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?
How the Heart Works
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
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 amyloid)

- **For ATTR** – PYP scan may sometimes 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?
What is Ejection Fraction?
A not very important number!
Ejection Fraction is a PERCENTAGE

• Top 2 examples: heart is pumping the same amount of blood out with each heart beat but ejection fraction (%) is twice as high on the first one

• 2nd one has weak heart muscle (EF 30%) but heart has enlarged to compensate, so amount of blood pumped is maintained

• 3rd 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 **PERCENTAGE**

- **Absolute amount** 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 **cardiac index**? 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

BP: 126/70

ACE
FPS: 57
f: 1.7 MHz/3.3 MHz
Measuring Heart Wall Thickness

Septum = 17 mm
Posterior wall = 17 mm
Measuring Heart Wall Thickness

Septum = 12 mm
Posterior wall = 11 mm
Measuring Heart Wall Thickness

Septum = 17 mm
Posterior wall = 17 mm

Septum = 12 mm
Posterior 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

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

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 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 *and* 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

http://bit.ly/1YuV19m
Thank you!