

# Amyloidosis and the Heart

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## **Cardiac Amyloidosis**

- Normal Heart Function
- How Amyloid affects the heart
- Explanation of Heart Tests
- How do I know if my heart is better, worse, the same?
- What can I do to help my heart?



#### **Right Atrium**

#### **Right Ventricle**

#### Left Atrium

#### Left Ventricle



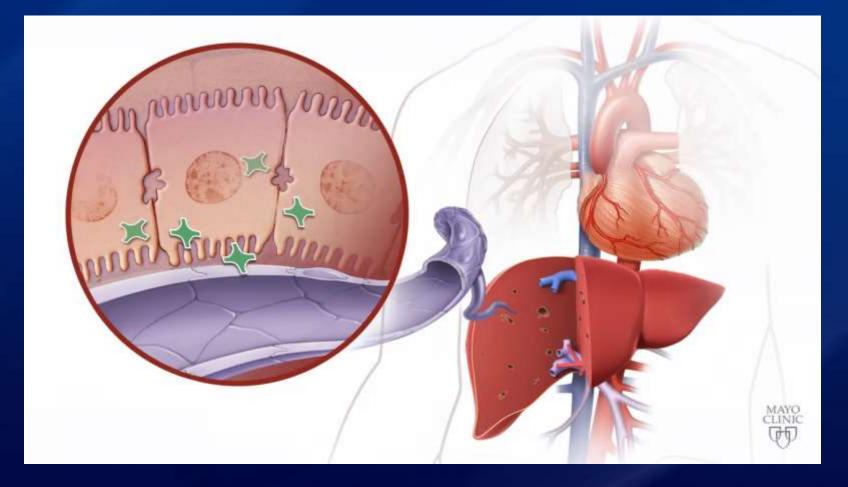
## How the Heart Works





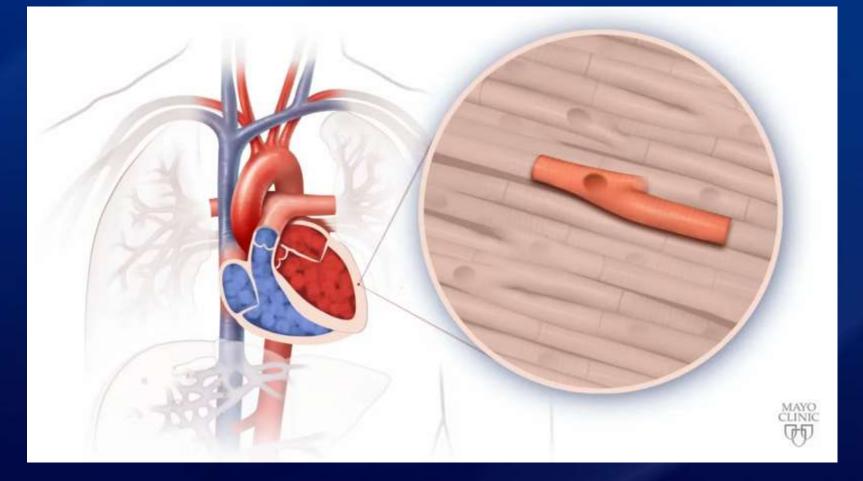
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## Transthyretin (TTR) Amyloid



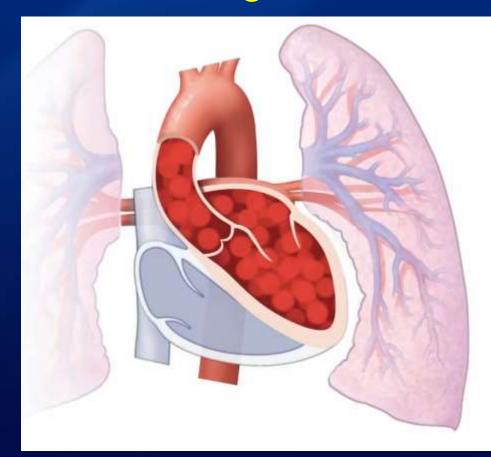


## Heart Muscle with Amyloid





## Fluid in Lungs due to Heart Failure

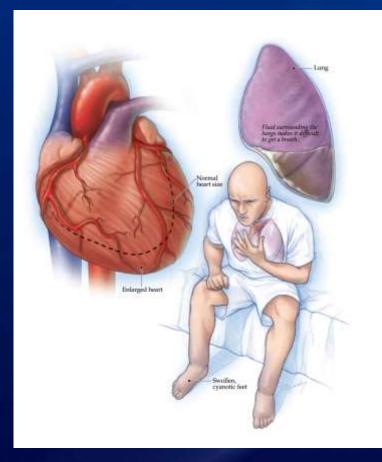






## Symptoms and Signs of Heart Failure

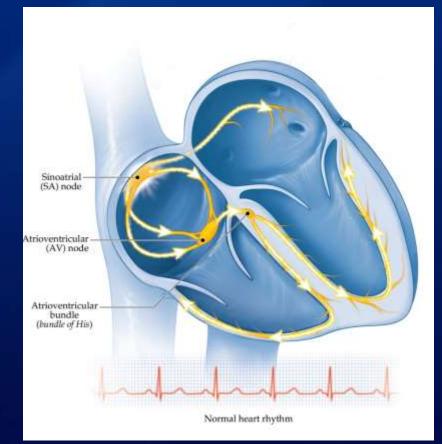
- Fatigue
- Shortness of Breath
- Swelling (edema)
- Unable to lie down due to shortness of breath
- Waking up gasping for air
- Cough, often at night

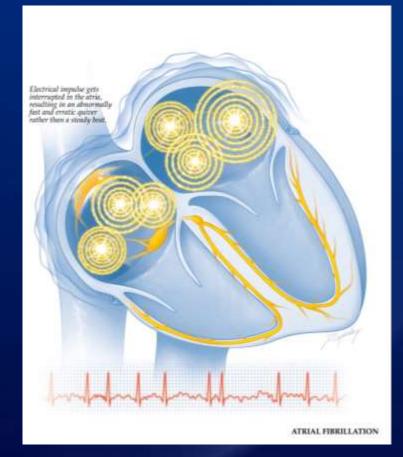






## Heart Rhythm problems (Arrhythmias)





### Normal Rhythm

#### **Atrial Fibrillation**



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## Heart Rhythm Problems in Amyloid

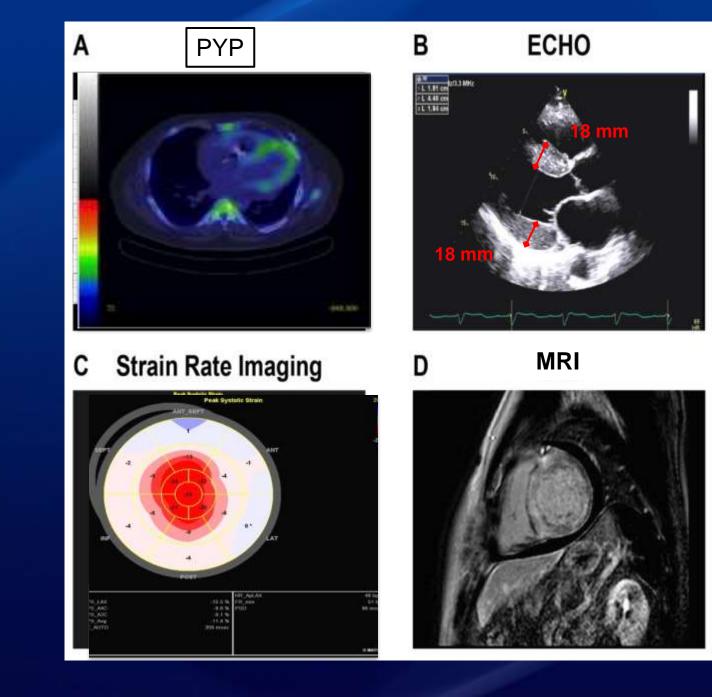
- Bradycardia too slow may need pacemaker
- Tachycardia too fast –
- Atrial fibrillation irregular rhythm from upper chambers
  - Medications
  - Electrical shock (cardioversion)
  - Risk of blood clot stroke need blood thinners
- Defibrillator for arrhythmias from ventricles



## Heart Tests to Diagnose Cardiac Amyloid

- Echo often first suspicion of amyloid
  - Measure thickness, pumping function, stiffness, valve function, pressure in lungs
- MRI certain patterns suggest amyloid
- Biopsy Heart (or fat, other organ, if echo suggests amy)
- For ATTR PYP scan may <u>sometimes</u> replace biopsy
  - Only if your blood and urine tests show no evidence of light chain (AL) amyloid
  - If you did not have a biopsy, only PYP ask your doctor these questions: what are my free light chains, what did the immunofixation of my serum and urine show, is there any monoclonal protein?

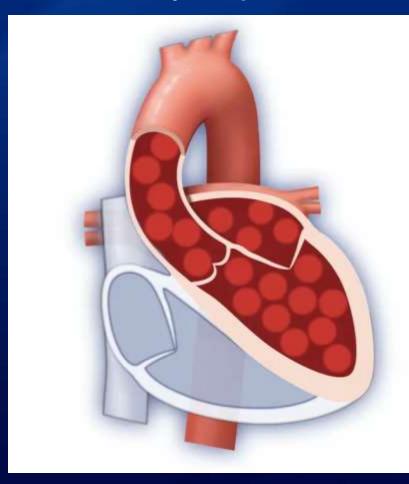




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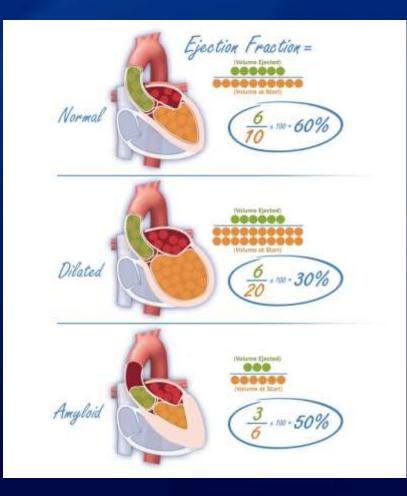
## What is Ejection Fraction? A not very important number!







## **Ejection Fraction is a PERCENTAGE**



- Top 2 examples: heart is pumping same amount of blood out with each heart beat but ejection fraction (%) is twice as high on the first one
- 2<sup>nd</sup> one has weak heart muscle (EF 30%) but heart has enlarged to compensate, so amount of blood pumped is maintained
- 3<sup>rd</sup> one is stiff heart due to amyloid- it can pump fine but does not fill well, so it is not pumping as much blood around



Normal EF Does Not Mean That Heart Function Is Normal!



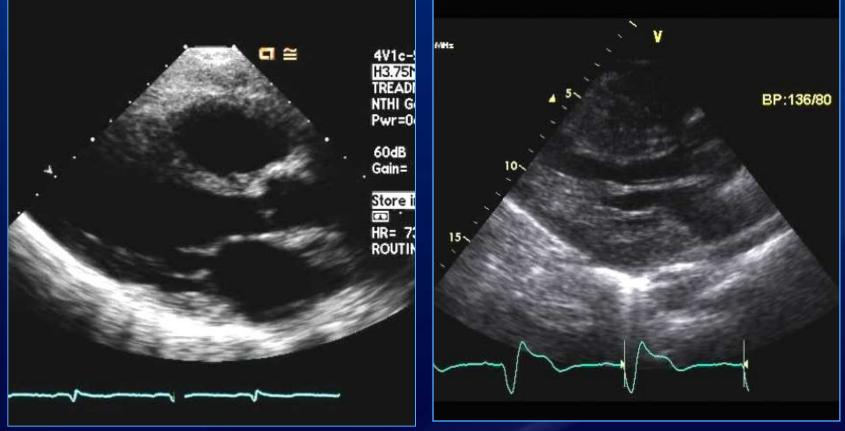
## Ejection Fraction (EF) is a <u>PERCENTAGE</u>

- <u>Absolute amount</u> of blood your heart is pumping is more important than the percentage
- In amyloid, the amount of blood pumped might be okay at rest, but your heart might not be able to keep up with exercise
- Instead of asking about EF, ask your doctor:
  - What is my <u>cardiac index</u>? Amount of blood pumped per minute for your size
  - What is my stroke volume index? Amount of blood pumped per heart beat for your size
  - These are available on most echo reports or from heart catheterization, but they need to be carefully measured



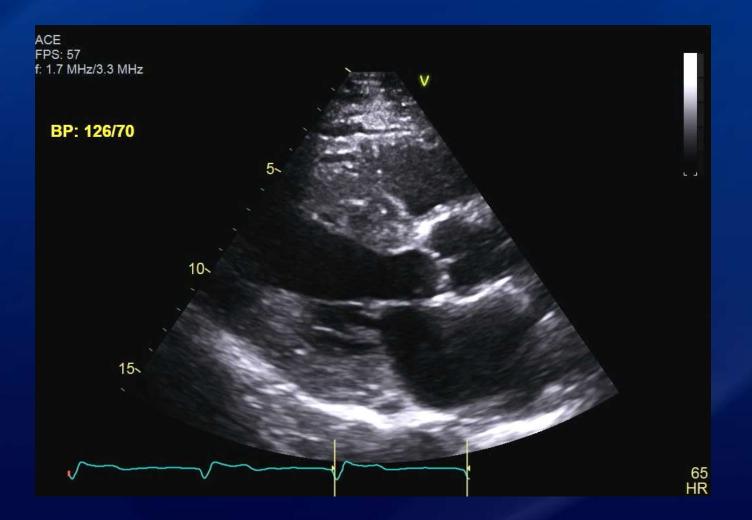
## Cardiac Amyloid Not all about wall thickening

#### AL: End stage Heart Failure TTR: Walking 3 miles/day





## **Echocardiogram in Amyloid**





# Measuring Heart Wall Thickness



Septum = 17 mm Posterior wall = 17 mm



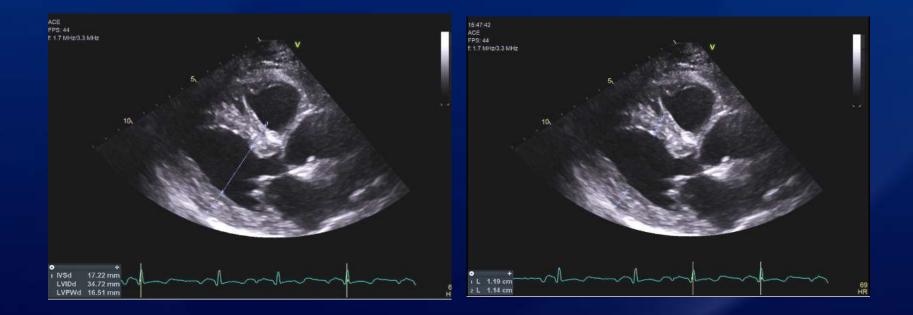
## Measuring Heart Wall Thickness



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#### Septum = 12 mm Posterior wall = 11 mm

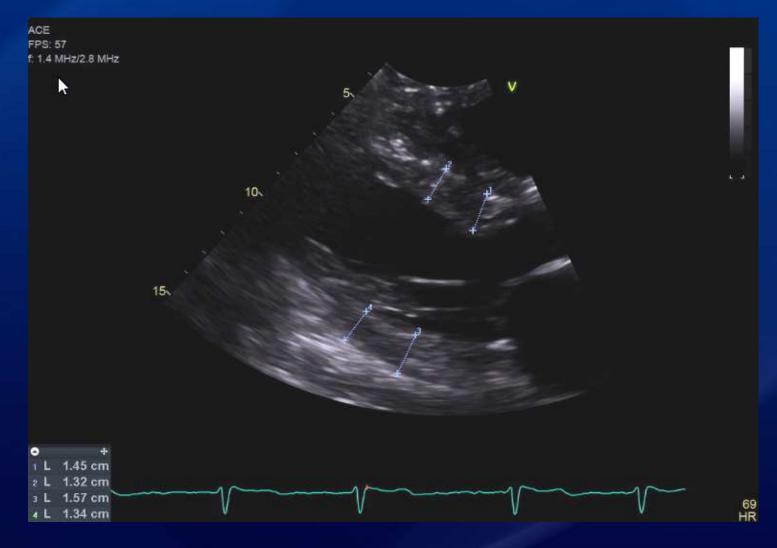
Measuring Heart Wall ThicknessSeptum = 17 mmSeptum = 12 mmPosterior wall = 17 mmPosterior wall = 11 mm



### Measurements on the same day!



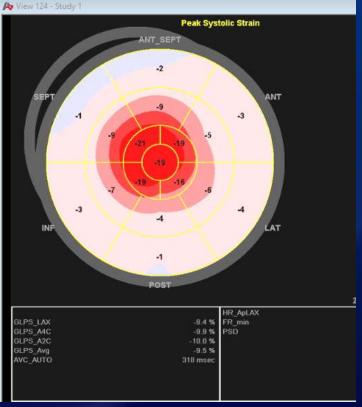
## Wall Thickness – Not Uniform Structures





## Strain- detailed look at heart function







## Cardiac Twist and Torsion Heart function is complex



#### Courtesy of Dr. Jae Oh

- There is not a single number that tells how your heart is doing
- You need a cardiologist who really understands amyloid to help youi interpret your heart function
- Pictures of the heart (echo, PYP, MRI) are best for <u>diagnosing</u> amyloid and don't help that much with follow up



## **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



## **Treatment of Cardiac Amyloid**

- Stop the source of amyloid –new drugs!
- Over time, the body can remove amyloid
- Medication to take amyloid out of heart?
  - Studies ongoing
- Diuretics (water pills) decrease shortness of breath and remove fluid
- Medications used for other type of heart failure often not helpful (beta-blockers, ACE-inhibitors)
  - Individualized treatment



# Follow-up for Cardiac Amyloid

- How do you feel?
- How far can you walk ?
- How often have you been hospitalized?
- Are you requiring more diuretic?
- What do simple blood tests show? Troponin, NT-BNP (BNP), creatinine



## Cardiac Amyloid – What can you do

- Make sure you have the right diagnosis
- Weigh yourself everyday look out for fluid
- Use compression stockings, if needed
- Limit your salt and fluid
- Exercise go at your own pace but get moving
- Do some light strength training
  - The heart likes to pump to muscle not flab



## What should patients <u>and</u> caregivers do?

- Keeping your whole body healthy is crucial
- Eat, Move, Sleep!
  - Eat: 5 Fruits/Veggies per day
  - Move: At least 10 minutes per day
  - Sleep: Eight hours for most

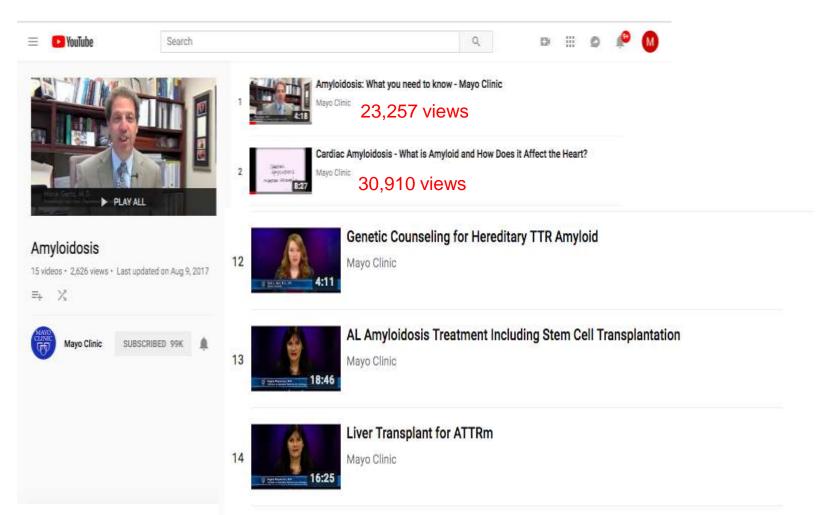


## Cardiac Amyloidosis - Summary

- 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
- Track your numbers
  - Steps and Reps!

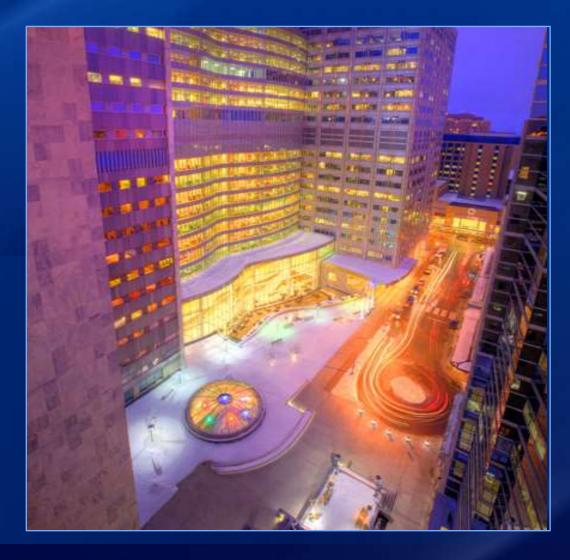


# Mayo Clinic Amyloid YouTube Channel <a href="http://bit.ly/1YuV19m">http://bit.ly/1YuV19m</a>





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## Thank you!