

Characteristics of Left-Dominant Arrhythmogenic Cardiomyopathy

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A 69-year-old woman was admitted to hospital with symptomatic heart failure accompanied by ventricular tachycardia (VT). The ECG displayed a sinus rhythm (**Supplementary Figure, Upper panel**); negative T waves were observed in the lateral precordial V5 and V6 leads. An epsilon wave was detected in leads II, III, and aVF, and the signal-averaged ECG showed positive late potentials (**Figure A**). The VT was characterized by a right bundle branch block (**Supplementary Figure, Lower panel**). Echocardiography showed left ventricular (LV) asynergy, with a 28% ejection fraction (**Supplementary Movie**). Late gadolinium enhancement-magnetic resonance imaging (LGE-MRI) was detected on the anterior wall, interventricular septum (IVS), and inferior-lateral wall (**Figure B**). An ECG-gated contrast computed tomography (CT) scan indicated linear hypointensity in the LV wall and the middle layer of the IVS, suggesting fatty degeneration (**Figure C,D, arrows**), and a scalloped appearance in the LV free wall.¹ ¹²³I- β -methyl-p-iodophenyl-pentadecanoic acid single-photon emission CT (BMIPP-SPECT) revealed a partial defect within the LV inferior-posterior-lateral wall (**Figure F, arrows**), and ¹⁸F-fluorodeoxyglucose positron emission tomography (FDG-PET) displayed mild uptake around the same lesion (**Figure E, arrows**). Electro-anatomical voltage mapping demonstrated that LV polarity was disrupted, but not right ventricular polarity (**Figure G**). Histopathological findings revealed moderate interstitial fibrosis in <50% of the residual myocardium (**Figure H**). Furthermore, genetic tests disclosed a desmoplakin mutation (c.944G>C, p.R315P) related to arrhythmogenic cardiomyopathy (ACM) with LV involvement. The patient

Received June 27, 2021; accepted July 1, 2021; J-STAGE Advance Publication released online August 11, 2021 Time for primary review: 4 days

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ISSN-1346-9843



was diagnosed with ACM based on the Task Force Criteria,² but the LV lesions were insufficient for diagnosis. Therefore, specific characteristics of LV involvement and diagnostic strategies for ACM are required.

Conflict of Interest

None.

Disclosures

N.H. is a member of *Circulation Journal's* Editorial Team.

Reference

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Supplementary Files

Supplementary Movie. Echocardiogram.

Please find supplementary file(s);

<http://dx.doi.org/10.1253/circj.CJ-21-0571>

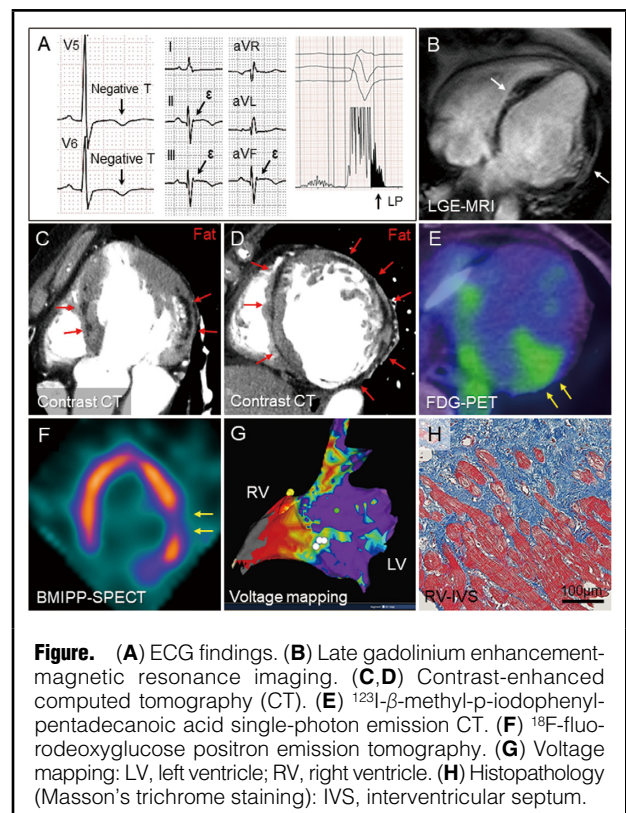


Figure. (A) ECG findings. (B) Late gadolinium enhancement-magnetic resonance imaging. (C,D) Contrast-enhanced computed tomography (CT). (E) ¹²³I- β -methyl-p-iodophenyl-pentadecanoic acid single-photon emission CT. (F) ¹⁸F-fluorodeoxyglucose positron emission tomography. (G) Voltage mapping; LV, left ventricle; RV, right ventricle. (H) Histopathology (Masson's trichrome staining): IVS, interventricular septum.