

/ Best of ESC 2022 /

Cardiac Imaging & Valvular Heart Disease

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No disclosures

Section 4. Recommendations on indications for surgery in severe aortic regurgitation				
Revised	Surgery is indicated in asymptomatic patients with resting ejection fraction $\leq 50\%$.	I	Surgery is recommended in asymptomatic patients with LVESD >50 mm or LVESD >25 mm/ m^2 BSA (in patients with small body size) or resting LVEF $\leq 50\%$.	I
	Surgery should be considered in asymptomatic patients with resting ejection fraction $>50\%$ with severe LV dilatation: LVEDD >70 mm or LVESD >50 mm (or LVESD >25 mm/ m^2 BSA in patients with small body size).	IIa		
New			Surgery may be considered in asymptomatic patients with LVESD >20 mm/ m^2 BSA (especially in patients with small body size) or resting LVEF $\leq 55\%$, if surgery at low-risk.	IIb
Revised	Heart Team discussion is recommended in selected patients in whom aortic valve repair may be a feasible alternative to valve replacement.	I	Aortic valve repair may be considered in selected patients at experienced centres when durable results are expected.	IIb

Surgery should be considered in **asymptomatic** patients with preserved LV function (LVESD <40 mm and LVEF >60%) and AF secondary to mitral regurgitation or pulmonary hypertension^c (SPAP at rest >50 mmHg). ^{285,289}

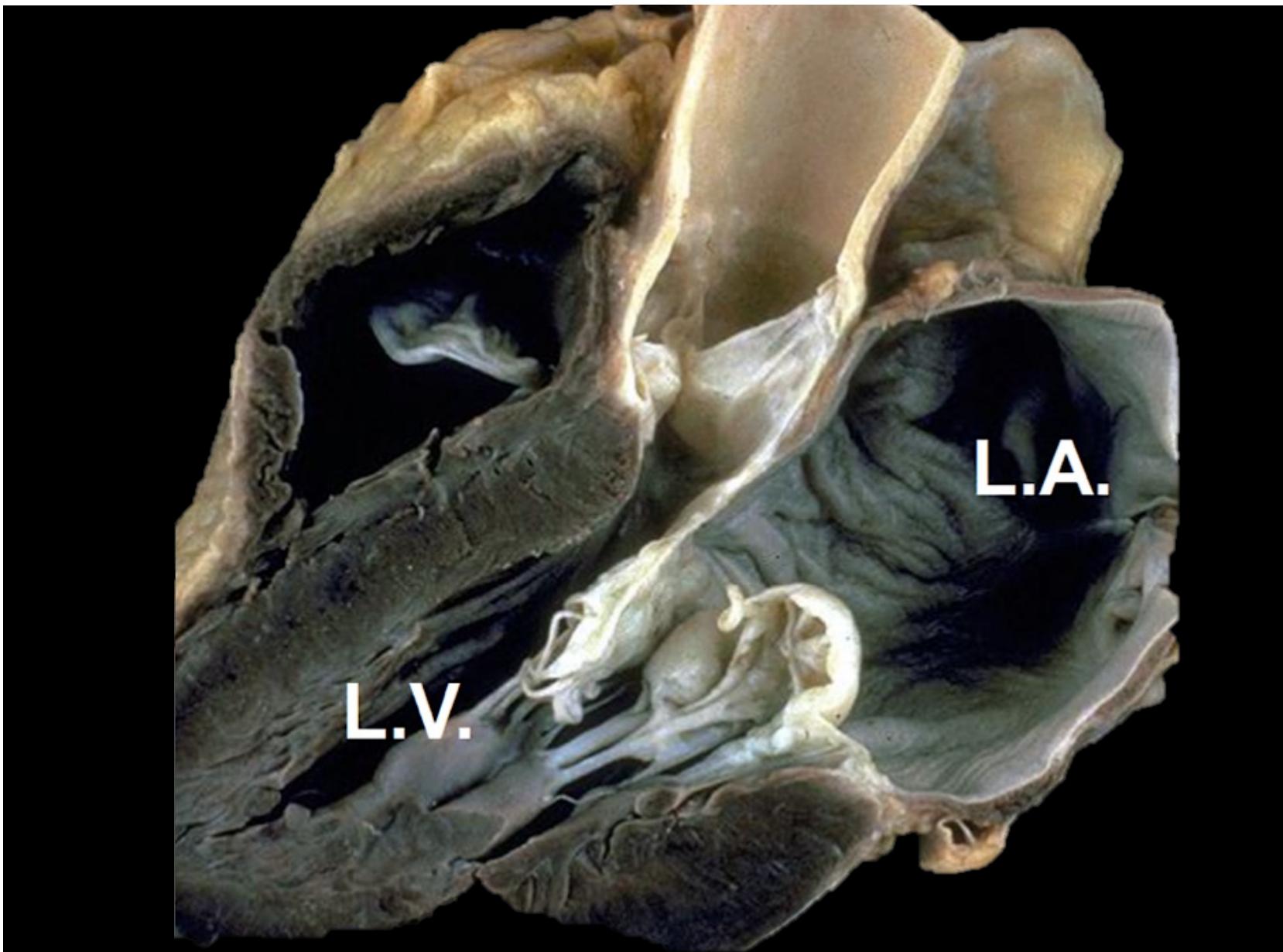
IIa**B**

Surgical mitral valve repair should be considered in low-risk **asymptomatic patients** with LVEF >60%, LVESD <40 mm^d and significant LA dilatation (volume index ≥60 mL/m² or diameter ≥55 mm) when performed in a Heart Valve Centre and a durable repair is likely. ^{285,288}

IIa**B**

EHRA expert consensus statement on arrhythmic mitral valve prolapse and mitral annular disjunction complex in collaboration with the ESC Council on valvular heart disease and the European Association of Cardiovascular Imaging endorsed by the Heart Rhythm Society, by the Asia Pacific Heart Rhythm Society, and by the Latin American Heart Rhythm Society

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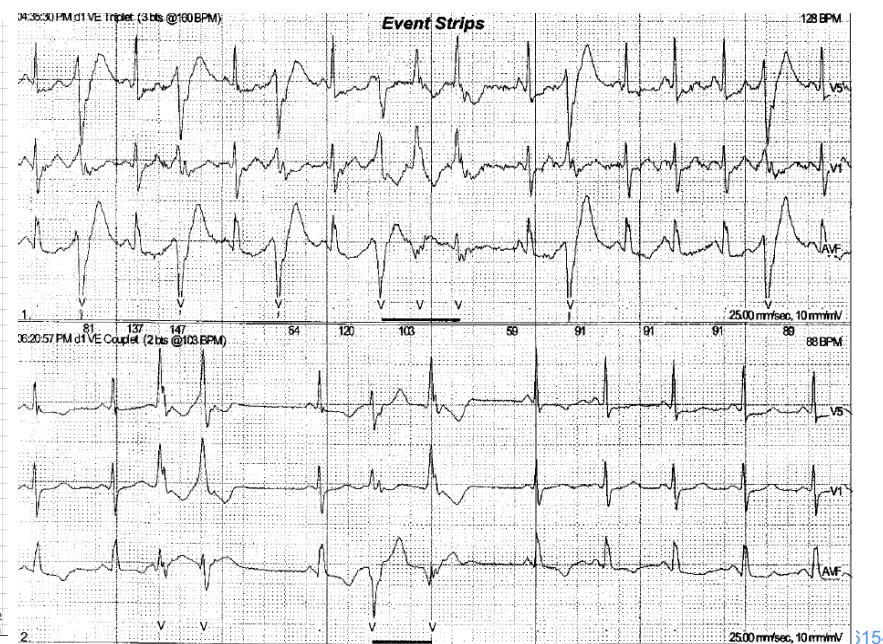
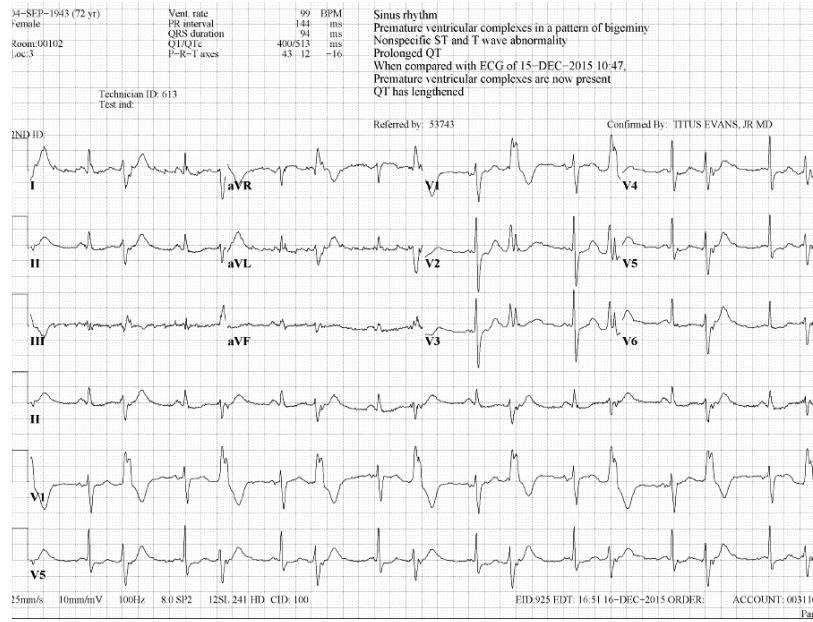
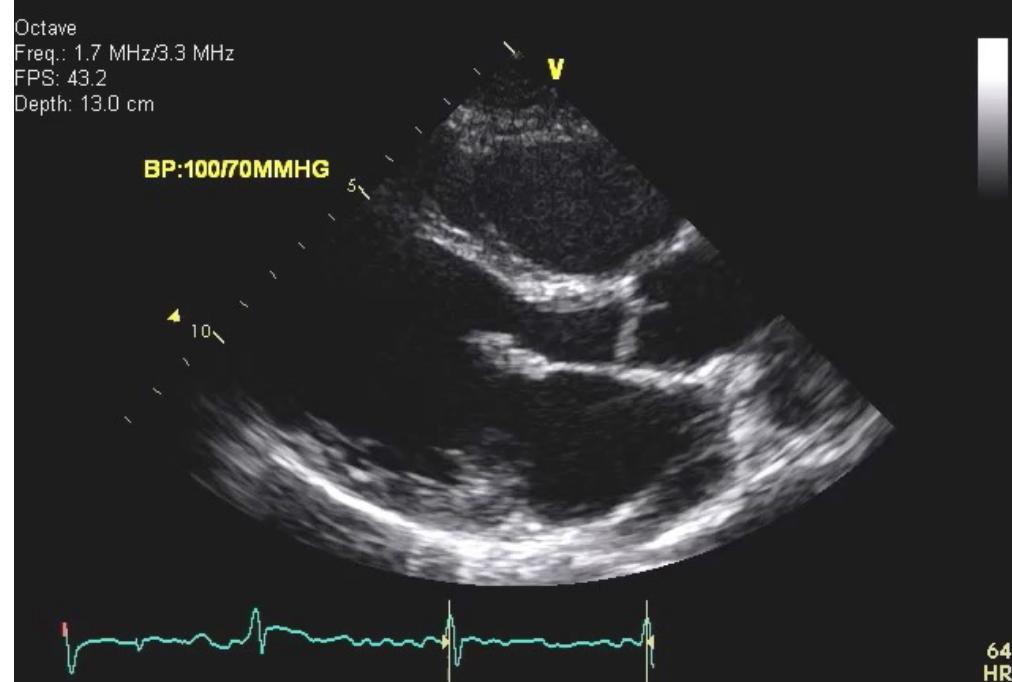
/ 65yo woman

/ non smoker

/ shortness of breath & palpitations

/ No murmur

/ Myxomatous MV



/ What is the arrhythmic mitral valve prolapse ? /

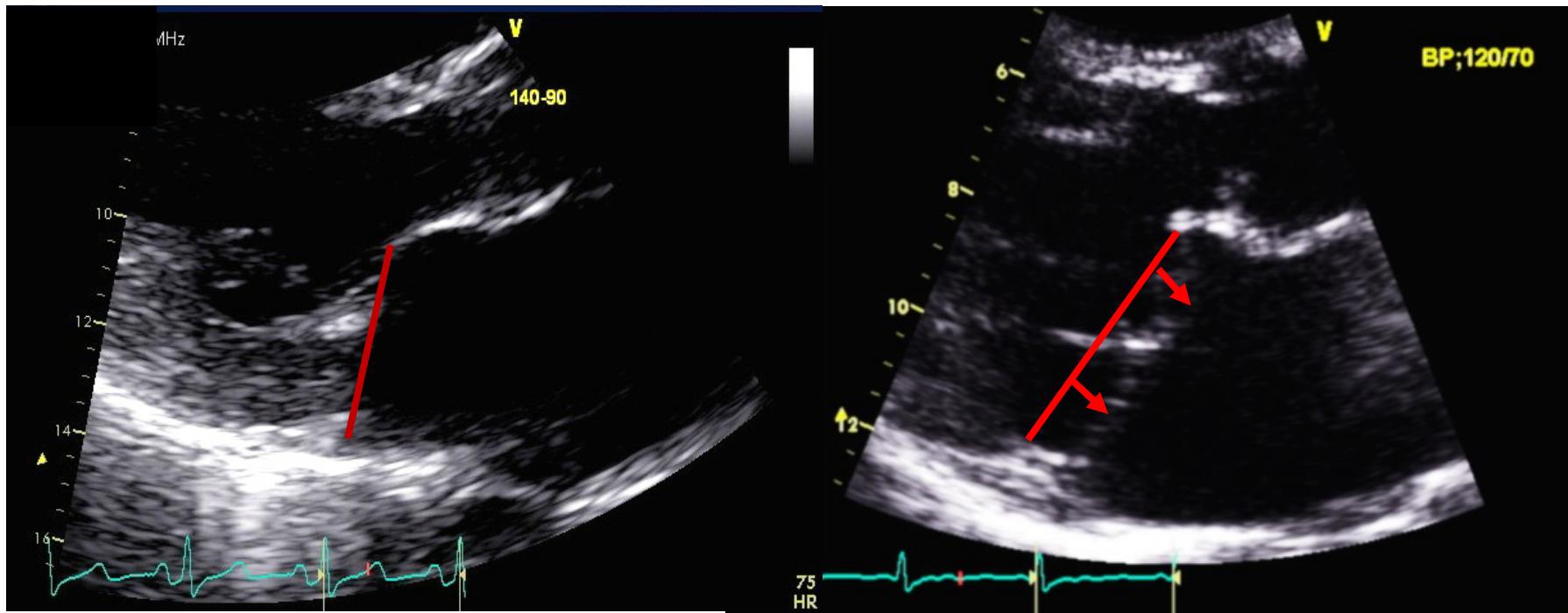
Mitral Valve Prolapse

What is the prevalence of
MVP ?

8 Million US citizens

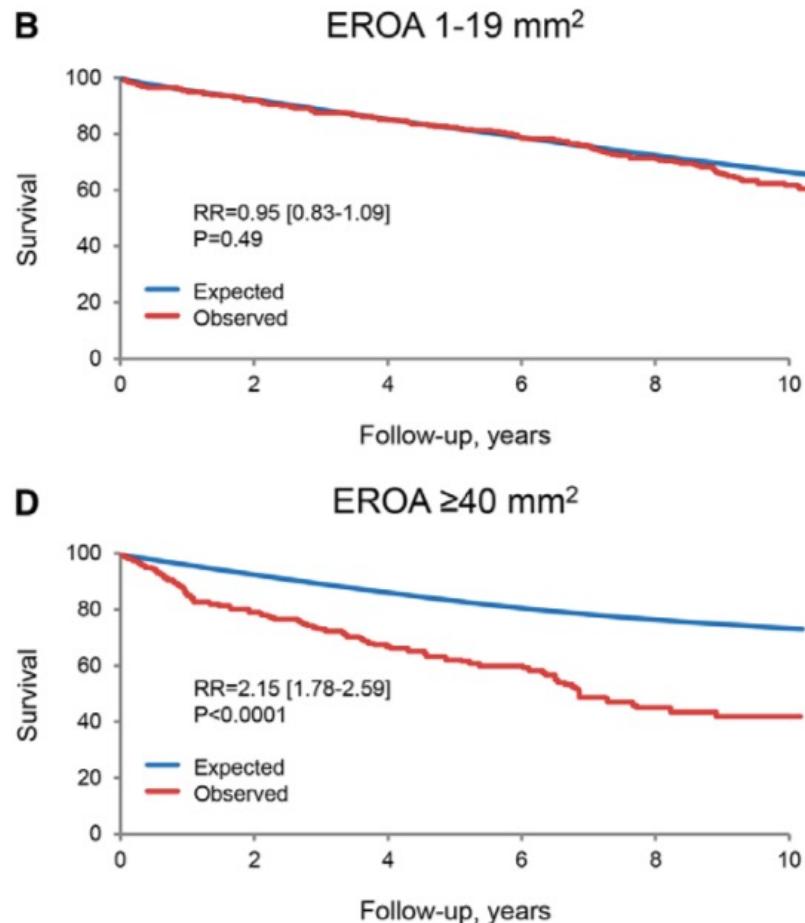
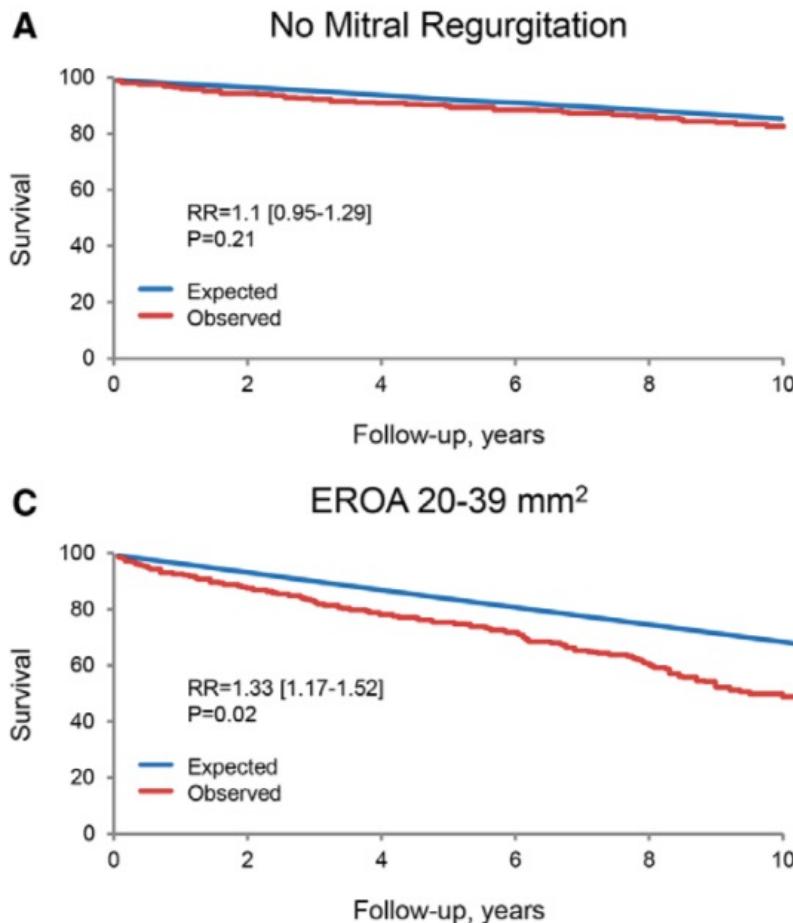
19 Million EU citizens

Mitral Valve Prolapse



Clinical Outcome of Degenerative Mitral Regurgitation

Critical Importance of Echocardiographic Quantitative Assessment in Routine Practice



Valve Disease

Sudden Death in Mitral Regurgitation Due to Flail Leaflet

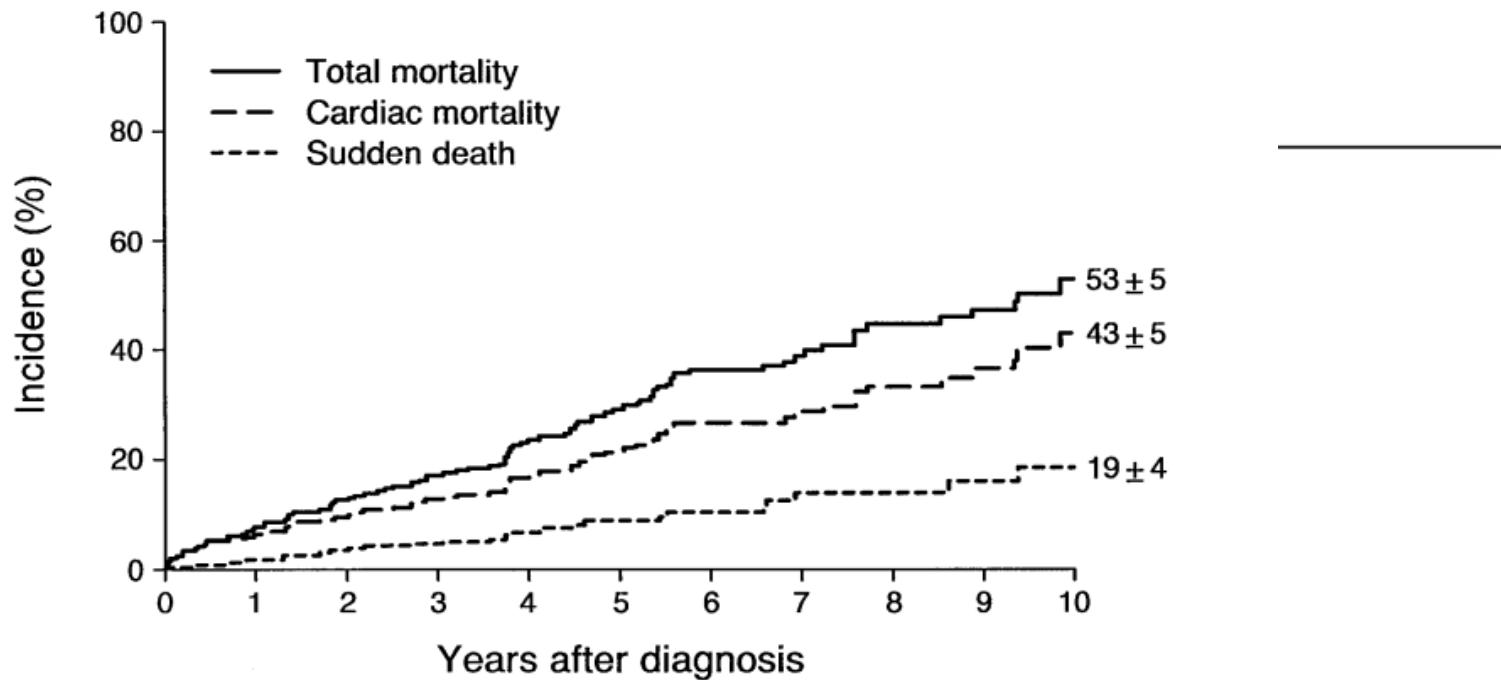
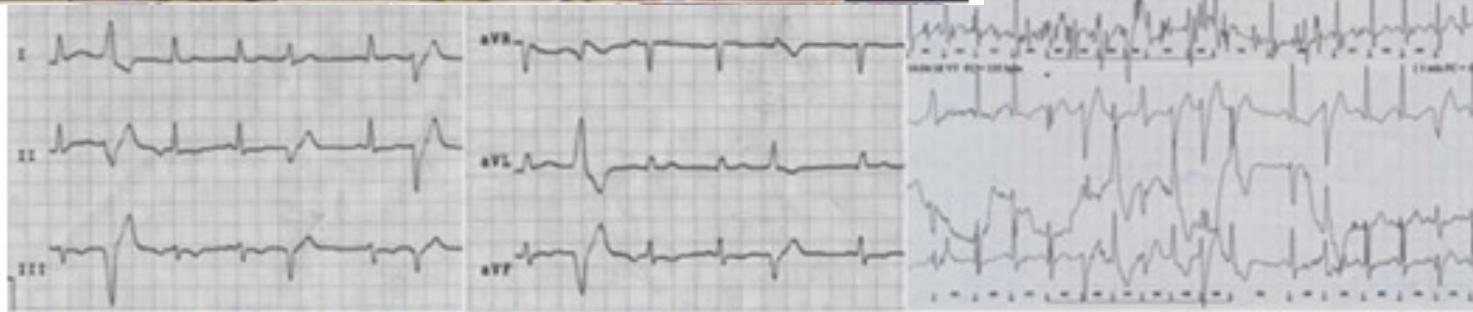


Figure 1. Incidence of total mortality, cardiac mortality and sudden death in patients with MR-FL. The event rates \pm SEE at 10 years are indicated.

Arrhythmic Mitral Valve Prolapse and Sudden Cardiac Death



hD*; Stefania Rizzo, MD, PhD;
Alberto Cipriani, MD;
erico Migliore, MD, PhD;
a Cacciavillani, MD, PhD;
no Thiene, MD; Sabino Iliceto, MD



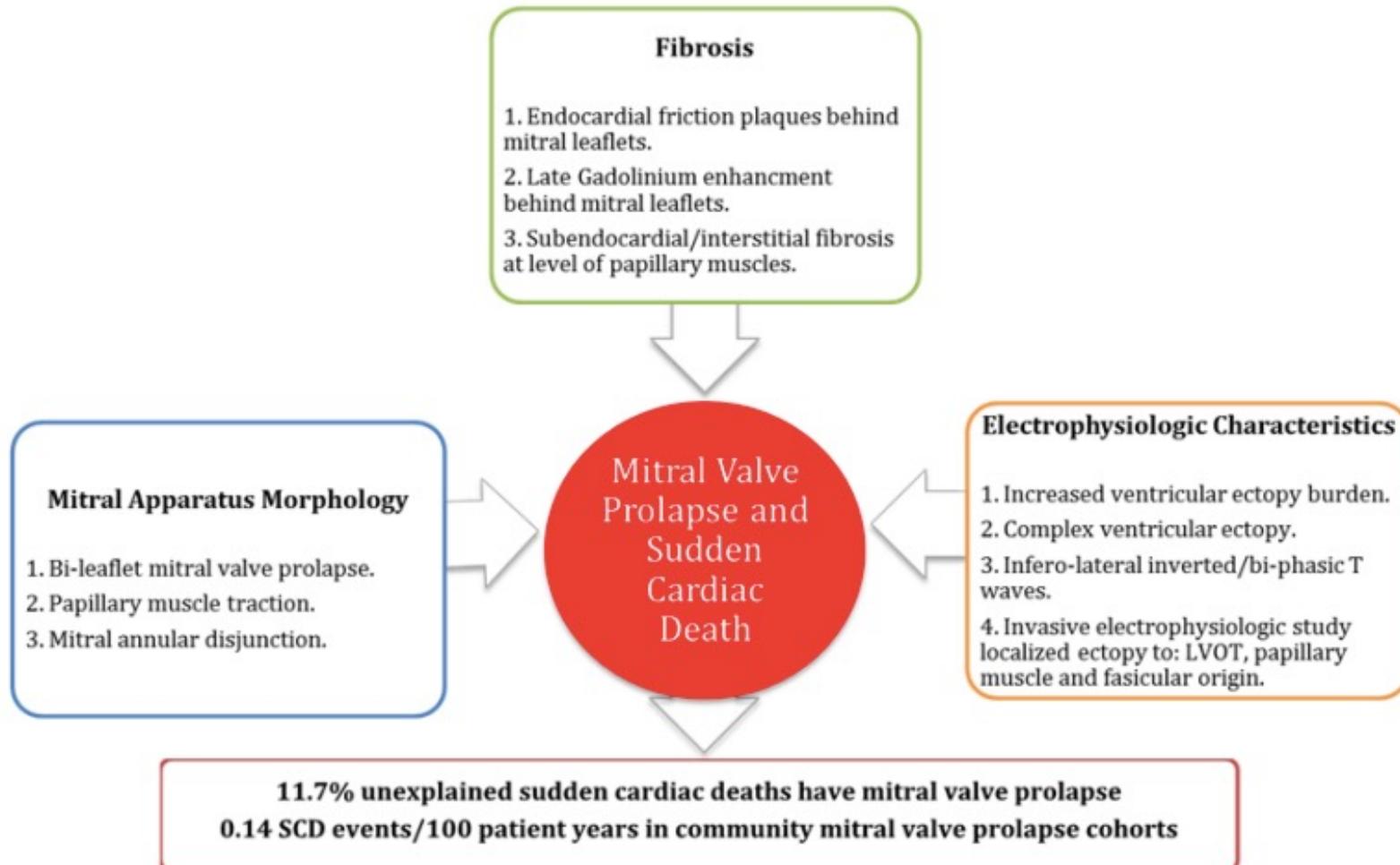
RESEARCH LETTER

Common Phenotype in Patients With Mitral Valve Prolapse Who Experienced Sudden Cardiac Death

Jérôme Hourdain, MD*, Marie Annick Clavel, DVM, PhD*, Jean-Claude Deharo, MD, Samuel Asirvatham, MD, Jean François Avierinos, MD, Gilbert Habib, MD, Frederic Franceschi, MD, PhD, Vincent Probst, MD, PhD, Nicolas Sadoul, MD, Raphael Martins, MD, PhD, Christophe Leclercq, MD, PhD, Michel Chauvin, MD, Jean Luc Pasquie, MD, PhD, Philippe Maury, MD, Gabriel Laurent, MD, PhD, Michael Ackerman, MD, PhD, David O. Hodge, MS, and Maurice Enriquez-Sarano, MD

Mitral valve prolapse (MVP) is a prevalent valvular condition with heterogeneous outcomes.^{1,2} Excess mortality is associated with moderate-to-severe mitral regurgitation (MR) and reduced left ventricular ejection fraction.² Whereas MVP is considered benign in patients without such risk factors,^{1,2} some reports have described ventricular arrhythmias and sudden death (SD) in apparently uncomplicated MVP.^{3,4} Moreover, MVP was identified in 42% of patients who experienced idiopathic out-of-hospital cardiac arrest.³

Mitral valve prolapse and sudden cardiac death: a systematic review and meta-analysis



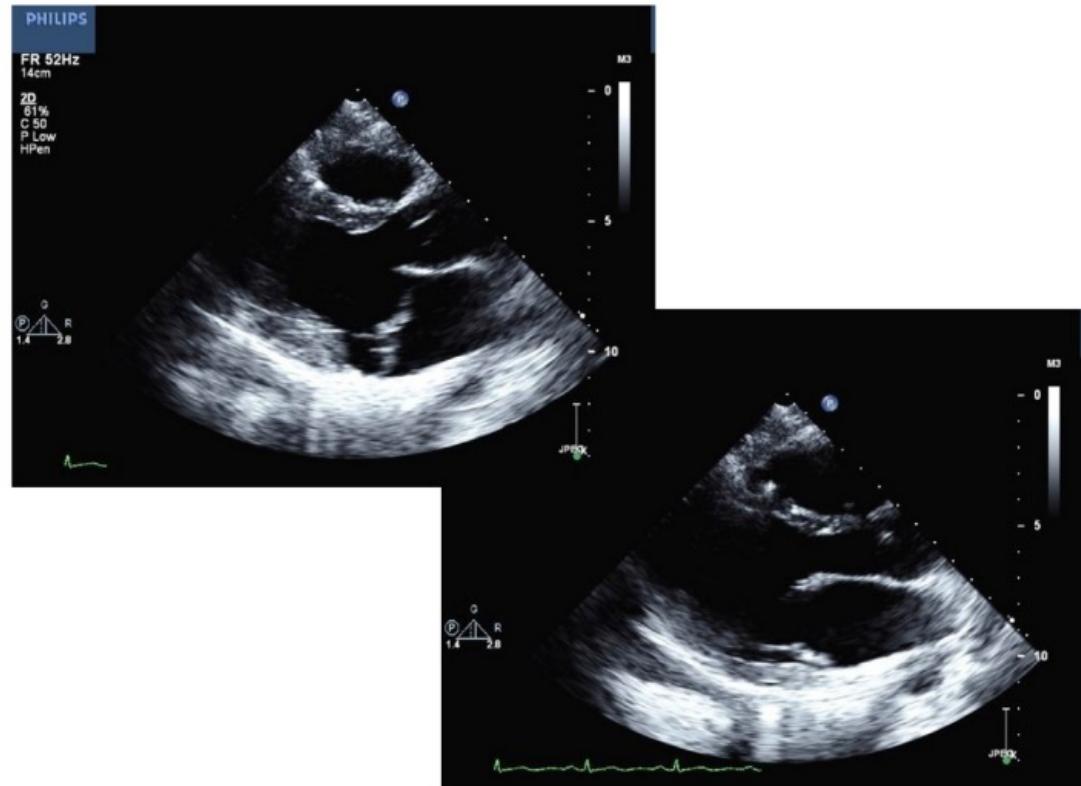
Arrhythmic MVP phenotype

What are the characteristics of
the AMVP phenotype ?

Bi-leaflet MVP..?

Malignant Bileaflet Mitral Valve Prolapse Syndrome in Patients With Otherwise Idiopathic Out-of-Hospital Cardiac Arrest

Chenni S. Sriram, MBBS,* Faisal F.
Jonathan N. Johnson, MD,* Maurice
Bryan C. Cannon, MD,* Samuel J. A
Rochester, Minnesota



...but very frequent !

Prognostic Implications of Left Atrial Enlargement in Degenerative Mitral Regurgitation

Benjamin Essayagh, MD,^a Clémence Antoine, MD,^a Giovanni Benfari, MD,^a David Messika-Zeitoun, MD,^a Hector Michelena, MD,^a Thierry Le Tourneau, MD,^c Sunil Mankad, MD,^a Christophe M. Tribouilloy, MD,^a Prabin Thapa, BSc,^a Maurice Enriquez-Sarano, MD,^a

TABLE 1 Baseline Characteristics

	Overall Population (N = 5,769)	LAVI <40 ml/m ² (n = 3,154)	LAVI 40 to <60 ml/m ² (n = 1,606)	LAVI ≥60 ml/m ² (n = 1,008)
Clinical characteristics	--	--	--	--
Mitral characteristics	--	--	--	--
No/trivial MR	20	33	8	1
Mild MR	30	40	23	9
Moderate MR	22	19	30	19
Severe MR	28	8	39	71
ERO, mm ²	19 (0-40)	0 (0-18)	28 (16-43)	44 (28-55)
RVol, ml	34 (0-65)	0 (0-32)	47 (28-72)	72 (49-95)
Flail leaflet	12	3	16	34
Bileaflet	39	39	39	38
Posterior	44	39	47	52

Risk, Determinants, and Outcome Implications of Progression of Mitral Regurgitation After Diagnosis of Mitral Valve Prolapse in a Single Community

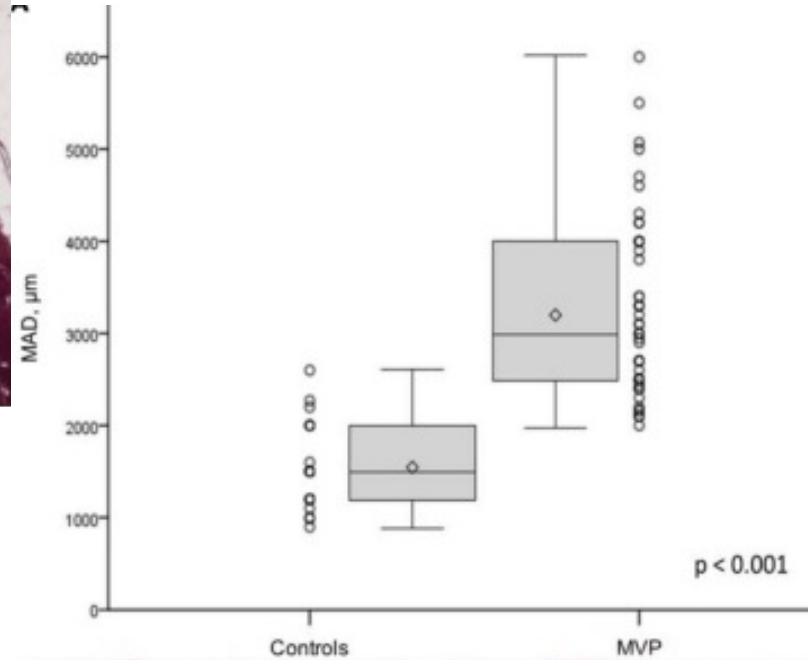
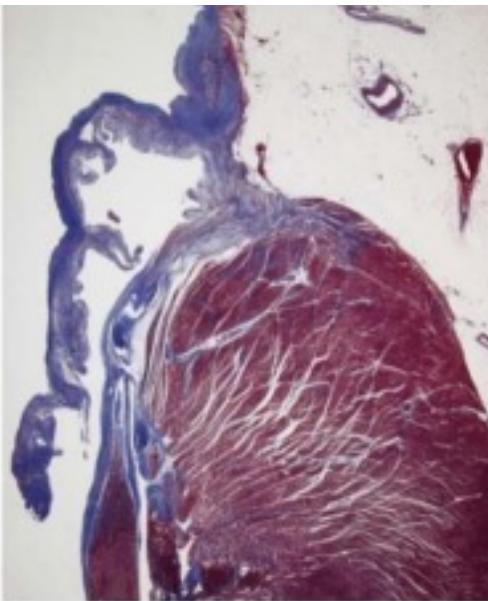
Jean-Francois Avierinos, MD,^{a,b,*} Delphine Detaint, MD,^a David Messika-Zeitoun, MD,^a Dania Mohty, MD,^a and Maurice Enriquez-Sarano, MD^a

Table 1
Baseline clinical and echocardiographic characteristics of 285 residents of Olmsted County, Minnesota, diagnosed with mitral valve prolapse and their association with the progression of mitral regurgitation during follow-up on univariate analysis

Variable	Value	HR (95% CI)	p Value
Age (yrs)	56 ± 22	1.02 (1.01-1.03)	<0.01
Women	162 (57%)	0.8 (0.5-1.09)	0.1
Body surface area (m ²)	1.8 ± 0.3	3.5 (1.6-7.8)	0.01
Previous ischemic neurologic event	23 (8%)	1.7 (0.9-3.5)	0.1
NYHA class III or IV or congestive heart failure	23 (8%)	5.3 (3.0-9.4)	<0.01
Systolic blood pressure at initial echocardiography (mm Hg)	135 ± 23	1.0 (0.99-1.01)	0.8
Diastolic blood pressure at initial echocardiography (mm Hg)	76 ± 10	0.98 (0.96-1.00)	0.08
Atrial fibrillation	35 (12%)	1.8 (1.06-3.2)	0.03
Co-morbidity index	1.0 ± 1.8	1.2 (1.1-1.3)	0.01
Echocardiographic variables			
Left ventricular ejection fraction (%)	60 ± 9	0.96 (0.94-0.99)	0.01
Indexed end-systolic diameter (mm/m ²)	18 ± 4	1.03 (0.98-1.09)	0.3
Indexed end-diastolic diameter (mm/m ²)	29 ± 5	0.98 (0.93-1.03)	0.5
Indexed left atrial diameter (mm/m ²)	23 ± 5	1.05 (1.02-1.09)	0.01
Posterior leaflet prolapse	56 (24%)	1.3 (0.8-2.1)	0.3
Anterior leaflet prolapse	88 (38%)	1.0 (0.6-1.8)	0.9
Bileaflet prolapse	90 (38%)	Reference	
Mitral valve thickening	141 (49%)	0.7 (0.5-1.0)	0.05
Flail leaflets	3 (1%)	3.7 (1.2-11.9)	0.03
Degree of MR at diagnosis			
None	207 (72%)	1.5 (1.2-1.8)	<0.01
Mild	35 (12%)		
Moderate	43 (15%)		

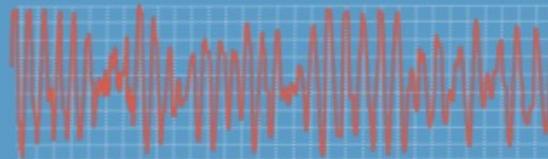
MAD..?

Morphofunctional Abnormalities of Mitral Annulus and Arrhythmic Mitral Valve Prolapse



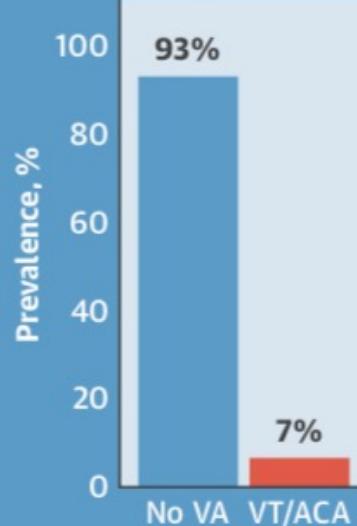
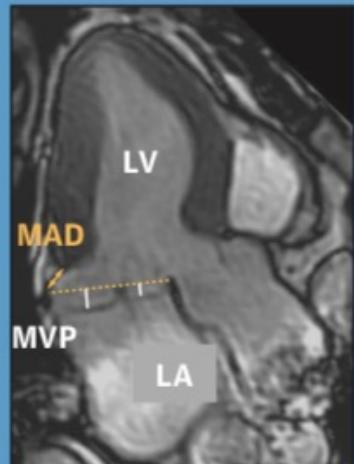
...but

116 Patients with Mitral Annulus Disjunction (MAD)

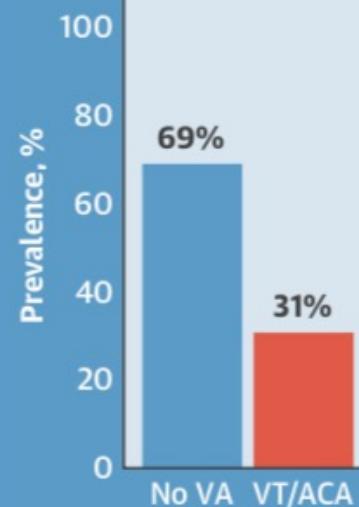
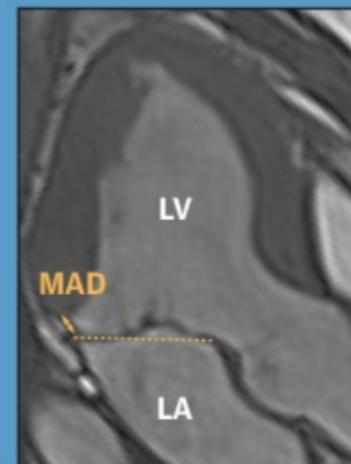


14 with aborted cardiac arrest or sustained ventricular tachycardia

90 MAD with Mitral Valve Prolapse



26 MAD without Mitral Valve Prolapse



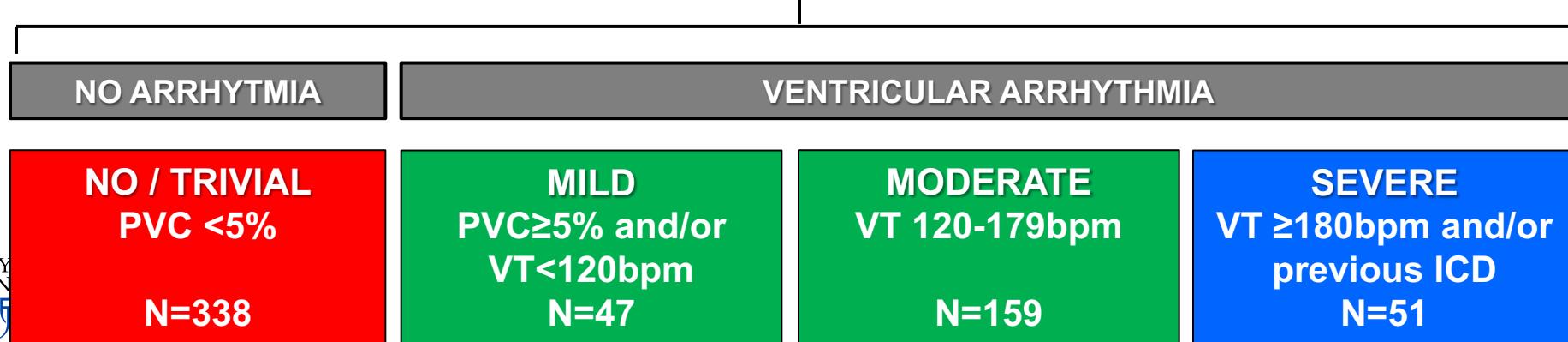
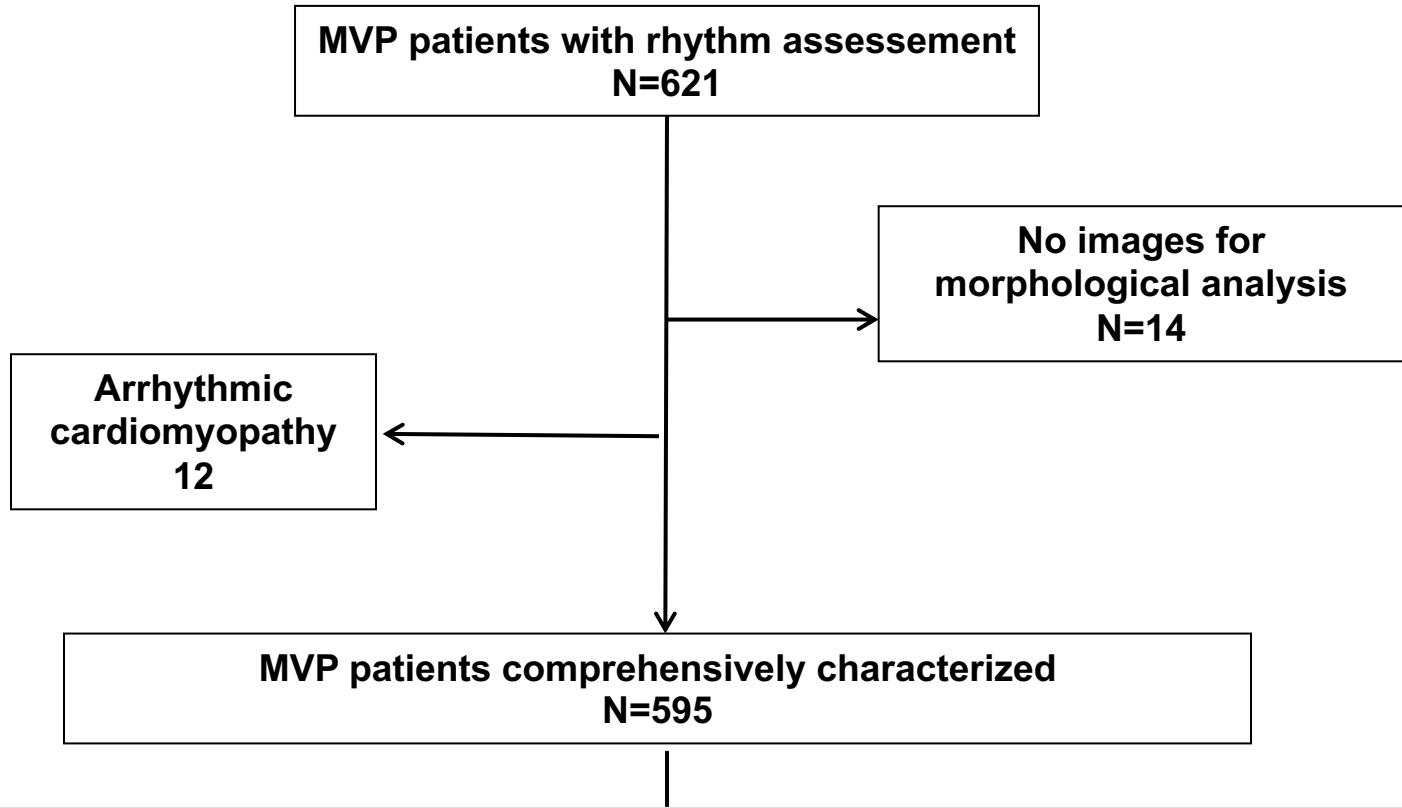
/ Why is the AMVP characteristics & outcome so uncertain ? /

AMVP Phenotype Uncertainty

Reasons

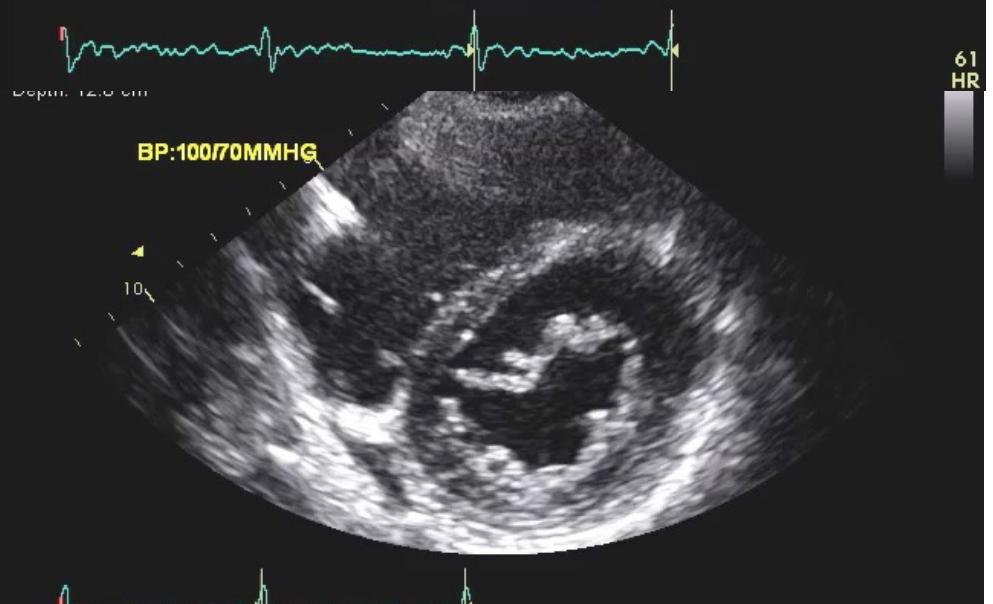
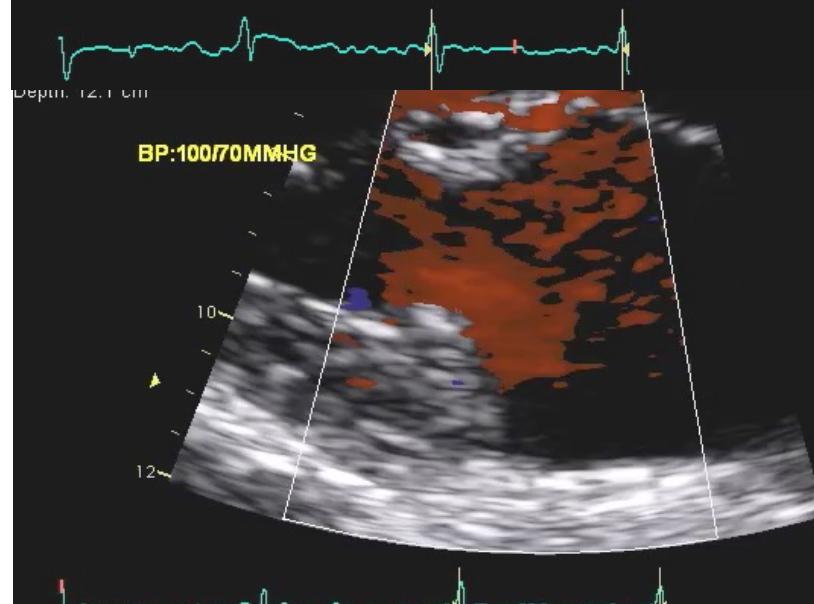
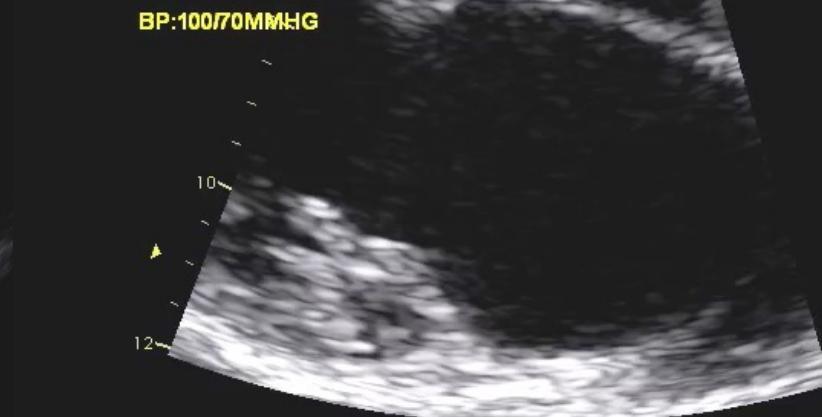
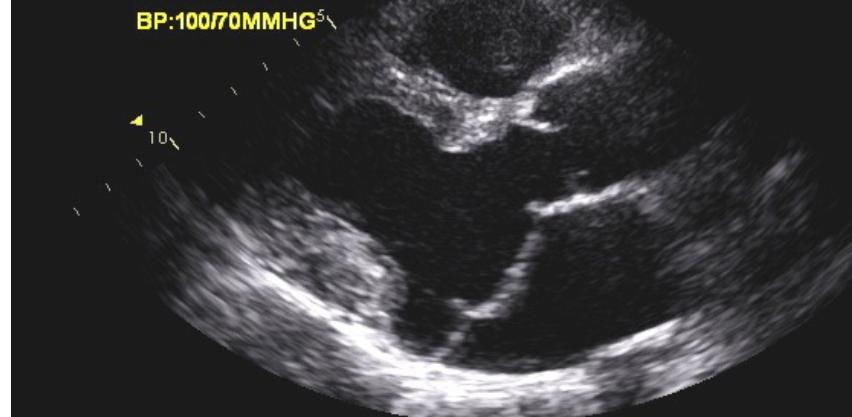
- 1-Small size studies**
- 2-Case-report**
- 3-No comprehensive characterization**
- 4-No follow-up**

The AMVP Phenotype

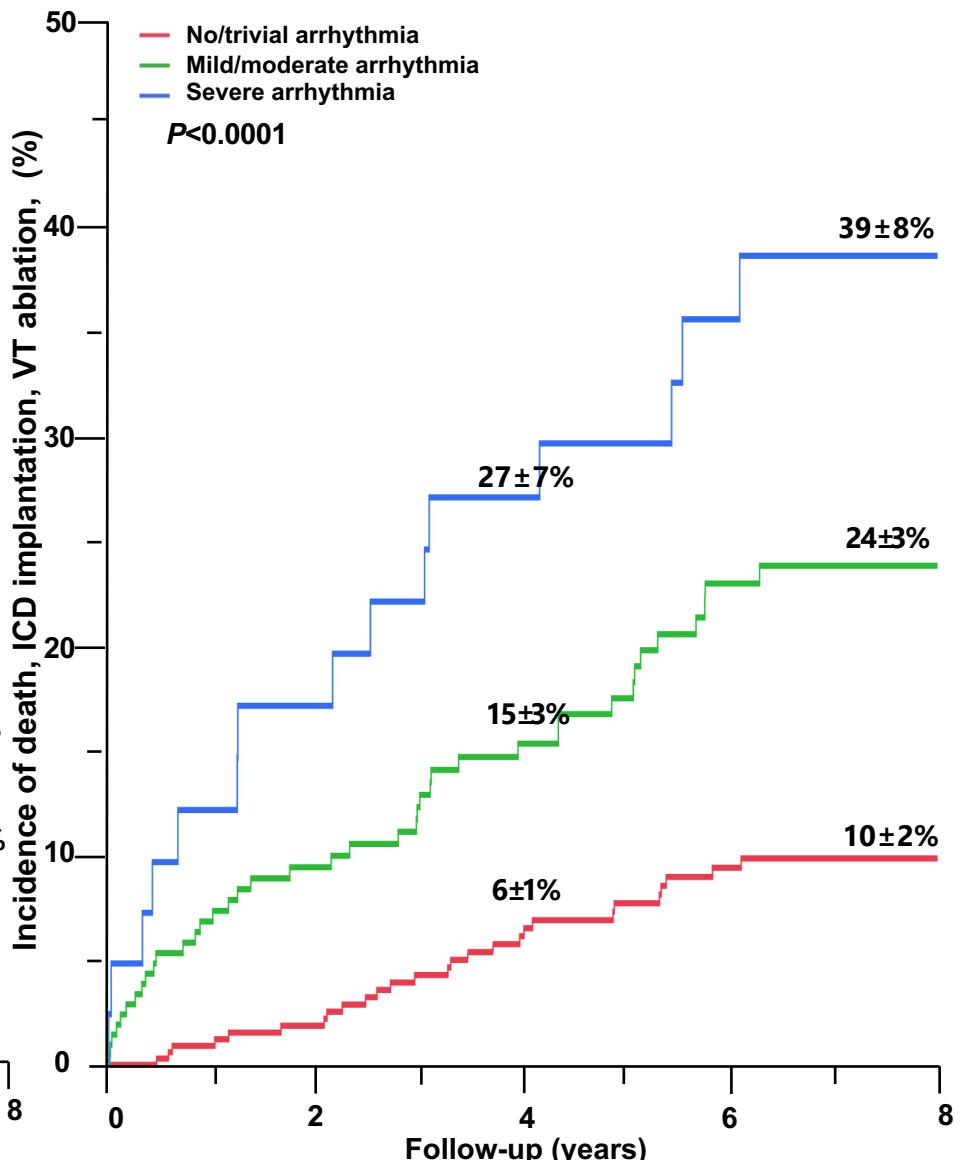
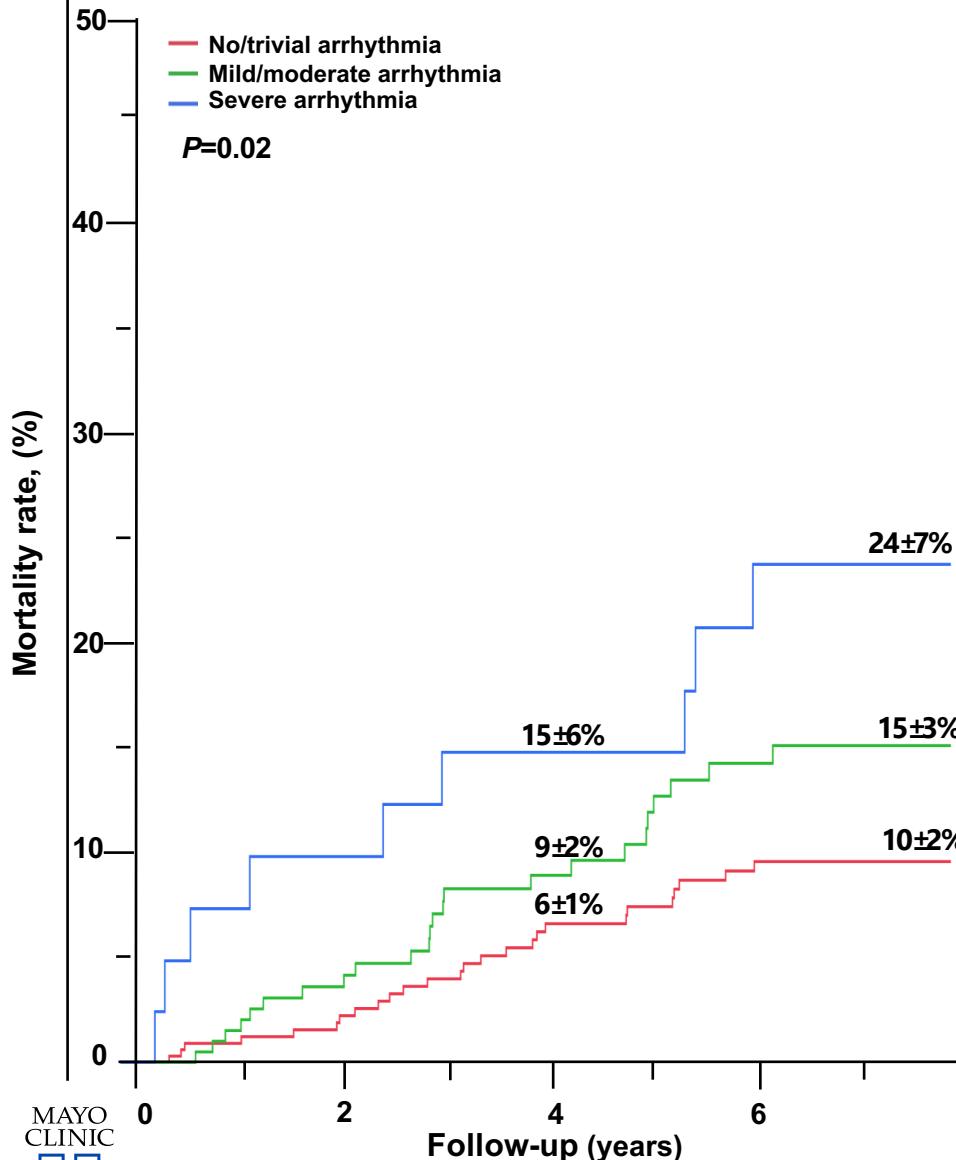


Octave
Freq: 1.7 MHz-3.3 MHz
FPS: 43.2
Depth: 13.0 cm

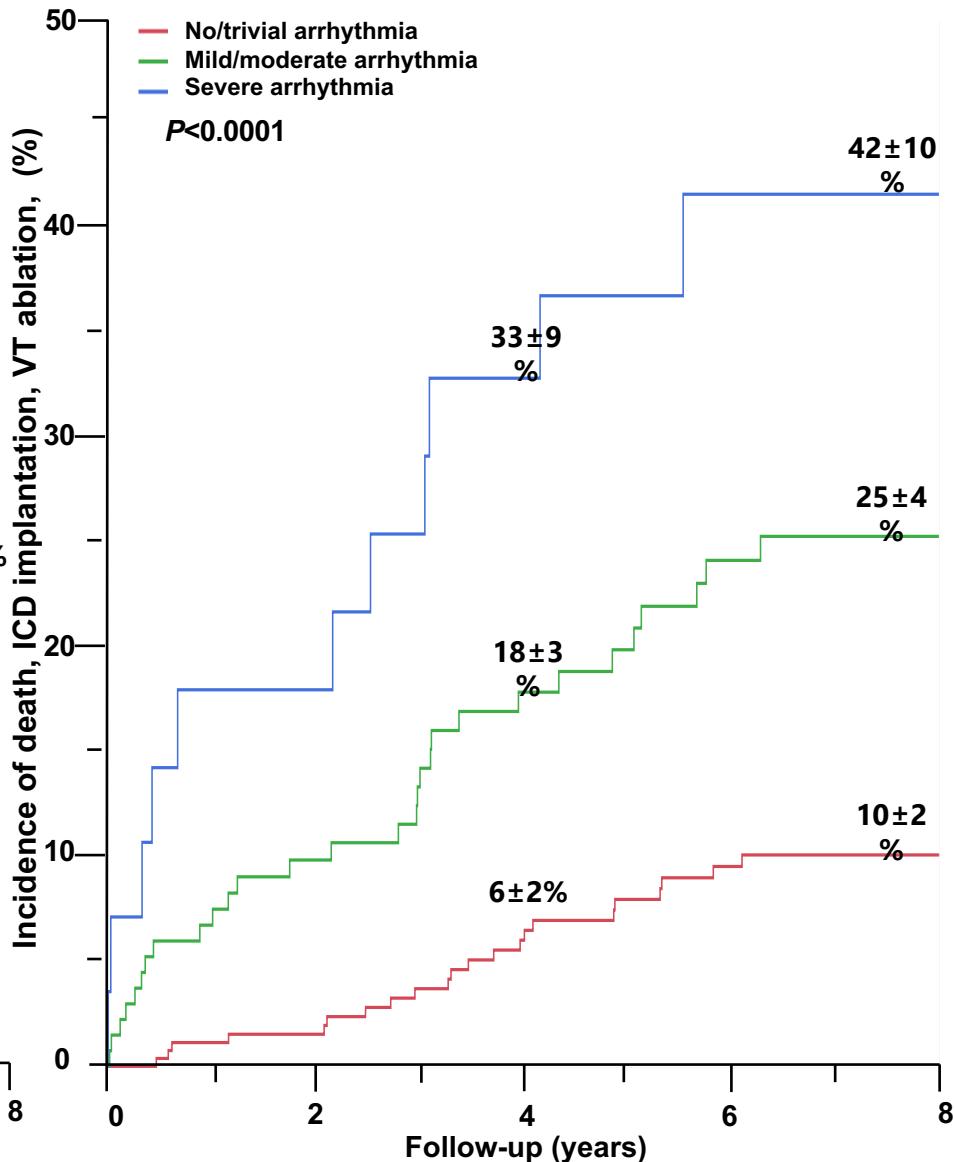
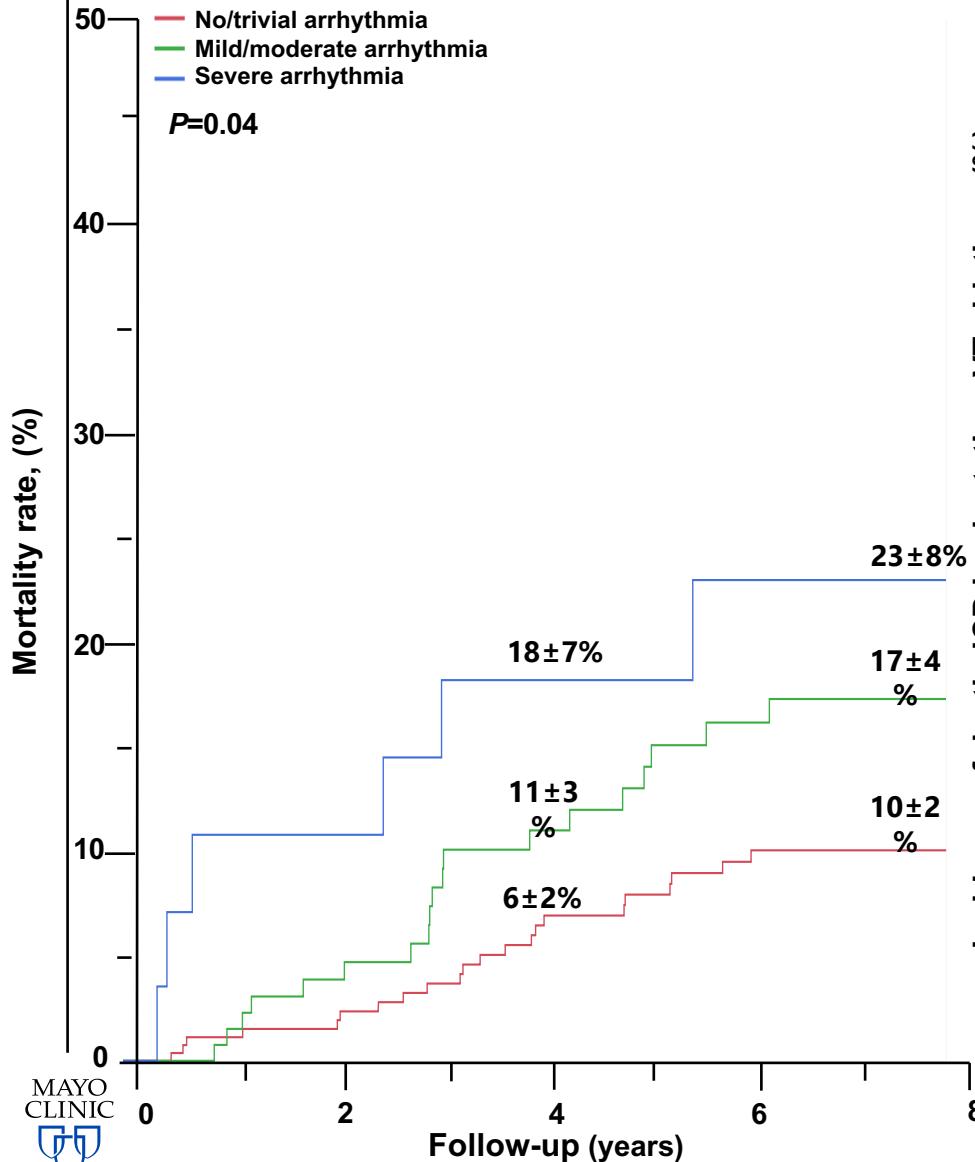
Octave
Freq: 1.7 MHz-3.3 MHz
PS: 100 /
Depth: 11.1 cm



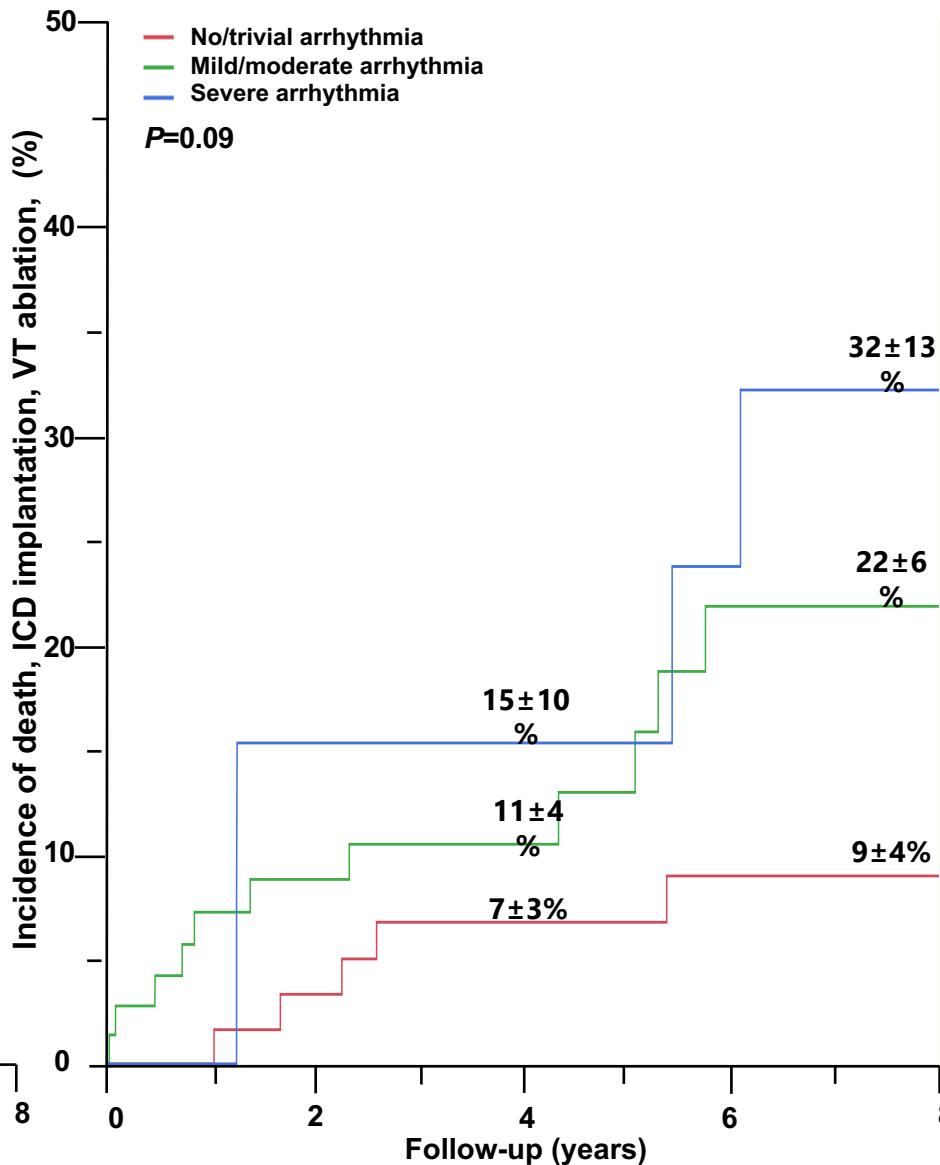
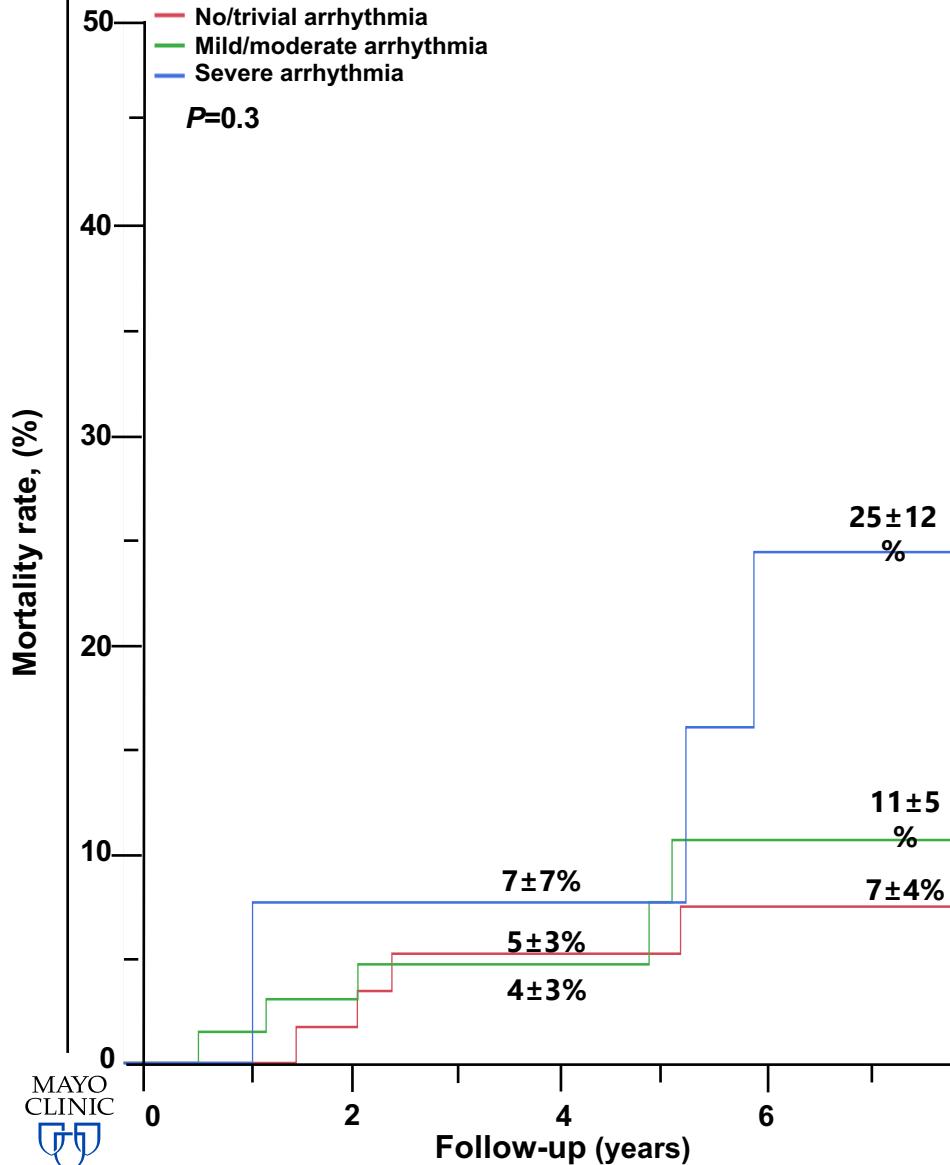
AMVP Outcome



Under medical management



Post MV surgery



THE ARRHYTHMIC MITRAL VALVE PROLAPSE

PHENOTYPE

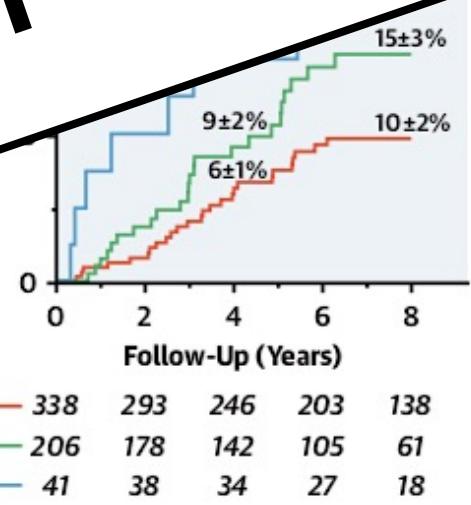
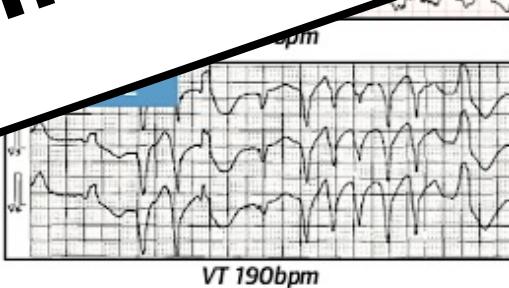
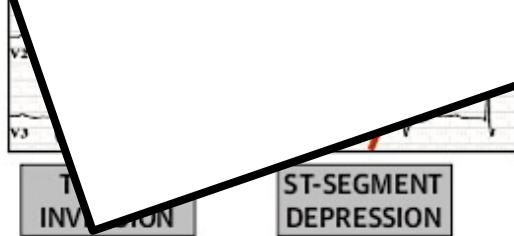


ARRHYTHMIA SEVERITY

NO/TRIVIAL

ECG

Message #1
Look for AMVP
in any MVP



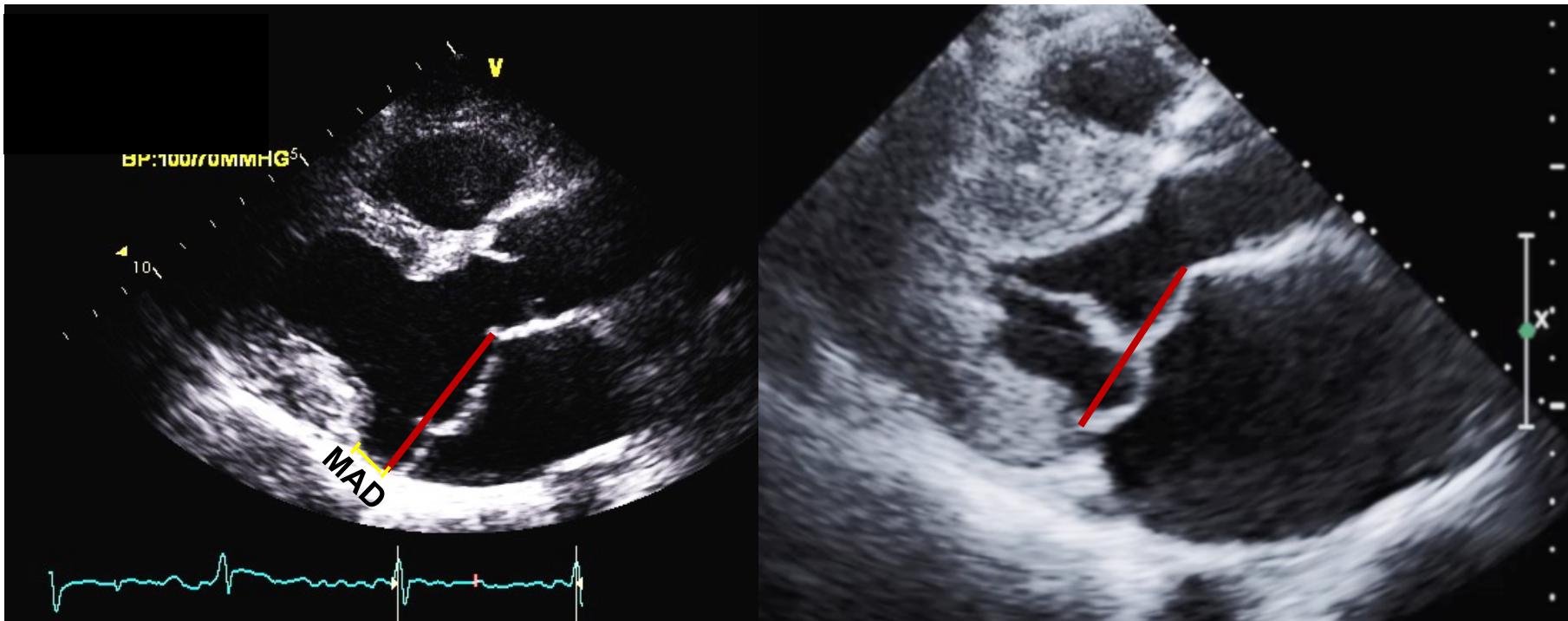
- No/Trivial Arrhythmia
- Mild/Moderate Arrhythmia
- Severe Arrhythmia

What about MAD ?

MAD = SCD ?

But first what is MAD and
how to diagnose MAD ?

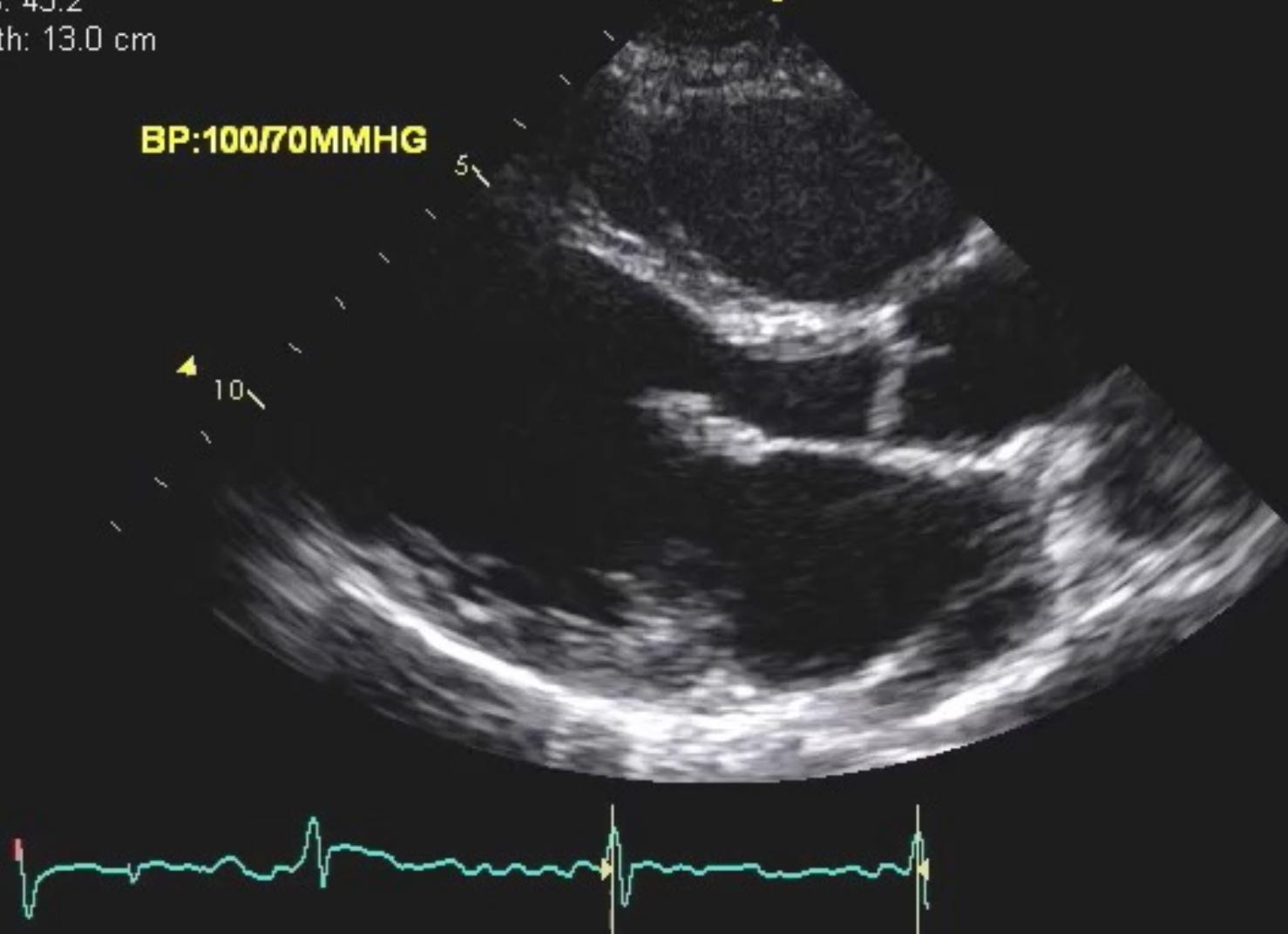
MAD DIAGNOSIS

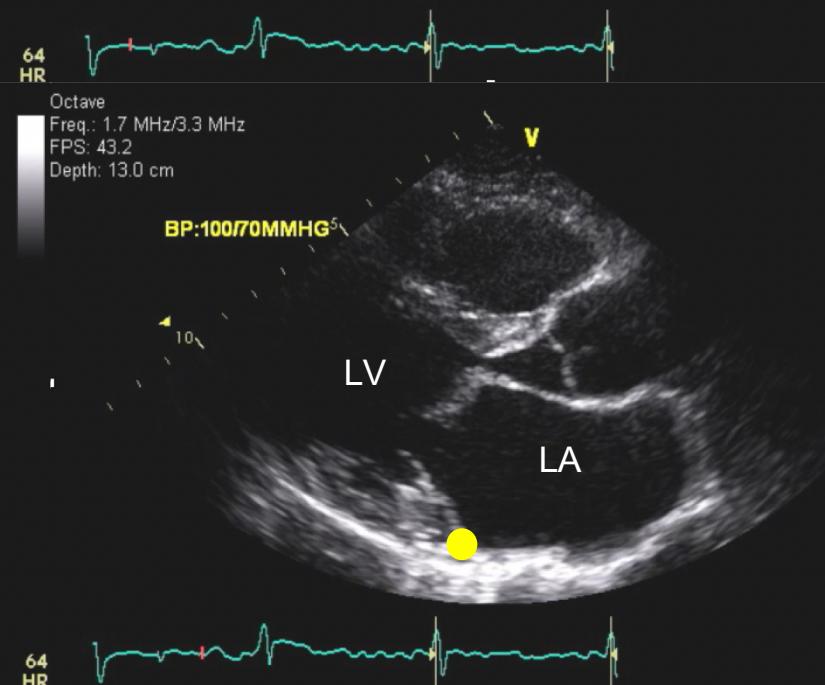
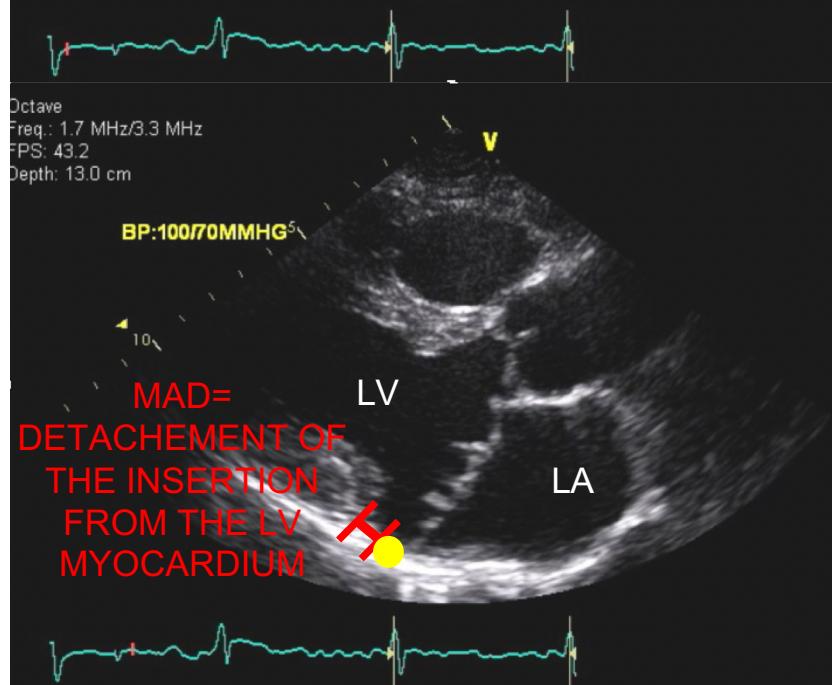
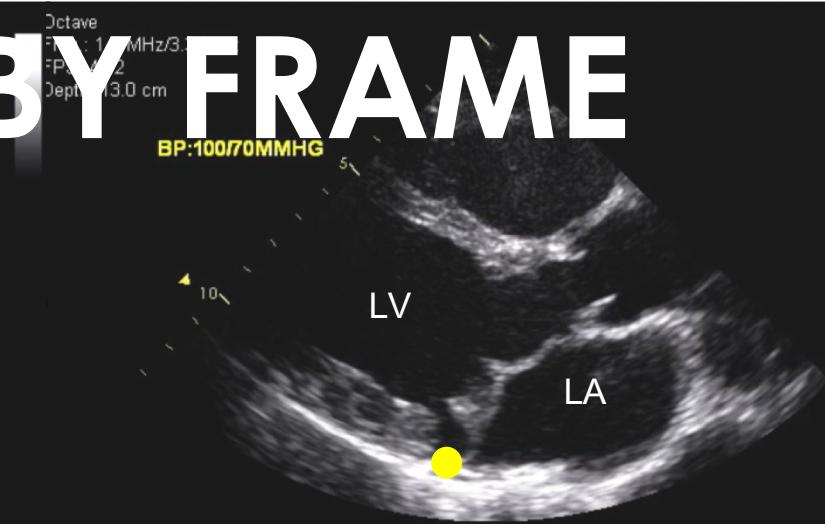
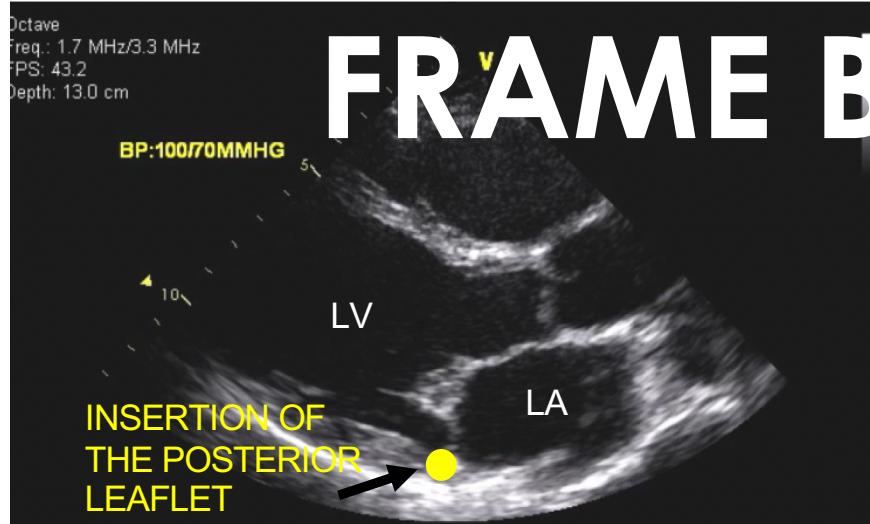


Octave
Freq.: 1.7 MHz/3.3 MHz
FPS: 43.2
Depth: 13.0 cm

MAD DIAGNOSIS

BP:100/70MMHG





freq: 1.5 MHz / 20.0 mm/12

FPS: 69.0/
Depth: 12.1 cm

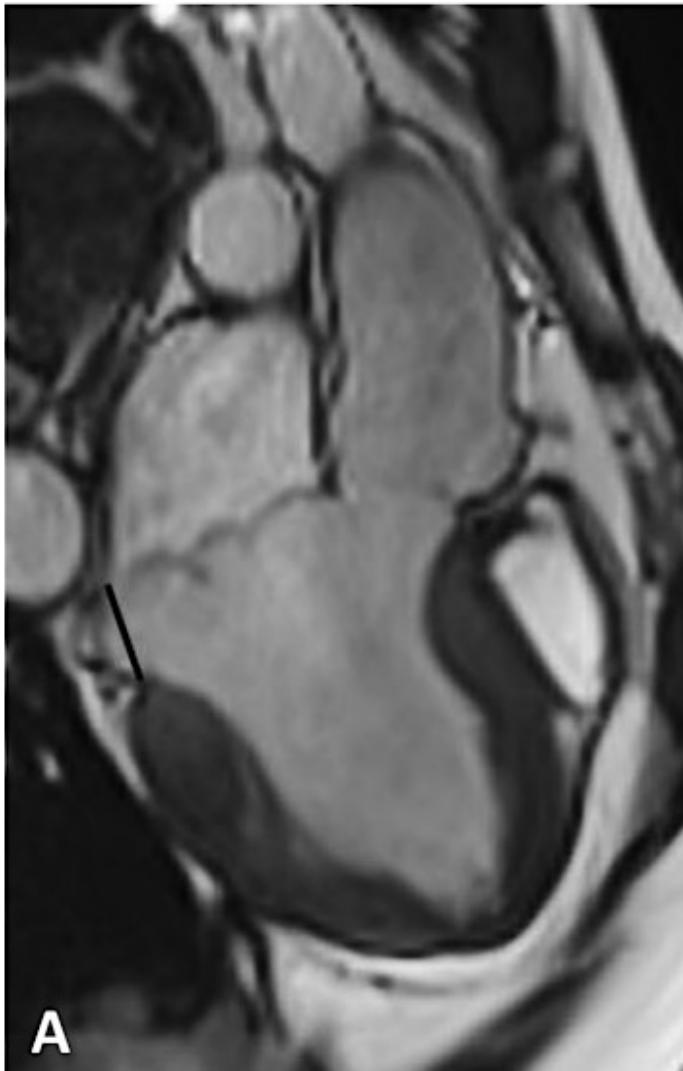
FRAME BY FRAME

BP:100/70MMHG

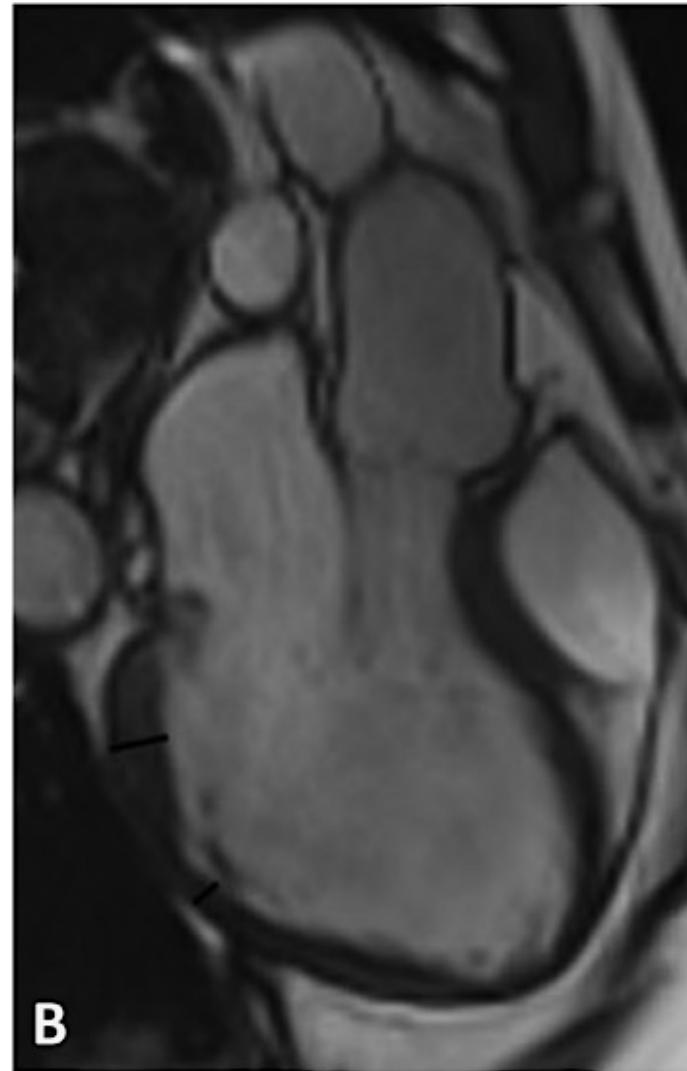


MAD DIAGNOSIS

Jean-

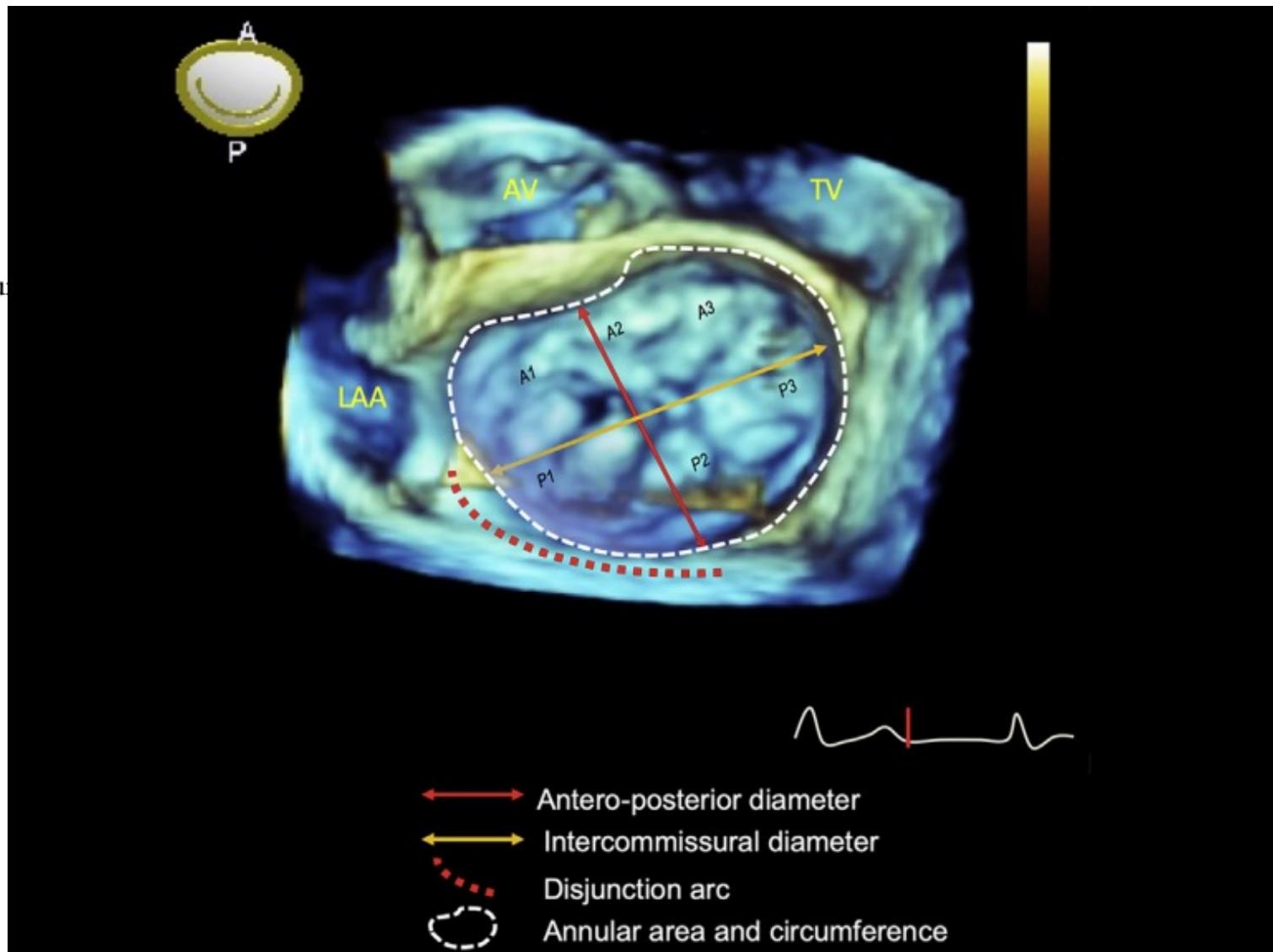


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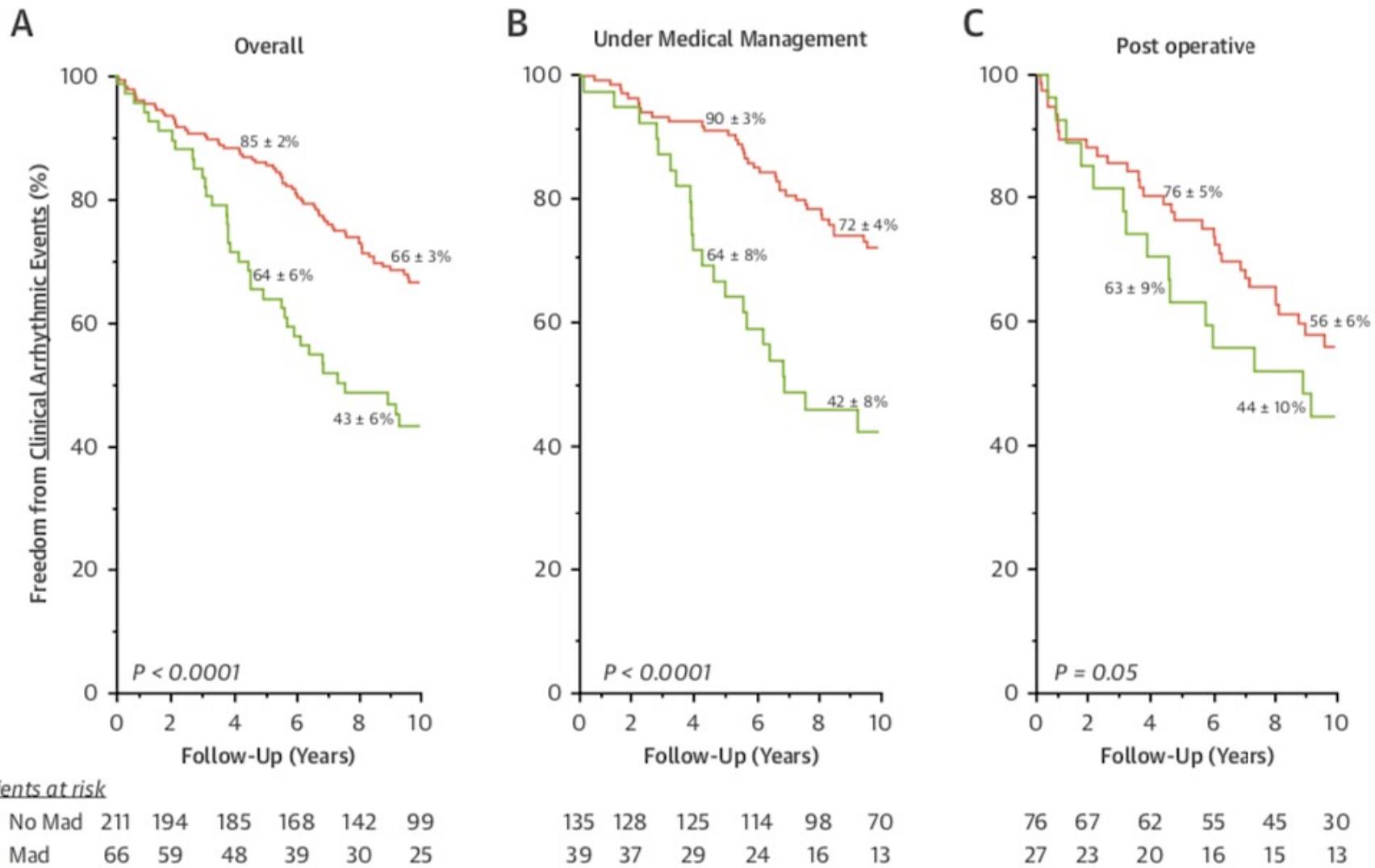


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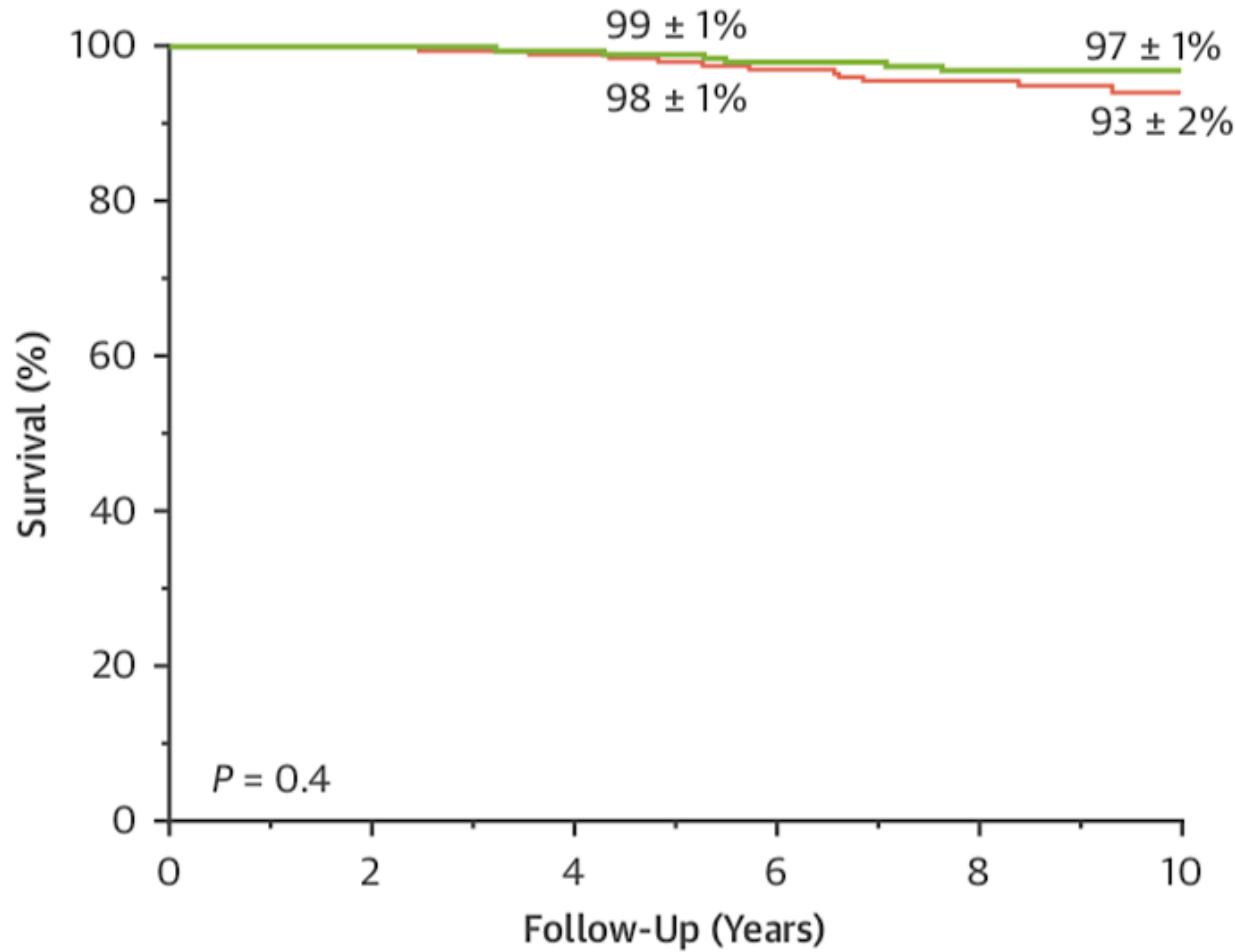
MAD DIAGNOSIS



MAD Outcome



MAD Outcome



Patients at risk

No Mad	185	185	184	180	145	87
Mad	179	179	179	176	147	90

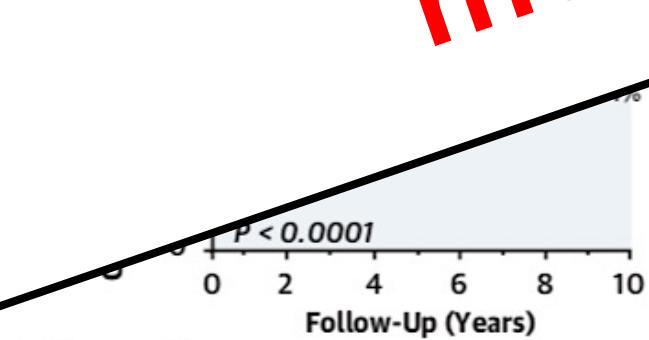
MAD Outcome

MAD PHENOTYPE

Younger age
Larger LV
Reduced LVEF

MAD

Message #2
MAD = arrhythmia monitoring

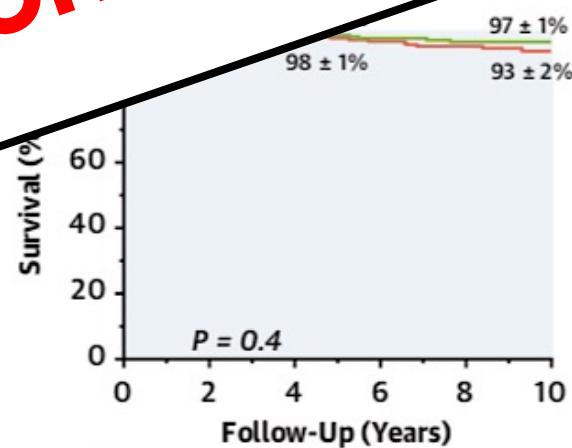


Patients at risk

No MAD	300	280	268	249	211	151
MAD	135	123	107	92	77	57

Patients at risk

No MAD	185	185	184	180	145	87
MAD	179	179	179	176	147	90



**/ So now what should we do
in daily practice ? /**

AMVP Management



Europace (2022) 00, 1–23
European Society of Cardiology
<https://doi.org/10.1093/europace/euac125>

CONSENSUS DOCUMENT

EHRA expert consensus statement on arrhythmic mitral valve prolapse and mitral annular disjunction complex in collaboration

Avi Sabbag (chair) ^{1*}, Benjamin Essayagh ^{2,3}, Juan David Ramírez Barrera ⁴, Cristina Basso ⁵, Ana Berni ⁶, Bernard Cosyns ⁷, Jean-Claude Deharo ⁸, Thomas Deneke ⁹, Luigi Di Biase¹⁰, Maurice Enriquez-Sarano ¹¹,

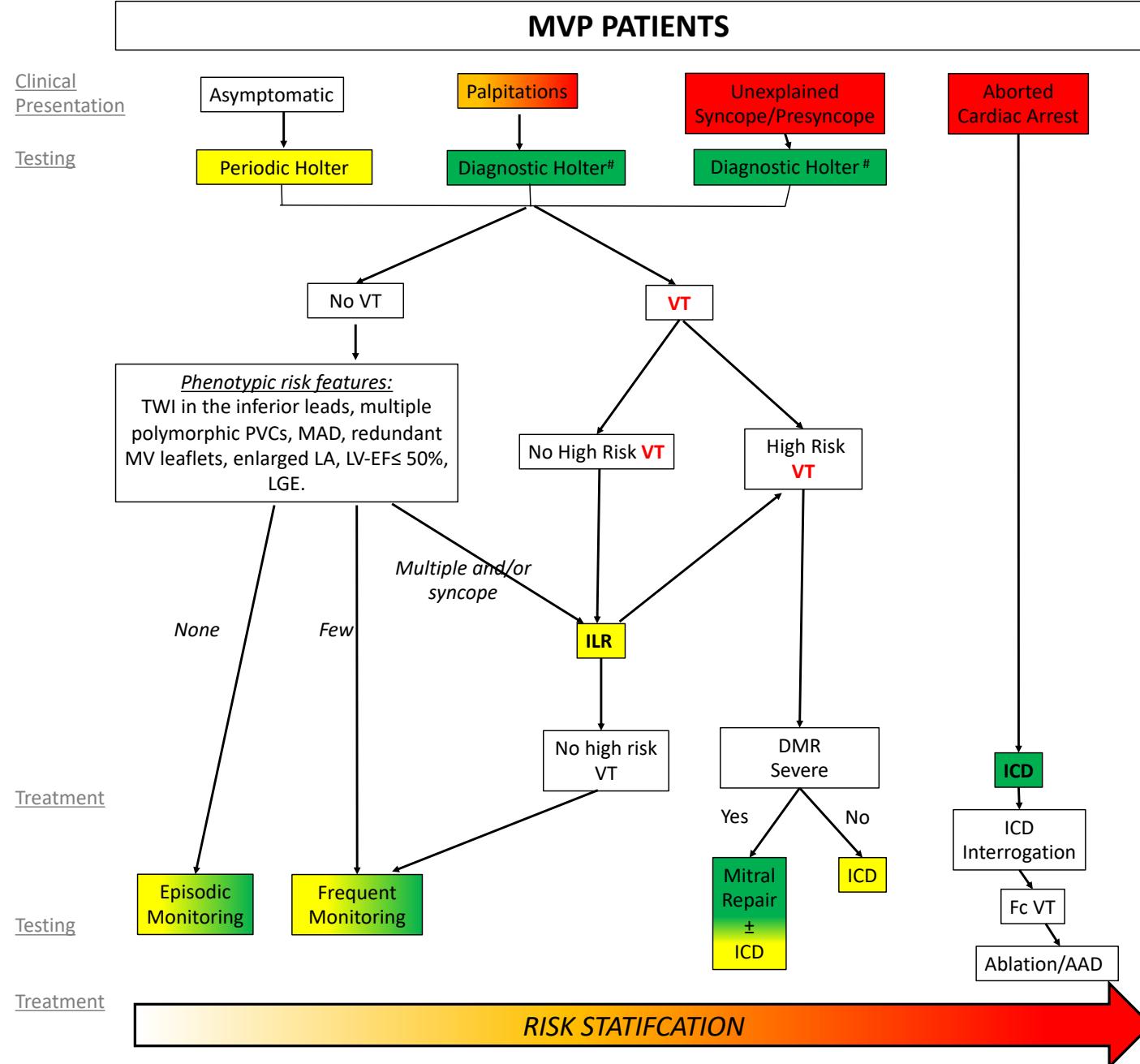
Box 2 The diagnosis of arrhythmic MVP requires:

- The presence of MVP (with or without MAD)
- The presence of ventricular arrhythmia that is
 - Frequent ($\geq 5\%$ total PVC burden) or
 - Complex (NSVT, VT, VF)
- The absence of any other well-defined arrhythmic substrate

Table 3 Arrhythmia severity classification

Severity	Arrhythmia burden/rate	Risk of mortality HR [95% CI]
Mild	PVC $\geq 5\%$ and/or ventricular arrhythmia <120 bpm	1.20 [0.68–2.14], $P=0.5$
Moderate	VT runs 120–179 bpm	
Severe	VT runs ≥ 180 bpm and/or history of sustained VT/VF	2.94 [1.36–6.36], $P=0.006$

HR, hazard ratio; PVC, premature ventricular contractions; VT, ventricular tachycardia.



MVP PATIENTS

Clinical Presentation

Asymptomatic

Palpitations

Unexplained
Syncope/Presyncope

Testing

Periodic Holter

Diagnostic Holter#

Message #3

AMVP = arrhythmia monitoring

Testing

Episodic Monitoring

Frequent Monitoring

No high risk
VTDMR
Severe

ICD

Mitral Repair
± ICD

ICD

ICD
Interrogation

Fc VT

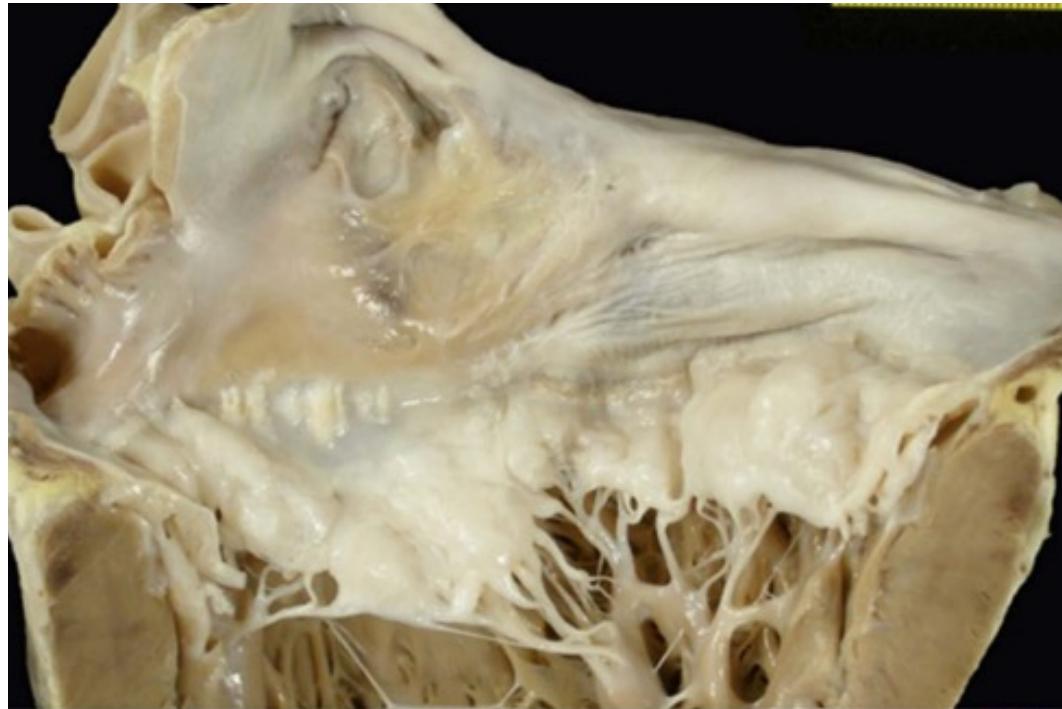
Ablation/AAD

Treatment

RISK STRATIFICATION

/ MVP in 2023 /

An imperious need to
look for AMVP and arrhythmia



/ Thank you /