Amyloidosis and the Heart

Martha Grogan, MD

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Cardiac Amyloidosis

- Normal Heart Function
- How Amyloid affects the heart
- Symptoms
- Explanation of Heart Tests
- Treatment Options
Normal Heart
Transthyretin (TTR) Amyloid
Normal Heart Muscle Cells
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%
• 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
Percentage of blood that leaves (is ejected) with each heart beat.

\[
\text{Ejection Fraction} = \frac{\text{(Volume Ejected)}}{\text{(Volume at Start)}} \times 100
\]

\[
\frac{6}{10} \times 100 = 60\%
\]
Ejection Fraction in Cardiac Amyloid

The heart is stiff so it does not enlarge

\[
\text{Ejection Fraction} = \frac{\text{Volume Ejected}}{\text{Volume at Start}} \times 100
\]

\[
\frac{3}{6} \times 100 = 50\%
\]
Weak heart muscle, the heart enlarges to compensate

\[ \text{Ejection Fraction} = \frac{\text{Volume Ejected}}{\text{Volume at Start}} \]

\[ \frac{6}{20} \times 100 = 30\% \]
Ejection Fraction = \[
\frac{\text{Volume Ejected}}{\text{Volume at Start}} \times 100
\]

**Normal**

\[
\frac{6}{10} \times 100 = 60\%
\]

**Amyloid**

\[
\frac{3}{6} \times 100 = 50\%
\]
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.
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 amyloid is first suspected due to abnormal echo
  - Measure thickness, pumping function, stiffness, valve function, pressure in lungs
- MRI – certain patterns suggest amyloid
- Biopsy
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 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
- 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
Advanced Therapy for Heart Failure

• Artificial heart pumps
  • Ventricular assist device (VAD)
  • Total artificial heart

• Heart Transplant
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