Acute Aortic Syndromes

Michael H. Picard, M.D.
Massachusetts General Hospital
Harvard Medical School

No disclosures

For everything you need to know about the aorta see

Circulation 2010;121:e266-e369
Normal range for size of aorta

Recommendations for aortic imaging techniques to determine the presence and progression of thoracic aortic disease

- Measurements of aortic diameter should be taken at reproducible anatomic landmarks, perpendicular to the axis of flow
- For CT and MR measurements, the external diameter should be measured. For aortic root measurements, the widest diameter, typically at the mid sinus level should be used.
- For echo, the internal diameter should be measured. For aortic root measurements, the widest diameter, typically at the mid sinus level should be used.

Acute aortic syndromes

- Aortic dissection
- Intramural hematoma
  - Variant of classic aortic dissection
  - Typically no intimal tear
- Penetrating atherosclerotic ulcer
- Rapidly expanding aortic aneurysm
- Rupture or contained rupture of an aortic aneurysm
- Traumatic aortic transection
Acute aortic dissection

- Incidence = 3/100,000 per year
- Life-threatening condition
  - Early mortality 1% per hour
  - Survival improved with prompt therapy
- Presentation may be non-specific and risk factors may not be recognized
  - Presentation can range from chest pain (?MI, PE, pericarditis) to syncope to paralysis to stroke

Chest Pain of aortic dissection

- Acute chest and/or infrascapular back pain
- Pain is abrupt in onset
- Pain is severe, worst ever
- Worst at onset not crescendo
- Can be described as tearing or stabbing in quality
- Pain may migrate
- Patients are restless, cannot get comfortable

Classic features of aortic dissection

- Hypertension
- BP differential between the 2 arms
- Murmure of AI
- Wide mediastinum on CXR
- Older patients have history of hypertension
- Younger patients have evidence of Marfan’s syndrome
Pathophysiology of Aortic Dissection: Intimal Tear + Disease of the Aortic Media

- Smooth muscle cell drop-out
- Elastic fiber degeneration
- Presence in the media of cystic spaces filled with mucoid material
- Weakening of the aortic wall

Cystic medial necrosis

Pathophysiology of Aortic Dissection: Intimal Tear + Disease of the Aortic Media

Classification of Aortic Dissection: Based on Involvement of the Ascending Aorta

DeBakey

Stanford

<table>
<thead>
<tr>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td>A (62%)</td>
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<td>B (38%)</td>
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Etiology of Cystic Medial Degeneration:
Common Underlying Abnormality of the Aortic Wall

- Acquired
  - Occurs to some extent with aging
  - Accelerated by longstanding hypertension
  - Great vessel vasculitis
- Congenital conditions causing a structural/functional defect of connective tissue
  - Connective tissue disorders
    - Marfan’s syndrome
    - Ehlers-Danlos syndrome
    - Loey-Dietz syndrome (TGF receptor mutations)
  - Congenital bicuspid aortic valve
  - Familial thoracic aortic aneurysm syndrome

Causes of Acquired Cystic Medial Degeneration:
Chronic Hypertension

- Most common cause of dissection in the older population
- Common misconception that arterial hypertension must be severe
  - Typical hypertension is most common
- Late consequences include intimal thickening, degradation of the extracellular matrix, loss of elastic fibers, smooth muscle cell necrosis
- Aortic wall becomes weak and stiff
- Aorta dilates or becomes aneurysmal

Causes of Congenital Cystic Medial Degeneration:
Marfan’s Syndrome

- Autosomal dominant, heritable disorder
- Due to mutations in one of the genes for fibrillin-1
  - A structural protein that is the major component of microfibrils of elastin
- Mutations result in a decrease in the amount of elastin in the aortic wall and a loss of elastin's normally highly organized structure
- The aorta exhibits markedly abnormal elastic properties
  - Over time the aorta becomes stiff and dilated
- ACE inhibitors and ARBs for treatment
Causes of Congenital Cystic Medial Degeneration: Bicuspid Aortic Valve

- Those with bicuspid aortic valve (BAV) have a significantly increased risk of aortic dilatation, aneurysm, and dissection
- Echo studies of young people with normally functioning BAV have shown that ~50% have dilatation of ascending aorta
- Cystic medial degeneration appears to be the culprit
  - Of those with BAV undergoing AVR, 75% had biopsy proven cystic medial degen., vs. 14% among tricuspid aortic valves
- No single gene responsible for BAV has been identified, and it is likely genetically heterogenous


Causes of Congenital Cystic Medial Degeneration: Familial Thoracic Aortic Aneurysm Syndrome

- Cystic medial degeneration can be seen in patients with ascending aneurysms or dissection who do not have overt connective tissue disorders, BAV, or other risk factors
- Such cases may be sporadic, but they are often familial in nature are now referred to as the familial thoracic aortic aneurysm syndrome
  - At least 19% of those with thoracic aortic aneurysms in Yale database had a family history of a thoracic aneurysm
  - Screening 1st degree relatives should be advised
- Several mutations have been identified
  - Dominant inheritance
  - Marked variability in the expression and penetrance


Identified Risk Factors in the International Registry of Aortic Dissection

- Advanced age 63 yr (mean)
- History of hypertension 72%
- Marfan’s syndrome 5%
- Bicuspid aortic valve 5%
- Peripartum period of pregnancy 1%
- Cocaine use 1%
- Cardiac catheterization 2%
- Prior cardiac surgery 18%

Non invasive imaging for diagnosis
all have high sensitivity and specificity but each have other advantages and disadvantages

• TEE
  – Bedside, rapid (if personnel available)
  – Can look at other cardiac structures

• Contrast enhanced multi-detector CT
  – Rapid scan, can also look at pulm embolism, coronary anatomy
  – Large dose of contrast, lots of radiation
  – Artifacts more likely with older devices

• cMR
  – High resolution images of various aortic pathologies
  – Difficult in an emergency situation or unstable patient
  – Concerns about gadolinium but can image without contrast

Local expertise should dictate

Echocardiographic findings

• Intimal flap
  – High frequency, low amplitude motion
  – flow respects the boundary
  – Artifacts
    • Motion: Low frequency (or same frequency as adjacent structure) and high amplitude
    • Color flow goes through

• +/- aortic dilation

• Aortic insufficiency

• Wall motion abnormality if flap obstructs coronary ostia

Echo for Aortic Dissection

TEE

• Sensitivity 98-100% and specificity 77-100% in different series

• False negative TEE is rare: few dissections are limited to the “blind spot” at the inferior portion of the arch

• False positive TEE: Artifacts are very common (23-55%), especially in the ascending aorta.
Type I aortic dissection

10 days post-partum after uneventful delivery of 2\textsuperscript{nd} child
- Sudden onset 10/10 back pain radiating to the chest while breast-feeding

PMH
- Migraines
- Allergic rhinitis

Family history
- Father with Marfan’s, s/p AVR at age 38
- Brother with Marfan’s, s/p AVR at age 28

TEE – dissection or artifact?
• CT: Type B dissection extending from just distal to L subclavian artery to aortic bifurcation
• Hospital course: Medically managed on beta-blocker and discharged home.

Complications of Acute Aortic Dissection: Aortic Rupture

• Type A
  – The ascending aorta is prone to rupture into the pericardium causing cardiac tamponade
  – 2/3 of all deaths from AoD
  – Surgery is indicated to prevent rupture
• Type B
  – Into the pleural space or mediastinum
  – Rupture is uncommon
  – Preventative surgery is not necessary

Specific Complications of Acute Aortic Dissection

- Rupture into pericardium
- Acute MI 2%
- Aortic insufficiency 32%
- Mesenteric ischemia 5%
- Lower extremity ischemia 3%
- Stroke 5%
- BP differential 15%
- Paraplegia <1%
- Renal ischemia 5-8%
Complications of Acute Aortic Dissection: Branch Artery Compromise

Dissection extends into branch artery
Distension of the false lumen compresses true lumen

Mechanisms of AI in acute type A aortic dissection

- leaflet thickening *
- bicuspid aortic valve *
- incomplete leaflet closure *
- aortic leaflet prolapse *
- intimal flap prolapse
- multiples of above

* not specific to dissection

Movsowitz et al, JACC 2000;36:884-90

Artifacts

- Side-by-side:
  - lateral resolution, side lobe, lens effect
- Behind, parallel motion:
  - Reverberation
- Behind, opposite motion:
  - Mirror image
Linear TEE artifacts artifacts in the aorta seen in the presence of dilated aortas

Artifact distance = 2 times that of interface

Transverse TEE image of dilated ascending aorta compressing LA. Note the artifact crosses borders

Appelbe et al, JACC 1993;21:754-60

Clues to artifacts

- Cross borders
- Motion identical to another real structure
  - Amplitude and frequency at a multiple of the real structure
- Indistinct edges
- Not reproduced in an orthogonal or other view
- Color flow passes through it
- Clues - foreign materials present
  - Catheters, prosthetic valves, grafts
  - These are not always in the plane of view

Clues to real structures

- Respects borders
- Distinct edges (unless thrombus)
- Motion
  - Intimal flaps - amplitude and frequency of motion different than cardiac cycle, respiration
- Seen in multiple views
- Color flow respects true borders
- Intimal flaps keep company other pathologic processes
Late Complications of Aortic Dissection

- Progressive expansion of dissected portions of aorta
  - Large aneurysms
  - Aortic rupture
- Development of new aneurysms elsewhere in non-dissected segments of the thoracic aorta
- Aortic insufficiency from aortic root dilatation
- Careful medical follow-up and surveillance imaging of aorta is essential

Acute Intramural Hematoma: Variant of Aortic Dissection

- Risk factors and the presenting signs/symptoms are the same as classic aortic dissection
- Natural history of acute IMH (in the West) appears to be similar to classic aortic dissection
  - Type A is prone to rupture and death → Urgent surgery
    - However, data suggests that Asian populations may have a more favorable outcome from Type A → ? No surgery
  - Type B is at low risk of rupture → Medical management

Intramural Hematoma: Variant of Aortic Dissection

- Unlike classic aortic dissection it is not caused by a visible tear in the intima
- Two possible causes:
  - Rupture of the vasa vasorum within the aortic media, producing a hematoma within the aortic wall
  - A small tear in the intima that permits only transient blood flow from the lumen into the aortic wall, which then thromboses
- No active communication between the aortic lumen and the hematoma and therefore no flow within the aortic wall
- Has a distinctly different and less obvious appearance on aortic imaging studies (especially TEE)
Classic Aortic Dissection vs Intramural Hematoma

Aortic Dissection

Intramural Hematoma

TEE imaging of Intramural Hematoma

65 yo F with CP radiating to back

TEE on admission
MR and CT

TEE in OR 14 hours after admission – note progression of IMH

Surgery: aortic root replacement. IMH extended into RCA so SVG to RCA.

Penetrating Aortic Ulcers (PAU)

- Atherosclerotic lesions with ulceration that penetrate the internal elastic media and allow hematoma in media
- Can lead to IMH, dissection, vessel rupture
- Most common in descending aorta
- Risk factors
  - Elderly, Htn, diffuse atherosclerosis
- On cross-sectional TEE image
  - Ulcer crater with or without thrombus
Penetrating aortic ulcers with superimposed thrombus

Ascending aortic thrombus due to aortic ulcer
37 yo M with stroke, TTE notes aortic clot
Anticoagulated and BP controlled
2 days later develops CP and STE. In OR clot in RCA

Ascending aortic thrombus due to atherosclerotic ulcer
57 yo F presents with RUE arterial insufficiency. Emboli extracted from upper extremity. TEE showed an clot. Recurrence of emboli so taken to surgery
Acute aortic syndromes

- Presentations can mimic other conditions
- High early mortality but prompt recognition improves survival
- Location of dissection determines complications and should dictate therapy
- Hypertension most common etiology
- Dissection and hematoma
  - Risk factors, presenting signs/symptoms, natural history are similar
- Early imaging (TEE, CT, MR) critical for quick diagnosis