

Prolasso mitralico aritmico

Il ruolo dell'imaging ecocardiografico

***Pellegrino Ciampi, MD
Policlinico Casilino
Rome, Italy***

A Historical Perspective

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

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

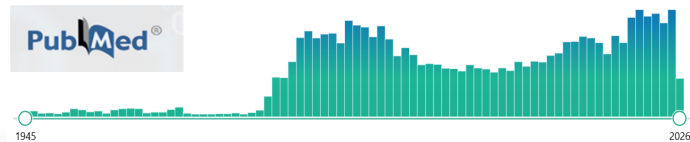
An Analysis of 90 Patients

J. B. BARLOW, C. K. BOSMAN, W. A. POCKOCK, AND P. MARCHAND

From the C.S.I.R. Cardio-Pulmonary Research Unit and the Cardiovascular Research Unit, Departments of Medicine and Thoracic Surgery, University of the Witwatersrand; and the Cardiac Clinic, General Hospital, Johannesburg, South Africa

In his early description of the condition, Dr. Barlow stated:

"The prognosis of this syndrome is uncertain, and sudden death may occur"



ESC

European Society of Cardiology

Europace (2022) **24**, 1981–2003
<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 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

Avi Sabbag (Chair) ^{1*}, Benjamin Essayagh ^{2,3}, Juan David Ramírez Barrera⁴, Cristina Basso ⁵, Ana Berni ⁶, Bernard Cosyns ⁷, Jean-Claude Deharo⁸, Thomas Deneke ⁹, Luigi Di Biase¹⁰, Maurice Enriquez-Sarano ¹¹, Erwan Donal ¹², Katsuhiko Imai ¹³, Han S. Lim¹⁴, Nina Ajmone Marsan ¹⁵, Mohit K. Turagam¹⁶, Petr Peichl ¹⁷, Sunny S. Po¹⁸, and Kristina Hermann Haugaa ¹⁹

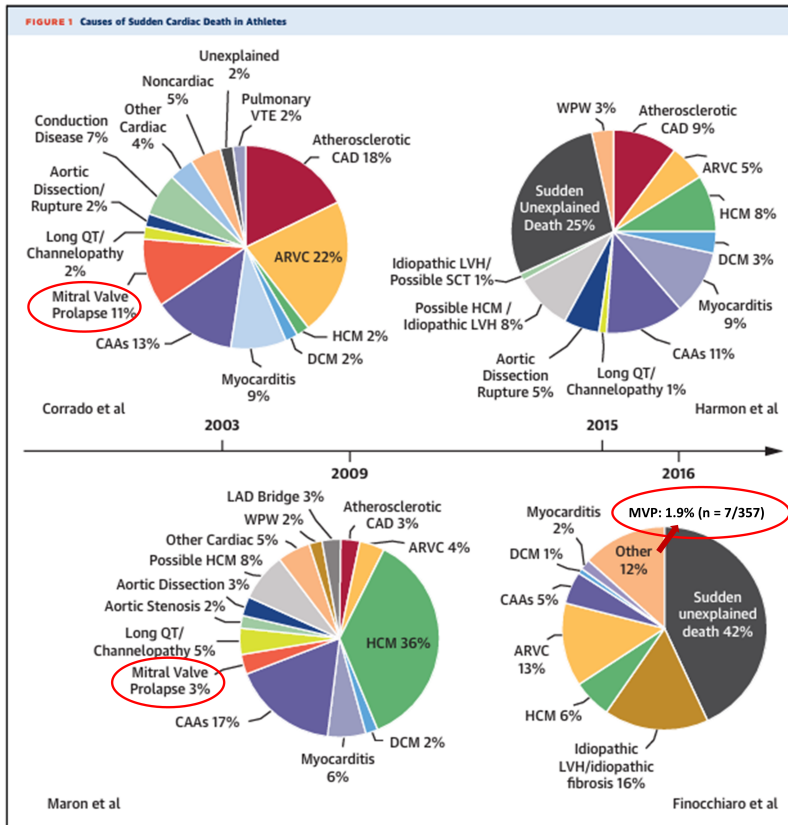
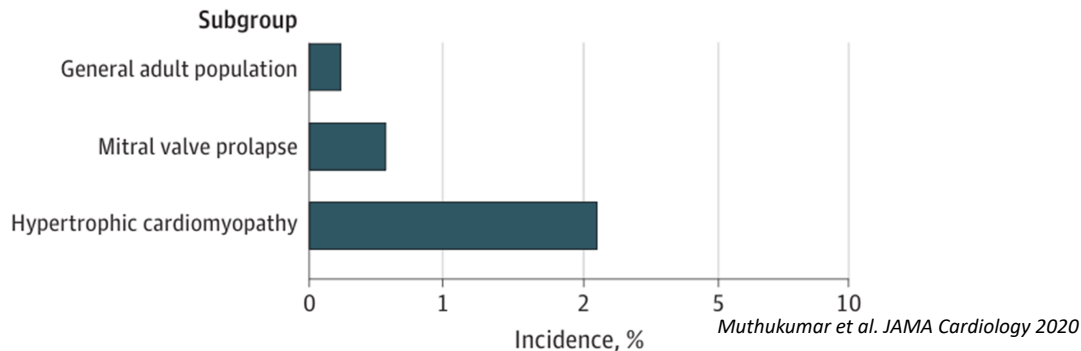
Epidemiology

Estimated prevalence in the general population **2-3%**

MVP-Related SCD:

- **3x** risk of general population
- Estimated risk is **0.2% to 1.8%**
- Implicated in **1.9%** of SCD in a recent meta-analysis (*Nalliah CJ et al. Heart 2019*)

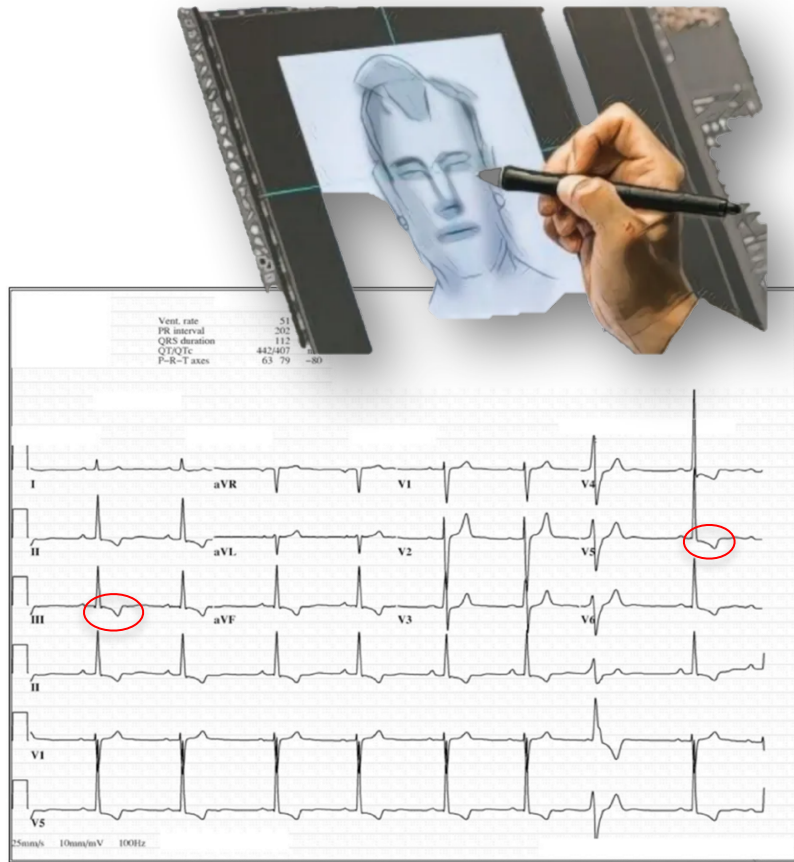
A Annual incidence of sudden cardiac death



aMVP: Can we draw a patient identikit?

Demographic, clinical and electrocardiographic risk factors

- Female prevalence (young age – 30 years)
- Chest pain, palpitations, dyspnoea on exercise comparable incidence in pts with and without MVP (Framingham study) – unexplained syncope may have high discriminative value
- T wave inversion or biphasic T wave in inferior and lateral leads
- QT prolongation
- Fragmented QRS



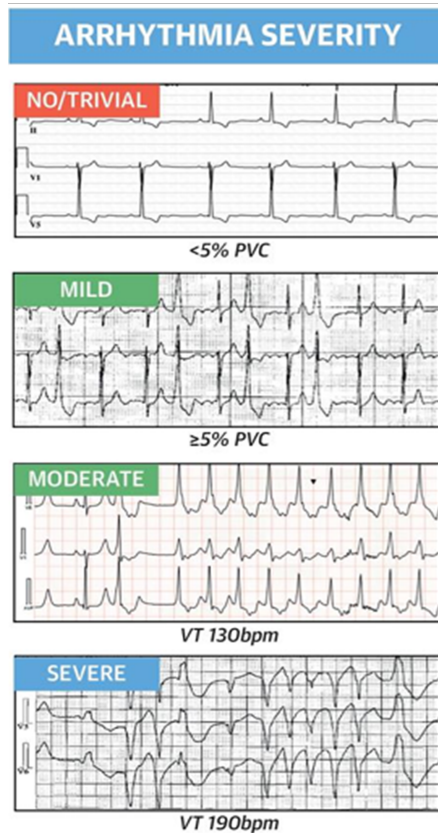
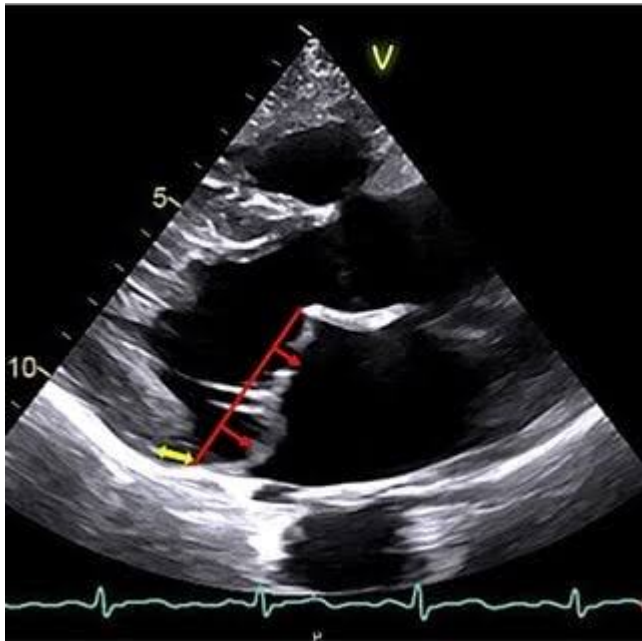
Arrhythmic mitral valve prolapse EHRA expert consensus statement definition

The presence of **MVP** (with or without MAD, severe degenerative mitral valve regurgitation or severe myxomatous MVP independent of MR severity)

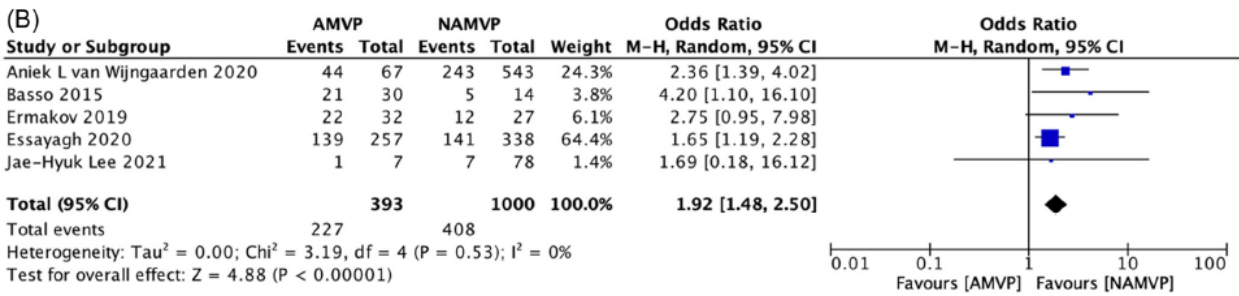
The presence of **ventricular arrhythmia** that is

- **Frequent** ($\geq 5\%$ total PVC burden)
- or
- **Complex** (NSVT, VT, VF)

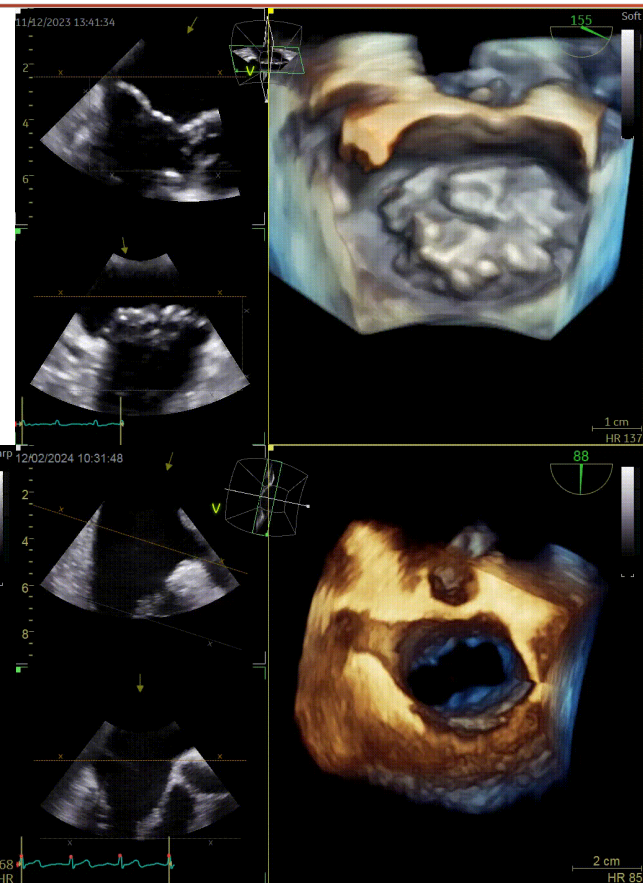
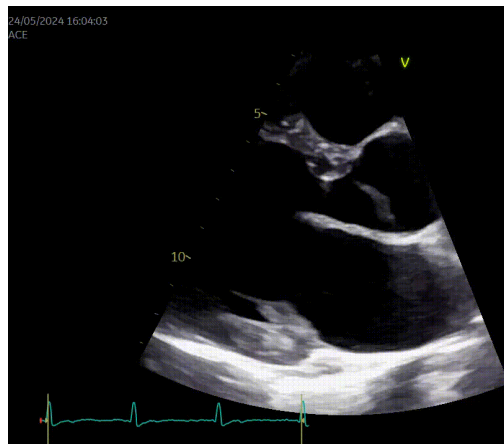
The absence of any other well-defined arrhythmic substrate



Echocardiographic Predictors of Arrhythmias: Mitral Valve Morphology



- **Bileaflet MVP**, quite prevalent among patients with MVP, **is not independently an SCD trigger** in large cohorts or population-based studies (Avierinos JF et al, Circulation 2002)
- Five studies enrolling 1393 patients compared prevalence of bileaflet prolapse between AMVP and NAMVP. **AMVP was associated with a higher prevalence of bileaflet prolapse** (OR: 1.92; 95% CI: 1.48–2.5; $p < .0001$)

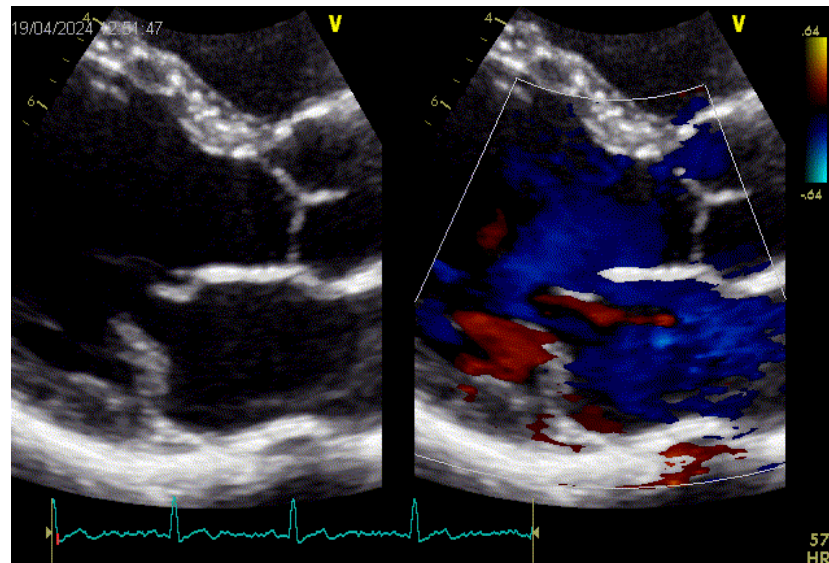


Arrhythmic Mitral Valve Prolapse and severe degenerative mitral regurgitation

- Patients with MVP and severe MR showed **high-risk of excess mortality** including excess SCD, irrespective of valvular morphology
- Atrial arrhythmias, reduced LV systolic function** and **severe heart failure symptoms** are associated with an increased SCD risk, but **even without such risk factors**, MVP with at least moderate to severe MR yields excess mortality and SCD

Table 10 Grading the severity of MR

MR severity classes	Mild	Moderate		Severe
MR Severity sub-classes ¹	Mild (Grade 1 or 1+)	Mild-to-moderate (Grade 2 or 2+)	Moderate-to-severe (Grade 3 or 3+)	Severe (Grade 4 or 4+)
Qualitative parameters				
MV morphology	None or mild leaflet abnormality or minimal tenting	Moderate leaflet abnormality or moderate tenting	Moderate leaflet abnormality or moderate tenting	Flail leaflet/large coaptation defect/severe tenting
Colour flow MR jet	Small, central (usually <4 cm ² or <20% of LA area)	Intermediate (usually 4–6 cm ² or 20–30% of LA area)	Intermediate (usually 6–8 cm ² or 30–40% of LA area)	Large central jet (usually >8 cm ² or >50% of LA area) or eccentric jet swirling and reaching the posterior wall of the LA
Flow convergence zone ^a CW signal of MR jet	No or small faint/parabolic	Dense, partial or parabolic	Dense, parabolic or triangular	Large throughout systole Holosystolic/dense/triangular
Semi-quantitative parameters				
VC width (mm)	<3	3 to <5	5 to <7	≥7 (≥8 for biplane) ^b
Pulmonary vein flow	Systolic dominance ^c	Normal or systolic blunting	Systolic blunting	Minimal to no systolic flow/ systolic flow reversal^d
Mitral inflow	A wave dominant ^b	Variable	E-wave dominant (Peak E > 1.2 m/s) ^f	E wave dominant (Peak E > 1.2 m/s) ^f
VTI mitral/VTI LVOT	<1	Intermediate	>1.2 ^f	>1.4 ^f
Quantitative parameters ^{h,k}				
EROA (mm ²)	<20	20–29	30–39	≥40
RVol (mL) ^j	<30	30–44	45–59	≥60
RF (%)	<30	30–39	40–49	≥50
CMR parameters				
RF (%)	<30	30–39	40–49	≥50
Structural parameters				
LV and LA size ^g	Usually normal	Normal or dilated	Usually dilated	Usually dilated
PA pressures ^e	Usually normal	Normal or elevated	Normal or elevated	Usually elevated

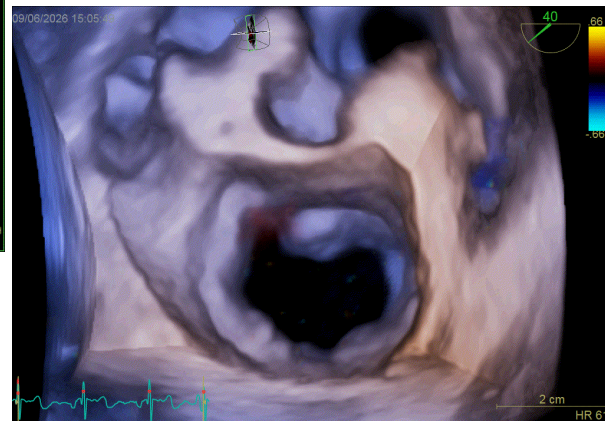
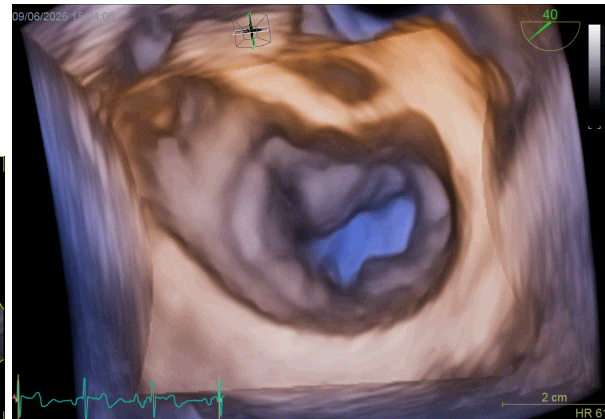
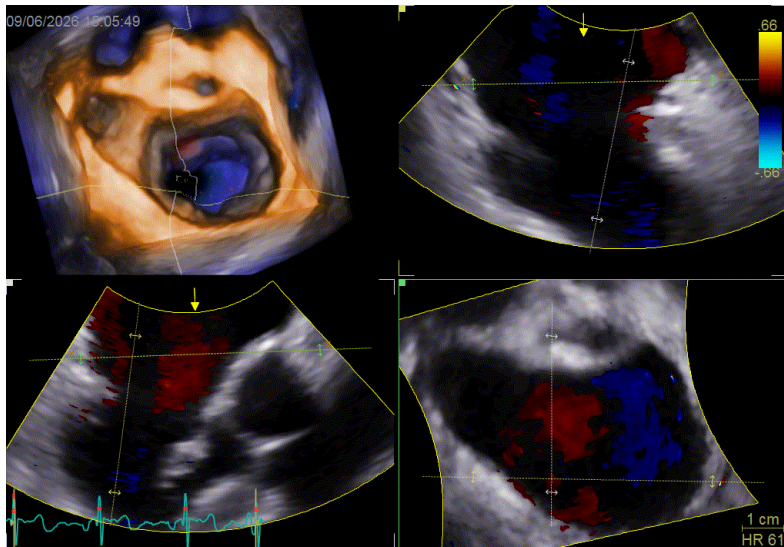


Arrhythmic Mitral Valve Prolapse: The Added Value of 3D Echocardiography

➤ **Accurate localization of mitral valve pathology**

- Involved scallops
- Chordal rupture
- MR severity
- MAD extent
- Clefts and cleft-like indentations

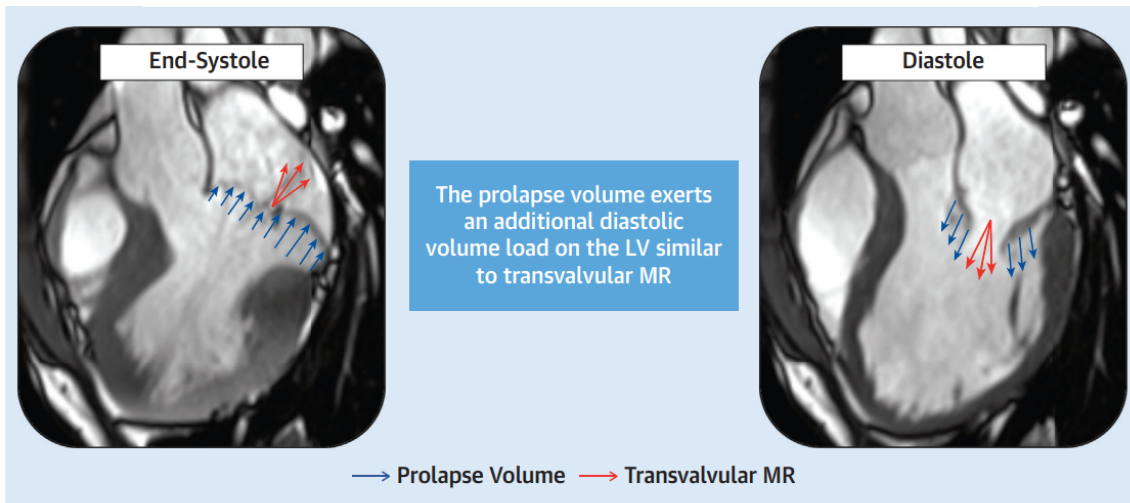
➤ **Gold standard for surgical planning in severe degenerative MR**



Arrhythmic Mitral Valve Prolapse and disproportionate LV remodeling

Additional Loading Effect of the Prolapse Volume on the Left Ventricle

- Assessment of LA and **LV remodeling** and both **regional** and **global LV function** (disproportionate LV enlargement in Barlow disease → consider **prolapse volume**)



El-Tallawi, K.C. et al. J Am Coll Cardiol Img. 2021;14(3):573-84.



Arrhythmic Mitral Valve Prolapse and disproportionate LV remodeling

- High prevalence of LGE, abnormal LV dilatation, > 1% PVC, %NSVT/VT/VF in patients with trace-mild MR! **MVP-associated cardiomyopathy?**

RESEARCH ARTICLE

Originally Published 1 April 1974 |

Check for updates

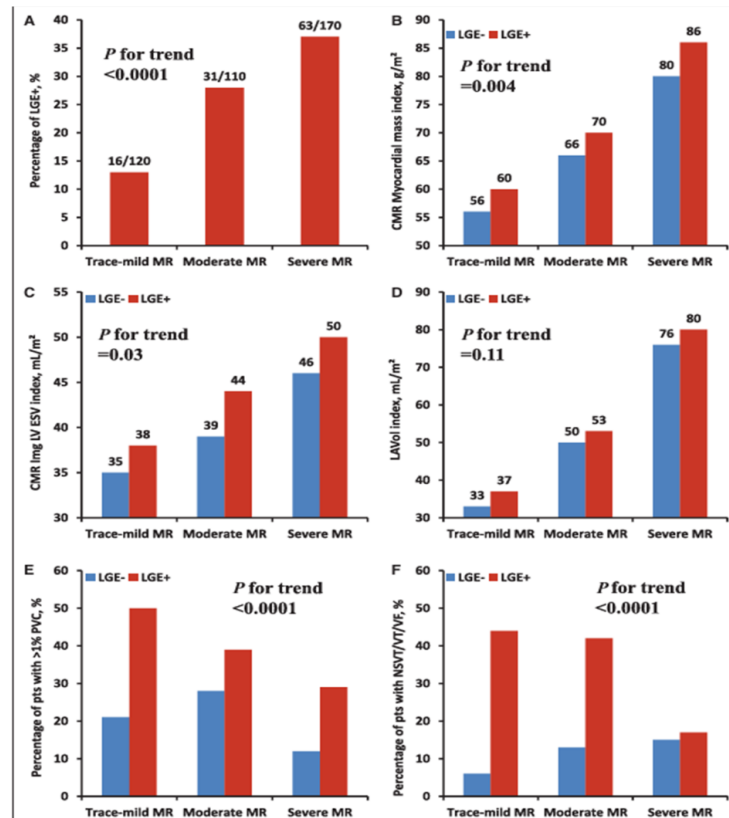
The Syndrome of Systolic Click, Murmur, and Mitral Valve Prolapse-A Cardiomyopathy?

STEPHEN J. GULOTTA, M.D., LOUIS GULCO, M.D., VELLORE PADMANABHAN, M.D., and STUART MILLER, M.D. | [AUTHOR INFO & AFFILIATIONS](#)

СТЕПЕНЪ ТЪ ГУЛОТЪЪ МЪДЪ ГЪЛОЪ ГЪЛЪОЪ МЪДЪ ВЪЛЪОРЕ ПЪДЪМЪНЪВЪНЪНЪ МЪДЪ ИЪ МЪЛЪЪ МЪЛЪЪ МЪДЪ | [AUTHOR INFO & AFFILIATIONS](#)

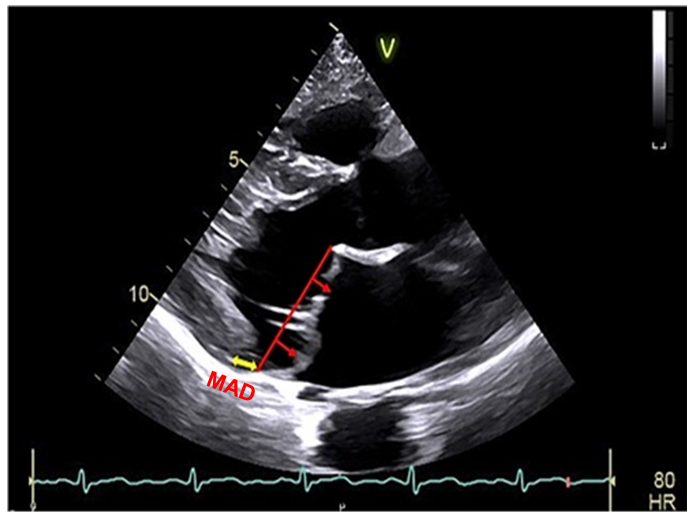
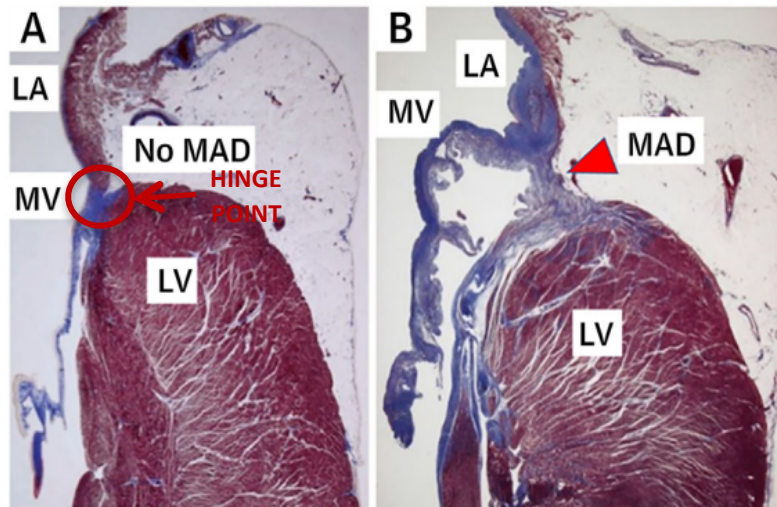
Cardiomyopathy

Circulation



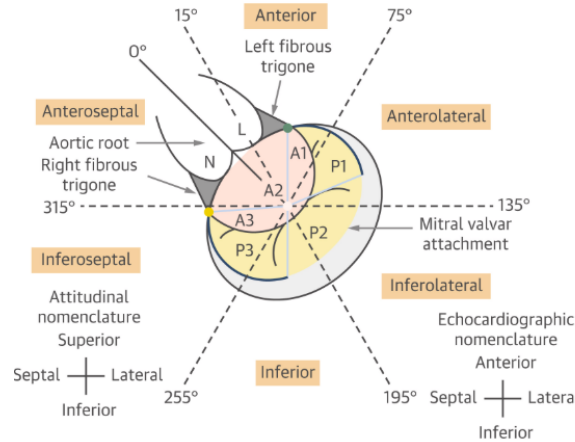
Arrhythmic Mitral Valve Prolapse and Mitral Annular Disjunction

Dislocation of the hinge line of the posterior leaflet to the posterior wall of the left atrium away from the crest of the left ventricle wall, in the direction of the roof of the left atrium (**> 8.5 mm** strong predictor for NSVT)

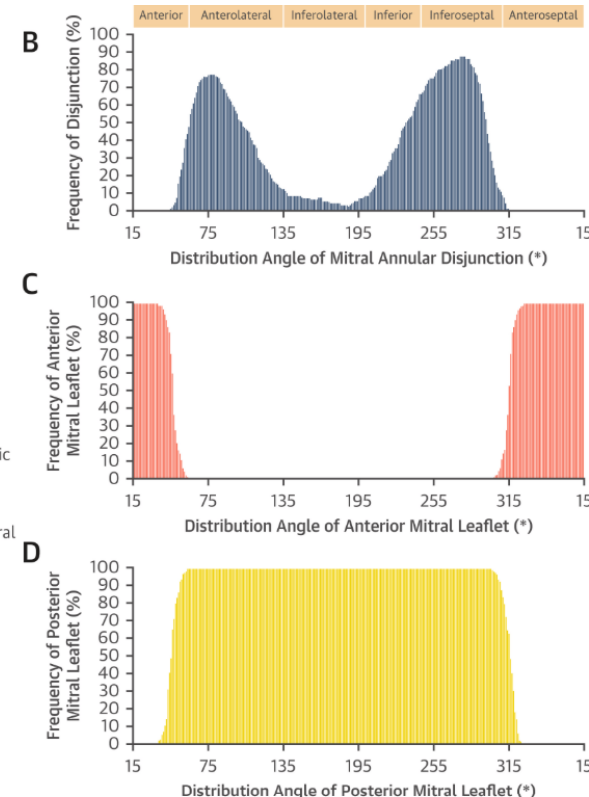


Arrhythmic Mitral Valve Prolapse and Mitral Annular Disjunction

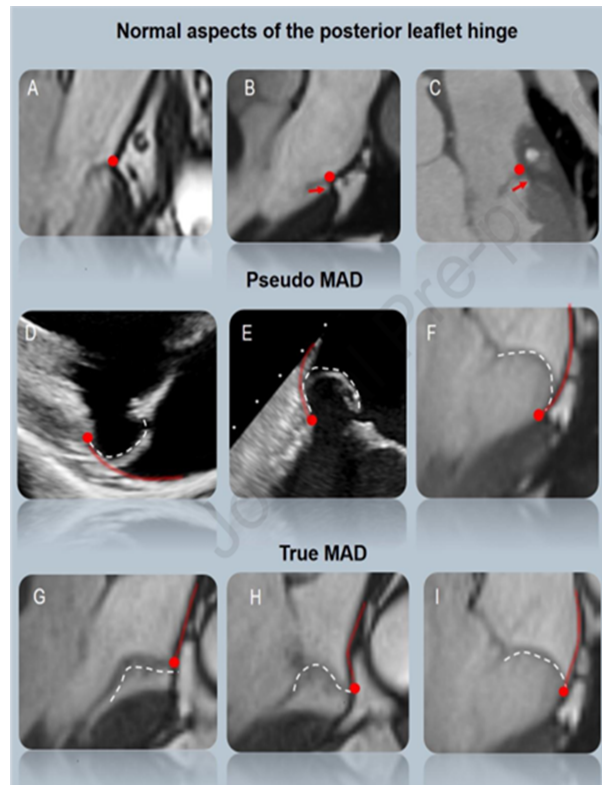
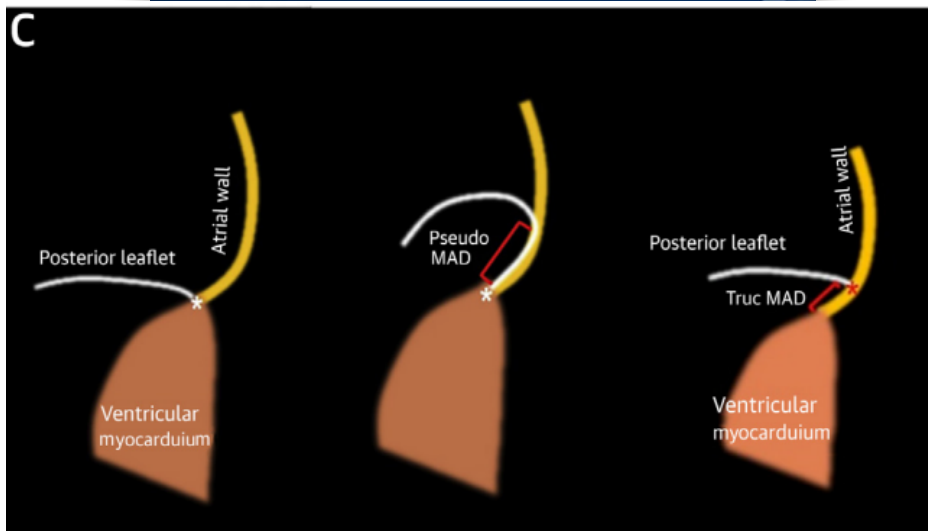
- **Variable prevalence in different MVP cohort studies**
- **A significant prevalence in structurally normal hearts**
- **MAD is infrequently observed in the posterior wall and is therefore likely often missed when the diagnosis relies on a parasternal long-axis view only (evolutional MAD vs pathogenic MAD)**



Toh H. et al *Eur Heart J Cardiovasc Imaging*. 2021

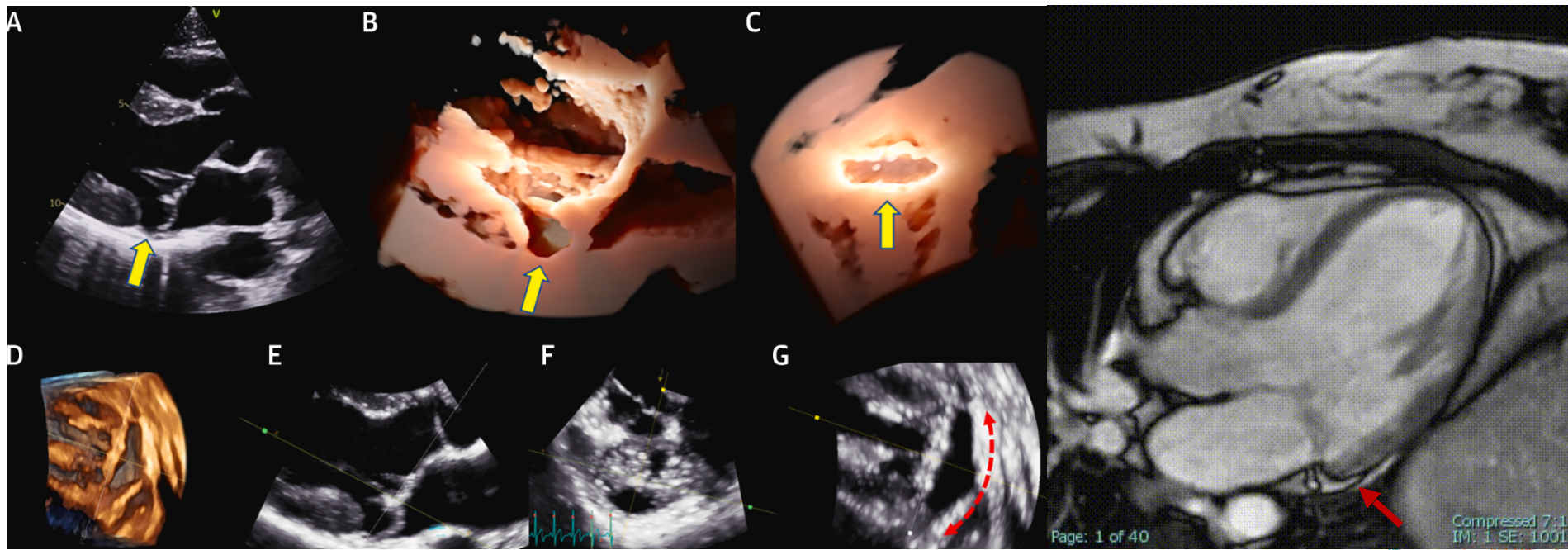


True-MAD vs Pseudo-MAD

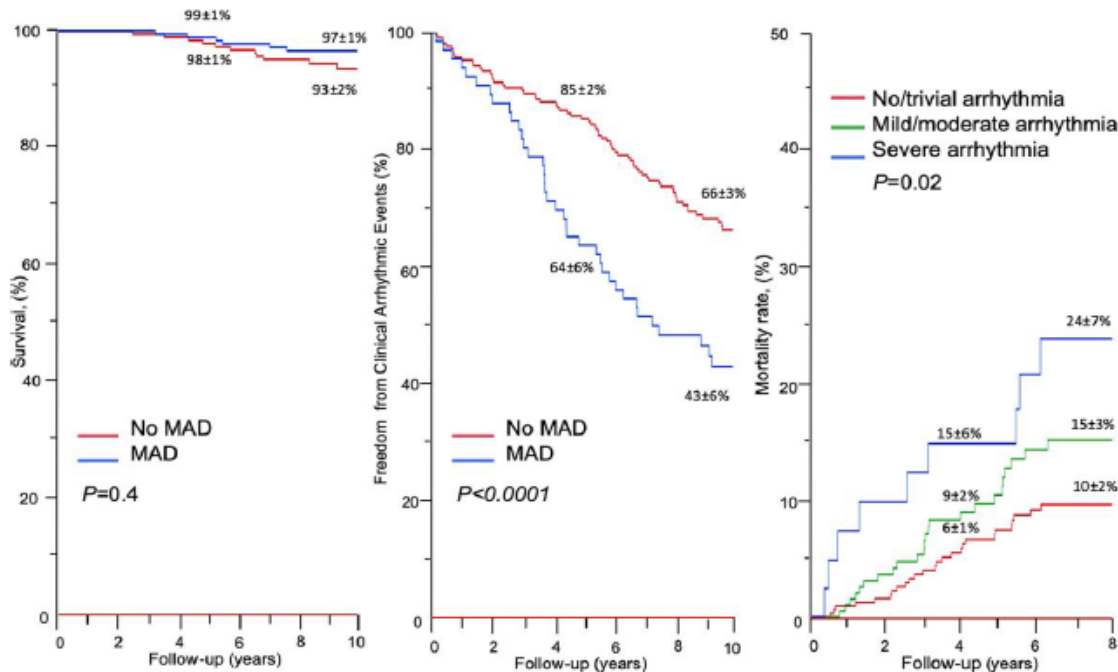


Multimodality Imaging for the Detection of True vs Pseudo MAD:

The Need for High Spatial Resolution



Arrhythmic Mitral Valve Prolapse and Mitral Annular Disjunction



MAD is not associated with excess-mortality

MAD is associated with excess-arrhythmic events

VA are associated with excess-mortality

Essayagh B Enriquez-Sarano M. Eur Heart J. 2023

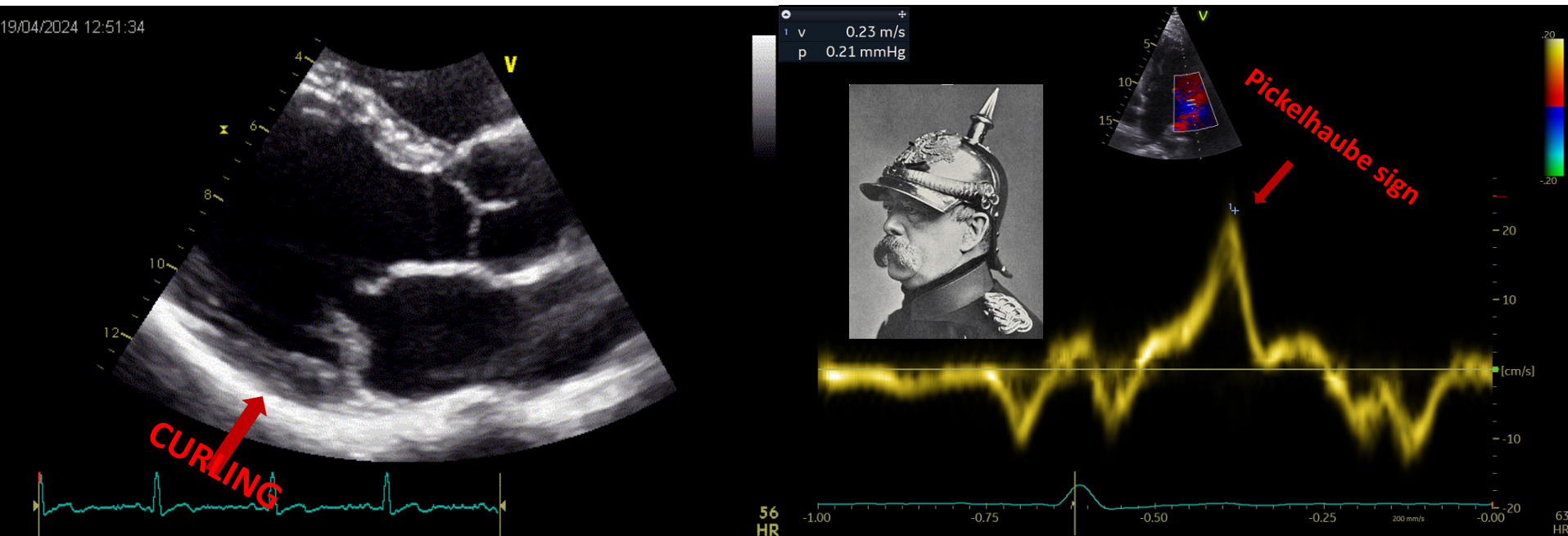


Echocardiographic predictors of arrhythmias: novel echo findings

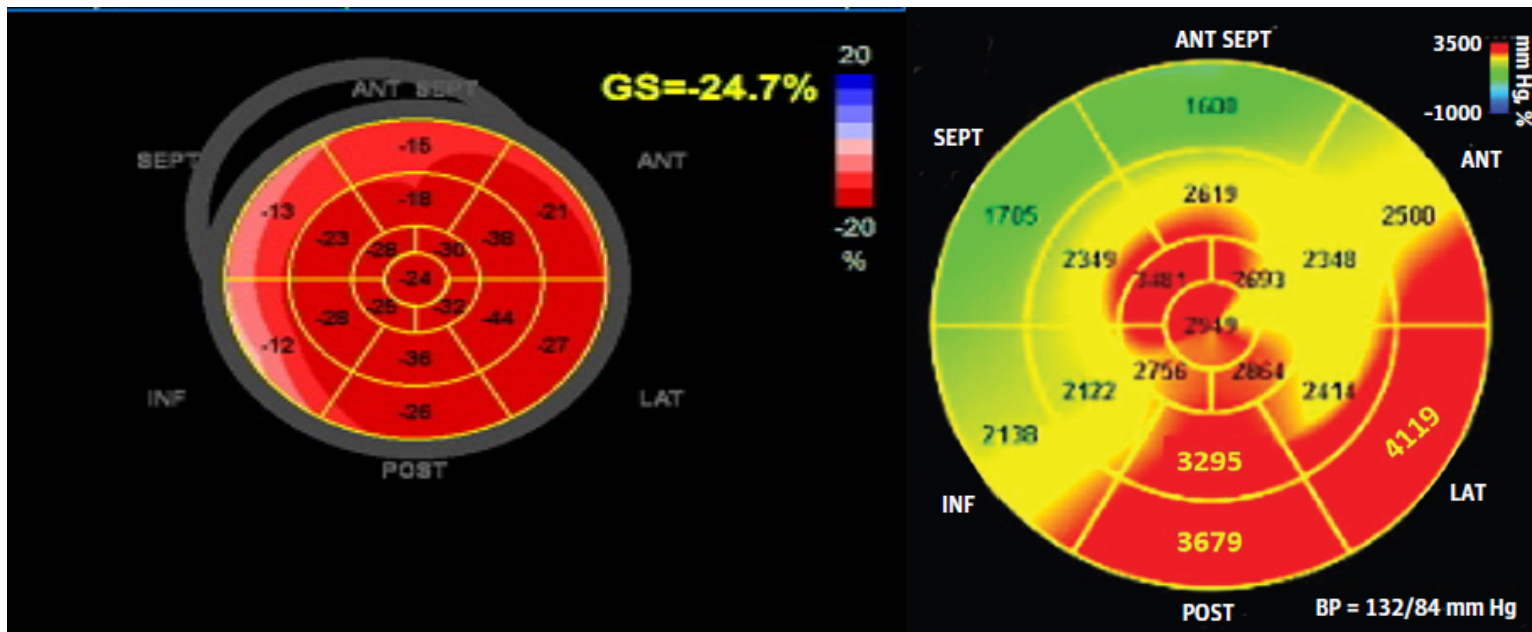


The Pickelhaube Sign: Novel Echocardiographic Risk Marker for Malignant Mitral Valve Prolapse Syndrome

19/04/2024 12:51:34



Echocardiographic predictors of arrhythmias: pathological GLS and MWI

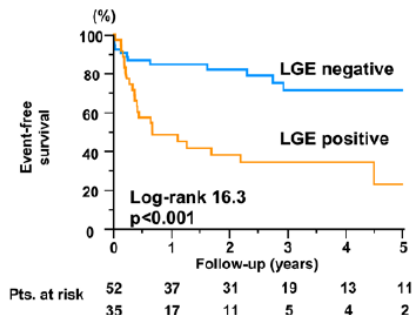
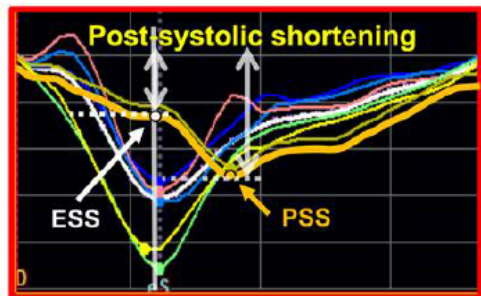


Danilowicz-Szymanowicz L. J Cardiovasc Dev Dis. 2023



Myocardial deformation in malignant mitral valve prolapse

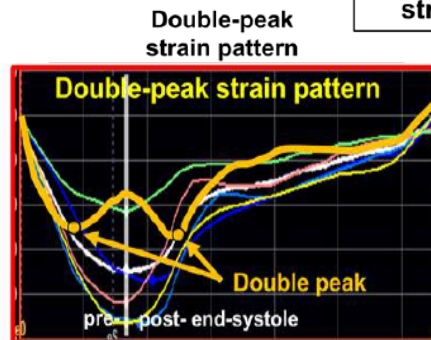
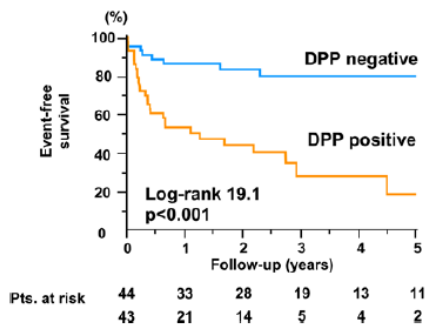
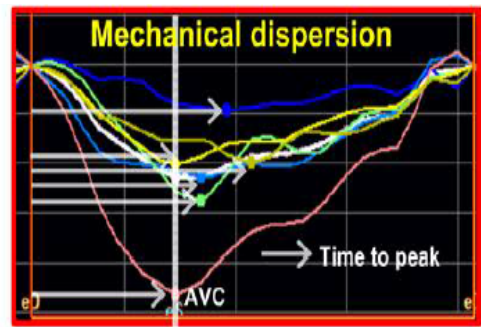
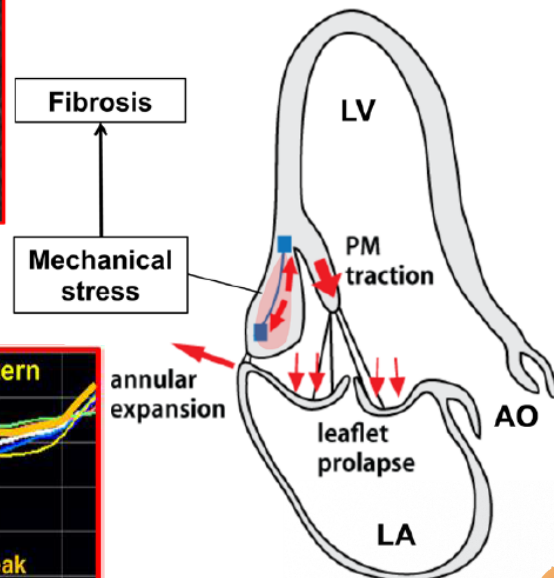
Arrhythmic events



LGE



Potential mechanism of mechanical stress



Yasufumi Nagata et al, Circulation: CVI 2023



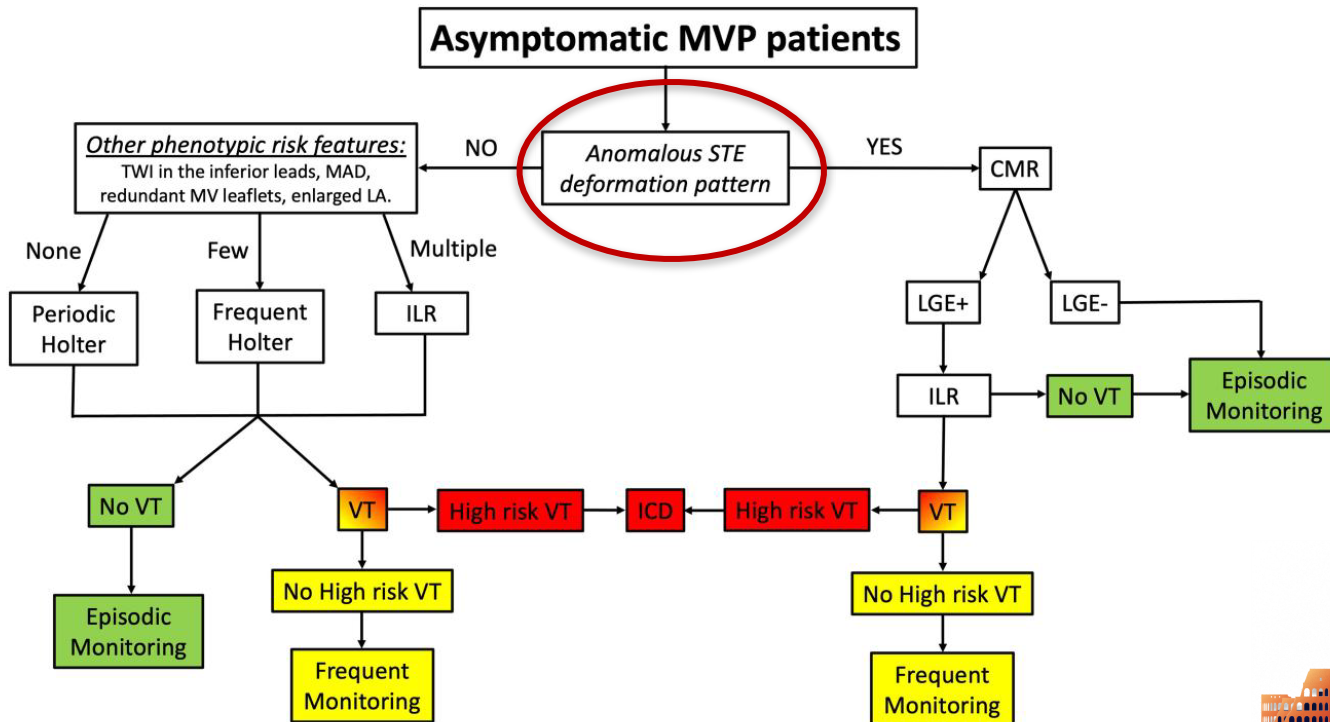


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Arrhythmic mitral valve prolapse: a practical approach for asymptomatic patients

Alessandro Malagoli ^{1*}, Alessandro Albini², Giovanni Benfari³, Federica Ilardi⁴, Matteo Lisi⁵, Giulia Elena Mandoli⁶, Maria Concetta Pastore⁶, Simona Sperlongano⁷, Matteo Cameli⁶, and Antonello D'Andrea ^{7,8}; on behalf of Working Group of Echocardiography



Take home messages

- The aMVP phenotype is best identified through an **integrated approach combining clinical history, ECG, and Holter monitoring with echocardiographic findings**
- **Transthoracic echocardiography is the cornerstone of risk stratification**, allowing recognition of key high-risk features (bileaflet prolapse, MAD, leaflet redundancy, systolic curling, Pickelhaube sign, LA/LV remodeling, reduced LVEF)
- Echocardiography also acts as a **gatekeeper for CMR**, helping select patients who warrant further imaging
- Echocardiographic and clinical risk markers should guide the **intensity of arrhythmia surveillance, follow-up strategy, and therapeutic decisions**, in a context of **progressive and dynamic disease expression over time**

