Brigham and Women's Hospital
Founding Member, Mass General Brigham

Aortic Diseases

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HARVARD MEDICAL SCHOOL
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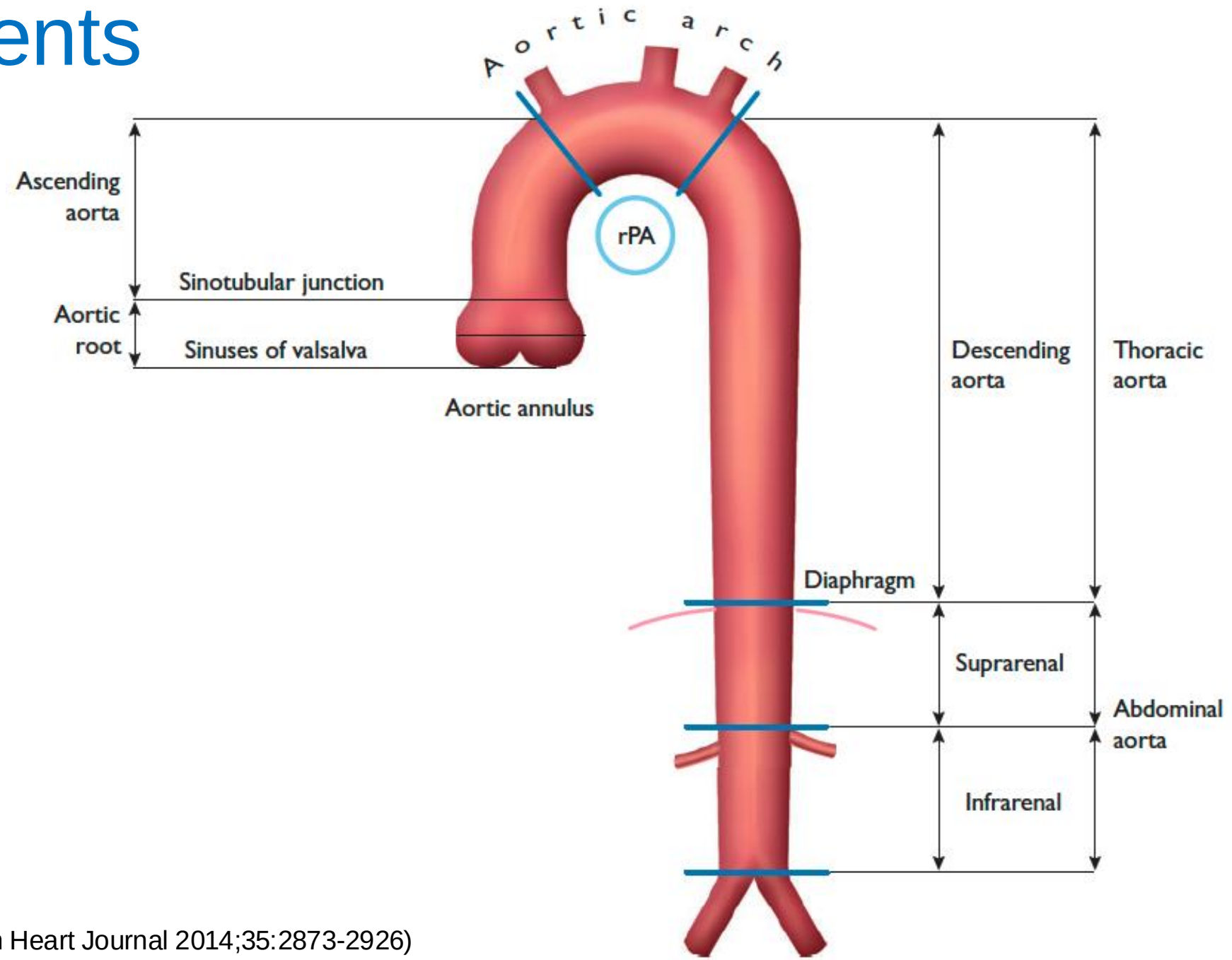
Learning Objectives

1. Familiarize with the breadth of aortic diseases
2. Review the clinical presentation of aortic disease
3. Familiarize with the outpatient management of stable aortic diseases
4. Familiarize with with the management of acute aortic syndromes

Aortic Segments



(Erbel R al. European Heart Journal 2014;35:2873-2926)

Definition

- The normal size of the aorta depends on sex, body size, and age
- For adult patients, aortic diameters are normalized using a ratio of aortic diameter to BSA or aortic diameter to height
- An aneurysm is defined as a dilatation of an artery that is ≥1.5 times the expected normal diameter
- This threshold can be used to define aneurysms of the descending and abdominal aorta

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

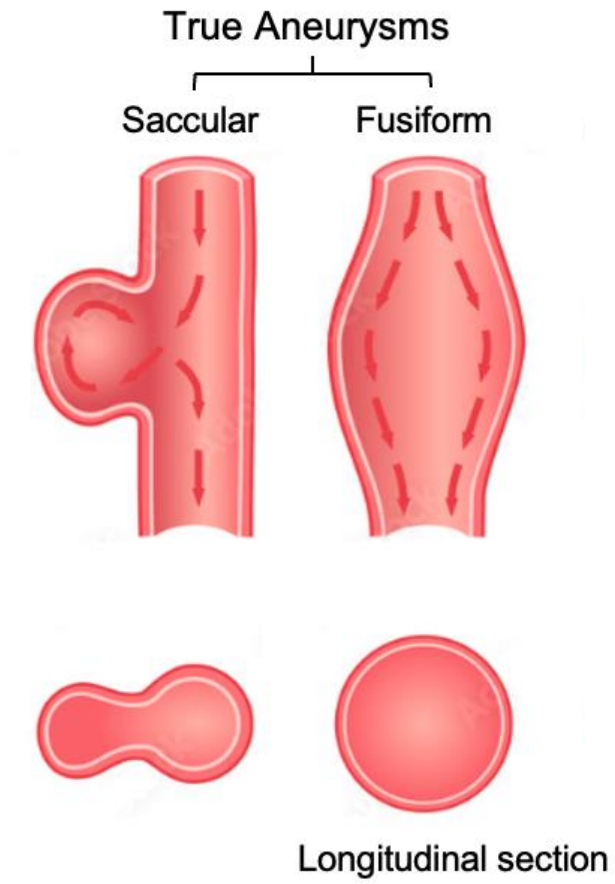
Normal Size of the Aorta

- A different definition is necessary for the ascending aorta, because the risk of dissection increases substantially at diameters below the 1.5 times threshold
- The most recent guidelines define an aortic root or ascending aortic diameter of 4.0-4.4 cm as dilatation and 4.5 cm or higher diameter as aneurysm

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

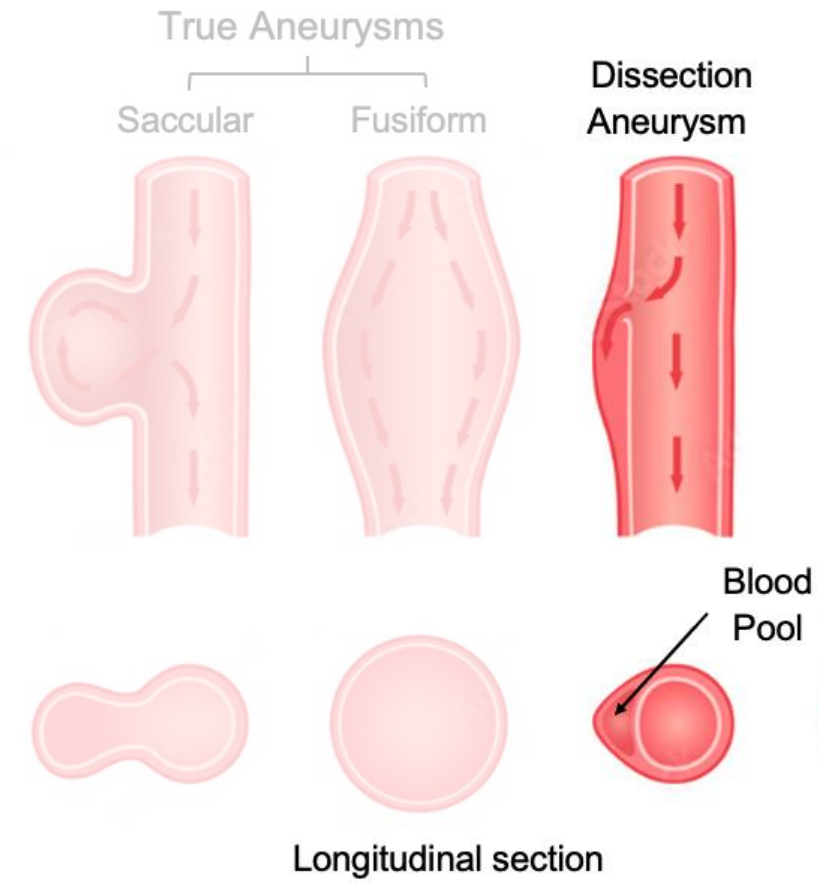
Types of Aneurysms

- True aneurysms include all 3 wall layers
- Fusiform aneurysms involve the entire circumference of the artery
- Saccular aneurysms (aka berry aneurysms) are spherical in shape and involve only a portion of the wall



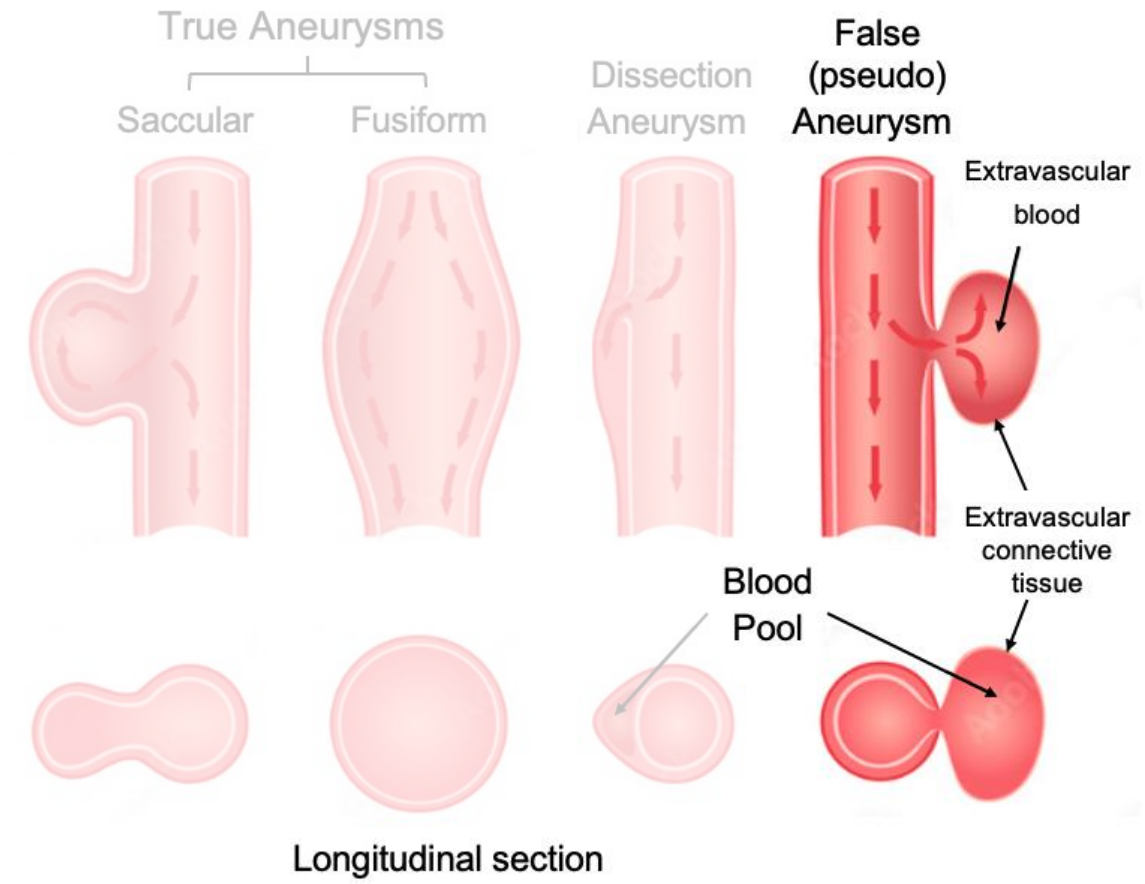
Types of Aneurysms

- Dissection aneurysm develop after the occurrence of an intimal tear and formation of a false lumen



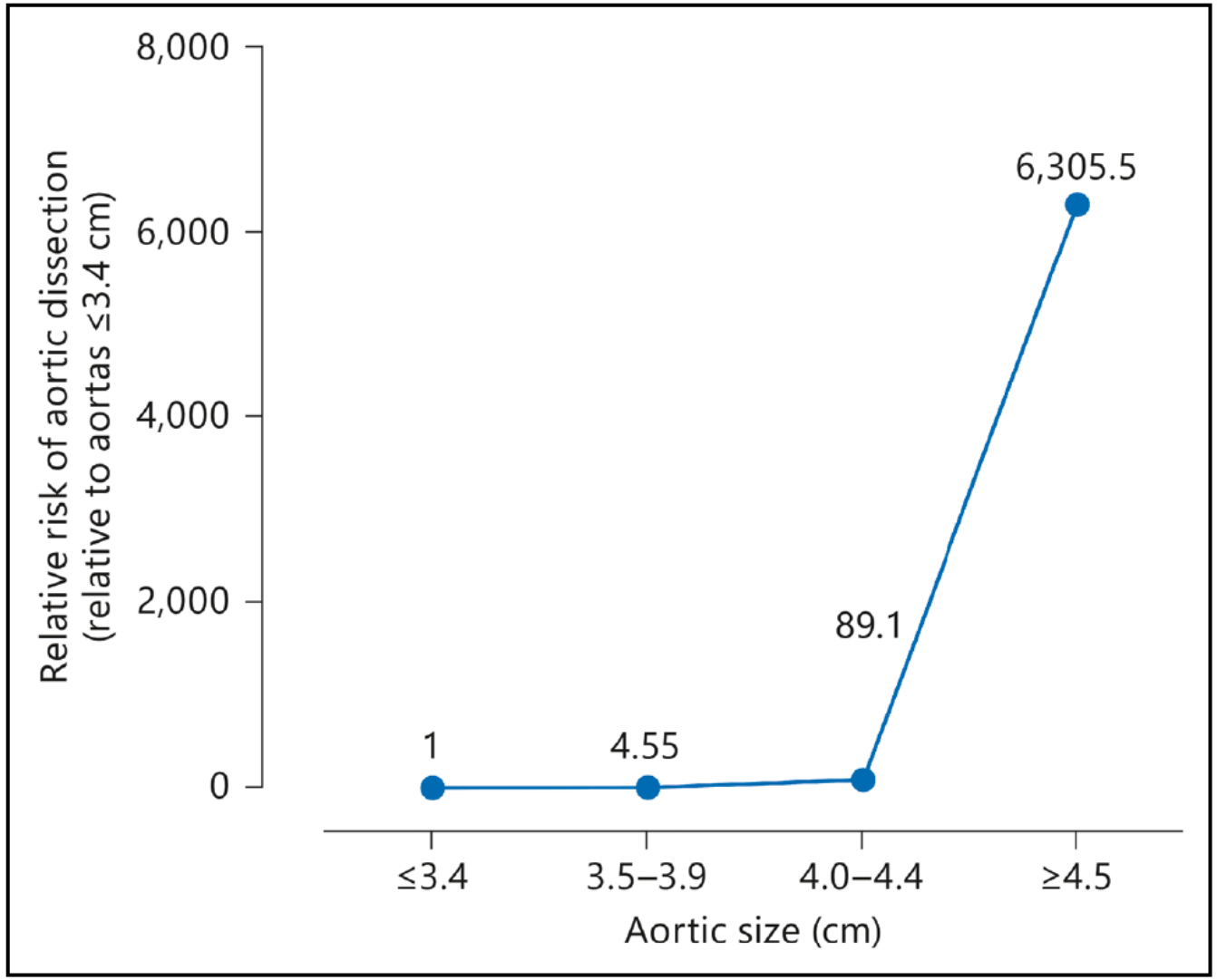
Types of Aneurysms

- False (pseudo) aneurysms are extravasations of blood from a vessel wall perforation that is contained by surrounding tissue
- Therefore, they are not contained within the arterial wall



Ascending Aorta Size and Risk of Dissection

- The natural history of aortic aneurysms is of a progressive dilatation over time. This is associated with an increased risk of aortic dissection and, eventually, rupture
- The relative risk of aortic dissection begins to increase appreciably at an ascending aortic diameter of 4.0 cm to 4.4 cm, and then increases dramatically at a diameter of ≥4.5 cm



(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Modalities for Aortic Imaging

Table 5. Diagnostic Performance of Aortic Imaging Modalities

Parameter	CT	MRI	TTE	TEE	US
Availability	+++	++	+++	++	+++
Portability	-	-	+++	+++	+++
Speed of acquisition	+++	+	++	++	++
Spatial resolution	+++	++	++	+++	++
Temporal resolution	+	++	+++	+++	+++
Three-dimensional data set	+++	++	+	+	+
Arch branch vessel evaluation	+++	+++	++	+	NA
Evaluation of valve and ventricular function	+	++	+++	+++	NA

CT indicates computed tomography; MRI, magnetic resonance imaging; NA, not applicable; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography; US, abdominal aortic ultrasound; +++ excellent results; ++ good results; + fair results; and -, not available.

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Thoracic Aortic Aneurysms

Thoracic Aortic Aneurysms (TAA) Epidemiology

- The true prevalence of TAA is unknown, as most are asymptomatic. However, TAA have been reported to occur in 5 to 10 per 100,000 person years
- Unfortunately, TAA's first presentation is often a life threatening acute aortic syndrome: dissection, intramural hematoma, impending rupture, or rupture

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Causes and risk factors for TAA

- Hypertension, hypercholesterolemia, smoking, and heritable genetic variants are risk factors for TAA
- Causes of TAA include heritable disorders, congenital conditions, degenerative conditions, previous aortic dissection, inflammatory and infectious diseases
- Many aneurysms of the root and ascending thoracic aorta are sporadic and idiopathic
- Patients with TAA have a modestly increased incidence of AAA and cerebral aneurysms

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Heritable Thoracic Aortic Disease: Syndromic Marfan syndrome Loeys-Dietz syndrome Vascular Ehlers-Danlos syndrome Smooth muscle dysfunction syndrome Others: attributable to pathogenic variants in FLNA, BGN, LOX
Heritable Thoracic Aortic Disease: Non-syndromic ACTA2, MYH11, PRKG1, MYLK, and others Familial thoracic aortic aneurysm without identified pathogenic variants in a known gene for HTAD
Congenital conditions Bicuspid aortic valve Turner syndrome Coarctation of the aorta Complex congenital heart defects (tetralogy of Fallot, transposition of the great vessels, truncus arteriosus)
Hypertension
Atherosclerosis
Degenerative
Previous aortic dissection
Inflammatory Aortitis Giant cell arteritis Takayasu arteritis Behçet disease Immunoglobulin G4-related disease, antineutrophil cytoplasmic antibody-related, sarcoidosis
Infectious Aortitis Bacterial, fungal, syphilitic
Previous traumatic aortic injury

Medical Management of TAA

Sporadic Aneurysms

Recommendations for BP Management in TAA
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	1. In patients with TAA and an average systolic BP (SBP) of ≥130 mm Hg or an average diastolic BP (DBP) of ≥80 mm Hg, the use of antihypertensive medications is recommended to reduce risk of cardiovascular events.[1-3]
2a	C-LD	2. In patients with TAA, regardless of cause and in the absence of contraindications, use of beta blockers to achieve target BP goals is reasonable.[1,4,5]
2a	C-EO	3. In patients with TAA, regardless of etiology and in the absence of contraindications, ARB therapy is a reasonable adjunct to beta-blocker therapy to achieve target BP goals.[6]

Recommendations for Treatment of TAA With Statins

COR	LOE	Recommendations
2a	C-LD	1. In patients with TAA and imaging or clinical evidence of atherosclerosis, statin therapy at moderate or high intensity is reasonable.[1,2]
2b	C-LD	2. In patients with TAA who have no evidence of atherosclerosis, the use of statin therapy may be considered.[3-6]

Recommendation for Smoking Cessation in TAA

COR	LOE	Recommendation
1	C-LD	1. In patients with TAA who smoke cigarettes, smoking cessation efforts are recommended.[1,2]

Recommendation for Antiplatelet Therapy in TAA

COR	LOE	Recommendation
2a	C-EO	1. In patients with atherosclerotic TAA and concomitant aortic atheroma or PAU, the use of low-dose aspirin is reasonable, unless contraindicated.[1,2]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Medical Management of TAA

Marfan Syndrome

COR	LOE	Recommendations
1	A	1. In patients with Marfan syndrome, treatment with either a beta blocker or an ARB, in maximally tolerated doses (unless contraindicated), is recommended to reduce the rate of aortic dilation.[1,2]
2a	C-LD	2. In patients with Marfan syndrome, the use of both a beta blocker and an ARB, in maximally tolerated doses (unless contraindicated), is reasonable to reduce the rate of aortic dilation.[3,4]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

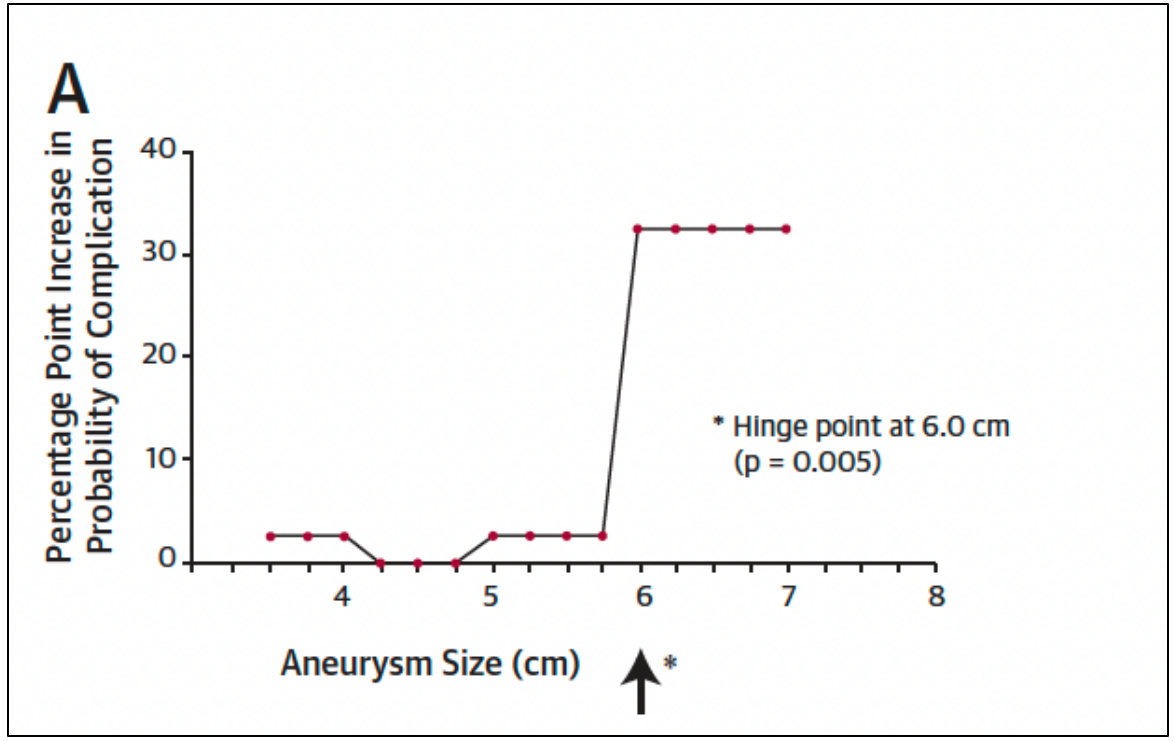
Surveillance of Thoracic Aortic Dilatation and Aneurysm

COR	LOE	Recommendations
1	C-LD	1. In patients with a dilated thoracic aorta, a TTE is recommended at the time of diagnosis to assess aortic valve anatomy, aortic valve function, and thoracic aortic diameters.[1-4]
2a	C-LD	2. In patients with a dilated thoracic aorta, a CT or MRI at the time of diagnosis is reasonable to assess thoracic aortic anatomy and diameters.[1,3,5-7]
2a	C-LD	3. In patients with a dilated thoracic aorta, follow-up imaging (with TTE, CT, or MRI, as appropriate based on individual anatomy) in 6 to 12 months is reasonable to determine the rate of aortic enlargement; if stable, surveillance imaging every 6 to 24 months (depending on aortic diameter) is reasonable.[1,3,4]

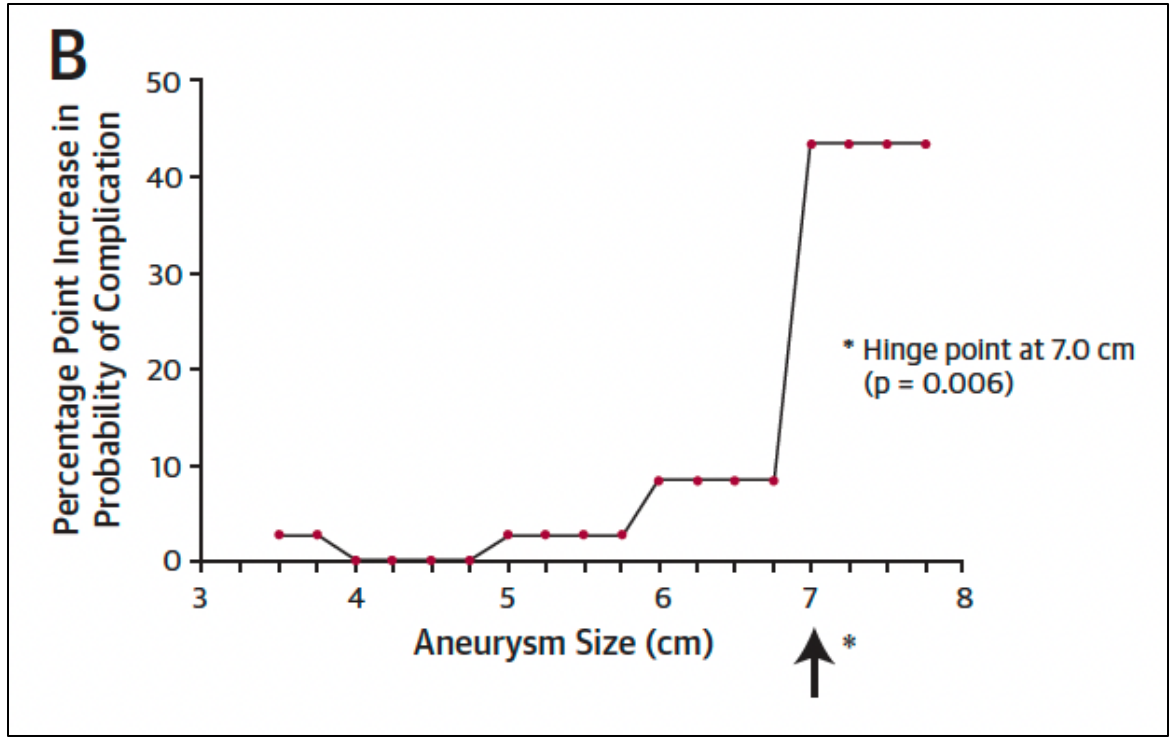
(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

TAA Size and Risk of Complications

ASCENDING AORTIC ANEURYSM



DESCENDING AORTIC ANEURYSM



(Goldfinger JZ et al. JACC 2014;64(16):1725-39)

Ascending TAA Repair Threshold

Sporadic Aneurysms

COR	LOE	Recommendations
1	C-LD	1. In patients with aneurysms of the aortic root and ascending aorta who have symptoms attributable to the aneurysm, surgery is indicated.[1,2]
1	B-NR	2. In asymptomatic patients with aneurysms of the aortic root or ascending aorta who have a maximum diameter of ≥5.5 cm, surgery is indicated.[3-9]
1	C-LD	3. In patients with an aneurysm of the aortic root or ascending aorta of <5.5 cm, whose growth rate confirmed by tomographic imaging is ≥0.3 cm/y in 2 consecutive years, or ≥0.5 cm in 1 year, surgery is indicated.[10-13]
2a	B-NR	4. In asymptomatic patients with aneurysms of the aortic root or ascending aorta who have a maximum diameter of ≥5.0 cm, surgery is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[14-17]
2a	B-NR	5. In patients undergoing repair or replacement of a tricuspid aortic valve who have a concomitant aneurysm of the ascending aorta with a maximum diameter of ≥4.5 cm, ascending aortic replacement is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[18-21]
2a	B-NR	In patients undergoing repair or replacement of a tricuspid aortic valve who have a concomitant aneurysm of the ascending aorta with a maximum diameter of ≥5.0 cm, ascending aortic replacement is reasonable.[18-21]
2b	C-LD	In patients undergoing cardiac surgery for indications other than aortic valve repair or replacement who have a concomitant aneurysm of ascending aorta with a maximum diameter of ≥5.0 cm, ascending aortic replacement may be reasonable.[18]
2a	C-LD	6. In patients with a height >1 standard deviation above or below the mean who have an asymptomatic aneurysm of the aortic root or ascending aorta and a maximal cross-sectional aortic area/height ratio of ≥10 cm^2/m, surgery is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[14,15,22]
2b	C-LD	7. In asymptomatic patients with aneurysms of the aortic root or ascending aorta who have either an ASI of ≥3.08 cm/m^2 or AHI of ≥3.21 cm/m, surgery may be reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[23]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Ascending TAA Repair Threshold

Marfan Syndrome

COR	LOE	Recommendations
1	B-NR	1. In patients with Marfan syndrome and an aortic root diameter of ≥5.0 cm, surgery to replace the aortic root and ascending aorta is recommended.[1-4]
2a	B-NR	2. In patients with Marfan syndrome, an aortic root diameter of ≥4.5 cm, and features associated with an increased risk of aortic dissection (see Table 10), surgery to replace the aortic root and ascending aorta is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[1,3,4]
2a	C-LD	3. In patients with Marfan syndrome and a maximal cross-sectional aortic root area (cm2) to patient height (m) ratio of ≥10, surgery to replace the aortic root and ascending aorta is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[5]
2b	C-LD	4. In patients with Marfan syndrome and an aortic diameter approaching surgical threshold, who are candidates for valve-sparing root replacement (VSRR) and have a very low surgical risk, surgery to replace the aortic root and ascending aorta may be reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[2-4]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

BAV-Associated Aneurysms

COR	LOE	Recommendations
1	B-NR	1. In patients with a BAV and a diameter of the aortic root, ascending aorta, or both of ≥5.5 cm, surgery to replace the aortic root, ascending aorta, or both is recommended.[1-3]
2a	B-NR	2. In patients with a BAV and a cross-sectional aortic root or ascending aortic area (cm^2) to height (m) ratio of ≥10 cm^2/m, surgery to replace the aortic root, ascending aorta, or both is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[3,4]
2a	B-NR	3. In patients with a BAV, a diameter of the aortic root or ascending aorta of 5.0 cm to 5.4 cm, and an additional risk factor for aortic dissection (Table 14), surgery to replace the aortic root, ascending aorta, or both is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[1,5]
2a	B-NR	4. In patients with a BAV who are undergoing surgical aortic valve repair or replacement, and who have a diameter of the aortic root or ascending aorta of ≥4.5 cm, concomitant replacement of the aortic root, ascending aorta, or both is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[1,6]
2b	B-NR	5. In patients with a BAV, a diameter of the aortic root or ascending aorta of 5.0 cm to 5.4 cm, no other risk factors for aortic dissection (Table 14), and at low surgical risk, surgery to replace the aortic root, ascending aorta, or both may be reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[1,2,5]

Abdominal Aortic Aneurysms

Abdominal Aortic Aneurysms Epidemiology

- Abdominal aortic aneurysm (AAA) is defined as an abdominal aortic diameter >3.0 cm
- In the US, the estimated prevalence of AAA is 1.4% among people between 50 and 84 years of age, or 1.1 million adults
- Strong risk factors for AAA include older age, male sex, and smoking
- Family history (1st degree), hypertension, and hypercholesterolemia, represent additional risk factors

(Schanzer AS et al. N Engl J Med 2021;385:1690-8)

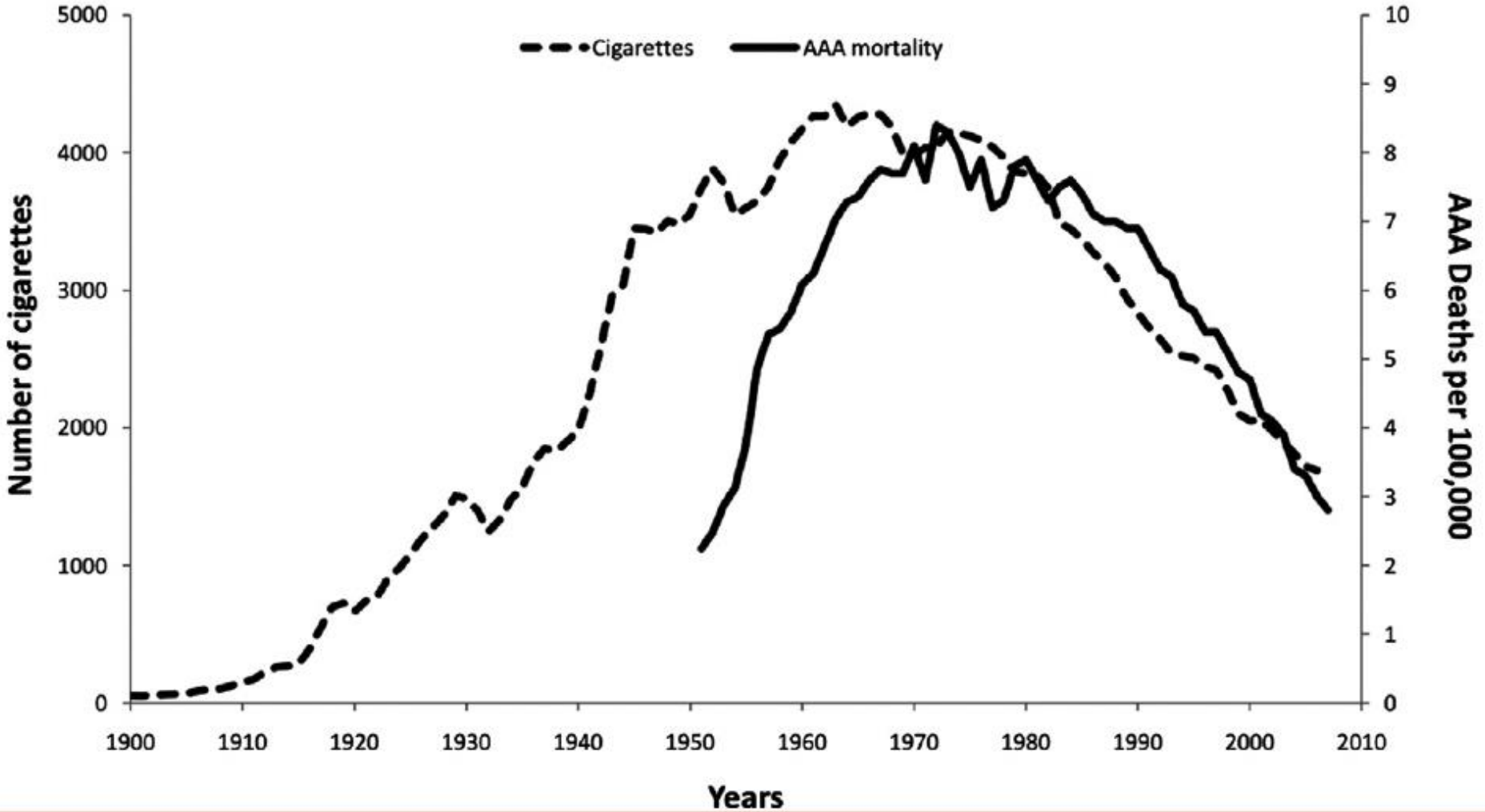


Fig 1. The annual adult per capita cigarette consumption and age-adjusted abdominal aortic aneurysm (*AAA*) deaths per 100,000 white men by year in the United States. (From Lederle FA. The rise and fall of abdominal aortic aneurysm. Circulation 2011;124:1097-9.).

(Chaikof EL et al. J Vasc Surg 2018;67(1):2-79

Abdominal Aortic Aneurysms Epidemiology

- The primary concern for AAA is the risk of rupture and death from hemorrhage
- The single most important predictor of rupture is the diameter of the aneurysm, with the risk increasing with larger aneurysms

(Schanzer AS et al. N Engl J Med 2021;385:1690-8)

Abdominal Aortic Aneurysms Screening

- Screening for AAA has led to a reduction in AAA-specific mortality in men
- Ultrasonography (US) of the abdomen is generally considered the gold standard for AAA diagnosis and monitoring in asymptomatic patients

COR	LOE	Recommendations
1	B-R	1. In men who are ≥65 years of age who have ever smoked, ultrasound screening for detection of AAA is recommended.[1]
1	C-LD	2. In men or women who are ≥65 years of age and who are first-degree relatives of patients with AAA, ultrasound screening for detection of AAA is recommended.[2,3]
2a	C-EO	3. In women who are ≥65 years of age who have ever smoked, ultrasound screening for detection of AAA is reasonable.[4,5]
2b	C-LD	4. In men or women <65 years of age and who have multiple risk factors (Table 15) or a first-degree relative with AAA, ultrasound screening for AAA may be considered.[5,6]
3: No Benefit	B-NR	5. In asymptomatic men or women >75 years who have had a negative initial ultrasound screen, repeat screening for detection of AAA is not recommended.[1]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

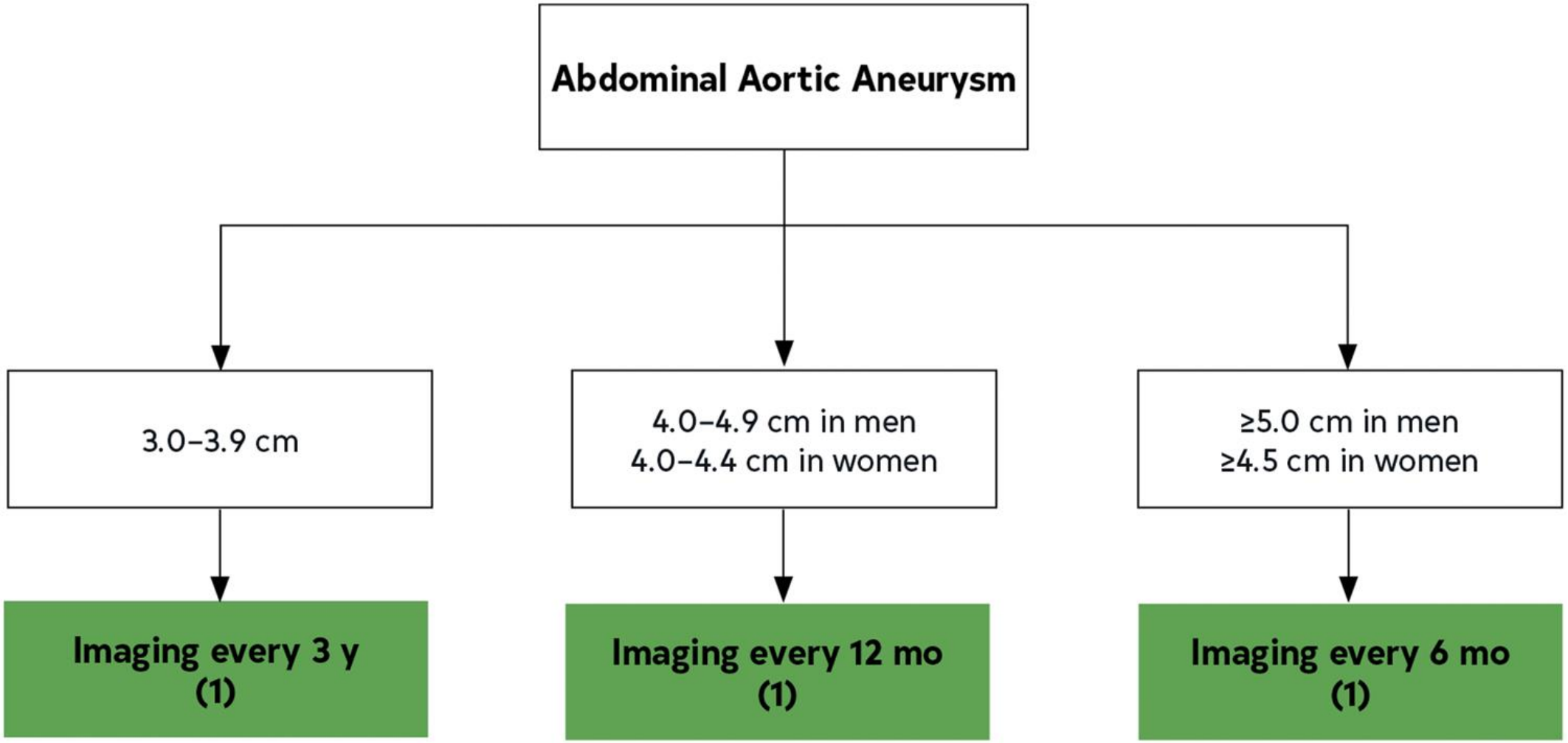
Abdominal Aortic Aneurysms Surveillance

- Multiple studies have established that US surveillance of AAA helps to prevent rupture and mortality
- The frequency of surveillance US depends on AAA size and the risk of rupture: the larger the size the shorter is the interval
- CT (or MRI) angiography should be used as an alternative when US imaging is not adequate or preoperatively

1	B-NR	1. In patients with an AAA of 3.0 cm to 3.9 cm, surveillance ultrasound is recommended every 3 years to assess for interval change.[1-8]
1	B-NR	2. In men with an AAA of 4.0 cm to 4.9 cm and in women with an AAA of 4.0 cm to 4.4 cm, surveillance ultrasound is recommended annually to assess for interval change.[1-8]
1	B-NR	3. In men with an AAA of ≥5.0 cm and women with an AAA of ≥4.5 cm, surveillance ultrasound is recommended every 6 months to assess for interval change.[1-8]
1	C-EO	4. In patients with an AAA that is inadequately defined with ultrasound, surveillance CT is recommended. In such patients, when there is a contraindication to CT or to lower cumulative radiation risk, surveillance MRI is reasonable.[9,10]
2a	C-LD	
1	C-EO	5. In patients with an AAA that meets criteria for repair, CT is recommended for preoperative planning.

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Abdominal Aortic Aneurysms Surveillance



(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Medical Management of AAA

Recommendation for BP Management in AAA
Referenced studies that support the recommendation are summarized in the Online Data Supplement.

COR	LOE	Recommendation
1	B-NR	1. In patients with AAA and an average SBP of ≥130 mm Hg, or an average DBP of ≥80 mm Hg, the use of antihypertensive medication is recommended to reduce risk of cardiovascular events.[1-3]

Recommendations for Treatment of AAA With Statins
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	1. In patients with AAA and evidence of aortic atherosclerosis, statin therapy at moderate or high intensity is recommended.[1-3]
2b	C-LD	2. In patients with AAA but no evidence of atherosclerosis, statin therapy may be considered.[4,5]

Recommendation for Smoking Cessation in AAA

COR	LOE	Recommendation
1	C-LD	1. In patients with AAA who smoke cigarettes, smoking cessation efforts are recommended.[1-4]

Recommendation for Antithrombotic Therapy in AAA

COR	LOE	Recommendation
2b	C-LD	1. In patients with AAA with concomitant atheroma and/or PAU, the use of low-dose aspirin may be considered, unless contraindicated.[1]

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Aortitis

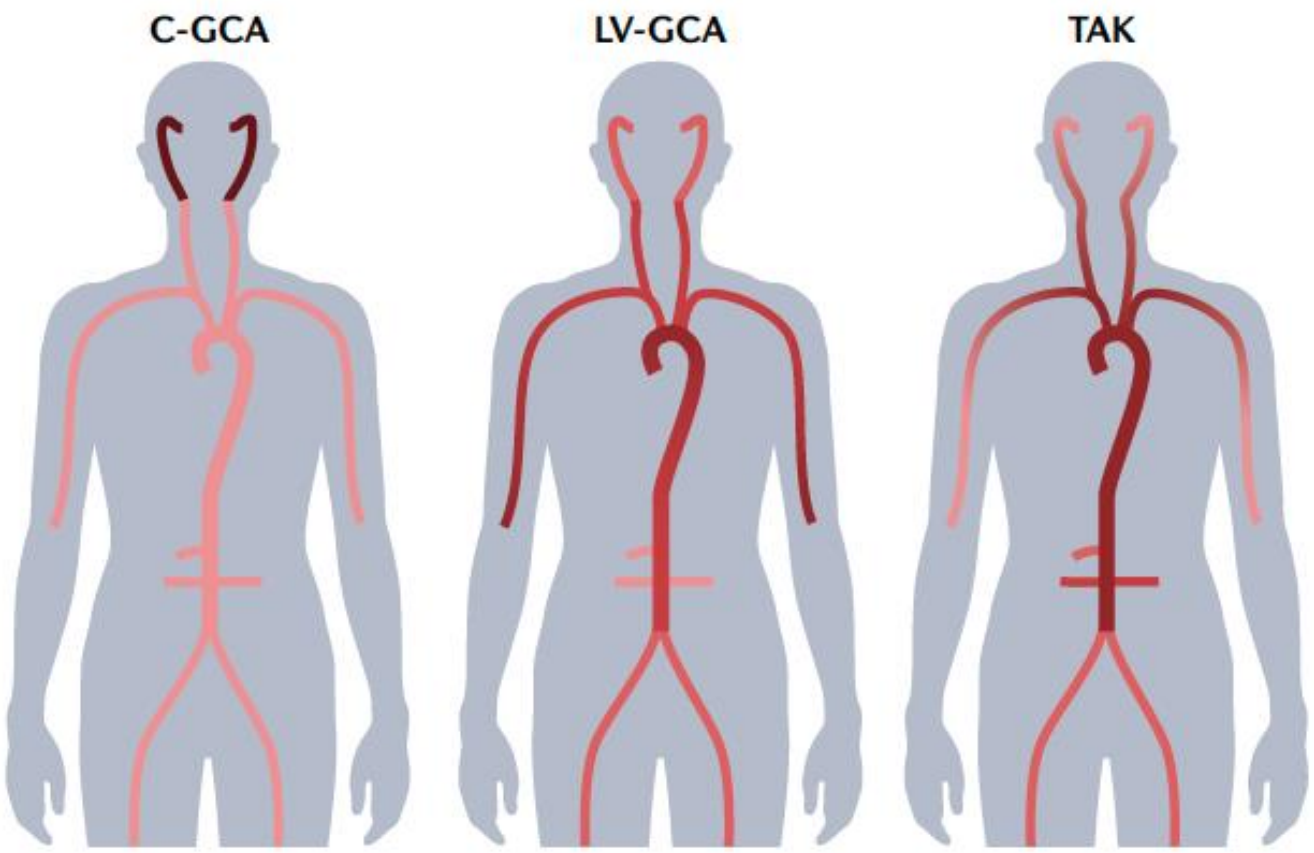
Aortitis Classification

- Aortitis refers to inflammatory or infectious processes of the aorta and its main branches
- Inflammatory aortitis include:
  - Large vessel vasculitis (Takayasu arteritis and giant cell arteritis)
  - IgG4-related disease, ANCA-related vasculitis, sarcoidosis, Behçet's disease, relapsing polycondritis, and granulomatosis with polyangiitis
- Infectious aortitis
  - Bacterial
  - Syphilitic
  - Fungal
  - Tuberculous

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Large Vessel Vasculitis Definition

- Large vessel vasculitis (LVV) include Takayasu arteritis (TAK) and giant cell arteritis (GCA)
- Both TAK and GCA are defined by granulomatous inflammation of the arterial wall
- Arteritis of the temporal artery and systemic symptoms of polymyalgia rheumatica define the cranial variant of GCA



(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

Large Vessel Vasculitis Presentation

Box 3 | **Clinical features of giant cell arteritis and Takayasu arteritis**

Systemic symptoms

- Anorexia
- Arthralgia
- Fatigue
- Lethargy
- Low-grade fever
- Myalgia
- Sweats
- Weight loss

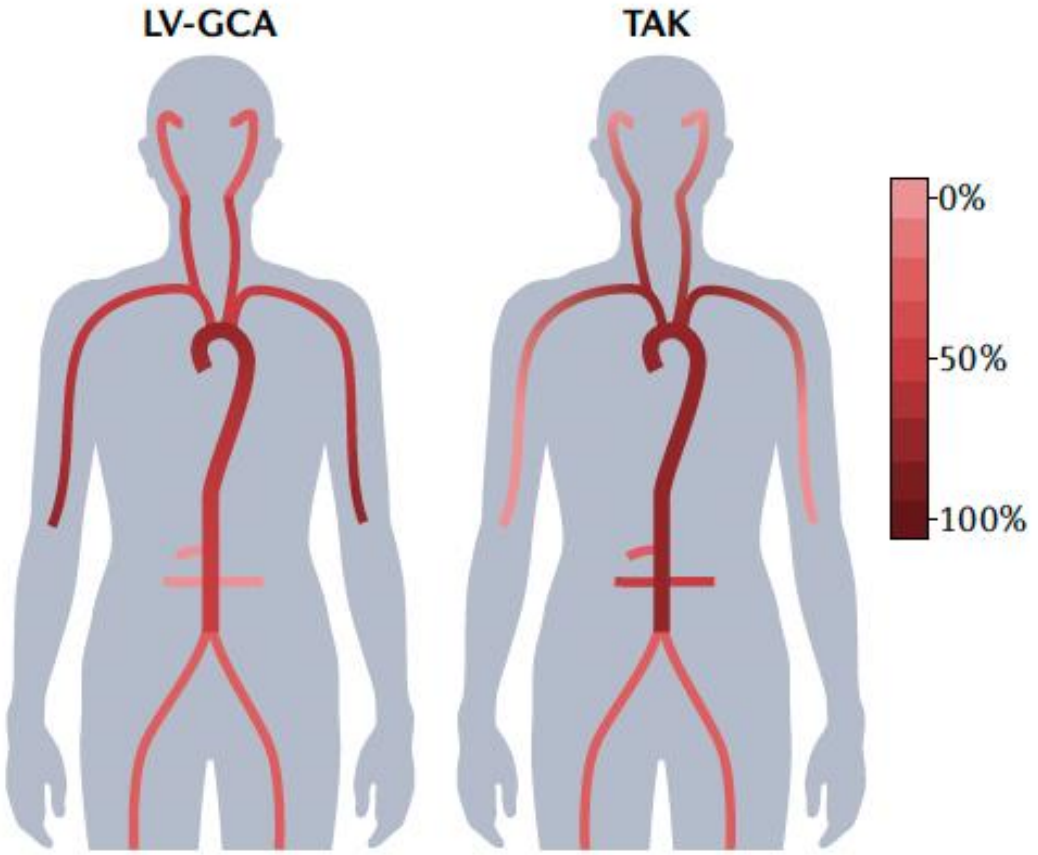
Symptoms of tissue/organ ischaemia

- Abdominal pain[a]
- Chest pain[a]
- Cough[b]
- Dyspnoea[a]
- Headache[b]
- Jaw claudication[b]
- Lightheadedness[a]
- Limb claudication[a]
- Neck pain[b]
- Neurological deficit
- Scalp tenderness[b]
- Tongue claudication[b]
- Vision disturbance[b]

Examination findings

- Aortic regurgitation[a]
- Carotidynia[a]
- Discrepancy between right and left arm blood pressure
- Hypertension[a]
- Ophthalmic abnormalities[b]
- Reduced or absent pulses[a]
- Scalp tenderness[b]
- Tender and/or thickened temporal arteries[b]
- Vascular bruits

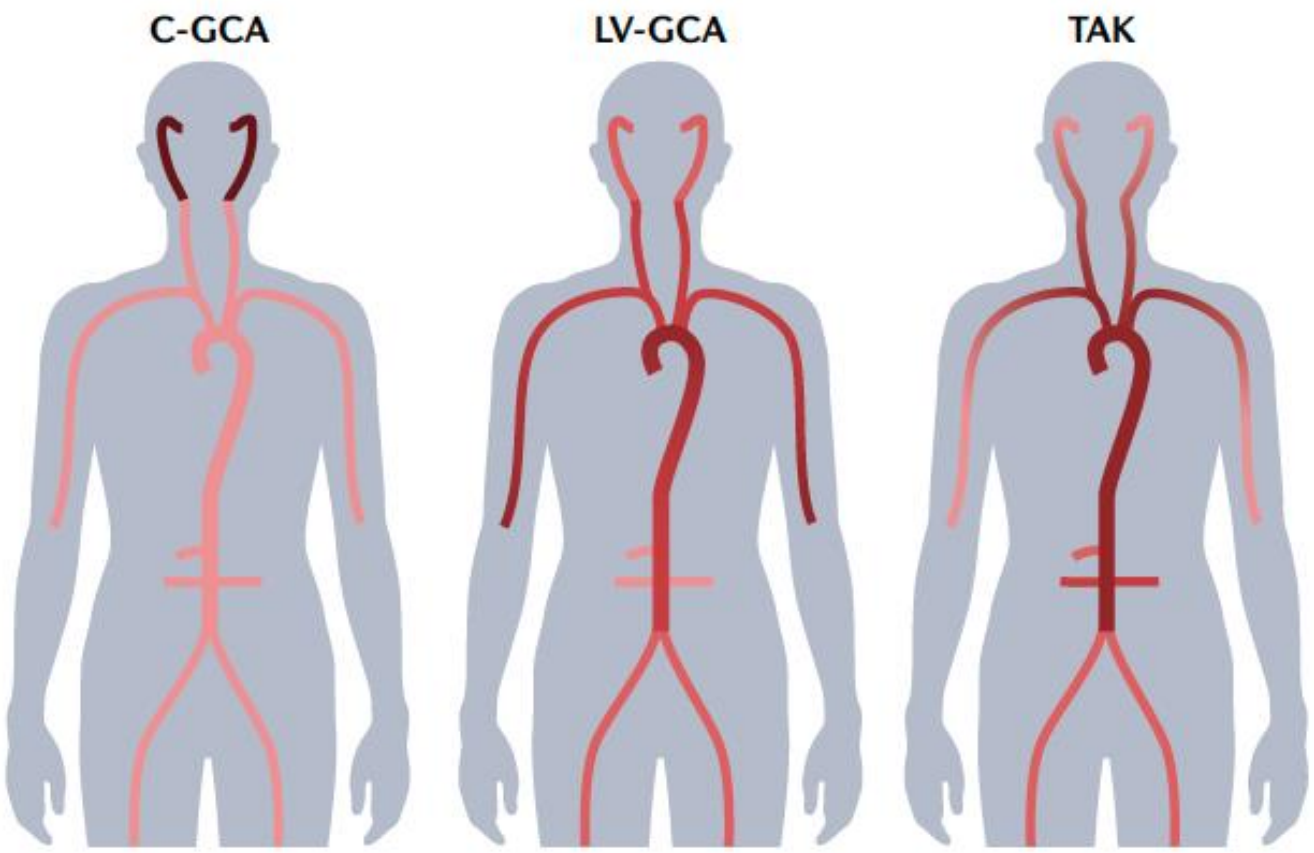
[a]More prevalent in Takayasu arteritis.
[b]More prevalent in giant cell arteritis[286].



(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

Large Vessel Vasculitis Diagnosis

- Large vessel vasculitis (LVV) include Takayasu arteritis (TAK) and giant cell arteritis (GCA)
- Both TAK and GCA are defined by granulomatous inflammation of the arterial wall
- Arteritis of the temporal artery and systemic symptoms of polymyalgia rheumatica define the cranial variant of GCA



(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

Large Vessel Vasculitis Diagnosis

- No validated diagnostic criteria exist for GCA and TAK
- Historically, diagnosis of GCA was based on a constellation of symptoms, ideally with histological confirmation of vasculitis
- Multimodality vascular imaging has equivalent diagnosis accuracy as temporal artery biopsy

(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

Table 35. Diagnostic Criteria for Inflammatory Aortitis

Names	Criteria Used in Diagnosis
Takayasu arteritis	Age of onset <40 y Intermittent claudication
	Diminished brachial artery pulse
	Subclavian artery or aortic bruit
	Systolic BP variation of >10 mm Hg between arms
	Aortographic evidence of aorta or aortic branch stenosis
Giant cell arteritis	Age >50 y Recent-onset localized headache
	Temporal artery tenderness or pulse attenuation
	Elevated erythrocyte sedimentation rate >50 mm/h
	Arterial biopsy shows necrotizing vasculitis
Diagnosis is established when ≥3 criteria are present	

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Large Vessel Vasculitis Imaging

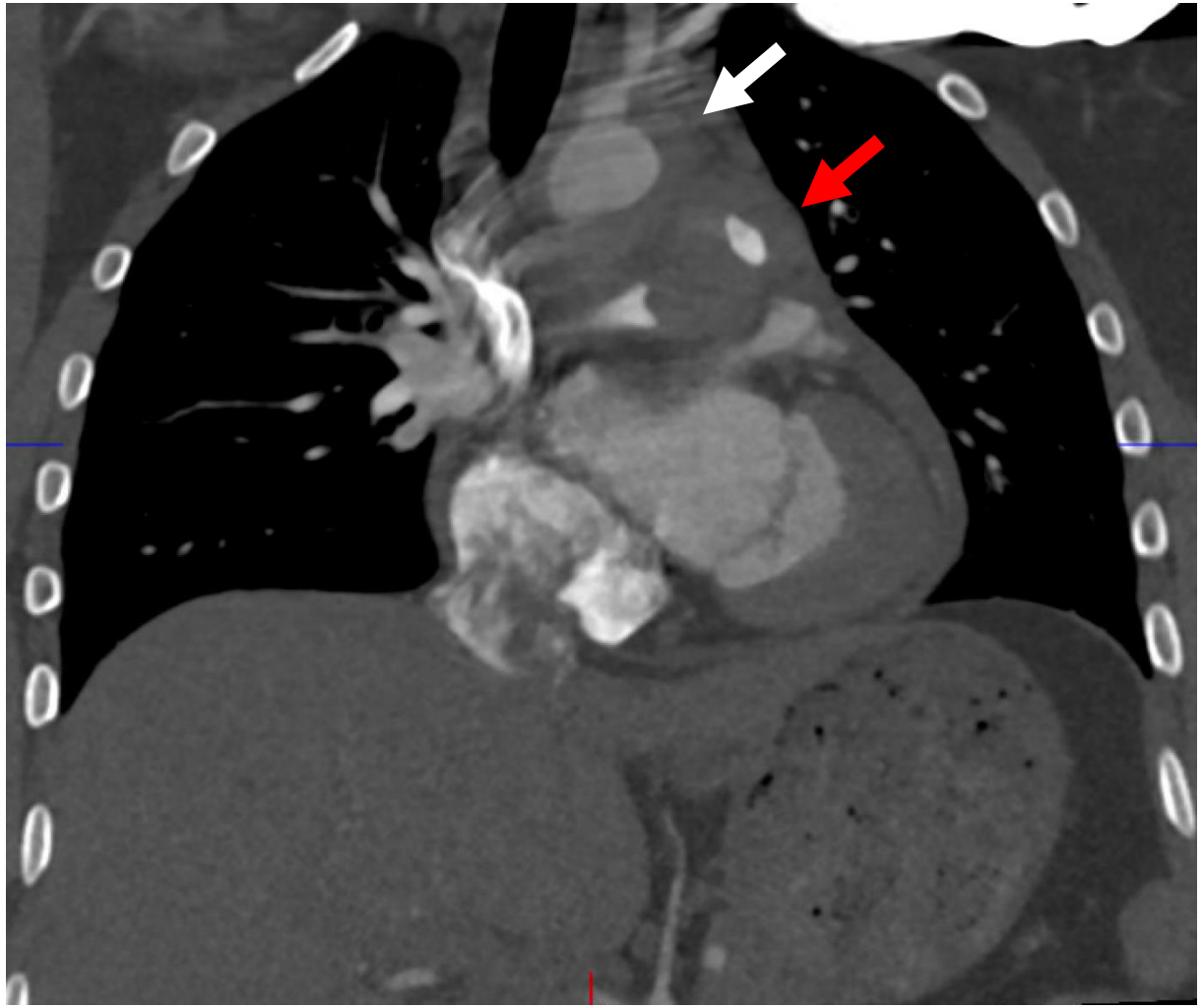
- Multiple imaging modalities are available to assess the extent and severity of LVV, including US, MRI, CT and 18F- fluorodeoxyglucose (FDG)- PET
- Each modality has advantages and disadvantages, and use is typically guided by the clinical scenario and local expertise
- Imaging of the aorta and major branches should be considered in all patients, even in those with a primarily cranial presentation
- Diagnostic accuracy of imaging declines quickly following treatment with glucocorticoids and imaging is best performed within 1 week of starting therapy

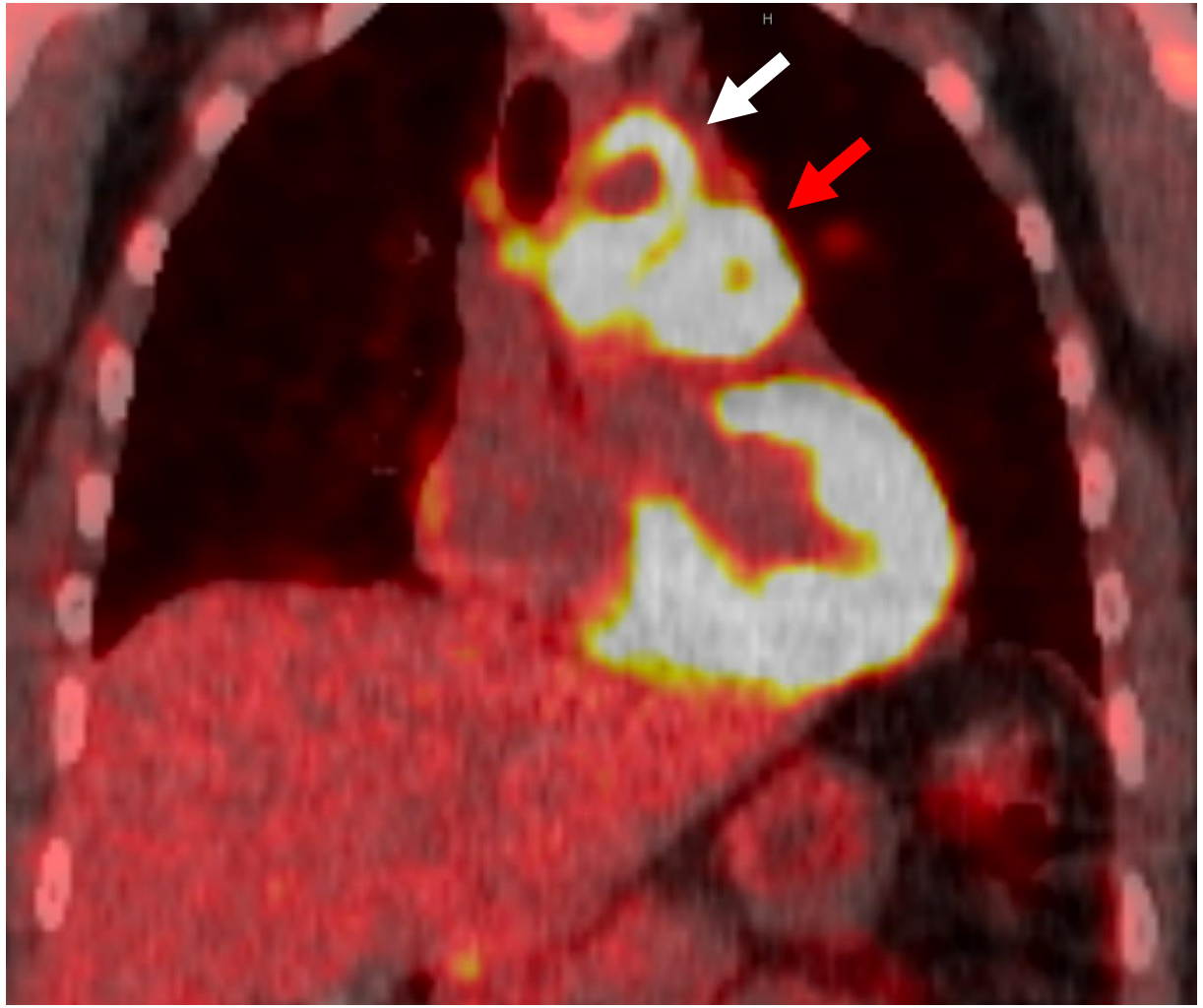
COR	LOE	Recommendations
Diagnosis		
1	C-LD	1. In patients with large vessel vasculitis (LVV), prompt evaluation of the entire aorta and branch vessels with MRI or CT, with or without 18F-FDG positron emission tomography (FDG-PET), is recommended.[1-6]

(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

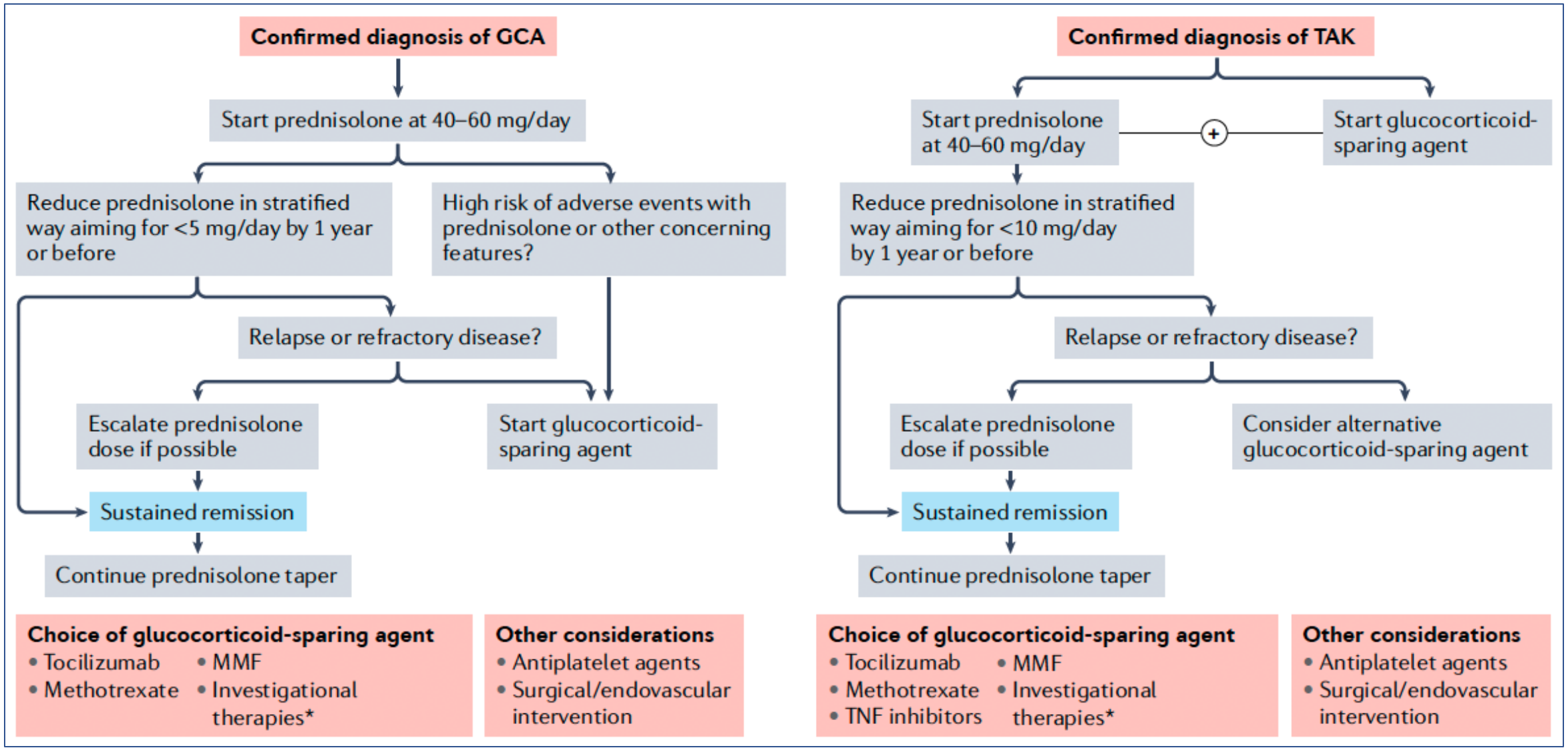
(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

Large Vessel Vasculitis Imaging





Large Vessel Vasculitis Treatment



(Pugh D et al. Nat Reviews Dis Primer 2021;7:93)

Infectious Aortitis

- The term “infectious aortitis” refers to an infection of the aorta and has supplanted the older term “mycotic aneurysm,” which was used broadly but actually implies a fungal cause
- Aortic infections arise from either contiguous spread from adjacent structures or septic emboli and hematogenous spread to the aortic wall via a vulnerable plaque or preexisting aneurysm
- Staphylococcus aureus, Pneumococcus, Escherichia coli, and Salmonella are the pathogens identified in most reports
- Syphilitic aortitis, typically occurs 10 to 25 years after systemic Treponema pallidum infection, is now rare. Fungal aortitis (mainly Candida or Aspergillus) and tuberculous aortitis are uncommon and typically arise in immunocompromised hosts

(Isselbacher EM et al. Circulation. 2022;146:e334–e482)

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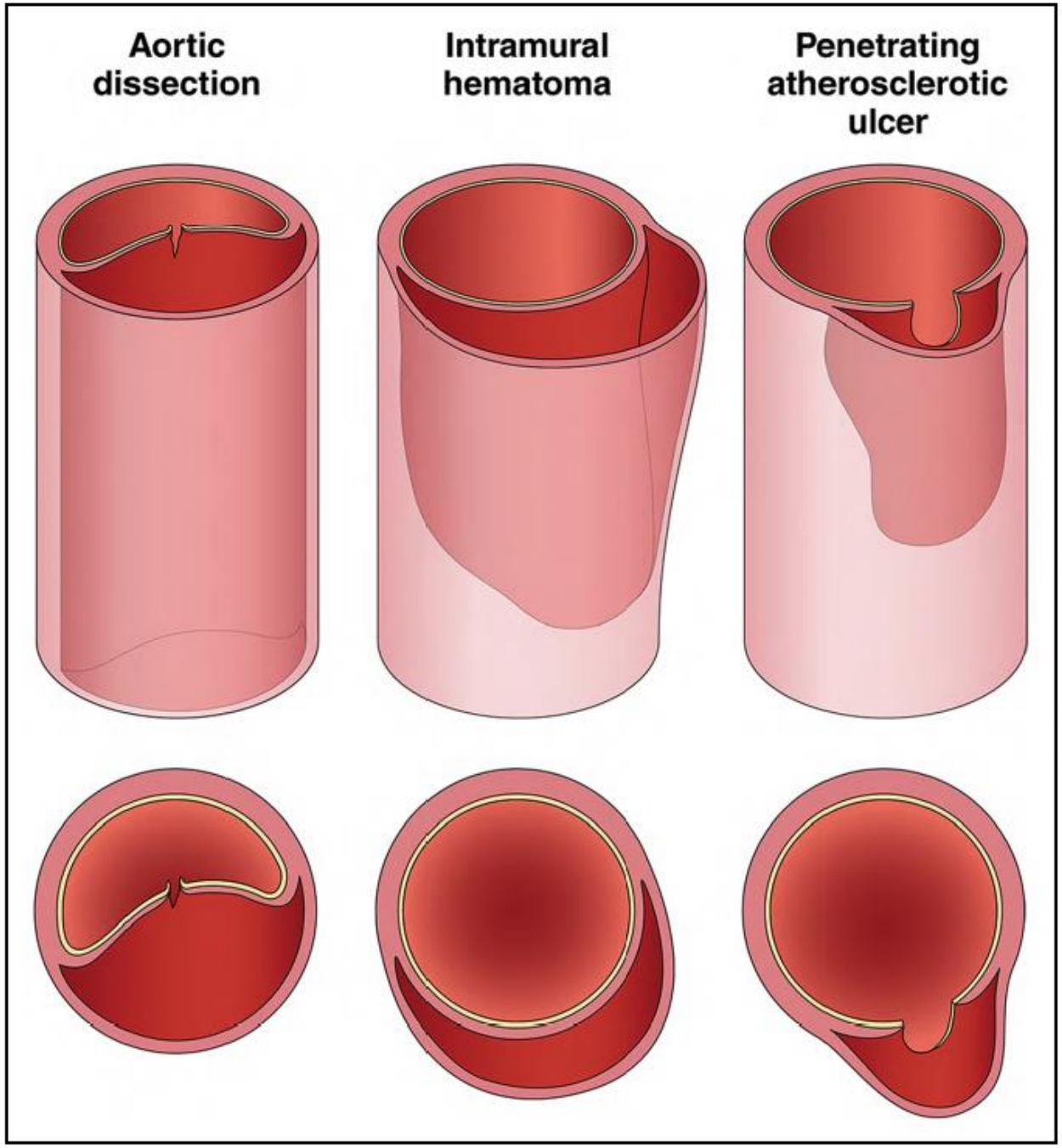
Acute Aortic Syndromes

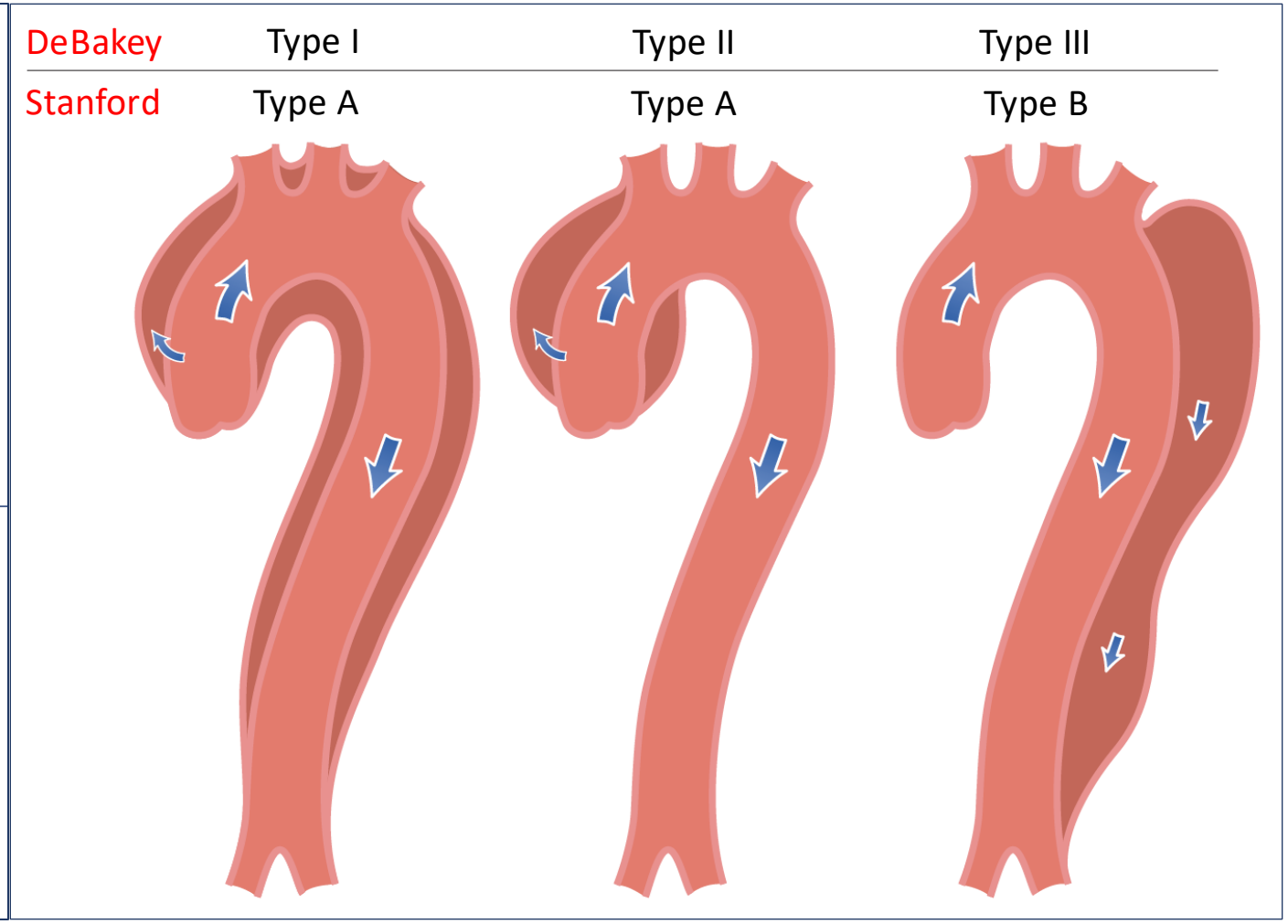
- ✓ Acute aortic syndromes (AAS) are life threatening conditions characterized by a breach in the integrity of the aortic wall
- ✓ The spectrum of AAS includes acute aortic dissection (AAD), intramural hematoma (IMH), penetrating atherosclerotic ulcer (PAU), and aortic rupture



(Isselbacher EM et al. *Circulation. 2022;146:e334–e482)*

Aortic Dissection Classification

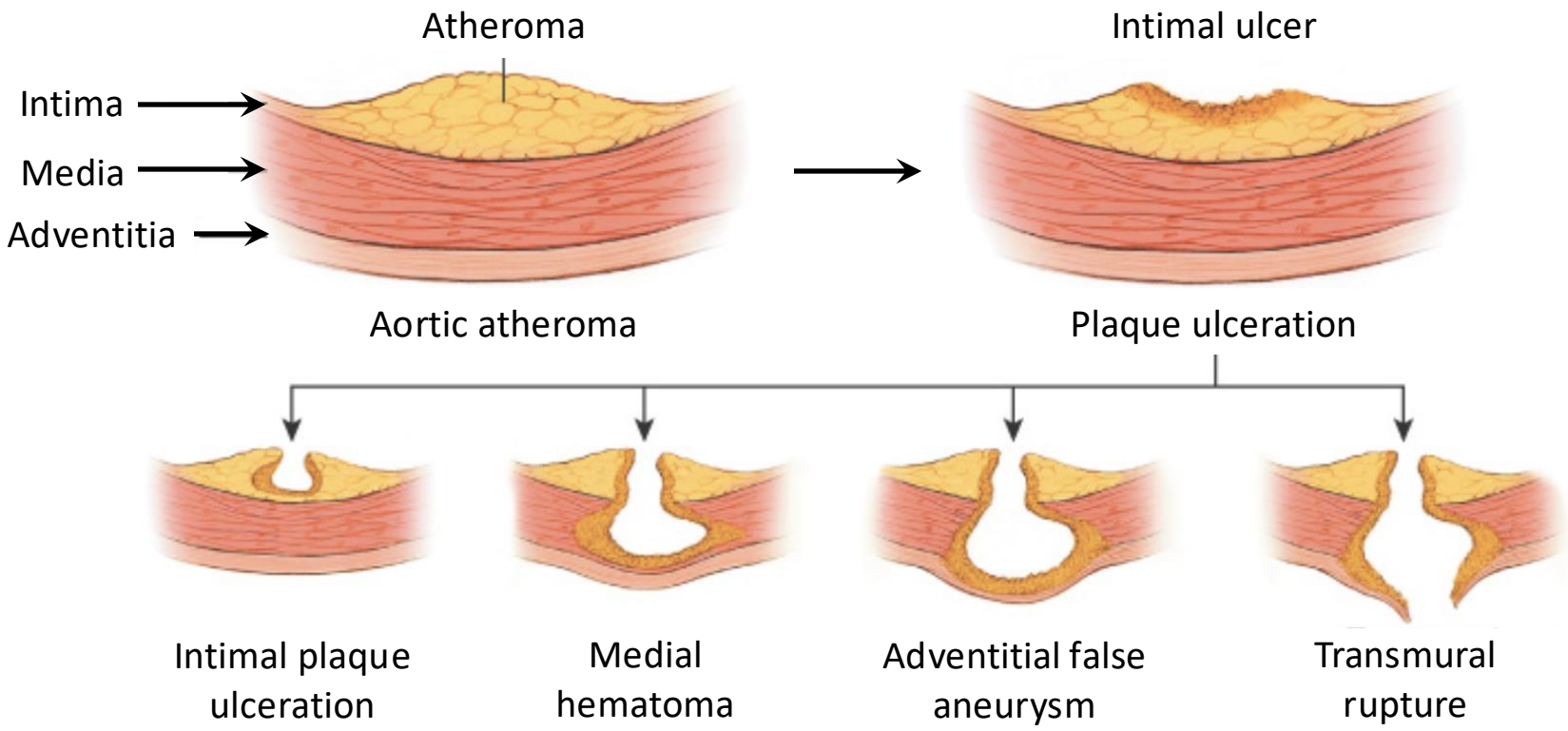
DeBakey	
Category I	Dissection tear in the ascending aorta propagating distally to include at least the aortic arch and typically the descending aorta
Category II	Dissection tear only in the ascending aorta
Category III	Dissection tear in the descending aorta propagating most often distally
Category IIIa	Dissection tear only in the descending thoracic aorta
Category IIIb	Tear extending below the diaphragm
Stanford	
Type A	All dissections involving the ascending aorta irrespective of the site of tear
Type B	All dissections that do not involve the ascending aorta; note that involvement of the aortic arch without involvement of the ascending aorta in the Stanford classification is labelled as Type B



(Modified from Erbel R et al. European Heart Journal 2014;35:2873-2926)

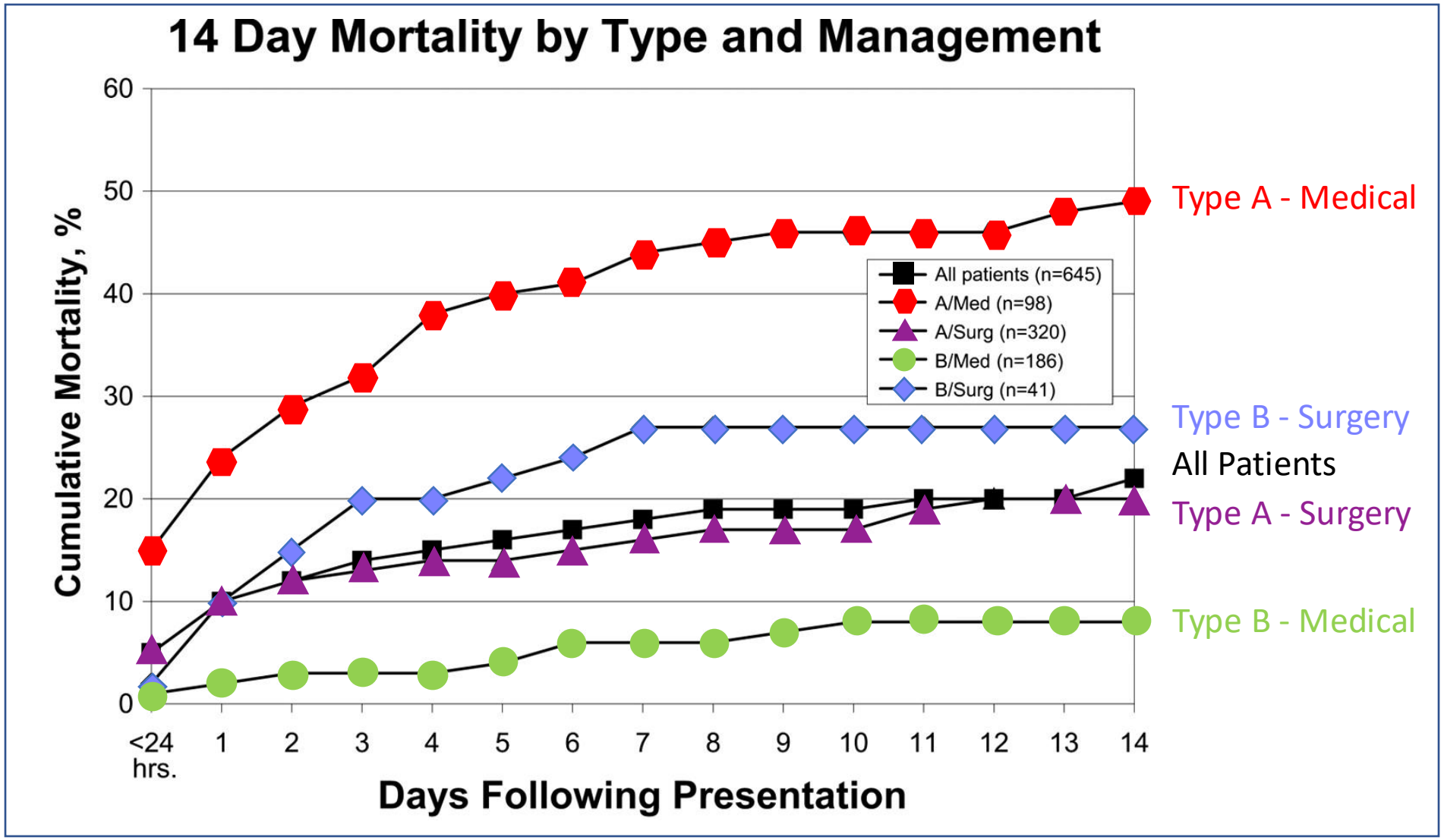
PENETRATING ATHEROSCLEROTIC ULCER

- ✓ Deep ulceration of atherosclerotic aortic plaques can present as an acute syndrome and lead to IMH, AAD or rupture



(Harris JA et al. J Vasc Surg 1994;19:90–98)

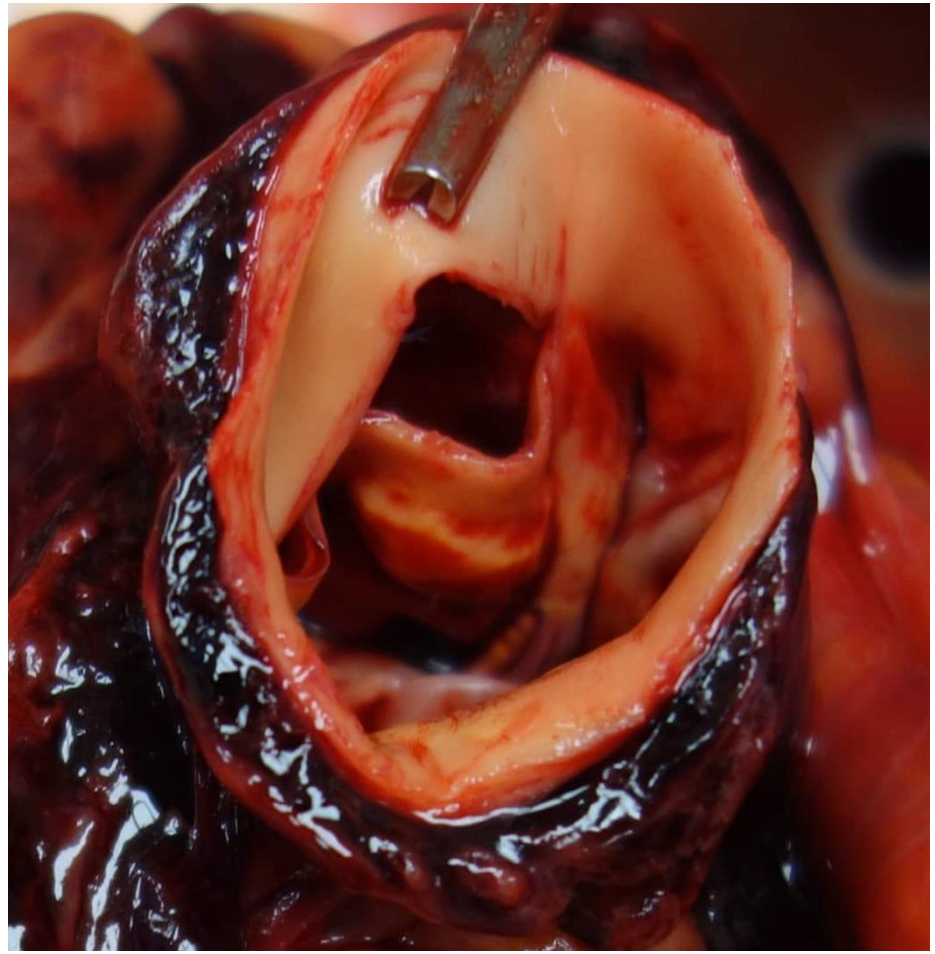
Aortic Dissection Mortality in IRAD



(Tsai TT et al. Circulation 2005;112:3802-3813)

Pathology of Acute Aortic Syndromes

- ✓ Disrupted aortic integrity is a fundamental component of the underlying pathology of AAS
- ✓ This can be due to an inherent instability of the aortic wall (e.g. inherited connective tissue disease) or an acquired condition, such as atherosclerotic degeneration, inflammation or infection



https://pikdo.net/p/anatomyandforensicpathology

(Nienaber CA et al. Nature Reviews Disease Primers 2016;2:16053)

Risk Factors for Acute Aortic Syndromes

Lifestyle factors

- Long-term arterial hypertension
- Smoking
- Dyslipidaemia
- Cocaine, crack cocaine or amphetamine use

Connective tissue disorders

- Marfan syndrome
- Loeys–Dietz syndrome
- Ehlers–Danlos syndrome
- Turner syndrome

Hereditary vascular disease

- Bicuspid aortic valve
- Coarctation of the aorta

Vascular inflammation

- Autoimmune disorders
  - Giant-cell arteritis
  - Takayasu arteritis
  - Beçhet disease
  - Ormond disease
- Infection
  - Syphilis
  - Tuberculosis

Deceleration trauma

- Car accident
- Fall from height

Iatrogenic factors

- Catheter or instrument intervention
- Valvular or aortic surgery
  - Side-clamping, cross-clamping or aortotomy
  - Graft anastomosis
  - Patch aortoplasty

(Nienaber CA et al. Eur Heart Journal 2012;33,1:26-25b)

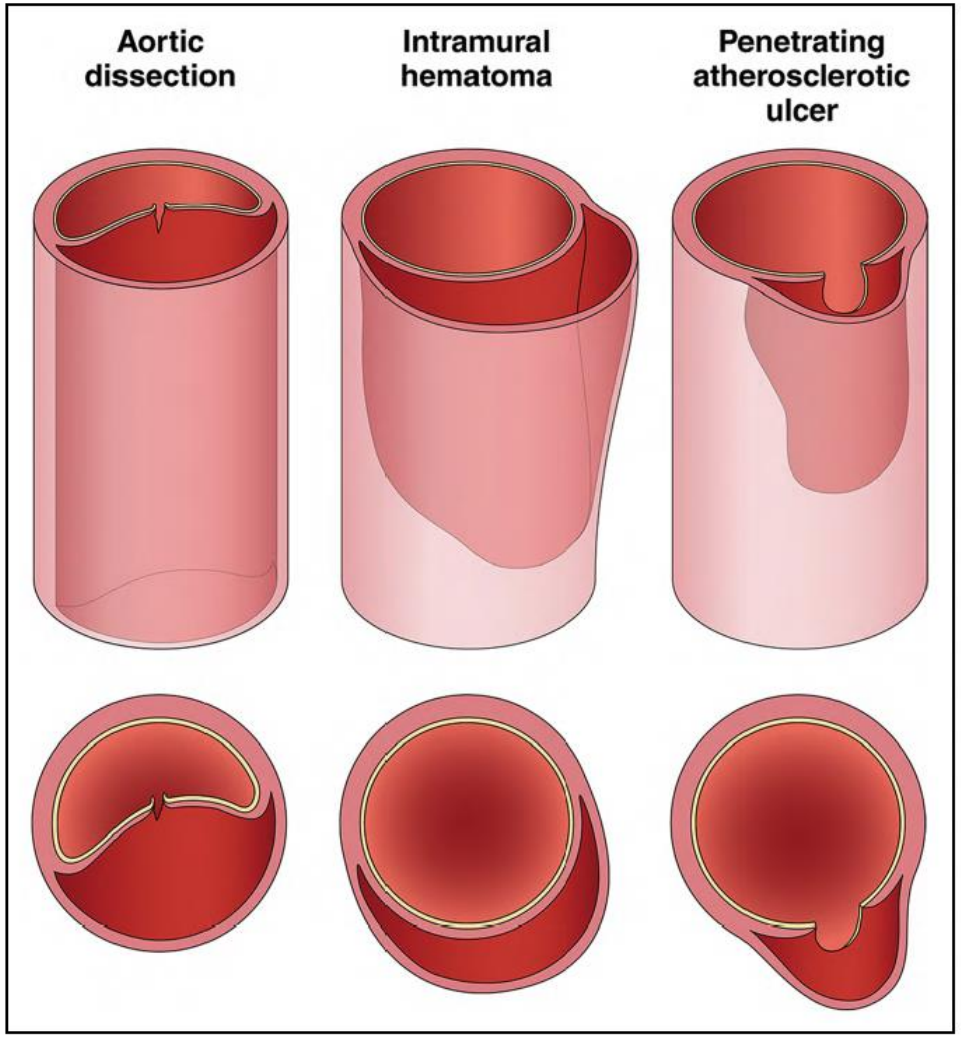
Genes Associated with Acute Aortic Syndromes

Gene	Key functions	Consequences of mutations
FBN1	• Encodes fibrillin 1 • Involved in the formation of microfibrils and elastogenesis • Promotes TGFβ2 bioavailability • Confers a smooth muscle cell phenotype on cells that express it	• Increases the risk of dissection in the ascending and thoracic aorta • Marfan syndrome (OMIM #154700)
EFEMP2	• Encodes fibulin 4 • Involved in the formation of elastic fibres	• Increases the risk of dissection in the ascending aorta • Cutis laxia autosomal recessive IIA (OMIM #219200)
TGFB1	• Encodes a cytokine with multiple functions	• Increases the risk of aortic dissection
TGFBR1 and *TGFBR2*	• Encode receptors involved in TGFβ signalling	• Increases the risk of dissection in the thoracic aorta • Loeys–Dietz syndrome (OMIM #609192)
MYH11	• Encodes smooth muscle myosin heavy chain • Involved in smooth muscle cell contraction	• Increases the risk of dissection in the thoracic aorta • Familial thoracic aortic aneurysm with patent ductus arteriosus (OMIM #132900)
ACTA2	• Encodes aortic smooth muscle actin • Involved in smooth muscle cell contraction	• Increases the risk of dissection in the thoracic aorta • Familial thoracic aortic aneurysm (OMIM #611788)
COL3A1	• Encodes type III collagen • A component of connective tissue	• Increases the risk of dissection in the thoracic aorta • Alters the composition of the extracellular matrix • Ehlers–Danlos vascular type IV (OMIM #130050)

(Nienaber CA et al. Eur Heart Journal 2012;33,1:26-25b)

Clinical Presentation of Acute Aortic Syndromes

- ✓ The presentation of patients with AAS is often similar, regardless of the underlying condition, whether AAD, IMH, or PAU
- ✓ In an analysis of IRAD, AAD presentation was described as abrupt pain by 84% of patients, with initially severe intensity in 90% of the cases

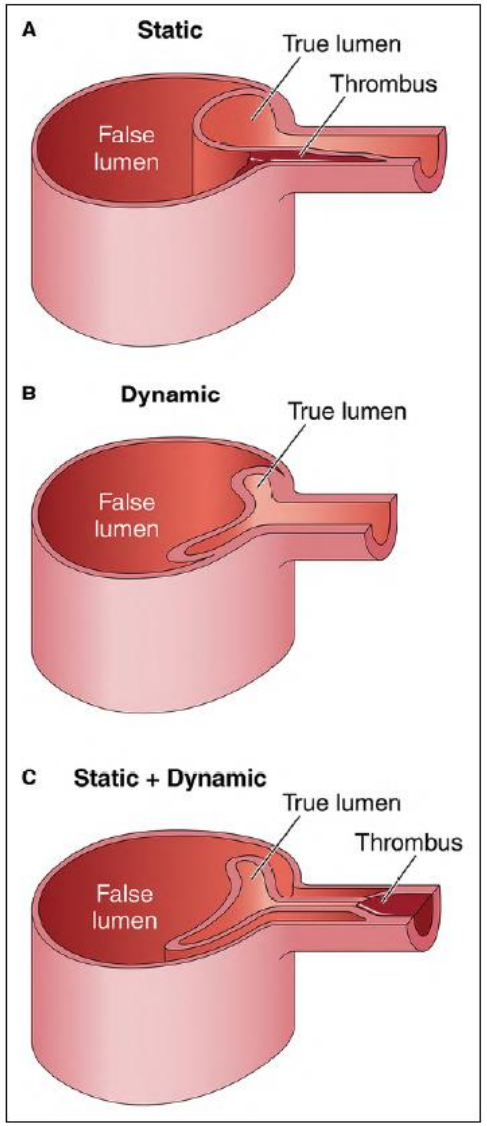


(Hagan PG et al. Jama 2000;283:897-903)

(Isselbacher EM et al. *Circulation. 2022;146:e334–e482)*

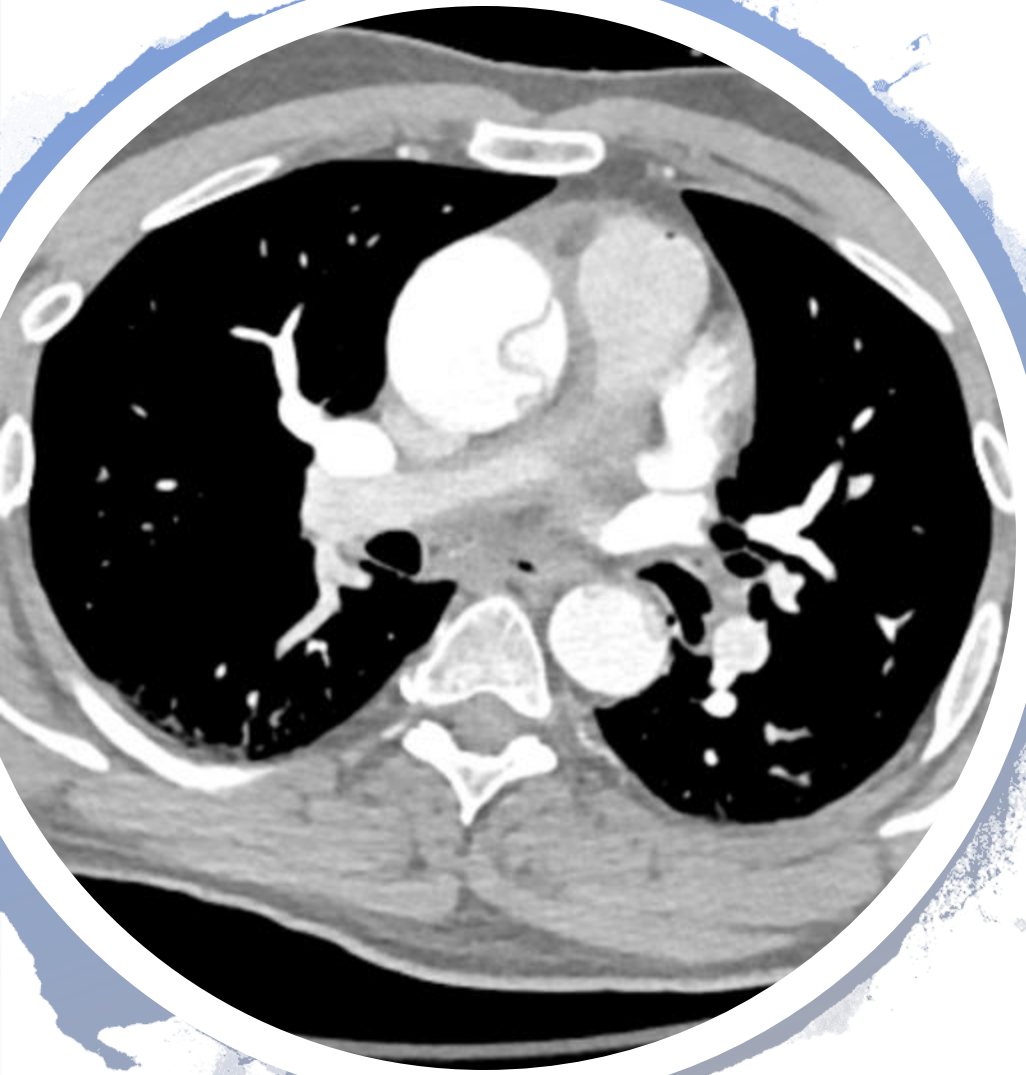
Clinical Presentation of Acute Aortic Syndromes

SYNDROME	CLINICAL PRESENTATION
Type A dissection	Syncope, **tamponade**, severe chest pain, **aortic insufficiency**, collapse, **arm systolic blood pressure differential**, **myocardial ischemia**, neurologic signs
Type B dissection	Severe chest or back pain, migrating pain, **distal pulse differential**, mesenteric ischemia, kidney ischemia/infarct, claudication, distal malperfusion
Intramural hematoma	Chest or back pain, tamponade (rare), high blood pressure, malperfusion (rare)
Penetrating ulcer	Painless or low intensity pain, pain located in back or abdomen, high blood pressure



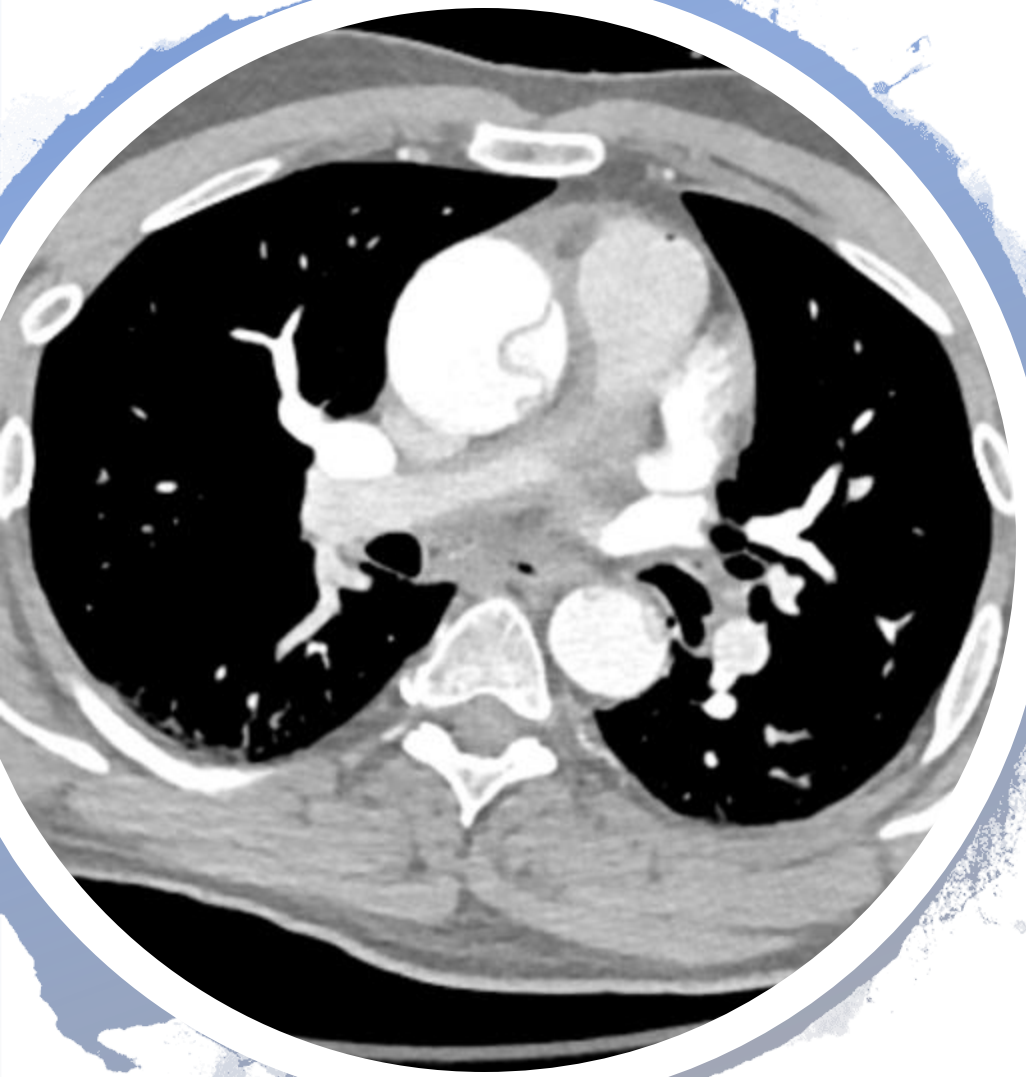
(Isselbacher EM et al. *Circulation. 2022;146:e334–e482*

Diagnosis of Acute Aortic Syndromes



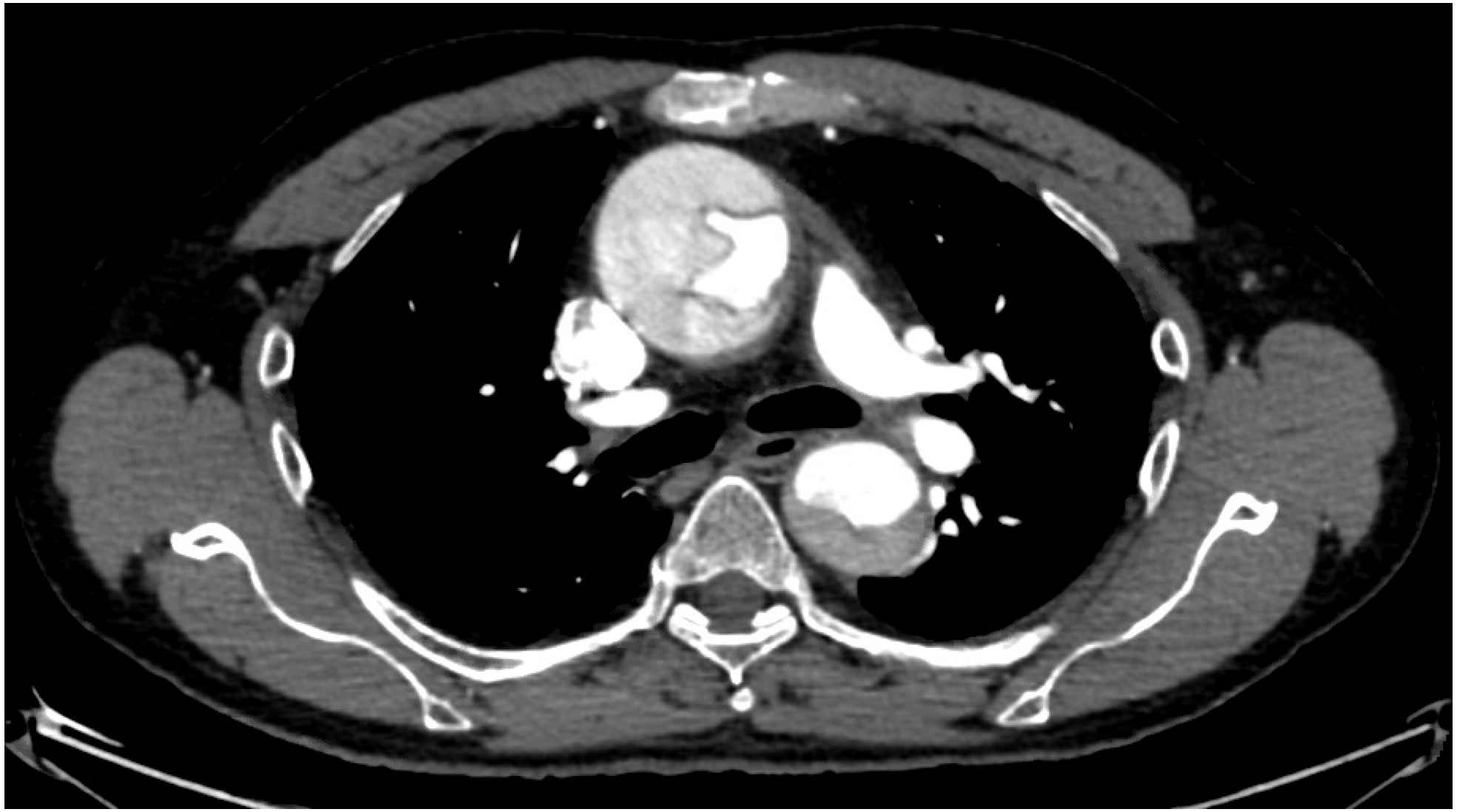
- ✓ The diagnosis of AAS is primarily based on appropriate imaging studies
- ✓ Primary goals of imaging include:
  - confirmation of diagnosis
  - anatomical classification of the AAS
  - assessment of the extent of aortic involvement
  - assessment of complications

Diagnosis of Acute Aortic Syndromes

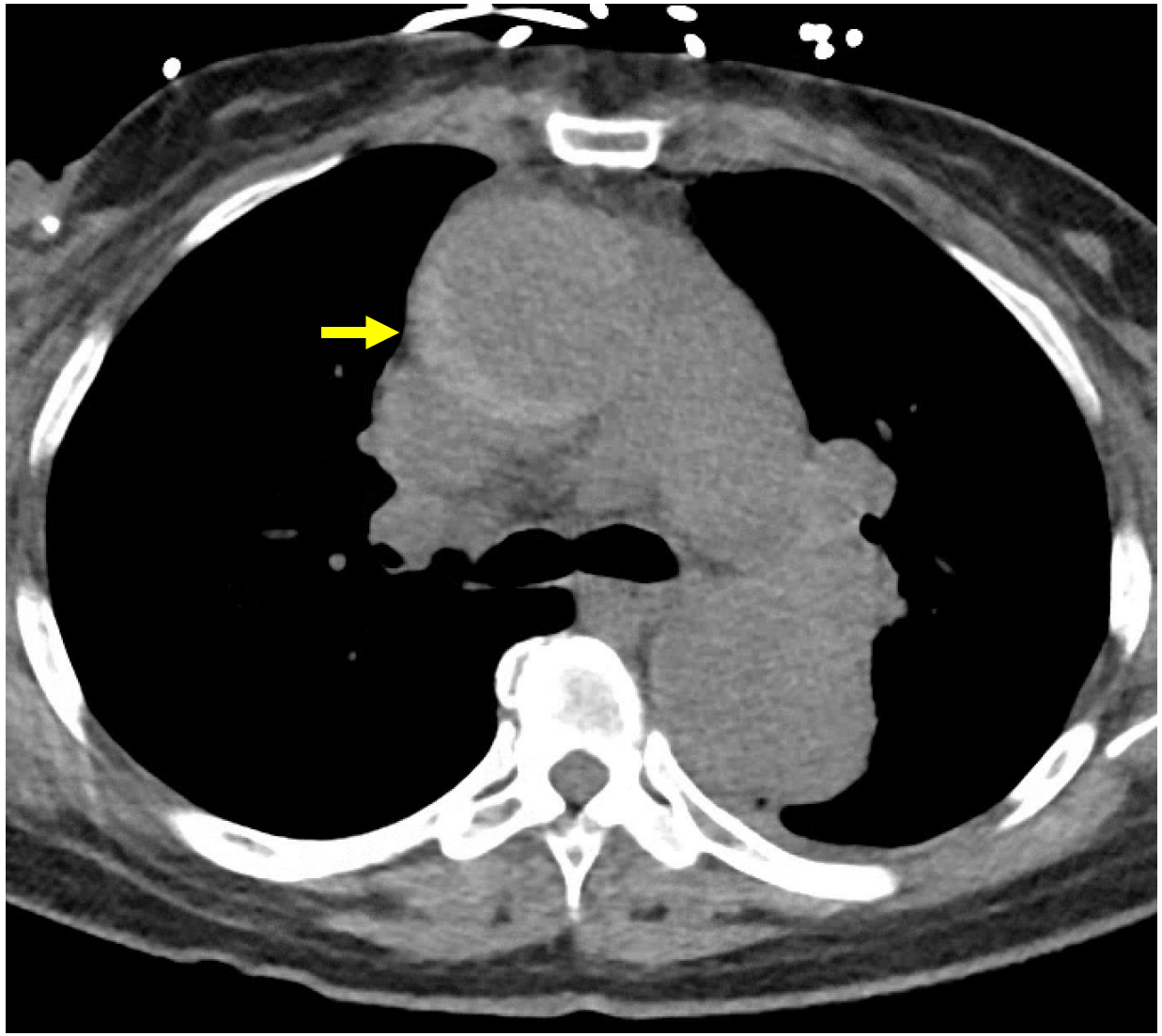


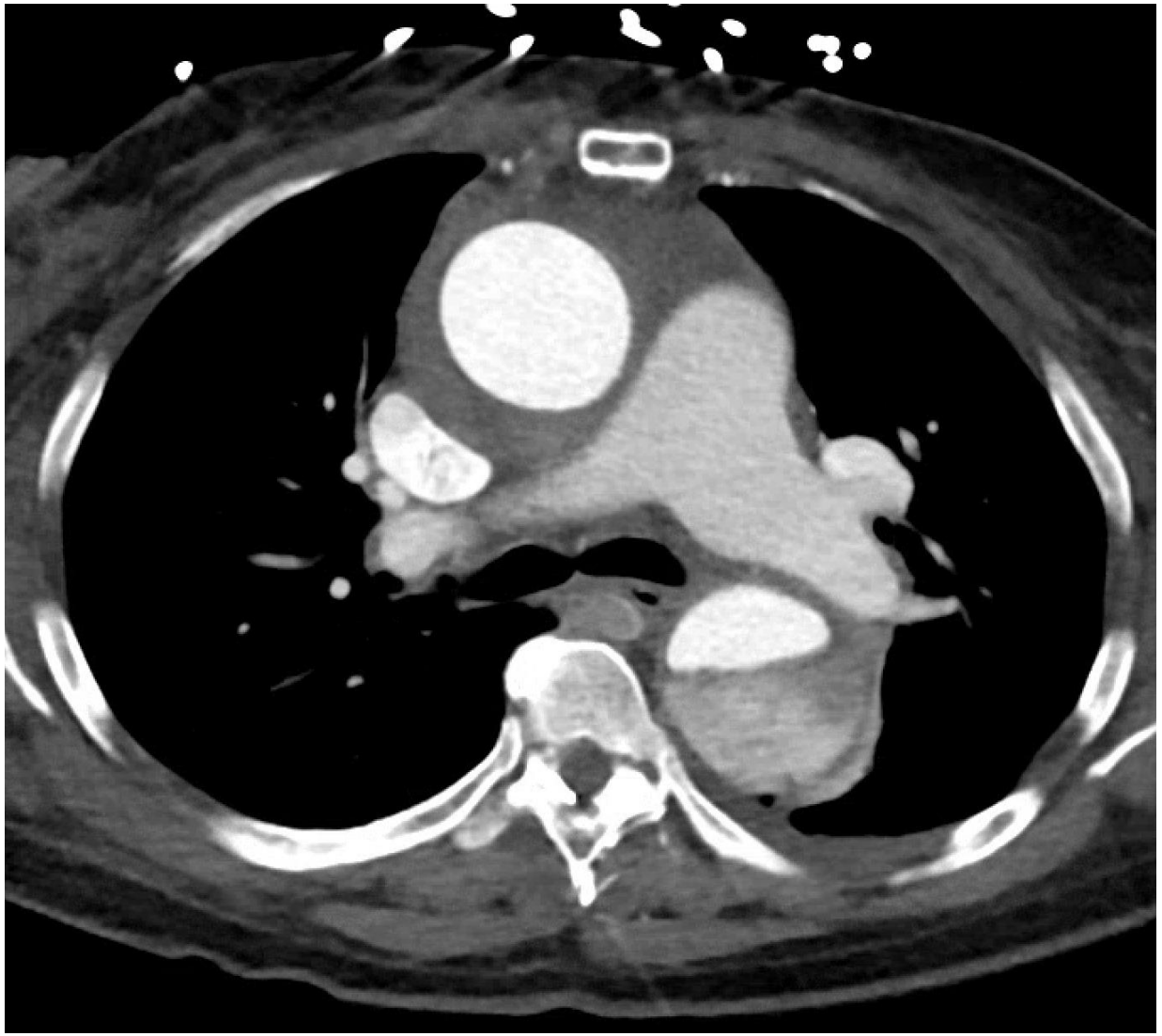
- ✓ CT and MRI angiography, and transesophageal echocardiography (TEE) are similarly reliable for the diagnosis of AAD
- ✓ CT and MRI angiography are superior to TEE for the assessment of AAD extension and branch involvement, as well as for the diagnosis of IMH, and PAU
- ✓ TEE may be preferred in unstable patients

Diagnosis of Acute Aortic Syndromes: Dissection



Diagnosis of Acute Aortic Syndromes: IMH





COR	LOE	Recommendations
1	B-NR	1. In patients presenting to the hospital with AAS, prompt treatment with anti-impulse therapy with invasive monitoring of BP with an arterial line in an ICU setting is recommended as initial treatment to decrease aortic wall stress.[1-5]
1	C-LD	2. Patients with AAS should be treated to an SBP <120 mm Hg or to lowest BP that maintains adequate end-organ perfusion, as well as to a target heart rate of 60 to 80 bpm.[3,6]
1	B-NR	3. In patients with AAS, initial management should include intravenous beta blockers, except in patients with contraindications.[2,5,7]
2a	B-NR	In those with contraindications or intolerance to beta blockers, initial management with an intravenous non-dihydropyridine calcium channel blocker is reasonable for heart rate control.[1,2,5]
1	C-LD	4. In patients with AAS, initial management should include intravenous vasodilators if the BP is not well controlled after initiation of intravenous beta-blocker therapy.[8]
1	C-EO	5. Patients with AAS should be treated with pain control, as needed, to help with hemodynamic management.

Acute Medical Management of All Patients With AAS

- ✓ Anti-impulse therapy with invasive monitoring of arterial pressure
- ✓ Target SBP <120 mmHg and target HR 60-80 bpm
- ✓ Intravenous BB unless contraindicated, can use non-dihydropyridine CCB as an alternative
- ✓ Intravenous vasodilator can be used after start of BB
- ✓ Pain control management as needed

(Isselbacher EM et al. *Circulation. 2022;146:e334–e482*

COR	LOE	Recommendations
1	B-NR	1. In patients presenting with suspected or confirmed acute type A aortic dissection, emergency surgical consultation and evaluation and immediate surgical intervention is recommended because of the high risk of associated life-threatening complications.[1,2]
2a	B-NR	2. In patients presenting with acute type A aortic dissection, who are stable enough for transfer, transfer from a low- to a high-volume aortic center is reasonable to improve survival.[3,4]
2a	B-NR	3. In patients presenting with nonhemorrhagic stroke complicating acute type A aortic dissection, surgical intervention is reasonable over medical therapy to reduce mortality and improve neurologic outcomes.[5,6]

Surgical Management of Type A AAS

- ✓ Emergency surgical evaluation and intervention is recommended
- ✓ In patients stable enough for transport, transfer to a high-volume aortic center is reasonable
- ✓ In patients with type A dissection complicated by stroke, surgery is reasonable

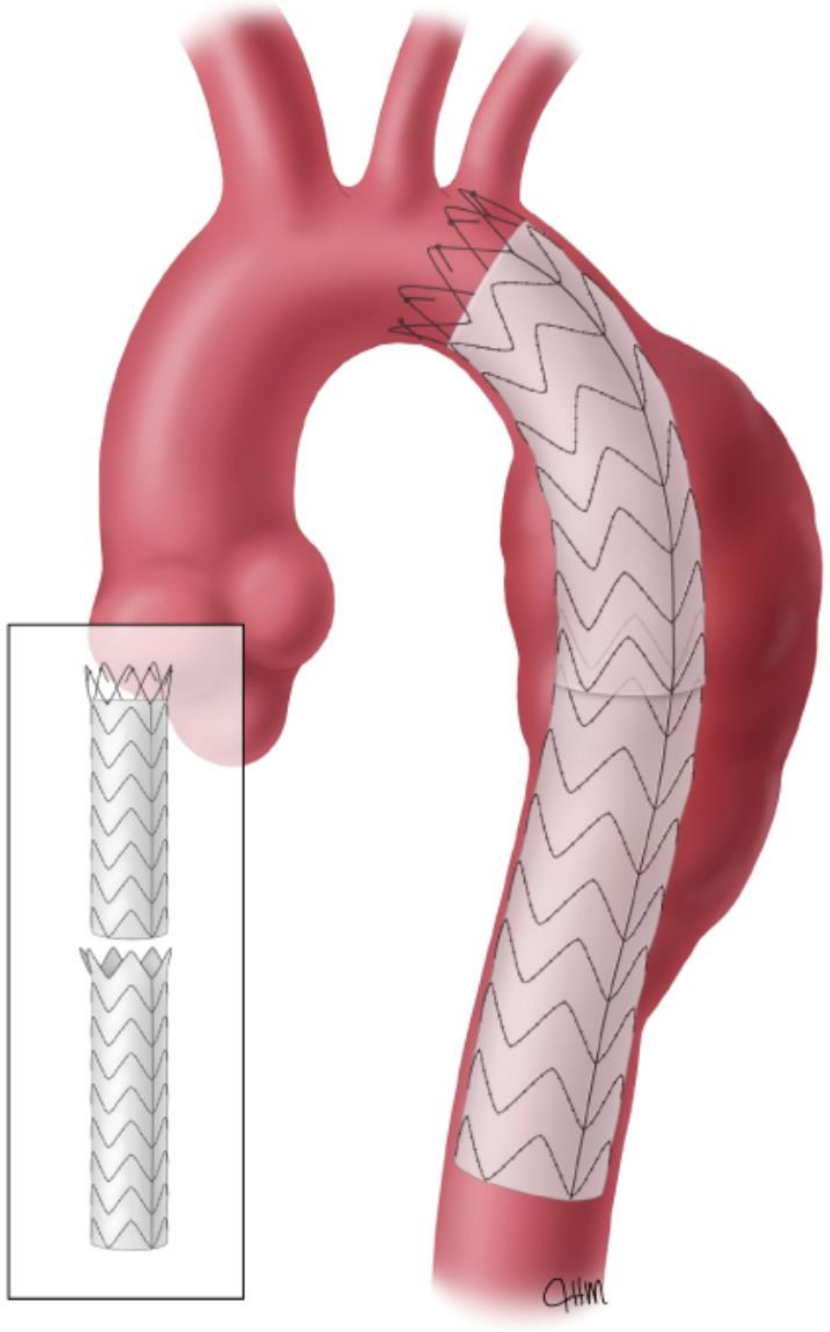
(Isselbacher EM et al. *Circulation. 2022;146:e334–e482*

COR	LOE	Recommendations
1	B-NR	1. In patients with acute type A aortic dissection presenting with renal, mesenteric, or lower extremity malperfusion, it is recommended to proceed to immediate operative repair of the ascending aorta.[1,2]
2a	C-LD	2. In patients with acute type A aortic dissection presenting with clinically significant mesenteric (celiac, SMA) malperfusion, either immediate operative repair of the ascending aorta or immediate mesenteric revascularization via endovascular or open surgical intervention by those with this expertise before ascending aortic repair is reasonable.[3-6]

Surgical Management of Malperfusion in Type A AAS

- ✓ In patients with acute type A AAD and mesenteric (celiac, superior mesenteric artery), renal, or lower extremity malperfusion, immediate surgical repair of the ASCENDING aorta is recommended
- ✓ In patients with type A dissection and mesenteric malperfusion, immediate mesenteric revascularization is reasonable before ascending aortic repair

(Isselbacher EM et al. *Circulation. 2022;146:e334–e482*



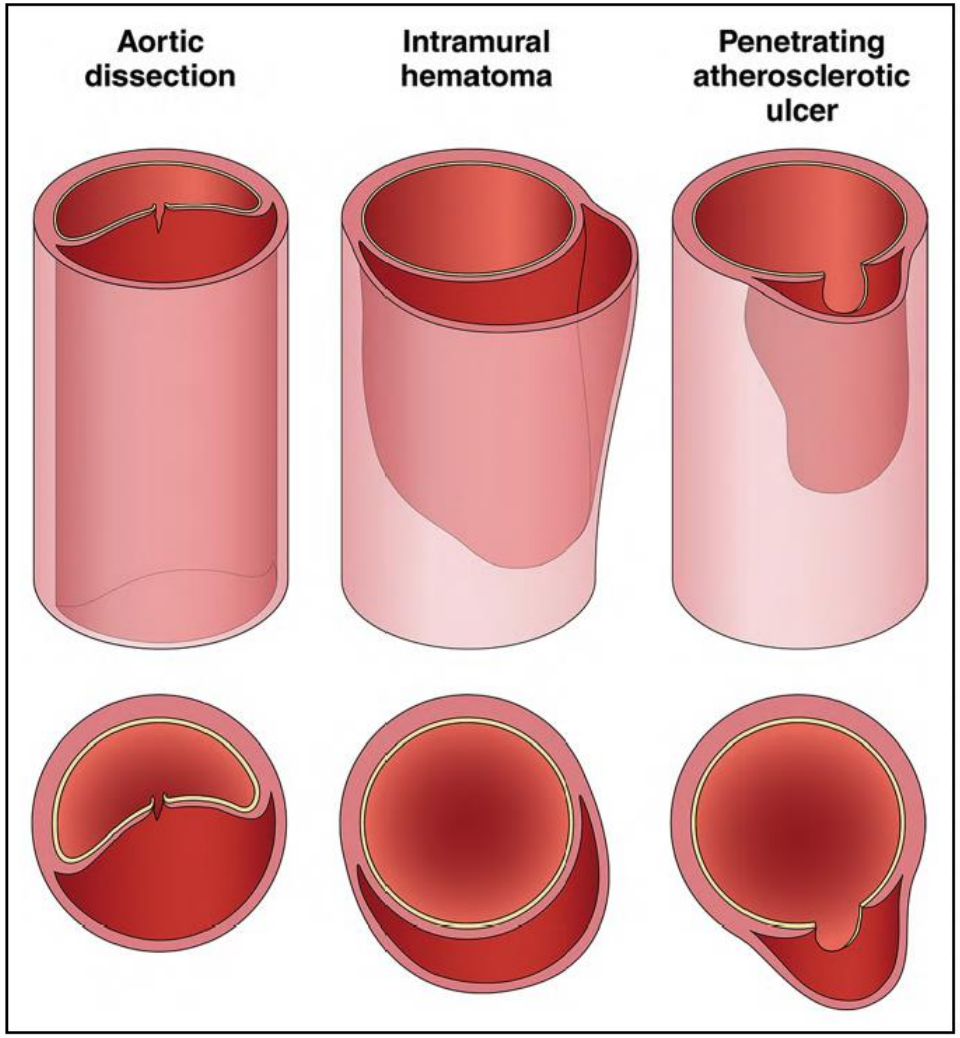
Management of Patients With Type B AAD

- ✓ In uncomplicated type B AAD, medical therapy is recommended
- ✓ In patients with complicated type B AAD, intervention is recommended
- ✓ In ruptured type B AAD, endovascular grafting is recommended if possible
- ✓ In patients with other complications, endovascular grafting is reasonable
- ✓ In patients with high risk anatomy, endovascular grafting may be considered

(Isselbacher EM et al. *Circulation. 2022;146:e334–e482*

Long-Term Management of AAS Survivors

- ✓ The 10-year actuarial survival rate among patients with AAS who survive their initial hospitalization ranges from 30% to 60%
- ✓ AAS should be considered a lifelong disease involving the entire aorta and its branches, which remain at high risk for re-dissection, aneurysm formation, and rupture, even after successful treatment of the acute index event



(Isselbacher EM et al. *Circulation. 2022;146:e334–e482)*

Long-Term Management of AAS Survivors

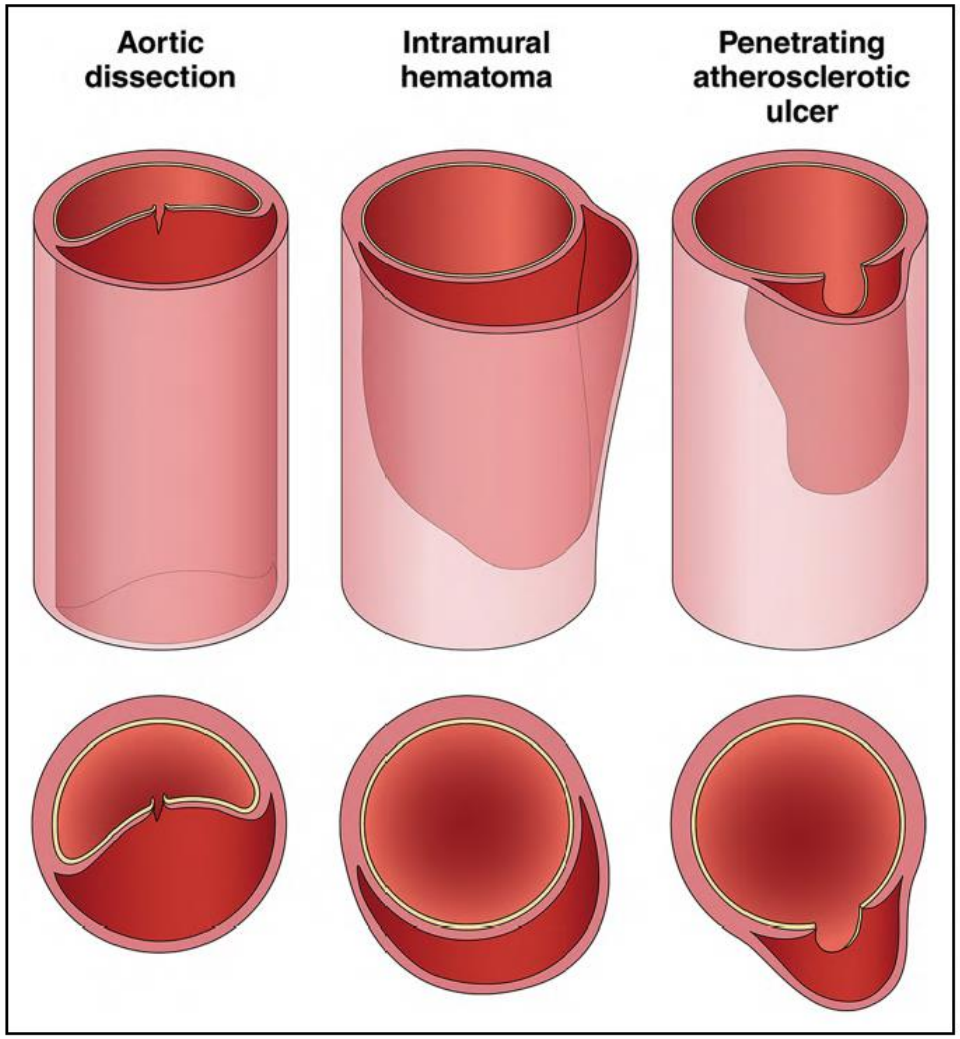
- ✓ Patients with AAS, regardless of the initial therapeutic strategy, require lifelong clinical and imaging monitoring, patient education, and, if appropriate, screening of family members for aortic disease
- ✓ CT or MRI angiography are recommended for surveillance at 1, 6, and 12 months, and annually thereafter, depending on clinical conditions, aortic size, and increase in size over time

COR	LOE	Recommendations
1	B-NR	1. In patients who have had an acute aortic dissection and IMH treated with either open or endovascular aortic repair and have residual aortic disease, surveillance imaging with a CT (or MRI) is recommended after 1 month, 6 months, and 12 months and then, if stable, annually thereafter.[1-6]
1	B-NR	2. In patients who have had an acute aortic dissection and IMH that was managed with medical therapy alone, surveillance imaging with a CT (or MRI) is recommended after 1 month, 6 months, and 12 months and then, if stable, annually thereafter.[7]

(Isselbacher EM et al. *Circulation. 2022;146:e334–e482)*

Long-Term Management of AAS Survivors

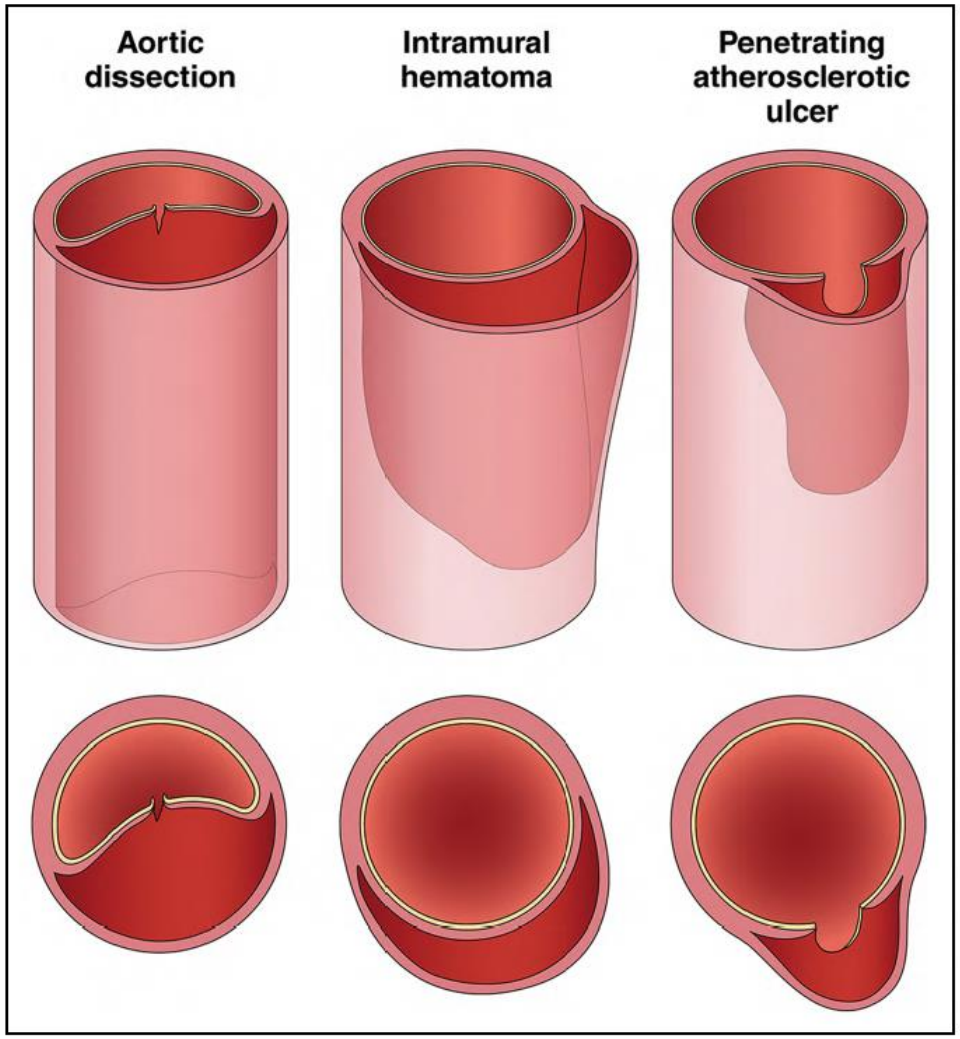
- ✓ Intensive blood pressure (<120/80 mmHg) and heart rate control (<60 bpm) remain the key goals of medical therapy
- ✓ Atherosclerotic aortic disease should be considered a coronary risk equivalent and treated with statins as appropriate. Statins may reduce thoracic aortic aneurysm growth rate and progression to dissection, rupture or death



(Isselbacher EM et al. *Circulation. 2022;146:e334–e482)*

Long-Term Management of AAS Survivors

- ✓ Target heart rate can be achieved with the use of BB, or with diltiazem or verapamil in patients who are intolerant to BB
- ✓ Management of blood pressure may require the addition of thiazide diuretics, ACE-inhibitors, ARBs, dihydropyridine calcium antagonists (amlodipine, nifedipine), and other second line agents (hydralazine, alpha blockers)



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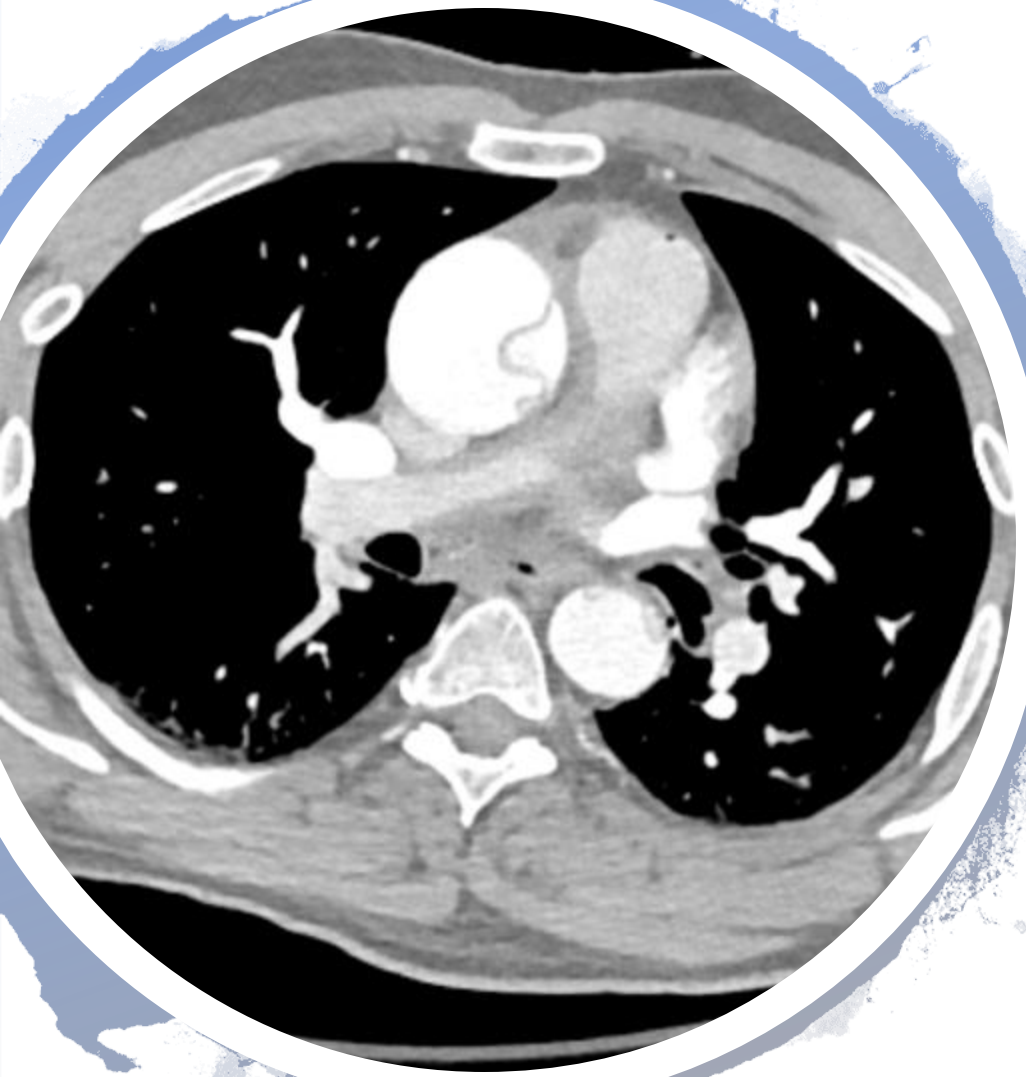
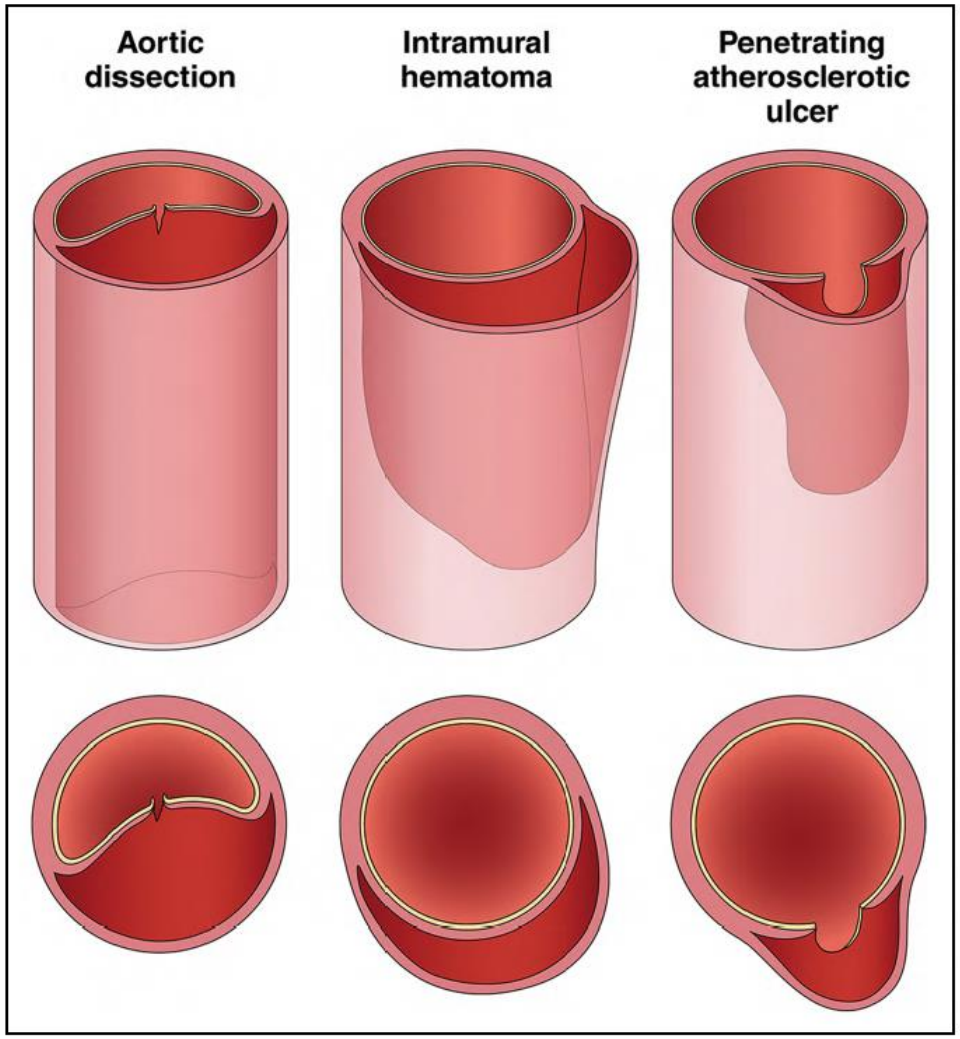
Long-Term Management of AAS Survivors



- ✓ MRI angiography should be preferred to CT angiography, particularly in younger patients, due to increased cancer risk from ionizing radiation
- ✓ Surveillance imaging with consistent use of the same modality is preferable to allow direct comparison of serial studies along with standardized reports and measurements at given landmarks

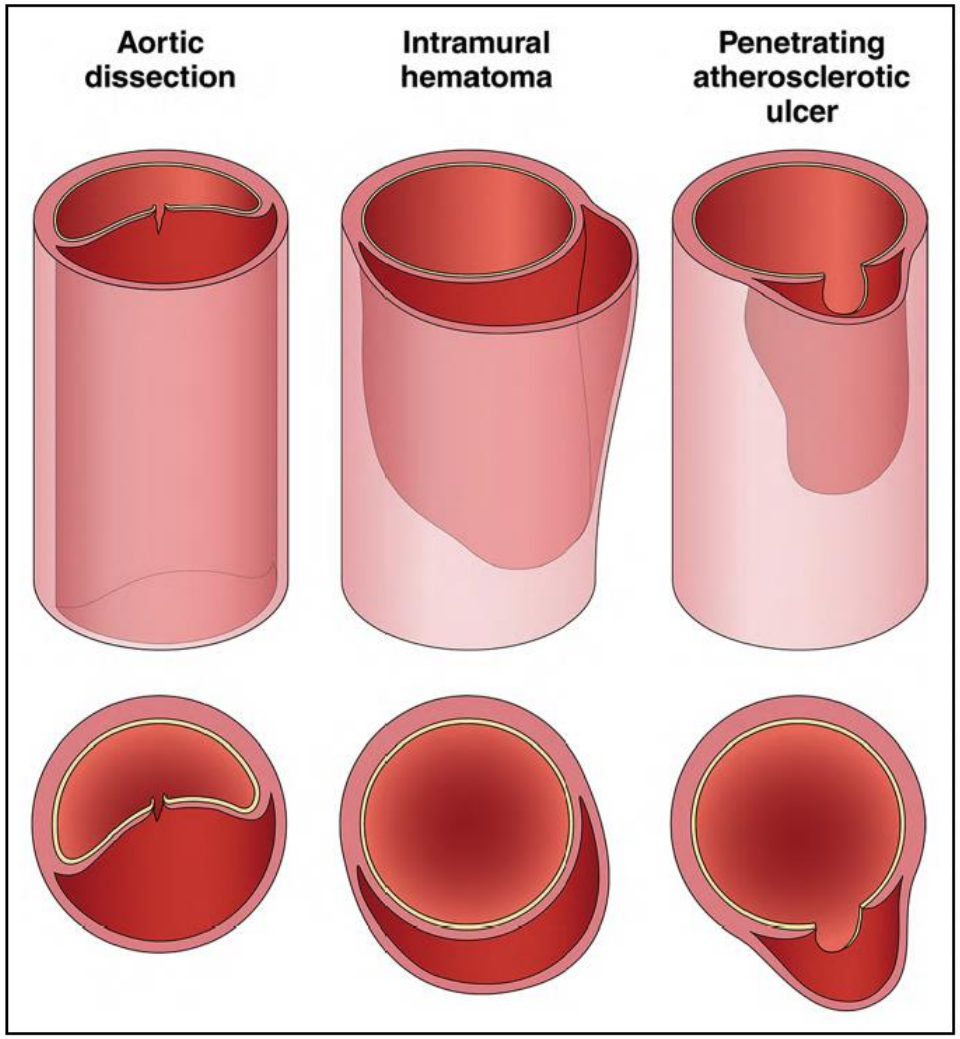
SUMMARY

- ✓ AAS are relatively uncommon aortic pathologies but are associated with high morbidity and mortality if not promptly diagnosed and treated
- ✓ If an AAS is suspected, emergent imaging must be obtained to confirm/rule out the diagnosis
- ✓ Patients with Type A AAS should undergo emergent surgical repair



SUMMARY

- ✓ Patients with type B AAS should be admitted to an ICU for blood pressure and heart rate control, and close monitoring for the occurrence of early complications
- ✓ Survivors of AAS should be closely followed for intensive blood pressure and heart rate control and for regular surveillance imaging



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