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Atrial Tachyarrhythmia in Arrhythmogenic Right Ventricular Cardiomyopathy: A Case Report

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ABSTRACT

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a heritable myocardial condition that mostly affects the right ventricle (RV). Atrial involvement is poorly understood and the evidence for atrial involvement remains limited. In this case report, we describe an 18-year-old woman who had ARVC with the atrial tachyarrhythmia and initially presented with palpitations. As a rare disorder, ARVC requires a careful diagnosis to prevent unnecessary delays in patient care. In our case, the definitive diagnosis of ARVC was made as per Revised International Task Force Criteria 2010.

1 | Introduction

Marcus and colleagues first described arrhythmogenic right ventricular cardiomyopathy or dysplasia (ARVC) in 1982 [1]. This inherited condition which is characterized by progressive degeneration and replacement of the right ventricular (RV) myocardium with fibrofatty tissues, which forms the anatomic basis for the reentry ventricular arrhythmias [2]. Patients with ARVC may exhibit symptoms at any age, but younger-onset is more prevalent, and men are more likely to be affected than women (ratio 3:1). The most frequently seen symptoms are syncope, tiredness, and palpitations [3]. Presentation varies from ventricular arrhythmia or congestive heart failure, and sometimes sudden cardiac death [4]. We present a unique case of ARVC with an atrial tachyarrhythmia/supraventricular tachycardia.

2 | Case History/Examination

An 18-year-old female presented to our hospital with chief complaints of palpitation for 1 month. At first, it was exertional and sometimes at rest lasting for 10–15 s which subsided on its own, but later, it was persistent throughout the day. There was no history of shortness of breath, chest pain, bluish discoloration of hands and lips, swelling of limbs, fever, loss of consciousness, blurring of vision, weakness of a body part, loss of appetite, and easy fatigability. There was no past medical history of tuberculosis, recurrent sore throat, lower respiratory tract infection, and any other chronic illness.

On general examination, she was conscious, cooperative and well oriented to time, place and person. The vital signs were

stable; pulse rate was 80 beats/min, regular, with normal volume and character, blood pressure was 100/60 mm of Hg in bilateral upper limbs, jugular venous pressure was normal, temperature was 98°F and respiratory rate was 18 breaths per min. Multiple erythematous macules were also present in bilateral hands and feet. On physical examination, apical impulse was present 12 cm from midsternal line in 5th intercostal space with lateral retraction, left parasternal heave was absent and no added sounds were heard on chest auscultation.

TABLE 1 | Lab test results of the patients.

Lab tests	Results	Reference values
Hemoglobin	14	12–15 g/dL
White blood count	5.6	$4.5–11.0 \times 10^9$/L
Neutrophil count	60	60%–70%
Lymphocyte count	38	20%–40%
Eosinophil count	2	1%–4%
Platelets count	226,000	150,000—450,000/μL
ESR	10	< 20 mm/h
CRP	0.6	< 1 mg/dL
RBS	70	60–140 mg/dL
Calcium	9	8–11 mg/dL
Magnesium	2	1.3–2.2 meq/L
TSH	3.6	0.5–5 mIU/L
Urea	29	20–40 mg/dL
Creatinine	0.9	0.7–1.2 mg/dL
Sodium	140	135–145 mmol/L
Potassium	4.6	3.5–5.5 mmol/L
AST	30	9–40 U/L
ALT	20	7–60 U/L

Abbreviations: ALT, alanine aminotransferase; AST, aspartate aminotransferase; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; RBS, random blood sugar; TSH, thyroid stimulating hormone.

3 | Differential Diagnosis, Investigation, and Treatment

The differential diagnosis of our case includes atrial fibrillation, Wolff–Parkinson White syndrome, hyperthyroidism, and acute anxiety. On investigation, blood work showed normal results (Table 1). Electrocardiogram (EKG) showed an atrial rate of 300 bpm, a ventricular rate of 75 bpm, regular rhythm without axis deviation and *p*-waves showed 4:1 counterclockwise conducted flutter wave (Figure 1). Twenty-four hour Holter ECG monitoring showed atrial flutter with atrial rate of 300 bpm, ventricular rate of 100 bpm, with variable AV block with few epsilon waves (Figure 2).

On imaging, chest X-ray showed cardiomegaly (Figure 3). Echocardiography revealed cardiomegaly with dilated right ventricular outflow tract (RVOT) (Figure 4). A cardiac gadolinium MRI was performed which showed dilated aneurysmal dyssynchronous right ventricular wall motion with ill-defined patchy enhancement along the RV wall and its RV outflow tract (Figure 5). Non-enhancing low signal intensity thrombus was also noted in the RV cavity. The left ventricle (LV) was non-dilated with left ventricular ejection fraction (LVEF) 29% and right ventricular ejection fraction (RVEF) 28% with minimal pericardial effusion. There was mild mitral, aortic, and pulmonary valve regurgitation. The patient also underwent a cardiac biopsy, which showed extensive right ventricular scarring on histopathological examination (Figure 6). These findings were suggestive of Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) with atrial flutter and right ventricular thrombus.

4 | Outcome and Follow-Up

Subsequently, the patient was managed with intravenous furosemide, oral digoxin, and anticoagulated with subcutaneous enoxaparin and oral warfarin. Patient was medically stable after

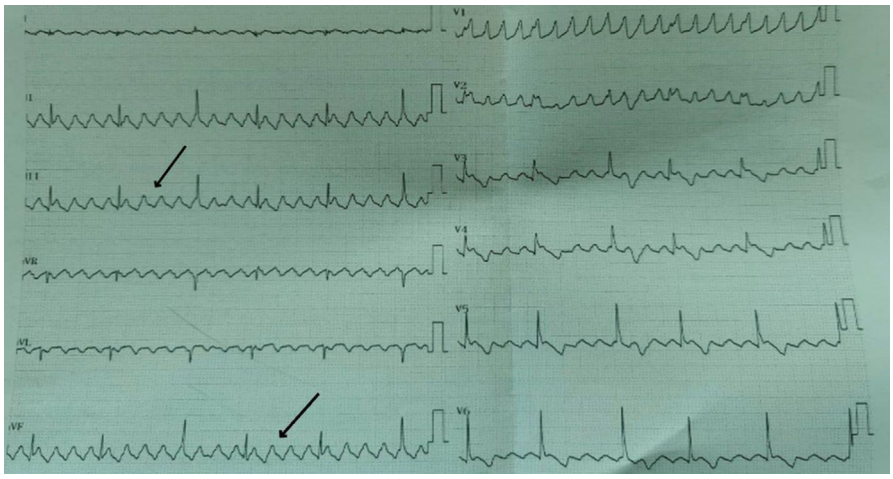

FIGURE 1 | ECG showing atrial flutter with atrial rate of 300 bpm and ventricular rate of 75 bpm, regular rhythm without axis deviation and *f*-wave with conduction rate through AV node of 4:1. Arrow showing typical counterclockwise 4:1 conducted flutter waves.

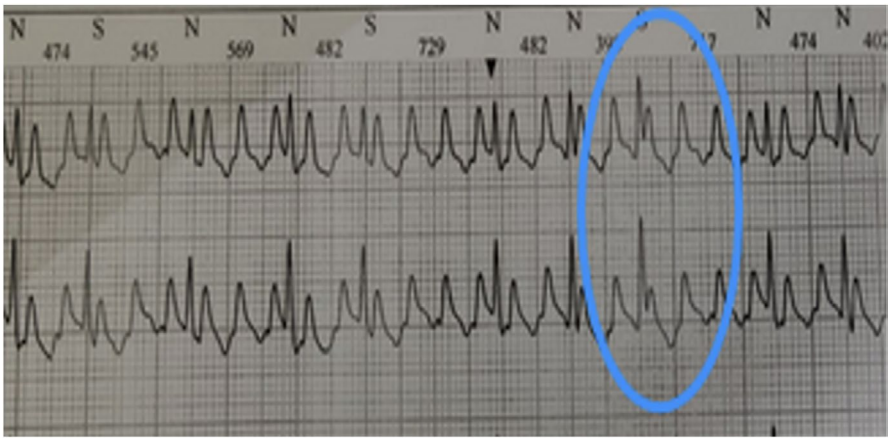

FIGURE 2 | Twenty-four hour Holter monitoring showing irregular R-R interval with epsilon waves. Oval ring shows epsilon waves.

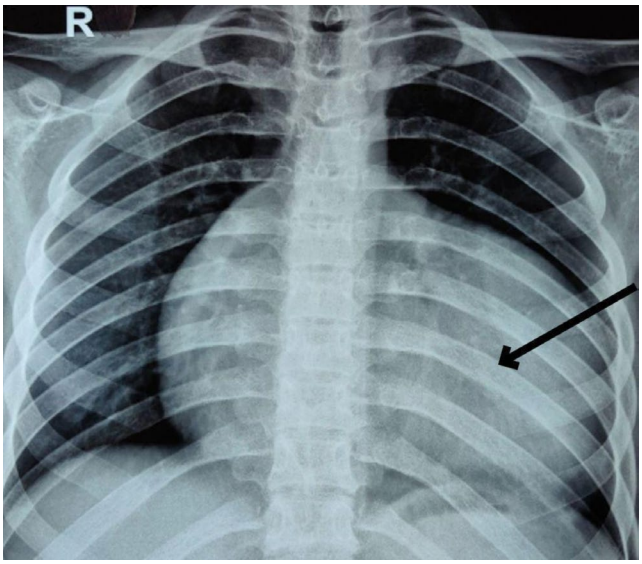

FIGURE 3 | Chest X-ray with posterior–anterior view. Arrow showing cardiomegaly.

5 days after which enoxaparin was discontinued and the patient was discharged with oral metoprolol and enalapril. She was advised to avoid high intensity exercise as it has been associated with severe ventricular dysfunction, arrhythmias and heart failure. Patient was scheduled for long term follow-up plans along with assessment for implantable cardioverter defibrillator if the symptoms reoccur. Considering her reproductive age, the patient was educated on pregnancy along with regular supervision under a multidisciplinary team.

5 | Discussion

ARVC is characterized by fibrofatty infiltration in RV myocardium. The triangle of dysplasia (diaphragmatic, apical, and infundibular region) is the area where infiltration occurs and can progress to aneurysm or dilatation [3]. The right ventricle is predominantly affected with frequent involvement of the left ventricle (LV). However, according to recent data, left ventricle involvement results into 13% of ARVC related death and remaining 87% of those are related to right ventricular involvement [5].

ARVC is predominantly a disease of the cardiac desmosome, a complex of structural proteins, which support myocytes mechanically. Impaired desmosome proteins lead to detachment of myocytes during mechanical stress and results in inflammation and repair, leading to fibrofatty replacement of injured myocytes or even cell death. Desmosomes are present throughout the cardiac system, including the atria, however there is still no evidence for direct atrial involvement with ARVC [6]. There is some data that suggests ARVC may directly affect the atrial chambers, but the human atrial myocardium has thin walls, which restricts the accuracy of standard imaging to detect fibrofatty infiltration [7]. Recently, atrial tachyarrhythmias including atrial fibrillation, atrial flutter and atrial tachycardia, have been reported to be relatively frequently seen in patients with ARVC however, the clinical significance of these atrial arrhythmias, and the factors that predict atrial fibrillation and atrial flutter, are not well understood in these patients [8].

Significant clinical challenges remain in making an early diagnosis of ARVC. No single test can definitively confirm or rule out ARVC. ECG abnormalities are seen in almost 90% of patients. *T* wave, inversions in the V1–V3 leads (a minor diagnostic criterion, but one of the most prevalent ECG abnormalities) is seen in 85% of patients, and QRS duration 110 ms in the V1–V3 [9]. In 30% of instances of ARVC, epsilon waves- post-excitation, electrical potentials that develop at the end of the QRS complex- result from delayed RV activation and are highly specific to ARVD [10]. RV dilation, localized aneurysms, enlarged LA, dilated RV outflow tract, increased reflectivity of moderator

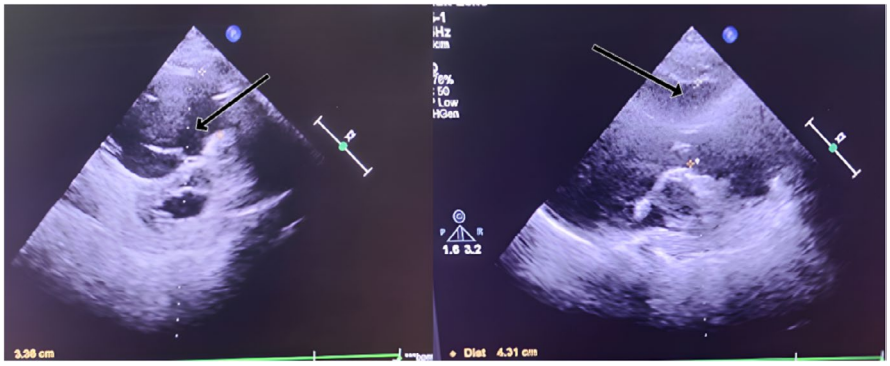

FIGURE 4 | Echocardiography showing cardiomegaly with dilated right ventricular outflow tract (RVOT), pulmonary hypertension, and mild tricuspid regurgitation. Arrow showing dilated RVOT.

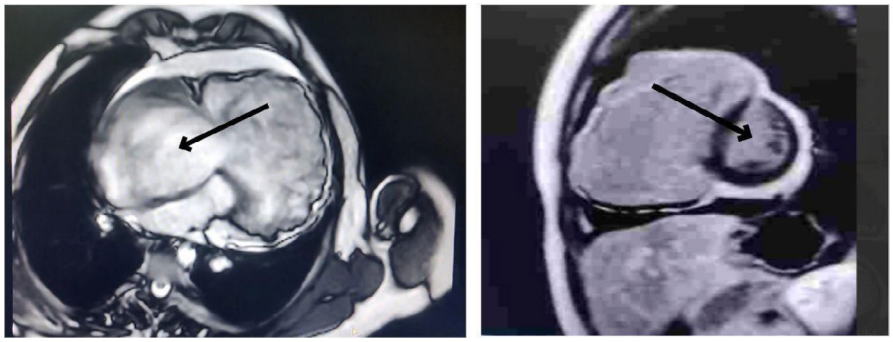

FIGURE 5 | Cardiac MRI showing Aneurysmal dilated right sided cardiac chambers with dyssynchronous right ventricular (RV) wall motion. Arrow showing dilated RV with aneurysm.

band, prominent trabeculations of RV apex, and inferobasal dyskinesis are the main echocardiographic findings. RV end-systolic and end-diastolic diameters, as well as the ratio of RV to LV end-diastolic diameters greater than 0.5, are additional crucial characteristics with 86% sensitivity, 93% specificity, and 86% positive predictive value [10].

The diagnosis of functional impairments, anatomical abnormalities, and the detection of fat or fibrous tissue after they have already occurred can all be greatly aided by Cardiac Magnetic Resonance (CMR) [11]. The histological diagnosis is conclusive, but endomyocardial biopsy is debatable due to the segmental nature of the disease and the fact that samples are typically taken from the septum. Tamponade and perforation are some of the complications that can happen [9]. The definitive diagnosis of ARVC was made according to The 2020 Padua Criteria (Table 2). In our case, the patient met two major criteria thus making a definitive biventricular involvement (Figure 7).

As a rare disorder, ARVC requires a careful diagnosis to prevent unnecessary delays in patient care. Sudden cardiac death (SCD) is the most dreaded ARVC complication, early diagnosis and treatment could save lives. CMR has become the main diagnostic tool for ARVC [11]. Thus, recognizing atrial arrhythmias in arrhythmogenic right ventricular cardiomyopathy reduces the incidence of cardiac comorbidities and mortality [14].

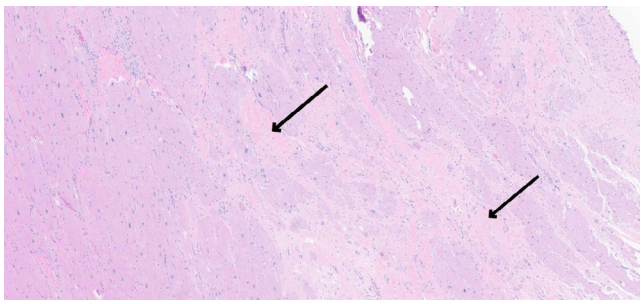

FIGURE 6 | Endomyocardial biopsy. Arrow showing extensive right ventricular fibrosis.

6 | Conclusion

Although ARVC remains a challenging disorder to diagnose, CMR has evolved into a valuable adjunctive diagnostic tool. Atrial arrhythmias are clinically significant in ARVC patients due to their association with inappropriate implanted cardioverter-defibrillator shocks, increased mortality, and heart failure. Recognizing atrial involvement is crucial for comprehensive management of arrhythmogenic right ventricular cardiomyopathy. The patient's life, as well as the lives of their family members and future generations, are affected by this condition further emphasizing the need for thorough evaluation and awareness in these patients.

TABLE 2 | The "Padua criteria" with ACM-arrhythmogenic cardiomyopathy.

	Criteria for RV involvement	Criteria for LV involvement
1. Morpho-functional ventricular abnormalities	By 2D echocardiogram, CMR or angiography: *Major* • Regional RV akinesia, dyskinesia or bulging + any one of the following: ○ Global RV dilatation (increase in RV EDV) or ○ Global RV systolic dysfunction (from image test specific monograms for age and sex) *Minor* Regional RV akinesia, dyskinesia or aneurysm of RV wall	By 2D echocardiogram, CMR or angiography: *Minor* • Global LV systolic dysfunction, with or without LV dilatation (increase in LV EDV) *Minor* • Regional LV hypokinesia or akinesia of LV wall, septum or both
2. Structural myocardial abnormalities	By CECMR: *Major* • Transmural LGE shows stria pattern of ≥ 1 RV regions (inlet, outlet, and apex in two orthogonal views)	By CECMR: *Major* • LV LGE shows stria pattern of ≥ 1 Bull's Eye segments (in two orthogonal views) of the free wall (subepicardial or mid myocardial), septum or both (excluding septal junctional LGE)
3. ECG repolarization abnormalities	*Major* • Inverted T waves in the right precordial leads (V1, V2 and V3) or beyond in individuals with complete pubertal development (with no RBBB) *Minor* • Inverted *T* waves in leads V1 and V2 within individuals with completed pubertal development (in the absence of complete RBBB) • Inverted *T* waves in V1–V4 in individuals with completed pubertal development with the presence of complete RBBB	*Minor* • Inverted *T* waves in left precordial leads (V4–V6) without complete LBBB
4. ECG depolarization abnormalities	*Minor* • Epsilon wave that is, reproducible low amplitude signals between the end of QRS complex to onset of the *T* wave, in right precordial leads (V1–V3) • Terminal activation duration of QRS ≥ 55 ms measured from the nadir of the S wave to the end of the QRS, in V1–V3 (in the absence of complete RBBB)	*Minor* • Low QRS voltages (< 0.5 mV peak to peak) in limb leads
5. Ventricular arrhythmias	*Major* • Frequent ventricular extrasystoles (> 500 per 24 h)/ non-sustained/sustained ventricular tachycardia of LBBB morphology *Minor* • Frequent ventricular extrasystoles (> 500 per 24 h)/ non-sustained/sustained ventricular tachycardia of LBBB morphology with the inferior axis	*Minor* • Frequent ventricular extrasystoles (> 500 per 24 h)/non-sustained/sustained ventricular tachycardia with an RBBB morphology
6. Family history/genetics	*Major* • History of disease in a first-degree relative • Confirmed pathologically at autopsy/ surgery in a first-degree relative • Identification of a pathogenic or likely pathogenetic mutation in the patient *Minor* • History in a first-degree relative in whom it is not possible or practicable to determine whether the family member meets diagnostic criteria or not • Premature sudden death (< 35 years of age) due to suspected disease in a first-degree relative • Confirmed pathologically or by diagnostic criteria in second-degree relative	

Abbreviations: ARVC, arrhythmogenic left ventricular cardiomyopathy; BSA, body surface area; CECMR, contrast-enhanced cardiac magnetic resonance; CMR, cardiac magnetic resonance; EDV, end diastolic volume; EF, ejection fraction; EMB, endomyocardial biopsy; LBBB, left bundle branch block; LGE, late gadolinium enhancement; LV, left ventricle; RBBB, right bundle branch block; RV, right ventricle; RVOT, right ventricular outflow tract.
Source: Adapted from Corrado et al. [12, 13].

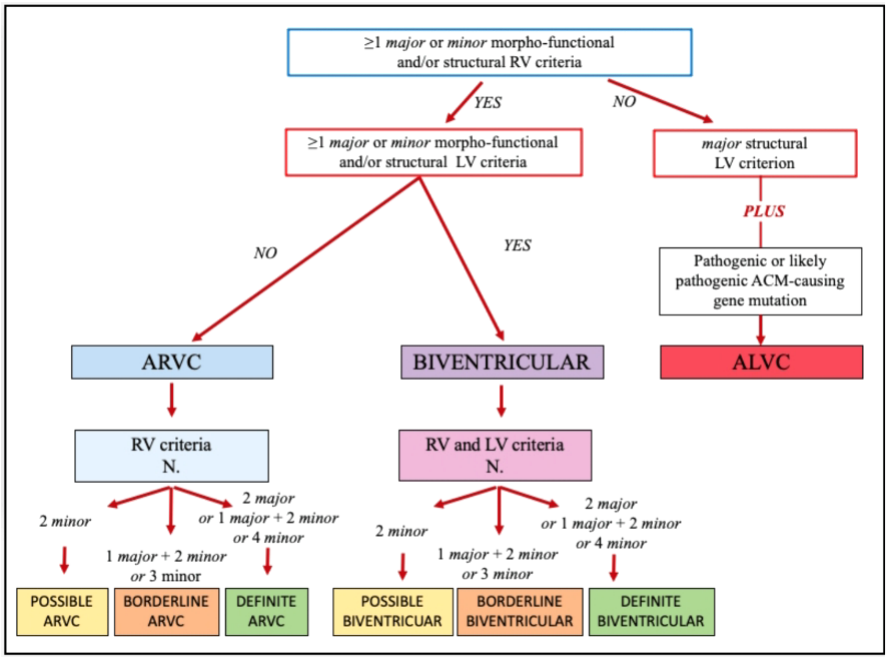

FIGURE 7 | Phenotypic classification of arrhythmogenic cardiomyopathy. Adopted from Graziano et al. [13].

Author Contributions

Shriya Sharma: conceptualization, formal analysis, resources, supervision, writing – original draft. **Ganesh Giri:** conceptualization, writing – original draft, writing – review and editing. **Sadiksha Khadka:** formal analysis, writing – review and editing. **Rohan Goswami:** conceptualization, formal analysis, supervision, visualization.

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Ethics Statement

The authors have nothing to report.

Consent

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

References

1. F. I. Marcus, G. H. Fontaine, G. Guiraudon, et al., "Right Ventricular Dysplasia: A Report of 24 Adult Cases," *Circulation* 65 (1982): 384, https://doi.org/10.1161/01.cir.65.2.384.

2. I. Altun, F. Akin, C. Sahin, and H. Beydilli, "An ECG Changed the Life of a Young Boy: A Case of Arrhythmogenic Right Ventricular Dysplasia," *BML Case Reports* 2014 (2014): bcr2014204703, https://doi.org/10.1136/bcr-2014-204703.

3. R. F. da Silva, K. Morgarbel, C. M. Luize, C. G. Rosa, M. Romano, and I. M. Liguori, "Displasia Arritmogênica Do Ventrículo Direito," *Arquivos Brasileiros de Cardiologia* 91 (2008): e38, https://doi.org/10.1590/S0066-782X2008001600016.

4. N. Güdücü, S. S. Kutay, E. Ozenç, et al., "Management of a Rare Case of Arrhythmogenic Right Ventricular Dysplasia in Pregnancy: A Case Report," *Journal of Medical Case Reports* 5 (2011): 1, https://doi.org/10.1186/1752-1947-5-300.

5. B. Belhassen, H. Shmilovich, E. Nof, et al., "A Case Report of Arrhythmogenic Ventricular Cardiomyopathy Presenting With Sustained Ventricular Tachycardia Arising From the Right and the Left Ventricles Before Structural Changes Are Documented," *European Heart Journal - Case Reports* 4 (2020): 1, https://doi.org/10.1093/ehjcr/ytz239.

6. T. Zghaib, M. Bourfiss, J. F. van der Heijden, et al., "Atrial Dysfunction in Arrhythmogenic Right Ventricular Cardiomyopathy," *Circulation. Cardiovascular Imaging* 11 (2018): e007344, https://doi.org/10.1161/CIRCIMAGING.117.007344.

7. C. Klein, Z. Souissi, F. Brigadeau, and D. Lacroix, "Arrhythmogenic Right Ventricular Cardiomyopathy and Left Atrial Tachycardia: A Case Report," *European Heart Journal - Case Reports* 1 (2017): ytx012, https://doi.org/10.1093/ehjcr/ytx012.

8. A. M. Saguner, S. Ganahl, A. Kraus, et al., "Clinical Role of Atrial Arrhythmias in Patients With Arrhythmogenic Right Ventricular Dysplasia," *Circulation Journal* 78 (2014): 2854, https://doi.org/10.1253/circj.cj-14-0474.

9. Diez ['diego Perez, Brugada'] josep, Diagnosis and Management of Arrhythmogenic Right Ventricular Dysplasia, https://www.escardio.org/Journals/E-Journal-of-Cardiology-Practice/Volume-7/Diagnosis-and-management-of-arrhythmogenic-right-ventricular-dysplasia.

10. U. Rao, S. Agarwal, and T. J. Gilbert, "Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC): Case Report and Review of Literature," *Heart Asia* 6 (2014): 145–149, https://doi.org/10.1136/heartasia-2014-010569.

11. K. H. C. Li, G. Bazoukis, T. Liu, et al., "Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D) in Clinical Practice,"

Journal of Arrhythmia 34 (2018): 11–22, https://doi.org/10.1002/joa3.12021.

12. D. Corrado, M. M. Perazzolo, A. Zorzi, et al., "Diagnosis of Arrhythmogenic Cardiomyopathy: The Padua Criteria," *International Journal of Cardiology* 319 (2020): 106, https://doi.org/10.1016/j.ijcard.2020.06.005.

13. F. Graziano, A. Zorzi, A. Cipriani, et al., "The 2020 'Padua Criteria' for Diagnosis and Phenotype Characterization of Arrhythmogenic Cardiomyopathy in Clinical Practice," *Journal of Clinical Medical Research* 11 (2022): 279, https://doi.org/10.3390/jcm11010279.

14. C. F. Camm, C. A. James, C. Tichnell, et al., "Prevalence of Atrial Arrhythmias in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy," *Heart Rhythm* 10 (2013): 1661, https://doi.org/10.1016/j.hrthm.2013.08.032.