

Predictors of and outcomes in arrhythmic mitral valve prolapse

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Background: Recently, the European Heart Rhythm Association (EHRA) defined the phenotype of the arrhythmic mitral valve prolapse (AMVP) complex and proposed management approaches. Assessing the incidence of SCD in this small subset of patients is challenging due to the low event-rate and longitudinal real-world data remain scarce.

Purpose: To determine the predictors of AMVP and study the clinical outcome of patients stratified by ICD indication as proposed by the EHRA consensus statement.

Methods: All consecutive patients with MVP who underwent cardiac magnetic resonance imaging (CMR) were enrolled in a single-center retrospective registry. AMVP was defined as the presence of frequent or complex ventricular arrhythmias (>5% total PVC burden, non-sustained ventricular tachycardia (VT), VT or ventricular fibrillation) in a patient with MVP without alternative arrhythmic substrates. The medical records of these patients included a comprehensive clinical, rhythmic and echocardiographic characterization allowing to collect the key date of transition to AMVP. A forward conditional multivariable logistic regression was performed to identify clinical risk factors for AMVP reporting the odds ratio (OR) and corresponding 95% confidence interval (95% CI). Incidence rates were calculated for the composite endpoint of SCD, sustained VT, ICD shocks, and all-cause mortality.

Results: A total of 169 patients (52.1% male, median age 51.4 years) were included, of which 99 (58.6%) were classified as AMVP. Clinical characteristics are presented in Image 1. Patients with non-arrhythmic MVP and AMVP had comparable left ventricular function, mitral valve regurgitation grade and extent of MVP. Multivariable logistic regression identified the presence of late gadolinium enhancement (LGE, OR 2.82, 95%CI 1.45–5.50, p=0.002) and mitral annular disjunction (MAD, OR 1.98, 95%CI 1.02–3.86, p=0.045) on CMR as only predictors of AMVP. Cardiac CMR results are presented in Image 2. The median overall follow-up since first cardiac contact was 8.0 years (IQR 5.0–15.6), while this was 5.0 years (IQR 2.1–7.4 years) after transition to AMVP. The incidence for the composite endpoint in the overall population was 0.4%/year (95%CI 0.2 – 0.9). The 5-year cumulative event rate was 1.8%. According to the EHRA risk stratification the implantation of an ICD would have been reasonable in 69 patients (69.7%). In total, 5 patients were implanted with an ICD for secondary prevention, but all those patients did not classify as AMVP prior to the life threatening arrhythmic event.

Conclusion: CMR is an important tool in the challenging risk stratification of AMVP given that the presence of MAD and LGE enhancement are predictors of AMVP. There is a lack of longitudinal data to determine when patients transition from MVP to AMVP and how this modifies their outcome.

Variable	All patients (N=169)	Non-arrhythmic MVP (N=70)	Arrhythmic MVP (N=99)	p-value
Age – first cardiac contact (y)	44.3 (29.8 – 56.9)	42.1 (19.3 – 55.7)	46.3 (33.3 – 57.6)	0.140
Age – CMR (y)	51.4 (36.5 – 62.4)	47.2 (31.3 – 61.9)	53.3 (39.0 – 63.1)	0.083
Male	88 (52.1%)	37 (52.9%)	51 (51.5%)	0.863
BMI (kg/m ²)	22.8 (20.6 – 24.9)	22.4 (20.3 – 24.8)	23.1 (20.9 – 25.0)	0.381
Arterial hypertension	27 (16.0%)	11 (15.7%)	16 (16.2%)	0.938
Dyslipidemia	52 (30.8%)	25 (35.7%)	27 (27.3%)	0.242
Active smoking	35 (20.7%)	12 (17.1%)	23 (23.2%)	0.336
Marfan	18 (10.7%)	12 (17.1%)	6 (6.1%)	0.023
Familial SCD	22 (13.0%)	10 (14.3%)	12 (12.1%)	0.680
Familial MVP	15 (8.9%)	7 (10.0%)	8 (8.1%)	0.666
Palpitations	115 (68.0%)	42 (60.0%)	73 (73.7%)	0.059
Syncope	23 (13.6%)	4 (5.7%)	19 (19.2%)	0.012
SCD	4 (2.4%)	0 (0.0%)	4 (4.0%)	0.089
Barlow's disease	56 (33.3%)	19 (27.1%)	37 (37.8%)	0.150
Heart rate (bpm)	65.0 (57.0 – 78.0)	71.0 (59.5 – 88.5)	62.0 (56.0 – 70.0)	<0.001
PR interval (ms)	158.0 (140.0 – 178.0)	160.0 (140.0 – 176.0)	158.0 (140.0 – 182.5)	0.804
QRS duration (ms)	94.0 (86.0 – 103.0)	96.0 (86.0 – 102.0)	94.0 (88.0 – 140.5)	0.834
QTc duration (ms)	413.0 (403.0 – 426.0)	407.0 (399.3 – 419.8)	417.0 (405.0 – 430.0)	0.005
PVC on any ECG	68 (41.5%)	13 (19.4%)	55 (56.7%)	<0.001
nsVT on any ECG	3 (1.8%)	0 (0.0%)	3 (3.1%)	0.146
Fragmented QRS	24 (14.6%)	9 (13.4%)	15 (15.5%)	0.718
Inferior TWI	61 (36.1%)	21 (30.0%)	40 (40.4%)	0.165
Holter performed	127 (75.1%)	37 (52.9%)	90 (90.9%)	<0.001
VPB burden (%)	2.5 (0.4 – 6.7)	0.2 (0.0 – 1.2)	1.3 (1.2 – 9.6)	<0.001
nsVT on Holter	73 (53.7%)	0 (0.0%)	73 (79.3%)	<0.001
CPET performed	143 (84.6%)	57 (81.4%)	86 (86.9%)	0.334
nsVT on CPET	22 (15.5%)	0 (0.0%)	22 (25.9%)	<0.001

Clinical characteristics and ECG results

Variable	All patients (N=169)	Non-arrhythmic (N=70)	Arrhythmic (N=99)	p-value
MVP extent				
Anterior MVP	5 (3.0%)	1 (1.4%)	4 (4.0%)	0.107
Posterior MVP	74 (43.8%)	37 (52.9%)	37 (37.4%)	
Bileaflet MVP	90 (53.3%)	32 (45.7%)	58 (58.6%)	
Anterior MVP in mm	5.7 (4.0 – 7.6)	6.0 (4.0 – 7.5)	5.2 (4.0 – 7.8)	0.792
Posterior MVP in mm	7.2 (5.2 – 9.6)	6.8 (4.6 – 9.6)	7.7 (6.0 – 9.6)	0.143
MAD	94 (56.0%)	31 (44.3%)	63 (64.3%)	0.010
MAD length (mm)	6.0 (5.0 – 7.9)	6.0 (5.0 – 7.8)	6.0 (5.0 – 8.0)	0.489
LVEF (%)	55.0 (51.3 – 59.3)	56.0 (53.0 – 60.0)	55.0 (49.9 – 59.0)	0.108
LVEF < 50%	33 (19.8%)	9 (13.0%)	24 (24.5%)	0.067
LV GLS	19.9 (17.4 – 23.3)	19.8 (18.3 – 23.4)	19.9 (16.8 – 23.3)	0.399
LV EDV (mL/m ²)	101.3 (85.1 – 113.6)	99.6 (82.6 – 112.8)	102.1 (87.6 – 116.9)	0.179
LV mass (g/m ²)	51.0 (43.0 – 61.6)	52.0 (43.0 – 60.8)	51.0 (43.6 – 62.0)	0.866
RVEF (%)	56.2 (50.3 – 60.0)	57.0 (50.2 – 61.0)	56.0 (50.3 – 60.0)	0.733
RV EDV (mL/m ²)	85.7 (72.8 – 97.6)	83.0 (65.0 – 95.5)	87.9 (76.3 – 98.6)	0.073
TAPSE abs (mm)	25.2 (21.3 – 29.6)	25.7 (21.4 – 29.5)	25.0 (21.1 – 29.8)	0.809
LA area (cm ² /m ²)	12.5 (9.8 – 15.4)	12.5 (9.3 – 15.1)	12.9 (9.9 – 15.5)	0.706
LA enlarged	45 (27.8%)	19 (28.4%)	26 (27.4%)	0.890
MR severity				
None	35 (22.9%)	14 (22.6%)	21 (23.1%)	0.519
Mild (≤1)	43 (28.1%)	21 (33.9%)	22 (24.2%)	
Moderate (≤2)	48 (31.4%)	16 (25.8%)	32 (35.2%)	
Severe (3 or 4)	27 (17.6%)	11 (17.7%)	16 (17.6%)	
LGE present	91 (56.5%)	26 (40.6%)	65 (65.7%)	<0.001
Myocardium	41 (25.5%)	9 (14.1%)	32 (33.0%)	0.007
Papillary muscle	79 (49.4%)	24 (37.5%)	55 (57.3%)	0.014
Anterior	10 (5.9%)	3 (4.3%)	7 (7.1%)	0.364
Posterior	25 (14.8%)	5 (7.1%)	20 (20.2%)	
Both	44 (26.0%)	16 (22.9%)	28 (28.3%)	

Overview of CMR measurements