68-year-old male with a 1-month history of dyspnea on exertion

Brandon Byrd, MS3

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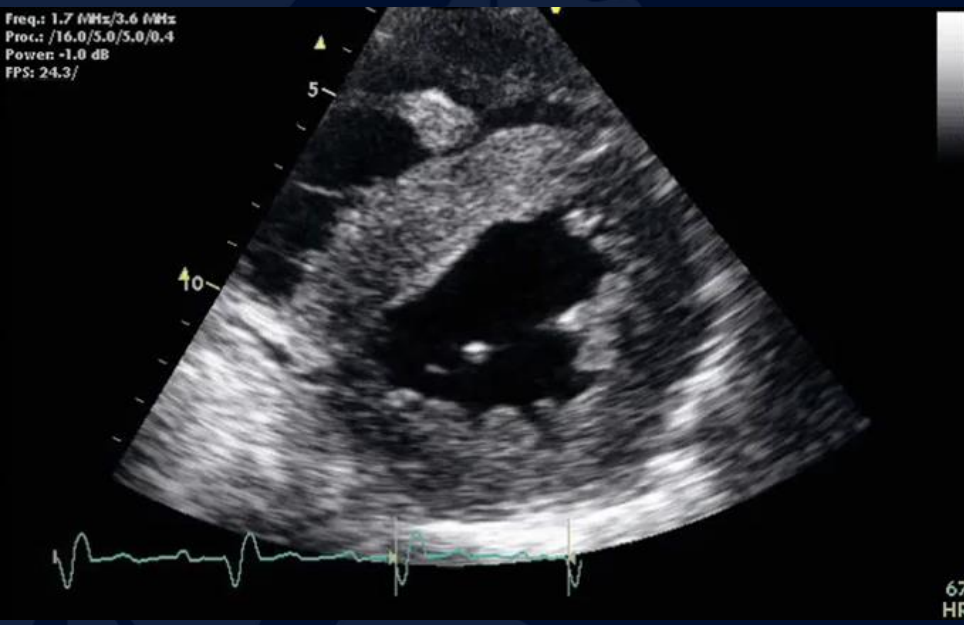
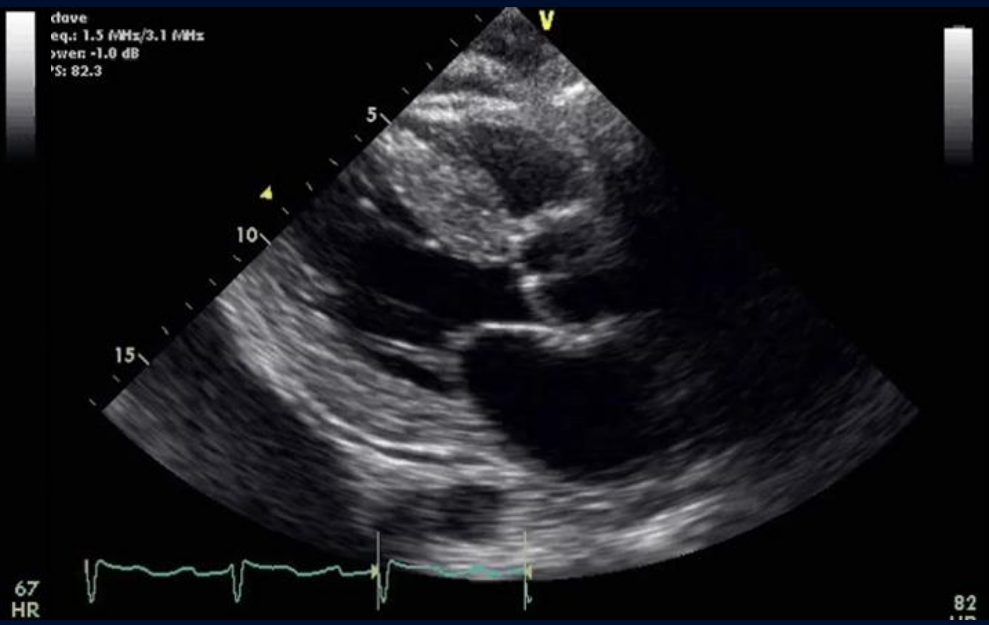
Echocardiogram





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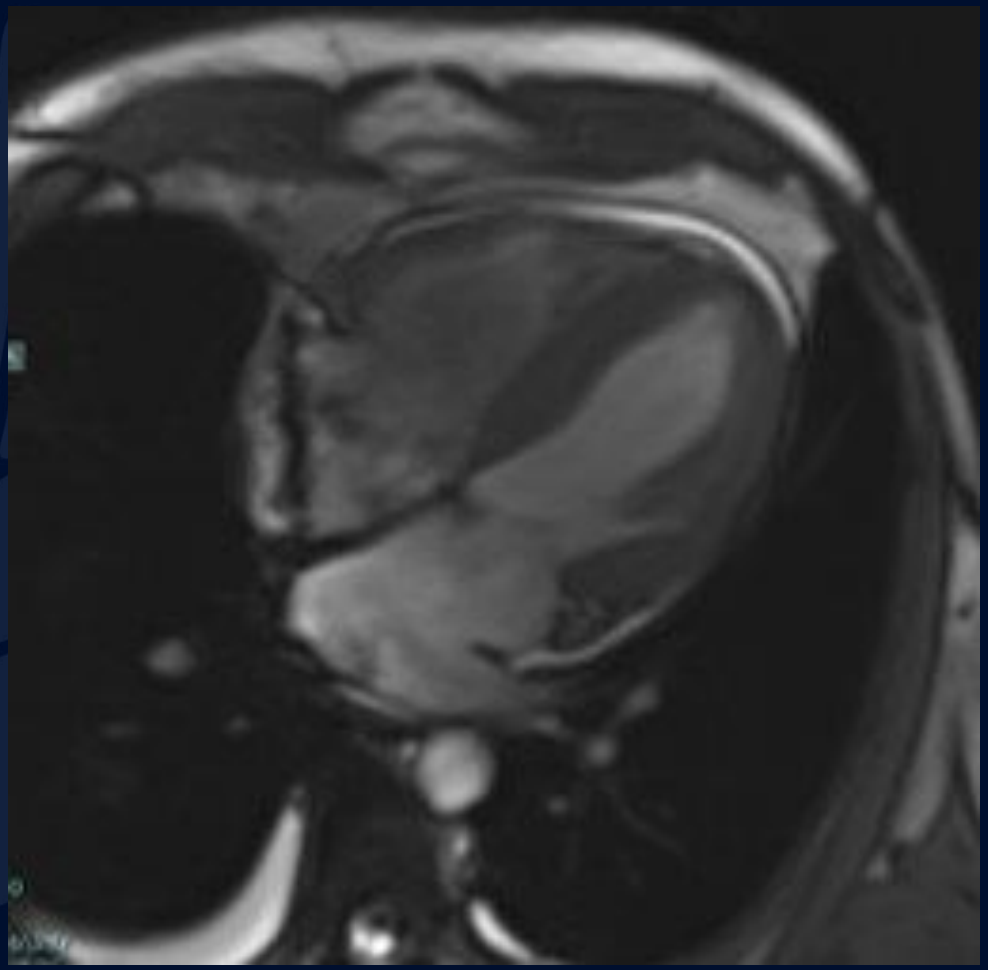
Cardiac MRI



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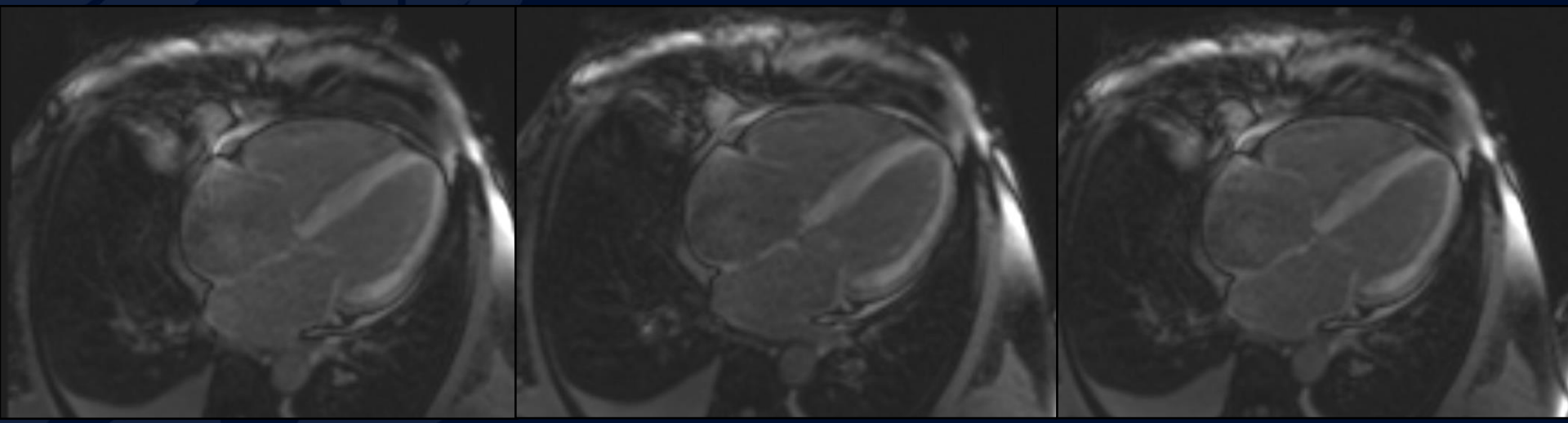
Cardiac MRI



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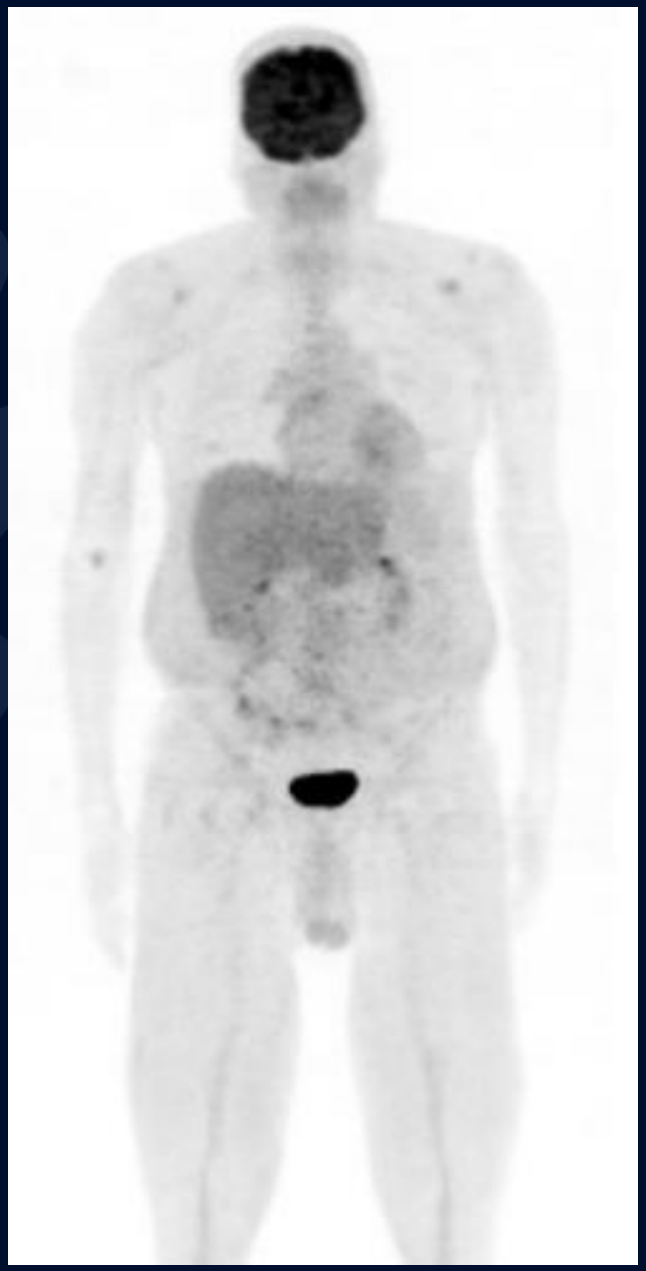
PET Scan



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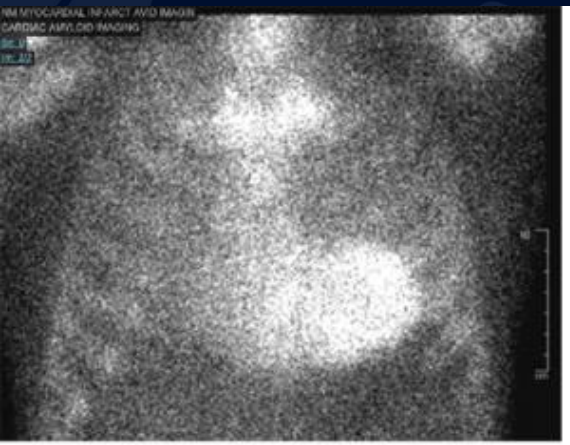
SPECT



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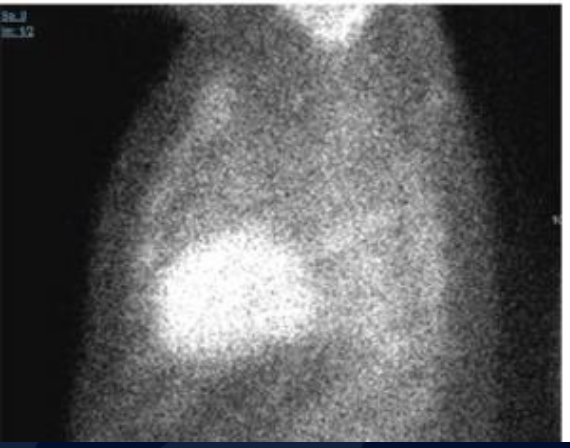

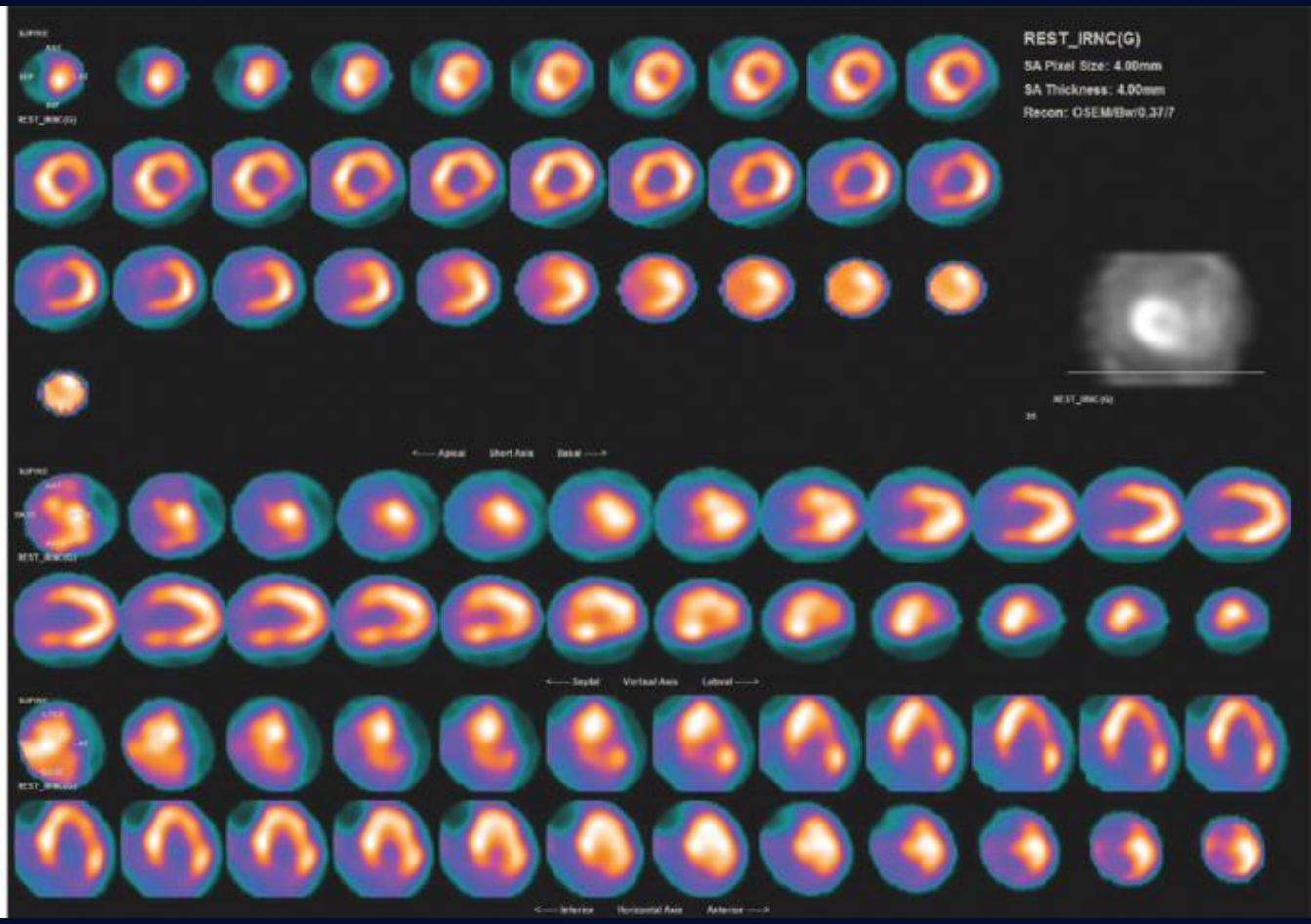

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Cardiac Amyloidosis

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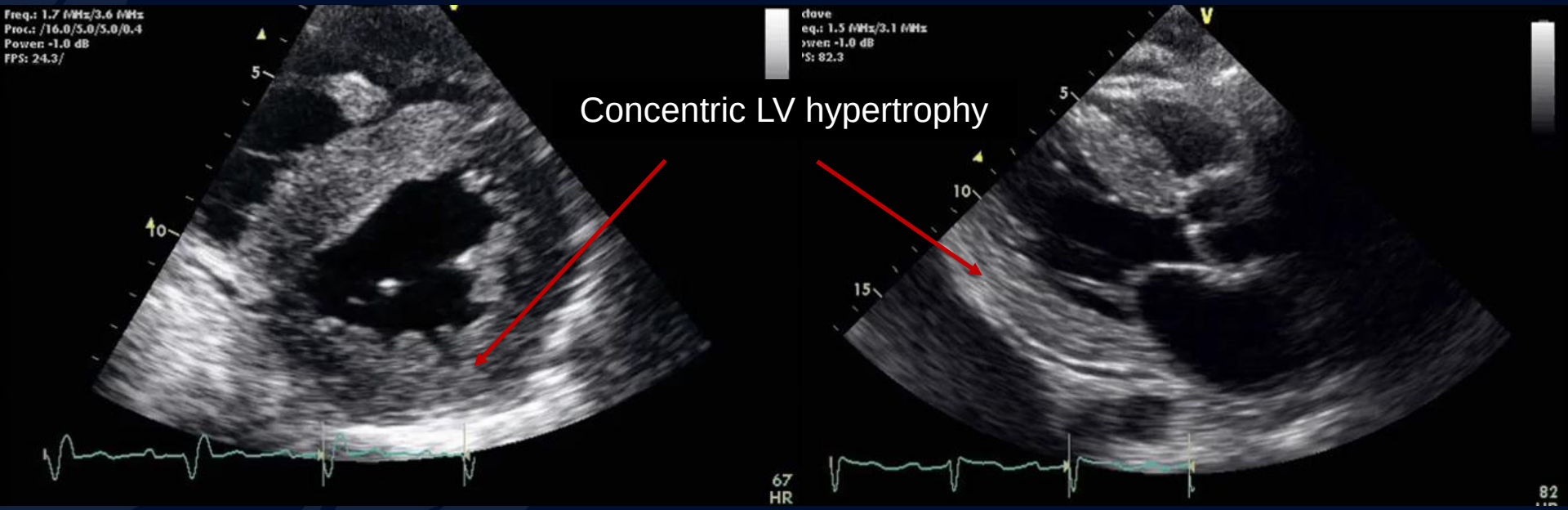
Echocardiogram



Parasternal short-axis view

Parasternal long-axis view

Concentric left ventricular wall thickening with increased echogenicity of the myocardium

Reduced systolic and diastolic function

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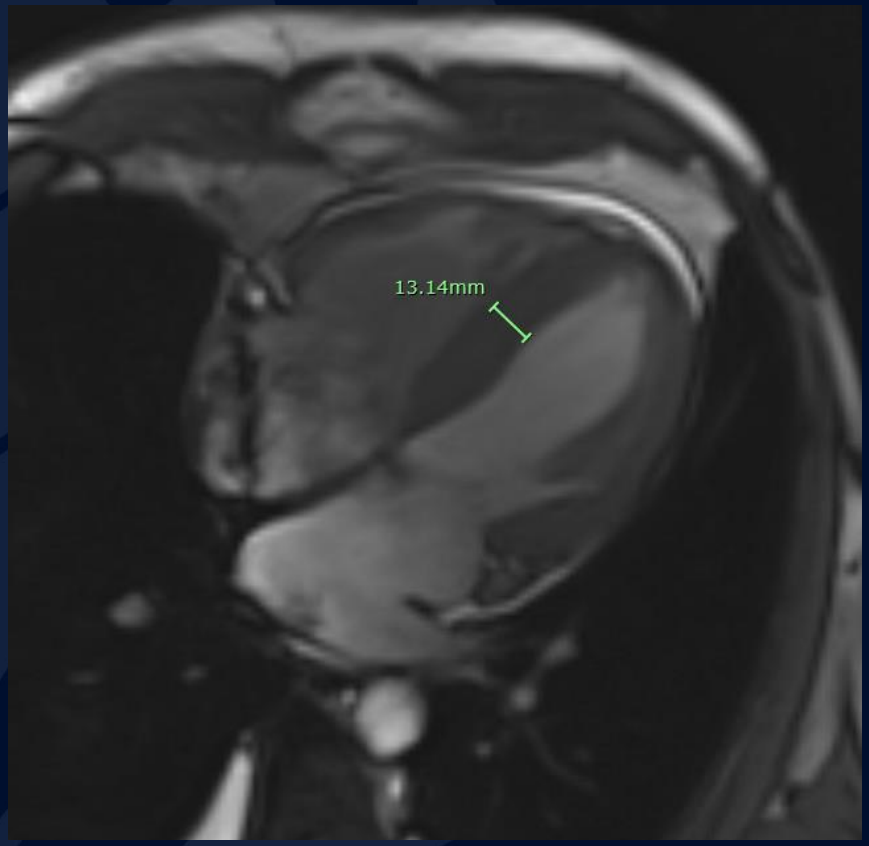
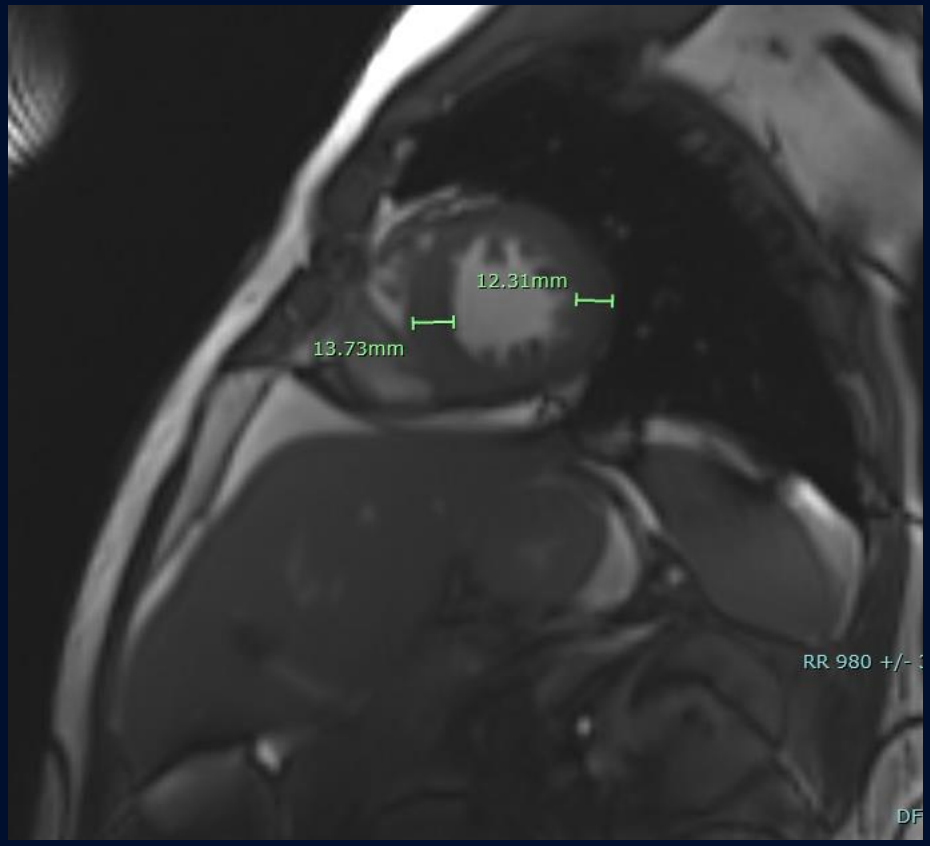
Cardiac MRI





Widened interventricular septum

Left ventricular hypertrophy

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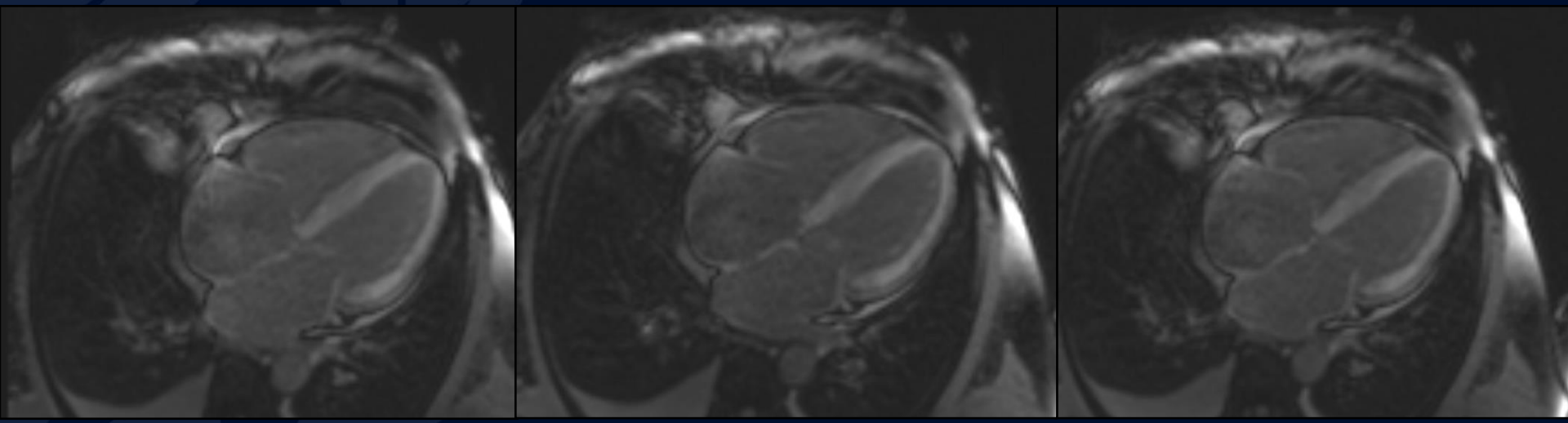
Cardiac MRI



Late gadolinium enhancement

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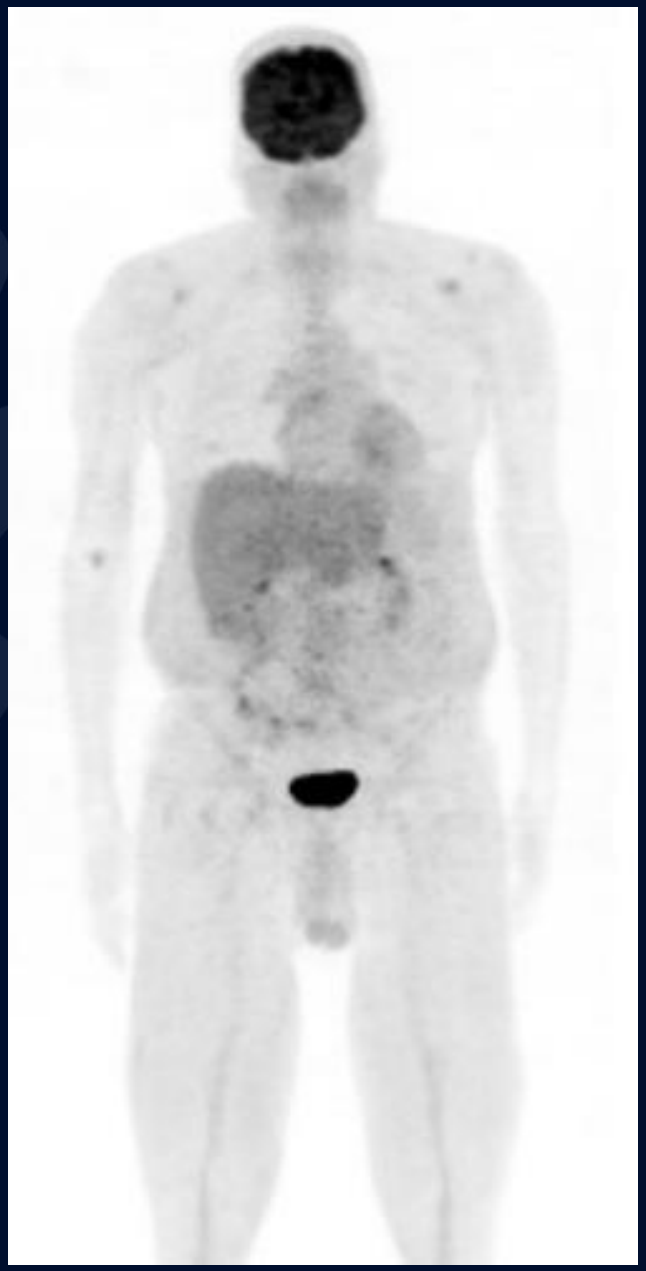
PET Scan



Mild increased radiotracer cardiac uptake

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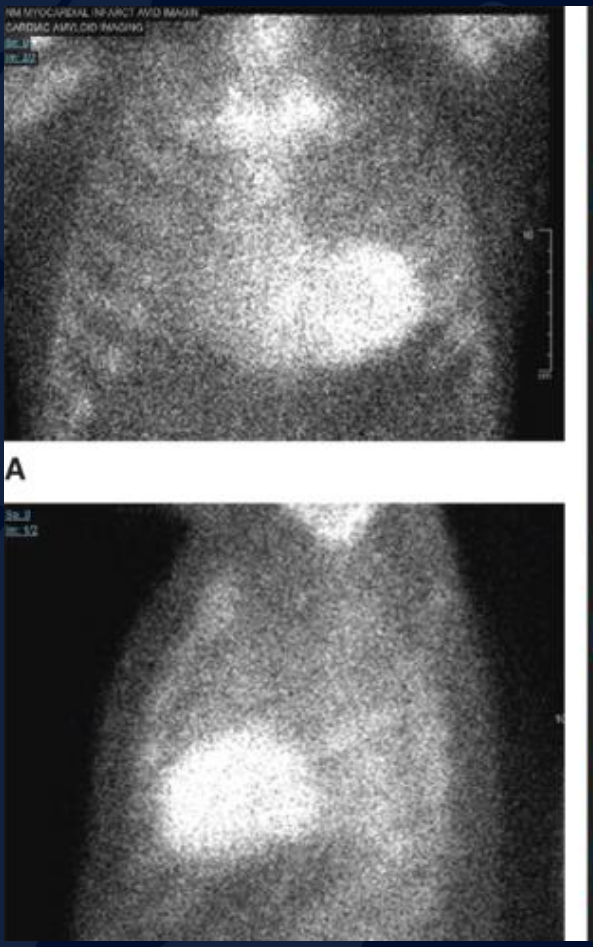
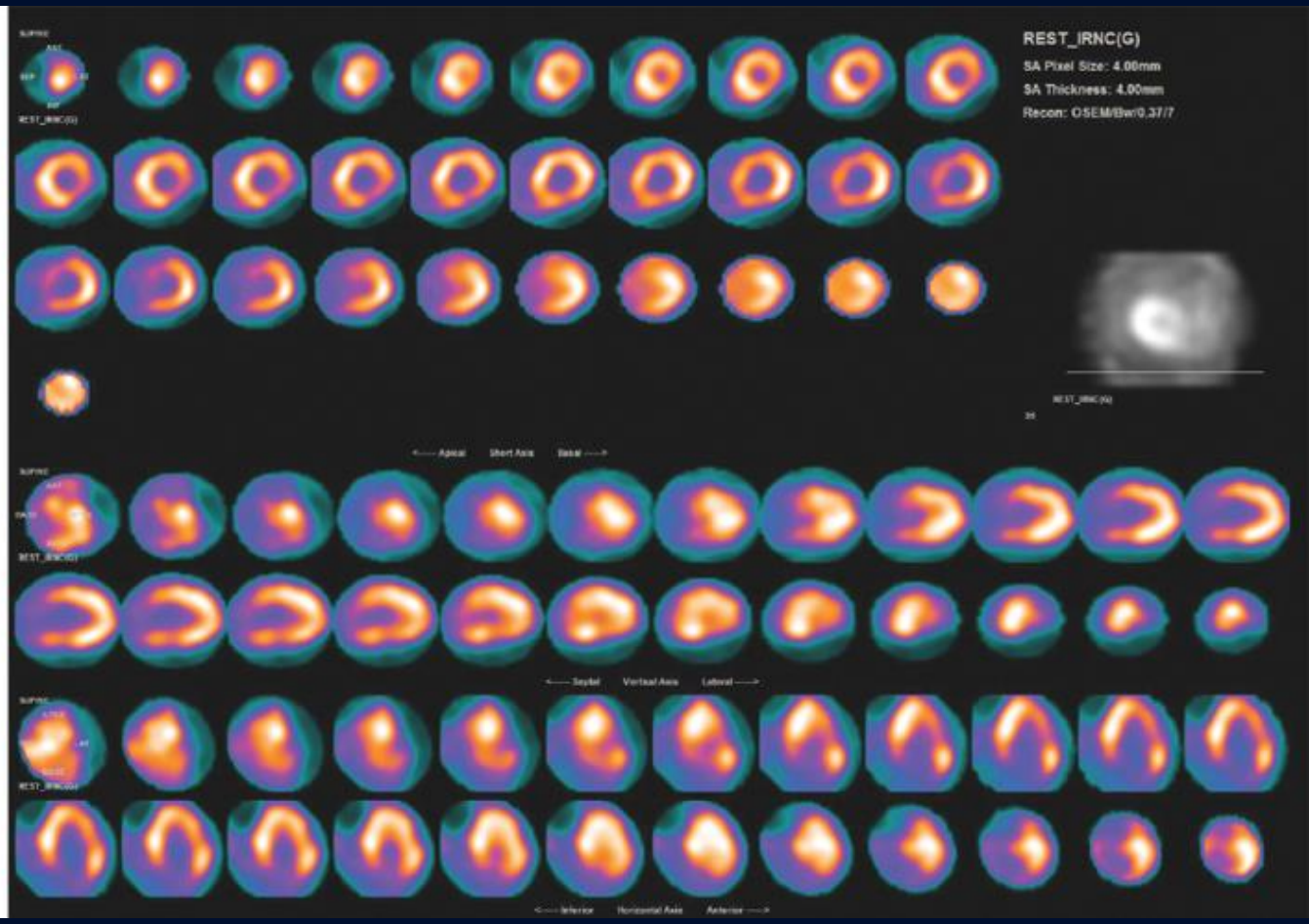
SPECT





Diffuse myocardial tracer uptake

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Cardiac Amyloidosis

Amyloidosis: A heterogeneous group of diseases characterized by extracellular accumulation of abnormal fibrillar protein deposits (amyloid)

- Types of amyloidosis that affect the heart
  - Immunoglobulin light chain amyloidosis (AL),
  - Familial transthyretin-related amyloidosis (ATTR)
  - Senile systemic amyloidosis (wild-type TTR)
  - Secondary amyloidosis (AA)
  - Isolated atrial amyloidosis (atrial natriuretic peptide)

Prognosis depends on type of amyloid and extent of cardiac and systemic involvement

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Imaging Findings

Echocardiogram

- Concentric thickening of the left ventricle, an abnormal bright echotexture, and/or asymmetric thickening of the septum
- Bi-atrial enlargement, valvular thickening, right ventricular hypertrophy, and small pericardial effusion may be observed
- With a small left ventricular cavity, low-gradient aortic stenosis is suspicious for hereditary amyloidosis
- +/- systolic and diastolic dysfunction

Contrast-enhanced cardiac MRI

- Helpful in infiltrative cardiomyopathies; gadolinium-based contrast can reveal abnormal patterns suggestive of cardiac amyloidosis
  - T1 signal may significantly be increased with amyloid deposition
- Inability to “null” or blacken the myocardium has been suggestive of cardiac amyloidosis.
- Left ventricular late gadolinium enhancement is very common
- Other features include, patchy myocardial scarring, right ventricular thickening, bi-atrial enlargement, and delayed enhancement of respective structures

SPECT (single-photon emission computed tomography)

- Tc-99m PYP bone tracer uptake graded visually with scores ranging from 0 to3
  - 0 – no cardiac uptake
  - 1 – cardiac uptake < rib uptake
  - 2 – cardiac uptake = rib uptake
  - 3 – cardiac uptake > rib uptake
- Contralateral lung uptake ration > 1.5 at 1 hour is also diagnostic
- Planar SPECT confirms myocardial tracer retention

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References

Echo - Di Carli, Marcelo F., et al. "Atlas of Noninvasive Imaging." *Harrison's Principles of Internal Medicine, 21e* Eds. Joseph Loscalzo, et al. McGraw Hill, 2022

MRI - Chapter 107 Immunoglobulin Light Chain Amyloidosis, Kaushansky K, Prchal JT, Burns LJ, Lichtman MA, Levi M, Linch DC. *Williams Hematology, 10e;* 2021

SPECT - Chapter 43 Cardiac Amyloidosis, Fuster V, Narula J, Vaishnava P, Leon MB, Callans DJ, Rumsfeld J, Poppas A. *Fuster and Hurst's The Heart, 15e;* 2022.

Gertz, Morie A., et al. "Cardiac Amyloidosis." *Fuster and Hurst's The Heart, 15e* Eds. Valentin Fuster, et al. McGraw Hill, 2022

Gertz, Morie A., et al. "Immunoglobulin Light Chain Amyloidosis." *Williams Hematology, 10e* Eds. Kenneth Kaushansky, et al. McGraw Hill, 2021Kaufman, Gregory P., et al. "Systemic Immunoglobulin Light Chain Amyloidosis." *The MD Anderson Manual of Medical Oncology, 4e* Eds. Hagop M. Kantarjian, et al. McGraw Hill, 2022,

Munshi, Nikhil C., et al. "Plasma Cell Disorders." Harrison's Principles of Internal Medicine, 21e Eds. Joseph Loscalzo, et al. McGraw Hill, 2022

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