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Arrhythmogenic right ventricular cardiomyopathy — a disease that ends athletic careers

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Abstract: Arrhythmogenic right ventricular cardiomyopathy, commonly detected between the second and fifth decades of life, is caused by mutations that result in the dysfunction of the junctions between cardiomyocytes. However, in 40% of patients, the causative mutation cannot be identified. The detection of arrhythmogenic right ventricular cardiomyopathy is particularly important in endurance athletes, as dangerous ventricular arrhythmias and symptoms of heart failure occur at a younger age in this patient group. The course of arrhythmogenic right ventricular cardiomyopathy involves four stages: no macroscopic changes in the myocardium, clinically overt ARVC, progressive dysfunction of the right ventricle and subsequently the left ventricle and end-stage. Transthoracic echocardiography is a widely available cardiac imaging study. This examination allows for the visualization of segmental or global wall motion abnormalities of the right ventricle. Cardiac magnetic resonance imaging, supplemented by late gadolinium enhancement, is the preferred examination for patients with suspected arrhythmogenic right ventricular cardiomyopathy. An alternative diagnostic study in cases of contraindications is multidetector-row computed tomography. Once a diagnosis of arrhythmogenic right ventricular cardiomyopathy is established, it is recommended to cease sports participation. Only moderate recreational physical activity is permissible.

Keywords: arrhythmogenic right ventricular cardiomyopathy, ARVC, sports medicine, medical examination of athletes.

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Background

Various factors associated with competitive sports can provoke life-threatening arrhythmias in the presence of underlying myocardial diseases, conduction system disorders, ion channelopathies, and heart defects. An example is the effect of ambient pressure changes in individuals

with an atrial septal defect (for instance, during diving or skydiving). It is crucial to perform systematic, periodic examinations of athletes to detect even subtle changes in the subjective assessment (new symptoms not previously reported, such as palpitations or fainting), objective assessment (a new heart murmur for example), and electrocardiographic (ECG) readings by identifying patterns that suggest the emergence of a medical condition which could potentially be life-threatening.

Objective

To define cardiomyopathy and the diagnosis of arrhythmogenic right ventricular cardiomyopathy.

Cardiomyopathies — definition

Cardiomyopathies are a heterogeneous group of pathologies characterized by structural and functional alterations of the heart. [1] Patients with these diseases can be symptomatic or asymptomatic. [2] Presented below are some examples of cardiomyopathies:

— dilated cardiomyopathy, which is one of the main causes of heart failure [3, 4]
— hypertrophic cardiomyopathy (HCM), which is the most common inherited cardiomyopathy caused by mutations in numerous genes
— restrictive cardiomyopathy (RCM), which is a heart-muscle disease characterized by stiffness of the ventricular walls and leads to diastolic dysfunction, raised end-diastolic pressure, and dilated atria [1]
— arrhythmogenic cardiomyopathy (ARCV), which is a pathology characterized by the substitution of the myocardium with fibro-fatty tissue, leading to the development of arrhythmias, reduced systolic function, and sudden cardiac death, especially in young patients [1]
— stress-induced cardiomyopathy, also known as broken-heart syndrome or Takotsubo cardiomyopathy, is connected with an abrupt onset of left ventricular dysfunction in response to severe emotional or physiologic stress. [1, 5]

Arrhythmogenic right ventricular cardiomyopathy — prevalence and clinical course

Arrhythmogenic right ventricular cardiomyopathy is caused by mutations that result in the dysfunction of the junctions between cardiomyocytes (connexons), specifically, impairment of desmosomes, gap junctions, adherens junctions, and ion channels. This leads to a loss of connectivity between cardiomyocytes and, ultimately, to their apoptosis. [6] Consequently, fibro-fatty remodelling of the myocardium occurs. In the final stage, this leads to thinning of the ventricular wall and the formation of aneurysms. [7] However, in 40% of patients, the causative mutation cannot be identified. [8]

Arrhythmogenic right ventricular cardiomyopathy occurs with a frequency of approximately 1:1,300 people. [9] It is most commonly detected between the second and fifth decades of life (and manifests very rarely in childhood). The risk is higher in first-degree relatives, especially siblings. It is more prevalent in males and follows a more severe clinical course in men, which is attributed to the influence of testosterone on the expression of defective genes and typically higher intensity of physical exertion. [7, 8]

The detection of arrhythmogenic right ventricular cardiomyopathy is particularly important in endurance athletes, as dangerous ventricular arrhythmias and symptoms of heart failure occur at a younger age in this patient group. [8, 10]

The course of arrhythmogenic right ventricular cardiomyopathy (ARVC) involves four stages:

I. no macroscopic changes in the myocardium, though arrhythmic episodes leading to sudden cardiac death cannot be ruled out [7, 8]

II. clinically overt ARVC, in which functional and structural changes in the heart muscle are present; patients may report palpitations, dyspnoea, or non-specific chest pain; syncope occurs rarely during this period

III. progressive dysfunction of the right ventricle, and subsequently the left ventricle

IV. end-stage, characterized by the presence of biventricular dilation and systolic dysfunction (which may be misdiagnosed as dilated cardiomyopathy). At this stage, the risk of sudden cardiac death is highest at approximately 4% per year. The primary cause of death is the worsening of heart failure symptoms, which increases the risk of complex ventricular arrhythmias. [7, 8]

The stable phases of ARVC can be interrupted by exacerbations in the form of myocarditis (the so-called "hot phase"). These exacerbations may present as acute coronary syndrome, featuring ECG changes (involving the ST-segment and T-wave) and an increase in markers of myocardial necrosis. [7, 8]

Diagnosis of arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy — ECG changes

ECG changes in various heart diseases are not visible from birth. The electrocardiographic recording of a healthy child changes with age and differs significantly from that of an adult. Certain ECG patterns that are considered normal for children—including young competitive and amateur athletes—would serve as a basis for further diagnostic testing for life-threatening diseases in adults. A primary example is the presence of negative T-waves in three precordial leads among adolescents under the age of 16; in this age group, these are common and, if they occur as isolated findings, are not indicative of pathology. Thorough, repeatable ECG analysis at annual intervals for individuals regularly training in various sports disciplines allows for the identification of those who require more in-depth cardiological diagnostics. This gives us the chance to prevent disease progression or even forestall sudden cardiac arrest resulting from, for instance, life-threatening heart rhythm disorders. Furthermore, athletes periodically face factors that can increase the risk of dangerous arrhythmias, such as electrolyte imbalances after intense exertion and high-stress situations.

Approximately 81–88% of patients with arrhythmogenic right ventricular cardiomyopathy exhibit abnormalities in their resting ECG, which manifest as the disease progresses. [11] If the diagnosis of ARVC is established using other diagnostic modalities, the ECG recording remains normal in half of the patients. [12] However, six years after the first episode of ventricular arrhythmia, abnormal ECG changes are present in every patient. [13]

ECG Abnormalities in patients with arrhythmogenic right ventricular cardiomyopathy:

I. Changes related to right ventricular depolarization disorders:

1. prolonged S-wave upstroke duration (greater than or equal to 55 ms) in leads V1–V3, measured from the nadir of the S-wave to the isoelectric line; due to possible QRS complex fragmentation or the presence of an epsilon wave, the Terminal Activation Delay (TAD) is used. TAD is measured from the nadir of the S-wave to the end of all QRS complex deflections; this parameter is not applicable in the presence of a right bundle branch block (RBBB). [14]
2. QRS complex fragmentation in leads V1–V3. [15]
3. epsilon wave — a low-amplitude, repetitive signal appearing as a notch on the ascending limb of the S-wave in leads V1–V3 (this finding can also occur in other conditions, such as Brugada syndrome, right ventricular myocardial infarction, sarcoidosis, and others). [16]
4. QRS complex widening greater than or equal to 110 ms in leads V1–V3; however, this is not a formal diagnostic criterion.
5. incomplete intraventricular conduction block, characterized by a QRS duration in leads V1–V3 that is 25 ms longer than the QRS duration in leads V4–V6; this is also not a formal diagnostic criterion. [17]
6. r'/S amplitude ratio < 1 in lead V1, proposed for patients with RBBB. This is considered the most reliable diagnostic parameter in cases of intraventricular conduction disturbances. [17]

II. Abnormalities resulting from right ventricular repolarization disorders:
   1. T-wave inversion (TWI) in leads V1–V3 — this is the most useful diagnostic ECG parameter. The presence of negative T-waves in a greater number of leads may indicate right ventricular dilation and increases the risk of ventricular arrhythmia (however, isolated T-wave inversion occurring only in leads V4–V6 is considered less significant).

III. Low QRS voltage in limb leads: this is characterized by low-amplitude QRS complexes in the limb leads (after excluding other causes such as sarcoidosis, pericardial effusion, emphysema, or obesity). This finding is associated with the presence of subepicardial fibro-fatty scarring located within the wall of the left ventricle. [18]

Holter ECG monitoring in patients with arrhythmogenic right ventricular cardiomyopathy

In Holter ECG recordings, the most common supraventricular tachyarrhythmia is atrial fibrillation, [9] the presence of which worsens the prognosis.

Ventricular arrhythmias may occur with varying degrees of severity, ranging from isolated premature ventricular contractions to complex arrhythmias. Characteristically, these ventricular arrhythmias exhibit a left bundle branch block morphology.

A recording of more than 500 premature ventricular contractions per 24 hours during Holter monitoring is a recognized, sensitive diagnostic criterion, useful in the screening of patients with suspected arrhythmogenic right ventricular cardiomyopathy. [19]

Sudden cardiac death in patients with arrhythmogenic right ventricular cardiomyopathy

Sudden cardiac death may be the first symptom of the disease, which is linked to myocardial dysplasia. ARVC is responsible for approximately 4–23% of sudden cardiac deaths occurring in young individuals under the age of 35. [20] This results from ventricular fibrillation during periods of disease exacerbation. In about 75% of cases, sudden cardiac death occurred during daily

activities unrelated to exertion, while in 3–5% of cases, it occurred during sports participation. [21] In the older patient group, monomorphic ventricular tachycardias (VT) are more common. These are triggered by a re-entry mechanism around areas of damaged myocardium during the chronic phase of the disease. [22]

Imaging studies

Transthoracic echocardiography

Transthoracic echocardiography is a widely available cardiac imaging study. This examination allows for the visualization of segmental or global wall motion abnormalities of the right ventricle. Additionally, it is possible to measure the dimensions of the right ventricular inflow and outflow tracts, which can be useful in prognosticating the course of the disease. [23]

Cardiac magnetic resonance imaging

Cardiac magnetic resonance imaging (MRI), supplemented by late gadolinium enhancement (LGE), is the preferred examination for patients with suspected arrhythmogenic right ventricular cardiomyopathy. An alternative diagnostic study in cases of contraindications, such as the presence of certain metal implants, is multidetector-row computed tomography (MDCT). [18] Cardiac magnetic resonance imaging has allowed for the genotypic differentiation of arrhythmogenic cardiomyopathy into right-dominant (ARVC), biventricular, and left-dominant (ALVC) forms. Cardiac MRI enables precise assessment of tissue morphology, thereby eliminating the need for endomyocardial biopsy. [23]

Genetic testing

Genetic testing is recommended for patients diagnosed with arrhythmogenic right ventricular cardiomyopathy, [9] as it enables targeted screening of patients' relatives. Mutations associated with the occurrence of ARVC are present in one in five individuals with a structurally healthy heart. [24] A negative genotyping result does not rule out a diagnosis of the disease.

Due to the lack of a pathognomonic sign for arrhythmogenic right ventricular cardiomyopathy, establishing a diagnosis is based on scoring systems. These scales evaluate the overall clinical picture, taking into account ECG changes in the form of repolarization, depolarization, and conduction disorders, heart rhythm disturbances, as well as significant family history and genetic testing. [18]

Management of arrhythmogenic right ventricular cardiomyopathy according to current standards

In cases where a diagnosis of arrhythmogenic right ventricular cardiomyopathy is established, it is recommended to cease participation in sports. Only moderate recreational physical activity is permissible.

Symptomatic treatment primarily involves the use of beta-blockers or amiodarone. Radiofrequency catheter ablation of arrhythmogenic sites and ICD (implantable cardioverter-defibrillator) implantation are also utilized. [25, 26]

Summary

A thorough patient history (including interviews with athletes attending scheduled appointments with sports medicine physicians to obtain medical clearance for continued training and competition) allows for targeted diagnostics based on reported symptoms (such as palpitations, chest discomfort, or syncope).

A meticulous ECG analysis, repeated at least once a year, enables the detection of new changes in the ECG recording.

If ECG abnormalities occur or heart disease is suspected, the athlete should be referred to a cardiology clinic. There, an echocardiogram, stress test, and, if necessary, cardiac magnetic resonance imaging or other essential diagnostic tests will be performed. Once a diagnosis of arrhythmogenic right ventricular cardiomyopathy is established, it is recommended to cease sports participation. Only moderate recreational physical activity is permissible. Thus, obtaining information about diseases occurring in the family of candidates for regular sports training and athletes that may cause sudden cardiac death is important, because it allows for early detection of potential threats. [27]

Conflict of interest

None declared.

Informed consent

Not applicable.

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