

REVISED TASK FORCE CRITERIA FOR THE DIAGNOSIS OF ARVC/D¹

Definite diagnosis: 2 major or 1 major and 2 minor criteria or 4 minor from different categories

Borderline: 1 major and 1 minor or 3 minor criteria from different categories

Possible: 1 major or 2 minor criteria from different categories

Acronyms within the following table are explained on the 2nd page of this document.

I. Global and/or Regional Dysfunction and Structural Alterations*

Major (by 2D echo)

- Regional RV akinesia, dyskinesia, or aneurysm
- *and* 1 of the following (end diastole):
 - PLAX RVOT ≥ 32 mm (corrected for body size [PLAX/BSA] ≥ 19 mm/m²)
 - PSAX RVOT ≥ 36 mm (corrected for body size [PSAX/BSA] ≥ 21 mm/m²)
 - *or* FAC $\leq 33\%$

Major (by MRI)

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
 - Ratio of RVEDV to BSA (RVEDV/BSA) ≥ 110 mL/m² (male) or ≥ 100 mL/m² (female)
 - *or* RVEF $\leq 40\%$

Major (by RV angiography)

- Regional RV akinesia, dyskinesia, or aneurysm

Minor (by 2D echo)

- Regional RV akinesia or dyskinesia
- *and* 1 of the following (end diastole):
 - PLAX RVOT ≥ 29 to < 32 mm (corrected for body size [PLAX/BSA] ≥ 16 to < 19 mm/m²)
 - PSAX RVOT ≥ 32 to < 36 mm (corrected for body size [PSAX/BSA] ≥ 18 to < 21 mm/m²)
 - *or* FAC $> 33\%$ to $\leq 40\%$

Minor (by MRI)

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
 - Ratio of RVEDV to BSA (RVEDV/BSA) ≥ 100 to < 110 mL/m² (male) or ≥ 90 to < 100 mL/m² (female)
 - *or* RVEF $> 40\%$ to $\leq 45\%$

II. Tissue Characterization of Wall

Major

- Residual myocytes $< 60\%$ by morphometric analysis, (or $< 50\%$ if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

Minor

- Residual myocytes 60% to 75% by morphometric analysis, (or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

III. Repolarization Abnormalities

Major

- Inverted T waves in right precordial leads (V1, V2, and V3) or beyond in individuals > 14 years of age (in the absence of complete RBBB QRS ≥ 120 ms)

Minor

- Inverted T waves in leads V1 and V2 in individuals > 14 years of age (in the absence of complete RBBB) or in V4, V5, or V6
- Inverted T waves in leads V1, V2, V3 and V4 in individuals > 14 years of age in the presence of complete RBBB

IV. Depolarization/Conduction Abnormalities

Major

- Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3)

Minor

- Late potentials by SAECG in ≥ 1 of 3 parameters in the absence of a QRS duration of ≥ 110 ms on the standard ECG
- Filtered QRS duration (fQRS) ≥ 114 ms
- Duration of terminal QRS < 40 μ V (low-amplitude signal duration) ≥ 38 ms
- Root-mean-square voltage of terminal 40 ms ≤ 20 μ V
- Terminal activation duration of QRS ≥ 55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V1, V2, or V3, in the absence of complete RBBB

V. Arrhythmias

Major

- Nonsustained or sustained VT of LBBB** morphology with superior axis (negative or indeterminate QRS in leads II, III, aVF and positive in lead aVL)

Minor

- Nonsustained or sustained VT of RV outflow configuration, LBBB morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis
- > 500 ventricular extrasystoles per 24 hours (Holter)

VI. Family History

Major

- ARVC/D confirmed in a first-degree relative who meets current Task Force criteria
- ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative
- Identification of a pathogenic mutation† categorized as associated or probably associated with ARVC/D in the patient under evaluation

Minor

- History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria
- Premature sudden death (< 35 years of age) due to suspected ARVC/D in a first-degree relative
- ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative

* Hypokinesia is not included in this or subsequent definitions of RV regional wall motion abnormalities for the proposed modified criteria.

† A pathogenic mutation is a DNA alteration associated with ARVC/D that alters or is expected to alter the encoded protein, is unobserved or rare in a large non-ARVC/D control population, and either alters or is predicted to alter the structure or function of the protein or has demonstrated linkage to the disease phenotype in a conclusive pedigree.

** The original document says "left bundle-branch" or LBB. Dr. Frank Marcus has confirmed this should read "left bundle-branch block" or LBBB.

Acronyms:

aVF: augmented voltage unipolar left foot lead	LBBB: left bundle-branch block	RVEDV: right ventricular end diastolic volume
aVL: augmented voltage unipolar left arm lead	PLAX: parasternal long axis view	RVEF: right ventricular ejection fraction
BSA: body surface area	PSAX: parasternal short axis view	RVOT: right ventricular outflow track
ECG: electrocardiogram	RBBB: right bundle-branch block	SAECG: signal averaged electrocardiogram
FAC: fractional area change	RV: right ventricular	VT: ventricular tachycardia

"Modifications of the original criteria have been proposed to facilitate clinical diagnosis in first-degree relatives who often have incomplete expression of the disease.

According to these recommendations, in the context of proven ARVC/D in a first-degree relative, the diagnosis of familial ARVC/D is based on the documentation of one of the following in a family member:

1. T-wave inversion in right precordial leads V1, V2, and V3 in individuals over the age of 14 years.
2. Late potentials by signal-averaged ECG (SAECG).
3. Ventricular tachycardia of left bundle-branch block morphology on ECG, Holter monitor, or during exercise testing or > 200 premature ventricular contractions in 24 hours.
4. Either mild global dilatation or reduction in RV ejection fraction with normal LV or mild segmental dilatation of the RV or regional RV hypokinesia."¹

¹ Circulation, Apr 2010; 121: 1533 - 1541; "SPECIAL REPORT: Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia: Proposed Modification of the Task Force Criteria" (Frank I. Marcus, William J. McKenna, Duane Sherrill, Cristina Basso, Barbara Bauce, David A. Bluemke, Hugh Calkins, Domenico Corrado, Moniek G.P.J. Cox, James P. Daubert, Guy Fontaine, Kathleen Gear, Richard Hauer, Andrea Nava, Michael H. Picard, Nikos Protonotarios, Jeffrey E. Saffitz, Danita M. Yoerger Sanborn, Jonathan S. Steinberg, Harikrishna Tandri, Gaetano Thiene, Jeffrey A. Towbin, Adalena Tsatsopoulou, Thomas Wichter, and Wojciech Zareba)