

Supplemental Material

Table S1. All Cases Included in Study.

Year	Author	Cases	Description
1968	Barlow ¹	1	Review of 90 patients with non-ejection systolic click and late systolic murmurs. 1 case of 39M with SCD. (iMVP)
1970	Trent ²	1	Report of 63F with MVP and SCD. (non-iMVP)
1971	Jeresaty ³	1	Review of 24 patients with mitral ballooning on angiography. 1 case of 62F with SCD. (iMVP)
1973	Jeresaty ⁴	1	Review of 100 patients with non-ejection click or MVP on left ventriculography. 1 case of 44F with SCD. (iMVP) 1 case of 62F with SCD (repeat case).
1973	Shappell ⁵	1	Report of 27F with MVP and SCD. (non-iMVP)
1974	Marshall ⁶	1	Report of 2 cases (27F and 36F) with MVP and SCD. (1 case iMVP and 1 case non-iMVP) Case of 27F (repeat case).
1975	Shappell ⁷	1	Series of 4 patients with MVP. 1 case of 23F with VF. (iMVP) 2 cases (27F and 36F) of SCD (repeat cases). 1 case of NSVT (not included).
1976	Jeresaty ⁸	2	Summary of 12 cases of MVP and SCD. 2 cases (39F and 40M) included. (both iMVP) 7 cases previously reported. 3 cases of personal communication without individual age or gender (not included).
1976	Kleid ⁹	1	Report of 38F with MVP and SCD. (iMVP)
1976	Ritchie ¹⁰	1	Report of 56M with MVP and VF. (non-iMVP)
1976	Winkle ¹¹	5	Series of 7 patients with MVP and VAs. 3 cases with VF and 2 cases with VT. (4 cases iMVP and 1 case non-iMVP) 2 cases excluded (1 with unmonitored cardiac arrest and 1 NSVT).
1977	Cobbs ¹²	1	Report of 39F with MVP and VF. (iMVP)
1977	Mills ¹³	2	Follow-up of 53 patients with MVP. 1 case of 58M with SCD. (non-iMVP) 1 case of 26F with VF. (iMVP)
1978	Davies ¹⁴	13	Review of 90 cases of MVP at autopsy. 13 cases with MVP and SCD (12 cases iMVP and 1 case non-iMVP).
1979	Forbes ¹⁵	1	Report of 25F with MVP and VF on anaesthesia induction. (non-iMVP)
1979	Watts ¹⁶	1	Report of 26F with MVP and VF. (iMVP)
1980	Anderson ¹⁷	2	Report of 2 cases (both 21F) with MVP and SCD. (both iMVP)
1980	Bennett ¹⁸	1	Report of 15F with MVP and TDP. (iMVP)
1980	Mair ¹⁹	3	Series of 3 cases (25F, 29F and 35F) with MVP and SCD. (all iMVP)
1980	Mautner ²⁰	2	Review of 22 patients with MVP and PVCs. 1 case of 51F with VF. (iMVP) 1 case of 50M with VT during anesthesia induction. (iMVP)
1981	Bharati ²¹	1	Report of 45M with MVP and SCD. (iMVP)
1981	Salmela ²²	1	Report of 27M with MVP and SCD. (non-iMVP)
1982	Noneman ²³	1	Report of 29M with MVP and VF. (iMVP)
1982	Vesterby ²⁴	3	Series of 3 cases (23F, 68M, 55M) with MVP and SCD. (1 case iMVP and 2 cases non-iMVP)
1982	Virmani ²⁵	1	Review of 30 autopsies in joggers. 1 case of 27M with MVP and SCD. (iMVP)
1983	Bharati ²⁶	2	Series of 3 cases of SCD in teenagers. 2 cases (17M and 19F) with MVP. (both iMVP)

1983	Chesler ²⁷	14	Series of 14 cases of MVP and SCD. (non-iMVP)
1983	Conklin ²⁸	1	Report of 22F with MVP and VT during labor. (iMVP)
1983	Morady ²⁹	2	Series of 31 patients with VAs undergoing EPS. 2 patients (28F and 39F) with MVP and VF. (both iMVP)
1984	Kempf ³⁰	1	Series of 27 cases with SCD on ambulatory ECG monitoring. 1 case of 31F with MVP. (non-iMVP)
1984	Pocock ³¹	1	Report of 24F with MVP and SCD. (non-iMVP)
1985	Andre-Fouet ³²	1	Report of 19M with MVP and SCD. (iMVP)
1985	Rosenthal ³³	5	Series of 20 patients with MVP and VAs. 5 patients with VF. (all iMVP)
1985	Sakuma ³⁴	1	Report of 54M with MVP, coronary vasospasm and VF. (non-iMVP)
1986	Casthely ³⁵	1	Report of 7M with MVP and VF during anaesthesia induction. (iMVP)
1986	Higgins ³⁶	1	Report of 36F with MVP and VT. (non-iMVP)
1986	Hoffman ³⁷	1	Report of 32F with MVP and VF. (iMVP)
1987	Broustet ³⁸	1	Report of 28F with MVP and SCD. (non-iMVP)
1988	Goldhammer ³⁹	1	Report of 46M with MVP and asystole. (iMVP)
1988	Scala-Barnett ⁴⁰	4	Series of 4 cases of MVP and SCD. (2 cases iMVP and 2 cases non-iMVP)
1988	Strasberg ⁴¹	1	Report of 27M with MVP and VF. (iMVP)
1988	Vlay ⁴²	1	Report of 24F with MVP and SCD. (iMVP)
1989	Abraham ⁴³	1	Report of 33F with MVP and asystole during anesthesia. (iMVP)
1989	Topaz ⁴⁴	2	Series of 22 patients with cardiac arrest. 2 patients [19M (also anomalous RCA) and 28M] with MVP. (1 case iMVP and 1 case non-iMVP)
1989	Martini ⁴⁵	2	Series of 6 cases with VF. 2 cases (14F and 35M) with MVP. (both iMVP)
1990	Boudoulas ⁴⁶	9	Series of 9 patients with MVP and cardiac arrest. (8 cases iMVP and 1 case non-iMVP)
1990	Corrado ⁴⁷	2	Series of 22 athletes with SCD. 2 cases (17F and 23M) with MVP. (both iMVP)
1990	Nelson-Piercy ⁴⁸	1	Report of 67M with MVP, anomalous RCA and VF. (non-iMVP)
1990	Sadaniantz ⁴⁹	0	Report of 27M with MVP and SCD (repeat case).
1991	Dollar ⁵⁰	15	Review of 56 cases of MVP at autopsy. 15 cases of SCD related to MVP. (14 cases iMVP and 1 case non-iMVP)
1993	Vohra ⁵¹	2	Series of 7 patients with MVP and VAs. 2 cases (28M and 45M) with SCD. (both non-iMVP)
1995	Martini ⁵²	1	Report of 42F with MVP, ARVC and SCD. (non-iMVP)
1997	Moritz ⁵³	1	Report of 6F with MVP and VF during anaesthesia induction. (iMVP)
1997	Wilde ⁵⁴	1	Report of 34M with MVP and VF. (iMVP)
1998	Ronneberger ⁵⁵	1	Report of 8M with MVP and SCD. (non-iMVP)
2000	Nolte ⁵⁶	1	Report of 26F with MVP, diffuse CAD due to PXE and SCD. (non-iMVP)
2001	Cannon ⁵⁷	1	Report of 25F with MVP and SCD. (iMVP)
2003	Abello ⁵⁸	1	Report of 28F with MVP and VF during pregnancy. (iMVP)
2003	Ciancarmerla ⁵⁹	1	Report of 49M with MVP and SCD. (iMVP)
2003	Nishida ⁶⁰	1	Series of 3 cases of SCD and alcohol abuse. 1 case of 37F with MVP. (non-iMVP)
2004	Chirachariyavej ⁶¹	1	Report of 38M with MVP and SCD. (iMVP)
2004	Frassati ⁶²	3	Series of 14 cases of SCD in psychiatric patients. 3 cases (22M, 51M and 57M) with MVP. (1 case iMVP and 2 cases non-iMVP)
2005	Zeidan ⁶³	1	Report of 21F with MVP and VF during anaesthesia reversal. (iMVP)
2007	Anders ⁶⁴	6	Series of 6 cases of MVP and SCD. (iMVP)
2007	Kesavan ⁶⁵	1	Report of 75F with MVP, CAD and VT. (non-iMVP)

2007	Knackstedt ⁶⁶	1	Report of a 54M with MVP and VF. (iMVP)
2010	Franchitto ⁶⁷	1	Report of 25F with MVP and SCD. (iMVP)
2010	Oliviera ⁶⁸	1	Report of 57F with MVP and SCD (also heart failure on trastuzumab). (non-iMVP)
2011	Rordorf ⁶⁹	1	Report of 32F with MVP and VF (also DCM post-partum with PJRT). (non-iMVP)
2014	Abbadj ⁷⁰	1	Report of 26F with MVP and VF. (iMVP)
2014	Rajani ⁷¹	1	Report of 27F with MVP and TDP. (iMVP)
2015	Lin ⁷²	1	Report of 30F with MVP and VT during pregnancy. (iMVP)
2015	Desai ⁷³	1	Report of 55M with MVP and SCD. (iMVP)
2015	Fais ⁷⁴	1	Report of 47F with MVP and SCD. (iMVP)
2016	Ahmed ⁷⁵	1	Report of 45M with MVP and VT. (iMVP)
2016	Vaidya ⁷⁶	5	Series of 5 patients with MVP, ICD and history of MV surgery (1 case also had HCM). (2 cases iMVP and 3 cases non-iMVP)
2017	Cacko ⁷⁷	1	Report of 28F with MVP and VF. (iMVP)
2017	Martini ⁷⁸	1	Report of 58M with MVP and VF (also Brugada ECG). (non-iMVP)
2017	Saha ⁷⁹	1	Report of 26F with MVP and VF during pregnancy. (iMVP)

SCD, sudden cardiac death; MVP, mitral valve prolapse; VF, ventricular fibrillation; NSVT, non-sustained ventricular tachycardia; VAs, ventricular arrhythmias; EPS, electrophysiology study; VT, ventricular tachycardia; TDP, torsade de pointes; PVCs, premature ventricular complexes; RCA, right coronary artery; ARVC, arrhythmogenic right ventricular cardiomyopathy; PXE, pseudoxanthoma elasticum; CAD, coronary artery disease; DCM, dilated cardiomyopathy; PJRT, persistent junctional reciprocating tachycardia, ICD, implantable cardiac defibrillator; MV, mitral valve; HCM, hypertrophic cardiomyopathy

Table S2. Predictors of Ventricular Arrhythmias or Sudden Cardiac Death.

	Author	Year	Study	N (% Female)	Age range	Study population	Diagnostic criteria	Predictor/association	Outcome/Endpoint
Clinical									
	Gaffney ⁸⁰	1979	Prospective Cohort	19 (100)*	19-46	MVP	M-mode or auscultation	Higher heart rate Lower cardiac index	Clinical severity (combination of symptoms and VAs)
	Puddu ⁸¹	1983	Prospective Cohort	15 (67)	NR	MVP	Echo (NS)	Plasma catecholamine level	QTc (supine)
	Snieszek ⁸²	1992	Prospective Cohort	53 (58)	19-52	MVP	Echo (LAX)	Adrenaline excretion	Complex VAs (Lown grade ≥3)
	Zuppiroli ⁸³	1994	Prospective Cohort	119 (47)	12-78	MVP	Echo (LAX)	Female	Complex VAs (Lown grade ≥3) [†]
	Babuty ⁸⁴	1994	Prospective Cohort	58 (50)	NR	MVP	Echo (LAX or A4C)	Age (older)	Complex VAs (Lown grade ≥3)
	Naksuk ⁸⁵	2016	Retrospective Cohort	32 (53)	NR	BiMVP with MV surgery	N/A	Age (younger)	Reduction in PVCs post MVR in BiMVP
	Fulton ⁸⁶	2017	Retrospective Cohort	18 (61)	NR	MVP	Echo (LAX)	Female	PVCs from PM
Electrical									
	Campbell ⁸⁷	1976	Prospective Cohort	20 (65)	12-61	MVP	Auscultation	Inferolateral T-wave changes	VT (>100bpm for 3 beats) or VF changes
	Babuty ⁸⁴	1994	Prospective Cohort	58 (50)	NR	MVP	Echo (LAX or A4C)	Late potentials	Non-sustained VT (≥3 beats and <30 seconds)
	Bobkowski ⁸⁸	2002	Prospective Cohort	151 (77)*	5-18	MVP	Echo (NS)	Late potentials	VAs (Lown grade ≥1) Non-sustained VT (>120bpm for ≥4 beats and <30 seconds)
	Akcay ⁸⁹	2010	Retrospective Case control	60 (72)	NR	MVP (with vs without VT)	Echo (NS)	QTc dispersion	VT (>120bpm for ≥3 beats) [†]
Imaging									
	Shah ⁹⁰	1982	Retrospective Cohort	88 (60)	12-84	MVP	M-mode	MR	Complex VAs (Lown grade ≥3)
	Nishimura ⁹¹	1985	Prospective Cohort	237 (60)	10-69	MVP	Echo (NS)	Redundant leaflets	Sudden death [†]
	Kligfield ⁹²	1985	Prospective Cohort	80 (65)*	19-72	MVP	Echo (NS)	MR	>1% PVC frequency Exercise induced PVCs and VT

									Complex VAs (Lown grade 4)
	Sanfilippo ⁹³	1989	Retrospective Cohort	22 (55)*	NR	MVP	Echo (LAX or A4C)	Anterior leaflet thickness MR Leaflet displacement	VAs (≥ 10 PVCs/hr or NSVT at ≥ 100 bpm for ≥ 3 beats)
	Zuppiroli ⁸³	1994	Prospective Cohort	119 (47)	12-78	MVP	Echo (LAX)	Anterior leaflet thickness	Complex VAs (Lown grade ≥ 3)†
	Babuty ⁸⁴	1994	Prospective Cohort	58 (50)	NR	MVP	Echo (LAX or A4C)	MR	Complex VAs (Lown grade ≥ 3) on Holter and exercise test
	Zouridakis ⁹⁴	2001	Prospective Cohort	89 (71)	NR	MVP	Echo (LAX or A4C)	MVP degree Anterior leaflet thickness	QT dispersion†
	Turker ⁹⁵	2010	Prospective Cohort	58 (55)	16-68	MVP	Echo (LAX)	Moderate-severe MR	VAs (Lown grade ≥ 1)†
	Carmo ⁹⁶	2010	Retrospective Cohort	38 (47)	NR	MVP	Echo (LAX)	Mitral annular disjunction	Non-sustained VT
	Han ⁹⁷	2010	Retrospective Cohort	16 (44)*	NR	MVP	Echo (NS)	LGE in PM	Complex VAs (Lown grade ≥ 4)
	Akcay ⁸⁹	2010	Retrospective Case control	60 (72)	NR	MVP (with vs without VT)	Echo (NS)	Anterior leaflet length	VT (>120 bpm for ≥ 3 beats)†
	Sriram ⁹⁸	2013	Retrospective Cohort	24 (67)	5-60	Idiopathic OHCA	Echo (NS)	BiMVP	Appropriate ICD therapies at follow-up
	Basso ⁹⁹	2015	Prospective Cohort	44 (66)	24-64	MVP	Echo (LAX)	LGE (PM, inferobasal wall and total percentage)	Complex VA (Lown grade ≥ 4 b or VF)
	Nordhues ¹⁰⁰	2016	Retrospective Case control	11338 (43)*	NR	BiMVP vs SiMVP	Echo (NS)	BiMVP	All-cause mortality (lower in BiMVP)
	Bui ¹⁰¹	2017	Retrospective Cohort	32 (34)*	NR	MVP	CMR	Myocardial T1 time	Complex VAs (Lown grade ≥ 3)
	Fulton ⁸⁶	2017	Retrospective Cohort	18 (61)	NR	MVP	Echo or CMR	BiMVP LGE in PM	PVCs from PM

A4C, apical 4 chamber; bpm, beats per minute; BiMVP, bileaflet MVP; ICD, implantable cardiac defibrillator; LAX, long axis; LGE, late gadolinium enhancement; NR, not reported; NS, not specified; OHCA, out of hospital cardiac arrest; PM, papillary muscle; SiMVP, single leaflet MVP.

*Studies also included normal control groups which are not presented

†Significant result on multivariate analysis; significant univariable predictors not presented

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