

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 7: Aortic Diseases, Including Marfan Syndrome



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

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Acute aortic dissection or rupture in Marfan syndrome or other aortopathies is an important cause of sudden death in athletes (1). Increased blood pressure and aortic stress during intense physical exertion place the patient with Marfan syndrome, Loeys-Dietz syndrome, familial thoracic aortic aneurysm (TAA) and dissection syndrome, bicuspid aortic valve (BAV) aortopathy, aortic aneurysm, or other genetically triggered aortic diseases at risk for aortic catastrophe

from aortic dissection or rupture or may accelerate aneurysm formation. Therefore, for people with aortic disease or a condition associated with aortic disease, discussion about safe levels of low-intensity, noncompetitive exercise should be emphasized beginning at a young age. This is important for a healthy lifestyle and to prevent social stigmatization, which may occur when physical activity is restricted excessively in young people.

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Marfan syndrome, an autosomal dominant disorder of connective tissue with an estimated prevalence of 1 in 5,000 to 10,000, is caused by abnormal fibrillin-1 attributable to mutations in the *FBN1* gene (2). Manifestations involve multiple organ systems, including the aorta, heart and valves, skeleton, eye, lungs, and dura. *FBN1* mutations can be identified in the vast majority of patients satisfying the revised Ghent criteria for Marfan syndrome (2). The diagnosis of Marfan syndrome is made by use of clinical criteria, imaging, family history, and genetic testing as outlined in the revised Ghent criteria (Tables 1 and 2) (2). Cardiovascular features of Marfan syndrome include mitral valve prolapse, mitral regurgitation, aortic root dilatation (most pronounced at the sinuses of Valsalva), and aortic dissection (2). The descending aorta, although less commonly involved in young patients, is also at risk for aneurysm formation and dissection.

Other genetically triggered aortic aneurysm syndromes and conditions associated with aortopathy may increase the risk of aortic dissection in competitive athletes. Loeys-Dietz syndrome is caused by mutations in *TGFBR1* and *TGFBR2* and is characterized by craniofacial features, arterial tortuosity, and aneurysms of the aorta and branch vessels, as well as increased risk of dissection at relatively small arterial dimensions (3). Vascular Ehlers-Danlos syndrome, caused by mutations in *COL3A1*, is associated with dissection and rupture of the aorta and branch vessels, even at relatively normal arterial dimensions.

TAA or dissection may be familial and is inherited as an autosomal dominant trait with decreased penetrance and variable expression. Mutations in several genes have been recognized as causing TAA disease, including *ACTA2*, *TGFBR1*, *TGFBR2*, *FBN1*, *MYH11*, *SMAD3*, *MLCK*, and *TGFB2*. Familial TAA syndromes may be associated with cerebral aneurysms or BAV; some patients have nonvascular manifestations (4,5).

BAV, which affects ~1% of the general population, may be associated with dilatation of the aortic root or

ascending aorta (6). BAV with or without TAA may be familial, and the specific gene loci responsible are yet to be determined. The prevalence of BAV in first-degree relatives of a person with BAV has been demonstrated to be ~9% (6). Cystic medial degeneration and abnormal aortic wall stress accompany BAV aortic disease independent of the valvular lesion (6). BAV with aortic aneurysm is a risk factor for aortic dissection (7). The risk of aortic dissection differs among genetically triggered aortopathies, being higher in those with Loeys-Dietz syndrome and Marfan syndrome than in BAV aortopathy.

MEASURING THE AORTIC ROOT AND ASCENDING AORTA

The ascending aorta may be divided into 2 segments, the aortic root and the upper ascending aorta. The aortic root begins at the aortic valve, includes the sinuses of Valsalva, and extends to the sinotubular junction. The upper portion of the ascending aorta begins at the sinotubular junction and rises to join the aortic arch. The normal aortic root diameter is dependent on multiple factors, including age, sex, body size, location of the aortic measurement, particular type of imaging modality used, and accuracy of measurement ascertainment (4,8). In adults, aortic diameters are larger in men than in women by 1 to 3 mm, whereas studies in children have not consistently

TABLE 1 Revised Ghent Criteria for the Diagnosis of Marfan Syndrome

1. In the absence of a family history of Marfan syndrome, any of the following:
 5. Dilated aorta (z score >2) and ectopia lentis = Marfan syndrome*
 2. Dilated aorta (z score >2) and *FBN1* mutation = Marfan syndrome
 3. Dilated aorta (z score >2) and systemic score >7 (see Table 2) = Marfan syndrome*
 4. Ectopia lentis and *FBN1* associated with known aortic dilatation = Marfan syndrome
- In the presence of a family history of Marfan syndrome, any of the following:
 5. Ectopia lentis and family history of Marfan syndrome = Marfan syndrome
 6. Systemic score >7 and family history of Marfan syndrome = Marfan syndrome*
 7. Dilated aorta (z score >2 at age ≥20 y; z score >3 at <20 y of age) and family history of Marfan syndrome = Marfan syndrome*

*Caveat: Without discriminating features of another connective tissue disorder such as Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, or Shprintzen-Goldberg syndrome, and after mutation analysis for *TGFBR1*, *TGFBR2*, *TGFB2*, *SMAD3*, *SKI*, *COL3A1*, or other genes as appropriate. Other genes/conditions will emerge with time. Modified with permission from Loeys et al. (2) Copyright © 2010, British Medical Journal Publishing Group.

TABLE 2 Scoring of Systemic Features in the Marfan Syndrome*

Feature	Points
Wrist and thumb sign	3
Wrist or thumb sign	1
Pectus carinatum deformity	2
Pectus excavatum or chest asymmetry	1
Hindfoot deformity	2
Plain pes planus	1
Pneumothorax	2
Lumbosacral dural ectasia	2
Protrusion acetabuli	2
Reduced upper-segment to lower-segment ratio (<0.85 in white adults; <0.78 in black adults) and increased arm span-to-height ratio (>1.05) and no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension	1
Facial features (3 of 5): dolichocephaly, enophthalmos, down-slanting palpebral fissures, malar hypoplasia, retrognathia	1
Skin striae	1
Myopia (>3 diopters)	1
Mitral valve prolapse	1

Maximum total, 20 points; score >7 indicates systemic involvement. *A detailed explanation of the systemic score and nosology may be found at <http://www.marfandx.org>. Modified with permission from Loeys et al. (2) Copyright © 2010, British Medical Journal Publishing Group.

demonstrated a sex difference in aortic diameter when corrected for body surface area (BSA) (8). Ascending aortic dimensions in adults are also related to age, sex, and BSA (9).

Variability in the measured aortic diameter may result from the type of imaging modality used, whether contrast is used, and whether internal or external aortic diameters are recorded. For example, transthoracic echocardiographic nomograms have reported aortic root diameters using sinus-to-sinus measurements from a leading-edge technique at end diastole (10), whereas *z*-score determinations validated in children have used maximal end-systolic diameter at the sinuses of Valsalva using inner-edge-to-inner-edge measurements (11). Measurements should be taken perpendicular to the axis of blood flow and should include the largest measured aortic diameter (whether at the sinuses of Valsalva or the ascending aorta) (4). Images taken from echocardiography, computed tomography (CT), or magnetic resonance imaging may overestimate the true aortic diameter if oblique slices are obtained. CT and magnetic resonance imaging measurements from sinus to commissure are generally smaller than echocardiographic measurements from sinus to sinus (8). CT or magnetic resonance imaging techniques are used when the extent of aortic enlargement is not adequately or completely visualized by the echocardiogram. Imaging techniques that avoid or minimize radiation are recommended whenever possible, particularly when serial assessment is anticipated. Regardless of which imaging technique is used, it is important that serial measurements be made at the same location by the same method for appropriate clinical correlation.

Older nomograms that predict normal and abnormal aortic dimensions are limited by such factors as failure to account for sex differences, limited age ranges of subjects studied (especially teenagers), marked jumps in “normal” aortic diameter based on age-range strata, and the use of small sample sizes (8,10).

***z* Scores**

Notably, *z* scores that incorporate height, weight, age, and sex are now preferred for determination of normal aortic diameter as opposed to a single aortic dimension (8). The *z* score describes how many standard deviations above or below a size or age-specific population mean a given measurement lies (12). They are especially useful for evaluation of cardiac dimensions in the young, whose normal values change during growth.

Aortic dilatation is recognized when the difference between the observed sinus of Valsalva diameter and the value predicted for age, sex, and BSA (*z* score) is >2.0, which corresponds to approximately the 98th percentile of the general population (8). A *z* score of 3 corresponds to the 99.9th percentile. Mild, moderate, and severe aortic

dilatation may be defined by *z*-score values of 2 to 3, 3.01 to 4.0, and >4.0, respectively (8,13). Reference values for ascending aortic diameter assessed by echocardiography are also available from large databases (14).

A formula for calculating aortic sinus of Valsalva diameter *z* scores was derived recently from a data set of 1207 healthy subjects >15 years old, in whom aortic root diameter ranged from 2.1 to 4.3 cm (8). Aortic dimensions were calculated by echocardiogram at end diastole from sinus to sinus using a leading-edge-to-leading-edge technique (Figure). *z* Scores are calculated from this database using the following equation (8) (online-only Data Supplement aortic *z*-score calculator):

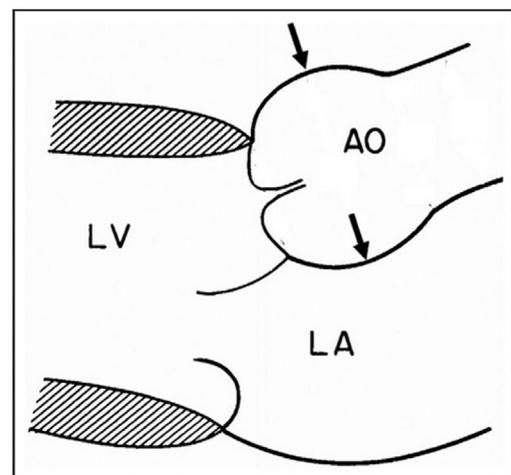
$$\begin{aligned} \text{Expected aortic root size: } & 2.423 + (\text{age [years]} \times 0.009) \\ & + (\text{BSA [square meters]} \times 0.461) \\ & - (\text{sex [1 = man, 2 = woman]} \times 0.267); \\ \text{standard error of estimate} & = 0.261 \text{ cm} \end{aligned}$$

$$\begin{aligned} z \text{ Score} = & (\text{observed aortic root size} \\ & - \text{expected aortic root size}) / 0.261 \end{aligned}$$

For example, a 22-year-old man with a BSA of 2.0 m² has an aortic root diameter of 4.1 cm at the sinuses of Valsalva. Thus, his expected aortic root size is calculated as follows:

$$\begin{aligned} & 2.423 + (22 \times 0.009) + (2.0 \times 0.461) - 0.267 = 3.276 \\ & 4.1 (\text{observed aortic root size}) \\ & - 3.28 (\text{expected aortic root size}) = 0.824 \\ & 0.824 / 0.261 (\text{standard error of estimate}) = 3.16 \end{aligned}$$

FIGURE Schematic of the Aortic Root Showing Measurement of the Aortic Root Diameter at Maximum Width Parallel to the Aortic Annular Plane by American Society of Echocardiography Leading-Edge Convention (Arrows)



AO indicates aorta; LA, left atrium; and LV, left ventricle. Reproduced with permission from Devereux et al. (8) Copyright © 2012, Elsevier Inc.

Thus, the *z* score is 3.16, which is significantly abnormal for this patient.

AORTIC DIMENSIONS IN ATHLETES

Intense physical exertion is associated with hemodynamic changes that increase aortic wall tension and may increase aortic dimension (15-17). Chronic intense weight training may influence aortic dimension (18). Furthermore, elite athletes have slightly larger aortas at the sinuses of Valsalva than nonathletic control subjects (19). Although mild aortic enlargement may be a normal adaptation to intense training, large increases in aortic size are unusual in athletes and when present are more consistent with an underlying pathological aortopathy, which may be exacerbated by exercise training (19).

TALL ATHLETES

Although increasing BSA is associated with larger aortic diameters, there is a nonlinear relationship, with a plateau, between aortic root dimensions and height (>189 cm or 74.5 inches in men; >175 cm or 69 inches in women) and BSA (>2.3 m²) in very tall people (20). A small proportion of athletes will have an aortic dimension slightly greater than the diameter considered to be at the upper limits of normal (i.e., >2 standard deviations above the mean, or *z* score >2) (15,16). Therefore, it is important to avoid attributing the enlarged aorta in tall (or large) athletes solely to height, BSA, or a physiological response to exercise (19). Mild aortic dilation in an athlete should trigger evaluation to determine whether an underlying aortopathy is present and whether the aortic size conveys an increased risk to the athlete.

We underscore that for athletes with aortic *z* scores above the normal range for age, sex, and BSA (i.e., *z* score >2 to 2.5), evaluation by a knowledgeable specialist, and often by a multidisciplinary team that includes a medical geneticist and cardiologist, is recommended to exclude an underlying disorder associated with aortic dilatation (such as Marfan syndrome, familial TAA syndrome, or BAV disease). Indeed, systemic features of some disorders may be subtle and often overlap with those in the general population. Referral to a specialized center with expertise in the clinical and genetic evaluation of genetic aortic disease may be necessary in some instances. In selected cases, we recognize that it may not be possible to distinguish pathological aortic dilatation from a nonpathological aortic size when the aortic measurement mildly exceeds the normal range in very tall people or in those with large BSA, especially when there is only a single evaluation at only 1 point in time.

OUTCOME AND RISK OF AORTIC DISSECTION AND RUPTURE

There is a paucity of data examining the long-term outcome of athletes with unexplained aortic dilatation (16,17). Of 2317 Italian athletes, 17 males (ages 25 ± 7 years; height 188 ± 10 cm; BSA 2.17 ± 0.25 m²) had aortic diameters >40 mm and were allowed to continue participation (16,17). Over an 8 ± 5 year follow-up, the aortic root increased mildly in diameter from 40.9 ± 1.3 to 42.9 ± 3.6 mm in these 17 athletes, and none experienced acute aortic dissection. Two athletes had progressive aortic dilation to 50 mm by ages 38 and 50 years, respectively (17).

The risk of aortic dissection in the general population is related to many factors, foremost of which is the severity of aortic dilation, and is sometimes triggered acutely by heavy weight lifting or strenuous exercise, including competitive sports (21-22a). However, some patients with acute aortic dissection do not have a markedly dilated aorta at the time of dissection (23,24). In a series of 177 patients without the Marfan syndrome phenotype or BAV who incurred an acute type A dissection, aortic diameter was <50 mm in 42% and <45 mm in 21% at the time of dissection. Furthermore, 12% of women had a dissection at an aortic diameter <40 mm (23). Similarly, in the International Registry of Acute Aortic Dissection, 40% of acute type A dissections occurred with aortic diameters <50 mm (24). There is no evidence that β-blockers, angiotensin receptor blockers, or angiotensin-converting enzyme inhibitors protect athletes from aortic dissection or rupture during intense competitive sports.

There are no prospective data available regarding the risks of competitive athletics in patients who have undergone surgical correction for aortic aneurysm or dissection; however, after aortic root replacement, patients with Marfan syndrome, Loeys-Dietz syndrome, and familial TAA disease remain at risk for distal aortic complications (3,5,25,26). Additionally, BAV aortopathy may involve aortic segments distal to the root (27).

PRIOR RECOMMENDATIONS FOR ATHLETES

The 36th Bethesda Conference Report (2005) recommended that athletes with “unequivocal aortic root enlargement” (therein defined as >40 mm in adults, >2 standard deviations beyond the mean for BSA in children and adolescents, or a *z* score of >2) only participate in low-intensity competitive sports (class IA sports) (28). Characterizing an aortic diameter of >40 mm as enlarged in males is an arbitrary but also useful definition, because very few apparently healthy young male athletes have been reported with aortic root diameters >40 mm (17,19). For example, in a study of >2,000 Italian athletes, the 99th

percentile value of aortic diameter by echocardiogram was 40 mm in males and 34 mm in females (16). In a Japanese study, only 6 of 1,562 male athletes (0.38%) had an aortic root dimension >40 mm, and 2 of these also had phenotypic features of the Marfan syndrome (29). In an evaluation of >1000 female Italian athletes, the 99th percentile for aortic size at the sinuses of Valsalva was 34 mm, and no woman had an aortic diameter >36 mm (16,17).

Guidelines for participation in competitive sports have been lacking for those patients with mildly dilated aortic dimensions (i.e., z scores of 2 to 2.5, or 1 to 2 mm above the normal ranges described above) and no diagnosis of an underlying connective tissue disorder, family history of aortic disease, or pathogenic gene mutation associated with aneurysm disease (17). In this situation, there are difficult individual choices regarding sports participation to be made on a case-by-case basis. Discussion with the athlete, parents (when appropriate), and coaches/trainers should include full disclosure and transparency regarding the potential risks of further training and competition. For instance, the mildly dilated aorta may represent a pathological aortic condition, and the aorta may dilate further with continued exercise and athletic participation, or it may dilate years later. In such a person, absence of a pathogenic genetic mutation does not exclude risk. Furthermore, although the absolute risk of aortic dissection or rupture in this clinical situation is unknown, it is not zero. If participation in competitive sports is continued, close aortic surveillance (i.e., every 6 to 12 months) with echocardiography or magnetic resonance angiography (MRA) should be performed to assess aortic dimension (17). The frequency of imaging is dependent on the absolute size of the aorta, the z score, stability of the aortic size, and the intensity of the sport. In the athlete with a mildly dilated aorta, continued aortic enlargement should not be regarded as physiological but rather consistent with an underlying aortopathy; disqualification from competition should result if the aorta continues to enlarge. Because some athletes identified with only a mildly dilated aortic root have required aortic aneurysm surgery several years later, long-term aortic surveillance is recommended even after engagement in the competitive athletic lifestyle has terminated (17,29).

Recommendations

1. Athletes with Marfan syndrome should undergo echocardiographic (and in some instances MRA or CT) measurement of the aortic root dimension every 6 to 12 months, depending on aortic size (Class I; Level of Evidence C).
2. Athletes with unexplained TAA, familial TAA syndrome, or known pathogenic mutation leading to a familial TAA syndrome (*ACTA2*, *MYH11*, *FBN1*, *TGFBR1*, *TGFBR2*, *MLCK*, *SMAD3*, *TGFB2*, and others) should undergo echocardiographic and (depending on the diagnosis) MRA or CT surveillance every 6 to 12 months to evaluate for progression of aortic or branch vessel disease (Class I; Level of Evidence C).
3. Athletes with aortic dimensions mildly above the normal range (z scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 36 to 38 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, or familial TAA syndrome should undergo echocardiographic or MRA surveillance every 6 to 12 months, with imaging frequency dependent on aortic size and stability of measurements (Class I; Level of Evidence C).
4. 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 (see Task Force 5 on valvular heart disease [30]) (Class I; Level of Evidence C).
5. Athletes with BAV and aortic dimensions above the normal range (scores 2 to 3 or aortic diameters measuring 40 to 42 mm in men or 36 to 39 mm in women) should undergo echocardiographic or MRA surveillance of the aorta every 12 months, with more frequent imaging recommended for increasing aortic z score (Class I; Level of Evidence C).
6. It is reasonable for athletes with Marfan syndrome to participate in low and moderate static/low dynamic competitive sports (classes IA and IIA; see definition of sports classification in Task Force 1 report [31]) if they do not have ≥ 1 of the following (Class IIa; Level of Evidence C):
 - 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)
 - b. Moderate to severe mitral regurgitation
 - c. Left ventricular systolic dysfunction (ejection fraction <40%)
 - d. Family history of aortic dissection at an aortic diameter <50 mm
7. It is reasonable for athletes with an unexplained TAA, familial TAA syndrome, or known pathogenic mutation leading to familial TAA syndrome (*ACTA2*, *MYH11*, *FBN1*, *TGFBR1*, *TGFBR2*, *MLCK*, *SMAD3*, *TGFB2*, and others) to participate in low static, low dynamic competitive sports (class IA) if they do not have ≥ 1 of the following (Class IIa; Level of Evidence C):
 - a. Aortic root dilatation (i.e., score >2, or aortic diameter >40 mm, or >2 standard deviations from the mean relative to BSA for children and adolescents <15 years old)

- b. Moderate to severe mitral regurgitation
 - c. Family history of aortic dissection
 - d. Cerebrovascular disease
 - e. Branch vessel aneurysm or dissection
8. 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 (*Class IIa; Level of Evidence C*):
- a. Aortic enlargement (score >2) or dissection, or branch vessel enlargement
 - b. Moderate to severe mitral regurgitation
 - c. Extracardiac organ system involvement that makes participation hazardous
9. 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 (*Class IIa; Level of evidence C*).
10. For athletes with a BAV and a mild to moderately dilated aorta (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 (*Class IIb; Level of Evidence C*).
11. 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 (*Class IIb; Level of Evidence C*).
12. 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 (*Class IIb; Level of Evidence C*).
13. 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 (*Class IIb; Level of Evidence C*).
14. 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 (*Class III; Level of Evidence C*).
15. 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 (*Class III; Level of Evidence C*).
16. Athletes with BAV and a markedly dilated aorta (>45 mm) should not participate in any competitive sports (*Class III; Level of Evidence C*).
17. Athletes with chronic aortic dissection or branch vessel arterial aneurysm or dissection should not participate in any competitive sports (*Class III; Level of Evidence C*).

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