

**Table S2-- Comparison of task force criteria and patients' conditions**

2010 Task force criteria [1]	ARVC patient
<p>I. Global or regional dysfunction and structural alterations</p> <p><b>Major</b></p> <p>By 2D echo:</p> <ul style="list-style-type: none"> <li>• Regional RV akinesia, dyskinesia, or aneurysm and one of the following (end diastole):               <ul style="list-style-type: none"> <li>— PLAX RVOT <math>\geq 32</math> mm (corrected for body size [PLAX/BSA] <math>\geq 19</math> mm/m<sup>2</sup>)</li> <li>— PSAX RVOT <math>\geq 36</math> mm (corrected for body size [PSAX/BSA] <math>\geq 21</math> mm/m<sup>2</sup>)</li> <li>— or fractional area change <math>\leq 33\%</math></li> </ul> </li> </ul> <p>By MRI:</p> <ul style="list-style-type: none"> <li>• Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following:               <ul style="list-style-type: none"> <li>— Ratio of RV end-diastolic volume to BSA <math>\geq 110</math> mL/m<sup>2</sup> (male) or <math>\geq 100</math> mL/m<sup>2</sup> (female)</li> <li>— or RV ejection fraction <math>\leq 40\%</math></li> </ul> </li> </ul> <p>By RV angiography:</p> <ul style="list-style-type: none"> <li>• Regional RV akinesia, dyskinesia, or aneurysm</li> </ul> <p><b>Minor</b></p> <p>By 2D echo:</p> <ul style="list-style-type: none"> <li>• Regional RV akinesia or dyskinesia and one of the following (end diastole):               <ul style="list-style-type: none"> <li>— PLAX RVOT <math>\geq 29</math> to <math>&lt; 32</math> mm (corrected for body size [PLAX/BSA] <math>\geq 16</math> to <math>&lt; 19</math> mm/m<sup>2</sup>)</li> <li>— PSAX RVOT <math>\geq 32</math> to <math>&lt; 36</math> mm (corrected for body size [PSAX/BSA] <math>\geq 18</math> to <math>&lt; 21</math> mm/m<sup>2</sup>)</li> <li>— or fractional area change <math>&gt; 33\%</math> to <math>\leq 40\%</math></li> </ul> </li> </ul> <p>By MRI:</p>	<p><b>Major</b></p> <p>The patient had regional RV dyskinesia and PLAX/BSA RVOT <math>\geq 21</math> mm/m<sup>2</sup> by 2D echo.</p>

<ul style="list-style-type: none"> <li>• Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following: <ul style="list-style-type: none"> <li>— Ratio of RV end-diastolic volume to BSA <math>\geq 100</math> to <math>&lt; 110</math> mL/m<sup>2</sup> (male) or <math>\geq 90</math> to <math>&lt; 100</math> mL/m<sup>2</sup> (female)</li> <li>— or RV ejection fraction <math>&gt; 40\%</math> to <math>\leq 45\%</math></li> </ul> </li> </ul> <p>PLAX indicates parasternal long-axis view; RVOT, RV outflow tract; BSA, body surface area; PSAX, parasternal short-axis view; aVF, augmented voltage unipolar left foot lead; and aVL, augmented voltage unipolar left arm lead.</p>	
<p>II. Tissue characterization of wall</p> <p><b>Major</b></p> <ul style="list-style-type: none"> <li>• Fibrofatty replacement of myocardium on endomyocardial biopsy</li> <li>• Residual myocytes <math>&lt; 60\%</math> by morphometric analysis (or <math>&lt; 50\%</math> if estimated), with the fibrous replacement of the RV free wall myocardium in <math>\geq 1</math> sample, with or without fatty replacement of tissue on endomyocardial biopsy</li> </ul> <p><b>Minor</b></p> <ul style="list-style-type: none"> <li>• Residual myocytes <math>60\%</math> to <math>75\%</math> by morphometric analysis (or <math>50\%</math> to <math>65\%</math> if estimated), with the fibrous replacement of the RV free wall myocardium in <math>\geq 1</math> sample, with or without fatty replacement of tissue on endomyocardial biopsy</li> </ul>	<p><b>Undetected</b></p>
<p>III. Repolarization abnormalities</p> <p><b>Major</b></p> <ul style="list-style-type: none"> <li>• Inverted T waves in the right precordial leads (V<sub>1</sub>, V<sub>2</sub>, and V<sub>3</sub>) or beyond in individuals <math>&gt; 14</math> years of age (in the absence of complete right bundle-branch block QRS <math>\geq 120</math> ms)</li> </ul> <p><b>Minor</b></p> <ul style="list-style-type: none"> <li>• Inverted T waves in leads V<sub>1</sub> and V<sub>2</sub> in individuals <math>&gt; 14</math> years of age (in the absence of complete right bundle-branch block) or in V<sub>4</sub>, V<sub>5</sub>, or V<sub>6</sub></li> <li>• Inverted T waves in leads V<sub>1</sub>, V<sub>2</sub>, V<sub>3</sub>, and V<sub>4</sub> in individuals <math>&gt; 14</math> years of age in the presence of complete right bundle-branch block</li> </ul>	<p><b>No match</b></p> <p>Inverted T waves in leads V<sub>1</sub>, V<sub>2</sub>, V<sub>3</sub>, in individuals <math>&gt; 14</math> years of age in the presence of complete right bundle-branch block</p>
<p>IV. Depolarization/conduction abnormalities</p> <p><b>Major</b></p>	<p><b>Major</b></p> <p>Epsilon wave (reproducible low-amplitude signals)</p>

<ul style="list-style-type: none"> <li>• Epsilon wave (reproducible low-amplitude signals between the end of QRS complex to the onset of the T wave) in the right precordial leads (V<sub>1</sub> to V<sub>3</sub>)</li> </ul> <p><b>Minor</b></p> <ul style="list-style-type: none"> <li>• Late potentials by SAECG in ≥1 of 3 parameters in the absence of a QRS duration of ≥110 ms on the standard ECG</li> <li>• Filtered QRS duration (fQRS) ≥114 ms</li> <li>• Duration of terminal QRS &lt;40 μV (low-amplitude signal duration) ≥38 ms</li> <li>• Root-mean-square voltage of terminal 40 ms ≤20 μV</li> <li>• Terminal activation duration of QRS ≥55 ms 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</li> </ul>	<p>between end of QRS complex to onset of the T wave) in the right precordial leads (V<sub>1</sub> to V<sub>3</sub>)</p>
<p>V. Arrhythmias</p> <p><b>Major</b></p> <ul style="list-style-type: none"> <li>• Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with a superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)</li> </ul> <p><b>Minor</b></p> <ul style="list-style-type: none"> <li>• Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with the inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of an unknown axis</li> <li>• &gt; 500 ventricular extrasystoles per 24 hours (Holter)</li> </ul>	<p><b>Minor</b></p> <p>Sustained ventricular tachycardia of unknown axis</p>
<p>VI. Family history</p> <p><b>Major</b></p> <ul style="list-style-type: none"> <li>• ARVC/D confirmed in a first-degree relative who met current Task Force criteria</li> <li>• ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative</li> <li>• Identification of a pathogenic mutation† categorized as associated or probably associated with ARVC/D in the patient under evaluation</li> </ul> <p><b>Minor</b></p>	<p><b>No match</b></p>

<ul style="list-style-type: none"> <li>• History of ARVC/D in a first-degree relative in whom it was not possible or practical to determine whether the family member met current Task Force criteria</li> <li>• Premature sudden death (&lt;35 years of age) due to suspected ARVC/D in a first-degree relative</li> <li>• ARVC/D confirmed pathologically or by current Task Force Criteria in a second-degree relative</li> </ul>	
<p><b>Definite diagnosis:</b> 2 major, or 1 major and 2 minor, or 4 minor criteria from different categories; <b>borderline diagnosis:</b> 1 major and 1 minor, or 3 minor criteria from different categories; <b>possible diagnosis:</b> 1 major, or 2 minor criteria from different categories.</p>	<p><b>2 major and 1 minor criteria</b></p>

#### References

1. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. Eur Heart J. 2010; 31: 806-14.

**Table S3. Primers for real-time PCR**

<b>Gene</b>	<b>Forward (5' to 3')</b>	<b>Reverse (5' to 3')</b>
<i>GAPDH</i>	GGTGAAGGTCGGAGTCAACGGATTTGGTCG	GGATCTCGCTCCTGGAAGATGGTGATGGG
<i>OBSCN</i>	GCTGAGCTCCGTGTGGACT	CAGTGGTGGACTCTGTTCCT
<i>ANK1.5</i>	CCAGATGAATGGTTACTCCTCAC	CAAGGGGATGGCGTCTAGG
<i>POU5F1</i>	CAGTGCCCGAAACCCACAC	GGAGACCCAGCAGCCTCAAA
<i>NANOG</i>	CAGAAGGCCTCAGCACCTAC	ATTGTTCCAGGTCTGGTTGC
<i><math>\alpha</math>-actinin</i>	TCCATCGGAGCCGAAGAAATC	GTGTCGGTGGATCAAAGCACA
<i>plakoglobin</i>	TCAGCAGCAAGGGCATCAT	TGGGTGTAAGTGGTGGTTTTCTT
<i>plakophilin2</i>	GCAAATGGTTTGCTCGATTT	GGCTGGTAATCTGCAATGGT
<i>N-cadherin</i>	CCTGCTTCAGGCGTCTGTAG	CTGCCTTTGTAGGTGGCCAC
<i>connexin43</i>	GGAATGCAAGAGAGGTTGAAAG	GGCATTGGAGAACTGGTAGA
<i>desmoplakin</i>	CAGTGGTGTCAGCGATGATGT	TGACGCTGGATATGGTGGAA
<i>C/EBP<math>\alpha</math></i>	CACGAAGCACGATCAGTCCAT	CGCACATTCACATTGCACAAG
<i>FABP4</i>	TGGTGGTGGAAATGCGTCAT	GGTCAACGTCCCTTGGCTTA
<i>PPAR<math>\gamma</math></i>	AGGCCATTTTCTCAAACGAG	CCATTACGGAGAGATCCACG

**Figure S1-Recording of implantable cardioverter defibrillator**



