Diseases of the Aorta

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Governor-Elect/President-Elect, Michigan Chapter, ACC

No relevant disclosures
Examination Content Outline

I. Physical Principles, Instrumentation, Examination Principles
   A. Routine Doppler Examination
   B. Transesophageal Echocardiography, Intraoperative Echocardiography, and Catheter-Based Echocardiography (ICE)
   C. Physical Principles of Ultrasound
   D. Cross-Sectional Echocardiographic Examination
   E. Principles of Doppler Flow Measurement
   F. Cross-Sectional Scanning: Technical Principles and Instrumentation
   G. Standard Plane Positions—Standard Imaging Planes
   H. Doppler Instrumentation
   I. Principles of Flow
   J. Principles of Color Flow Mapping
   K. M-Mode Echocardiography
   L. Digital Image Processing
   M. Doppler Signal Processing, Tissue Characterization
   N. Three-Dimensional Echocardiography
   O. Place (Role) of Echocardiography
   P. Hand-Held Echo
   Q. Laboratory Accreditation

II. Vascular Heart Disease
    A. Aortic Valve, Aorta, and Subaortic Outflow Tract
    B. Mitral Valve
    C. Echo-Doppler Assessment of Prosthetic Heart Valves
    D. Echocardiographic Findings in Infective Endocarditis
    E. Fluid Dynamics of Regurgitant Jets
    F. Tricuspid Valve
    G. Pulmonic Valve
    H. Pulmonary Hypertension

III. Chamber Size and Function
    A. Coronary Artery Disease, Stress Echocardiography
    B. General Considerations, Assessment of Chamber Size and Function
    C. Echocardiographic Assessment of the Cardiomyopathies
    D. Diastolic Function
    E. Left Atrium, Pulmonary Veins, and Coronary Sinus
    F. Right Ventricle
    G. Right Atrium
    H. Interatrial and Interventricular Septum
    I. Inferior and Superior Vena Cava
    J. Doppler Estimation of Volumetric Flow
    K. Coronary Arteries

IV. Congenital Heart Disease
    A. Complex Congenital Heart Disease
    B. Aortic Valve, Heart and Subaortic Outflow Tract
    C. Tricuspid Valve Anomalies
    D. Mitral Valve
    E. Doppler Estimation of Volumetric Flow
    F. Pulmonic Valve Anomalies
    G. Coronary Arteries Anomalies
    H. Fetal Echocardiography
    I. Terminology and Anatomic and Physiologic Basis of CHD
    J. Principles of Medical and Surgical Management
    K. Echo Evaluation of Post-Op Congenital Heart Disease

V. Cardiac Masses, Pericardial Disease, Contrast and New Applications
    A. Pericardial Disease
    B. Cardiac Tumors and Masses
    C. Contrast Echocardiography
    D. Assessment of Myocardial Perfusion with Contrast
    E. Echocardiography in Disorders of Cardiac Rhythm and Conduction
    F. Echocardiography in Cardiac Transplantation

VI. Miscellaneous Topics (Role of Echo)
    A. Heart Failure
    B. Cardiac Sources of Embolism (PFO, ASA, SEC, Aortic Atheroma, etc)
    C. Pulmonary Heart Disease
    D. Systemic Diseases
    E. Atrial Fibrillation
    F. Tricuspid Valve
    G. Athlete's Heart
    H. Right Pathologies
    I. Pregnancy
    J. Interventional Echocardiography
    K. Digital Lab
    L. Quality in the Echo Lab

Reference Statement
JBE does not endorse or recommend any third-party review course or material. Any text in cardiovascular techniques and evaluation, cardiac patient care and management may be used. Current standards and guidelines endorsed by professional societies are also appropriate.
Suggested reading

Echocardiography in aortic diseases: EAE recommendations for clinical practice

Arturo Evangelista, Frank A. Flachskampf, Raimund Erbel, Francesco Antonini-Canterin, Charalambos Vlachopoulos, Guido Rocchi, Rosa Sicari, Petros Nihoyannopoulos, and Jose Zamorano on behalf of the European Association of Echocardiography

Guidelines and Standards

Multimodality Imaging of Diseases of the Thoracic Aorta in Adults: From the American Society of Echocardiography and the European Association of Cardiovascular Imaging

Endorsed by the Society of Cardiovascular Computed Tomography and Society for Cardiovascular Magnetic Resonance

(J Am Soc Echocardiogr 2015;28:119-82.)
Aortic disease

Atherosclerotic
  Aneurysm
  Atheroembolic disease
  Rupture
  Pseudoaneurysm
  Penetrating ulcer
  Dissection
  Intramural hematoma

Non-atherosclerotic
  Cystic medial necrosis
  Aneurysm
  Aortic dissection
  Intramural hematoma
  Anuloaortic ectasia

Inflammatory/infectious
  Takayasu arteritis
  Giant cell arteritis
  Endocarditis

Miscellaneous
  Trauma
  Intraluminal thrombus
  Poststenotic dilation
  Hypertension
  Aortic insufficiency/Stenosis
  Iatrogenic injury
Objectives

Kick off Ques
Aortic anatomy
Basic Measurements by echo
Brief comparison of modalities
Dissection (acute and chronic)
Aneurysms
False alarms
Bicuspid aortic valve/aortopathy
Coarctation
Marfan
Loeys-Dietz
Atherosclerotic Disease
Kick off Ques (Ans)
The following measurement of the aortic root on 2D TTE follows ASE guidelines

a true
b false
In the provided arch view choose the most appropriate response for this movie clip

1. normal suprasternal view
2. arch dissection
3. arch aneurysm
4. anomalous venous drainage
Question 3

Which of the following conditions associated with aortic aneurysms is called by TGFbeta or SMAD 3 mutation with systemic connective tissue involvement?

a. Bicuspid Aortopathy
b. Loey Dietz syndrome
c. Marfan syndrome
d. Ehlers-Dalos syndrome
Question 4

Which of the following characteristics is true for identifying a false lumen (FL) from true lumen (TL) in aortic dissection?

1. FL is smaller in size than true
2. Systolic compression is usually seen with FL
3. Color flow can be seen from FL to TL
4. Flow is brisk in FL
The aortic annulus represents the junction of the proximal ascending aorta with LVOT.

It is part of the fibrous skeleton of the heart and is contiguous with the anterior mitral valve leaflet and perimembranous septum.

Relatively resistant to dilation.

Annulus dimension is very close to LVOT dimension.
The geometry of the sinotubular junction is a crucial feature of normal aortic valve coaptation.

Insertion of aortic valve cusps is continuous from the level of the annulus up through the sinuses to the level of the sinotubular junction.

Dilation of the sinotubular junction can result in malcoaptation of the aortic cusps resulting in secondary aortic insufficiency.
Normal Aortic Anatomy

Ascending aorta

Aortic arch
22–36 mm

Tubular ascending aorta
22–36 mm
(15 ± 2 mm/m²)

Sinotubular junction
22–36 mm
(15 ± 1 mm/m²)

Sinuses of Valsalva
29–45 mm
(19 ± 1 mm/m²)

Aortic annulus
20–31 mm
(13 ± 1 mm/m²)

Descending aorta
20–30 mm

Diaphragm

Left common carotid artery
Left subclavian artery
Ligamentum arteriosum

Modified from
Normal Aortic Root Measurements

Male

<table>
<thead>
<tr>
<th>Age (y)</th>
<th>15–29</th>
<th>30–39</th>
<th>40–49</th>
<th>50–59</th>
<th>60–69</th>
<th>≥70</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean normal (cm)</td>
<td>3.3</td>
<td>3.4</td>
<td>3.5</td>
<td>3.6</td>
<td>3.7</td>
<td>3.8</td>
</tr>
<tr>
<td>Upper limit of normal (cm) (95% CI)</td>
<td>3.7</td>
<td>3.8</td>
<td>3.9</td>
<td>4.0</td>
<td>4.1</td>
<td>4.2</td>
</tr>
</tbody>
</table>

Add 0.5 mm per 0.1 m² BSA above 2.0 m² or subtract 0.5 mm per 0.1 m² BSA below 2.0 m².⁶

Female

<table>
<thead>
<tr>
<th>Age (y)</th>
<th>15–29</th>
<th>30–39</th>
<th>40–49</th>
<th>50–59</th>
<th>60–69</th>
<th>≥70</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean normal (cm)</td>
<td>2.9</td>
<td>3.0</td>
<td>3.2</td>
<td>3.2</td>
<td>3.3</td>
<td>3.4</td>
</tr>
<tr>
<td>Upper limit of normal (cm)</td>
<td>3.3</td>
<td>3.4</td>
<td>3.6</td>
<td>3.6</td>
<td>3.7</td>
<td>3.9</td>
</tr>
</tbody>
</table>

Add 0.5 mm per 0.1 m² BSA above 1.7 m² or subtract 0.5 mm per 0.1 m² BSA below 1.7 m².⁶

age : 1-15 16-39 ≥40
Aorta size is related most strongly to body surface area (BSA) and age.

Indexing by height avoids the influence of overweight on BSA (limitation of large data).

Avoid overestimation with oblique segments, perform measurement only when circular sections are obtained.

Measurements of descending thoracic aorta in short axis and of the aortic arch in long axis are recommended.
# Typical Echo Views for Aorta Interrogation

<table>
<thead>
<tr>
<th>View</th>
<th>Part of aorta</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Transthoracic echo</strong></td>
<td></td>
</tr>
<tr>
<td>Parasternal long + short axis</td>
<td>Ascending + descending thoracic</td>
</tr>
<tr>
<td>Apical four-chamber</td>
<td>Descending thoracic</td>
</tr>
<tr>
<td>Apical two-chamber and/or long axis</td>
<td>Descending thoracic</td>
</tr>
<tr>
<td>Suprasternal</td>
<td>Arch, descending + ascending thoracic</td>
</tr>
<tr>
<td>Subcostal</td>
<td>Abdominal (+ascending thoracic)</td>
</tr>
<tr>
<td><strong>Transoesophageal echo</strong></td>
<td></td>
</tr>
<tr>
<td>Upper oesophageal long + short axis</td>
<td>Ascending thoracic</td>
</tr>
<tr>
<td>Aortic (long + short axis)</td>
<td>Descending thoracic + arch</td>
</tr>
</tbody>
</table>
Although TTE is not the ideal tool for visualizing all aortic segments, important information can always be gained by careful use of all echo windows.

In all patients with suspected aortic disease, the right parasternal view is recommended for estimating the true size of the ascending aorta.

Underutilized: Right parasternal
The aorta can be visualized from the annulus through the ascending and arch portions and the descending thoracic aorta to the level of the gastroesophageal junction.

Has several advantages:

1- Proximity of the oesophagus and the thoracic aorta permits high resolution images.
2- Availability of multiplane imaging permits improved incremental assessment of the aorta from its root to the descending aorta.
A short segment of the distal ascending aorta, just before the innominate artery, remains unvisualized owing to interposition of the right bronchus and trachea (blind spot)!!

TEE views of innominate and left carotid are usually suboptimal
Aortic dissection

- Clinical presentation can be identical to other conditions in (Acute Aortic Syndrome).

- Typically in the setting of aortic dilatation.

- 40% in aortas smaller than 5 cm!

- Usually there is more than one communication point between true and false lumen.

- Classically begins at the ligamentum arteriosum or Ascending Ao.
<table>
<thead>
<tr>
<th>Percentage</th>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>60%</td>
<td>DeBakey I</td>
<td>Stanford A (Proximal)</td>
</tr>
<tr>
<td>10–15%</td>
<td>DeBakey II</td>
<td></td>
</tr>
<tr>
<td>25–30%</td>
<td>DeBakey III</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Stanford B (Distal)</td>
</tr>
</tbody>
</table>
**Echo Assessment in Aortic Dissection**

<table>
<thead>
<tr>
<th>Task</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Identify presence of a dissection flap</td>
<td>Flap dividing two lumens</td>
</tr>
<tr>
<td>Define extension of aortic dissection</td>
<td>Extension of the flap and true/false lumens in the aortic root (ascending/arch/descending abdominal aorta)</td>
</tr>
<tr>
<td>Identify true lumen</td>
<td>Systolic expansion, diastolic collapse, systolic jet directed away from the lumen, absence of spontaneous contrast, forward systolic flow</td>
</tr>
<tr>
<td>Identify false lumen</td>
<td>Diastolic diameter increase, spontaneous contrast and or thrombus formation, reverse/delayed or absent flow</td>
</tr>
<tr>
<td>Identify presence of false luminal thrombosis</td>
<td>Mass separated from the intimal flap and aortic wall inside the false lumen</td>
</tr>
<tr>
<td>Localize entry tear</td>
<td>Disruption of the flap continuity with fluttering or ruptured intimal borders; color Doppler shows flow through the tear</td>
</tr>
<tr>
<td>Assess presence, severity and mechanisms of AR</td>
<td>Anatomic definition of the valve (bicuspid, degenerated, normal with/without prolapse of one cusp); dilation of different segments of the aorta; flap invagination into the valve; severity by classic echocardiographic criteria</td>
</tr>
<tr>
<td>Assess coronary artery involvement</td>
<td>Flap invaginated into the coronary ostium; flap obstructing the ostium; absence of coronary flow; new regional wall motion abnormalities</td>
</tr>
<tr>
<td>Assess side-branch involvement</td>
<td>Flap invaginated into the aortic branches</td>
</tr>
<tr>
<td>Detect pericardial and/or pleural effusion</td>
<td>Echo-free space in the pericardium/pleura</td>
</tr>
<tr>
<td>Detect signs of cardiac tamponade</td>
<td>Classic echocardiographic and Doppler signs of tamponade</td>
</tr>
</tbody>
</table>
## True vs False Lumen: Key findings

<table>
<thead>
<tr>
<th></th>
<th>True lumen</th>
<th>False lumen</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Size</strong></td>
<td>True &lt; false</td>
<td>Most often: false &gt; true lumen</td>
</tr>
<tr>
<td><strong>Pulsation</strong></td>
<td>Systolic expansion</td>
<td>Systolic compression</td>
</tr>
<tr>
<td><strong>Flow direction</strong></td>
<td>Systolic antegrade flow</td>
<td>Systolic antegrade flow reduced or absent, or retrograde flow</td>
</tr>
<tr>
<td><strong>Communication flow</strong></td>
<td>From true to false lumen in systole</td>
<td></td>
</tr>
<tr>
<td><strong>Contrast echo flow</strong></td>
<td>Early and fast</td>
<td>Delayed and slow</td>
</tr>
</tbody>
</table>
TTE views of massive aortic dilation and dissection
Aortic Dissection Complications

- Rupture
- Tamponade
- Aortic regurgitation
- Coronary artery involvement
- Other branch vessel involvement
Mechanisms of Aortic Regurgitation in Dissection
Echo Insights

(A) effacement of dilation of the sinotubular junction resulting in malcoaptation of the aortic valve
(B) intrinsic aortic valve disease
(C) disruption of the insertion of an aortic cusp
(D) prolapse of a portion of the intimal dissection flap through the aortic valve, which serves as a conduit for aortic regurgitation (D).
Coronary Artery Involvement on TEE with AD
Comparison of Imaging Modalities for Imaging Aortic Dissection

n=1139, MGH database

<table>
<thead>
<tr>
<th>Imaging Modality</th>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Helical CT</td>
<td>100%</td>
<td>98%</td>
</tr>
<tr>
<td>TEE</td>
<td>98%</td>
<td>95%</td>
</tr>
<tr>
<td>MRI</td>
<td>98%</td>
<td>98%</td>
</tr>
<tr>
<td>Aortogram</td>
<td>88%</td>
<td>94%</td>
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</table>

IRAD registry

<table>
<thead>
<tr>
<th>Imaging Modality</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>CT</td>
<td>93%</td>
</tr>
<tr>
<td>TEE</td>
<td>88%</td>
</tr>
<tr>
<td>MRI</td>
<td>100%</td>
</tr>
<tr>
<td>Aortogram</td>
<td>87%</td>
</tr>
</tbody>
</table>

real world
### Comparative Strengths of Modalities for Acute Aortic Syndrome

<table>
<thead>
<tr>
<th>Advantages of modality</th>
<th>CTA</th>
<th>TTE</th>
<th>TEE</th>
<th>MRA</th>
<th>Angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>Readily available</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Quickly performed</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Performed at bedside</td>
<td>−</td>
<td>+++</td>
<td>+++</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Noninvasive</td>
<td>+++</td>
<td>+++</td>
<td>+</td>
<td>+++</td>
<td>−</td>
</tr>
<tr>
<td>No iodinated contrast</td>
<td>−</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>−</td>
</tr>
<tr>
<td>No ionizing radiation</td>
<td>−</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>−</td>
</tr>
<tr>
<td>Cost</td>
<td>++</td>
<td>+</td>
<td>++</td>
<td>++</td>
<td>+++</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Diagnostic performance</th>
<th>CTA</th>
<th>TTE</th>
<th>TEE</th>
<th>MRA</th>
<th>Angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>+++</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
</tr>
<tr>
<td>Specificity</td>
<td>+++</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
</tr>
<tr>
<td>Ability to detect IMH</td>
<td>+++</td>
<td>+</td>
<td>++</td>
<td>+++</td>
<td>−</td>
</tr>
<tr>
<td>Site of intimal tear</td>
<td>+++</td>
<td>−</td>
<td>++</td>
<td>+++</td>
<td>++</td>
</tr>
<tr>
<td>Presence of AR</td>
<td>−</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>+++</td>
</tr>
<tr>
<td>Coronary artery involvement</td>
<td>+</td>
<td>−</td>
<td>++</td>
<td>+</td>
<td>+++</td>
</tr>
<tr>
<td>Presence of pericardial effusion</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>−</td>
</tr>
<tr>
<td>Branch vessel involvement</td>
<td>++</td>
<td>−</td>
<td>+</td>
<td>++</td>
<td>+++</td>
</tr>
</tbody>
</table>

Ciggarora et al
Isselbacher et al
All that Glitters in Not A Flap on Echo

- Not every line in the Aorta is dissection!
- Reverberation artifacts are very common in the Aorta (40%).
- Usually from the posterior wall of the ascending aorta or the posterior wall of the right pulmonary artery.
- Clues to differentiate:
  1. Color Doppler
  2. Moving in parallel with the reverberating structures
  3. Double the distance from the probe
Pseudo Dissection Flaps
Intra Mural Hematoma (IMH)

- Variant of Aortic dissection, usually result from rupture of vasa vasorum, resulting in hematoma in the medial space BUT no communication with the Lumen.
- 16% will progress to AD.
- Clinical presentation, prognosis and treatment is the same as AD.
Intra Mural Hematoma (IMH)

Localized thickening of aortic wall
- Usually crescentic
- Occasionally circumferential
- Echo-lucent spaces common

Relatively smooth luminal surface

Absence of dissection flap

Maintenance of circular lumen
Marfan Syndrome

- It usually begins with dilatation of the aortic sinuses, which progresses into the sinotubular junction and ultimately into the aortic annulus.
- Compared with atherosclerotic aortic aneurysms, aortic aneurysms in Marfan syndrome commonly occur in younger patients and enlarge more rapidly.
- Main mechanism for AI is dilatation of the ST junction.
Marfan Syndrome Aorta Surveillance

**NATIVE PROXIMAL AORTA**

- **Baseline TTE**
  - Aortic diameter (aortic root, ascending aorta)
  - Repeat TTE at 6mo
  - Rate of growth

  - Max diameter <45mm
    - Stable
    - Annual imaging (TTE)*

  - Max diameter ≥45mm
    - Significant growth
    - Imaging at 6-12mo intervals (TTE)*

- **Baseline MRI (or CT)**
  - Aortic diameter (entire aorta)

  - Normal aortic diameter beyond root
    - 5-yearly MRI (or CT)
    - Annual MRI (or CT)

  - Aneurysmal aorta beyond root

**AFTER PROXIMAL AORTIC REPLACEMENT**

- **Baseline MRI (or CT)**
  - Aortic diameter distal to graft

  - No previous dissection
    - Stable diameter
    - 2-yearly MRI (or CT)
  
  - Previous dissection
    - Significant growth
    - MRI (or CT) at 6-12mo intervals
27 year old male with Loeys Dietz Syndrome, SMADD3 mutation and severe MVPS MR s/p MV replacement for followup for MR
MRI surveillance
<table>
<thead>
<tr>
<th>Imaging recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Annual CTA or MRA performed from head to at least the pelvis [19, 32]</td>
</tr>
<tr>
<td>Run-off studies may be considered to evaluate for iliac aneurysms [30, 32]</td>
</tr>
<tr>
<td>3- to 6-month serial echocardiograms to evaluate valvular disease and aortic root dilation if initial measurements are abnormal; echo at 6 months and then annually if initial measurements are normal with minimal progression [5, 32]</td>
</tr>
<tr>
<td>Serial echocardiograms every 3 months for 1 year after aortic root grafting, then every 6–12 months thereafter if warranted based on severity of phenotype [32]</td>
</tr>
<tr>
<td>Cervical spine plain films with flexion and extension views to assess for instability, particularly prior to general anesthesia and/or surgery [3, 32]</td>
</tr>
<tr>
<td>Thoracolumbar spinal imaging to assess for scoliosis and spondylolisthesis [3]</td>
</tr>
<tr>
<td>DEXA scan to assess for osteopenia</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Surgical recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Children</em>: if clinical phenotype is severe, surgery once ascending aorta exceeds 99th percentile and annulus is at least 1.8 cm, or rapid expansion (&gt;0.5 cm/year); timing depends on presence of TGFBR mutation, family history and ability to place graft of sufficient size to permit growth, among other factors [5, 32]</td>
</tr>
<tr>
<td><em>Adolescents and adults</em>: 4.0-cm threshold for aortic root grafting, or rapid expansion (&gt;0.5 cm/year) [17, 19]; some data to suggest 5.0-cm threshold in adult patients with less severe clinical phenotype [57]</td>
</tr>
<tr>
<td><em>Pregnancy</em>: Elective early cesarean section may be recommended, with referral to high-risk OB/GYN clinic and prenatal diagnosis if disease-causing mutation is known [2]</td>
</tr>
</tbody>
</table>
Bicuspid Aortic Valve and Aortopathy

BAV is most common congenital disorder 0.5-1.5% of population

sporadic AD transmission : M: F = 3: 1
Types of BAV fusion

A

B

R

R

TV

RVOT

PA

LA

P

P

N

L

75%

20-25%

<3%
Aorta issues in BAV

- At autopsy up to 35% reported to have aortic involvement
- Can occur at any level mainly unto ascending aorta
- AS does not need to be present for aorta enlargement
- Intrinsic wall abnormalities (elastin fragmentation, fibrillin issues, cellular apoptosis, MMP overexpression, location of the outflow jet hitting aortic wall all contribute)
BAV - Aorta surveillance

Diagnostic/Baseline TTE*
Valve morphology/function
Ascending aortic diameter

Aortic diameter
<40mm
40-45mm
≥45mm

Rate of growth
<3mm/y ↔ ≥3mm/y

Family history of dissection
NO ↔ YES

5-yearly TTE*
2-yearly TTE*
Annual TTE*
Baseline MRI + Annual TTE*
BAV/Aortopathy

- Shones complex
- Williams syndrome
- Turner syndrome

9% prevalence of BAV in 1st degree relatives

ACC guidelines recommend echo screening in all first degree relatives

1/3 of first degree relatives of BAV patients have aorta dilation despite trileaflet valves
40 year old male with HTN and with family history of bicuspid aortic valve and heart murmur

50-70% of Coarctation cases have associated BAV
< 10% of cases of BAV have associated Coarctation
Remember Shone complex association /Turner syndrome
Mild Coarctation
Coarctation of the Aorta

- Usually just distal to the Left SC artery (Preductal or post ductal).
- Best seen on suprasternal windows.
- Doppler:
  - Peak gradient can be obtained,
  - Look for persistent diastolic flow.
- Account for Proximal velocity if >1.5 m/s.
- Peak gradient calculation (remember prox velocity) as part of Bernoulli equation.
Severe Coarctation
**Sinus of Valsalva ANEURYSM**

- most often arise from the right sinus.
- highly variable in size and by definition communicate with the sinus by a relatively wide mouth.
- typically protrude down into the right atrium
- rupture typically causes acute decompensation
- continuous murmur
Aortic Atheroma

- DA>Arch>Aao.
- The morphology of atheromatous plaques is dynamic, with frequent formation and resolution of mobile components.
- Highly prevalent (51% of the population over 45 years)
- Simple vs Complex
Aortic Atheromas

Simple Atheromas
- Protruding < 4 mm
- No mobile debris
- No surface ulceration

Complex Atheroma
- Protruding > 4 mm
- Mobile debris
- > 2mm ulceration

Grading of aortic atheroma

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Percentage of incidence of stroke (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal aorta</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>Extensive intimal thickening &lt; 3 mm</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>Protrudes &lt; 5 mm into aortic lumen</td>
<td>5</td>
</tr>
<tr>
<td>4</td>
<td>Protrudes &gt; 5 mm into aortic lumen</td>
<td>10.5</td>
</tr>
<tr>
<td>5</td>
<td>Mobile atheroma</td>
<td>46.5</td>
</tr>
</tbody>
</table>

Katz et al JACC 1992
Complex Atheroma
Pregnancy In Aortopathies

- **PREGNANCY (WISH)**
- **Counselling**
  - Women with aortic dilatation and/or genetic aortopathy contemplating pregnancy should be counselled about the risk for aortic dissection and the hereditary nature of the disease. Pregnancy should be discouraged if aortic diameter has reached the threshold for prophylactic surgery (table 3). Prepregnancy aortic surgery should be considered

- **Pharmacological treatment**
  - Strict blood-pressure control is indicated
  - Start β-blocker therapy in women with genetic aortopathy syndrome, aortic dilatation and/or (history of) type B aortic dissection
  - Angiotensin receptor blockers/ACE inhibitors are contraindicated: switch to β-blockers as soon as contraceptives are stopped

- **Imaging**
- **Prepregnancy**
  - MRI (or CT) imaging of the entire aorta advisable

- **During pregnancy**
  - Aortic root/ascending aortic dilatation and/or genetic aortopathy syndrome: transthoracic echocardiography per 4–8 weeks
  - Dilatation beyond distal ascending aorta: MRI (without gadolinium)
Pregnancy and Surgery Recs In Aortopathies

**During pregnancy**
- Indications: type A dissection; type B dissection, if complicated by malperfusion or aortic rupture (otherwise, medical treatment with MRI monitoring); prophylactic, aortic diameter =50 mm with rapid growth
- First/second trimester (=28 weeks), fetus not viable: aortic repair with intensive fetal monitoring
- Third trimester (=29 weeks), fetus viable: urgent caesarean section directly followed by aortic surgery

**DELIVERY**

**At centre where cardiothoracic surgery is available**

**Mode of delivery**
- Aortic root/ascending aortic diameter <40 mm: vaginal delivery favoured
- Aortic root/ascending aortic diameter 40–45 mm: consider vaginal delivery with epidural anaesthesia and expedited second stage (eg, forceps, vacuum delivery) or caesarean delivery
- Aortic root/ascending aortic diameter >45 mm: consider caesarean delivery

**Timing**
- Consider elective caesarean delivery in Loeys-Dietz syndrome, vEDS or non-syndromic familial thoracic aortic aneurysms and dissections and aortic dilatation
### Recommendations for Surgical Repair of Thoracic Aortic Aneurysms

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Size Threshold for Repair</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic aneurysm (regardless of etiology)</td>
<td>Any size causing symptoms (dissection, pain, compression)</td>
</tr>
<tr>
<td>Degenerative</td>
<td>5.5 cm</td>
</tr>
<tr>
<td>BAV</td>
<td>5-5.5 cm; aortic area/height &gt;10 cm²/m</td>
</tr>
<tr>
<td>Marfan</td>
<td>4-5 cm; aortic area/height &gt;10 cm²/m</td>
</tr>
<tr>
<td>Loeys-Dietz</td>
<td>4.2-4.6 cm</td>
</tr>
<tr>
<td>Turner</td>
<td>2.5 cm/m²</td>
</tr>
<tr>
<td>Requiring other cardiac surgery</td>
<td>4.5 cm</td>
</tr>
</tbody>
</table>
Question 1

The following measurement of the aortic root on 2D TTE follows ASE guidelines

a true
b false

Measurement tips
should be made perpendicular to long axis of aorta
largest diameter from right coronary sinus to non coronary sinus
leading edge to leading edge ; preferable in diastole
Contrast that with LVOT measurement : mid systole / inner edge to inner edge
Question 2

Correct answer: normal suprasternal view

brachiocephalic vein

rt PA

normal suprasternal view
arch dissection
arch aneurysm
anomalous venous drainage
Question 3

Which of the following conditions associated with aortic aneurysms is caused by TGF-Beta or SMADD 3 mutation with systemic connective tissue involvement?

a. Bicuspid Aortopathy
b. Loey Dietz syndrome
c. Marfan syndrome
d. Ehlers-Dalos syndrome

**Surveillance**

- annual or biannual echocardiograms
- annual computed tomography angiography (CTA) or magnetic resonance angiography (MRA)
- brain to pelvis
- cervical spine X-rays
Question 4

Which of the following characteristics is true for identifying a false lumen (FL) from true lumen (TL) in aortic dissection?

1. FL is smaller in size than true
2. Systolic compression is usually seen with FL
3. Color flow can be seen from FL to TL
4. Flow is brisk in FL
Thank you