

Introduction

- Mitral annular disjunction (MAD) is an under-reported cause of sudden cardiac death
- MAD is the aberrant attachment of the posterior leaflet of the mitral valve (MV) directly onto the left atrial wall, with a demarcation between the MV attachment and the atrium-MV junction¹
- Here we present a case of a woman with cardiac arrest of unknown origin, later found to have MAD.

Case description

- A 59-year-old woman with history of MV prolapse and mitral regurgitation sustained a ventricular fibrillation arrest in the field
- She was treated with DC cardioversion, epinephrine, and lidocaine with ROSC after 45 minutes
- She was hospitalized and required repeat DC cardioversion for recurrent ventricular fibrillation
- Physical Exam: Grade 2 Systolic Murmur (Loudest at Apex)
- EKG: Sinus Rhythm with Prolonged QT (550ms) and 2mm ST depressions on anteroseptal leads
- Transthoracic Echo: LVEF 15-20% with global hypokinesis
- Cardiac catheterization: Normal coronary artery anatomy and hemodynamics
- EKG (4d s/p arrest): NSR w/normal QT (400ms) and no ST depressions
- TTE (5d s/p arrest): Improved LVEF 55% and MAD
- Cardiac MRI (CMRI): Localized separation of the MV annulus with 10mm migration of the annulus to the left atrial wall, diagnostic for MAD. Dyskinesia of the posterolateral attachment of the MV leaflet to the left atrium seen on MRI was thought to cause her sudden cardiac arrest
- Amiodarone was initiated for ectopy and a single chamber ICD implanted for secondary prevention of SCD.

Figure

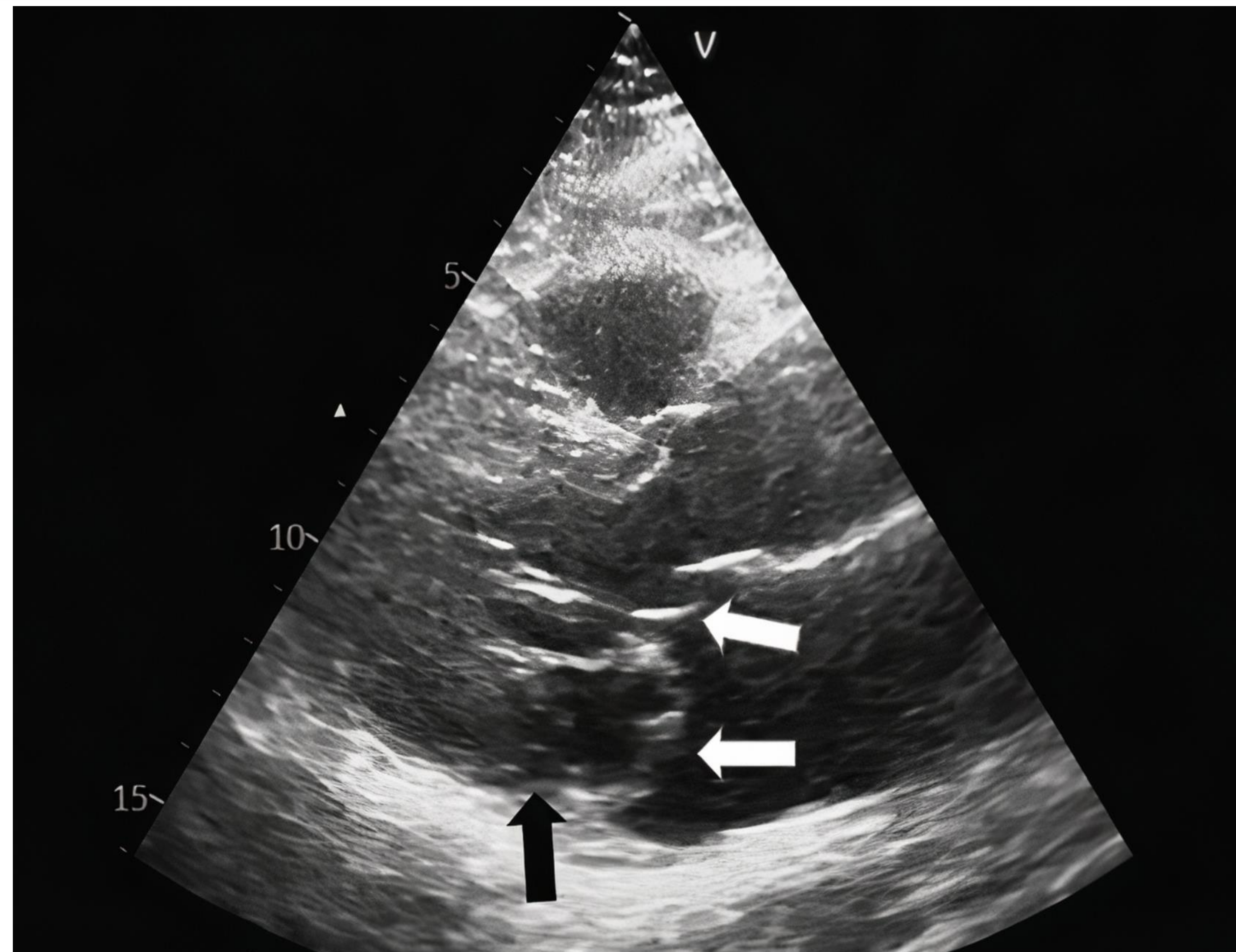


Figure 1 : Transthoracic Echocardiogram: Parasternal long-axis view of the left ventricle during systole. Elongated and thickened mitral valve leaflets prolapsing into the left atrium (white arrows). Disjunction area (black arrows)⁴

Discussion

- MAD is an uncommon cause of SCD, particularly in young women²
- 1/10 of MAD patients experience potentially fatal arrhythmias
- In this case, MAD causing SCD was evident after performing a repeat echocardiogram. Initial LV dysfunction, ST depressions, and prolonged QT likely occurred due to myocardial “stunning” from cardiac arrest.
- For higher-risk patients, further CMRI findings of MAD >8.5 mm or left ventricular scarring may be relevant in identifying patients requiring aggressive therapy²
- The role of ICD for primary prevention in this population is ill-defined. Electrophysiology studies may guide risk assessment, but there is insufficient data to support primary preventive ICD in other specific demographics or official risk calculators³
- Further research is needed to investigate to assess primary prevention ICDs for malignant ventricular arrhythmias and SCD in MAD

Conclusion

- This case report highlights the importance of keeping the uncommon diagnosis of MAD in mind when considering differential diagnoses for causes of SCD in women with MVP
- Furthermore, it poses the question of SCD primary prevention in MAD

References

