

Decision-making in Aneurysmal Aortic Disease

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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

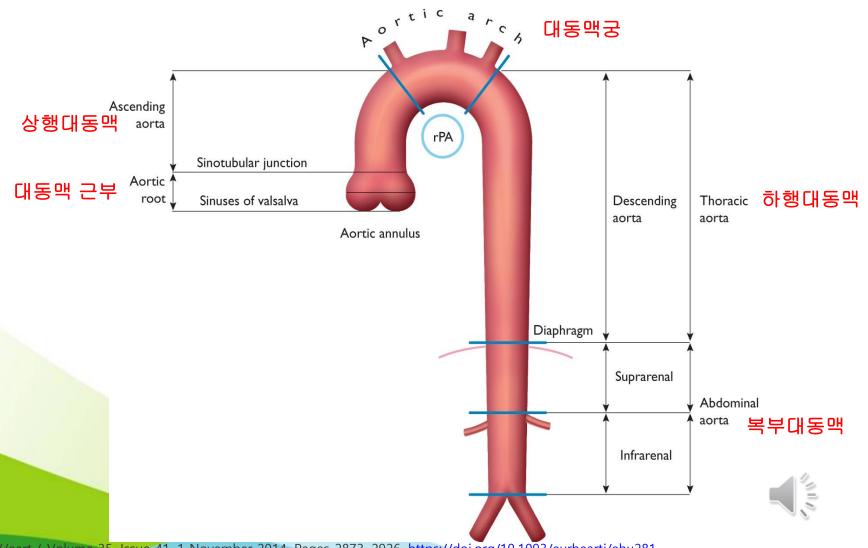
The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC)

ACC/AHA CLINICAL PRACTICE GUIDELINE

2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines



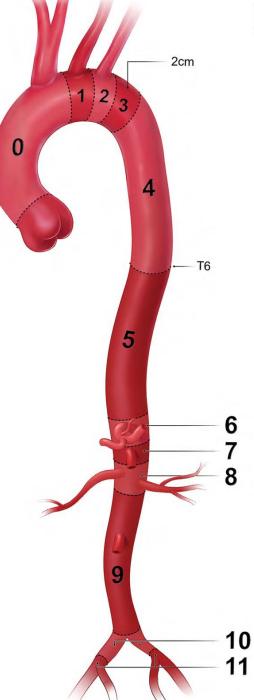
Anatomy of aorta



Fur Heart J, Volume 35, Issue 41, 1 November 2014, Pages 2873–2926, https://doi.org/10.1093/eurheartj/ehu281



Classification of Aortic Anatomic Segments by 11 Landing Zones







Normal aorta

- 3 layer (intima, media, adventia)
- Conduit function, control of the systemic vascular resistance and HR
- Healthy aorta < 40mm and taper gradually downstream
- Rate of aortic expansion : 0.9mm(men),0.7mm(women) for each decade





Aortic aneurysm

- Aortic root & ascending aorta aneurysm
- Aortic arch aneurysm
- Descending thoracic aortic aneurysm
- Thoraco-abdominal aortic aneurysm
- Abdominal aortic aneurysm



Broad range of symptoms

- Cough, shortness of breath, or difficult or painful swallowing → large TAAs.
- Constant or intermittent abdominal pain or discomfort, a pulsating feeling in the abdomen, or feeling of fullness after minimal food intake → Large AAAs.
- Stroke, transient ischemic attack, or claudication → secondary to aortic atherosclerosis.
- Hoarseness due to left laryngeal nerve palsy in rapidly progressing lesions. → Large TAA



Image modalities

Table 3 Comparison of methods for imaging the aorta

Advantages/disadvantages	TTE	TOE	СТ	MRI	Aortography
Ease of use	+++	++	+++	++	+
Diagnostic reliability	+	+++	+++	+++	++
Bedside/interventional use ^a	++	++	-	-	++
Serial examinations	++	+	++(+) ^b	+++	-
Aortic wall visualization ^c	+	+++	+++	+++	-
Cost	-	-			
Radiation	0	0		_	
Nephrotoxicity	0	0			

 $^{+\} means\ a\ positive\ remark\ and \\ --means\ a\ negative\ remark.\ The\ number\ of\ signs\ indicates\ the\ estimated\ potential\ value$

CT = computed tomography; MRI = magnetic resonance imaging; TOE = transoesophageal echocardiography; TTE = transthoracic echocardiography.



^aIVUS can be used to guide interventions (see web addenda)

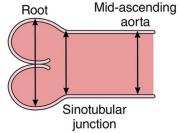
b+++ only for follow-up after aortic stenting (metallic struts), otherwise limit radiation

^cPET can be used to visualize suspected aortic inflammatory disease

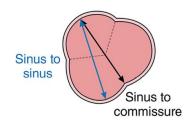


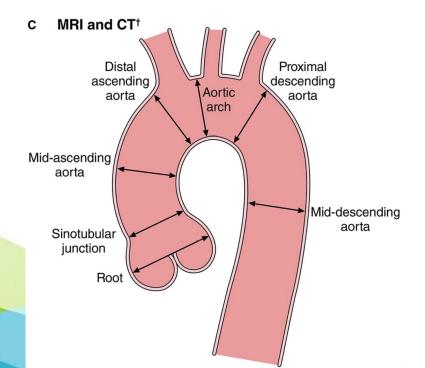
Aortic Imaging Techniques

Echocardiography*



Sinus measurement









Treatment options

- Medical therapy
 - Reducing BP(130/80) & cardiac contractility
 - b-blocker ,ARB, statin, low dose aspirin

- Endovascular therapy
 - TEVAR(thoracic endovascular aortic repair)
 - EVAR(abdominal endovascular aortic repair)







Thoracic aortic aneurysms (TAA)

- 5 to 10 per 100 000 person years
- Risk factors
 - ; Hypertension, smoking, hypercholesterolemia, and heritable genetic variants are risk factors
- TAA \uparrow → AAA & Cerebral aneurysms \uparrow
- Aortic root and ascending thoracic aorta
 - ; Heritable influence and present at younger ages, BAV
- Descending thoracic aorta
 - ; degenerative and present at older age



Cause of TAA

Recommendations for HTAD: Genetic Testing and Screening of Family Members for TAD

Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations		
1	B-NR	 In patients with aortic root/ascending aortic aneurysms or aortic dissection, obtaining a multigenerational family history of TAD, unex- plained sudden deaths, and peripheral and intracranial aneurysms is recommended.¹⁻³ 		
1	B-NR	 In patients with aortic root/ascending aortic aneurysms or aortic dissection and risk factors for HTAD (Table 8, Figure 17), genetic testing to identify pathogenic/ likely pathogenic variants (ie, mutations) is recommended.⁴⁻⁶ 		
1	B-NR	3. In patients with an established pathogenic or likely pathogenic variant in a gene predisposing to HTAD, it is recommended that genetic counseling be provided and the patient's clinical management be informed by the specific gene and variant in the gene. ⁷⁻⁹		
1	B-NR	4. In patients with TAD who have a pathogenic/ likely pathogenic variant, genetic testing of at- risk biological relatives (ie, cascade testing) is recommended. ^{6,10,11} In family members who are found by genetic screening to have inherited the pathogenic/likely pathogenic variant, aortic imaging with TTE (if aortic root and ascending aorta are adequately visualized, otherwise with CT or MRI) is recommended. ^{4,5,12}		
1	B-NR	5. In a family with aortic root/ascending aortic aneurysms or aortic dissection, if the disease-causing variant is not identified with genetic testing, screening aortic imaging (as per recommendation 4) of at-risk biological relatives (ie, cascade testing) is recommended. ¹³⁻¹⁵		

HTAD (see Table 7): syndromic

Marfan syndrome

Loeys-Dietz syndrome

Vascular Ehlers-Danlos syndrome

Smooth muscle dysfunction syndrome

Others: attributable to pathogenic variants in FLNA, BGN, LOX

HTAD (see Table 7): nonsyndromic

ACTA2, MYH11, PRKG1, MYLK, and others

Familial thoracic aortic aneurysm without identified pathogenic variants in a known gene for HTAD

Congenital conditions

Bicuspid aortic valve

Turner syndrome

Coarctation of the aorta

Complex congenital heart defects (tetralogy of Fallot, transposition of the great vessels, truncus arteriosus)

Hypertension

Atherosclerosis

Degenerative

Previous aortic dissection

Inflammatory aortitis

Giant cell arteritis

Takayasu arteritis

Behçet disease

Immunoglobulin G4-related disease, antineutrophil cytoplasmic antibody-related, sarcoidosis

Infectious aortitis

Bacterial, fungal, syphilitic

Previous traumatic aortic injury

HTAD indicates heritable thoracic aortic diseases; and TAA, thoracic aortic aneurysms.

Definitions of aortic aneurysm

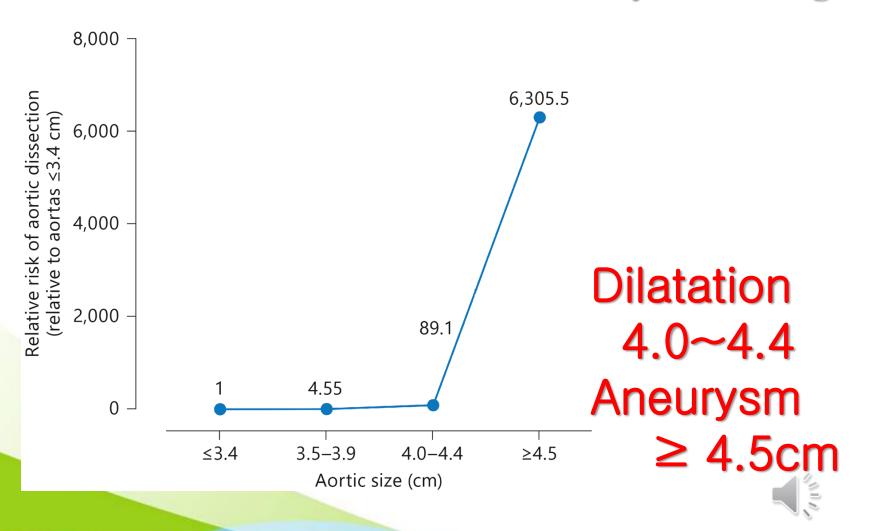
- The conventional definition
 - ; Dilated to at least 1.5 times its expected normal diameter

- Defining aneurysms of the aortic root and ascending thoracic aorta
 - ; 40's male 3.5cm, \rightarrow 1.5 times ; 5.25cm
 - ; Marfan syndrome or a familial thoracic aortic aneurysm (< 5.0cm. Op indication)





Relative Risk of Aortic Dissection by Size Range





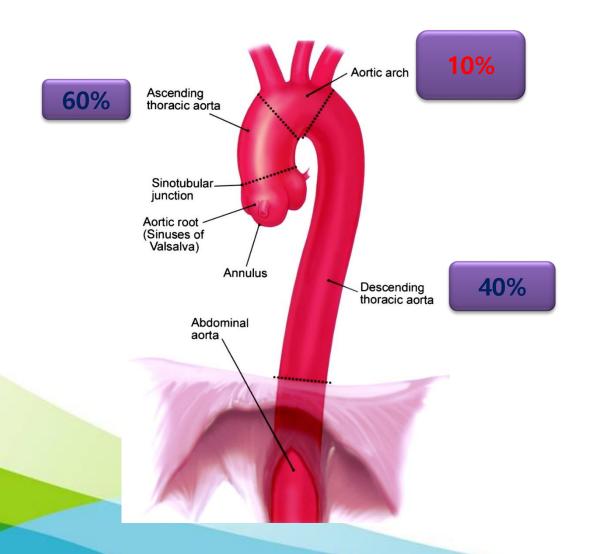
Normalizing Aortic Root and Ascending Aortic Diameters for Body Size

- The Z-Score
 - ; Pediatric population
- The Aortic Size Index and Aortic Height Index
- The Cross-Sectional Area to Height Ratio
 - ; Ratio of the cross-sectional area of the aorta(cm²) to the patient's height (m), >10 cm²/m high risk





Thoracic aortic Aneurysm







Aortic root & ascending aortic aneurysm

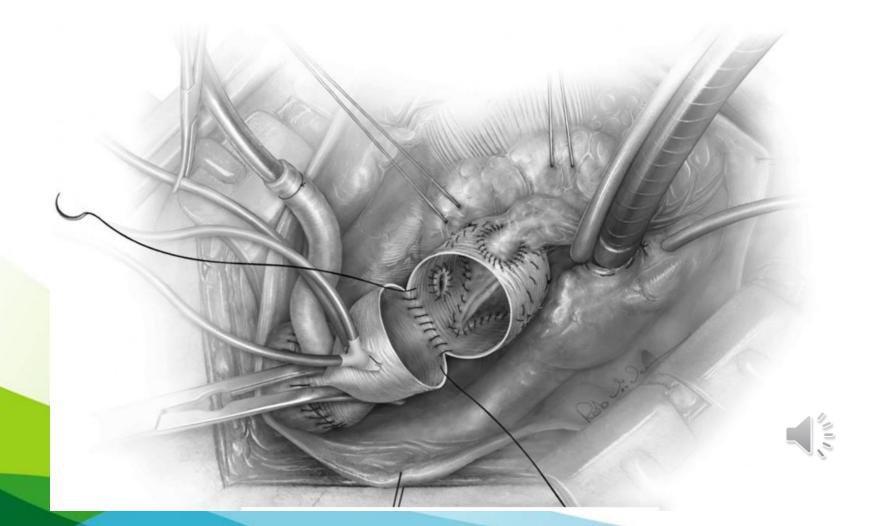
Recommendations on interventions on ascending aortic aneurysms

Recommendation	Recommendations		
aortic root aneurys aortic diameter ^c ≥5	Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter ^c ≥50 mm for patients with Marfan syndrome.		
who have aortic roomaximal ascending at 245 mm Marfan factors • ≥50 mm bicuspi factors • ≥55 mm	n for patients with syndrome with risk of for patients with risk of the valve with risk of the valve with risk	lla	U
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	С



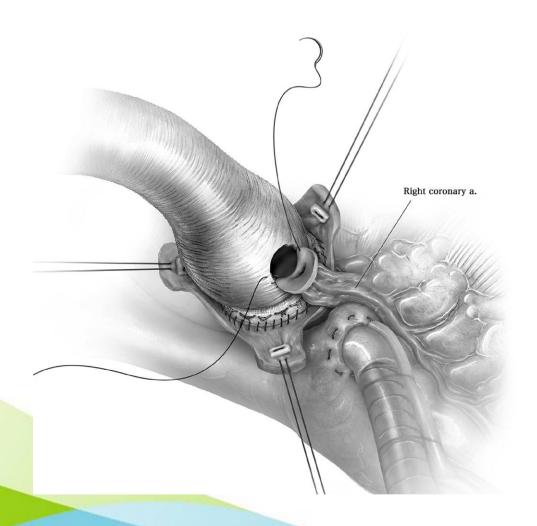


Valve sparing David operation





The Button Bentall Operation

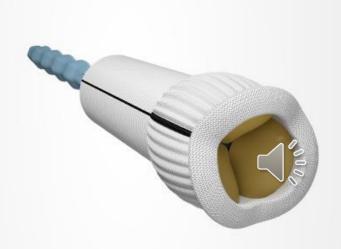






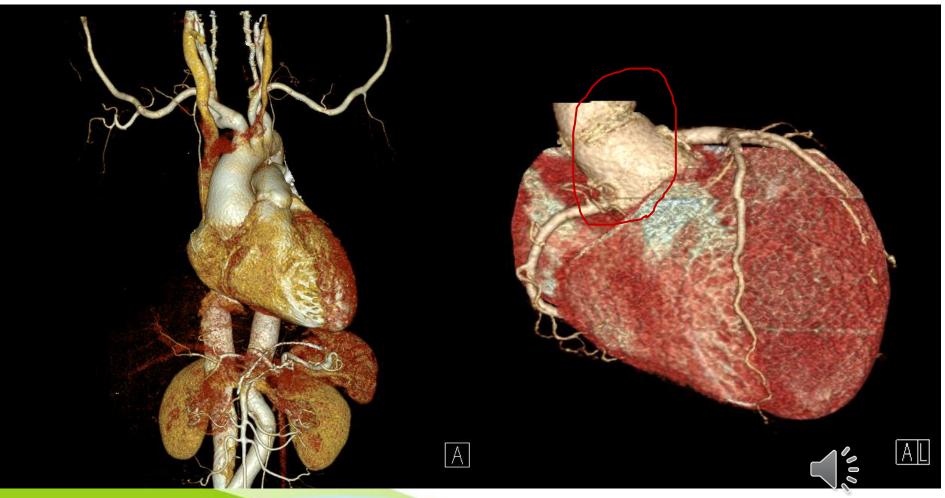
Valved Graft







Aortic root aneurysm



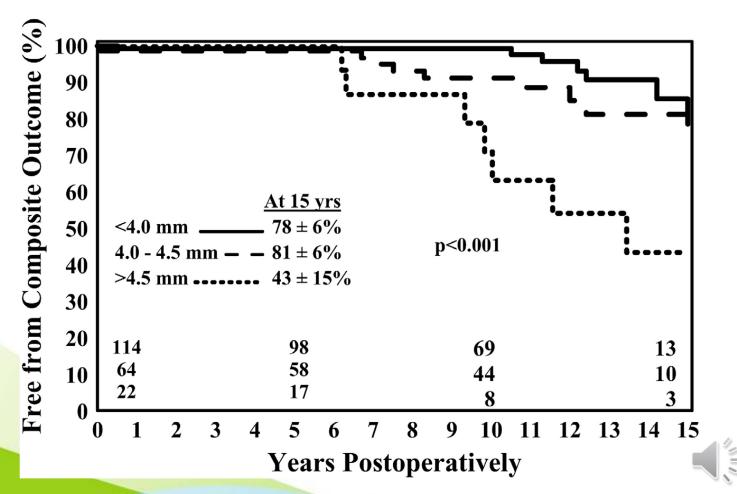
Bio-bentall OP (Handmade AVG: CE magna ease 23mm+ Graft 28mm)

Aortic diseases associated with bicuspid AV

- Common congenital cardiac defect : 1~2%
- Male > female (2:1~4:1)
- LCC and RCC fusion (70%)
- 27% will require cardiovascular surgery within20 years among all BAV patients.
- Notch1 gene mutations : aortopathy, unknown
- When surgery is indicated for BAV(AS, AR, aortic root replacement) should be considered if the root is larger than 45 mm in diameter,

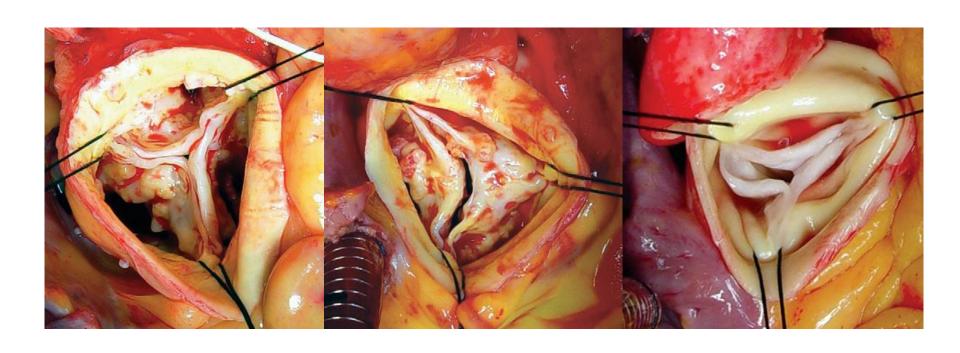


Aortic aneurysm in bicuspid AV



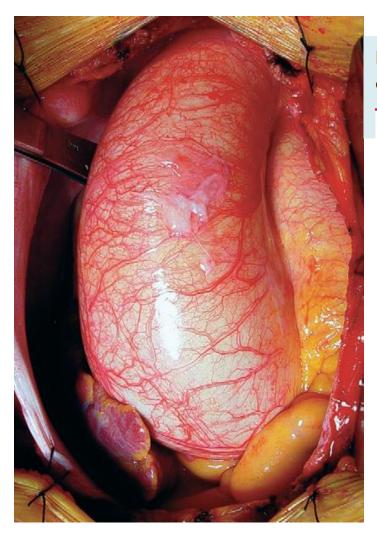
Aortic complications (aneurysm, dissection, or sudden death)

Aortic valve disease(AS, AR)





Ascending aorta in bicuspid AV



Recommendations for the management of aortic root dilation in patients with bicuspid aortic valve

Recommendations	Class ^a	Levelb
In cases of BAV, surgery of the ascending aorta is indicated in case of: • aortic root or ascending aortic diameter >55 mm. • aortic root or ascending aortic diameter >50 mm in the presence of other risk factors.	ı	С
aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled.		



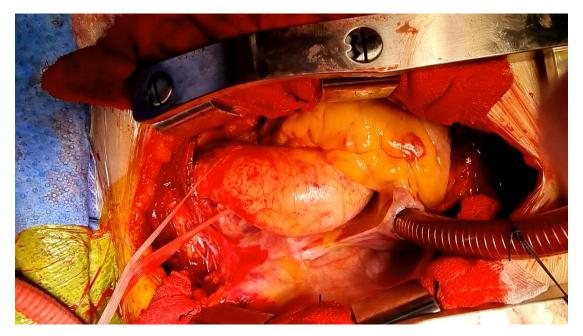
Recommendations for BAV Aortopathy Interventions: Replacement of the Aorta in Patients With BAV

Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	 In patients with a BAV and a diameter of the aortic root, ascending aorta, or both of ≥5.5 cm, surgery to replace the aortic root, ascending aorta, or both is recom- mended.¹⁻³
2a	B-NR	 In patients with a BAV and a cross-sectional aortic root or ascending aortic area (cm²) to height (m) ratio of ≥10 cm²/m, surgery to replace the aortic root, ascending aorta, or both is reasonable, when performed by expe- rienced surgeons in a Multidisciplinary Aortic Team.^{3,4}
2a	B-NR	3. In patients with a BAV, a diameter of the aortic root or ascending aorta of 5.0 cm to 5.4 cm, and an additional risk factor for aortic dissection (Table 14), surgery to replace the aortic root, ascending aorta, or both is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team. ^{1,5}
2a	B-NR	4. In patients with a BAV who are undergoing surgical aortic valve repair or replacement, and who have a diameter of the aortic root or ascending aorta of ≥4.5 cm, concomitant replacement of the aortic root, ascending aorta, or both is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team. ^{1,6}
2b	B-NR	5. In patients with a BAV, a diameter of the aortic root or ascending aorta of 5.0 cm to 5.4 cm, no other risk factors for aortic dissection (Table 14), and at low surgical risk, surgery to replace the aortic root, ascending aorta, or both may be reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team. ^{1,2,5}











Pre op. vs Post op.





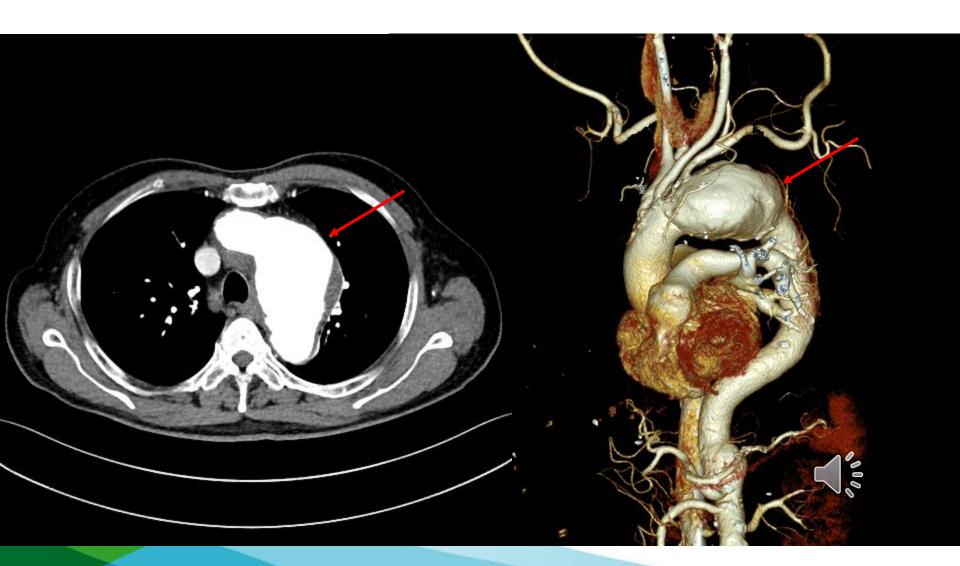
Arch & descending aneurysm (ESC 2014)

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



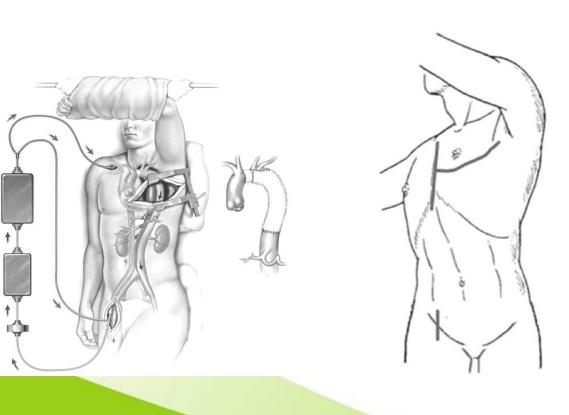


Aortic arch aneurysm





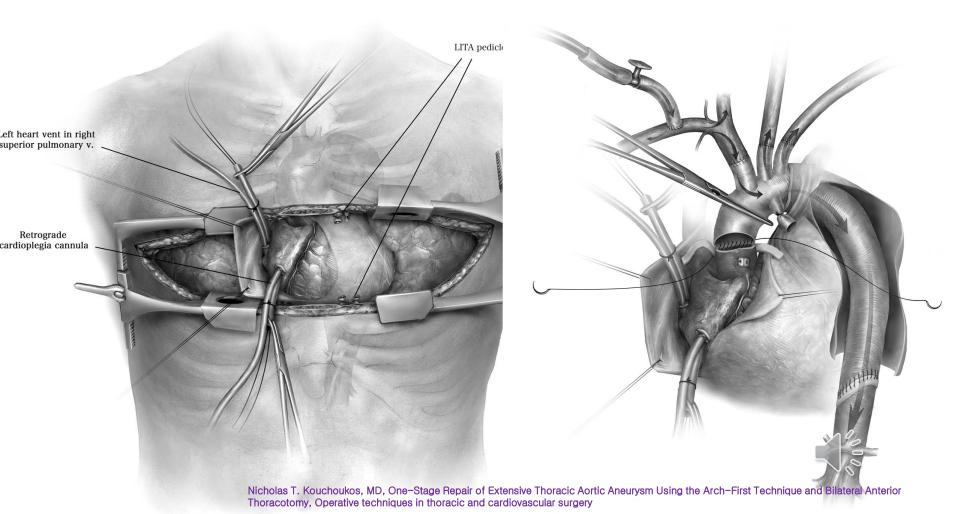
Various approach for aortic arch surgery







One-Stage Repair of Extensive Thoracic Aortic Aneurysm



New and expanded classification system (1)



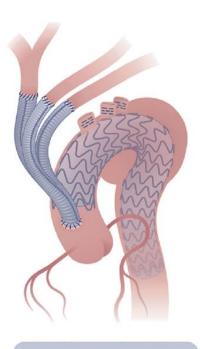
Zone 0 with innominate snorkel (0_s)



Zone 0 with branched endograft (0_B)



Zone 1



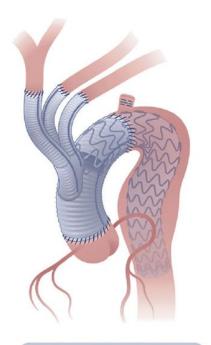
Type I_N

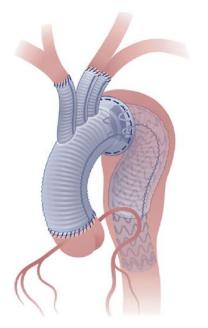
Hughes GC, Vekstein A. Current state of hybrid solutions for aortic arch aneurysms. Ann Cardiothorac Surg 2)21:10(6):731-743.

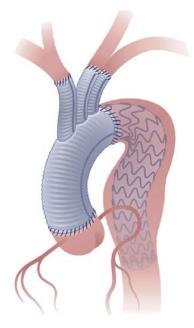
Hybrid aortic arch repair

New and expanded classification system (2)









Type II

Type III with conventional elephant trunk (III_c)

Type III with frozen elephant trunk (III_F)

Hughes GC, Vekstein A. Current state of hybrid solutions for aortic arch aneurysms. Ann Cardiothorac Surg 2021:10(6):731-743.



Type III c elephant trunk + Antegrade TEVAR

Hybrid TEVAR after TAR with elephant trunk

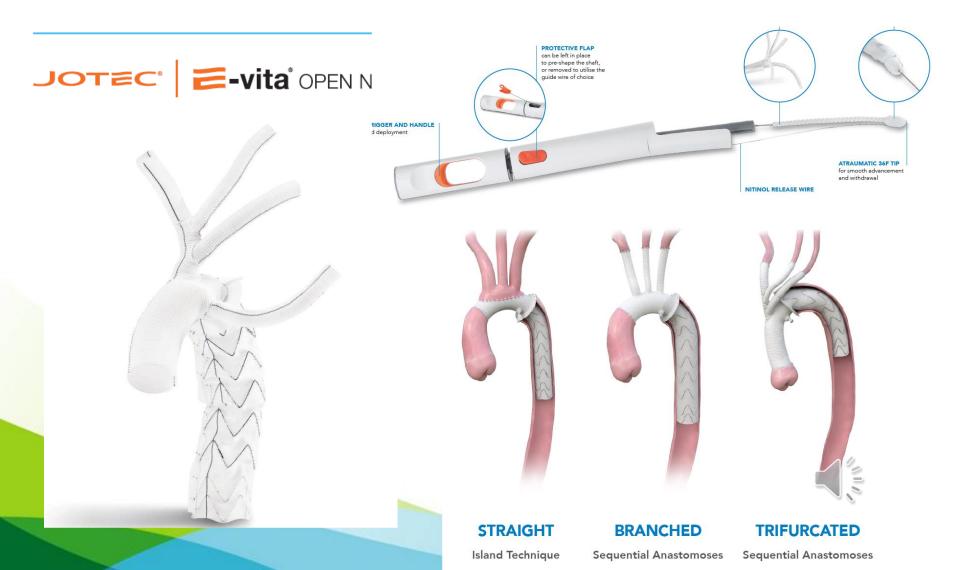
Seoul St. Mary's Hospital

Dept. Cardiovascular surgery





Frozen Elephant Trunk in Aortic Arch Disease





TAR c FET (E-vita neo)

TAR c FET (E-vita neo)

70yrs/male, Aneurysm of Arch to proximal aorta

The catholic University of Korea

Seoul St. Mary's Hospital

Dept. Cardiovascular surgery





2022 ACC/AHA Guideline

6.5.3.1. Size Thresholds for Repair of Descending TAA

Recommendations for Size Thresholds for Repair of Descending TAA Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	 In patients with intact descending TAA, repair is recommended when the diameter is ≥5.5 cm.^{1,2}
2 b	B-NR	2. In patients with intact descending TAA and risk factors for rupture (Table 17), repair may be considered at a diameter of <5.5 cm. ²⁻⁶
2 b	B-NR	3. In patients at increased risk for perioperative morbidity and mortality (Table 18), it may be reasonable to increase the size threshold for surgery accordingly. ⁷



Table 17. Risk Factors for Aortic Rupture Among Patients With Descending TAA

Aneurysm growth of ≥0.5 cm/y³

Symptomatic aneurysm4

Marfan, Loeys-Dietz, or vascular Ehlers-Danlos syndrome, or HTAD (see Section 6.1.2, "Genetic Aortopathies")²

Saccular aneurysm⁵

Female sex²

Infectious aneurysm⁶

HTAD indicates heritable thoracic aortic disease; and TAA, thoracic acrtic aneurysm.



2022 ACC/AHA Guideline

Recommendations for Endovascular Versus Open Repair of Descending TAA

Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	1. In patients without Marfan syndrome, Loeys- Dietz syndrome, or vascular Ehlers-Danlos syndrome, who have a descending TAA that meets criteria for intervention and anatomy suitable for endovascular repair, TEVAR is recommended over open surgery. ¹⁻⁴
1	B-NR	2. In patients with a descending TAA that meets criteria for repair with TEVAR, who have smaller or diseased access vessels, considerations for alternative vascular access are recommended. ⁵
2a	B-NR	3. In patients with a descending TAA that meets criteria for intervention, who have anatomy unsuitable for endovascular repair, and who are without significant comorbidities and have a life expectancy of at least 10 years, open surgical repair is reasonable. ⁶⁻⁹



Zone 0: ascending aorta (Ao) to innominate artery (innom.)

ascending

Zone 1: innominate artery to left common carotid (LCC)

Zone 2: LCC to left subclavian artery (LSA)

Zone 3: first 2 cm distal to LSA

Zone 4: Zone 3 to mid descending Ao (~T6)

Zone 5: mid descending Ao to celiac artery

Zone 6: celiac artery to superior mesenteric artery (SMA)

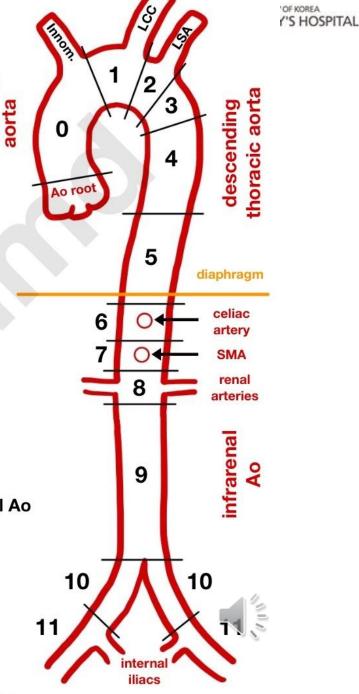
Zone 7: SMA to renal arteries

Zone 8: renal to infra-renal abdominal Ao

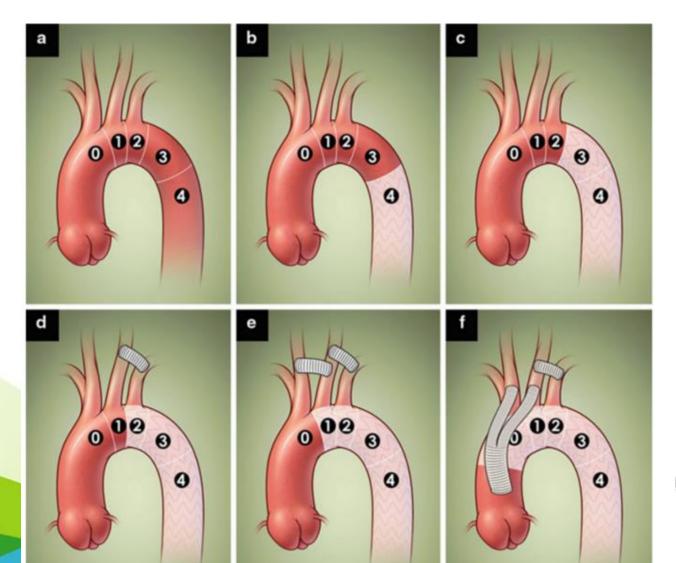
Zone 9: infrarenal abdominal Ao

Zone 10: common iliac arteries

Zone 11: external iliac arteries



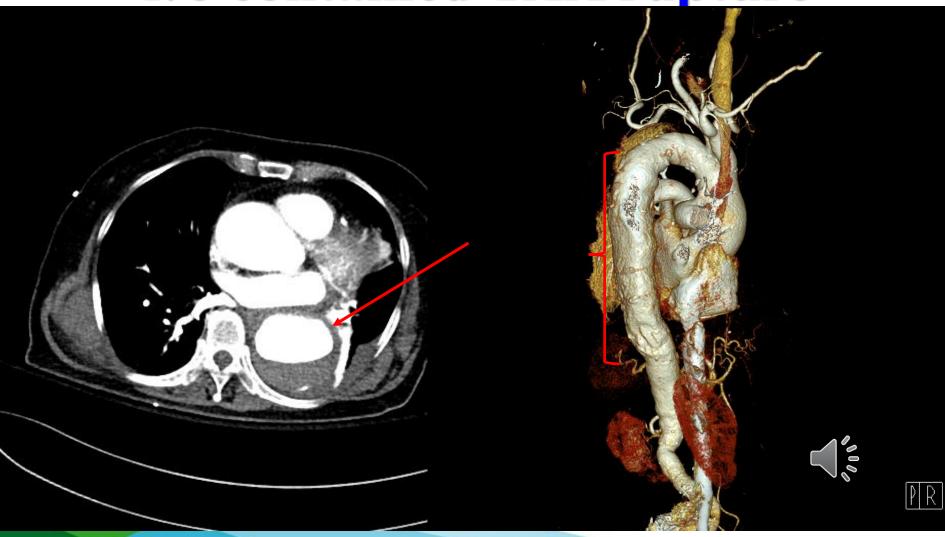
TEVAR

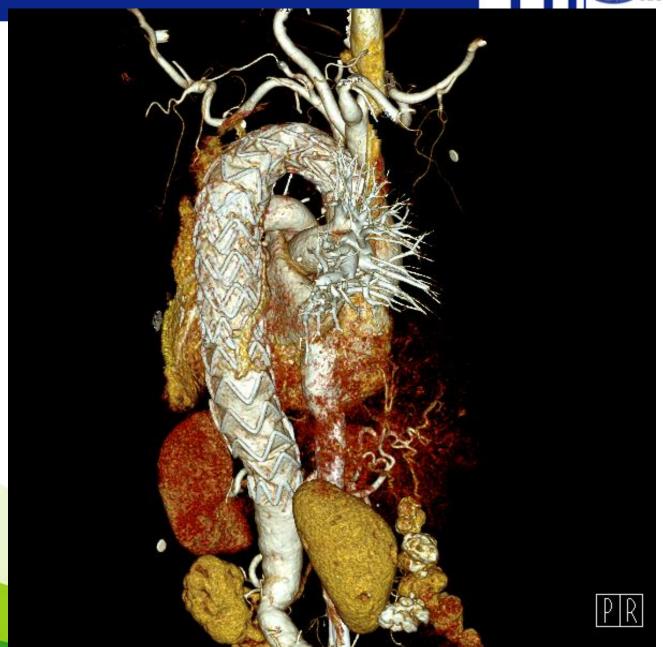






Zone 3 TEVAR in Type B IMH or r/o contained TAA rupture









Zone 2 TEVAR (LSA to LCC bypass)

- 80 years old male
- Huge Saccular aneurysm of DTA (4.7cm)
- Severe AS (high risk patient)
 - → One stage TAVI and TEVAR





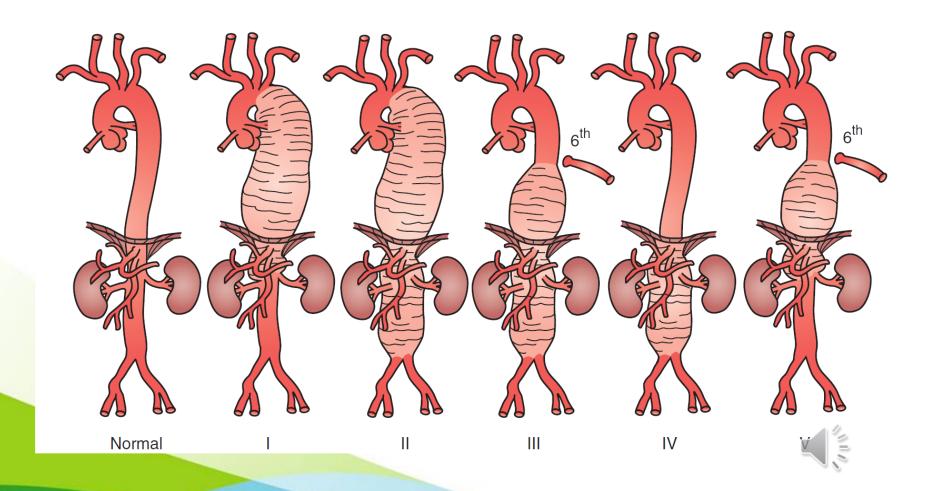
Zone 2 TEVAR (LSA to LCC bypass)







Crawford classification of TAAA





2022 ACC/AHA Guideline

Recommendations for Size Thresholds for Open Surgical Repair of TAAA

Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	 In patients with intact degenerative TAAA, repair is recommended when the diameter is ≥6.0 cm.¹⁻³
2 a	B-NR	2. In patients with intact degenerative TAAA, repair is reasonable when the diameter is ≥5.5 cm and the repair is performed by experienced surgeons in a Multidisciplinary Aortic Team. ¹⁻³
2 a	B-NR	3. In patients with intact degenerative TAAA who have features associated with an increased risk of rupture (Table 19), repair is reasonable when the diameter is <5.5 cm.4



2022 ACC/AHA Guideline

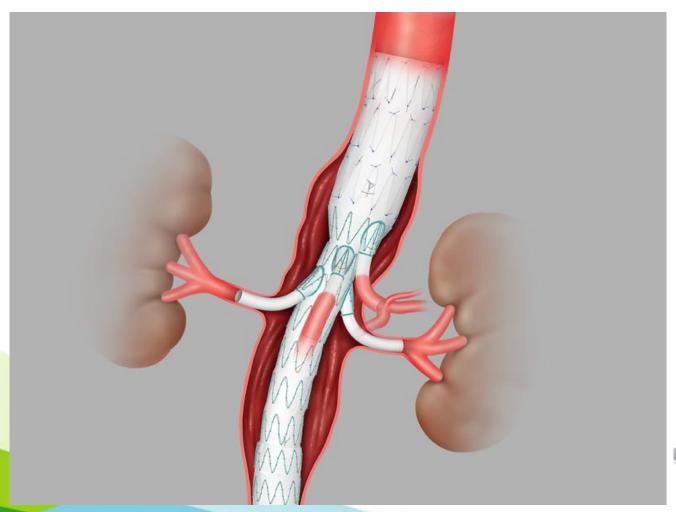
Recommendations for Open Versus Endovascular Repair of TAAA Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
Ruptured TA	AA	
1	B-NR	1. In patients with ruptured TAAA requiring intervention, open repair is recommended. ¹⁻⁵
2 b	C-LD	2. In patients with ruptured TAAA requiring intervention, provided that the patient is hemodynamically stable, endovascular repair may be reasonable in centers with endovascular expertise and access to appropriate endovascular stent grafts. ⁶
Intact TAAA		
1	C-LD	3. In patients with Marfan syndrome, Loeys-Dietz syndrome, or vascular Ehlers-Danlos syndrome and intact TAAA requiring intervention, open repair is recommended over endovascular repair. ⁷⁻⁹
2b	B-NR	4. In patients with intact degenerative TAAA and suitable anatomy, endovascular repair with fenestrated stent grafts, branched stent grafts, or both may be considered in centers with endovascular expertise and access to appropriate endovascular stent grafts. ¹⁰⁻¹³





Zenith® t-Branch® Thoracoabdominal Endovascular Graft





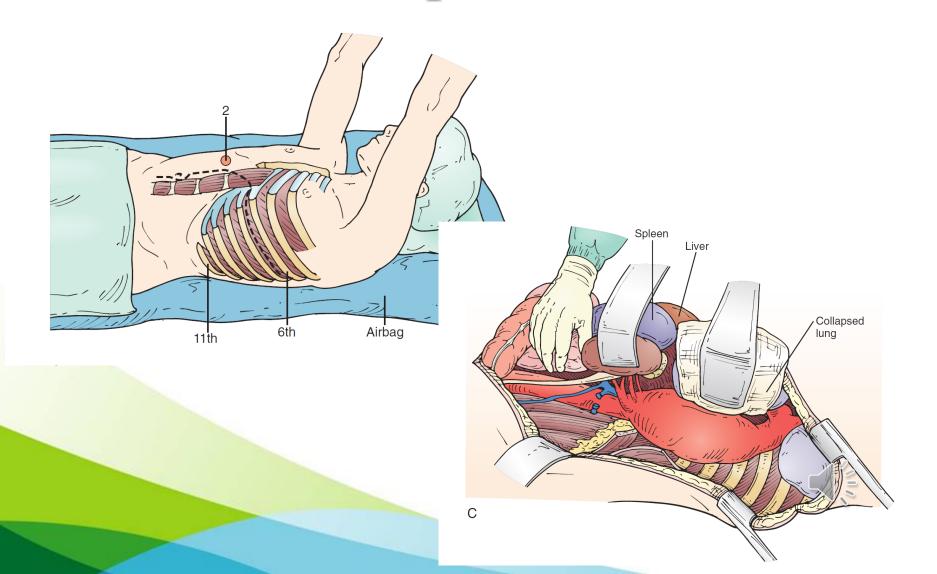


Recommendations for TAAA Spinal Cord Protection Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	A	 In patients undergoing open TAAA repair who are at high risk for SCI, cerebrospinal fluid drainage is recommended to reduce the inci- dence of temporary SCI, permanent SCI, or both.¹⁻⁷
1	B-NR	 In patients who experience delayed spinal cord dysfunction after either open or endo- vascular TAAA repair, timely measures to optimize spinal cord perfusion and decrease intrathecal pressure are recommended (Table 20).^{1-4,8}

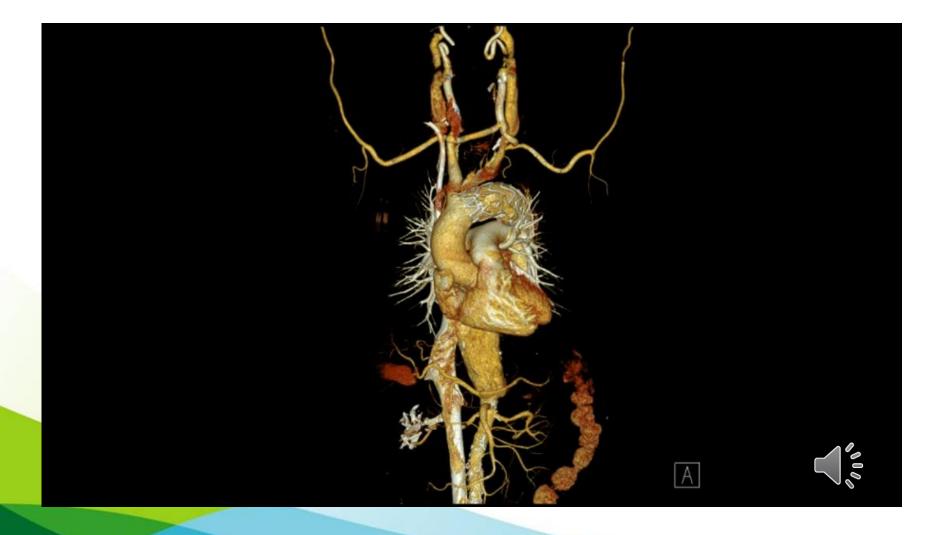


TAAA repair incision





Thoracoabdominal Aortic Aneurysms



Abdominal aortic aneurysm

Recommendations for AAA: Cause, Risk Factors, and Screening Referenced studies that support the recommendations are summarized in the Online Data Supplement.

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

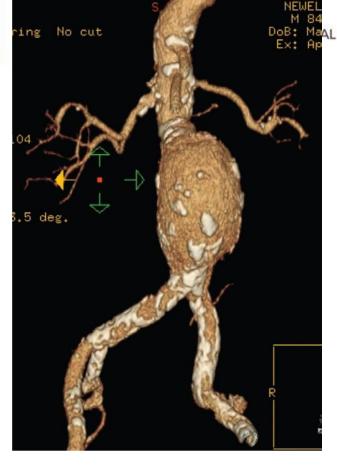
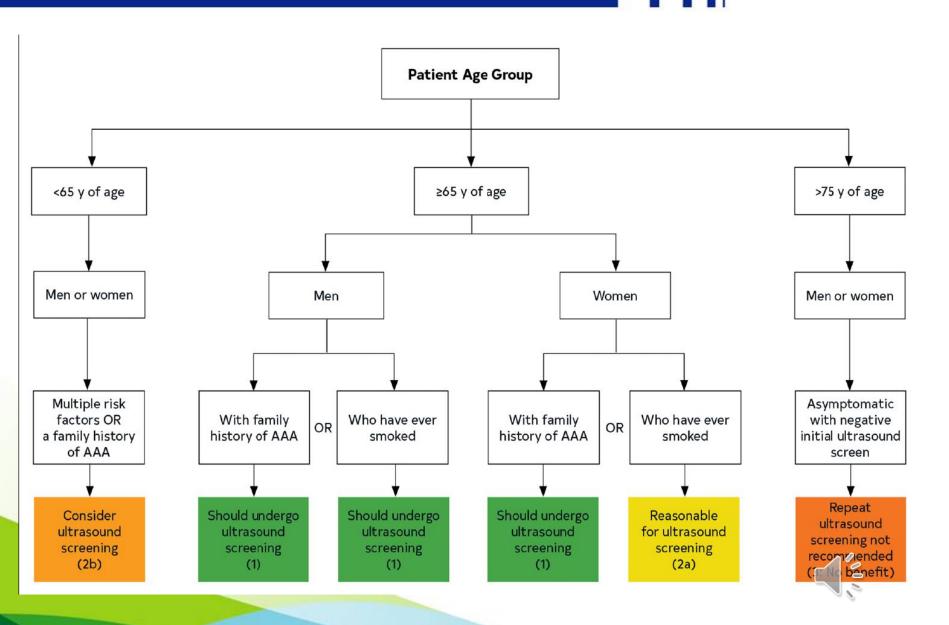


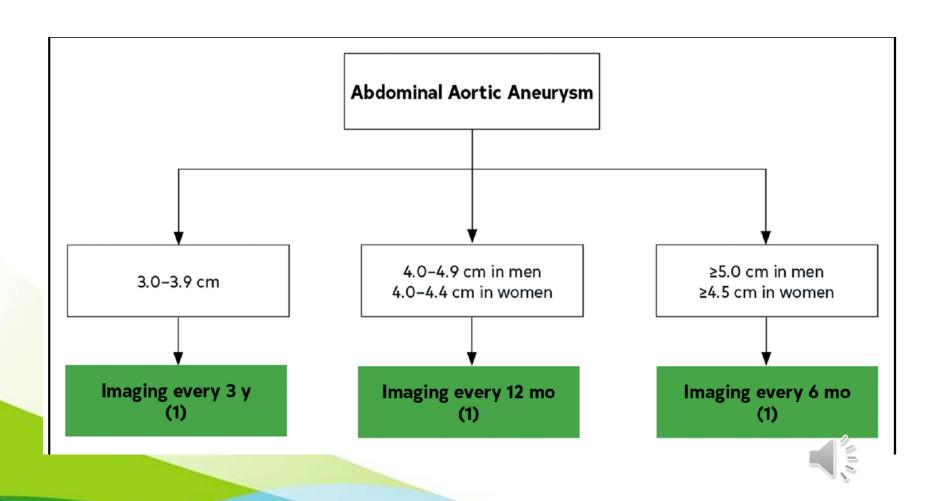
Table 15. Risk Factors for Abdominal Aortic Aneurysm

Strong Risk Factors	Additional Risk Factors
Smoking history	Hypertension
Older age	Hyperlipidemia
Male sex	White race
Family history of abdominal aortic aneurysm	Inherited vascular connective tissue disorcer
	Atherosclerotic cardiovascular disease





Surveillance of AAA





Open Versus Endovascular Repair of AAA

Recommendations for the Threshold for AAA Repair Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations	
1	A	 In patients with unruptured AAA, repair is recommended in those with a maximal aneurysm diameter of ≥5.5 cm in men or ≥5.0 cm in women.¹⁻⁶ 	
COR	LOE	Recommendations	
1	B-NR	2. In patients with unruptured AAA who have symptoms that are attributable to the aneurysm, repair is recommended to reduce the risk of rupture. ^{7,8}	
2b	C-LD	3. In patients with unruptured saccular AAA, intervention to reduce the risk of rupture may be reasonable.9	
2b	C-LD	 In patients with unruptured AAA and aneurysm growth of ≥0.5 cm in 6 months, repair to reduce the risk of rupture may be reasonable.¹⁻⁵ 	

Recommendations for Open Versus Endovascular Repair of AAA Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	Α	 In patients with nonruptured AAA with low to moderate operative risk and who have anatomy suitable for either open or EVAR, a shared decision-making process weighing the risks and benefits of each approach is recom- mended.¹⁻¹¹
1	B-NR	 In patients undergoing elective endovascular repair for nonruptured AAA, adherence to manufacturer's instructions for use is recom- mended.¹²⁻¹⁶
2a	B-NR	3. In patients with nonruptured AAA and a high perioperative risk, EVAR is reasonable to reduce the risk of 30-day morbidity, mortality, or both. ^{9,10}
2a	B-NR	4. For patients with nonruptured AAA, a moderate to high perioperative risk, and anatomy suitable for an FDA-approved fenestrated endovascular device, endovascular repair is reasonable over open repair to reduce the risk of perioperative complications. 10,11,17,18



Repair of Ruptured AAA

Recommendations for Repair of Ruptured AAA Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-R	 In patients presenting with ruptured AAA who are hemodynamically stable, CT imag- ing is recommended to evaluate whether the AAA is amenable to endovascular repair.¹⁻³
1	B-R	2. In patients presenting with ruptured AAA who have suitable anatomy, endovascular repair is recommended over open repair to reduce the risk of morbidity and mortality. ^{1,4-6}
2a	B-NR	 In patients undergoing endovascular repair for ruptured AAA, local anesthesia is preferred to general anesthesia to reduce risk of periop- erative mortality.⁷⁻⁹
2a	C-LD	4. In patients with ruptured AAA, permissive hypotension can be beneficial to decrease the rate of bleeding. ^{1,3,10-12}





Treatment of Concomitant Common Iliac Aneurysms

Recommendations for the Treatment of Concomitant Common Iliac Aneurysms

Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	C-LD	 For patients with asymptomatic small AAA and concomitant common iliac artery aneurysm(s) ≥3.5 cm, elective repair of both abdominal and iliac aneurysms is recom- mended.¹⁻⁴
1	B-NR	2. When treating common iliac artery aneurysms or ectasia as part of AAA repair, preservation of at least 1 hypogastric artery is recommended, if anatomically feasible, to decrease the risk of pelvic ischemia. ^{5,6}





EVAR





Open repair of AAA



