The arrhythmic mitral valve prolapse: presentation and outcome

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Topic(s):
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Background: The majority of patients with of Mitral-valve-prolapse (MVP) have a excellent prognosis. Until recently most cases of mortality were thought to be related to mitral regurgitation and left ventricular dysfunction. The concept of the arrhythmic MVP emerged to describe cases of sudden cardiac death (SCD) in the presence of isolated MVP yet it’s phenotype remains incompletely and inconsistently defined.

Purpose: To analyze the prevalence, severity and characteristics of ventricular-arrhythmia (VA), to determine it’s phenotypical context and independent impact on outcome in patients with MVP.

Methods: A cohort of 595 (65±16 years, 278 female) consecutive patients with MVP and comprehensive clinical, arrhythmia (24hour-Holter) and Doppler-echocardiographic characterization, was identified and long-term outcome analyzed.

Results: VA was frequent, present in 43% of patients with at least ventricular ectopy=5%, but was most often moderate (ventricular-tachycardia—VT 120-179bpm) in 27% and rarely severe (VT=180/min) in 8.6%. Presence of VA was associated with older age, male sex, bileaflet-prolapse, marked leaflet redundancy, mitral-annulus-disjunction (MAD), larger left-atrium and left ventricular end-systolic diameter, and T-wave-inversion/ST-depression (all P=0.001). Severe VA was independently associated with presence of MAD, leaflet-redundancy and T-wave-inversion/ST-depression (all P<0.0001) but not with mitral regurgitation severity or ejection-fraction. Outcome primary endpoint of overall survival after arrhythmia diagnosis (8-year 87±2%) was strongly associated with arrhythmia-severity (8-year 90±2% for no/trivial arrhythmia, 85±3% for mild/moderate and 76±7% for severe arrhythmia. P=0.02, Figure). Excess-mortality was substantial for severe-arrhythmia (univariate-hazard-ratio 2.70[1.27-5.77], P=0.01 vs. no/trivial arrhythmia), even adjusted comprehensively including for MVP-characteristics (adjusted-hazard-ratio 2.94[1.36-6.36], P=0.006).

Conclusions: This large cohort of isolated consecutive MVP characterized with 24-hour-Holter monitoring, clinical and Echocardiographic assessment, demonstrates that VA are frequent with MVP but rarely severe. The arrhythmic MVP was independently and strongly associated with specific ECG and morphologic patterns, particularly ST-T changes, MAD presence and marked leaflet redundancy, suggestive of a specific arrhythmic MVP phenotype, independently of MR-severity. Arrhythmia, particularly severe, is associated with long-term excess-mortality, independently of any other characteristics, including MR severity and LVEF. These findings lay the foundation for novel risk-stratification of MVP for the conduct of prospective controlled studies evaluating the management of MVP high-risk patients.

Figure – Impact on survival of ventricular arrhythmia
Overall survival of MVP stratified by ventricular arrhythmia (Panel A) or ventricular arrhythmia severity (Panel B) throughout follow-up.
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