

2014 version

# 2014 ESC Guidelines on the Diagnosis and Treatment of Aortic Diseases



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# 2014 ESC Guidelines on the Diagnosis and Treatment of Aortic Diseases

Chairpersons

**Raimund Erbel (Germany) and Victor Aboyans (France)**

# 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**

Chairpersons:

**Raimund Erbel (Germany), Victor Aboyans (France)**

Authors/Task Force members:

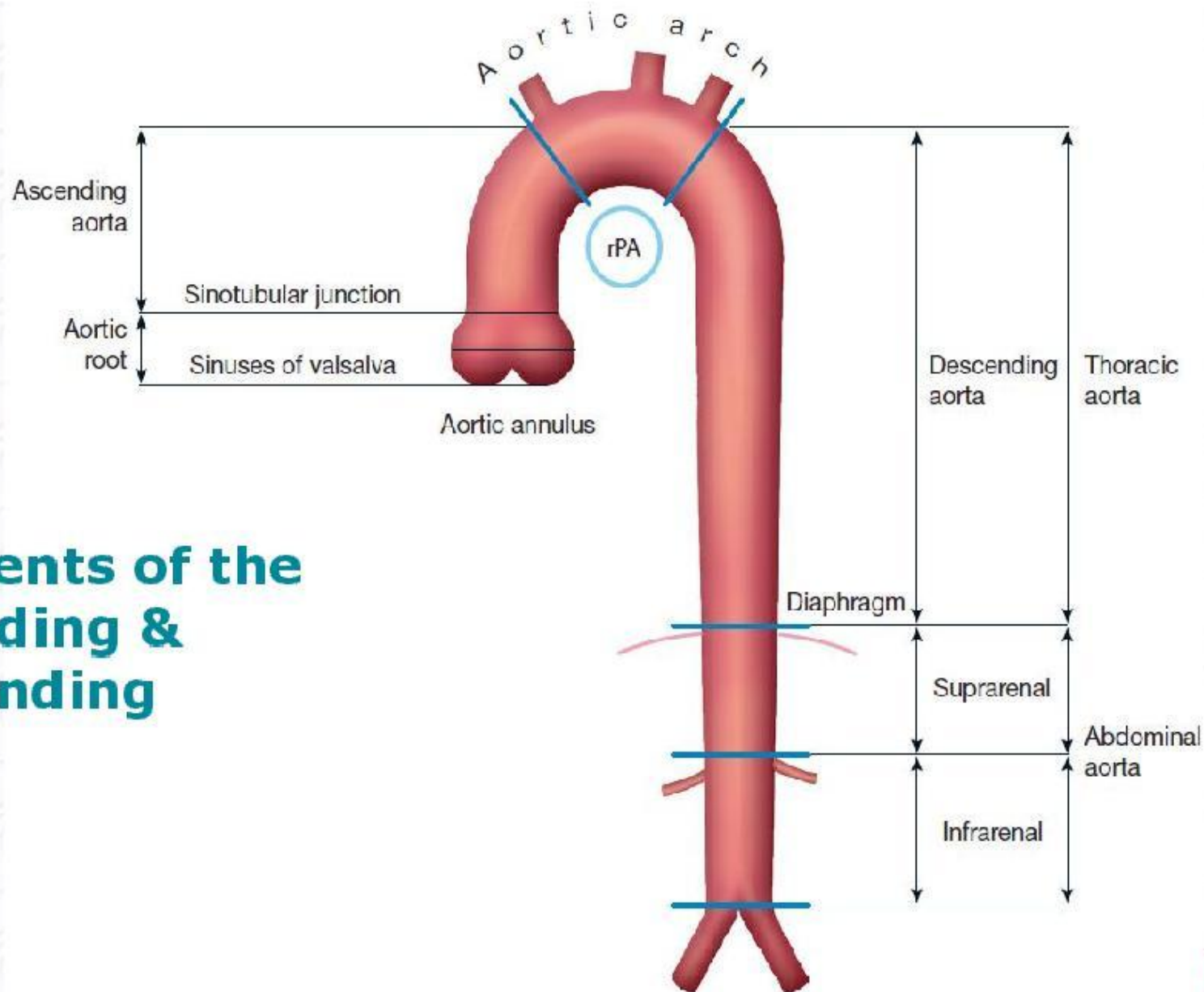
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# Classes of recommendations

Classes of recommendations	Definition	Suggested wording to use
<b>Class I</b>	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.	Is recommended/ is indicated.
<b>Class II</b>	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.	
<b>Class IIa</b>	<i>Weight of evidence/opinion is in favour of usefulness/efficacy.</i>	<i>Should be considered.</i>
<b>Class IIb</b>	<i>Usefulness/efficacy is less well established by evidence/opinion.</i>	May be considered.
<b>Class III</b>	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.	Is not recommended.

# Levels of evidence

<b>Level of Evidence A</b>	<b>Data derived from multiple randomized clinical trials or meta-analyses.</b>
<b>Level of Evidence B</b>	<b>Data derived from a single randomized clinical trial or large non-randomized studies.</b>
<b>Level of Evidence C</b>	<b>Consensus of opinion of the experts and/or small studies, retrospective studies, registries.</b>



## Segments of the ascending & descending aorta

## Comparison of methods for imaging the aorta

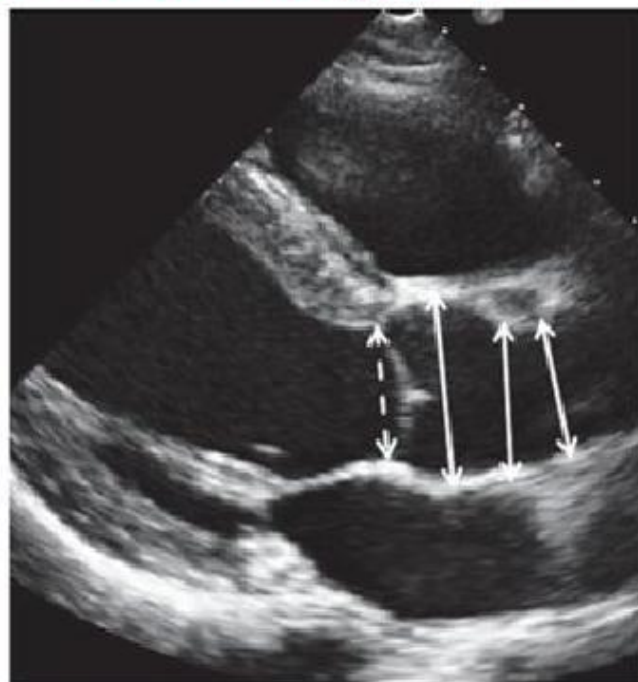
Advantages/disadvantages	TTE	TOE	CT	MRI	Aortography
Ease of use	+++	++	+++	++	+
Diagnostic reliability	+	+++	+++	+++	++
Bedside/interventional use	++	++	-	-	++
Serial examinations	++	+	++(+)	+++	-
Aortic wall visualization	+	+++	+++	+++	-
Cost	-	-	--	---	---
Radiation	0	0	---	-	--
Nephrotoxicity	0	0	---	--	---

+ means a positive remark and — means a negative remark. The number of signs indicates the estimated potential value

++(+) only for follow-up after aortic stenting (metallic struts), otherwise limit radiation

Parasternal long-axis and suprasternal imaging of the aorta indicating the points of diameter measurements of the aortic root and aortic arch for transthoracic echocardiography. Sinuses of Valsalva; sinotubular junction; ascending aorta. Also shown, the measurement of the aortic valvular ring.

**Parasternal long-axis view**



**Suprasternal long-axis view**



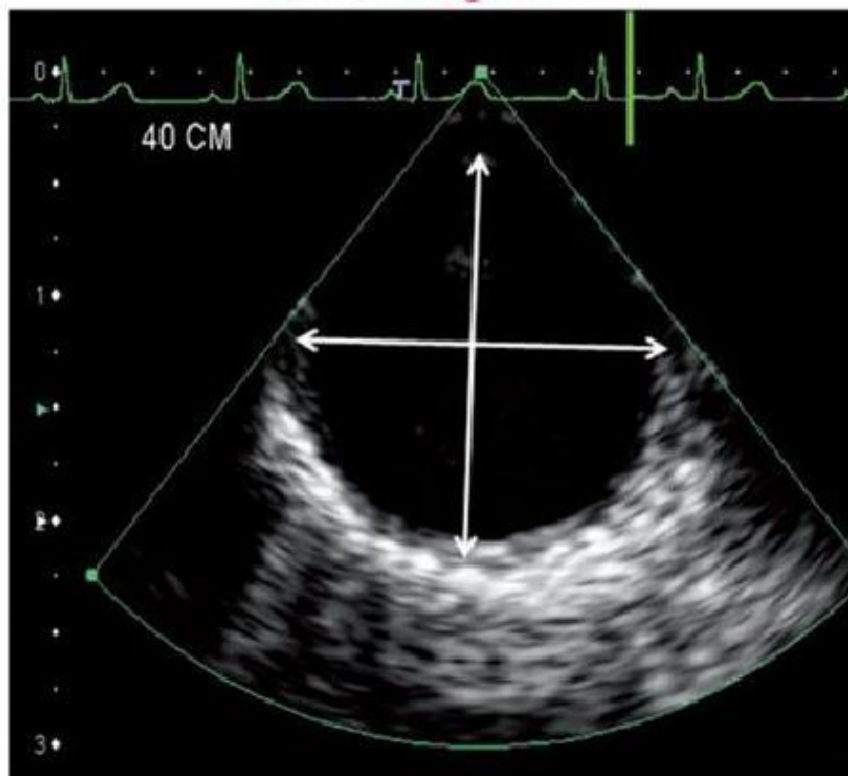
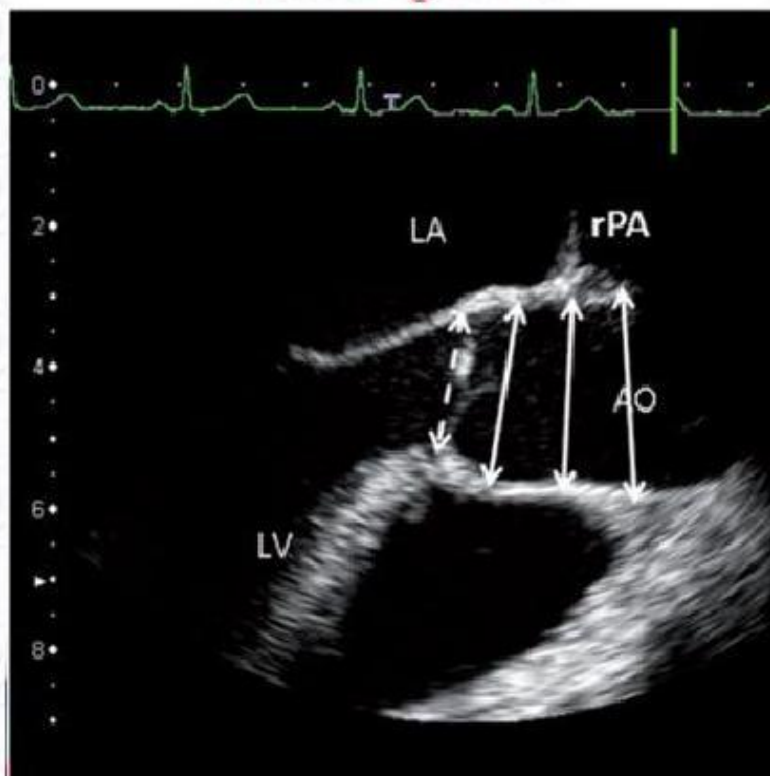


Transoesophageal echocardiographic long-axis and cross-sectional image of the ascending and descending aorta, indicating the points of diameter measurements: sinus of Valsalva, beginning of the ascending aorta, ascending aorta at the level of the right pulmonary artery. Also shown, the measurement of the aortic valvular ring.

### Transoesophageal Echocardiogram

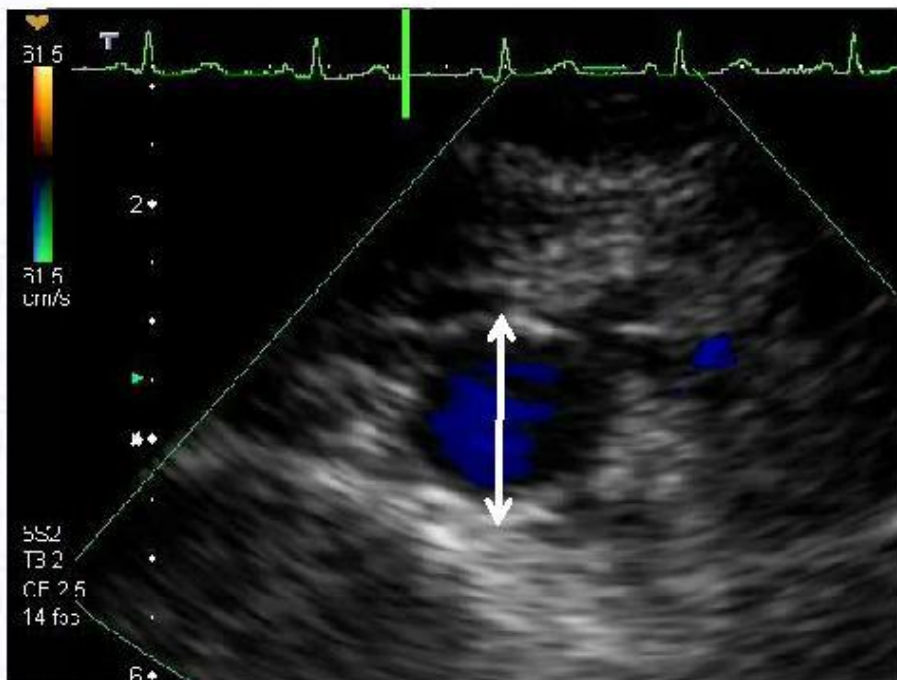
ascending aorta

descending aorta

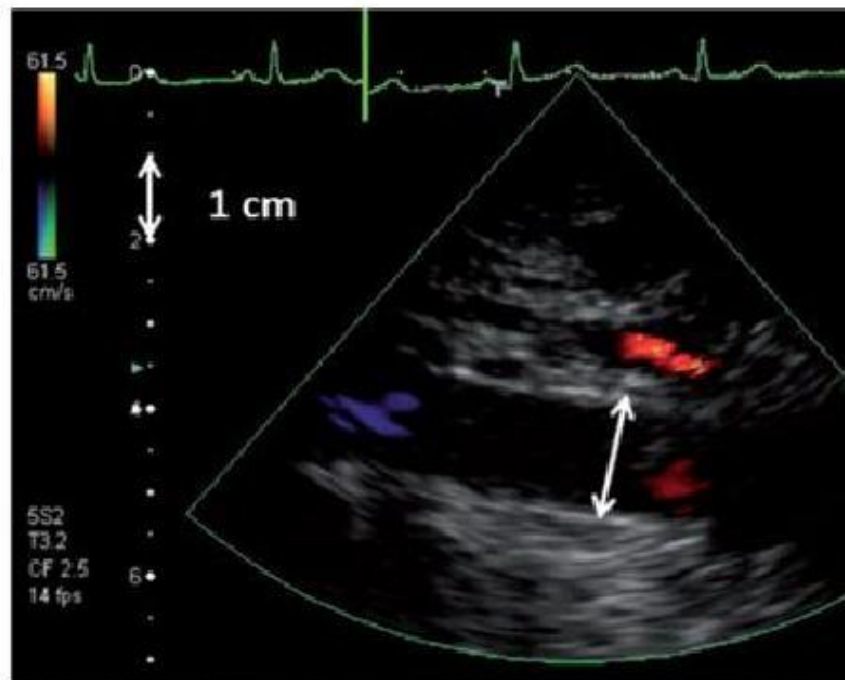


# Cross-sectional and long-axis imaging of the abdominal aorta indicating the points of diameter measurements for ultrasound

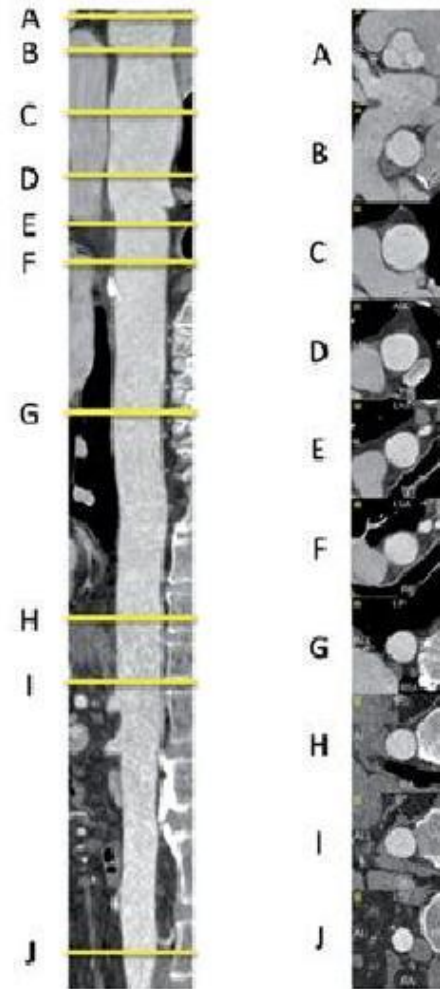
## Abdominal short-axis view



## Abdominal long-axis view



Thoracic and abdominal aorta in a three-dimensional reconstruction (left lateral image), parasagittal multiplanar reconstruction (MPR) along the centreline (left middle part), straightened-MPR along the centreline with given landmarks (A – J) (right side), orthogonal to the centreline orientated cross-sections at the landmarks (A – J)



# Imaging the aorta

Recommendations	Class	Level
It is recommended to measure diameters at pre-specified anatomical landmarks, perpendicular to the longitudinal axis.	<b>I</b>	<b>C</b>
In case of repetitive imaging of the aorta over time to assess change in diameter, it is recommended to use the imaging modality with the lowest iatrogenic risk.	<b>I</b>	<b>C</b>
In case of repetitive imaging of the aorta over time to assess change in diameter, it is recommended to use the same imaging modality with a similar method of measurement.	<b>I</b>	<b>C</b>
It is recommended to report all relevant aortic diameters and abnormalities according to the aortic segmentation.	<b>I</b>	<b>C</b>
It is recommended to assess renal function, pregnancy, and history of allergy to contrast media in order to select the optimal imaging modality of the aorta with minimal radiation exposure, except for emergency cases.	<b>I</b>	<b>C</b>
The risk of radiation exposure should be assessed, especially in younger adults and in those undergoing repetitive imaging.	<b>IIa</b>	<b>B</b>
Aortic diameters may be indexed to the body surface area, especially for the outliers in body size.	<b>IIb</b>	<b>B</b>

## (Thoracic) endovascular aortic repair ((T)evar)

Recommendations	Class	Level
It is recommended to decide the indication for TEVAR or EVAR on an individual basis, according to anatomy, pathology, comorbidity and anticipated durability, of any repair, using a multidisciplinary approach.	<b>I</b>	<b>C</b>
A sufficient proximal and distal landing zone of at least 2 cm is recommended for the safe deployment and durable fixation of TEVAR.	<b>I</b>	<b>C</b>
In case of aortic aneurysm, it is recommended to select a stent-graft with a diameter exceeding the diameter of the landing zones by at least 10-15% of the reference aorta.	<b>I</b>	<b>C</b>
During stent-graft placement, invasive blood pressure monitoring and control, either pharmacologically or by rapid pacing, is recommended.	<b>I</b>	<b>C</b>
Preventive cerebrospinal fluid (CSF) drainage should be considered in high-risk patients.	<b>IIa</b>	<b>C</b>

# classification of Endoleaks

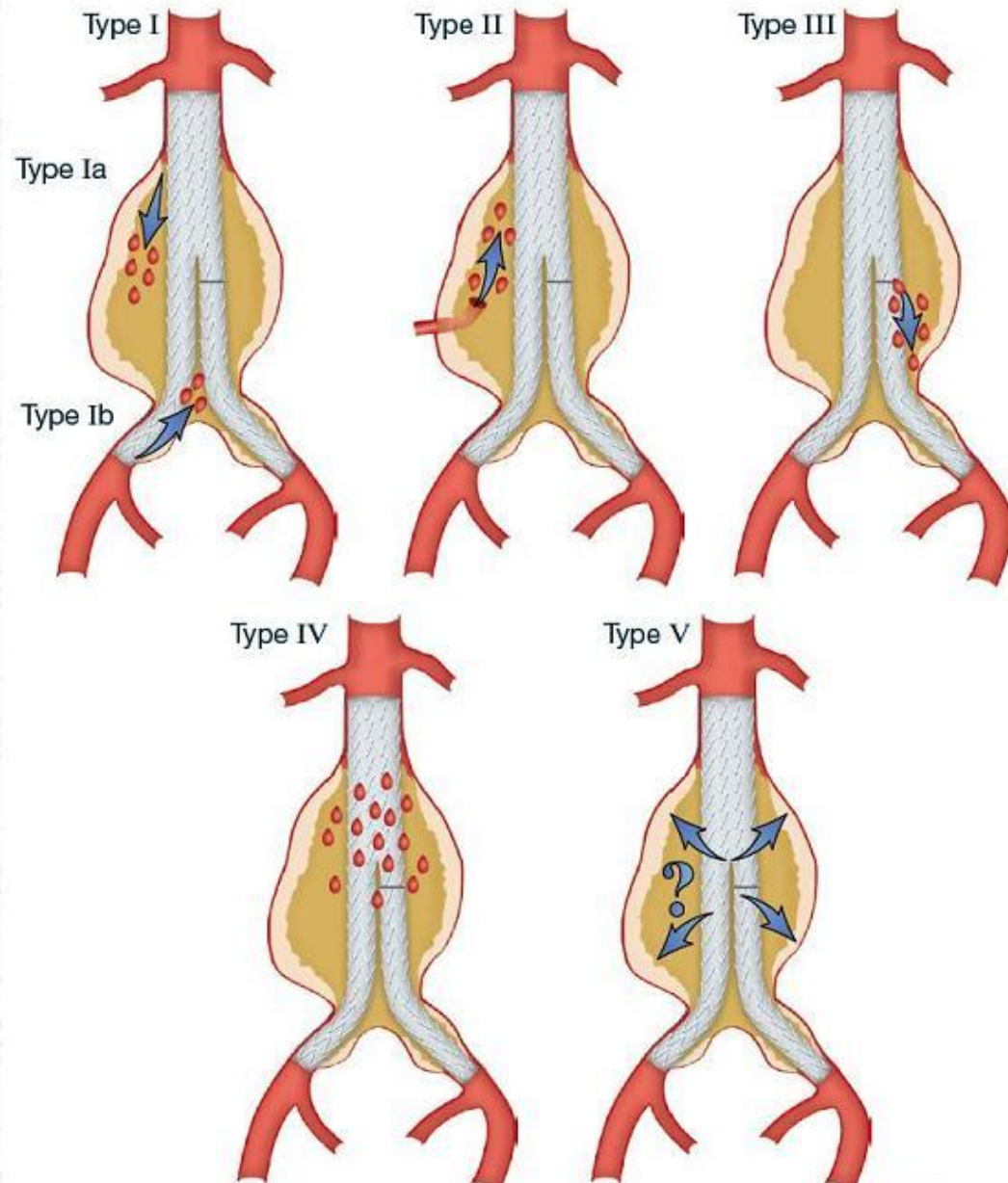
**Type I:** Leak at graft attachment site above, below, or between graft components (Ia: proximal attachment site; Ib: distal attachment site).

**Type II:** Aneurysm sac filling retrogradely via single (IIa) or multiple branch vessels (IIb).

**Type III:** Leak through mechanical defect in graft, mechanical failure of the stent-graft by junctional separation of the modular components (IIIa), or fractures or holes in the endograft (IIIb).

**Type IV:** Leak through graft fabric as a result of graft porosity.

**Type V:** Continued expansion of aneurysm sac without demonstrable leak on imaging (endotension, controversial).



# Surgical techniques in aortic disease

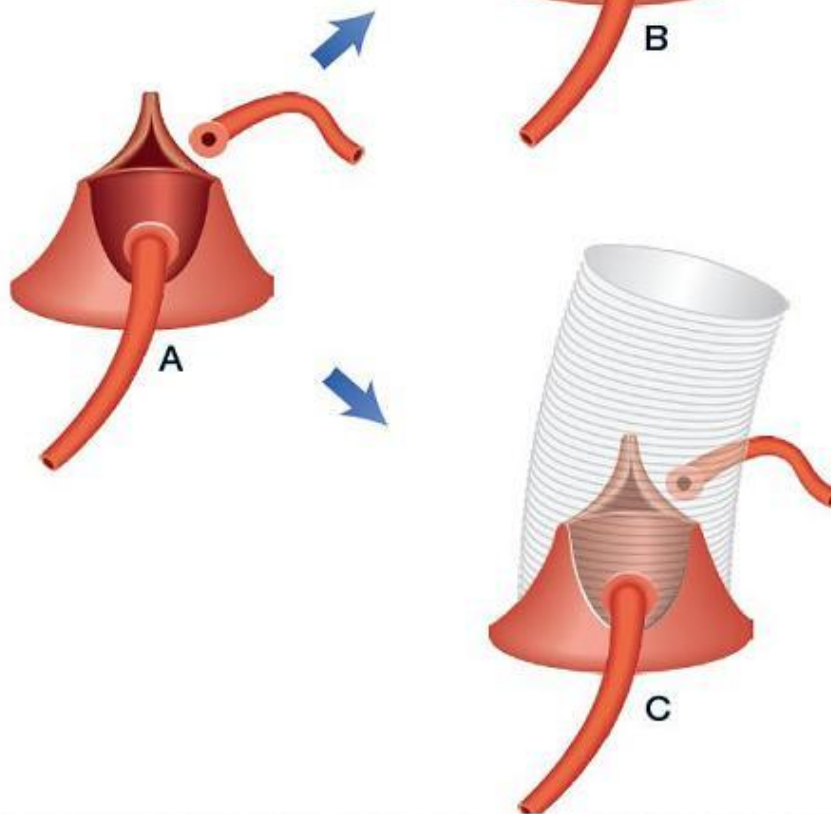
Recommendations	Class	Level
Cerebrospinal fluid drainage is recommended in surgery of the thoracoabdominal aorta to reduce the risk of paraplegia.	<b>I</b>	<b>B</b>
Aortic valve repair using the reimplantation technique or remodelling with aortic annuloplasty is recommended in young patients with aortic root dilatation and tricuspid aortic valves.	<b>I</b>	<b>C</b>
For repair of acute type-A aortic dissection (AD), an open distal anastomotic technique avoiding aortic clamping (hemiarch/complete arch) is recommended.	<b>I</b>	<b>C</b>
In patients with connective tissue disorders requiring aortic surgery, the replacement of aortic sinuses is indicated.	<b>I</b>	<b>C</b>
Selective antegrade cerebral perfusion should be considered in aortic arch surgery to reduce the risk of stroke.	<b>IIa</b>	<b>B</b>
The axillary artery should be considered as first choice for cannulation for surgery of the aortic arch and in aortic dissection.	<b>IIa</b>	<b>C</b>
Left heart bypass should be considered during repair of the descending aorta or the thoracoabdominal aorta to ensure distal organ perfusion.	<b>IIa</b>	<b>C</b>

# Aortic root surgery in aortic dissection

(A) Concept of valve-sparing aortic root repair, excision of diseased aorta, and isolation of coronary ostia.

(B) Reimplantation technique supporting the aortic annulus by the Dacron prosthesis – David

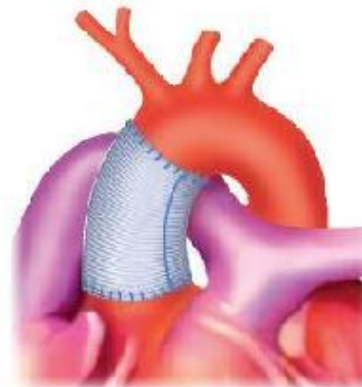
(C) Remodelling technique without annular support – Yacoub



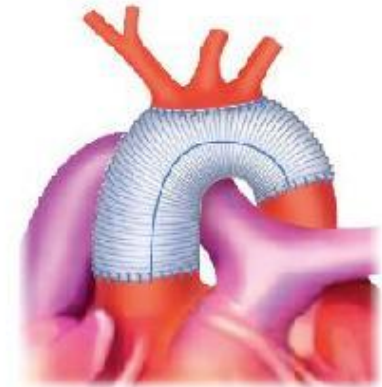


# Ascending aortic & aortic arch surgery

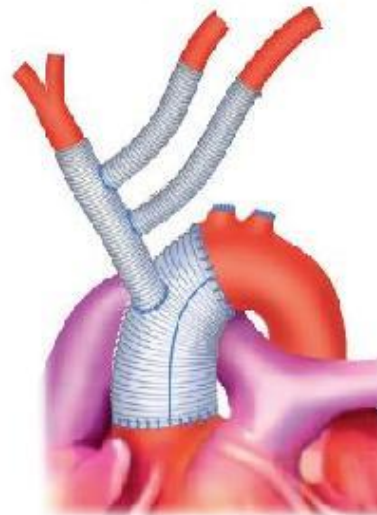
- (A) Ascending aortic replacement from sinotubular junction to cranial ascending aorta.
- (B) Hemi-arch replacement encompassing the concavity of the aortic arch.
- (C) Total arch replacement using a trifurcated technique for the supra-aortic vessels.
- (D) Frozen elephant trunk technique including total arch replacement using the island technique.



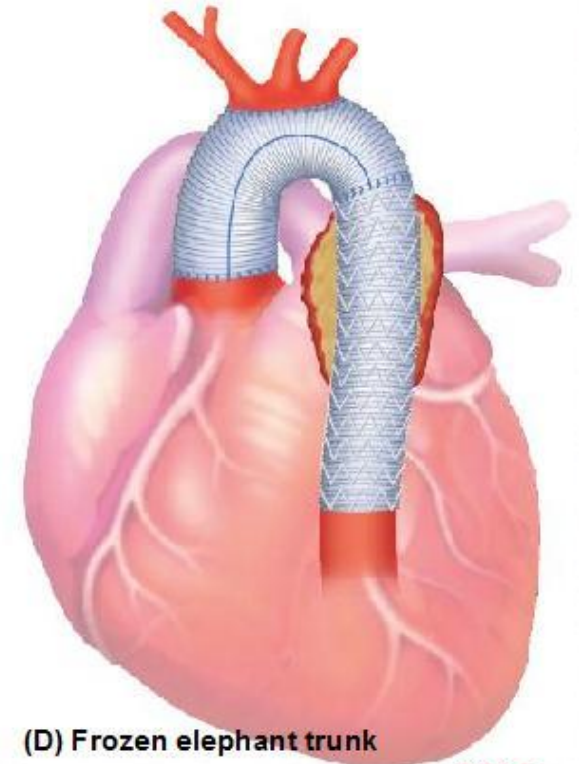
A - Supracommissural ascending aortic replacement



C - Total arch replacement

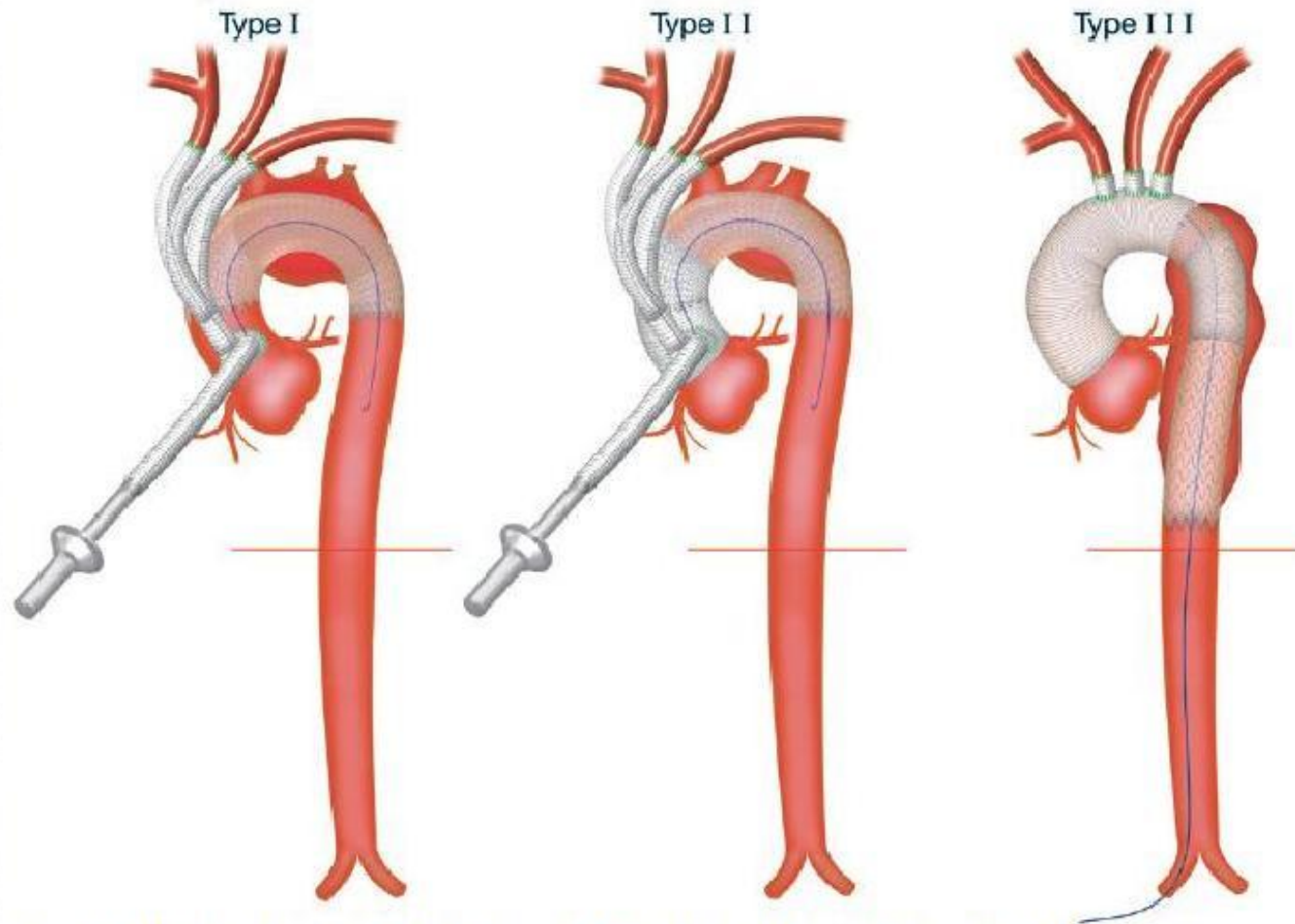


B - Hemiarch replacement with rebranching of supra-aortic vessels (trifurcated graft)



(D) Frozen elephant trunk

# Different methods for aortic arch debranching

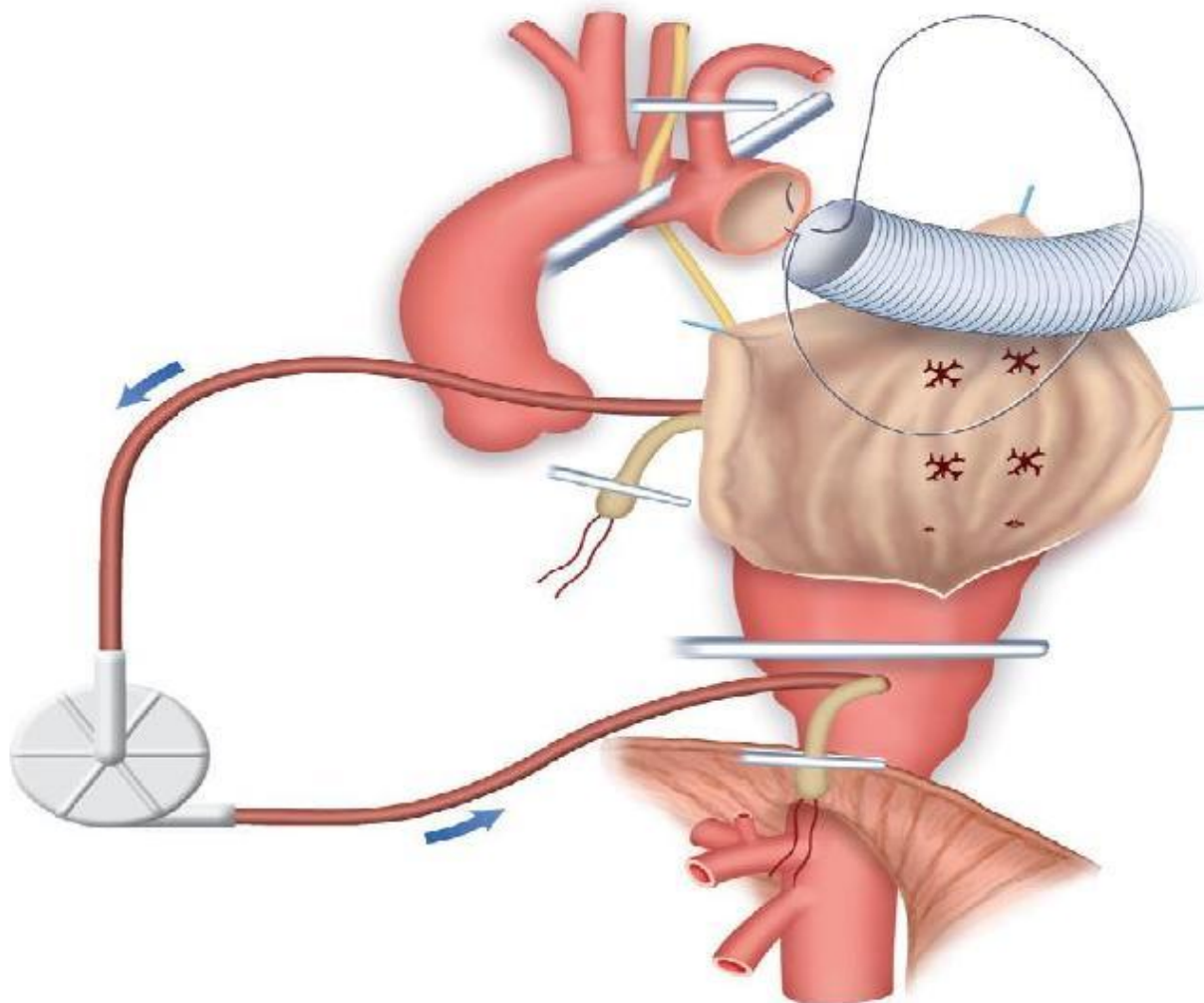


**Type I:** Total aortic arch debranching and TEVAR for off-pump total arch repair (use of beating heart cardiopulmonary bypass optional).

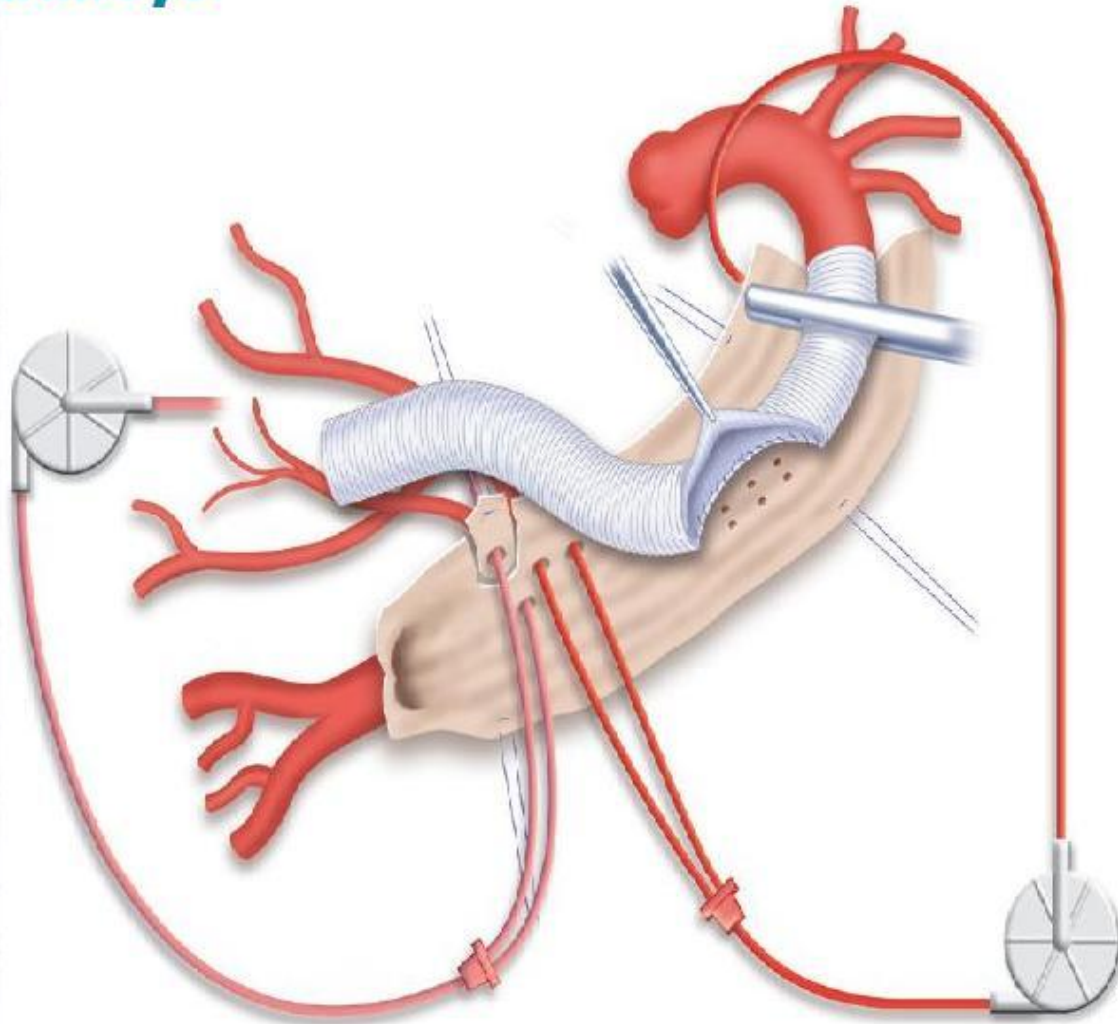
**Type II:** Total aortic arch debranching and TEVAR in combination with ascending aortic replacement in patients with proximal disease extension for total thoracic aortic repair

**Type III:** Total aortic arch replacement with conventional elephant trunk technique and distal extension by TEVAR in patients with distal disease extension, for total thoracic aortic repair.

# Illustration of left heart bypass for thoracic & thoraco-abdominal aortic replacement, inflow via left-sided pulmonary veins, & arterial return via any downstream segment

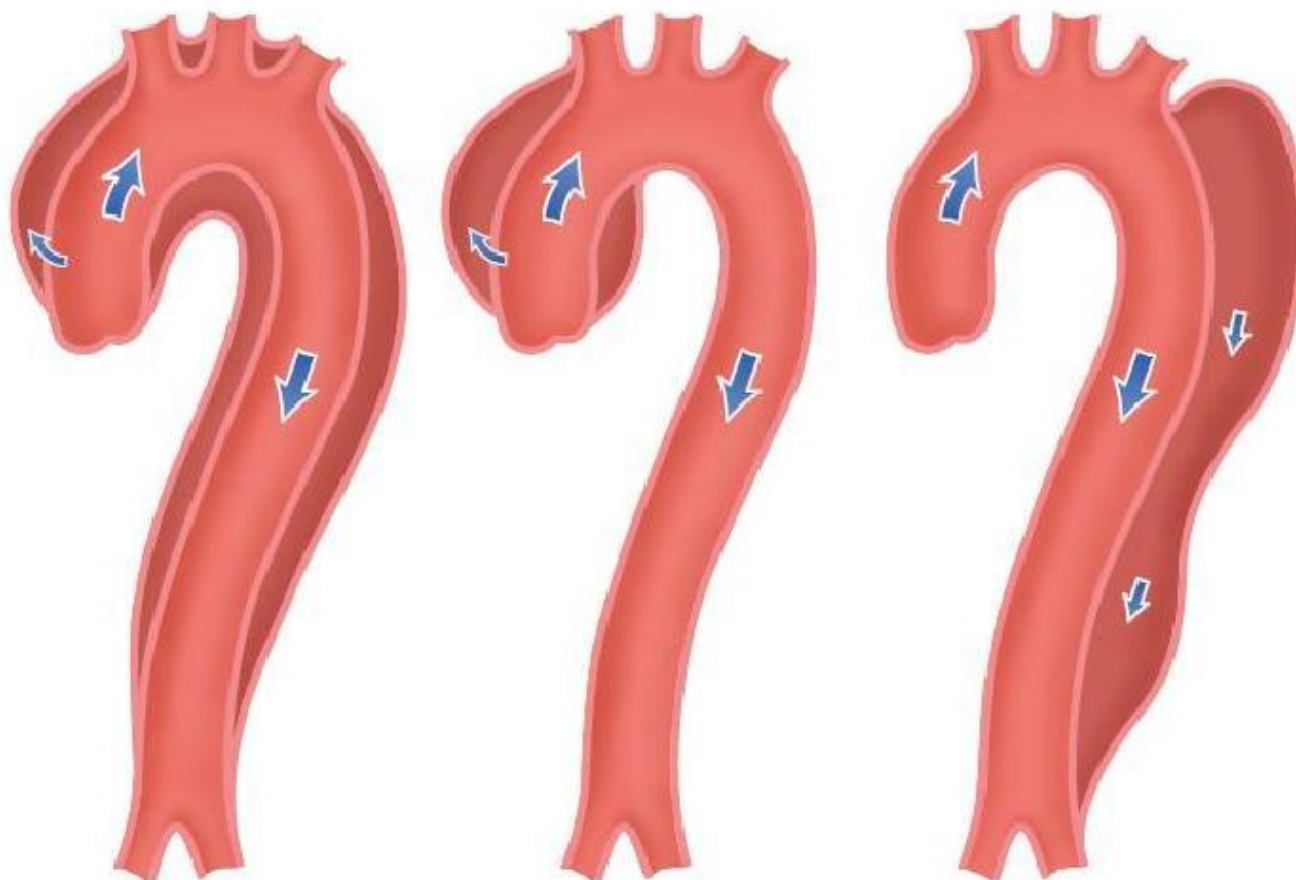


# Illustration of left heart bypass for thoracic & thoraco-abdominal aortic replacement showing selective visceral blood perfusion as well as selective bilateral cold saline perfusion of kidneys



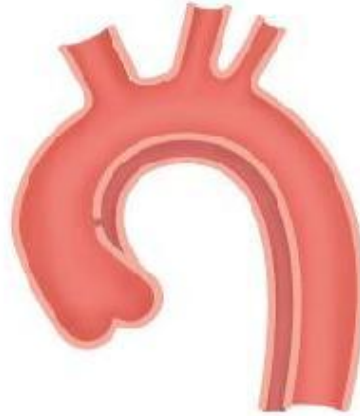
# Classification of aortic dissection localization

De Bakey	Type I	Type II	Type III
Stanford	Type A	Type A	Type B

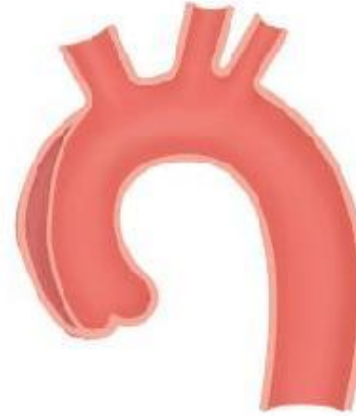


# Classification of acute aortic syndromes (AAS)

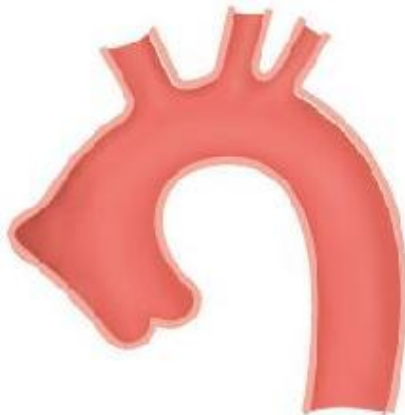
Class 1



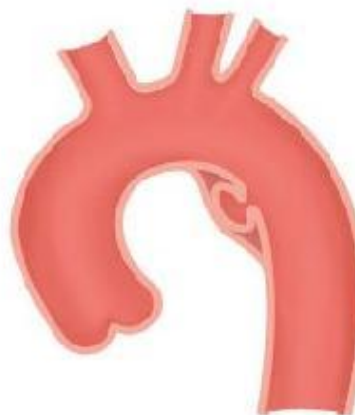
Class 2



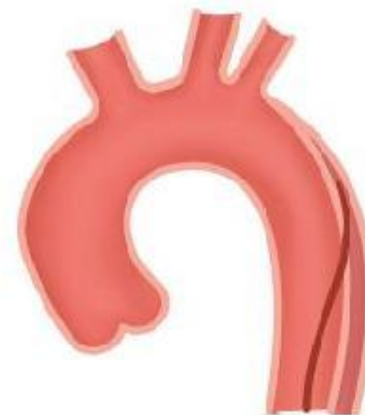
Class 3



Class 4



Class 5



**Class 1:** Classic AD with true and FL with or without communication between the two lumina.

**Class 2:** Intramural haematoma.

**Class 3:** Subtle or discrete aortic dissection with bulging of the aortic wall.

**Class 4:** Penetrating aortic ulcer following plaque rupture.

**Class 5:** Iatrogenic or traumatic AD, illustrated by a catheter-induced separation of the intima.

## Main clinical presentations and complications of patients with acute aortic dissection

	Type A	Type B
Chest pain	80%	70%
Back pain	40%	70%
Abrupt onset of pain	85%	85%
Migrating pain	<15%	20%
Aortic regurgitation	40-75%	N/A
Cardiac tamponade	<20%	N/A
Myocardial ischaemia or infarction	10-15%	10%
Heart failure	<10%	<5%
Pleural effusion	15%	20%
Syncope	15%	<5%
Major neurological deficit (coma/stroke)	<10%	<5%
Spinal cord injury	<1%	N/R
Mesenteric ischaemia	<5%	N/R
Acute renal failure	<20%	10%
Lower limb ischaemia	<10%	10%

# Laboratory tests required for patients with acute aortic dissection

Laboratory tests	To detect signs of:
Red blood cell count	Blood loss, bleeding, anaemia
White blood cell count	Infection, inflammation (SIRS)
C-reactive protein	Inflammatory response
ProCalcitonin	Differential diagnosis between SIRS and sepsis
Creatine kinase	Reperfusion injury, rhabdomyolysis
Troponin I or T	Myocardial ischaemia, myocardial infarction
D-dimer	Aortic dissection, pulmonary embolism, thrombosis
Creatinine	Renal failure (existing or developing)
Aspartate transaminase / alanine aminotransferase	Liver ischaemia, liver disease
Lactase	Bowel ischaemia, metabolic disorder
Glucose	Diabetes mellitus
Blood gases	Metabolic disorder, oxygenation



# Details required from imaging in acute aortic dissection *(Cont.)*

## **Intramural haematoma**

Localization and extent of aortic wall thickening

Co-existence of atheromatous disease (calcium shift)

Presence of small intimal tears

## **Penetrating aortic ulcer**

Localization of the lesion (length and depth)

Co-existence of intramural haematoma

Involvement of the peri-aortic tissue and bleeding

Thickness of the residual wall

## **In all cases**

Co-existence of other aortic lesions: aneurysms, plaques, signs of inflammatory disease, etc.

# Diagnostic value of various imaging modalities for acute aortic syndromes

Lesion	TTE	TOE	CT	MRI
Ascending aortic dissection	++	+++	+++	+++
Aortic arch dissection	+	+	+++	+++
Descending aortic dissection	+	+++	+++	+++
Size	++	+++	+++	+++
Mural thrombus	+	+++	+++	+++
Intramural haematoma	+	+++	++	+++
Penetrating aortic ulcer	++	++	+++	+++
Involvement of aortic branches	+	(+)	+++	+++

+++ = excellent; ++ = moderate; + = poor; (+) = poor and inconstant.

# Diagnostic work-up of acute aortic syndrome (AAS)

Recommendations	Class	Level
<b>History and clinical assessment</b>		
In all patients with suspected AAS, pretest probability assessment is recommended, according to the patient's condition, symptoms, and clinical features.	<b>I</b>	<b>B</b>
<b>Laboratory testing</b>		
In case of suspicion of AAS, the interpretation of biomarkers should always be considered along with the pretest clinical probability.	<b>IIa</b>	<b>C</b>
In case of low clinical probability of AAS, the determination of D-dimers levels should be considered to rule-out the diagnosis when negative.	<b>IIa</b>	<b>B</b>
In case of intermediate clinical probability of AAS with a positive (point-of-care) D-dimer test, further imaging tests should be considered.	<b>IIa</b>	<b>B</b>
In patients with high probability (risk score 2 or 3) of aortic dissection, testing of D-dimers is not recommended.	<b>III</b>	<b>C</b>

# Diagnostic work-up of acute aortic syndrome (AAS) (cont.)

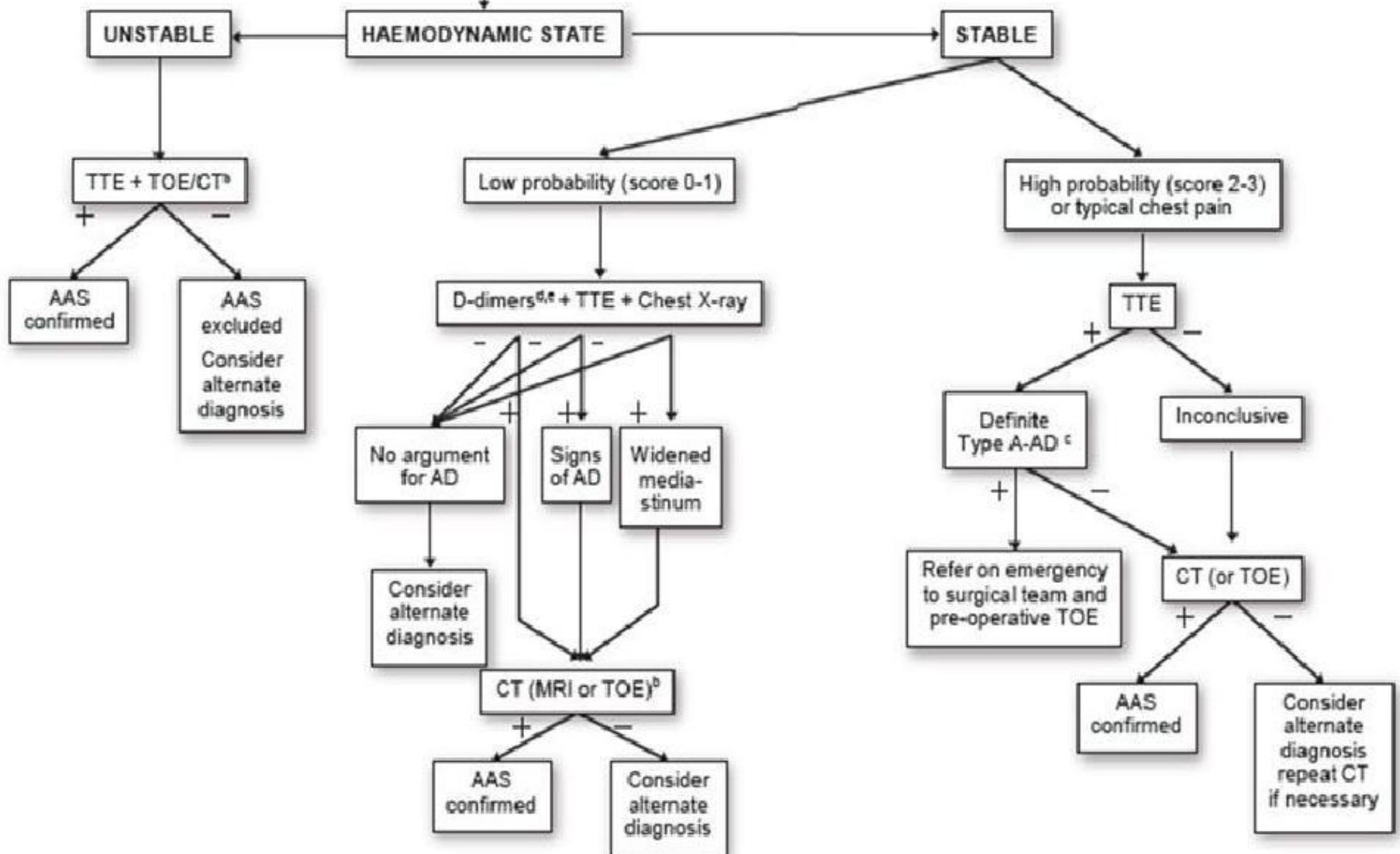
Recommendations	Class	Level
<b>Imaging</b>		
TTE is recommended as an initial imaging investigation.	<b>I</b>	<b>C</b>
In unstable patients with a suspicion of AAS, the following imaging modalities are recommended according to local availability and expertise:		
• TOE	<b>I</b>	<b>C</b>
• CT	<b>I</b>	<b>C</b>
In stable patients with a suspicion of AAS, the following imaging modalities are recommended (or should be considered) according to local availability and expertise:		
• CT	<b>I</b>	<b>C</b>
• MRI	<b>I</b>	<b>C</b>
• TOE	<b>IIa</b>	<b>C</b>
In case of initially negative imaging with persistence of suspicion of AAS, repetitive imaging (CT or MRI) is recommended.	<b>I</b>	<b>C</b>
Chest X-ray may be considered in case of low clinical probability of AAS.	<b>IIb</b>	<b>C</b>
In case of uncomplicated type-B aortic dissection treated medically, repeated imaging (CT or MRI) during the first days is recommended.	<b>I</b>	<b>C</b>

# Clinical data useful to assess the *a priori* probability of acute aortic syndromes

High-risk conditions	High-risk pain features	High-risk examination features
<ul style="list-style-type: none"><li>• Marfan syndrome (or other connective tissue diseases)</li><li>• Family history of aortic disease</li><li>• Known aortic valve disease</li><li>• Known thoracic aortic aneurysm</li><li>• Previous aortic manipulation (including cardiac surgery)</li></ul>	<ul style="list-style-type: none"><li>• Chest, back, or abdominal pain described as any of the following:<ul style="list-style-type: none"><li>– abrupt onset</li><li>– severe intensity</li><li>– ripping or tearing</li></ul></li></ul>	<ul style="list-style-type: none"><li>• Evidence of perfusion deficit:<ul style="list-style-type: none"><li>– pulse deficit</li><li>– systolic blood pressure difference</li><li>– focal neurological deficit (in conjunction with pain)</li></ul></li><li>• Aortic diastolic murmur (new and with pain)</li><li>• Hypotension or shock</li></ul>

# ACUTE CHEST PAIN

Medical history + clinical examination + ECG → STEMI<sup>a</sup>: see ESC guidelines



# Treatment of aortic dissection

Recommendations	Class	Level
In all patients with AD, medical therapy including pain relief and blood pressure control is recommended.	<b>I</b>	<b>C</b>
In patients with type-A AD, urgent surgery is recommended.	<b>I</b>	<b>B</b>
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.	<b>IIa</b>	<b>B</b>
In uncomplicated type-B AD, medical therapy should always be recommended.	<b>I</b>	<b>C</b>
In uncomplicated type-B AD, TEVAR should be considered.	<b>IIa</b>	<b>B</b>
In complicated type-B AD, TEVAR is recommended.	<b>I</b>	<b>C</b>
In complicated type-B AD, surgery may be considered.	<b>IIb</b>	<b>C</b>

# Predictor of intramural haematoma complications

Persistent and recurrent pain despite aggressive medical treatment

Difficult blood-pressure control

Ascending aortic involvement

Maximum aortic diameter  $\geq 50$  mm

Progressive maximum aortic wall thickness ( $> 11$  mm)

Enlarging aortic diameter

Recurrent pleural effusion

Penetrating ulcer or ulcer-like projection secondary to localized dissections in the involved segment

Detection of organ ischaemia (brain, myocardium, bowels, kidneys, etc.)



# Diagnostic work-up of intramural haematoma (IMH)

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

# Management of penetrating aortic ulcer (PAU)

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

## (Contained) rupture in the thoracic aortic aneurysm

Recommendations	Class	Level
In patients with suspected rupture of the TAA, urgent CT angiography for diagnosis confirmation is recommended.	I	C
In patients with acute contained rupture of TAA, urgent repair is recommended.	I	C
If the anatomy is favourable and the expertise available, endovascular repair (TEVAR) should be preferred over open surgery.	I	C

# Traumatic aortic injury

Recommendations	Class	Level
In case of suspicion of TAI, CT is recommended.	<b>I</b>	<b>C</b>
If CT is not available, TOE should be considered.	<b>IIa</b>	<b>C</b>
In case of TAI with suitable anatomy requiring intervention, TEVAR should be preferred to surgery.	<b>IIa</b>	<b>C</b>

## Work-up for Patients with aortic aneurysm

Recommendations	Class	Level
When an aortic aneurysm is identified at any location, assessment of the entire aorta and aortic valve is recommended at baseline and during follow-up.	<b>I</b>	<b>C</b>
In case of aneurysm of the abdominal aorta, duplex ultrasound for screening of peripheral artery disease and peripheral aneurysms should be considered.	<b>IIa</b>	<b>C</b>
Patients with aortic aneurysm are at increased risk of cardiovascular disease: general principles of cardiovascular prevention should be considered.	<b>IIa</b>	<b>C</b>

# Interventions of thoracic aortic aneurysm (TAA)

Recommendations	Class	Level
<b>Interventions on ascending aorta</b>		
Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter $\geq 50$ mm for patients with Marfan syndrome.	<b>I</b>	<b>C</b>
Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameter: $\geq 45$ mm for patients with Marfan syndrome with risk factors. $\geq 50$ mm for patients with bicuspid valve with risk factors. $\geq 55$ mm for other patients with no elastopathy.	<b>IIa</b>	<b>C</b>
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.	<b>IIb</b>	<b>C</b>
<b>Interventions on aortic arch aneurysms</b>		
Surgery should be considered in patients who have isolated aortic arch aneurysm with maximal diameter $\geq 55$ mm.	<b>IIa</b>	<b>C</b>
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.	<b>IIb</b>	<b>C</b>

# Diagnostic work-up of thoracic aortic aneurysm (TAA)

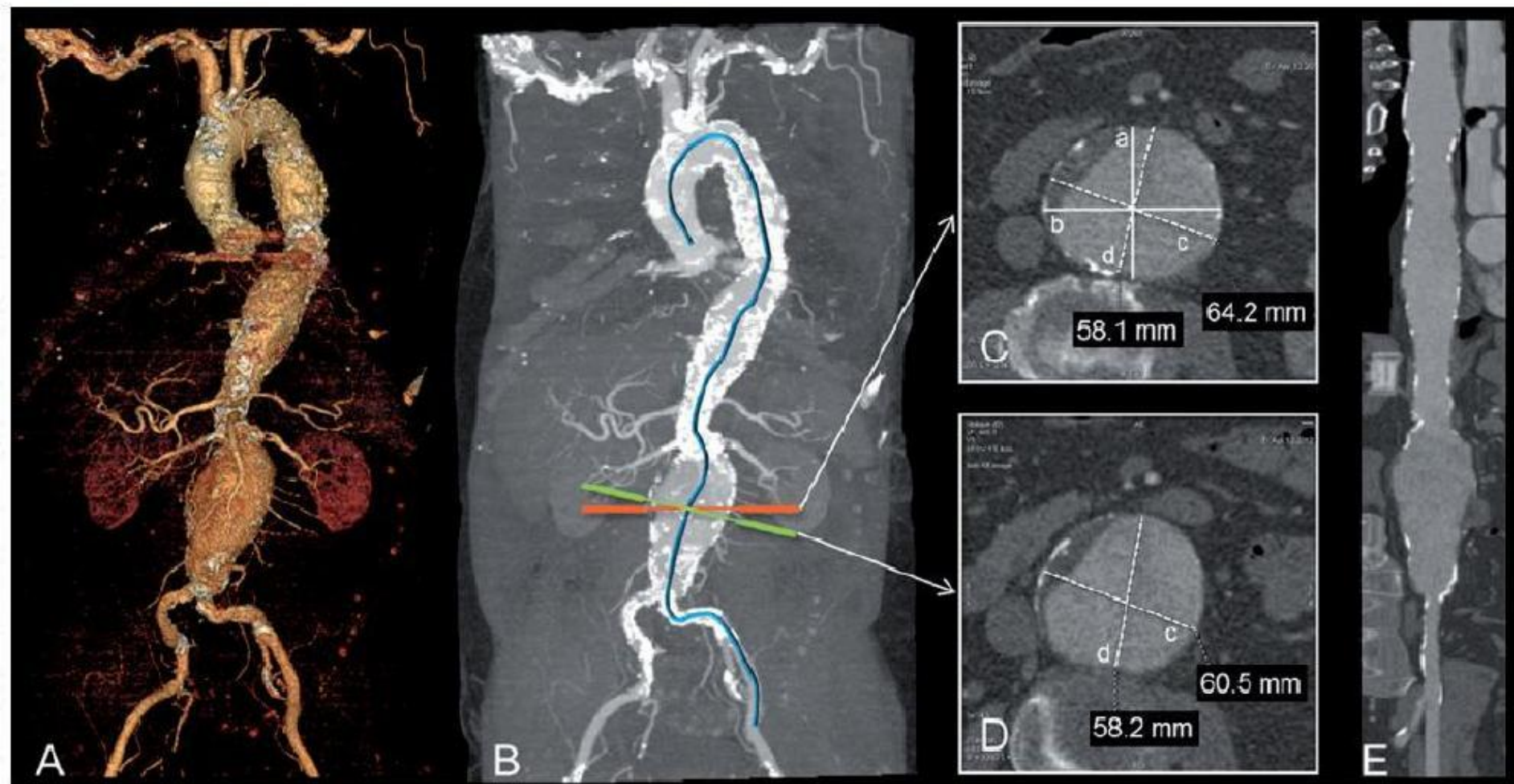
Recommendations	Class	Level
<b>Interventions on descending aortic aneurysms</b>		
TEVAR should be considered rather than surgery when anatomy is suitable.	<b>IIa</b>	<b>C</b>
TEVAR should be considered in patients who have descending aortic aneurysm with maximal diameter $\geq 55$ mm.	<b>IIa</b>	<b>C</b>
When TEVAR is not technically possible, surgery should be considered in patients who have descending aortic aneurysm with maximal diameter $\geq 60$ mm.	<b>IIa</b>	<b>C</b>
When intervention is indicated, in case of Marfan syndrome or other elastopathies, surgery should be indicated rather than TEVAR.	<b>IIa</b>	<b>C</b>

# Abdominal aortic aneurysm (AAA) screening

Recommendations	Class	Level
Population screening for AAA with ultrasound:		
Is recommended in all men >65 years of age.	<b>I</b>	<b>A</b>
May be considered in women >65 years of age with history of tobacco smoking.	<b>IIb</b>	<b>C</b>
Is not recommended in female non-smokers without familial hystory.	<b>III</b>	<b>C</b>
Targeted screening for AAA with ultrasound should be considered in first-degree siblings of a patient with AAA.	<b>IIa</b>	<b>B</b>
Opportunistic screening for AAA during TTE:		
Should be considered in all men >65 years of age.	<b>IIa</b>	<b>B</b>
May be considered in women >65 years with a history of current/past smoking.	<b>IIb</b>	<b>C</b>



# CT evaluation of aortic aneurysm



Thoracic and abdominal aorta in a three-dimensional reconstruction (left lateral image), parasagittal multiplanar reconstruction (MPR) along the centreline (left middle part), straightened-MPR along the centreline with given landmarks (A-I) (right side), orthogonal to the centreline orientated cross-sections at the landmarks (A-J). Landmarks A-J should be used to report aortic diameters: (A) sinuses of Valsalva; (B) sinotubular junction; (C) mid ascending aorta (as indicated); (D) proximal aortic arch (aorta at the origin of the brachiocephalic trunk); (E) mid aortic arch (between left common carotid and subclavian arteries); (F) proximal descending thoracic aorta (approximately 2 cm distal to left subclavian artery); (G) mid descending aorta (level of the pulmonary arteries as easily identifiable landmarks, as indicated); (H) at diaphragm; (I) at the celiac axis origin; (J) right before aortic bifurcation. (Provided by F Nensa, Institute of Diagnostic and Interventional Radiology, Essen.)

## Management of asymptomatic patients with enlarged aorta or abdominal aortic aneurysm (AAA)

Recommendations	Class	Level
In patients with abdominal aortic diameter of 25–29 mm, new ultrasound imaging should be considered 4 years later.	<b>IIa</b>	<b>B</b>
Surveillance is indicated and safe in patients with AAA with a maximum diameter of <55 mm and slow (<10 mm/year) growth.	<b>I</b>	<b>A</b>
In patients with small (30–55 mm) AAAs, the following time interval for imaging should be considered: Every 3 years for AAA of 30–39 mm diameter. Every 2 years for AAA of 40–44 mm diameter. Every year for AAA >45 mm diameter.	<b>IIa</b>	<b>B</b>

## Management of asymptomatic patients with enlarged aorta or abdominal aortic aneurysm (AAA) (cont.)

Recommendations	Class	Level
Smoking cessation is recommended to slow the AAA growth.	<b>I</b>	<b>B</b>
To reduce aortic complications in patients with small AAAs, the use of statins and ACE-inhibitors may be considered.	<b>IIb</b>	<b>B</b>
AAA repair is indicated if: AAA diameter exceeds 55 mm. Aneurysm growth exceeds 10 mm/year.	<b>I</b>	<b>B</b>
If a large aneurysm is anatomically suitable for EVAR, either open or endovascular aortic repair is recommended in patients with acceptable surgical risk.	<b>I</b>	<b>A</b>
If a large aneurysm is anatomically unsuitable for EVAR, open aortic repair is recommended.	<b>I</b>	<b>C</b>
In patients with asymptomatic AAA who are unfit for open repair, EVAR along with best medical treatment may be considered.	<b>IIb</b>	<b>B</b>

## Management of patients with symptomatic abdominal aortic aneurysm (AAA)

Recommendations	Class	Level
In patients with suspected rupture of AAA, immediate abdominal ultrasound or CT is recommended.	<b>I</b>	<b>C</b>
In case of ruptured AAA, emergency repair is indicated	<b>I</b>	<b>C</b>
In cas of symptomatic but non-ruptured AAA, urgent repair is indicated.	<b>I</b>	<b>C</b>
In case of symptomatic AAA anatomically suitable for EVAR, either open or endovascular aortic repair is recommended.	<b>I</b>	<b>A</b>

# Genetic testing in aortic diseases

Recommendations	Class	Level
It is recommended to investigate first-degree relatives (siblings and parents) of a subject with TAAD to identify a familial form in which relatives all have a 50% chance of carrying the family mutation/disease.	<b>I</b>	<b>C</b>
Once a familial form of TAAD is highly suspected, it is recommended to refer the patient to a geneticist for family investigation and molecular testing.	<b>I</b>	<b>C</b>
Variability of age of onset warrants screening every 5 years of 'healthy' at-risk relatives until diagnosis (clinical or molecular) is established or ruled out.	<b>I</b>	<b>C</b>
In familial non-syndromic TAAD, screening for aneurysm should be considered not only in the thoracic aorta but also throughout the arterial tree (including cerebral arteries).	<b>IIa</b>	<b>C</b>

# Management of aortic root dilatation in patients with bicuspid aortic valve (BAV)

Recommendations	Class	Level
Patients with known BAV should undergo an initial TTE to assess the diameters of the aortic root and ascending aorta.	<b>I</b>	<b>C</b>
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.	<b>I</b>	<b>C</b>
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.	<b>I</b>	<b>C</b>
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.	<b>I</b>	<b>C</b>
In the case of aortic diameter >50 mm or an increase >3 mm/year measured by echocardiography, measurement confirmation using another imaging modality (CT or MRI) is indicated.	<b>I</b>	<b>C</b>

# Management of aortic root dilatation in patients with bicuspid aortic valve (BAV)

Recommendations	Class	Level
In case of BAV, surgery of the ascending aorta:		
is indicated in case of aortic root or ascending aortic diameter >55 mm.	<b>I</b>	<b>C</b>
is indicated in case of aortic root or ascending aortic diameter >50 mm in the presence of other risk factors.	<b>I</b>	<b>C</b>
is indicated in case of aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled.	<b>I</b>	<b>C</b>
$\beta$ -blockers may be considered in patients with BAV and dilated aortic root >40 mm.	<b>IIb</b>	<b>C</b>
Because of familial occurrence, screening of first-degree relatives should be considered.	<b>IIa</b>	<b>C</b>
In patients with any elastopathy or BAV with dilated aortic root (>40 mm), isometric exercise with a high static load (e.g. weightlifting) is not indicated and should be discouraged.	<b>III</b>	<b>C</b>

## Interventions in coarctation of the aorta

Recommendations	Class	Level
In all patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), abnormal blood pressure response during exercise, or significant left ventricular hypertrophy, an intervention is indicated.	<b>I</b>	<b>C</b>
Independent of the pressure gradient, hypertensive patients with >50% aortic narrowing relative to the aortic diameter at the diaphragm level (on MRI, CT, or invasive angiography) should be considered for intervention.	<b>IIa</b>	<b>C</b>
Independent of the pressure gradient and presence of hypertension, patients, with >50% aortic narrowing relative to the aortic diameter at the diaphragm level (on MRI, CT, or invasive angiography) may be considered for intervention.	<b>IIb</b>	<b>C</b>



# Semi-quantitative grading of severity of aortic atherosclerosis

Grade	
Grade I	Normal aorta
Grade II	Increased intimal thickening without luminal irregularities
Grade III	Single or multiple protruding atheromas
Grade IV	Atheroma with mobile or ulcerated (complicated) structure

# Management of patients with aortic plaque

Recommendations	Class	Level
In the presence of aortic atherosclerosis, general preventive measures to control risk factors are indicated.	<b>I</b>	<b>C</b>
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.	<b>IIa</b>	<b>C</b>
Prophylactic surgery to remove high-risk aortic plaque is not recommended.	<b>III</b>	<b>C</b>

# Inflammatory diseases associated with aortitis

Disease	Diagnostic criteria	Definitive diagnosis
Giant cell arteritis	<ul style="list-style-type: none"> <li>• Age at onset &gt;50 years</li> <li>• Recent-onset localized headache</li> <li>• Temporal artery tenderness or pulse attenuation</li> <li>• Elevated erythrocyte sedimentation rate &gt;50 mm/h</li> <li>• Artery biopsy showing necrotizing vasculitis</li> </ul>	Three or more criteria are present (sensitivity >90%; specificity >90%)
Takayasu arteritis	<ul style="list-style-type: none"> <li>• Age at onset &lt;40 years</li> <li>• Intermittent claudication</li> <li>• Diminished brachial artery pulse</li> <li>• Subclavian artery or carotid bruit</li> <li>• Systolic blood pressure variation of &gt;10 mmHg between arms</li> <li>• Aortographic evidence of aorta or aortic branch stenosis</li> </ul>	Three or more criteria are present (sensitivity 90.5%; specificity 97.8%)
Behçet disease	<ul style="list-style-type: none"> <li>• Oral ulceration</li> <li>• Recurrent genital ulceration</li> <li>• Uveitis or retinal vasculitis</li> <li>• Skin lesion, erythema nodosum, pseudofolliculitis or pathergy</li> </ul>	Oral ulceration plus two of the other three criteria
Ankylosing spondylitis	<ul style="list-style-type: none"> <li>• Onset of pain at age &lt;40 years</li> <li>• Back pain for &gt;3 months</li> <li>• Morning stiffness</li> <li>• Subtle symptom onset</li> <li>• Improvement with exercise</li> </ul>	Four of the diagnostic criteria are present

# Follow-up and management of chronic aortic diseases

Recommendations	Class	Level
<b>Chronic aortic dissection</b>		
Contrast CT or MRI is recommended to confirm the diagnosis of chronic aortic dissection.	<b>I</b>	<b>C</b>
Initial close imaging surveillance of patients with chronic aortic dissection is indicated to detect signs of complications as soon as possible.	<b>I</b>	<b>C</b>
In asymptomatic patients with chronic dissection of the ascending aorta, elective surgery should be considered.	<b>IIa</b>	<b>C</b>
In patients with chronic aortic dissection, tight blood pressure control <130/80 is indicated.	<b>I</b>	<b>C</b>
Surgery repair or TEVAR is recommended for complicated type-B aortic dissection (aortic diameter >60 mm, >10 mm/year growth, malperfusion, recurrent pain).	<b>I</b>	<b>C</b>

# Follow-up management of chronic aortic diseases (cont.)

Recommendations	Class	Level
<b>Follow-up after endovascular treatment for aortic diseases</b>		
After TEVAR or EVAR, surveillance is recommended after 1 month, 6 months, 12 months, and then yearly. Shorter intervals can be proposed in the event of abnormal findings requiring closer surveillance.	<b>I</b>	<b>C</b>
CT is recommended as the first-choice imaging technique for follow-up after TEVAR or EVAR.	<b>I</b>	<b>C</b>
If neither endoleak nor AAA sac enlargement is documented during first year after EVAR, colour DUS, with or without contrast agents, should be considered for annual postoperative surveillance, with non-contrast CT imaging every 5 years.	<b>IIa</b>	<b>C</b>
For patients with TAA <45 mm, annual imaging is recommended; while in patients with TAA ≥45 mm and <55 mm, imaging every 6 months is recommended, unless the stability of the lesions is evidenced after serial imaging.	<b>I</b>	<b>C</b>
For follow-up after (T)EVAR in young patients, MRI should be preferred to CT for magnetic resonance-compatible stent-grafts, to reduce radiation.	<b>IIa</b>	<b>C</b>
Long-term surveillance of open abdominal aortic repair may be considered at loose (5-year) intervals using colour DUS or CT imaging.	<b>IIb</b>	<b>C</b>



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