# REVISED TASK FORCE CRITERIA

I. Global and/or Regional Dysfunction and Structural Alterations <sup>*</sup> Major (by 2D echo)		
Regional RV akinesia, dyskinesia or aneurysm.		
And one of the following (end diastole):		
Parasternal long axis view RVOT (PLAX)	$\geq$ 32 mm	
Parasternal short axis view RVOT (PSAX)	$\geq$ 36 mm	
Corrected for body size(PSAX/BSA)	$\geq 21 \text{ mm/m}^2$	
or		
Fractional area change (FAC)	$\leq$ 33%	
Major (by MRI)		
Regional RV akinesia or dyskinesia or dyssynchronous RV contraction		
And one of the following:		
Right ventricular end diastolic volume (RVEDV/BSA)	$\geq 110 \text{ ml/m}^2 \text{male}$	
	$\geq 100 \text{ ml/m}^2 \text{female}$	
OR		
Right ventricular ejection fraction (RVEF)	<i>≤</i> 40%	
Major (by RV angiography)		
Regional RV akinesia, dyskinesia or aneurysm		
Minor (by 2D echo)		
Regional RV akinesia or dyskinesia		
And one of the following (end diastole):		
Parasternal long axis view RVOT (PLAX)	$\ge$ 29 - < 32 mm	
Corrected for body size (PLAX/BSA)	$\geq$ 16 - < 19 mm/m <sup>2i</sup>	
Parasternal short axis view RVOT (PSAX)	$\ge$ 32 - < 36 mm	
Corrected for body size (PSAX/BSA)	$\geq$ 18 - < 21 mm/m <sup>2</sup>	
or		
Fractional area change (FAC)	$>$ 33% - $\leq$ 40%	
Minor (by MRI)		
Regional RV akinesia or dyskinesia or dyssynchronous RV contraction		
And one of the following:		
Right ventricular end diastolic volume/BSA	$\geq$ 100 - < 110 ml/m <sup>2</sup> male $\geq$ 90 - < 100 ml/m <sup>2</sup> female	

#### REVISED TASK FORCE CRITERIA

## OR

Right ventricular ejection fraction (RVEF)

>40% -  $\leq45\%$ 

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 at least 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy.

# Minor

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

III. Repolarization Abnormalities

# Major

Inverted T waves in right precordial leads (V<sub>1</sub>, V<sub>2</sub> and V<sub>3</sub>) or beyond in individuals > 14 years of age (in the absence of complete right bundle branch block QRS  $\ge$  120 msecs).

# Minor

Inverted T waves in leads  $V_1$  and  $V_2$  in individuals > 14 years of age (in the absence of complete right bundle branch block), or in  $V_4$ ,  $V_5$ , or  $V_6$ .

Inverted T waves in leads  $V_1$ ,  $V_2$ ,  $V_3$  and  $V_4$  in individuals > 14 years of age in the presence of complete right bundle branch block.

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 ( $V_1$  to  $V_3$ )

## Minor

Late potentials by signal averaged ECG in at least one of three parameters in the absence of a QRS duration of  $\geq 110$  msecs on the standard ECG.

Filtered QRS duration (fQRS)	$\geq$ 114 msecs
Duration of terminal QRS $< 40 \ \mu V (LAS)$	≥38 msecs
RMS voltage of terminal 40 msecs	${\geq}20~\mu V$

Terminal activation duration of QRS  $\geq$  55ms measured from the nadir of the S wave to the end of the QRS, including R', in V<sub>1</sub>, V<sub>2</sub> or V<sub>3</sub>, in the absence of complete right bundle branch block.

## V. Arrhythmias

# Major

Non-sustained or sustained VT of left bundle branch morphology with superior axis (negative or

#### REVISED TASK FORCE CRITERIA

indeterminate QRS in II, III, AVF and positive in AVL)

#### Minor

Greater than 500 ventricular extrasystoles/24 hours by 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  $^{\pm}$  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 if the family member meets current task force criteria.

Premature sudden death (<35 years) due to suspected ARVC/D in a first degree relative.

ARVC/D confirmed pathologically or by current Task Force Criteria in second degree relative.

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

<sup>†</sup>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.

Diagnostic terminology for original criteria Diagnostic terminology for revised criteria This diagnosis is fulfilled by the presence of two major, or one major plus two minor criteria or four minor criteria from different groups. 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