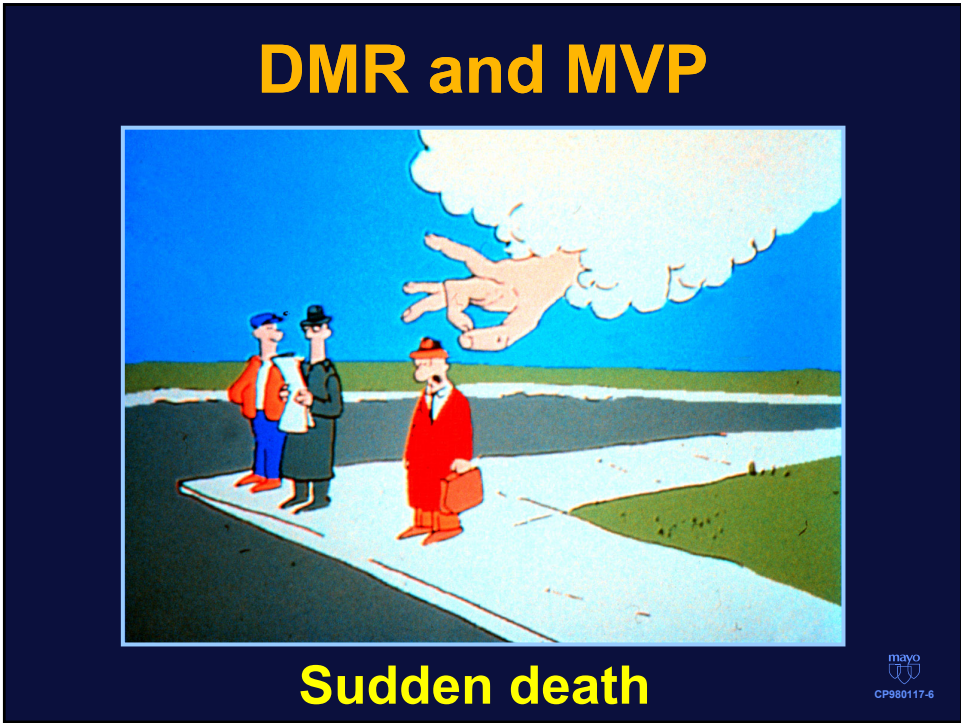
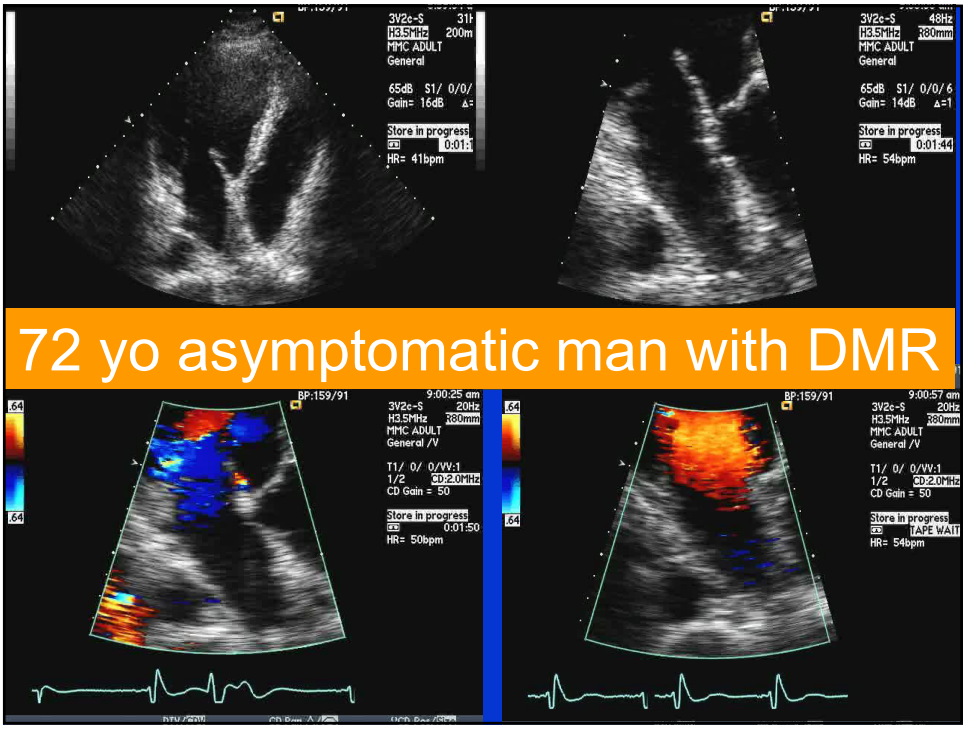




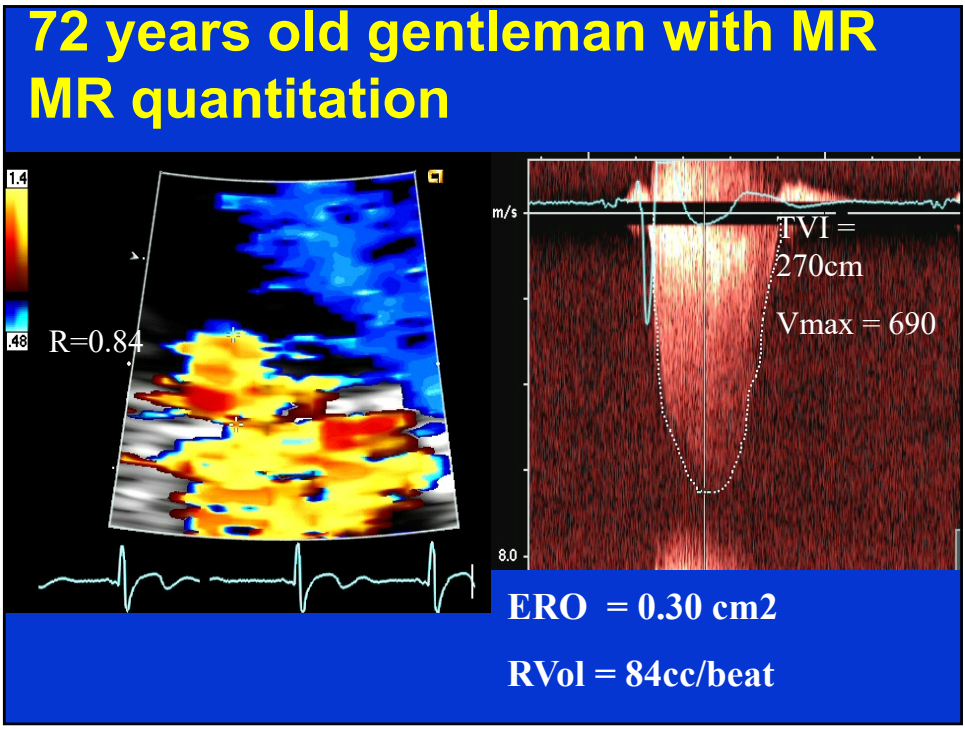
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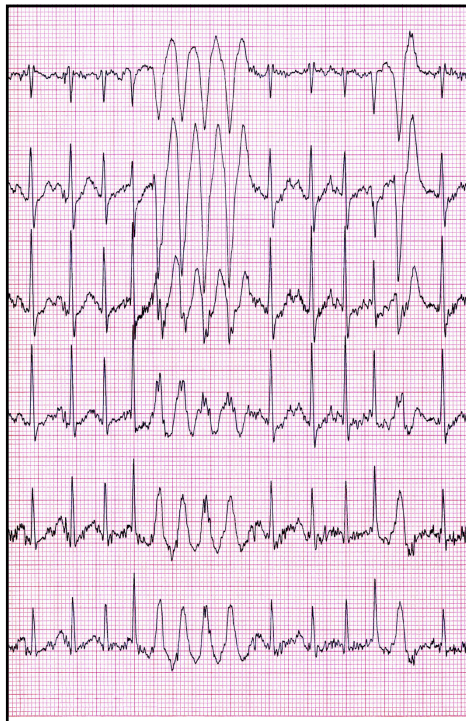
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3



4



- Ex testing:
12 mins on O₂
protocol
- Double product:
31,000
- Peak VO₂:
26 ml/kg/min or
107 % of expected
Peak exercise VT

Patient still not
interested in surgery

5

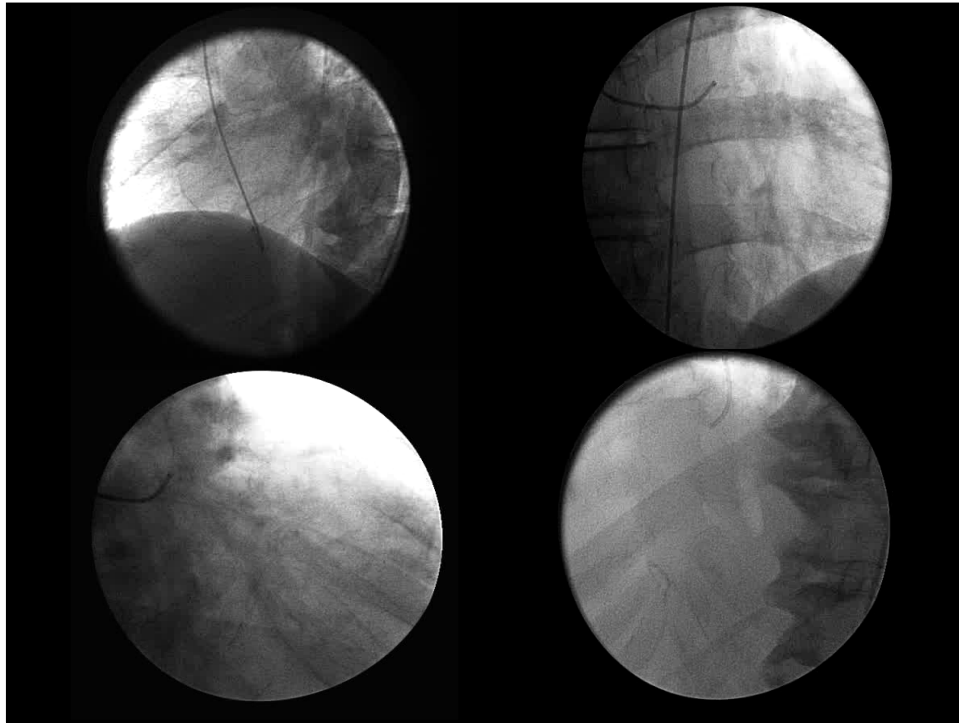
1 month after last visit: Cardiac arrest

- *'He was sitting on packing box, then slumped over and was found on the floor unresponsive.'*
- *911 was called, Patient found in VFib.*
- *Patient was shocked twice. Returned to sinus rhythm*

In Hospital

- **EKG**: sinus rhythm, 86/mn
- **Physical examination**: O₂sat 97% on room air, BP147/84, no sign of pulmonary edema, systolic murmur 3/6
- **CT** : No evidence of hemorrhage, with a small lacunar infarct in the right subinsular region
- **Coronary and LV angiograms**

6



7

CORONARY Summary

- middle LAD is 40% obstructed by a single discrete lesion
- First marginal branch is 60% obstructed by a single lesion
- middle RCA by a single mild discrete lesion

LV Gram

- Mild LV dilatation, EF 59%
- Moderate MR ?

CARDIAC SURGERY

- Mitral valve repair
- Coronary artery bypass grafting IMA to LAD, SVG OM
- Then ICD implantation.....no further discharge!

8

Valve Disease

Sudden Death in Mitral Regurgitation Due to Flail Leaflet

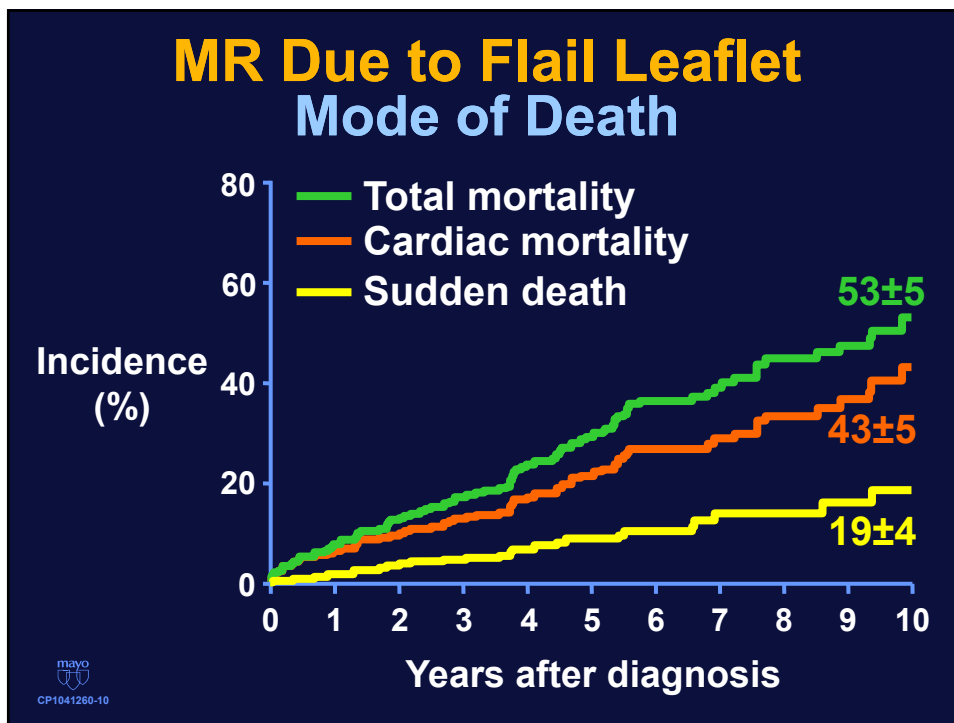
Francesco Grigioni, MD,* Maurice Enriquez-Sarano, MD, FACC,* Lieng H. Ling, MD,*
Kent R. Bailey, PhD,† James B. Seward, MD, FACC,* A. Jamil Tajik, MD, FACC,*
Robert L. Frye, MD, FACC*

Rochester, Minnesota

SUDDEN DEATH IN MR-FL 25/348 Patients, Age 71±9 Yrs

- Male 84%
- Atrial fibrillation (36%)
- Hypertension (28%)
- Posterior leaflet (87%)

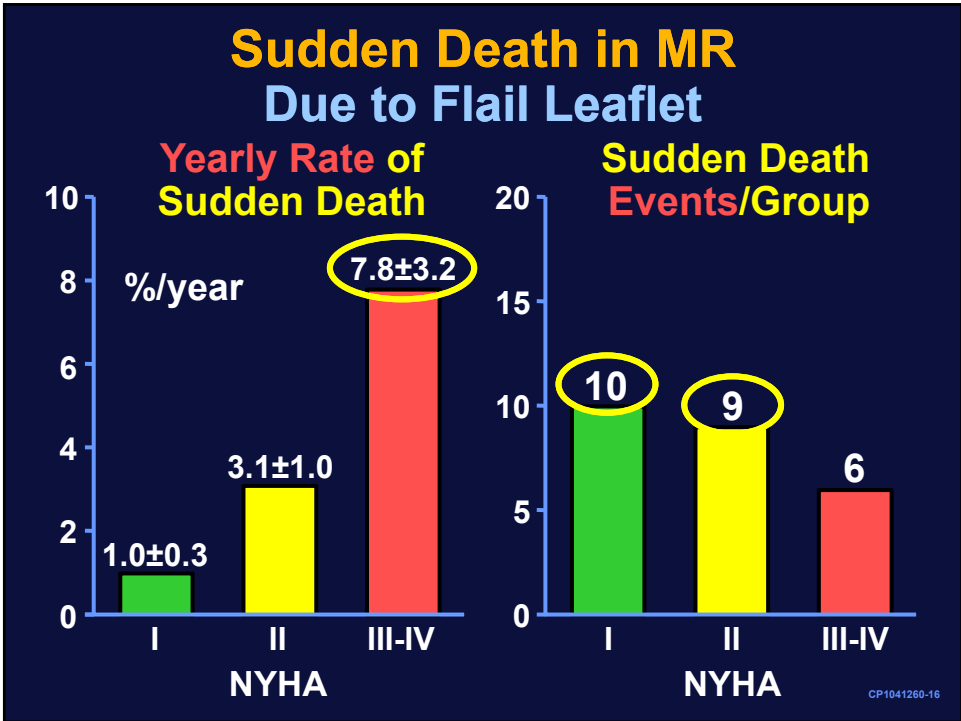
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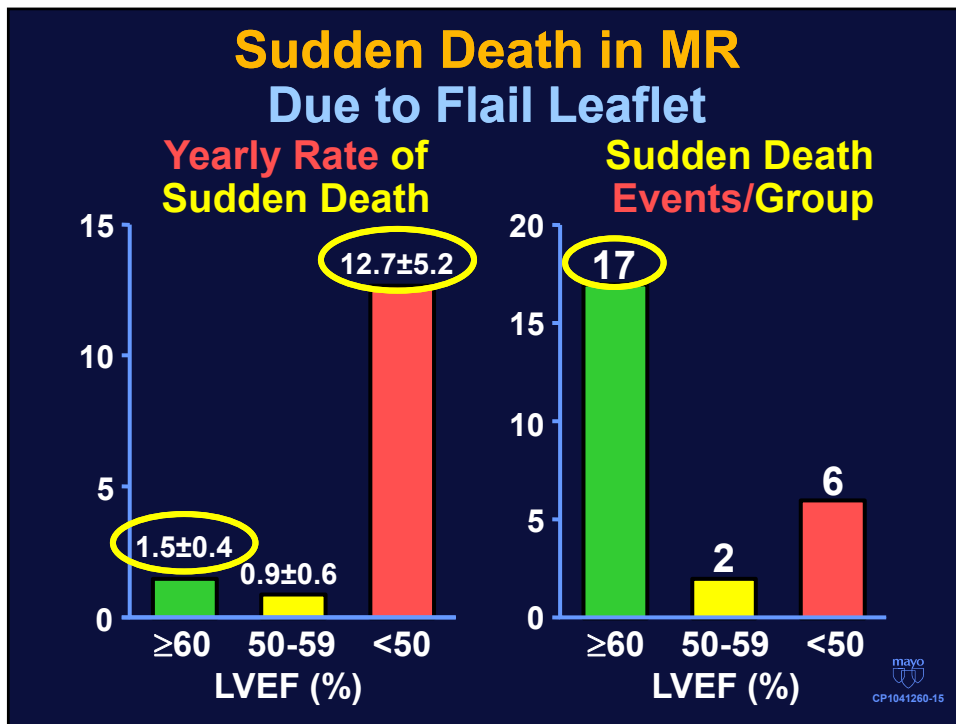
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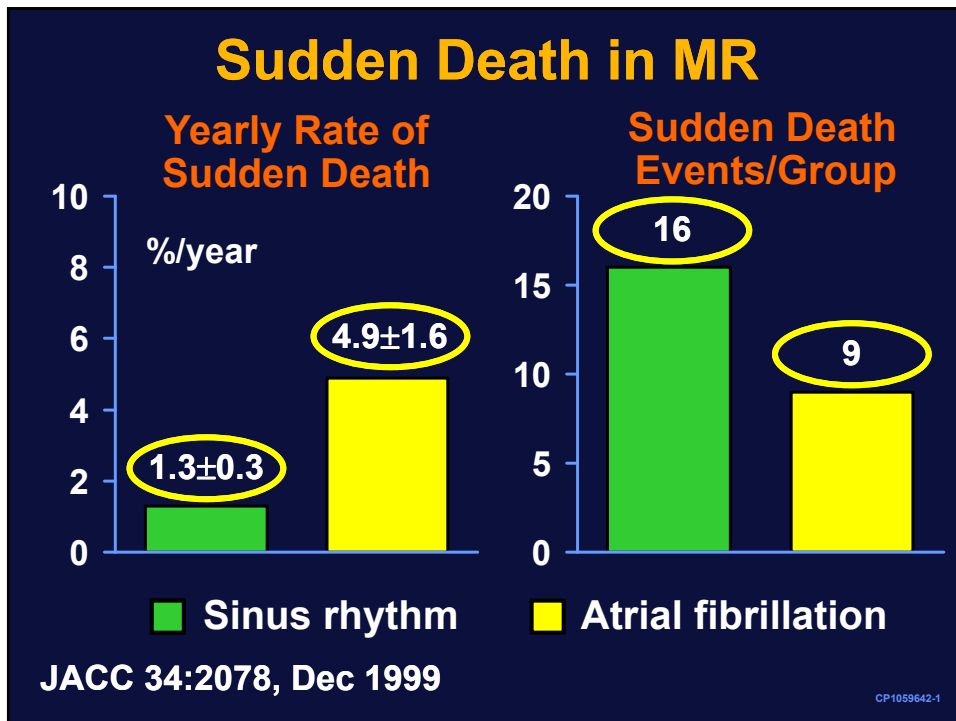
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14

Mitral Regurgitation
Sudden Death Rate

No Risk Factor
0.8% per year

Effect of Surgery

Hazard Ratio
0.29 [0.11-0.72]



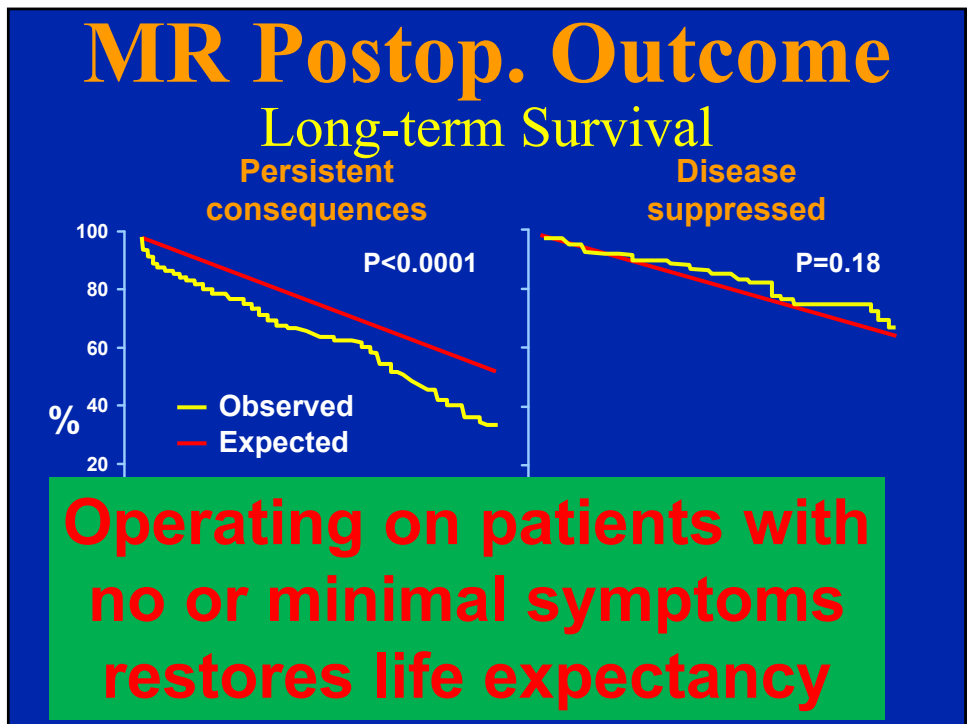
15

Degenerative MR

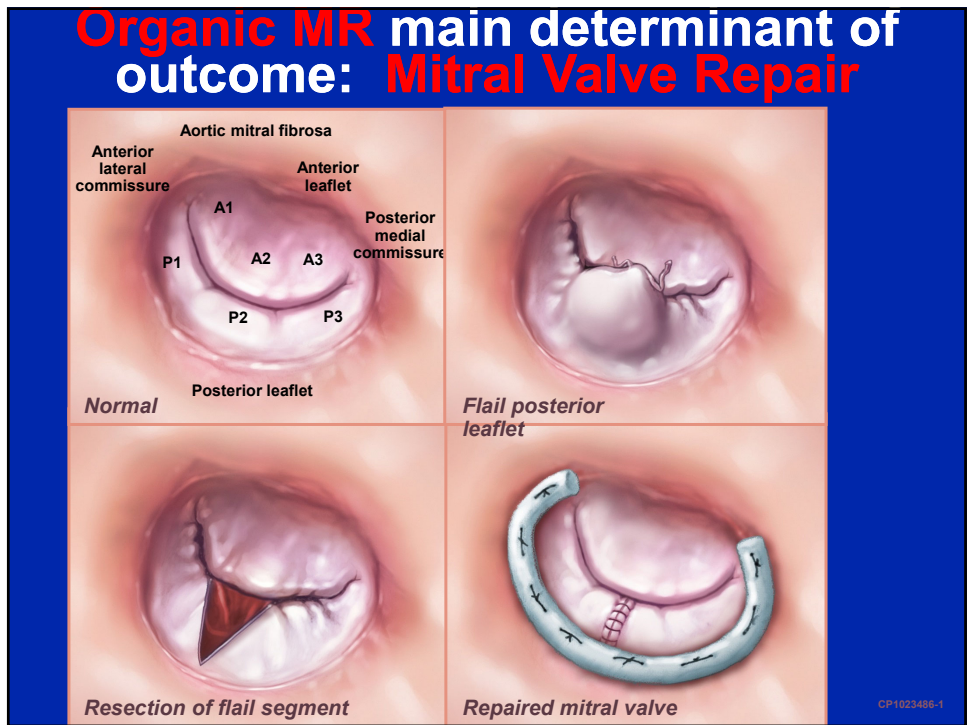
Sudden Death

- Notable proportion of overall mortality of DMR
- More frequent with Sx, low EF, AF
- SD events may occur without risk factors
- SD rate is markedly reduced by Mitral Surgery

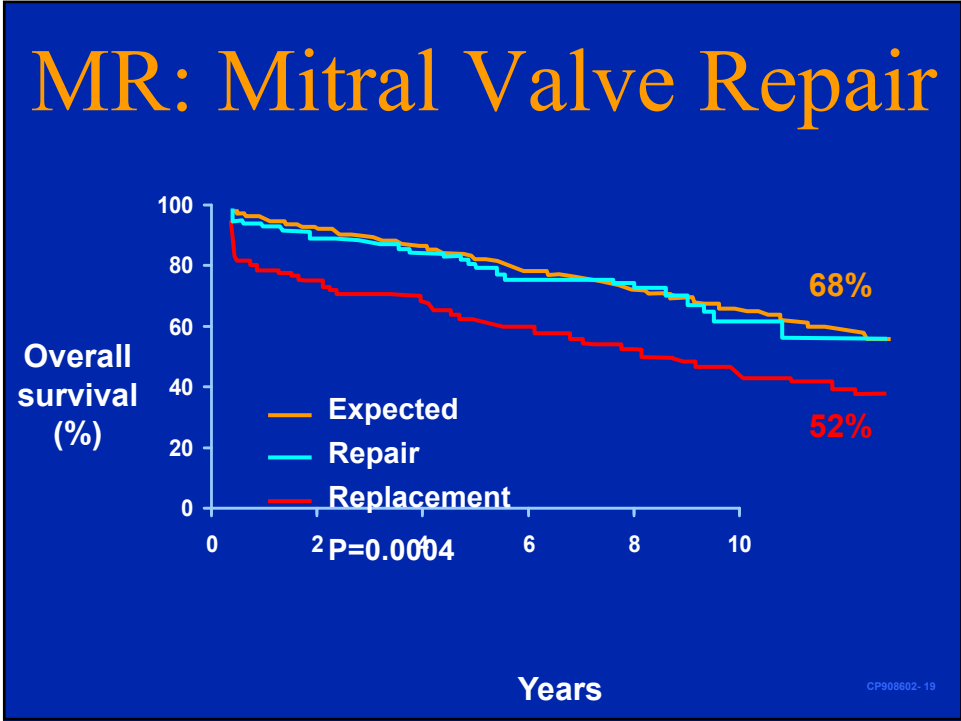
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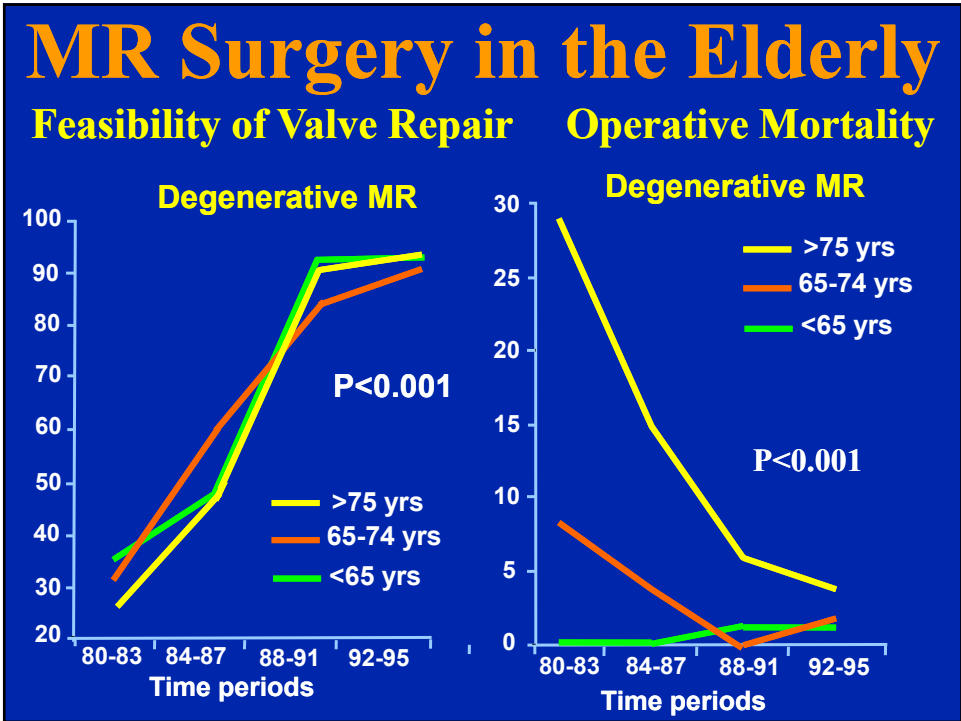
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Research

Original Investigation

Association Between Early Surgical Intervention vs Watchful Waiting and Outcomes for Mitral Regurgitation Due to Flail Mitral Valve Leaflets

Rakesh M. Suri, MD, DPhil; Jean-Louis Vanoverschelde, MD; Francesco Grigioni, MD, PhD; Hartzell V. Schaff, MD; Christophe Tribouilloy, MD; Jean-Francois Avierinos, MD; Andrea Barbieri, MD; Agnes Pasquet, MD; Marianne Huebner, PhD; Dan Rusinaru, MD; Antonio Russo, MD; Hector I. Michelena, MD; Maurice Enriquez-Sarano, MD

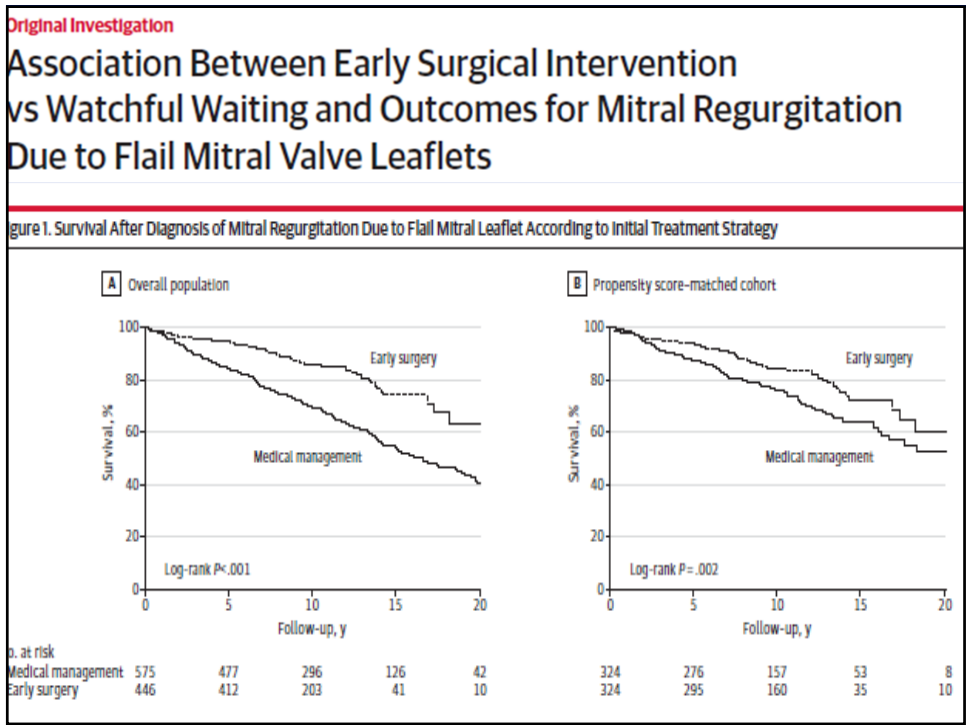
IMPORTANCE The optimal management of severe mitral valve regurgitation in patients without class I triggers (heart failure symptoms or left ventricular dysfunction) remains controversial in part due to the poorly defined long-term consequences of current management strategies. In the absence of clinical trial data, analysis of large multicenter registries is critical.

OBJECTIVE To ascertain the comparative effectiveness of initial medical management (nonsurgical observation) vs early mitral valve surgery following the diagnosis of mitral regurgitation due to flail leaflets.

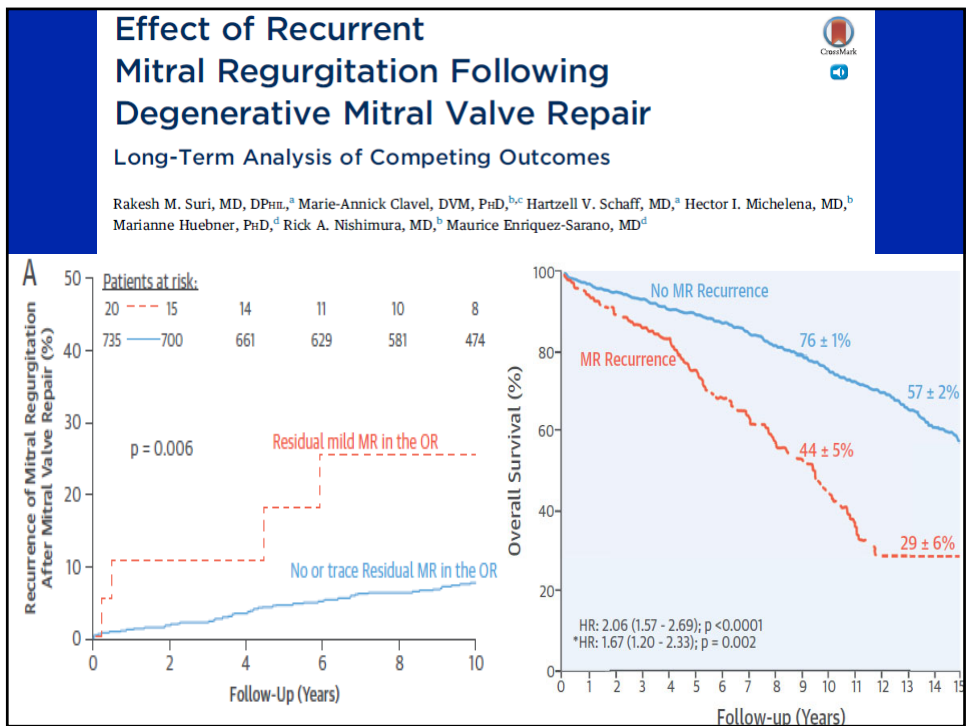
DESIGN, SETTING, AND PARTICIPANTS The Mitral Regurgitation International Database (MIDA) registry includes 2097 consecutive patients with flail mitral valve regurgitation (1980-2004) receiving routine cardiac care from 6 tertiary centers (France, Italy, Belgium, and the United States). Mean follow-up was 10.3 years and was 98% complete. Of 1021 patients with mitral regurgitation without the American College of Cardiology (ACC) and the American Heart Association (AHA) guideline class I triggers, 575 patients were initially medically managed and

← Editorial page 587
 + Supplemental content at jama.com

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Degenerative MR

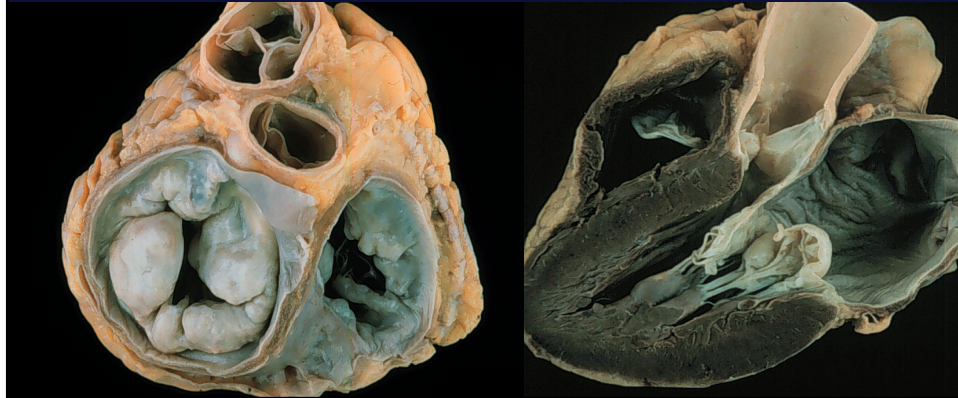
Sudden Death

In patients with DMR moderate or severe, the best prevention of SD is:

- Elective mitral valve **repair**
- Performed at **low-risk**
- With requirement of **“Perfection”**

24

Mitral Valve Prolapse Outcome is a great source of **Confusion**



25

Brit. Heart J., 1968, 30, 203.

Few Complications

Late Systolic Murmurs and Non-Ejection ("Mid-Late") Systolic Clicks

An Analysis of 90 Patients

J. B. BARLOW, C. K. BOSMAN, W. A. ...

From the C.S.I.R. Cardio-Pulmonary Laboratory, Departments of Medicine and the Cardiovascular Laboratory, University of Chicago, Chicago, Ill.

The conduction system in mitral valve prolapse syndrome with sudden death

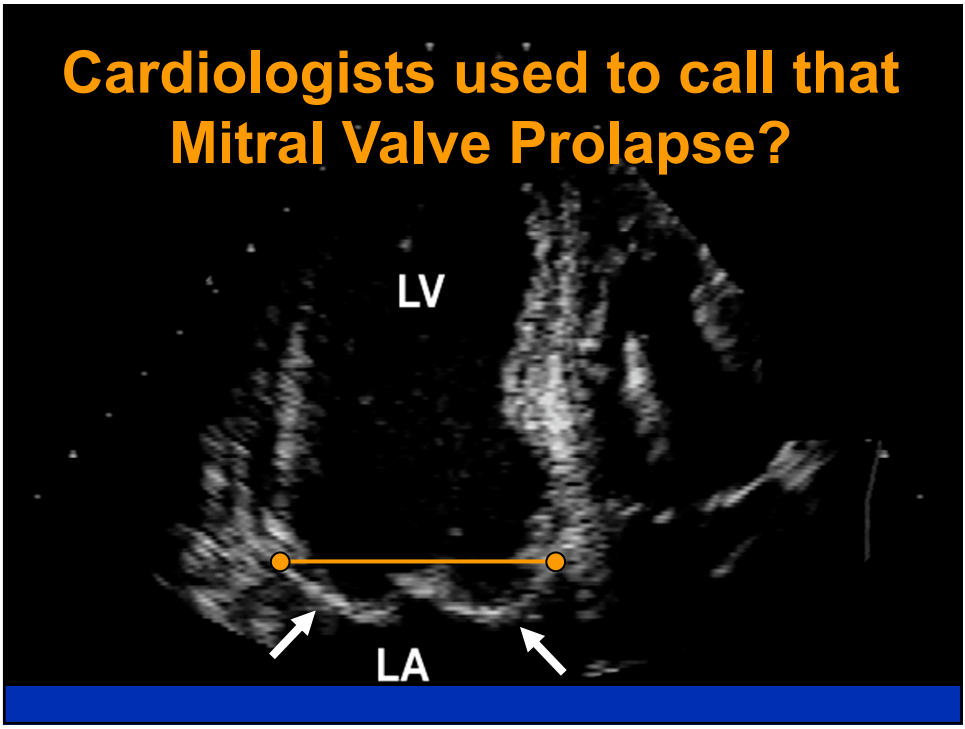
Saroja Bharati, M.D., Arthur S. Granston, M.D., Philip R. Liebson, M.D., Henry S. Loeb, M.D., Kenneth M. Rosen, M.D., and Maurice Lev, M.D.*
Chicago, Ill.

Sudden death

Center, Faculty of Medicine, Health Sciences, University of Toronto, Toronto, Ontario, Canada

We are confused by discordant data on MVP outcome

26



27

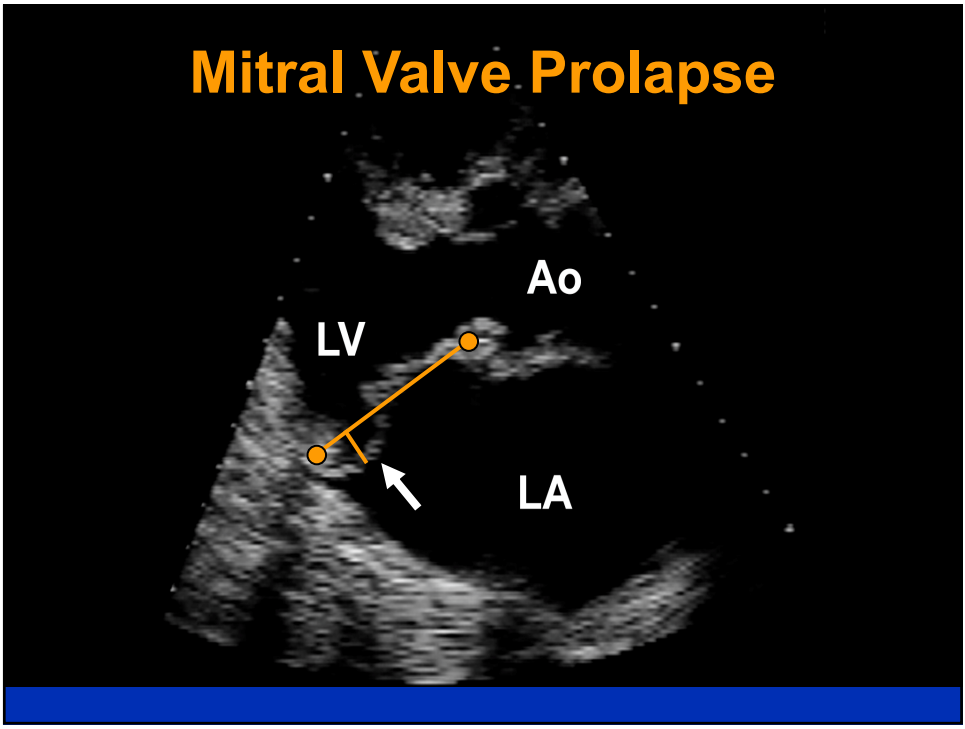
The Saddle-shape of the Mitral annulus

The diagram illustrates the heart with several scan planes and a SPARK GAP ARRAY. A red arrow points from the title to two cross-sectional views of the mitral valve annulus. The left view shows the annulus in a cross-section with the right atrium (RA) and left atrium (LA) labeled, and a 1 cm scale bar. The right view shows the annulus in a longitudinal section with the aorta (AO), left atrium (LA), and left ventricle (LV) labeled.

Three-Dimensional Echocardiographic Reconstruction of the Mitral Valve, With Implications for the Diagnosis of Mitral Valve Prolapse

Robert A. Levine, MD, Mark D. Handschumacher, BS,
Anthony J. Sanfilippo, MD, Albert A. Hagege, MD, Pamela Harrigan, RDMS,

28



29

Prevalence of MVP

Previously overestimated	5-17%
With current criteria	0.6-2.4%

30

Sudden Death in the Mitral Valve Prolapse-Click Syndrome

ROBERT M. JERESATY
 Hartford, Connecticut

The mitral valve prolapse-click syndrome continues. Murmurs were heard and recorded in 1965. The electrocardiogram was normal.

Refractory Ventricular Tachycardia and Fibrillation in a Patient With the Prolapsing Mitral Leaflet Syndrome: Successful Control With Overdrive Pacing

RHYTHM STRIPS (MCL I)

5:00 PM
 5:30 PM
 6:00 PM

S₁ C S₂

31

The New England Journal of Medicine

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Volume 313 NOVEMBER 21, 1985 Number 21

ECHOCARDIOGRAPHICALLY DOCUMENTED MITRAL-VALVE PROLAPSE Long-Term Follow-up of 237 Patients

Probability

Years

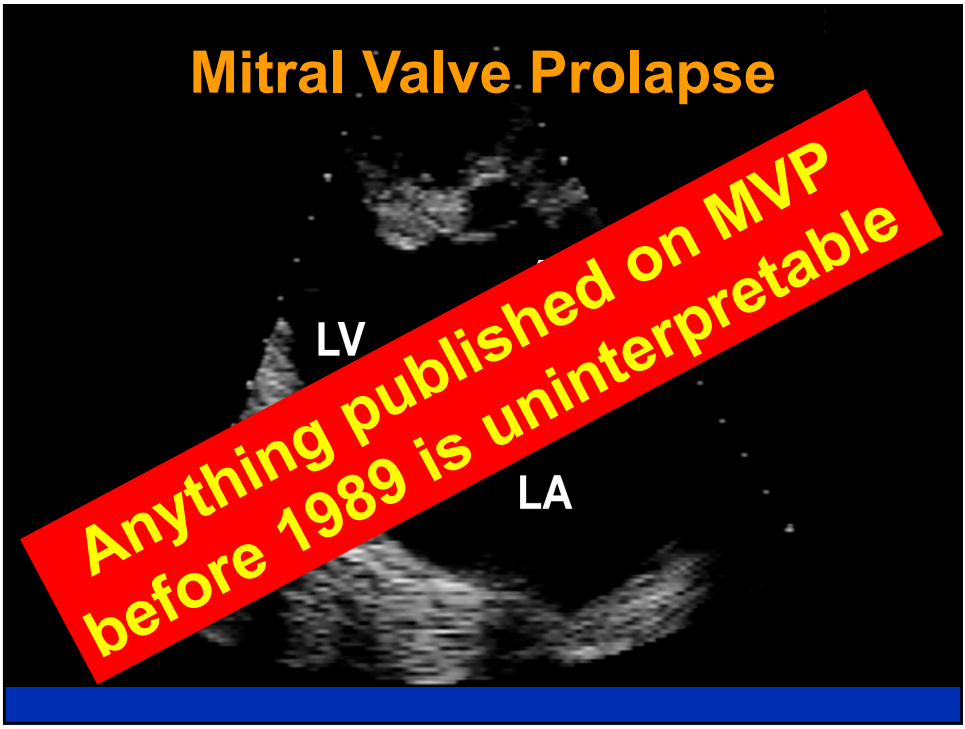
(237) (224) (223) (220) (215) (210) (176) (69) (22)

Table 2. Association of Complications with Redundancy of Mitral-Valve Leaflets in 237 Patients with Mitral-Valve Prolapse.

COMPLICATION	LEAFLETS	
	REDUNDANT (N = 97)	NONREDUNDANT (N = 140)
Sudden death	6	0
Infective endocarditis	3	0
Cerebral embolic event*	1	1
Total (%)	10 (10.3)	1 (0.7)

*In patients in normal sinus rhythm.

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Prevalence and Clinical Outcome of Mitral Valve Prolapse

The New England Journal of Medicine July 1st, 1999

TABLE 2. PREVALENCE OF VARIOUS CLINICAL FINDINGS ACCORDING TO THE PRESENCE OR ABSENCE OF MITRAL-VALVE PROLAPSE.

CLINICAL FINDING	MITRAL-VALVE PROLAPSE (N=84)	NO MITRAL-VALVE PROLAPSE (N=3407)
	no. (%)	
Congestive heart failure	0	25 (0.7)
Atrial fibrillation	1 (1.2)	58 (1.7)
Cerebrovascular disease*	1 (1.2)	52 (1.5)
Syncope	3 (3.6)	103 (3.0)

*Cerebrovascular disease refers to stroke or transient ischemic attack.

Freed, LA et al. *N Engl J Med.* 1999;341:1-7.

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Mitral Valve Problems Are Not So Serious After All
 As many as 28 million Americans who were told they are in danger of stroke, heart failure and other problems because of mitral-valve prolapse probably do not have to worry. A new study showed that about 2.4 percent of the population — not the 5 percent to 15 percent previously estimated — have the condition abnormally long that can cause the heart

have it, it was found no more likely to cause the feared complications than those who do not.
 HOLCOM

Independent Dies

Is MVP Really and Uniformly Benign?

... of a week with a
 ... he pleaded
 ... count
 ... regulators
 ... misdemeanor
 ... count of failure to pay taxes, and avoided jail time even though he has already served time for a felony.
 But Kenneth W. Starr, the independent counsel who struck the deal, seemed even

TH.
 SU,

... Republican and Democratic Presi-

Webster L. Hubbell.

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Natural History of Asymptomatic Mitral Valve Prolapse in the Community

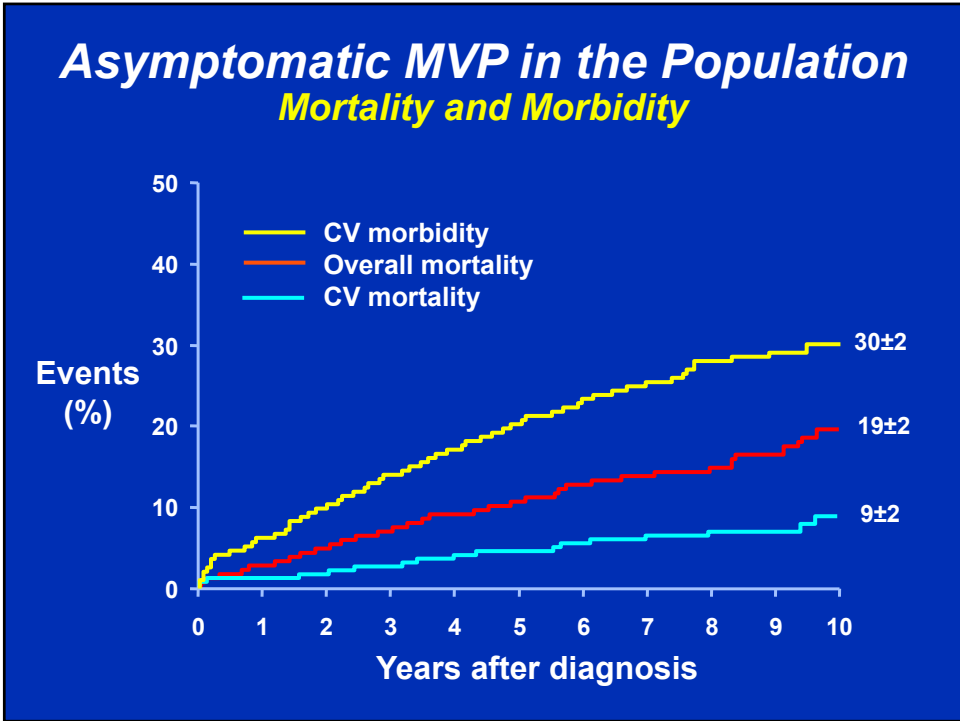
Jean-François Avierinos, MD; Bernard J. Gersh, MB, ChB, DPhil; L. Joseph Melton III, MD; Kent R. Bailey, PhD; Clarence Shub, MD; Rick A. Nishimura, MD; A. Jamil Tajik, MD; Maurice Enriquez-Sarano, MD

Background—The outcome of mitral valve prolapse (MVP) is controversial, with marked discrepancies in reported complication rates.

Methods and Results—We conducted a community study of all Olmsted County, Minn, residents first diagnosed with asymptomatic MVP between 1989 and 1998 (N=833). Diagnosis, motivated by auscultatory findings (n=557) or incidental (n=276), was always confirmed by echocardiography with the use of current criteria. End points analyzed during 4581 person-years of follow-up were mortality (n=96, 19±2% at 10 years), cardiovascular morbidity (n=171), and MVP-related events (n=109, 20±2% at 10 years). The most frequent primary risk factors for cardiovascular mortality were mitral regurgitation from moderate to severe (P=0.002, n=131) and, less frequently, ejection fraction <50% (P=0.003, n=31). Secondary risk factors independently predictive of cardiovascular morbidity were slight mitral regurgitation, left atrium ≥40 mm, flail leaflet, atrial fibrillation, and age ≥50 years (all P<0.01). Patients with only 0 or 1 secondary risk factor (n=430) had excellent outcome, with 10-year mortality of 5±2% (P=0.17 versus expected), cardiovascular morbidity of 0.5%/y, and MVP-related events of 0.2%/y. Patients with ≥2 secondary risk factors (n=250) had mortality similar to expected (P=0.20) but high cardiovascular morbidity (6.2%/y, P<0.01) and notable MVP-related events (1.7%/y, P<0.01). Patients with primary risk factors (n=153) showed excess 10-year mortality (45±9%, P=0.01 versus expected), high morbidity (18.5%/y, P<0.01), and high MVP-related events (15%/y, P<0.01).

Conclusions—Natural history of asymptomatic MVP in the community is widely heterogeneous and may be severe. Clinical and echocardiographic characteristics allow separation of the majority of patients with excellent prognosis from subsets of patients displaying, during follow-up, high morbidity or even excess mortality as direct a consequence of MVP. (*Circulation*. 2002;106:1355-1361.)

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Asymptomatic MVP Risk Stratification

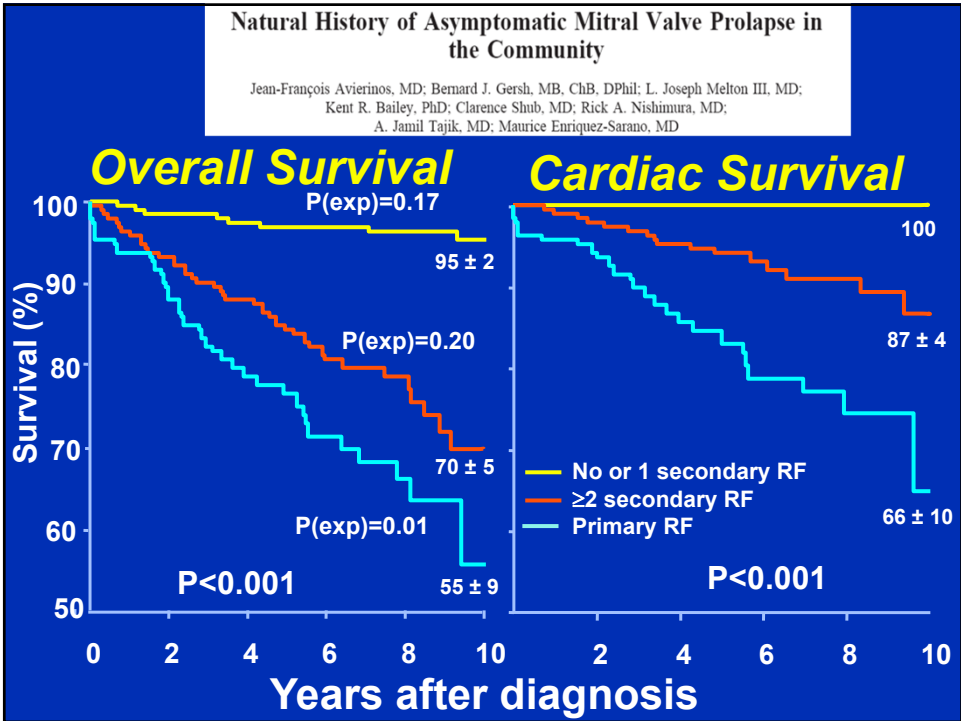
Primary Risk Factors (mort)	Secondary Risk Factors (morb)
<ul style="list-style-type: none">• EF < 50%• MR ≥ moderate	<ul style="list-style-type: none">• Age ≥ 50 years• A Fib• Slight MR• Flail leaflet• LA ≥ 40 mm

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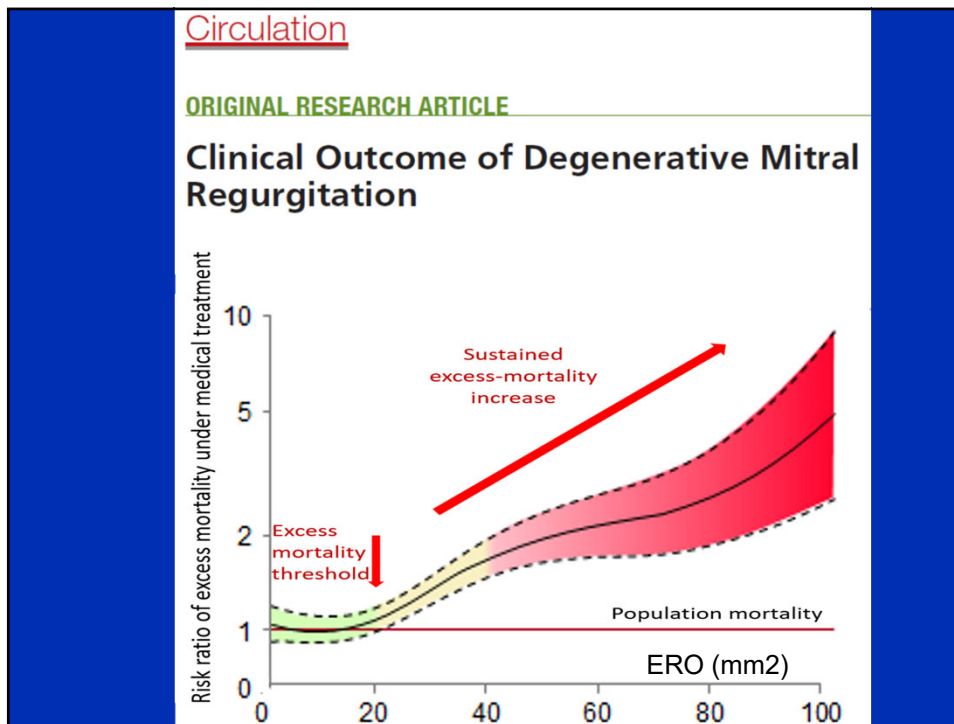
Outcome of MVP – Risk Stratification

3 groups	No.	%
No or 1 secondary RF	430	52
≥2 secondary RF	250	30
Primary RF	153	18
• MR ≥ moderate	131	
• EF <50%	31	

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MVP
in the current era
All studies confirm:
-MR severity is the major determinant of MVP outcome
-Poor outcome in the group with severe MR
-Benign outcome in the group with no/mild MR

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Mitral Valve Prolapse and Sudden Death

“...Our son, Crick, had mitral valve prolapse. I think he is the only one who died suddenly of thousands of patients I’ve seen with mitral valve prolapse.”

*W. Proctor Harvey, MD, FACC:
 Conversations with the Editor
 William C. Roberts, MD*

Am J Cardiol 89:435, Feb 15, 2002

CP1058261-1

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Journal of the American College of Cardiology
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 Published by Elsevier Inc. Vol. 62, No. 3, 2013
 ISSN 0735-1097/\$36.00
<http://dx.doi.org/10.1016/j.jacc.2013.02.066>

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

Chenni S. Sriram, MBBS,* Faisal F. Syed, MBCrB,† M...
 Jonathan N. Johnson, MD,* Maurice Enriquez-Sar...
 Bryan C. Cannon, MD,* Samuel J. Asirvatham, MD...
 Rochester, Minnesota

Table 1. Demographics of OHCA Cohort (n = 24)

Demographic	With MVP (n = 10)	No MVP (n = 14)	p Value
n	10	14/24 (58%)	—
Age at sentinel event (years)	34.7 (median 34.7; range: 5-60)	32 ± 14 (median 29.4; range: 14-51)	0.84
Women	9/10 (90%)	7/14 (50%)	0.04
QTc interval (ms)	434 ± 22	428 ± 28	0.52
Cardiac arrest at home	8/10 (80%)	14/14 (100%)	0.16
Activity at time of arrest	5 wakeful rest, 7 acute emotional or physical stress, 0 sleep	8 wakeful rest, 2 acute emotional or physical stress, 0 sleep	9 wakeful rest, 5 acute emotional or physical stress, 0 sleep
ICD implantation	24/24 (100%)	10/10 (100%)	—
Follow-up after ICD placement (days)	3.8 ± 4.1 (median 1.8; range: 0.1-11.9)	7.2 ± 3.5 (median 7.3; range: 2.4-11.9)	0.0003
Patients with appropriate ICD therapies	13/24 (54%)	8/10 (80%)	0.04

Did they die with MVP Or Did they die because of MVP

44



45

Arrhythmic Mitral Valve Prolapse and Sudden Cardiac Death

Cristina Basso, MD, PhD*; Martina Perazzolo Marra, MD, PhD*; Stefania Rizzo, MD, PhD;
 Manuel De Lazzari, MD; Benedetta Giorgi, MD; Alberto Cipriani, MD;
 Anna Chiara Frigo, MSc; Ilaria Rigato, MD, PhD; Federico Migliore, MD, PhD;
 Kalliopi Pilichou, PhD; Emanuele Bertaglia, MD; Luisa Cacciavillani, MD, PhD;
 Barbara Bauce, MD, PhD; Domenico Corrado, MD, PhD; Gaetano Thiene, MD; Sabino Iliceto, MD

Circulation. 2015;132:556-566.

650 SCD age<40

MVP =43, 6.6%

Table 1. Clinical and Pathological Features of 43 Patients Who Died Suddenly With Isolated MVP

Variables	SCD Resulting From MVP (n=43)	Control Subjects (n=15)	P Value
MVP leaflet involvement			
Posterior, n (%)	13 (30)	0	...
Bileaflet, n (%)	30 (70)	0	...
Endocardial fibrous plaque, n (%)	25 (58)	0	...
Histology features, n (%)			
LV scar			
PM, n (%)	43 (100)	0	...
Inferobasal wall	38 (88)	0	...
Fibrous tissue /myocardium, % area			
PM, mean±SD	30.5±10.7	6.3±1.6	<0.0001
Inferobasal wall, mean±SD	33.1±7.6	6.4±1.4	<0.0001

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Morphofunctional Abnormalities of Mitral Annulus and Arrhythmic Mitral Valve Prolapse

Martina Perazzolo Marra, MD, PhD*; Cristina Basso, MD, PhD*; Manuel De Lazzari, MD; Stefania Rizzo, MD, PhD; Alberto Cipriani, MD; P. Letta Giorgi, MD; Carmelo Lacognata, MD; Ilaria Rigato, MD, PhD; ... Migliore, MD, PhD; Kalliopi Pilichou PhD; Luisa Cacciavillani, MD; ... Bertaglia, MD, PhD; Anna Chiara Frigo, MSc; Barbara Baucce; ... Torrado, MD, PhD; Gaetano Thiene

52 arrhythmic ... MR
 MRI for I

MAD is a feature of Arrhythmic MVP and MVP with SCD

MAD	31%	Autopsy SCD
Bi MVP	39%	MAD=
Complex VA	6%	MVP 3mm
		NoMVP 1mm

Circ Cardiovasc Imaging. 2016;9:e005030.

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Circulation

RESEARCH LETTER

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

Isolated MVP and SUD

There a typical phenotype found in SD associated with isolated MVP:

- Clinically: Syncope/presyncope
- Echo: severe myxomatous disease with annular disjunction
- ECG: PVC-VT from PPM or annulus

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ESC European Society of Cardiology
European Heart Journal (2023) 44, 3121–3135
<https://doi.org/10.1093/eurheartj/ehad491>

STATE OF THE ART REVIEW
Valvular heart disease

Arrhythmic mitral valve prolapse and mitral annular disjunction: pathophysiology, risk stratification, and management

Benjamin Essayagh ^{1,2}, Avi Sabbag ³, Edward El-Am ⁴, João L. Cavalcante ⁴, Hector I Michelena ¹, and Maurice Enriquez-Sarano ^{4*}

26 y.o. with hx of presyncope works as nursing aid SCA due to VF requiring 8 defibrillation for SR return

The top right image shows an ECG with a regular rhythm. The middle right image is a color Doppler echocardiogram showing mitral regurgitation. The bottom left image shows two parasternal short-axis views of the mitral valve with yellow arrows indicating prolapse and a dashed line indicating annular disjunction. The bottom right image shows two apical four-chamber views of the heart with a yellow arrow pointing to the mitral annular disjunction.

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Circulation

IN DEPTH

Mitral Valve Prolapse, Ventricular Arrhythmias, and Sudden Death

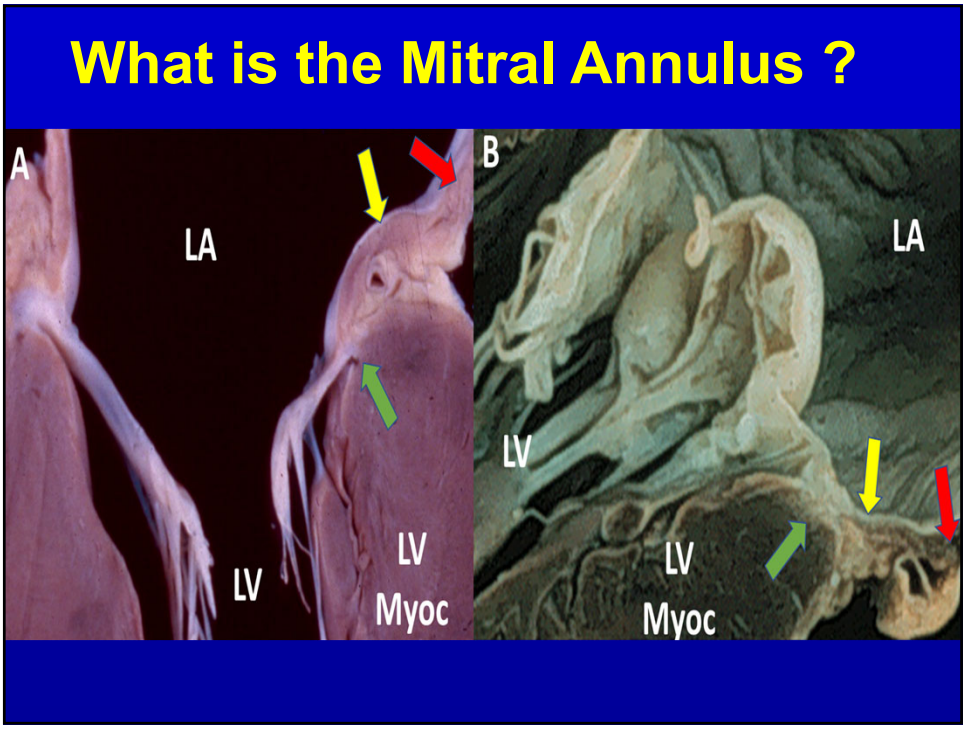
ABSTRACT: Despite a 2% to 3% prevalence of echocardiographically defined mitral valve prolapse (MVP) in the general population, the actual burden, risk stratification, and treatment of the so-called arrhythmic MVP are unknown. The clinical profile is characterized by a patient, usually female, with mostly bileaflet myxomatous disease, mild mitral regurgitation, and a high prevalence of ventricular arrhythmias.

Cristina Basso, MD, PhD
Sabino Iliceto, MD
Gaetano Thiene, MD
Martina Perazzolo Marra, MD, PhD

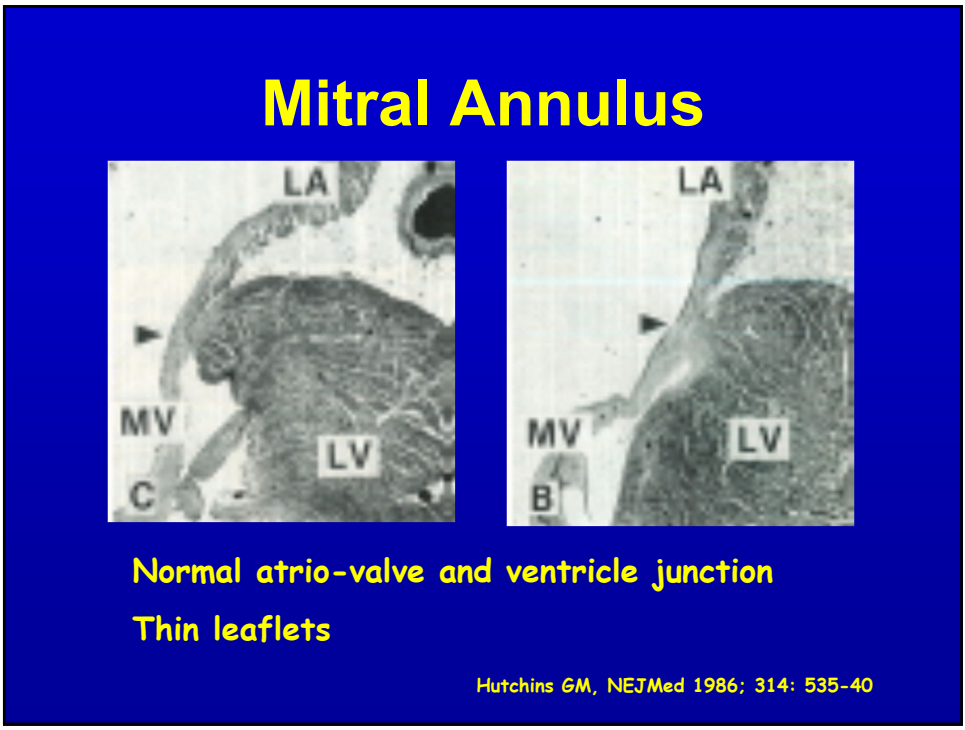
MAD, the red alert ?

To prevent the exponential increase in costs, referrals, and false-positive results, only MVP patients with red flags, particularly MAD and systolic curling, besides arrhythmic presentation, will undergo further investigation, including contrast-enhanced or T1 mapping CMR and a strict arrhythmia surveillance for proper management and SCD prevention.

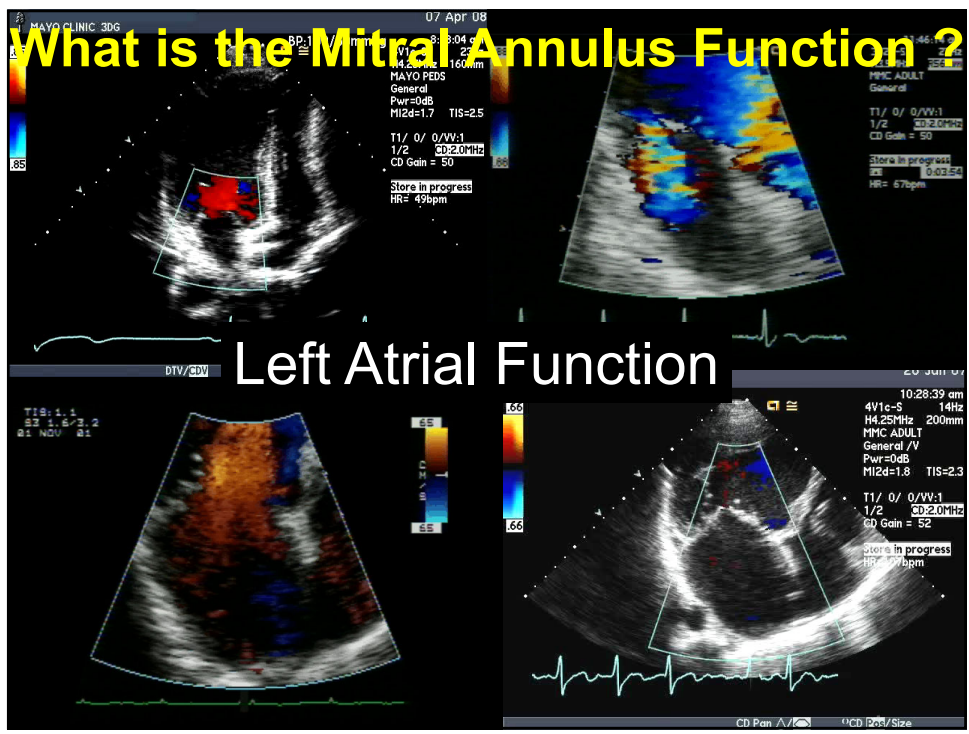
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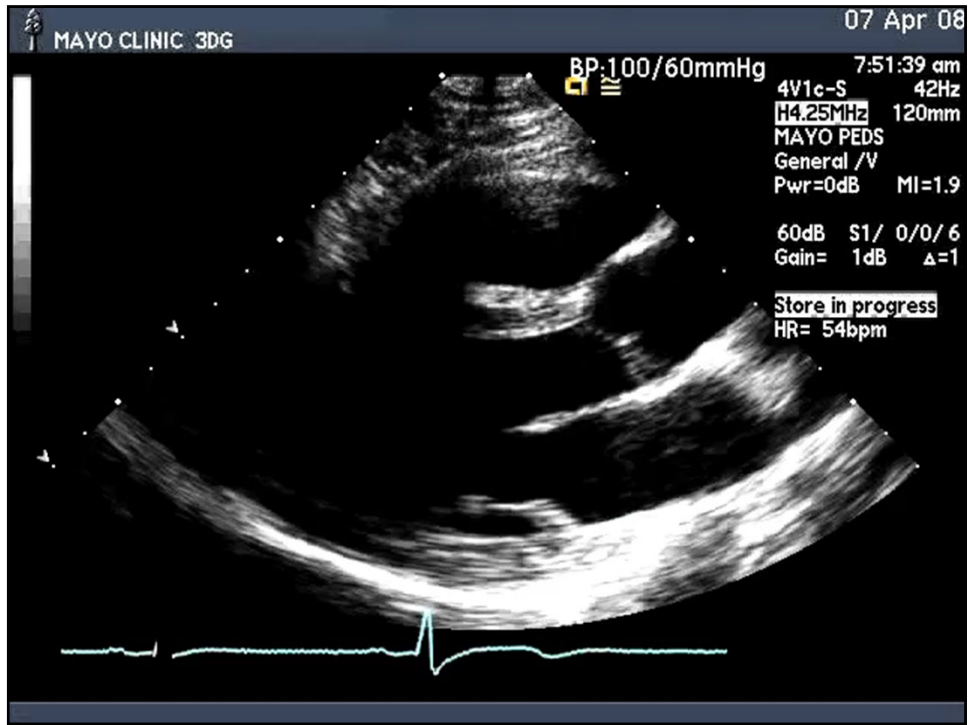
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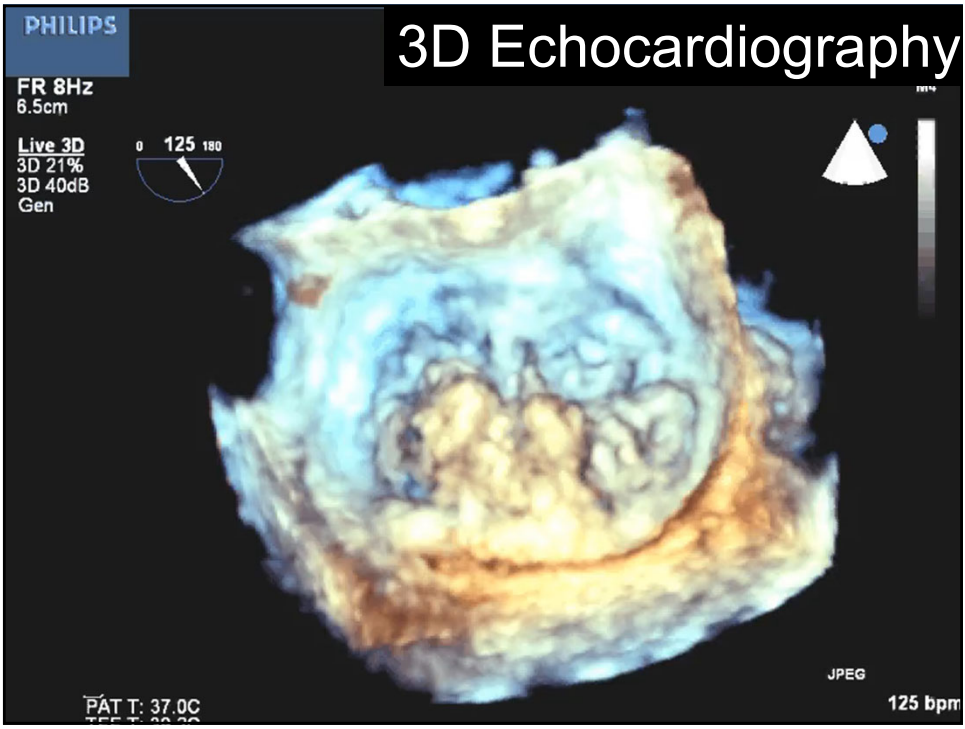
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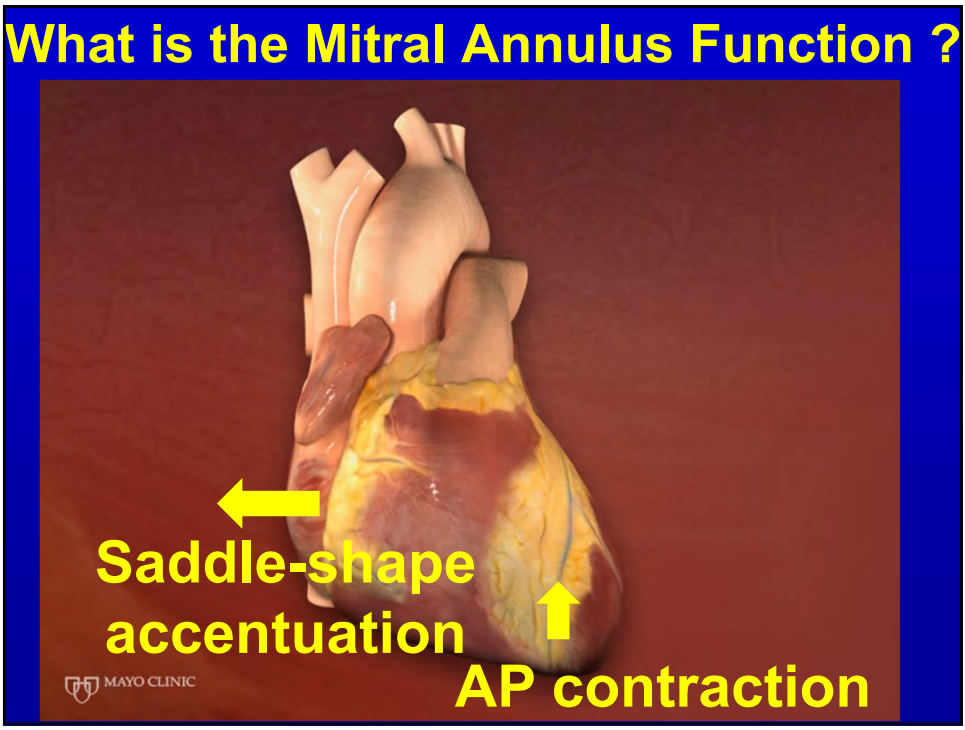
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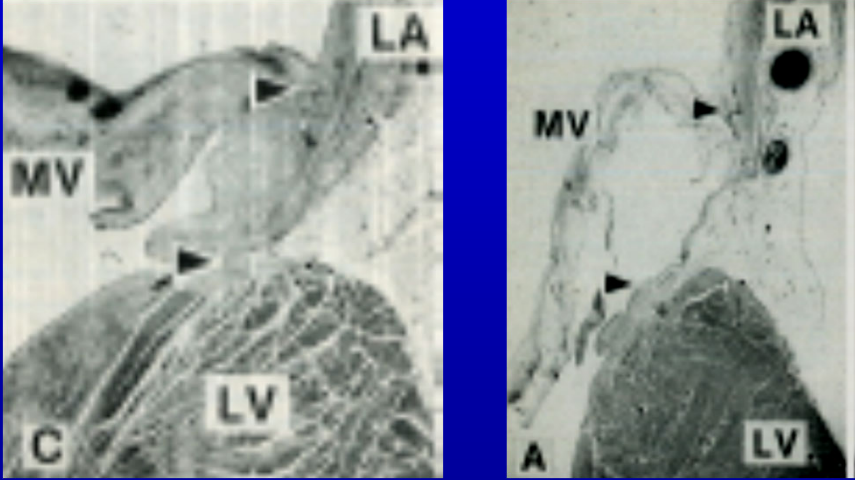


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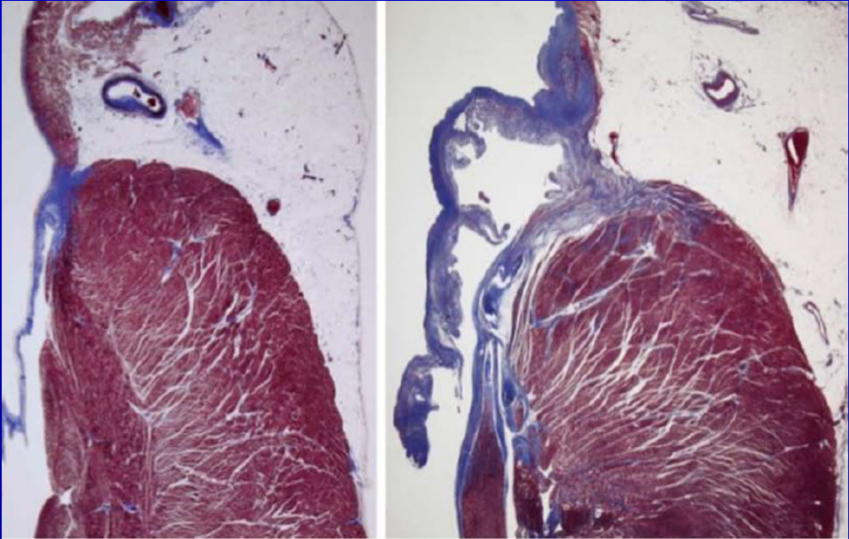
Mitral Annulus Disjunction



Hutchins GM, NEJMed 1986; 314: 535-40

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Mitral Annulus Disjunction

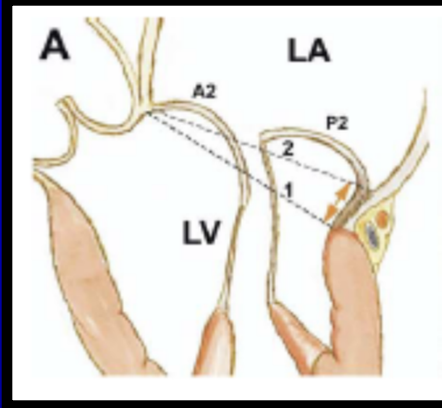
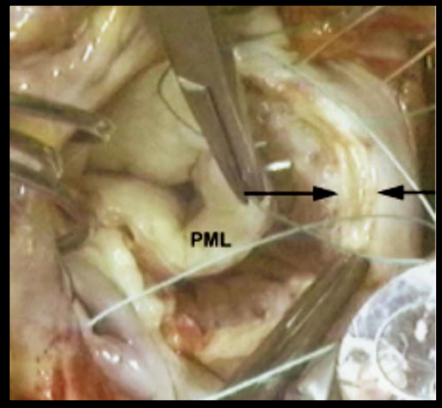


Perrazolo-Mara M, Circ Img 2016;9:e005030.

58

Mitral Annulus Disjunction

Surgical inspection



Eriksson MJ, JASE 2005; 18; 1014-22

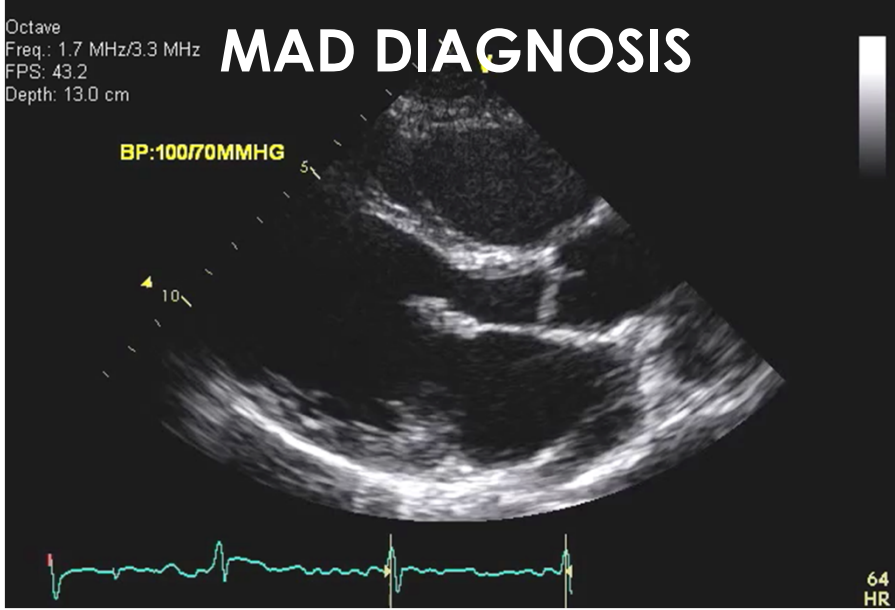
59

/ #2 MAD

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

MAD DIAGNOSIS

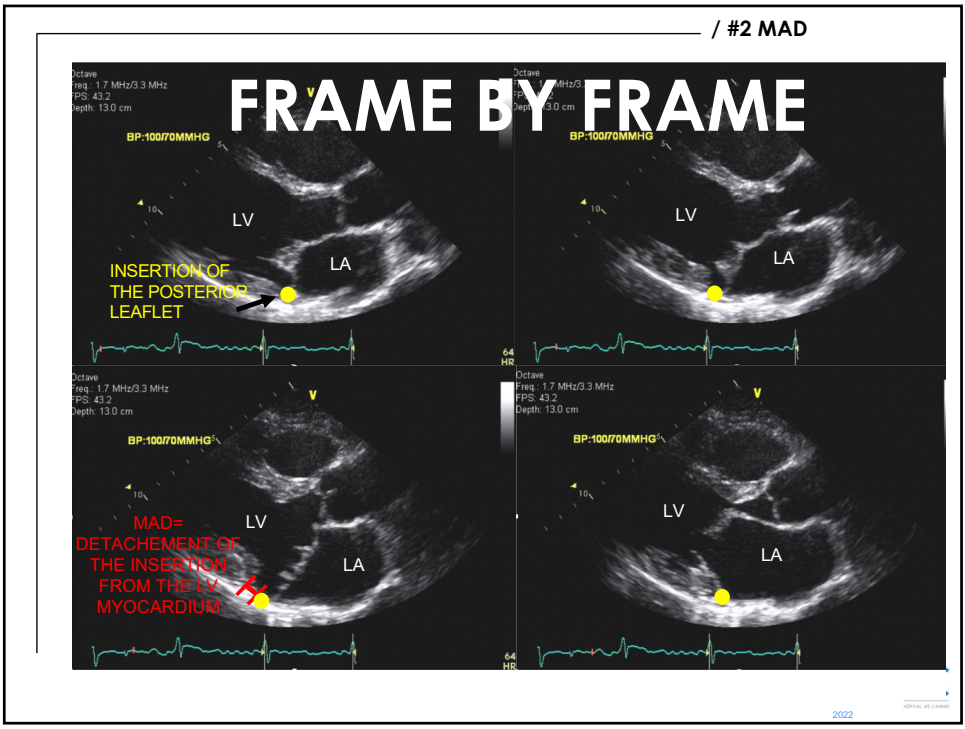
BP: 100/70MMHG



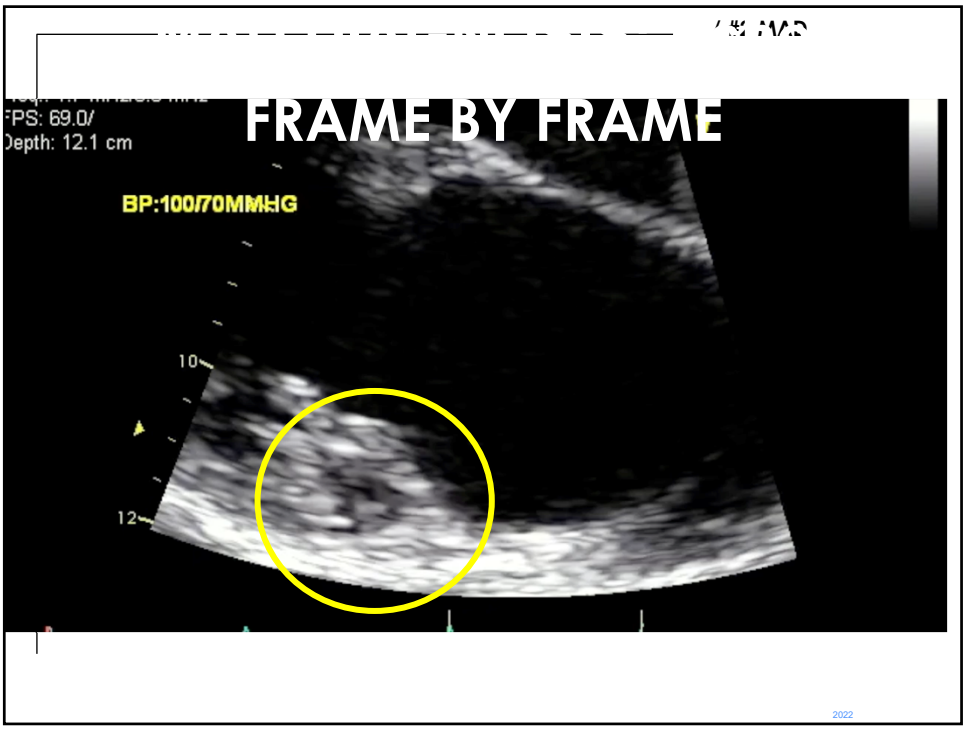
64
HR

2022

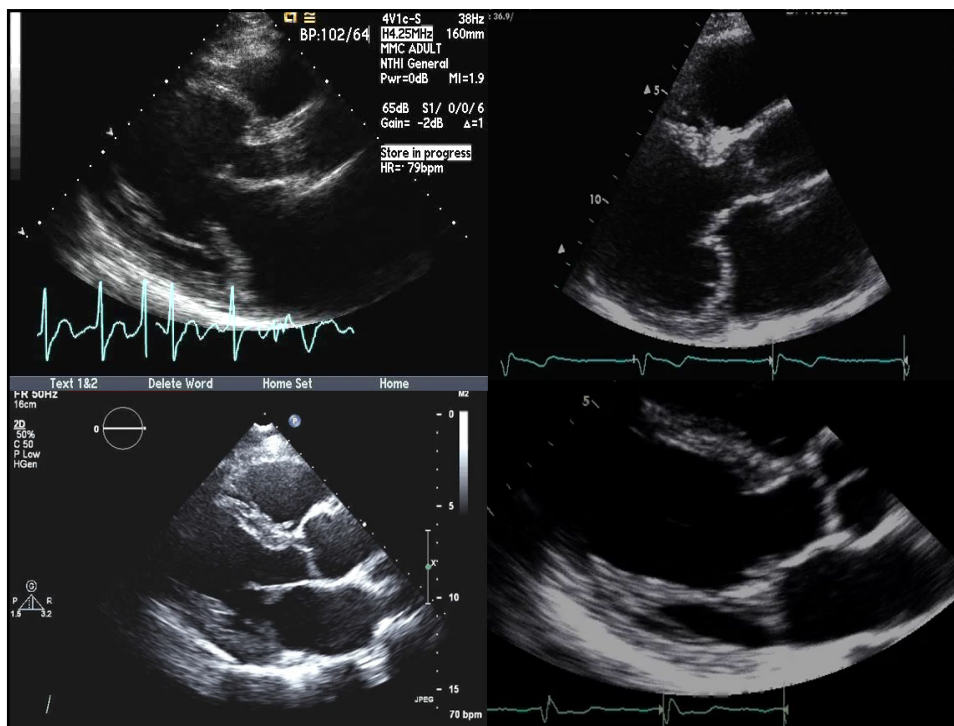
60



61



62



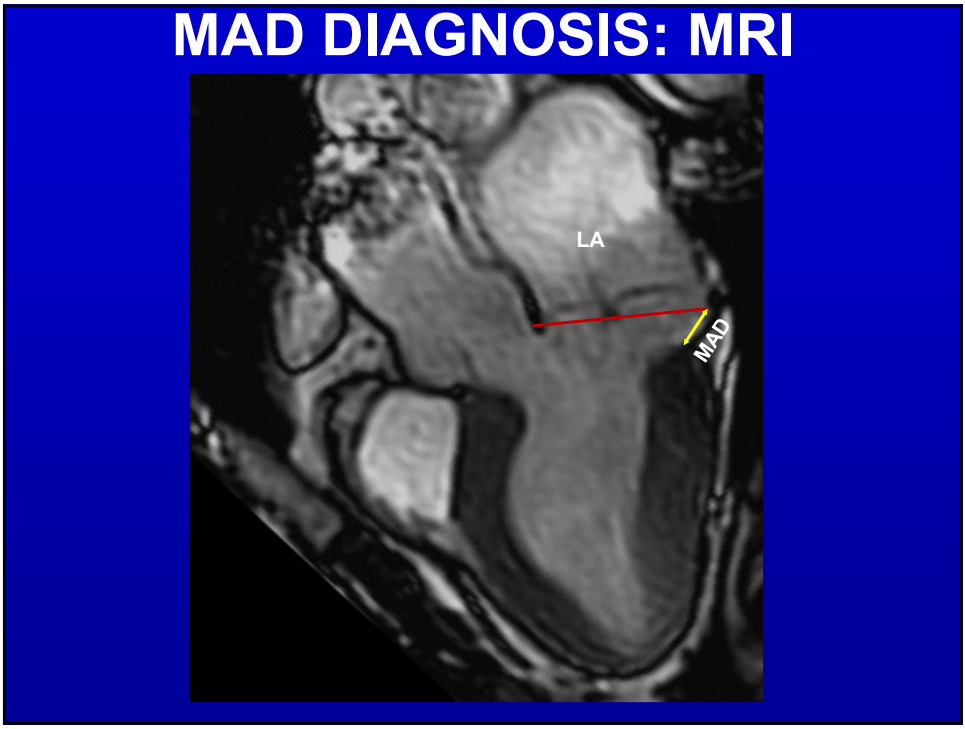
63

MAD diagnosis

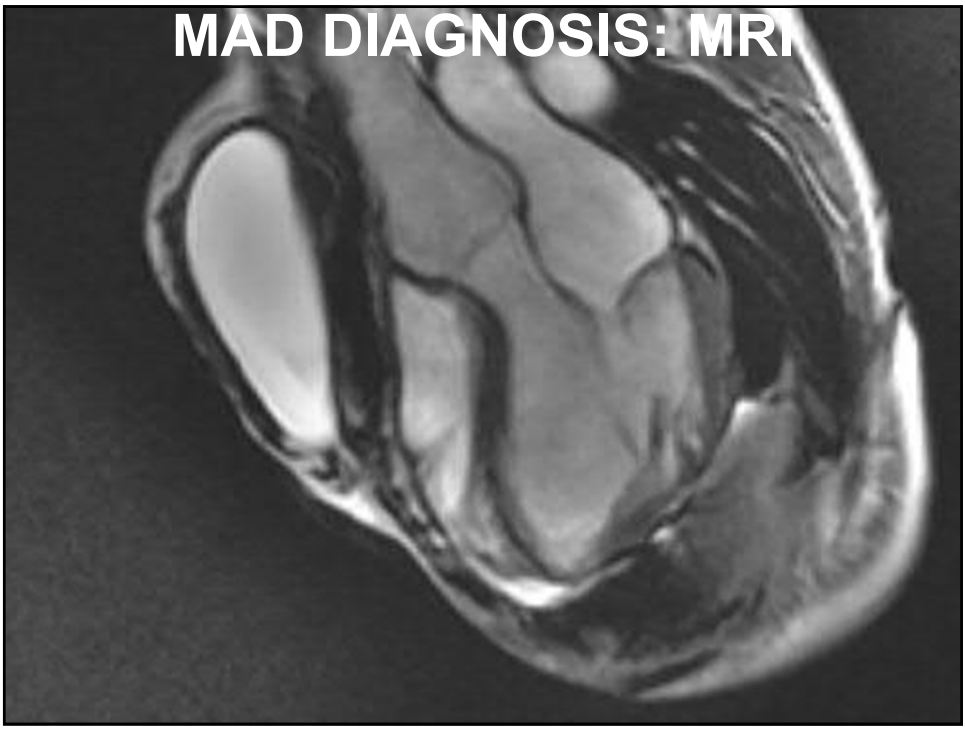
MAD diagnosis requires:

- **Recognition of the mitral annulus** position frame by frame in LAX views throughout systole, then
- **Detection of separation annulus-LV** myocardium at mid and late-systole
- **Determination of MVP depth** from the annulus position

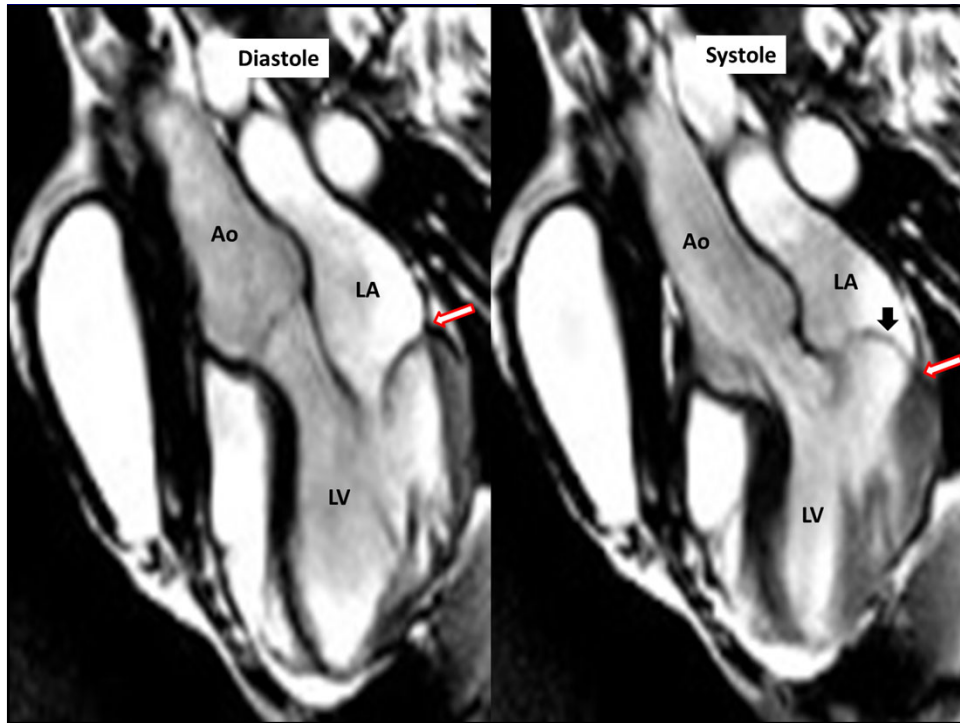
64



65



66



67

MAD diagnosis requires:

- **Recognition of the mitral annulus** position frame by frame in LAX views throughout systole, then
- **Detection of separation annulus-LV** myocardium at mid and late-systole
- **Determination of MVP depth** from the annulus position

Imaging must provide sufficient spatial and temporal resolution to quantify MAD and MVP

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Context & Physiologic Consequences of MAD ?

69

Mitral Annular Disjunction of Degenerative Mitral Regurgitation: Three-Dimensional Evaluation and Implications for Mitral Repair

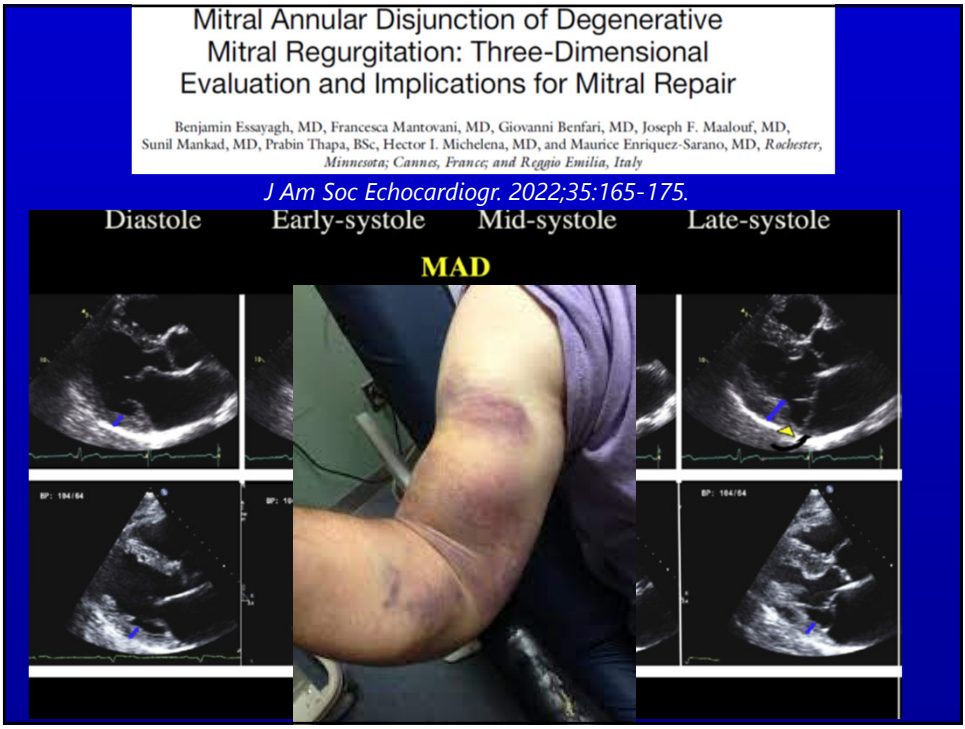
Benjamin Essayagh, MD, Francesca Mantovani, MD, Giovanni Benfari, MD, Joseph F. Maalouf, MD, Sunil Mankad, MD, Prabin Thapa, BSc, Hector I. Michelena, MD, and Maurice Enriquez-Sarano, MD, Rochester, Minnesota; Cannes, France; and Reggio Emilia, Italy

J Am Soc Echocardiogr. 2022;35:165-175.

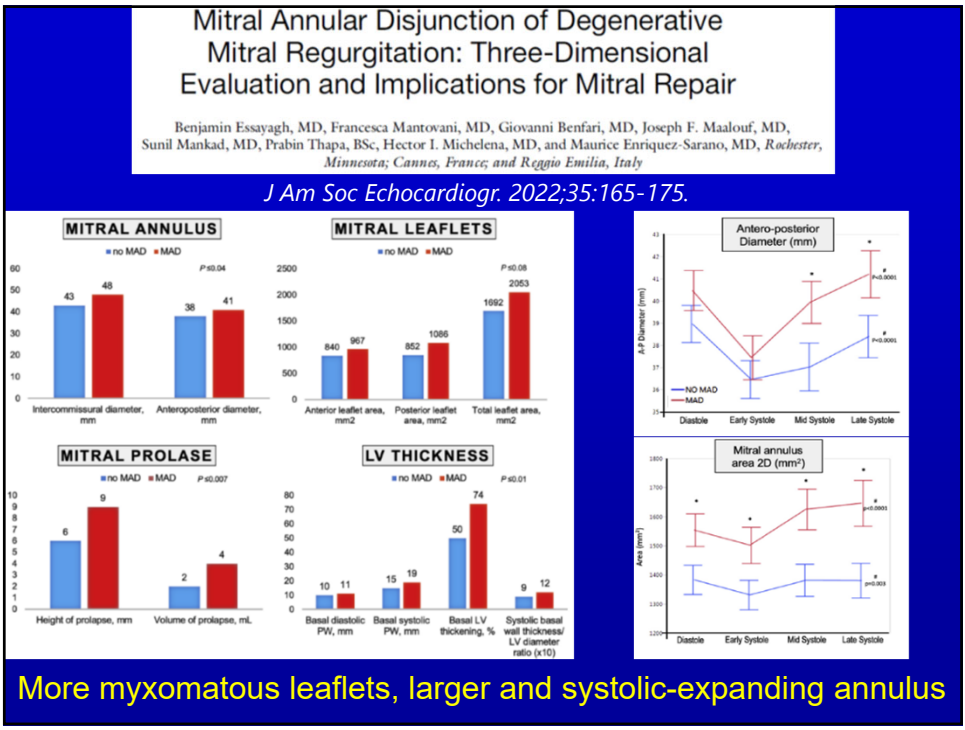
The image shows a 3D echocardiogram of the mitral annulus. A dashed white line outlines the annular area and circumference. A red dotted line indicates the disjunction arc. Two diameters are shown: the antero-posterior diameter (red line) and the intercommissural diameter (yellow line). Labels include A1, A2, A3, P1, P2, P3, LAA, AV, and TV. A legend at the bottom identifies the diameters, disjunction arc, and annular area/circumference. A small inset shows a cross-section of the annulus with 'A' and 'P' labels. A small ECG trace is visible at the bottom right.

- Antero-posterior diameter
- Intercommissural diameter
- Disjunction arc
- Annular area and circumference

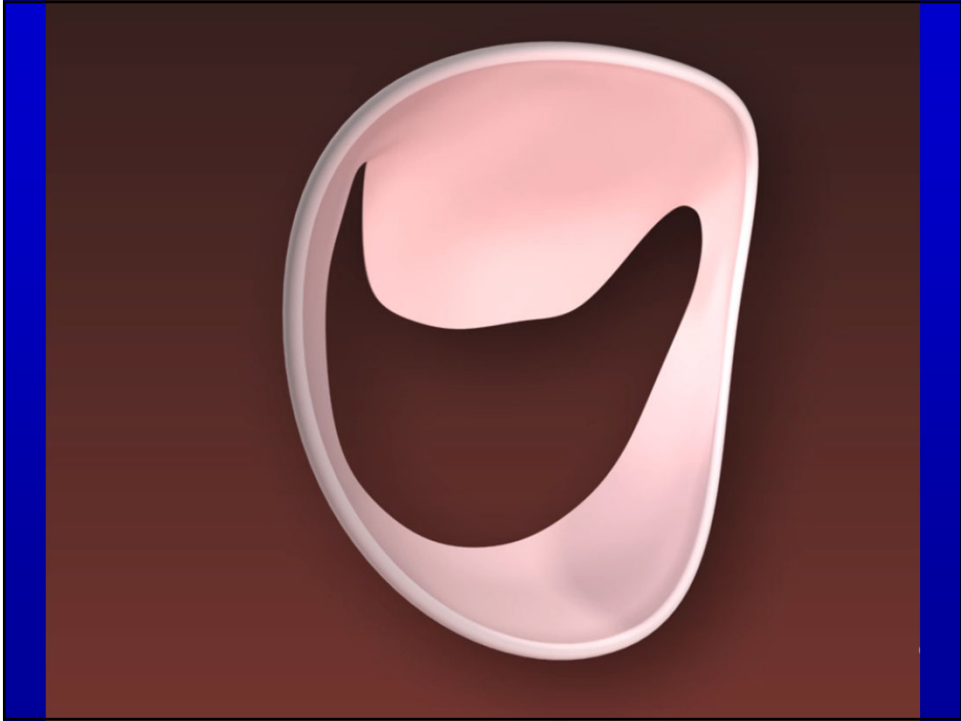
70



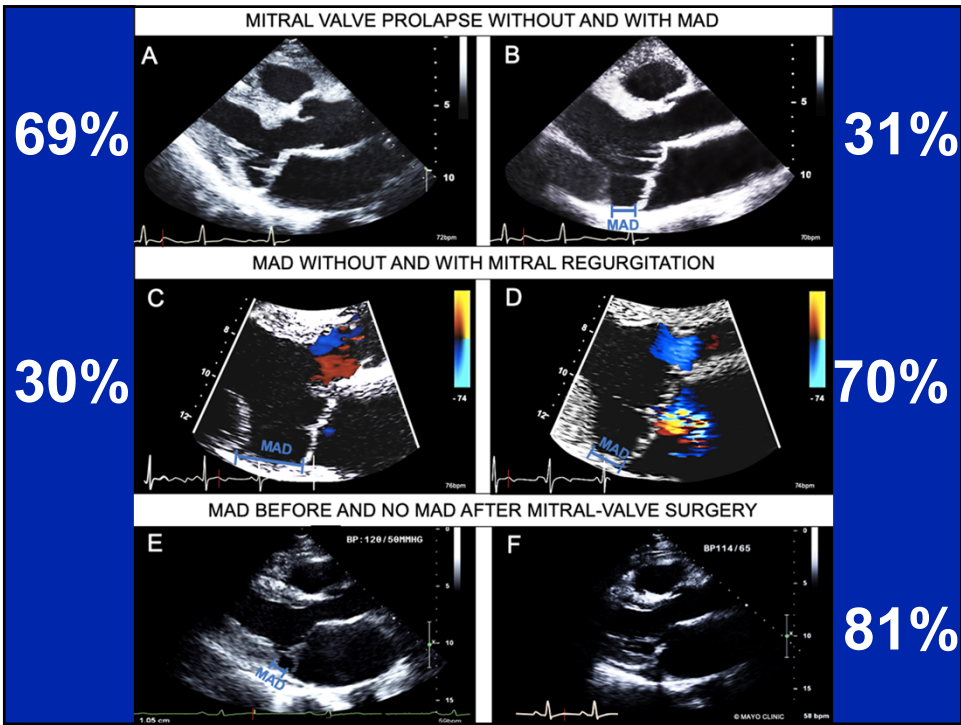
71



72



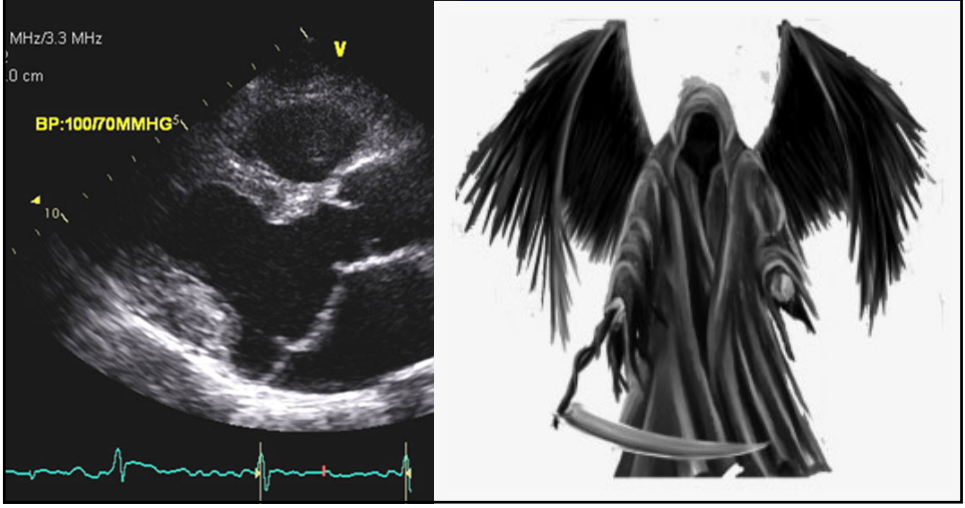
73



74

Mitral Valve Prolapse

is MAD.....the Angel of Death?



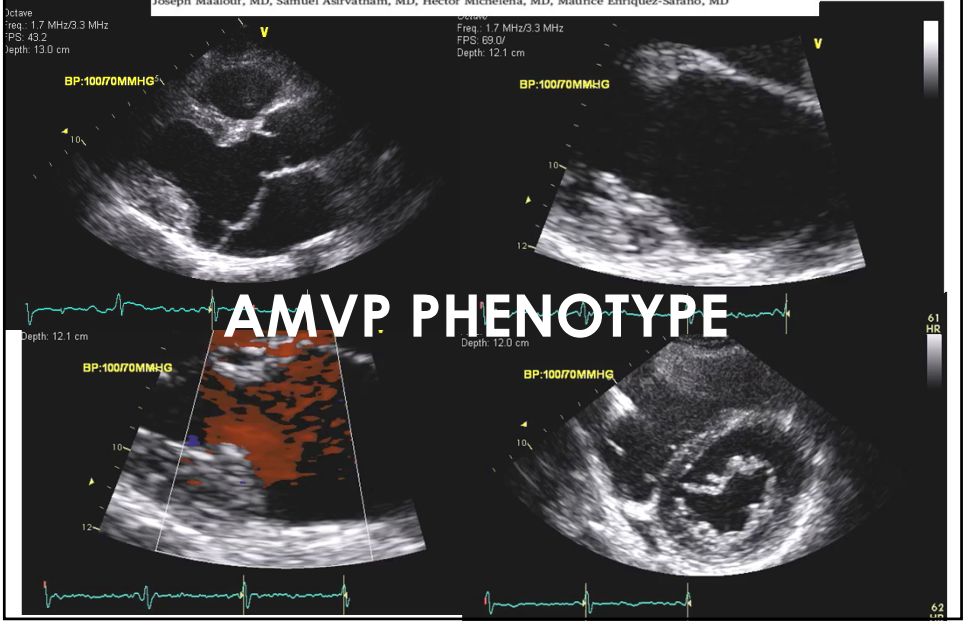
The image contains two side-by-side panels. The left panel is a parasternal short-axis echocardiogram of the mitral valve. It shows the leaflets prolapsing into the left atrium. Technical details include: MHz/3.3 MHz, 0 cm, BP:100/70MMHG, and a 10x magnification. A yellow 'V' is marked on the image. The right panel is a black and white illustration of the Grim Reaper, a figure with large, dark wings, wearing a hooded robe, and holding a scythe.

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Presentation and Outcome of Arrhythmic Mitral Valve Prolapse

Phenotype

Benjamin Essayagh, MD,* Avi Sabbag, MD,* Cl mence Antoine, MD, Giovanni Benfari, MD, Li-Tan Yang, MD, Joseph Maalouf, MD, Samuel Asirvatham, MD, Hector Michelena, MD, Maurice Enriquez-Sarano, MD



The image displays four echocardiographic views of the mitral valve. The top-left view is a parasternal short-axis view with technical details: Octave, 1.7 MHz/3.3 MHz, FPS: 43.2, Depth: 13.0 cm, BP:100/70MMHG, and a 10x magnification. The top-right view is a parasternal short-axis view with technical details: 1.7 MHz/3.3 MHz, FPS: 69.0, Depth: 12.1 cm, BP:100/70MMHG, and a 10x magnification. The bottom-left view is a parasternal short-axis view with technical details: Depth: 12.1 cm, BP:100/70MMHG, and a 10x magnification. The bottom-right view is a parasternal short-axis view with technical details: Depth: 12.0 cm, BP:100/70MMHG, and a 10x magnification. A central text overlay reads "AMVP PHENOTYPE". A heart rate of 61 HR is shown on the right side.

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Phenotypic risk features:
 TWI in the inferior leads, multiple polymorphic PVCs, MAD, redundant MV leaflets, enlarged LA, LV-EF ≤ 50%, LGE.

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Circulation

ORIGINAL RESEARCH ARTICLE

Replacement Myocardial Fibrosis in Patients With Mitral Valve Prolapse
 Relation to Mitral Regurgitation, Ventricular Remodeling, and Arrhythmia

BACKGROUND: Mitral valve prolapse (MVP) is a frequent disease that can be complicated by mitral regurgitation (MR), heart failure, arterial embolism, rhythm disorders, and death. Left ventricular (LV) replacement myocardial fibrosis, a marker of maladaptive remodeling, has been described in patients with MVP, but the implications of this finding remain scarcely explored. We aimed at assessing the prevalence, pathophysiological and prognostic significance of LV replacement myocardial fibrosis through late gadolinium enhancement (LGE) by cardiac magnetic resonance in patients with MVP.

METHODS: Four hundred patients (53±15 years of age, 55% male) with MVP (trace to severe MR by echocardiography) from 2 centers, who

Authors: Anne-Laure Constant Dit Beaufils, MD*, Olivier Huttin, MD*, Jean-Michel Serfaty, MD, PhD†, Christine Selton-Suty, MD†, Thierry Le Tourneau, MD, PhD†

Echocardiographic characteristics	Overall n=400	LGE+ n=110	LGE- n=290	P value
Mitral regurgitation grade, n (%)				<0.0001
Trace-mild	120 (30)	16 (15)	104 (36)	
Moderate	110 (28)	31 (28)	79 (27)	
Severe	170 (43)	63 (57)	107 (37)	
Effective regurgitant orifice area, mm ²	32±20	37±20	30±20	0.005
Regurgitant volume, mL	48±31	56±30	45±31	0.002
Anterior leaflet length, mm	26.6±3.9	27.3±3.7	26.3±4.0	0.04
Posterior leaflet length, mm	18.6±4.0	19.4±4.0	18.2±4.0	0.03
Mitral annulus, mm	38.5±5.6	40.3±5.1	37.7±5.6	<0.0001
Mitral annulus disjunction, n (%)	206 (52)	61 (55)	145 (50)	0.39
Flail leaflet, n (%)	138 (35)	49 (45)	89 (31)	0.02
Bileaflet mitral valve prolapse, n (%)	201 (50)	56 (51)	145 (50)	0.96

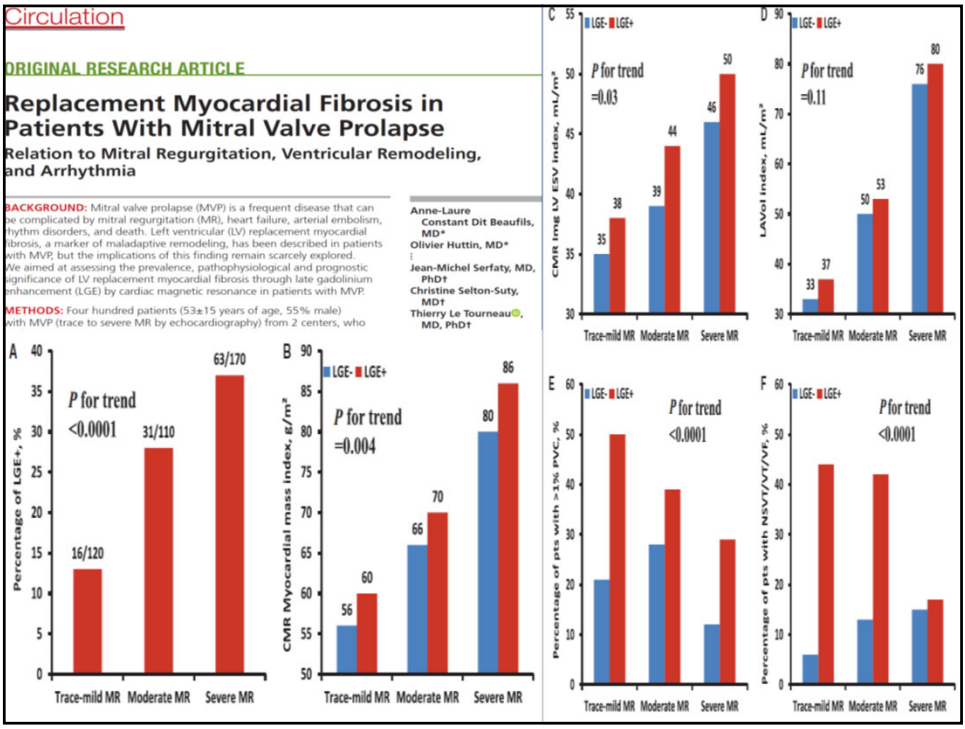
Baseline cardiac MRI	All n=400	LGE+ n=110	LGE- n=290	P value
LV end-diastolic volume index, mL/m ²	106±29	115±30	103±27	0.0001
LV end-systolic volume index, mL/m ²	42±15	46±17	40±13	<0.0001
LV ejection fraction, %	61±7	61±7	61±7	0.24
Myocardial mass index, g/m ²	67±20	74±28	65±17	0.002
RV end-diastolic volume index, mL/m ²	82±23	84±22	81±23	0.14
RV end-systolic volume index, mL/m ²	39±15	42±15	38±15	0.004
RV ejection fraction, %	53±8	51±8	54±8	<0.0001
Regurgitant volume, mL	41±29	50±29	38±28	0.0004
Regurgitant fraction, %	33±17	38±16	31±17	0.0007

LGE+ 28%

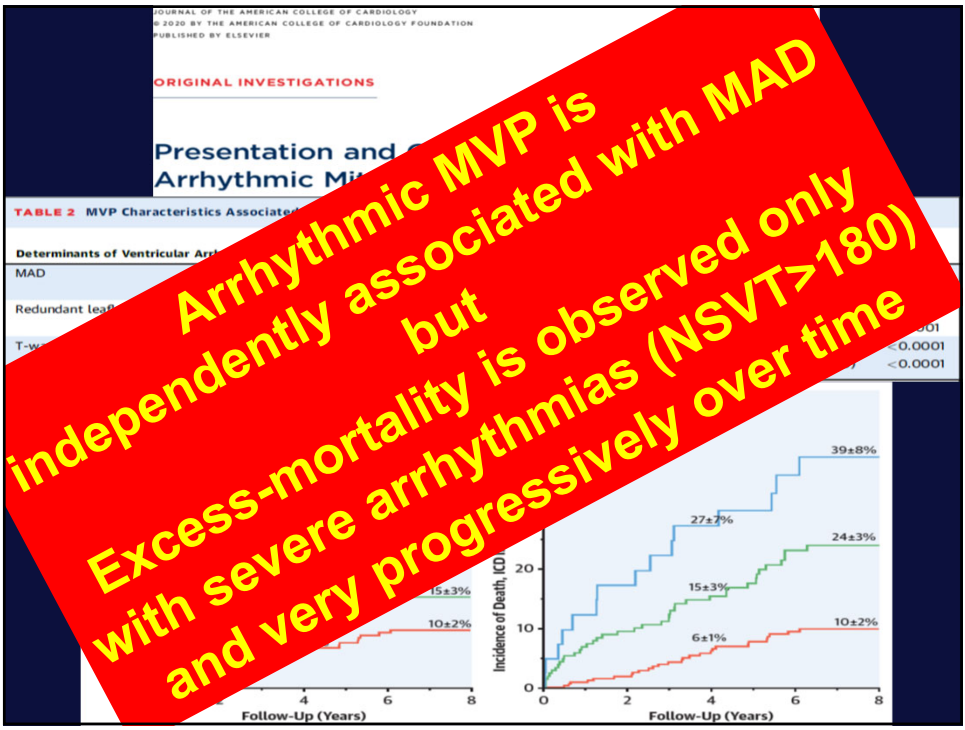
Localization of LGE+, %

Localization	n/N
Basal IL wall	71/110
Other wall	28/110
PM	29/110
Hinge Point	9/110
Base of PM	6/110

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79



80

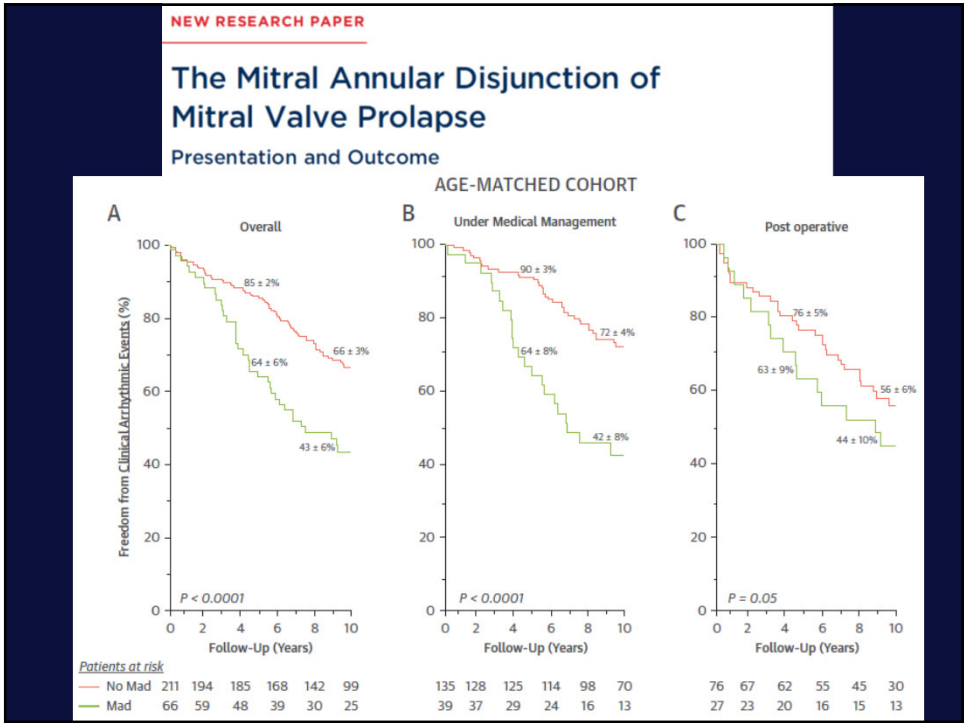
NEW RESEARCH PAPER

The Mitral Annular Disjunction of Mitral Valve Prolapse

Presentation and Outcome

Benjamin Essayagh, MD,^{a,b} Avi Sabbag, MD,^{a,c,d} Clémence Antoine, MD,^a Giovanni Benfari, MD,^{a,e}
 Roberta Batista, MD,^a Li-Tan Yang, MD,^a Joseph Maalouf, MD,^a Prabin Thapa, MSc,^a Samuel Asirvatham, MD,^a
 Hector I. Michelena, MD,^a Maurice Enriquez-Sarano, MD^a

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NEW RESEARCH PAPER

The Mitral Annular Disjunction and Mitral Valve Prolapse: Presentation and Outcome

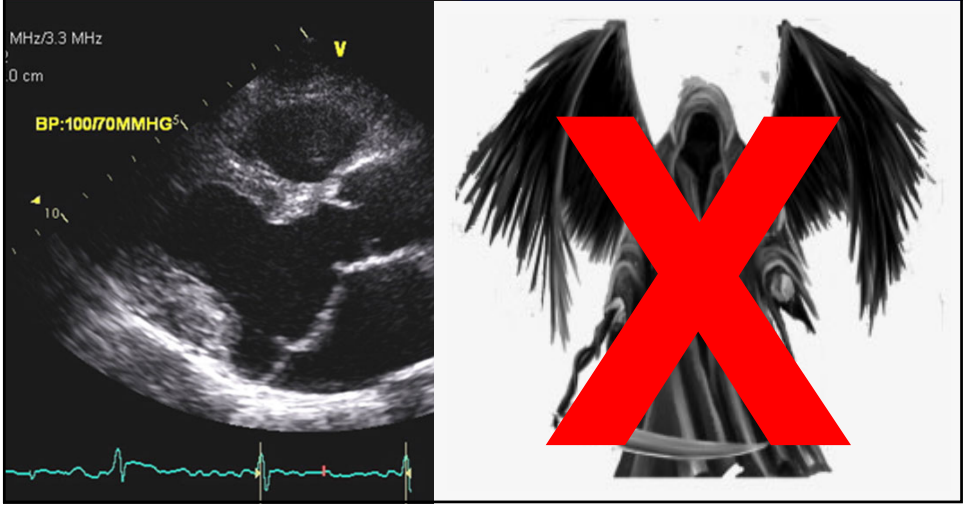
100

**Mitral Annular Disjunction is:
A manifestation of severely
myxomatous MVP,
An independent determinant of
AMVP, but
Survival is maintained within the
first 10-year FU**

Follow-Up (Years)	6	8	10	
185	184	180	145	87
179	179	176	147	90

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Mitral Valve Prolapse
MAD is not the Angel of Death
but advocates for AMVP monitoring



MHz: 3.3 MHz
0 cm
BP: 100/70 mmHg
10

The image shows an echocardiogram on the left and a black and white illustration of a grimacing figure with wings on the right. A large red 'X' is superimposed over the figure, indicating that the 'Angel of Death' metaphor is incorrect.

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Sudden death-DMR MVP

SD is an important issue in myxomatous diseases

DMR: Notable SD rate. Approach to prevent SD is **early repair**

Isolated MVP: SD is infrequent but risk is recognized by the **triad**
Syncope/MAD/High-risk VT

85

ESC European Society of Cardiology | European Heart Journal (2023) 44, 3121–3135 | <https://doi.org/10.1093/eurheartj/ehad491> | STATE OF THE ART REVIEW | Valvular heart disease

Arrhythmic mitral valve prolapse and mitral annular disjunction: pathophysiology, risk stratification, and management

Benjamin Essayagh^{1,2}, Avi Sabbag³, Edward El-Am¹, João L. Cavalcante⁴, Hector I Michelena¹, and Maurice Enriquez-Sarano^{4*}

Arrhythmias for treatment: No/trivial, Mild, Moderate, Severe.

Phenotype for monitoring: MAD, Severe myxomatous disease, Mitral regurgitation, Fibrosis.

Phenotypic risk features: TWI in the inferior leads, multiple polymorphic PVCs, MAD, redundant MV leaflets, enlarged LA, LV-EFs 50%, LGE.

Decision: Episodic monitoring, Frequent monitoring, Mitral repair ± ICD, ICD, Ablation/AAD.

Risk stratification: Indicated by a red arrow at the bottom.

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AMVP is a real entity

Detect the phenotype with highest propensity for development of Ventricular Arrhythmias

Detect the Arrhythmias by monitoring, if necessary prolonged, if necessary repeated

Treat Arrhythmias based on their severity and association to DMR

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89



90



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