

Figure S1: Instrumental findings in Proband IV-4 of the Family A. **A** At ECG sinus rhythm with 65 bpm, mild right ventricular conduction delay and negative T wave V1-V3 were present. **B** Cardiac magnetic resonance showing a mild right ventricular dilation with kinetic abnormality of the anterior wall. **C** SAECG showing presence of late potentials at 40 Hz filter.

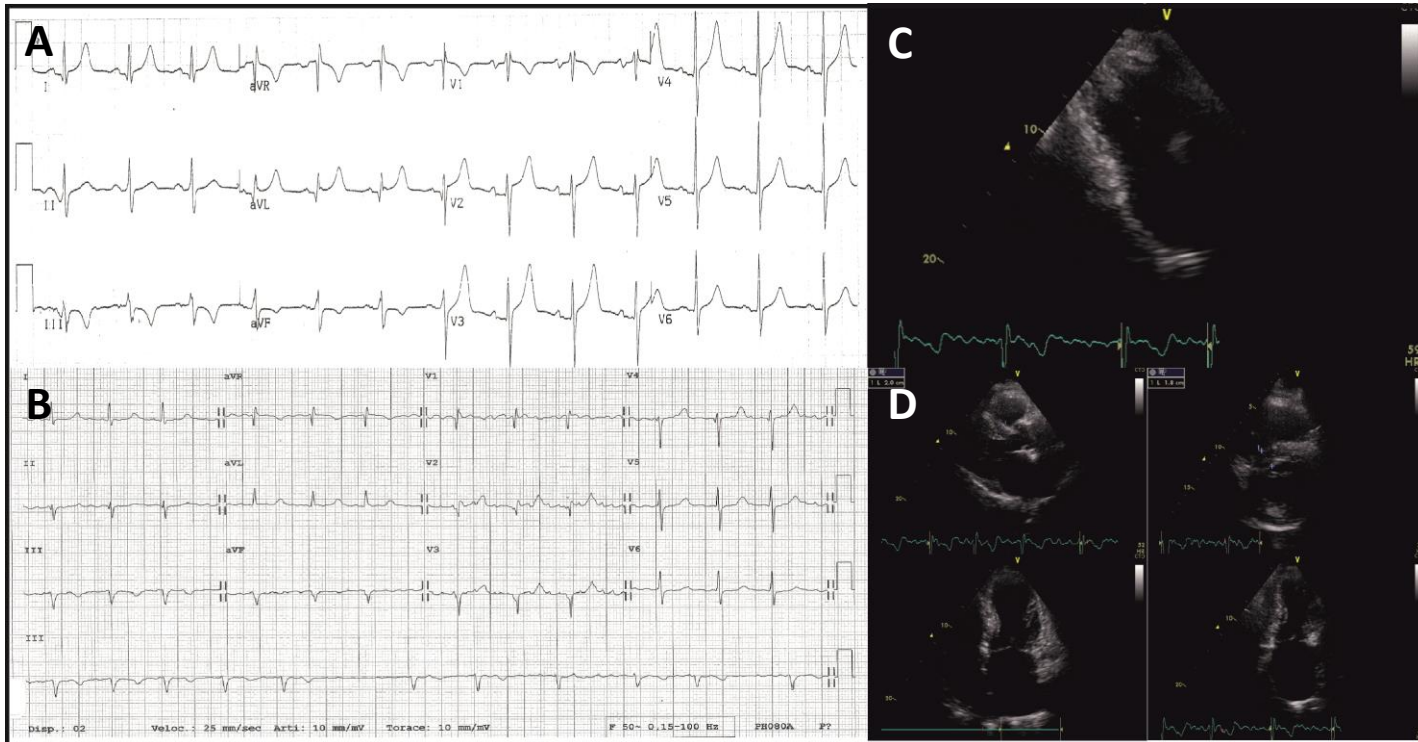


Figure S2: Instrumental findings in Patient III-6 of the Family A. **A** First ECG showing sinus rhythm, inferior negative T waves and a slight right intraventricular delay. **B** During follow-up ECG changed showing left axis deviation and septal pseudonecrosis (absence of R waves in V1-V3). **C** Echocardiogram of the right ventricle (RV) indicating normal dimensions, wall thickening, hypokinesia of the RV basal to mid free wall and dilated atria. **D** Echocardiogram focused on left ventricle (LV) revealed an asymmetric hypertrophy with a maximum ventricular thickness of 18mm.

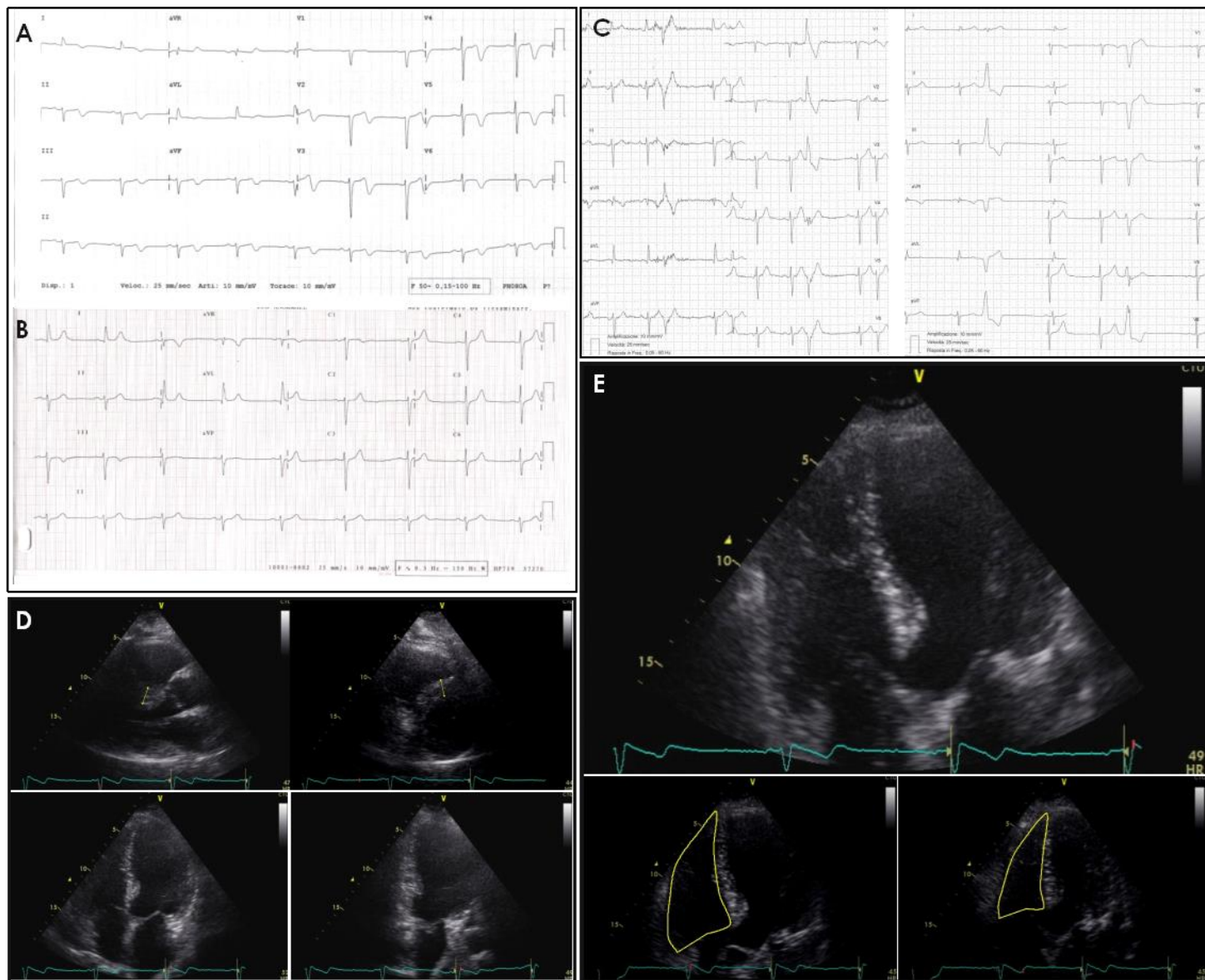


Figure S3: Instrumental findings in Patient III-4 of the Family A. **A** First ECG showing left axis deviation and repolarization abnormalities. **B** Last ECG showing persistent left axis deviation but absence of repolarization abnormalities. **C** 24-hours Holter revealing sporadic isolated polymorphic PVCs both from LV and RV. **D** Echocardiogram identifying an asymmetric septal wall thickening. **E** Echocardiogram showing RV mild dilation and increase trabeculation with a normal regional and global systolic function.