### I. Global/regional dysfunction/structural alterations

<table>
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<tr>
<td>2 major, 1 major + 2 minor, or 4 minor</td>
<td>Definite = 2 major OR 1 major + 2 minor</td>
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<td>Borderline = 1 major + 1 minor OR 3 minor</td>
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<td>Possible = 1 major OR 2 minor</td>
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#### Major
- Severe dilatation and reduction of RVEF w/no (or only mild) LV impairment
- Localized RV aneurysms (akineti or dyskinetic 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 ≤ 33%

- By MRI:
  - Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
  - and 1 of the following:
    - Ratio of RV end-diastol vol to BSA ≥ 110 mL/m² (male) or ≥ 100 mL/m² (female)
    - or RV ejection fraction ≤ 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 ≤ 40%

- By MRI:
  - Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
  - and 1 of the following:
    - Ratio of RVEDV to BSA ≥ 100 to <110 mL/m² (male) or ≥ 90 to <100 mL/m² (fem)
    - or RV EF ≥ 40% to ≤ 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/o 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/o w/o fatty replacement of tissue on endomyocardial biopsy

### III. Repolarization abnormalities

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

#### Minor
- TWI in V1 and V2; ≥ 14 yrs; in absence of complete RBBB or in V6, V5, or V4
- TWI in V1, V2, or V3; ≥ 14 yrs; in presence of complete RBBB

### IV. Depolarization/conduction abnormalities

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

- Epsilon wave (reproducible low-amp signals b/t end of QRS complex to onset of T wave) in right precordial leads (V1, V2, V3)

#### Minor
- late potentials (SAECG)

- LP by SAECG in ≥ 1 of 3 parameters in absence of QRS duration of ≥ 110ms on ECG
  - Filtered QRS duration (QRS) ≥ 114ms
  - Duration of terminal QRS ≥ 40µV (LAS duration) ≥ 38ms
  - RMS voltage of terminal 40ms ≤ 20µV
  - TAD of QRS ≥ 55ms 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 (<35yrs) 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