# An unusual thrombus in a patient with arrhythmogenic cardiomyopathy

Arda Güler[1], Sinem Aydin[1], Songul Ustundag[2], Ekrem Güler[3], and Gamze Babur Guler[1]

[1]Istanbul Mehmet Akif Ersoy Gogus Kalp Ve Damar Cerrahisi Egitim Ve Arastirma Hastanesi
[2]Medicine Hospital Istanbul Turkey
[3]Istanbul Medipol Universitesi Tip Fakultesi

August 22, 2022

**Abstract**

Herein we present a case of a right ventricular (RV) thrombus in a patient with arrhythmogenic cardiomyopathy (ACM). The 24-year old female patient was diagnosed with ACM after echocardiography and cardiac magnetic resonance imaging. Interestingly, at echocardiography, an unusal thrombus formation was detected at RV lateral wall. Also, CMR confirmed the thrombus and oral anticoagulant therapy was started. Even in arrhythmogenic right ventricular cardiomyopathy patients thrombi are rarely reported. However, the development of imaging techniques may enable more frequent detection and effective treatment of thrombi in these patients.

## Introduction

Left ventricular (LV) thrombi are frequently encountered in our clinical practice and usually associated with the underlying myocardial disease. However, right ventricular (RV) thrombi are rare and more difficult to diagnose (1). Imaging of the right ventricle in transthoracic echocardiography (TTE) can be difficult and the importance given to right ventricular pathologies is lower than that of the left ventricle. Although RV thrombi can be observed in the presence of severe RV dilatation, wall motion defect and cardiomyopathy, the cases reported in the literature are very limited (2–4). Arrhythmogenic cardiomyopathy (ACM) is a rare disease of genetic origin that can present with life-threatening arrhythmias with RV dilatation and wall motion abnormalities (5). Due to the presence of RV dilatation, aneurysms, and wall motion abnormalities, we may rarely detect RV thrombi in the course of ACM. Hence, in this case, we will try to present a patient with ACM with atypically localized RV thrombus formation.

## Case report

A 24-year-old female patient was admitted to our tertiary center with chest pain. She had no family history of cardiac disease or sudden cardiac death. Her ECG was sinus rhythm and T wave negativity was detected in leads V1-5 . TTE showed a slight decrease in LV ejection fraction as 45%, prominently dilated right heart chambers, decreased RV functions (TAPSE: 11, tricuspid annular peak systolic velocity: 7.7), severe tricuspid regurgitation (Video-1). Additionally, an image of 13.6 x 9.3 mm, hyperechoic, consistent with thrombus was observed in the lateral wall of the RV (Figure-1). The patient underwent cardiac magnetic resonance imaging (CMR) for morphological and functional assessment of the heart. The CMR images showed enlarged RV and right atrium, severe hypokinesia of the RV, dyskinesia in the free wall of RV and paradoxical movement in interventricular septum, and wall irregularity in both ventricles (Figure-2). The late gadolinium images depicted diffuse enhancement in the right ventricular free wall and inferior wall. The left ventricular inferior septum showed midwall enhancement. A nodular mass was noted on the lateral wall of

RV in the vicinity of the tricuspid valve which did not uptake gadolinium and was compatible with thrombus (Figure-3). Based on these findings, the patient was diagnosed with arrhythmogenic cardiomyopathy with biventricular involvement. ICD implantation was planned for primary protection.

Regarding the RV thrombus, we started warfarin and followed up with the effective INR value.

**Discussion**

RV thrombus formation is rare, even in ACM with dominantly RV involvement. Wlodarska et al. reported an annual incidence of 0.5 thromboembolic events per 100 patients in 126 confirmed Arrhythmogenic right ventricular cardiomyopathy (ARVC) patients at a mean follow-up of 99 months (6). In ARVC patients, slowed blood flow with dilated and hypokinetic RV are important risk factors for thromboembolism. Also, a study by Wu et al. showed that female gender and low LVEF were also associated with high thrombus prevalence (7).

In previously reported cases, most of the RV thrombi were located in the RV apex (2,8). In our case, interestingly, we detected the thrombus on the lateral wall. Since RV evaluation with TTE may be insufficient, evaluation of ACM, especially with RV involvement, patients with CMR is of great importance. CMR is an important diagnostic tool in showing wall motion abnormalities, and thanks to its high resolution, it enables the detection of structures such as thrombus that may be missed in TTE.

Treatment strategy in the presence of RV thrombus has been defined for pulmonary embolism, but there are no guidelines for treatment in the presence of cardiomyopathy (9). In a study by Akdis et al., it was showed that RV thrombi were regressed with anticoagulation and the importance of early and adequate anticoagulation was emphasized (8). Surgical thrombectomy may rarely be considered in selected cases that do not resolve despite anticoagulant therapy. Likewise, although the optimal duration for anticoagulation has not been determined, life-long therapy seems to be a reasonable option. There is no study on which anticoagulant agent to choose. Although it is claimed that NOACs can be effective in this regard, we preferred VKA with the classical approach and determined the follow-up under the control of INR as a more accurate approach.

**Conclusions**

To sum up, ACM is a rare cardiac disease associated with sudden cardiac death, where RV thrombi can also rarely be seen. Therefore, it is of great importance that the echocardiographic evaluation is carried out in detail and meticulously, and, if available, to add CMR for diagnostic workup. In patients with intracardiac thrombus, early and effective use of oral anticoagulants and close follow-up are crucial in terms of prognosis and prevention of thromboembolism.

**References**

1. Egolum UO, Stover DG, Lenihan D, Damp JB, Anthony R, Wasserman AM, vd. Intracardiac Thrombus: Diagnosis, Complications and Management. Am J Med Sci. 01 Mayıs 2013;345(5):391-5.

2. Low QJ, Siaw C, Cheo SW, Kim HS, Benjamin Leo CL, Norliza O, vd. A case of arrhythmogenic right ventricular cardiomyopathy with right ventricle thrombus: A case report. Med J Malaysia. Temmuz 2020;75(4):452-4.

3. Nakano M, Yamaguchi Y, Kutsuzawa D, Kumagai K. Rapidly formed right ventricular thrombus detected by intracardiac echocardiography before catheter ablation in a case of arrhythmogenic right ventricular cardiomyopathy. Hear Case Rep. 01 Eylül 2015;1(5):384-5.

4. Kemmner S, Lesevic H, Reents T, Schunkert H, Burgdorf C. Right ventricular thrombus formation in a patient with arrhythmogenic right ventricular dysplasia following radiofrequency ablation. Clin Case Rep. 2016;4(6):554-7.

5. Corrado D, van Tintelen PJ, McKenna WJ, Hauer RNW, Anastastakis A, Asimaki A, vd. Arrhythmogenic right ventricular cardiomyopathy: evaluation of the current diagnostic criteria and differential diagnosis. Eur

Heart J. 07 Nisan 2020;41(14):1414-29.

6. Wlodarska EK, Wozniak O, Konka M, Rydlewska-Sadowska W, Biederman A, Hoffman P. Thromboembolic complications in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. Eur Eur Pacing Arrhythm Card Electrophysiol J Work Groups Card Pacing Arrhythm Card Cell Electrophysiol Eur Soc Cardiol. Ağustos 2006;8(8):596-600.

7. Wu L, Yao Y, Chen G, Fan X, Zheng L, Ding L, vd. Intracardiac thrombosis in patients with arrhythmogenic right ventricular cardiomyopathy. J Cardiovasc Electrophysiol. Aralık 2014;25(12):1359-62.

8. Akdis D, Chen K, Saguner AM, Stämpfli SF, Chen X, Chen L, vd. Clinical Characteristics of Patients with a Right Ventricular Thrombus in Arrhythmogenic Right Ventricular Cardiomyopathy. Thromb Haemost. Ağustos 2019;119(8):1373-8.

9. Barrios D, Chavant J, Jiménez D, Bertoletti L, Rosa-Salazar V, Muriel A, vd. Treatment of Right Heart Thrombi Associated with Acute Pulmonary Embolism. Am J Med. Mayıs 2017;130(5):588-95.

**Figure legends**

**Figure-1:** TTE 4-chamber view shows the thrombus image (white arrow) at RV lateral wall. (TTE: Transthoracic Echocardigraphy, RA: Right atrium, RV: Right ventricle, LV: Left ventricle)

**Figure-2:** CMR 3-chamber view shows the wall irregularity in both right and left ventricles. (CMR: Cardiac magnetic resonance, LA: Left atrium, RV: Right ventricle, LV: Left ventricle)

**Figure-3:** CMR 4-chamber (a) and short-axis (b) LGE views showing non contrast enhancing thrombus (arrows). (CMR: Cardiac magnetic resonance, LGE: Late gadolinium enhancement, RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle)