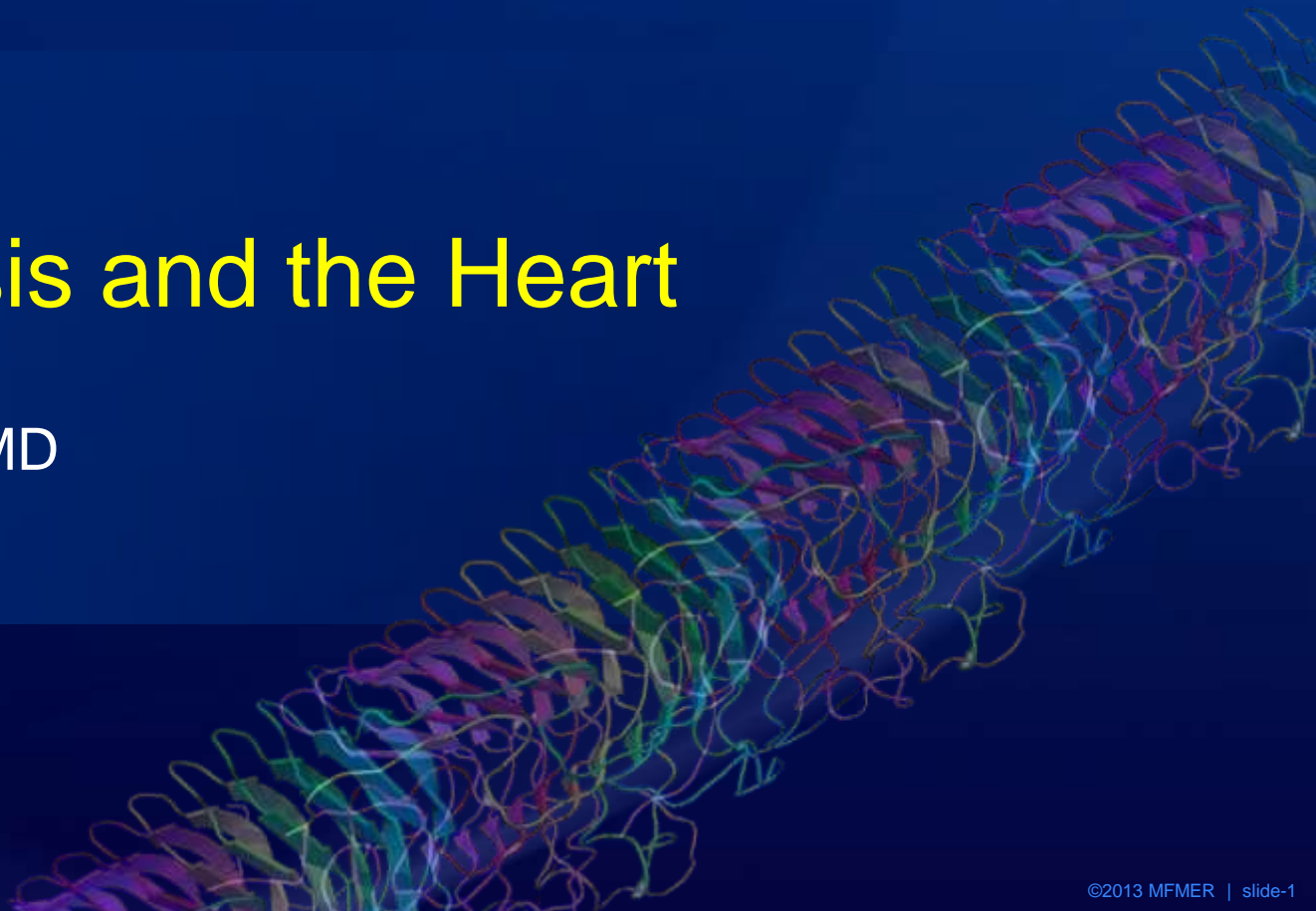




# Amyloidosis and the Heart

Martha Grogan, MD



# Cardiac Amyloidosis

- Amyloid is a disorder of *misfolded* proteins
- Proteins circulate in your blood stream to perform many different functions
- They should be dissolved - in other words – liquid
- In amyloid they become solid and deposit in the organs and tissues of your body and cause problems

# What is Transthyretin?

- Transthyretin - a protein that:

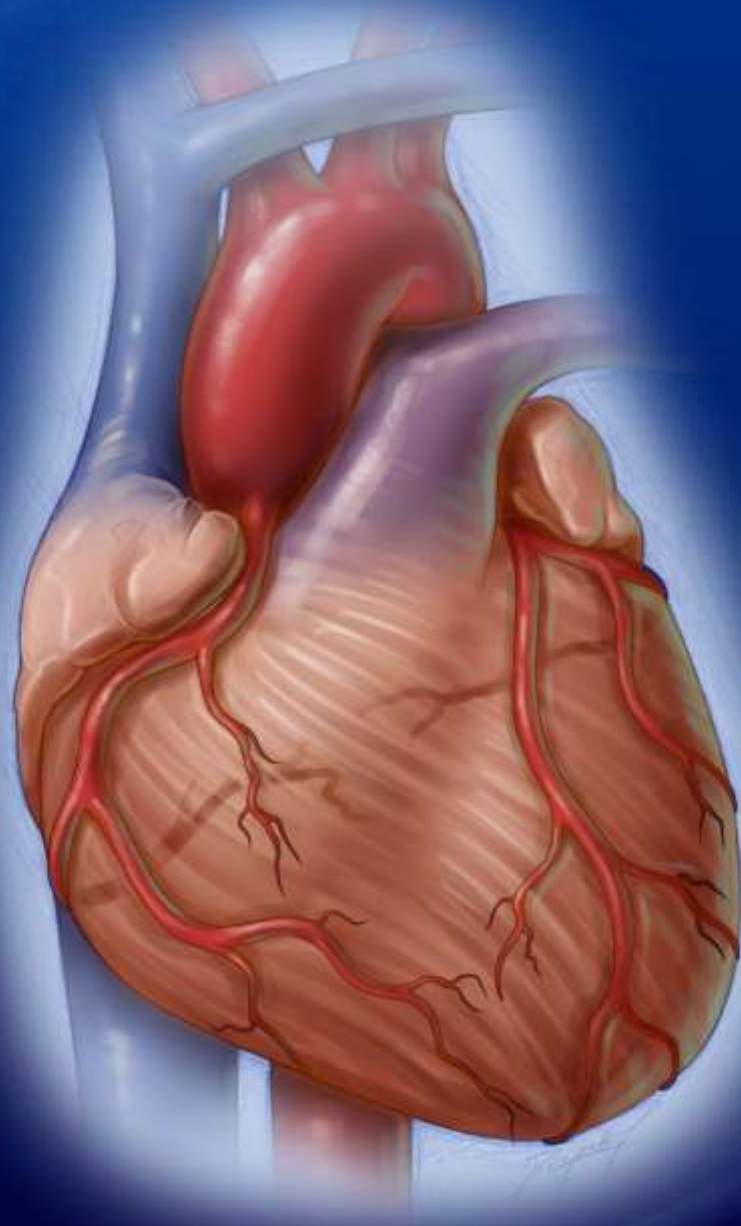
Transports **T**hyroxine and **R**etinol (vitamin A)

a. Hereditary (familial)

deposits in nerves, heart, or both

b. Wild type (“age related” or “senile)

deposits in heart, carpal tunnel

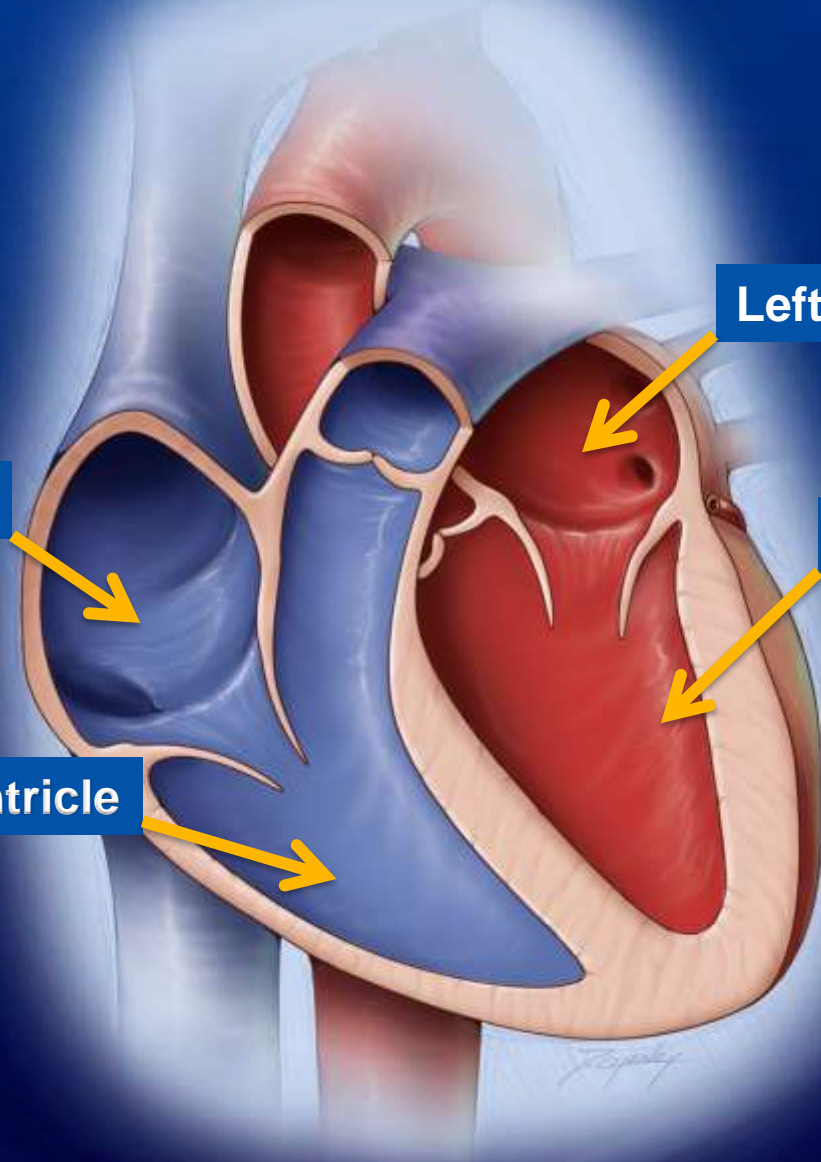


**Right Atrium**

**Right Ventricle**

**Left Atrium**

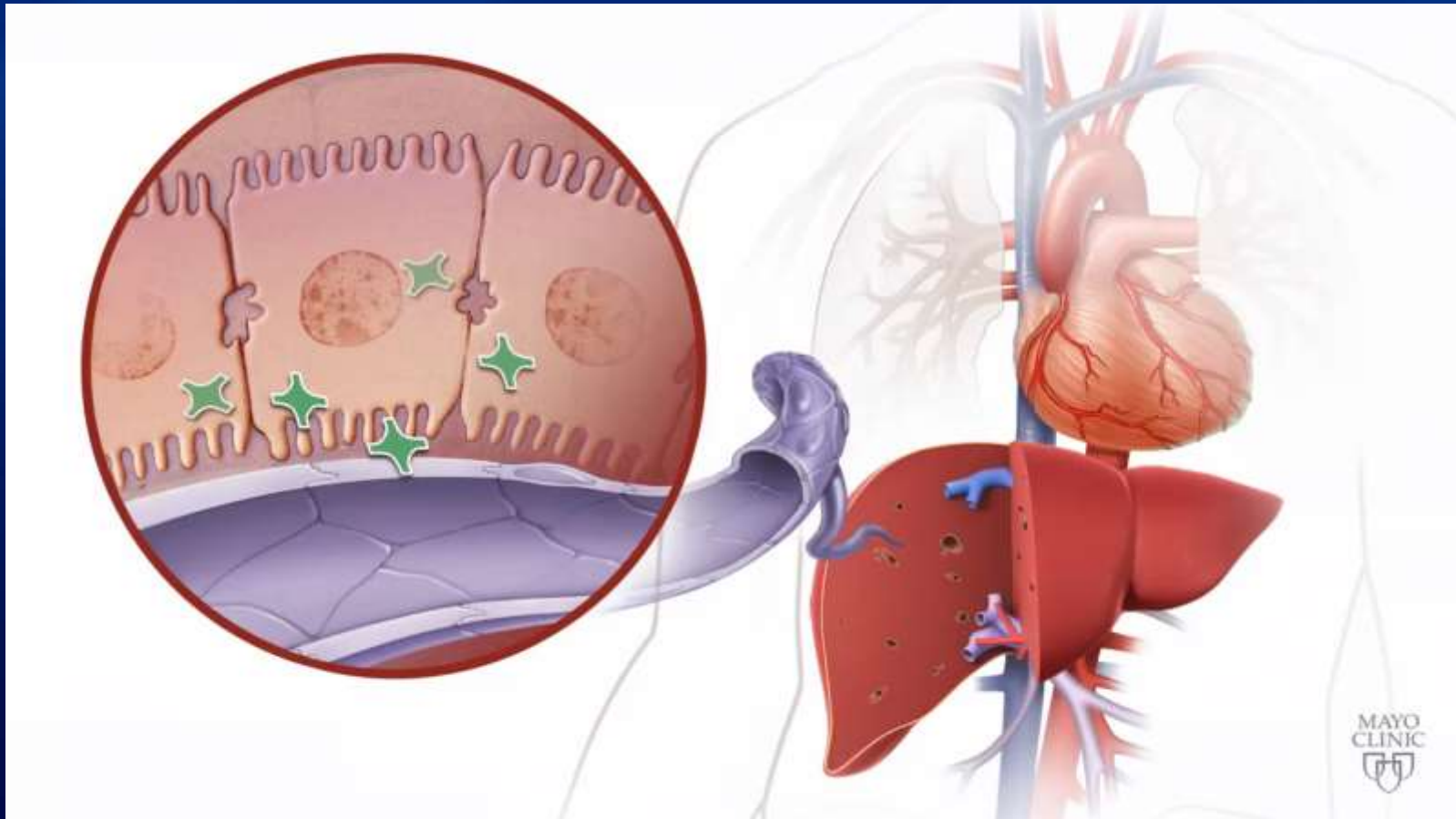
**Left Ventricle**



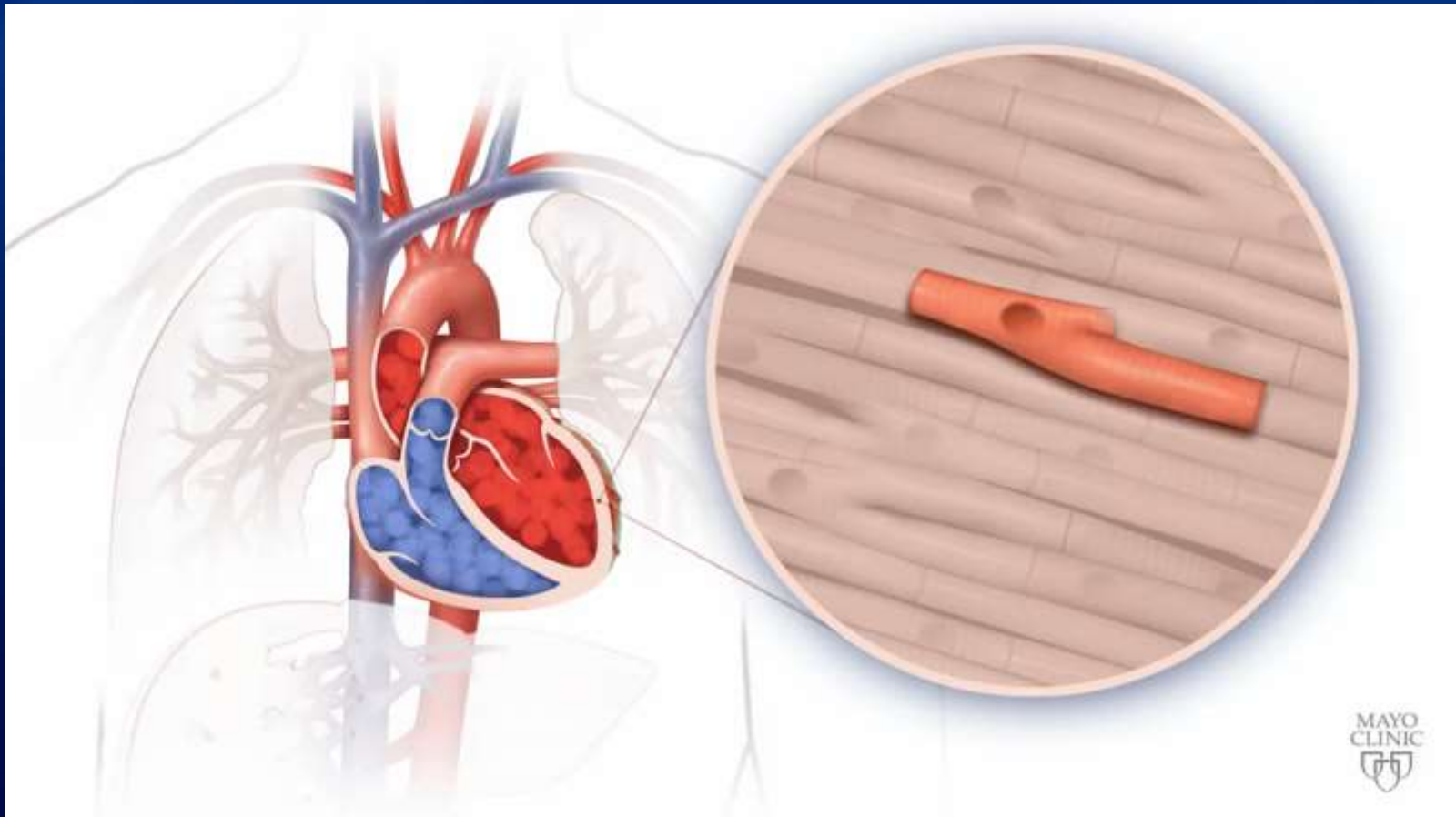
# How the Heart Works



# Transthyretin (TTR) Amyloid



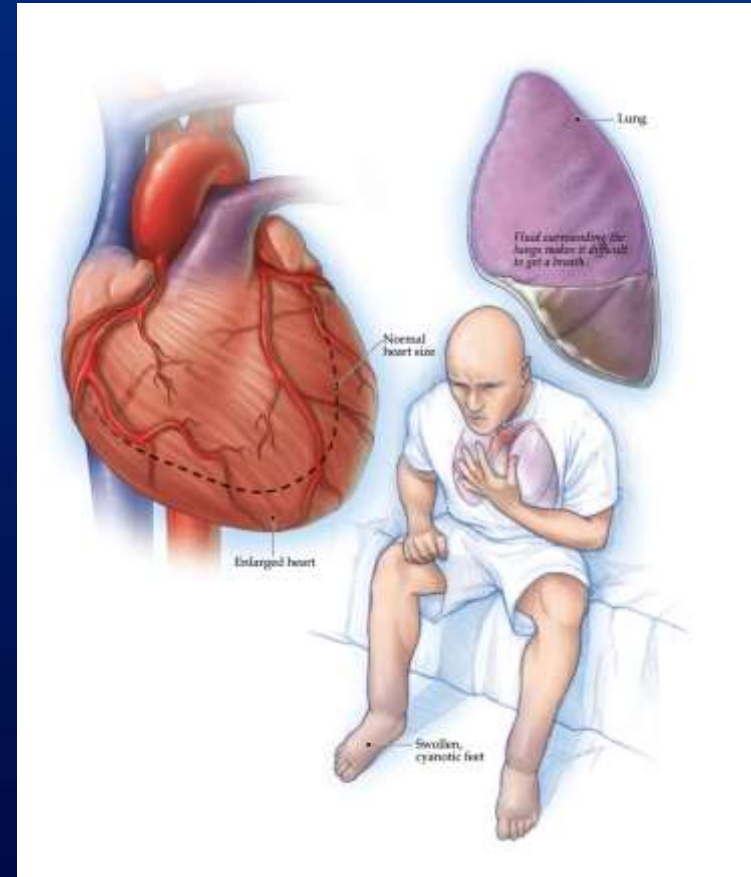
# Heart Muscle with Amyloid



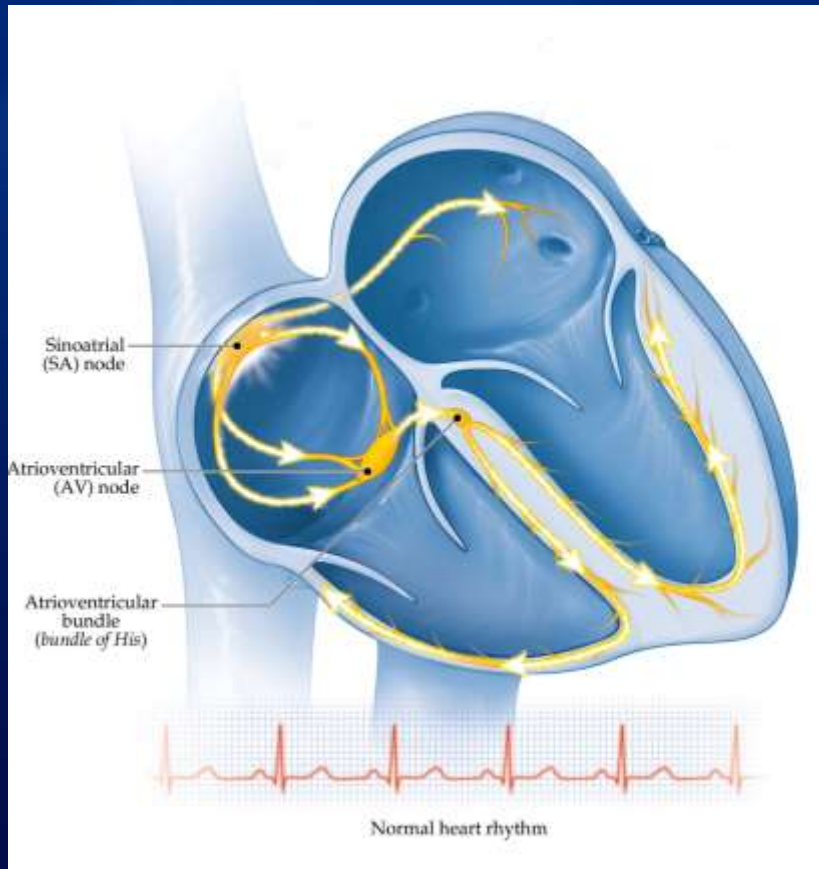


# Symptoms and Signs of Heart Failure

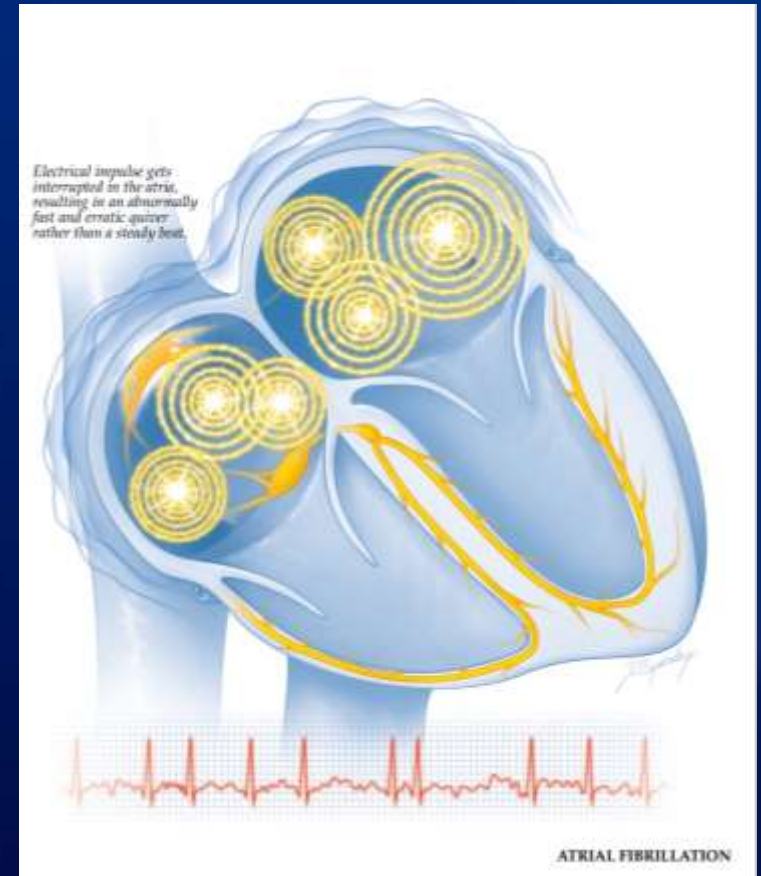
- Fatigue
- Shortness of Breath
- Swelling (edema)
- Unable to lie down due to shortness of breath
- Waking up gasping for air
- Cough, often at night



# Heart Rhythm problems (Arrhythmias)



Normal Rhythm

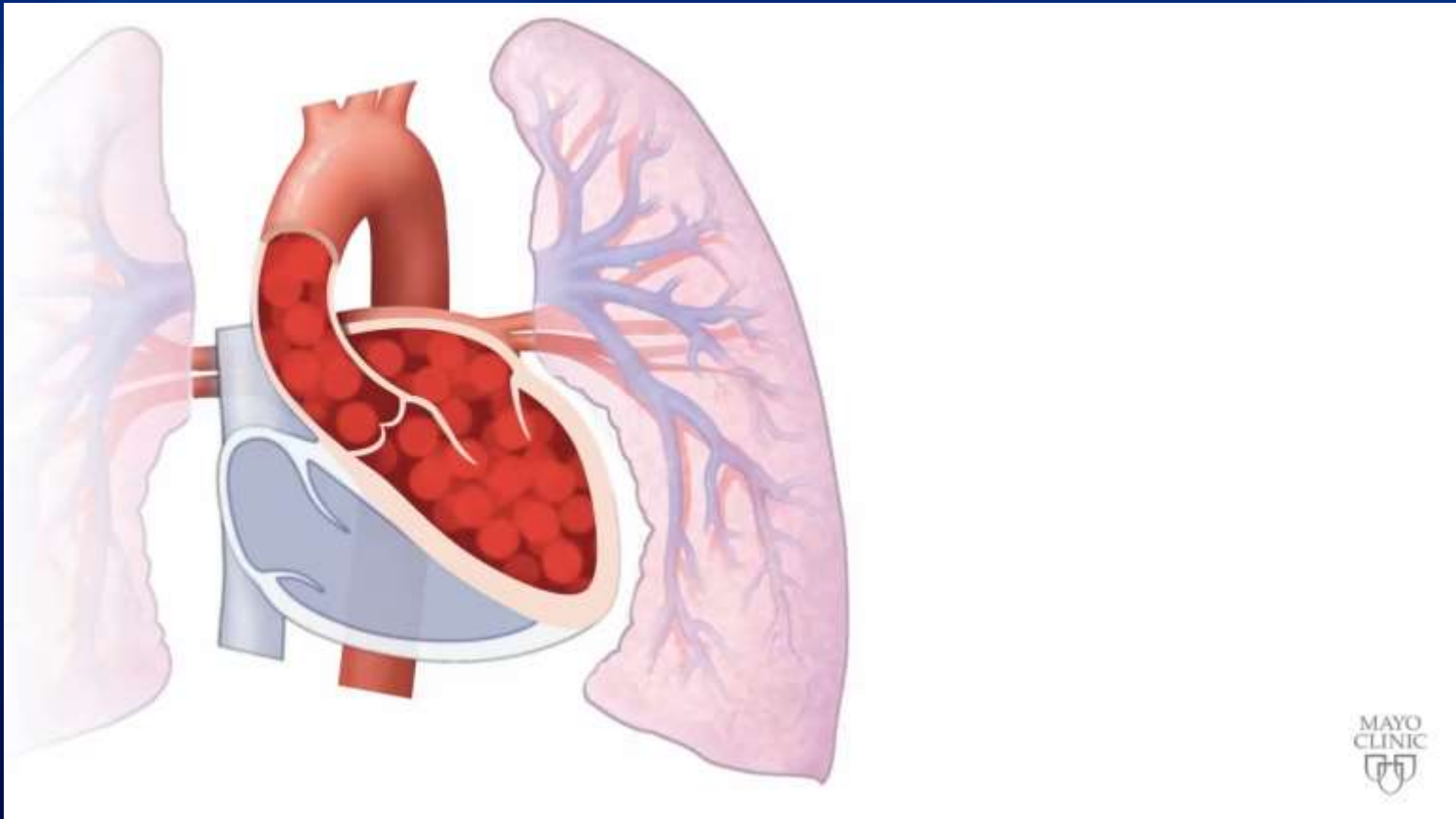


Atrial Fibrillation

# Heart Rhythm Problems in Amyloid

- Bradycardia – too slow – may need pacemaker
- Tachycardia – too fast –
- Atrial fibrillation – irregular rhythm from upper chambers
  - Medications
  - Electrical shock (cardioversion)
  - Risk of blood clot – stroke – need blood thinners
- Defibrillator – for arrhythmias from ventricles

# Fluid in Lungs due to Heart Failure



# Heart Tests to Diagnose Cardiac Amyloid

- Echo – often amyloid is first suspected due to abnormal echo
  - Measure thickness , pumping function, stiffness, valve function, pressure in lungs
- MRI – certain patterns suggest amyloid
- PYP – for TTR amyloid
- Biopsy

# Blood Tests in Cardiac Amyloid

- **Troponin** – protein released from heart muscle, usually due to heart attack; often increased in amyloid- but not heart attack
- **BNP or NT pro-BNP** – another protein from heart, released in response to higher pressure in heart
  - Varies up to 40% over a week
  - Trend is more important than one number

# Ejection Fraction =

Normal



$$\frac{6}{10} \times 100 = 60\%$$

Dilated



$$\frac{6}{20} \times 100 = 30\%$$

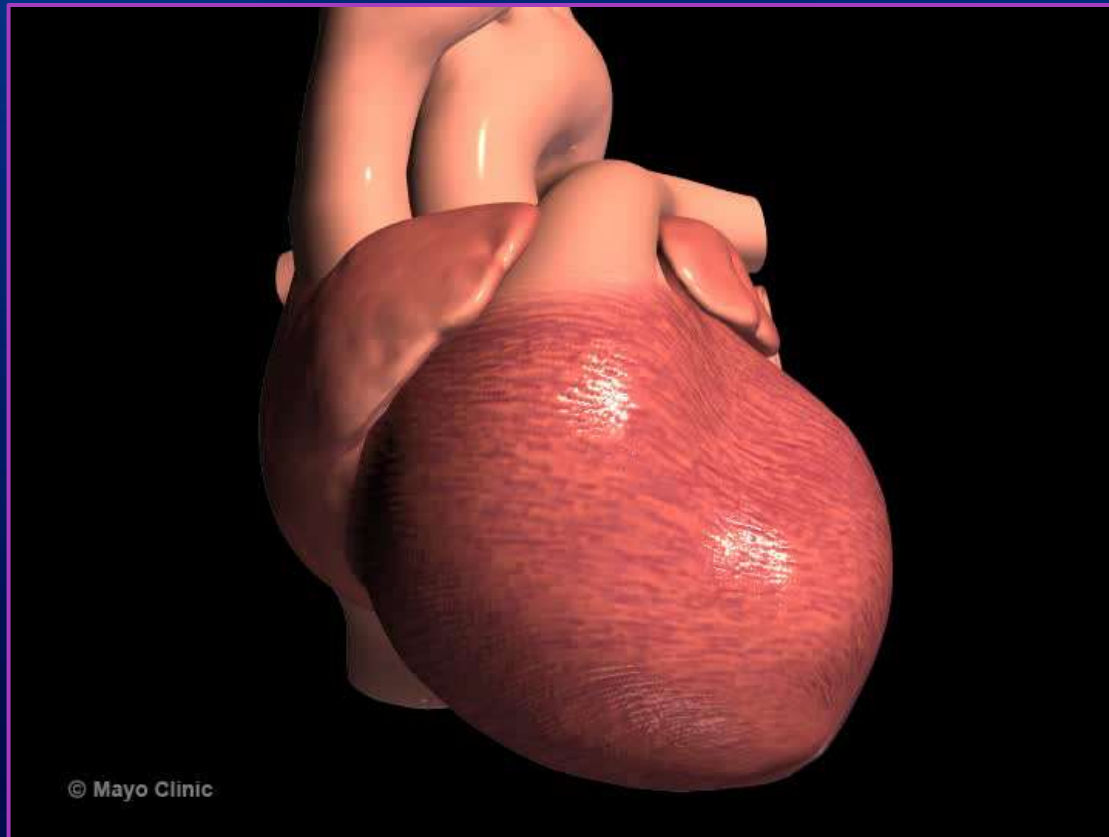
Amyloid



$$\frac{3}{6} \times 100 = 50\%$$

# Cardiac Twist and Torsion

Heart function is complex, like wringing a towel\*



Courtesy of Dr. Jae Oh



# What is Myocardial Strain?

# Myocardial Strain

- Heart contracts - muscle cells shorten

Echo strain reflects how much the muscle of the heart shortens

negative number, more negative is better

example -18% is better than -8%

- “Bulls-eye” pattern - typical for amyloid
- Great test to suggest amyloid
- Not widely available, technically challenging

# Treatment of Cardiac Amyloid

- Stop the source of amyloid
- No medication to take amyloid out of heart (yet)
- Diuretics to decrease shortness of breath and get rid of fluid
- Medications used for other type of heart failure often not helpful (beta-blockers, ACE-inhibitors)
  - Individualized treatment

# Cardiac Amyloidosis - Summary

- Amyloid - stiff heart - hard to fill
- Heart Failure and Rhythm problems
- Heart function is complex - *a single number* does not tell you how your heart is doing
- Treatment options are expanding