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Amyloidosis and the Heart

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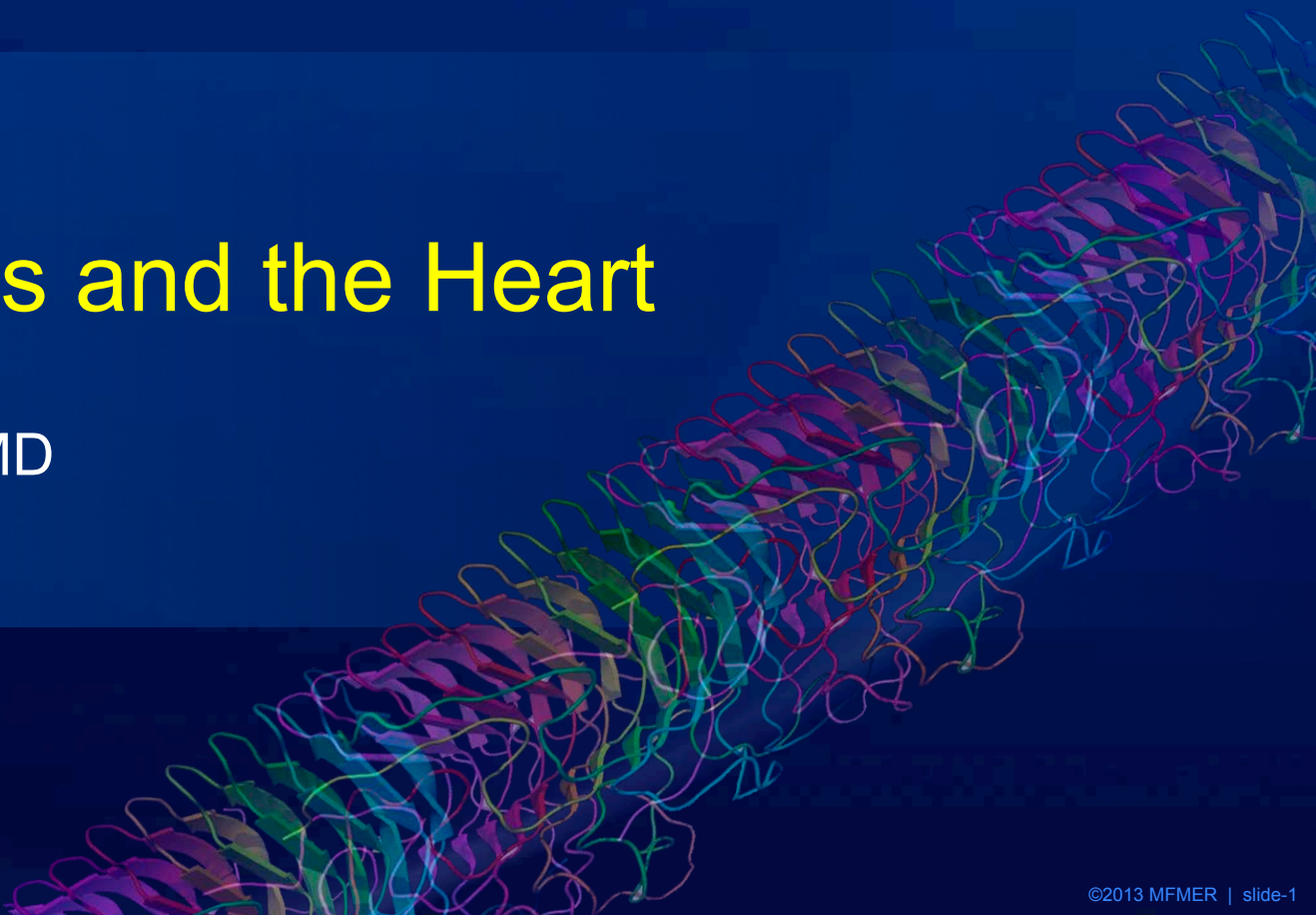
October 26, 2013



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Cardiac Amyloidosis

- Normal Heart Function
- How Amyloid affects the heart
- Symptoms
- Explanation of Heart Tests
- Treatment Options

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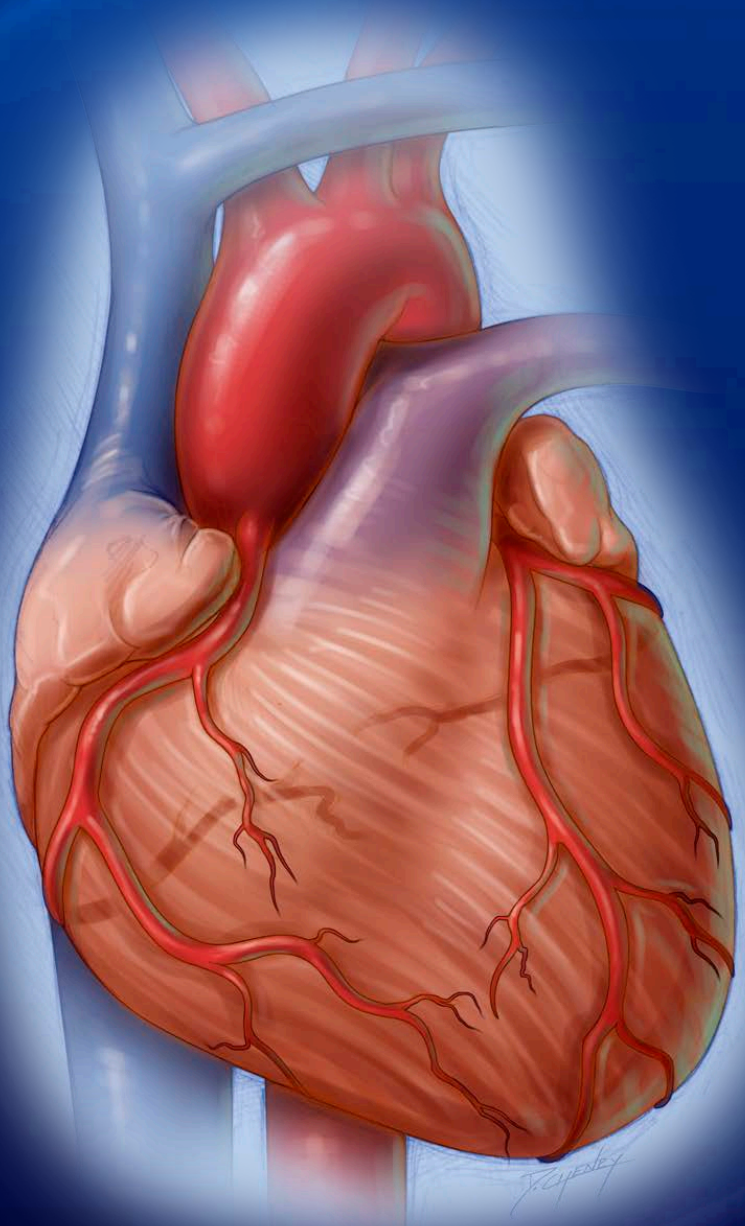

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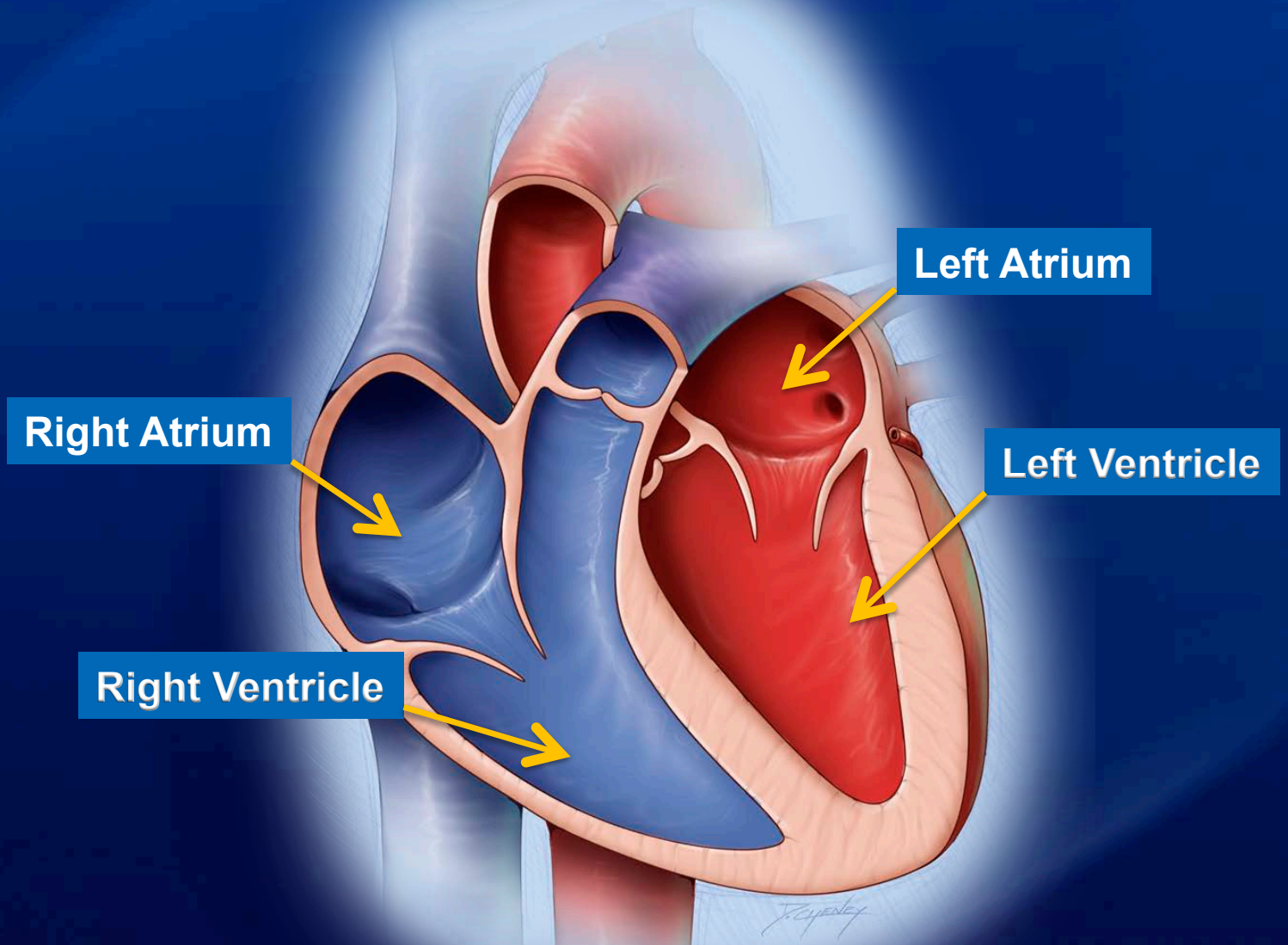

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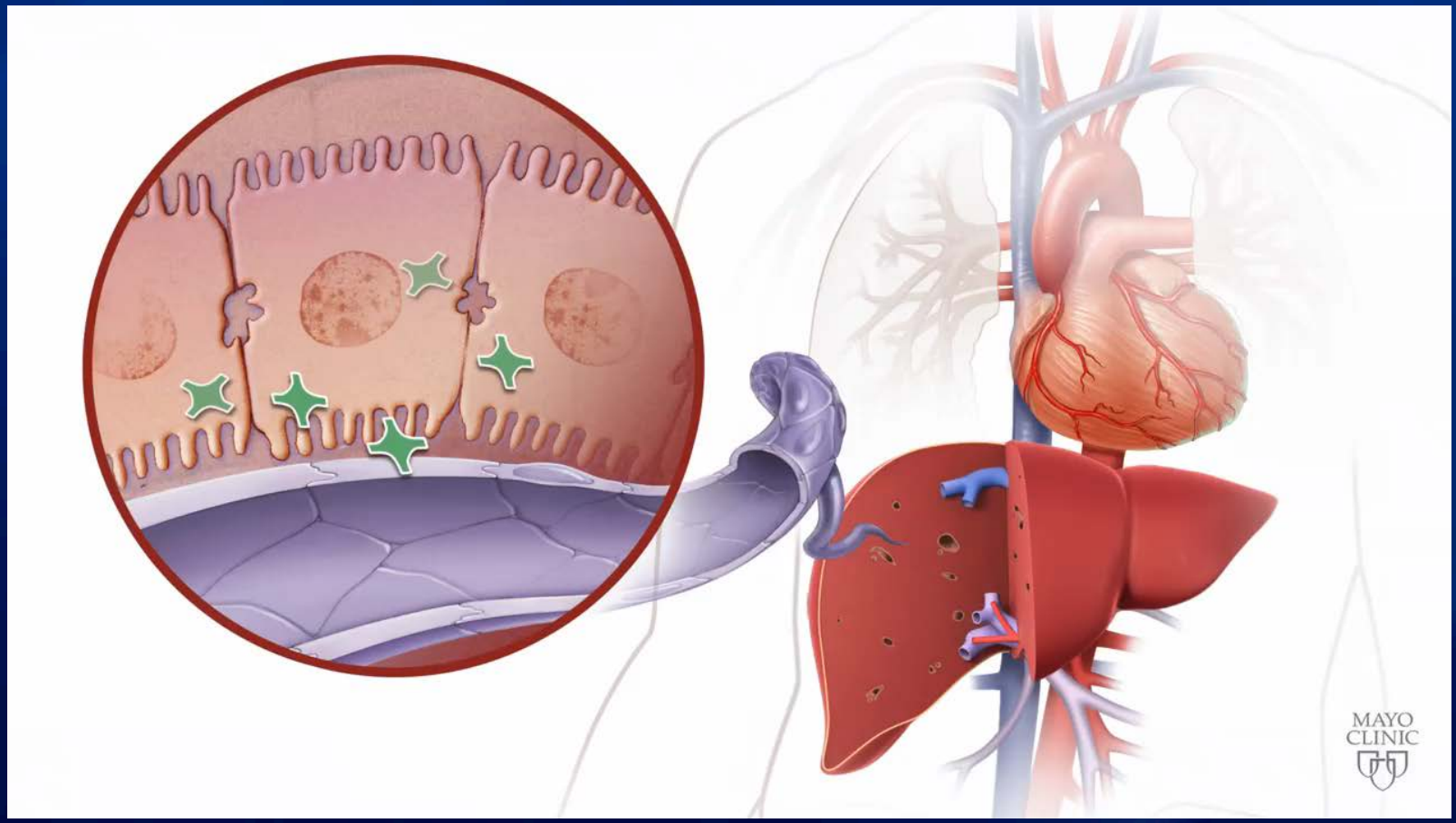
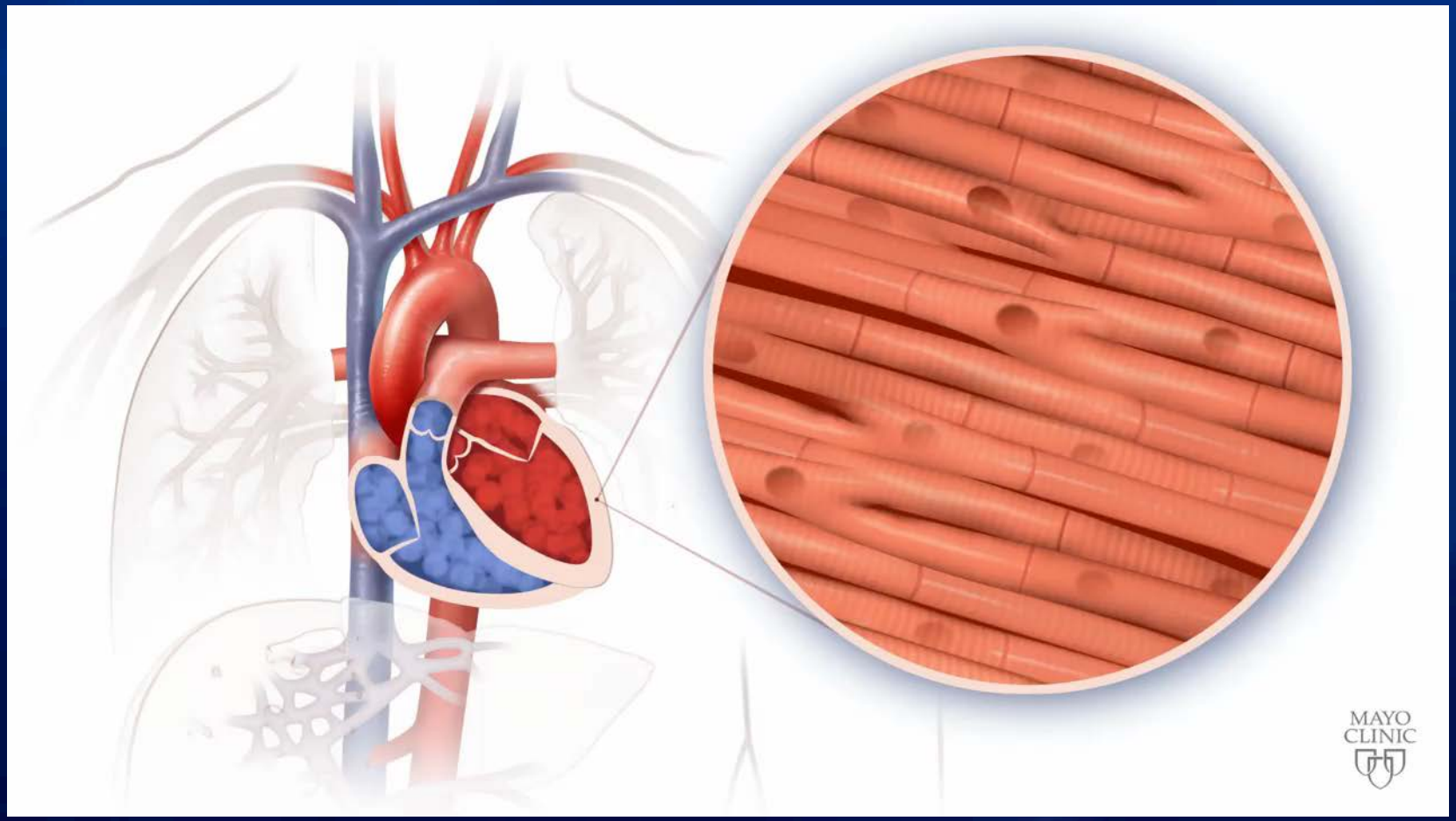
Normal Heart

Transthyretin (TTR) Amyloid



Normal Heart Muscle Cells



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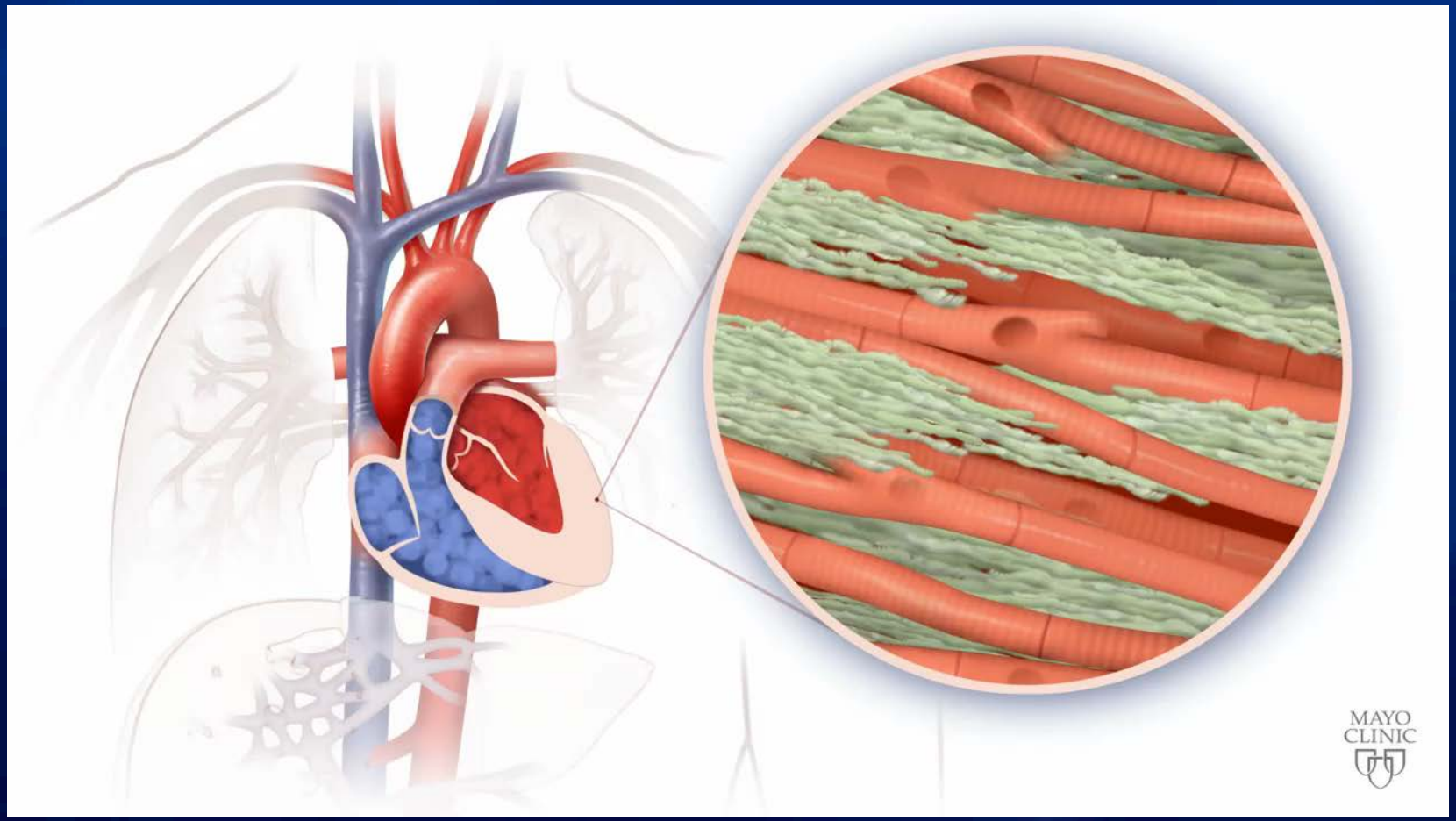
Heart Muscle Infiltrated by Amyloid fibrils (green)



What is Ejection Fraction

- The percentage of blood that is ejected with each heart beat
- It is not 100% because the heart needs to stay primed, so it never empties completely
- Normal ejection fraction is 55-70%

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- In amyloid the ejection fraction is often normal, but the heart is stiff – because of amyloid infiltration
- The heart should be very elastic, and able to relax (recoil) after it contracts
- In amyloid the stiff heart muscle doesn't relax well, making it difficult to fill
- Because the heart does not fill well, it does not pump much blood around
- Pressure builds up in the heart chambers because the body is trying to fill it with blood

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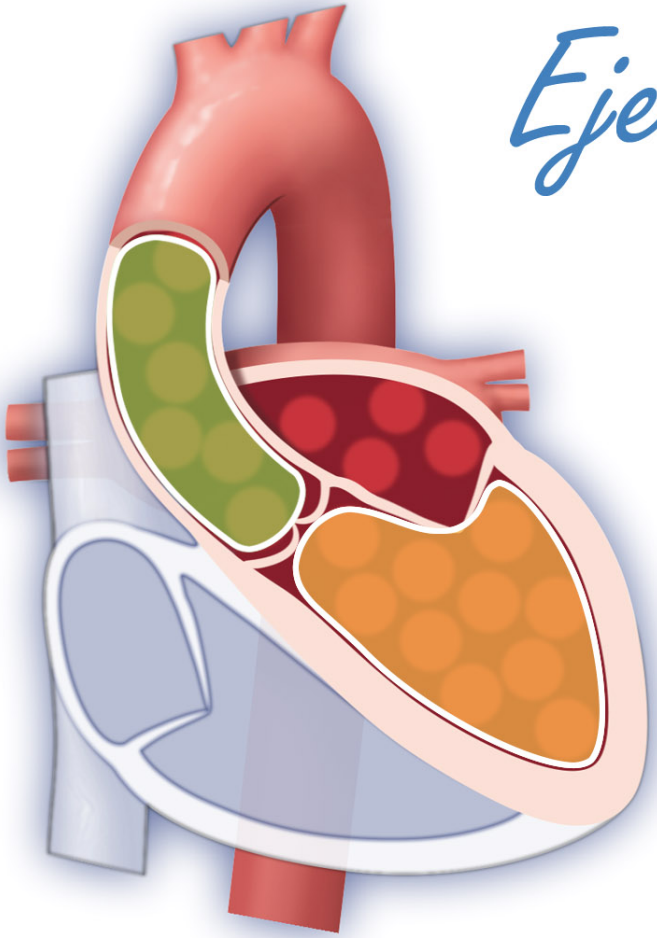
Percentage of blood that leaves (is ejected) with each heart beat.



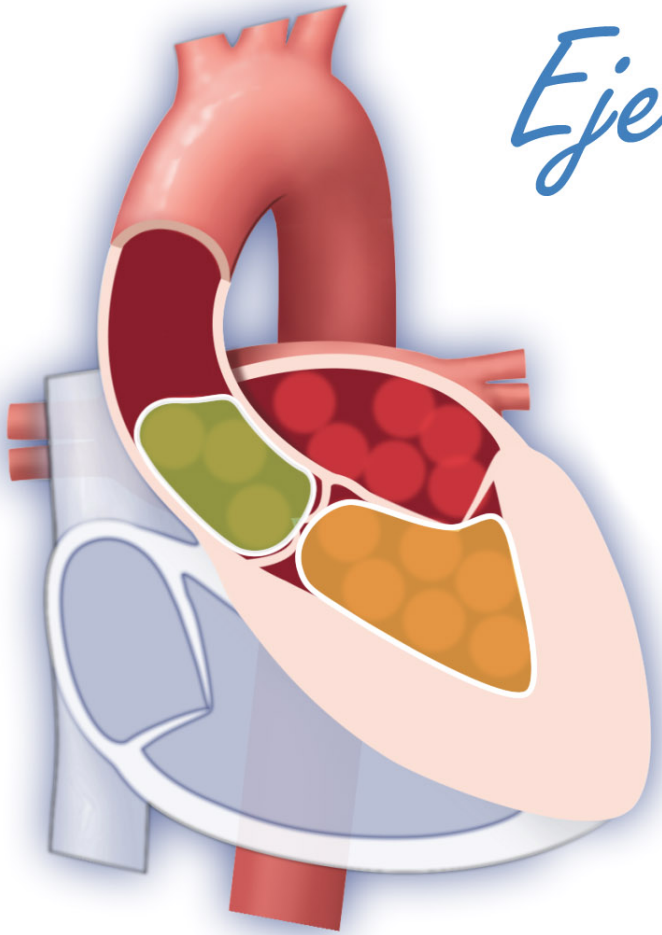
Ejection Fraction =

(Volume Ejected)

(Volume at Start)

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

Ejection Fraction in Cardiac Amyloid
The heart is stiff so it does not enlarge



Ejection Fraction =

(Volume Ejected)

(Volume at Start)

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

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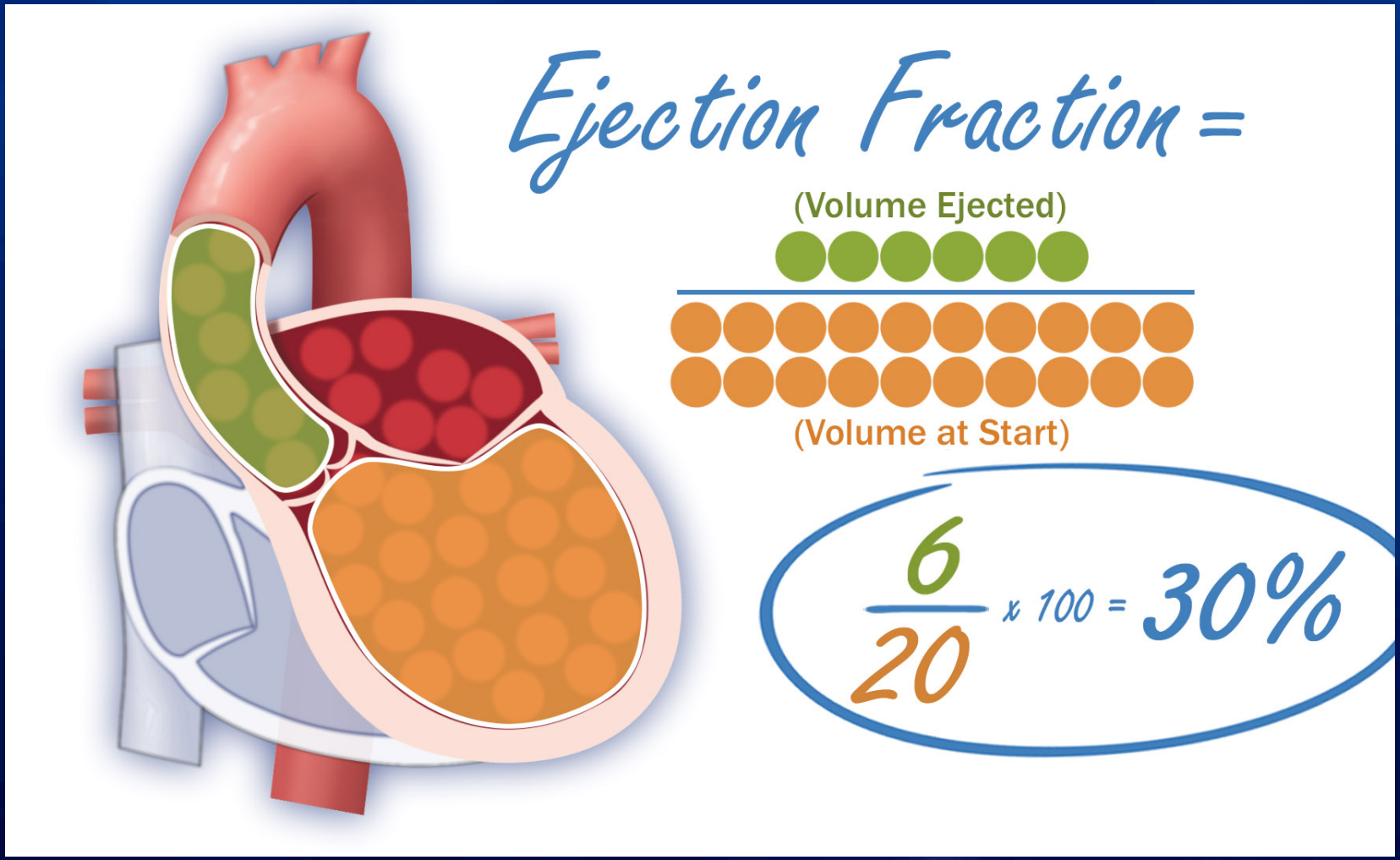
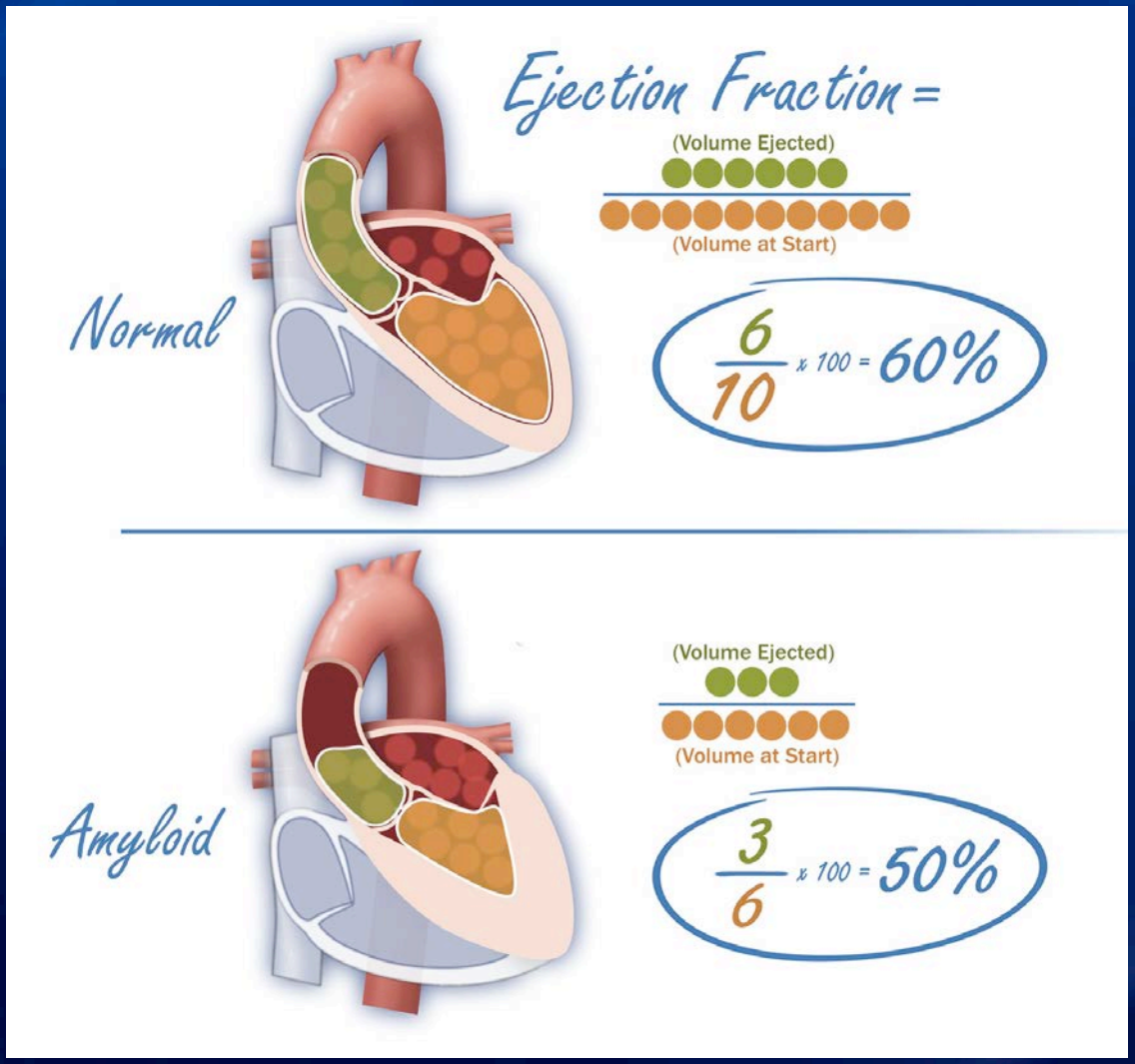
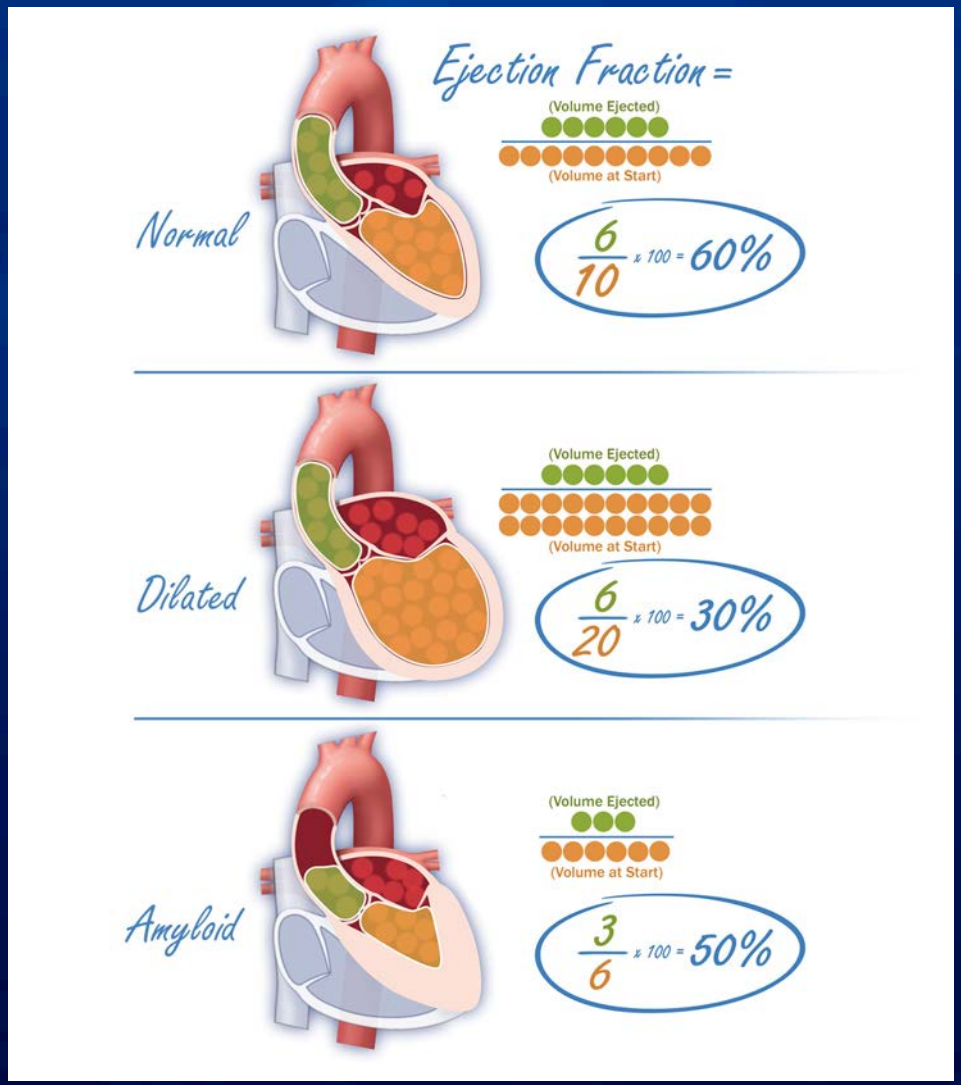
Weak heart muscle, the heart enlarges to compensate




Ejection Fraction =
(Volume Ejected)
(Volume at Start)
Normal
6/10 x 100 = 60%
(Volume Ejected)
(Volume at Start)
Amyloid
3/6 x 100 = 50%



Notice that the dilated heart with an ejection fraction of 30% pumps as much blood around as the normal heart. The amyloid heart has an ejection fraction of 50% but is pumping only half as much blood around because it is not filling well.

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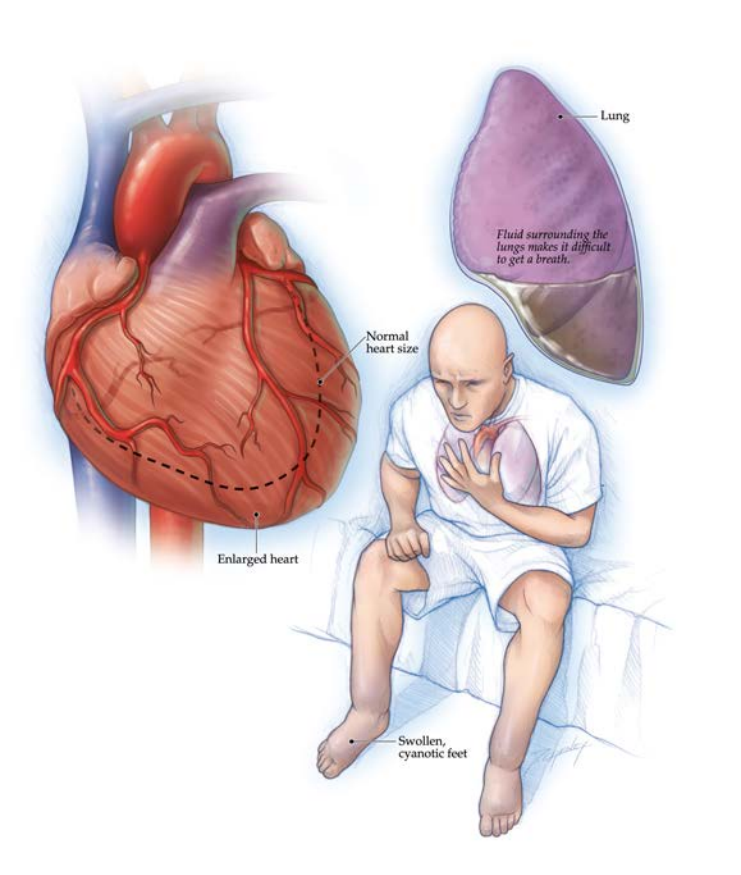
Symptoms and Signs of Heart Failure

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



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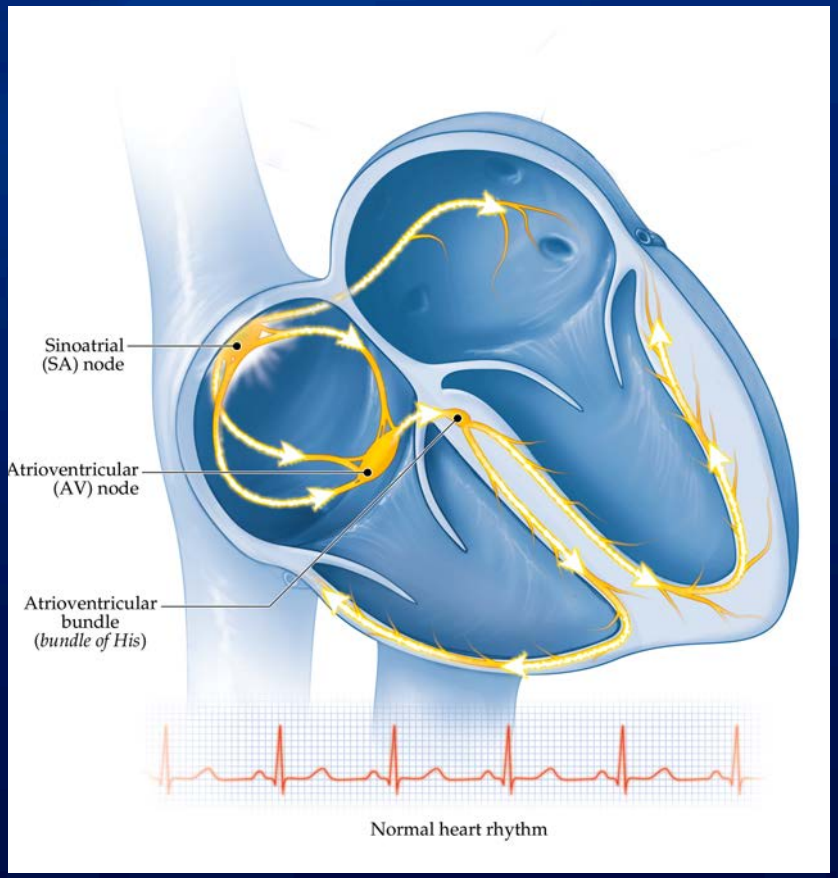
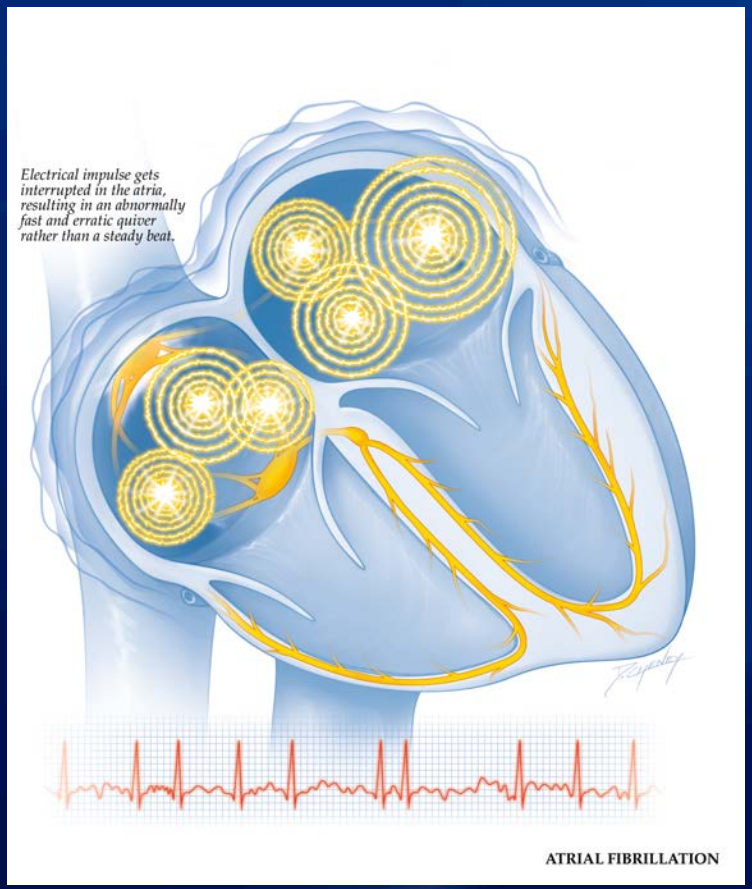
Heart Rhythm problems (Arrhythmias)





Normal Rhythm

Atrial Fibrillation

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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

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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
- Biopsy

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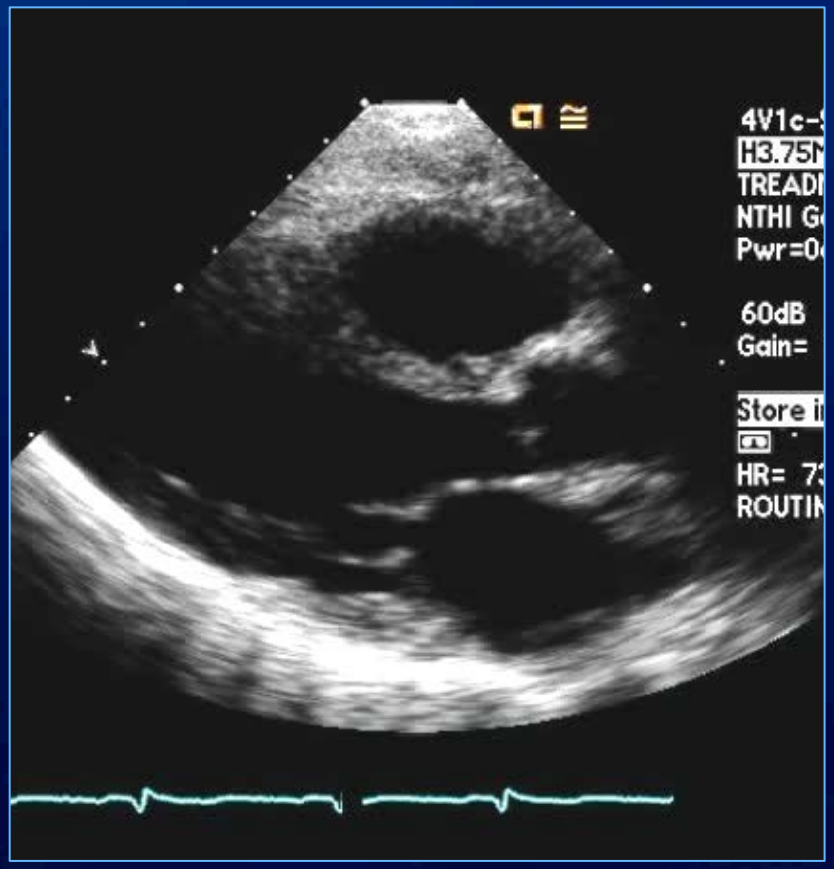
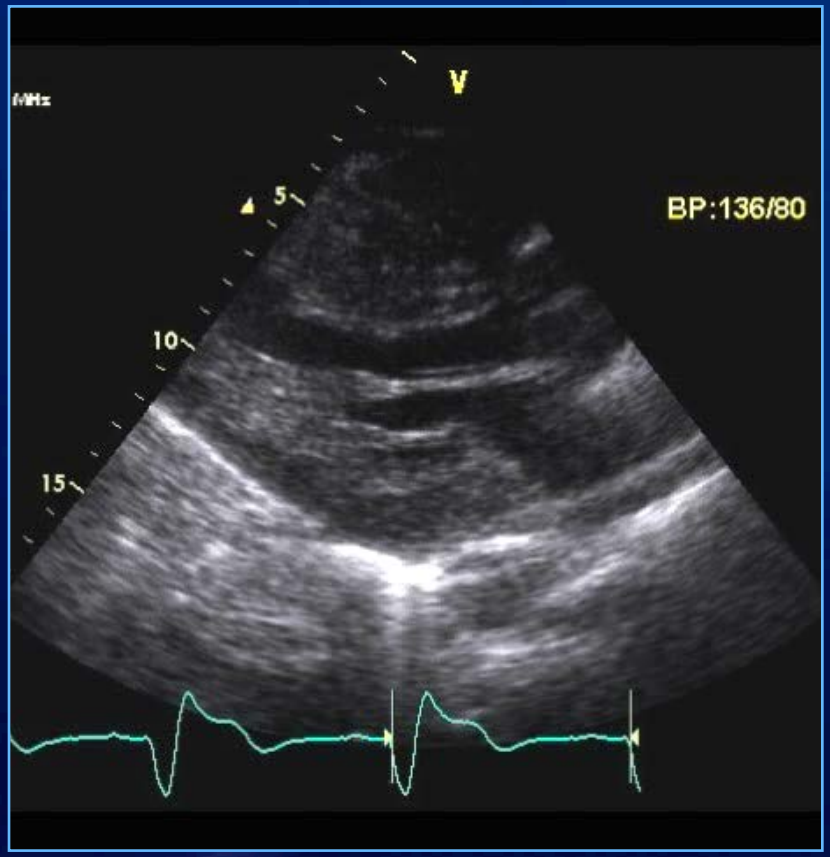
Cardiac Amyloid
Not all about wall thickening

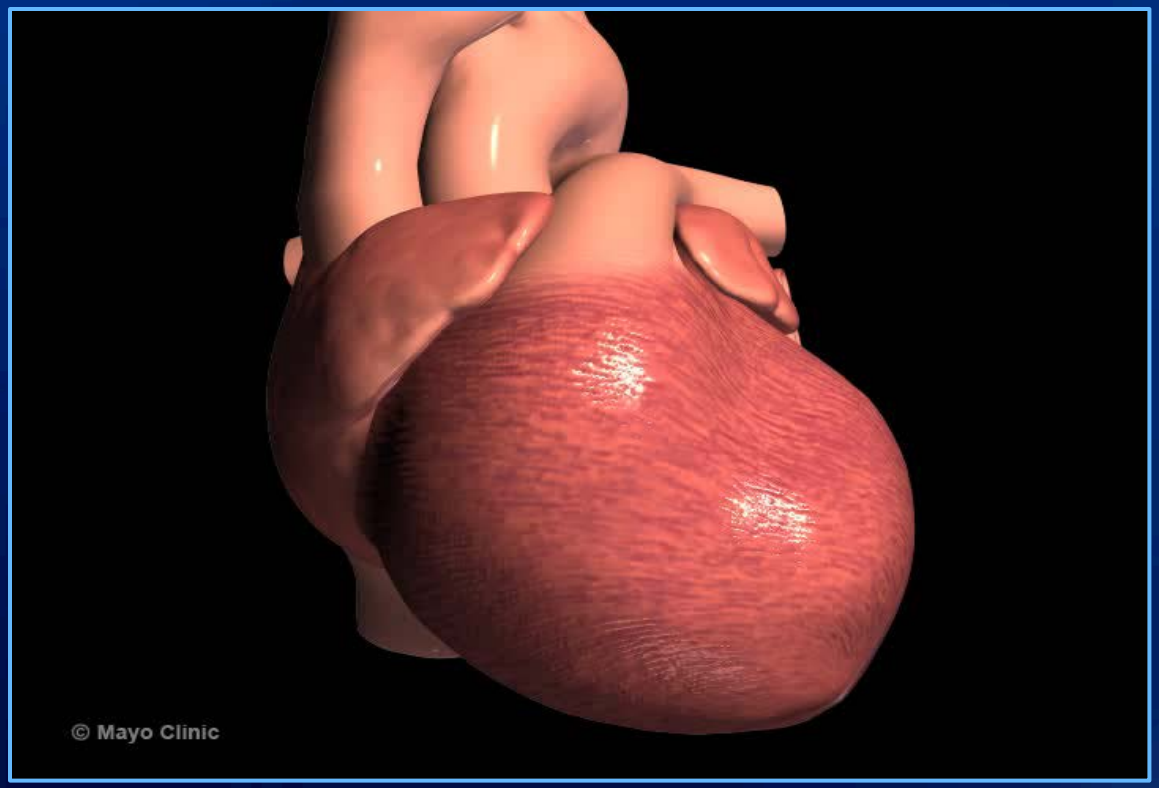
AL: End stage Heart Failure



TTR: Walking 3 miles/day



Cardiac Twist and Torsion
Heart function is complex



Courtesy of Dr. Jae Oh

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 presure in heart
  - Varies up to 40% over a week
  - Trend is more important than one number

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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

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Advanced Therapy for Heart Failure

- Artificial heart pumps
  - Ventricular assist device (VAD)
  - Total artificial heart
- Heart Transplant

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1st Pacemaker

1st Artificial Heart

Cardiac Amyloidosis

- 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

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