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Valvular Heart Disease (J Dal-Bianco, Section Editor)

# Valvular Heart Disease in Athletes

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## Abstract

*Purpose of Review* Valvular heart disease is prevalent in older athletes with primarily degenerative valvular disorders and younger athletes with congenital or genetic syndromes. Limited data exist on the risks and benefits of exercise for athletes with underlying valvular disorders, so current guidelines are primarily based upon expert consensus. This review focuses on the current data, guideline recommendations, and emerging clinical conundrums for athletes with common valvular heart conditions including aortic stenosis, bicuspid aortic valve (BAV), mitral regurgitation (MR), mitral valve prolapse (MVP), and thoracic aortic aneurysms.

*Recent Findings* Aortic growth appears similar in athletes compared to non-athletes with BAV. Return to exercise following mitral valve repair for primary MR does not seem to lead to significant valve deterioration or adverse outcomes in short-term follow-up. Longitudinal cohort studies of athletes with MVP have suggested that ventricular arrhythmias can be common, but sudden cardiac death is rare. Aortic dilation is uncommon in young otherwise healthy athletes, but can commonly be found in older endurance and strength athletes.

*Summary* Valvular heart conditions in athletes are prevalent in clinical practice; however, there are limited data on the outcomes in these patients to drive guideline development and clinical decision-making. Future research should focus on defining the risks of continual exercise on outcomes in patients with known valvular disease, the optimal time for valve repair/replacement, and the risks of returning to exercise following valvular intervention.

## Introduction

Competitive athletes and highly active people are a growing population that is increasingly encountered in clinical practice. Valvular heart disease is commonly found in older athletes with valvular degeneration and in young athletes with congenital or genetic anomalies. While there has recently been an increasing awareness of the importance of individualized care for athletic populations [1], data on the clinical relevance of valvular heart disease in athletes are lacking. Current 2015 American Heart Association/American College of Cardiology (AHA/ACC) and 2020 European Society of Cardiology (ESC) guidelines provide recommendations on the management of specific valvular disorders in athletic cohorts [2, 3••]. However, virtually all of these recommendations are based on expert consensus rather than peer-reviewed scientific evidence. In this review, we focus on the current data, guideline recommendations, and emerging clinical conundrums for athletes with common valvular heart conditions including aortic stenosis (AS), bicuspid aortic valve (BAV), mitral regurgitation (MR), mitral valve prolapse (MVP), and thoracic aortic aneurysms (TAAs).

# Exercise-induced cardiovascular remodeling

Routine physical activity leads to specific changes in cardiovascular structure and function, which are commonly referred to as exercise-induced cardiovascular remodeling (EICR). Longitudinal studies have shown that EICR is sport-specific, and the typical structural and functional cardiovascular adaptations caused by exercise have been characterized among elite and recreational athlete cohorts [4–14]. In general, endurance training requires a high metabolic demand over long periods of time. To meet this demand, the heart increases cardiac output to ensure adequate blood delivery to metabolically active tissues. Prolonged and repetitive increases in cardiac output represent a volume challenge for the heart, which stimulates biventricular dilation with variable increases in left ventricular (LV) wall thickness as determined by the specific endurance sport discipline (eccentric remodeling and hypertrophy) [15]. In contrast, strength exercise requires brief sequential bursts of high-intensity skeletal muscle contraction which increases systolic blood pressure without significantly increasing cardiac output. Therefore, strength training represents an increased pressure load on the LV and aorta, which leads to increased LV wall thickness without a significant increase in LV chamber dimensions (concentric hypertrophy). While pure dynamic (e.g., cycling) and pure static (e.g., weightlifting) sporting disciplines can each lead to unique forms of EICR, sports often contain a mixture of static and dynamic components (e.g., rowing). Accordingly, the accurate interpretation of cardiac imaging in an athlete, specifically differentiating EICR from pathologic hypertrophy, requires careful consideration of a specific athlete's predominant sporting discipline [16]. The physiologic increase in chamber dimensions secondary to EICR has also been associated with an increased incidence of mild valvular regurgitation. In a study comparing 45 athletes with 26 matched sedentary controls, athletes had an overall higher rate of valvular regurgitation (91% vs. 38%, $p<0.001$), and specifically a higher prevalence of mitral (69% vs. 27%) and tricuspid regurgitation (76% vs. 15%) [17].

## Aortic stenosis

AS is a common form of valvular heart disease that increases in prevalence with age [18, 19]. The most common causes of valvular aortic stenosis include congenital abnormalities (e.g., bicuspid or unicuspid valves), valvular calcification, and rheumatic heart disease. With an aging population, the global morbidity and mortality attributable to calcific aortic stenosis are rising [20]. Calcific AS is a progressive condition characterized by steady increases in LV afterload that parallel reductions in the functional valve orifice area, leading to concentric hypertrophy, diastolic dysfunction, and eventually LV systolic dysfunction. Normally active patients with calcific AS rarely have symptoms until the stenosis becomes severe (aortic valve area < 1 $cm^2$, aortic velocity > 4 ms, and/or mean AV gradient > 40 mmHg). Aortic valve replacement (AVR) is recommended by current ACC/AHA guidelines in symptomatic patients with severe AS, LVEF < 50% with dobutamine stress echo with aortic velocity > 4 ms, or aortic valve area (AVA) < 0.6 $cm^2$ and stroke volume index (SVI) < 35 (class I recommendation) [21••]. For asymptomatic patients with echocardiographic indices consistent with severe AS, current guidelines give a class I indication for AVR in patients with LVEF < 50% or another indication for cardiac surgery, and a class IIa recommendation for patients with an abnormal exercise treadmill test or for patients with low surgical risk in the setting of multiple alternative indications (aortic velocity ≥ 5 ms, brain natriuretic peptide > 3 × normal, or rapid progression of disease) [21••].

Calcific AS is commonly encountered during the care for master's athletes, a patient population that maintains high levels of physical activity into old age. While high levels of physical activity confer a favorable impact on numerous determinants of atherosclerotic heart disease (i.e., plasma lipoprotein levels, systolic blood pressure, glucose metabolism, etc.), it does not appear to reduce the likelihood of valvular heart disease. In a study assessing the impact of physical activity on the prevalence of AS in 69,288 adults (mean follow-up 15.3 years), there was no association between leisure-time exercise and AS (≥ 4 h/week vs. < 1 h/week: hazard ratio 1.18, 95% CI 0.97–1.43) [22]. To date, we are unaware of data defining the temporal progression of calcific AS among master's athletes. In our experience, general population-based estimates of a reduction in functional valve area of 0.1 $cm^2$/year appear to be similarly applicable to master's athletes. Current 2015 AHA/ACC and 2020 ESC disqualification guidelines for athletes have similar recommendations advising no sport restriction for athletes with mild AS, low–moderate-intensity exercise in athletes with moderate AS who have a normal response to exercise (normal BP response, no arrhythmias, and no signs of ischemia), and athletes with severe AS should avoid sports unless asymptomatic then they may consider low-intensity exercise (Table 1) [2, 3]. Despite these guidelines, many asymptomatic master's athletes with moderate to severe calcific AS and no imminent indication for surgical valve replacement will elect to continue with unrestricted exercise. We routinely support this decision

**Table 1 Comparison of the 2015 AHA/ACC vs. 2020 ESC guidelines for the management of athletes with aortic stenosis**

| Severity | 2015 AHA/ACC | 2020 ESC—leisure sport | 2020 ESC—competitive sports |
|---|---|---|---|
| **Mild** | Class IIa, LOE C<br>Athletes with mild AS (stage B) and a normal maximal exercise response can participate in all sports | Class I, LOE C<br>Participation in all recreational sports, if desired, is recommended | Class I, LOE C<br>Participation in all competitive sports, if desired, is recommended |
| **Moderate** | Class IIa, LOE C<br>Athletes with moderate AS (stage B) can 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 and the training regimen demonstrates satisfactory exercise capacity without symptoms, ST-segment depression, or ventricular tachyarrhythmias, and with a normal blood pressure response | Class IIa, LOE C<br>Participation in all recreational sports involving low to moderate intensity, if desired, should be considered in individuals with LVEF≥50%, good functional capacity, and normal exercise test | Class IIb, LOE C<br>Participation in all competitive sports involving low to moderate effort, if desired, may be considered in individuals with LVEF≥50%, good functional capacity, and normal BP response during exercise |
| **Severe** | Class III, LOE C<br>Asymptomatic athletes with severe AS (stage C) should not participate in competitive sports, with the possible exception of low-intensity (class IA) sports | Class IIb, LOE C<br>Participation in all recreational sports/exercise involving low intensity, if desired, may be considered in individuals with LVEF≥50% and normal BP response during exercise | Class IIb, LOE C<br>Participation in low-intensity skill sports may be considered in a select group of individuals with LVEF≥50% |
| | Class III, LOE C<br>Symptomatic patients with AS (stage D) should not participate in competitive sports | Class III, LOE C<br>Participation in competitive or recreational sports/exercise of moderate and high intensity is not recommended | Class III, LOE C<br>Participation in sports or exercise of moderate or high intensity is not recommended |

*AS*, aortic stenosis; ***LOE***, level of evidence; *LVEF*, left ventricular ejection fraction.

when coupled with serial transthoracic echocardiography and maximal effort-limited exercise testing that will provide both the patient and the clinician with a timely opportunity to detect the symptom onset of myocardial pathology.

An emerging clinical conundrum among middle-aged athletes (e.g., 40–65 years old) with a guideline driven indication for surgical aortic valve replacement (AVR) [21••,23] is whether to insert a mechanical or bioprosthetic valve. Traditionally, guidelines have recommended mechanical AVR placement for all patients < 60 years without a clear and conventional contraindication to anticoagulation. The advent of valve-in-valve transcatheter AVR (TAVR) and improved durability with modern heart valves have changed recommendations which now advise mechanical AVR for patients < 50 years, and shared decision-making for bioprosthetic vs. mechanical AVR in patients 50–65 years old [21••]. Careful consideration of the risk associated with lifelong systemic anticoagulation (AC) is an important component of this shared-decision-making discussion with master's athletes. While "contact sports" (i.e., American-style football, hockey, martial arts) pose a significant risk of traumatic bleeding in the setting of AC, ostensibly "non-contact" sports (i.e., cycling, rock climbing, sky diving, etc.) also carry a non-trivial risk of adverse outcomes secondary to bleeding. The valve selection discussion should prioritize consideration of the individual patient's sporting discipline, post-operative athletic goals, risk tolerance, and desire to avoid future valvular interventions. Methods of intermittent AC have been proposed to reduce the risks of bleeding in athletes, particularly in the treatment of venous thromboembolic disease [24]. However, these methods can only be used with the short acting direct oral anticoagulant (DOAC) medications which are not yet in widespread use in the setting of mechanical valve prostheses. Accordingly, athletes with mechanical valve replacements require warfarin-based anticoagulation for which an intermittent dosing program is neither safe nor practical. A limited literature suggests that structured exercise training programs are safe and capable of increasing exercise capacity, muscular strength, and quality of life after valve replacement [25–27]. While some master's athletes choose to retrain independently following surgery, we routinely encourage all to participate in structured cardiac rehabilitation. Maximal effort-limited exercise testing at 3, 6, and 12-month post-operative intervals may provide valuable physiologic and clinical data for the patient and clinician.

## Bicuspid aortic valve

Bicuspid aortic valves are one of the most common congenital heart defects affecting ~ 0.5–2% of the population with an approximately 3:1 male to female predominance [28]. The phenotypes of BAV are highly variable with some patients presenting with 2 aortic valve cusps, and others presenting with 3 cusps and fusion of two of the leaflets. The most common BAV phenotype involves fusion of the right and left coronary leaflets with a single raphe [29]. BAVs most commonly occur as an isolated defect but have also been associated with complex genetic syndromes (i.e., Turner's syndrome, Marfan's

syndrome, Loeys-Dietz syndrome) [30–32] and other congenital heart lesions [33]. Complications arising from BAVs include AS, aortic regurgitation, infective endocarditis, and aortopathy with aneurysmal dilation of the ascending aorta and aortic root. All athletes with BAVs should undergo serial echocardiograms to assess valve function and aortic dimensions [21••,23]. The frequency of surveillance can be determined by the presence and severity of pathology. Current guidelines also recommend screening of all first-degree relatives of athletes diagnosed with BAV. Indications for AVR in patients with BAV without aortopathies are similar to those discussed about for patients with trileaflet aortic valves [21••,23]. At present, surgical AVR continues to represent the preferred standard of care over TAVR for athletes with BAV but emerging data are challenging this paradigm [34, 35].

Aortopathy is common among athletes with BAV. At present, there are limited data characterizing the impact of exercise on aortic dilation and/or valve deterioration in athletes with BAV. Small studies have suggested that there is no significant difference in baseline aortic dimensions or aortic regurgitation between athletes and non-athletes with BAV, and the rate of aortic growth over intermediate follow-up (7 years) appears similar between athletes and non-athletes [36, 37]. In a recent study comparing matched athletes with BAV ($n = 41$), non-athletes with BAV ($n = 41$), and athletes with a tricuspid AV ($n = 41$), athletes with a tricuspid AV had smaller aortic dimensions than both matched athletes and non-athletes with BAV [36]. The only study that has assessed extended duration exercise training was performed by Spataro et al. in 81 Olympic athletes with BAV (73 male, 8 female, $22.7 \pm 5.6$ years) [38]. They divided their cohort into low-risk athletes ($n = 51$) who were allowed to continue training and high-risk athletes ($n = 30$) who were immediately disqualified. Among low-risk athletes, 6/51 (12%) developed symptoms or worsening of bicuspid aortic valve disease (e.g., aortic dilation, aortic stenosis, LV dilation, arrhythmias) over the mean 13-year follow-up. Of the high-risk athletes, 2/11 (18%) with follow-up available (mean 10 years) required AVR for worsening aortic regurgitation with LV dysfunction, and 1 of these athletes had a sudden cardiac death event 1 year after AVR.

Both the 2015 AHA/ACC and 2020 ESC disqualification guidelines for athletes recommend similar management strategies for BAV valvular dysfunction as those proposed for tricuspid AS and aortic regurgitation (Table 1) [2, 3]. Both guidelines recommend against any form of sport restriction for athletes with BAV and aortic root and ascending aorta < 40 mm, but recommend that contact sports should be avoided if aortic dimensions are > 40 mm (Table 2). Guidelines differ however in their recommendations for athletes with BAV and an aortic dimension > 45 mm. The 2015 AHA/ACC guidelines recommend such athletes be restricted from all competitive sports [39], whereas the ESC guidelines recommend that athletes with BAV and aortic dimension 45–50 mm participate in only skill sports or mixed or endurance sports at low intensity [3]. In a recent study assessing the impact of the 2015 AHA/ACC guidelines on 123 pediatric patients with BAV but no genetic syndrome or complex congenital disease (age 5–22 years old), 1/3 of children were restricted from some competitive activity during their school years or adult years [40]. The most common indication for sports restriction was aortic dilation (34%, 42/123) with the majority of patients meeting criteria for mild

**Table 2 Comparison of the 2015 AHA/ACC vs. 2020 ESC guidelines for the management of athletes with thoracic aortic aneurysm in bicuspid aortic valve and hereditary thoracic aortic disease**

| Guideline topic | 2015 AHA/ACC | 2020 ESC |
|---|---|---|
| **Bicuspid aortic valve** | Class I, LOE C<br>Athletes with BAV can participate in all competitive athletics if the aortic root and ascending aorta are not dilated (i.e., z score<2, or<2 standard deviations from the mean, or<40 mm in adults). The function of the BAV (whether stenotic or regurgitant) is also important in determining participation recommendations | Low risk<br>Aorta<40 mm in BAV<br>-All sports permitted with preference for endurance sports over power sports |
| | Class IIb, LOE C<br>For athletes with a BAV and a mild to moderately dilated aorta (z score 2 to 3.5 or aortic root or ascending aortic diameters measuring 40 to 42 mm in men or 36 to 39 mm in women) and no features of associated connective tissue disorder or familial TAA syndrome, participation in low and moderate static and dynamic competitive sports with a low likelihood of significant bodily contact (classes IA, IB, IC, IIA, IIB, and IIC) may be considered. For these athletes, avoidance of intense weight training should be considered | Low–intermediate risk<br>Aorta 40–45 mm in BAV<br>-Avoid high and very high-intensity exercise, contact, and power sports<br>-Preference for endurance over power sports |
| | Class IIb, LOE C<br>For athletes with a BAV and a dilated aorta measuring 43 to 45 mm, participation in low-intensity competitive sports (class IA) with a low likelihood of bodily contact may be considered | Intermediate risk<br>Aorta 45–50 mm in BAV<br>-Only skill sports or mixed or endurance sports at low intensity |
| | Class III, LOE C<br>Athletes with BAV and a severely dilated aorta (score>3.5 to 4 or>43 mm in men or>40 mm in women) should not participate in any competitive sports that involve the potential for bodily collision | High risk<br>Aorta>50 mm in BAV<br>-Sports are (temporarily) contraindicated |
| | Class III, LOE C<br>Athletes with BAV and a markedly dilated aorta (>45 mm) should not participate in any competitive sports | |

**Table 2** (continued)

| Guideline topic | 2015 AHA/ACC | 2020 ESC |
|---|---|---|
| **Marfan syndrome or other HTAD (e.g., Loeys-Dietz, vascular Ehlers-Danlos)** | Class IIa, LOE C<br>It is reasonable for athletes with Marfan syndrome to participate in low and moderate static/low dynamic competitive sports (classes IA and IIA) if they do not have 1 of the following:<br>a.Aortic root dilatation (i.e., z score>2, or aortic diameter>40 mm, or>2 standard deviations from the mean relative to BSA in children or adolescents<15 years old)<br>b.Moderate to severe mitral regurgitation<br>c.Left ventricular systolic dysfunction (ejection fraction<40%)<br>d.Family history of aortic dissection at an aortic diameter<50 mm | Low–intermediate risk<br>MFS or other HTAD without aortic dilation<br>-Avoid high and very high-intensity exercise, contact, and power sports<br>-Preference for endurance over power sports |
| | Class IIa, LOE C<br>It is reasonable for athletes with Loeys-Dietz syndrome or vascular Ehlers-Danlos syndrome to participate in low static, low dynamic sports (class IA) if they do not have any of the following:<br>a.Aortic enlargement (z score>2) or dissection, or branch vessel enlargement<br>b.Moderate to severe mitral regurgitation<br>c.Extracardiac organ system involvement that makes participation hazardous | Intermediate risk<br>Aorta 40–45 mm in MFS or other HTAD<br>-Only skill sports or mixed or endurance sports at low intensity |
| | Class III, LOE C<br>Athletes with Marfan syndrome, familial TAA syndrome, Loeys-Dietz syndrome, unexplained aortic aneurysm, vascular Ehlers-Danlos syndrome, or a related aortic aneurysm disorder should not participate in any competitive sports that involve intense physical exertion or the potential for bodily collision | High risk<br>Aorta>45 mm in in MFS or other HTAD<br>-Sports are (temporarily) contraindicated |

*BAV*, bicuspid aortic valve; ***BSA***, body surface area; *HTAD*, hereditary thoracic aortic disease; ***LOE***, level of evidence; *MFS*, Marfan syndrome; ***TAA***, thoracic aortic aneurysm.

dilation (Z score 2–3). Interestingly, the authors also found that 7% (9/123) of participants who were restricted from sport had a subsequent echo that did not meet criteria for sports restriction. It must be acknowledged that the application of both US and European-based guidelines among BAV athletes with aortopathy will render a significant number of athletes "too sick to play, but not sick enough to fix." This common scenario, defined by a degree of aortic dilation not sufficient to meet criteria for surgical aortic intervention but of ample severity to merit sport restriction, represents a formidable challenge with no clear best answer. We routinely evaluate such athletes and apply a shared-decision-making process that acknowledges the known risks and benefits, individualized to each athlete, of both continuing and discontinuing competitive sport participation.

## Mitral regurgitation

MR is a common valvular disease which can be caused either by a primary structural/functional abnormality of the mitral valvular apparatus (leaflets, chordae tendineae, papillary muscles, and/or annulus) or secondary to non-valvular myocardial pathology (e.g., left ventricular dilation and/or systolic dysfunction). Chronic MR leads to eccentric LV remodeling/hypertrophy in response to the volume challenge imparted by rapid early diastolic filling and in an attempt to augment forward stroke volume, thereby preserving cardiac output in the setting of high regurgitant flow. Over time, this initial compensatory process can transition into maladaptive pathology as progressive LV dilation gives rise to systolic dysfunction, and concomitant left atrial dilation reduces atrial pump function and increases the risk of atrial fibrillation. The ultimate goal in the management of athletes with chronic MR is to determine the need for and optimal timing of surgical intervention as pharmacotherapy, aside from tight control of arterial hypertension, is of limited value. The approach to surgical intervention in athletes is similar to that proposed for use in the general public. Specifically, MV surgery is recommended for all symptomatic patients with severe MR (vena contracta ≥ 0.7 cm, regurgitant volume ≥ 60 ml, regurgitant fraction ≥ 50%, effective regurgitant orifice ≥ 0.4 $cm^2$) and for asymptomatic patients with severe MR and the presence of LV systolic dysfunction (LVEF ≤ 60%, LV end systolic diameter ≥ 40 mm) [21••]. In general, surgical MV repair performed by an experienced surgeon is preferred over MV replacement. It is critical that the surgeon understands if a patient desires to return to competitive athletics after surgical recovery. This knowledge should be used in the selection of annular ring size, ideally favoring a larger ring size in athletes who want to return to sport. A larger annular ring may cause trace to mild post-operative MR after implantation, but may prevent an inadequately sized functional diastolic orifice which can limit ventricular filling and thus cardiac output during physical exertion. In patients who undergo surgical repair for primary MR, return to exercise does not appear to correlate with risks of future adverse outcomes (recurrent moderate or worse MR, mean transmitral gradient ≥ 8 mmHg, heart failure

or late onset atrial fibrillation > 3 months) on short-term follow-up (median 34 months) [41]. At the present time, percutaneous mitral valve repair and replacement, technologies being developed in other populations, are not relevant to otherwise healthy competitive athletes with chronic MR.

Athletes undergoing imaging tests for other indications are frequently found to have trace to mild MR which can be considered benign and likely secondary to EICR [17]. While there are limited studies on the effect of MR on athletic performance, mild MR does not seem to significantly affect cardiopulmonary exercise capacity in small studies [42]. Both the 2015 AHA/ACC and 2020 ESC disqualification guidelines for athletes recommend participation in all sports for athletes in sinus rhythm with mild or moderate MR in absence of LV enlargement beyond what can be attributed to EICR, systolic dysfunction, or pulmonary HTN (Table 3). Asymptomatic athletes with severe MR may also be permitted to continue low to moderate-intensity exercise.

Acute severe MR is uncommon but does occur among competitive athletes. Acute severe MR among otherwise healthy athletes typically presents as acute decompensated heart failure and constitutes a surgical emergency. Common etiologies include acute papillary muscle rupture, a condition that can be triggered by intense isometric activity, and acute LV dysfunction in the setting of coronary insufficiency or fulminant inflammatory heart disease. This diagnosis often proves challenging as rapid elevation of left atrial pressure typically eliminates or markedly reduces the severity of the regurgitation murmur, thereby masking the presence of severe MR.

## Mitral valve prolapse

Mitral valve prolapse, a common congenital defect, is found in approximately 2–3% of the population [43]. Current diagnostic criteria define MVP as systolic billowing of any portion of the mitral leaflets ≥ 2 mm past the mitral annular plane in a parasternal long axis or apical 3-chamber view. MVP can be caused by a multitude of congenital or acquired leaflet, chordae, or papillary muscle abnormalities. The presence of MVP has also been associated with multiple connective tissue diseases such as Marfan syndrome or Ehlers-Danlos syndrome. MVP can lead to numerous cardiac complications including mitral regurgitation, infective endocarditis, arrhythmias, and sudden cardiac death. Given that the spectrum of disease for MVP can range from a benign imaging finding to fatal ventricular arrhythmias, there has been a recent interest in characterizing risk factors for arrhythmic MVP. Associated risk factors for ventricular arrhythmias in MVP include female sex, myocardial fibrosis, mitral annular disjunction, leaflet redundancy, bileaflet prolapse, Pickelhaube sign on echo (spiked systolic lateral mitral annular velocities), moderate–severe MR, complex ventricular ectopy, and T-wave inversion/ST-segment depression on a resting 12-lead ECG [44–51]. It must be emphasized that none of these clinical features is sufficiently sensitive or specific to differentiate electrically benign from electrically high-risk MVP in isolation, and that the vast majority of competitive athletes with MVP will be detected incidentally and at no

**Table 3 Comparison of the 2015 AHA/ACC vs. 2020 ESC guidelines for the management of athletes with mitral regurgitation**

| Severity | 2015 AHA/ACC | 2020 ESC—leisure sport | 2020 ESC—competitive sports |
|---|---|---|---|
| **Mild** | Class I, LOE C<br>Athletes with mild to moderate MR who are in sinus rhythm with normal LV size and function and with normal pulmonary artery pressures (stage B) can participate in all competitive sports | Class I, LOE C<br>Participation in all sports, if desired, is recommended | Class I, LOE C<br>Participation in all competitive sports, if desired, is recommended |
| **Moderate** | Class IIa, LOE C<br>It is reasonable for athletes with 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 [LVEDD < 60 mm or < 35 mm/m$^2$ in men or < 40 mm/m$^2$ in women]) to participate in all competitive sports (stage B) | Class IIa, LOE C<br>Participation in all recreational sports, if desired, should be considered in individuals fulfilling the following:<br>•LVEDD < 60 mm or < 35.3 mm/m$^2$ in men and < 40 mm/m$^2$ in women<br>•LVEF ≥ 60%<br>•Resting sPAP < 50 mmHg<br>•Normal exercise test | Class IIa, LOE C<br>Participation in all competitive sports, if desired, should be considered in individuals fulfilling the following:<br>•LVEDD < 60 mm or < 35.3 mm/m$^2$ in men and < 40 mm/m$^2$ in women<br>•LVEF ≥ 60%<br>•Resting sPAP < 50 mmHg<br>•Normal exercise test |
| **Severe** | Class IIb, LOE C<br>Athletes with severe 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 [LVEDD < 60 mm or < 35.3 mm/m$^2$ in men or < 40 mm/m$^2$ in women]) can participate in low-intensity and some moderate-intensity sports (classes IA, IIA, and IB) (stage C1) | Class IIb, LOE C<br>Participation in all recreational sports involving low and moderate intensity, if desired, may be considered in individuals fulfilling the following:<br>•LVEDD < 60 mm or < 35.3 mm/m$^2$ in men and < 40 mm/m$^2$ in women<br>•LVEF ≥ 60%<br>•Resting sPAP < 50 mmHg<br>•Normal exercise test | Class IIb, LOE C<br>Participation in competitive sports involving low exercise intensity, if desired, may be considered in individuals fulfilling the following:<br>•LVEDD < 60 mm or < 35.3 mm/m$^2$ in men and < 40 mm/m$^2$ in women<br>•LVEF ≥ 60%<br>•Resting sPAP < 50 mmHg<br>•Normal exercise test |
| | Class III, LOE C<br>Athletes with MR and definite LV enlargement (LVEDD ≥ 65 mm or ≥ 35.3 mm/m$^2$ [men] or ≥ 40 mm/m$^2$ [women]), pulmonary hypertension, or any degree of LV systolic dysfunction at rest (LV ejection fraction < 60% or LVESD > 40 mm) should not participate in any competitive sports, with the possible exception of low-intensity class IA sports | | Class III, LOE C<br>Participation in competitive sports is not recommended in individuals with a LVEF < 60% |

*LOE*, level of evidence; ***LV***, left ventricular; *LVEDD*, left ventricular end diastolic dimension; ***LVEF***, left ventricular ejection fraction; *LVESD*, left ventricular end systolic dimension; ***sPAP***, systolic pulmonary arterial pressure.

clinical risk. Current valvular disease guidelines recommend surgical repair or mitral valve replacement (MVR) in patients with MVP according to the general guidelines for surgical intervention of MR [21••,23].

Athletes will most frequently present with asymptomatic MVP without significant regurgitation detected as an incidental finding on screening echocardiography or as detected by a mid to late systolic click at the apex on cardiac auscultation. The prevalence of MVP in athletes has been reported as similar to estimates in the general population (1–3%) [52, 53]. In a recent study of 215 athletes with MVP (age 30 ± 13, 67% male), a total of 10 (5%) were found to have moderate/severe mitral regurgitation and 62 (29%) had ventricular arrhythmias (VAs) [52]. The athletes with VAs were older, had higher systolic blood pressure, larger LV size and mass, and larger left atrial size. There were a total of 8 clinical events (8 ± 2-year follow-up) which included 6 mitral valve surgeries (***n*** = 2 flail leaflet, *n* = 2 dyspnea, ***n*** = 2 progressive MR with LV dilation), 1 ischemic stroke, and 1 episode of atrial fibrillation requiring hospitalization. Importantly, there were no episodes of SCD. However, MVP has been adjudicated as a very rare cause of SCD in previous young competitive athlete registries [54]. Current 2015 AHA/ACC and 2020 ESC disqualification guidelines for athletes do not provide specific recommendations for MVP but recommend management be dictated by the severity of concomitant MR (Table 3). In general, athletes with asymptomatic MVP without significant regurgitation do not require restriction from sport, though care should be taken to acquire a comprehensive personal and family medical history to screen for features suggestive of prior ventricular arrhythmias and/or a high-risk family pedigree.

## Thoracic aortic aneurysm

TAAs are most frequently degenerative in etiology and often occur with aging in association with risk factors for atherosclerosis (particularly hypertension). Less common but important causes of TAAs, particularly in young competitive athletes, include connective tissue diseases (e.g., Marfan's, Loeys-Dietz, Ehlers-Danlos syndromes), inflammatory disorders (e.g., vasculitis), infection, and other genetic syndromes. In general, TAAs usually expand slowly over years with an average expansion rate of 0.1 cm/year for ascending TAAs. As the risk of aortic dissection/rupture increases in parallel with aortic size, the absolute risk of an acute aortic syndrome should integrate aortic dimensions with other risk factors including HTN, age, and the underlying cause of aortopathy [55]. Current guidelines recommend operative intervention for aortic diameters ≥ 5.5 cm, aortic growth rate > 0.5 cm/year, and aortic diameter ≥ 4.5 cm in patients undergoing aortic valve replacement or repair including those with BAV, and recommend lower surgical intervention thresholds for patients with genetically mediated syndromes (e.g., Loeys-Dietz) [56].

Studies of young competitive athlete cohorts have consistently shown that aortic dilation, most often defined by an aortic root or ascending aorta ≥ 40 mm in males and ≥ 34 mm in females, is uncommon in this population [57, 58]. Given these findings, young athletes with aortic dimensions

outside of these limits may have an underlying pathological condition and should undergo appropriate evaluation as studies of college athletes have found that aortic dissection/rupture accounts for 5–6% of sudden cardiac death [54, 59]. Until recently, little was known about the prevalence of aortic enlargement in master's athletes or aging former competitive athletes. In a recent study examining aortic size in 206 former National Football League athletes (mean age 57.1 ± 10.3 years), 30% had an aortic diameter > 40 mm, with former NFL athletes showing significantly larger ascending aortas compared to a control group after adjusting for known risk factors [60]. Another study assessing aortic dimensions among master's runners and rowers (age 50–75) found that 21% of athletes had an aortic diameter ≥ 40 mm, and 24% had a z score ≥ 2, indicating a measurement greater than 2 standard deviations above the population mean [61]. The clinical implications of mild to moderate aortic dilation among former elite athletes and active master's athletes remain uncertain. On-going clinical surveillance studies will be required to determine rates of progression and corollary adverse outcomes.

Current 2015 AHA/ACC and 2020 ESC disqualification guidelines recommend no restriction from sports for athletes with aortic dimensions < 40 mm and no known hereditary thoracic aortic disease (HTAD) [3, 39] (Table 4). The 2015 AHA/ACC guidelines state that athletes with mildly increased aortic dimensions (z scores 2–2.5 or aortic root diameters measuring 40–41 mm in tall men or 35–37 mm in tall women) with no evidence of Marfan syndrome, Loeys-Dietz syndrome, familial TAA syndrome, or BAV may consider sports after genetic evaluation for aortopathy (class IIb; level of evidence C) [39]. There are no further recommendations for otherwise healthy athletes (without a known genetic aortopathy or disease associated with aortopathy) with aortic diameter between 40 and 50 mm in the 2015 AHA/ACC guidelines [39]. The 2020 ESC guidelines differ in that for athletes with aortic dimension between 40 and 45 mm with BAV or tricuspid aortic valve, they recommend endurance sports over power sports and avoidance of high-intensity or contact sports [3]. For athletes with BAV or tricuspid aortic valve and aortic dimension 45–50 mm, the 2020 ESC guidelines recommend only skill sports or mixed or endurance sports at low intensity. Given the variation between guidelines and the limited data to drive these recommendations, otherwise healthy athletes with aortic dimension from 40 to 50 mm and no known genetic or HTAD syndrome represent a gray-zone area. These athletes may often be restricted from sport but do not meet criteria for surgical intervention given that current guidelines in the general population recommend surgical management for otherwise healthy patients (in the absence of a BAV or other known secondary cause of aortopathy) with aortic diameters ≥ 5.5 cm, rapid growth, or ≥ 4.5 cm undergoing aortic valve repair or replacement [56]. As discussed above, this "too sick to play, but not sick enough to fix" phenotype represents a formidable clinical challenge as limited data defining the risk of aortic dissection/rupture in this cohort render shared-decision-making discussions difficult. This remains an important area of scientific uncertainty and should be considered in future investigation and guideline development.

Athletes who have had a history of TAA repair or a have a known HTAD are generally restricted from sports at lower thresholds than otherwise healthy athletes with non-hereditary TAA (Tables 2 and 4). For athletes with prior TAA

**Table 4 Comparison of the 2015 AHA/ACC vs. 2020 ESC guidelines for the management of athletes with thoracic aortic aneurysm without known hereditary thoracic aortic disease**

| Guideline topic | 2015 AHA/ACC | 2020 ESC |
|---|---|---|
| **TAA** | Class IIb, LOE C<br>For athletes with aortic dimensions mildly above the normal range (scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 35 to 37 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, familial TAA syndrome, or BAV, participation in all competitive athletics may be considered after a comprehensive evaluation for an underlying genetic condition associated with aortopathy is performed. This may include analysis for mutations in FBN1 and other genes associated with aortopathies in certain circumstances | Low risk<br>Aorta < 40 mm in tricuspid valve<br>-All sports permitted with preference for endurance sports over power sports<br>Low–intermediate risk<br>Aorta 40–45 mm in tricuspid valve<br>-Avoid high and very high-intensity exercise, contact, and power sports<br>-Preference for endurance over power sports<br>Intermediate risk<br>Aorta 45–50 mm in tricuspid valve<br>-Only skill sports or mixed or endurance sports at low intensity |
| | Class IIb, LOE C<br>For athletes with aortic dimensions mildly above the normal range (scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 35 to 37 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, familial TAA syndrome, or BAV, avoidance of intense weight training may be considered | High risk<br>Aorta > 50 mm in tricuspid valve<br>-Sports are (temporarily) contraindicated |
| **TAA following surgical repair** | Class IIa, LOE C<br>It is reasonable for athletes with surgical correction of the aortic root or ascending aorta for aneurysm disease or dissection and no evidence of residual aortic enlargement or dissection to participate in low static, low dynamic sports (class IA) that do not include the potential for bodily collision | Low–intermediate risk<br>After thoracic aortic surgery<br>-Avoid high and very high-intensity exercise, contact, and power sports<br>-Preference for endurance over power sports<br>High risk<br>After thoracic aortic surgery with sequelae<br>-Sports are (temporarily) contraindicated |

*BAV*, bicuspid aortic valve; ***BSA***, body surface area; *LOE*, level of evidence; ***TAA***, thoracic aortic aneurysm.

repair without significant residual sequelae (e.g., aortic enlargement, dissection), the 2015 AHA/ACC guidelines recommend low static, low dynamic sports (class IA) that do not include the potential for bodily collision, and the 2020 ESC guidelines provide a similar recommendation of preferring endurance exercise and to avoid high and very high-intensity exercise, contact, and power sports (Table 4). The risk for dissection following aortic repair in athletes, and the safety of specific sporting disciplines, is another area with limited data and should be a focus of future scientific inquiry.

## Conclusion

There are limited primary data to guide risk stratification and clinical management decisions for competitive athletes with valvular heart disease. Given these inherent limitations, current guidelines are based almost exclusively on expert consensus and extrapolation from data derived from the study of general population cohorts. Future research should focus on defining the risks of continual exercise in athletes with known valvular disease, the optimal time for valve repair/replacement, and the risks of returning to exercise following valvular intervention.

## Compiance with Ethical Standards

**Conflict of Interest**

Dr. Petek declares that he has no conflict of interest. Dr. Baggish has received funding from the National Institute of Health/National Heart, Lung, and Blood Institute, the National Football Players Association, and the American Heart Association and receives compensation for his role as team cardiologist from the US Olympic Committee/US Olympic Training Centers, US Soccer, US Rowing, the New England Patriots, the Boston Bruins, the New England Revolution, and Harvard University.

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## Publisher's Note