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AORTIC DISEASES

ESC GUIDELINES ON THE DIAGNOSIS AND TREATMENT OF AORTIC DISEASES

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ESC ESSENTIAL MESSAGES

2014 ESC GUIDELINES ON THE DIAGNOSIS AND TREATMENT OF AORTIC DISEASES*

The Task Force on diagnosis and treatment of aortic diseases of the European Society of Cardiology (ESC)

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Table of contents

European Heart Journal (2014) 35, 2873–2926 - doi: 10.1093/eurheartj/ehu281

Take home messages

1. The holistic view to the aorta as "whole organ"

The guidelines on diagnosis and treatment of aortic diseases highlight the value of a holistic approach, viewing the aorta as the whole organ; indeed, in many cases tandem lesions of the aorta may exist, as illustrated by the increased probability of thoracic aortic aneurysm in the case of abdominal aortic aneurysm, making a distinction between the two regions inadequate. In addition thorako-abdominal aortic diseases are overwriting this separation.

2. Diagnostic Imaging

Whereas a clinical examination and laboratory testing play a minor role in the diagnosis and treatment of aortic diseases, imaging techniques, particularly modern images techniques, play a major role yielding a view of the total aorta, which requires standardized reports and measurements at given landmarks.

Comparison of methods for imaging the aorta

Advantages/disadvantages	TTE	TOE	CT[c]	MRI[c]	AORTOGRAPHY
Ease of use	+++	++	+++	++	+
Diagnostic reliability	+	+++	+++	+++	++
Bedside/interventional use[a]	++	++	–	–	++
Serial examinations	++	+	++(+)[b]	+++	–
Aortic wall visualization[c]	+	+++	+++	+++	–
Cost	–	–	– –	– – –	– – –
Radiation	0	0	– – –	–	– –
Nephrotoxicity	0	0	– – –	– –	– – –

CT = computed tomography; MRI = magnetic resonance imaging; TOE = transoesophageal echocardiography; TTE = transthoracic echocardiography.
+ means a positive aspect and – means a negative point. The number of signs indicates the estimated potential value.
[a]IVUS can be used to guide interventions (see web addenda art www.escardio.org/guidelines).
[b]+++ only for follow-up after aortic stenting (metallic struts), otherwise limit radiation.
[c]PET can be used to visualize suspected aortic inflammatory disease.

In the daily work-up transthoracic echocardiography plays a major role including transoesophageal echocardiography as well as ultrasonography for the abdominal aorta.

It is recommended to measure diameters at anatomical landmarks perpendicular to the longitudinal axis. In case of repetitive imaging of the aorta the imaging should be used, with the lowest iatrogenic risk. In addition, it is recommended to use the same imaging modality with the similar method of measurement. All relevant parameters are recommended to be reported recording to the aortic segmentation. It is recommended to assess renal function, pregnancy and history of allergy to contrast agents in order to select the optimal imaging modality with minimal radiation exposure.
Complete list of normal values for all discussed imaging techniques are found in the Full Text and Web Addenda.

Take home messages

3. The acute aortic syndrome

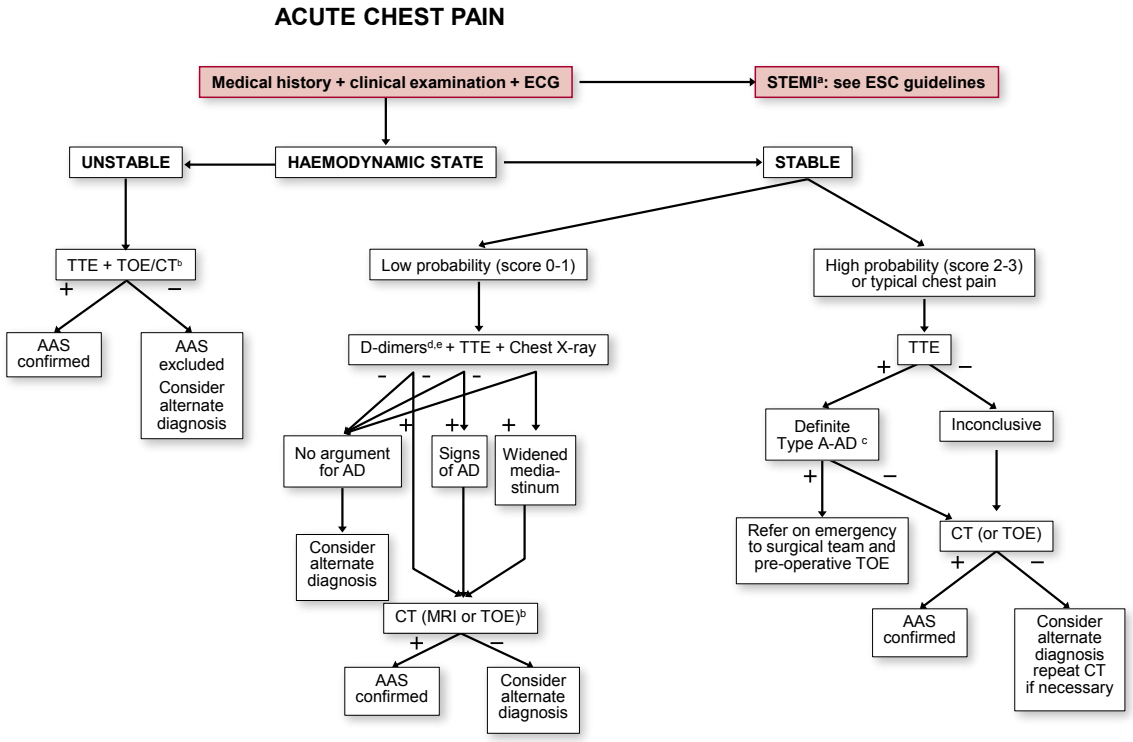
Acute aortic syndromes (AAS) are defined as emergency conditions with similar clinical characteristics involving the aorta: aortic dissection, intramural haematoma, penetrating aortic ulcer, complete rupture of the aorta, traumatic aortic injury, iatrogenic aortic dissection. A flowchart for the emergency room has been developed in order to enhance the standardization of decision making in acute aortic syndromes, because survival is strongly related to time. In the diagnostic work-up clinical data are useful to assess the priori probability of AAS including the high-risk patient conditions, high-risk pain features and signs of high-risk examination features.

Clinical data useful to assess the *a priori* probability of acute aortic syndromes		
High-risk conditions	**High-risk pain features**	**High-risk examination features**
▪ Marfan syndrome (or other connective tissue diseases) ▪ Family history of aortic disease ▪ Known aortic valve disease ▪ Known thoracic aortic aneurysm ▪ Previous aortic manipulation (including cardiac surgery)	▪ Chest, back, or abdominal pain described as any of the following: - abrupt onset - severe intensity - ripping or tearing	▪ Evidence of perfusion deficit: - pulse deficit - systolic blood pressure difference - focal neurological deficit (in conjunction with pain) ▪ Aortic diastolic murmur (new and with pain) ▪ Hypotension or shock

Table modified from Rogers AM et al Circulation 2011;123:2213-8.
Risk score varies from 0–3 according the number of positive categories (1 point per column).

Based on the probability of acute aortic syndromes the decision making can be based according to developed flow-chart.

Flowchart for decision-making based on pretest sensitivity of AAS



AAS = acute aortic syndrome; AD = aortic dissection; ESC = European Society of Cardiology; CT = computed tomography; ECG = electrocardiogram; MRI = magnetic resonance imaging; STEMI = ST-segment elevation myocardial infarction; TTE = transthoracic echocardiography; TOE = transoesophageal echocardiography.
[a]STEMI can be associated with AAS in rare cases. - [b]Pending local availability, patient characteristics, and physician experience.
[c]Proof of Type A AD by the presence of flap, aortic regurgitation, and/or pericardial effusion. - [d]Preferably point-of-care, otherwise classical.
[e]Also troponin to detect non-ST-segment elevation myocardial infarction.

Take home messages

4. Treatment of acute aortic syndrome

4.1 Medical management

A lot of patients with aortic diseases have comorbidities such as coronary artery disease, chronic kidney disease, diabetes, dyslipidaemia, hypertension and others. Therefore treatment and prevention strategies have to be similar to those indicated for the above diseases. Specific treatments in different aortic diseases are addressed in each specific chapter.

4.2 (Thoracic) endovascular aortic repair ((T)EVAR)

- It is recommended to decide the indication of endovascular repair on individual basis according to anatomy, pathology, comorbidity and anticipated durability, of any repair using multidisciplinary approach (Class I C).
- A sufficient proximal and distal landing zone of at least 2 cm is recommended for the safe deployment and durable fixation of TEVAR (Class I C).
- 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 (Class I C).
- During stent graft placement, invasive blood pressure monitoring and control either pharmacologically or by rapid pacing is recommended (Class I C).
- For complicated type B aortic dissection, TEVAR is recommended (Class I C).
- If the anatomy is suitable and the expertise available endovascular repair should be preferred over open surgery in contained rupture of thoracic aortic aneurysm (Class I C).
- For uncomplicated type B aortic dissection endovascular therapy should be considered (Class IIa B) as well as in complicated type B intramural haematoma, complicated type B penetrating aortic ulcer and traumatic aortic injury (Class IIa C)

4.3 Surgery in acute aortic syndrome

- In patient with type A aortic dissection urgent surgery is recommended (Class I B).
- Surgery is also indicated in typ A intramural haematoma (Class I C).
- In case of type B penetrating aortic ulcer, surgery should be considered (Class IIa C).
- For complicated type B aortic dissection, intramural haematoma and penetrating aortic ulcer surgery may be considered (Class IIb C).

5. Aortic aneuryms

- 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 (Class I C).
- In case of aneurysm of the abdominal aorta, duplex ultrasound for screening of peripheral artery disease and peripheral aneurysms should be considered (Class IIa C).
- Patients with aortic aneurysms are at increased risk of cardiovascular disease, general principles of cardiovascular prevention should be considered (Class IIa C).

5.1 Indication for intervention for ascending and arch aortic aneuryms

- Surgery is indicated in patients who have aortic root aneurysms with maximal aortic diameter ≥50 mm for patients with Marfan syndrome (Class I C).
- Surgery should be considered in patients who have aortic root aneurysm with maximal ascending aortic diameter:
  - ≥45 mm for patients with Marfan syndromes with risk factors (family history of aortic dissection and/or aortic diameter increase >3 mm/year
  - ≥50 mm for patients with bicuspid valve with risk factors
  - ≥55 mm for patients with no elastopathy (Class IIa C).

Take home messages

- Surgery should be considered in patients who have isolated aortic arch aneurysm with maximal diameter ≥55 mm (Class IIa C).
- Lower levels thresholds for intervention may be considered according to body surface area in patients of small stature or in case of rapid progression, aortic valve regurgitation, planned pregnancy, and patients preference (Class IIb C).
- Aortic arch repair may be considered in patient with aortic arch aneurysm who already have an indication for surgery of an adjacent aneurysm located in the ascending aorta or descending aorta (Class IIb C).

5.2 Intervention for descending aortic aneurysms

Valvular problems associated with bicuspid aortic valve (BAV) are covered in the 2012 ESC/EACTS guidelines on management of valvular diseases *(European Heart J 2012;33:2451-2496)*.

6. Abdominal aortic aneurysm

- Abdominal aortic aneurysms (AAA) have a prevalence of about 2% and are particularly found in men >65 years and women who are smoking >65 years. Aortic AAA are usually asymptomatic until rupture occurs. The aortic diameter relates to risks of rupture. As a screening tool ultrasound is recommended in all men >65 years (Class I A) and considered in women >65 years and tobacco smoking (Class IIb C).
- Very new is the advice, to use a 2 minutes extra time during TTE to check for existence of an asymptomatic AAA in men >65 years (Class IIa B) and women >65 years who are smoking (Class IIb C).
- Target screening should be considered in first degree siblings of AAA patients (Class IIa B).
- Please check 2014 ESC/ESA Guidelines on non-cardiac surgery for cardiovascular risk assessment and management *(European Heart J 2014:35:2383-2431)*. Additional information concerning reduction of cardiac risk in case of intervention and surgery are given.

6.1 Endovascular aortic repair and open vascular surgery

About 60% of all AAA are suitable for endovascular therapy. In randomized controlled studies endovascular aortic repair (EVAR) reduced mortality threshold. But long-term result were similar due to high re-intervention rates.

- Smoking cessation is recommended to slow the AAA growth (Class I B).
- AAA repair is indicated if AAA diameter exceeds 55 mm (Class I B).
- If the anatomy is suitable for EVAR, either open or endovascular aortic repair is recommended (Class I A).
- If the aneurysm is anatomically not suitable for EVAR, open endovascular aortic surgery is recommended (Class I C).

6.2 Management of symptomatic abdominal aortic aneurysms

In case of rupture of abdominal aortic aneurysms two randomized controlled trials are available demonstrating similar 30 days mortality results (30.4% versus 37.4%). Based on these results following recommendations are given.

- In suspected ruptured AAA, immediate abdominal ultrasound is recommended (Class I C).
- In case of rupture, AAA emergency repair is indicated (Class I C).
- In case of symptomatic but non rupture AAA, urgent repair is indicated (Class I C).
- In case of symptomatic AAA anatomical suitable for EVAR, either open or endovascular repair is recommended (Class I A).

Take home messages

7. Long-term follow-up for chronic aortic dissection

- Contrast CT or MRI is recommended to confirm the diagnosis of chronic aortic dissection (Class I C).
- Close imaging surveillance in aortic dissection is indicated to detect signs of complications (Class I C).
- In patients with chronic aortic dissection, tight blood pressure control (> 130/80 mmHg) is indicated (Class I C).
- After TEVAR or EVAR surveillance is recommended after 1 month, 6 months, 12 months and then yearly (Class I C).
- CT is recommended as the first choice imaging technique for follow-up after TEVAR or EVAR (Class I C).
- In AAA Doppler ultrasound with or without contrast agents should be considered for annual postoperative surveillance, with non-contrast CT imaging over 5 years (Class IIa C).
- For follow-up in young patients MRI should be preferred to CT for imaging magnetic resonance-compatible stent grafts (Class IIa C).

8. Genetic diseases affording the aorta

Chromosomal and inherited syndromic thoracic aneurysms

During the last years more insight into chromosomal aortic diseases have been given not only for the Marfan syndrome, but also for the Loeys-Dietz syndrome, the Turner syndrome, the Ehlers Danlos syndrome Typ IV, non syndromic familiar aortic aneurysms and even aneurysms – osteoarthritis syndrome and arterial tortuosity syndrome.

- It is recommended to investigate first degree relatives (siblings and parents) of a subject with thoracic aortic disease to indentify a familiar form in which relatives all have a 50% chance of carrying the familiar mutation-disease (Class I C).
- Once a familial form of thoracic acute aortic dissection (TAAD) is highly suspected, it is recommended to refer the patient to geneticist for family investigation and molecular testing (Class I C).
- Variable of age of oncet warrants screening every 5 years of "healthy" at-risk relatives until diagnosis (clinical or molecular) is established or ruled out (Class I C).
- 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) (Class IIa C).

8.1 Medical therapy in genetic diseases

- In Marfan syndrome beta-blockers are prescribed to reduce the progression rate.
- Angiotensin 2 receptor blockers demonstrated attenuation of the dilatation rate of aortic aneurysms.
- In Ehlers-Danlos syndrome beta-blockers reduce arterial complications.
- No specific data are available for other genetic diseases.

9. Management of bicuspid aortic valve

BAV have a prevalence of about 1%. Fusion of the right and left coronary cusp or fusion of the right and none coronary cusp or found combined with normal size aorta, supra-coronary dilatation or cylindric aortic shape. The maximal aortic dilatation rate does not differ for bicuspid aortic valve and Marfan syndrome and is maximal in the tubular aorta (0.42 ± 0.6 and 0.49 ± 0.5 mm/year). BAV have a high heritability with about one quarter with bicuspid aortic valve found in the first degree relatives. The aortic root dilatation is found in about one third in first degree relatives.

Take home messages

- In case of BAV, surgery is indicated when the aortic root or thoracic aorta diameter is >55 mm, >50 mm in presence of other risk factors, >45 mm when surgical aortic valve replacement is scheduled (Class I C).
- Because of the familial occurence screening of first degree relatives should be considered (Class IIa C).

10. Future developments

The installation of hybrid rooms have been shown to be pacemakers for the development of new diagnostic and treatment options like thoracic endovascular aortic repair, debranching or aortic arch surgery, and the frozen elephant trunk. This can lead in the future to further paradigma changes in the diagnosis and treatment of aortic diseases.

It is time to form aortic teams and centers in order to provide full access to experts in the field of cardiology, radiology, pediatric cardiology, genetics, aortic and cardiovascular surgery, which is needed not only for the acute but also for the intense follow-up of patients with aortic diseases.

Major gaps in evidence

- The 2014 ESC Guidelines on diagnosis and treatment of aortic diseases contains in 118 recommendations 4% Class I A recommendations, and in 15% Class I-II B recommendations. Thus, 80% are consensus decision.
- We need more epidemiological data on acute aortic syndrome in Europe.
- More evidence needed on the caseload-outcome relationship in the field aortic diseases.
- The implantation and efficacy of aortic centers in Europe should be assessed. The establishment of a European network of aortic centers should be encouraged along with establishment of large registries and multicenter studies.
- The value of biomarkers should be clarified.
- More data of accuracy and reproducibility of aortic measurements are needed.
- The knowledge on relationship between aortic size and outcome should be improved and the superiority of 3D on 2D better documented.
  It has to been investigated, if there are difference of aortic diameters related to age, gender and body-size taken into account outcome of interventions.
- Data for female patients with aortic diseases are lacking.
- The lack of evidence on the efficacy of medical therapy and the role of antihypertensive drugs, statins, inflammatory drugs an non-syndromic aortic diseases like aortic dissection, thoracic aortic aneurysms and AAA, genetic diseases is present.
- For thoracic aortic aneurysms, randomized studies are needed and the optimal timing for preventive intervention according to lesion size and other characteristics as well as individual patient features.
- The optimal timing and techniques of intervention in chronic aortic dissection is still unclear.

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