Aortic Imaging and Follow-up
"There is no condition more conducive to clinical humility than aneurysm of the aorta.”

William Osler
55 yo F suddenly develops ventricular fibrillation

– Defibrillated in the field with an AED

– Brought to ER in Sinus Tachycardia
  • Diaphoretic, SBP= 94 mmHG
  • No ST deviations by EKG
  • “Widened Mediastinum” suggested by CXR

STAT transesophageal echo (TEE):
What would you do next?
(1) Send patient to the O.R. for acute aortic dissection
(2) Bolus with heparin and consider lytics
(3) Admit for observation
(4) None of the above
She is started on IV heparin.

24 HOURS LATER
SBP drops to 85 requiring 2mcg dopamine.

Repeat PCXR suggests, once again, **widened mediastinum**

CTA OF CHEST
No ‘widened mediastinum’
Faced with indecision, Osler told his trainees:

“Look wise, say nothing, and grunt.”
Imaging is critically important to diagnosis and subsequent outcomes.
Is the heart big? Mediastinum? Aorta?
‘Widening of the mediastinum’ is dependent on many anatomic and imaging variables.
sensitivity only 67% for an ascending aortic dissection, even lower specificity-- many conditions & imaging variables can cause a widening of the mediastinum.
38 yo Female with several years of cough.

Self-referred to Aortic Clinic due to report of “tortuous aorta” on most recent CXR.
The cardiac contour is unremarkable. No infiltrate is identified. No effusion or pneumothorax.

No acute cardiopulmonary process.
Length of the ascending aorta (aortic annulus to apex arch) increases 80% between the ages of 20 and 80 years of age.
For the lengthening aorta to continue to fit in the prison of the thorax, it becomes more C shaped and tortuous (rightward unfolding of the aorta)
Aortic Surgery

Replacement of the diseased aortic section with a Dacron tube graft, sparing the aortic valve if possible.
Critical Issues

If we wait for an Acute aortic syndrome, Late risk of death: Even young patients with Acute dissection have a 5-year survival of 50% after surgery due to residual events related to unresected dissected aorta - improved to 70% in a formal aortic clinic.

This stresses the importance of:

– identification those at risk and treatment prior to an acute presentation
– Long-term close follow-up with specialists after surgery
Thoracic Aneurysm: Increasing incidence with population aging

True increasing incidence, not due to increased imaging
Incidence of thoracic aortic aneurysms and dissection is up

Operations for thoracic aortic aneurysms and dissection up

Olsson C. et al., Circulation 2006;114:2611-2618
Acute Cardiopulmonary symptoms (chest pain, dysnea,)

**ACS**
- STEMI
- NSTEMI
- Unstable Angina

**AAS**
- Acute Aortic Syndromes
- Aortic Dissection
- IMH
- PAU
- PAU

**VTE**
- Venothromboembolism

**Tamponade**
- Esophageal Rupture
- Spontaneous PTX

**ACS**
- STEMI
- NSTEMI
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Two aortic imaging studies in the first 24 hours are necessary in the majority (70%) of patients
ECG-gated CT reveals the great vessels without artifacts & Coronaries
ECG-gated CT reveals the great vessels without artifacts & Coronaries
# Advantages and disadvantages

<table>
<thead>
<tr>
<th>Imaging modality</th>
<th>Advantages</th>
<th>Limitations</th>
</tr>
</thead>
</table>
| **TTE**          | • Available and cost effective  
                  • Lack of radiation or contrast  
                  • Good visualization of proximal aorta | • Limited visualization of distal aorta  
                  • May not account for asymmetry  
                  • Image quality dependent on operator and patient factors |
| **TEE**          | • Visualization of more distal segments  
                  • Lack of radiation or contrast  
                  • Can be used intra-operatively | • Invasive procedure  
                  • Sector width of 90° limits orientation  
                  • Blind spot may limit distal assessment |
| **CT**           | • Excellent resolution  
                  • True short-axis measurements  
                  • Assessment of aortic wall pathology | • Requires ionizing radiation  
                  • Contrast dye exposure  
                  • Limited valvular assessment |
| **MRI**          | • Good spatial and temporal resolution  
                  • True short-axis measurements  
                  • Allows tissue characterization and valvular/ventricular function assessment  
                  • Aortic imaging can be done without contrast | • Cannot be performed in patients with cardiac implantable electronic devices  
                  • More prone to artifacts  
                  • Dependent on patient factors (ability to breath hold and heart rate) |
Question: A CT chest report returns “The ascending aorta measures “4.2 cm X 5.6 cm”

Which number do you use to determine whether to refer the patient for surgery?
Question: A CT chest report returns “The ascending aorta measures “4.2 cm X 5.6 cm”

Which number do you use to determine whether to send the patient to surgery?

A. Average the two numbers together
B. Use the lower of the two numbers in management, to prevent sending the patient for surgery too early
C. Use the higher of the two numbers, to prevent sending the patient for surgery too late
D. None of the above
Insist on repeat measurements perpendicular to flow at multiple points within the aorta, indexed to body surface area to refine the risk
ACCF/AHA Guideline

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

A Report of the American College of Cardiology Foundation/American Heart Association
Task Force on Practice Guidelines, American Association for Thoracic Surgery, American
College of Radiology, American Stroke Association, Society of Cardiovascular
Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of
Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine

Endorsed by the North American Society for Cardiovascular Imaging

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1. The location at which the aorta is abnormal (see Section 2).

2. The maximum diameter of any dilatation, measured from the external wall of the aorta, perpendicular to the axis of flow, and the length of the aorta that is abnormal.

3. For patients with presumed or documented genetic syndromes at risk for aortic root disease measurements of aortic valve, sinuses of Valsalva, sinotubular junction, and ascending aorta.

4. The presence of internal filling defects consistent with thrombus or atheroma.

5. The presence of IMH, PAU, and calcification.

6. Extension of aortic abnormality into branch vessels, including dissection and aneurysm, and secondary evidence of end-organ injury (eg, renal or bowel hypoperfusion).

7. Evidence of aortic rupture, including periaortic and mediastinal hematoma, pericardial and pleural fluid, and contrast extravasation from the aortic lumen.

8. When a prior examination is available, direct image to image comparison to determine if there has been any increase in diameter.

IMH indicates intramural hematoma; and PAU, penetrating atherosclerotic ulcer.
Careful imaging and measuring is time-consuming BUT accurate size is, beyond symptoms, the best intervention criterion in 2018.
<table>
<thead>
<tr>
<th>Aortic Location</th>
<th>Measurement (orthogonal to axis of flow)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic Annulus</td>
<td></td>
</tr>
<tr>
<td>Sinus of Valsalva</td>
<td></td>
</tr>
<tr>
<td>Sinotubular Ridge</td>
<td></td>
</tr>
<tr>
<td>Distal Ascending prior to 1st arch vessel</td>
<td></td>
</tr>
<tr>
<td>Proximal Descending after last arch vessel (Isthmus)</td>
<td></td>
</tr>
<tr>
<td>Mid descending at level of left atrium</td>
<td></td>
</tr>
<tr>
<td>Distal descend at aortic hiatus</td>
<td></td>
</tr>
<tr>
<td><strong>Maximum Aorta Dimension:</strong></td>
<td></td>
</tr>
<tr>
<td>Ascending Aorta</td>
<td></td>
</tr>
<tr>
<td>Descending Aorta</td>
<td></td>
</tr>
<tr>
<td>Body Surface Area (Dubois)</td>
<td>m²</td>
</tr>
<tr>
<td>Aortic Size Index (ASI)</td>
<td>cm/m²</td>
</tr>
</tbody>
</table>

**ASI Interpretation Guide**

<table>
<thead>
<tr>
<th>Risk Category (combined endpoint of rupture, dissection, or death)</th>
<th>Aortic Size Index (ASI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low risk (4% per year)</td>
<td>&lt;2.75 cm/m²</td>
</tr>
<tr>
<td>Moderate risk (8% per year)</td>
<td>2.75-4.24 cm/m²</td>
</tr>
<tr>
<td>High risk (20% per year)</td>
<td>&gt;4.25 cm/m²</td>
</tr>
</tbody>
</table>

## 2010 Guidelines on Thoracic Aortic Disease

### Table 17. Suggested Follow-Up of Aortic Pathologies After Repair or Treatment

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Interval</th>
<th>Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute dissection</td>
<td>Before discharge, 1 mo, 6 mo, yearly</td>
<td>CT or MR, chest plus abdomen TTE</td>
</tr>
<tr>
<td>Chronic dissection</td>
<td>Before discharge, 1 y, 2 to 3 y</td>
<td>CT or MR, chest plus abdomen TTE</td>
</tr>
<tr>
<td>Aortic root repair</td>
<td>Before discharge, yearly</td>
<td>TTE</td>
</tr>
<tr>
<td>AVR plus ascending</td>
<td>Before discharge, yearly</td>
<td>TTE</td>
</tr>
<tr>
<td>Aortic arch</td>
<td>Before discharge, 1 y, 2 to 3 y</td>
<td>CT or MR, chest plus abdomen</td>
</tr>
<tr>
<td>Thoracic aortic stent</td>
<td>Before discharge, 1 mo, 2 mo, 6 mo, yearly 0 or 30 days*</td>
<td>CXR, CT, chest plus abdomen</td>
</tr>
<tr>
<td>Acute IMH/PAU</td>
<td>Before discharge, 1 mo, 3 mo, 6 mo, yearly</td>
<td>CT or MR, chest plus abdomen</td>
</tr>
</tbody>
</table>
Where is the Future?

Gene Expression Profiles

Clinical Data
- Treatments
- Family History
- Demographics
- Environmental

Genomic Data
- SNP’s
- Genome-scale sequence

Metabolic Data
Proteomic Data

Patterns Integration Models

Predictions:
- Risk
- Individualized Prognosis & Diagnosis
- Drug Response
- Environment (e.g. Diet) Response

Imaging
Hypertension is one risk factor for aortic aneurysms and for **acute aortic syndromes**

- Risk factors for descending aorta aneurysm & abdominal AAA is similar to those for atherosclerosis

- Risk factors for ascending aneurysms are 95% time unknown
# Etiology of Thoracic Aortic Aneurysms

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Connective tissue disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>Mutation in the gene <em>FBN1</em>; aneurysms typically involve the aortic root at the sinus level.</td>
</tr>
<tr>
<td><strong>Ehlers-Danlos syndrome type IV, vascular type</strong></td>
<td>Associated with mutations in the gene for type III procollagen, <em>COL3A1</em>.</td>
</tr>
<tr>
<td><strong>Loeys-Dietz syndrome</strong></td>
<td>Mutations in the genes <em>TGFBR1</em> or <em>TGFBR2</em>; associated with arterial tortuosity and multiple arterial aneurysms.</td>
</tr>
<tr>
<td><strong>Congenital conditions</strong></td>
<td></td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>Aneurysms most often involve the ascending aorta, less often the root at the sinus level.</td>
</tr>
<tr>
<td>Turner syndrome</td>
<td>Often associated with a bicuspid aortic valve.</td>
</tr>
<tr>
<td><strong>Genetic</strong></td>
<td></td>
</tr>
<tr>
<td>Familial thoracic aortic aneurysm syndrome</td>
<td>Known mutations are identified in 20% of families, and include <em>ACTA2</em> (14%), <em>TGFBR2</em> (4%), and <em>MYH11</em> (1%).</td>
</tr>
<tr>
<td><strong>Atherosclerosis</strong></td>
<td>Typically involves the descending aorta.</td>
</tr>
<tr>
<td><strong>Vasculitis</strong></td>
<td></td>
</tr>
<tr>
<td>Takayasu arteritis</td>
<td>May cause stenotic lesions as well as aneurysms. Affects younger population. Near universal left subclavian artery involvement.</td>
</tr>
<tr>
<td>Giant cell arteritis</td>
<td>Affects older population, particularly women, and may include symptoms of polymyalgia rheumatica.</td>
</tr>
<tr>
<td><strong>Nonspecific (idiopathic) aortitis</strong></td>
<td>Found on pathological analysis of aortic specimen at time of surgery.</td>
</tr>
<tr>
<td><strong>Other autoimmune conditions (e.g., Behçet disease, systemic lupus erythematosus)</strong></td>
<td>Rare causes of thoracic aortic aneurysm.</td>
</tr>
<tr>
<td><strong>Infectious</strong></td>
<td></td>
</tr>
<tr>
<td>Bacterial aneurysm (&quot;mycotic&quot; aneurysm)</td>
<td>Usually the result of circulating bacteria seeding an abnormal aortic wall.</td>
</tr>
<tr>
<td>Syphilitic (luetic) aortitis</td>
<td>Aneurysms are a late consequence of the infection.</td>
</tr>
<tr>
<td><strong>Aortic injury</strong></td>
<td></td>
</tr>
<tr>
<td>Prior acute aortic syndrome (aortic dissection, intramural hematoma, or penetrating atherosclerotic ulcer)</td>
<td>Sometimes the antecedent acute aortic syndrome was clinically silent or went undiagnosed.</td>
</tr>
<tr>
<td>Trauma</td>
<td>Aortic transsection typically results in formation of a pseudoaneurysm.</td>
</tr>
</tbody>
</table>
Question Which aortopathy is associated with a bifid uvula?

A. Marfan’s syndrome

B. Loeys-Dietz syndrome

C. Turner’s Syndrome

D. Bicuspid Aortopathy / Bicuspid Aortic Valve Syndrome

E. Haven’t seen a penlight in years nor a uvula
Bicuspid Aortopathy

- Worldwide quantitatively the cause of most congenital cardiovascular morbidity and mortality: 15% of patients with dissection have bicuspid valves, more than Marfan’s

- Can be inherited as autosomal dominant—15% family members can have aneurysms even if that family member has a normal aortic valve
AHA/ACC guidelines suggest echo screening of 1st degree relatives of those with Bicuspid Aortic valve
Patient Education

• We recommend those with aortic aneurysms, even post-surgery, remain physically active, but with some restrictions.
  – Avoid any isometric exercises that might cause one to grunt (Valsalva) including weightlifting in daily living: lifting furniture, heavy suitcases, sit-ups, push-ups, pull-ups, bags of mulch, etc.
• For some this may include weight as low as 25 pounds.
Valsalva maneuver with aortic aneurysms: straining from constipation, weightlifting, chopping wood, shoveling snow: SBP can reach 300mmHg

Class IIa

1. For patients with a current thoracic aortic aneurysm or dissection, or previously repaired aortic dissection, employment and lifestyle restrictions are reasonable, including the avoidance of strenuous lifting, pushing, or straining that would require a Valsalva maneuver. (Level of Evidence: C)
Patient Education

Cardio exercise such as jogging, swimming, tennis, hiking, and cycling is recommended, up to the point of being able to maintain a conversation during exercise.
Maintain low to normal blood pressure.

Avoid blood pressure spikes:

• Decongestants, both over the counter (OTC) and prescription, containing pseudoephedrine, phenylephrine, phenylpropanolamine, and oxymetazoline. This includes Claritin D and Robitussin D

• Treat HTN with Toprol XL every 12, Bystolic, Losartan, Valsartan, Lisinopril; avoid amlodipine
USB drives

• Maintain all operative reports plus the actual CT and MRI Scan images, especially when traveling.
• These help providers in emergency situations rapidly understand the repair, prior areas of hematoma, graft locations, comparing with new scans.
Conclusions

Recognize

– the increasing incidence of Aortic aneurysms and probably AAS as our population ages

– asymptomatic disease in high risk groups requires imaging to recognize dilation before catastrophic events occur

– All societies (STS, ACC, SVM, ATS) recommend careful measuring of the aorta perpendicular to flow (true short axis measurement)

– Careful imaging and measuring is time-consuming but accurate size is, beyond symptoms, the best intervention criterion in the 2018
Conclusions

Recognize

– Can not rely on CXR for aortic disease exclusion – neither sensitive or specific

– ‘Tortuosity’ in younger patients deserves investigation
Remember

– Because of frequency, **Bicuspid Aortic valve** is the cause of most congenital cardiovascular morbidity and mortality

– Care of patients with aortic diseases falls into a grey zone between our current specialties, emphasizing its importance.
Mohit Bhasin MD

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