

DESCENDING THORACIC AORTIC, THORACOABDOMINAL, AND ABDOMINAL AORTIC ANEURYSMS: ETIOLOGY AND INDICATIONS TO OPERATE

AORTIC ANEURYSMS AND DISSECTIONS: WHAT EVERY CLINICIAN SHOULD KNOW

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Disclosures

- No relevant disclosures

Circulation

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

Developed in collaboration with and endorsed by the American Association for Thoracic Surgery, American College of Radiology, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Thoracic Surgeons, and Society for Vascular Surgery

Endorsed by the Society of Interventional Radiology and Society for Vascular Medicine

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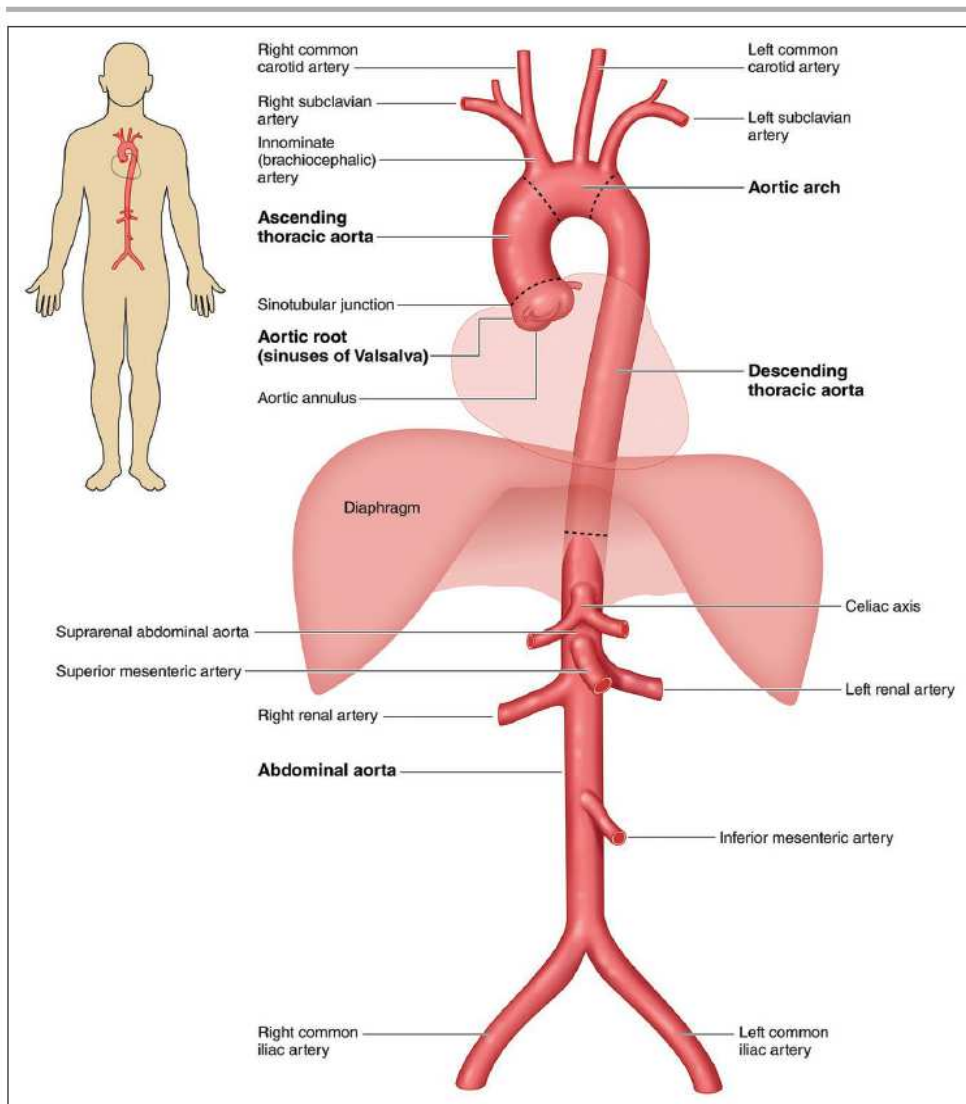
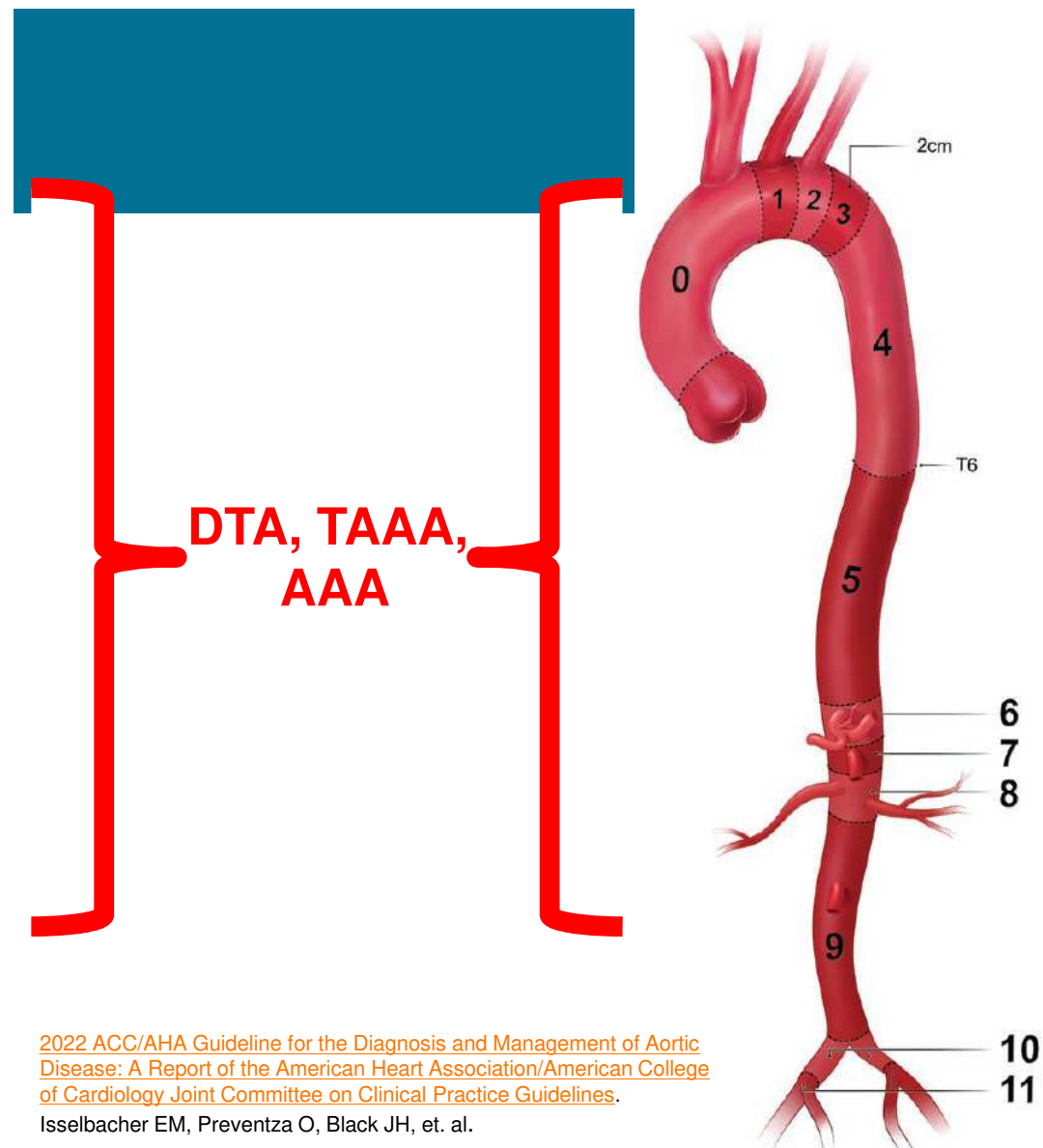
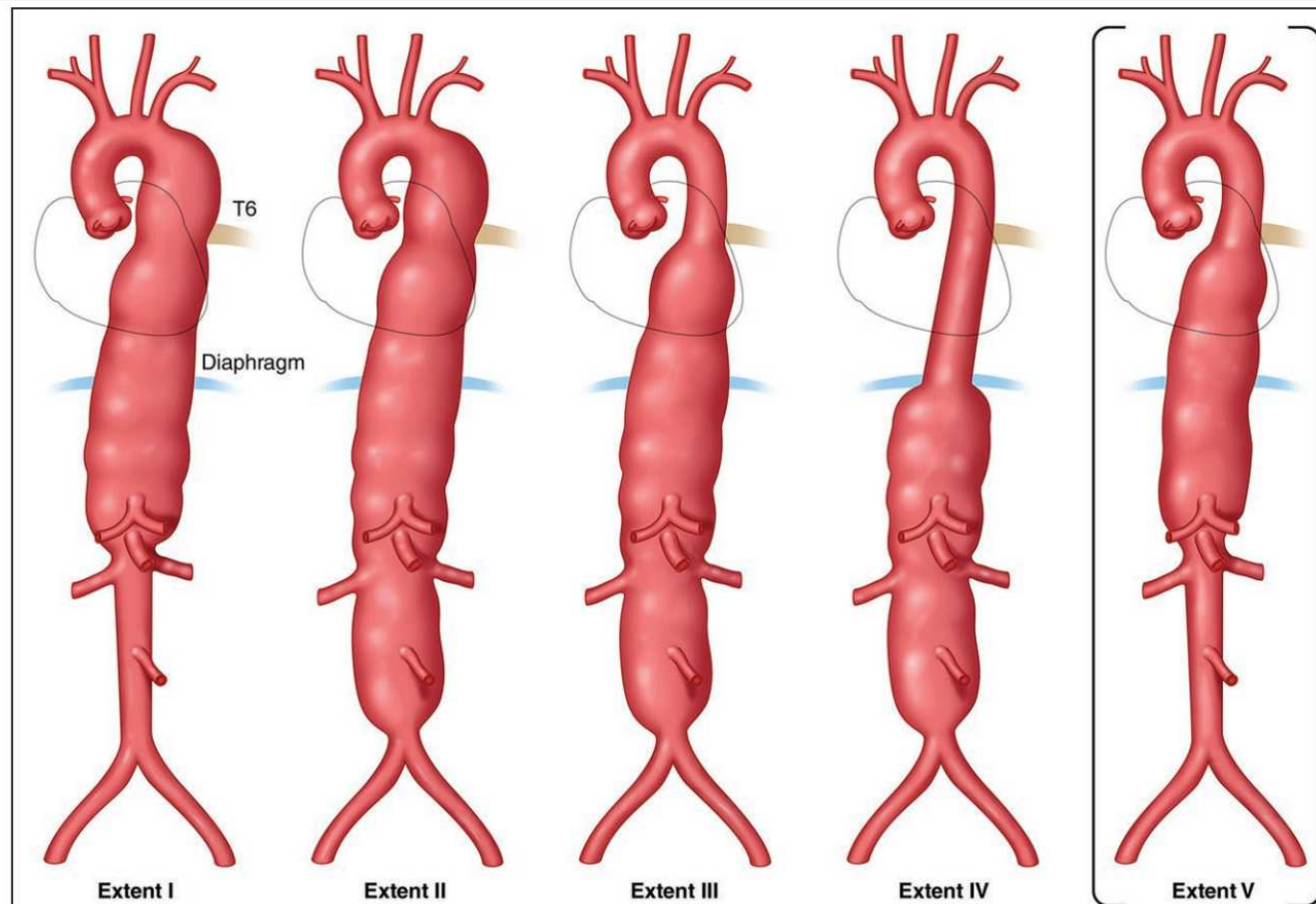
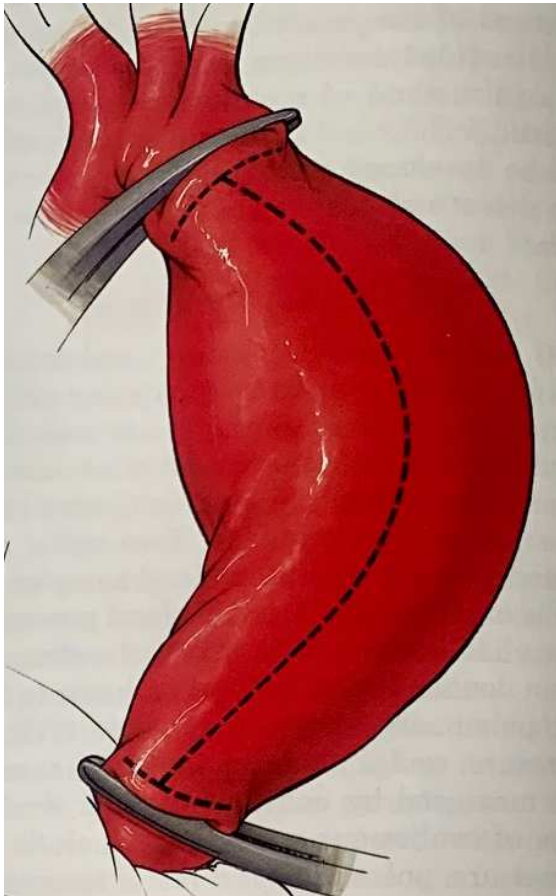


Figure 1. The Anatomy of the Aorta and Its Main Branches.



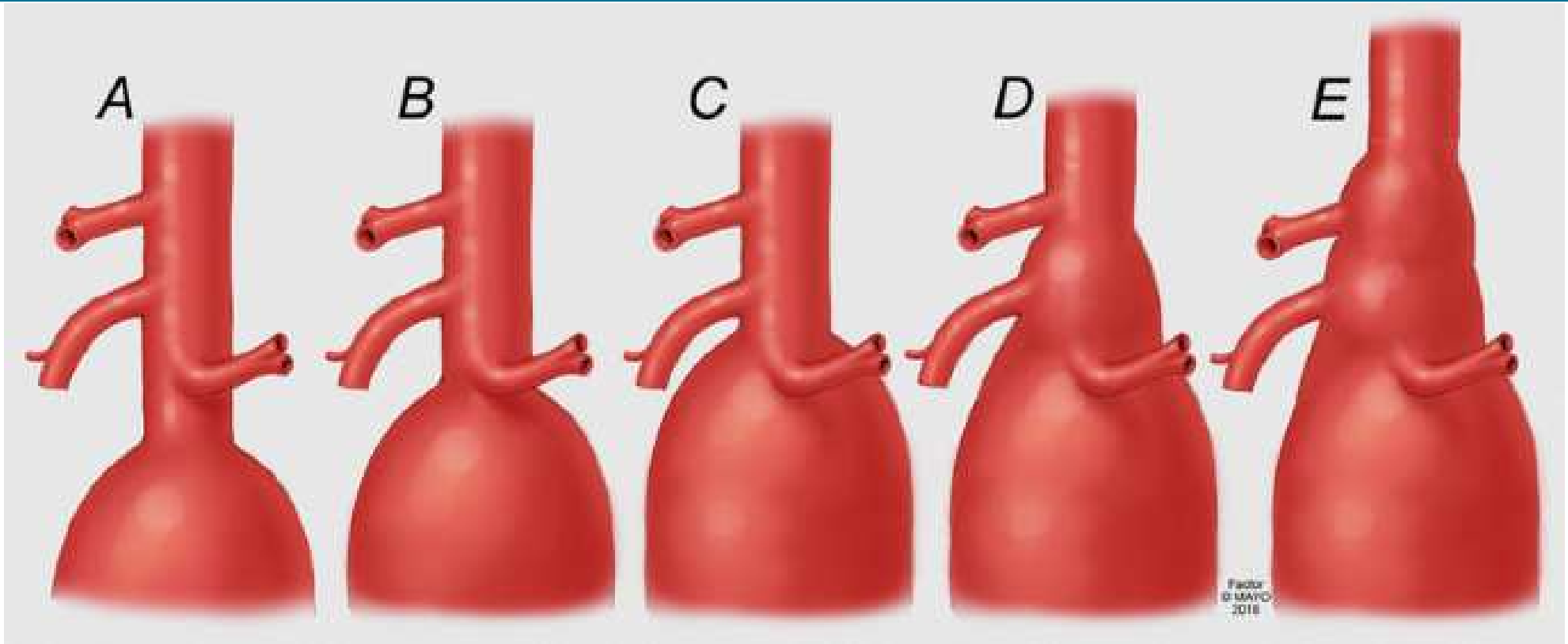
Definition and Classification Review



Infrarenal —————> Type IV TAAA



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Endovascular Aortic Repair, 2017, ISBN : 978-3-319-15191-5, Gustavo S. Oderich, Tiziano Tallarita



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Etiology and Pathogenesis

- Degenerative
 - “Standard” – HLD, HTN, smoking
 - Previous Dissection
 - Trauma
 - Connective Tissue Disorder/Genetic
- Aortitis
 - Infectious
 - Inflammatory

Degenerative

- Standard risk factors for atherosclerosis
 - Smoking
 - HTN
 - HLD
 - Negative association with diabetes
- Presence of other large artery aneurysms



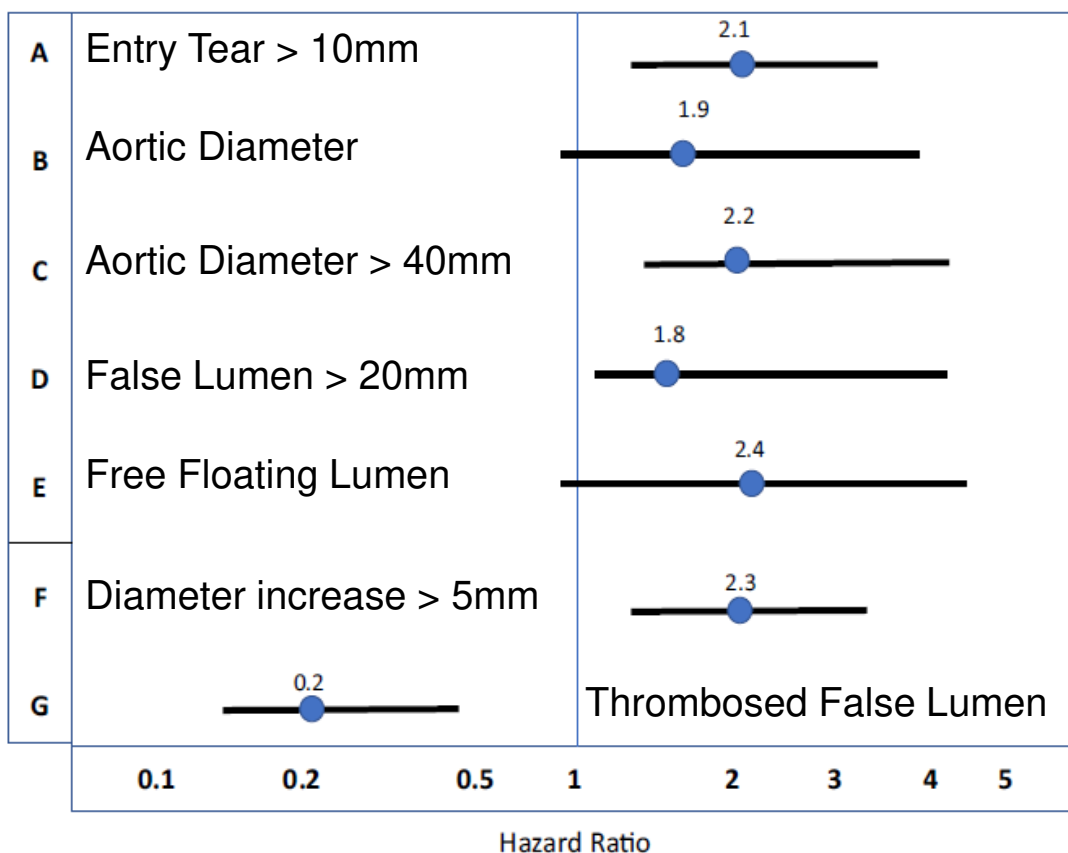
Predictors of late aortic intervention in patients with medically treated type B aortic dissection



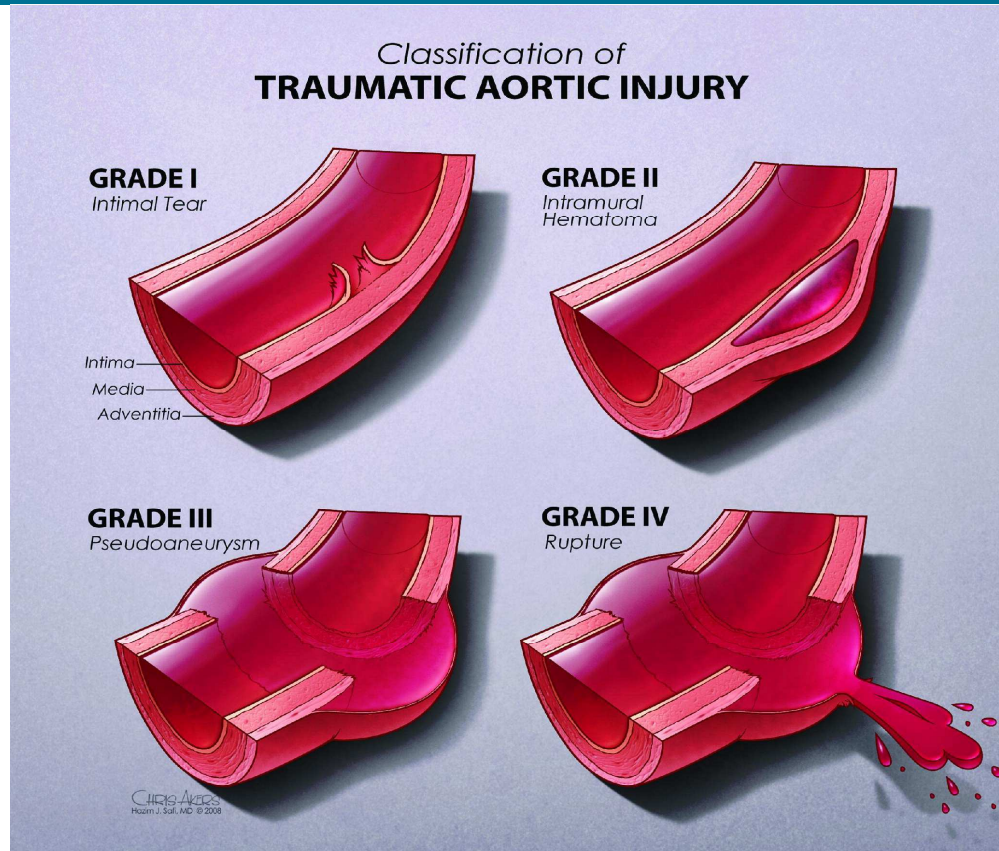
Samuel I. Schwartz, MD, Christopher Durham, MD, W. Darrin Clouse, MD, Virendra I. Patel, MD, MPH, R. Todd Lancaster, MD, MPH, Richard P. Cambria, MD, and Mark F. Conrad, MD, MMSc, *Boston, Mass*

Aortic anatomy (initial scan)			
Entry tear >10 mm	13	34	.02
Total aortic diameter, mm	31 ± 8.7	41 ± 9.4	.01
Total aortic diameter >0 mm	21	58	.01
False lumen diameter >20 mm	20	35	.02
Free-floating adventitia	2	4	.19
Aortic anatomy (serial scan)			
Increase in aortic diameter by >5 mm	33	67	.01
Completely thrombosed false lumen	37	8	.001

Categorical variables are presented as %. Continuous variables are presented as mean ± standard deviation.

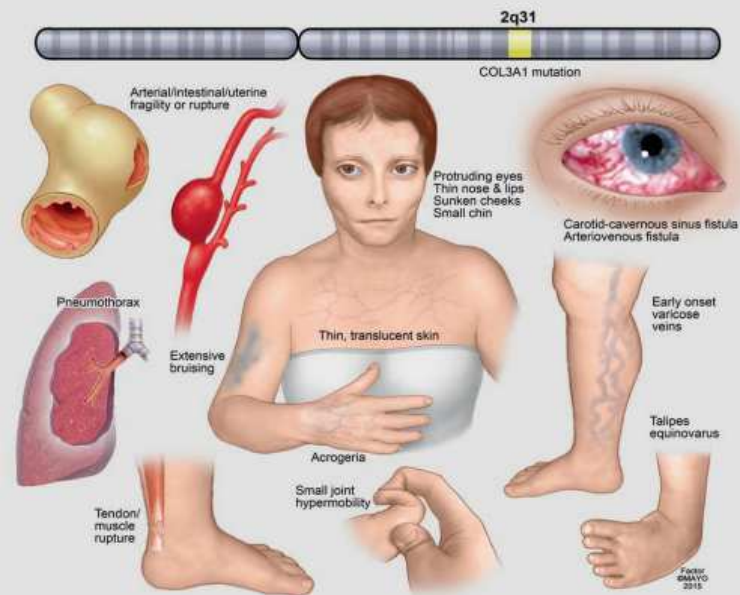


Trauma

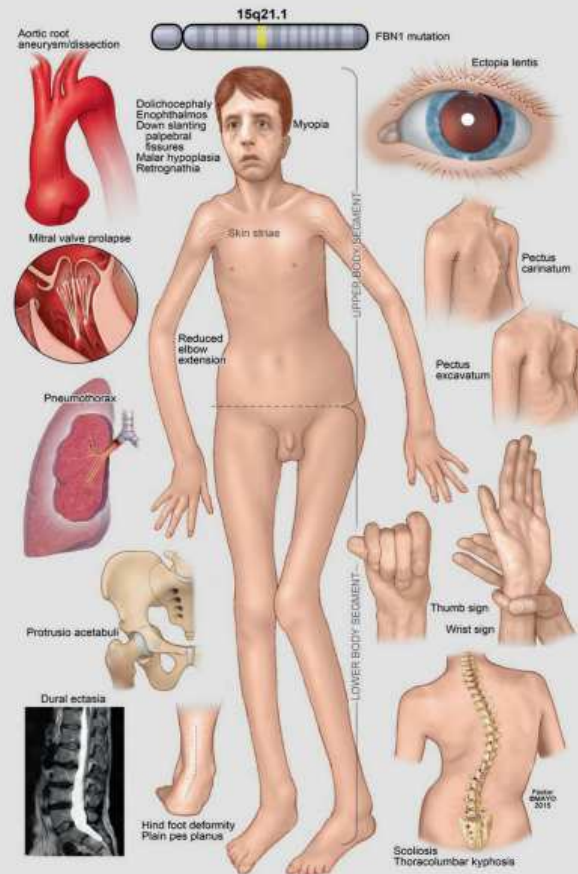


Connective Tissue Disorders

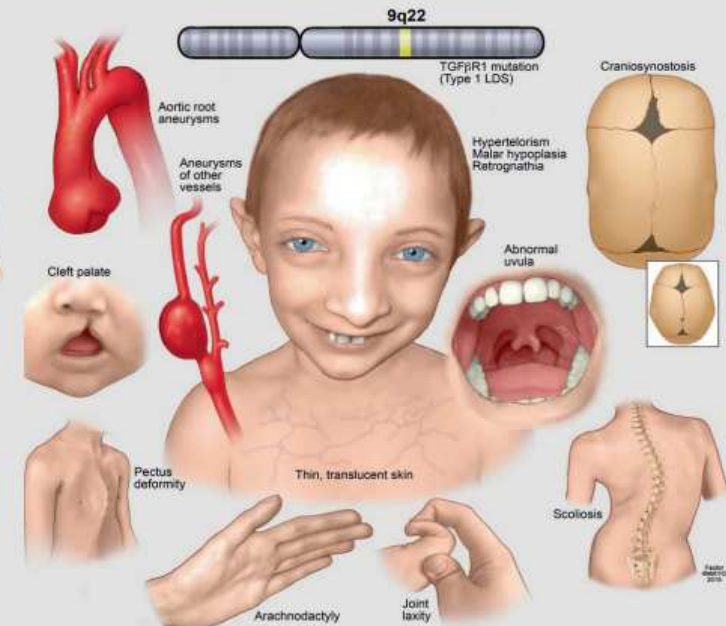
Vascular Ehlers-Danlos Syndrome



Marfan Syndrome



Loeys Dietz Syndrome



Turner Syndrome

Shalhoub, Genetic considerations in patients with aortic disease
Endovascular Aortic Repair, Oderich ed, 2017

Familial TAAD

- Family history
- Tall stature
- Stretch marks
- Joint hypermobility
- Sunken or protruding chest
- Hernia
- Scoliosis
- Livedo reticularis

Table 7. TAA Syndromes and Conditions Attributable to a Heritable or Genetic Cause

Condition	Gene	Clinical Features
Syndromic HTAD*		
Marfan syndrome	<i>FBN1</i>	Aortic root aneurysm, aortic dissection, TAA, MVP, long bone overgrowth, arachnodactyly, dolichostenomelia, scoliosis, pectus deformities, ectopia lentis, myopia, tall stature, pneumothorax, dural ectasia
Loeys-Dietz syndrome	<i>TGFBR1, TGFBR2, SMAD3, TGFBR3</i>	TAA, branch vessel aneurysms, aortic dissection, arterial tortuosity, MVP, craniosynostosis, hypertelorism, bluish sclera, bifid/broad uvula, translucent skin, visible veins, club feet, dural ectasia, and premature osteoarthritis and peripheral neuropathy†
Vascular Ehlers-Danlos syndrome	<i>COL3A1</i>	TAA, AAA, arterial rupture, aortic dissection, MVP, bowel and uterine rupture, pneumothorax, translucent skin, atrophic scars, small joint hypermobility, easy bruising, carotid-cavernous fistula
Arterial tortuosity syndrome	<i>SLC2A10</i>	Tortuous large and medium sized arteries, aortic dilation, craniofacial, skin and skeletal features
Shprintzen-Goldberg syndrome	<i>SKI</i>	Craniosynostosis, skeletal features, aortic dilation
Ehlers-Danlos syndrome with periventricular nodular heterotopia	<i>FLNA</i>	X-linked, periventricular nodular heterotopia, TAA, BAV, MV disease, PDA, VSD, seizures, joint hypermobility
Meester-Loeys syndrome	<i>BGN</i>	X-linked, TAA, aortic dissection, MV disease
LOX-related TAA	<i>LOX</i>	TAA, BAV, aortic dissection, Marfanoid habitus in some
Smooth muscle dysfunction syndrome	<i>ACTA2</i>	TAA, moyamoya-like cerebrovascular disease, pulmonary hypertension, pulmonary disease, hypoperistalsis, hypotonic bladder, congenital mydriasis ¹¹
Nonsyndromic HTAD (Familial TAA)		
FTAA	<i>ACTA2</i>	TAA, aortic dissection, premature CAD and moyamoya-like cerebrovascular disease, livedo reticularis, iris flocculi
FTAA	<i>MYH11</i>	TAA, aortic dissection, PDA
FTAA	<i>MYLK</i>	Aortic dissection at relatively small aortic size
FTAA	<i>PRKG1</i>	Aortic dissection at young ages at small aortic sizes
FTAA	<i>MAT2A</i>	TAA, aortic dissection, BAV
FTAA	<i>MFAP5</i>	TAA, aortic dissection, skeletal features may be present
FTAA	<i>FOXE3</i>	TAA, aortic dissection
FTAA	<i>THSD4</i>	TAA, aortic dissection

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(Continued)

Aortitis

Infectious

- Septic embolism
- Bacterial inoculation/seeding
- Contiguous infection
 - Fistula
 - Bronchogenic
 - Esophageal
 - Enteric

Inflammatory

- Arteritis
 - Giant cell
 - Takayasu
 - IgG4-mediated
- Ankylosing spondylitis
- SLE
- Behcet's

AAA (Infrarenal)

- Most common aortic site of true arterial aneurysms
- Male Sex
- Older age
- Smoking
- Positive family history (First-degree relative)



AAA – Why most common?

- Multi-factorial
 - Embryo-histologic → Decreased elastin and collagen
 - Hemodynamically susceptible
 - Standing reflected pulse wave
 - Turbulent flow → High and low shear stress
 - Alterations in growth factor and degradatory matrix metalloproteinase activity

Indications to operate

- “The purpose of surgical or endovascular intervention is to reduce the risk of adverse aortic events (ie, aortic dissection, rupture, and aortic-related death)...the optimal timing of intervention requires a careful anatomic assessment, followed by *weighing the risk of future adverse aortic events against the risk of intervention.*”

[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](#)

Isselbacher EM, Preventza O, Black JH, et. al.

Indications to Operate



- Based on static images
- Based on statistical averages across hundreds of thousands of patients
- Sometimes based on clinical/anecdotal experience

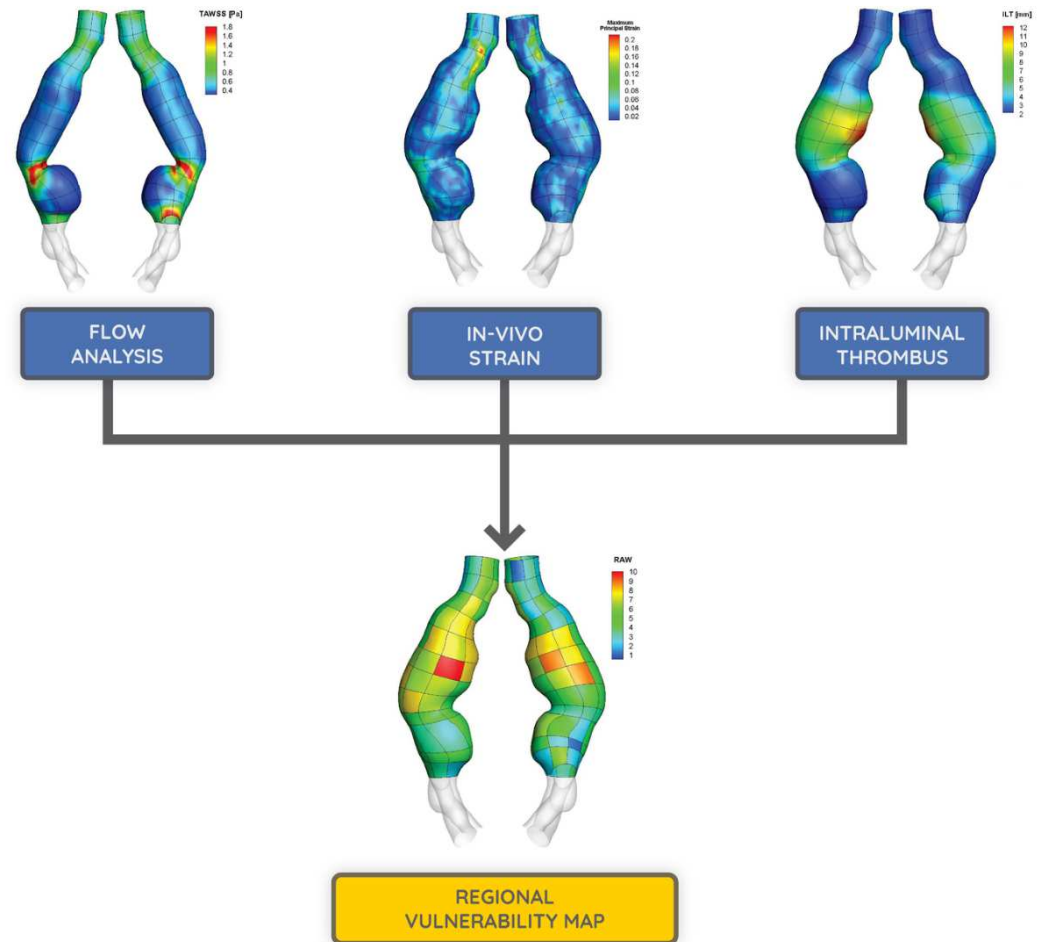


Table 2. Applying American College of Cardiology/American Heart Association Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care* (Updated May 2019)

CLASS (STRENGTH) OF RECOMMENDATION	LEVEL (QUALITY) OF EVIDENCE†
CLASS 1 (STRONG) Benefit >>> Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> Is recommended Is indicated/useful/effective/beneficial Should be performed/administered/other Comparative-Effectiveness Phrases‡: <ul style="list-style-type: none"> Treatment/strategy A is recommended/indicated in preference to treatment B Treatment A should be chosen over treatment B 	LEVEL A <ul style="list-style-type: none"> High-quality evidence‡ from more than 1 RCT Meta-analyses of high-quality RCTs One or more RCTs corroborated by high-quality registry studies
CLASS 2a (MODERATE) Benefit >> Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> Is reasonable Can be useful/effective/beneficial Comparative-Effectiveness Phrases‡: <ul style="list-style-type: none"> Treatment/strategy A is probably recommended/indicated in preference to treatment B It is reasonable to choose treatment A over treatment B 	LEVEL B-R (Randomized) <ul style="list-style-type: none"> Moderate-quality evidence‡ from 1 or more RCTs Meta-analyses of moderate-quality RCTs
CLASS 2b (WEAK) Benefit ≥ Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> May/might be reasonable May/might be considered Usefulness/effectiveness is unknown/unclear/uncertain or not well-established 	LEVEL B-NR (Nonrandomized) <ul style="list-style-type: none"> Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies Meta-analyses of such studies
CLASS 3: No Benefit (MODERATE) Benefit = Risk (Generally, LOE A or B use only) Suggested phrases for writing recommendations: <ul style="list-style-type: none"> Is not recommended Is not indicated/useful/effective/beneficial Should not be performed/administered/other 	LEVEL C-LD (Limited Data) <ul style="list-style-type: none"> Randomized or nonrandomized observational or registry studies with limitations of design or execution Meta-analyses of such studies Physiological or mechanistic studies in human subjects
Class 3: Harm (STRONG) Risk > Benefit Suggested phrases for writing recommendations: <ul style="list-style-type: none"> Potentially harmful Causes harm Associated with excess morbidity/mortality Should not be performed/administered/other 	LEVEL C-EO (Expert Opinion) <ul style="list-style-type: none"> Consensus of expert opinion based on clinical experience

COR and LOE are determined independently (any COR may be paired with any LOE).

A recommendation with LOE C does not imply that the recommendation is weak. Many important clinical questions addressed in guidelines do not lend themselves to clinical trials. Although RCTs are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

* The outcome or result of the intervention should be specified (an improved clinical outcome or increased diagnostic accuracy or incremental prognostic information).

† For comparative-effectiveness recommendations (COR 1 and 2a; LOE A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

‡ The method of assessing quality is evolving, including the application of standardized, widely-used, and preferably validated evidence grading tools; and for systematic reviews, the incorporation of an Evidence Review Committee.

COR indicates Class of Recommendation; EO, expert opinion; LD, limited data; LOE, Level of Evidence; NR, nonrandomized; R, randomized; and RCT, randomized controlled trial.

6.5.3. Descending TAA

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	1. In patients with intact descending TAA, repair is recommended when the diameter is ≥ 5.5 cm. ^{1,2}
2b	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
2b	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

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

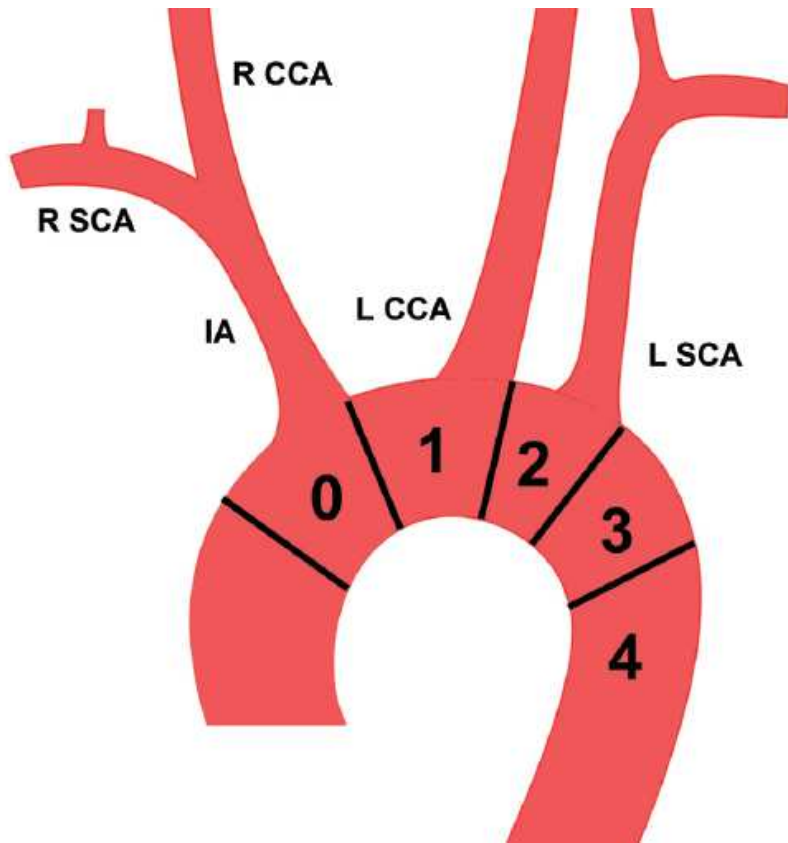
High-Risk Features for Rupture
Aneurysm growth of ≥ 0.5 cm/y ³
Symptomatic aneurysm ⁴
Marfan, Loeys-Dietz, or vascular Ehlers-Danlos syndrome, or HTAD (see Section 6.1.2, "Genetic Aortopathies") ²
Saccular aneurysm ⁵
Female sex ²
Infectious aneurysm ⁶

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

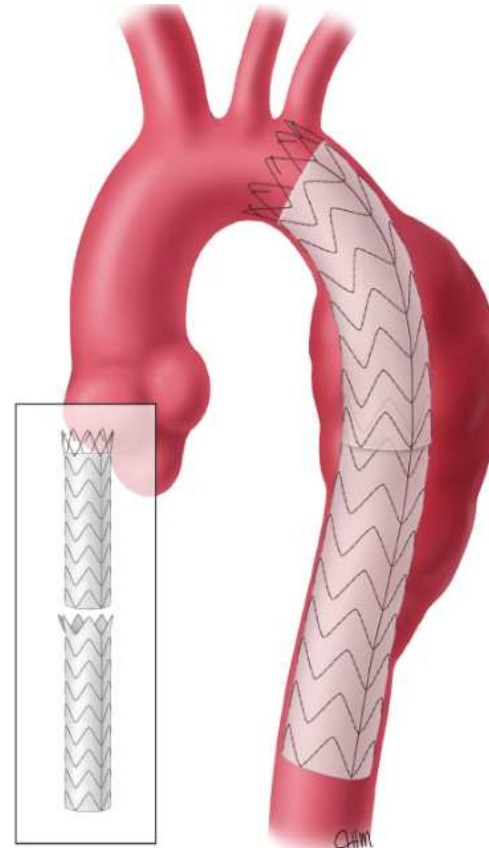
Table 18. Patient Characteristics Associated With Increased Perioperative Morbidity and Mortality After Open and Endovascular Repair of Descending TAA

Open Surgical Repair	Endovascular Repair
Advanced age ⁸	Functional dependence
65–74 y (OR, 1.8; 95% CI, 1.4–2.4; $P < 0.001$)	
≥ 75 y (OR, 2.6; 95% CI, 2.0–3.5; $P < 0.001$)	
Preoperative renal insufficiency (stage 3 or greater CKD) or hemodialysis	Thoracoabdominal aortic aneurysm extent
COPD and FEV1 $\leq 50\%$ predicted	Pulmonary disease
Previous stroke ⁹	Need for iliac access
	Zone 1/2 landing for thoracic stent graft ⁷

TEVAR (Thoracic Aortic Endovascular Repair)



<https://evtoday.com/articles/2016-nov/expanding-the-landing-zone-for-tevar>

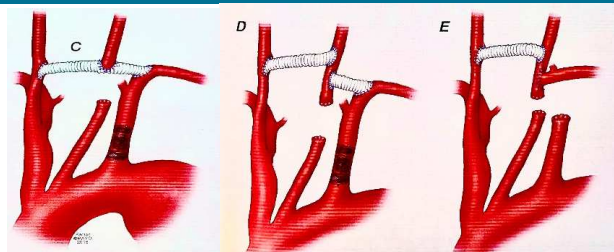


<http://dqsendovascular.blogspot.kr/2012/10/tevar-devices-summary.html>

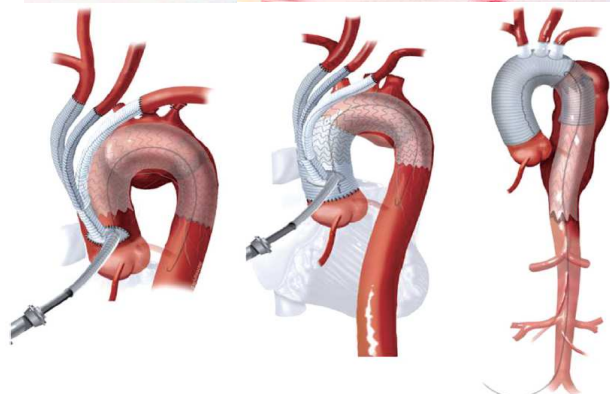
Largest Seal
Zone Intended
Aortic Diameter:
42mm for most
companies

TEVAR Landing Zone Adjuncts = Greater Morbidity/Risk

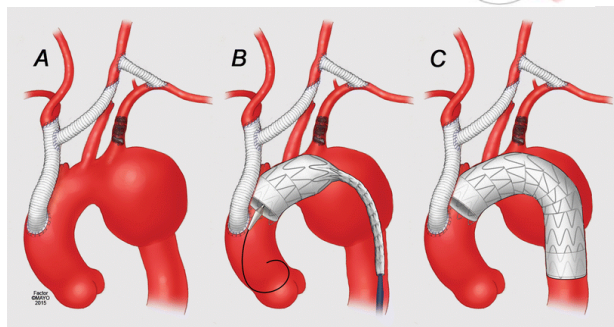
- Transcervical
- Transthoracic
- Hybrid



Rana, M.A., Oderich, G.S., Pochettino, A. (2017). Techniques and Results of Aortic Arch Hybrid Repair. In: Oderich, G. (eds) Endovascular Aortic Repair. Springer, Cham. https://doi.org/10.1007/978-3-319-15192-2_36



Vallabhajosyula P, Szeto WY, Desai N, Komlo C, Bavaria JE. Type II arch hybrid debranching procedure. Ann Cardiothorac Surg. 2013 May;2(3):378-8. doi: 10.3978/j.issn.2225-319X.2013.05.08. PMID: 23977611; PMCID: PMC3741848.



Rana, M.A., Oderich, G.S., Pochettino, A. (2017). Techniques and Results of Aortic Arch Hybrid Repair. In: Oderich, G. (eds) Endovascular Aortic Repair. Springer, Cham. https://doi.org/10.1007/978-3-319-15192-2_36

TAAA

6.5.4. Thoracoabdominal Aortic Aneurysms

6.5.4.1. Size Thresholds for Open Surgical Repair of TAAA

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	1. In patients with intact degenerative TAAA, repair is recommended when the diameter is ≥ 6.0 cm. ¹⁻³
2a	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
2a	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

9.3-19% (2-4 times median operative mortality)

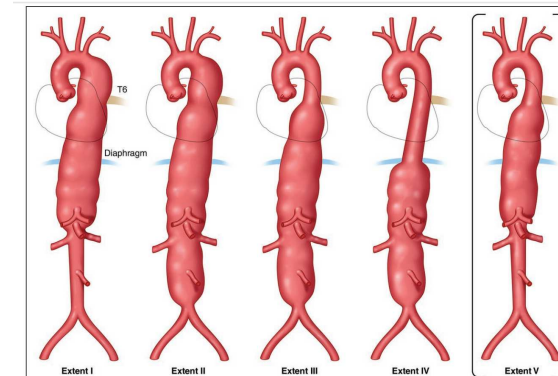


Table 19. Features Associated With an Increased Risk of TAAA Rupture

Rapid growth (confirmed increase in diameter of ≥ 0.5 cm/y)
Symptomatic aneurysm
Significant change in aneurysm appearance
Saccular aneurysm or presence of penetrating atherosclerotic ulcers

TAAA indicates thoracoabdominal aortic aneurysm.

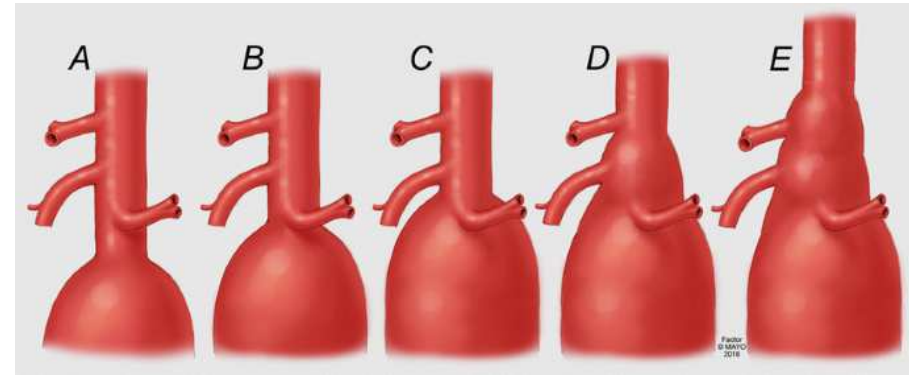


6.5.5.3. Threshold for AAA Repair

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	1. 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
	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	4. 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



Thank you

