

doi:10.1016/j.jacc.2005.02.010

TASK FORCE 3 REFERENCES

1. Zoghbi WA, Enriquez-Sarano M, Foster E, et al. Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography. J Am Soc Echocardiogr 2003;16:777–802.
2. Kostucki W, Vandenbossche JL, Friart A, Englert M. Pulsed Doppler regurgitant flow patterns of normal valves. Am J Cardiol 1986;58:309–13.
3. Richards KL, Cannon SR, Crawford MH, Sorensen SG. Noninvasive diagnosis of aortic and mitral valve disease with pulsed-Doppler spectral analysis. Am J Cardiol 1983;51:1122–7.
4. Douglas PS, Berman GO, O'Toole ML, Hiller WD, Reichek N. Prevalence of multivalvular regurgitation in athletes. Am J Cardiol 1989;64:209–12.
5. Bonow RO, Carabello B, de Leon AC, et al. ACC/AHA guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Management of Patients With Valvular Heart Disease). J Am Coll Cardiol 1998;32:1486–582.
6. Otto CM, Burwash IG, Legget ME, et al. Prospective study of asymptomatic valvular aortic stenosis: clinical, echocardiographic, and exercise predictors of outcome. Circulation 1997;95:2262–70.
7. Enriquez-Sarano M, Basmadjian AJ, Rossi A, Bailey KR, Seward JB, Tajik AJ. Progression of mitral regurgitation: a prospective Doppler echocardiographic study. J Am Coll Cardiol 1999;34:1137–44.
8. Rahimtoola SH, Durairaj A, Mehra A, Nuno I. Current evaluation and management of patients with mitral stenosis. Circulation 2002;106:1183–8.
9. Faletra F, Pezzano A J, Fusco R, et al. Measurement of mitral valve area in mitral stenosis: four echocardiographic methods compared with direct measurement of anatomic orifices. J Am Coll Cardiol 1996;28:1190–7.
10. Carabello BA. Progress in mitral and aortic regurgitation. Curr Probl Cardiol 2003;28:549–84.
11. Dujardin KS, Enriquez-Sarano M, Bailey KR, Nishimura RA, Seward JB, Tajik AJ. Grading of mitral regurgitation by quantitative Doppler echocardiography: calibration by left ventricular angiography in routine clinical practice. Circulation 1997;96:3409–15.
12. Pelliccia A, Culasso F, Di Paolo FM, Maron BJ. Physiologic left ventricular cavity dilatation in elite athletes. Ann Intern Med 1999;130:23–31.
13. Galan A, Zoghbi WA, Quinones MA. Determination of severity of valvular aortic stenosis by Doppler echocardiography and relation of findings to clinical outcome and agreement with hemodynamic mea-

surements determined at cardiac catheterization. Am J Cardiol 1991;67:1007–12.
14. Carabello BA. Evaluation and management of patients with aortic stenosis. Circulation 2002;105:1746–50.
15. Rosenhek R, Binder T, Porenta G, et al. Predictors of outcome in severe, asymptomatic aortic stenosis. N Engl J Med 2000;343: 611–7.
16. Dehmer GJ, Firth BG, Hillis LD, et al. Alterations in left ventricular volumes and ejection fraction at rest and during exercise in patients with aortic regulation. Am J Cardiol 1981;48:17–27.
17. Nistri S, Sorbo MD, Marin M, Scognamiglio R, Thiene G. Aortic root dilatation in young men with normally functioning bicuspid aortic valves. Heart 1999;82:19–22.
18. Svensson LG, Kim KH, Lytle BW, Cosgrove DM. Relationship of aortic cross-sectional area to height ratio and the risk of aortic dissection in patients with bicuspid aortic valves. J Thorac Cardiovasc Surg 2003;126:892–3.
19. Rahimtoola SH. Choice of prosthetic heart valve for adult patients. J Am Coll Cardiol 2003;41:893–904.
20. Borer JS, Herrold EM, Hochreiter C, et al. Natural history of left ventricular performance at rest and during exercise after aortic valve replacement for aortic regurgitation. Circulation 1991;84:III133–9.

Appendix 1

The authors of this report declared they have no relationships with industry pertinent to this topic.

Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome

Barry J. Maron, MD, FACC, *Chair*
Michael J. Ackerman, MD, PhD, FACC, Rick A. Nishimura, MD, FACC, Reed E. Pyeritz, MD, PhD, Jeffrey A. Towbin, MD, FACC, James E. Udelson, MD, FACC

HYPERTROPHIC CARDIOMYOPATHY

General considerations. Hypertrophic cardiomyopathy (HCM) is a relatively common form of genetic heart disease (0.2%; 1:500 in the general population) (1), and the most common cause of sudden unexpected cardiac death in young people, including competitive athletes (2). Sudden death may occur at any age, but is most common in individuals 30 years of age or less. At present, 12 mutant genes (most encoding sarcomeric proteins) and over 400 specific mutations in these genes have been implicated in the pathogenesis of clinically diagnosed HCM (3).

The disease is characterized by heterogeneous presentation and natural history in which the most consistent diagnostic feature demonstrated by echocardiography is otherwise unexplained and usually asymmetric hypertrophy associated with a non-dilated left ventricle (LV) (3–5). Clinical diagnosis of HCM is made by recognition of the disease phenotype with LV hypertrophy (3,4). In this regard, a maximal LV end-diastolic wall thickness of 15 mm or more (or on occasion, 13 or 14 mm) is the absolute dimension generally accepted for the clinical diagnosis of HCM in an adult athlete (in children, 2 or more standard deviations from the mean relative to body surface are; *z*-score of 2 or more); however, any LV wall thickness (including normal) is theoretically compatible with the presence of a mutant HCM gene (3,4). Of note, individuals of virtually any age (but usually less than 14 years old) harboring a HCM-causing mutant gene may not manifest LV hypertrophy (3,4).

In a disease such as HCM, extrapolation of risk level from non-athletes to highly trained competitive athletes is tenuous. This relates to the unstable electrophysiologic substrate and propensity for potentially lethal ventricular tachyarrhythmias in HCM, interacting with the physiologic stresses inherent in athletic training and competition (i.e., alterations in blood volume, hydration, and electrolytes). Furthermore, no single clinical, morphologic, or electrophysiologic factor has emerged as the reliable predictor of risk in HCM (3,4). Therefore, because the panel could not precisely stratify sudden death risk specifically for all athletes with HCM, the present recommendations for sports eligibility remain conservative and homogeneous for those athletes within the diverse HCM clinical spectrum.

Given the inability to precisely stratify risk on clinical grounds in individual young patients with HCM, a broad recommendation to exclude such individuals from competitive sports will, by definition, deny participation to some unnecessarily. However, given the frequency with which HCM is associated with sudden death in young athletes (2), and recent data showing that athletic activity per se is associated with higher risk in those with underlying cardiovascular abnormalities (6), the present recommendations are viewed as prudent. That is, the goal is to encompass all preventable sudden deaths in young persons with HCM, while acknowledging that other athletes who may not be destined for sudden death will also be subjected to the same recommendations.

Preclinical diagnosis. With the availability of preclinical genetic diagnosis, a relatively small number of youthful family members have been identified as affected by a HCM-causing mutant gene solely on the basis of laboratory