Thoracic Aortic Aneurysm: when to intervene?

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No Disclosures
Normal Adult Thoracic Aortic Diameters

<table>
<thead>
<tr>
<th>Thoracic Aorta</th>
<th>Range of Reported Mean (cm)</th>
<th>Reported SD (cm)</th>
<th>Assessment Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>Root (female)</td>
<td>3.50 to 3.72</td>
<td>0.38</td>
<td>CT</td>
</tr>
<tr>
<td>Root (male)</td>
<td>3.63 to 3.91</td>
<td>0.38</td>
<td>CT</td>
</tr>
<tr>
<td>Ascending (female, male)</td>
<td>2.86</td>
<td>NA</td>
<td>CXR</td>
</tr>
<tr>
<td>Mid-descending (female)</td>
<td>2.45 to 2.64</td>
<td>0.31</td>
<td>CT</td>
</tr>
<tr>
<td>Mid-descending (male)</td>
<td>2.39 to 2.98</td>
<td>0.31</td>
<td>CT</td>
</tr>
<tr>
<td>Diaphragmatic (female)</td>
<td>2.40 to 2.44</td>
<td>0.32</td>
<td>CT</td>
</tr>
<tr>
<td>Diaphragmatic (male)</td>
<td>2.43 to 2.69</td>
<td>0.27 to 0.40</td>
<td>CT, arteriography</td>
</tr>
</tbody>
</table>

2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease: Executive Summary
Definition

• **Aneurysm (or true aneurysm):** a permanent localized dilatation of an artery, having at least a 50% increase in diameter compared to the expected normal diameter of the artery in question. Although all 3 layers (intima, media, and adventitia) may be present, the intima and media in large aneurysms may be so attenuated that in some sections of the wall they are undetectable.
Definition

- **Pseudoaneurysm (or false aneurysm):** contains blood resulting from disruption of the arterial wall with extravasation of blood contained by periarterial connective tissue and not by the arterial wall layers.

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- **Ectasia**: arterial dilatation less than 50% of normal arterial diameter.

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Locations

• 60% Root/Ascending Aorta
• 40% Descending Aorta
• 10% Aortic Arch
• 10% Thoracoabdominal Aorta

Etiology

- Degenerative
- Bicuspid Aortic Valve Aortopathy
- Marfan Syndrome
- Familial Thoracic Aortic Aneurysm
- Non-Marfan Genetic Aortopathy

Canadian Cardiovascular Society Position Statement on the Management of Thoracic Aortic Disease
Risk Factors

• Smoking
• Hypertension
• Atherosclerosis
• Genetic Connective Tissue Disorders (Marfan and Ehlers-Danlos syndromes)
• Bicuspid/Unicuspid aortic valves
• Syphilis

Cardiac Surgery in the Adult 2nd edition, 2003
Incidence

5.9 cases/100,000 person years (13\textsuperscript{th} cause of death)

Cardiac Surgery in the Adult, 2\textsuperscript{nd} edition, 2003
Should We Be Concerned?

• CDC:
  – Average of 43,199 deaths/yr (could be > 46,000)
  – Breast Cancer, Pancreatic Cancer, Colon Cancer, Homicides, MVA

• Very little research and even less funding dedicated to this disease

Thoracic Aneurysm is a lethal disease

Average yearly rate of rupture/dissection/death

Aneurysm size has a profound impact on rupture/dissection/death

Kaplan Meier Cumulative Survival at 5 yrs

Elective surgical repair restores survival to near normal.
Screening

• Asymptomatic
• Not easily detectable.....until **acute** and often **catastrophic complication** occurs (rupture/dissection/death)
Bicuspid Aortic Valve and Congenital Variants

- Prevalence of Aortic dilatation of ascending aorta in patients with Bicuspid aortic valve ranges 20-84%  
- All patients with bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation  
- All first-degree relatives of patients with bicuspid aortic valve, familial thoracic aneurysm, premature thoracic disease and dissection should be evaluated for bicuspid aortic valve and asymptomatic aortic disease

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Genetic Syndromes

• **Marfans:**
  – Echocardiogram at time of initial diagnosis
  – Annual imaging of aorta to document diameter stability

• **Loeys-Dietz or genetic mutation:**
  – Complete aortic imaging at initial diagnosis and 6 months thereafter.
  – MRI yearly from cerebrovascular circulation to pelvis

• **Turner Syndrome:**
  – Imaging of heart and aorta- Bicuspid valve/coarctation/dilatation of aorta

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Familial Thoracic Aortic Aneurysm and Dissection

- Aortic Imaging for first-degree relatives
- Specific gene mutation (FBN1, TGFBR1, TGFBR2, etc) genetic counseling recommended for first-degree relatives
- Aortic Imaging of second-degree relatives if first-degree relatives are known to have aneurysm and/or dissection.

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When do we operate?
Symptomatic Patients with Thoracic Aortic Aneurysm

- Symptoms suggestive of expansion/rupture and/or dissection require prompt surgical intervention
Asymptomatic Patients With Ascending Aortic Aneurysm

- Degenerative thoracic aneurysm, chronic dissection, intramural hematoma, penetrating ulcer in whom ascending aorta or aortic sinus diameter >5.5 cm----Evaluated for surgical repair.
- Growth rate of more than 0.5cm/y in an aorta less than 5.5 cm--Evaluated for repair.
- Patients undergoing aortic valve repair/replacement with ascending aorta or aortic root > 4.5 cm----Evaluated for repair
Asymptomatic Patients With Ascending Aortic Aneurysm (Genetic/Familial)

- Elective operations at smaller diameters: **4.0-5.0 cm**
- Loeys-Dietz syndrome or confirmed TGFBR1/TGFBR2 mutation to undergo aortic repair when aortic diameter reaches **4.2 cm**
- Women with Marfan syndrome contemplating pregnancy, reasonable to replace aortic root and ascending aorta if diameter exceeds **4.0 cm**
Asymptomatic Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysm

• Chronic dissection in low risk pts, especially those with connective tissue disorder if aortic diameter **exceeds 5.5 cm, open repair recommended**
• Degenerative or traumatic aneurysm **exceeding 5.5 cm**, saccular aneurysms, pseudoaneurysms—**consider endovascular repair**
• Thoracoabdominal aneurysm with limited endovascular options, **elective open surgery recommended** when aortic diameter **exceeds 6.0 cm.**
Asymptomatic Patient with Ascending Aortic Aneurysm and Bicuspid Valve

• Operative intervention is indicated in patients with BAV if the diameter of the aortic root or ascending aorta is **5.5 cm or greater** (I-B-NR)

• Operative intervention is reasonable in patients with BAV if the diameter of the aortic root or ascending aorta is **5.0 cm or greater** and an additional risk factor for dissection is present. (IIa-B-NR)

• Replacement of the ascending aorta is reasonable in patients with BAV undergoing AVR because of severe aortic stenosis or aortic regurgitation when the diameter of the ascending aorta is greater than **4.5 cm** (IIa-C)

Surgery for Aortic Dilatation in Patients With Bicuspid Aortic Valves
A Statement of Clarification From the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines
Table 2. Recommended size thresholds for intervention for asymptomatic thoracic aortic aneurysms*

<table>
<thead>
<tr>
<th>Condition</th>
<th>Aortic root</th>
<th>Ascending</th>
<th>Arch</th>
<th>Descending</th>
</tr>
</thead>
<tbody>
<tr>
<td>Degenerative</td>
<td>5.5 cm</td>
<td>5.5 cm</td>
<td>6.0 cm</td>
<td>6.5 cm</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>5.0-5.5 cm</td>
<td>5.0-5.5 cm</td>
<td>5.5 cm</td>
<td>6.5 cm</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>5.0 cm†</td>
<td>5.0 cm</td>
<td>5.5-6.0 cm</td>
<td>5.5-6.0 cm</td>
</tr>
<tr>
<td>Familial aortopathy</td>
<td>4.5-5.0 cm</td>
<td>4.5-5.0 cm</td>
<td>5.5-6.0 cm</td>
<td>5.5-6.0 cm</td>
</tr>
<tr>
<td>Other genetic syndromes</td>
<td>4.0-5.0 cm</td>
<td>4.2-5.0 cm</td>
<td>5.5-6.0 cm</td>
<td>5.5-6.0 cm</td>
</tr>
<tr>
<td>Undergoing cardiac surgery</td>
<td>—</td>
<td>4.5 cm</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

*Canadian Cardiovascular Society Position Statement on the Management of Thoracic Aortic Disease*
Employment and Lifestyle in patients with Thoracic Aortic Disease

• Employment and lifestyle restrictions are reasonable, including avoidance of strenuous lifting, pushing or straining.
• Little data
• Aerobic exercise (dynamic exercise) is associated with modest increase in mean arterial pressure.
• Isometric exercise (weight lifting) is associated with substantial increase in intra-thoracic pressure and dramatic increase in mean arterial pressure (systolic > 300 mmHg).
Conclusion

- Thoracic Aneurysm is a lethal disease
- Aneurysm size has a profound impact on rupture/dissection/death
- Elective surgical repair restores survival to near normal.

Thank You