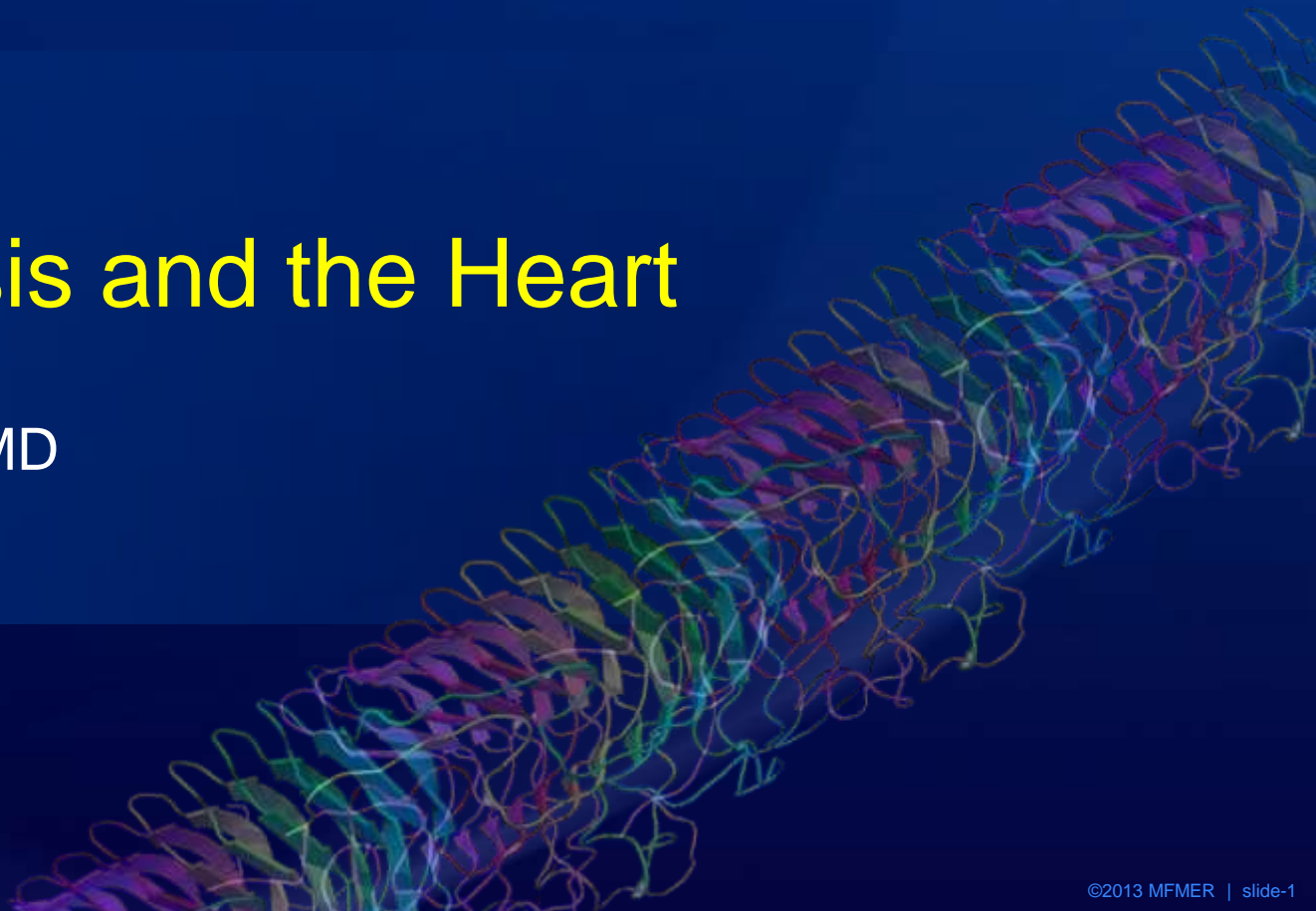




Amyloidosis and the Heart

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:

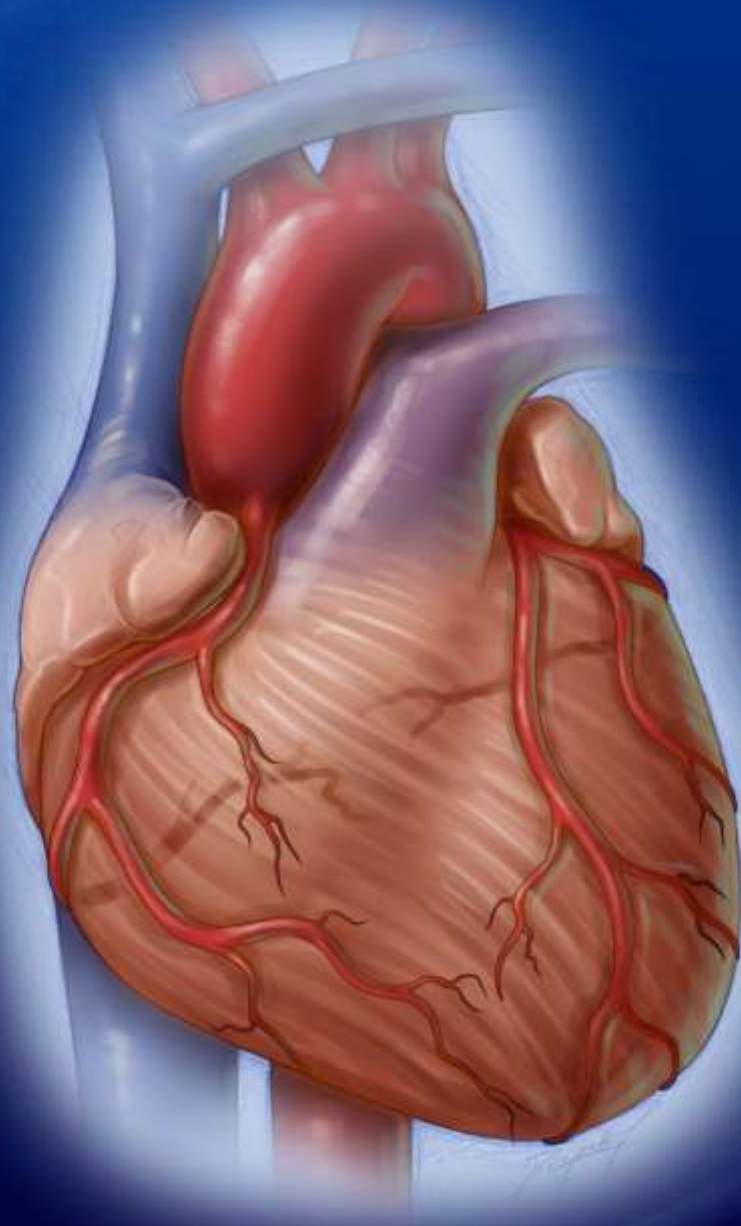
Transports **T**hyroxine and **R**etinol (vitamin A)

a. Hereditary (familial)

deposits in nerves, heart, or both

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

deposits in heart, carpal tunnel

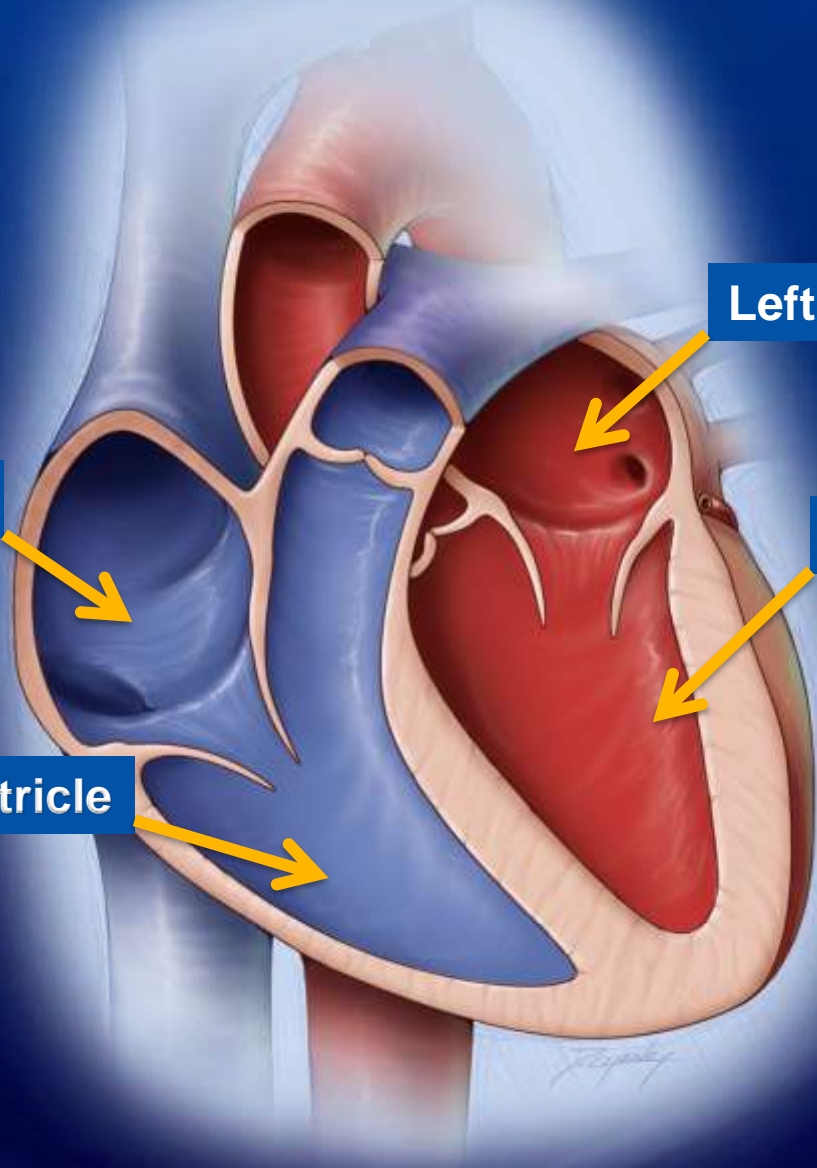


Right Atrium

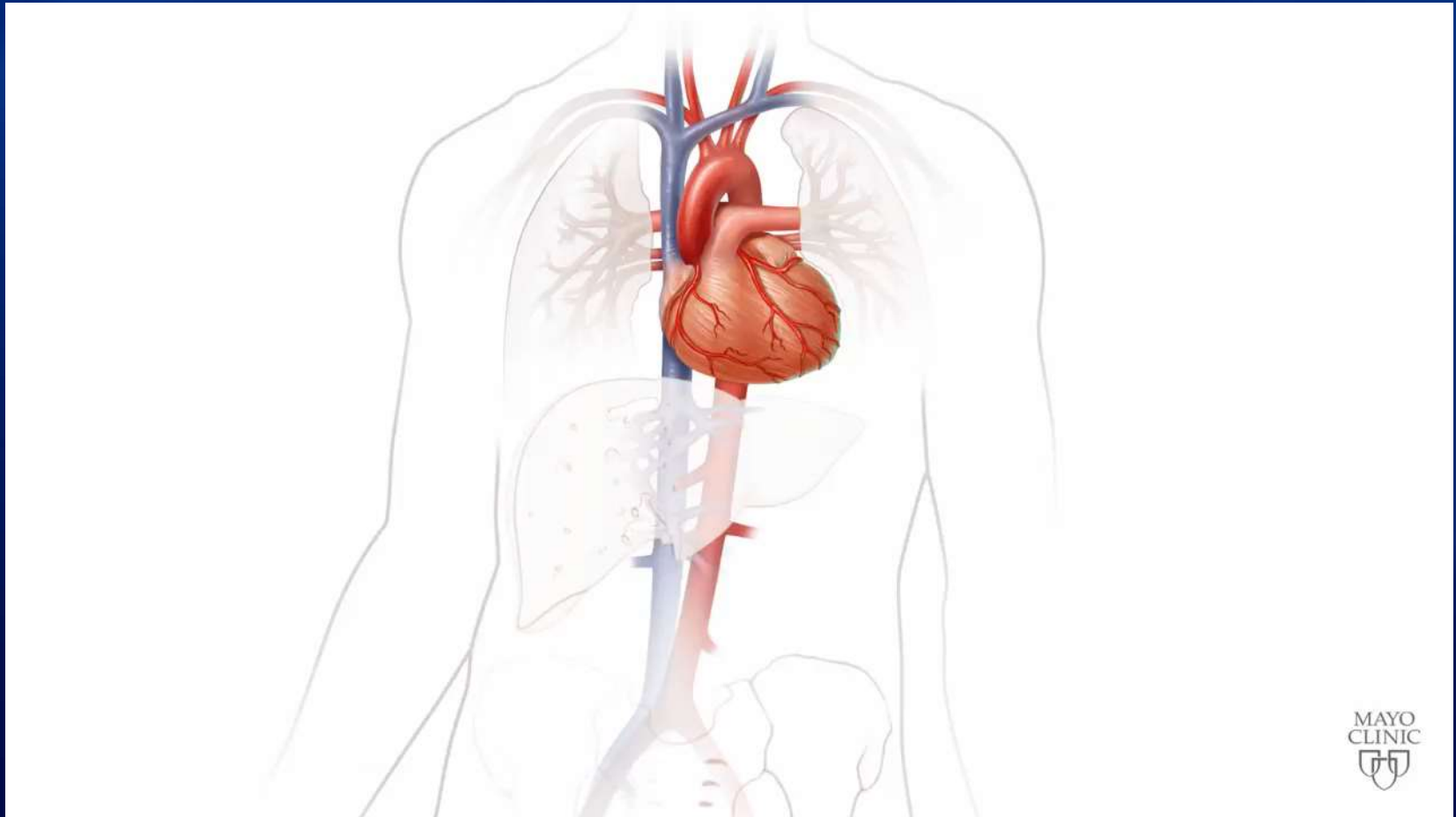
Left Atrium

Left Ventricle

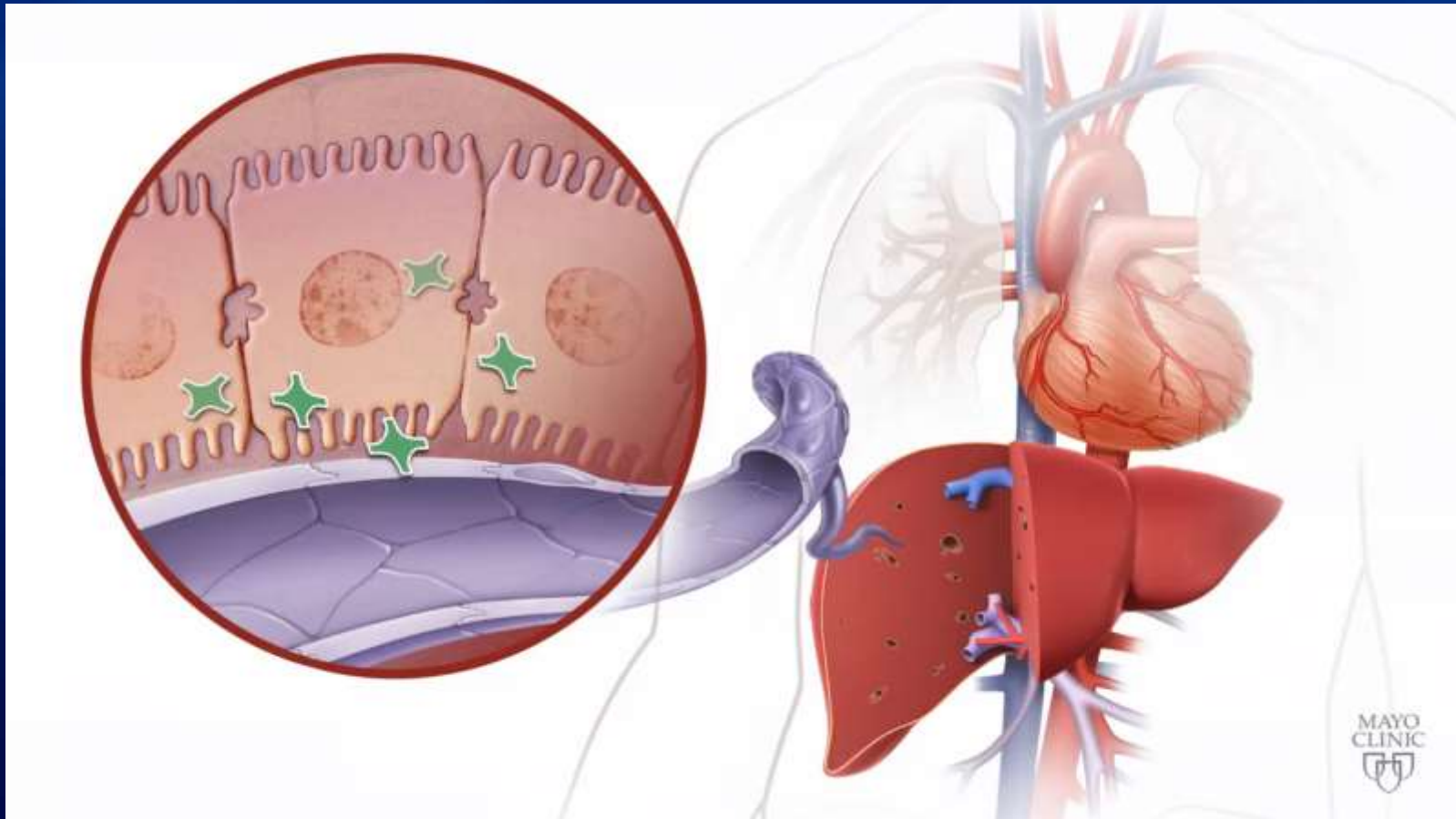
Right Ventricle



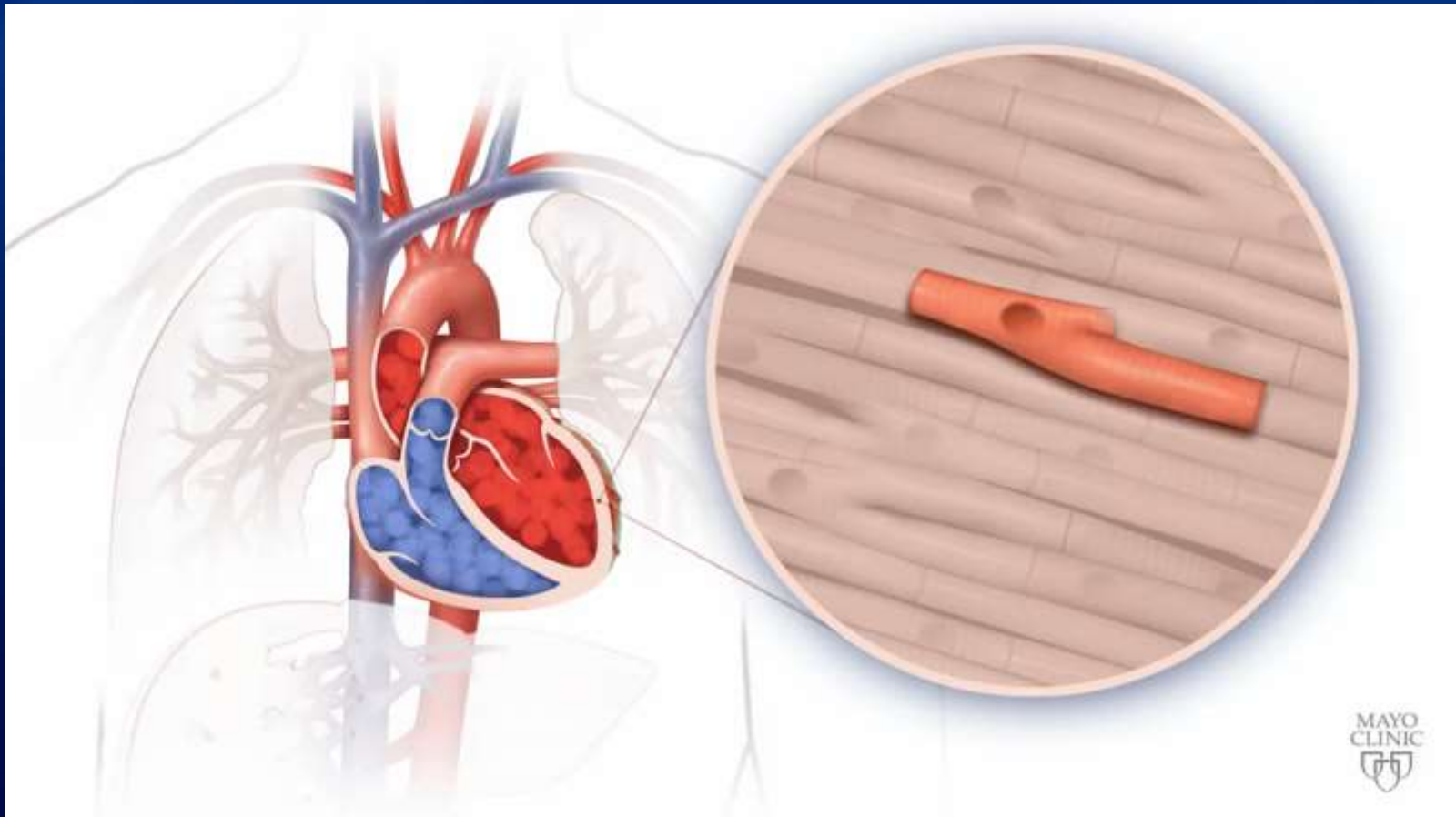
How the Heart Works



Transthyretin (TTR) Amyloid

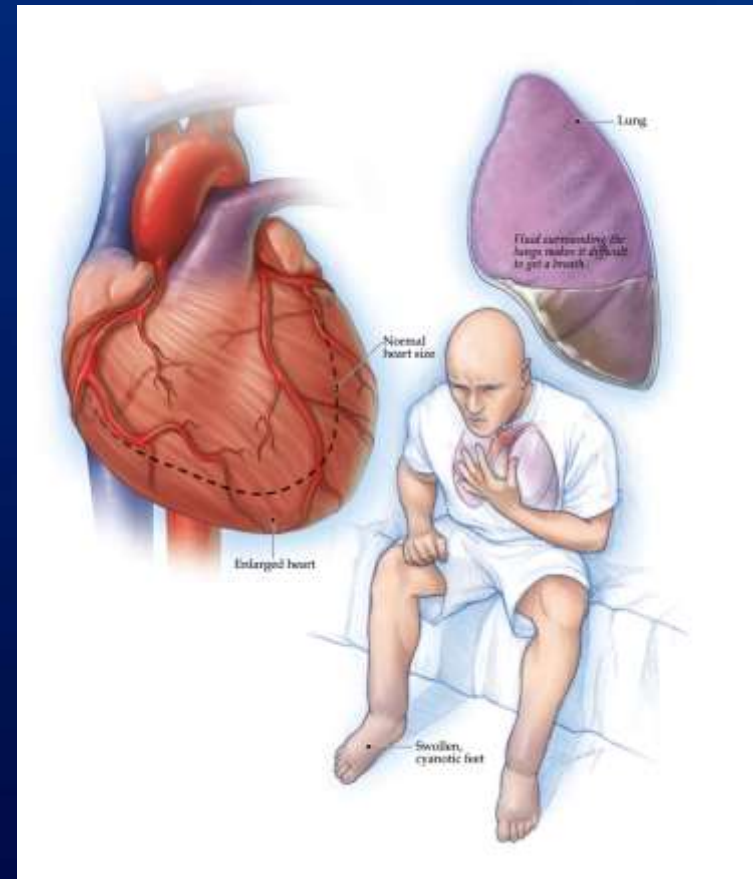


Heart Muscle with Amyloid

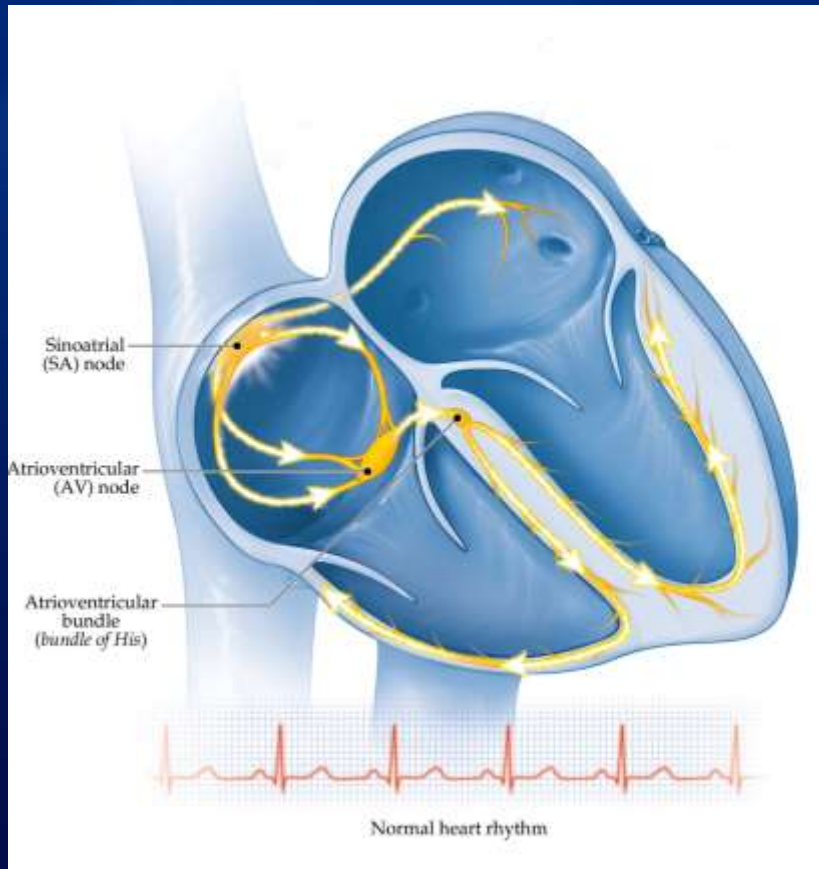


Symptoms and Signs of Heart Failure

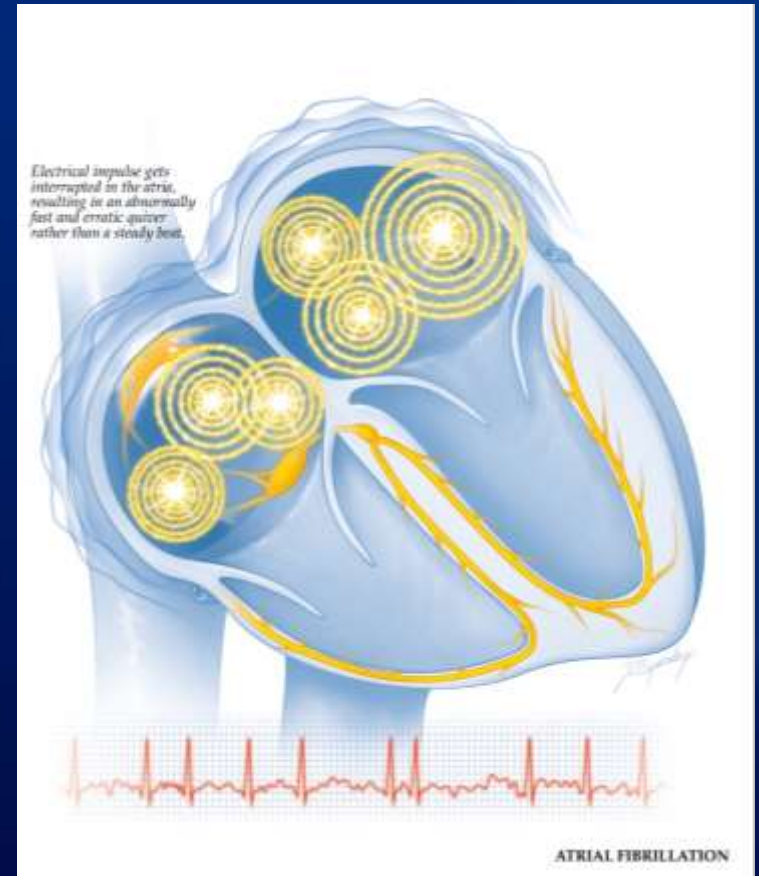
- Fatigue
- Shortness of Breath
- Swelling (edema)
- Unable to lie down due to shortness of breath
- Waking up gasping for air
- Cough, often at night



Heart Rhythm problems (Arrhythmias)



Normal Rhythm

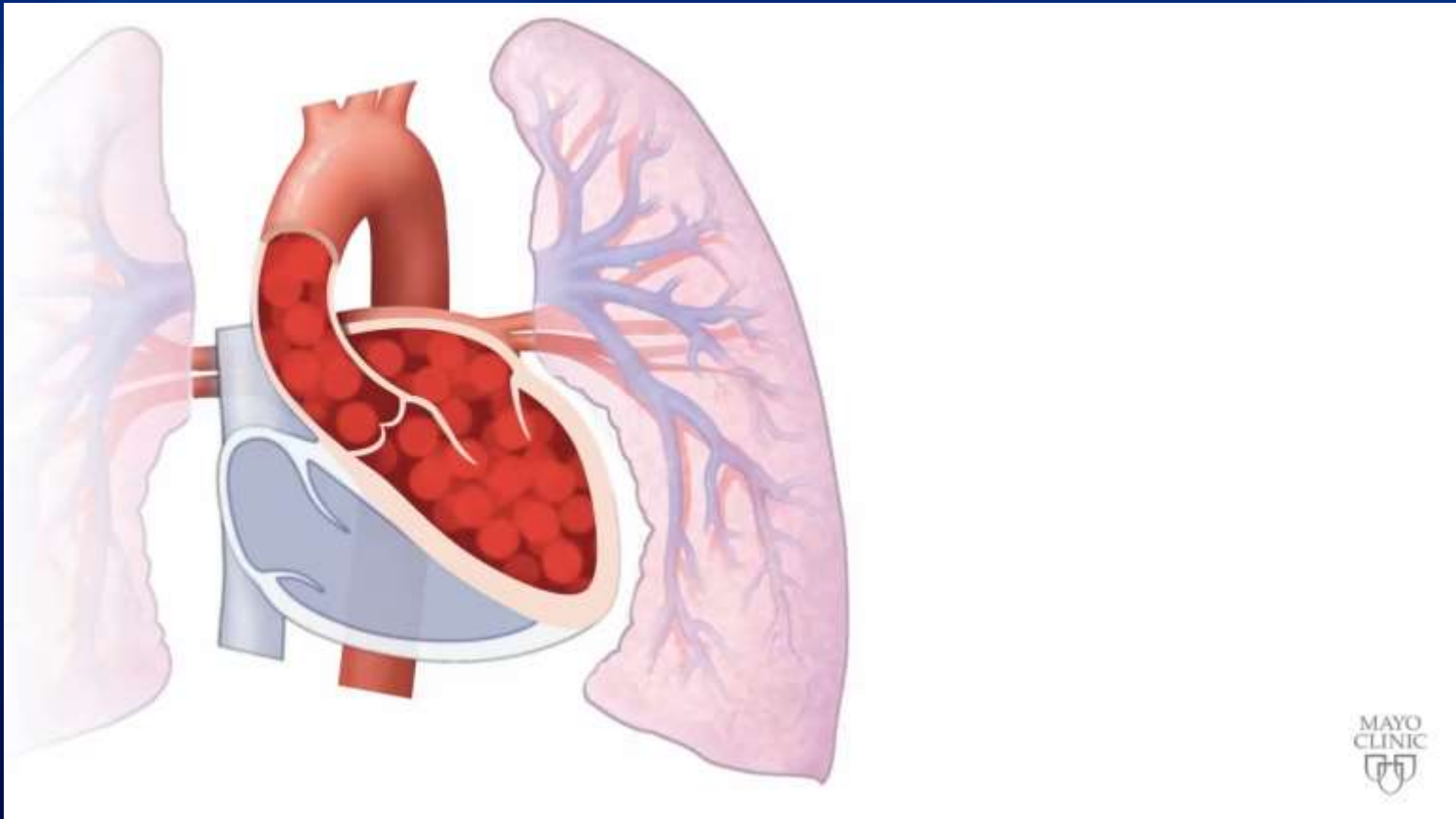


Atrial Fibrillation

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



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

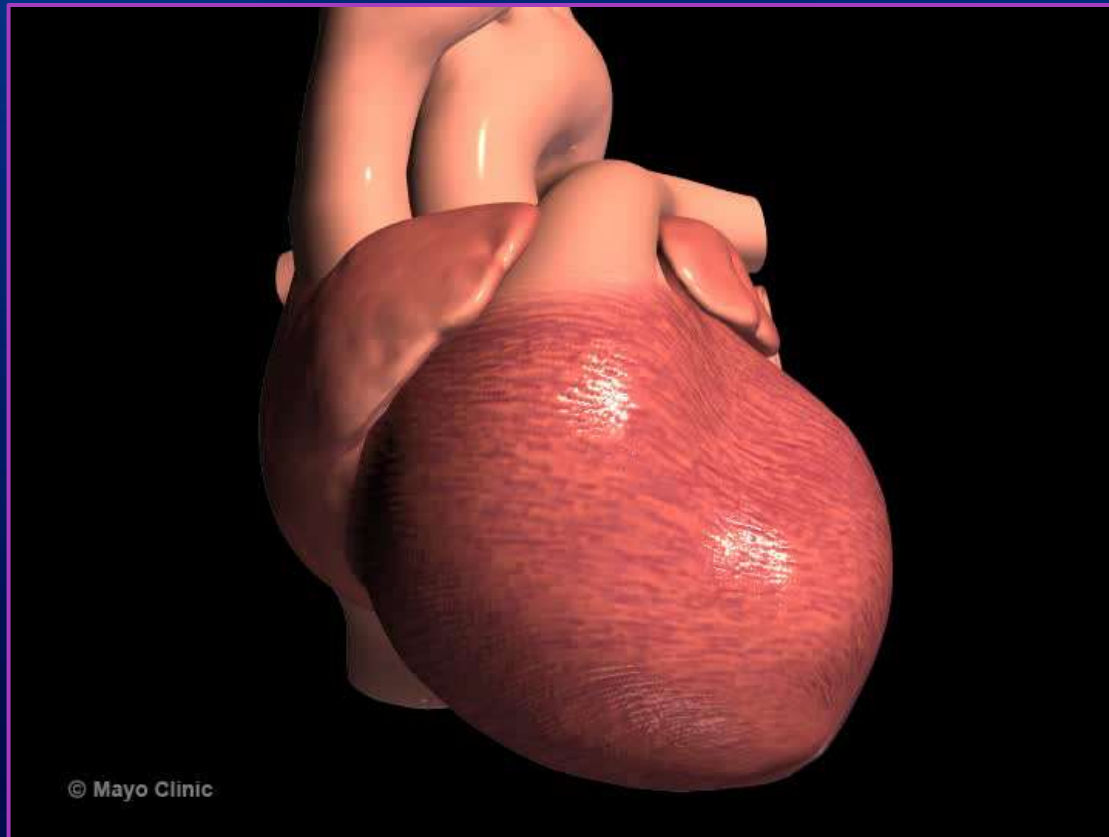
Amyloid



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

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