

The Gene-sis of Heart Failure

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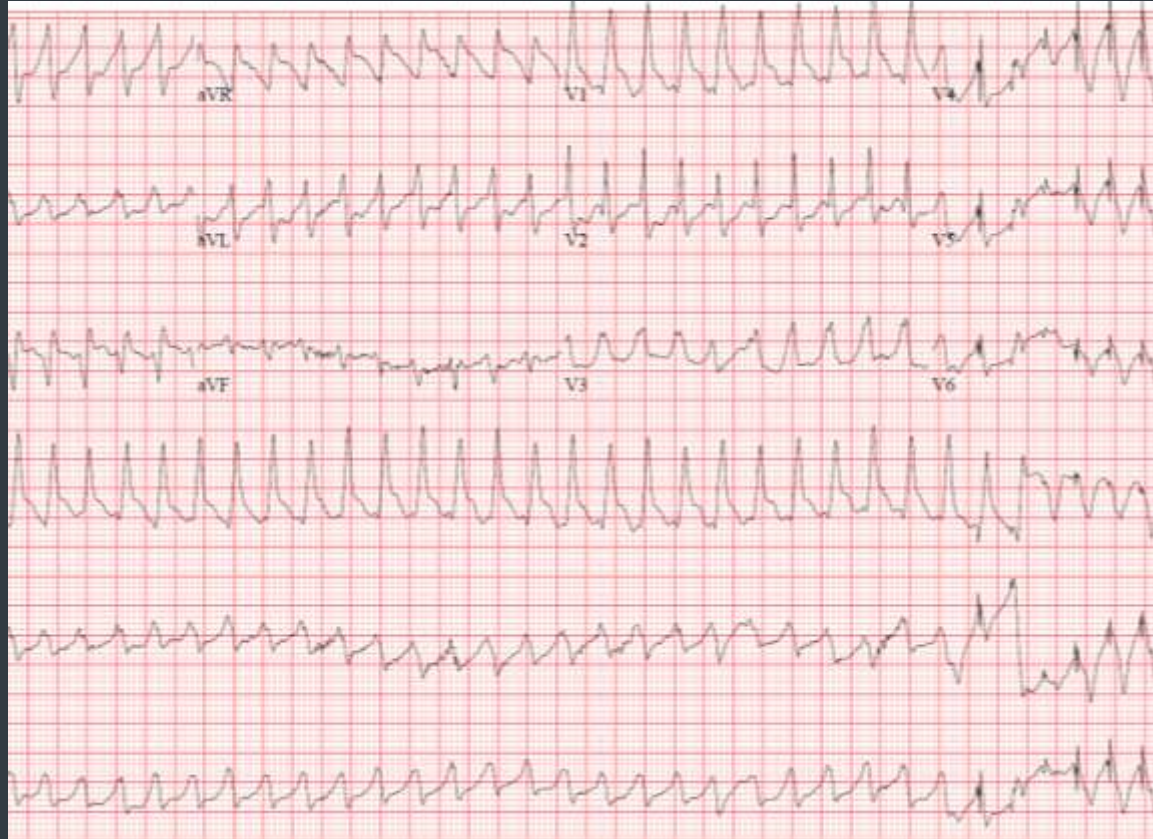
Disclosures

- Nothing to disclose



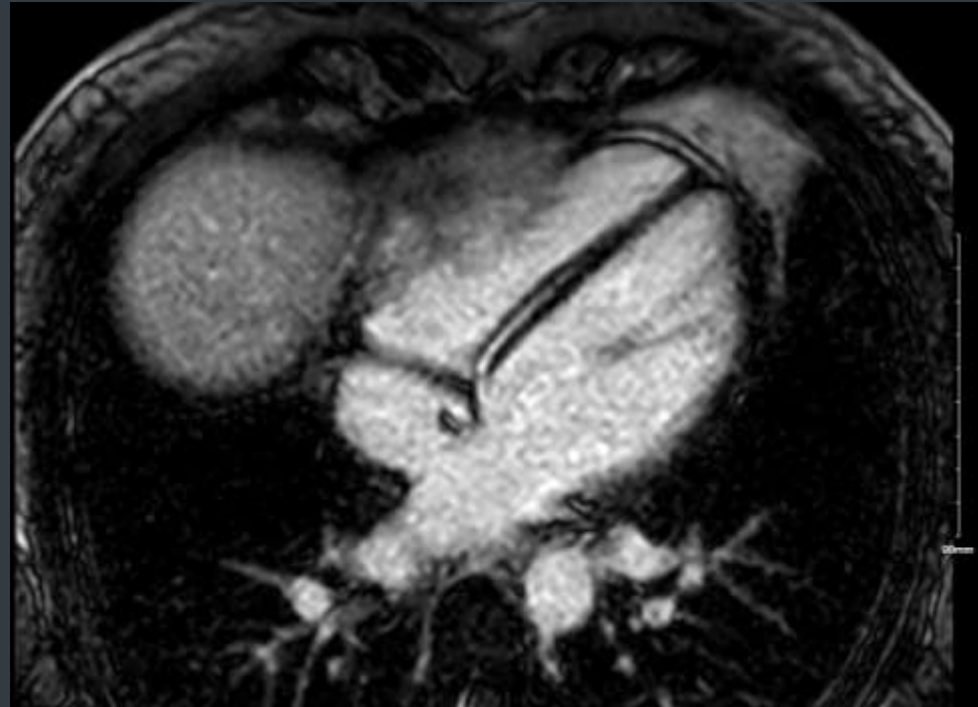
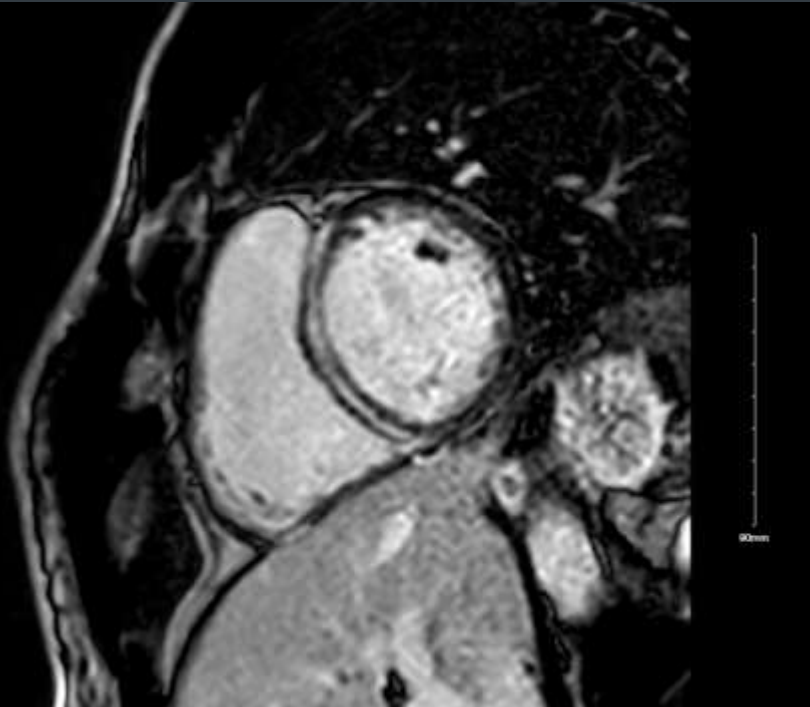
A 38-year-old male..

- Abrupt onset of severe lightheadedness
- In the ER, noted to have a HR 250 bpm
- Shock First, Talk Later



A 38-year-old male..

- Underwent cardiac MRI

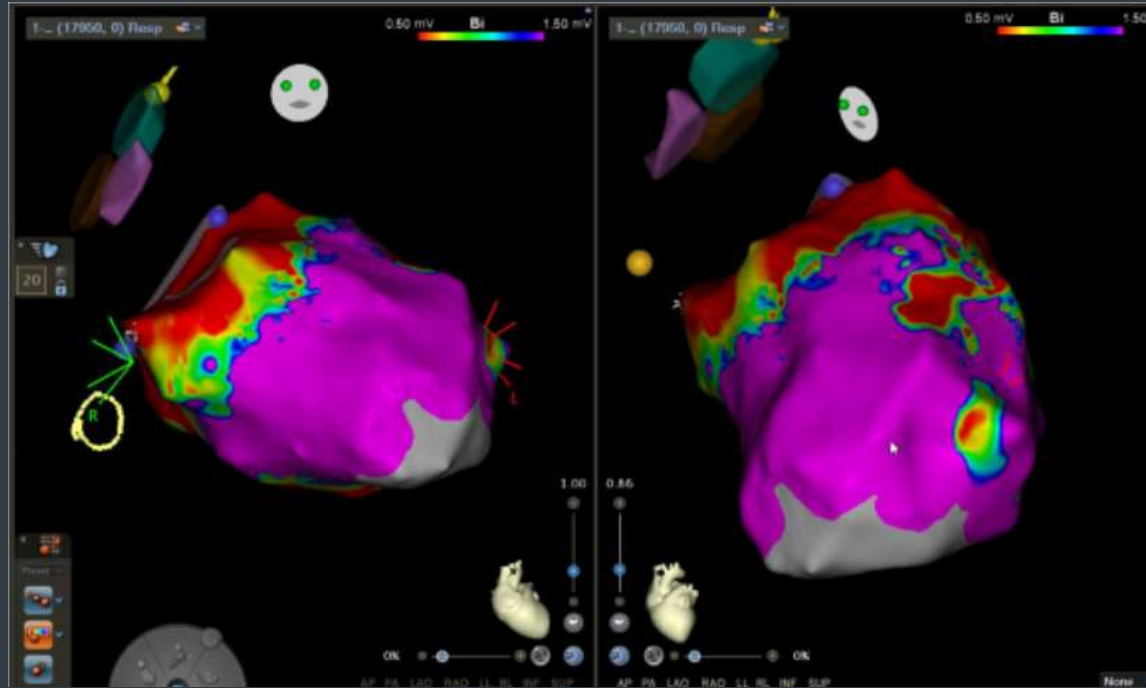


Reduced LVEF of 49%; late Gadolinium enhancement involving the base of the heart



A 38-year-old male..

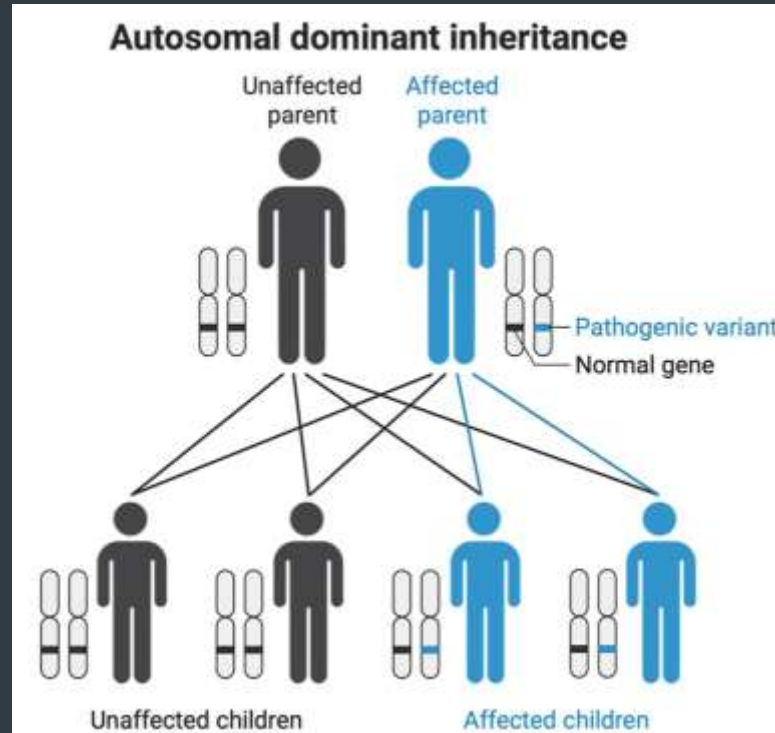
- **One year later**, had sudden onset palpitations → 2 ICD shocks
- Underwent VT ablation
- **One week later**, presented to the ER with Ventricular Tachycardia



Lamin A/C Cardiomyopathy

Autosomal Dominant

- ❖ Lamin A and C are structural proteins in cells



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- Atrial and ventricular arrhythmias
- Dilated Cardiomyopathy
- AV Blocks

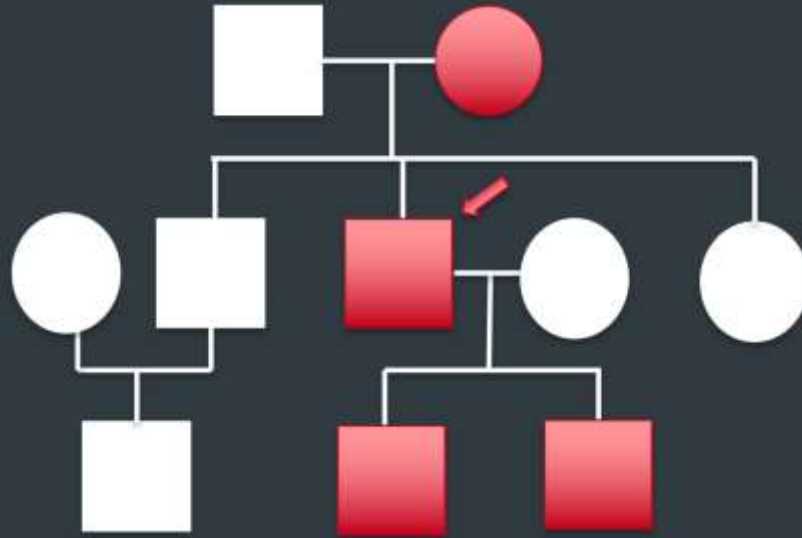


- Muscular dystrophy
- Presents after 2nd decade of life
- Hutchinson–Gilford progeria syndrome; rare



Lamin A/C Cardiomyopathy

- Identification of Proband and Cascade testing is key



- There are no targeted therapies



What happened next?

Admitted with Sustained Ventricular tachycardia

➤ Refractory VT despite 2+ antiarrhythmics

➤ Listed for Heart Transplant

➤ Post-op Cardiogenic shock

➤ VA ECMO + Impella for 48 hours

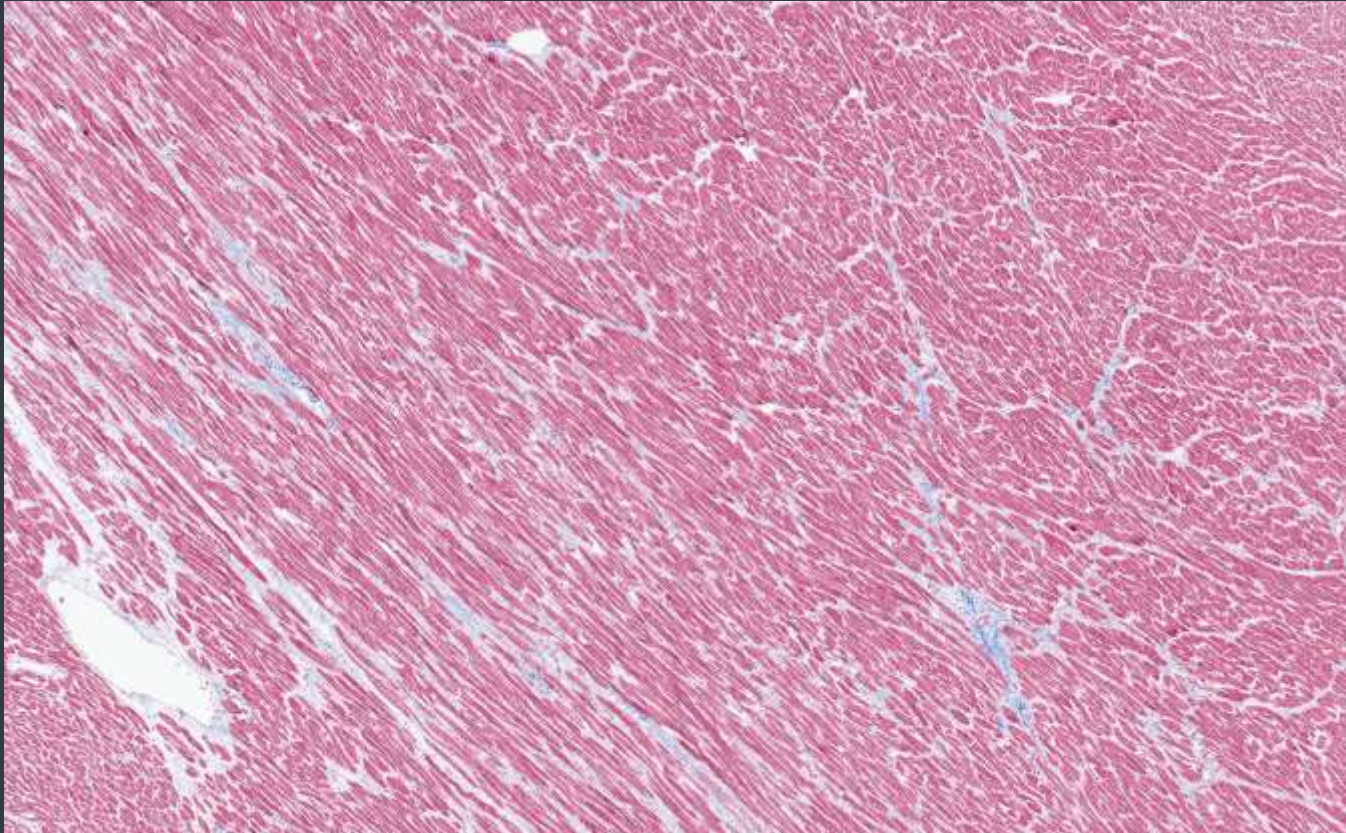
➤ Immunosuppression, Inpatient PT/OT



Native Heart- Gross specimen



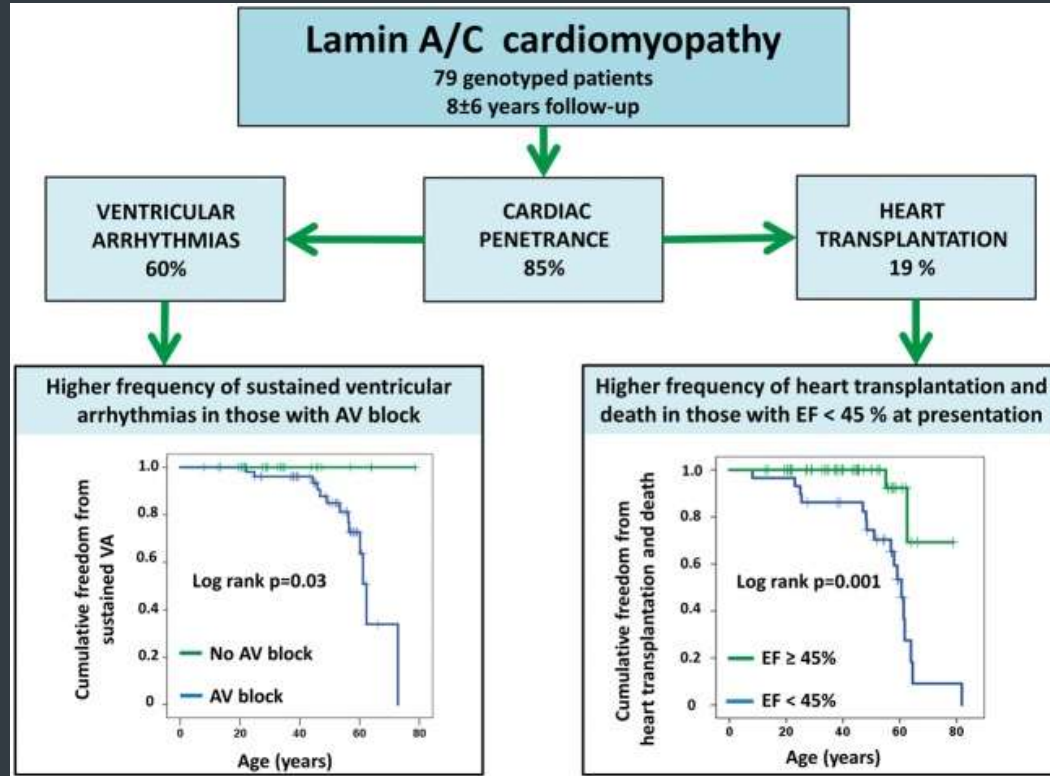
Histology



Mild diffuse interstitial fibrosis in the left ventricle (trichrome stain)



Lamin A/C Cardiomyopathy



Hasselberg NE et al. Lamin A/C cardiomyopathy: young onset, high penetrance, and frequent need for heart transplantation. *Eur Heart J.* 2018



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