

## Mitral Valve Disease 2020: *A Couple of Quandries*

James D. Thomas, MD, FACC, FASE  
Director, Center for Heart Valve Disease  
Bluhm Cardiovascular Institute  
Professor of Medicine, Feinberg School  
of Medicine, Northwestern University  
Chicago, Illinois

Conflicts of interest: GE, Abbott, Edwards, Caption Health (honoraria)  
Spouse employment: Caption Health

1

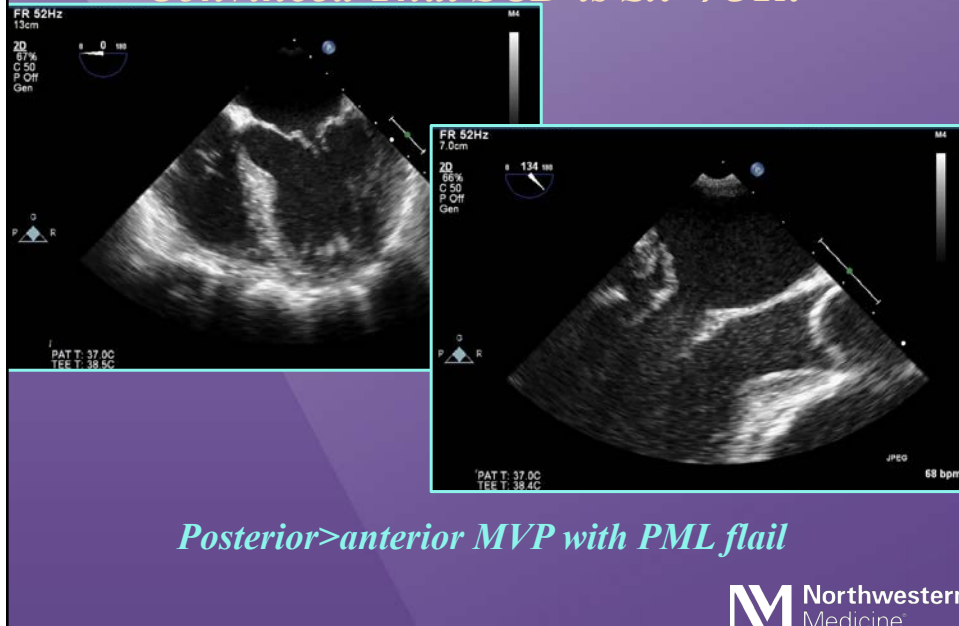
## 64yo Man w/ Bi-MVP and Severe Asx MR *Avid Swimmer (10 Mile Races in the Lake)*



- *Flail PML, ROA 0.5 cm<sup>2</sup>; LV/LA enlargement; surgery recommended (IIa), but patient was reluctant.*
- *While swimming in a pool, suffered VF arrest, fortunately in front of an anesthesiologist.*
- *Therapeutic hypothermia with full recovery; ICD placed.*

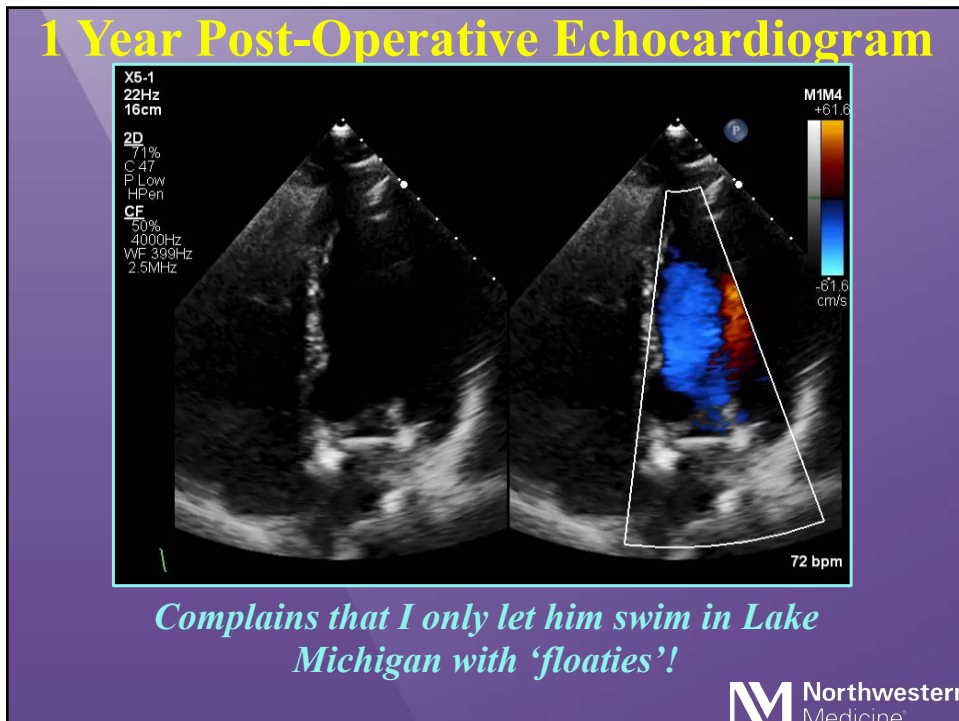
3

**Still Reluctant (“I Have No Symptoms”)  
Convinced That SCD is Sx ⇒ OR!**



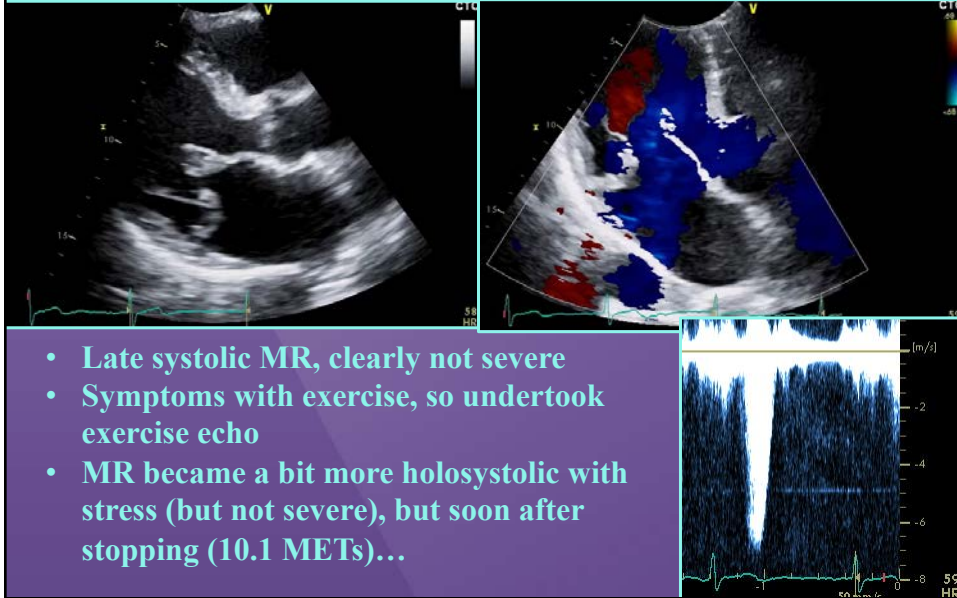
4

**1 Year Post-Operative Echocardiogram**



5

## 58yo Man with MVP and no Fxn Limitations *Palpitations and Lightheaded w/ Exercise*



6

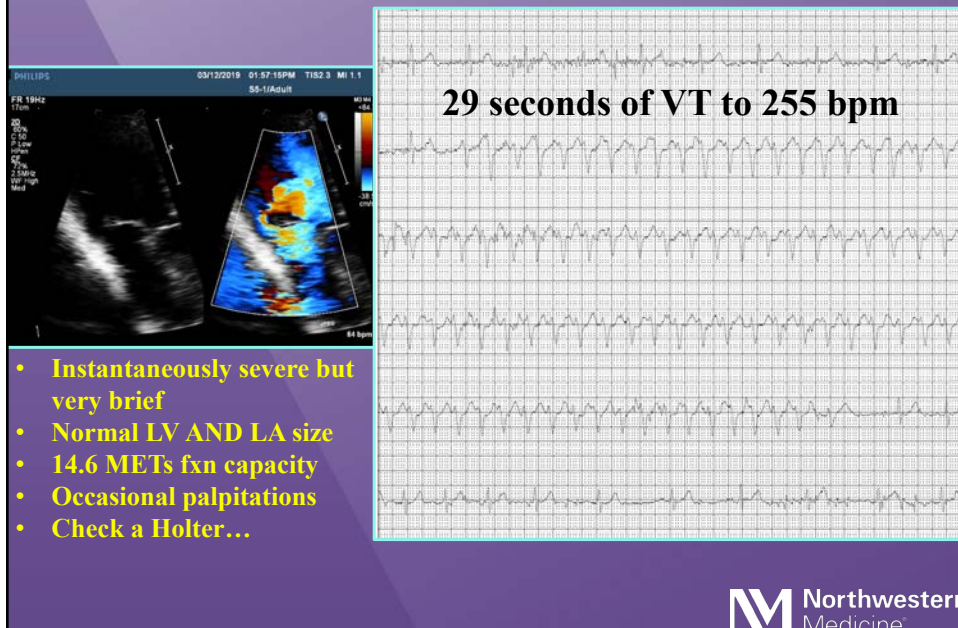
## Runs of Multifocal Ventricular Tachycardia



Admitted for monitoring, underwent MV repair

7

## 60yo Active Man with Murmur



The image displays two medical diagnostic tools. On the left is an echocardiogram showing a cross-section of the heart with a color Doppler overlay indicating blood flow. On the right is an ECG tracing showing a rapid, irregular rhythm. The text '29 seconds of VT to 255 bpm' is overlaid on the ECG. Below the echocardiogram is a list of clinical findings.

- Instantaneously severe but very brief
- Normal LV AND LA size
- 14.6 METs fxn capacity
- Occasional palpitations
- Check a Holter...

PHILIPS 03/12/2019 01:57:16PM T182.3 MI 1.1  
55-1/Adult  
FR 18Hz  
2D  
30cm  
2.5D  
3.0cm  
4.0cm  
5.0cm  
6.0cm  
7.0cm  
8.0cm  
9.0cm  
10.0cm  
11.0cm  
12.0cm  
13.0cm  
14.0cm  
15.0cm  
16.0cm  
17.0cm  
18.0cm  
19.0cm  
20.0cm  
21.0cm  
22.0cm  
23.0cm  
24.0cm  
25.0cm  
26.0cm  
27.0cm  
28.0cm  
29.0cm  
30.0cm  
31.0cm  
32.0cm  
33.0cm  
34.0cm  
35.0cm  
36.0cm  
37.0cm  
38.0cm  
39.0cm  
40.0cm  
41.0cm  
42.0cm  
43.0cm  
44.0cm  
45.0cm  
46.0cm  
47.0cm  
48.0cm  
49.0cm  
50.0cm  
51.0cm  
52.0cm  
53.0cm  
54.0cm  
55.0cm  
56.0cm  
57.0cm  
58.0cm  
59.0cm  
60.0cm  
61.0cm  
62.0cm  
63.0cm  
64.0cm  
65.0cm  
66.0cm  
67.0cm  
68.0cm  
69.0cm  
70.0cm  
71.0cm  
72.0cm  
73.0cm  
74.0cm  
75.0cm  
76.0cm  
77.0cm  
78.0cm  
79.0cm  
80.0cm  
81.0cm  
82.0cm  
83.0cm  
84.0cm  
85.0cm  
86.0cm  
87.0cm  
88.0cm  
89.0cm  
90.0cm  
91.0cm  
92.0cm  
93.0cm  
94.0cm  
95.0cm  
96.0cm  
97.0cm  
98.0cm  
99.0cm  
100.0cm

Northwestern  
Medicine

8

## Mitral Valve Disease and Arrhythmias *Questions Remaining*

- *What is the relationship between MVP and VEA?*
- *Does it change your threshold to intervene?*
- *Does fixing the MR lower the VT/VF risk?*
- *Are you doing more CMR to look for fibrosis?*

9



## Sudden Cardiac Death in MVP A Checkered History

- Association first reported decades ago, based on small retrospective studies
- Estimation of annual SCD risk among MVP is unclear – range: 0.2-2% per year
- Framingham study (Freed et al, NEJM 1999) found no excess arrhythmias in MVP
- Further investigation is needed

**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)
<b>Syncope</b>	<b>3 (3.6)</b>	<b>103 (3.0)</b>

\*Cerebrovascular disease refers to stroke or transient ischemic attack.

Freed et al. NEJM 1999; 341: 1-7

10

## Risk Factors for SCD in MVP



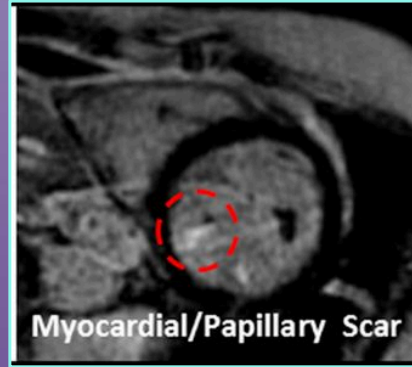
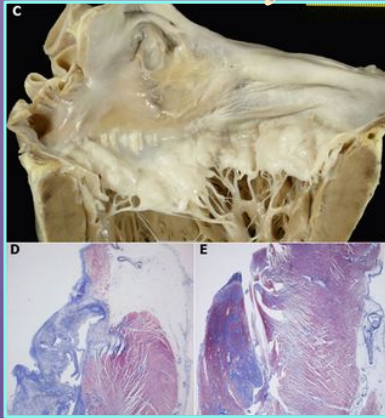
- All SCDs in Veneto Region in Italy autopsied over 21 years
- MVP was 3<sup>rd</sup> most frequent cause of SCD (after ARVD and CAD) in 12% of cases
- Most striking association was papillary muscle fibrosis

MVP leaflet involvement	
Posterior, n (%)	13 (30)
Bileaflet, n (%)	30 (70)
Endocardial fibrous plaque, n (%)	25 (58)
Histology features, n (%)	
LV scar	
PM, n (%)	43 (100)
Inferobasal wall	38 (88)
Fibrous tissue /myocardium, % area	
PM, mean±SD	30.5±10.7
Inferobasal wall, mean±SD	33.1±7.6
Cardiomyocytes diameter, mean±SD, μm	19.2±6.0

Basso et al. Circulation 2015; 132: 556–566

11

## Risk Factors for SCD Myocardial Fibrosis



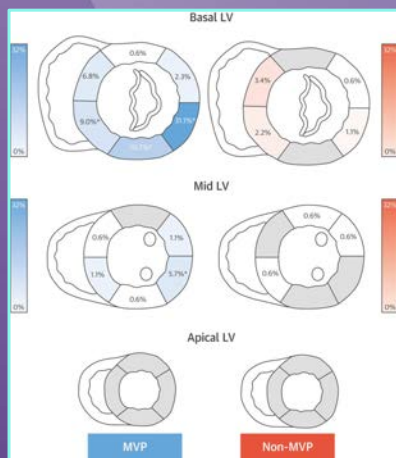
- Compared with MVP without arrhythmia, MVP patients with ventricular arrhythmia had more myocardial fibrosis by cMRI (93% vs. 14%,  $P < 0.01$ ).
- Fibrosis distribution: 83% in pap muscle, 73% in inferobasal wall

Basso et al. *Circulation* 2015; 132: 556–566

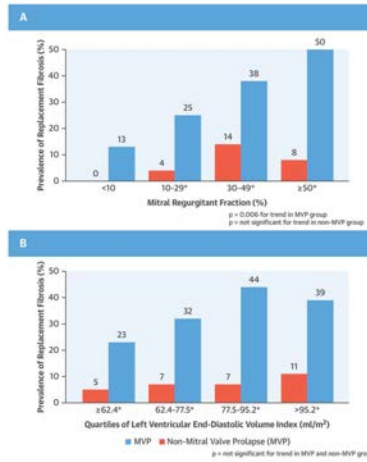


12

## Is Myocardial Fibrosis Specific to MVP Related MR?



CENTRAL ILLUSTRATION: Prevalence of Replacement Fibrosis in MVP and Non-MVP Patients



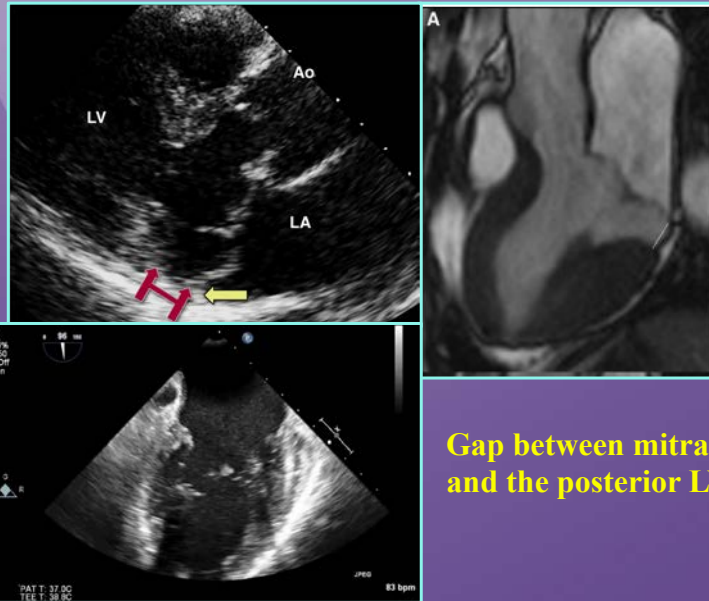
Kitkungvan, D. et al. *J Am Coll Cardiol*. 2018;72(8):823-34.

Kitkungvan D. *J Am Coll Cardiol* 2018; 72: 823-834



13

## Mitral Annular Disjunction



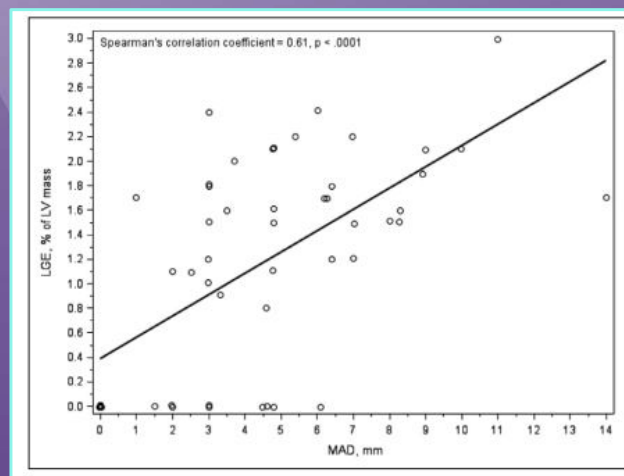
Gap between mitral PML and the posterior LV wall

Marra et al. *Circ Cardiovasc Imaging* 2016.  
Enriquez-Sarano. *JACC CV Imaging* 2017.



14

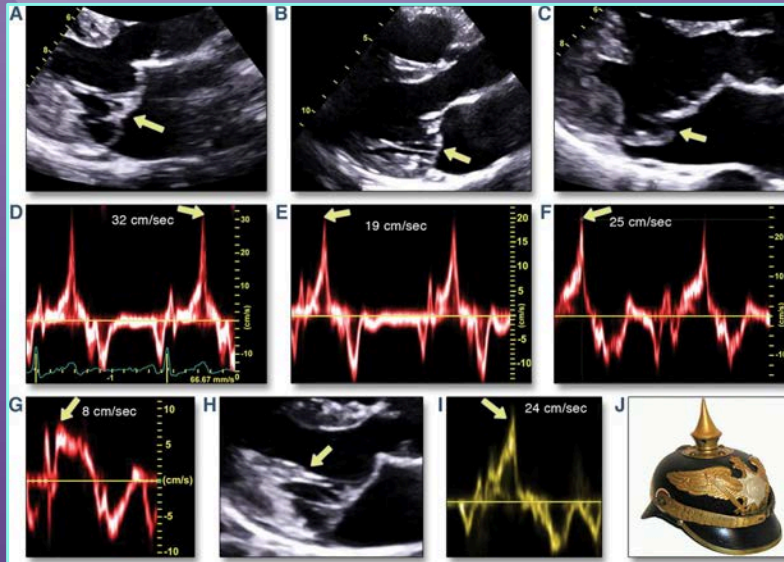
## Annular Disjunction and LV Fibrosis



Perazzolo Marra et al. *Circ Cardiovasc Imaging* 2016 Aug; 9(8): e005030

15

## Specific Sign of Mitral Annular Disjunction *The Pickelhaube Sign!*



Muthukumar et al. JACC Imaging 2017; 10: 1078-80

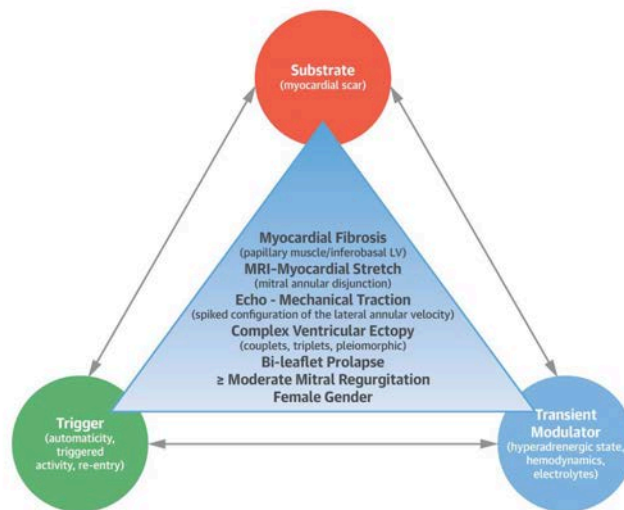
16 Northwestern  
Medicine

16

## Arrhythmic Mitral Valve Prolapse

JACC Review Topic of the Week

**CENTRAL ILLUSTRATION: Mitral Valve Prolapse-Related Sudden Cardiac Death: Interactions and Risk Factors**



Miller, M.A. et al. J Am Coll Cardiol. 2018;72(23PA):2904-14.

western  
e

17



*Many thanks!*

