

1994 Task Force Criteria

2010 Task Force Criteria

2 major, 1 major + 2 minor, or 4 minor

Definite = 2 major OR 1 major + 2 minor
 Borderline = 1 major + 1 minor OR 3 minor
 Possible = 1 major OR 2 minor

I. Global/regional dysfunction/structural alterations**Major**

- Severe dilatation and reduction of RVEF w/no (or only mild) LV impairment
- Localized RV aneurysms (akinetic or dyskinesic areas w/diastolic bulging)
- Severe segmental dilatation of the RV

By 2D Echo

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

By MRI:

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

By RV Angiography:

- Regional RV akinesia, dyskinesia, or aneurysm

Minor

- Mild global RV dilatation and/or EF reduction with normal LV
- Mild segmental dilatation of the RV
- Regional RV hypokinesia

By 2D Echo:

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

By MRI:

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

II. Tissue characterization of wall**Major**

- Fibrofatty replacement of myocardium on endomyocardial biopsy

- Residual myocytes $< 60\%$ by morphometric analysis (or $< 50\%$ if estimated), w/fibrosis replacement of RV free wall myocardium in ≥ 1 sample, w/ or w/o fatty replacement of tissue on endomyocardial biopsy

Minor

- Residual myocytes 60% to 75% by morphometric analysis (or 50% to 60% if est.) w/fibrous replacement of the RV free wall in ≥ 1 sample, w/ or w/o fatty replacement of tissue on endomyocardial biopsy

III. Repolarization abnormalities**Major**

- TWI (V₁, V₂, V₃) or beyond; > 14 yrs; in absence of complete RBBB QRS ≥ 120 ms

Minor

- TWI in right precordial leads (V₂ and V₃) (people age > 12 yrs, in absence of RBBB)

- TWI in V₁ and V₂; > 14 yrs; in absence of complete RBBB or in V₄, V₅, or V₆
- TWI in V₁ -V₄; > 14 yrs; in presence of complete RBBB

IV. Depolarization/conduction abnormalities**Major**

- Epsilon waves or localized prolongation (> 110 ms) of QRS complex in right precordial leads (V₁ to V₃)

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

Minor

- late potentials (SAECG)

- LP by SAECG in ≥ 1 of 3 parameters in absence of QRS duration of ≥ 110 ms on ECG
 ___ Filtered QRS duration (fQRS) ≥ 114 ms
 ___ Duration of terminal QRS $< 40\mu$ V (LAS duration) ≥ 38 ms
 ___ RMS voltage of terminal 40 ms $\leq 20\mu$ V
- TAD of QRS ≥ 55 ms measured from nadir of S wave to end of QRS, including R', in V₁, V₂, or V₃, in absence of complete RBBB

V. Arrhythmias**Major**

- LBS NSVT or sustained VT (neg or indet QRS in II, III, and aVF and pos in aVL)

Minor

- LBBB sustained or NSVT (ECG, Holter, ETT)
- > 1000 ventricular extrasystoles per 24 hours (Holter)

- NSVT or sustained VT of RV outflow configuration, LBI (pos QRS in II, III, and aVF and neg in aVL) or of unknown axis
- > 500 ventricular extrasystoles per 24 hours (Holter)

VI. Family History**Major**

- Familial disease confirmed at necropsy or surgery

- ARVC/D confirmed in FDR who meets TFC
- ARVC/D confirmed pathologically at autopsy or surgery in FDR
- Pathogenic mutation (assoc or probably assoc w/ ARVC/D) in pt under eval

Minor

- Fam hx of SD (< 35 yrs) due to suspected ARVC/D
- Familial hx (clinical dx based on present criteria)

- Hx of ARVC in FDR in whom not poss or pract to determine if FM meets TFC
- Premature SD (< 35 yrs) due to suspected ARVC/D in FDR
- ARVC/D confirmed pathologically or by current TFC in 2ndDR