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Normal Size of Right Ventricle in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C)

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COLUMN ARTICLE

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is determined by progressive degeneration of the right ventricular myocardium, serious ventricular arrhythmias, fibrous-fatty replacement, and increased risk of sudden death.

The actual prevalence of ARVD/C is difficult to assess; accordingly, the initial studies show that up to 12% of sudden cardiac deaths in patients less than 35 years of age may be related to ARVD/C (2), also disease is more common in male slightly. Aborted or completed sudden death (23%), palpitations (27%) and syncope (26%) are the most common sign and symptom; so, due to these clinical features of the disease, diagnosis is difficult too [1,2].

ARVD/C have distinguished with four distinct clinico-pathologic stages. The earliest stage or "concealed phase," is revealed ultra-structural or microscopic abnormalities of the myocardium. The patient in this stage is asymptomatic. The next phase is characterized by ventricular arrhythmias with origin of the right ventricle, and patients in this phase often complain with palpitations, syncope, or sudden cardiac death. Structural abnormalities of the myocardium are now evident on noninvasive imaging but are localized to the right ventricle. Imaging abnormalities include right ventricular dilation with systolic dysfunction and wall motion defects including akinesia or dyskinesia or localized aneurysms. The third phase is revealed by progressive right ventricle dysfunction, often with subtle left ventricle involvement. This is ultimately followed by the final phase in which the degenerative process results in severe global dilation with biventricular systolic failure in some patients [2-4].

ARVD/C is determined based on the presence of major and minor criteria encompassing genetic, electrocardiographic, pathophysiologic, and histo-pathologic factors. The imaging modalities performed to evaluate right ventricular abnormalities include conventional angiography, echocardiography, radionuclide angiography, ultrafast computed tomography, and cardiac magnetic resonance (CMR) imaging. MR imaging is the optimal technique for diagnose and follow-up of clinically suspected ARVD/C.

Revised Task Force Criteria for Echocardiography contains:

Major

Regional right ventricular akinesia, dyskinesia, or aneurysm and one of the following criteria:

a. Right ventricular outflow tract (RVOT) size in PLAX ≥ 32 mm (≥ 19 mm/m^2)
b. RVOT size in PSAX ≥ 36 mm (≥ 21 mm/m^2)
c. Right ventricular FAC ≤ 33%

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Minor

Regional right ventricular akinesia or dyskinesia and one of the following criteria:

a. RVOT size in PLAX: 29 - 32 mm
b. RVOT size in PSAX: 32 - 36 mm
c. Right ventricular FAC: 33% - 40%

The sensitivity and specificity for the diagnosis of ARVD/C are 75% and 95% for the PLAX view and 62% and 95% for the PSAX view [5].

However, in spite of this fact that RVOT dilatation in PLAX and PSAX are diagnostic criteria for evaluation of ARVD/C, we had three patients with normal right ventricle in four chamber echocardiographic view and also RVOT size in PLAX or PSAX views but had RV dysfunction with RV free wall akinesia or dyskinesia in echocardiography and ARVD/C was confirmed in all three patients by CMR, and even right ventricle and RVOT sizes were normal in CMR too.

So, we suggest a new attention to ARVD/C size criteria in echocardiography and CMR views.

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