Supplementary files

„Clinical characteristics of patients with arrhythmic mitral valve prolapse in a single tertiary center: prevalence of electrocardiographic and myocardial abnormalities"

Echocardiography

All participants underwent echocardiography using a *GE VIVID9* ultrasound system (*GE Ultrasound, Horten, Norway*) equipped with a phased-array transducer (M5S). Standard echocardiographic parameters were obtained according to the recommendations [1]. All echocardiograms were digitally stored and analyzed offline using a commercial workstation (*EchoPAC version 201; GE Healthcare Horten, Norway*). Left atrial (LA) and LV volumes were measured using biplane methods as averaged values from four- and two-chamber views. LA volume was indexed by body surface area. LVEF was measured using biplane Simpson's method [1].

MVP was defined as superior displacement >2 mm of any part of the mitral leaflet beyond the mitral annulus according to the American Society of Echocardiography guidelines [2]. MR was quantified according to guidelines [2,3]. MAD distance was measured from the LA wall - mitral valve leaflet junction to the top of the LV wall during end-systole in the parasternal long-axis view and was defined as the longitudinal MAD distance in the posterolateral wall [4]. The circumferential extent of MAD was not assessed. Curling was defined as an unusual systolic motion of the posterior mitral ring on the adjacent myocardium and assessed quantitatively in millimeters by tracing a line between the top of LV inferobasal wall and the LA wall–posterior MV leaflet junction and from this line a perpendicular line to the lower limit of the mitral annulus [5]. Echocardiographic analyses were performed by two echocardiography experts blinded to all clinical data.

ECG

Complex VA were defined as a history of ventricular fibrillation (VF) and/or the presence of multiform premature ventricular complexes (PVC), ventricular couplets, ventricular bigeminy, non-sus-

tained (≥3 consecutive ventricular beats lasting <30 s with heart rate >100 beats/min; VTns), or sustained ventricular tachycardia (VTs). Data on VA were collected from Holter ECG recordings, exercise test ECGs, cardiac devices, telemetry, and medical records.

CMR imaging

CMR was performed on a 1.5T scanner (*Aera, Siemens, Erlangen, Germany*) or 3T scanner (*Achieva, Philips Healthcare, Best, the Netherlands*). On CMR, a series of morphofunctional parameters were carefully evaluated. Standard volumes and function parameters were measured based on long-axis cine, and a short-axis cine stack covering the entire LV. MAD was defined as a separation between the LA-valve junction and the atrial margin of the LV free wall. Hypertrabeculation was recognized based on Petersen's criterion i.e. non-compacted [NC] to compacted [C] myocardial thickness ratio of 2.3 or more in at least one segment in end-diastolic long axis cine images [6]. The presence of LGE was visually assessed independently by two experienced readers based on the 17 segment model [7]. Any discrepancy was solved by consensus. CMR was not performed in patients with an implanted ICD.

References

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4. Dejgaard LA, Skjølsvik ET, Lie ØH, et al. The Mitral Annulus Disjunction Arrhythmic Syndrome. J Am Coll Cardiol. 2018; 72: 1600–1609.

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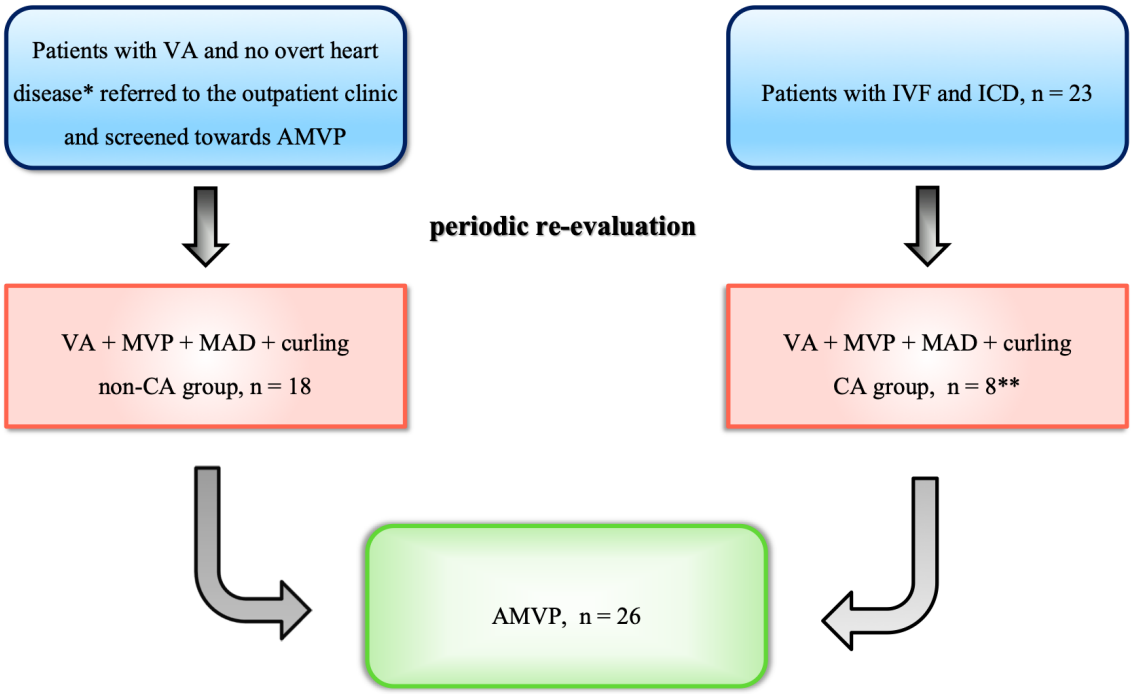
6. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. J Am Coll Cardiol. 2005; 46: 101–5.
7. Cerqueira MD, Weissman NJ, Dilsizian V, et al. Standardized myocardial segmentation and nomenclature for tomographic imaging of the heart: a statement from the Cardiac Imaging Committee of the Council on Clinical Cardiology of the American Heart Association. Circulation. 2002; 105: 539–42.

Figure S1. Flowchart of the study patients



*Except for MVP; **The patient with symptomatic polymorphic VT/VF and DDD pacemaker was also included in this group.

Abbreviations: AMVP, arrhythmic mitral valve prolapse; CA, cardiac arrest; ICD, implantable cardioverter-defibrillator; IVF, idiopathic ventricular fibrillation; MAD, mitral annular disjunction; MVP, mitral valve prolapse; VA, ventricular arrhythmias.

Figure S2. (A) Demographic of the patients (age and sex) included in the study; (B) Age of first presentation to a cardiologist.

Abbreviations: CA, cardiac arrest; VA, ventricular arrhythmia; VF, ventricular fibrillation.

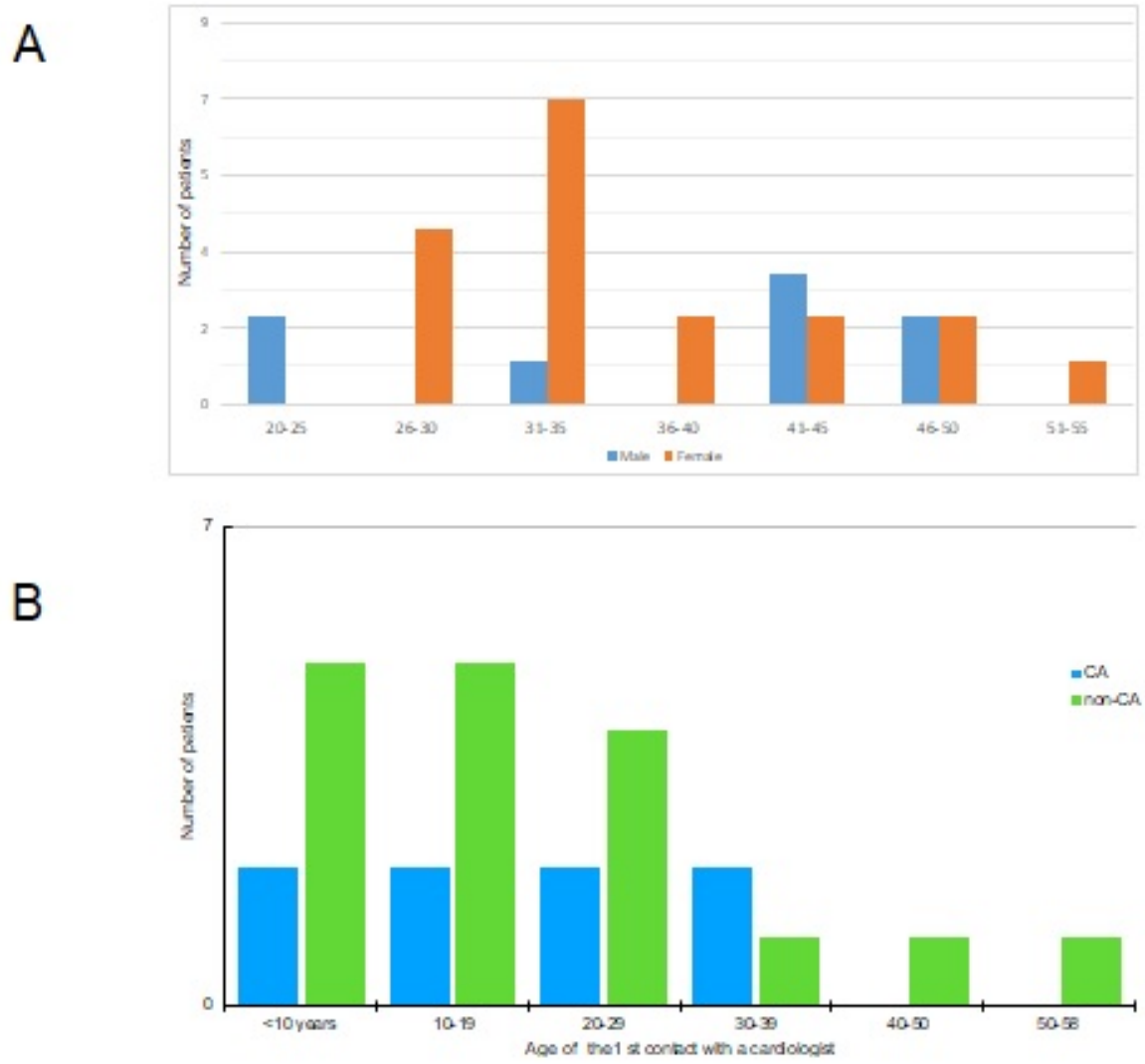
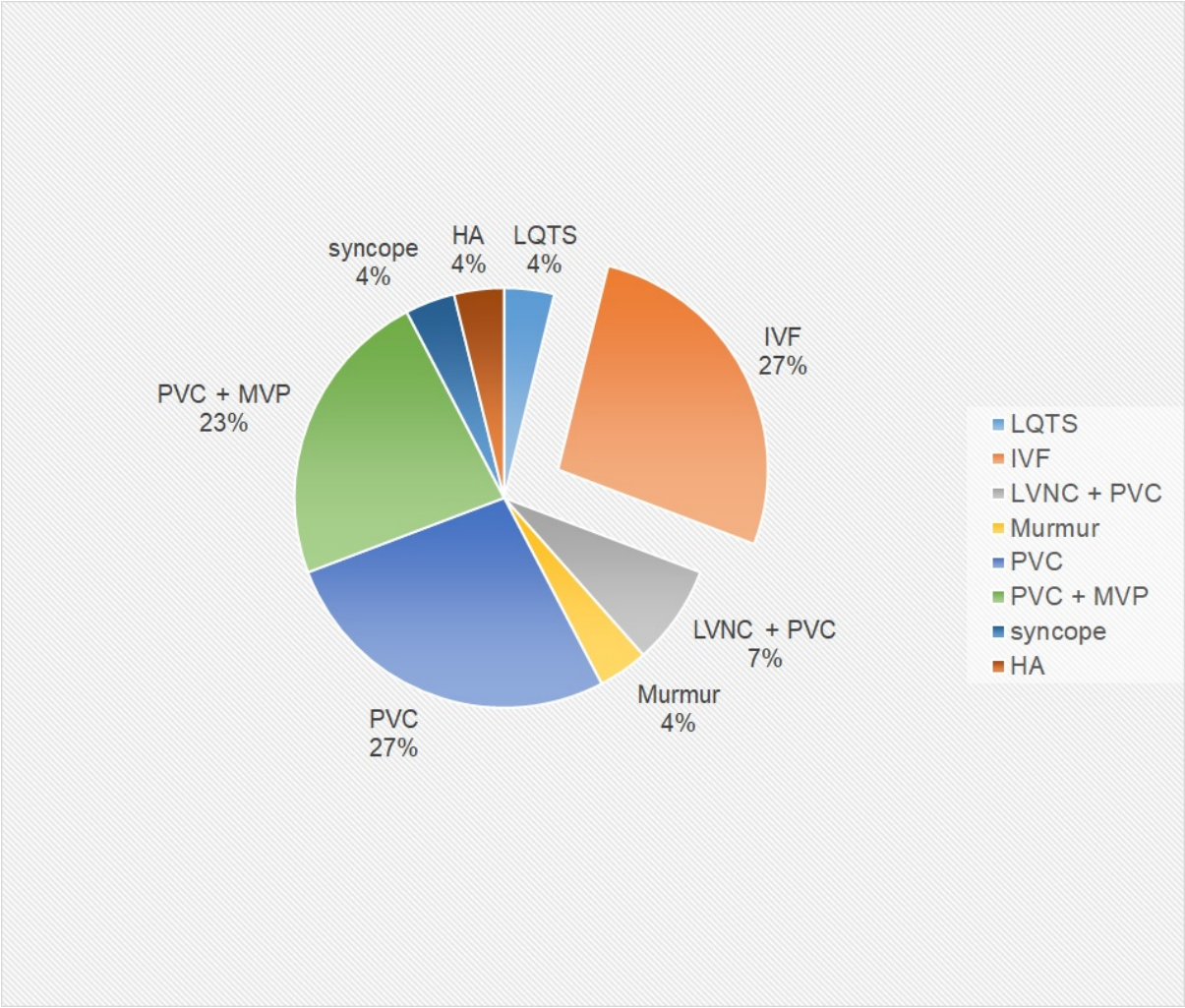


Figure S3. Referral indications/primary diagnoses in the study patients (the percentages do not add up to 100).

Abbreviations: HA, arterial hypertension; IVF, idiopathic ventricular fibrillation; LQTS, long QT syndrome; LVNC, left ventricular non-compaction; MVP, mitral valve prolapse; PVC, premature ventricular contraction.



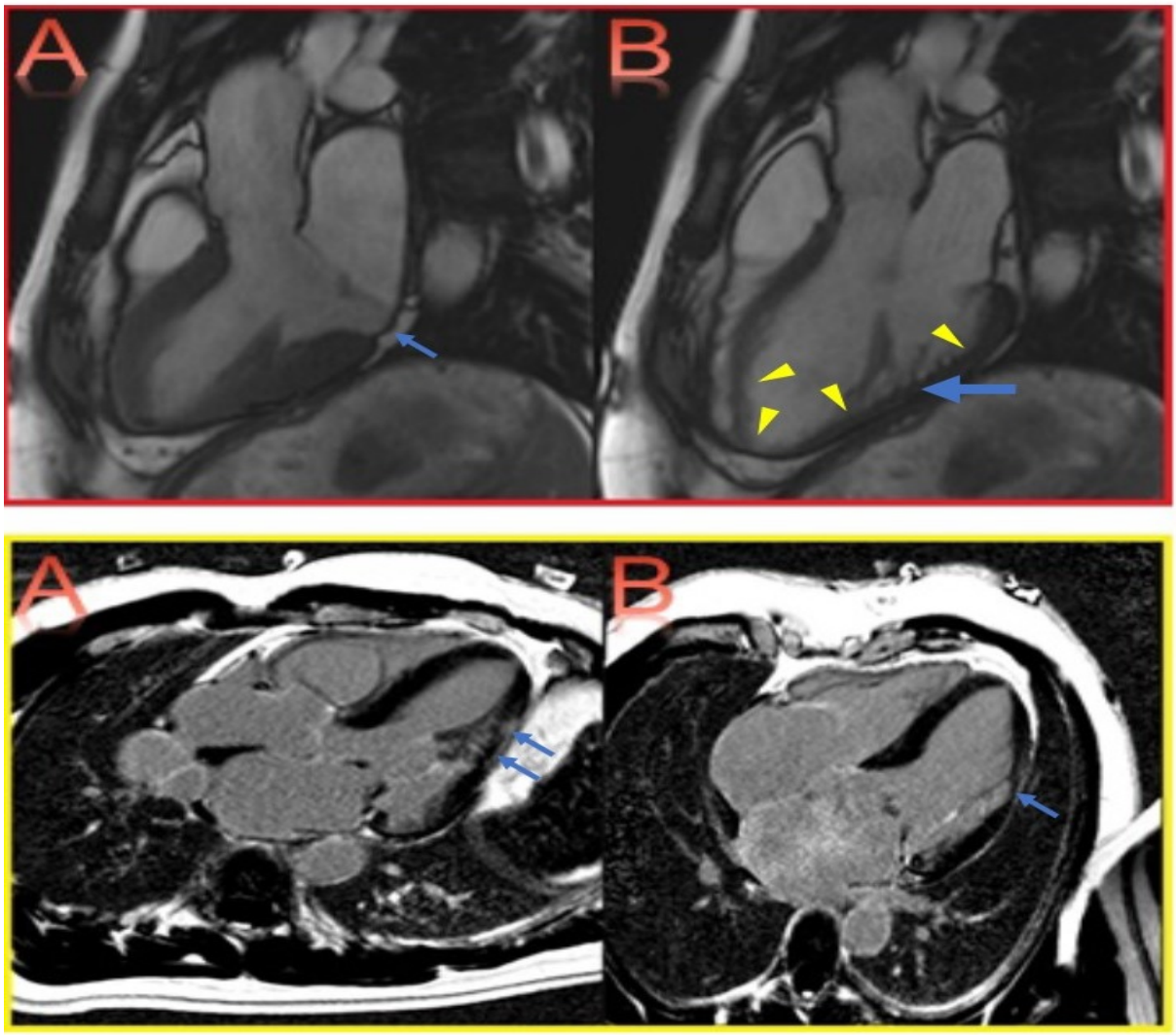
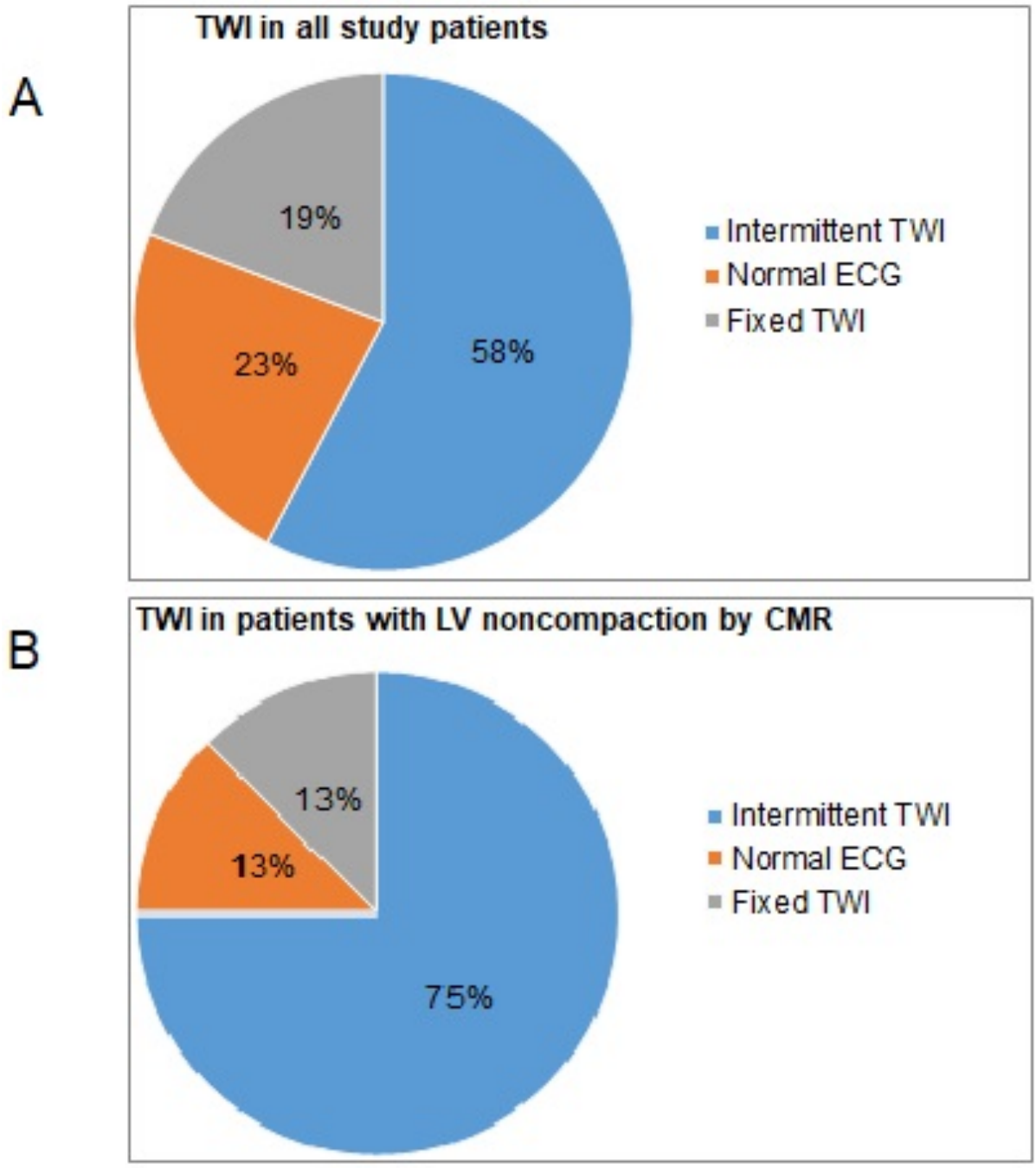


Figure S4. Red frame: Long axis (3CH) systolic (A) and long axis (3CH) diastolic (B) still frames from routine balanced steady state free precession cine sequence, showing mitral annulus disjunction (blue arrow) , relative thinning of compacted layer in several LV segments (arrowheads) and fragmented base of the PM (thick arrow).

Yellow frame: Typical example of late gadolinium enhancement (LGE) extent and distribution in AMVP patients. Minimal, diffuse subendocardial / intramural LGE in the mid-infero-lateral segment, involving the trabeculations and/ or fragmented PM base (blue arrows). (Siemens Aera, 1,5T Erlangen, Germany).

Abbreviations: AMVP, arrhythmic mitral valve prolapse; LV, left ventricle.

Figure S5. Variability of inferior TWI in (A) all study patients, (B) patients with LV non-compaction.



Abbreviations: LV, left ventricle; TWI, T-wave inversion.