J Cardiovasc Imaging. 2019 Apr;27(2):102-104
https://doi.org/10.4250/jcvi.2019.27.e24
pISSN 2586-7210·eISSN 2586-7296

Journal of Cardiovascular Imaging JCVI

Editorial

Check for updates

Comparison of CMR Findings according to the Presence or Absence of Isolated Focal Right Ventricular Dyskinetic Segments in Patients with Clinical Suspicion of ARVC

Sung Min Ko, MD, PhD

Department of Radiology, Konkuk University Medical Center, Konkuk University School of Medicine, Seoul, Korea

▸ See the article "Patients with Isolated Focal Right Ventricular Dyskinetic Segments: Toward a Better Understanding of This Cohort" in volume 27 on page 93.

OPEN ACCESS

Received: Mar 10, 2019
Accepted: Mar 27, 2019

Address for Correspondence:
Sung Min Ko, MD, PhD
Department of Radiology, Konkuk University Medical Center, Konkuk University School of Medicine, 4-12 Hwayang-dong, Gwangjin-gu, Seoul 05030, Korea.
E-mail: ksm9723@yahoo.co.kr

Copyright © 2019 Korean Society of Echocardiography
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (https://creativecommons.org/licenses/by-nc/4.0/) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ORCID iDs
Sung Min Ko
https://orcid.org/0000-0002-7420-6269

Conflict of Interest
The authors have no financial conflicts of interest.

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by progressive fibrofatty replacement of primarily the right ventricular (RV) myocardium that predisposes to left-threatening ventricular tachycardia and RV dysfunction. ARVC is usually inherited in an autosomal dominant manner and linked to mutations in genes encoding desmosomes or desmosome-related proteins. The prevalence of ARVC has been reported to be 1:1000-1:5000. The disease often affects the RV apex, the base of the right ventricle, and the RV outflow tract (RVOT). Pathology in ARVC may also extend to involve the left ventricle, particularly the posterolateral region. Generally, patients with ARVC are asymptomatic, and unexplained syncope or sudden cardiac death can be the first clinical manifestation in young adults, even in early childhood, and in athletes. ARVC is most commonly diagnosed after an individual presents with arrhythmia findings.[1-3]

Diagnosis of ARVC is based on the presence of major and minor criteria from the Task Force Criteria revised in 2010 (2010 TFC), which includes evaluation of findings from six different diagnostic categories using a combination of electrocardiograms (ECGs) and signal averaged ECGs, imaging studies that include echocardiography, cardiovascular magnetic resonance (CMR), or RV angiography, and arrhythmia presence documented by telemetric monitoring, tissue characterization, genetic data, and family history. Based on this, patients are classified as having a definite, borderline, or possible ARVC.[1]

Imaging findings of ARVC with multi-imaging modalities include localized RV akinesia, dyskinesia, dyssynchronous RV contractions, or aneurysm, RVOT dilatation, RV enlargement, RV dysfunction, trabecular prominence and derangement, scalloped appearance of the RV free wall, abundant epicardial adipose tissue, and intramyocardial fat deposits. These imaging findings may occur after electrical alterations and ventricular arrhythmias. Diagnostic imaging criteria are scored if a regional RV wall motion abnormality is present in combination with RV dilatation or global RV systolic dysfunction.[1-4] Echocardiography is a key imaging modality for diagnosing ARVC, and facilitates the recognition of typical RV morphological and functional abnormalities. However, the evaluation of subtle regional structural changes of the right ventricle with echocardiography is challenging and requires

Generated by xmlinkpress

a high level of expertise.[4][5] CMR can have an important role in diagnosing ARVC and related pathologies. CMR provides more precise definition of the endocardial and myocardial layers in any desired image orientation in terms of the assessment of RV regional wall motion abnormalities. In addition, CMR allows for accurate and reproducible measurement of RV quantitative volumes.[6-8] Borgquist et al.[9] demonstrated that 36 (50%) out of 72 CMR-positive patients fulfilled ARVC criteria by echocardiography among 102 patients with definite ARVC who had undergone both echocardiography and CMR. The diagnostic performance for echocardiography when compared with steady state free precession (SSFP) cine CMR was low; sensitivity was 50% and specificity 70%, positive predictive value 80%, and negative predictive value 37%. Based on study reports, SSFP cine CMR has a higher diagnostic value for the evaluation of RV regional wall abnormalities compared to echocardiography in patients with ARVC. Myocardial fibrosis related to electroanatomic scarring on delayed-enhancement CMR imaging are complementary findings to the radiologic diagnosis of ARVC. However, subjective assessment of RV wall thinning and intramyocardial fat by CMR is prone to false-positive findings. In addition, delayed-enhancement CMR for characterization of an RV myocardium is problematic and unreliable because of the thin wall of the RV and possible confusion with fat.[4][7][8] Accordingly, fatty filtration, wall thinning, delayed-enhancement, and LV involvement are not included from the 2010 TFC.[1]

It remains unclear whether CMR findings with the only focal dyskinetic segments with normal RV volume and/or ejection fraction (EF) are associated with ARVC as part of the disease spectrum or as a disease precursor in patients with palpitation and arrhythmia. In addition, it remains difficult to differentiate normal variations in RV wall motion near the moderator band insertion and pathological RV wall motion abnormalities by CMR.[7] In this issue of the *Journal of Cardiovascular Imaging*, Mansour MJ et al.[10] assessed the CMR findings of 65 patients with clinically suspected ARVC (definite or borderline ARVC in 5 [7.7%] patients, isolated focal dyskinetic RV segments in 27 [41.5%] patients, and no RV dyskinetic segments in 33 [50.8%] patients). They found that 27 patients with isolated RV dyskinetic segments had slightly lower RVEF (55 ± 7% vs. 57 ± 5%), larger RV end-diastolic volume indices (82 ± 12 ml/m^2 vs. 72 ± 12 ml/m^2, p = 0.0127), and a trend for higher odds of dilated right ventricle (odds ratio 3.0 [0.81-11], p = 0.09) compared to 33 patients with no ARVC. However, it is still insufficient to ascertain whether these abnormal CMR findings are associated with part of the ARVC spectrum in patients with clinical suspicion of ARVC in spite of the study results. It is due to several major limitations in this study including the absence of echocardiographic and histological data and clinical and imaging follow-up, lack of ECG and/or Holter monitor findings, the small number of patients, and a single observer. The clinical and prognostic implications of these CMR findings need to be followed up prospectively in larger cohorts with clinical suspicion of ARVC.

REFERENCES

1. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. *Eur Heart J* 2010;31:806-14.
PUBMED | CROSSREF
2. Haugaa KH, Haland TF, Leren IS, Saberniak J, Edvardsen T. Arrhythmogenic right ventricular cardiomyopathy, clinical manifestations, and diagnosis. *Europace* 2016;18:965-72.
PUBMED | CROSSREF
3. Corrado D, Link MS, Calkins H. Arrhythmogenic right ventricular cardiomyopathy. *N Engl J Med* 2017;376:61-72.
PUBMED | CROSSREF

4. Te Riele ASJM, Tandri H, Sanborn DM, Bluemke DA. Noninvasive multimodality imaging in ARVD/C. *JACC Cardiovasc Imaging* 2015;8:597-611.
PUBMED | CROSSREF
5. Mast TP, Teske AJ, Doevendans PA, Cramer MJ. Current and future role of echocardiography in arrhythmogenic right ventricular dysplasia/cardiomyopathy. *Cardiol J* 2015;22:362-74.
PUBMED | CROSSREF
6. Yoo SJ, Grosse-Wortmann L, Hamilton RM. Magnetic resonance imaging assessment of arrhythmogenic right ventricular cardiomyopathy/dysplasia in children. *Korean Circ J* 2010;40:357-67.
PUBMED | CROSSREF
7. Rastegar N, Burt JR, Corona-Villalobos CP, et al. Cardiac MR findings and potential diagnostic pitfalls in patients evaluated for arrhythmogenic right ventricular cardiomyopathy. *Radiographics* 2014;34:1553-70.
PUBMED | CROSSREF
8. te Riele AS, Tandri H, Bluemke DA. Arrhythmogenic right ventricular cardiomyopathy (ARVC): cardiovascular magnetic resonance update. *J Cardiovasc Magn Reson* 2014;16:50.
PUBMED | CROSSREF
9. Borgquist R, Haugaa KH, Gilljam T, et al. The diagnostic performance of imaging methods in ARVC using the 2010 Task Force criteria. *Eur Heart J Cardiovasc Imaging* 2014;15:1219-25.
PUBMED | CROSSREF
10. Mansour MJ, Hamoui O, Asmar J, et al. Patients with isolated focal right ventricular dyskinetic segments: toward a better understanding of this cohort. *J Cardiovasc Imaging* 2019;27:93-101.
CROSSREF