Electrocardiogram and Imaging

An Integrated Approach to Arrhythmogenic Cardiomyopathies

Ketty Savino, MD[a,*], Giuseppe Bagliani, MD[b], Federico Crusco, MD[c], Margherita Padeletti, MD[d], Massimo Lombardi, MD, FESC[e]

KEYWORDS

• Electrocardiography • Cardiac imaging • Echocardiography • Cardiac magnetic resonance
• Arrhythmogenic cardiomyopathies • Arrhythmias

KEY POINTS

- Cardiovascular imaging has improved the management of patients with cardiac arrhythmias.
- Using echocardiography and magnetic resonance, the anatomic pattern of cardiomyopathies can be classified as hypertrophic, dilated, inflammatory, or right ventricular arrhythmogenic.
- A correct approach to the electrocardiogram (basal and during arrhythmias) should be used to identify the anatomic predictors of arrhythmias and sudden cardiac death by imaging techniques.

INTRODUCTION

Cardiovascular imaging radically changes the diagnosis of heart diseases. These tools accurately visualize heart morphology and function, study cardiac hemodynamics, stratify the cardiovascular risk, and address the most effective treatment.

Echocardiography is the most used imaging technique, because of its accuracy, availability, portability, safety, and cost. The transthoracic approach is the first diagnostic step: 2-dimensional (2D) and mono-dimensional (M-mode) examination provide qualitative and quantitative information about chambers size, volumes, and ventricular function. Pulsed, continuous, and color-Doppler studies allow an accurate analysis of cardiac hemodynamics. More recently, tissue Doppler imaging (TDI), speckle tracking, and 3-dimensional (3D) echocardiography have allowed the study of the earliest phases of systolic and diastolic ventricular dysfunction.

Cardiac magnetic resonance (CMR) is a gold-standard technique. Its strengths are the excellent image resolution, the intrinsic high contrast, absence of interference from lungs and bone, the 3D tomographic images, and multiple imaging techniques in a single system. Its disadvantages are the high cost, not widespread diffusion, long

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[a] Cardiology and Cardiovascular Physiopathology, University of Perugia, Piazza Menghini, 1, Perugia 06129, Italy; [b] Arrhythmology Unit, Cardiology Department, Foligno General Hospital, Via Massimo Arcamone, Foligno 06034, Italy; [c] Radiology, Foligno Hospital, Via Massimo Arcamone, Foligno 06034, Italy; [d] Cardiology, Mugello Hospital, Viale della Resistenza, 60, 50032 Borgo San Lorenzo FI, Italy; [e] Multimodality Cardiac Imaging Section, Policlinico San Donato, San Donato Milanese, Piazza Edmondo Malan, 2, 20097 San Donato Milanese MI, Italy
* Correspondig author. Via Beata Chiara Luce Badano 4, Perugia 06125, Italy.
E-mail address: ketty.savino@unipg.it

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acquisition time, and certain contraindications, such as claustrophobia and pacemakers.

Although imaging plays a key role in the setting of cardiomyopathies, it is "only" the integration with clinical history and electrocardiogram (ECG) that provides a comprehensive view of heart disease.

This article discusses the role of imaging in the diagnosis of heart disease that may cause arrhythmias and sudden cardiac death (SCD), integrated with the ECG.

In practical ways, some clinical cases are exposed and are analyzed for ECG and cardiovascular imaging.

From a practical point of view, the authors divided arrhythmogenic heart diseases into 4 groups:

- *Cardiomyopathies with hypertrophic pattern* (hypertrophic cardiomyopathy [HCM], restrictive cardiomyopathies, athlete's heart)
- *Cardiomyopathies with dilative pattern* (ischemic, nonischemic cardiopathy)
- *Inflammatory heart disease* (myocarditis)
- *Right ventricular diseases* (arrhythmogenic right ventricular cardiopathy) (**Table 1**).

Finally, the authors discuss predictors of arrhythmias and SCD, which are highlighted by cardiovascular imaging.

CARDIOMYOPATHIES WITH HYPERTROPHIC PATTERN

The hypertrophic pattern is present in many cardiopathies that have same phenotype but very different causes and physiopathologies.

Table 1
Different pattern in arrhythmogenic cardiomyopathies

Hypertrophic pattern	Hypertrophic cardiomyopathy Restrictive cardiomyopathy • Amyloidosis • Anderson-Fabry disease • Primitive restrictive cardiomyopathy • Sarcoidosis • Athlete's heart
Dilated pattern	Ischemic dilated cardiomyopathy Nonischemic dilated cardiomyopathy
Inflammatory heart disease	Myocarditis
Right ventricular disease	Arrhythmogenic right ventricular cardiomyopathy

Hypertrophic Cardiomyopathy

In HCM, the ECG has a wide spectrum of modifications ranging from normal or near normal to the most frequent changes in ST and T wave, and pathologic Q waves. These modifications can be isolated or combined.[1] In some cases, they are very suggestive of HCM, but they must be interpreted with an imaging tool for a definitive diagnosis.

Echocardiography is the crucial test for the definition of heart anatomy and function, offers a good prognostic stratification, and is recommended for the evaluation of patients with known or suspected HCM.[2] The transthoracic approach identifies the presence, location, and severity of ventricular hypertrophy; in most, the increased thickness is segmental and preferentially involves the basal interventricular septum, lateral wall, and apex, but it can be found at any location of the left ventricle (LV) and right ventricle (RV).[3] The increase of left ventricular mass depends on the extension of thickness segments. Other abnormalities are the systolic anterior movement (SAM) of the mitral valve, the left ventricular outflow tract obstruction (LVOTO), papillary muscles abnormalities, such as thickness, implant, muscle elongation, and accessory muscles. Color Doppler shows the presence and severity of mitral regurgitation; a midsystolic eccentric jet is suggestive of LVOTO.[4,5]

Atrial dilatation due to the increased left ventricular pressure and mitral regurgitation is important prognostic information.[6] Diastolic dysfunction aids in the understanding of symptoms and the stage of the disease.[7]

Generally, at the 2D echocardiography, radial contractile function is normal (normal ejection fraction [EF]), but left ventricular systolic function is reduced at global longitudinal strain.[8,9]

In most cases, echocardiography can correctly identify HCM diagnosis, all associated abnormalities (valvular regurgitation, presence, location, and grade of LVOTO), and studies of systolic and diastolic dysfunction. Hypertrophy severity, atrial enlargement, and diastolic dysfunction are good prognostic tools.

Unfortunately, there are some cases in which transthoracic echocardiography is limited from a suboptimal acoustic window and cannot accurately identify hypertrophy location or distribution; in these cases, other imaging tools can be useful.[2]

CMR is the gold-standard technique for providing information on ventricular function and morphology; it is particularly indicated for studying segmental thickness, ventricular function, left