PLACE
PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

ROMA 2024
- 12 Giugno MINICORSI Precongressuali
- 13-15 Giugno CONGRESSO

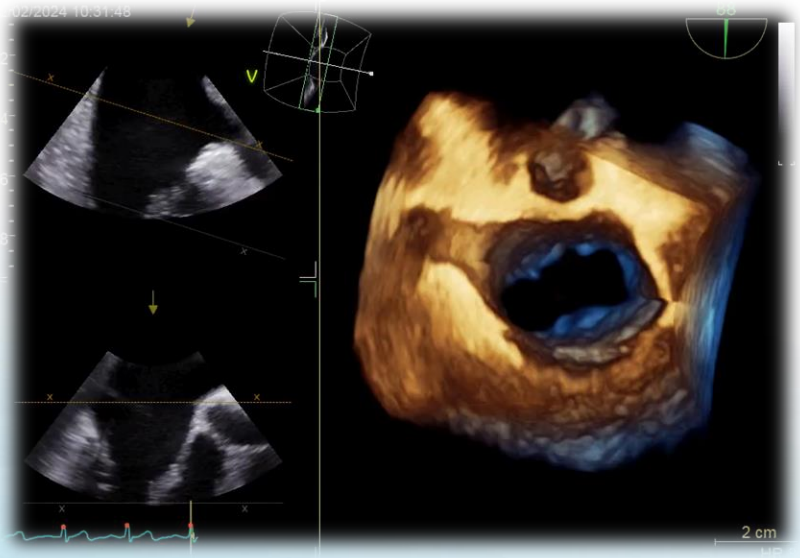
CARDIOMIOPATIA DILATATIVA NON ISCHEMICA E PROLASSO VALVOLARE ARITMICO

Prolasso mitralico aritmico: un approccio imaging guidato nel paziente asintomatico

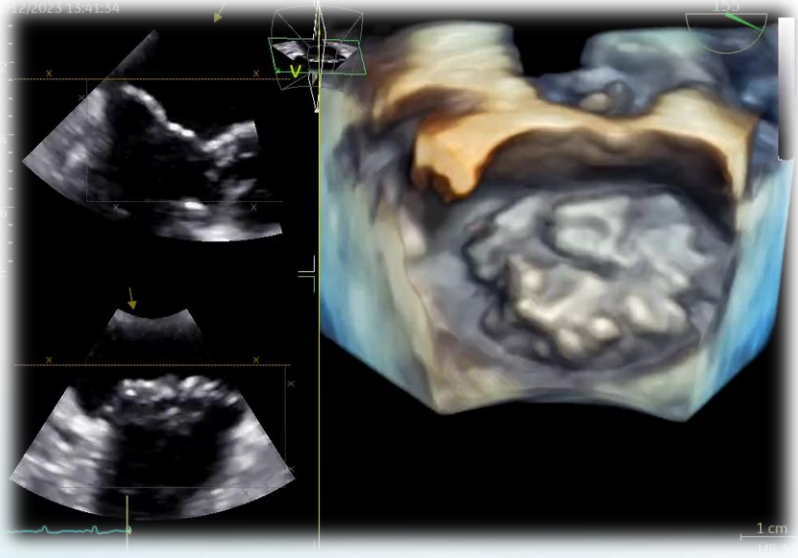
Pellegrino Ciampi, MD
Policlinico Casilino
Roma

Mitral valve prolapse: anatomopathological definition

Degenerative mitral valve disease characterized by pathological remodeling of the leaflets with two distinct phenotypes, fibroelastic deficiency (FED) and Barlow's disease



FED



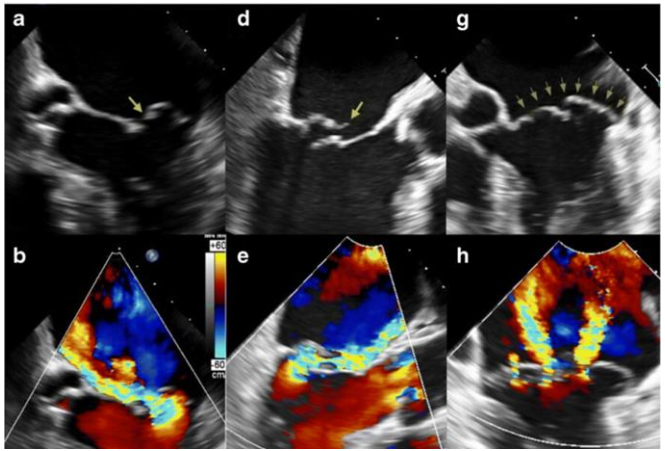
Barlow's disease

Clinical implications

Leading cause of **degenerative mitral regurgitation** (DMR) in the Western world

Congestive heart failure (volume overload)

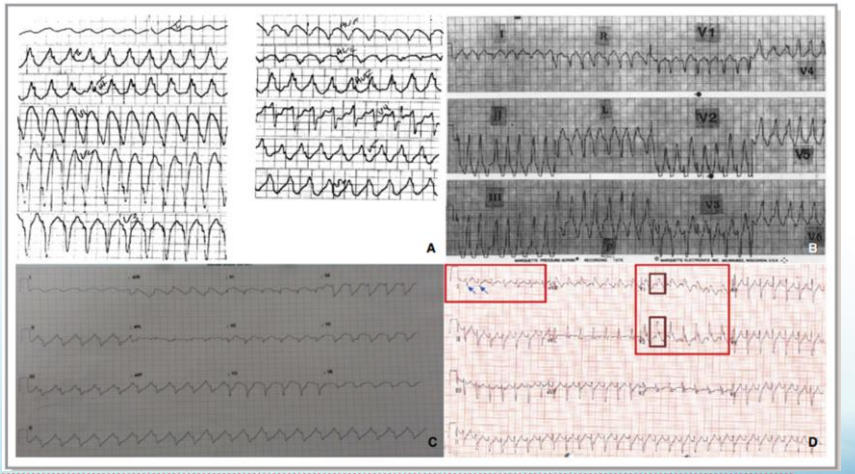
Endocarditis (arterial embolism)



Rhythm disorder (from premature ventricular contractions through arrhythmic sudden death) – "**arrhythmic and malignant MVP**"

- AMVP due to severe DMR

- AMVP with severe myxomatous disease irrespective of DMR

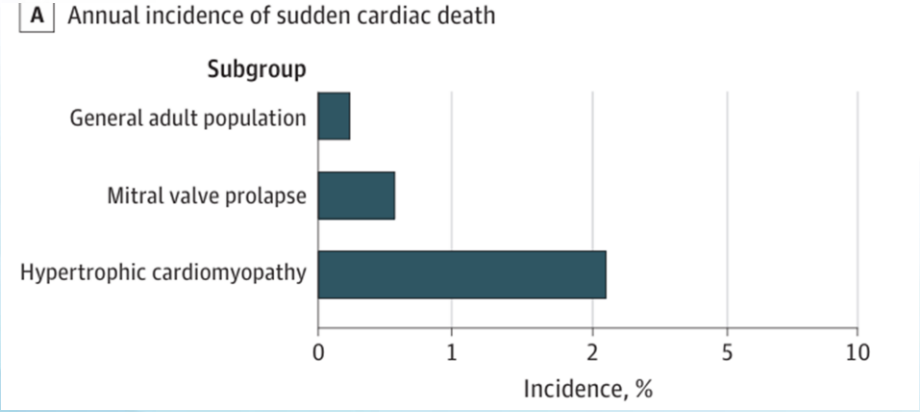


Epidemiology

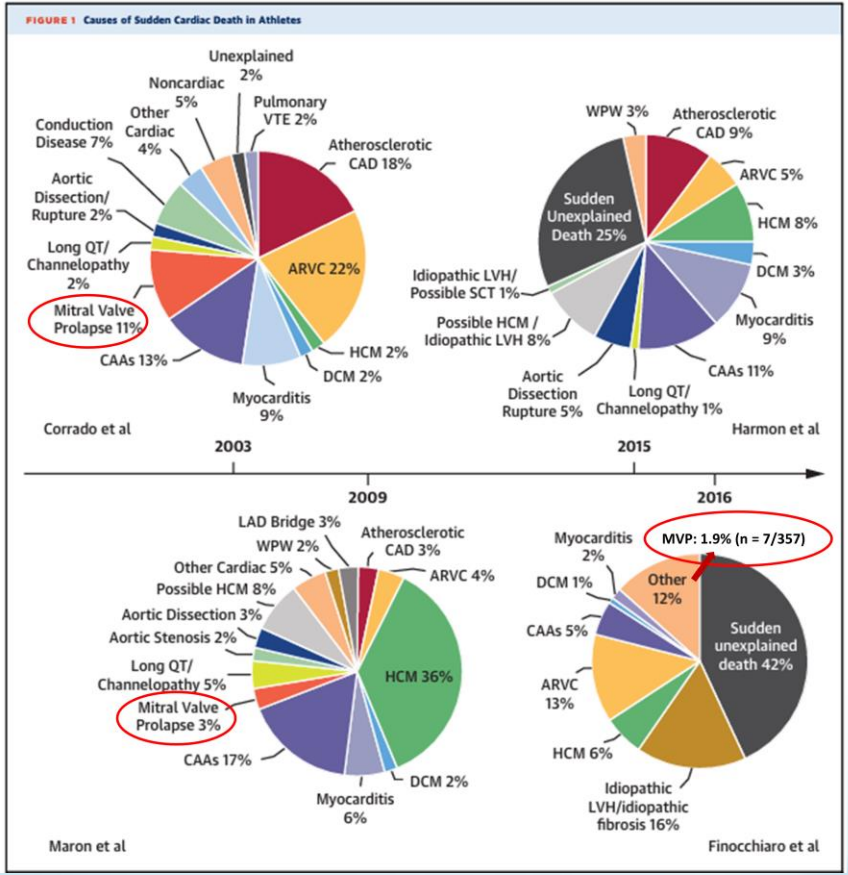
Estimated prevalence in the general population **2-3%**

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)*



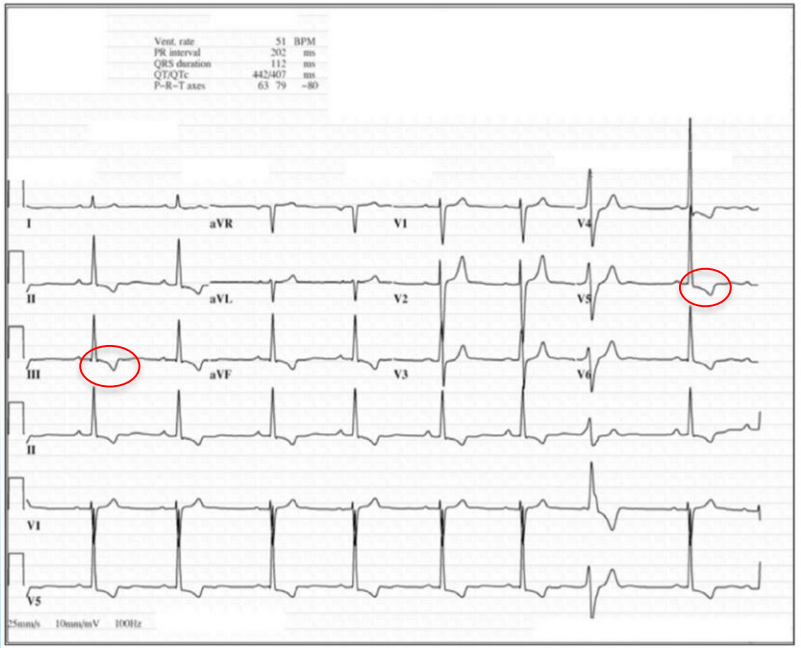
Muthukumar et al. JAMA Cardiology 2020



aMVP: Can we draw a patient identikit?

Demographic, **clinical** and **electrocardiographic risk factors**

- Female prevalance (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



Premature ventricular contractions and **Holter monitoring**

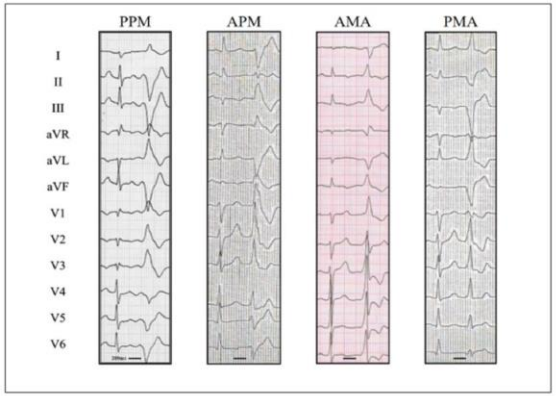
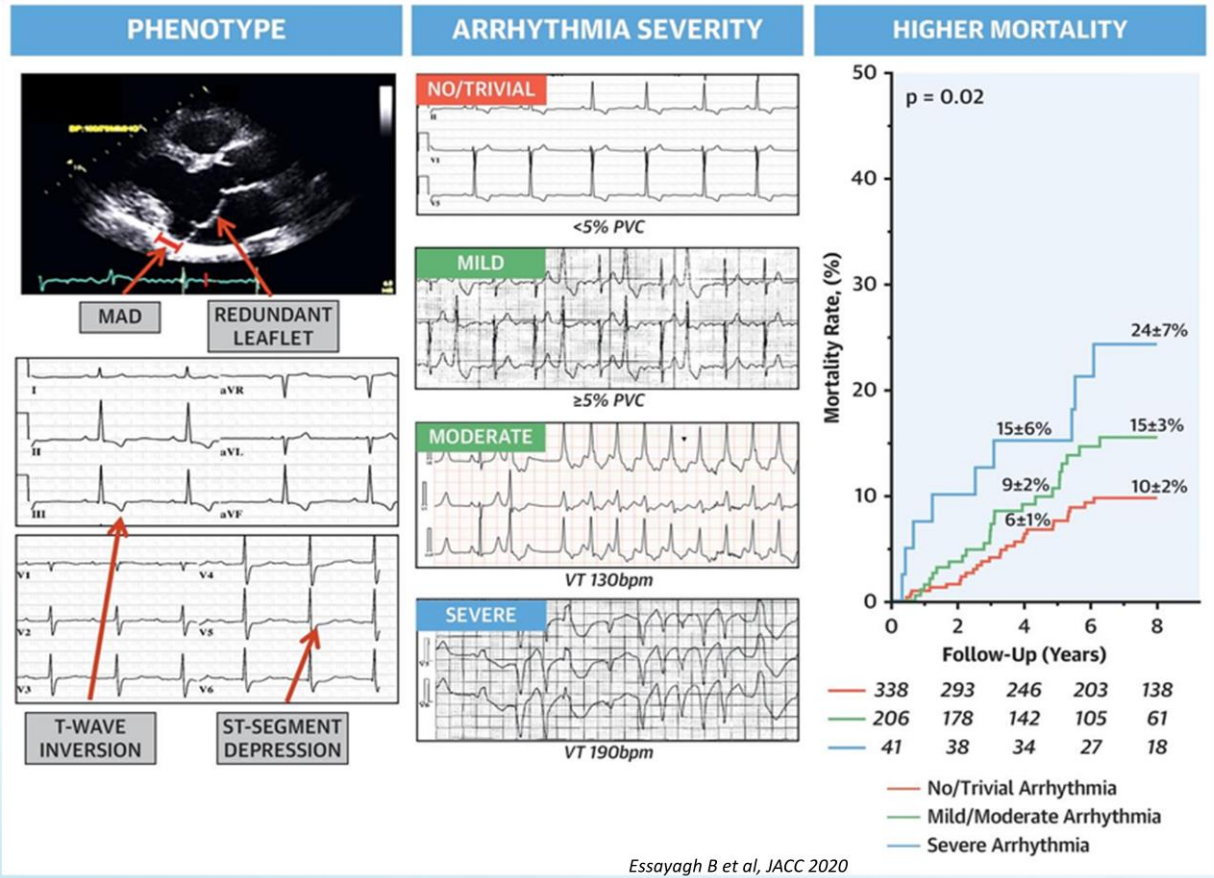


Table 3 **Arrhythmia severity classification**

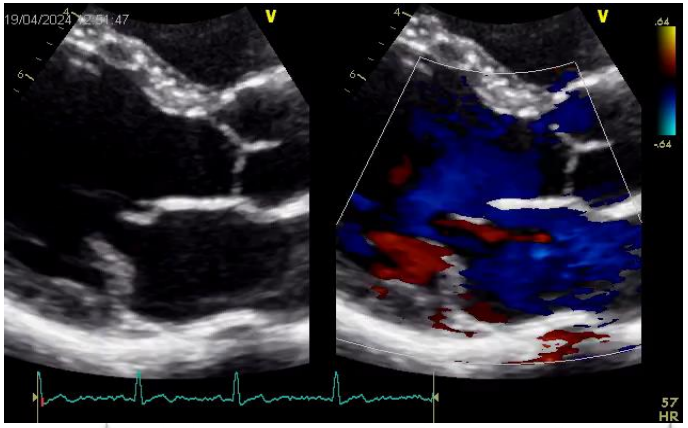
Severity	Arrhythmia burden/rate	Risk of mortality HR [95% CI]	References
Mild ventricular arrhythmia	PVC≥5% and/or VT runs <120 bpm	1.20 [0.68–2.14], $P=0.5$	[14]
Moderate ventricular arrhythmia	VT runs 120–179 bpm		
Severe ventricular arrhythmia	VT runs ≥180 bpm and/or history of sustained VT/VF	2.94 [1.36–6.36] $P=0.006$	

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



Essayagh B et al, JACC 2020

Identifying MVP at risk for ventricular arrhythmia and SCD: do imaging tools help?

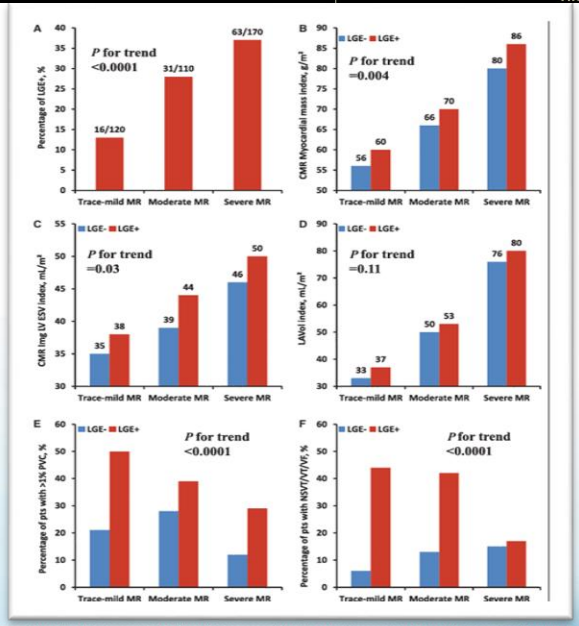


- superior systolic **displacement** of the mitral valve leaflet of **> 2mm**

- Quantification of **MR severity** (integration of qualitative, semi-quantitative and quantitative parameters)

- Assessment of **LV remodelling** and both **regional** and **global LV function**

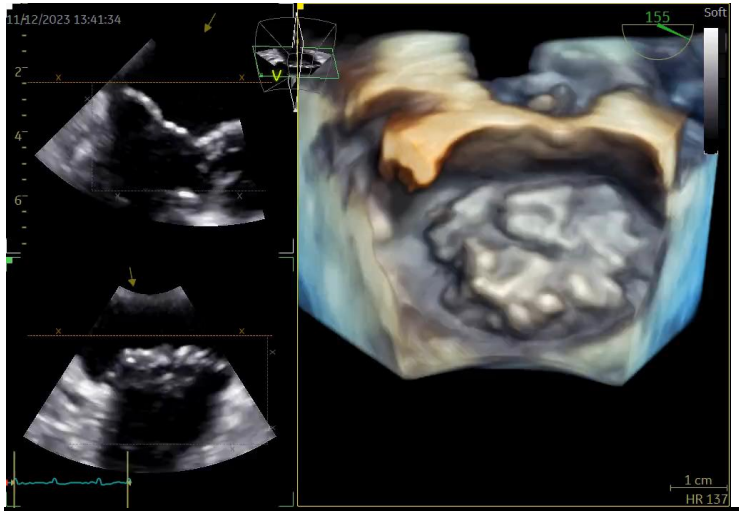
- High prevalence of LGE, abnormal LV dilatation, > 1% PVC, %NSVT/VT/VF in patients with trace-mild MR! **MVP-associated cardiomyopathy?**



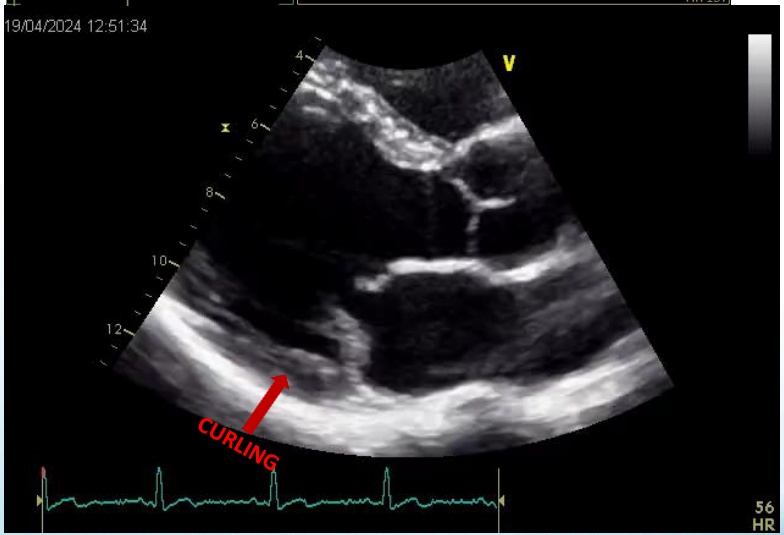
Constant Dit Beaufils AL et al. Circulation. 2021

Ehocardiographic predictors of arrhythmias

- severe myxomatous degeneration (thick and redundant leaflets with multi-segment bileaflet MVP)



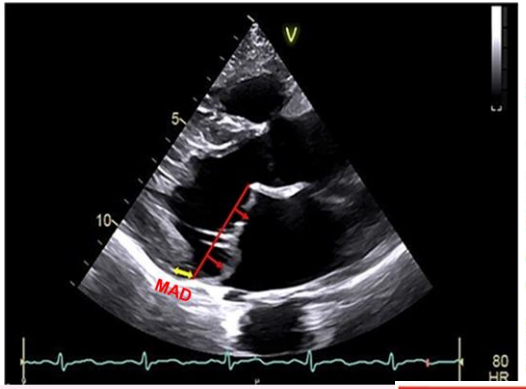
- Systolic Curling (ballerina foot deformity)



- Mitral annulus disjunction (MAD)

MAD or **MADNESS**

- Systolic separation between the ventricular myocardium and the mitral annulus supporting the PML (**> 8.5 mm** strong predictor for NSVT)
- **Variable prevalence in** different **MVP** cohort studies
- A **significant prevalence** in structurally **normal hearts**
- **Association** with a **higher risk of arrhythmic** events **but not with a higher mortality**
- Association of MAD with VT/aborted cardiac arrest in patients without MVP: a **MAD arrhythmic syndrome?**



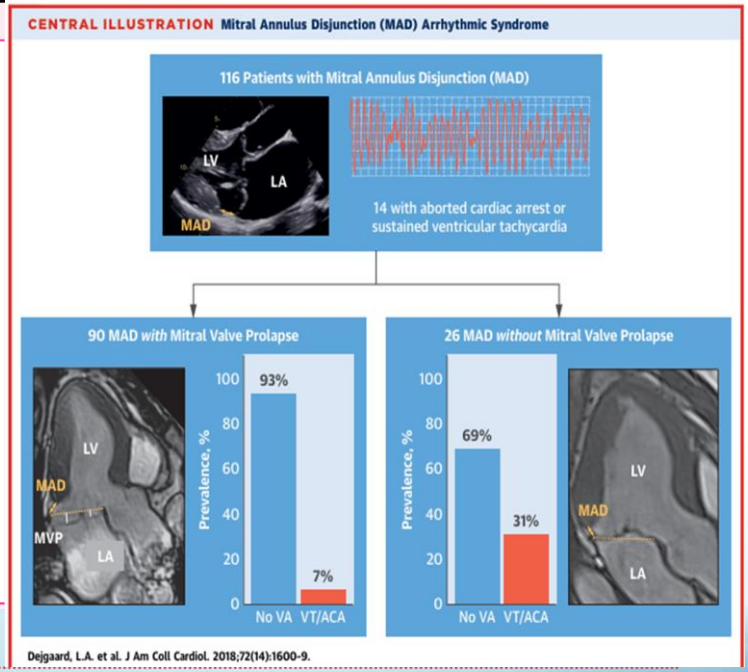
ESC European Society of Cardiology
European Heart Journal - Cardiovascular Imaging (2021) 22, 614–622
doi:10.1093/ehjci/jeab022

Prevalence and extent of mitral annular disjunction in structurally normal hearts: comprehensive 3D analysis using cardiac computed tomography

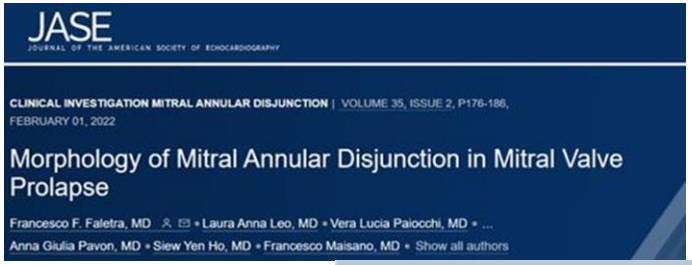
Hiroyuki Toh[1], Shumpei Mori[2]*, Yu Izawa[1], Hiroshi Fujita[1], Keisuke Miwa[1], Masataka Suzuki[1], Yu Takahashi[1], Takayoshi Toba[1], Yoshiaki Watanabe[3], Atsushi K. Kono[3], Justin T. Tretter[4], and Ken-Ichi Hirata[1]

Table 1 Prevalence of mitral annular disjunction

Study	Year	Population	Prevalence of MAD, *n*/*N* (%)
TTE			
Carmo *et al.*[22]	2010	Myxomatous mitral valve prolapse	21/38 (55)
Lee *et al.*[27]	2017	Mitral valve prolapse	42/156 (27)
Konda *et al.*[20]	2017	Patients referred for TTE	125/1439 (9)
Mantegazza *et al.*[28]	2019	Mitral valve prolapse, severe MR	103/979 (16)
Torras *et al.*[29]	2019	Mitral valve prolapse	22/101 (22)
Essayagh *et al.*[19]	2021	Mitral valve prolapse	186/596 (31)
Essayagh *et al.*[30]	2021	Mitral valve prolapse and severe MR undergoing repair	27/61 (44)
CMR			
Christiansen *et al.*[31]	2010	Mitral valve prolapse undergoing CMR	18/31 (58)
Perazzolo Marra *et al.*[16]	2016	Arrhythmic myxomatous mitral valve prolapse	37/52 (71)
Essayagh *et al.*[32]	2019	Mitral valve prolapse	31/89 (35)
Zugwitz *et al.*[33]	2022	UK Biobank imaging study participants	1990/2607 (76)
Figliozzi *et al.*[26]	2023	Mitral valve prolapse	321/474 (68)

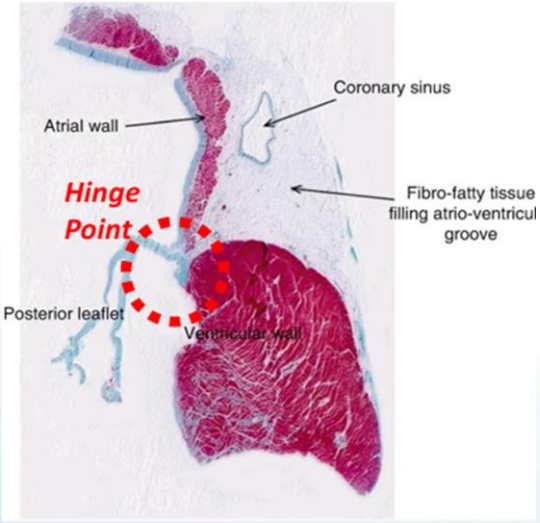


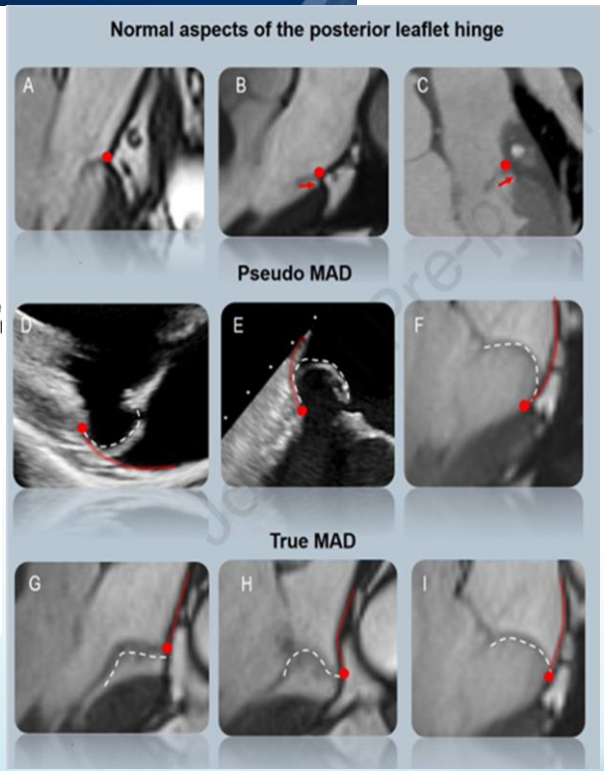
True MAD vs the «new» concept of pseudo-MAD



- The «**systolic MAD**» or **pseudo-MAD**, most common, result of a juxtaposition of the belly of the billowing PL on the adjacent LA wall

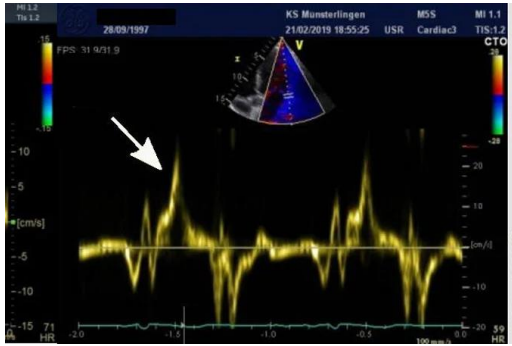
- The «**true MAD**», more rare, when the insertion of PL at hinge line is clearly displaced on the atrial wall either in **diastole** or in **systole**





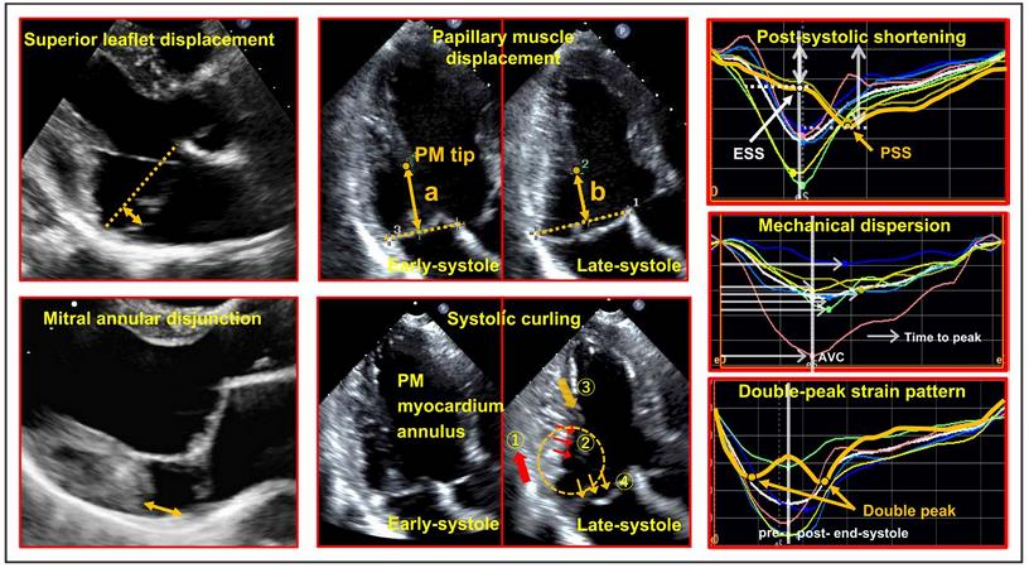
Ehocardiographic predictors of arrhythmias: novel echo findings

- **Pickelhaube sign**: marker of abrupt forceful myocardial stretch (**>16 cm/s** associated with VA, highest signal velocity in the **posterolateral annulus**)



- **Speckle tracking echocardiography**: ventricular repolarization heterogeneity – electromechanical dissynchrony, ventricular hyperexcitability

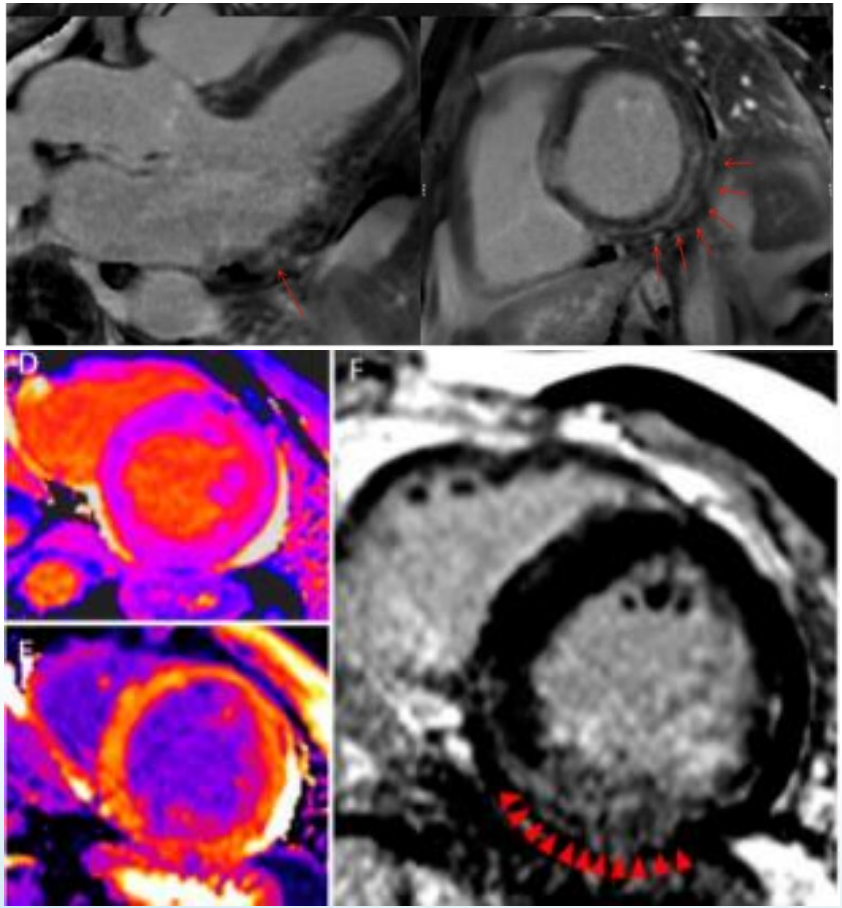
  - Supranormal peak longitudinal strain in infero-lateral mid-basal ventricular segments
  - Mechanical dispersion
  - Postsystolic strain index (PSI)
  - Double peak strain pattern



Nagata Y, Bertrand PB, Baliyan V, Kochav J, et al. Circ Cardiovasc Imaging. 2023 Apr

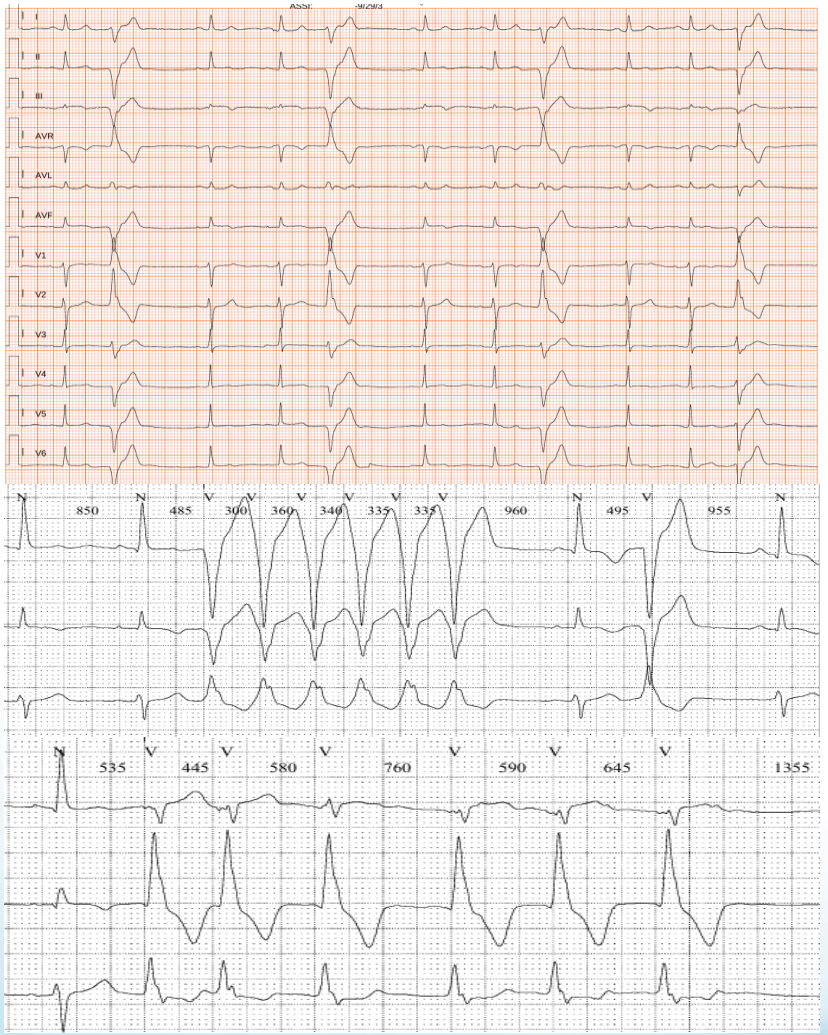
CMR predictors of arrhythmias

- **variable prevalence** rates of different pattern of **replacement fibrosis** (mid wall, patchy, or subendocardial) of the **PPM** and **infero basal LV wall** by LGE in patients with arrhythmic MVP (**28-37%**)
- strong correlation of LGE and arrhythmic events/SCD despite MR severity (**25%** of LGE in pts with **mild MR**)
- Future studies will determine whether burden/patterns of LGE enhance arrhythmic risk stratification in pts with MVP
- Most patients with MVP and VA have scar, but not all!
- In a retrospective analysis by Garbi et al of SCD cases with MVP, **81%** had either **diffuse interstitial fibrosis** or **none** (role of **T1 mapping** to detect arrhythmic risk in MVP pts)



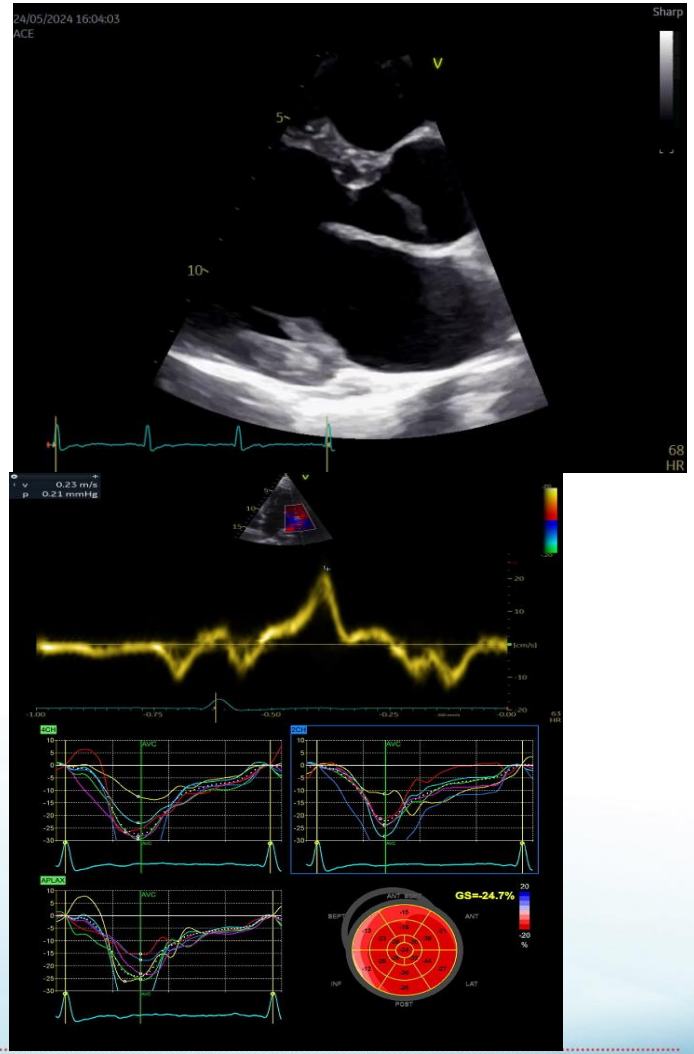
Clinical Case

- Female, 37 y/o
- neither CV risk factors nor family history of CM and SCD
- Episodic palpitations, nor syncope or presyncope
- EKG: SR, inverted T waves in DIII, flattened T waves in lateral leads, PVCs RBBB superior axis morphology
- Holter EKG: 2884 PVC (2.9%), 2 NSVT



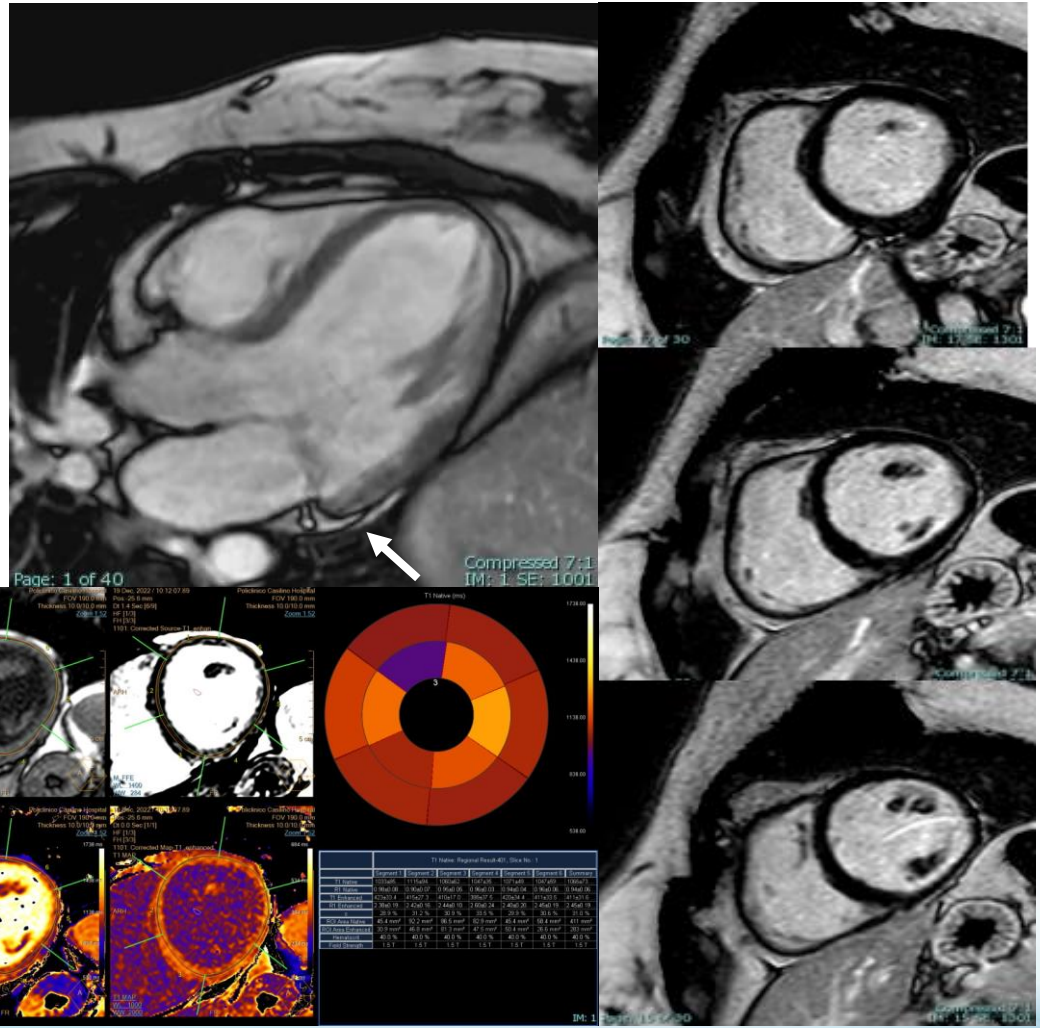
Echocardiographic findings

- severe myxomatous degeneration (thick and redundant leaflets with multi-segment bileaflet MVP)
- Mild to moderate MR
- Severe LA dilatation, mild LV dilatation with normal LVEF
- Curling, Pickelhaube sign (TDI S’ 23 cm/s), systolic MAD 6 mm
- Excessively increased (more negative) segmental LS in basal and mid lateral segments with increased electro-mechanical dyssyncrony (MD and PSI ↑↑)



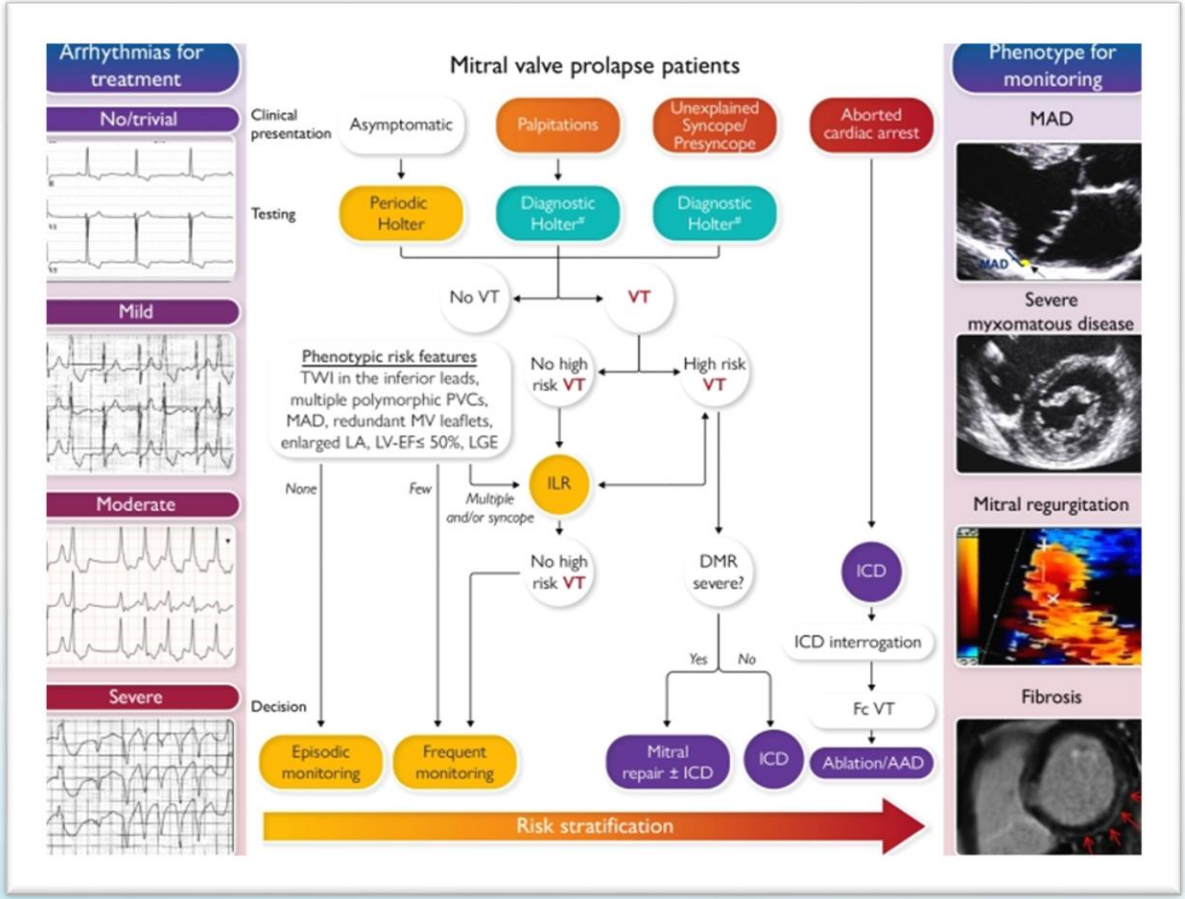
CMR findings

- LV and LA dilatation, preserved LV function;
- Mild MR;
- Basal curling;
- True MAD 5 mm;
- No LGE, normal native myocardial T1, ↑ ECV in basal infero-lateral segment



Treatment and follow up

- ILR implantation
- Medical therapy with Nadolol and Flecainide
- No symptoms
- Reduction of arrhythmic burden without high risk VT at follow up



Sabbag A, Essayagh B, Barrera et al, Heart Rhythm Society. Europace. 2022.

Take Home Messages

- When diagnosing MVP, it is important to consider an aMVP phenotype, also irrespective of DMR severity

- MVP diagnosis requires a 24h Holter-ECG also in asymptomatic patients without severe VA at index monitoring because arrhythmic risk in MVP is progressive over time

- Multimodality imaging plays a pivotal role in aMVP phenotypic characterisation and should be advised as part of routine follow up

- Longitudinal studies and prospective outcome data are needed to determine the predictive value of all the currently available risk stratification imaging tools