

# **New Diagnostic Criteria**

## **Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)**

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# Characteristics of ARVC

- **Fibrofatty infiltration of RV myocardium**
- **Ventricular arrhythmia**
- **RV dilatation, aneurysm, dysfunction, ultimately RV and LV failure**
- **Sudden death in youth or athletes**



**JOHANNIS MARIAE  
LANCISII**

*A Secretiori Cubiculo, & Archiatro  
Pontificii*

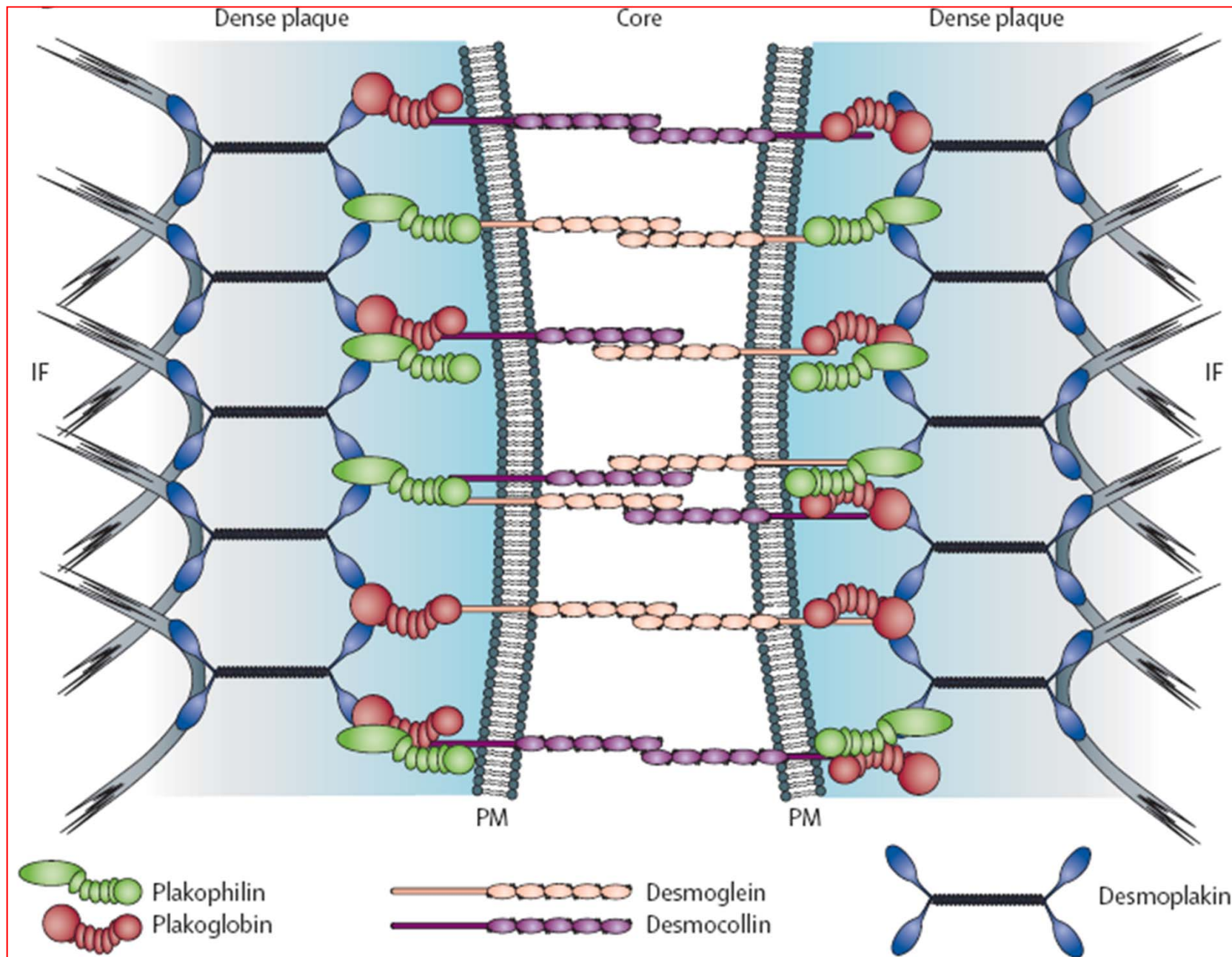
**DE MOTU CORDIS  
ET ANEURYSMATIBUS  
OPUS POSTUMUM  
IN DUAS PARTES DIVISUM.**



**NEAPOLI ANNO MDCCXXXVIII.**  
Excudebat FELIX-CAROLUS MUSCA  
**SUPERIORUM FACULTE.**



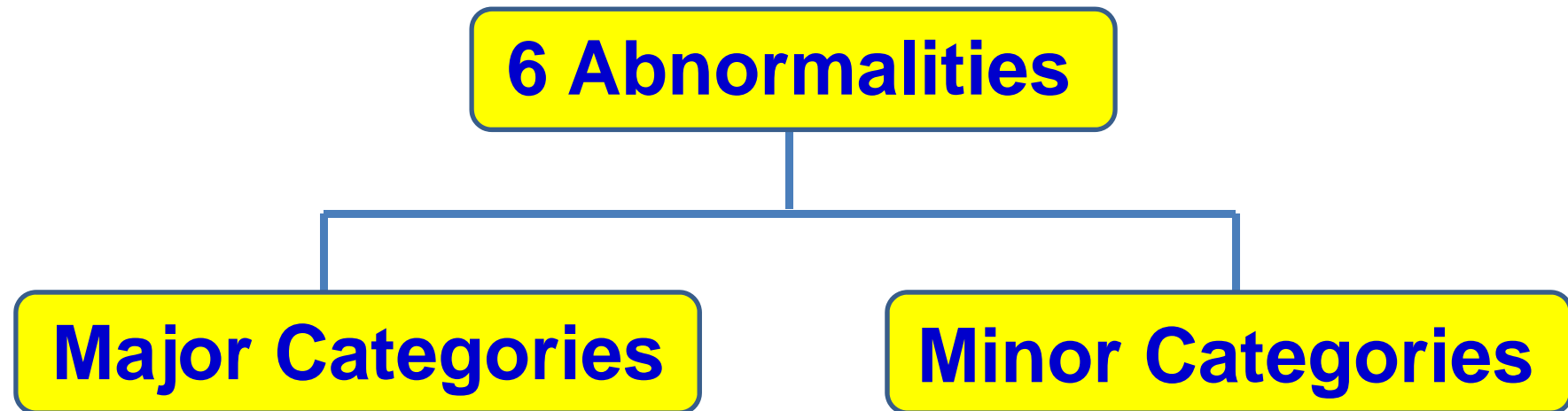
# Intracellular Mechanical Junction (Desmosome) of Cardiomyocyte



# 1994 Task Force Criteria for ARVC

- I. Global and/or regional dysfunction and structural alterations**
- II. Tissue characterization of wall (Biopsy)**
- III. Repolarization abnormalities**
- IV. Depolarization/conduction abnormalities**
- V. Arrhythmias**
- VI. Family History**

# Diagnosis of ARVC



- **Two Major criteria**
- **One Major + two Minor criteria**
- **Four Minor criteria**

# **Criteria for ARVC (1994)**

## **Structural and Functional Changes**

### **Major Criteria**

- **Severe RV dilatation and reduction of RV EF with no (or only mild) LV impairment**
- **Localized RV aneurysms (akinetic or dyskinetic areas with diastolic bulging)**
- **Severe segmental dilatation of RV**

# **Criteria for ARVC (1994)**

## **Structural and Functional Changes**

### **Minor Criteria**

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



# **Limitation of the 1994 Task Force Criteria**

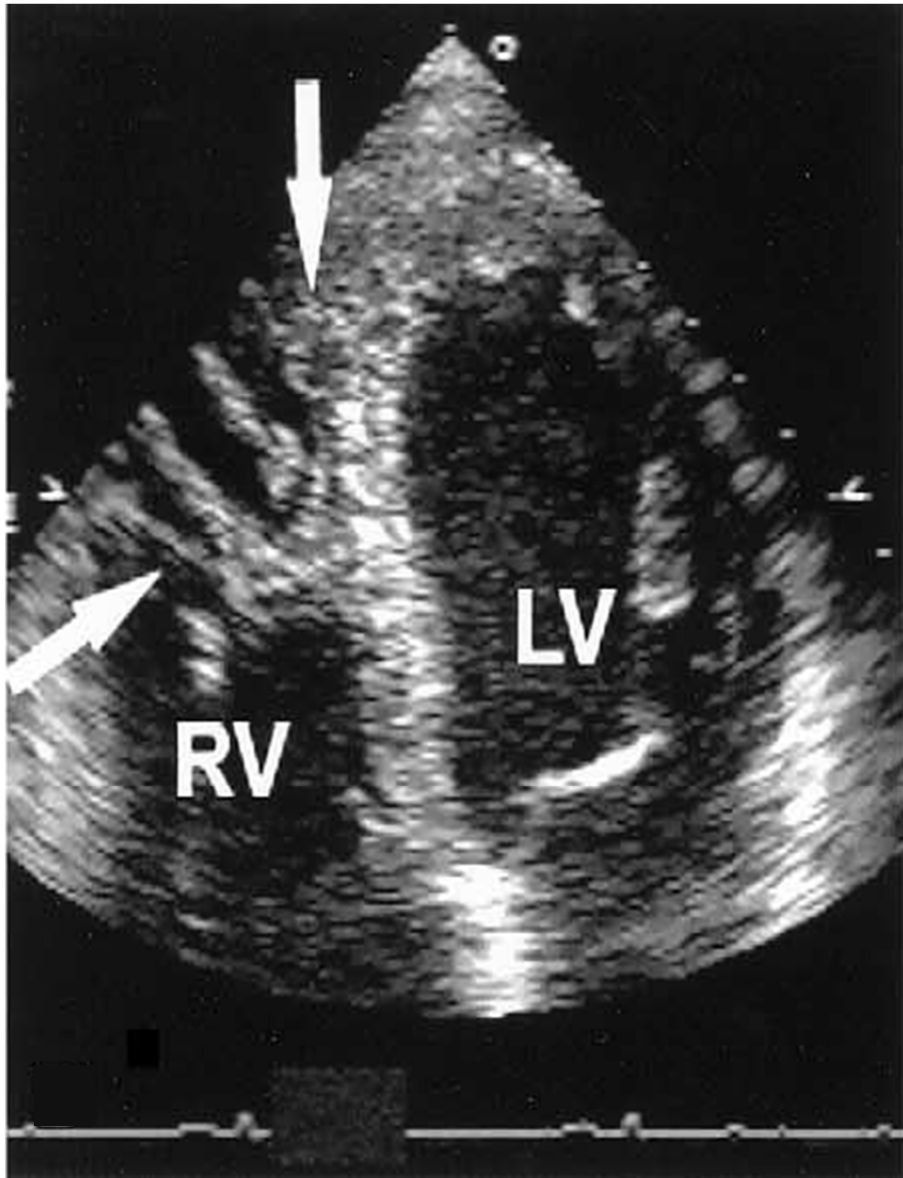
- **Overt or severe end of disease spectrum**
- **Highly specific, but lack of sensitivity for early and familial disease**
- **Qualitative and subjective criteria for ventricular structure and function**

# **Echo Findings from Multidisciplinary Study of ARVC**

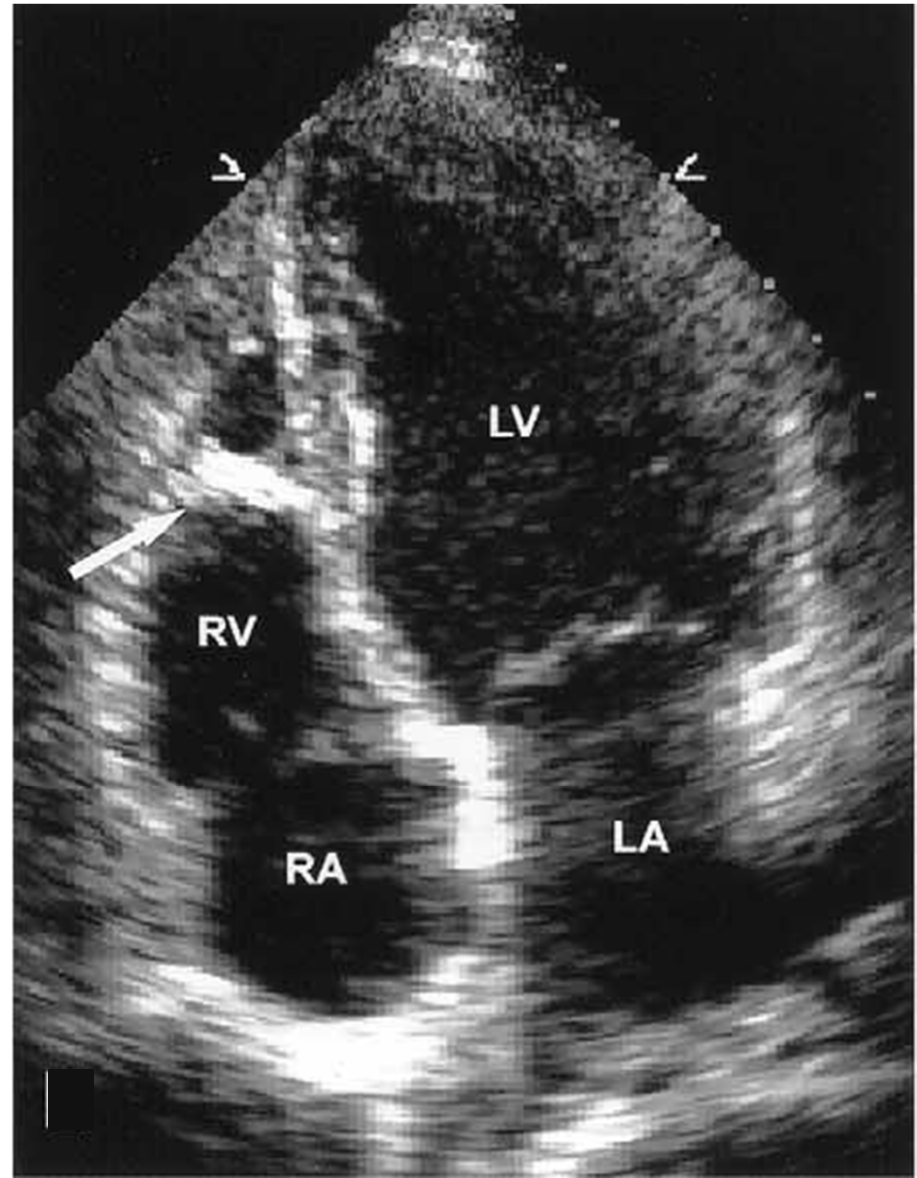
- **To quantify echo abnormalities in newly diagnosed probands with ARVC**
- **29 ARVC probands from 11 centers (1994 Task Force Criteria): compared with 29 normal controls**
- **37.0 ± 14.2 yr-old; 17 males**
- **Dimensions from parasternal and apical views**
- **RV systolic function by FAC**
- **Segmental analysis**
  - **RVOT, anterior, anteroseptal, apical, septal, inferobasal, inferoapical segments**

## Frequency of Qualitative Echo Findings in Probands of ARVC

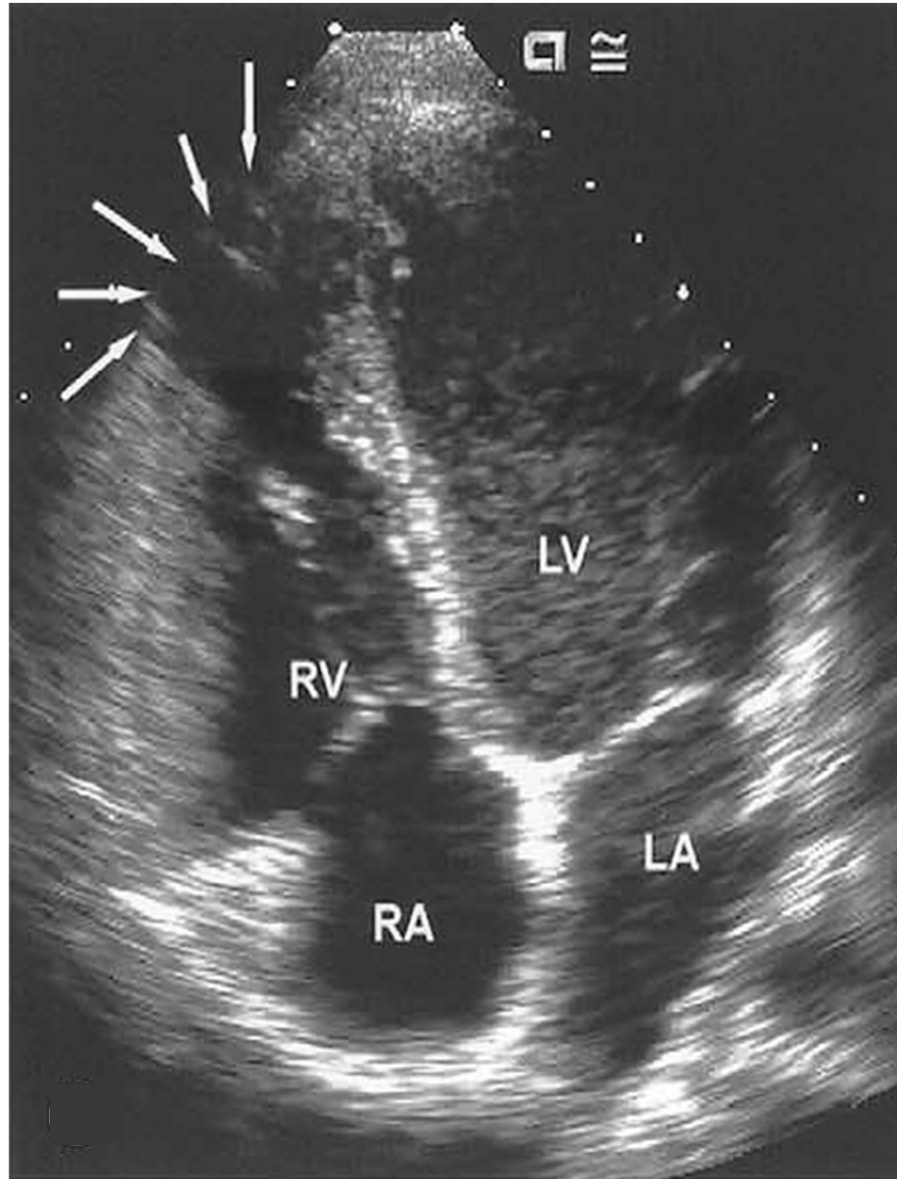
	Number	Percent
RV global function		
Normal	11	38
Mildly reduced	8	28
Severely reduced	10	34
RV regional WMA	23	79
RVOT	13	45
Anteroseptal	16	55
Anterior	20	70
Apex	21	72
Septal	16	55
Inferior basal	17	59
Inferior apical	15	52
Hyperreflective moderator band	9	31
Excessive/abnormal trabeculations	15	54
Sacculations	5	17



**Excessive trabeculation**



**Hyper-reflective moderator band**



**Aneurysm of RV**

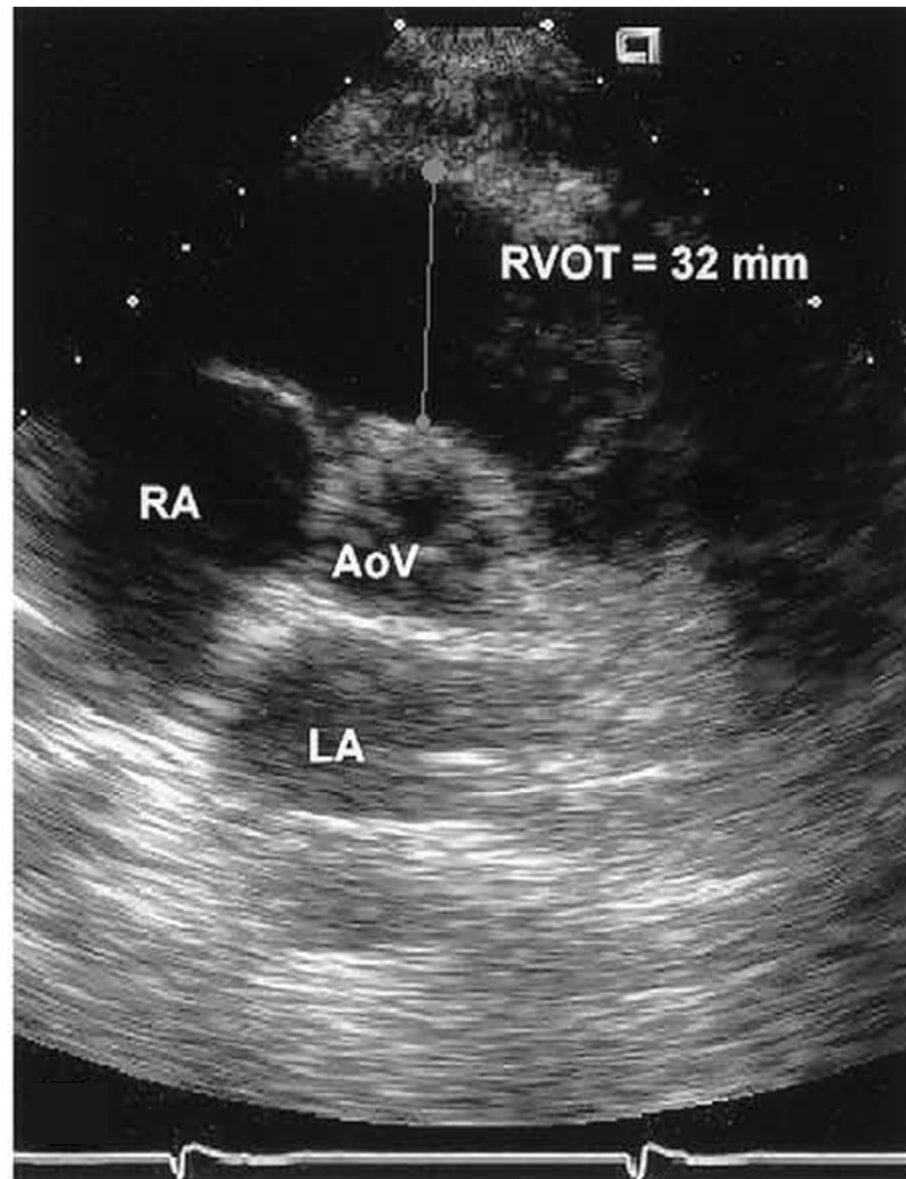
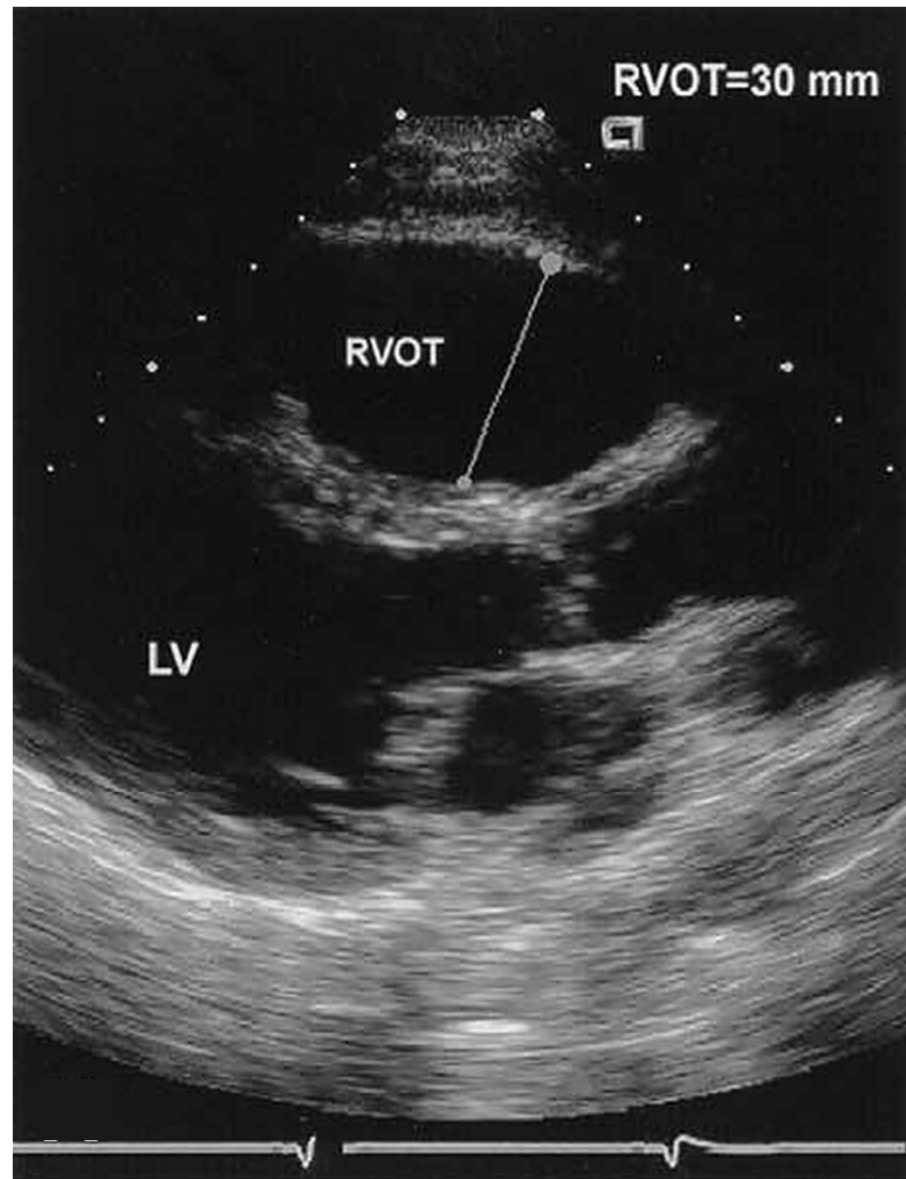
# Quantitative Echo Findings in Probands with ARVC

## Dimension of Right Atrium and Ventricle

Right Heart Dimensions	ARVD Probands (mm ± SD)	Matched Controls (mm ± SD)	p Value
RA medial-lateral (mm)	44.8 ± 11.4	36.6 ± 6.9	0.0035
RA superior-inferior (mm)	51.3 ± 10.6	45.7 ± 5.8	0.023
RVOT-PLAX diastole (mm)	37.9 ± 6.6	26.2 ± 4.9	0.00001
RVOT-PLAX systole (mm)	32.8 ± 7.2	20.1 ± 4.0*	0.00001
RVOT-PSAX diastole (mm)	38.9 ± 4.7	31.1 ± 4.7*	0.00001
RVOT-PSAX systole (mm)	28.3 ± 6.1	19.0 ± 5.1*	0.00001
RVOT/aortic valve	1.28 ± 0.2	1.04 ± 0.2*	0.0001
RVIT PLAX diastole (mm)	57.0 ± 12.2	49.2 ± 8.8	0.0065
RVIT PLAX systole (mm)	46.5 ± 12.6	34.2 ± 6.8*	0.0001
RVIT PSAX diastole (mm)	37.3 ± 8.5	28.1 ± 5.2*	0.0004
RVIT PSAX systole (mm)	32.3 ± 8.5	21.6 ± 5.6*	0.0004
RV medial-lateral-apical 4-chamber diastole (mm)	34.0 ± 8.9	25.1 ± 4.0*	0.00001
RV medial-lateral-apical 4-chamber systole (mm)	27.26 ± 9.8	17.6 ± 3.9*	0.00001
RV LAX length-apical 4-chamber diastole (mm)	79.2 ± 15.6	76.1 ± 7.6	0.2281
RV LAX length-apical 4-chamber systole (mm)	66.7 ± 15.8	61.2 ± 6.0	0.0802

\*p < 0.003.





# Diagnostic Accuracy of RVOT-PLAX Dimension during Diastole

ARVC Probands (mm±SD)	Matched Controls (mm±SD)	Reference Values (mm±SD)	% of probands Enlarged
37.9±6.6	26.2±4.9	22±1.5	100%

↓  
2SD = 25 mm

**ROC curve analysis; when > 30 mm**  
**Sensitivity 89%, Specificity 86%**

# RV Function by Echo in Probands with ARVC

	ARVC Probands (mm±SD)	Matched Controls (mm±SD)
RV end-dia area (cm <sup>2</sup> )	25.2 ± 7.7	17.9 ± 3.5
RV end-sys area (cm <sup>2</sup> )	18.9 ± 8.4	10.5 ± 2.3
RV FAC (%)	27.2 ± 16	41.0 ± 7.1
% with FAC >32%	35%	97%
% with FAC 26-32%	24%	3%
% with FAC <26%	41%	0%

# **Revised Criteria for ARVC (2010)**

## **Purpose and Contents**

- **To incorporate new knowledge and technology improving diagnostic sensitivity**
- **To maintain diagnostic specificity**
- **To include quantitative parameter**

# **Revised Criteria for ARVC (2010)**

## **Global or Regional Dysfunction and Structural Alterations**

- **Echo: 69 probands vs 450 normal**
- **MRI: 44 probands vs 462 normal**

### **Selection of criteria for echo**

**Major Criteria: 95% specificity**

**Minor Criteria: Specificity = Sensitivity**

## Sensitivity and Specificity of Proposed Echo Criteria

Echo Parameters	Value	Sensitivity %	Specificity %
<b>PLAX RVOT (by BSA)</b>	<b><math>\geq 32</math> mm <math>\geq 19</math> mm/m<sup>2</sup></b>	<b>75</b>	<b>95</b>
<b>PSAX RVOT (by BSA)</b>	<b><math>\geq 36</math> mm <math>\geq 21</math> mm/m<sup>2</sup></b>	<b>62</b>	<b>95</b>
<b>FAC</b>	<b><math>\leq 33</math> %</b>	<b>55</b>	<b>95</b>
<b>PLAX RVOT (by BSA)</b>	<b>29 - 31 mm 16 - 18 mm/m<sup>2</sup></b>	<b>87</b>	<b>87</b>
<b>PSAX RVOT (by BSA)</b>	<b>32 - 35 mm 18 - 20 mm/m<sup>2</sup></b>	<b>80</b>	<b>80</b>
<b>FAC</b>	<b><math>\leq 40</math> %</b>	<b>76</b>	<b>76</b>



# **Revised Criteria for ARVC (2010)**

## **Global or Regional Dysfunction and Structural Alterations**

### **Major Criteria**

#### **By 2D echo:**

- **Regional RV akinesia, dyskinesia, or aneurysm**
- **and 1 of the following (end-diastole)**
  - **PLAX RVOT  $\geq 32$  mm ( $\geq 19$  mm/m<sup>2</sup>)**
  - **PSAX RVOT  $\geq 36$  mm ( $\geq 21$  mm/m<sup>2</sup>)**
  - **Fractional Area Change (FAC)  $\leq 33\%$**

# **Revised Criteria for ARVC (2010)**

## **Global or Regional Dysfunction and Structural Alterations**

### **Minor Criteria**

**By 2D echo:**

- **Regional RV akinesia or dyskinesia**
- **and 1 of the following (end-diastole)**
  - **PLAX RVOT: 29 - 31 mm (16 - 18 mm/m<sup>2</sup>)**
  - **PSAX RVOT: 32 - 35 mm (18 - 20 mm/m<sup>2</sup>)**
  - **Fractional Area Change (FAC) : 33 - 40%**

# **Revised Criteria for ARVC (2010)**

## **Global or Regional Dysfunction and Structural Alterations**

### **Major Criteria**

#### **By MRI:**

- **Regional RV akinesia, dyskinesia, or dyssynchronous RV contraction**
- **and 1 of the following (end-diastole)**
  - **$RVEDV/BSA \geq 110 \text{ mL/m}^2(\text{M})$  or  $\geq 100 \text{ mL/m}^2(\text{F})$**
  - **$RV \text{ EF} \leq 40\%$**

#### **By RV angiography:**

- **Regional RV akinesia, dyskinesia, or aneurysm**

# **Revised Criteria for ARVC (2010)**

## **Global or Regional Dysfunction and Structural Alterations**

### **Minor Criteria**

#### **By MRI:**

- **Regional RV akinesia, dyskinesia, or dyssynchronous RV contraction**
- **and 1 of the following (end-diastole)**
  - **RVEDV/BSA: 100-110 mL/m<sup>2</sup>(M) or  $\geq$  90-100 mL/m<sup>2</sup>(F)**
  - **RV EF: 40 - 45%**

# **Arrhythmogenic RV cardiomyopathy/dysplasia clinical presentation and diagnostic evaluation: Results from the North American Multidisciplinary Study**

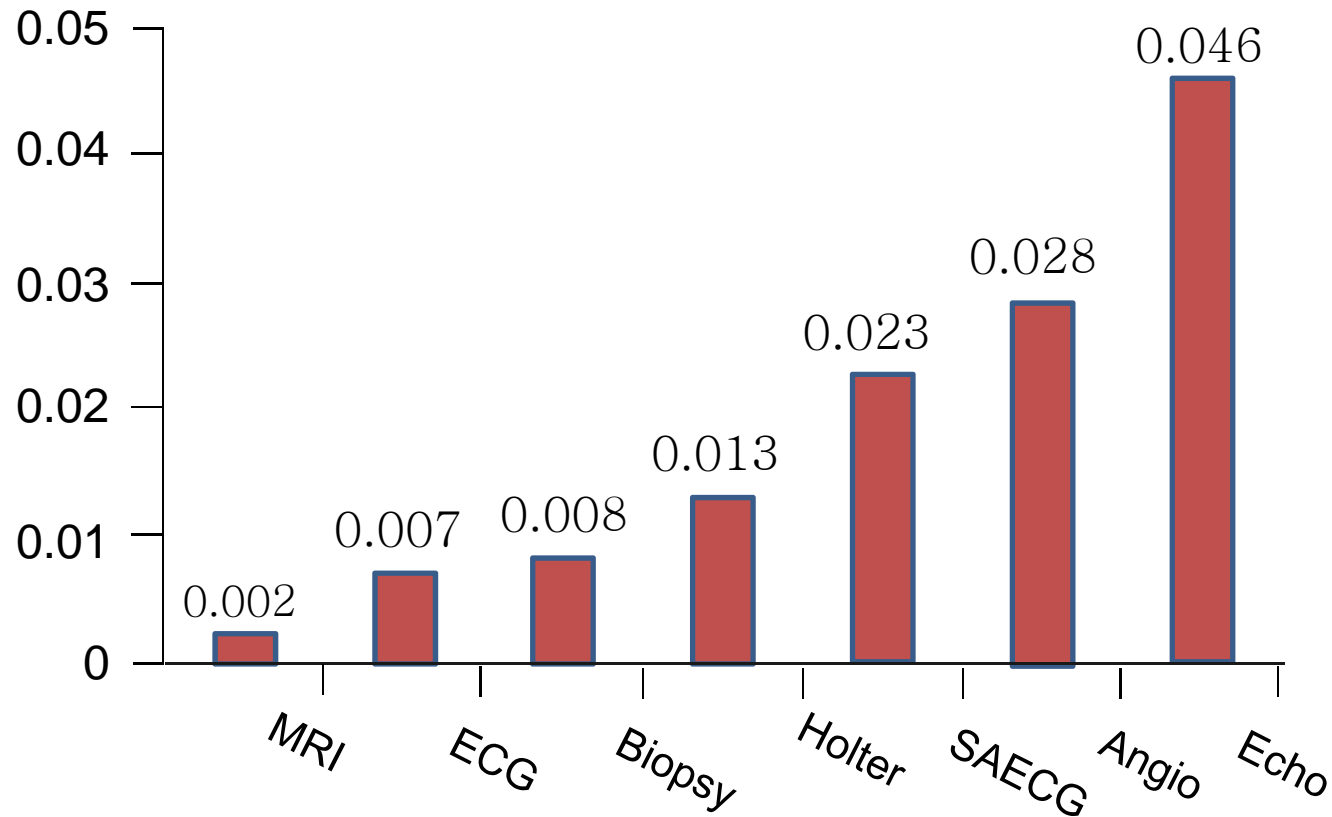
Frank I. Marcus, MD, Wojciech Zareba, MD, Hugh Calkins, MD, Jeffrey A. Towbin, MD, Cristina Basso, MD, David A. Bluemke, MD, PhD, N.A. Mark Estes, III, MD, Michael H. Picard, MD, Danita Sanborn, MD, Gaetano Thiene, MD, Thomas Wichter, MD, David Cannom, MD, David J. Wilber, MD, Melvin Scheinman, MD, Henry Duff, MD, James Daubert, MD, Mario Talajic, MD, Andrew Krahn, MD, Michael Sweeney, MD, Hasan Garan, MD, Scott Sakaguchi, MD, Bruce B. Lerman, MD, Charles Kerr, MD, Jack Kron, MD, Jonathan S. Steinberg, MD, Duane Sherrill, PhD, Kathleen Gear, RN, Mary Brown, MS, Patricia Severski, BS, Slava Polonsky, MS, Scott McNitt, MS

- **Purpose: to assess the clinical characteristics and diagnostic evaluation of ARVC patients**
- **This study proposed an important combination of diagnostic modalities for optimal clinical evaluation of patients suspected ARVC**

# Which Tests do show a better diagnostic performance ?

Analysis of 108 new patients with suspected ARVC:  
Definite 73, Borderline 28, no ARVC 7

C-Statistics Decline





## Comparison of Predictive Models for Correct Classification of ARVC

Variables included in predictive model	Variable removed	Score ( $\chi^2$ )
Echo, Angio, SAECG, Holter, ECG, Biopsy, MRI	none	61.44
Echo, Angio, SAECG, Holter, ECG, Biopsy	MRI	61.24
Echo, Angio, SAECG, Holter, ECG	Biopsy, MRI	59.34*
Echo, Angio, SAECG, Holter,	ECG, Biopsy, MRI	57.11*
Echo, Angio, SAECG	Holter, ECG, Biopsy, MRI	52.26*

# Conclusions

- **Modification of the original Task Force criteria represent a working framework to improve diagnosis of ARVC**
- **Echocardiography is the most important diagnostic modality to assess ARVC**
- **Understanding the molecular and genetic background and the development of newer imaging techniques will improve the diagnosis of ARVC in the future.**