CHAPTER 14

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Diseases of the Aorta

CHAPTER OUTLINE

ABBREVIATIONS

AAA	Abdominal aortic aneurysm
ACC/AHA	American College of Cardiology/American Heart Association
ACEI	Angiotensin converting enzyme inhibitor
ACS	Acute coronary syndrome
AI	Aortic insufficiency
AS	Aortic stenosis
AV	Aortic valve
AVR	Aortic valve replacement
BAV	Bicuspid aortic valve
BP	Blood pressure
bpm	Beats per minutes
CABG	Coronary artery bypass graft
cm	Centimeter
CT(A)	Computed tomogram (angiography)
CXR	Chest X-ray
dP/dt	Change in pressure over change in time, which is a measure of left ventricle ejection impulse, an index of shear stress on the aortic wall
DTAA	Descending thoracic aortic aneurysm
EVAR	Endovascular aortic aneurysm repair
GCA	Giant cell arteritis
HF	Heart failure
HR	Heart rate
IMA	Inferior mesenteric artery
IMH	Intramural hematoma of the aorta
IRAD	International Registry of Acute Aortic Dissection
LDL	Low density lipoprotein
LR	Likelihood ratio
M:F	Male to female ratio
mmHg	Millimeters of mercury
MMP	Matrix metalloproteinase
MR	Magnetic resonance

PAD	Peripheral artery disease
PAU	Penetrating atherosclerotic ulcer of the aorta
SBP	Systolic blood pressure
SD	Standard deviation
SMA	Superior mesenteric artery
STS	Society of Thoracic Surgeons
SVC	Superior vena cava
TAA	Thoracic aortic aneurysm
TAAA	Thoracoabdominal aortic aneurysm
TEE	Transesophageal echocardiography
TEVAR	Thoracic endovascular aortic repair
TGF	Transforming growth factor
TIA	Transient ischemic attack
TTE	Transthoracic echocardiography
USPSTF	United States Preventive Services Task Force

AORTIC ANATOMY

- The largest artery in the body; muscular; retroperitoneal
- Histologically contains three layers:
  - Intima (endothelium supported by internal elastic lamina)
  - Media (smooth muscle cells and numerous elastic fibers that give the aorta remarkable tensile strength)
  - Adventitia (collagenous support matrix with external elastic lamina; site of entry of the vasa vasorum externae)
- The segments of the aorta are differentiated by their anatomic location, size, and branch vessels (Table 14-1). Dimensions in males are slightly larger than in females, and the aortic diameters generally increase with age.

TABLE 14-1

AORTA ANATOMY

AORTIC SEGMENTS	DIVISIONS	APPROXIMATE DIAMETERS	MAJOR BRANCH ARTERIES
Ascending			
	Root (sinuses of Valsalva)	≤4.0 cm	Right and left main coronary arteries
	Ascending aorta	≤3.5 cm	None
Arch		≤3.0 cm	Brachiocephalic (innominate), left subclavian, and left common carotid arteries
Descending		≤2.5 cm	Intercostal, spinal, bronchial arteries
Abdominal			
	Suprarenal	≤2.0 cm	Celiac axis, SMA, and renal arteries
	Infrarenal	≤2.0 cm	IMA, common iliac arteries

GENERAL HISTORY AND PHYSICAL EXAMINATION

- **Relevant history**:
  - Due to aorta: Look for chest, back, abdominal, flank pain or discomfort.
  - Due to complications of aortic disease:
    - Depends on the segment of the aorta affected and branch vessels and distal organ territories that are impacted.
    - Look for neurologic symptoms, syncope, heart failure, myocardial infarction, renal failure, thromboembolic disease, compression of adjacent structures such as nerves, esophagus or tracheobronchial tree.
- **Past medical history**: aortic disease, vascular disease, hypertension (HTN), thromboembolic events, trauma.
- **Family history**: aortic disease.
- **Physical examination**:
  - Check bilateral blood pressure (BP) and pulses (radial, carotid, femoral), pulsus paradoxus.
  - Look for evidence of aortic insufficiency (AI), tamponade, heart failure, neurologic deficits, and/or pulsatile abdominal mass.

IMAGING MODALITIES

Chest Radiography (CXR)

- CXR has overall limited sensitivity (~30–60%) for aortic diseases, and alone cannot be used to exclude acute or chronic aortopathy.
- Calcification or tortuosity of the ascending, arch, and descending thoracic aorta may be visualized, but this is a non-specific finding in the elderly.
- Opacification of the aorticopulmonary window, enlargement of the thoracic aorta, increased mediastinal width, displacement of trachea from midline, or obscured/irregular aortic margin may indicate thoracic aortic aneurysm, dissection, or rupture.
- Displaced intimal calcium and pleural effusion may indicate dissection.

Echocardiography and Ultrasonography

- Portable, avoids radiation and contrast media, and can be deployed intra-operatively.
- **Transthoracic Echocardiography (TTE)**
  - TTE cannot provide a comprehensive exam of the aorta, but certain regions can be visualized: aortic valve and root, ascending aorta, arch, descending, and abdominal aorta.
  - TTE is reasonable for assessing aortic valve disorders and monitoring aortic root and ascending aortic dilatation (e.g. especially in Marfan syndrome). It is not sensitive enough to rule out thoracic aortic dissection (sensitivity 70%).
- **Transesophageal Echocardiography (TEE)**
  - TEE can visualize the ascending aorta, transverse arch, and entire descending thoracic aorta. The distal ascending aorta and proximal aortic arch may be obscured by the trachea.

  - TEE, in contrast to other modalities, can provide functional information such as flow dynamics in true and false lumens, detection of AI, detection of cardiac tamponade, and assessment of left ventricular function.

- **Abdominal ultrasonography** is the technique of choice for screening for infrarenal abdominal aortic aneurysm (AAA), but is less accurate as applied to the suprarenal aorta or branch vessels.

Computed Tomography (CT)

- CT is a highly accurate, rapid, reproducible, and readily available technique for detecting and sizing aortic aneurysms and for the diagnostic evaluation of suspected aortic dissection.
- CT is also helpful at mapping branch vessels, and for detecting mimics of aortic disease (e.g. pericardial disease, gastrointestinal disease).

Magnetic Resonance (MR) Imaging

- MR is also a highly accurate technique for aortic imaging. However, the study time is lengthy and the patient is relatively inaccessible, making this modality unsuitable for acute or unstable patients.
- MR is most often performed with intravenous gadolinium as a contrast agent, but the "black-blood" technique with spin-echo sequences can provide satisfactory images without the need for gadolinium.

Aortography

- Catheter-based aortography is an invasive technique that can demonstrate the full extent of aneurysmal disease and dissection, map branch vessel involvement, and demonstrate the presence of AI.
- However, aortography is not readily available in most settings, requires an expert physician operator, and requires that potentially unstable patients undergo a prolonged procedure.

AORTIC ANEURYSMS

Definitions

- Aneurysm = dilatation of the aorta involving all three vessel wall layers.
- Pseudoaneurysm = contained leak of blood in communication with vessel.
- Fusiform = symmetric circumferential bulging of the aorta.
- Saccular = asymmetric localized bulging of the aortic wall.

Abdominal Aortic Aneurysms (AAA)

- **Epidemiology**:
  - Up to 3% prevalence >50 years, and 5% >65 years, with M:F ratio up to 10:1 [1].
  - Infrarenal AAA represents the most common location.
- **Etiology** [2]:
  - Chief pathophysiologic factors are atherosclerosis and smoking. Male gender, advanced age, dyslipidemia, and family history also contribute.
  - Inflammation, both primary or secondary to atherosclerosis, is increasingly recognized as a key factor that results in oxidative stress in the aortic media, deterioration of aortic tensile properties, and apoptosis of smooth muscle cells.

- There is an increased prevalence of AAA among first-degree relatives of affected individuals. Genetic basis is still unclear (may include structural proteins, proteases such as matrix metalloproteinases (MMP), or immunomodulatory genes).
- Bacteria and mycobacteria can generate infectious (also known as mycotic) aneurysms.

■ **History and Examination** [3, 4]

- Most AAA are asymptomatic, and diagnosed on physical examination or incidentally on imaging.
- Patients may have persistent pain in the lower abdomen or lower back, with a "gnawing" character.
- New or worsening pain may herald AAA expansion or rupture.
- Classic triad of AAA rupture = pain, hypotension, and pulsatile abdominal mass.
- Space-occupying effects of AAA include extremity ischemia, gastrointestinal or ureteral obstruction.
- Palpation of a pulsatile mass may help detect AAAs large enough to merit repair (sensitivity 68%, positive predictive value 43%), but alone is not sufficient to exclude AAA.
  - ■ Sensitivity correlates with AAA diameter (61% for 3.0–3.9 cm, 82% for > 5.0 cm), but sensitivity decreases with obesity.
  - ■ Palpation maneuvers for AAA are not believed to cause rupture.
- Auscultation of bruits does not help diagnose AAA.

■ **Screening and Diagnosis**

- Screening by exam and ultrasound is recommended by American College of Cardiology (ACC)/American Heart Association (AHA) (class I in 2006 guidelines) for males above age 60 who are siblings or offspring of parents with AAA.
- The United States Preventive Services Task Force (USPSTF) recommends abdominal ultrasonography screening for infrarenal AAA in all males age 65–74 who have ever smoked (ACC/AHA 2006 guidelines IIa recommendation)
- There are no recommendations for screening females or older males.

■ **Prognosis**:

- Risk of rupture varies with size. Annual risks are 0.3% for AAA diameter <4.0 cm, 1.5% for 4.0–4.9 cm, and 6.5% for 5.0–5.9 cm [5].
- Females with AAA have a greater risk of rupture than males, and experience rupture at smaller AAA diameters.
- Overall mortality from AAA rupture >50–80%.
- Mural thrombus within an AAA is associated with increased rates of growth and cardiovascular events.

■ **Medical Treatment**:

- Smoking cessation and lipid control (LDL goal <70 mg/dL) is essential.
  - ■ Studies of statins in AAA suggest possible reduction in AAA growth.
- Aspirin, reduction in BP, and reduction of dP/dt are reasonable.
- Beta-blockers carry a IIa recommendation for reducing the rate of AAA growth. For repair of atherosclerotic AAA, perioperative beta-blockade has a class I indication.
- Angiotensin converting enzyme inhibitors (ACEI) may, in addition to BP reduction, reduce rate of AAA rupture.
- Several studies have hinted a role for macrolide and tetracycline antibiotics based on a possible effect on Chlamydia (previously thought to be important in AAA pathogenesis), and for anti-inflammatory and anti-metalloproteinase properties. However, such therapies are not yet recommended for clinical use.

- **AAA Repair** [3]:
  - **Indications**: given high mortality from ruptured AAA, **prophylactic repair should be undertaken when infrarenal AAA is ≥5.5 cm. Infrarenal AAA of 4.0–5.4 cm should be re-imaged every 6–12 months.**
    - Size threshold may be smaller in females, in those with small body habitus, or those with family history of AAA or rupture.
    - Growth velocity >0.5 cm/year may be an indication for repair.
    - Symptoms always constitute an indication for repair.
    - Suprarenal AAA (or thoracoabdominal aneurysms, see below) may be repaired at sizes of 5.5–6.0 cm.
  - **Surgical repair of AAA**
    - Resection of aneurysm and replacement with a synthetic graft.
  - **Endovascular aortic aneurysm repair (EVAR)** [6–8]
    - Percutaneous fluoroscopically-guided deployment of an expanding endovascular stent inside the aneurysm and attached to the aorta at the proximal and distal aneurysm margins, thereby excluding the aneurysm from aortic blood flow.
    - Only about half of AAA patients have anatomy suitable for EVAR: anatomic considerations include aneurysm length, proximal and distal landing zones, tortuosity, aneurysm thrombus or calcium, iliac artery diameter.
    - EVAR reduces peri- and immediate post-procedure morbidity and mortality and post-operative hospitalization, but whether the long-term outcomes are improved or are as durable as open surgical repair remains under investigation. Randomized trials suggest no difference in long-term mortality (EVAR-1 and DREAM trials), although a single retrospective Medicare analysis from 2012 suggests higher all-cause mortality at 2.5 years from open repair vs. EVAR [9].
    - Endoleak: EVAR is associated with endoleak, or persistent blood flow into the aneurysm sac due to inability to completely exclude it from circulation.
    - Post-EVAR patients require imaging surveillance at 1, 6, and 12-months, in order to monitor for endoleaks, assess graft position, check aneurysm sac size, and gauge need for reintervention (class I).
    - Repair endoleaks that leak into aneurysm sac around an imperfect seal at proximal and/or distal anastomosis of stent graft OR structural defect, e.g. tear, stent fracture, etc.
    - EVAR patients have an approximately 10% higher reintervention rate at 6 years compared to open repair.
  - The 2011 ACC/AHA guidelines on peripheral artery disease (PAD) give a class I recommendation for "open or endovascular repair of infrarenal AAAs" in "good surgical candidates."
    - There is a class IIa recommendation for open AAA repair in good surgical candidates who could not comply with surveillance imaging post-EVAR.
  - Due to short-term advantages, EVAR has been considered for higher risk patients (e.g. older, high perioperative risks). However, the EVAR-2 trial studied patients deemed "physically ineligible" to undergo open repair and found no improvement in all-cause mortality versus medical therapy alone.
    - The 2011 ACC/AHA PAD guidelines give EVAR a class IIb recommendation in high risk surgical patients (uncertain benefit in this group).

Thoracic Aortic Aneurysm (TAA)

- **Epidemiology**:
  - TAA is believed to be about one-third as common as AAA. Because TAA is a clinically silent disease, the incidence is estimated from autopsy series at 3–4%.
  - TAA is most commonly seen > age 50; the age of onset is earlier than for AAA.
  - Male:Female ratio is ~2:1, as compared to AAA which has a much higher ratio.
- **Anatomic location:**
  - Ascending aorta: 60%
    - Root aneurysms are associated with Marfan syndrome
    - Ascending aortic aneurysms are associated with bicuspid aortic valve or sporadic aneurysms.
  - Arch: 10%
  - Descending thoracic aorta: 40%
  - Thoracoabdominal (see below): <10%
  - Multiple aneurysms: <10%
- **Etiology** [10–12]:
  - HTN and atherosclerosis are the primary risk factor for non-syndromic descending and thoracoabdominal aneurysms.
  - For root and ascending aneurysms, medial degeneration is the final common etiopathologic pathway.
    - Medial degeneration may be acquired (e.g. HTN) or congenital (e.g. Marfan syndrome).
    - Medial degeneration (previously called cystic medial necrosis), involves smooth muscle cell apoptosis, elastic fiber degeneration (particularly important in Marfan syndrome), and subintimal spaces infiltrated with mucoid proteoglycan.
    - MMP's are also implicated.
  - Bicuspid aortic valve (AV) is the most common cardiac congenital anomaly (~1–2% population prevalence; Male:Female 3:1), and is associated with TAA, dissection, and coarctation [13].
    - Bicuspid AV is associated with aortic medial degeneration.
  - Genetic TAA syndromes:
    - *Marfan syndrome:* Besides thoracic aortopathy, manifestations include valvular, skeletal, and ocular pathology. The etiology is an autosomal dominant defect in fibrillin-1, a structural glycoprotein in the extracellular matrix of the aorta media. Fibrillin-1 is also involved in downregulating the activity of TGFβ.
      - Aortic root dilatation is present in 80% of Marfan adults. Aneurysms may also appear in carotid/other cerebral arteries and the abdominal aorta.
    - *Loeys-Dietz syndrome:* Mutations in the TGFβ receptor cause a syndrome of arterial tortuosity with hypertelorism, bifid uvula, and cleft palate.
    - *Ehlers-Danlos syndrome, type IV*: This autosomal dominant defect in type III procollagen affects large and medium arteries, causing carotid and vertebral dissections. It also causes characteristic facial features and also marked weakness of skin, gastrointestinal, and uterine structures.
    - *Familial thoracic aortic aneurysm syndrome:* These are found in 20% of those with unexplained TAAs. There is no one genetic defect to screen for.

- Syphilis, once a common infectious cause of saccular TAA, is now a rarity.
- Autoimmune conditions may be associated with aortitis and secondary TAA (see below).

■ **History and Examination** [11, 12]

- TAAs are in general asymptomatic, but patients may have chronic back or chest pain, with the location of pain related to anatomic location of TAA. A pulsating sensation may be reported.
- The space-occupying TAA can cause symptoms depending on location and the structure affected (Table 14-2).
- Aortic root or ascending aortic aneurysms may present with AI or even HF.
- TAA may erode into the spine or esophagus (ask about hemoptysis).
- TAA may present with thromboembolic phenomena, e.g. to cerebral, spinal, visceral, or extremity arteries.
- Acute pain may herald dissection or impending rupture.
  - ■ Location of pain has some correlation with anatomy: anterior pain with ascending TAAs, neck pain with arch aneurysms, interscapular pain with DTAA.

■ **Screening and Diagnosis**:

- Most TAA are recognized incidentally on CXR, CT, or TTE.
- TTE is recommended as the first test for assessment of the ascending aorta in known or suspected connective tissue disorders, or genetic conditions that predispose to TAA (2011 ACC/AHA Appropriate Use Criteria for Echocardiography) [14].
- There are no consensus guidelines on population screening for TAA.
- For first-degree relatives of patients with TAA, screening with echocardiography or CT is recommended.
- In patients with bicuspid AV, ACC/AHA 2006 valve guidelines give a class I recommendation for an initial TTE for assessment of aortic root and ascending aortic dimensions [15].
  - ■ If the TTE is insufficient to document morphology and dimensions, or ascending aortic dilatation is present, then CT or MR is reasonable.
  - ■ Bicuspid AV patients with root or ascending aorta >4.0 cm should have yearly imaging (though the size cutoff can be reduced for smaller stature patients).

■ **Prognosis** [10–12]:

- Growth velocity of TAA is variable but approximated at 0.5–5 mm/year; growth rates are higher for larger aneurysms and descending TAA.
- Annual risks of dissection or rupture vary with TAA size, from <2% for diameter <5.0 cm, 3% for 5.0–5.9 cm, and 6.9% for >6.0 cm.
  - ■ Patients with underlying connective tissue disease such as Marfan, Loeys-Dietz, and Ehlers-Danlos syndromes experience rupture at smaller sizes.
- TAA rupture causes ~75% mortality at 24 h.

TABLE 14-2

SPACE-OCCUPYING SYMPTOMS OF TAA

AFFECTED STRUCTURE	RESULTANT SYMPTOMS
Coronary arteries	Chest pain
Trachea, bronchioles	Dyspnea, stridor, wheeze, cough
Esophagus	Dysphagia
Superior vena cava (SVC)	SVC syndrome
Recurrent laryngeal nerve	Hoarseness (Ortner's syndrome)
Spinal cord compression	Horner's syndrome, paraplegia

- **Surveillance imaging**
  - Once thoracic aortic pathology is detected, a full imaging evaluation of the thoracic aorta should be performed to document extent of disease and baseline aortic diameters.
  - ACC/AHA guidelines recommend annual imaging for following most aneurysms.
    - It is also reasonable to obtain the first follow-up imaging exam at 6 months after diagnosis to exclude rapid growth (as may be seen in aortitis).
    - Once a TAA growth trajectory is stable, ACC/AHA guidelines consider imaging every 2–3 years for smaller TAA in older patients.
  - Re-imaging should be considered for a change in clinical status or physical exam.
- **Medical management** [12]
  - The primary goals are to reduce dP/dt and BP in order to reduce aortic wall tension and reduce the risk of aortic dissection or rupture
  - Goal heart rate <60 beats per minute (bpm) and systolic BP <110–120 mmHg.
  - Trial data is limited [25, 26]:
    - Beta-blockers have been the mainstay of medical treatment for TAA. While beta-blockers have been demonstrated to reduce the rate of aneurysm growth in Marfan patients with large aneurysms, their efficacy in aneurysms of other etiologies has not been proven.
    - Losartan, an angiotensin receptor blocker, is also a TGFβ antagonist, and has been proven to dramatically reduce the rate of aneurysm growth in a mouse model of Marfan syndrome. Angiotensin receptor blockers have previously been shown to slow aneurysm growth in a small non-randomized study of children with Marfan syndrome. However, a recent randomized trial of losartan versus atenolol in children and young adults with Marfan's syndrome found no significant difference between rates of aortic root dilatation between the two treatment groups.
  - Burst precautions:
    - Patients should avoid activity or exertion that can acutely raise aortic wall stress.
    - Guidelines suggest avoiding heavy lifting or straining, i.e., isometric activities that would require the Valsalva maneuver.
- **TAA Repair** [11, 12]
  - **Indications**: Suggested criteria for elective TAA repair based on ACC/AHA guidelines include the following diameter thresholds:
    - **Repair of ascending TAA at a diameter of ≥5.5 cm, but at a lower diameter of ≥5.0 cm for those with Marfan syndrome or a familial thoracic aortic aneurysm syndrome, and at a diameter of ≥4.4 cm for Loeys-Dietz syndrome.**
      - For those with Marfan syndrome and bicuspid AV who have either a small or large body habitus, surgery is recommended when the ratio of the maximal root or ascending aortic cross-sectional area (in square centimeters) to patient height (in meters) is >10.
      - The 2016 revised ACC AHA guidelines raised the threshold for TAA repair back to **≥5.5 cm** for asymptomatic patients with bicuspid AV. Repair is still indicated at **≥5 cm** for bicuspid AV patients with additional risk factors (family history of dissection or rapid growth rate) for patients at low surgical risk [16].
    - Arch diameter ≥5.5 cm.
    - Descending TAA diameter ≥5.5–6.0 cm.
    - Rapid growth rate of a TAA >0.5 cm/year.

- In patients undergoing cardiothoracic surgery for another indication (e.g., coronary artery bypass graft (CABG), AV repair), an aortic root or ascending aortic diameter **≥4.5 cm** may be repaired.
- TAA symptoms are an indication for repair.

– **Open surgical approach**

- Ascending and arch TAA require median sternotomy, while descending TAA and thoracoabdominal aortic aneurysms (TAAA) are approached via left thoracotomy.
  - Surgical repair of descending TAA and TAAA are associated with significant morbidity, including risk of spinal cord ischemia and paraplegia. Various neuroprotective strategies help reduce spinal cord ischemia.
- Repair of arch aneurysms is usually performed with insertion of prefabricated branched graft and supported by antegrade cerebral perfusion.
- Root aneurysms used to require sacrificing the aortic valve and insertion of a valved-conduit (composite aortic graft or Bentall procedure). Now the aortic valve can usually be preserved and resuspended within the prosthetic graft (valve-sparing root repair or David procedure).
- In recent decades, overall surgical mortality has declined from 10–20% to approximately 5%.

– **Thoracic endovascular aortic repair (TEVAR)**

- For descending TAA, TEVAR is an alternative to open repair when anatomy is conducive.
- Akin to EVAR for AAA, TEVAR provides an upfront reduction in morbidity and mortality, but long-term mortality benefits are not proven.
- Although there are no randomized trials of TEVAR versus open repair, large registry and metaanalysis data (a mix of aneurysms and dissections) have been favorable. TEVAR is therefore now recommended for descending TAA ≥5.5 cm; the Society of Thoracic Surgeons (STS) recommends TEVAR as class IIa if there are comorbidities, and class IIb if no comorbidities.
- TEVAR also results in endoleak (12–18% incidence), and surveillance is therefore indicated at 1, 3, 6, and 12-months post-procedure.

Prevalence of Concurrent Aneurysms [17]

The presence of an aortic aneurysm is associated with increased prevalence of aneurysm at another location.

- About 1/4 of TAA patients have an AAA.
- Similarly, about 1/4 of AAA patients will have a TAA.
- Consider pan-aortic imaging initially in any patient with either AAA or TAA.
- Popliteal and iliac artery aneurysms are common in AAA/TAA patients.

Thoracoabdominal Aortic Aneurysms (TAAA)

TAAA involve the descending thoracic aorta and extend to the abdominal aorta.

- TAAA and its repair are classified by Crawford types.
  - Types I and II each involve the majority of descending thoracic aorta from the left subclavian artery, with Type I involving the proximal portion of the abdominal aorta and type II extending to the infrarenal abdominal aorta.

- Type III involves the distal descending thoracic aorta and below the diaphragm a large part of the abdominal aorta.
- Type IV involves the diaphragmatic aorta and most of the abdominal aorta.

■ TAAA repair is indicated at a diameter ≥5.5–6.0 cm.

ACUTE AORTIC SYNDROMES: AORTIC DISSECTION, INTRAMURAL HEMATOMA (IMH), PENETRATING ATHEROSCLEROTIC ULCER (PAU)

Patients with these syndromes present similarly regardless of mechanism.

Aortic Dissection

■ **Definition**: Penetration of blood from the aortic lumen into the medial layer of the aortic wall, due to a tear in the intimal layer, resulting in splitting the media into layers and creating a second channel for blood flow called the false lumen.

■ **Classification**: Because morbidity, mortality, and management are dictated by whether or not the dissection involves the ascending thoracic aorta, classification schemes have been designed to distinguish those that involve the ascending aorta from those that do not (see Fig. 14-1).

- *DeBakey Classification*: type I originates in ascending aorta and progresses to arch ± descending aorta; type II is confined to ascending aorta; type III originates in and is generally localized to the descending aorta.
- *Stanford*: type A involves ascending aorta regardless of origin; type B is distal to the ascending thoracic aorta.

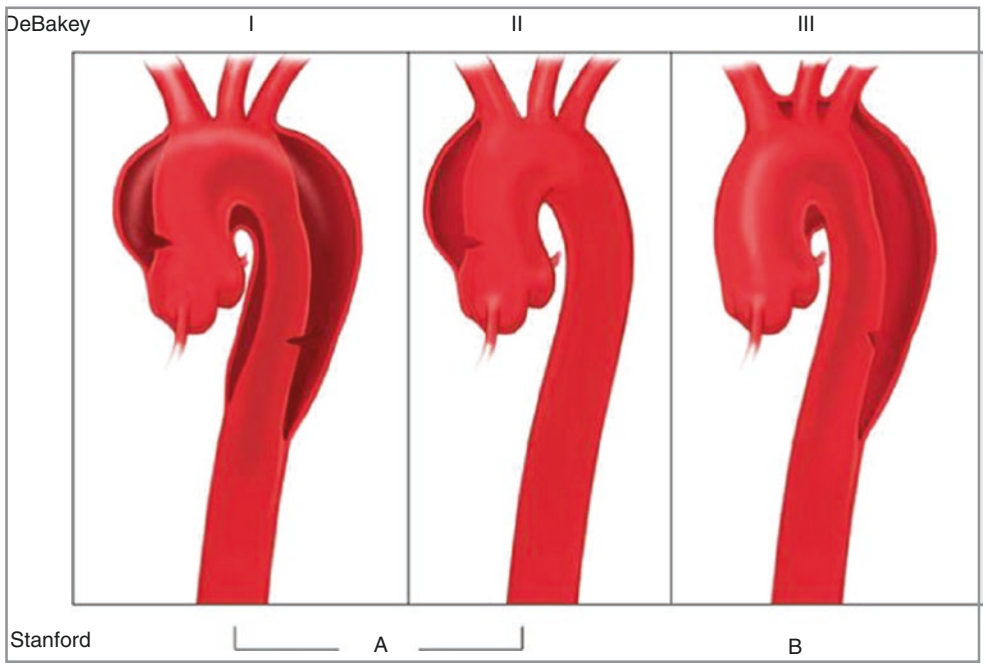

FIGURE 14-1

Classification systems for aortic dissection. © Massachusetts General Hospital Thoracic Aortic Center, used with permission

- **Epidemiology**: Estimated incidence of 2–16 cases per 100,000 person-years, but incidence likely underestimated due to early rapid mortality (out-of-hospital deaths attributed to other causes; misdiagnosis of aortic dissection).
  - Type A dissection is twice as common as type B.
  - Marfan syndrome accounts for approximately 5% of dissections, and up to 50% of patients under age 40.
  - Bicuspid AV accounts for more cases of dissection than Marfan syndrome; even though the risk of dissection per patient is less than in Marfan syndrome, the fact bicuspid AV has a prevalence of 1–2% makes it quantitatively an important cause of aortic dissection.
  - When aortic dissection occurs in women less than 40 years of age, 12% occur in the peripartum period of pregnancy.
- **Etiology**
  - Dissection is common at sites of aneurysm, and thus risk factors for aneurysm and dissection are similar, e.g. HTN, BAV, Marfan syndrome, family history of TAA or dissection.
    - However, dissection may occur in non-aneurysmal aortic segment.
    - Procedures that involve manipulation of the aorta (e.g., catheterization, intra-aortic balloon pumps, and cardiac surgery) may cause iatrogenic dissection.
    - Cocaine may trigger dissection.
    - Traumatic aortic injury typically causes aortic transsection but can cause dissection.
- **History and Examination** [11, 18, 19] (see Table 14-3)
  - Pain is the most common symptom of dissection (~90%). Evaluating characteristics of the pain by questions about onset, intensity, quality, and radiation can be very helpful, as physicians that asked these questions correctly identified dissection in ~90% of cases versus ~50% when a question was skipped.
    - Abrupt onset is reported in 84% of cases. It is typically maximal at onset, which is not typical in acute coronary syndromes.
    - Pain is characterized as severe in 90% of cases.

TABLE 14-3

SIGNS AND SYMPTOMS OF AORTIC DISSECTION

SIGNS AND SYMPTOMS	POSITIVE LR	SENSITIVITY (%)
Severe pain		90
Enlarged aorta or wide mediastinum on CXR	2.0–3.4	90
Sudden onset	1.6–2.6	84
AI murmur		45
Ripping or tearing	1.2–10.8	39
Radiating or migrating	1.1–7.6	31
Pulse or BP differential	5.7	26–31
Diastolic murmur	1.4	28
Neurological deficit	6.6–33.0	17
Syncope		13
Shock		13
Lower extremity ischemia		10
Stroke		6–8
Tamponade		5
Spinal cord ischemia		2

- The quality is classically thought to be "ripping" or "tearing" but "sharp" (51%) or "stabbing" (64%) pain is more often reported.
- Location of pain may correlate with that of the dissection, and migrates as the dissection propagates.
  - Type A dissections are twice as likely to have anterior versus posterior chest or back pain.
  - Type B dissections have back pain in 2/3 of cases, but chest pain may also occur.
  - Abdominal pain may be the sole manifestation of dissection, and pain may also be localized in neck, lower back, or extremities.
- The pain is often persistent, but it can abate.
- When pain is absent, patients usually present with syncope, stroke, or heart failure.

– Blood pressure

- At presentation, approximately one third of patients are hypertensive, while one seventh are hypotensive and one seventh are in shock.
- HTN is a more common presentation for Type B dissections.
- Hypotension in a type A dissection suggests severe AI, tamponade, or coronary ischemia.
- BP must be measured in both arms, and frequently needs to measured in both arms and both legs, in order to recognize pseudohypotension, or a spuriously low BP measurement due to dissection affecting the a branch artery.
- A BP differential of > 20 mmHg is considered significant.

– Syncope is seen commonly in type A dissection (19%) and uncommonly in type B (3%).
– Focal neurologic deficit is present in ~1/6 cases of thoracic aortic dissection.
– Whenever a constellation of cardiovascular, neurologic, and abdominal symptoms is otherwise unexplained, an acute aortic syndrome must be considered as a potential unifying diagnosis.

■ **Sequelae of Aortic Dissection**

– Hemopericardium resulting in pericardial effusion and potentially cardiac tamponade and shock.
– Acute AI, potentially causing heart failure or shock.
– Acute myocardial infarction (inferior more common than anterior, due to predilection of dissection to extend into right coronary artery)
– Dissection involving other branch arteries: Carotid (stroke or transient ischemic attack), spinal (paraplegia), renal (acute renal insufficiency), mesenteric (abdominal pain, mesenteric ischemia), or iliac arteries (lower extremity pain, ischemia).

■ **Diagnosis** [11, 18, 20]

– ECG and CXR are quick tests (especially for patients at low and intermediate risk of dissection) that may reveal an alternate explanation for the presenting symptoms. However, they cannot be used to rule in or out aortic dissection.
– To definitively diagnose or exclude aortic dissection, one should obtain a CT angiogram, a TEE, or an MR angiogram. In most settings CT is preferred. The ultimate choice of diagnostic tests depends on which modalities are readily available at a given institution and the expertise with which a test can be performed and interpreted.
– When clinical suspicion for dissection is quite high, one negative imaging modality (CT, TEE, or MR) may not be sufficient to fully exclude the diagnosis, and the evaluation should include a second confirmatory test (ACC/AHA guideline, class I).
– Biomarkers: D-dimer alone cannot be used to exclude aortic dissection as the negative predictive value is only ~97% however it can be helpful in cases where the probability of dissection is already low. The D-dimer can be normal in causes of intramural hematoma.

- **Prognosis**
  - Type A
    - Immediate death rate may be as high as 40%.
    - Mortality is estimated at 1–2% per hour after dissection.
    - Death is commonly related to hemopericardium and tamponade, rupture, or propagation of dissection. Survival can be improved with early recognition and treatment.
  - Type B
    - Overall 30-day mortality is 10% for uncomplicated patients managed medically but rises to 30% among patients with complications who require surgical treatment.
- **Treatment of type A dissection** [11, 21]
  - Urgent surgical repair is indicated for acute type A dissections.
  - The goal of surgery is to replace the dissected ascending aorta, in order to prevent death from aortic rupture.
  - The aortic arch is typically not repaired in the acute setting, unless the arch is significantly dilated or the intimal tear is located within the arch.
  - Preoperative coronary angiography is typically not indicated as it causes an unnecessary delay in aortic repair.
  - Medical therapy should be instituted while awaiting operative repair.
  - dP/dt reduction and HR goal <60 represent primary goals, with secondary goal SBP <100–120 mmHg (or to the minimum level that preserves perfusion).
  - IV beta-blockade should be started first, so as to avoid reflex tachycardia (and thus increased dP/dt) from vasodilator therapy. Propranolol, metoprolol, esmolol, or labetalol are all reasonable; when beta-blockers are contraindicated, IV diltiazem or verapamil should be considered.
  - Sodium nitroprusside can also be used for rapid reduction and careful titration of BP.
  - Analgesia is necessary to blunt pain-related increases in HR and BP.
  - Hypotension: If due to tamponade, the treatment is volume resuscitation and emergent surgery; pericardiocentesis is not helpful and in fact associated with increased mortality (and should only be considered en route to the operating room or after cardiopulmonary bypass is established, unless the patient is in arrest).
- **Treatment of type B dissection**
  - Uncomplicated type B dissection is managed medically with reduction of dP/dt and BP, as detailed above.
  - Intervention is indicated for complications, including malperfusion syndromes, refractory pain, refractory HTN, enlarging aneurysms, or rupture.
  - Endovascular intervention is now generally preferred over open surgical repair to treat complicated type B dissections because they are associated with a significant lower 30-day mortality.
    - However, the long term impact of stent-grafting an acutely dissected descending aorta remains unknown.
    - In has been theorized that early stent-grafting of uncomplicated type B dissections might serve to prevent potential late complications or aneurysm expansion, and a randomized trial is underway, but at present there are no data to support such a strategy.
- **Late complications**: Patients are at risk and must be followed for aneurysm formation, recurrent dissection, rupture, AI (for those involving the root or ascending aorta), and endoleak (following TEVAR procedures).

- The highest risk is in the first 1–2 years.
- Medications to reduce dP/dt and control HTN can reduce the rate of complications. The goal is a HR of <60 and a SBP of <120.
  - Beta-blockers have been shown to improve late outcomes and therefore represent the mainstay of chronic therapy.
  - Calcium channel blockers, ACE inhibitors, and angiotensin receptor blockers may also be of benefit but the data are unclear.
- Because of the risk of progressive aortic growth following acute aortic dissection, patients should undergo serial surveillance imaging at intervals of 1, 3, 6, and 12-months and then annually thereafter, if stable.

Intramural Hematoma (IMH) [22]

- **Definition**: Bleeding contained within the media of the aortic wall but without communication with the aortic lumen.
- **Etiology**: Some may be due to rupture of the vasa vasorum within the aortic wall, whereas others appear secondary to microscopic intimal tears.
- **Epidemiology**: The risk factors are similar to those of aortic dissection, although it is less commonly seen among those with Marfan syndrome.
  - IMH accounts for 6–10% of acute aortic syndromes.
  - The descending aorta is affected in 60% of cases.
- **History and Examination:** clinical presentation is similar to classic aortic dissection.
- **Diagnosis**: IMH is diagnosed using CT, MR, or TEE.
  - Unlike classic aortic dissection, there is no intimal flap or blood flow within the aortic wall. Instead, IMH appears as a crescentic or circumferential thickening of the aortic wall. The presence of thrombus in the aortic wall has higher intensity on CT scanning, and does not enhance with contrast. Aortography can easily miss IMH.
- **Treatment and prognosis** and similar to classic aortic dissection.
  - IMH may convert to classic dissection in ~10% of cases.
  - Surveillance imaging after an acute IMH is similar to classic dissection.

Penetrating Atherosclerotic Ulcer for the Aorta (PAU)

- **Definition**: An atherosclerotic plaque breaches the internal elastic lamina, allowing blood to penetrate the wall to a varying degree.
- **Epidemiology**: PAUs are most prevalent in older patients with a history of atherosclerosis, HTN, and smoking.
- PAUs appear most often in the mid-to-distal descending thoracic aorta (90%) due to the prevalence of atherosclerosis in this segment.
- PAU may convert to typical dissection, IMH, a saccular aneurysm, or pseudoaneurysm, which in turn may lead to rupture.
- **Treatment**: Small PAUs are managed medically with antihypertensives and surveillance imaging. Large or expanding PAUs, or ulcers that have caused pseudoaneurysms, may require intervention, and TEVAR is generally preferred given that this population tends to be older and at high risk from open repair.

Aortic Transsection

- **Definition**: Through-and-through tear involving all three layers of the aortic wall. It may be partial or complete, in which case the aorta is completely severed. Such injuries may lead to fatal exsanguination, but in some cases the patient survives because the bleeding is contained by a pseudoaneurysm.
- Traumatic aortic tears result from deceleration injuries, and therefore typically occur near anatomic sites at which the aorta is anchored in the chest, i.e., the ligamentum arteriosum and aortic root.
- **Diagnosis** is most readily and accurately made by CT angiography.
- **Treatment**: intervention is required: For tears near the ligamentum, TEVAR is the treatment of choice; for tears in the aortic root open cardiac, surgery is required.

VASCULITIDES INVOLVING THE AORTA [23]

Giant Cell Arteritis (GCA)

GCA, also known as temporal arteritis, occurs most often in those older than age 50 (especially among those >75 years old). Women outnumber men 2:1, and it is more common among those with northern European ethnicity.

- It affects elastic arteries, including the aorta and extracranial (but not intracranial) arteries.
- Polymyalgia rheumatica, an inflammatory condition characterized by constitutional symptoms and shoulder and hip pain and stiffness, is found in about half of patients with GCA.
  - Approximately 15% of patients with polymyalgia rheumatica have GCA.
- Constitutional symptoms including malaise, anorexia, weight loss, and fevers are common.
- Aortic aneurysms: More than 10% can develop thoracic aneurysm as a late manifestation.
- Therefore surveillance imaging is recommended for up to 10 years after onset.
- **History and Examination**: mainly due to arterial occlusion:
  - Temporal artery: Ocular symptoms (double or blurry vision may precede permanent blindness), temporal headache, jaw claudication, scalp tenderness.
  - Vertebral artery: Symptoms of vertigo, dizziness, cerebellar signs, stroke.
  - Axillosubclavian arteries: Symptoms of arm claudication, absent pulse.
- American College of Rheumatology diagnostic criteria include: age >50, new headache, temporal artery tenderness or diminished pulse, erythrocyte sedimentation rate >50 mm/h, necrotizing vasculitis on biopsy.
- **Treatment**:
  - Corticosteroids are the principal treatment of GCA; therapy over 1-2 years is used to prevent recurrence.
  - Biopsy should not delay initiation of steroids when GCA is suspected; the biopsy will have reasonable yield even within a few days of starting steroids.
  - Other immunomodulators are as efficacious as corticosteroids.

Takayasu Arteritis

- Takayasu arteritis, sometimes referred to as "pulseless disease," is an idiopathic granulomatous vasculitis affecting large and medium sized muscular arteries (aorta, brachiocephalic arteries, pulmonary artery).
- Takayasu arteritis is most common in women (Male:Female ~1:10) in the second to third decades. Takayasu arteritis is more common in East Asia and Africa than in Europe or North America.
- Takayasu arteritis follows an early inflammatory stage (marked by a non-specific systemic inflammatory state with fever, sweats, and weight loss) followed by a later sclerosing phase.
  - The non-specific initial stage results in delayed diagnosis so that 90% of patients present in the sclerotic phase at diagnosis.
  - Arteritis can manifest as stenosis or aneurysm, and aortic involvement may be patchy.
- **History and Examination**: vary by affected artery
  - Aorta: AI, myocardial ischemia or infarction due to stenoses of the coronary artery ostia.
  - Subclavian: Decreased upper extremity BP (or a BP differential), pain in upper extremities, bruits.
  - Carotid: Amaurosis fugax, stroke.
  - Renal: Marked HTN.
- **Diagnosis**:
  - Diagnostic criteria from the American College of Rheumatology include intermittent claudication, diminished pulses, subclavian bruits, BP differential, and angiographic evidence of aortic or branch vessel stenoses.
  - Angiography can reveal both stenoses and aneurysms.
- **Treatment**:
  - High dose corticosteroids are the mainstay of treatment and the treatment course may need to extend 1–2 years. ACC/AHA recommends periodic monitoring of disease activity by exam and/or inflammatory markers.
  - Cyclophosphamide, azathioprine, methotrexate, or tumor necrosis factor inhibitors may be used in cases where either systemic inflammatory symptoms recur, vascular disease continues to progress, or inflammatory markers rise.
  - Surgical bypass (or reconstruction) and balloon angioplasty are options to treat severe arterial stenoses.

IgG4-Related Disease [24]

- IgG4-related disease is an autoimmune condition characterized by overproduction of IgG4 with lymphoplasmacytic and eosinophil tissue infiltrate, obliterative phlebitis, and fibrosis.
- IgG4-related disease affects a number of glandular tissues but a lymphoplasmacytic aortitis has been described that may generate aneurysm and dissection.
- In a single center experience, IgG4-related disease caused approximately 9% of non-infectious thoracic aortitis.

TABLE 14-4

QUICK REVIEW

FINDINGS	IMPLICATION
Elderly male, abdominal pain, pulsatile mass	AAA rupture
Ehlers-Danlos syndrome type IV	Risk of ascending aortic aneurysms
Loeys-Dietz syndrome	Mutations in the TGFβ receptor; treat with losartan
dP/dt	Principle to reduce wall stress by reducing HR and BP
Marfan syndrome	High risk for aortic root aneurysms and dissection
DeBakey classification system	Types I and II involve the ascending aorta
Stanford classification system	Type A involves the ascending aorta
Histologic pattern in aneurysm wall	Medial degeneration
Laplace's Law	Wall tension proportional to product of pressure and radius
Type A aortic dissection	High risk of rupture, tamponade, and death
Genetic defect in Marfan syndrome	Mutation in *FBN-1*, the gene for fibrillin-1
Prevalence of BAV	1–2% of the general population
Pseudohypotension	Falsely low BP in due to compromise of branch artery in aortic dissection
Intramural hematoma	Blood in aortic media that does not communicate with aortic lumen
Syndrome associated with giant cell arteritis	Polymyalgia rheumatica
Hoarseness	Recurrent laryngeal nerve compression (Ortner's syndrome) by a large TAA
Myocardial infarction in aortic dissection	Type A dissection with compromise of the right coronary artery ostium
Paraplegia after descending thoracic aortic aneurysm repair	Major risk associated with surgery on the descending or thoracoabdominal aorta
Bicuspid valve repair indicated at ≥5.5 cm if asymptomatic	Repair safe to be delayed for asymptomatic patients
Aortic injury following a motor vehicle accident	Deceleration injury causing aortic transsection; most often occurs at ligamentum arteriosum
Circle of Willis aneurysm in aortic dissection patient	Coarctation of the aorta is the likely underlying lesion

QUICK REVIEW (TABLE 14-4)

Questions and Answers

1. A 41 year old female acquaintance was evaluated for a heart murmur. On exam she had a systolic ejection click and faint systolic ejection murmur. An echocardiogram reported a bicuspid aortic valve with a horizontal commissure and, but the valve functioned well and there was no stenosis and trace aortic insufficiency. Her aortic root diameter was 3.6 cm. Her cardiologist reassured her that there is nothing further to do and she should simply follow-up with her internist annually. She is anxious and has therefore called you for advice. What is the most prudent suggestion to offer?

A. The cardiologist's plan is sound, so she should follow up with her internist.
B. She should undergo genetic testing for a mutation in the gene for fibrillin-1.
C. Her first degree relative should be screened for thoracic aortic aneurysms.
D. She should undergo a CT or MR to determine if her ascending thoracic aorta is dilated.
E. She should undergo annual surveillance echocardiograms to monitor her bicuspid aortic valve function.

1. Answer D. The echocardiogram documented a normal aortic root diameter, but no mention was made of the size of the ascending thoracic aorta. The aortic root and ascending thoracic aorta are distinct anatomically, and one aortic segment may be enlarged while the other is normal in size. Therefore unless both diameters are documented in the report as normal, a dilated aorta cannot be excluded. Half of those with bicuspid aortic valve have a dilated proximal aorta, and the majority of such aneurysms involve the ascending aorta rather than the root, so in any patient diagnosed with a bicuspid aortic valve the ascending thoracic aortic diameter must be evaluated. Since this echocardiogram apparently did not exclude a dilated ascending aorta another imaging study, either a CT or MR, should be obtained. There is no indication to test for the fibrillin-1 mutation in the setting of bicuspid aortic valve, as this is an abnormality associated with Marfan syndrome. The patient has a bicuspid aortic valve but has not yet been found to have a TAA, so there is yet no indication to screen her first-degree relatives for aneurysms. Her bicuspid aortic valve functions well, so there is no indication for annual surveillance echocardiograms to monitor the valve.

2. A 66 year old male new to your practice has a past history of uncontrolled HTN and a descending thoracic aortic aneurysm, and four months ago suffered a type B aortic dissection that was managed with TEVAR. Which of the following is not endorsed in ACC/AHA guidelines for this patient?
   A. Chronic beta-blocker therapy
   B. Surveillance imaging of the thoracic aorta at 1, 3, 6 and 12 months following the aortic dissection.
   C. D-dimer to assess for degree of thrombosis of false lumen.
   D. Pan-aortic imaging to exclude e.g. a concurrent AAA.
   E. Burst precautions.
   F. All of the above are indicated or recommended.

2. Answer C. The 2010 ACC/AHA guidelines on thoracic aortic disease highlight beta-blockade as an integral therapy for patients following aortic dissection. Serial imaging is recommended at regular intervals following an acute dissection given the risk of rapid early growth of dissected segments of the aorta. Burst precautions, or avoiding heavy lifting, straining, or pushing that would raise aortic pressure, is prudent. Because of the prevalence of concurrent aneurysms, patients with either TAA or AAA should have the entire aorta imaged on at least one occasion. While degree of thrombosis of a false lumen of a type B dissection correlates with mortality, D-dimer testing is neither helpful nor indicated in this context.

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