ACUTE AORTIC SYNDROME

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OVERVIEW OF THIS EPISODE

- Scope of the problem
- Predisposing aortic disease
- TAA and its variants
- Acute aortic syndrome
- Aortic dissection; diagnosis and management
- Type B dissection and IMH
- Long term follow-up

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SCOPE OF THE PROBLEM

- Aortic diseases can be broadly classified as thoracic aortic aneurysm (TAA), abdominal aortic aneurysm (AAA) and acute aortic syndromes (AAS).
- These diseases are variously associated with one or more conditions leading to increased aortic wall stress (such as systemic hypertension, cocaine use or trauma) and/or aortic media abnormalities (such as Marfan syndrome, bicuspid aortic valve (BAV), inflammatory vasculitis and atherosclerosis).
- During the past two decades, the overall global death rates from aortic disease (including TAA, AAA and acute aortic dissection (AAD)) have increased from 2.49 per 100,000 (95% CI 1.78–3.27) in 1990 to 2.78 per 100,000 (95% CI 2.04–3.62) in 2010.
- This increase seems to be more evident in developing countries, with a median death rate of 0.71 (95% CI 0.28–1.40) compared with 0.22 (95% CI 0.10–0.33) in developed nations

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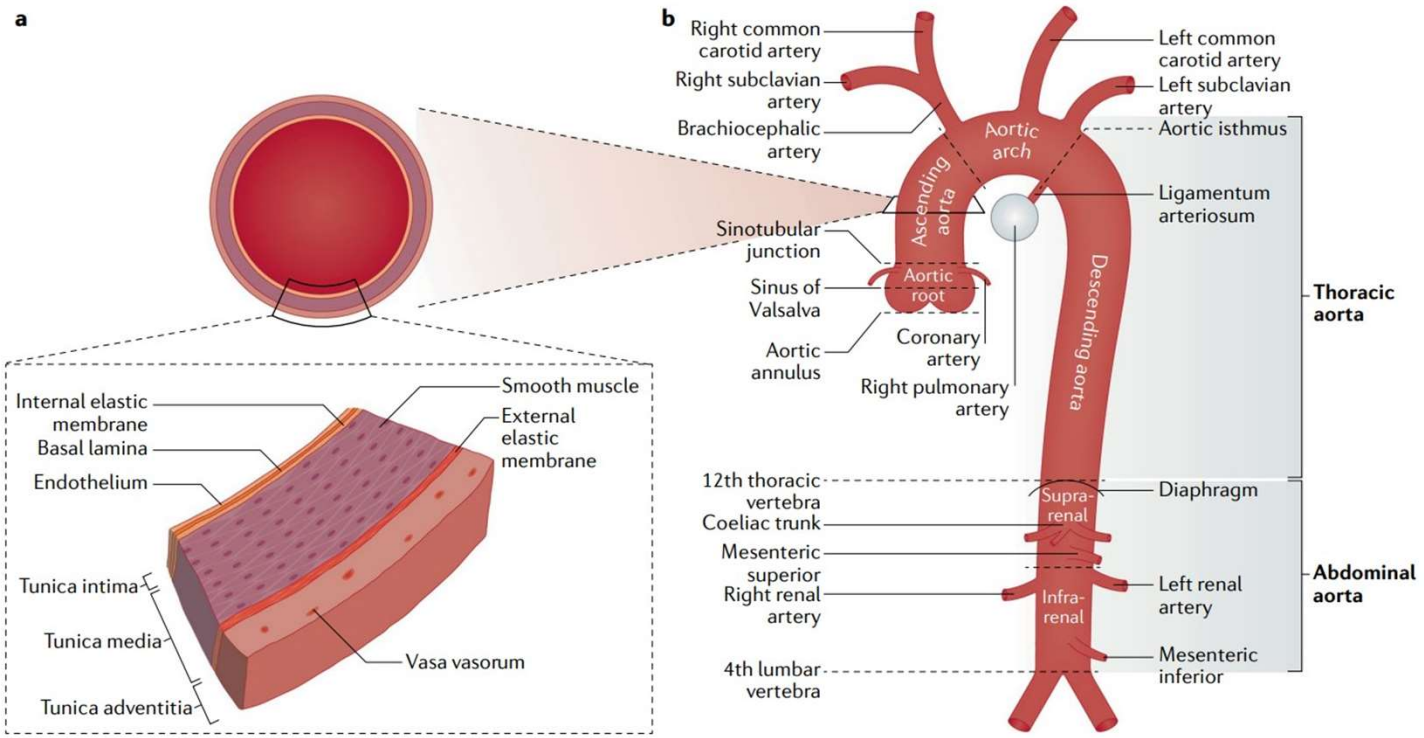

Fig. 1 | **The structure and anatomy of the aorta. a** | The aortic wall is composed of three layers: the thin tunica intima, the thickest musculoelastic tunica media and the outer fibrous tunica adventitia. **b** | Anatomically, the aorta is divided into thoracic and abdominal components, above and below the diaphragm, respectively. The thoracic aorta includes the aortic root and the ascending aorta, the aortic arch and the descending segment. The abdominal aorta includes the suprarenal and infrarenal segments; it then ends at the level of the fourth lumbar vertebra, where it bifurcates into the left and right common iliac arteries.

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Table 1 | **Global death rates from aortic disease stratified by age and sex**

Age (years)	Men		Women	
	1990	2010	1990	2010
25–29	0.18 (0.10–0.32)	0.16 (0.10–0.27)	0.10 (0.04–0.17)	0.07 (0.04–0.11)
30–34	0.29 (0.16–0.52)	0.25 (0.15–0.47)	0.18 (0.07–0.35)	0.11 (0.06–0.18)
35–39	0.48 (0.28–0.89)	0.43 (0.25–0.74)	0.30 (0.12–0.65)	0.19 (0.1–0.35)
40–44	0.84 (0.51–1.44)	0.78 (0.46–1.4)	0.53 (0.23–1.20)	0.35 (0.19–0.65)
45–49	1.54 (0.91–2.65)	1.42 (0.88–2.37)	0.82 (0.37–1.75)	0.58 (0.32–1.15)
50–54	2.86 (1.71–4.87)	2.64 (1.60–4.51)	1.44 (0.67–3.01)	1.02 (0.60–1.88)
55–59	5.44 (3.16–9.13)	5.02 (3.05–8.43)	2.20 (1.21–3.83)	1.69 (1.05–2.7)
60–64	10.37 (6.48–16.4)	9.41 (5.92–15.21)	4.35 (2.34–7.93)	3.31 (2.03–5.55)
65–69	19.79 (12.62–31.32)	17.19 (10.90–28.76)	8.47 (4.63–14.91)	6.37 (3.97–10.25)
70–74	33.52 (20.73–54.78)	29.11 (18.81–46.81)	17.09 (8.9–33.07)	12.33 (7.37–20.84)
75–79	53.62 (34.66–80.46)	46.70 (29.85–73.76)	26.63 (16.46–44.07)	21.02 (13.4–33.43)
≥80	88.93 (59.35–130.7)	86.82 (57.06–128.59)	59.65 (38.74–93.08)	52.46 (34.88–80.33)

Aortic disease includes aortic aneurysms and aortic dissections. Estimates are expressed as mean death rates per 100,000 of the general population, with 95% CIs. Adapted with permission from REF.[6]

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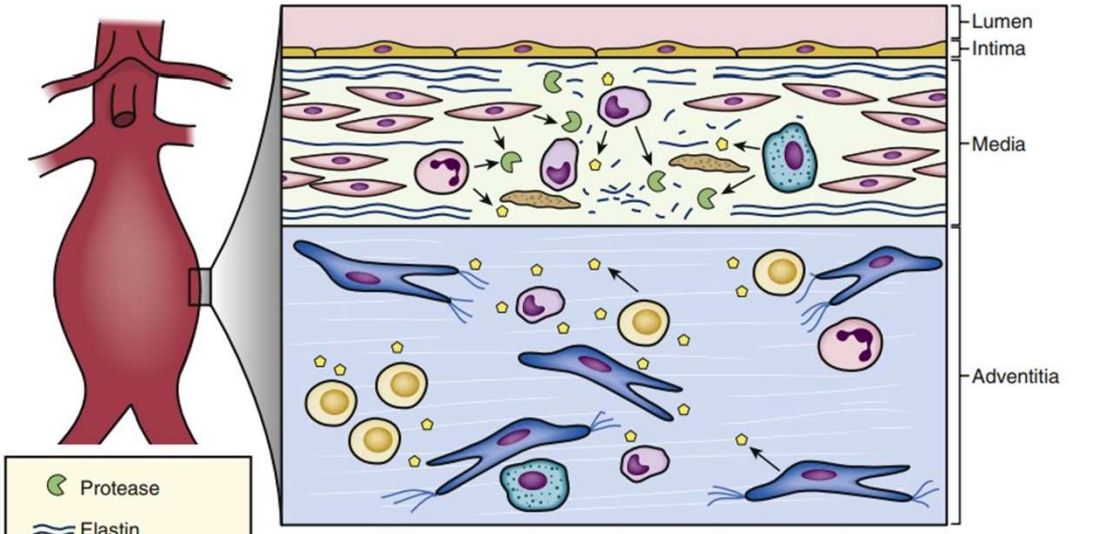

	Macrophage	Lymphocyte	Neutrophil	Mast cell	VSMC	Fibroblast
Cytokines and chemokines	TGF-β TNF-α IL-6 IL-23	IFN-γ IL-1β TGF-β IL-6 TNF-α IL-17 IL-23		IL-1β TNF-α IL-6	TGF-β MCP-1 IL-6	TGF-β MCP-1
Proteases	MMP-9	Cysteine Protease	Elastase	Chymase	MMP-2	

MicroRNAs	miR-21, miR-29b, and miR-712
RAS	Renin, ACE, and AT1R

EFIGURE 42.1 Pathophysiology and therapeutic targets of AAA. Progressive dilation of the aortic wall is associated with recruitment of leukocytes, macrophage activation, and production of proinflammatory cytokines. Over a period of years, apoptosis and cellular senescence of smooth muscle cells occurs in conjunction with infiltration of lymphocytes, mast cells, and neutrophils. Macrophages and VSMCs also produce proenzyme forms of proteases that are activated in the extracellular space and degrade extracellular matrix proteins (elastin and interstitial collagens). Adventitial fibroblasts are presumed to promote structural repair, but the interstitial collagen becomes disorganized. The table illustrates major cell types involved in AAA pathogenesis, as well as selected examples of future therapeutic targets that are involved in AAA pathogenesis. *AAA,* Abdominal aortic aneurysm; *AT1R,* angiotensin type 1 receptor; *IFN,* interferon; *IL,* interleukin; *MCP,* monocyte chemoattractant protein; *miR,* microRNA; *MMP,* matrix metalloproteinase; *RAS,* renin-angiotensin system; *TGF-β,* transforming growth factor-β; *TNF,* tumor necrosis factor; *VSMC,* vascular smooth muscle cell. (From Davis FM, Rateri DL, Daugherty A. Mechanisms of aortic aneurysm formation: translating preclinical studies into clinical therapies. *Heart*. 2014;100:1498–1505.)

Box 1 | **Conditions associated with aortic syndromes and aneurysms**

Conditions associated with increased aortic wall stress

- Hypertension, particularly if uncontrolled
- Phaeochromocytoma
- Cocaine or other stimulant use
- Weight lifting or other Valsalva manoeuvre
- Coarctation of the aorta
- Traumatic aortic injuries (partial or complete transection of the aorta)
  - High-speed motor vehicle accidents
  - Falling from a great height

Conditions associated with aortic media abnormalities

- Genetic syndromes
  - Marfan syndrome
  - Loeys–Diez syndrome
  - Ehlers–Danlos syndrome, type IV or vascular type
  - Turner syndrome
  - Arterial tortuosity syndrome
  - Aneurysms–osteoarthritis syndrome
  - Others
- Genetic, non-syndromic familial thoracic aortic aneurysm and dissection syndrome
  - Known genetic variants in *ACTA2*, *FBN1*, *MYH11*, *MYLK*, *PRKG1*, *SMAD3*, *TGFBR1* or *TGFBR2*
  - Unknown genetic variants
- Bicuspid aortic valve (including previous aortic valve replacement)
- Non-genetic conditions
  - Inflammatory vasculititis (Takayasu arteritis, giant cell arteritis or Behçet arteritis)
- Other
  - Atherosclerosis
  - Pregnancy
  - Polycystic kidney disease
  - Chronic corticosteroid or immunosuppression agent administration
  - Fluoroquinolone exposure
  - Infection involving the aortic wall from either bacteraemia or extension of adjacent infection

Iatrogenic

- Cardiac surgery
- Coronary angiography
- Coronary intervention

REF.[3].

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TAA

TABLE 42.2 Thoracic Aortic Aneurysm Syndromes and Conditions Due to a Heritable or Genetic Cause

CONDITION	GENE	CLINICAL FEATURES
Syndromic HTAD*		
Marfan syndrome	*FBN1*	Aortic root aneurysm, AD, TAA, MVP, long bone overgrowth, scoliosis, pectus deformities, ectopia lentis, myopia, tall stature, PTX, dural ectasia
Loeys-Dietz syndrome	*TGFBR1, TGFBR2, *SMAD3, TGFB2, TGFB3, SMAD2*	TAA, branch vessel aneurysms, AD, arterial tortuosity, MVP, craniosynostosis, hypertelorism, bluish sclera, bifid/broad uvula, translucent skin, visible veins, club feet, dural ectasia, *premature osteoarthritis
Vascular Ehlers-Danlos syndrome	*COL3A1*	TAA, AAA, arterial rupture, AD, MVP, bowel and uterine rupture, PTX, translucent skin, atrophic scars, small joint hypermobility, easy bruising, carotid-cavernous fistula
Arterial tortuosity syndrome	*SLC2A10*	Tortuous large-and medium-sized arteries, aortic dilatation, craniofacial, skin and skeletal features
Shprintzen-Goldberg syndrome	*SKI*	Craniosynostosis, skeletal features, aortic dilatation
Congenital contractural arachnodactyly (Beals syndrome)	*FBN2*	MVP, arachnodactyly, Marfanoid habitus, digital contractures, mild aortic dilatation
Cutis laxa	*EFEMP2* (Fibulin-4)	TAA, arterial tortuosity, arterial stenosis, hypertelorism, arachnodactyly
EDS with periventricular nodular heterotopia (PVNH)	*FLNA (filamin A)*	X-linked, PVNH, TAA, BAV, MV disease, PDA, VSD, seizures, joint hypermobility
Meester-Loeys syndrome	*BGN*	X-linked, TAA, AD, skeletal abnormalities
LOX-related TAA	LOX (lysyl oxidase)	TAA, BAV, AD, Marfanoid habitus in some
Nonsyndromic HTAD (Familial TAA)		
FTAA	*ACTA2* (α-smooth muscle actin)	TAA, AD, BAV, Moya-Moya, premature CAD and CVD, livedo reticularis, iris flocculi
FTAA	*MYH11* (Myosin heavy chain-11)	TAA, AD, PDA
FTAA	*MYLK* (Myosin light chain kinase)	AD at relatively small aortic size
FTAA	*PRKG1* (Protein kinase cGMP-dependent)	Aortic root aneurysm and AD
FTAA	*MAT2A* (MAT IIα)	TAA, AD, BAV
FTAA	*MFAP5* (microfibrillar-associated protein 5)	TAA, AD, skeletal features may be present
FTAA	*FOXE3* (forkhead transcription factor)	TAA, AD
Bicuspid Aortic Valve/Associated Ascending Aortic Aneurysm		
Familial BAV/AS and TAA	*NOTCH1* (NOTCH1)	Aortic stenosis, TAA
TGFBR1, TGFBR2, TGFB2, TGFB3, ACTA2, MAT2A, GATA5, SMAD6, LOX, ROBO4, TBX20	BAV with TAA	Syndromic and nonsyndromic FTAA with an increased frequency of BAV
Turner syndrome	XO, Xp	BAV, COA, TAA, AD, short stature, lymphedema, webbed neck, premature ovarian failure, affects 1 in 2500 live-born girls

*Some individuals with pathogenic variants in a gene which can lead to syndromic HTAD have very few or no syndromic features and variants in some genes causing syndromic HTAD may also lead to nonsyndromic HTAD.

AAA, Abdominal aortic aneurysm; *AAT*, aortic aneurysm syndrome; *AD*, aortic dissection; *BAV*, bicuspid aortic valve; *CAD*, coronary artery disease; *COA*, coarctation of the aorta; *CVD*, cerebrovascular disease; *EDS*, Ehlers-Danlos syndrome; *FTAA*, familial thoracic aortic aneurysm (and dissection) syndrome; *HTAD*, heritable TAA; *LDS*, Loeys-Dietz syndrome; *MFS*, Marfan syndrome; *MV*, mitral valve; *MVP*, mitral valve prolapse; *PDA*, patent ductus arteriosus; *PTX*, pneumothorax; *TAA*, thoracic aortic aneurysm; *TGF*, transforming growth factor; *VSD*, ventricular septal defect.

TABLE 42.3 Clinical Features Increasing the Likelihood of a Genetic Predisposition for Thoracic Aortic Aneurysm Disease

1. Aortic dilatation (especially >45 mm)
   a. Consideration for 40–45 mm (or z-score >3), especially when dilated sinuses of Valsalva, or in the young (<50 years old), or when associated with a +FH
2. Positive family history of TAA (and/or cerebral aneurysm disease)
   a. First-degree and/or second-degree relative with:
      i. TAA or aortic dissection
      ii. Aneurysm or dissection in the arterial tree, diagnosed below 60 years old*
      iii. Bicuspid aortic valve
      iv. Patent ductus arteriosus
      v. Sudden (unexplained) death below 50 years old
3. Syndromic features
   a. Craniofacial features (craniosynostosis, hypertelorism, cleft palate, bifid uvula)
   b. Ocular features (lens dislocation, retinal detachment, high myopia [-6 diopters or higher], iris flocculi
   c. Cardiovascular features (MVP, arterial tortuosity, multiple aneurysms/dissections, BAV, PDA)
4. Musculoskeletal features
   a. Pectus deformities
   b. Disproportionately elongated fingers, toes, arms, legs
   c. Joint dislocations, hypermobility, or contractures
   d. Severe, early-onset osteoarthritis
   e. Severe scoliosis or kyphosis
   f. Lumbosacral dural ectasia
5. Cutaneous features
   a. Translucent skin with visible veins
   b. Livedo reticularis
   c. Abnormal striae not related to weight gain
   d. Widened scars
   e. Facial milia
6. Other features
   a. Spontaneous pneumothorax
   b. Recurrent hernias
   c. Spontaneous rupture of internal organs

*There may be a wide variability in the age of onset or recognition of aneurysm disease in heritable thoracic aortic disease.
BAV, Bicuspid aortic valve; *FH,* family history; *MVP,* mitral valve prolapse; *PDA,* patent ductus arteriosus; *TAA,* thoracic aortic aneurysm.
Adapted from Verhagen JMA, Kempers M, Cozijnsen L, et al. Expert consensus recommendations on the cardiogenetic care for patients with thoracic aortic disease and their first-degree relatives. *Int J Cardiol*. 2018;258:243–248.

TABLE 42.4 Size Threshold for Prophylactic Aortic Root or Ascending Aortic Aneurysm Resection for Various Conditions

CONDITION	SIZE THRESHOLD FOR PROPHYLACTIC AORTIC ROOT OR ASCENDING ANEURYSM RESECTION*
Degenerative aneurysm	≥5.5 cm
Bicuspid aortic valve	≥5.5 cm
Bicuspid aortic valve with risk factors or low surgical risk†	≥5.0 cm
Bicuspid aortic valve requiring aortic valve replacement	≥4.5 cm
Marfan syndrome	≥5.0 cm
Marfan syndrome with risk factors‡	≥4.5 cm
Loeys-Dietz syndrome§	4.0–4.5 cm
Familial thoracic aortic aneurysm syndromes¶	4.5–5.0 cm
Turner syndrome**	>2.5 cm/m²

*Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid growth of the aorta. Age, body size, rapid growth, family history, risk of surgery, and patient and physician preferences may influence aortic size threshold.
†Family history of aortic dissection or aortic growth rate ≥0.5 cm per year or if the patient is at low surgical risk (<4%) and the surgery is performed by an experienced aortic surgical team in a center with established expertise in these procedures. Other risk factors for aortic dissection include coarctation of the aorta, hypertension, and the root phenotype of BAV.
‡Family history of aortic dissection or rapid aortic growth (>3 mm per year), severe aortic or mitral regurgitation. If pregnancy desired, consider prophylactic aortic surgery for aortic diameter of ≥4.0 to 4.5 cm.
§It is reasonable to consider surgical repair of the aorta in adults with Loeys-Dietz syndrome (LDS) or a confirmed *TGFBR1* or *TGFBR2* mutation with aortic diameter of ≥4.2 cm by transesophageal echocardiogram or ≥4.4 to 4.6 cm by CT or MRI. Aortic surgery at smaller diameters (>4 cm) may be recommended when there are severe craniofacial features, marked arterial tortuosity, rapid growth or a family history of aortic dissection. LDS due to *SMAD3* mutations may be treated similarly and less information is available for LDS related to T*GFB2* and T*GFB3* mutations.
¶Surgical thresholds vary depending upon the specific gene mutation involved. TAA due to *ACTA2*, *SMAD3*, and *MYLK* may lead to aortic dissection at relatively small aortic diameters and smaller size thresholds may be indicated.
**AHA Scientific Statement (Silberbach M, et al., 2018) recommends prophylactic surgery at ASI greater than 2.5 cm/m², whereas ESC 2014 Guidelines (Erbel R, et al., 2014) recommend prophylactic surgery at ASI ≥2.75 cm/m².
Adapted from Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). *Eur Heart J*. 2014;35:2873–2926; Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with Thoracic Aortic Disease. *Circulation*. 2010;121:e266–e369; Hiratzka LF, Creager MA, Isselbacher EM, et al. Surgery for aortic dilatation in patients with bicuspid aortic valves: a statement of clarification from the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2016;67:724–731. Silberbach M, Roos-Hesselink JW, Andersen NH, et al. Cardiovascular Health in Turner Syndrome: A Scientific statement from the American Heart Association. *Circ Genom Precis Med*. 2018 Oct;11(10):e000048.

Table 2 | **Features of TAAs and AAAs**[2,11–13]

Feature	TAA	AAA
Male-to-female ratio	2:1 to 4:1	4:1 to 5:1
Risk factors	Atherosclerosis, smoking, syndromic disorders (such as Marfan syndrome), non-syndromic disorders, bicuspid aortic valve, infectious or non-infectious aortitis, and traumatic injury	Smoking, age >60 years, atherosclerosis, hypertension, male sex, AAA in first-degree relatives, dyslipidaemia and obesity
Preventive measures	Healthy lifestyle, cardiovascular risk-factor modifications, and educational and screening programmes	Healthy lifestyle, cardiovascular risk-factor modifications, and educational and screening programmes
Screening	TTE	A-US
Signs and symptoms	Long asymptomatic phase; back, interscapular or left shoulder pain (descending TAA), or neck or jaw pain (aortic arch TAA); involvement of adjacent organs and/or structures In the event of rupture: hypotension and/or shock, gastrointestinal haemorrhage (rare) or haemoptysis (rare)	Long asymptomatic phase, pulsatile abdominal mass, atypical abdominal or back pain In the event of rupture: intense pain, hypotension and fast pulse
Diagnostic test	TTE plus CT angiography or MRI	A-US plus CT angiography or MRI
Surveillance	Using TTE plus MRI if needed, annually if TAA diameter is <45 mm, or every 6 months if TAA diameter is 45–55 mm	Using A-US, every 3 years if AAA diameter is 30–39 mm, annually if AAA diameter is 40–49 mm, or every 3–6 months if AAA diameter is ≥50 mm
Treatment	TAA repair is indicated if TAA diameter is ≥50 mm in women or in patients with connective tissue disorders; or if TAA diameter is ≥55 mm in patients with no connective tissue disorder TEVAR is often preferred to open surgery for anatomically suitable descending aorta aneurysms ≥55 mm in patients without connective tissue disorders	AAA repair is indicated if AAA diameter is >55 mm in men or >50 mm in women or aneurysm growth is >10 mm per year Among patients with an aneurysm that is anatomically suitable for EVAR and who are at an acceptable surgical risk, either open or endovascular aortic repair is recommended

AAA, abdominal aortic aneurysm; A-US, abdominal ultrasonography; EVAR, endovascular aneurysm repair; TAA, thoracic aortic aneurysm; TEVAR, thoracic endovascular aortic repair; TTE, 2D transthoracic colour–Doppler echocardiography.

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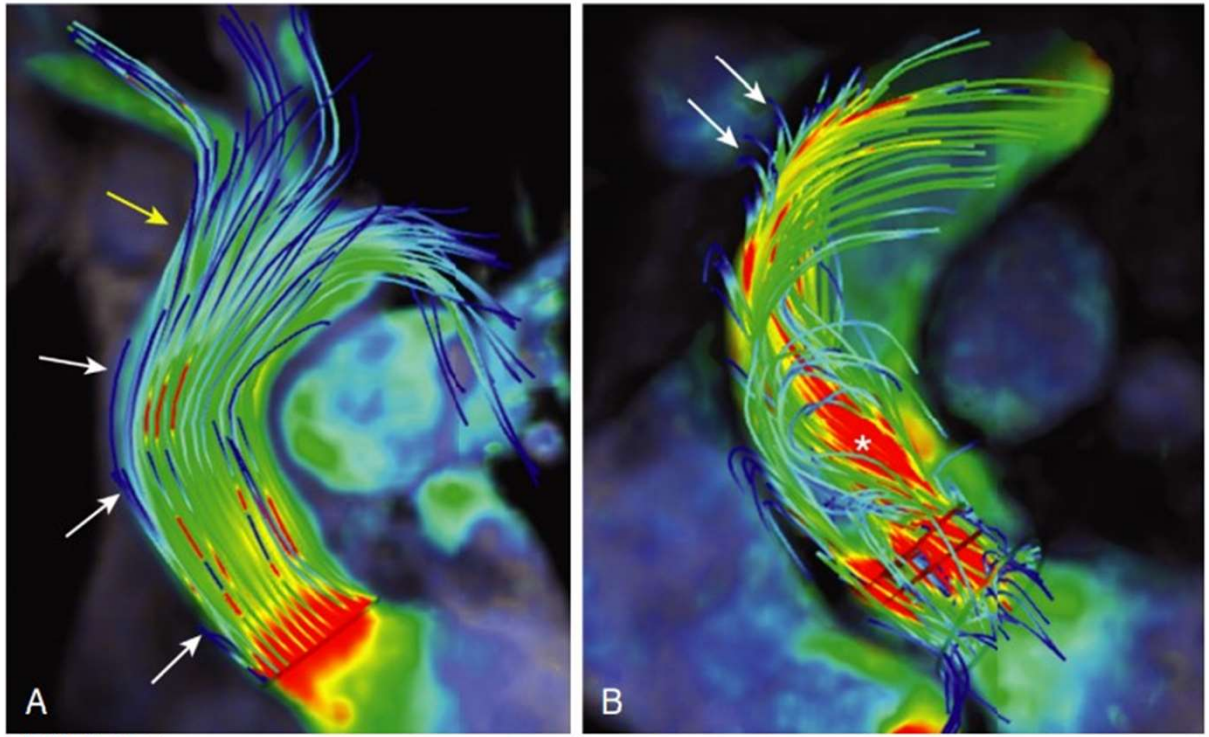
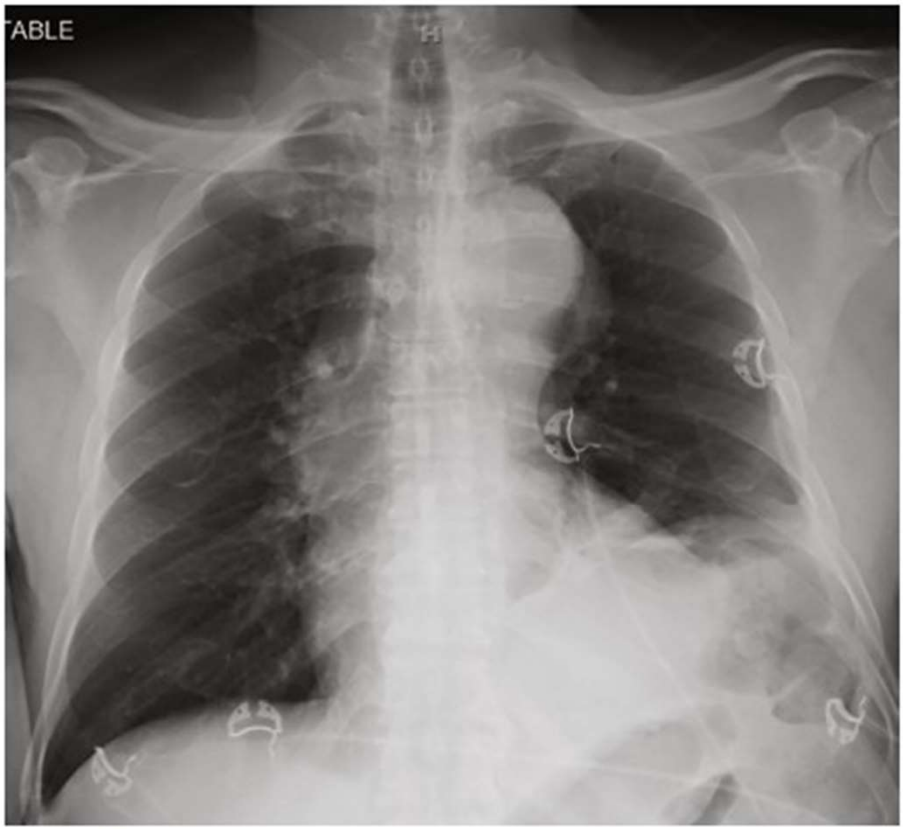
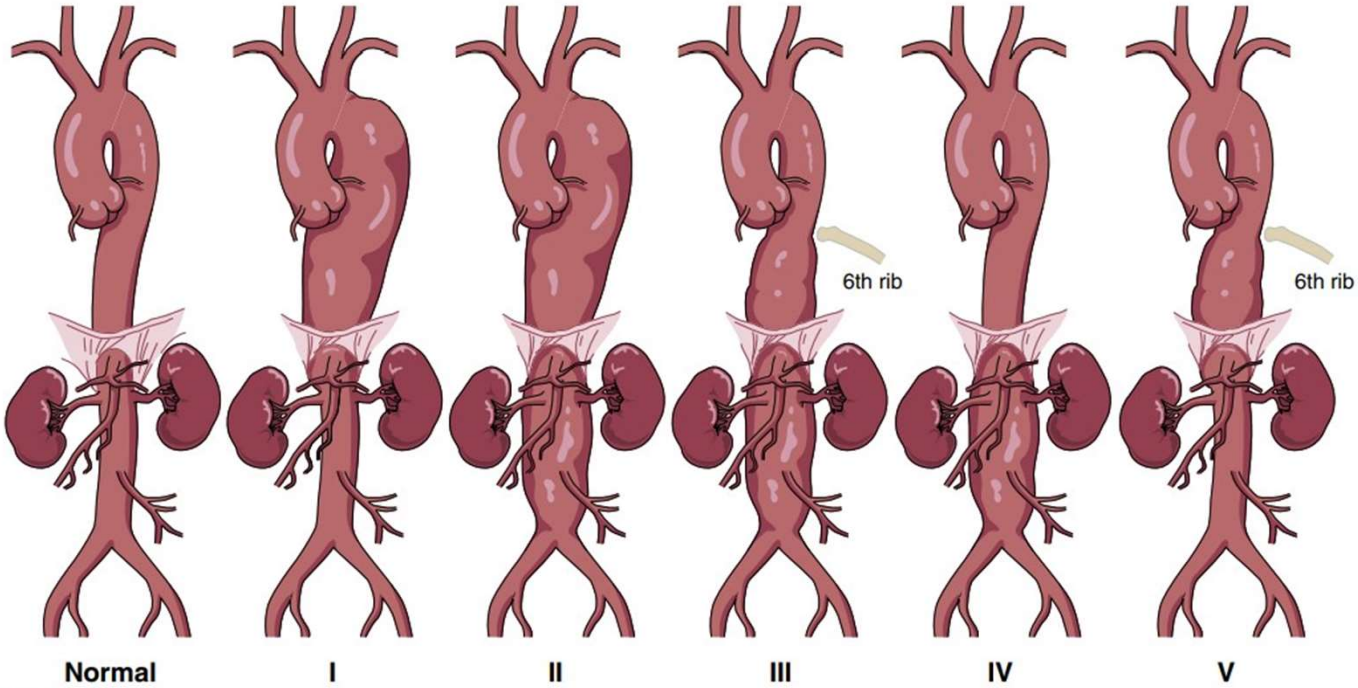


FIGURE 42.7 Abnormal ascending aortic flow patterns related to bicuspid aortic valve (BAV). **A,** Healthy subject with normal 3-cusp aortic valve. Streamline visualization demonstrates normal laminar flow in the ascending aorta *(white arrows)* and proximal arch *(yellow arrow)*, with color scale representing flow velocity (blue indicates low velocity, red indicates high velocity). **B,** Subject with BAV without stenosis or regurgitation or ascending aortic dilation. Streamlines illustrate eccentric, helical flow in the ascending aorta with high-velocity systolic flow jet (*asterisk*) impacting the aortic wall in the region of the greater curvature *(white arrows)*. (Courtesy Dr. Nicholas Burris. From Bhave NM, Nienaber CA, Clough RE, Eagle KA. Multimodality imaging of thoracic aortic diseases in adults. *JACC Cardiovasc Imaging*. 2018;11:902–919.)



EFIGURE 42.7 Chest radiograph demonstrating widening of the aortic knob and descending aortic shadow due to an aortic arch and proximal descending aortic aneurysm. Note the elevated left hemidiaphragm due to phrenic nerve involvement and atelectasis from left bronchial obstruction. This patient also had hoarseness of voice from laryngeal nerve compression.



EFIGURE 42.15 Classification for thoracoabdominal aortic aneurysms. (From Charlton-Ouw KM, Estrera AL. Thoracic endovascular aortic repair for aneurysm: How I teach it. *Ann Thorac*. 2018;106:646–650.)

Table 3 | **Genetic syndromes associated with thoracic aortic aneurysm and dissection**

Genetic syndrome	Prevalence; incidence[a]	Common clinical features	Genes involved	Samples for diagnostic testing	Comments on aortic disease
Marfan syndrome	15/100,000; 25/100,000[b]	Skeletal features, ectopia lentis, dural ectasia	*FBN1*[c]	Ghent diagnostic criteria; DNA for sequencing[d]	Surgical repair when the aorta reaches 50 mm unless the patient has a family history of aortic dissection at <50 mm, a rapidly expanding aneurysm or presence of clinically significant aortic valve regurgitation
Loeys–Dietz syndrome	Reported in 52 families	Bifid uvula or cleft palate, arterial tortuosity, hypertelorism, skeletal features similar to those in Marfan syndrome, craniosynostosis, aneurysms and dissections of other arteries	*TGFBR2* or *TGFBR1*	DNA for sequencing	Surgical repair recommended at an aortic diameter of >42 mm by TEE (internal diameter) or 44–46 mm by CT and/or MRI (external diameter)
Ehlers–Danlos syndrome type IV or vascular type	1/100,000[b]; 1/10,000–25,000	Thin translucent skin, gastrointestinal rupture, rupture of the gravid uterus, rupture of medium-sized and large arteries	*COL3A1*	DNA for sequencing, dermal fibroblasts for analysis of type III collagen	Surgical repair is complicated by friable tissues; non-invasive imaging is recommended
Turner syndrome	5.5/100,000 (birth prevalence)[b]; 1/2,000–2,500	Short stature, primary amenorrhoea, bicuspid aortic valve, aortic coarctation, webbed neck, low-set ears, low hairline, broad chest	45, X karyotype	Blood cells for karyotype analysis	Risk of aortic dissection is increased in patients with bicuspid aortic valve, aortic coarctation, hypertension or pregnancy
Arterial tortuosity syndrome	<1/100,0000; unknown (102 cases)	Tortuosity, stenosis and aneurysm of large-sized and medium-sized arteries; altered facial features; soft, hyperextensible skin; skeletal features similar to those in Marfan syndrome	*SLC2A10*	DNA for sequencing	Aortic tortuosity, stenosis and aneurysm; management requires baseline whole-body vascular imaging and follow-up should be individually tailored
Aneurysms–osteoarthritis syndrome	<1/100,0000; unknown	Tortuosity, aneurysms and dissections throughout the arterial tree; early-onset joint abnormalities; craniofacial, skin and skeletal features similar to those in Marfan syndrome and Loeys–Dietz syndrome	*SMAD3*	DNA for sequencing	Aortic aneurysms and dissections; some physicians suggest aggressive surgical management, as is recommended for Loeys–Dietz syndrome

TEE, transoesophageal echocardiography. [a]Data on prevalence and incidence are global unless otherwise indicated. [b]Data from Europe only (http://www.orpha.net). [c]Genetic variants in *TGFBR2* have also been associated with Marfan syndrome, but the clinical phenotype is debated. [d]Screening is recommended for first-degree relatives; all affected family members should undergo regular aortic imaging. Data from REFS[2,3].

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Table 4 | **Aortic diseases associated with BAV**

Prevalence (%)	Incidence (%)	Gene involved	Screening	Prevalence of BAV in aortic dissection (%)		BAV type	Percent of patients	Comments on aortic disease
				Type A	Type B			
1.0–2.0	0.5–2.0	*NOTCH1*	First-degree relatives, using transthoracic echocardiography	2–9	3	LCC–RCC	70	Ascending aortic dilatation is common; aortic root dilatation; aortic coarctation
						RCC–NCC	10–20	Aortic root is rarely affected; dilatation of the ascending aorta
						LCC–NCC	5–10	–

BAV, bicuspid aortic valve; LCC, left coronary cusp; NCC, non-coronary cusp; RCC, right coronary cusp. Data from REF.[2].

Table 5 | **Non-syndromic familial thoracic aortic aneurysm and dissection**

Gene involved	Contribution (%)	Associated clinical features	Comments on aortic disease
ACTA2	14	Livedo reticularis, iris floccule, patent ductus arteriosus, bicuspid aortic valve	Two of 13 patients with documented aortic dissections <50 mm; patients with thoracic aortic aneurysm and dissection also present with coronary artery disease, stroke and Moyamoya disease
TGFBR2	4	Thin translucent skin, arterial or aortic tortuosity, aneurysm of arteries	Multiple aortic dissections documented at aortic diameter of <50 mm
MYH11	1	Patent ductus arteriosus	Patients with documented aortic dissection at aortic diameter of 45 mm
MYLK	–	Abnormality of connective tissue, cutis marmorata, cystic medial necrosis of the aorta	Aortic dissection with little to no aortic enlargement
PRKG1	–	Erdheim cystic medial necrosis	Aortic aneurysm and acute aortic dissection at relatively young age

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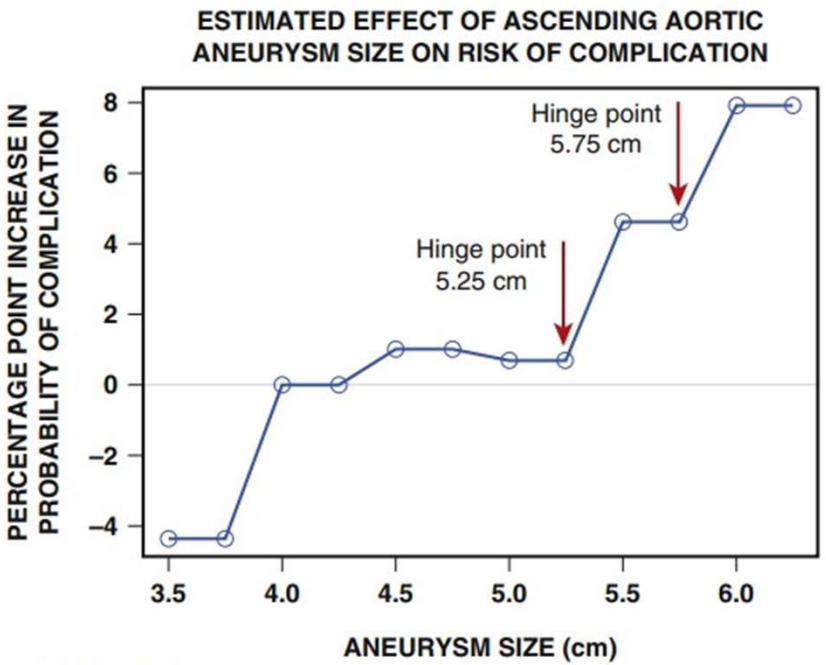

FIGURE 42.10 Estimated probability of rupture or dissection of the ascending aorta by aneurysm size. (From Zafar MA, Li Y, Rizzo JA, et al. Height alone, rather than body surface area, suffices for risk estimation in ascending aortic aneurysm. *J Thorac Cardiovasc Surg.* 2018;155[5]:1938–1950.)

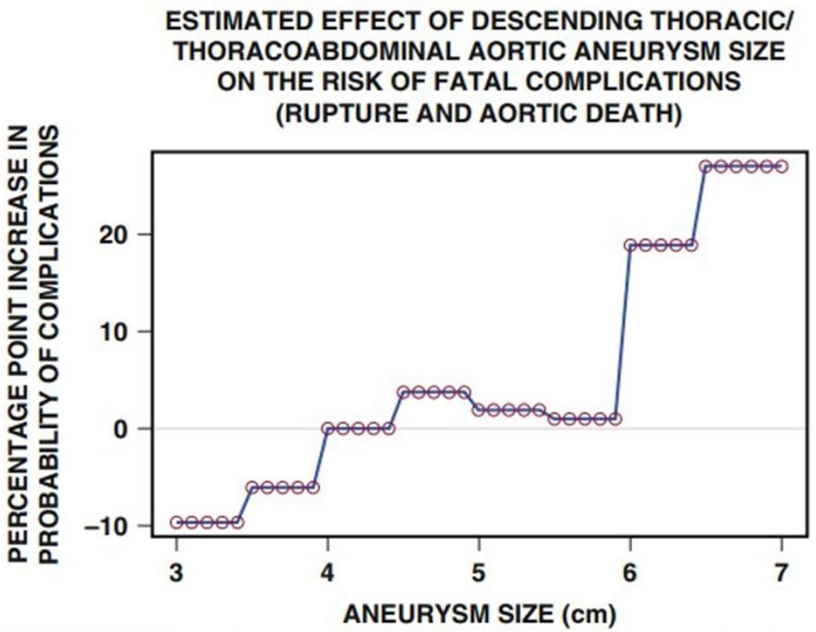

FIGURE 42.11 Probability of rupture or aortic death of the descending thoracic/ thoracoabdominal aorta by aneurysm size. Analysis of the predicted probability of fatal complications (i.e., rupture or aortic death) revealed that the risk increased sharply at 2 hinge points: 6.0 cm and 6.50 cm. (From Zafar MA, Chen JF, Wu J,et al. Natural history of descending thoracic and thoracoabdominal aortic aneurysms. *J Thorac Cardiovasc Surg.* 2019;161[2]:498–511.e1.)

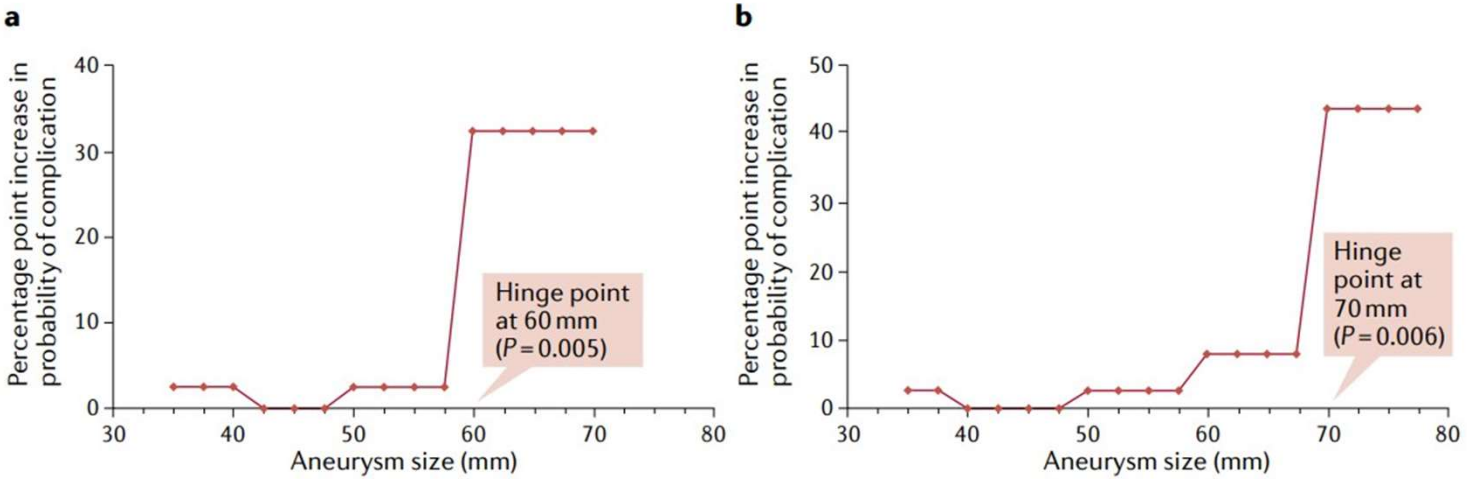

Fig. 3 | **Effect of aortic aneurysm size on the risk of complications.** Increasing thoracic aortic aneurysm (TAA) size confers an increased risk of dissection or rupture. Ascending TAAs with a diameter of ≥60 mm (part **a**) and descending TAAs with a diameter of ≥70 mm (part **b**) are widely regarded as being at very high risk of aortic dissection or rupture. However, in addition to the maximum aneurysm diameter, the risk of dissection or rupture is also related to the rate of aneurysm growth per year and the patient's demographic and clinical features, such as age, sex, smoking status, diastolic hypertension and aneurysm-related pain[11]. Adapted with permission from REF.[48], Elsevier.

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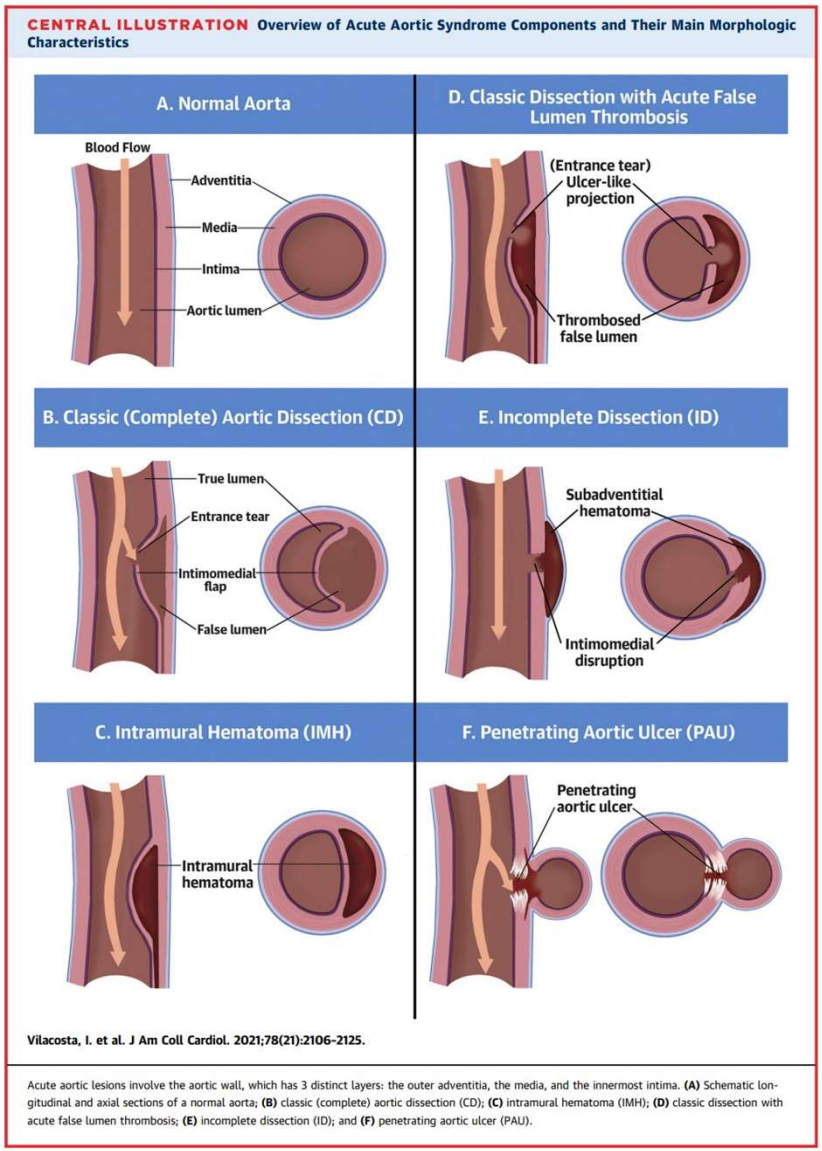
Acute aortic syndrome



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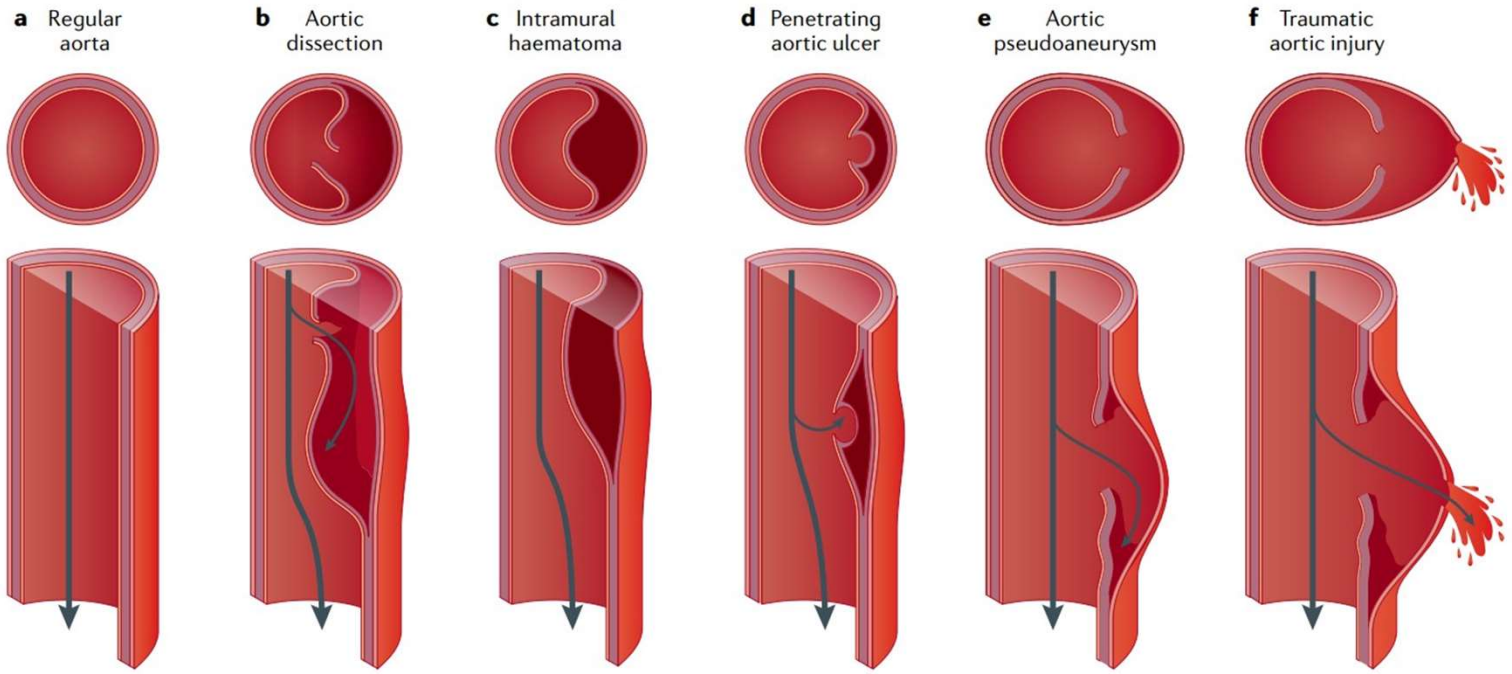

Fig. 4 | **Acute aortic syndromes. a** | Normal aorta. **b** | Classic aortic dissection: disruption of the medial layer results in separation of the aortic wall layers and subsequent formation of true and false lumens divided by an intimal flap. **c** | Intramural haematoma can develop in the media of the aortic wall in the absence of a false lumen and intimal tear. **d** | Penetrating aortic ulcer: ulceration of an aortic atherosclerotic plaque, penetrating through the internal elastic lamina into the media. **e** | Pseudoaneurysm: dilatation of the aorta owing to disruption of all the aortic wall layers, contained only by the periaortic connective tissue. **f** | Traumatic aortic injury: rupture of all aortic wall layers caused by a trauma.

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TABLE 42.7 Risk Factors for Aortic Dissection

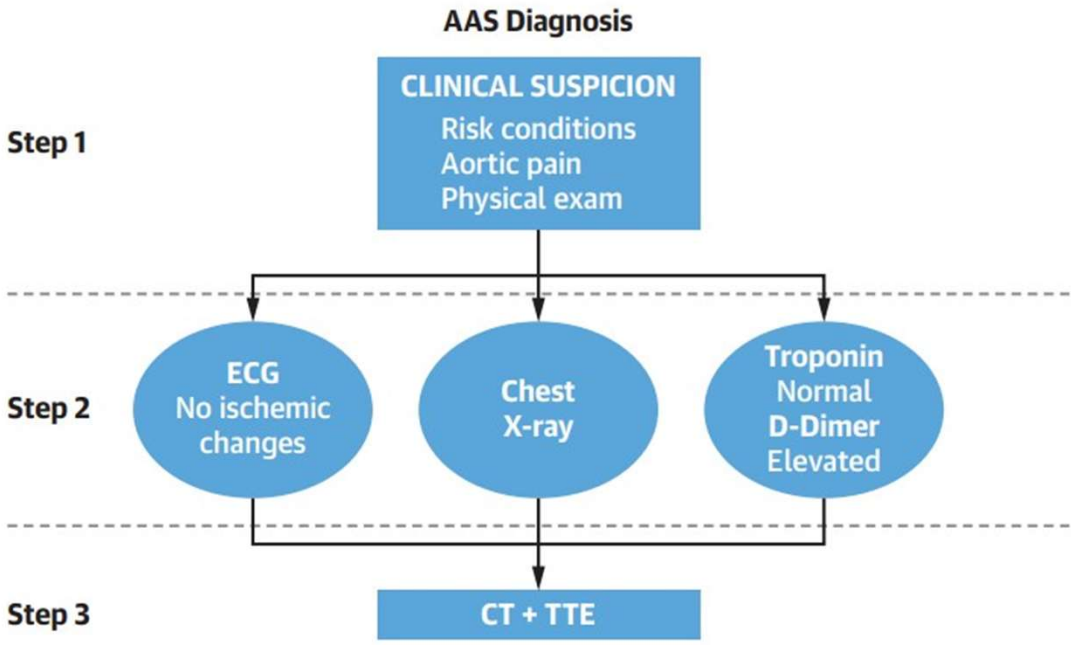
Hypertension
Heritable thoracic aortic conditions and syndromes (see Table 47.2)
Marfan syndrome
Loeys-Dietz syndrome
Vascular Ehlers-Danlos syndrome
Nonsyndromic heritable thoracic aortic disease
Congenital conditions
Bicuspid aortic valve
Coarctation of the aorta
Turner syndrome
Tetralogy of Fallot
Atherosclerosis
Penetrating atherosclerotic ulcer
Trauma, blunt or iatrogenic
Coronary artery bypass grafting/aortic valve replacement/TAVR
Endovascular aneurysm repair (EVAR, TEVAR)
Catheter/guidewire/intra-aortic balloon pump
Aortic/vascular surgery
Motor vehicle accident
Cocaine/methamphetamine use
Inflammatory/infectious diseases
Giant cell arteritis
Takayasu's arteritis
Behçet disease
Aortitis/IgG4-related disease
Syphilis
Pregnancy (with underlying aortopathy)
Weightlifting (with underlying aortopathy)

EVAR, Endovascular aneurysm repair; *TAVR,* transcatheter aortic valve replacement; *TEVAR,* thoracic endovascular aneurysm repair.

TABLE 42.8 Organ System Complications of Acute Aortic Dissection

Cardiovascular	Aortic regurgitation
	Congestive heart failure
	Syncope
	Coronary ischemia or acute myocardial infarction
	Hemopericardium or cardiac tamponade
	Pericarditis
	Cardiac arrest
Pulmonary	Pleural effusion
	Hemothorax
	Hemoptysis (from an aortotracheal or bronchial fistula)
Renal	Renal ischemia or infarction
	Acute renal failure
	Renovascular hypertension
Neurologic	Stroke
	Transient ischemic attack
	Paraparesis or paraplegia
	Encephalopathy
	Coma
	Ischemic neuropathy
Gastrointestinal	Mesenteric ischemia or infarction
	Ileus/abdominal pain
	Pancreatitis
	Hemorrhage (from mucosal ischemia or an aortoenteric fistula)
Peripheral vascular	Upper-or lower-extremity ischemia
Systemic	Fever

FIGURE 5 3-Step Diagnostic Algorithm for Patients With a Suspicion of AAS



Step 1 is devoted to calculating the patient's pretest probability of acute aortic syndrome (AAS) as recommended by the European and U.S. guidelines. Step 2 is a key stage: electrocardiography (ECG), chest x-ray, and laboratory testing including troponins and D-dimers are done. Notice that in patients with "aortic pain," the triad of normal ECG, normal troponins, and increased levels of D-dimers is a warning pattern of AAS. In step 3, the diagnosis of AAS is confirmed or excluded by performing a computed tomographic (CT) scan of the entire aorta. Focused transthoracic echocardiography (TTE) in the emergency room can be useful.

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Acute aortic Syndrome Diagnosis And management

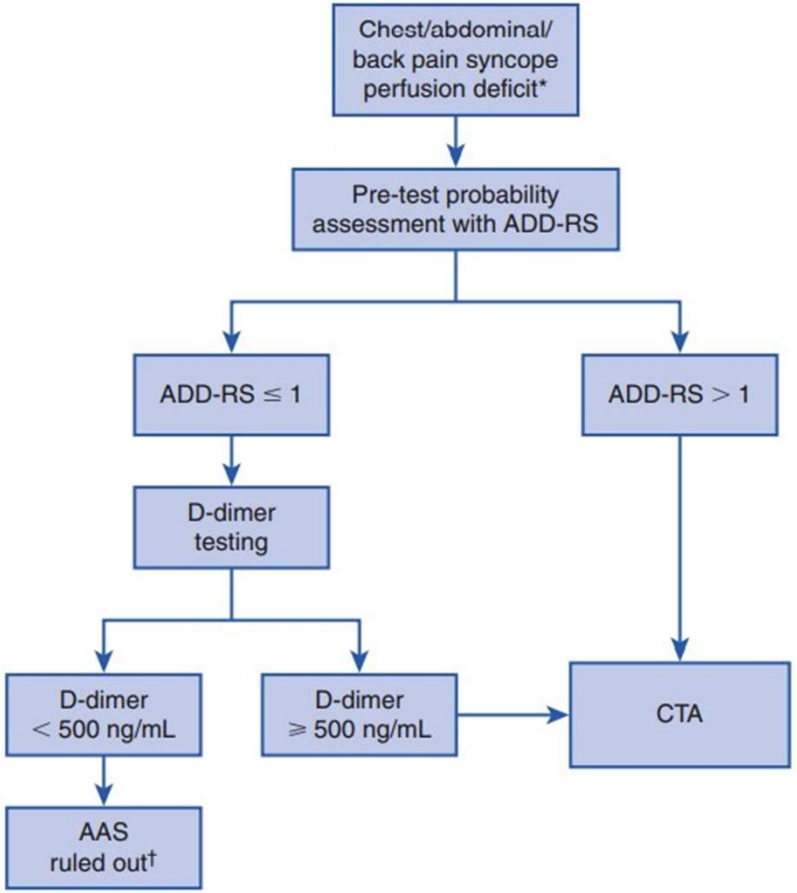

FIGURE 42.17 Proposed diagnostic algorithm for acute aortic dissection based on pretest probability assessment and D-dimer. *AAS*, Acute aortic syndrome; *ADD-RS*, aortic dissection detection risk score (see discussion under "Integrated Diagnostic Evaluation and Management Algorithms" in text); *CTA*, computed tomography angiography. *AAS in differential diagnosis. †Caution in patients with early presentation (≤2 hours) or long-lasting symptoms (≥1 week).

TABLE 1 Imaging Findings That May Influence Clinical Decision Making
Involvement of the ascending aorta
Site of the entrance tear
Severe pericardial effusion/cardiac tamponade
Significant aortic regurgitation and mechanism
Signs of aortic rupture
Signs of end-organ ischemia or malperfusion

TABLE 42.6 Aortic Dissection Classification Based Upon Duration from Symptom Onset

CLASSICAL DEFINITION	TAD GUIDELINES*	IRAD CLASSIFICATION†	ESC GUIDELINES‡	SVS/STS REPORT§
Acute: <14 days	Acute: <14 days	Hyperacute: <24 hr	Acute: <14 days	Hyperacute: <24 hr
Chronic: >14 days	Subacute: <2–6 weeks	Acute: 2–7 days	Subacute: 14–90 days	Acute: 1–14 days
	Chronic: >6 weeks	Subacute: 8–30 days	Chronic: >90 days	Subacute: 15–90 days
		Chronic: >30 days		Chronic: >90 days

*Hiratzka LF, Bakris GL, Beckman JA, et al. Guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. *Circulation*. 2010;121:e266–e369.

†Booher AM, Isselbacher EM, Nienaber CA, et al. The IRAD classification system for characterizing survival after aortic dissection. *Am J Med*. 2013;126:730 e19–e24.

‡Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). *Eur Heart J*. 2014;35:2873–2926.

§Lombardi JV, Hughes GC, Appoo JJ, et al. Society for Vascular Surgery (SVS) and Society of Thoracic Surgeons (STS) reporting standards for Type B aortic dissections. *Ann Thorac Surg*. 2020;109:959–981.

ESC, European Society of Cardiology; *IRAD,* International Registry of Acute Aortic Dissection; *TAD,* thoracic aortic diseases.

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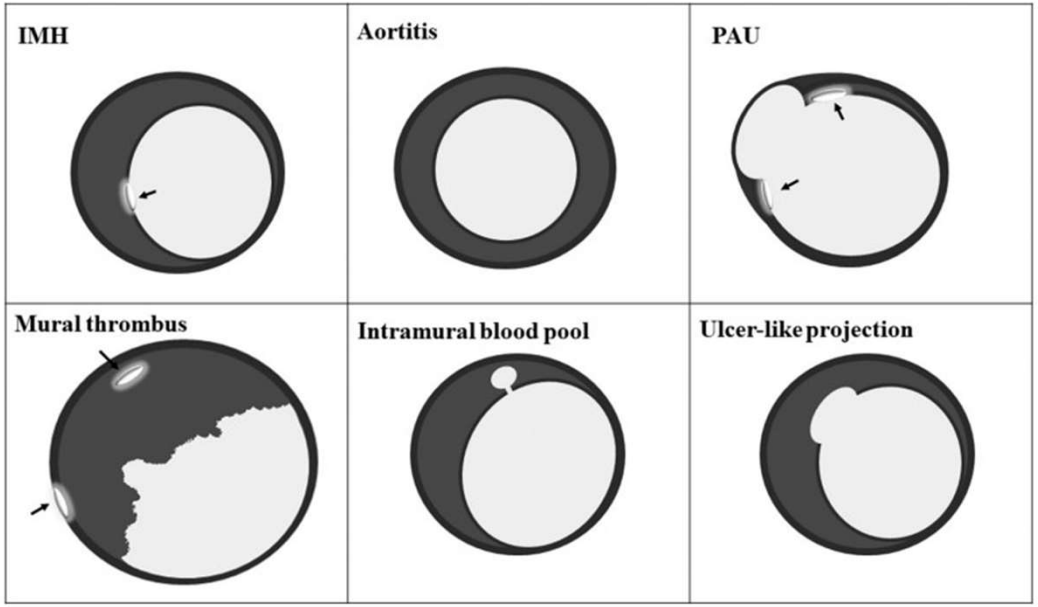
FIGURE 11 Acute Aortic Syndrome Computed Tomography Differential Diagnosis



Schematic representation of computed tomographic (CT) scans of the aorta of patients with acute aortic syndrome (AAS) and related entities. In intramural hematoma (IMH), calcium is displaced inwards, whereas in a dilated aorta with mural thrombus, calcium is located along the outer aorta border or within the thrombus. Aortic wall thickening is crescentic in IMH, whereas it is usually circumferential in aortitis. Ulcer-like projection (ULPs) have a wide communication with the aortic lumen, whereas intramural blood pools (IBPs) have a tiny communication. Both ULPs and IBPs are found in patients with IMH and are not usually accompanied by calcium deposits. Aortic wall deformation and calcified plaques are typically seen in patients with penetrating aortic ulcers (PAUs). Notice the calcium position **(arrows)** in each entity.

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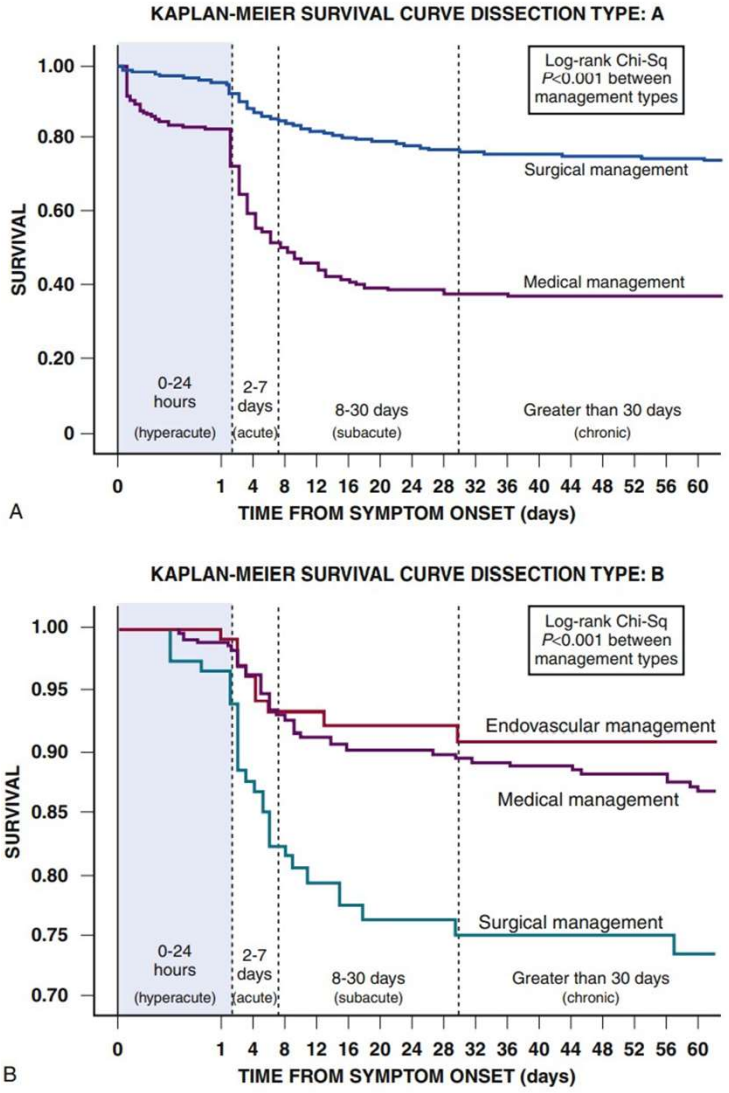

FIGURE 42.14 International Registry of Acute Aortic Dissection (IRAD) classification system of survival after aortic dissection. **A,** Kaplan-Meier survival curve for type A dissection stratified by treatment type. **B,** Kaplan-Meier survival curve for type B dissection stratified by treatment type. (From Booher AM, Isselbacher EM, Nienaber CA, et al. The IRAD classification system for characterizing survival after aortic dissection. *Am J Med*. 2013;126:730 e719–724.)

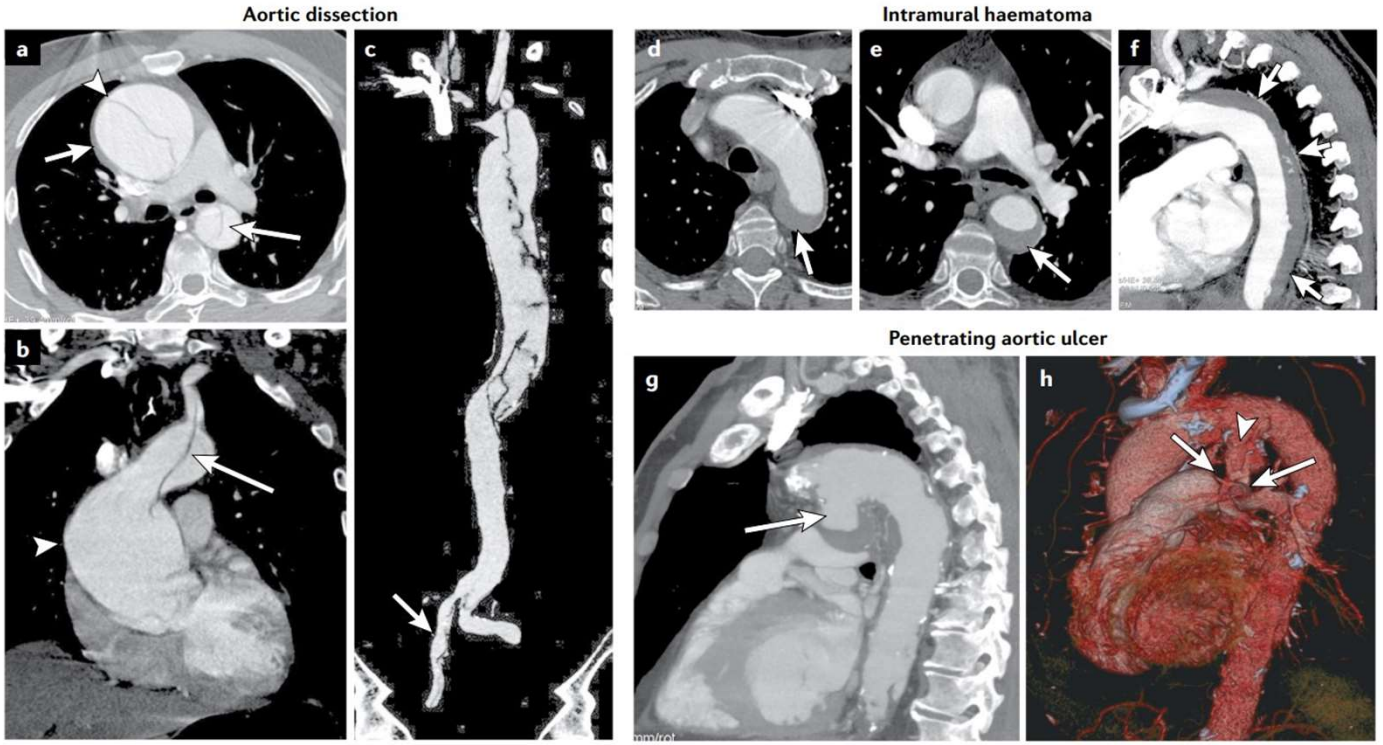

Fig. 5 | **Imaging of acute aortic syndromes. a** | Aortic dissection. Axial contrast-enhanced CT image showing an ascending thoracic aortic aneurysm (TAA; arrowhead) complicated by a type A dissection (arrows) caused by elastic tissue dystrophy in a patient with Marfan syndrome. **b** | Coronal contrast-enhanced CT reconstruction showing an ascending TAA (arrowhead) with an intimal dissection (arrow) that extends into the anonymous trunk lumen in a patient with Marfan syndrome. **c** | Coronal contrast-enhanced CT reconstruction showing an intimal flap inside the aortic lumen that extends into the common right iliac artery lumen (arrow). **d** | Intramural haematoma. Axial contrast-enhanced CT image showing a thoracic aortic arch haematoma with intramural blood collection (arrow). **e** | Axial contrast-enhanced CT image showing a descending thoracic aortic haematoma with intramural semilunar blood collection (arrow). **f** | Sagittal contrast-enhanced CT reconstruction showing a descending thoracic aortic haematoma with intramural blood collection (arrows) with a decreased diameter of the aortic lumen. **g** | Penetrating aortic ulcer. Oblique sagittal contrast-enhanced CT reconstruction showing an aortic arch ulcer (arrow) with out-pouching of contrast medium extending from the aortic lumen into an intramural haematoma. **h** | Volume-rendered, contrast-enhanced CT reconstruction showing an aortic arch ulcer (arrowhead) extending from the aortic lumen into the aortic–pulmonary window (arrows). Images courtesy of L. Romano (A. Cardarelli Hospital, Naples, Italy).

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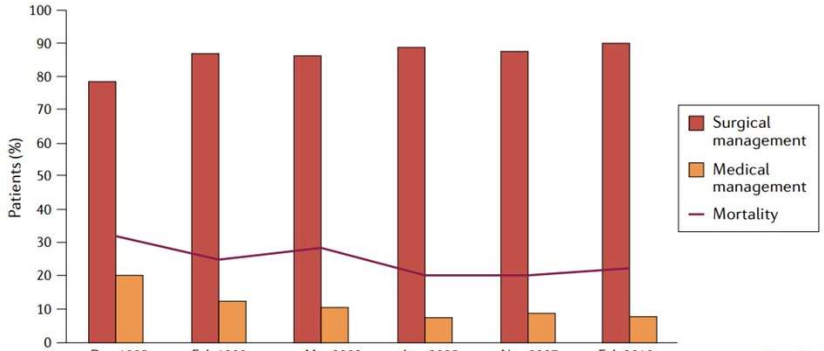
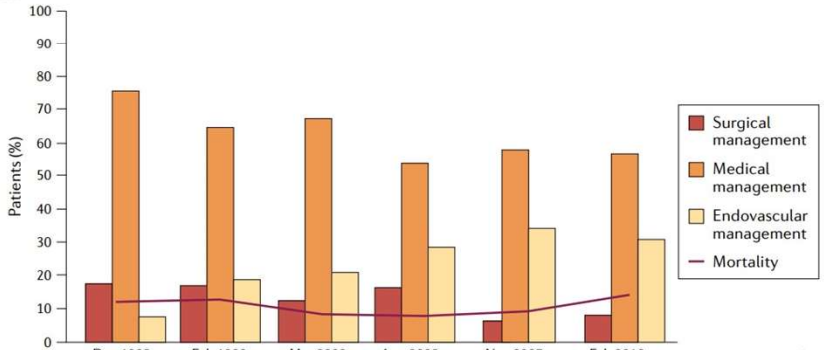
a Type A AAD



Mortality	Dec 1995–Feb 1999	Feb 1999–Mar 2002	Mar 2002–Aug 2005	Aug 2005–Nov 2007	Nov 2007–Feb 2010	Feb 2010–Feb 2013	*P* value	Trend *P* value
Overall	154 (31.4%)	119 (25.3%)	140 (27.6%)	99 (20.8%)	103 (19.8%)	106 (21.7%)	<0.001	<0.001
Surgical	96 (25.0%)	80 (19.6%)	97 (22.1%)	76 (17.9%)	72 (15.8%)	81 (18.4%)	0.015	0.003
Medical	54 (54.5%)	37 (64.9%)	34 (64.2%)	17 (48.6%)	24 (52.2%)	22 (56.4%)	0.512	0.626

b Type B AAD



Mortality	Dec 1995–Feb 1999	Feb 1999–Mar 2002	Mar 2002–Aug 2005	Aug 2005–Nov 2007	Nov 2007–Feb 2010	Feb 2010–Feb 2013	*P* value	Trend *P* value
Overall	30 (12.1%)	34 (12.7%)	19 (8.2%)	20 (7.7%)	20 (9.1%)	35 (14.1%)	0.103	0.915
Surgical	13 (30.2%)	12 (26.7%)	1 (3.6%)	3 (7.0%)	0 (0.0%)	4 (21.1%)	0.003	0.007
Medical	17 (9.1%)	16 (9.2%)	11 (7.1%)	10 (7.1%)	9 (7.1%)	17 (12.1%)	0.630	0.705
Endovascular	0 (0.0%)	6 (12.2%)	7 (14.6%)	7 (9.5%)	10 (13.3%)	12 (15.6%)	0.528	0.216

Fig. 8 | **Aortic dissection: management and mortality over time in the IRAD.** The International Registry of Acute Aortic Dissection (IRAD) analysed trends in mortality among patients with type A or type B acute aortic dissection (AAD) over 17 years (December 1995 to February 2013)[7,85]. **a** | In-hospital mortality for type A AAD decreased significantly from 31% to 22%, mainly owing to decreased surgical mortality (25% to 18%; $P = 0.003$). **b** | In-hospital mortality for type B AAD did not change significantly (12% to 14%). Adapted with permission from REF.[85], Elsevier.

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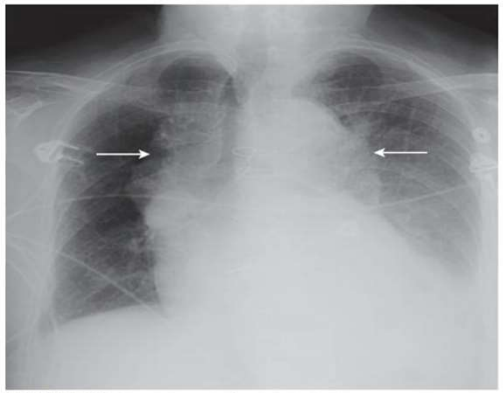


EFIGURE 42.19 Chest radiograph of an individual with acute type A aortic dissection demonstrating a widened mediastinum and enlargement of the ascending and descending aortic shadows *(arrows)*.

ETABLE 42.1 Comparison of CTA, MRA, TTE, and TEE for the Diagnosis of Aortic Dissection

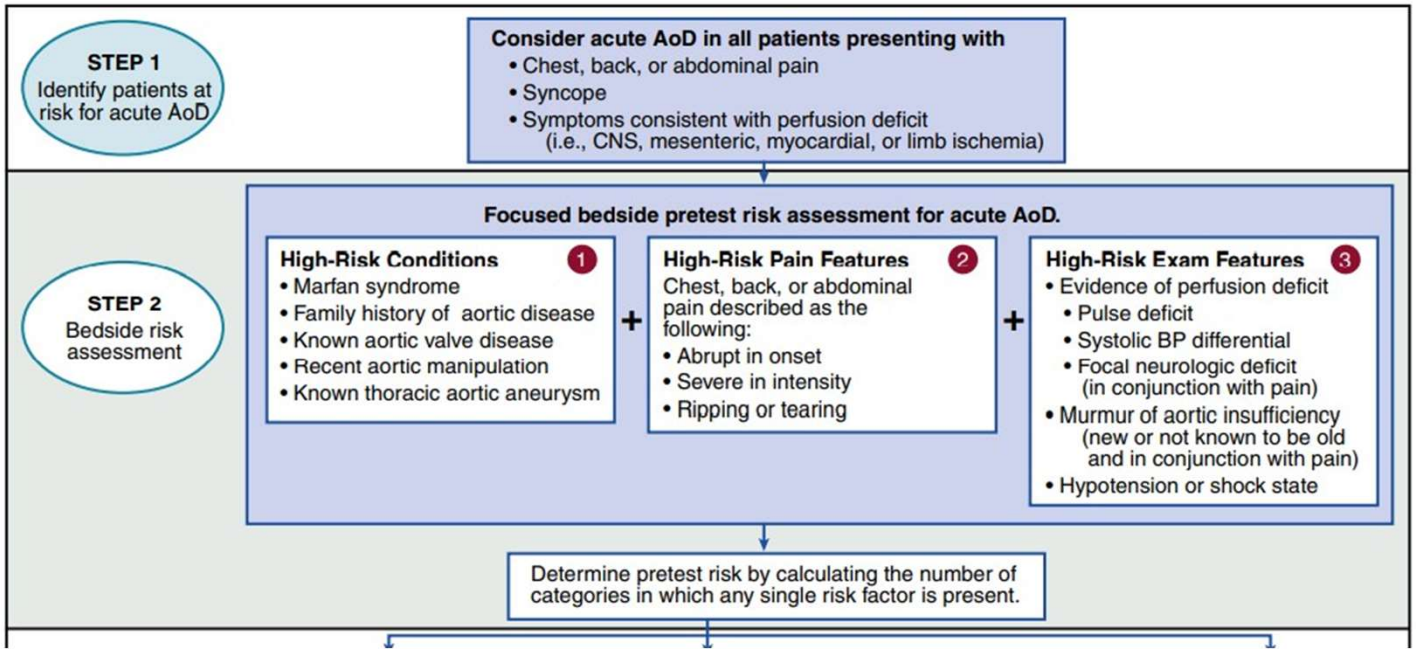
	CTA	MRA	TTE	TEE
Sensitivity	+++	+++	+	++
Specificity	+++	+++	++	+++
Patient tolerability	+++	++	+++	++
Speed	+++	+	+++	++
Risk of complications	Low to moderate	Low to moderate	Negligible	Low to moderate
Availability	+++	+	+++	++
Assessment of myocardial function	+	+++	+++	+++
Assessment of aortic regurgitation	N/A	+++	++	+++
Characterization of branch vessel involvement	+++	+++	+	+

+, fair; ++, good; +++, excellent; N/A, not applicable.
CTA, Computed tomographic angiography; *MRA,* magnetic resonance angiography; *TEE,* transesophageal echocardiography; *TTE,* transthoracic echocardiography.
From Bhave NM, Nienaber CA, Clough RE, Eagle KA. Multimodality imaging of thoracic aortic diseases in adults. *JACC Cardiovasc Imaging.* 2018;11(6):902–919.

ETABLE 42.2 Diagnostic Information from Imaging of Acute Aortic Dissection

1. Establish the presence of aortic dissection or variant (aortic IMH, PAU)
2. Location of the dissection (type A, B)
3. Anatomic features
   a. Extent of dissection
   b. Sites of entry and reentry, entry tear diameter
   c. False lumen patency and diameter
4. Complications of dissection
   a. Type A
      i. Aortic regurgitation
      ii. Coronary artery involvement
      iii. Pericardial effusion/hemopericardium
   b. Aortic rupture or leakage
   c. Branch vessel involvement
   d. Malperfusion
   e. Aneurysmal enlargement
   f. High-risk features of IMH (ulcer-like projections, hematoma diameter, hemopericardium)

IMH, Intramural hematoma; *PAU,* Penetrating atherosclerotic aortic ulcer.


STEP 1
Identify patients at risk for acute AoD
Consider acute AoD in all patients presenting with
• Chest, back, or abdominal pain
• Syncope
• Symptoms consistent with perfusion deficit (i.e., CNS, mesenteric, myocardial, or limb ischemia)
STEP 2
Bedside risk assessment
Focused bedside pretest risk assessment for acute AoD.
High-Risk Conditions 1
• Marfan syndrome
• Family history of aortic disease
• Known aortic valve disease
• Recent aortic manipulation
• Known thoracic aortic aneurysm
+
High-Risk Pain Features 2
Chest, back, or abdominal pain described as the following:
• Abrupt in onset
• Severe in intensity
• Ripping or tearing
+
High-Risk Exam Features 3
• Evidence of perfusion deficit
• Pulse deficit
• Systolic BP differential
• Focal neurologic deficit (in conjunction with pain)
• Murmur of aortic insufficiency (new or not known to be old and in conjunction with pain)
• Hypotension or shock state
Determine pretest risk by calculating the number of categories in which any single risk factor is present.

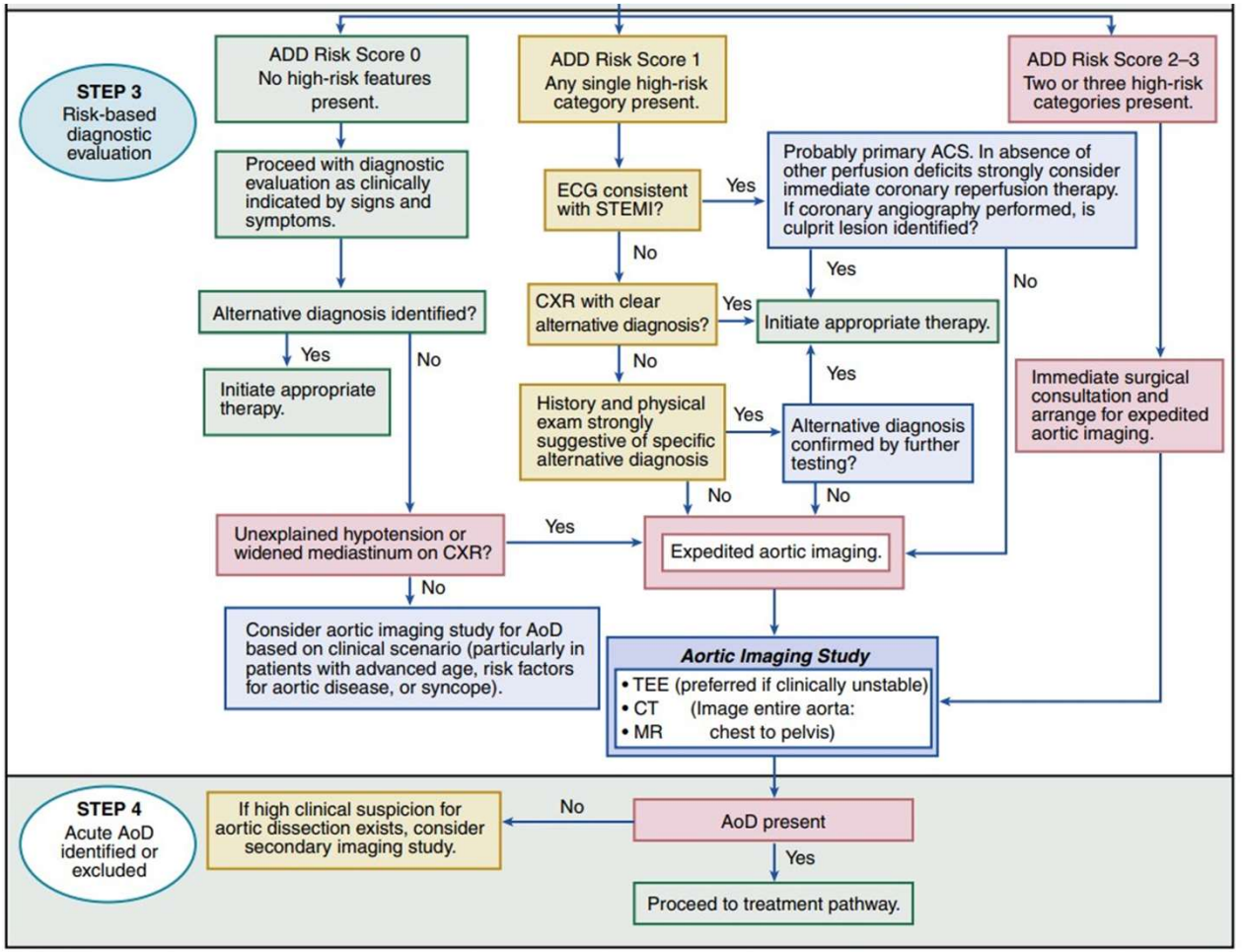
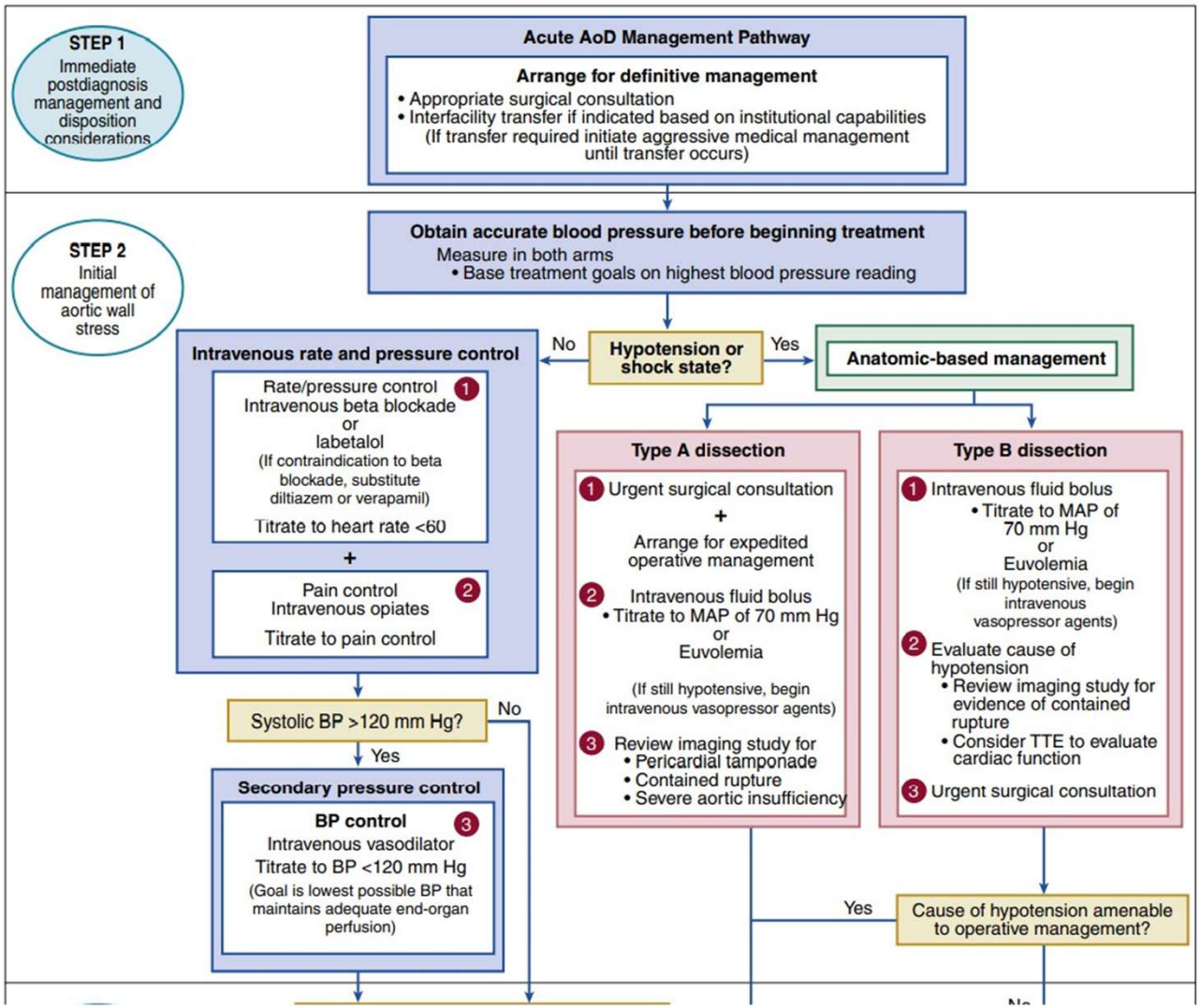


FIGURE 42.19 Evaluation pathway for aortic dissection (AoD). *ACS,* Acute coronary syndrome; *ADD,* aortic dissection detection; *BP,* blood pressure; *CNS,* central nervc system; *CXR,* chest x-ray; *STEMI,* ST-segment elevation myocardial infarction. (Modified from 2010 American College of Cardiology/American Heart Association Thoracic Aor disease guidelines. From Rogers AM, Hermann LK, Booher AM, et al. Sensitivity of the aortic dissection detection risk score, a novel guideline-based tool for identification of acu aortic dissection at initial presentation: results from the international registry of acute aortic dissection. *Circulation*. 2011;123:2213.)


STEP 1
Immediate postdiagnosis management and disposition considerations
Acute AoD Management Pathway
Arrange for definitive management
• Appropriate surgical consultation
• Interfacility transfer if indicated based on institutional capabilities
(If transfer required initiate aggressive medical management until transfer occurs)
STEP 2
Initial management of aortic wall stress
Obtain accurate blood pressure before beginning treatment
Measure in both arms
• Base treatment goals on highest blood pressure reading
Hypotension or shock state?
No
Yes
Anatomic-based management
Intravenous rate and pressure control
1 Rate/pressure control
Intravenous beta blockade
or
labetalol
(If contraindication to beta blockade, substitute diltiazem or verapamil)
Titrate to heart rate <60
+
2 Pain control
Intravenous opiates
Titrate to pain control
Systolic BP >120 mm Hg?
No
Yes
Secondary pressure control
3 BP control
Intravenous vasodilator
Titrate to BP <120 mm Hg
(Goal is lowest possible BP that maintains adequate end-organ perfusion)
Type A dissection
1 Urgent surgical consultation
+
Arrange for expedited operative management
2 Intravenous fluid bolus
• Titrate to MAP of 70 mm Hg
or
Euvolemia
(If still hypotensive, begin intravenous vasopressor agents)
3 Review imaging study for
• Pericardial tamponade
• Contained rupture
• Severe aortic insufficiency
Type B dissection
1 Intravenous fluid bolus
• Titrate to MAP of 70 mm Hg
or
Euvolemia
(If still hypotensive, begin intravenous vasopressor agents)
2 Evaluate cause of hypotension
• Review imaging study for evidence of contained rupture
• Consider TTE to evaluate cardiac function
3 Urgent surgical consultation
Yes
Cause of hypotension amenable to operative management?

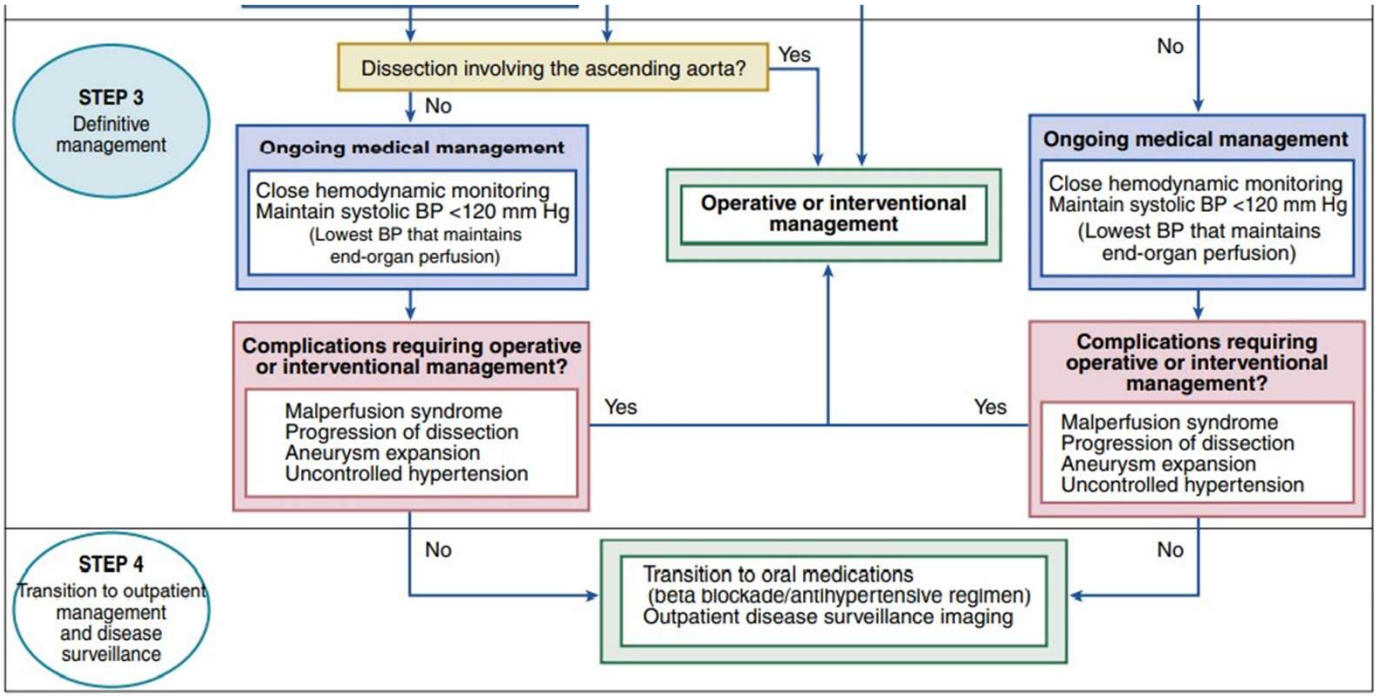


FIGURE 42.20 Management pathway for acute aortic dissection. *AoD,* Aortic dissection; *BP,* blood pressure; *MAP,* mean arterial pressure. (From Hiratzka LF, Bakris G Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: Executiv summary. A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surger American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. *J Am Coll Cardiol*. 2010;55:e27–e129.)

TABLE 42.9 Selected Pharmacologic Therapy for Acute Aortic Dissection*

Intravenous Beta-Blocker (Preferred Negative Inotrope)

- **Esmolol**: Bolus 250–500 μg/kg IV, then continuous IV infusion at 50–100 μg/kg/min, titrated to effect with maximum dose of 200 μg/kg/min
- **Labetalol**: Bolus 20 mg IV over 2 min, then 20–80 mg IV every 10 min until adequate response (maximum 300 mg), then continuous IV infusion at 0.5–2 mg/min IV, titrated to effect
- **Metoprolol**: Give 2.5–15 mg IV followed by 2.5–15 mg IV every 3–6 hr

Intravenous Calcium Channel Blocker (Secondary Negative Inotrope)

- **Diltiazem**: Initial bolus of 0.25–0.35 mg/kg IV, then continuous IV infusion of 5–20 mg/hr
- **Verapamil**: 5–10 mg IV and may repeat after 5–10 min

Intravenous Vasodilator (*After* Initiation of Negative Inotrope)

- **Clevidipine**: Give 1–2 mg/hr; can double dose up to maximum dose of 16–32 mg/hr
- **Sodium nitroprusside**: Start continuous infusion at 0.25–0.5 μg/kg/min, titrated to a maximum of 8–10 μg/min. Use only in presence of beta blockers
  - *Caution*: Thiocyanate toxicity may occur in patients with renal impairment or prolonged infusions
- **Nicardipine**: Give 2.5–5 mg/hr and titrate up to a maximum of 15 mg/hr
- **Nitroglycerin**: Initial 5 μg/min; up to 200 μg/min as an IV infusion
- **Enalaprilat**: Give 1.25 mg, then 1.25–5 mg IV every 6 hr, titrated to effect
- **Fenoldopam**: Give 0.1 μg/kg/min and titrate up to a maximum of 1.6 μg/kg/min

*Goal of therapy is heart rate less than 70 beats/min and blood pressure 100–120 mm Hg or as low as possible without compromising organ perfusion.

Type B dissection
And
IMH

TABLE 2 Main Complications in Type B Acute Aortic Syndrome
Hemodynamic instability (hypotension-shock)
Signs of rapid aortic expansion
Malperfusion syndrome
Signs of aortic rupture or impending rupture (periaortic hematoma)

TABLE 3 Risk Factors for Poor Outcome in Type A Intramural Hematoma
Existence of ulcer-like projection
Aortic diameter of ≥50 mm
Rapid enlargement of the aortic diameter
Hematoma thickness of ≥11 mm
Moderate-severe pericardial effusion/cardiac tamponade
Significant pleural effusion
Periaortic hematoma/signs of impending rupture
Significant aortic regurgitation
Hemodynamic instability (hypotension-shock)
Refractory pain
Malperfusion syndrome

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TABLE 42.10 Indications for Thoracic Endovascular Aortic Repair for Complicated Type B Aortic Dissection (or Open Surgical Repair if Anatomy Is Unsuitable for Thoracic Endovascular Aortic Repair)

Rupture/Impending rupture
Malperfusion
Hemothorax
Refractory pain
Refractory hypertension
Aneurysmal dilatation (>55 mm)
Rapid increase in aortic diameter
Recurrent symptoms

42 Diseases of the Aorta
ALAN C. BRAVERMAN AND MARC SCHERMERHORN

Cardio_Cast

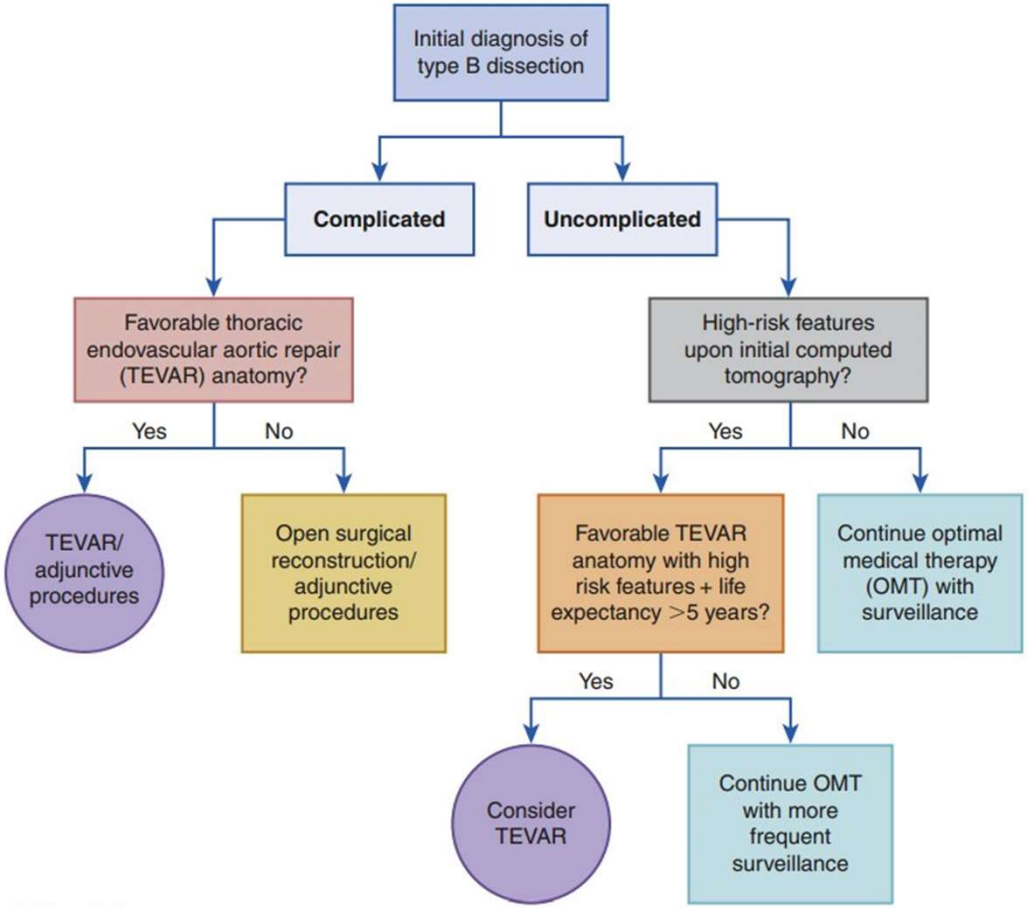
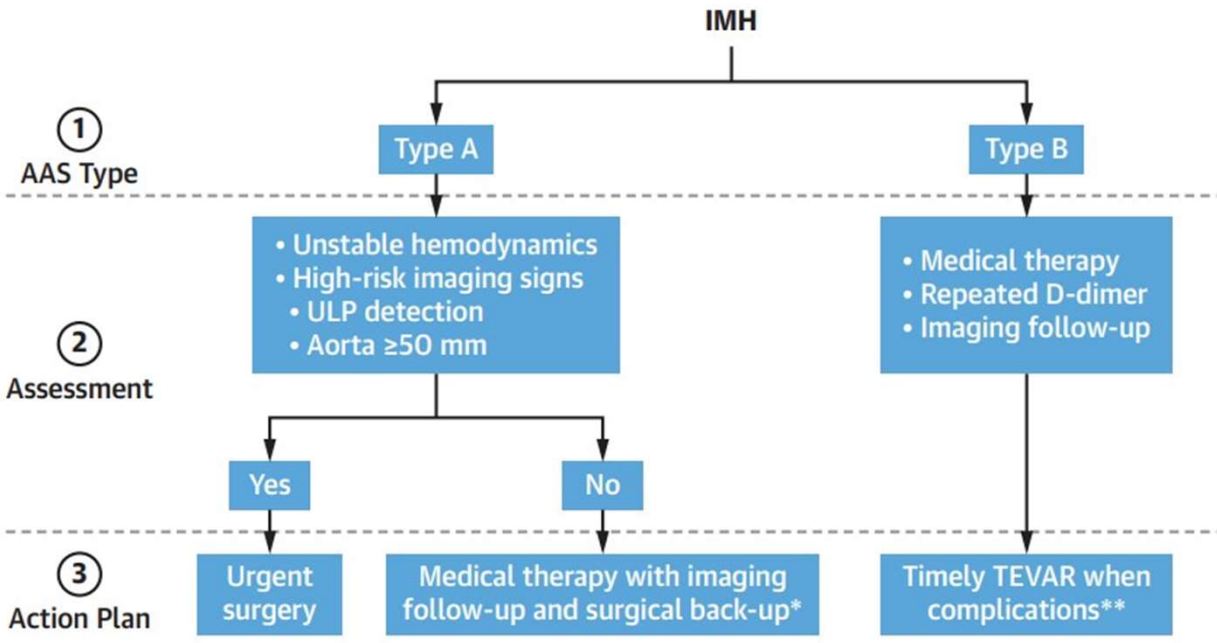


FIGURE 42.22 Management algorithm for acute type B aortic dissection. Adjunctive procedures are, for example, endovascular fenestration, surgical revascularization, coil embolization, and selective branch vessel stenting. High-risk features include primary entry tear diameter greater than 10 mm, initial aortic diameter greater than 40 mm, false lumen diameter greater than 22 mm, partially thrombosed false lumen, and saccular false lumen formation (see eTable 42.4). *OMT,* Optimal medical therapy; *TEVAR,* thoracic endovascular aortic repair. (From Tadros RO, Tang GHL, Barnes HJ, et al. Optimal treatment of uncomplicated Type B aortic dissection: JACC review topic of the week. *J Am Coll Cardiol.* 2019;74:1494–1504.)

FIGURE 13 Management of Patients With Intramural Hematoma



Sequential 3-step treatment algorithm for patients with intramural hematoma (IMH). The first step, to determine the involvement (type A) or not (type B) of the ascending aorta, is critical. In the second step, the clinical status, imaging findings, and laboratory results indicate the best therapeutic approach (third step). *Especially in the elderly and patients with comorbidities. **IMH expansion; progressive aortic dilation; periaortic hematoma; localized aortic dissection. TEVAR = thoracic endovascular aortic repair.

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AAS long-term follow-up

Box 2 | **Acute aortic syndromes and aneurysms: long-term follow-up**

Clinical and imaging follow-up

Acute aortic syndrome[2,3]

- 1, 3, 6, 9 and 12 months and yearly thereafter
- First choice: TTE[a] and MRI
- Second choice: TTE[a] and CT angiography

Thoracic aortic aneurysm[11]

- Open repair: CT angiography after 6 months then yearly (CT angiography or MRI); after 3 years of follow-up, the interval can be extended to every 2–3 years.
- Thoracic endovascular repair: CT angiography between 1 and 12 months after the procedure, then yearly (CT angiography or MRI); after 3 years of follow-up, the interval can be extended to every 2–3 years.

Abdominal aortic aneurysm[12,13]

- Open repair: abdominal and pelvic imaging (CT angiography) is recommend every 5 years.
- Endovascular aneurysm repair: abdominal and pelvic imaging (CT angiography and A-US) after 1 month; in the absence of an endoleak or sac enlargement, imaging should be repeated in 12 months (CT angiography or A-US); thereafter, abdominal and pelvic CT angiography should be performed every 5 years.

Cardiovascular risk-factor modifications and healthy lifestyle targets

Blood pressure and heart rate[91]

- Blood pressure <120/80 mmHg
- Heart rate <60 bpm
- First-line therapy: β-blockers
- Second-line therapy: angiotensin-converting enzyme inhibitors or angiotensin-receptor blockers
- Third-line therapy: calcium-channel blockers (long-acting dihydropyridines)

Plasma LDL-cholesterol level[122]

- Patients at very high risk: <1.4 mmol/l (<55 mg/dl) or a reduction by ≥50% from baseline
- First-line therapy: statins
- Second-line therapy: statins plus ezetimibe
- Third-line therapy: statins plus ezetimibe plus PCSK9 inhibitor

Educational programmes and psychological support[125–128]

- Optimization of and adherence to medical treatment
- Healthy diet (low in saturated fat, with a focus on whole-grain products, vegetables, fruit and fish)
- BMI 20–25 kg/m^2
- Waist circumference <94 cm (men) or <80 cm (women)
- Avoid cocaine and other stimulating drugs, such as methamphetamine
- Drive carefully and wear a seatbelt
- No exposure to tobacco in any form
- Avoid excessive alcohol intake
- Annual influenza vaccination
- Genetic counselling

Exercise training[125–127]

- Mild to moderate aerobic exercise (3–5 METs) can be performed for ≥30 min on most days of the week, for a total of 150 min per week (walking, slow jogging and recreational cycling).
- Avoid strenuous physical activities (isometric exercise, pushing or straining that would require a Valsalva manoeuvre, carrying heavy objects) and contact sports (such as competitive football or ice hockey).
- Common-sense approach to sexual activity: avoid straining and maximal exertion.
- Participation in a cardiac rehabilitation programme is advised (in-hospital, at a community centre and/or home-based).

A-US, abdominal ultrasonography; MET, metabolic equivalent of task; TTE, 2D transthoracic colour–Doppler echocardiography. [a]Multiview cardiovascular ultrasonography: transthoracic, suprasternal and abdominal.

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