Cardiovascular Screening and the Elite Athlete: Advances, Concepts, Controversies, and a View of the Future

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For centuries, the elite athlete has enjoyed a status of reverence and adoration in our culture. An early example was the first recognized Olympic Games organized in Greece in 776 BC. The legend of Pheidippedes began with his success as an Olympic champion around 500 BC. Ten years later his celebrated run from the plains of conflict ...

**KEYWORDS**
- Cardiovascular screening
- Elite athletes
- Athletic sudden death
- Athletic heart
- Hypertrophic cardiomyopathy

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Marathon to Athens to announce the Greek victory over the Persians and his sudden
death (SD) that followed marks the beginning of our celebration of the marathon race
and has permanently etched athletic SD into our consciousness. The cultural impor-
tance of the highly trained elite athlete has continued to develop and amplify into
modernity. Continuous media coverage on television, radio, Internet, and social
networking has elevated the athlete's status to unprecedented heights. Any well-
recognized elite athlete participating in sports at the intercollegiate, national team,
professional, or Olympic levels occupies an important place in the collective psyche
of fellow students, faculty, staff, alumni, and fans around the world. Depending on
the athlete and the sport, this effect ripples from thousands to millions of affected indi-
viduals. The unexpected death of a young and previously healthy individual from
natural causes, although a tragedy for family and friends, is ultimately reconcilable.
By contrast, the death of a celebrated athlete, at the summit of their invincibility,
extends to countless individuals beyond friends and family. It is generally perceived
as somehow preventable and thus it is ultimately irreconcilable. The well-known words
of the English poet and scholar A.E. Housman capture the impact of the death of
a young athlete on a small town:

The time you won your town the race
We chaired you through the market place;
Man and boy stood cheering by
And home we brought you shoulder-high.
Today, the road all runners come,
Shoulder-high we bring you home,
And set you at your threshold down,
Townsman of a stiller town.

A.E. Housman (1859–1936): To an Athlete Dying Young

Perhaps the most salient case of athletic SD in the modern era was that of Hank
Gathers, who was a senior forward on the Loyola Marymount Division I basketball
team in 1990. In the previous season, Gathers had become only the second player
in collegiate history to lead the nation in scoring and rebounding. During the 1990
season Loyola Marymount contended for a high national ranking and hoped to gain
the national title. Because of the small size of the school and the arc of the team
into national consciousness, Loyola Marymount became a favorite of National Colle-
giate Athletic Association basketball fans across the country. However, events
conspired, beginning with Gathers fainting on the court. Both sustained and nonsus-
tained ventricular arrhythmias were documented. Gathers was withdrawn for 3 weeks
and treated medically with propranolol. He was not disqualified, returned to competi-
tion later in the season, and during a tournament game, Gathers collapsed and died of
ventricular fibrillation.

The intricacies of this case, including the probable diagnosis of myocarditis, have
been elaborately discussed. The emotional cost to the Gathers family, the Loyola
Marymount University family, and the entire sports world is beyond measure. This
case also resulted in tremendous financial cost, including an in-court judgment against
the physician and an out-of-court settlement with the university. The case of Hank
Gathers in 1990 and the tragic timing of those events ushered in a new precedent
for those involved in the care and supervision of the elite athlete. Flo Hyman, an
Olympic volleyball player with the Marfan syndrome, died 4 years before Gathers
and other elite conditioned sports luminaries later suffered similar fates to Gathers,
including Pete Maravich, Reggie Lewis, and Jason Collier (basketball), Thomas Her-
rion (football), Jiri Fischer (hockey), Sergei Grinkov (ice skating), and athletes in Europe
and Africa including Marc-Vivian Foe, a soccer player from Cameroon who died of hypertrophic cardiomyopathy (HCM) during a televised international match.\(^4\) Although, the incidence of athletic SD is an uncommon event, the celebrity status of the elite athlete and the consequences of any such catastrophe have generated considerable worldwide interest in cardiovascular screening and programmatic evaluation of athletes to detect unrecognized and potentially life-threatening abnormalities in an effort to reduce this risk to a level as low as can possibly be achieved.

This article addresses programmatic cardiovascular screening and evaluation of the elite athlete at the intercollegiate, national team, professional, and Olympic levels. Although much of the content may apply to high-school and recreational sports at large, it is not specifically designed to address the athletes participating in the vast array of sports activities in today’s world.

**CONCEPTS AND CONTROVERSY IN CARDIOVASCULAR SCREENING OF THE ELITE ATHLETE**

The modern elite athlete is unique to our general population in a myriad of ways. The athlete tests the heart and vascular system to their limit both in training and in competition. The athlete’s cardiovascular engine is pushed to the extremes of exertion both in endurance (isotonic, dynamic, or aerobic) and in strength (isometric, static, or anaerobic). The intensity of the performance endured by elite athletes exposes the abnormal cardiovascular system, and any unrecognized underlying weakness could lead to compromised performance, untoward symptoms, and sudden cardiac death.

The following can support a strong argument for a comprehensive screening program for elite athletes:

- The occurrence of athletic SD during training or competition is unacceptable. Thus, all reasonable preventative measures should be undertaken.
- Detection of previously undiagnosed arrhythmias, congenital, or acquired heart disease that could affect the health, performance, and longevity of the athlete is of value.
- Detection of autosomal-dominant conditions: HCM, Marfan syndrome and related vascular disorders, long QT, and arrhythmogenic right ventricular cardiomyopathy (ARVC) could lead to evaluation and treatment of siblings and offspring, particularly in families with numerous athletes, who have a 50% chance of being affected.
- The extreme level of performance of elite athletes mandates cardiovascular screening that is not applicable to the general population.
- The prevalent usage of dextroamphetamines and amphetamines to treat attention-deficit/hyperactivity disorder\(^5\) and the widespread availability of these drugs, particularly at the intercollegiate level, mandates knowledge of underlying cardiovascular abnormalities in treated athletes as well as untreated athletes who have unfettered access to these drugs.

The causes of SD in athletes less than the age of 35 years are well known and were established in a landmark paper by Maron and colleagues.\(^6\) HCM, an autosomal-dominant genetic disorder of the cardiac sarcomere, is responsible for 48% of these deaths in the United States. HCM and other congenital cardiac defects account for approximately 80% to 90% of overall deaths, including congenital coronary artery anomalies and diseases of the aorta. Only a few cases were suspected or diagnosed before the athlete’s death. There are also many congenital heart conditions and acquired cardiovascular disorders that could affect the future health of the athlete.
and/or impair performance. Many of these conditions have subtle, if any, physical findings and remain undetected into late adolescence and young adulthood (Box 1).

The BAV is of particular importance. This is the most prevalent congenital defect in the general population (2%), especially in males, and may be overlooked because SD as a result of aortic valve disease is rare because the murmurs of significant aortic stenosis (AS) and regurgitation (AR) are easily audible, leading to disease detection and recognition. We encourage renewed attention to the congenital BAV because more than 50% of affected individuals have an associated abnormality of the ascending aorta at the sinuses of Valsalva or above the sinotubular junction. This finding includes asymptomatic young athletes with a functionally normal aortic valve (or mild AS/AR). Furthermore, there is evidence of a significant prevalence of

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<td>Congenital and acquired cardiovascular disorders often undetected in childhood</td>
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<td>• Bicuspid aortic valve (BAV): functionally normal or mild valve disorder</td>
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<td>50% have associated abnormality of ascending aorta (congenital aortopathy)</td>
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<td>• Atrial septal defect (ASD)/left to right shunts</td>
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<td>Secundum ASD, which includes atrial septal aneurysm with small or multiple ASD/patent foramen ovale</td>
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<td>Partial anomalous pulmonary venous return</td>
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<td>• HCM</td>
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<td>• Anomalous coronary arteries, coronary artery fistulae</td>
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<td>• Wolff-Parkinson-White syndrome</td>
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<td>• Long QT, short QT</td>
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<td>• Ventricular tachycardia, pathologic premature ventricular contractions (PVCs)</td>
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<td>• Arrhythmogenic right ventricular cardiomyopathy</td>
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<td>• Mitral valve prolapse</td>
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<td>• Acquired mild to moderate valvular heart disease: rheumatic, degenerative, radiation-induced, and traumatic (particularly traumatic tricuspid regurgitation)</td>
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ascending aortic dilation in first-degree relatives of patients with BAV, and this aortopathy is not necessarily accompanied by the BAV.\textsuperscript{9} It is logical that this may pose a potential risk to young athletes and that detection of these abnormalities would be favorable and would allow for special monitoring of aortic dimensions and disqualification in extreme cases.

The low frequency of SD events in athletes creates a problem inherent to undertaking any screening program.\textsuperscript{6,10} Data from the Minneapolis Heart Institute Foundation collected on athletic SD from public records (usually print or broadcast media) document approximately 125 cases of athletic SD per year.\textsuperscript{11} Although likely an underestimation of total cases of SD because of underreporting, this statistic may capture most cases of elite athlete SD. Broad-based testing of a large population with a low incidence of events creates the difficult epidemiologic problem of testing results containing more false-positive results than true-positive results. If screening errors for each athlete who has a true abnormality sideline several healthy athletes with normal cardiovascular systems, then clearly our screening protocol has failed.

Accordingly, any comprehensive screening program for elite athletes should be structured to maximize true-positive results and minimize false-positive results. The remaining sections of this article review past and current testing strategies and explore new testing strategies that could improve diagnostic yield and reduce unwanted false-positive results.

CARDIOVASCULAR SCREENING OF THE ELITE ATHLETE

Standards and guidelines for programs and individual health care providers involved in athletic cardiovascular screening are lacking, as are credentialing and recommendations for knowledgeable caregivers in this area. In practice, subspecialty health care providers with little practical experience may evaluate athletes and associated cardiovascular diagnostic studies with athletes, including the spectrum of the athlete’s heart. Furthermore, providers may have limited knowledge and exposure to congenital heart disease, HCM, arrhythmogenic right ventricular dysplasia, long QT, and anomalous coronary arteries. Practices vary from state to state and it is fair to assume the current system of screening is not ideally designed to maximize the yield of true-positive results in this population and simultaneously minimize undesirable false-positive results.

\textit{History and Physical Examination: Initiating the Process}

Ideally, the initial history and physical examination should be performed within a credentialed sports medicine facility. This strategy ensures the vital association of the athlete to a licensed sports medicine provider and connects the athlete to a medical home to begin a relationship between the athlete and provider that promotes trust and full disclosure of symptoms and past history from the athlete. The primary sports medicine provider, in turn, navigates the subspecialty referral process, including cardiovascular screening, supports the athlete, and integrates the medical plan in concert with sophisticated sports training staff. It is essential that the medical home of the athlete fully integrates the athletic training staff who have developed a sophisticated involvement in the complete health of athletes, with a wide array of knowledge and technology, including stethoscopes, blood pressure cuffs, and automatic external defibrillators. Athletic trainers also take an active role in coordinating medical appointments for athletes, and attending those appointments allows the athletic trainer to carry the medical information back to the athletic arena. This valuable resource can assist in execution of the plan; report on the consequences...
of the plan; and provide on-site eyes and ears that greatly assist in management and
diagnosis of cardiovascular disorders.

The history and physical examination includes screening for a family history of SD or
heart disease in relatives less than 50 years old, a detailed personal history, and phys-
ical examination. This strategy has been described in detail by Maron and
colleagues12 and was adopted by the American Heart Association consensus panel
recommendations for preparticipation screening.13 The history and physical examina-
tion are limited and cannot be expected to detect most dangerous cardiovascular
diseases. In a retrospective analysis, only 3% of athletes who suffered SD and had
participated in screening history and physical examination were considered to have
possible cardiovascular abnormalities, and none was disqualified.14

The physical examination is likely to be helpful in screening patients with the Marfan
syndrome and related vascular disorders (Loeys-Dietz syndrome15 and Ehlers-Danlos
syndrome), with careful attention paid to skin (laxity, striae) the musculoskeletal
system (arachnodactyly, scoliosis, anterior chest wall deformity), eyes (myopia, ecto-
pia lentis), and general assessment for any syndromic abnormalities (bifid uvula, cleft
palate, hypertelorism, micrognathia). Detection of these disorders is critical because
aortic dissection, although often not fatal, changes the remainder of that individual’s
life (Fig. 1). Careful auscultation of the heart is important and the systolic opening click
of the functionally normal BAV can be subtle and should not be overlooked. In general,
the physical examination may prove to be of greater value as sports medicine
continues to grow and experienced evaluators become more prevalent.

Family history is important but also limited. For example, Marfan syndrome and HCM
are complex genetic disorders and up to 25% to 33% of individuals affected have
spontaneous mutations that are new and private. Thus, a negative family history is
misleading.16,17 Any family history of SD before the age of 35 years mandates a referral
to cardiology to determine what further diagnostic testing is indicated because of the
prevalent genetic transmission of HCM, long QT, ARVC, and Marfan syndrome and
related vascular disorders (including familial BAV/congenital aortopathy).

Fig. 1. CT scan with contrast showing a spiral type B dissection of the descending thoracic
aorta (arrows indicate the intimal flap) in a female intercollegiate swimmer with the Marfan
syndrome. The dissection occurred during practice. The sinuses of Valsalva were only border-
line abnormal with an eccentric posterior sinus.
Diagnostic Testing

Routine diagnostic testing beyond the history and physical examination is controversial and not widely agreed on or standardized. Additional noninvasive testing strategies might include electrocardiography (ECG), transthoracic echocardiography (TTE), exercise testing (ETT), Holter or event recording, genetic testing, computed tomography (CT) angiography, and cardiac magnetic resonance imaging (CMR) with or without contrast.

For the purpose of screening, we believe that only tests that are noninvasive and pose no potential health threat to the athlete are acceptable. Accordingly, radiation (CT) and administration of intravenous contrast (CT and CMR) are unacceptable tools and should be used only when a significant disorder is suspected. Genetic testing for conditions like long QT, HCM, Marfan syndrome, and related disorders should be performed only by experienced providers under select circumstances. For the most part recognition and treatment of these disorders is clinical. Genetic testing is expensive and limited because a negative test does not exclude the condition being tested and a positive test does not always well predict the phenotypic expression of that condition within the individual of concern.

The 2 most widely used tests are the ECG and TTE and these are discussed separately. Routine use of ETT to screen young elite athletes is not of value because of the low incidence of coronary artery disease and because ETT is inconsistent and not generally helpful in detecting anomalous coronary artery variations that have been associated with SD. Selective use of ETT may be of value in symptomatic athletes with exercise-induced arrhythmias. Of particular concern is catecholaminergic polymorphic ventricular tachycardia (CPVT), which is a genetic disorder that can lead to SD. ETT may also benefit older athletes with risk factors present that could increase the risk of premature coronary artery disease.

In athletes more than the age of 35 years, the leading cause of SD shifts away from HCM and overwhelmingly favors coronary artery disease (80%). To reiterate, it is essential that the subspecialty cardiovascular professionals who are involved in any screening program and perform and interpret further diagnostic testing have a thorough understanding of the normal range of physiologic adaptations that comprise the athlete’s heart and also have experience with arrhythmias as well as a variety of conditions present in the population of adults with congenital heart disease. Otherwise, athletes with normal adaptations are sidelined unnecessarily and those with real cardiovascular issues may be underdiagnosed.

ECG

ECG is a safe, relatively inexpensive, and routinely available screening tool for the elite athlete and can provide useful diagnostic information about underlying electrical or structural abnormalities of the heart that could place the athlete at risk. Accordingly, the ECG has been the most widely used cardiovascular diagnostic test in athletic screening. However, this is a complicated diagnostic and interpretive process because most elite athletes manifest some degree of electrocardiographic aberration when compared with the normal population because of intense athletic training, which leads to important alterations in both structure and autonomic regulation of the heart. Most highly trained athletes have an abnormal resting ECG with well-documented training-related abnormalities, including sinus bradycardia, first-degree atrioventricular block, complete right bundle branch block (RBBB), early repolarization, and QRS voltage criteria for left ventricular hypertrophy (LVH). LVH poses a particular problem in the athlete because it may simply reflect the physiologic adaptation to
isometric training (LVH), or isotonic training (dilation with increased LV mass), or a combination of the two. The challenge for any screening program is to differentiate between this adaptive response and the pathologic condition of HCM.

Several factors influence the type and degree of electrocardiographic abnormalities in the elite athlete. Physiologic and structural changes that alter the ECG are more common in males and in athletes of African or Caribbean descent. The type of training undertaken by the athlete is also important, with physiologic abnormalities detected on ECG more common in endurance sports that also include significant amounts of static/isometric training, such as cycling, rowing, canoeing, and cross-country skiing. More recent data including sports prevalent in the United States suggest that ECG abnormalities are also more common in football. Race can accentuate this as well, with male black football players twice as likely to have ECG abnormalities as their white counterparts. Angiotensin gene polymorphisms may also predispose athletes to abnormal remodeling and thus more extensive increases in LV mass and wall thickness. Cardiovascular screening inclusive of ECG poses a more difficult interpretive challenge in the male athlete compared with the female athlete because female athletes are more likely to have a normal or only mildly abnormal ECG. Race affects the female athlete’s ECG in a fashion similar to that noted with males but the differences are less dramatic.

Corrado and colleagues published findings of a 25-year experience with elite Italian athletes participating in a wide array of sports. This study has helped to establish thorough guidelines for the interpretation of ECG in elite athletes. It also provides the modern screening process with the sophistication to discern the difference between physiologic adaptation and potentially dangerous electrophysiologic or structural abnormalities. From the Italian perspective this strategy has been shown to be cost-effective. Corrado and colleagues have also provided insights regarding the types of abnormalities that are “uncommon and training un-related ECG changes,” including T-wave inversion, ST segment depression, pathologic q waves, left atrial enlargement, left axis deviation/left anterior hemiblock, right axis deviation/left posterior hemiblock, right ventricular hypertrophy, ventricular preexcitation (Wolff-Parkinson-White), complete left bundle branch block (LBBB) and RBBB, long or short QT, and Brugada-like early repolarization. LVH by voltage criteria is a common ECG abnormality in athletes, reflecting increases in LV wall thickness or mass. This physiologic adaptation usually results in increased QRS amplitude alone, not in combination with ST segment depression, T-wave inversion, or prolongation of the QRS or shift in QRS axis. In the elite athlete these changes are adaptations in hypertrophy and mass and usually do not correlate with underlying HCM on TTE. However, caution is advised because the sensitivity of a negative ECG in the detection of HCM is limited by the 20% of individuals with HCM who have normal or only mildly abnormal ECG. Simply stated, ECG alone does not uncover all individuals with HCM.

ARVC has historically been a difficult clinical diagnosis but more recently criteria for diagnosis by CMR have become more specific. ECG may reveal T-wave inversion of greater than 2mm in 2 or more adjacent leads (a finding uncommon in older athletes) and this finding may warrant magnetic resonance imaging (MRI) screening for ARVC if TTE reveals no evidence of other structural heart disease. Examples of abnormal screening ECG are provided in Figs. 2–6.

The complexity of this interpretive process has understandably created controversy with regard to mandated ECG screening of athletes. In the United States, use of ECG in screening has been advocated, and the limitations and pitfalls inherent in widespread screening have also been elaborately argued. Mandated preparticipation
ECG screening has been discouraged and has not been endorsed by the American College of Cardiology, the American Heart Association, and most recently in 2005 at the 36th Bethesda Conference on Eligibility Recommendations for Competitive Athletes with Cardiovascular Disorders. For the past 27 years there has been an Atlantic divide on this subject, with the Italian experience including a federally subsidized preparticipation screening program inclusive of ECG in all athletes. Italian investigators have published observations suggesting a reduction in the overall rate of athletic SD after implementation of this more aggressive strategy and further argue that preparticipation screening with ECG led to the identification and disqualification of athletes with HCM and as a result, the demographics of athletic SD in the Veneto region of Italy have shifted away from HCM as the most common cause of SD and toward ARVC. In this last study of 269 pathologic cases of athletic SD only 1 athlete had HCM, with more cases attributable to ARVC (22%).

Fig. 2. Long QT in a patient with torsades de pointes. The QT interval indicated by the bar in V6 is prolonged to approximately 600 ms.

Fig. 3. T-wave inversion in 3 consecutive precordial leads (arrows) in an athlete with ARVC.
There will be continued controversy regarding the implementation of any complex interpretive process like ECG screening and we believe there is merit on both sides of the argument. In order for the benefit to exceed the harm and for any program to be cost-effective, it must be advanced in a sophisticated manner with knowledgeable providers involved at all levels. Although one can argue that the Italian system has merit and should be emulated, it can also be argued that logistically we are not ready, capable, nor can we afford implementation of widespread ECG screening of elite athletes in the United States.\(^46\)

**Fig. 4.** Brugada syndrome with coved type ST segment increase greater than 2 mm in V1 (arrow) followed by a negative/inverted T wave.

**Fig. 5.** Wolff-Parkinson-White syndrome. During sinus rhythm the delta wave is seen in lead V4 (thick arrow). There are also frequent salvos of rapid atrial fibrillation with a wide QRS, which indicates conduction via an accessory pathway (narrow arrows). With atrial fibrillation conducted at this rate (close to 300 beats per minute) this athlete is at risk for the degeneration of the rhythm to ventricular fibrillation and thus SD.
The debate over inclusion of TTE in athletic preparticipation screening is even more complex, and there are no guidelines in Europe or the United States endorsing routine use of TTE unless the history, physical examination, family history, or screening ECG warrants further testing. Much as with ECG, TTE has historically struggled to define normal with regard to physiologic versus pathologic hypertrophy. This process remains challenging and controversial, but great progress in this area has been made and with astute analysis the gray zone between normal adaptation to training and abnormal physiology has been clarified, giving us sharper and more powerful clinical tools to separate the two.4,51,52

The phenotypic expression of the pathologic condition of HCM is highly variable, with affected genotypes manifesting LV wall thickness ranging from normal (<12 mm) to mild to moderate hypertrophy (13–15 mm) to severe (30–50 mm).4,17,53 Normal physiologic adaptation to training frequently shows increases in LV wall thickness into the upper limit of normal and in some cases LV wall thickness extends into the gray zone of 13 to 15 mm.52,54 This finding characterizes the problem of overlap between the elite athlete and the spectrum of HCM, with cases disguised and difficult to recognize coming from both groups.

However, there are several features that careful echocardiographic analysis can assess to help unravel this complex overlap and separate normal adaptation from HCM. LV end-diastolic cavity dimensions of greater than 55 mm are common in trained athletes but rare in HCM. The LV cavity in HCM is usually small and dilation rarely occurs in the context of late-stage systolic dysfunction and progression to dilated cardiomyopathy.4,55 Systolic anterior motion (SAM) of the anterior leaflet of the mitral valve, although definitively part of the HCM spectrum, is not a feature of the athlete’s heart.4 SAM typically occurs with small LV cavity size and would be unlikely in an athlete whose training included significant amounts of isotonic/dynamic exercise,
with those effects of volume loading leading to a larger than normal cavity size. Again, this finding is not absolute because one could conceive of a scenario in which primarily isometric/static training could lead to hypertrophy and in the proper setting of dehydration could lead to SAM associated with smaller LV cavity size. Diastolic abnormalities of pulsed wave Doppler or tissue Doppler are absent in athletes,\textsuperscript{56,57} thus these findings suggest a pathologic condition such as HCM or restrictive/constrictive physiology. Areas of regional hypertrophy rather than concentric hypertrophy are also more suggestive of HCM. This is 1 area in which CMR has been shown to have greater sensitivity than TTE.\textsuperscript{58-60} Usage of TTE as a diagnostic tool to assess this complex area can be enhanced but if screening TTE is too limited then our ability to differentiate may also be compromised within the screening process.

Gender and race continue to be important in screening TTE. Female athletes are less likely to have LV cavity dilation, and of the 600 elite female athletes undergoing screening TTE, none had LV wall thickness greater than 12 mm.\textsuperscript{61} As noted earlier, race has a more limited impact on females but remains important to consider because black female athletes had LV wall thickness measurements 0.6 mm greater than their white female counterparts.\textsuperscript{34} Accordingly, the gray zone problem is more common among male elite athletes than female, and an astute screening echocardiographer would be concerned that LV wall thickness of greater than 12 mm associated with normal or small cavity size in a female athlete likely represents HCM.\textsuperscript{4} Examples of athletic heart and HCM are shown in Fig. 7.

CMR performed with gadolinium can detect fibrosis of the heart within the spectrum of HCM and late gadolinium enhancement (LGE) may also have important prognostic value (Fig. 8).\textsuperscript{62,63} There are cases that are difficult and perhaps impossible to resolve and it is prudent to reevaluate all athletes diagnosed by screening with pathologic hypertrophy or HCM after detraining to reassess wall thickness and determine if the hypertrophy persists or regresses. CMR can be of diagnostic value to assess acute myocarditis or persistent sequelae of myocarditis that could pose long-term risk to the athlete (Fig. 9). Genetic testing has a limited role and can only confirm a positive mutation; a negative test does not exclude HCM because of the frequency of spontaneous and yet undetected mutations.

The congenital anomalous coronary artery is responsible for a significant remaining percentage of athletic SD (14%),\textsuperscript{6} particularly the left coronary artery arising from the right sinus, with fewer cases involving the right coronary artery arising from the left sinus.\textsuperscript{18} This anomaly escapes detection by traditional screening methods in the United States because only a few have premonitory symptoms and of those all (9/9) had a normal ECG and, when performed, stress ECG with maximal exercise was also normal (6/6).\textsuperscript{18} The only safe and noninvasive opportunity to diagnose this anomaly in preparticipation screening is TTE, which can identify the right and left coronary arteries in most elite athletes (95%).\textsuperscript{64} However, recognition of abnormal coronary anomalies is another matter and in the practical context, coronary origins and courses are not routinely assessed in most adult echo laboratories. Accordingly, routine and broad-based screening using sonographers with limited experience with normal coronaries and even less experience with anomalous coronary arteries seems an inadequate solution to this difficult problem. By contrast, coronary origins and courses are routinely assessed in congenital echo laboratories because of the frequent occurrence of anomalous coronary arteries within the spectrum of congenital heart disease. Because both CMR and CT angiography are expensive and require contrast intravenously for definitive anatomic clarification, we believe that a careful congenital approach to TTE may be the only hope of having any impact with successful screening for this disorder.

TTE easily detects dilation of the ascending aorta at the sinuses of Valsalva, which could herald dissection or rupture in undetected Marfan syndrome or related diseases
Fig. 7. (A–D) ECG and TTE of the athletic heart (Fig. 7A, B) and HCM (Fig. 7C, D). (A) African American Division I basketball player. ECG shows diffusely increased QRS voltage without QRS widening (narrow arrows) and upwardly convex ST elevation followed by inverted T waves (broad arrows). This ECG shows the more pronounced abnormalities of the athletic heart seen in African American athletes. (B) This same athlete’s short-axis TTE image shows a mild increase in septal wall thickness to 1.25 cm (narrow arrows) with mild LV cavity dilation of 6.0 cm (broad arrows) and a corresponding increase in LV mass. This is a representative example of athlete’s heart in an elite African American athlete. (C) An 18-year-old high-school football player with HCM. The ECG reveals increased QRS voltage but with mild prolongation in QRS duration (narrow arrows) and prominent T-wave inversion (broad arrows). (D) Short-axis image of a TTE in an African American Division I basketball player with HCM and exertional angina. The septum is abnormally hypertrophied, measuring 1.6 cm (narrow arrows) and the LV cavity size is 5.0 cm, which is considerably smaller than noted with the athletic heart. Stress echocardiography revealed complete systolic LV cavity obliteration.
of the vascular system. The aortopathy associated with the BAV occurs in approximately 50% of all patients, including those with functionally normal and minimally abnormal valves that easily evade detection. At the University of Virginia, screening echocardiography has detected 3 male athletes with BAV of 854 athletes and one has an associated aortopathy with a root that is mild to moderately enlarged at 4.4 cm (Fig. 10). None of these diagnoses was known before TTE screening. The BAV aortopathy can involve the sinuses of Valsalva but also can isolate to the ascending aorta above the sinotubular junction, which can be overlooked with standard adult TTE imaging. With a complete congenital protocol it is routine to image this aspect of the aorta 1 interspace above the standard long-axis sinus view and rotate the transducer clockwise toward the short-axis view to ideally image this difficult anatomic area. Additional attention to the transverse arch and descending thoracic aorta from suprasternal views coupled with detailed subcostal imaging of the abdominal aorta provides additional diagnostic accuracy in forms of mild coarctation and other vascular disorders that affect the aorta remote from the sinuses of Valsalva.

Focused congenital imaging can also detect a host of other previously undetected conditions, as noted earlier (see Box 1). Although most are not life threatening, altered physiology could affect performance and lead to exercise-related symptoms of cardiovascular compromise.

CARDIOVASCULAR SCREENING IN THE ELITE ATHLETE: OUR VIEW OF THE FUTURE

Sports medicine and sports training have become increasingly more sophisticated, providing comprehensive care for all aspects of the athlete’s health. This is the
foundation of the medical home of the athlete and the subspecialty care of elite athletes with known or suspected abnormalities must follow this initiative, with providers experienced in the complex interplay between normal physiologic adaptation and pathologic conditions. Our current status with the cardiovascular care of elite athletes in many ways mirrors the problem facing the adult congenital heart disease (ACHD) population 20 years ago. It was widely recognized that a large population of patients was emerging with a unique variety of care issues without a sufficient workforce of knowledgeable providers available regionally to ensure the best possible care. It would be of great benefit to all elite athletes if centers of care including expertise in screening and evaluation of athletes could be developed regionally (similar to the programs currently developed for adults with congenital heart disease) to provide

Fig. 9. (A–C) 21-year-old African American Division I college basketball player with myocarditis. This athlete initially presented with syncope and had a second syncopal episode 5 months later. (A) ECG reveals persistent diffuse ST segment elevation (narrow arrows) followed by T-wave inversion (broader arrows). (B) Cardiac MRI shows LGE of the epicardial aspect of the anteroapex and inferoapex (arrows). (C) Late epicardial enhancement of the right ventricular free wall is also noted (leftward arrow). The ECG is suggestive of chronic pericarditis and myocarditis, and the typical MRI finding of epicardial LGE is characteristic of this disorder compared with the patchy LGE/fibrosis seen in HCM. Electrophysiologic study revealed easily inducible rapid ventricular tachycardia, and the athlete is currently restricted from competition.
Fig. 10. (A–F) TTE examples of a familial BAV syndrome in 2 brothers, both of whom are Division I lacrosse players, and one of whom has an associated congenital aortopathy. (A) Parasternal short-axis view of an 18-year-old athlete with a BAV (arrows) with fusion of the right and left coronary cusps and a horizontally oblique aperture. (B) Parasternal long-axis view with color Doppler showing moderate aortic insufficiency (AR) oriented directly toward the anterior leaflet of the mitral valve (arrow). This posterior orientation of AR is characteristic of prolapse of the anterior cusp. (C) Parasternal long-axis view showing the typical systolic doming of the aortic cusps (arrow) and measurement of the ascending aorta at the sinuses of Valsalva is normal for body surface area at 3.4 cm (red line bordered by x’s). (D) Parasternal short-axis view of 21-year-old brother with a near identical valve with the same fusion pattern and aperture (arrows). (E) Parasternal long-axis view showing a similar orientation of the AR by color Doppler (arrow). In this brother the AR is mild to moderate. (F) Parasternal long-axis view showing the presence of a congenital aortopathy in the older brother. Maximal dilation is mild to moderate at 4.4 cm when measured at the sinuses of Valsalva (red line bordered by x’s). Both brothers are eligible to play based on current guidelines. Because of the hemodynamic effects of elite training that could accelerate either the AR or the aortopathy, both athletes are being evaluated by TTE every 6 months.
colleges, universities, and national and professional teams with referral options for
thorough and experienced evaluations.

One can easily envision that larger universities particularly at the Division I level are
in a good position to develop this expertise and establish programs, with the primary
mission being the cardiovascular health and well-being of the athlete with secondary
but important goals of physician training and research. Providers with a broader expe-
rience in screening are in the best position to interpret screening diagnostic studies
and to guide the athlete through any further diagnostic testing. The relationship of
the physician provider and the athlete has received particular attention because of
potential personal, ethical, legal, or institutional conflicts of interest.66,67 Although
these conflicts can never be fully resolved, in general the physician should be
committed first to the health and safety of the athlete and simultaneously balance
the responsibility toward the school or team responsible for the athlete.

Mandated screening currently performed in Italy enjoys the financial support of the
Italian government and backing within Italian law. This strategy is not feasible in the
United States, where, for example, screening colonoscopy or breast mammography
is recommended but not mandated. Instead, the type of program and the choices
diagnostic testing are largely decided by the schools and teams that host the
athletes. We believe that institutions responsible for the well-being of elite athletes
are likely to have significant motivation to protect their athletes from any serious
adverse event. Future screening and ongoing care programs can learn from the large
body of work in this area to develop a more precise set of tools initiated at the moment
of contact with the athlete. Because of the developed depth of our understanding,
both ECG and TTE are useful and appropriate diagnostic tools for cardiovascular
screening of elite athletes.

ECG is useful in the diagnosis of both electrical and structural abnormalities of the
heart. However, ECG is inherently compromised by lack of sensitivity and specificity,
particularly in the detection of structural heart disease. There is considerable overlap
between physiologic and pathologic hypertrophy, and ECG cannot detect abnormal-
ities of the aorta or anomalous coronary artery, and may also be unrevealing in a host
of congenital and acquired conditions that could be dangerous or compromise perfor-
mance. For these reasons we believe that both ECG and TTE should be used together
to enhance diagnostic yield and reduce the rate of false-positive results that could
interrupt training and participation unnecessarily.

Inclusion of TTE into routine cardiovascular screening requires providers and
sonographers with expertise in athlete’s heart and congenital heart disease. We advo-
cate a more congenital approach to TTE screening, with more focus on the aorta
(ascending, arch, descending thoracic, and abdominal), coronary arteries, atrial
septum, and attention to the possibility of regional (especially apical) hypertrophy.
This recommendation supplements standard views with more subcostal and supra-
sternal imaging. We include standard TTE long-axis and short-axis views, and apical
4-chamber and 2-chamber views with careful measurement of LV wall thickness and
mass. All valves and great arteries are subject to pulsed, continuous, and color
Doppler. This strategy includes calculation of pulmonary artery and right atrial pres-
sure. Tissue Doppler is performed if there is LVH, any question of HCM, or left atrial
enlargement. Our early experience indicates that this assessment can be accom-
plished in less than 8 minutes and with progressive experience the duration required
will likely decrease. Although they are noble in intention and common in practice,
a word of caution is due regarding screening programs with limited TTE that do not
record images.68 This practice does not allow for retrospective analysis of data, later
analysis off-line, and, if a death were to occur, this could create legal vulnerability as
a result of the act of screening with no reviewable data. Whether our recommended approach to screening in this manner improves the positive yield of diagnostic testing and simultaneously reduces the rate of false-positive results remains to be seen. We are not aware of previous studies that have favored a more congenital approach to this problem.

If we can follow the lead of the regional programs in ACHD with similar regional programs developed for elite athletes, then a safe medical home will have been created. A reliable network can be established through trainers and athletic departments to market these programs and develop consultation for any athlete within the region. For example, at the University of Virginia there are approximately 700 collegiate athletes. In the remainder of the Commonwealth of Virginia there are more than 100 colleges and universities, with thousands of elite athletes. Those athletes suspected of cardiovascular abnormalities require additional testing including TTE, ETT, CMR/CT angiography, Holter and event monitoring, and electrophysiologic studies. Because most athletes are fully insured, financial models outlining success that are fiscally sustainable may qualify for other sources of internal or external funding that can support these programs. The success of this type of program could lead to more widespread and affordable screening expertise for neighboring institutions and athletes.

The future of cardiovascular ultrasonography (TTE) is rapidly moving toward a less expensive and more widely available technology. Portable laptop-sized echo machines are available at 25% of the cost of larger hospital-based machines. These machines are available for approximately $60,000, and image and data quality are sufficient for these purposes. The next generation is evolving further, with echo capability now available in a size slightly larger than a smart phone for approximately $8000. Accordingly, future screening packages could be offered at a fraction of previously incurred cost. However, caution is encouraged, particularly in the realm of echocardiography, because unlike ECG, TTE is acquired with dynamic imaging and is highly dependent on the skill and experience of both the ultrasonography technician and the interpreting cardiologist. A dynamic interaction between the two produces the most desirable results.

Potential sources of future cardiovascular providers in this area could easily draw from 2 important sources. The first source could come from the large pool of former and current athletes within the realm of the cardiovascular subspecialty. There would be increased job satisfaction among provider athletes motivated to remain involved in sports. It is clear in our program that athletes prefer their providers to have an athletic background. Although meeting this preference is clearly not required, athletes are more comfortable if the unique aspects of their existence are more completely understood. The second source could emerge from providers with expertise in the care of adults with congenital heart disease. These providers have the most extensive experience with the structural conditions likely to be encountered in the athlete, and this new clinical opportunity could provide an exciting and new supplement to their current practice, which, in many cases, is not enough to sustain full-time support of clinical staff.

We see this situation as a unique and exciting opportunity for providers. Athletes are a highly motivated and engaging population. All involved in the program at the University of Virginia find our participation with the athletes to be an energy source, rather than energy sink. We strive toward a 24-hour turnaround for athletes of concern to be evaluated so that training and participation are not interrupted by unnecessary delays. Our program is comprised of sports medicine staff, a wide array of athletic training staff, 5 cardiologists (areas of expertise include ACHD, electrophysiology, and pediatric cardiology), 1 cardiology nurse, and 2 congenital sonographers. Four senior cardiovascular fellows participate in screening, clinical evaluations, grant proposals, and clinical research. Thus far the enthusiasm to participate has exceeded
our ability to accommodate all who would like to be involved. If this article inspires new involvement and the development of new ideas and programs for our athletes then our goals will have been fulfilled.

REFERENCES


