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# Echocardiography in Athletes in Primary Prevention of Sudden Death

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## Abstract

Echocardiography is a noninvasive imaging technique useful to provide clinical data regarding physiological adaptations of athlete's heart. Echocardiographic characteristics may be helpful for the clinicians to identify structural cardiac disease, responsible of sudden death during sport activities. The application of echocardiography in preparticipation screening might be essential: it shows high sensitivity and specificity for identification of structural cardiac disease and it is the first-line imagining technique for primary prevention of SCD in athletes. Moreover, new echocardiographic techniques distinguish extreme sport cardiac remodeling from beginning state of cardiomyopathy, as hypertrophic or dilated cardiomyopathy and arrhythmogenic right ventricle dysplasia. The aim of this paper is to review the scientific literature and the clinical knowledge about athlete's heart and main structural heart disease and to describe the rule of echocardiography in primary prevention of SCD in athletes.

**Keywords:** Athlete's heart, cardiomyopathy, echocardiography, myocardial work, prevention, speckle tracking strain, sudden cardiac death

## INTRODUCTION

The European Society of Cardiology (ESC) definition for sudden death (SD) is a nontraumatic, unexpected fatal event occurring within 1 h of the onset of symptoms in an apparently healthy control. If death is not witnessed, the definition applies when the victim was in good health 24 h before the event.[1] Cardiovascular (CV) diseases are responsible for approximately 17 million deaths every year in the world and about 25% of which are sudden cardiac death (SCD).[2]

A SD may be defined as a SCD when a congenital, or acquired, potentially fatal cardiac condition was known to be present during life, or autopsy has identified a cardiac or vascular anomaly as the probable cause of the event, or no obvious extra cardiac causes have been identified by postmortem examination, and therefore, an arrhythmic event is a likely cause of death.[1]

In last years, the number of sport practitioners has increased by many times. The benefits of sport practice in improving CV health are unquestionable, but an increase in CV events has also been demonstrated during its practice. Therefore, the absolute number of people at risk of SCD during exercise is also increasing.[3]

SCD of an athlete is a rare, but tragic event, which devastates families, institutions, the community, and sports medicine team. It is widely publicized by the media with significant social implications, conveying the idea that such an event can be preventable.

Sport activity might play a trigger role of cardiac arrest in athletes with structural or electrical heart abnormalities, generating malignant arrhythmias, as ventricular fibrillation. The culprit diseases are often clinically silent and unlikely to be suspected or identified on the basis of spontaneous symptoms.[4]

Preparticipation screening (PPS) protocol proposed by the ESC focuses on three points: family and personal history, physical examination, and 12-lead electrocardiogram (ECG), showing

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a 70% sensitivity to detect the most frequent causes of SCD in young athletes.[5] However, some cardiac structural diseases, as incipient forms of cardiomyopathies and anomalous origin of coronary arteries (AOCA), can be missed on physical examination and ECG, but they may be easily identified with echocardiography.

Therefore the echocardiogram might be a useful, accessible, and noninvasive tool to increase sensitivity of screening, to identify cardiac disease, and to prevent SCD in athletes.

The purpose of this work is to define the echocardiographic structural characteristics of the main cardiac pathologies, which may be responsible for SCD in the athletes.

## EPIDEMIOLOGY OF SUDDEN CARDIAC DEATH IN ATHLETES

The assessment of the precise frequency of SCD is hindered by the retrospective nature of most analyses.

SCD related to sport accounts for a little but meaningful proportion of all SCD: recent studies reported around 5% of overall SCD occurring during sporting activity. Considering all sports, SCD occurred only in 6% in young competitive athletes, as compared with 94% among recreational sports participants. Most sports-related SCD occurs in the middle-age group of 35 years or older, mainly in recreational athletes and 80% of these deaths are due to atherosclerotic coronary artery disease (CAD).[6]

Prevalence of CV diseases that predispose to SCD during sports in young athletes is estimated to be 0.2%–0.7%.[7]

In literature, the reported incidence of SCD in young athletes is quite variable. Maron *et al*.[8] reported 0.6 SCD per 100,000 person-years in young competitive athletes in United States, Van Camp *et al*.[9] reported 0.4 SCD in 100,000 athletes per year, in high school and college athletes, while Harmon *et al*.[10] stated that the overall incidence of SCD was 1:53,703 athlete-years, with higher incidence in black athlete compared to white athletes.

Concerning Europe, especially among young Italian athletes, Corrado *et al*.[11] reported the incidence of SCD 3.6/100,000 person-years before routine PPS and 0.4/100,000 person-years after starting routine PPS.

Moreover, an Italian study[12] reported among the nonathletic young people an incidence of SCD 0.9/100,000/year, while the incidence of fatalities among young competitive athletes was estimated to be approximately 2.3/100,000 athletes per year; therefore, the incidence of SCD in young athletes was 2.5-fold greater, suggesting that physical activity rises the risk of SD in this age group.

## ETIOLOGY OF SUDDEN CARDIAC DEATH IN ATHLETES

Different causes of SCD in athletes are related to the age of onset. Atherosclerotic CAD is the most frequent etiology of SCD in adult athletes (35 years or older). In this age group, habitual physical activity may protect against CV events, preventing the development of atherosclerotic CAD by favorable effects on weight reduction, lipid and carbohydrate metabolism, and collateral circle development, reducing atherosclerotic plaques formation in the coronary circulation.[13]

The vast majority of young SD victims have an underlying structural heart disease. It is responsible of the majority of SCD in young athletes (age ≤35 years), providing a substrate for malignant arrhythmias, as ventricular tachycardia/fibrillation, leading to cardiac arrest.[14-18] A wide range of CV diseases (including congenital and inherited heart disorders) may underlie SCD: sport activities act as triggers of cardiac arrest in those athletes affected by CV conditions, which predispose to malignant ventricular arrhythmias during physical exercise[11] [Tables 1 and 2].

## PREPARTECIPATION SCREENING IN SPORT ACTIVITIES

PPS is essential to identify the presence of silent heart diseases and to reduce the risk of sports related SCD.

History, physical examination, and ECG are the least expensive tools of PPS and constitute the primary screening examinations in most guidelines.[19]

Nowadays, the American Heart Association recommendations for PPS of a competitive athlete include essential elements in history and physical examination, especially regarding familiar history of cardiomyopathy or SCD, while physical examination should include careful auscultation to detect any heart murmur, due to left ventricular outflow tract obstruction or valvular disease.[19]

The addition of 12-lead ECG may improve the sensitivity of PPS to identify significant silent cardiac abnormalities: Wolff–Parkinson–White syndrome, ion channelopathies (long QT syndrome and Brugada syndrome), hypertrophic cardiomyopathy (HCM), and arrhythmogenic right ventricular (RV) cardiomyopathy (ARVC).[20]

While in the USA, it is still controversial if the inclusion of an ECG in the regular PPS is cost-effective, in Europe, this recommendation was established more than 10 years ago,[5] and it has been adopted by the different sport committees and international federations.[21]

Italian studies[7,11] showed that PPS of athletes with ECG determined reduction of incidence of SCD from 3.6/100,000 person-years to 0.4/100,000 person-years (*P* < 0.001), representing 90% reduction in sports-related mortality [Figure 1].

It is estimated that 30% of lethal CV causes, like congenital coronary anomalies, aortic dilatation, 10%–30% cases of cardiomyopathies (HCM/ARVC), and premature CAD cannot be identified by PPS with ECG.[8]

## Table 1: Common cardiovascular diseases associated with sudden cardiac deaths in athletes

| Structural cardiac disease | Nonstructural cardiac disease |
|---|---|
| Congenital/genetic | |
| HCM | Brugada syndrome |
| Idiopated dilated cardiomyopathy | WPW syndrome |
| Arrhythmogenic right ventricle cardiomyopathy | Catecholaminergic polymorphic ventricular tachycardia |
| Other cardiomyopathy: Left ventricular non-compaction | Congenital long QT syndrome |
| AOCA | Primitive ventricular fibrillation |
| Valvular heart disease: Mitral valve prolapse | Other ion channelopathies |
| Aortopathy | |
| Acquired | |
| Premature coronary artery disease | Acquired long QT: Drug-induced |
| Myocarditis | Other substance ingestion or environmental factors: Hypo-or hyper-thermia |

WPW=Wolff-Parkinson-White, HCM=Hypertrophic cardiomyopathy, ARVC=Arrhythmogenic right ventricle cardiomyopathy, AOCA=Anomalous origin of coronary arteries

## Table 2: Distribution and prevalence of cardiovascular abnormalities associated with sudden cardiac deaths in young athletes (<35 years old)

| Young competitive athlete | % |
|---|---|
| Unexplained[8] | 36% |
| HCM[15] | 11% |
| Primitive ventricular fibrillation[8] | 8% |
| Premature coronary artery disease[16] | 6% |
| Congenital heart disease[8] | 6% |
| Possible HCM[15] | 4% |
| ARVC[56] | 4% |
| Myocarditis[17] | 5% |
| Long QT syndrome[8] | 4% |
| WPW syndrome[8] | 4% |
| Early repolarization syndrome[8] | 2% |
| Mitral valve prolapse[8] | 2% |
| Tunneled LAD coronary artery[18] | 2% |
| Ruptured ascending aorta[55] | 2% |
| Commotio cordis[8] | 2% |

HCM=Hypertrophic cardiomyopathy, WPW=Wolff–Parkinson–White, LAD=Left anterior descending, ARVC=Arrhythmogenic right ventricle cardiomyopathy

Nowadays, there are only few data[22] regarding the cost-effectiveness of adding an echocardiography in the screening: this tool provides higher sensitivity of the PPS, especially in some competitive athletes, practicing sports with long distance (endurance athletes), or high static component (power athletes).[23] Moreover, the echocardiography may be useful to detect cardiac disease that cannot be identified with a standard 12-lead ECG and to make a differential diagnosis between pathologic cardiac hypertrophies and athlete's heart remodeling. Nevertheless, it is less useful to recognize the arrhythmic cardiac diseases responsible for SCD.[24]
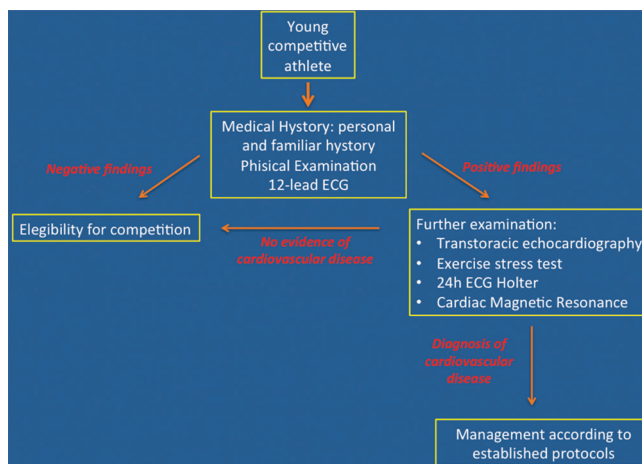


**Figure 1:** Athlete's preparticipation screening according to Italian Guidelines

## ECHOCARDIOGRAPHY IN ATHLETE'S HEART

Long-term physical training causes structural, functional, and electrical heart changes that are physiological responses to the hemodynamic demands of increased cardiac output during effort: this adaptive remodeling can be defined as "athlete's heart."[25] According to the Morganroth *et al.*'s original hypothesis,[26] two main models of training can be identified, which cause two distinct patterns of cardiac remodeling: eccentric hypertrophy in endurance training and concentric hypertrophy in strength training.

Concerning echocardiographic assessment, endurance athletes are characterized by increased left ventricle (LV) end-diastolic diameter, rarely >60 mm, even if ejection fraction (EF) is preserved.[27] Instead, concentric remodeling in power athletes involves all myocardial segments, with a maximal septal thickness <12 mm. Identification of HCM is challenging, when wall thickness is between 13 and 15 mm (the so-called gray zone of left ventricular hypertrophy [LVH]), but after a deconditioning period of at least 3 months, a reduction in wall thickness can be observed in athletes, but not in HCM[28] [Figure 2].

LV remodeling in athletes is associated with normal or increased myocardial relaxation; thus, LV diastolic function is often supranormal, presenting a transmitral E/A ratio often > 2, with typical low A velocity (late diastole). Pulsed tissue Doppler imaging (TDI)-derived early diastolic myocardial velocity (e') of basal septal and basal lateral wall is increased in athletes, responsible of a low E/e' ratio.[29]

Moreover, the athlete's heart can be considered an interesting model of strain variation at different loading conditions because there is a LV adaptation at rest and a load dependency of strain measurement.[30] Speckle tracking may be useful to distinguish physiologic or pathologic LVH, as HCM and amyloidosis [Figure 3].

Concerning the right sections, the RV shows greater inflow and outflow dimensions in athletes compared with sedentary

controls, with no significant difference in the systolic function: RV measures were all significantly greater in endurance athletes, compared to age- and sex-matched strength athletes. Moreover, in highly trained endurance athletes, resting RV global systolic function as measured by fractional area change

and tricuspid annular plane systolic excursion (TAPSE) seems to be lower; TDI and two-dimensional (2D)-strain-derived deformation indexes are reduced at rest, especially at the RV inlet and mid-free wall level.[31]

## ECHOCARDIOGRAPHY IN STRUCTURAL HEART DISEASE

### Hypertrophic cardiomyopathy

Echocardiography is central to the diagnosis and monitoring of HCM.

Measurements of LV wall thickness should be performed at end diastole, preferably in short-axis views and it is essential that all LV segments from base to apex be examined, recording the wall thickness at mitral, mid-LV, and apical levels. In an adult, HCM is defined by a wall thickness ≥15 mm in one or more LV myocardial segments, while in children, LV wall thickness more than two standard deviations greater than the predicted mean (*Z*-score >2) is essential for diagnosis.[32] The clinical diagnosis of HCM in first-degree relatives of patients with assessed HCM is based on the presence of unexplained increased LV wall thickness ≥13 mm in one or more LV myocardial segments.[32]

Often, these patients have resting or induced systolic anterior movement (SAM) of the mitral valve leaflets, resulting in obstruction to the LV outflow tract (LVOTO): it is defined as an instantaneous peak Doppler LVOTO pressure gradient ≥30 mmHg at rest or during physiological provocation (Valsalva maneuver). LVOTO is hemodynamically
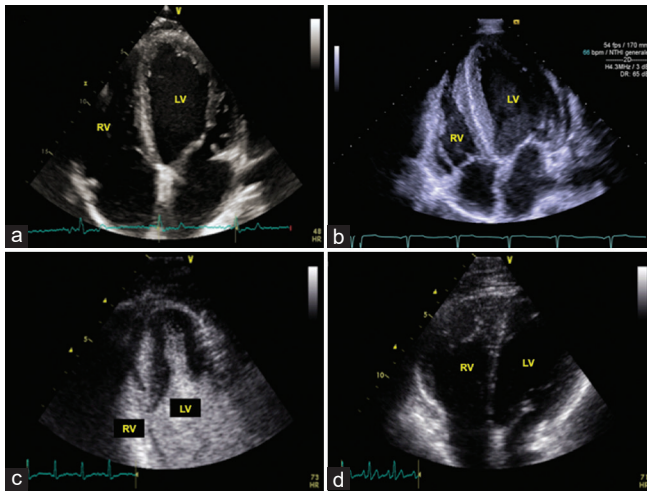


**Figure 2:** Differential diagnosis between physiological and pathological adaptation to training. (a) "bull's eyes" and "athletes" heart of a Caucasian cyclist, with parallel increase in cavity diameters and wall thickness; (b) end-stage hypertrophic cardiomyopathy with mild reduction of left ventricular systolic function and mild enlargement of cavity diameters; (c) apical hypertrophic cardiomyopathy well detected by contrast echocardiography; (d) arrhythmogenic right ventricular dysplasia with right ventricular dilatation and trabeculation. LV: left ventricle; RV: right ventricle
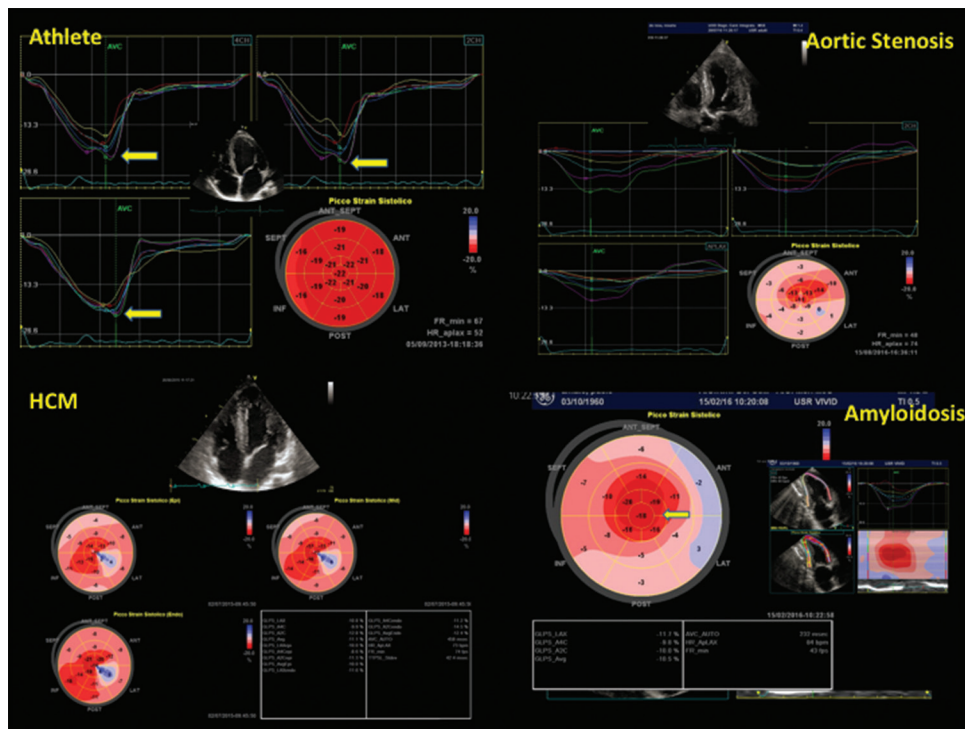


**Figure 3:** Speckle tracking strain bull's eyes eyes in different models of physiologic or pathologic left ventricular hypertrophy. Note the normal deformation in athlete, in contrast with diffuse impairment in aortic stenosis and hypertrophic cardiomyopathy. Conversely, in cardiac amyloidosis, a typical pattern of apical sparing is observed

important when gradient becomes ≥50 mmHg.[33] The left atrium (LA) is often enlarged because of SAM-related mitral regurgitation and elevated LV filling pressures. Patients with HCM often have diastolic dysfunction and the identification of a restrictive pattern (E/A ratio ≥ 2; E-wave deceleration time ≤ 150 ms), associated with high LV filling pressure (E/e' >12–15).[33]

Radial contractile function (EF) is typically normal or increased, while myocardial longitudinal velocities and deformation parameters (strain and strain rate) are often reduced despite a normal EF, especially at the site of hypertrophy; strain parameters may be reduced even before the development of increased wall thickness in genetically affected relatives[34] [Figure 4].

## Idiopathic dilated cardiomyopathy

The hallmark of the idiopathic dilated cardiomyopathy (IDC) is LV dilatation and/or dysfunction, and dilatation may precede dysfunction in many cases.[35]

A comprehensive analysis of regional, as well as global, systolic function is essential in establishing the diagnosis: the presence of regional wall motion abnormalities and the additional finding of echo bright, thinned myocardium may be suggestive of ischemic cardiomyopathy.
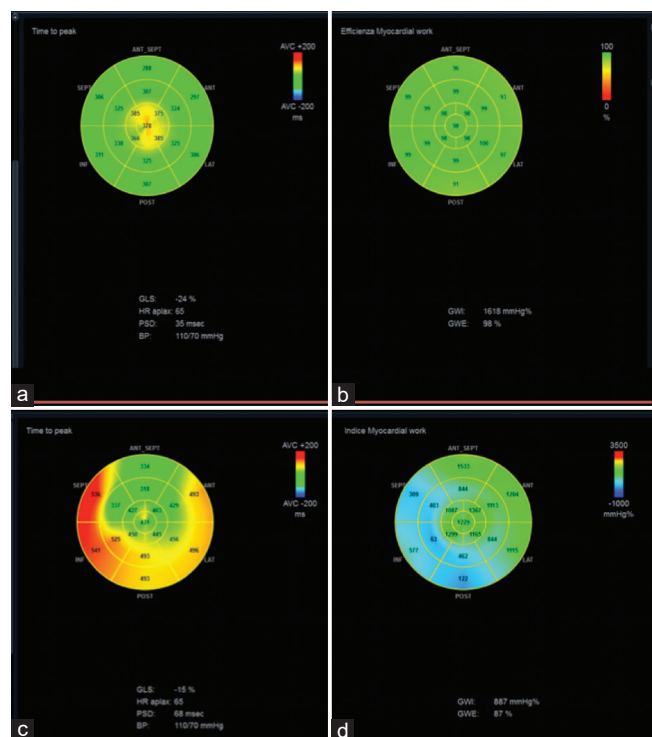


**Figure 4:** Strain and myocardial work analyses in physiological and pathological left ventricular hypertrophy. Upper panels: two-dimensional strain (a) and myocardial work (b) in a power athlete, with normal both global longitudinal strain and myocardial work efficiency. Lower panels: two-dimensional strain (c) and myocardial work (d) in a hypertrophic cardiomyopathy. Note the impairment in both global and regional strain (especially in the interventricular septal region) as well as the reduced myocardial efficiency

The biplane modified Simpson's rule is recommended to establish the EF, despite this technique is highly operator dependent (standard deviation of 8.5% around the mean EF): therefore, three-dimensional echocardiography may improve the reproducibility of LV volume calculation and EF. LV shape can provide additional information: the "sphericity index," ratio between the length (mitral annulus to apex in the apical view) and diameter (mid-cavity level in the short-axis view) is often abnormal in IDC.[36]

As stated before, LV cavity enlargement is part of the cardiac remodeling observed endurance athletes despite this LV dilatation is often mild and indexed LV cavity dimensions are below pathologic limits.

However, in the selected population of athletes, LV remodeling can be extreme: Pelliccia *et al*.[37] showed that more than 10% of elite ultraendurance athletes had LV cavity end-diastolic dimensions >60 mm and simulating forms of DCM, while Gilbert *et al*.[38] revealed a slight LV systolic dysfunction with LVEF around 45%–49% in marathoners.

New echocardiographic techniques may be useful in these cases, for differential diagnosis with IDC, because the adaptive cardiac remodeling shows normal or even supranormal values of strain and strain rate, while in IDC patients, these values are reduced. Finally, stress echocardiography is a further tool in extreme endurance athletes: a normal or supranormal contractile reserve confirm the physiological LV remodeling, while a reduced EF recovery during stress may guide toward the identification of IDC patients.[15]

## Other cardiomyopathies: left ventricular noncompaction

LV noncompaction cardiomyopathy (LVNC) is a rare cardiomyopathy, presenting with multiple deep ventricular trabeculations, secondary to the arrest of normal myocardial development: it is a wide clinical entity, from asymptomatic patients to advanced cases with heart failure and arrhythmias, associated to exercise-related SCD in athletes.[39]

Otherwise, a surprising high prevalence of LV hypertrabeculation has been founded in athletes: studies[40] reported trabeculations in 20% of the athletes, and about 8% complied conventional criteria for the diagnosis of LVNC; this prevalence increased to 13% when only black athletes were considered. This high prevalence suggests that it could be expression of cardiac adaptation to increased preload and afterload, influenced by genetic factors.

Echocardiographic characteristics might be useful to distinguish athletes from disease: the site of trabeculations (apical region in LVNC cardiomyopathy vs. mid-cavity region in athletes), systolic and diastolic function (reduced in LVNC vs. normal in athletes), and the preservation of contractile reserve at the stress echocardiography in athletes with hypertrabeculation.[40]

## Anomalous origin of coronary arteries

AOCA is a rare congenital disease, and it has been recognized as a frequent cause of sports-related SCD. *In vivo* identification

of AOCA is not easy because individuals with this anomaly are often asymptomatic, rarely with symptoms as exertional syncope or chest pain, without ECG signs of myocardial ischemia. Transthoracic 2D echocardiography (TTE) is the only noninvasive tool to visualize the ostia and first tracts of coronary arteries.[41]

The target of TTE is to demonstrate that both the proximal left and right coronary arteries actually originate from their usual coronary sinuses.

Main AOCA associated with SCD in athletes includes the origin of the left main coronary artery from the right aortic sinus or the origin of the right coronary artery from the left sinus[42] [Figure 5].

The execution of simple TTE in the parasternal short-axis projection in the plane of the aortic root may distinguish the two coronary ostia and even identify the initial course of the coronary arteries: Pelliccia *et al*.[43] studied 1360 elite athletes by means of TTE, visualizing the ostium and the proximal portion of left main coronary artery in 97% of the cases and right main coronary artery in 80%.

Moreover, TTE with color-Doppler techniques may be useful to identify the proximal course of the coronary arteries and to determine the flow direction when it is necessary to rule out the presence of an anomaly.[44] Finally, the ostium of the anomalous coronary artery would be smaller, with valve-like ridges, as compared to the normal circular ostium, because the anomalous vessel has to bend over itself to reach, from the opposite sinus of Valsalva, to reach its normal supply territory.[44]

The identification of the coronary ostium in young athletes with chest pain or exertional syncope should be carried out systematically in the TTE examination.

Definitive diagnosis relies on imaging tests such as cardiac magnetic resonance (CMR) or computed tomography (CT)-coronary angiography, which allow to accurately demonstrate the anomalous coronary artery origin and course.[45]

### Valvular heart disease: mitral valve prolapse

Mitral valve prolapse (MVP) is a common valve abnormality in general population. Despite the general belief of a benign
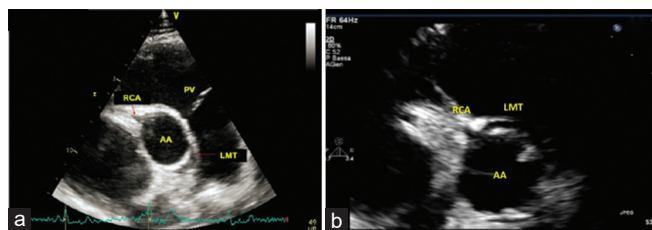


**Figure 5:** Standard echocardiography focused on coronary artery origin from ascending aorta. In a normal individual (a) the separate origin of left main trunk and right coronary artery is easily detected. In a pathologic individual (symptomatic for chest pain) (b) a common origin of left main trunk from right coronary artery is well evidenced

disorder, several studies report SCD in MVP patients, with a substantial percentage of asymptomatic young individuals.[46]

Echocardiography is essential for MVP diagnosis:[47] it is defined by abnormal systolic bulging of one or both leaflets toward the LA with displacement of coaptation point into the LA (>2 mm beyond a line connecting the annular hinge point, ideally in parasternal long-axis view). Leaflets are generally elongated and thickened: "Classic MVP" is characterized by a leaflet thickness ≥5 mm, while nonclassic MVP is defined by leaflet thickness <5 mm. Moreover, mitral annulus is usually enlarged over 28 mm, associated with annular flattening and mitral annular disjunction.

Mitral regurgitation (MR) evaluation is possible through color-Doppler (regurgitant jet area, vena contracta, and flow convergence Proximal Isovelocity Hemisferic Surface Area (PISA)), continuous wave Doppler, and pulsed wave Doppler; to evaluate the hemodynamics consequences of MR, the pulmonary veins flow, and the assessment of LA and LV volumes are necessary. MR is typically in mid-to-late systole and associated with progressive left cardiac chambers' dilatation.[48]

Ventricular and supraventricular arrhythmias are complications of MVP: moderate-to-severe MR has been demonstrated to be an independent risk factor for generating arrhythmias.[49]

Recently, Muthukumar *et al*.[50] described that the "Pickelhaube sign," the high-velocity systolic signal with TDI resembling a spiked helmet, was an indicator of a malignant phenotype in MVP: the tugging of the posteromedial papillary muscle in mid-systole by the myxomatous prolapsing leaflets causes the adjacent posterobasal LV wall to be pulled sharply toward the apex, resulting in the observed spiked configuration of the lateral annular velocities. It is possible that this mechanical traction is arrhythmogenic and may be responsible of SCD.

Finally, Spartalis *et al*.[46] concluded that the subset of patients with malignant MVP at higher risk for SCD is characterized by young women with bileaflet MVP, biphasic, or inverted T waves in the inferior leads and complex ventricular arrhythmias with a right bundle branch block morphology on ECG.

### Premature coronary artery disease

Premature CAD is an important substrate for SCD even in young people and athletes. In this population, SCD is often the first manifestation of the disease, without previous history of angina pectoris or previous myocardial infarction. Proximal left anterior descending coronary artery is the typical culprit vessel.[8]

Echocardiography and exercise testing may fail to show myocardial ischemia or arrhythmias:[12] rarely, an echo-inducible myocardial ischemia may be demonstrated by stress echocardiography.

Therefore, early identification of athletes with premature CAD at risk of ischemic cardiac arrest remains a challenge.

### Myocarditis

Myocarditis accounts for 5%–22% of SCDs in younger populations. There appears to be a higher incidence of SCD

occurring in the context of exercise in cases of myocarditis.[51] Myocarditis may provide a myocardial electrical substrate for life-threatening ventricular arrhythmias and SCD, both in acute or healed form.

In acute form, TTE may show global LV dysfunction; however, localized wall motion abnormalities with pericardial effusions are also common. Patients with the acute form may exhibit localized or diffuse myocardial edema, with an increased wall thickness on echo.[52]

In healed form, specificity of routine CV tests for this disease is limited because ECG changes and echocardiographic LV systolic dysfunction, either regional or global, are found in a minority of affected patients: myocardial fibrosis is often confined to small myocardial area and involves outer wall layers (without compromising myocardial thickening), thus the scar is usually undetectable by echocardiography.[53] A nonischemic pattern of delayed gadolinium enhancement on cardiac MR may be useful for scar identification.

In acute phase, prohibition of competitive activity for athletes is essential; however, after a 6-month restriction, the disease process should be evaluated for resolution.[54]

### Aortopathy

Acute aortic dissection or rupture in Marfan syndrome or other aortopathies are important etiologies of SCD in athletes.[18] Echocardiographic measurements should include the largest measured aortic diameter. *Z*-scores may be preferred for determination of normal aortic diameter as opposed to a single aortic dimension, especially in young patients. Aortic dilatation is identified when *Z*-score is >2.0. Mild, moderate, and severe aortic dilatation may be defined by *Z*-score values of 2–3, 3.01–4.0, and >4.0, respectively. Measurements of aortic diameter are different between CT scan (inner edge to inner edge) and echocardiography (leading edge to leading edge). Recently, echo inner edge to inner edge measurements have been adopted, especially for ascending aortic diameters, to minimize the difference between CT scan and echocardiography results.[55] Although mild aortic enlargement may be a normal adaptation to intense training, large increases in aortic size are unusual in athletes and related to an underlying pathological aortopathy, which may be exacerbated by exercise training.

### Arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy, characterized by progressive loss of myocytes and replacement by fibrofatty tissue, creating the substrate for ventricular arrhythmias. It typically involves the RV, but biventricular and left-dominant variants exist.[56]

Vigorous or long-term athletic exercise might facilitate the phenotypic expression of ARVC: competitive sports increase the risk of SCD by 5 fold in athletes with ARVC and ARVC is present in 4%–22% of athletes with SCD.[57]

A meta-analysis[58] regarding the echocardiographic assessment of RV in ARVC stated that patients with ARVC had larger RV outflow tract (RVOT) (mean ± standard deviation; 34 vs. 28 mm, $P < 0.001$), lower TAPSE (17 vs. 23 mm, $P < 0.001$), and myocardial strain (−17% vs. −30%, $P < 0.001$), compared with healthy controls.

Exercise remodeling related to endurance exercise is characterized by RV enlargement, especially in the basal segment, associated with a mild reduction of global RV peak systolic longitudinal strain values at rest; however, lower strain values are mostly an adaptive response, demonstrated by an increased reserve after exercise provocation;[58] ARVC patients show a disproportionate enlargement of the RVOT, associated with significative RV motion abnormalities, essential to fulfill the ARVC diagnostic criteria.[58-59]

A combination of clinical and family history, electrocardiographic, and echocardiographic features is recommended to differentiate both entities;[16] in doubtful cases, cardiac MR may offer an accurate structural and functional RV evaluation, distinguishing segments with fibrosis, dyskinesia, or outflow tract microaneurysms[17] [Figure 6 and Table 3].

### Conclusions

Echocardiography is an essential imaging technique to identify structural heart disease that may be responsible of SCD in athletes.

Differential diagnosis between athlete's heart remodeling and beginning stage of cardiomyopathies may be difficult, but development of new echocardiographic technique, like strain and myocardial deformation analysis, has improved this issue.

In next future, echocardiography will reach a major role in the beginning phases of athlete's heart screening, to detect
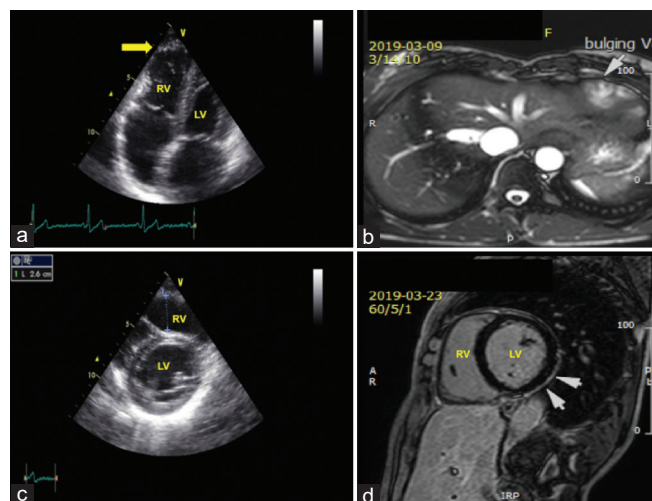


**Figure 6:** Master endurance athlete coming to our department for syncope during competition and nonsustained ventricular tachycardia by electrocardiogram. (a) Right ventricle apical bulging by standard echocardiography, confirmed (b) by cardiac magnetic resonance (see arrows); (c) normal left ventricle and right ventricle diameters, with normal left ventricle systolic function; (d) midventricular left ventricle late enhancement pattern in the inferolateral wall

## Table 3: Echocardiographic differential diagnosis athlete's heart versus cardiomyopathies

| | Athlete's heart | HCM | IDC | LVNC | ARVD |
|---|---|---|---|---|---|
| LV dimension | Normal-mild symmetric hypertrophy <13 mm (Power athlete)<br>Normal-mild dilatation <60 mm (Endurance athlete) | Moderate-severe asymmetric hypertrophy ≥15 mm (Gray zone 13–15 mm) | Moderate-severe dilatation ≥60 mm | Normal in early disease<br>Dilatation in advanced disease | - |
| Diastolic dysfunction (E/A) | Absent | Present | Present | Present | - |
| LV filling pressure (E/e') | Normal | High | High | - | - |
| Left atrial dimension | Normal-mild dilatation | Moderate-severe dilatation | Mild or moderate or severe dilatation | - | - |
| LV GLS | Normal-supranormal | Reduced | Reduced | Reduced | - |
| LV ejection fraction | Preserved (>50%) | Preserved (>50%) | Reduced (<50%) | Normal in early disease<br>Reduced in advanced disease | - |
| Stress echo LV contractile reserve | Normal >10% or supranormal | Normal | Reduced (<10%) | Reduced (<10%) in advanced disease | - |
| LV trabecular location | Midcavity | - | - | Apical | - |
| RV enlargement | Global or RVD1 mild dilatation (Endurance athlete) | - | - | - | Early dilatation |
| Ratio RV/LV volumes | <1 | <1 | <1 | - | ≥1 |
| RV regional motion abnormalities | Absent | Absent | - | - | Present |
| RV systolic function (TAPSE-FAC-GLS) | Normal or supranormal | Normal | - | - | Reduced |

HCM=Hypertrophic cardiomyopathy, IDC=Idiopathic dilated cardiomyopathy, LV=Left ventricle, LVNC=LV non-compaction cardiomyopathy, GLS=Global longitudinal strain, RV=Right ventricle, RVOT=RV outflow tract, FAC=Fractional area change, TAPSE=Tricuspid annular plane systolic excursion, ARVD=Arrythmogenic right ventricular cardiomyopathy, RVD1=Right ventricular diameter

structural cardiac disease in early stages and reduce SCD risk.

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## Conflicts of interest

There are no conflicts of interest.

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