

Aorta. Dissecção, Aneurismas, Inflamatórias 2018

- Aula apresentada no Curso Auxiliar Preparatório ao Título de Especialista em Cardiologia da Sociedade Brasileira de Cardiologia
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Aortopatias, dissecação e aneurisma

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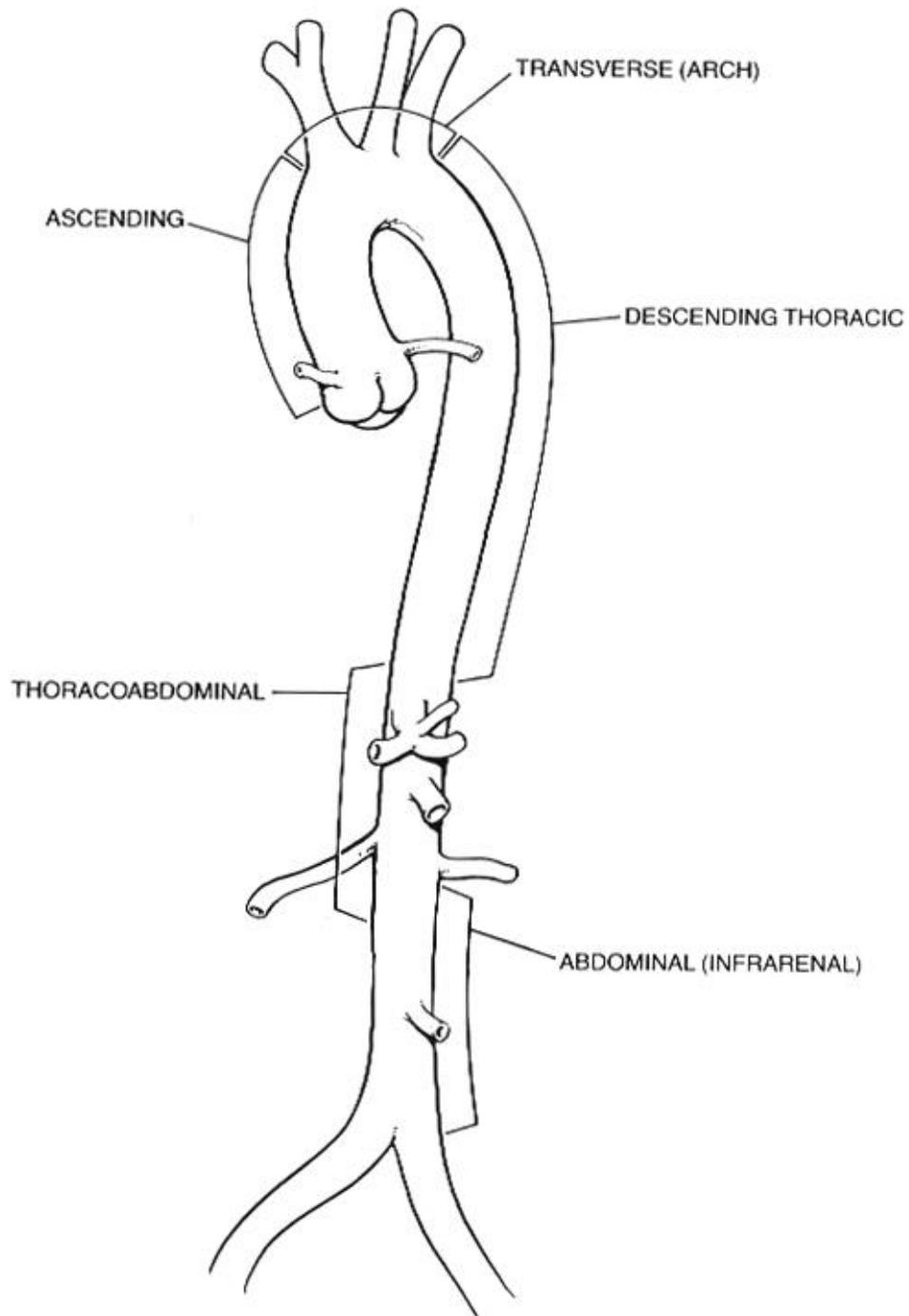
Declaração de potencial conflito de interesses

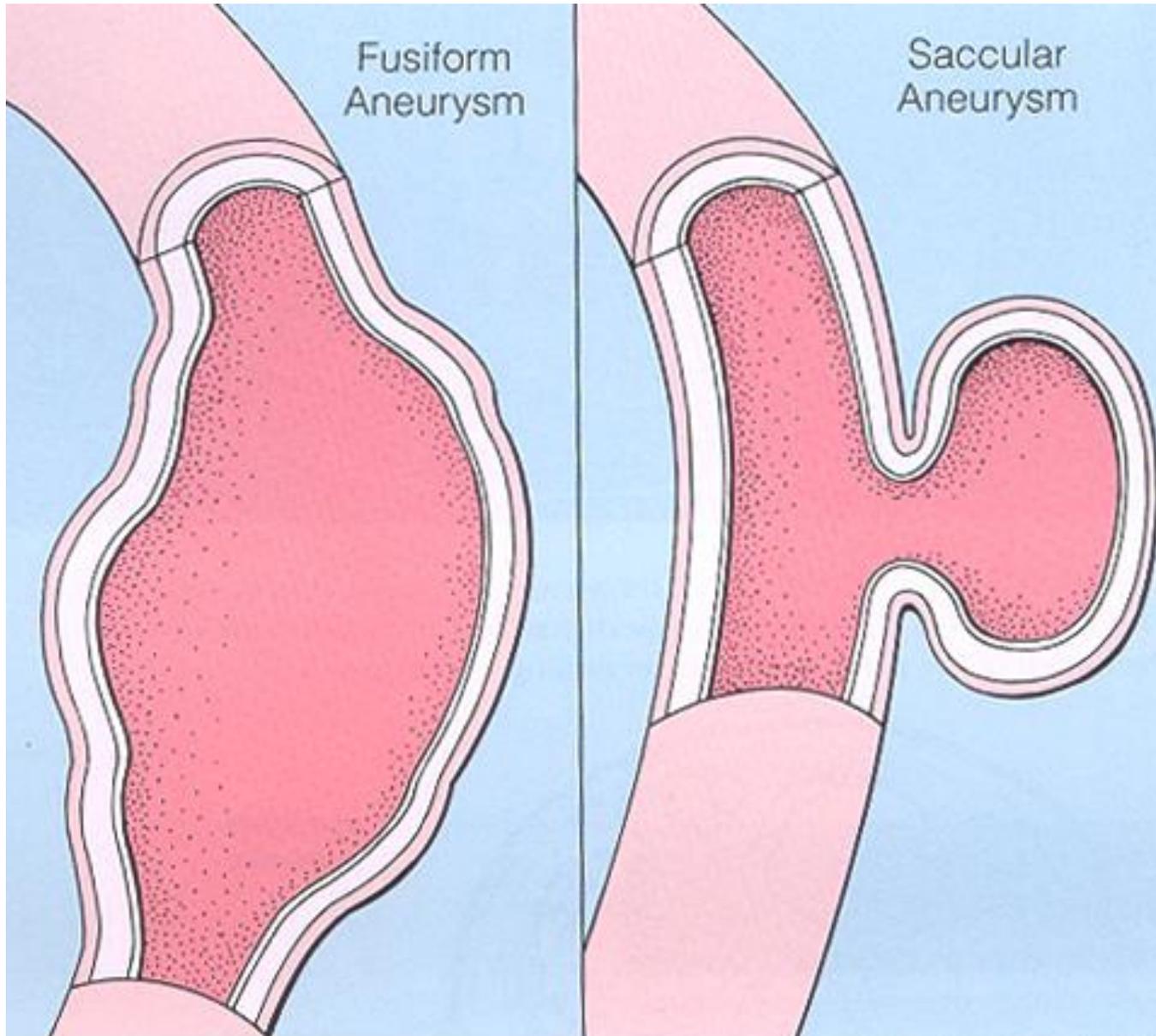
Aortopatias, dissecação e aneurisma

Não tenho conflitos de interesses relacionados a esta apresentação

Tópicos

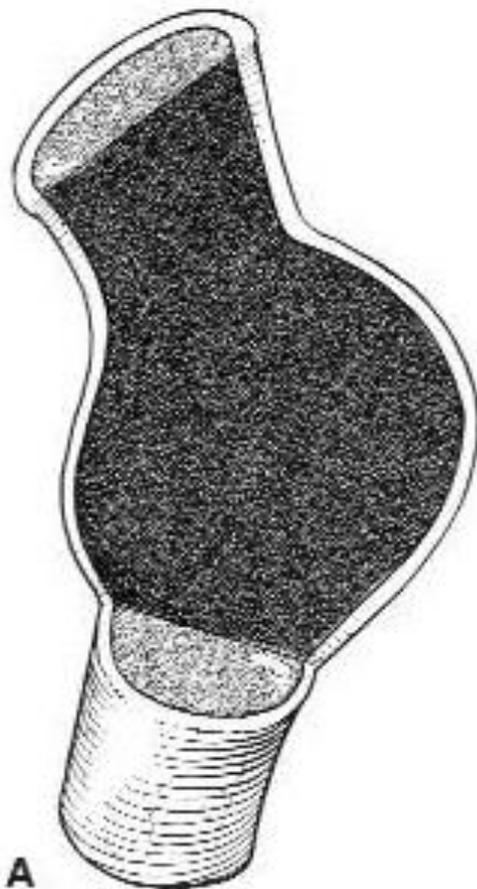
- *Síndromes aórticas agudas (dissecção da aorta, hematoma intramural e úlcera penetrante aterosclerótica)*
- *Aneurismas da aorta torácica*
- *Aortopatias inflamatórias (Takayasu, conectivopatias)*
- *Ateroembolismo*



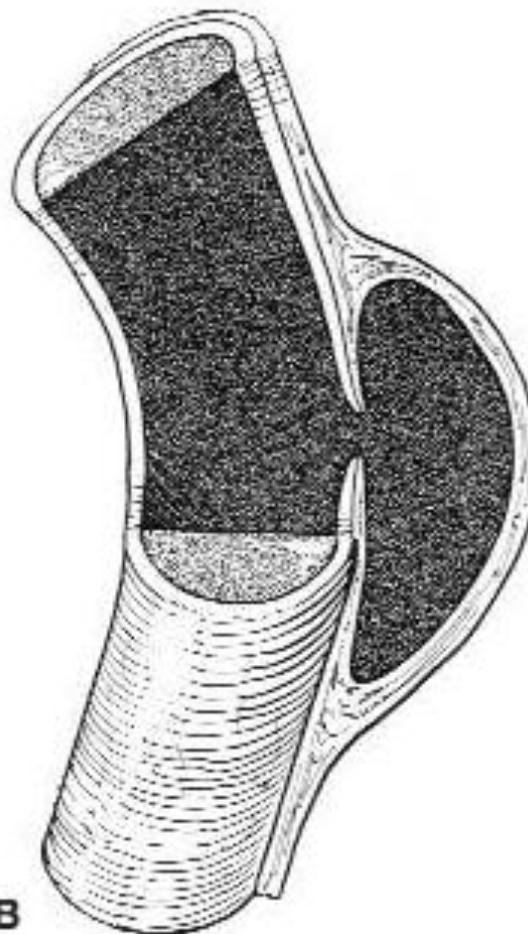


DISSECÇÃO

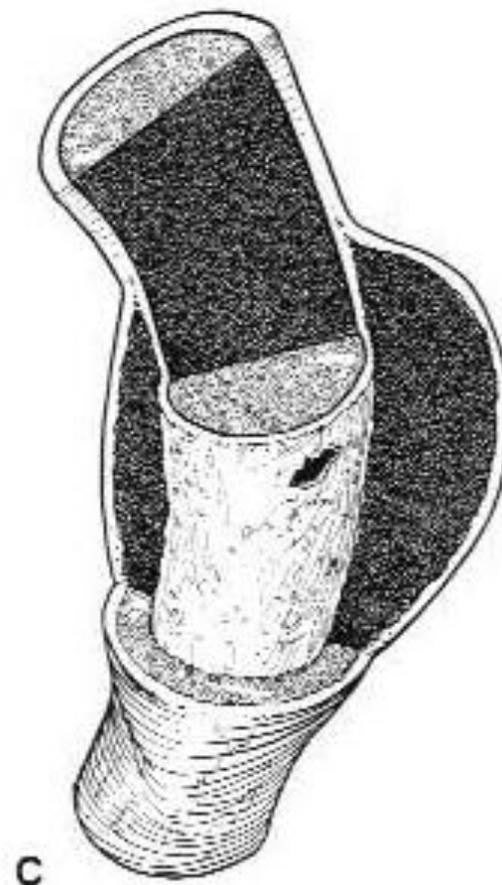
TRUE



FALSE



DISSECTING



Acute aortic syndromes

Acute aortic syndromes are a constellation of life-threatening medical conditions, including classic acute aortic dissection (AAD), intramural haematoma (IMH), penetrating atherosclerotic aortic ulcer (PAU) (even thoracic aortic rupture), which share common pathophysiological pathways (breakdown of the intima and media), clinical characteristics, and diagnostic and therapeutic challenges.^{2,9}

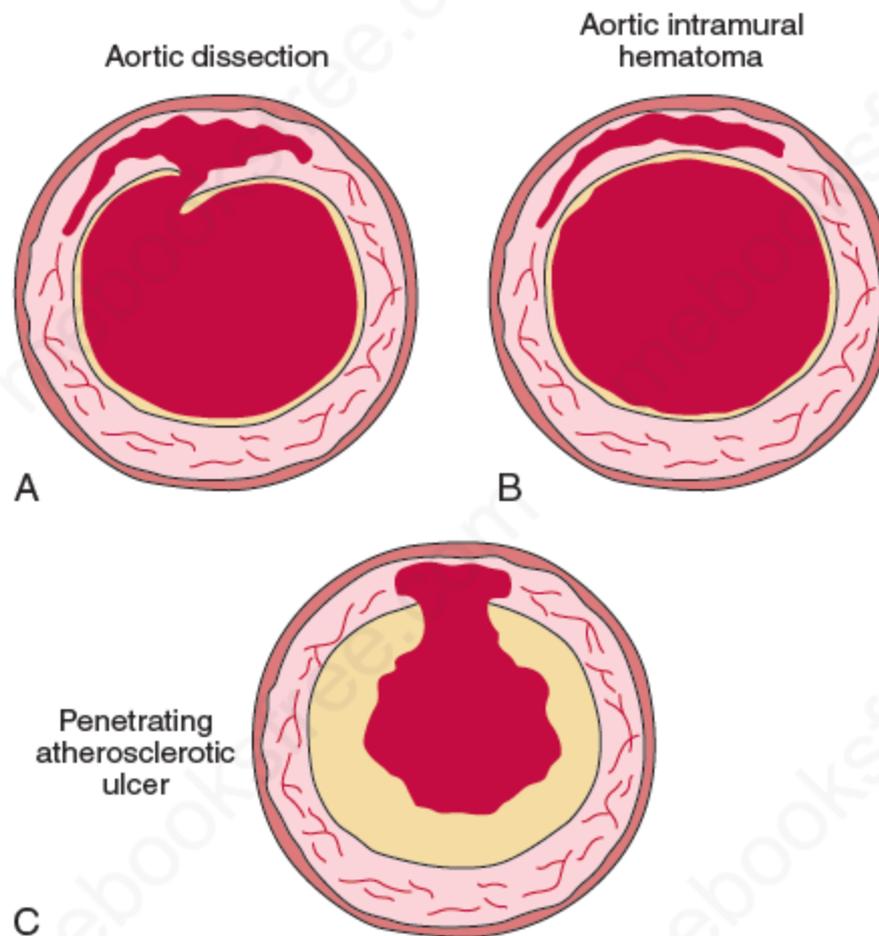
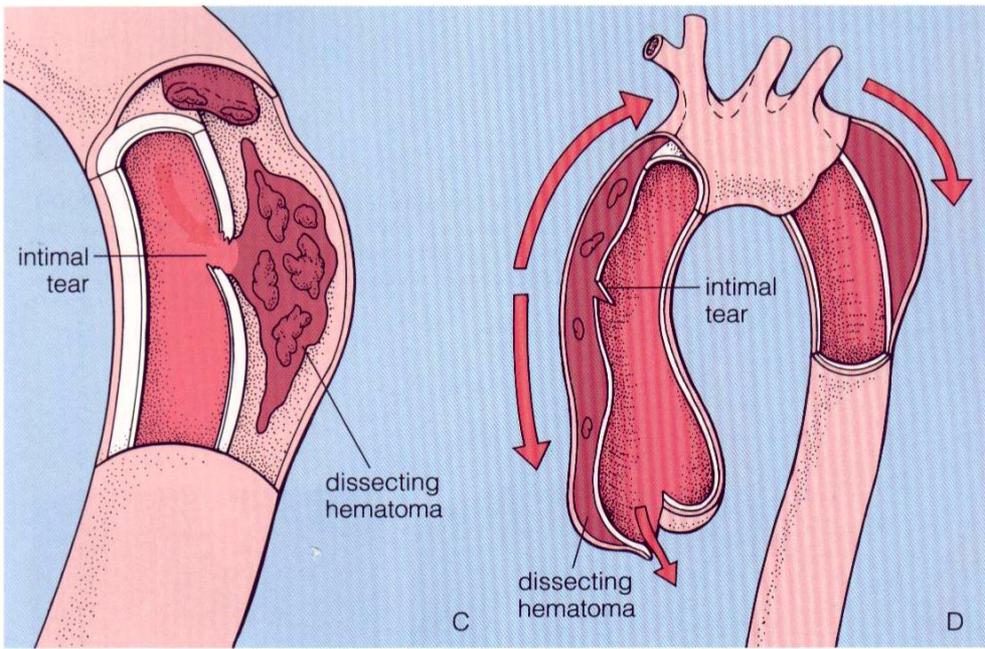
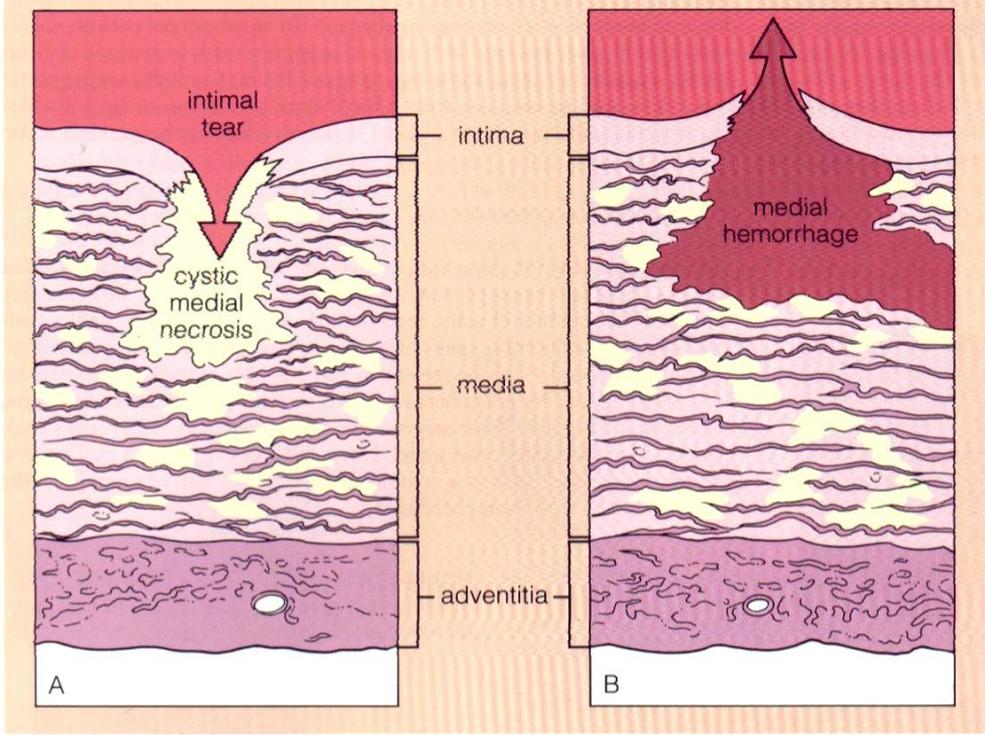
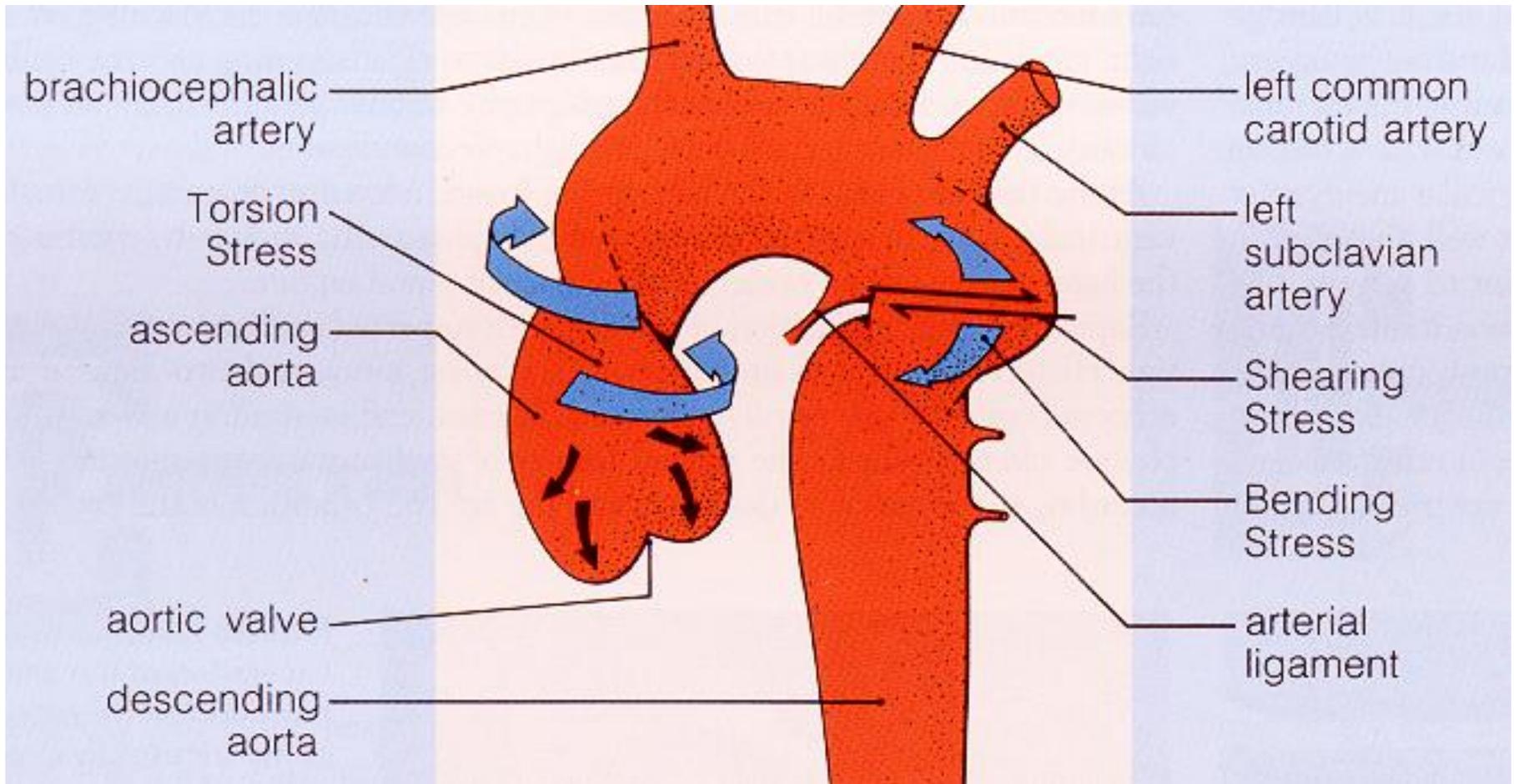


FIGURE 63.10 Acute aortic syndromes. **A**, Classic aortic dissection. There is a tear in the intima with blood entering the media and a dissecting cleavage plane propagating for variable distances anterograde (and occasionally retrograde) throughout the aortic wall. **B**, Aortic intramural hematoma (IMH). A spontaneous hemorrhage of the vasa vasorum leads to bleeding within the media in the absence of an intimal tear or intimal flap. **C**, Penetrating atherosclerotic aortic ulcer (PAU). An ulcerated aortic plaque ruptures into the media, leading to an outpouching or ulceration in the aortic wall. This may be associated with IMH formation, pseudoaneurysm, or a focal, thick-walled aortic dissection.





Síndromes Aórticas Agudas

DISSECÇÃO AGUDA DE AORTA

- ***Evento súbito, que se caracteriza pela separação das camadas da aorta, geralmente com ruptura da íntima, sendo o evento mais catastrófico que acomete a aorta***
- ***Exige diagnóstico ágil e intervenção adequada***

DISSECÇÃO AGUDA DE AORTA

EPIDEMIOLOGIA

- ***2-3x mais freqüente que ruptura de AnAoAb***
- ***9000 / ano nos EUA (10/100.000 hab/ano)***
- ***5:1 H / M***
- ***2 mortes / 100.000 hab / ano***
- ***5ª – 6ª década***

DISSECÇÃO AGUDA DE AORTA

FATORES PREDISPONENTES

- ***HAS, HAS, HAS***
- ***Doenças do tecido conjuntivo
(Marfan, Turner, Ehlers-Danlos)***
- ***Necrose cística da aorta***
- ***Rins policísticos***
- ***V. Ao bicúspide, CoAo, Aortite***
- ***Gravidez***
- ***Aterosclerose***
- ***Trauma / Iatrogenia***

Hypertension
Heritable or genetic thoracic aortic disease and syndromes (see Table 63.1)
Marfan syndrome
Loeys-Dietz syndrome
Familial thoracic aortic aneurysm syndromes
Vascular Ehlers-Danlos syndrome
Turner syndrome
Congenital diseases/syndromes
Bicuspid aortic valve
Coarctation of the aorta
Tetralogy of Fallot
Atherosclerosis
Penetrating atherosclerotic ulcer
Trauma, blunt or iatrogenic
Catheter/guidewire
Intra-aortic balloon pump
Aortic/vascular surgery
Motor vehicle accident
Coronary artery bypass grafting/aortic valve replacement/TAVR
Thoracic endovascular aneurysm repair (TEVAR)
Cocaine/methamphetamine use
Inflammatory/infectious diseases
Giant cell arteritis
Takayasu arteritis
Behçet disease
Aortitis
Syphilis
Pregnancy (with underlying aortopathy)
Weightlifting (with underlying aortopathy)

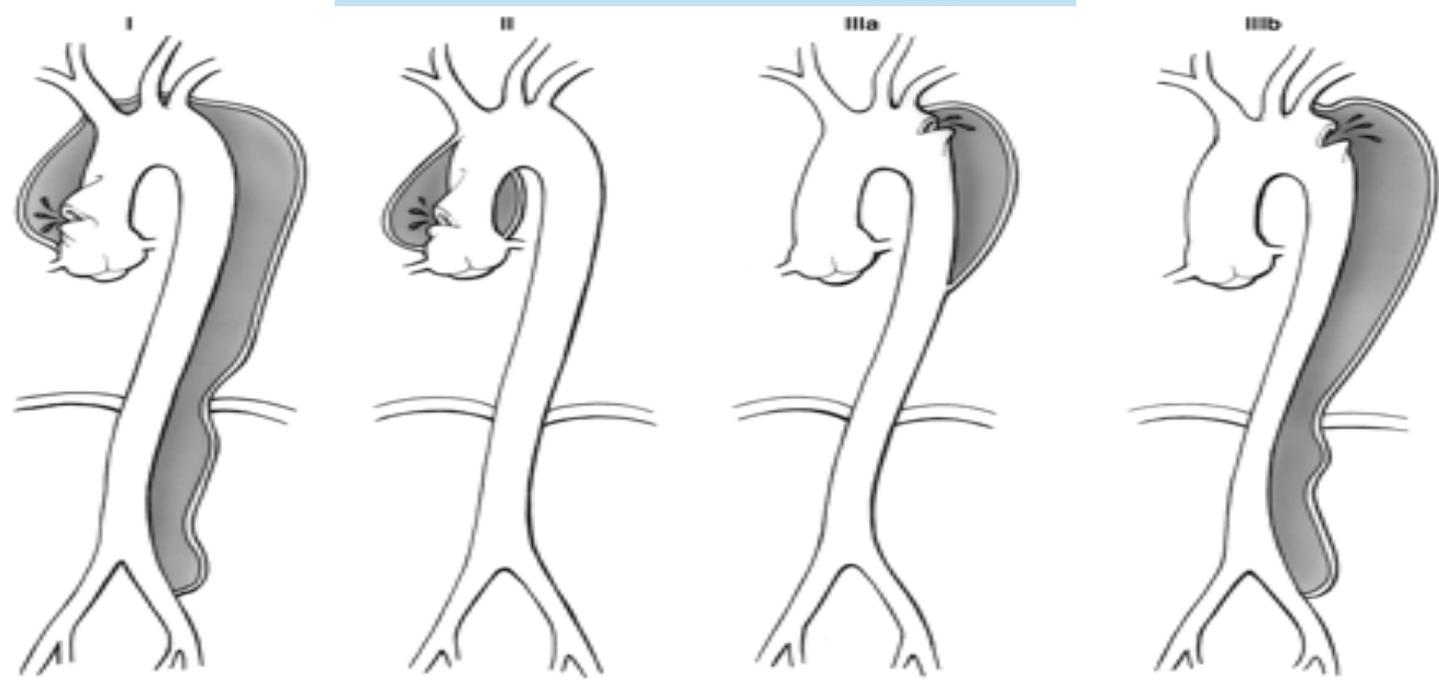
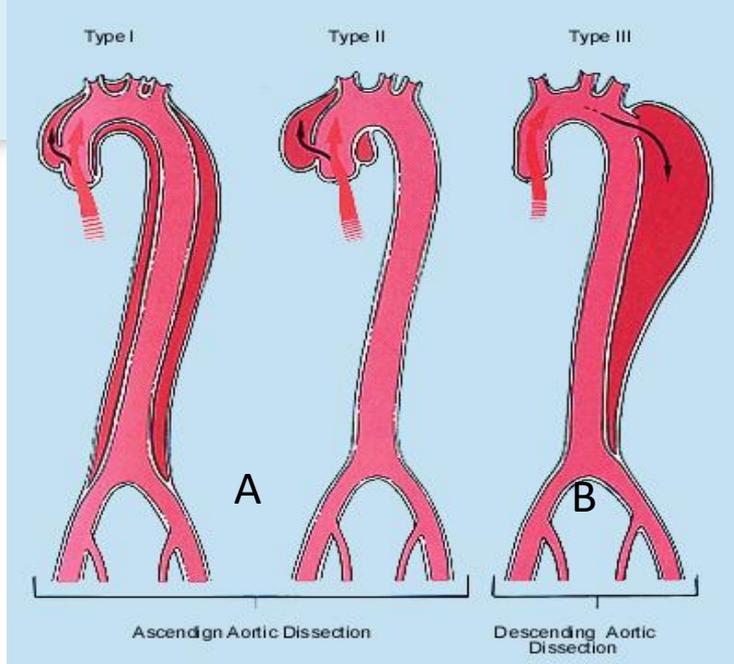
TAVR, Transcatheter aortic valve replacement.

Table 1 Classification schemes of aortic dissection

DeBakey (<i>Figure 1</i>)	
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 (<i>Figure 1</i>)	
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
Svensson (<i>Figure 2</i>)	
Class I	Classical dissection with true and false lumen
Class II	Intramural haematoma or haemorrhage
Class III	Subtle dissection without haematoma
Class IV	Penetrating atherosclerotic aortic ulcer
Class V	Iatrogenic or traumatic dissection

Modified from Nienaber and Clough.⁹

Qto a Duração:
Aguda
Crônica



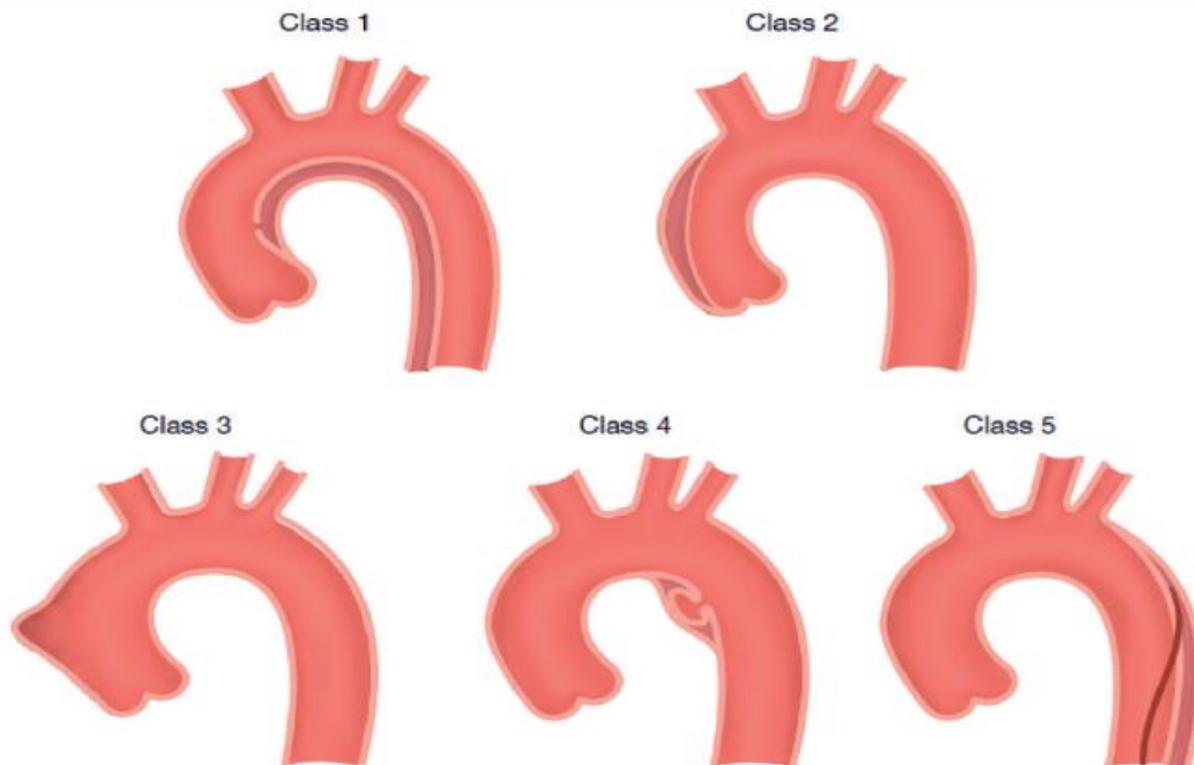


Figure 2 Classification of acute aortic syndromes by Svensson *et al.*¹¹ (see Table 1). Modified from Erbel *et al.*,² with copyright permission.

Figure 17. Classes of intimal tears. I. Classic dissection with intimal tear and double lumen separated by septum. Communication between lumens is typically in descending aorta at sheared-off intercostal arteries or distal reentry site. II. IMH. No intimal tear or septum is imaged but is usually found at surgery or autopsy. DeBakey Types II and IIIa are common extent of this lesion. III. Intimal tear without medial hematoma (limited dissection) and eccentric aortic wall bulge. Rare and difficult to detect by TEE or CT. Patients with Marfan syndrome prone to this type. May result in aortic rupture or extravasation. IV. PAU usually to the adventitia with localized hematoma or saccular aneurysm. May propagate to Class I dissection, particularly when involving ascending aorta or aortic arch. V. Iatrogenic (catheter

TABLE 63.4 Aortic Dissection Classification Based on Duration from Symptom Onset

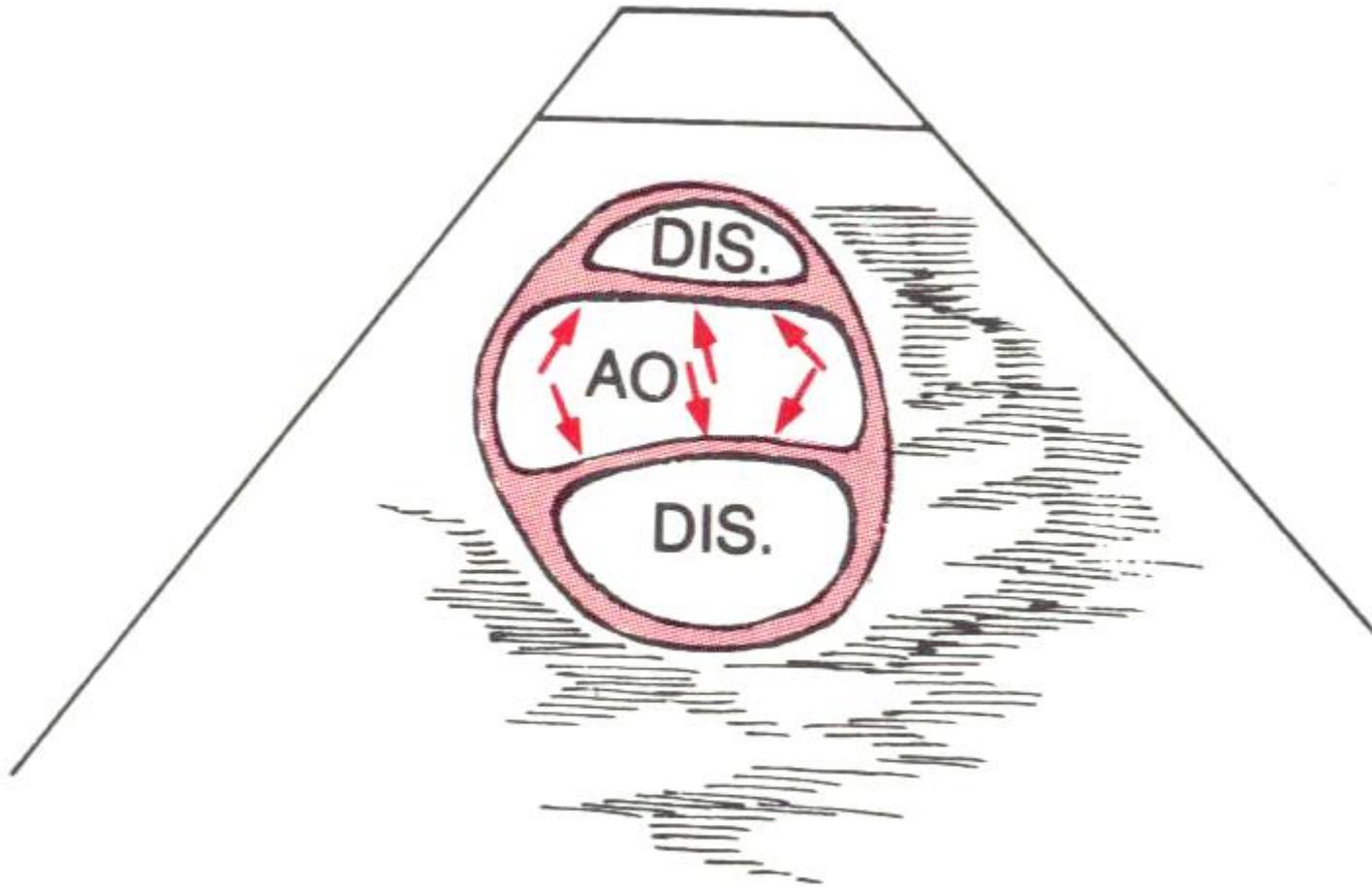
CLASSIC DEFINITION	TAD GUIDELINES ¹	IRAD CLASSIFICATION ²	ESC GUIDELINES ³
Acute: <14 days	Acute: <14 days	Hyperacute: <24 hours	Acute: <14 days
Chronic: >14 days	Subacute: <2-6 weeks	Acute: 2-7 days	Subacute: 14-90 days
	Chronic: >6 weeks	Subacute: 8-30 days	Chronic: >90 days
		Chronic: >30 days	

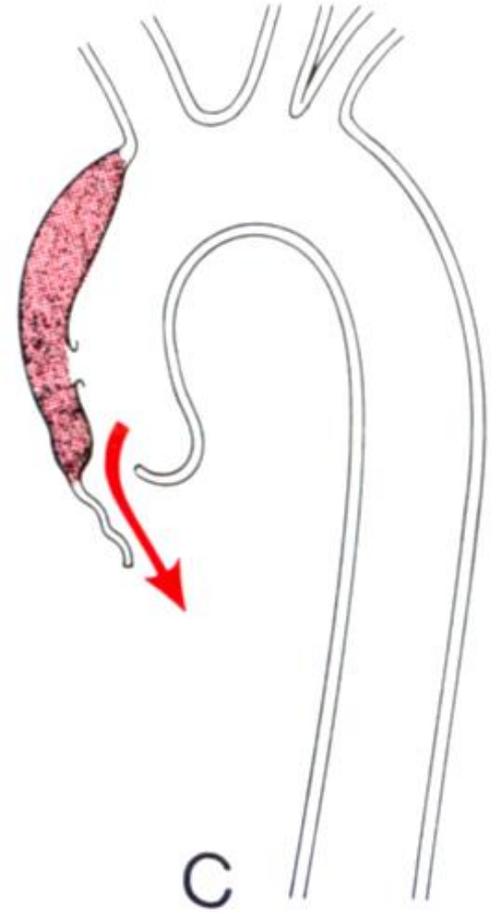
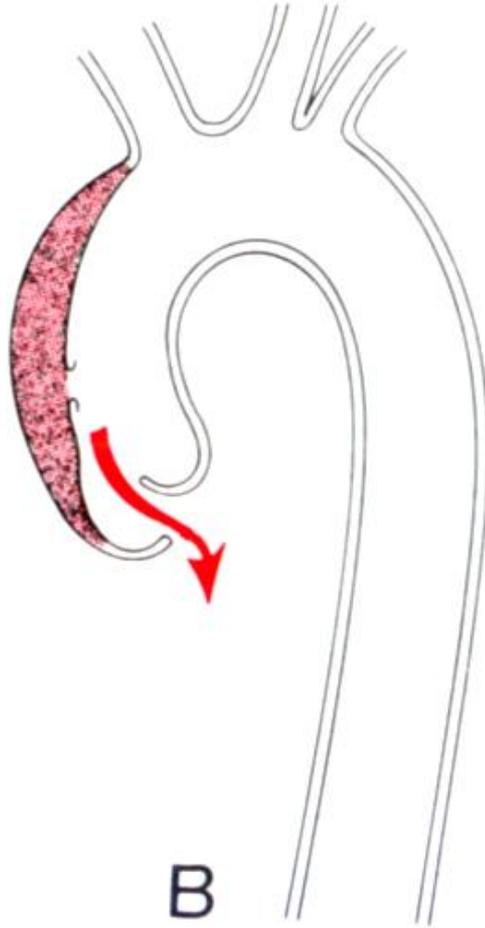
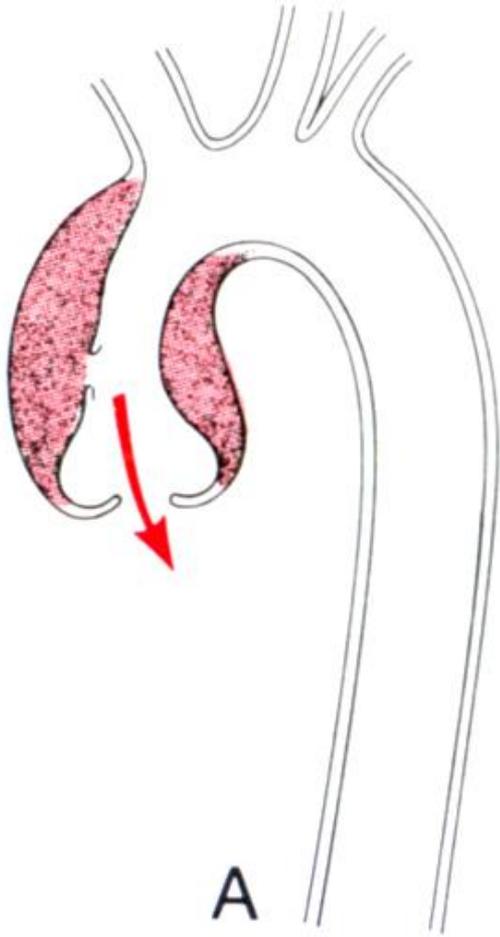
TAD, Thoracic Aortic Diseases; IRAD, International Registry of Acute Aortic Dissection; ESC, European Society of Cardiology.

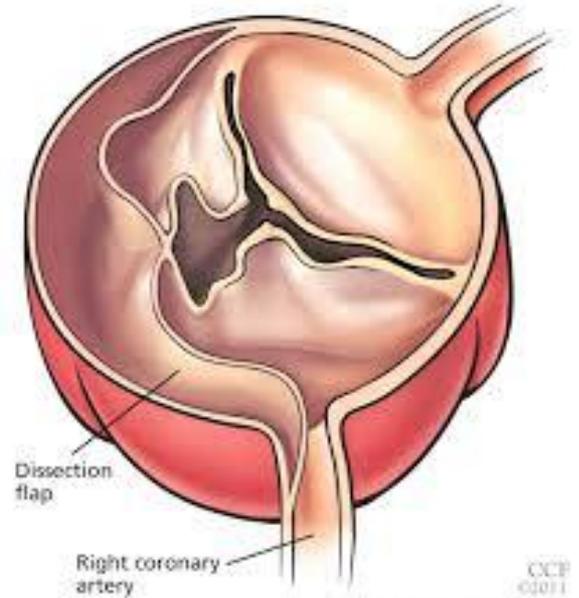
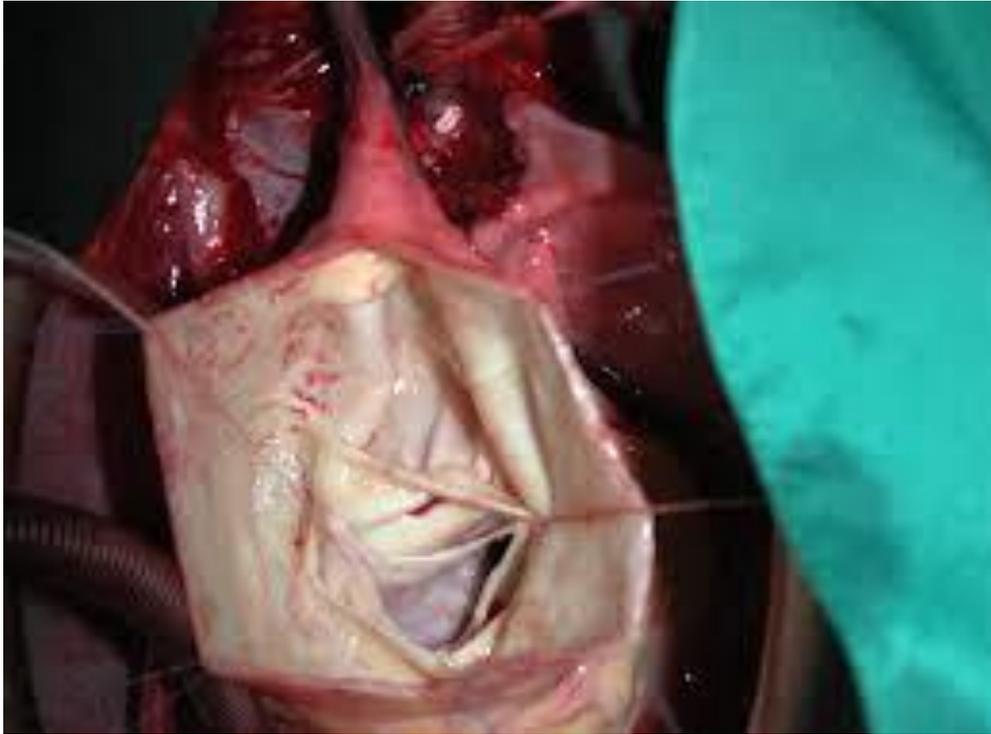
¹Hiratzka LF, Bakris GL, Beckman JA, et al. Guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American 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:e266-369.

²Booher AM, Isselbacher EM, Nienaber CA, et al. The IRAD classification system for characterizing survival after aortic dissection. *Am J Med* 2013;126:730 e19-24.

³Erbel 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;35:2873-926.







Dissection flap

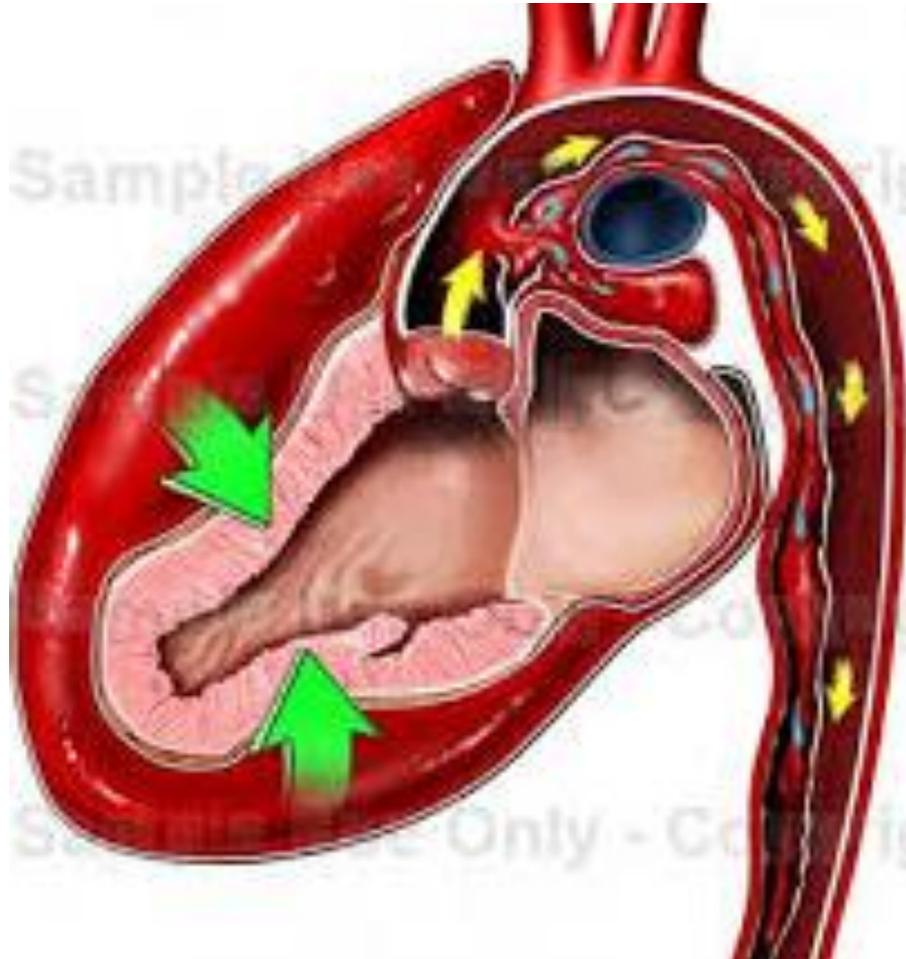
Right coronary artery

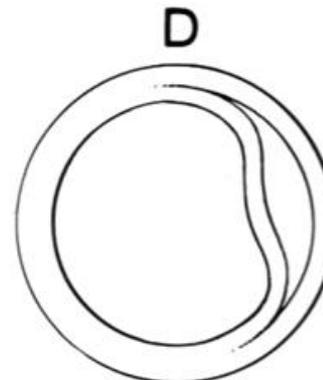
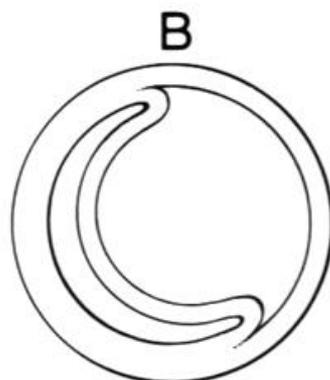
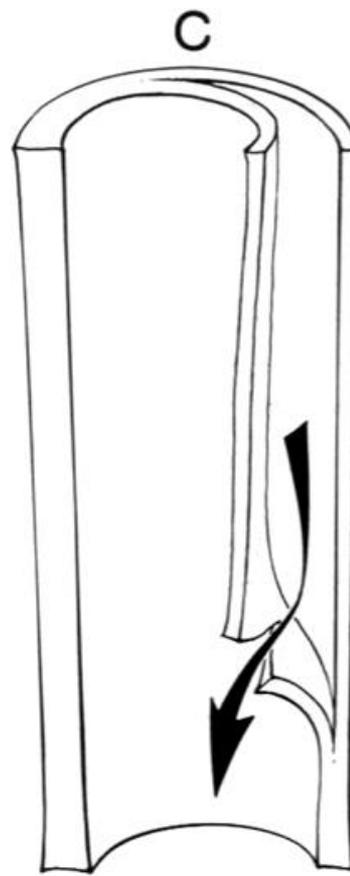
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Medical Illustrator: Joseph Pangrace

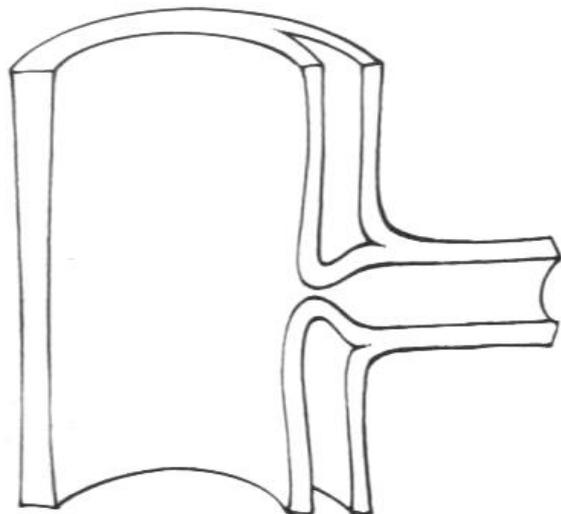


Riscos de puncionar ou drenar tamponamento cardíaco em dissecção de aorta

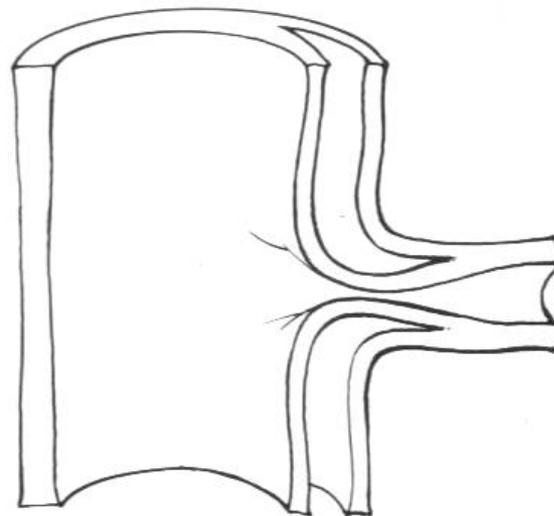




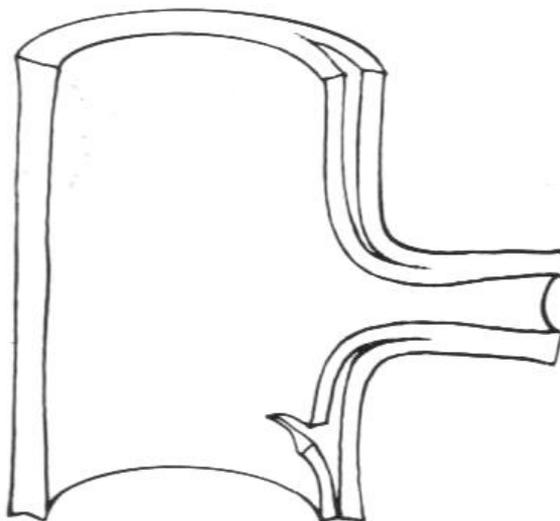
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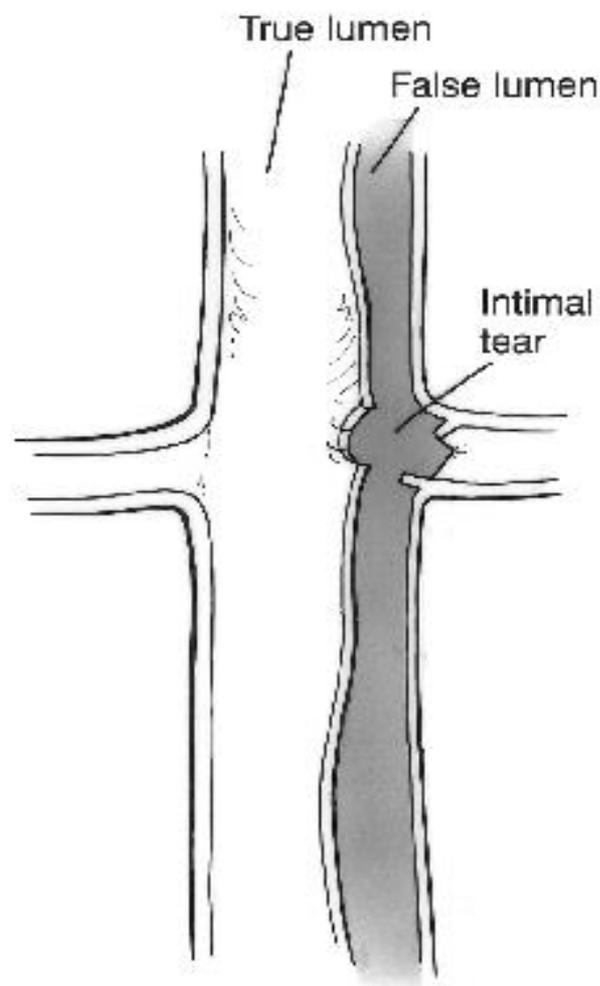
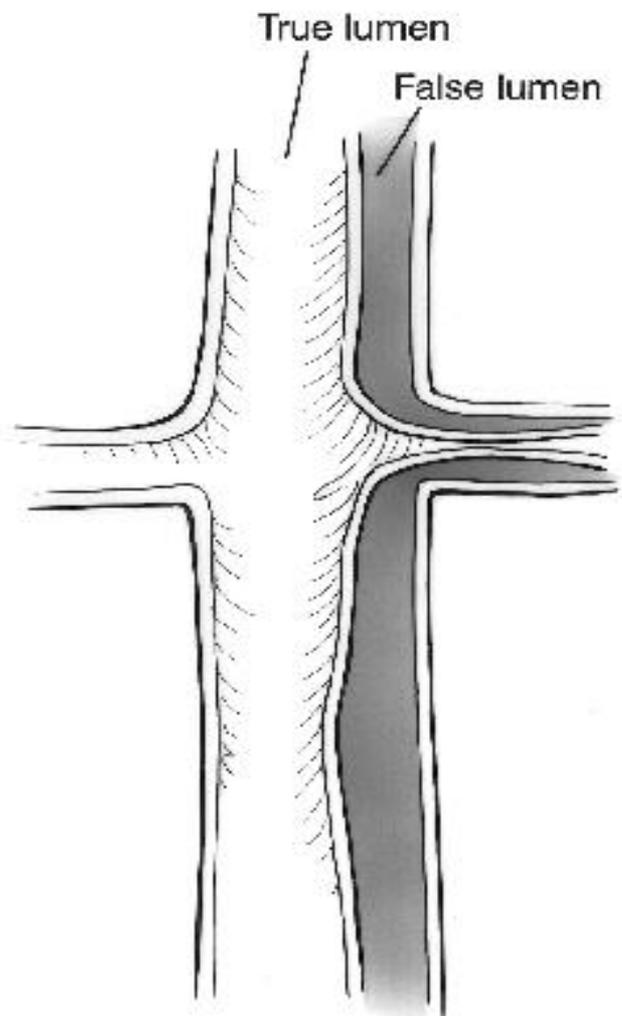


B



C





MANIFESTAÇÕES CLÍNICAS

	<i>Tipo A</i>	<i>Tipo B</i>
• <i>Frequência</i>	<i>60-75%</i>	<i>25-40%</i>
• <i>Dor</i>		
<i>Retroesternal</i>	<i>+++</i>	<i>+,-</i>
<i>Interescapular</i>	<i>+,-</i>	<i>+++</i>
• <i>Síncope</i>	<i>++</i>	<i>+,-</i>
• <i>AVC</i>	<i>+</i>	<i>-</i>
• <i>ICC</i>	<i>+</i>	<i>-</i>
• <i>I Ao</i>	<i>++</i>	<i>+,-</i>
• <i>IAM</i>	<i>+</i>	<i>-</i>
• <i>Derrame pericárdico</i>	<i>+++</i>	<i>-</i>
• <i>Derrame pleural</i>	<i>+,-</i>	<i>+++</i>
• <i>Isquemia MM/visceral</i>	<i>++</i>	<i>+</i>

Table 3 Presenting symptoms, signs, chest X-ray, and electrocardiographic features from the International Registry of Aortic Dissection registry

Category	Overall (n = 5638)	Type A AAD (n = 3747)	Type B AAD (n = 1891)	P-value
Symptoms and signs				
Chest or back pain	4692 (87.9%)	3113 (87.5%)	1579 (88.7%)	0.191
Severe or worst ever pain	4692 (87.9%)	3113 (87.5%)	1579 (88.7%)	0.191
Abrupt onset of pain	4220 (84.0%)	2789 (83.3%)	1431 (85.4%)	0.052
Migrating pain	664 (14.8%)	400 (13.7%)	264 (16.8%)	0.005
Pain presenting within 6 h of symptom onset	2950 (75.8%)	1700 (77.0%)	790 (73.1%)	0.015
Any focal neurological deficit	695 (13.7%)	575 (17.2%)	120 (7.0%)	<0.001
Hypotension, shock, or tamponade	1136 (23.4%)	1054 (32.6%)	82 (5.0%)	<0.001
Hypertension at presentation	1943 (40.0%)	893 (27.6%)	1050 (64.6%)	<0.001
Any pulse deficit	1170 (32.3%)	811 (35.9%)	359 (26.3%)	<0.001
Aortic regurgitation	1440 (38.7%)	1266 (51.8%)	174 (13.6%)	<0.001
Abdominal pain	1442 (30.5%)	766 (24.9%)	676 (41.1%)	<0.001
Chest radiography				
Normal	999 (28.1%)	588 (26.9%)	411 (30.2%)	0.031
Widened mediastinum	1509 (49.5%)	1016 (53.7%)	493 (42.6%)	<0.001
Abnormal aortic contour	1289 (43.7%)	760 (41.8%)	529 (46.6%)	0.011
Electrocardiography				
Normal	1763 (39.1%)	1147 (38.3%)	616 (40.7%)	0.120
Left ventricular hypertrophy	817 (23.1%)	490 (20.9%)	327 (27.3%)	<0.001
Myocardial ischemia or infarction	647 (18.0%)	526 (21.9%)	121 (10.1%)	<0.001

AAD, acute aortic dissection.

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.

Initial management of patients with suspected aortic dissection

Recommendation	Class I	II	III	Level of evidence
1. Detailed medical history and complete physical examination (whenever possible)	ⓐ			C
2. Intravenous line, blood sample (CK, TnT(I), myoglobin, WBC, D-dimer, haematocrit, LDH)	ⓐ			C
3. ECG: documentation of ischaemia	ⓐ			C
4. HR and blood pressure monitoring	ⓐ			C
5. Pain relief (morphine sulphate)	ⓐ			C
6. Reduction of systolic blood pressure using beta-blockers (i.v. propranolol, metoprolol, esmolol or labetalol)	ⓐ			C
7. Transfer to intensive care unit	ⓐ			C
8. In patients with severe hypertension additional vasodilator (i.v. sodium nitroprusside to titrate BP to 100-120 mmHg)	ⓐ			C
9. In patients with obstructive pulmonary disease, blood pressure lowering with calcium channel blockers		ⓑ		C
10. Imaging in patients with ECG signs of ischaemia before thrombolysis if aortic pathology is suspected		ⓑ		C
11. Chest X-ray			ⓐ	C

TABLE 63.6 Organ System Complications of Acute Aortic Dissection

Cardiovascular	<ul style="list-style-type: none"> Cardiac arrest Syncope Aortic regurgitation Congestive heart failure Coronary ischemia Myocardial infarction Cardiac tamponade Pericarditis
Pulmonary	<ul style="list-style-type: none"> Pleural effusion Hemothorax Hemoptysis (from an aortotracheal or bronchial fistula)
Renal	<ul style="list-style-type: none"> Acute renal failure Renovascular hypertension Renal ischemia or infarction
Neurologic	<ul style="list-style-type: none"> Stroke Transient ischemic attack Paraparesis or paraplegia Encephalopathy Coma Spinal cord syndrome Ischemic neuropathy
Gastrointestinal	<ul style="list-style-type: none"> Mesenteric ischemia or infarction Pancreatitis Hemorrhage (from aortoenteric fistula)
Peripheral vascular	<ul style="list-style-type: none"> Upper or lower extremity ischemia
Systemic	<ul style="list-style-type: none"> Fever

Zipes et al (Ed.)
 Braunwald's Heart Disease
 Elsevier 2018

Complicações / Má perfusão

Vascular system

Renal

Frequency

23%-75%

Extremities (upper and lower)

25%-60%

Mesenteric

10%-20%

Coronary

5%–11%

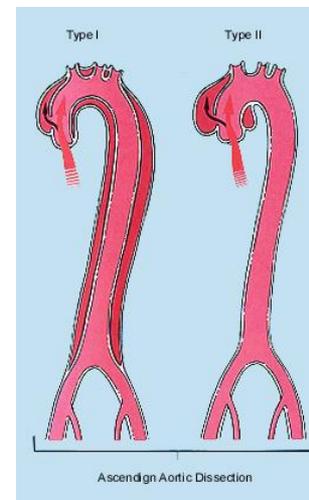
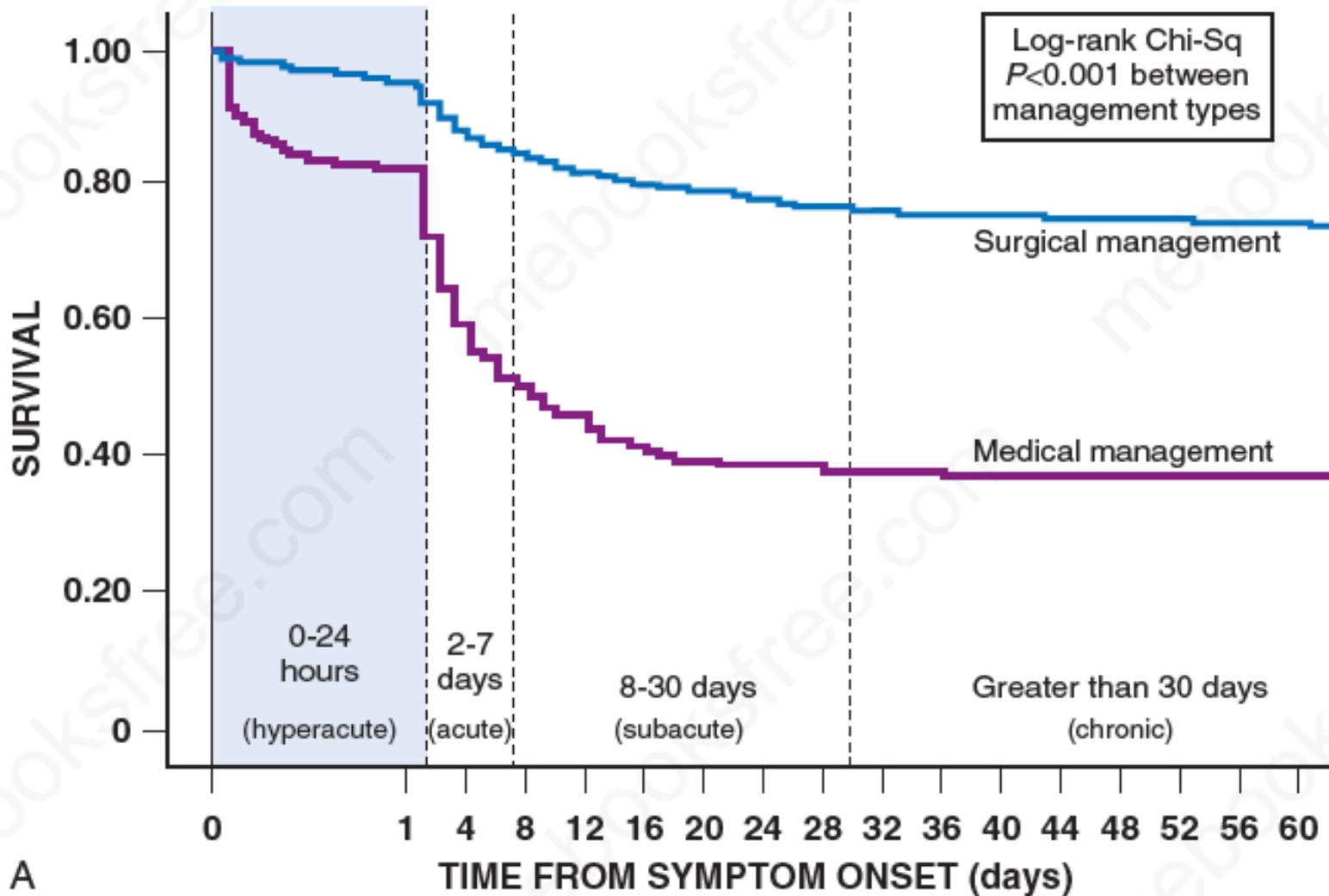
Cerebral

3%–13%

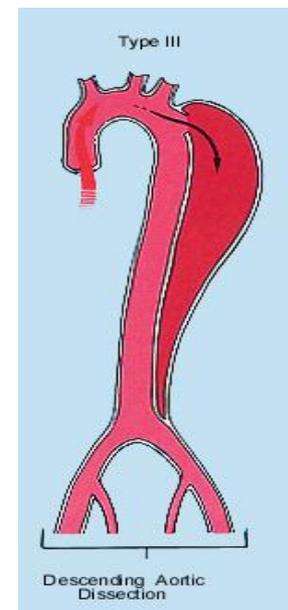
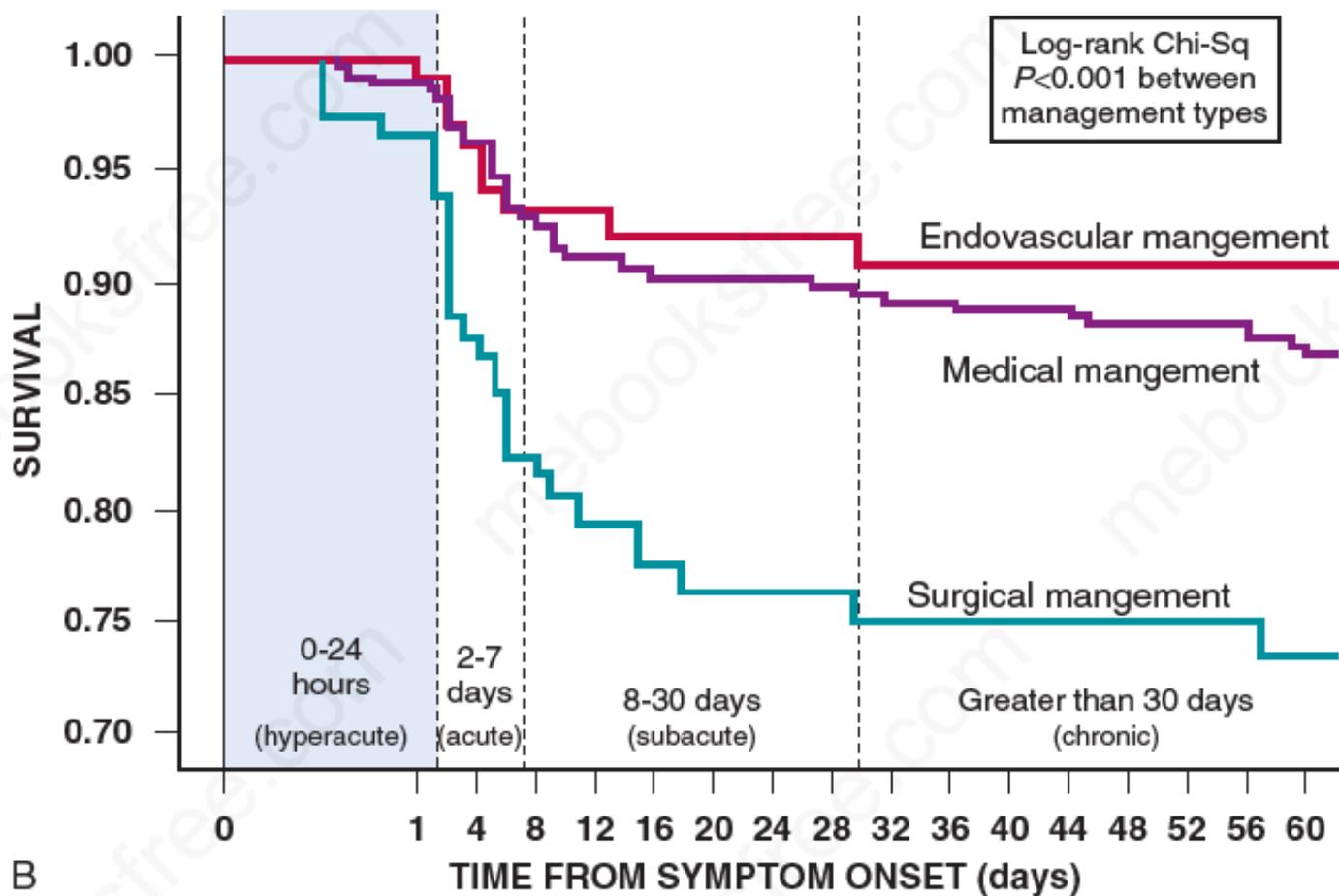
Spinal

2%–9%

KAPLAN-MEIER SURVIVAL CURVE DISSECTION TYPE: A



KAPLAN-MEIER SURVIVAL CURVE DISSECTION TYPE: B



B

FIGURE 63.12 International Registry of Acute Aortic Dissection (IRAD) classification system of survival after aortic dissection. Kaplan-Meier survival curves for type A dissection (A) and type B dissection (B) 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 e19-24.)

DIAGNÓSTICO DIFERENCIAL

- *Síndrome coronariana aguda*
- *Insuficiência aórtica aguda*
- *Aneurisma aórtico*
- *Dor músculo-esquelética*
- *Pericardite*
- *Embolia pulmonar*
- *Colecistite*
- *Tumores mediastinais*



EXAMES DE IMAGEM

	<i>Sensibilidade // Especificidade</i>	
• <i>ETT/ETE</i>	<i>77-85%</i>	<i>93-96%</i>
• <i>TOMOGRAFIA</i>	<i>83-94%</i>	<i>87-100%</i>
• <i>RESSONÂNCIA</i>	<i>> 98%</i>	<i>> 98%</i>
• <i>AORTOGRAFIA</i>	<i>88%</i>	<i>>95%</i>

Table 5 Relative strengths of imaging modalities for acute aortic syndromes

	TTE	TOE	MRI	CT
Imaging factors				
Comprehensive aortic assessment	+	++	+++	+++
Tomographic (3D reconstruction)	-	-	+++	+++
Functional data	+++	+++	++	+
Tissue characterization	-	-	+++	+++
Clinical factors				
Portability	+++	+++	-	-
Patient access/monitoring	+++	+++	+	++
Rapidity	+++	++	++	+++
Need for contrast	-	-	++	+++
Need for sedation	-	+++	-	-
Lack of radiation exposure	+++	+++	+++	-

3D, three-dimensional; CT, computed tomography; MRI, magnetic resonance imaging; TOE, transoesophageal echocardiography; TTE, transthoracic echocardiography.

Modified from Bossone et al³³

Imaging: diagnostic goals

- ***Confirm diagnosis***
 - ***Classify the dissection/delineate the extent***

 - ***Differentiate true and false lumen***
 - ***Localize intimal tears***

 - ***Distinguish between dissection, intimal tear, penetrating ulcer, intramural haematoma***

 - ***Assess side branch involvement (including coronary arteries)***

 - ***Detect and grade aortic regurgitation***

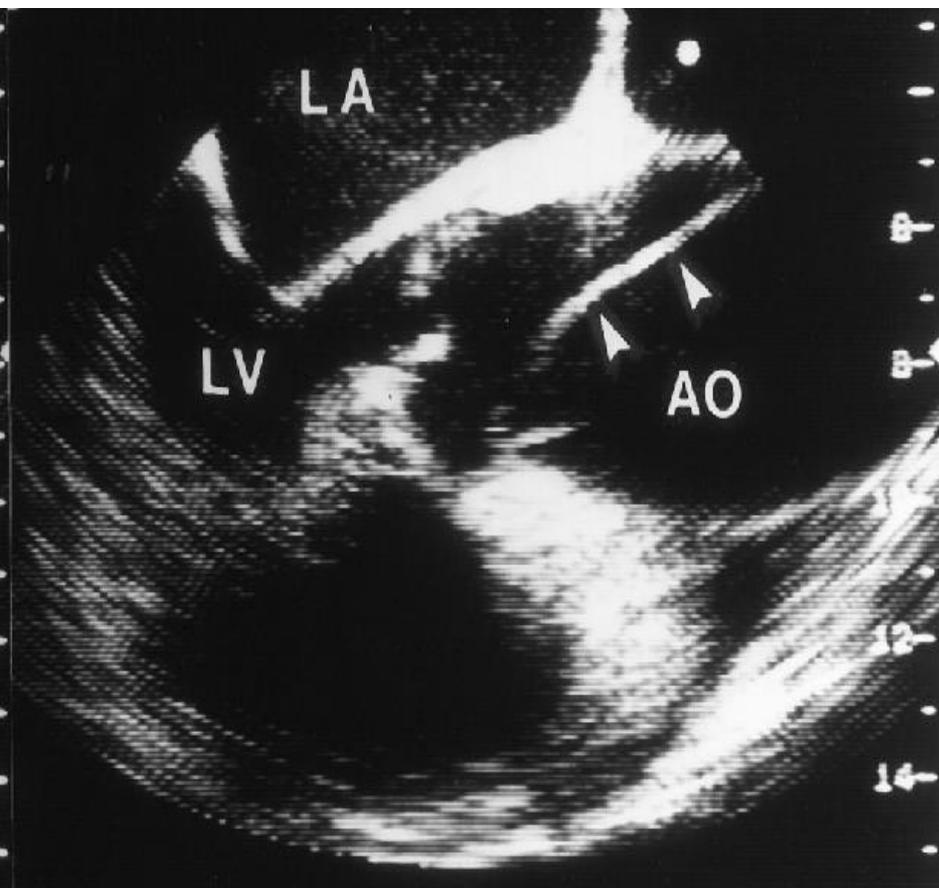
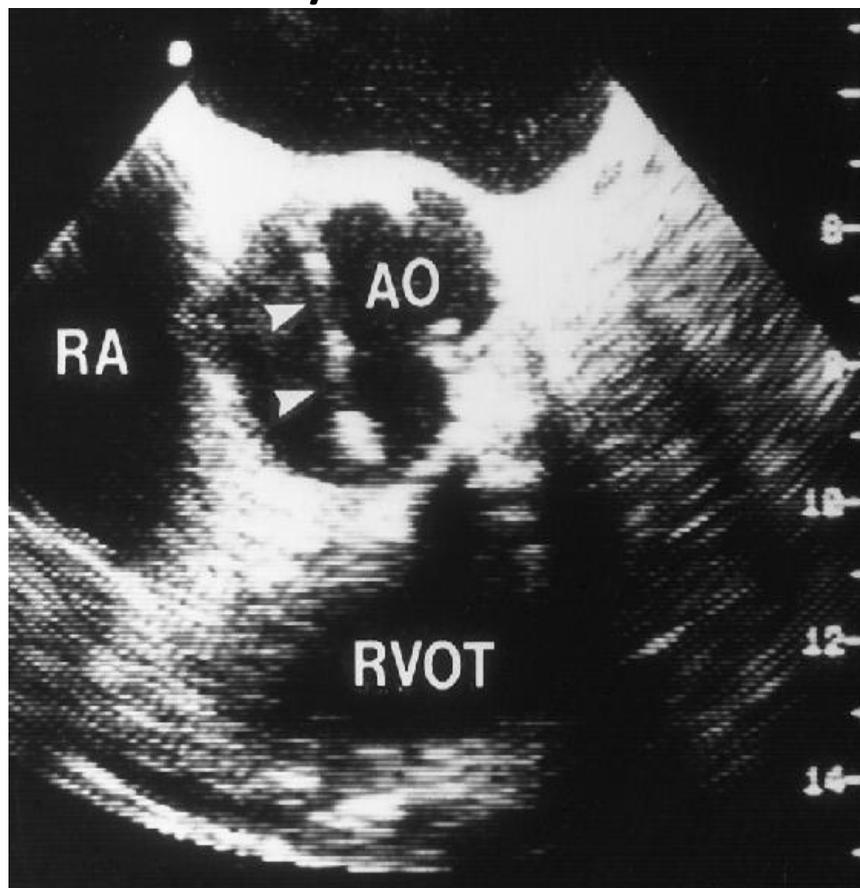
 - ***Detect extravasation (periaortic or mediastinal haematoma, pleural or pericardial effusion)***
-

EXAMES DE IMAGEM

ETT / ETE

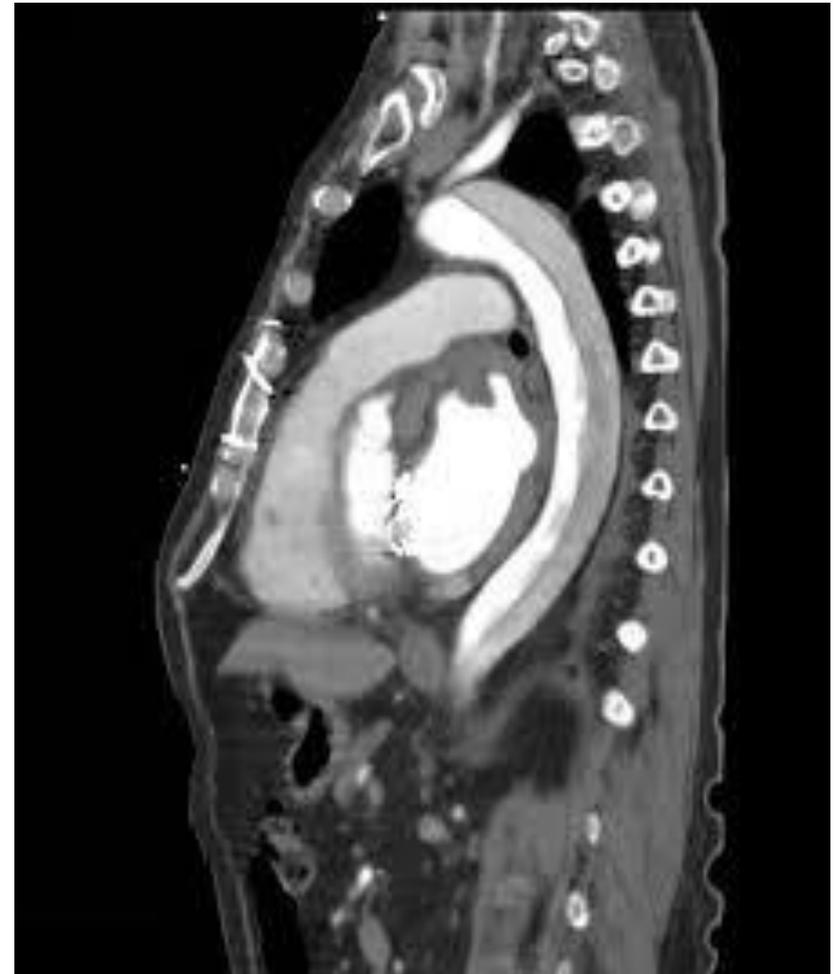
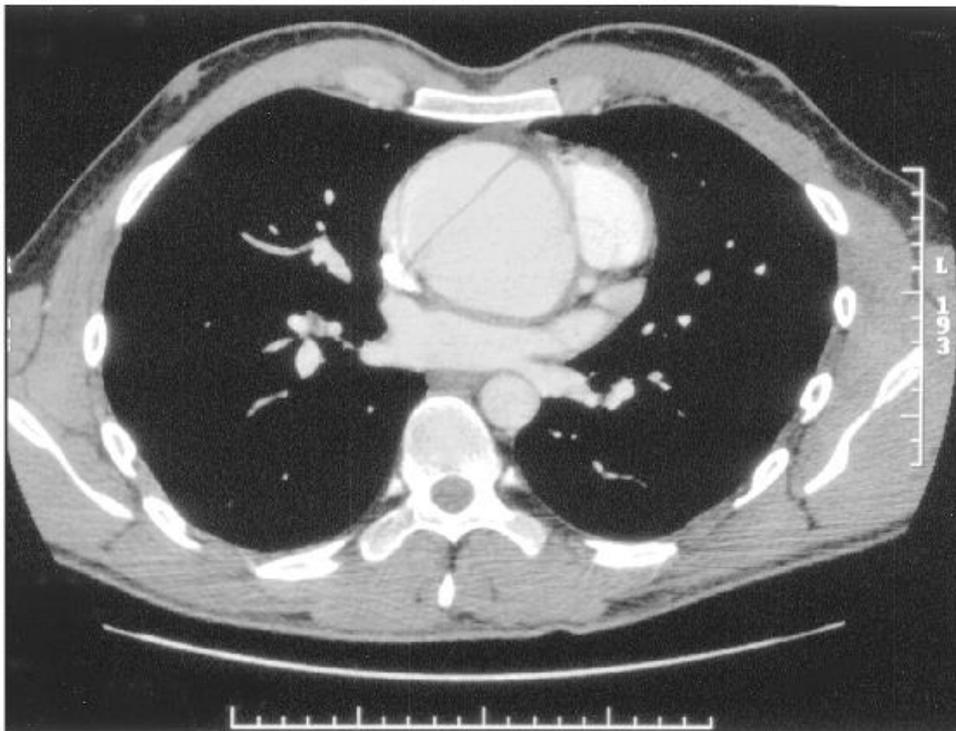
S 77-85%

E 93-96%



Angiotomografia

S 83-94% E 87-100%



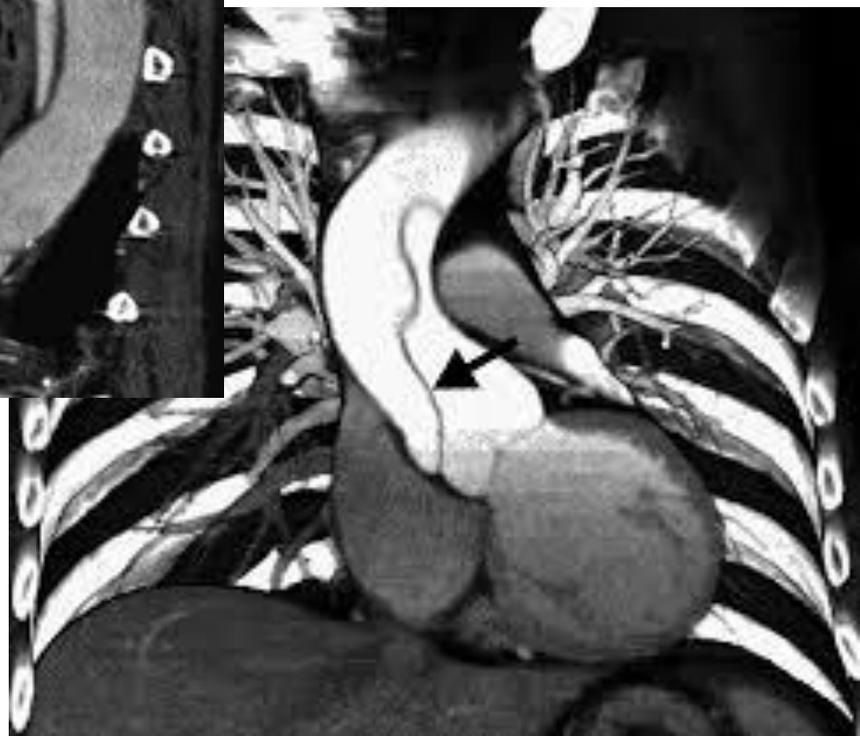
Angiotomografia

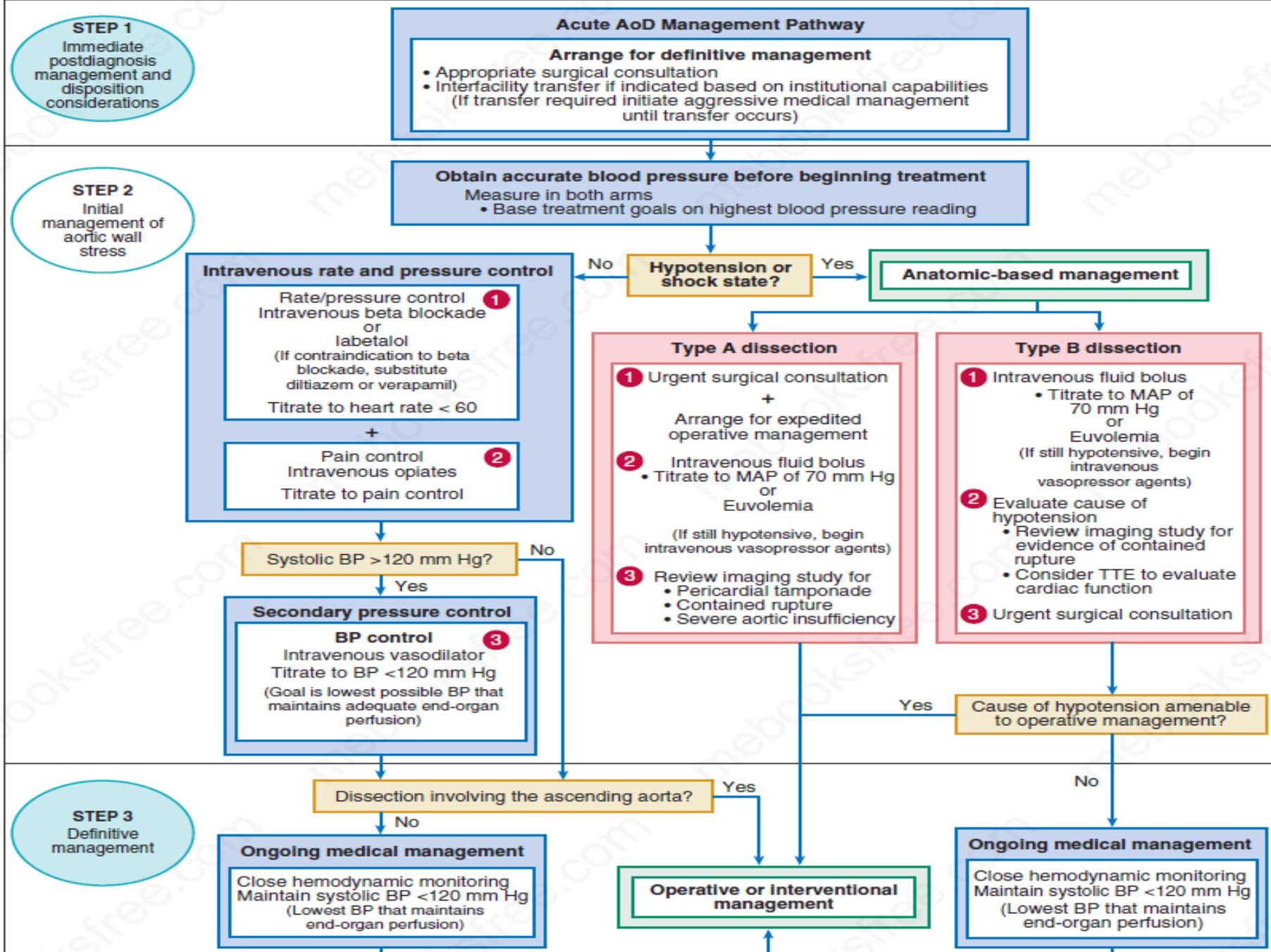
S 83-94% E 87-100%



Ressonância Magnética Nuclear

E > 98% **S > 98%**





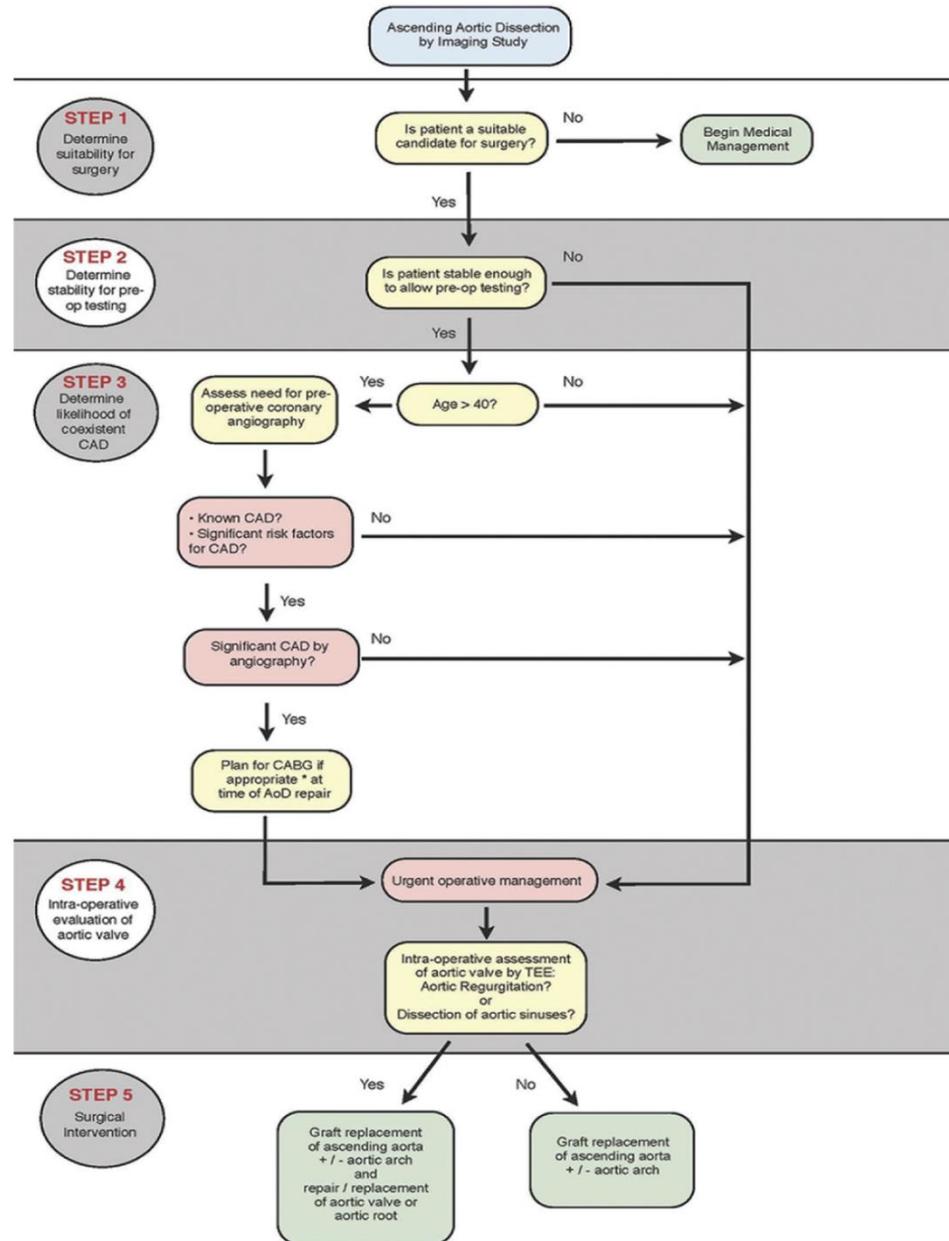


Figure 22. Acute surgical management pathway for AoD. *Addition of 'if appropriate' based on Patel et al. (226a) AoD indicates aortic dissection; CABG, coronary artery bypass graft surgery; CAD, coronary artery disease; TAD, thoracic aortic disease; and TEE, transesophageal echocardiogram.

DISSECÇÃO AGUDA DE AORTA

TRATAMENTO

- ***TIPO A (Tipo I e II)***

CIRÚRGICO

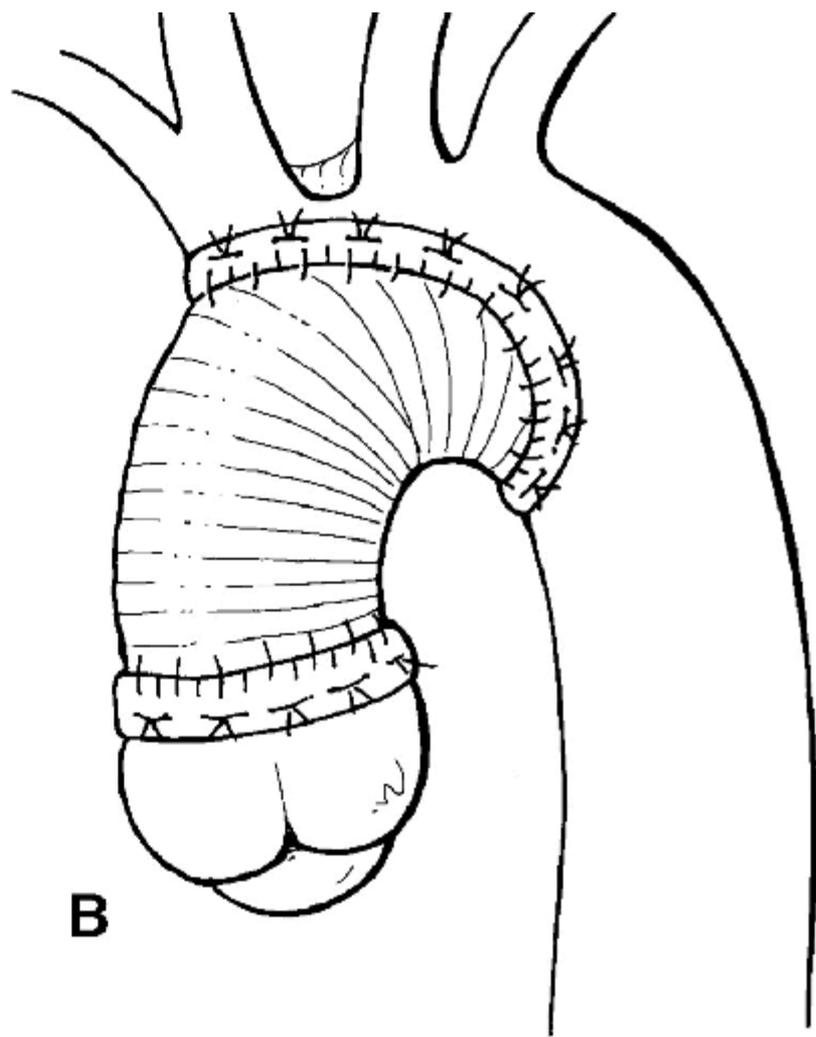
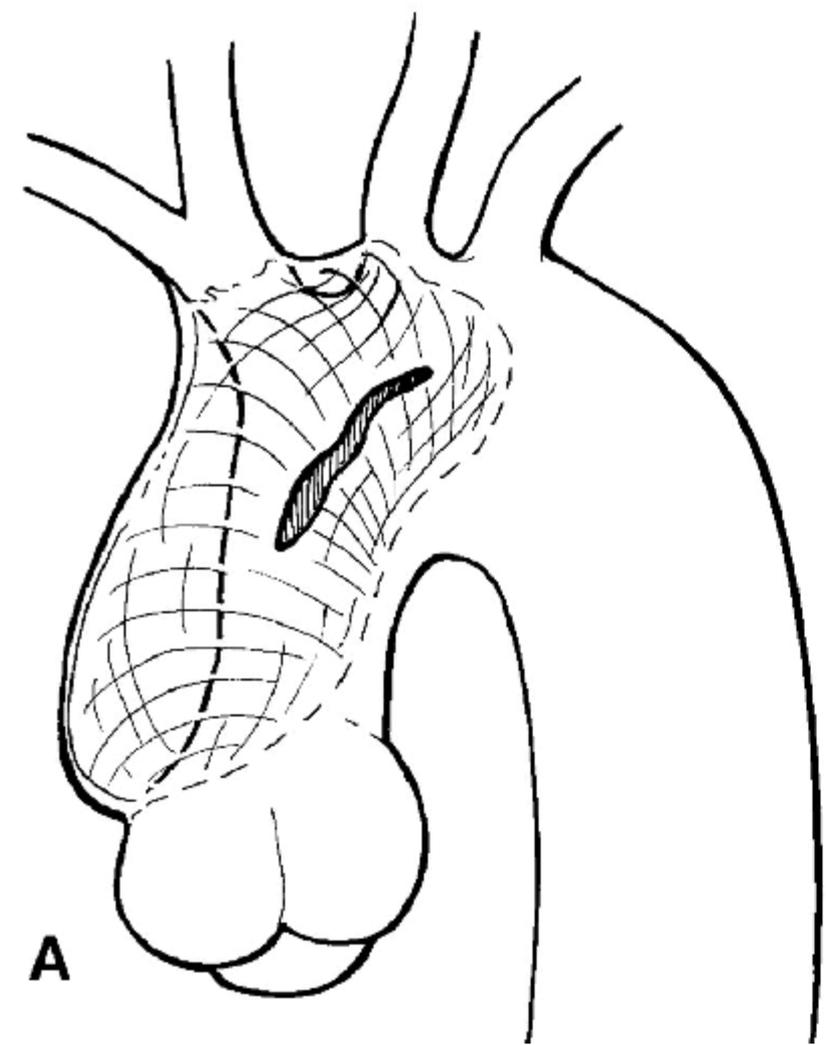
OBJETIVOS: Prevenir ruptura

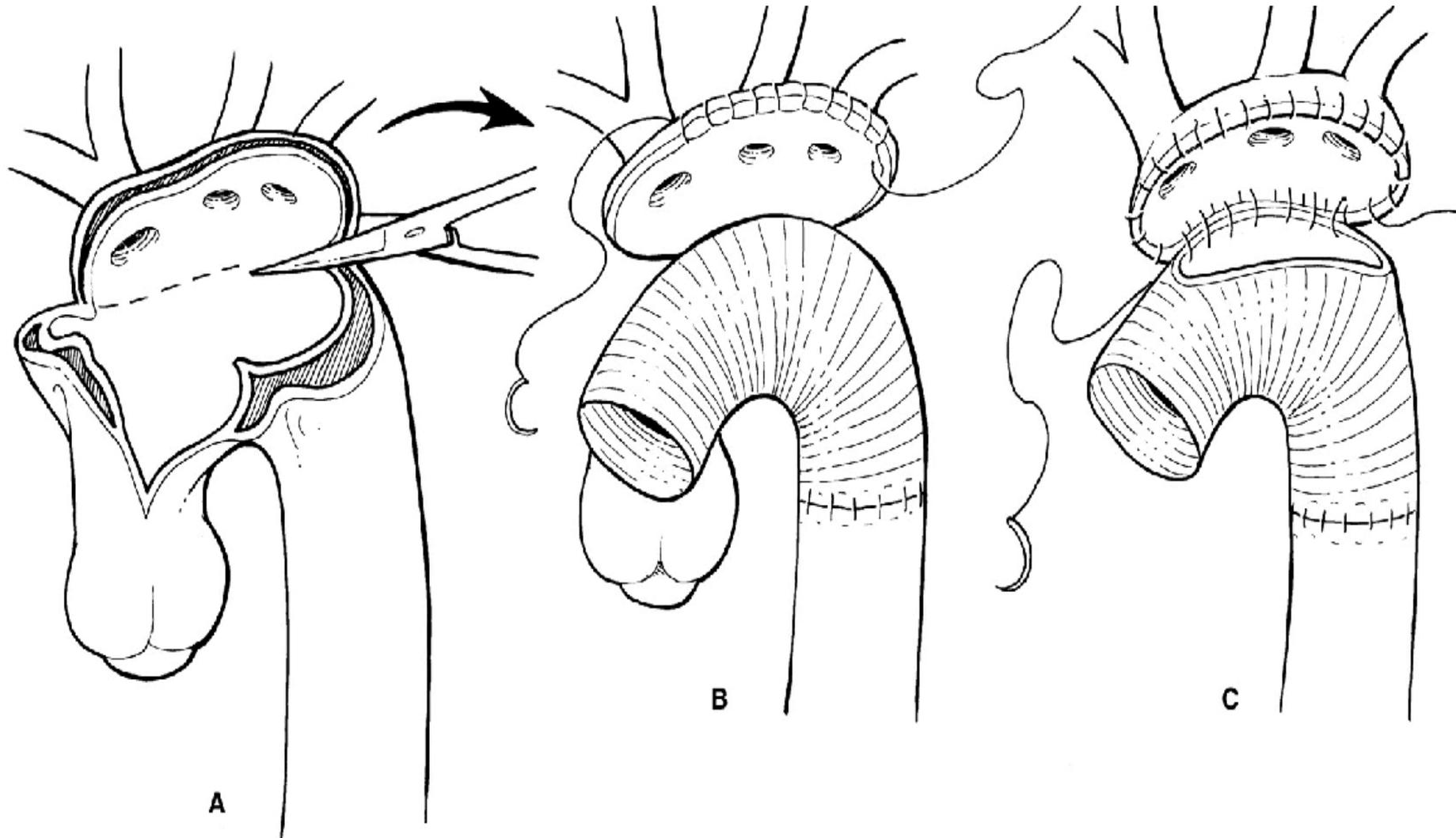
Prevenir tamponamento

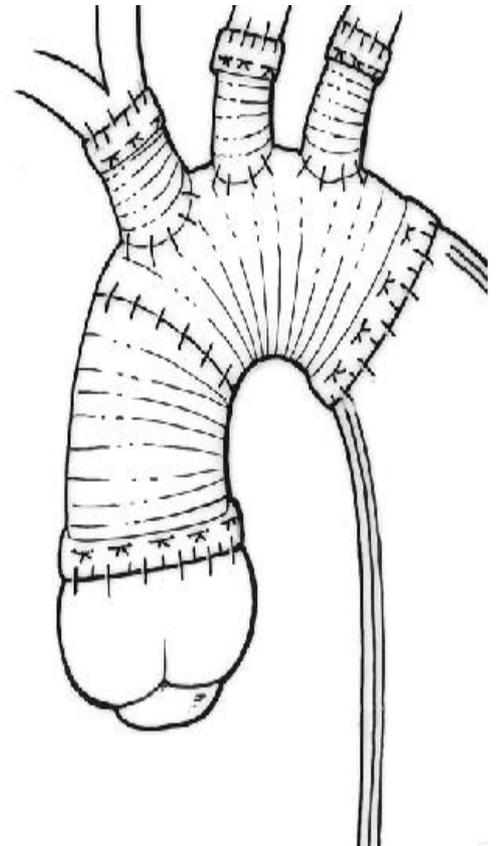
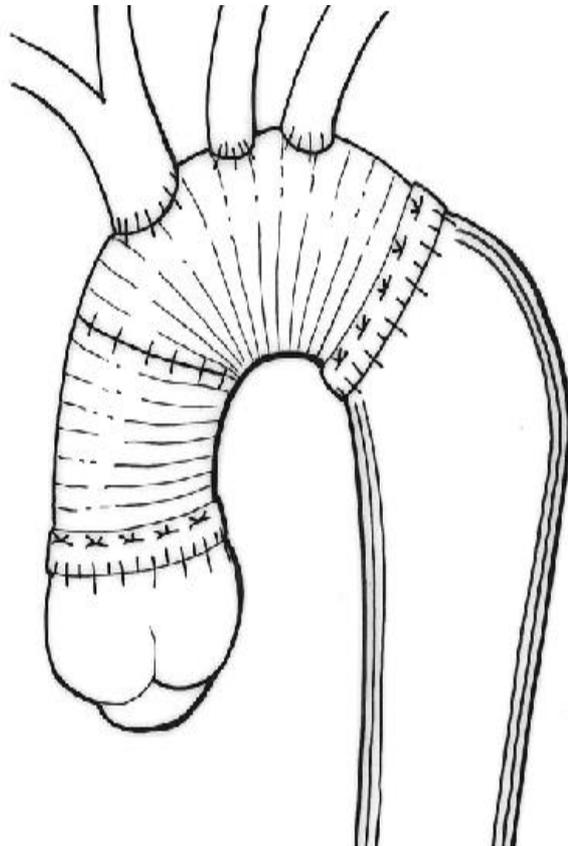
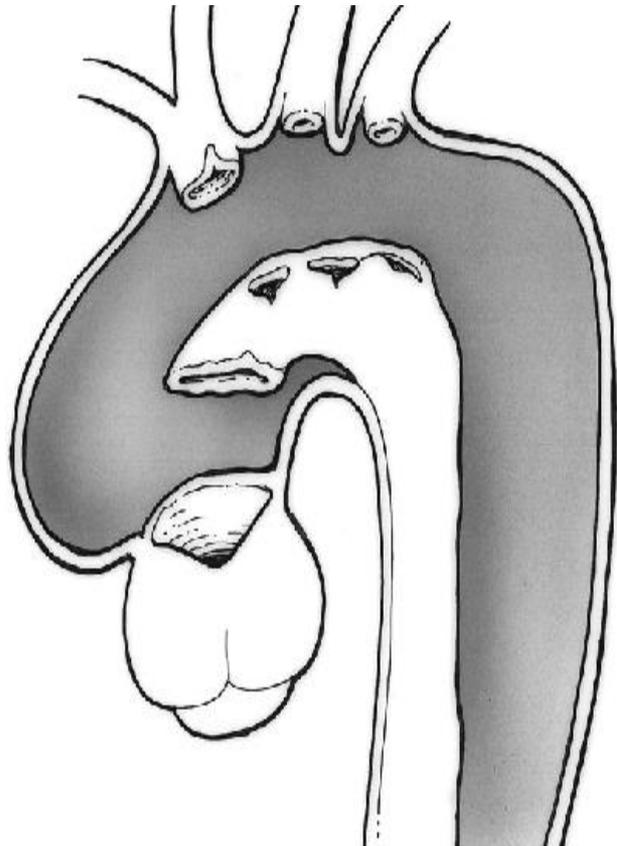
Tratar insuficiência aórtica

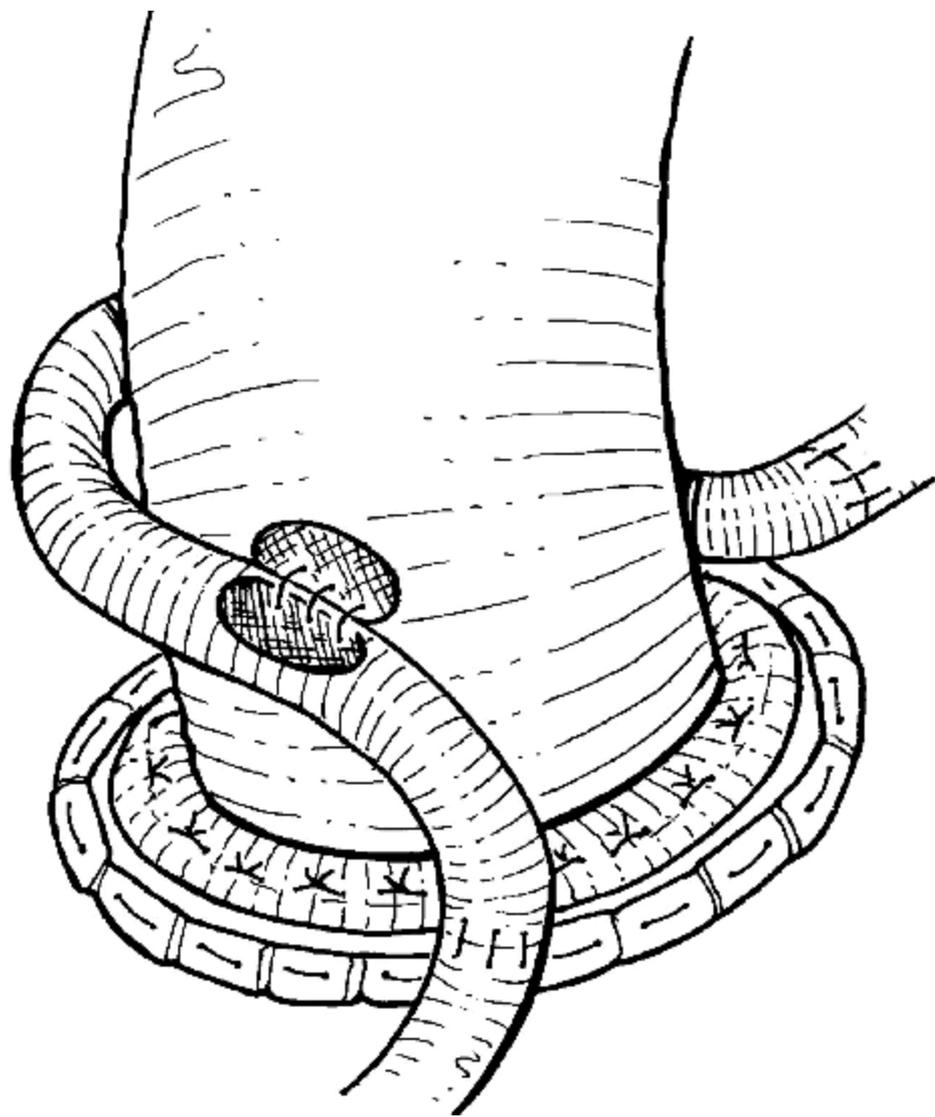
Evitar isquemia

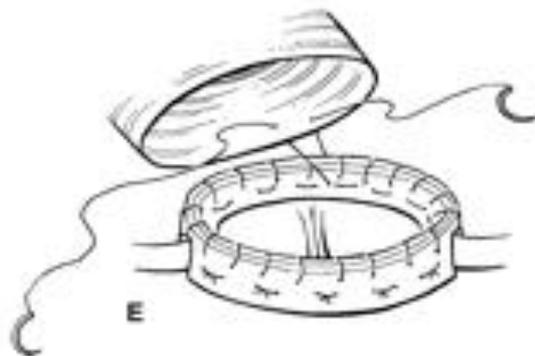
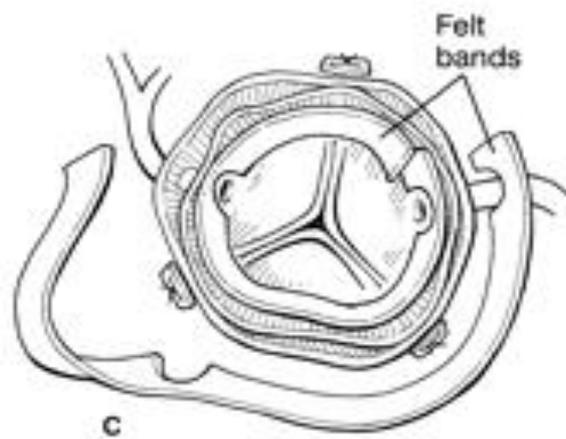
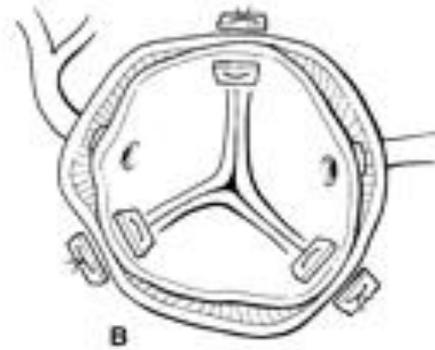
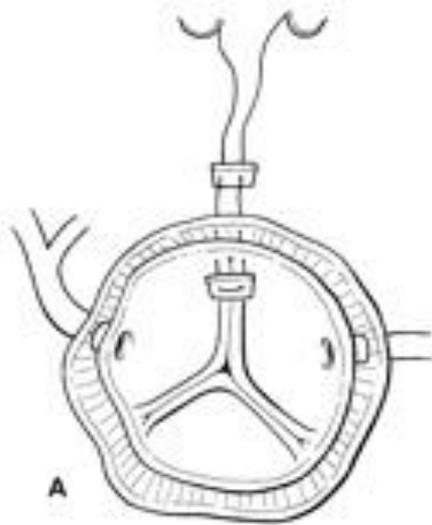
Tratar ruptura da íntima

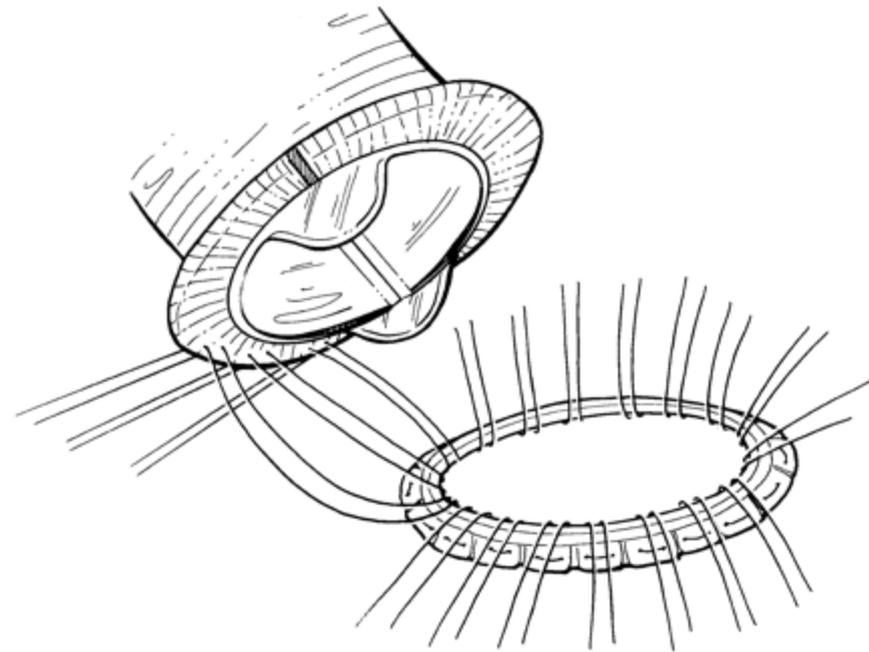


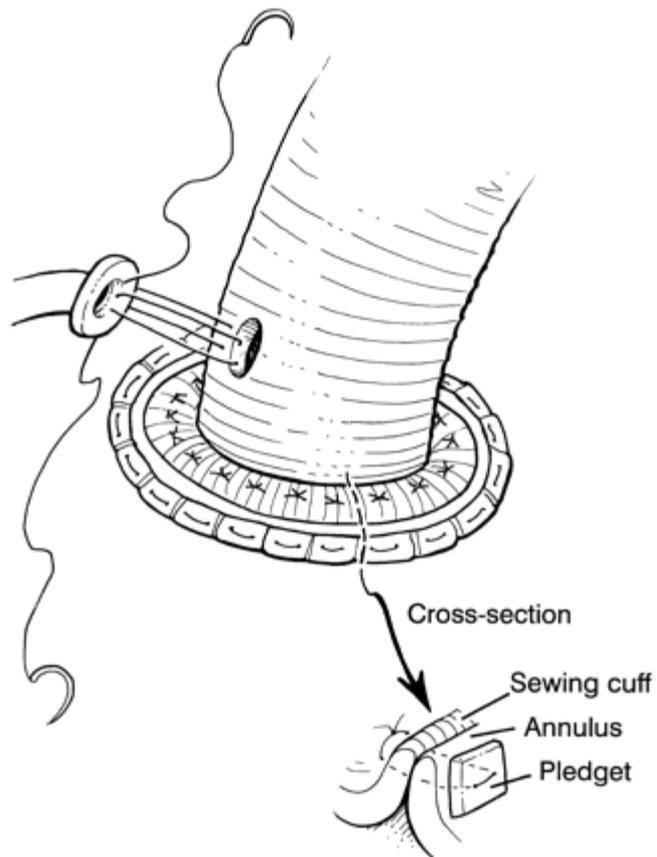












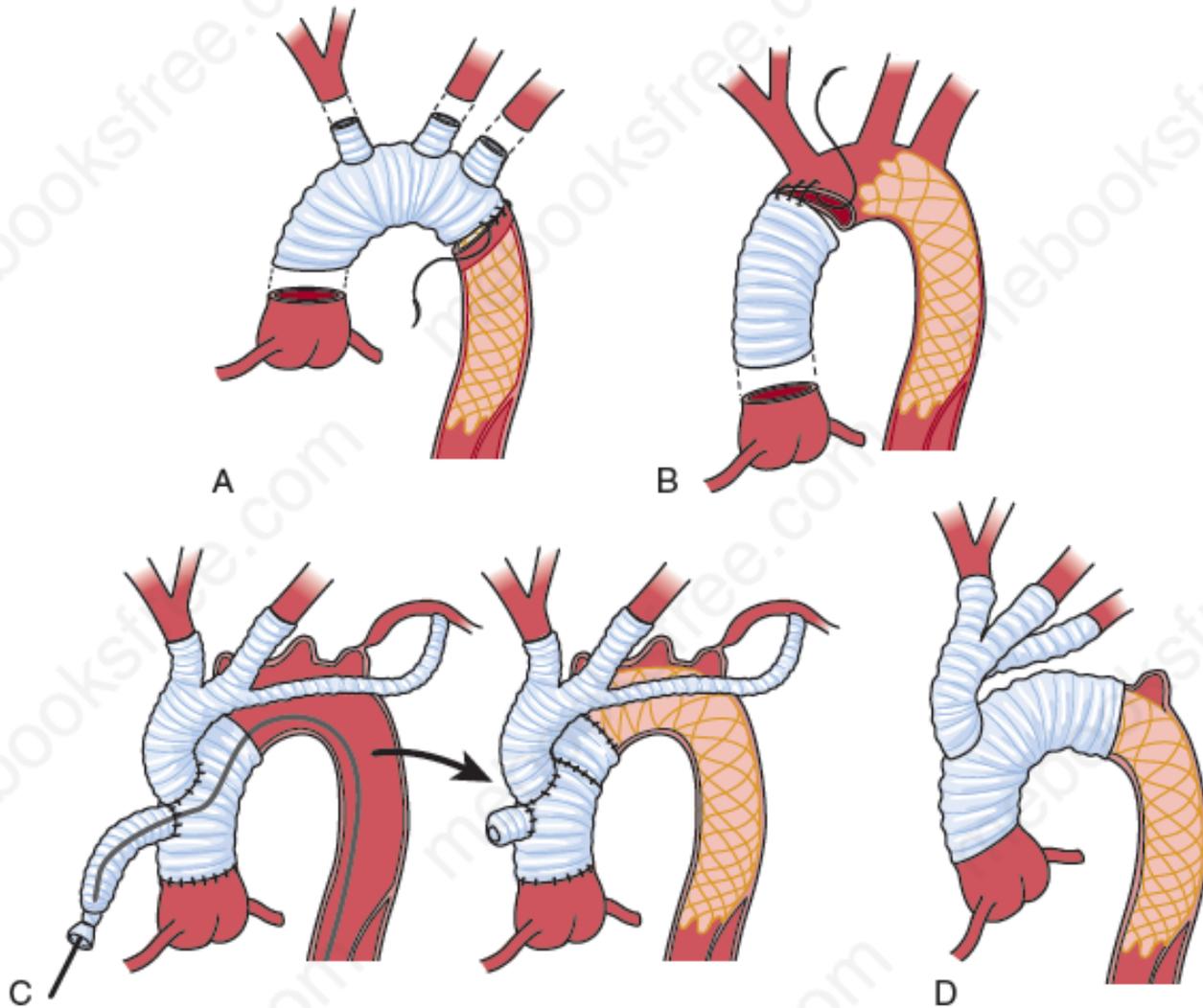
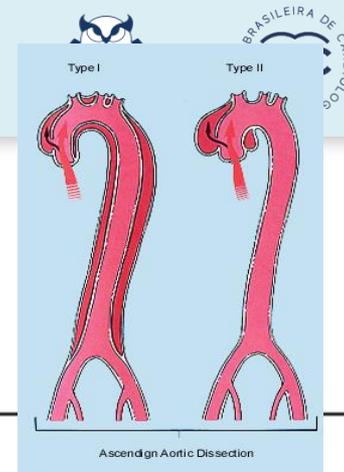


FIGURE 63.21 Approaches to extended distal repair for acute type A aortic dissection. **A**, Open stent graft and total arch replacement with antegrade stent-graft placed in descending thoracic aorta at circulatory arrest. **B**, Open stent-graft and hemiarch replacement with antegrade stent-graft placed in the descending thoracic aorta at circulatory arrest. **C**, Closed stent-graft with hybrid arch. Proximal rerouting of arch vessels to sinotubular junction and endovascular stent graft deployment into ascending aortic graft with fluoroscopy after weaning from cardiopulmonary bypass. **D**, Closed stent-graft with hybrid arch replacement. Arch replaced surgically to level of left subclavian artery and polyester proximal landing zone created for stent-graft in transverse arch. (From El-Hamamsy I, Ouzounian M, Demers P, et al. State-of-the-art surgical management of acute type A aortic dissection. *Can J Cardiol* 2016;32:100-9. Images courtesy Dr. Jehangir Appoo.)



Diretriz Brasileira para tratar Dissecção Aguda Aorta Tipo A

Tabela I – Recomendações para o tratamento das dissecções agudas tipo A.

Recomendações	Classe de evidência	Nível de evidência
1. Cirurgia imediata para evitar ruptura / tamponamento / morte	I	C
2. Enxerto reto na aorta ascendente, se raiz da aorta e válvula aórtica normais	I	C
3. Enxerto reto na aorta ascendente e ressuspensão valvar aórtica, se raiz da aorta normal e válvula insuficiente por perda de sustentação	I	C
4. Tubo valvado, se aorta ascendente dilatada ou ectasia ânulo/aórtica e válvula aórtica insuficiente	I	C
5. Auto ou homoenxerto, se (situação nº 4) associada à endocardite	IIA	C
6. Ressuspensão valvar aórtica e remodelamento da raiz da aorta em síndrome de Marfan	IIA	C
7. Reparo parcial do arco aórtico (<i>hemiaroh repair</i>), se dissecção compromete o arco, mas não há destruição ou lesão da íntima	I	C
8. Reconstrução total do arco, se há destruição ou lesão da íntima dentro do mesmo	I	C
9. Em caso de intervenção no arco, reconstrução aberta com método de proteção cerebral (PCC hipotérmica – retroperfusão cerebral – cerebroplegia – perfusão axilar)	I	C
10. Enxerto(s) de veia(s) safena(s), se óstios coronarianos comprometidos pela delaminação e não passíveis de reimplante		

DISSECÇÃO AGUDA DE AORTA

TRATAMENTO

- **TIPO B**

CLÍNICO

PERCUTÂNEO

CIRÚRGICO

Expansão

Hematoma

Isquemia visceral

Dor intratável

DISSECÇÃO AGUDA DE AORTA TIPO B TRATAMENTO ENDOVASCULAR

- Prevenir complicações
- Prevenir aumento da falsa luz
- Bom índice de sucesso (94,4%)
- Baixa mortalidade (0 - 6,3%)
- Exige acompanhamento por imagem
- Permanência da falsa luz

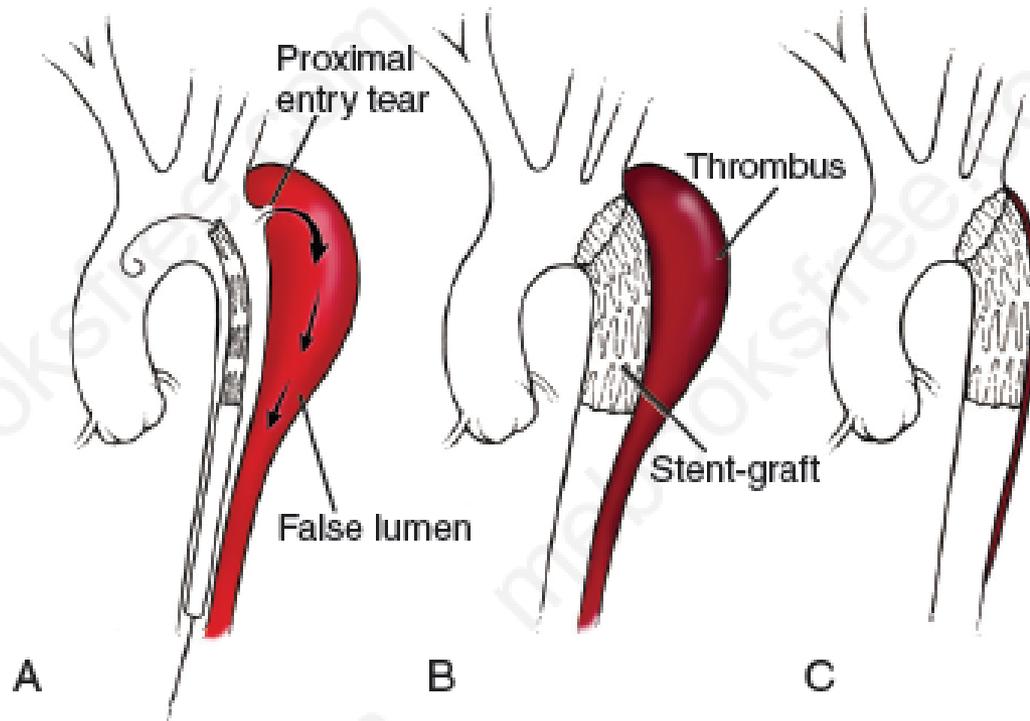
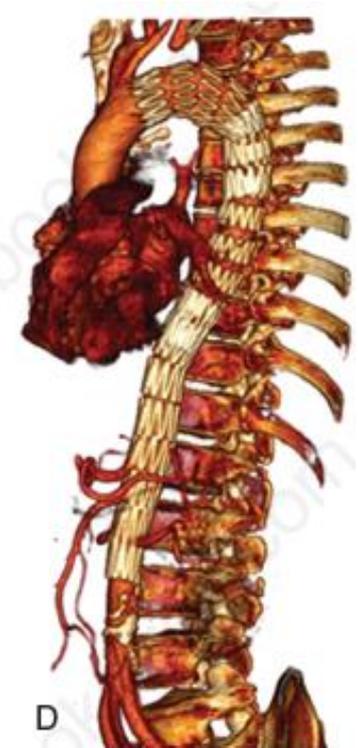


FIGURE 63.26 Thoracic endovascular aneurysm repair (TEVAR) after aortic dissection in the setting of aneurysmal enlargement of the false channel. **A**, Endograft is advanced to cover the proximal entry tear into the false channel. **B**, Sealing of the entry tear promotes thrombosis in the false lumen. **C**, Remodeling of the aorta occurs with expansion of the true lumen and a smaller, thrombosed false lumen.

TABLE 63.7 Indications for Thoracic Endovascular Aortic Repair for Type B Aortic Dissection*

- Rupture
- Impending rupture
- Malperfusion
- Hemorrhagic pleural effusion
- Refractory pain
- Refractory hypertension
- Aneurysmal dilation (>55 mm)
- Rapid increase in diameter
- Recurrent symptoms

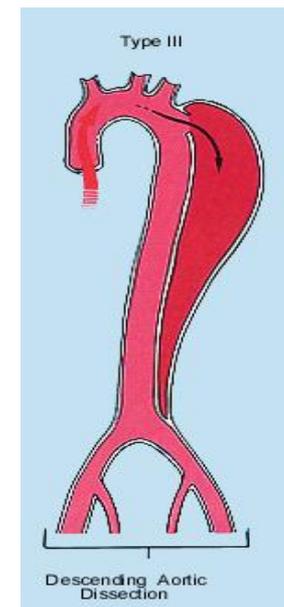
*Or open surgical repair if anatomy is unsuitable for TEVAR.



Diretriz Brasileira para tratar Dissecção Aguda Aorta Tipo B

Tabela 2. Recomendações para o tratamento das dissecções agudas tipo B.

Recomendações	Grau de Recomendação
1. Manejo clínico com analgesia e controle agressivo da PA.	A
2. Tratamento cirúrgico se dor persistente / recorrente, sinais de expansão, ruptura ou má perfusão de extremidades	A
3. Implante de endoprótese recoberta se dor persistente / recorrente, sinais de expansão, ruptura ou má perfusão de extremidades e anatomia favorável	A
4. Stent para desobstruir origem de ramo visceral, ou para manter fenestração aberta	C
5. Fenestração por balão e implante de stent se compressão grave da luz verdadeira, com ou sem reentrada distal	C
6. Implante de endoprótese recoberta na luz verdadeira para evitar dilatação aneurismática crônica da aorta	C
7. Implante de endoprótese recoberta na luz verdadeira para ocluir a lesão intimal e promover a trombose da falsa luz	C



Recommendations for treatment of aortic dissection

Recommendations	Class ^a	Level ^b	Ref. ^c
In all patients with AD, medical therapy including pain relief and blood pressure control is recommended.	I	C	
In patients with Type A AD, urgent surgery is recommended.	I	B	1,2
In patients with acute Type A AD and organ malperfusion, a hybrid approach (i.e. ascending aorta and/or arch replacement associated with any percutaneous aortic or branch artery procedure) should be considered.	IIa	B	2,118, 202–204, 227
In uncomplicated Type B AD, medical therapy should always be recommended.	I	C	
In uncomplicated Type B AD, TEVAR should be considered.	IIa	B	218,219
In complicated Type B AD, TEVAR is recommended.	I	C	
In complicated Type B AD, surgery may be considered.	IIb	C	

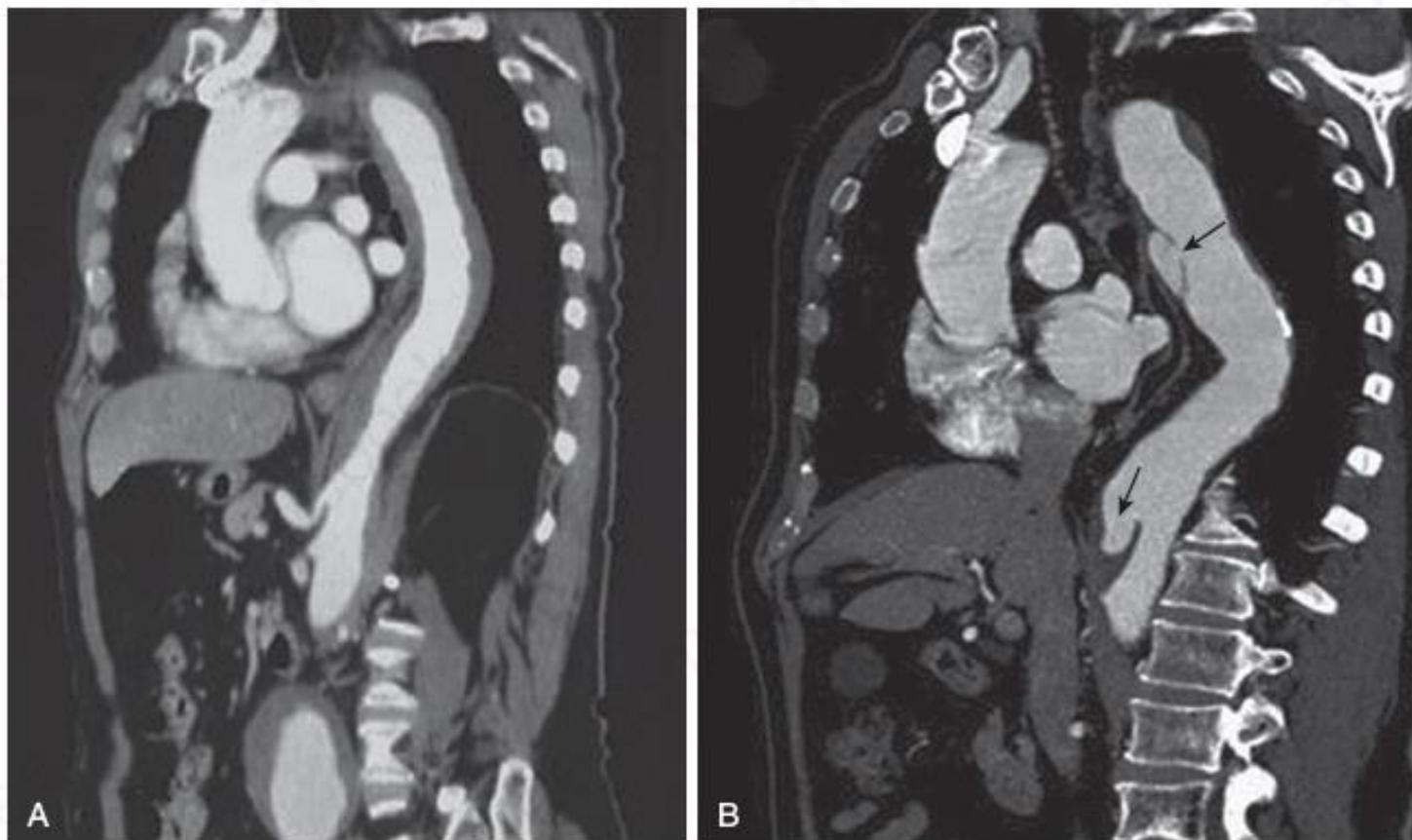
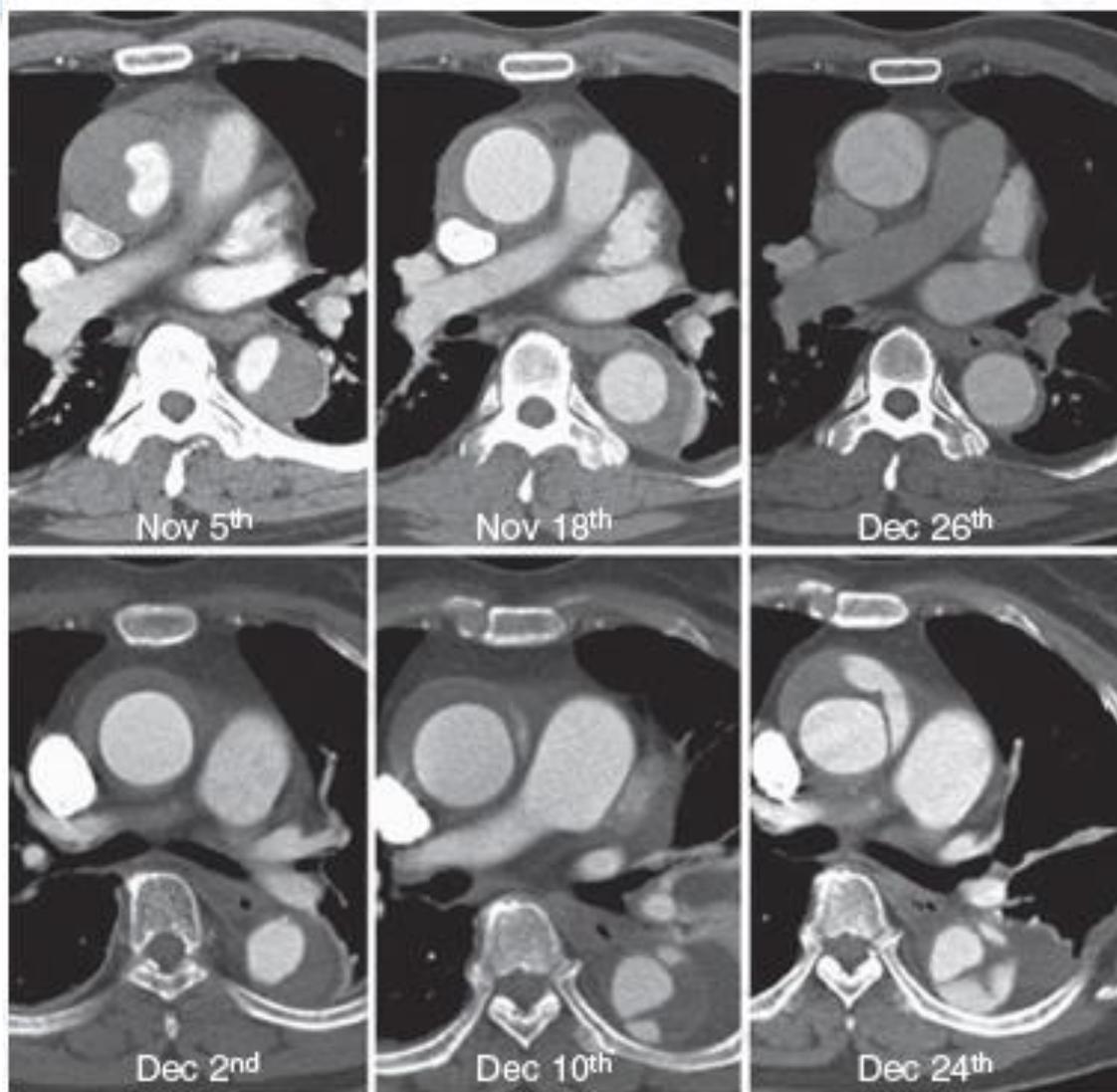


FIGURE 63.32 **A**, Computed tomography image of intramural hematoma (IMH) in descending thoracic aorta at diagnosis of acute aortic syndrome. **B**, Development of two ulcerlike projections in the same patient at 6 months (*black arrows*). (From Evangelista A, Czerny M, Nienaber C, et al. Interdisciplinary expert consensus on management of type B intramural haematoma and penetrating aortic ulcer. *Eur J Cardiothorac Surg* 2015;47:209-17.)



EFIGURE 63.33 Computed tomography images showing complete resorption of a proximal hematoma (**upper panels**) and development of typical aortic dissection during medical treatment (**lower panels**). (From Song JK. Aortic intramural hematoma: aspects of pathogenesis 2011. Herz 2011;36:488-97.)

Recommendations on the management of intramural haematoma

Recommendations	Class ^a	Level ^b
In all patients with IMH, medical therapy including pain relief and blood pressure control is recommended.	I	C
In cases of Type A IMH, urgent surgery is indicated.	I	C
In cases of Type B IMH, initial medical therapy under careful surveillance is recommended.	I	C
In uncomplicated ^c Type B IMH, repetitive imaging (MRI or CT) is indicated.	I	C
In complicated ^c Type B IMH, TEVAR should be considered.	IIa	C
In complicated ^c Type B IMH, surgery may be considered.	IIb	C

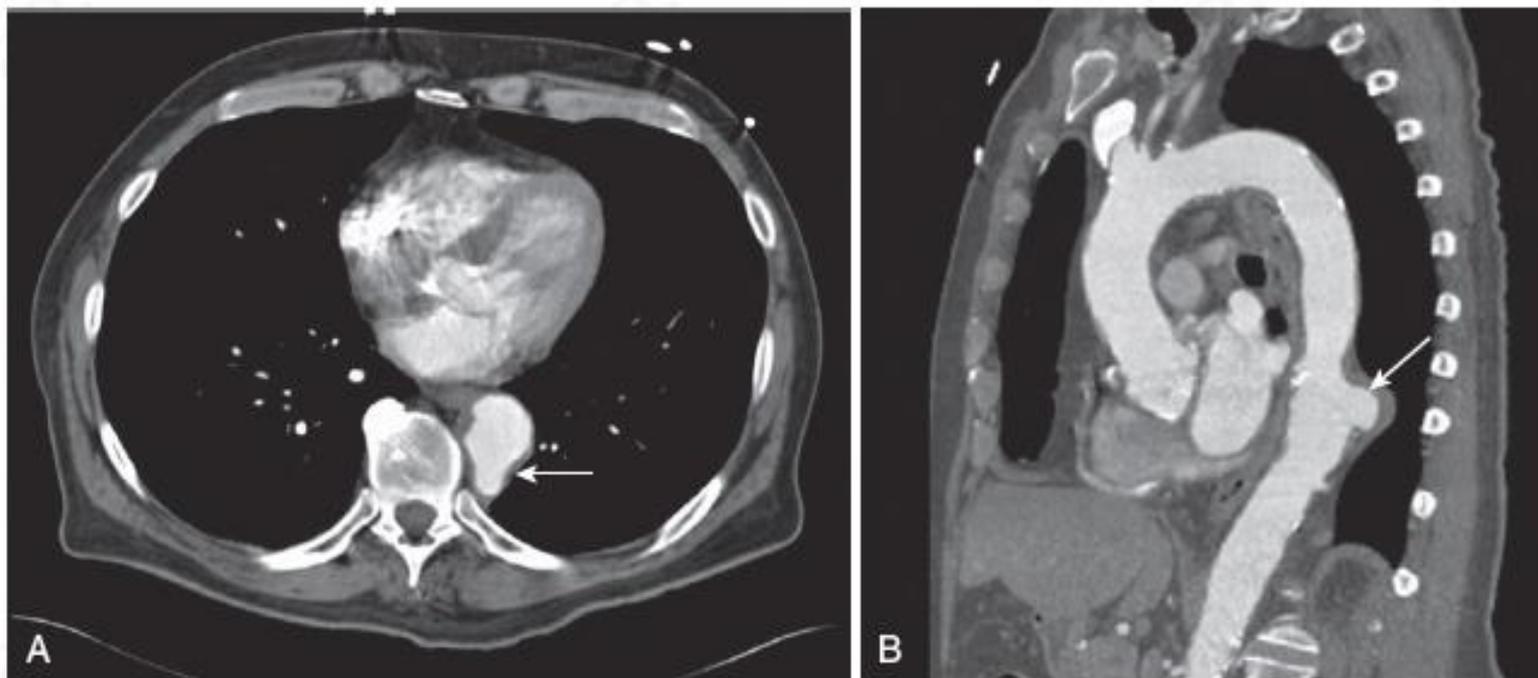


FIGURE 63.25 Contrast-enhanced computed tomography scan demonstrating an acute penetrating atherosclerotic aortic ulcer (PAU). **A**, Axial image demonstrating the typical focal outpouching of the aortic ulcer (*arrow*). **B**, Sagittal image demonstrating the PAU (*arrow*) with associated intramural hematoma. Symptomatic PAU has an increased risk of aortic rupture and is often amenable to endovascular repair. (From Braverman AC. Aortic dissection. In Levine GN, editor. Color Atlas of Cardiovascular Disease. New Delhi, India: Jaypee Brothers Medical Publishers; 2015, pp 895-903.)

Recommendations on management of penetrating aortic ulcer

Recommendations	Class ^a	Level ^b
In all patients with PAU, medical therapy including pain relief and blood pressure control is recommended.	I	C
In the case of Type A PAU, surgery should be considered.	IIa	C
In the case of Type B PAU, initial medical therapy under careful surveillance is recommended.	I	C
In uncomplicated Type B PAU, repetitive imaging (MRI or CT) is indicated.	I	C
In complicated Type B PAU, TEVAR should be considered.	IIa	C
In complicated Type B PAU, surgery may be considered.	IIb	C

Diretriz SBC para acompanhamento dissecação

Tabela III - Recomendações para seguimento de pacientes com dissecação aórtica.

Recomendações	Classe de evidência	Nível de evidência
Manejo contínuo da HAS com betabloqueadores	I	C
Exame de imagem (RMN, CT, ETC)	I	C
Restrição moderada da atividade física	I	C

Tópicos

- *Síndromes aórticas agudas (dissecção da aorta, hematoma intramural e úlcera penetrante aterosclerótica)*
- *Aneurismas da aorta torácica*
- *Aortopatias inflamatórias (Takayasu, conectivopatias)*
- *Ateroembolismo*

Aneurismas da Aorta Torácica

Incidência 5-10 / 100.000

Causa e Evolução natural dependem da localização

<i>Ao Ascendente</i>	<i>60%</i>
<i>Ao Descendente</i>	<i>35%</i>
<i>Arco Ao</i>	<i><10%</i>
<i>Toracoabdominal</i>	<i><10%</i>

Aneurismas da Aorta Torácica

Causas

Hereditárias, genéticas (congênitas)

Degenerativas (“ateroscleróticas”)

Mecânicas

Inflamatórias

Infeciosas

Fatores de Risco

Tabagismo

HAS

Idade

DPOC

DAC

História familiar

Degeneração relacionada a idade + HAS

Aneurismas da Aorta Torácica

Necrose ou Degeneração Cística da Camada Média

Síndromes de Marfan e outras de causa genética

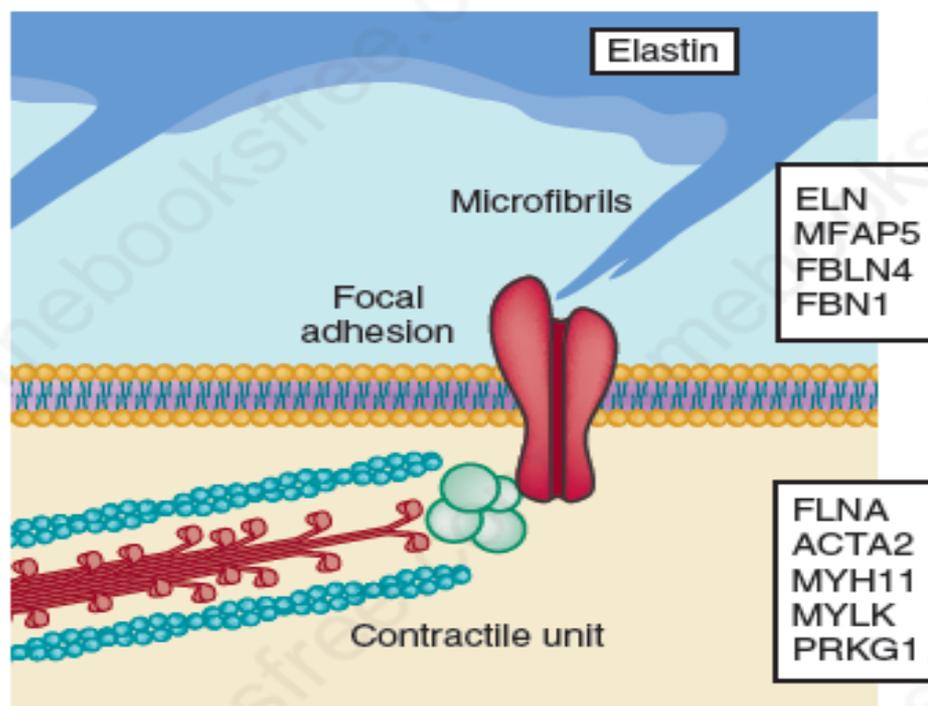


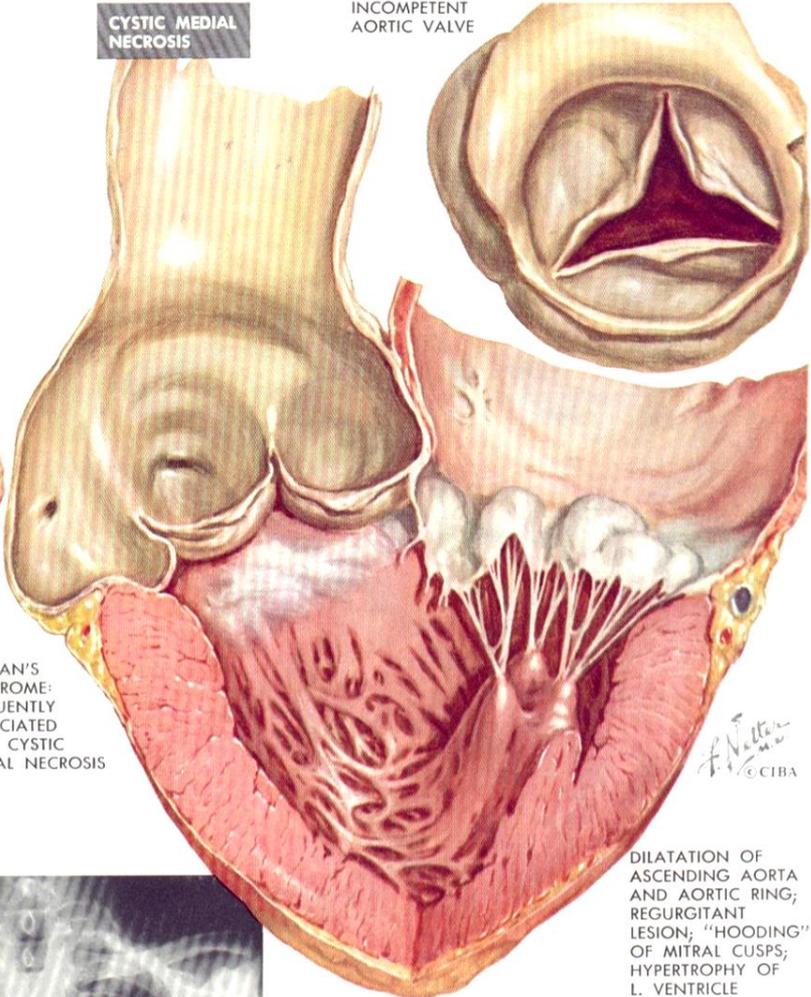
FIGURE 63.3 Elastin-contraction unit in smooth muscle cells. The altered genes that predispose to thoracic aortic aneurysms and acute aortic dissections are indicated (see Table 63.1). The location of the gene indicates where its protein product is located in the elastin-contraction unit. (From Karimi A, Milewicz D. Structure of the elastin-contraction units in the thoracic aorta and how genes that cause thoracic aortic aneurysms and dissections disrupt this structure. *Can J Cardiol* 2016;32:26-34.)



MARFAN'S SYNDROME: FREQUENTLY ASSOCIATED WITH CYSTIC MEDIAL NECROSIS

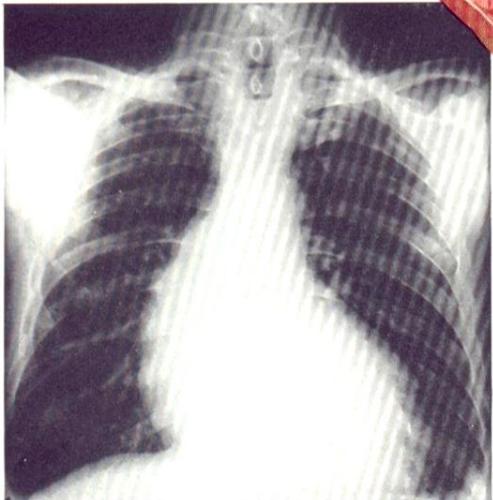
CYSTIC MEDIAL NECROSIS

INCOMPETENT AORTIC VALVE

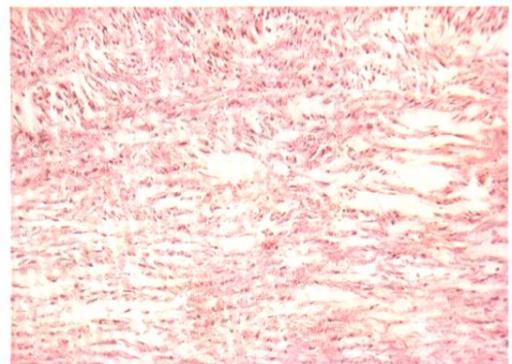


DILATATION OF ASCENDING AORTA AND AORTIC RING; REGURGITANT LESION; "HOODING" OF MITRAL CUSPS; HYPERTROPHY OF L. VENTRICLE

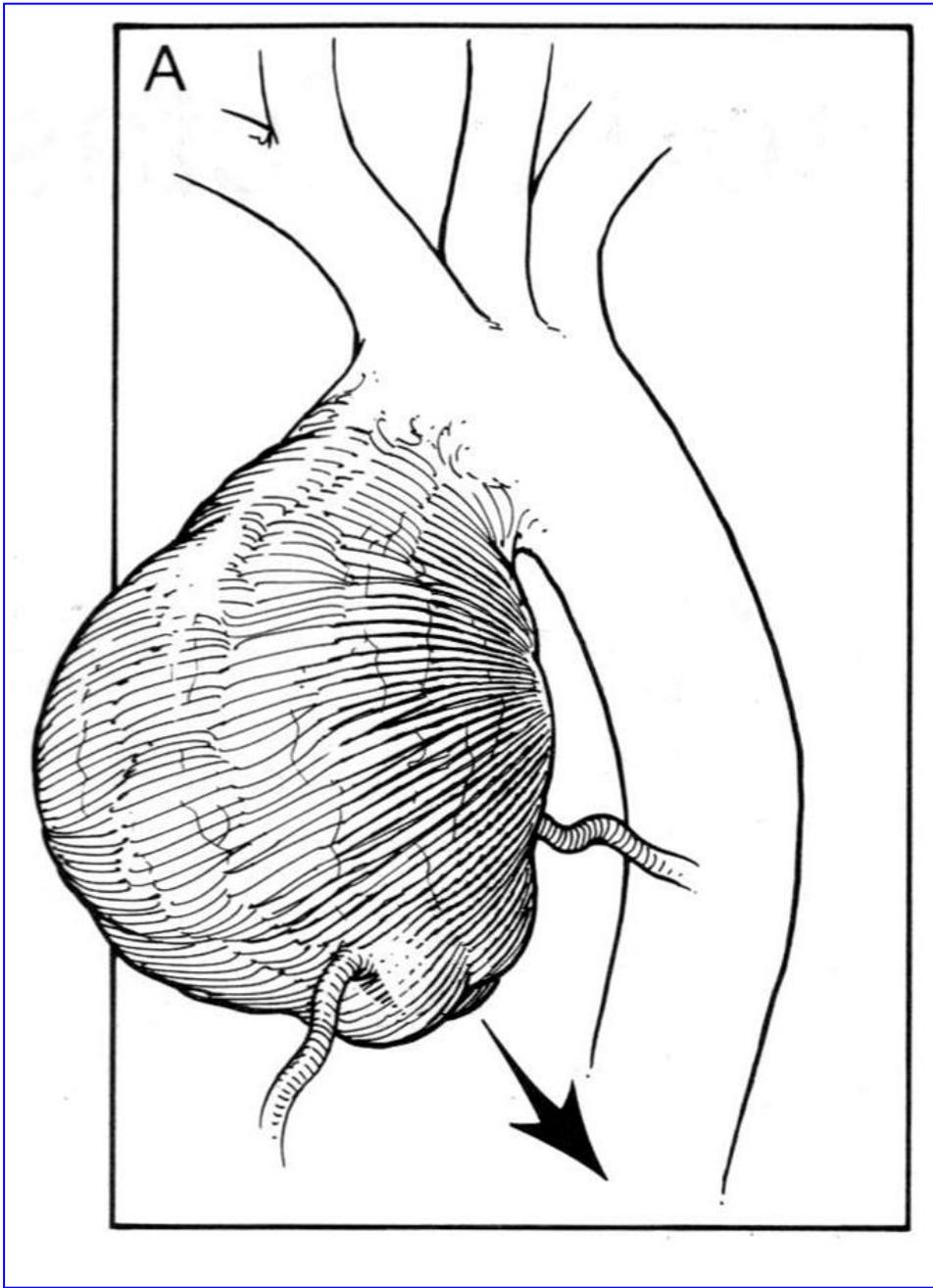
F. Netter
© CIBA



DILATION OF ASCENDING AORTA



CYSTIC MEDIAL NECROSIS OF AORTA



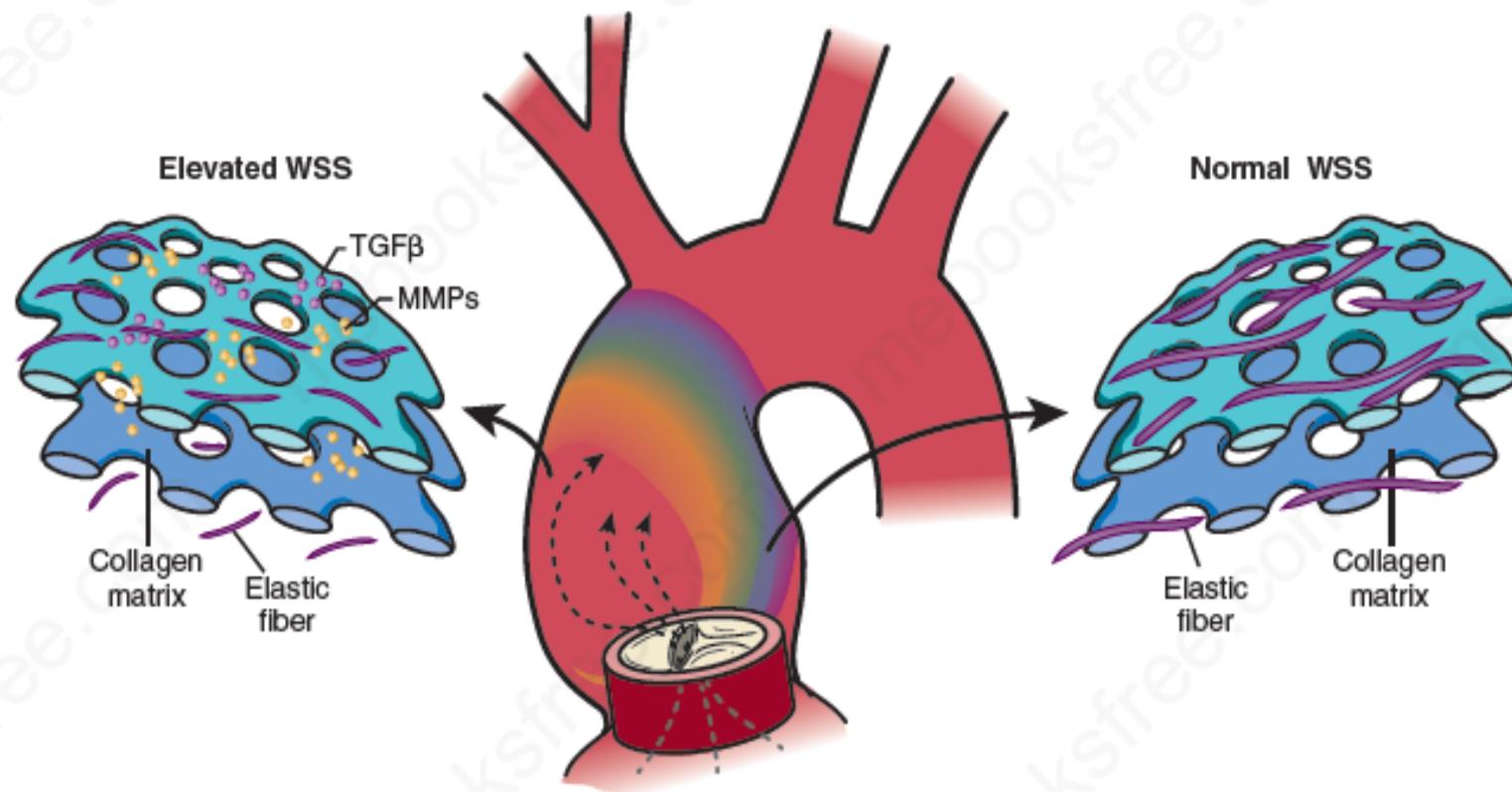


FIGURE 63.7 Bicuspid aortic valve (BAV) aortopathy is related to abnormal aortic flow and elevated wall shear stress. Four-dimensional flow cardiac magnetic resonance imaging (4D flow CMR) is used to assess the relation between wall shear stress (WSS) and regional aortic tissue remodeling in BAV patients. Elevated aortic WSS generated by aberrant flow from cusp fusion corresponds to more severe extracellular matrix (ECM) dysregulation than adjacent regions of normal WSS in the same patient's aorta. Elastic fiber degeneration is more severe in regions of elevated WSS (less elastin, thinner fibers, and greater distances between laminae), where higher concentrations of mediators of ECM dysregulation (matrix metalloproteinases [MMPs] and transforming growth factor beta [TGFβ]) are also observed. These data implicate valve-related hemodynamics as a contributing factor to BAV aortopathy. (From Guzzardi DG, Barker AJ, van Ooij P, et al. Valve-related hemodynamics mediate human bicuspid aortopathy insights from wall shear stress mapping. *J Am Coll Cardiol* 2016;66:892-900.)

Table 5. Risk of Complications by Aortic Diameter and Body Surface Area With Aortic Size Index Given Within Chart

	Aortic Size (cm)									
	3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.5	8.0
BSA										
1.30	2.69	3.08	3.46	3.85	4.23	4.62	5.00	5.38	5.77	6.15
1.40	2.50	2.86	3.21	3.57	3.93	4.29	4.64	5.00	5.36	5.71
1.50	2.33	2.67	3.00	3.33	3.67	4.00	4.33	4.67	5.00	5.33
1.60	2.19	2.50	2.80	3.13	3.44	3.75	4.06	4.38	4.69	5.00
1.70	2.05	2.35	2.65	2.94	3.24	3.53	3.82	4.12	4.41	4.71
1.80	1.94	2.22	2.50	2.78	3.06	3.33	3.61	3.89	4.17	4.44
1.90	1.84	2.11	2.37	2.63	2.89	3.16	3.42	3.68	3.95	4.22
2.00	1.75	2.00	2.25	2.50	2.75	3.00	3.25	3.50	3.75	4.00
2.10	1.67	1.90	2.14	2.38	2.62	2.86	3.10	3.33	3.57	3.80
2.20	1.59	1.82	2.05	2.27	2.50	2.72	2.95	3.18	3.41	3.64
2.30	1.52	1.74	1.96	2.17	2.39	2.61	2.83	3.04	3.26	3.48
2.40	1.46	1.67	1.88	2.08	2.29	2.50	2.71	2.92	3.13	3.33
2.50	1.40	1.60	1.80	2.00	2.20	2.40	2.60	2.80	3.00	3.20

□ = low risk (~1% per yr); ■ = moderate risk (~8% per yr); ■ = severe risk (~20% per yr).

White area indicates low risk, light gray area indicates moderate risk, and dark gray area indicates severe risk.

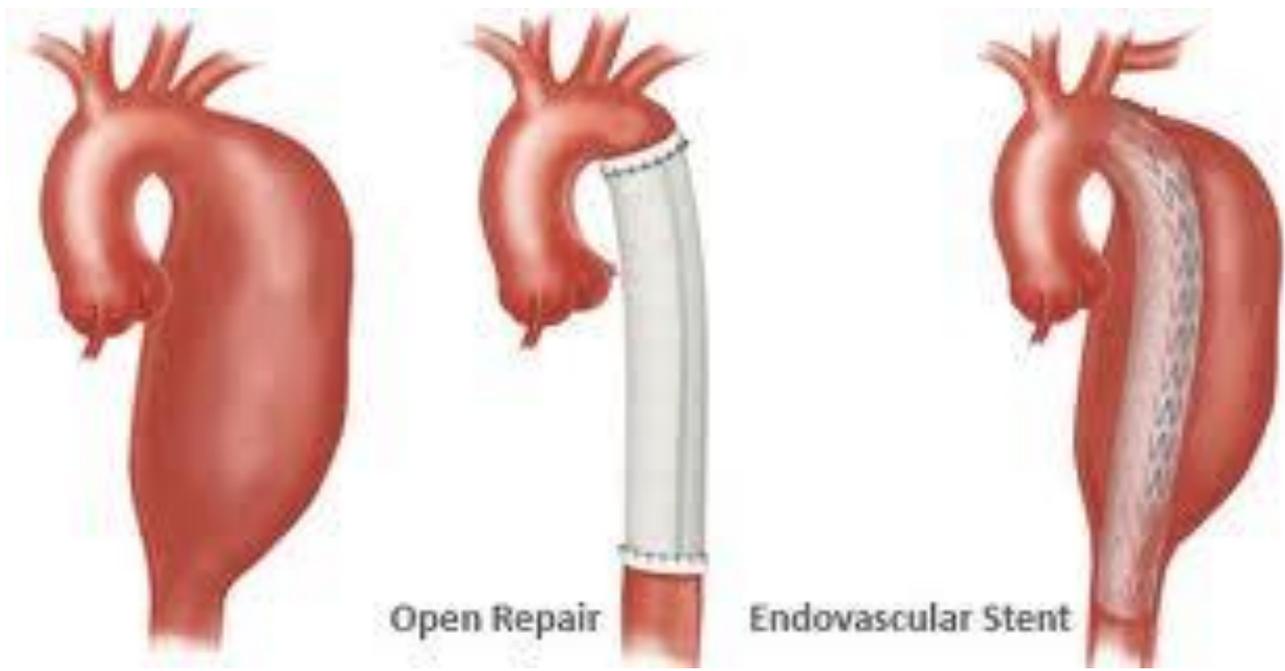
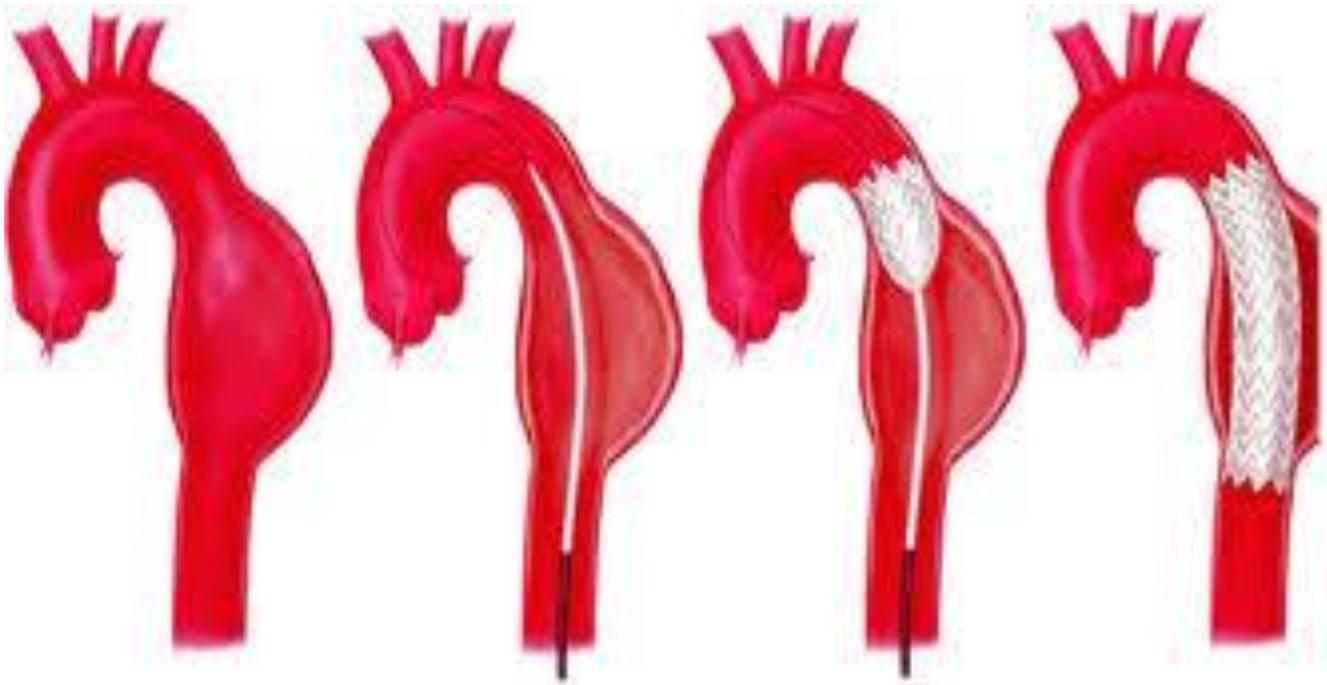
BSA = body surface area.

Recommendations	Class ^a	Level ^b
Patients with known BAV should undergo an initial TTE to assess the diameters of the aortic root and ascending aorta.	I	C
Cardiac MRI or CT is indicated in patients with BAV when the morphology of the aortic root and the ascending aorta cannot be accurately assessed by TTE.	I	C
Serial measurement of the aortic root and ascending aorta is indicated in every patient with BAV, with an interval depending on aortic size, increase in size and family history	I	C
In the case of a diameter of the aortic root or the ascending aorta >45 mm or an increase >3 mm/year measured by echocardiography, annual measurement of aortic diameter is indicated.	I	C
In the case of aortic diameter >50 mm or an increase >3 mm/year measured by echocardiography, confirmation of the measurement is indicated, using another imaging modality (CT or MRI).	I	C
In cases of BAV, surgery of the ascending aorta is indicated in case of: <ul style="list-style-type: none"> • aortic root or ascending aortic diameter >55 mm. • aortic root or ascending aortic diameter >50 mm in the presence of other risk factors.^c • aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled. 	I	C

Recommendations on interventions on ascending aortic aneurysms

Recommendations	Class ^a	Level ^b
Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter ^c ≥ 50 mm for patients with Marfan syndrome.	I	C
Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameters: <ul style="list-style-type: none"> • ≥ 45 mm for patients with Marfan syndrome with risk factors.^d • ≥ 50 mm for patients with bicuspid valve with risk factors.^{e,f} • ≥ 55 mm for other patients with no elastopathy.^{g,h} 	IIa	C
Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid progression, aortic valve regurgitation, planned pregnancy, and patient's preference.	IIb	C

Interventions on aortic arch aneurysms		
Surgery should be considered in patients who have isolated aortic arch aneurysm with maximal diameter ≥ 55 mm.	IIa	C
Aortic arch repair may be considered in patients with aortic arch aneurysm who already have an indication for surgery of an adjacent aneurysm located in the ascending or descending aorta.	IIb	C



Open Repair

Endovascular Stent

Interventions on descending aortic aneurysms		
TEVAR should be considered, rather than surgery, when anatomy is suitable.	IIa	C
TEVAR should be considered in patients who have descending aortic aneurysm with maximal diameter ≥ 55 mm.	IIa	C
When TEVAR is not technically possible, surgery should be considered in patients who have descending aortic aneurysm with maximal diameter ≥ 60 mm.	IIa	C
When intervention is indicated, in cases of Marfan syndrome or other elastopathies, surgery should be indicated rather than TEVAR.	IIa	C

Diretriz Brasileira para AAT

Tabela 4. Recomendações para tratamento cirúrgico dos aneurismas crônicos da aorta torácica/toráco-abdominal.

Recomendações	Grau de Recomendação
Aorta Ascendente 1	
Cirurgia se sintoma compressivos, insuficiência aórtica, ou diâmetro aórtico ≥ 6 cm ² .	A
Em síndrome de Marfan, cirurgia profilática se diâmetro $\geq 5,5$ cm ou $\geq 5,0$ cm em casos com história familiar de dissecação ou morte súbita.	D
Aorta Descendente 1	
Cirurgia se sintomas ou diâmetro aórtico ≥ 6 cm ² .	A
Implante de endoprótese se diâmetro aórtico $\geq 6,0$ cm e anatomia for favorável.	A

Tópicos

- *Síndromes aórticas agudas (dissecção da aorta, hematoma intramural e úlcera penetrante aterosclerótica)*
- *Aneurismas da aorta torácica*
- *Aortopatias inflamatórias (Takayasu, conectivopatias)*
- *Ateroembolismo*

Aortopatias Inflamatórias

Vasculites inflamatórias não infecciosas

Arterite de células gigantes (ou temporal)

Arterite de Takayasu

Doenças de Becet

Buerger

Kawasaki

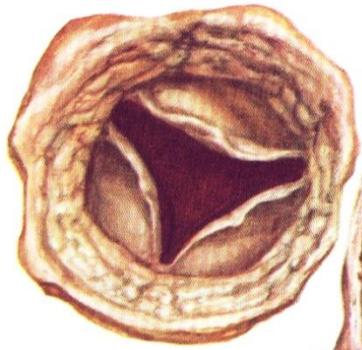
Espondilite anquilosante

Reiter

Menos comuns, infecciosas:

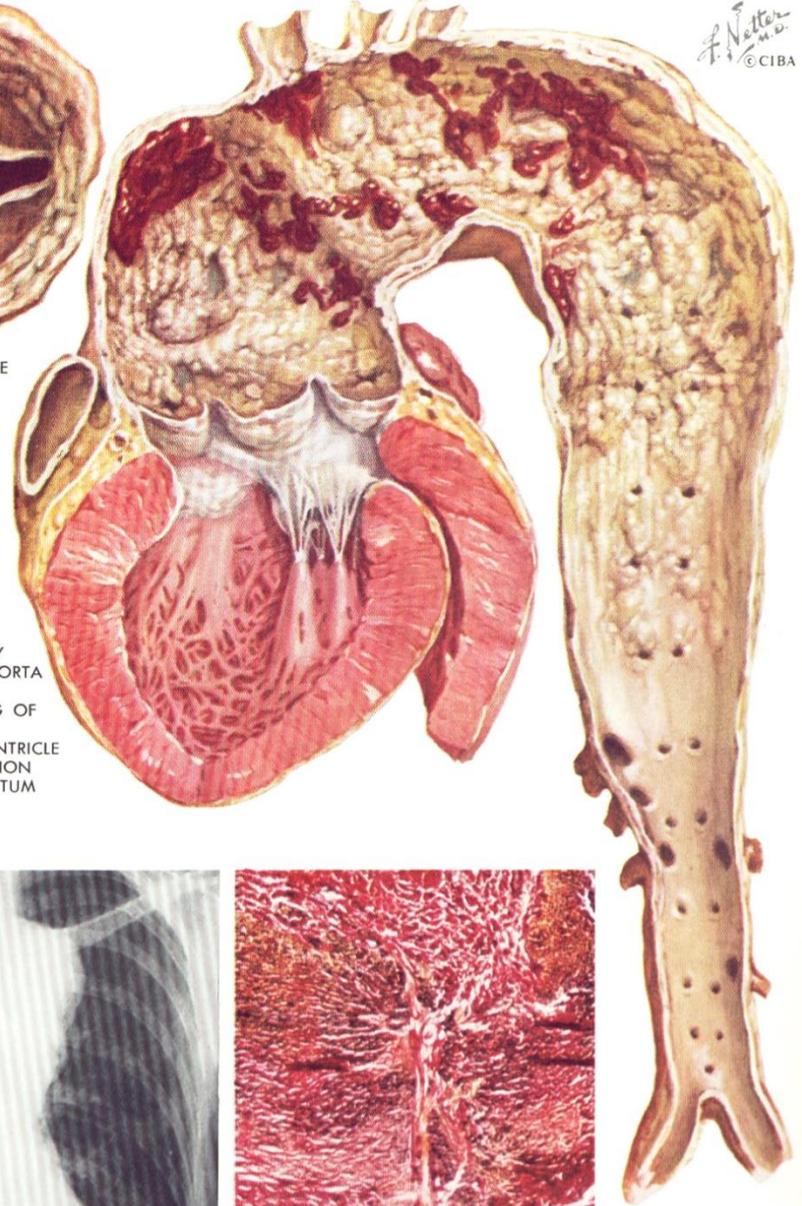
Infecções por Estafilococos, Salmonelas, micobactérias, Treponema pallidum

Aneurismas micóticos

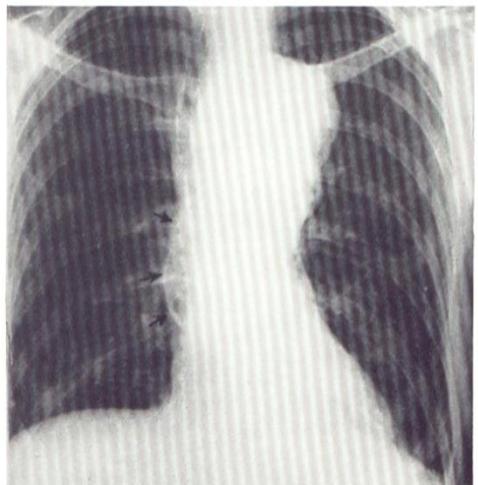


INCOMPETENT AORTIC VALVE WITH TAUT, SEPARATED CUSPS VIEWED FROM ABOVE

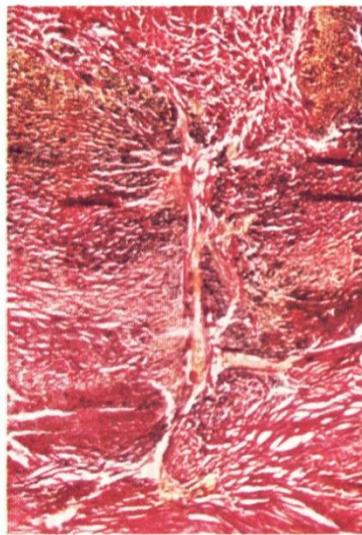
SYPHILITIC HEART DISEASE



DILATED AND MARKEDLY SCLEROTIC THORACIC AORTA WITH WIDENED AORTIC RING AND NARROWING OF CORONARY OSTIA; HYPERTROPHY OF L. VENTRICLE WITH REGURGITANT LESION UPON VENTRICULAR SEPTUM



CALCIFICATION OF ASCENDING AORTA AND DILATATION OF THORACIC AORTA



STELLATE SCAR IN MEDIA OF AORTA

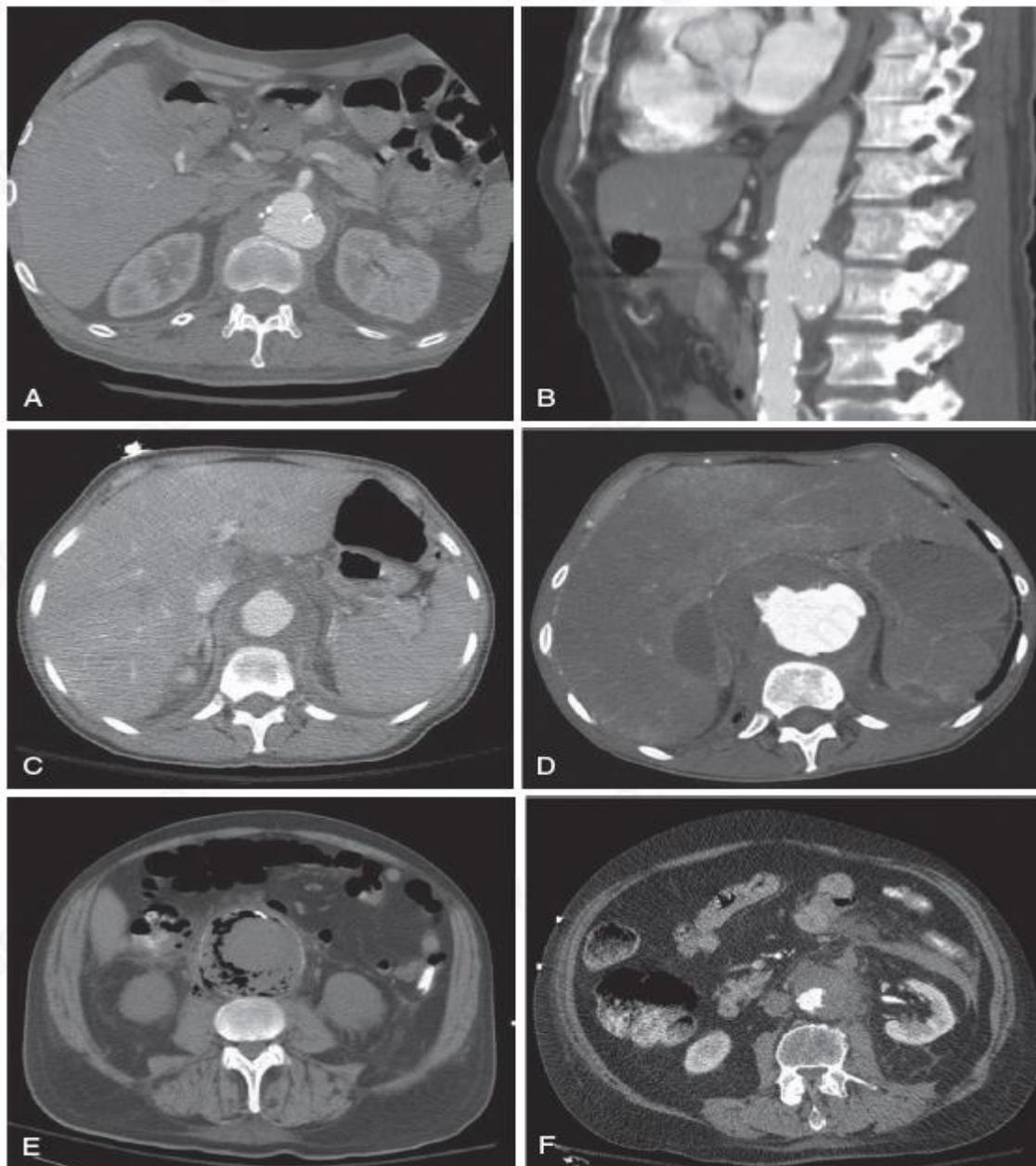


FIGURE 63.26 Typical features of mycotic aneurysms (MAs) on computed tomography. **A, B**, MAs classically present as focal outpouchings of the vascular wall, as seen on axial (**A**) and sagittal (**B**) images. In this case, there is calcification of the aortic wall, which may have been a nidus for infection. **C, D**, MAs tend to grow rapidly, as in this case, with images at initial presentation (**C**) and 2 weeks later (**D**). Also noted is irregularity of the aortic wall and mural thrombus. **E**, Inflammatory changes are common, as in this case with gas in the aortic wall in an MA due to infection by adjacent diverticulitis. **F**, MAs are prone to rupture. (From Deipolyi AR, Rho J, Khademhosseini A, Oklu R. Diagnosis and management of mycotic aneurysms. *Clin Imaging* 2016;40:256-62.)

Kawasaki disease

- Also called mucocutaneous lymph node syndrome
- Caused by systemic necrosis of vasculature from idiopathic (nonspecific) immune activation
- Affects small-medium arteries
- Affects children
- Presents as fever, red mucosal lesions (strawberry tongue), lymphadenopathy, and rash
- Treat with IVIG and aspirin



Takayasu's arteritis



Image from Wikipedia.org; licensing details available at wikipedia.org/wiki/File:Takayasu_Arteritis.jpg

Takayasu arteritis is a rare, large-vessel vasculitis of unknown aetiology,

Young women

Asian population

2.6 per million inhabitants

The thoracic aorta and its major branches are the most frequent locations

followed by the abdominal aorta.

Treatment: steroids

Secondline agents include methotrexate, azathioprine, and anti-tumour necrosis factor-alpha agents. (ESC Guidelines 2018)

Tópicos

- *Síndromes aórticas agudas (dissecção da aorta, hematoma intramural e úlcera penetrante aterosclerótica)*
- *Aneurismas da aorta torácica*
- *Aortopatias inflamatórias (Takayasu, conectivopatias)*
- *Ateroembolismo*

Ateroembolismo

- *Processo aterosclerótico: placas lipídicas na íntima*
- *Inflamação secundária, tecido fibroso, erosão, trombos:*

Complicações

Trombóticas (tromboembolismo)

Oclui artérias maiores: AVC, AIT, infarto renal, isquemias periféricas

Ateroscleróticas (colesterol/ateroembolismo)

Oclui artérias menores e arteríolas: isquemias MIs, Insuf. renal, isquemia mesentérica

Ateroembolismo

- *Tratamento*
- Antiplaquetários*
- Anticoagulantes*
- Estatinas (lipid lowering agents)*

Recommendations on management of aortic plaque

Recommendations	Class ^a	Level ^b
In the presence of aortic atherosclerosis, general preventive measures to control risk factors are indicated.	I	C
In the case of aortic plaque detected during the diagnostic work-up after stroke or peripheral embolism, anticoagulation or antiplatelet therapy should be considered. The choice between the two strategies depends on comorbidities and other indications for these treatments.	IIa	C
Prophylactic surgery to remove high-risk aortic plaque is not recommended.	III	C

Curso Auxiliar Preparatório para a Prova de Título de Especialista
em Cardiologia, edição 2019.

Aortopatias, dissecação e aneurisma

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Curso Auxiliar Preparatório para a Prova de Título de
Especialista em Cardiologia, edição 2019.

- Tema de sua aula: **Aortopatias, dissecação e aneurisma.**
 - Duração da aula: entre 20min. - 30min.
 - Data da gravação: **15 de setembro de 2018**
 - Horário: **17:00h**
 - Material para apresentação: **PowerPoint** (deverá ser entregue com antecedência por e-mail)
 - Material para avaliação do cardiologista: **Elaborar 10 questões de múltipla escolha no formato WORD.**

Descritivo

- Síndromes aórticas agudas (dissecção da aorta, hematoma intramural e úlcera penetrante aterosclerótica); Aneurismas da aorta torácica; Aortopatias inflamatórias (Takayasu, conectivopatias); Ateroembolismo.

- O senhor precisará preparar uma aula no tema que foi proposto, respeitando as referências indicadas.
-
- Haverá um estúdio no congresso preparado para sua gravação. Seu horário resercado é:
 -
 - **Data da gravação: 15 de setembro de 2018**
 - **Horário: 17:00h**
 -
- O senhor precisará preparar 10 questões sobre sua aula em formato word, multipla escolha com opções de A) à E) (5 opções)
-
- Toda sua aula precisará estar no slide padrão enviado em anexo.
-
- Envio também um arquivo com instruções.