2023 ACC India

Aortic Aneurysm Disease

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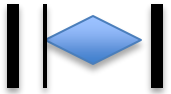
Disclosures

- None

Aortic Aneurysm Disease

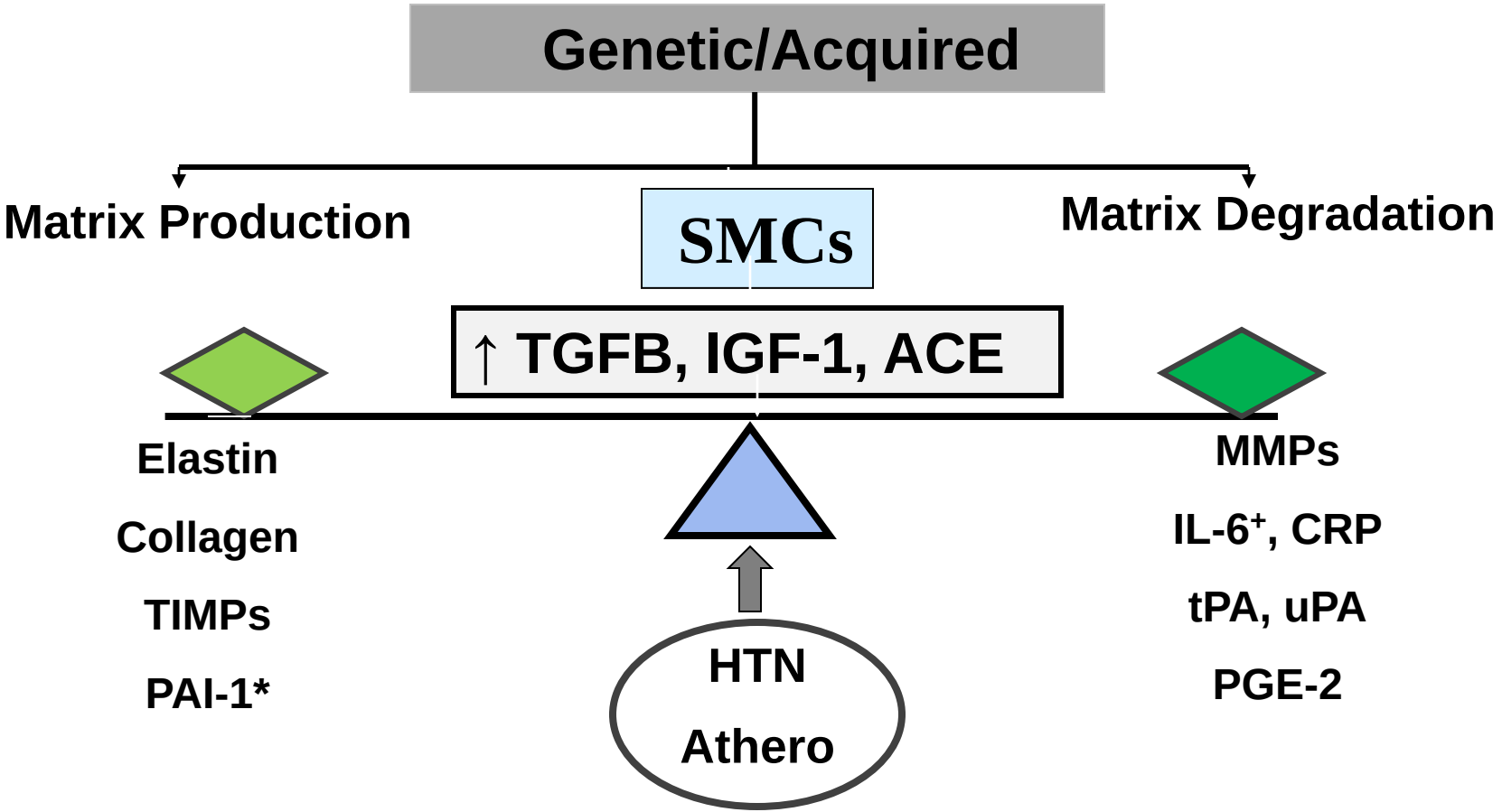
- **47-year-old man**
  - **Heart murmur noted 12/2021**
  - **TTE 3 months ago: BAV, mod AS/AR, normal LV, Ao 4.2cm sinus, 4.8cm ascending, no coarctation**
  - **Asymptomatic**
  - **BP 154/98, HR 90**
  - **BMI 30**



S1 **S2**

Aortic Aneurysm Disease

Aortic Aneurysm Disease



Thoracic Aortic Aneurysm Disease

- **Inherited CT disorders**
  - **MFS, EDS, Loey-Dietz, BAV, Turner's**
- **Atherosclerosis**
- **Inflammatory diseases**
- **Infection**
- **Trauma**
- **Chronic dissection**

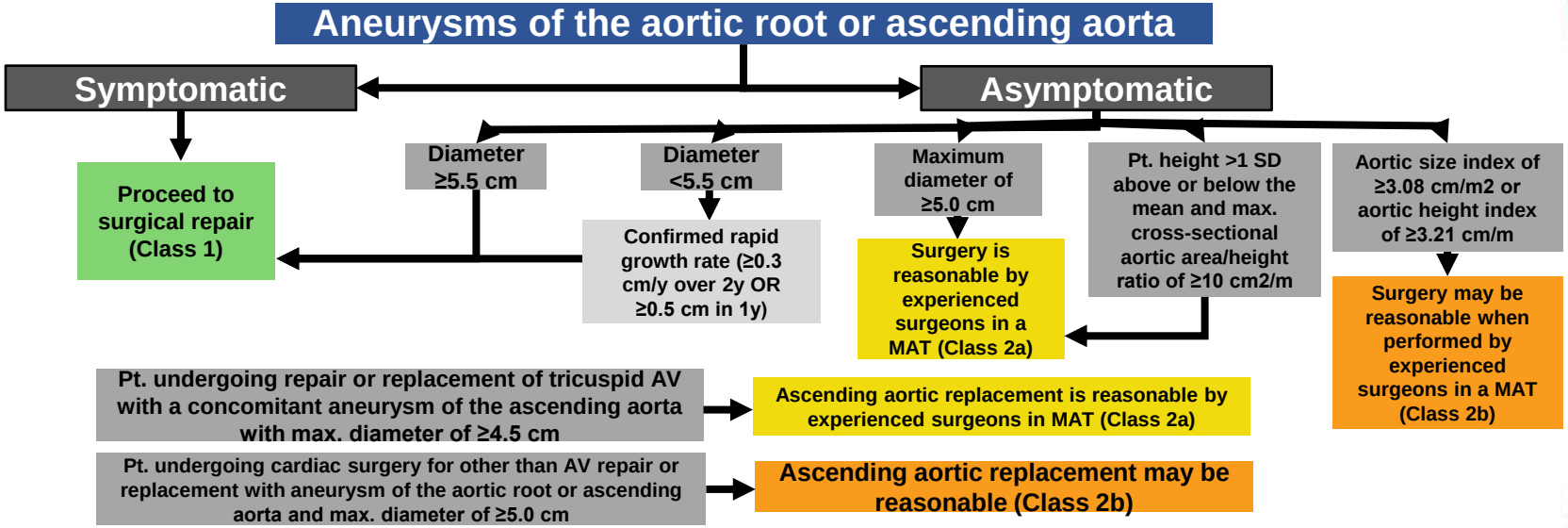
Genetic Disorders

Thoracic Aortic Disease

Genetic Syndrome	Common Clinical Features	Genetic Defect	Diagnostic Test
Marfan Syndrome	Skeletal features Ectopia lentis	*FBN1* mutations*	Rev Ghent Criteria DNA for sequencing
Loeys-Dietz Syndrome	Bifid uvula or cleft palate Arterial tortuosity Hypertelorism	*TGFBR2* or *TGFBR1* mutations	DNA for sequencing
Ehlers-Danlos Syndrome	Thin, translucent skin, GI rupture Rupture of visceral organs Rupture of medium to large arteries	*COL3A1* mutations	DNA for sequencing Dermal fibroblasts for analysis of type 3 collagen
Turner Syndrome	Short stature Primary amenorhea BAV disease Aortic coarctation	45 XO karyotype	Cells for karyotype analysis

*The defective gene at a second locus for MFS is TGFBR2 but the clinical phenotype as MFS is debated.

Recommendations for Surgery for Sporadic Aneurysms of the Aortic Root and Ascending Aorta

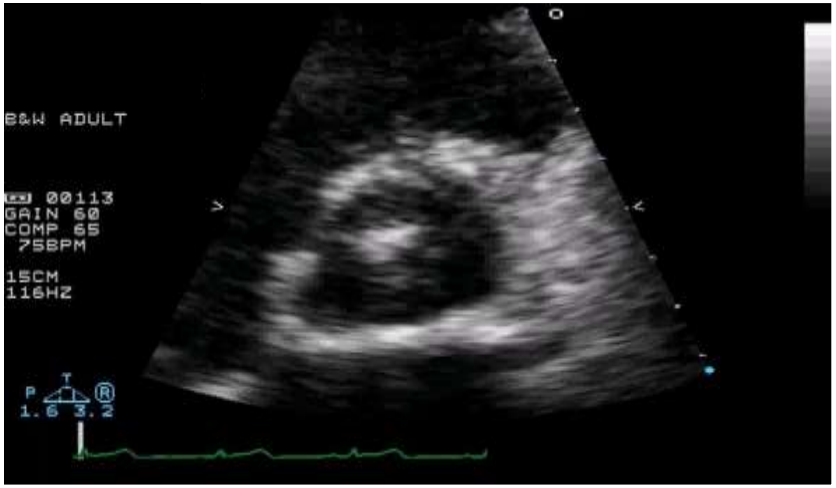


Abbreviations: AV indicates aortic valve; cm, centimeter; CT, computed tomography; y, year; MAT, multidisciplinary aortic team; max, maximal; pt, patient; SD, standard deviation; and y, year.

American Heart Association.

Isselbacher, E. M., et al. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease. *Circulation.*

BAV Disease


B&W ADULT
00113
GAIN 60
COMP 65
75BPM
15CM
116HZ
1.6 3.2


FR 39Hz
9.8cm
ZD
58%
C 50
P Med
HPen
M4
1.4 2.8
88 bpm

BAV Disease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58%
C 50
P Med
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M4
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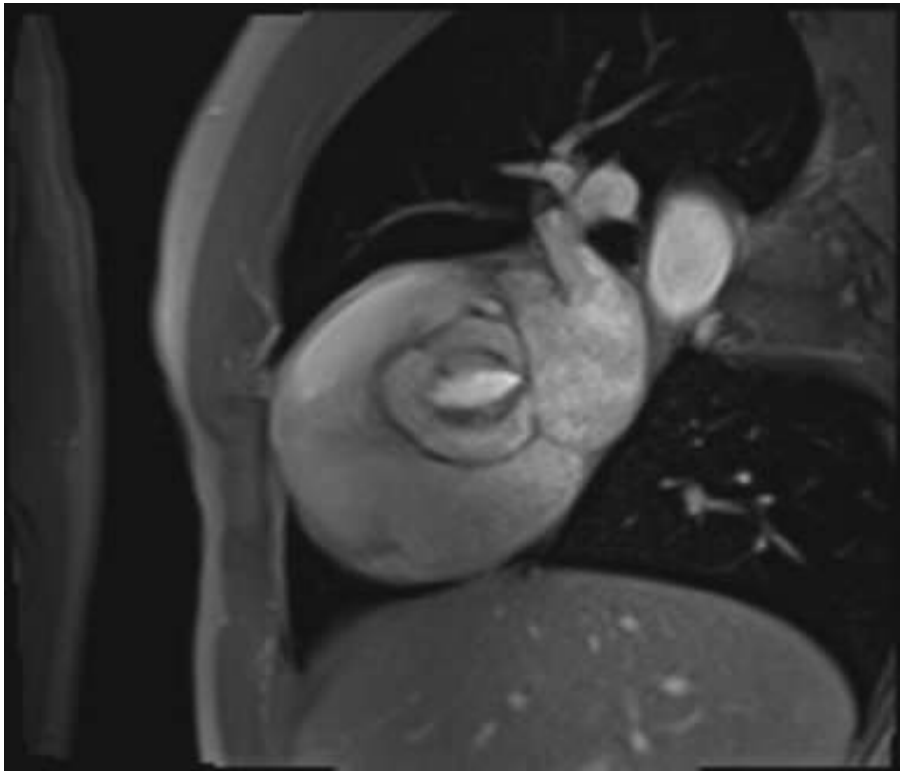
87 bpm

BAV Aortopathy

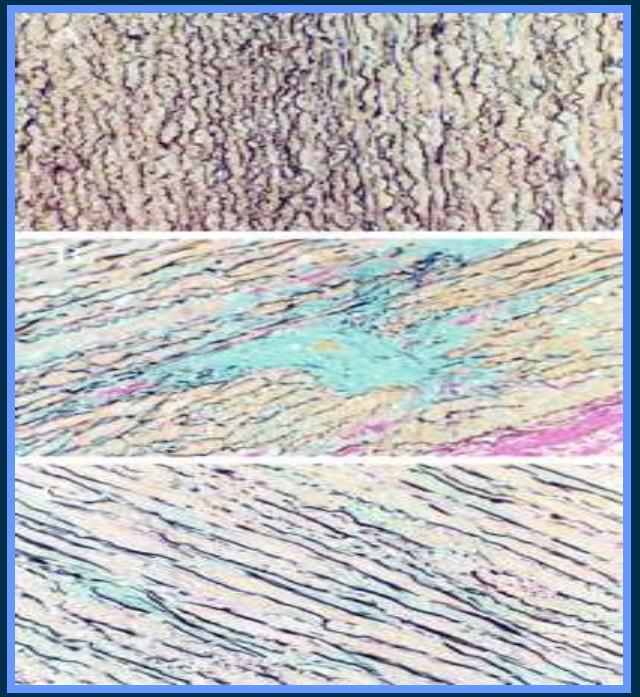


Bicuspid Aortic Valve Disease

Medial Degeneration

VSMC Apoptosis

Role of MMP-2



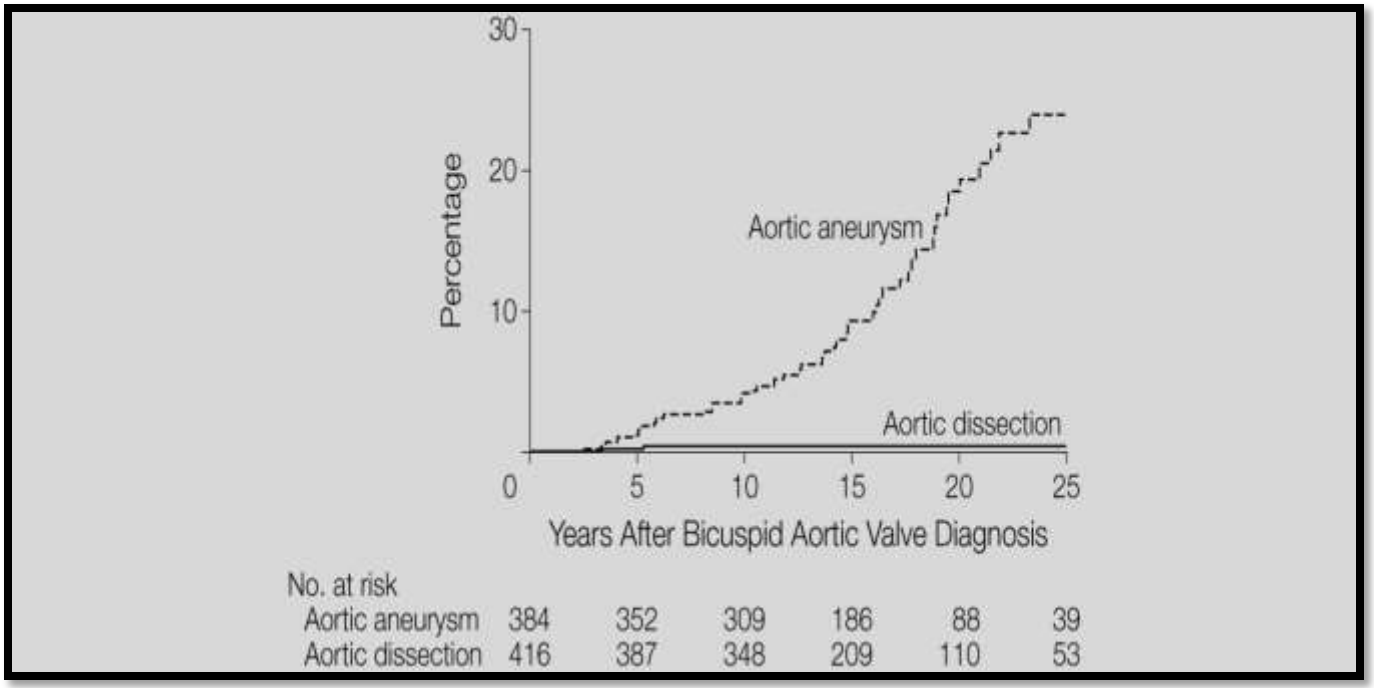
Normal

Marfan

Nataatmadja M et al. *Circulation* 2003; 108:II 329.

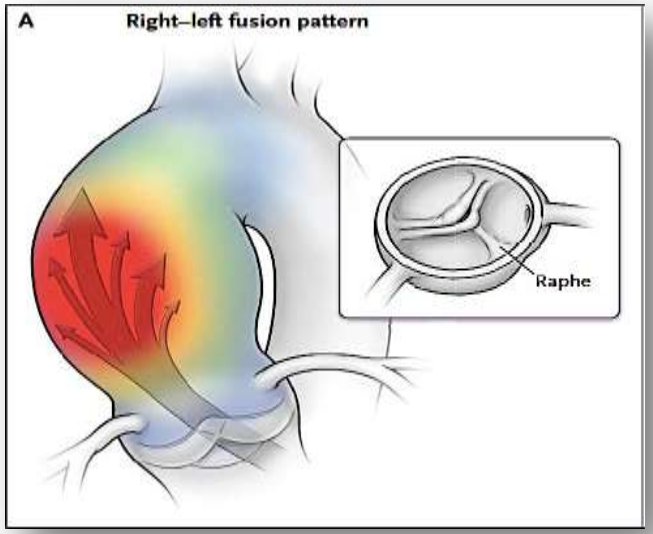
BAV

Natural History of Aortic Disease in Patients with BAV



JAMA. 2011;306(10):1104-1112. doi:10.1001/jama.2011.1286

BAV Aortopathy



Size Criteria for Surgery

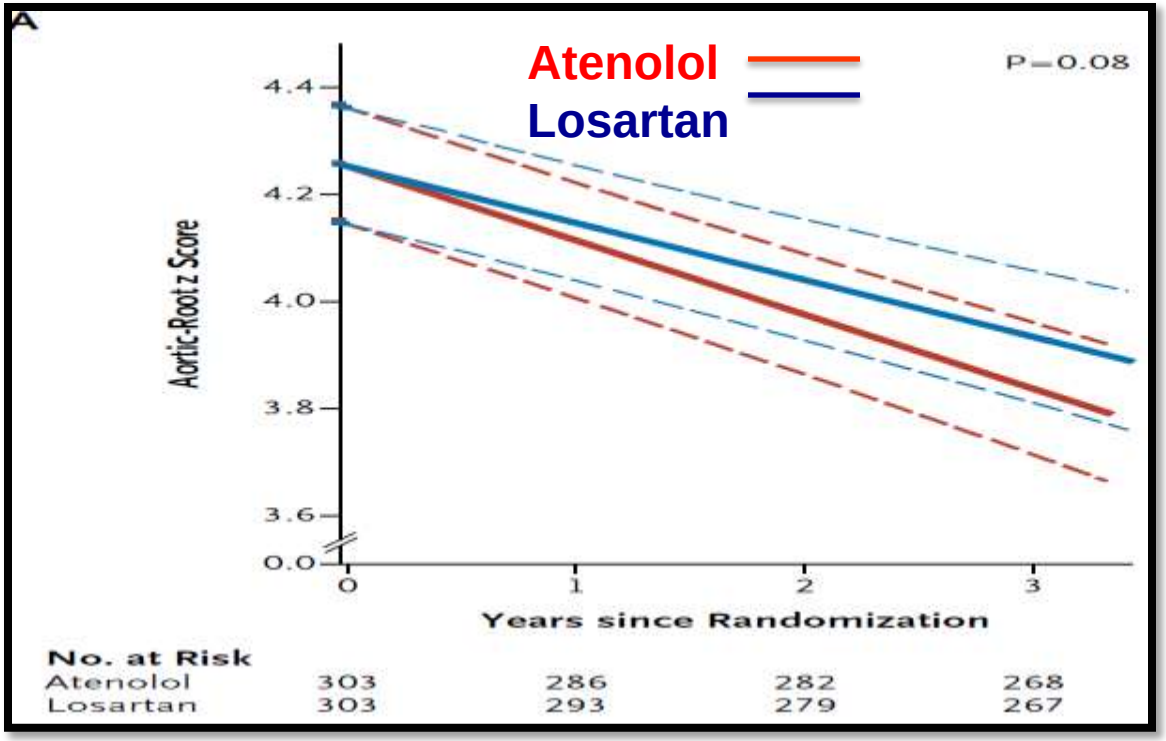
1. ≥5.5 cm (Class I)
2. Cross sectional root or ascending aortic area to height ratio >10cm^2/m (IIa)
3. 5.0-5.4 cm with RFs for dissection (IIa)
4. ≥4.5 cm if AVR indicated (IIa)
5. 5.0-5.4 cm at Center with MAT (IIb)

Verma S and Siu SC. NEJM 2014; 370:1920-9.

Medical Therapy in Marfan Syndrome

COR	LOE	Recommendations
1	A	1. In patients with Marfan syndrome, treatment with either a beta blocker or an ARB, in maximally tolerated doses (unless contraindicated), is recommended to reduce the rate of aortic dilation.
2a	C-LD	2. In patients with Marfan syndrome, the use of both a beta blocker and an ARB, in maximally tolerated doses (unless contraindicated), is reasonable to reduce the rate of aortic dilation.

MFS: ARB vs Atenolol



Lacro RV et al. NEJM 2014; 371:2061-71

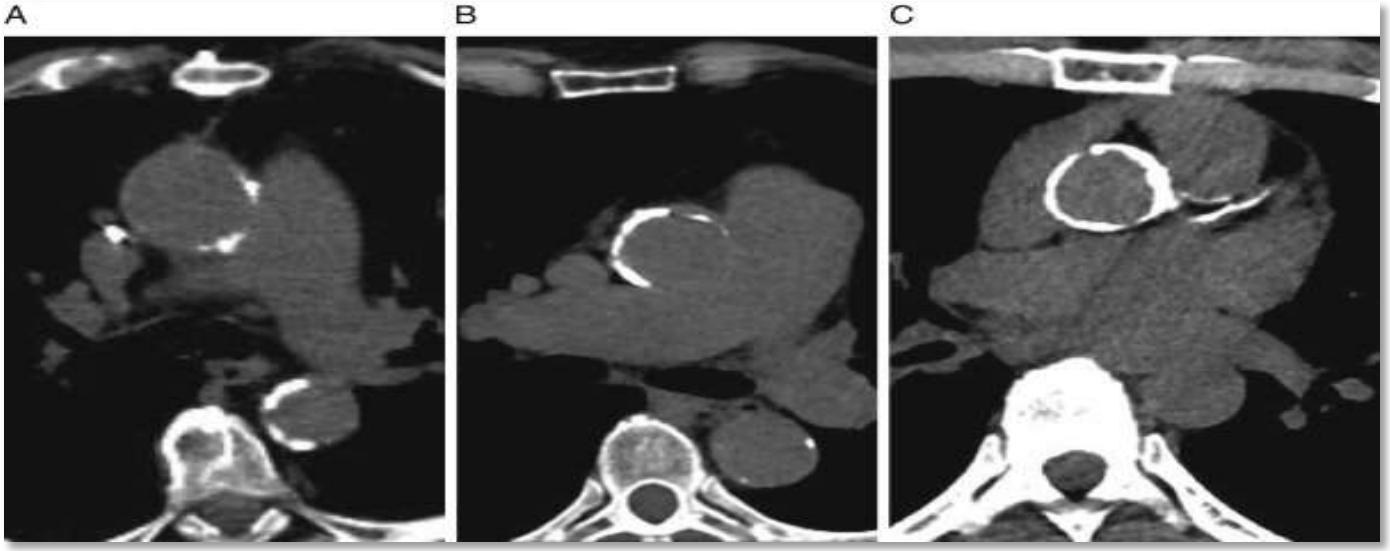
Recommendations for Replacement of the Aortic Root in Patients With Marfan Syndrome

COR	LOE	Recommendations
1	B-NR	1. In patients with Marfan syndrome and an aortic root diameter of ≥5.0 cm, surgery to replace the aortic root and ascending aorta is recommended.
2a	B-NR	2. In patients with Marfan syndrome, an aortic root diameter of ≥4.5 cm, and features associated with an increased risk of aortic dissection, surgery to replace the aortic root and ascending aorta is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.

Replacement of the Aortic Root in Patients With Marfan Syndrome (2)

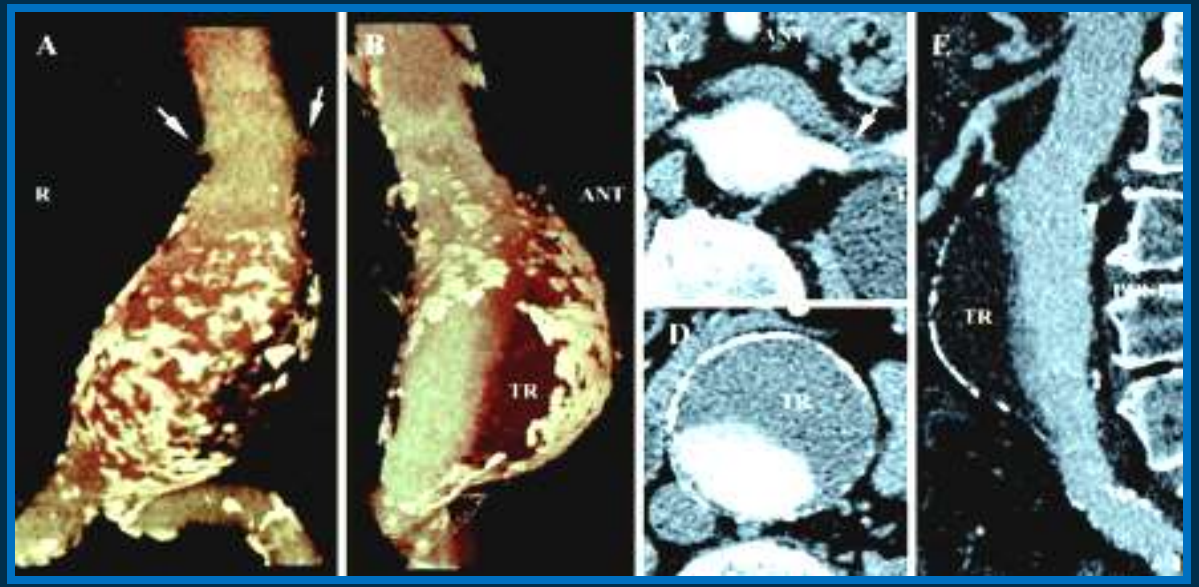
2a	**C-LD**	**3. In patients with Marfan syndrome and a maximal *cross-sectional aortic root area (cm^2) to patient height (m) ratio of ≥ 10*, surgery to replace the aortic root and ascending aorta is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.**
2b	**C-LD**	**4. In patients with Marfan syndrome and an aortic diameter approaching surgical threshold, who are candidates for valve-sparing root replacement (VSRR) and have a very low surgical risk, surgery to replace the aortic root and ascending aorta may be reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.**

Thoracic Aortic Atherosclerosis



Desai M et al. JACC CV Img 2018; 11:1012-26

Abdominal Aortic Aneurysm



Sianos G et al. *Circulation* 2001; 104: e10.

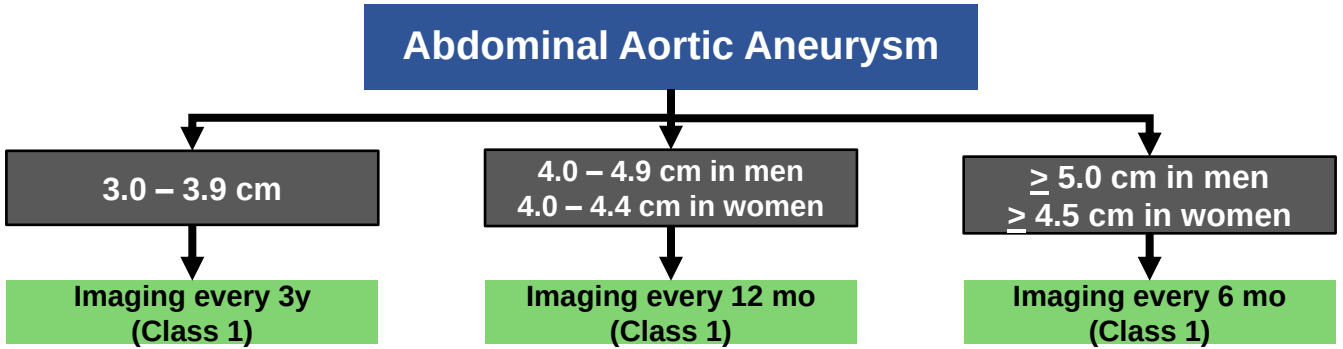
Background

Major Risk Factors for AAA Development#

- **Age > 65 yrs**
- **Male gender***
- **Smoking**
- **Affected first degree family relative**
- **Atherosclerotic risk factors, ASCVD**

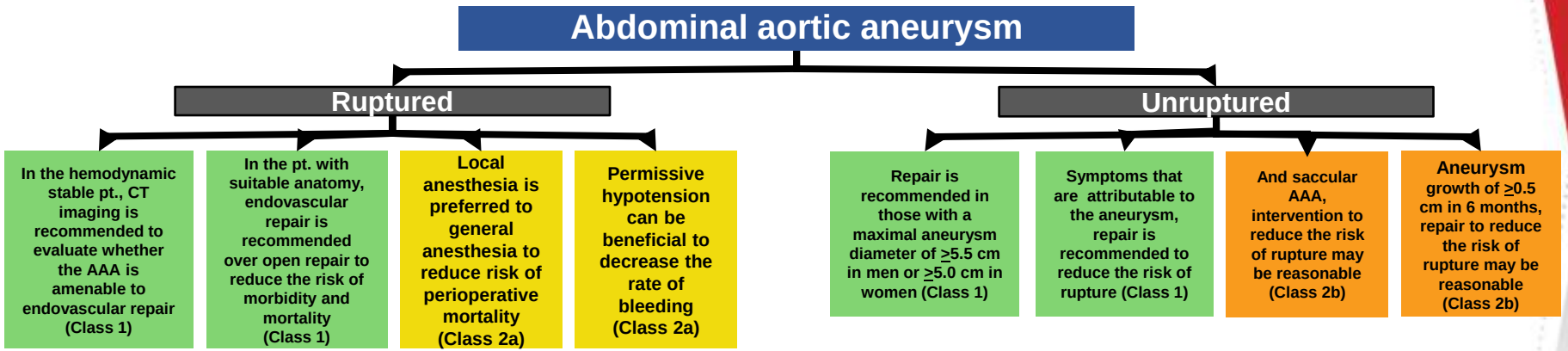
 # AAA less common in African Americans and patients with diabetes
*** Women have higher tendency to rupture at smaller Ao diameters**

Frequency of Surveillance Imaging of Abdominal Aortic Aneurysms Based on Current Aortic Diameter



Abbreviations: cm indicates centimeter; mo, month; and y, year.

Guidance for Repair of Abdominal Aortic Aneurysms

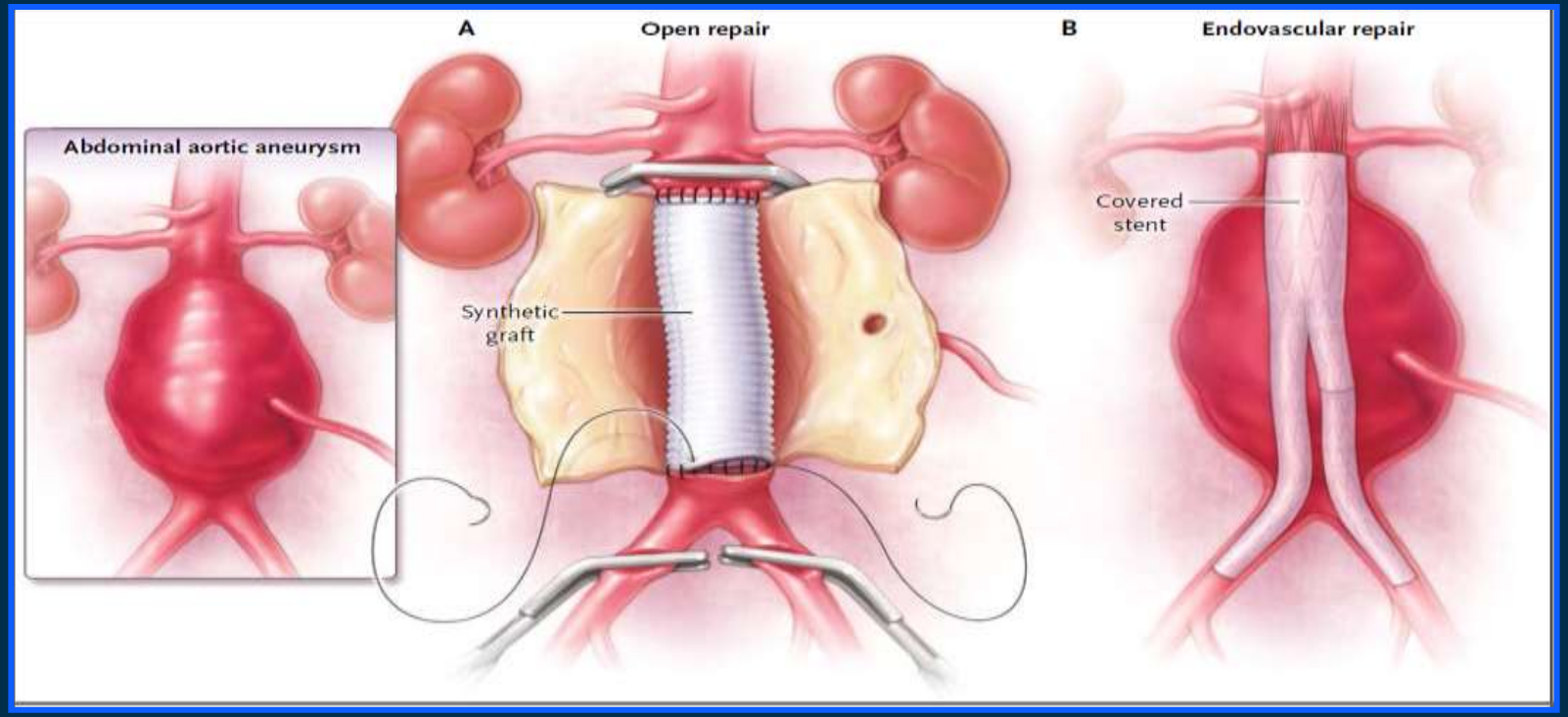


Abbreviations: AAA indicates abdominal aortic aneurysm; cm, centimeter; CT, computed tomography; and pt., patient.

American Heart Association.

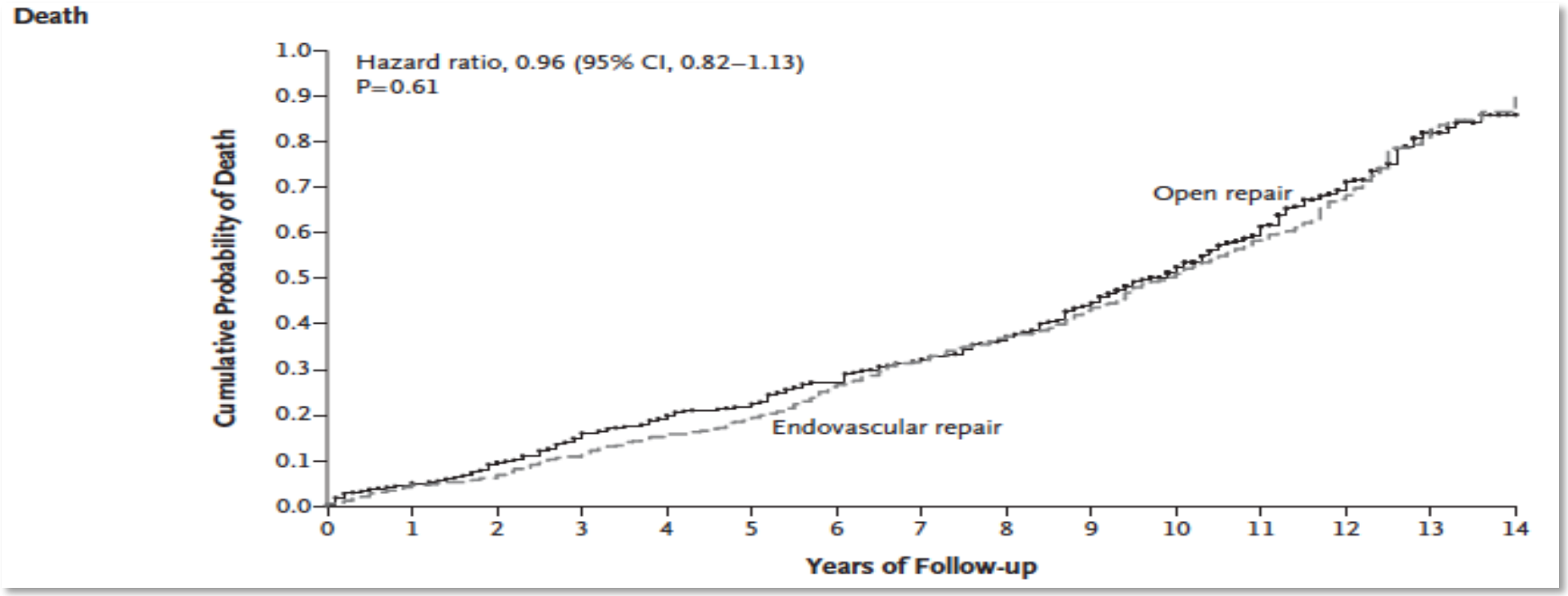
Isselbacher, E. M., et al. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease. *Circulation.*

AAA Repair



Kent KC. NEJM 2014; 371: 2101-8

Open vs Endovascular Repair AAA



Lederle FA et al. NEJM 2019;380:2126

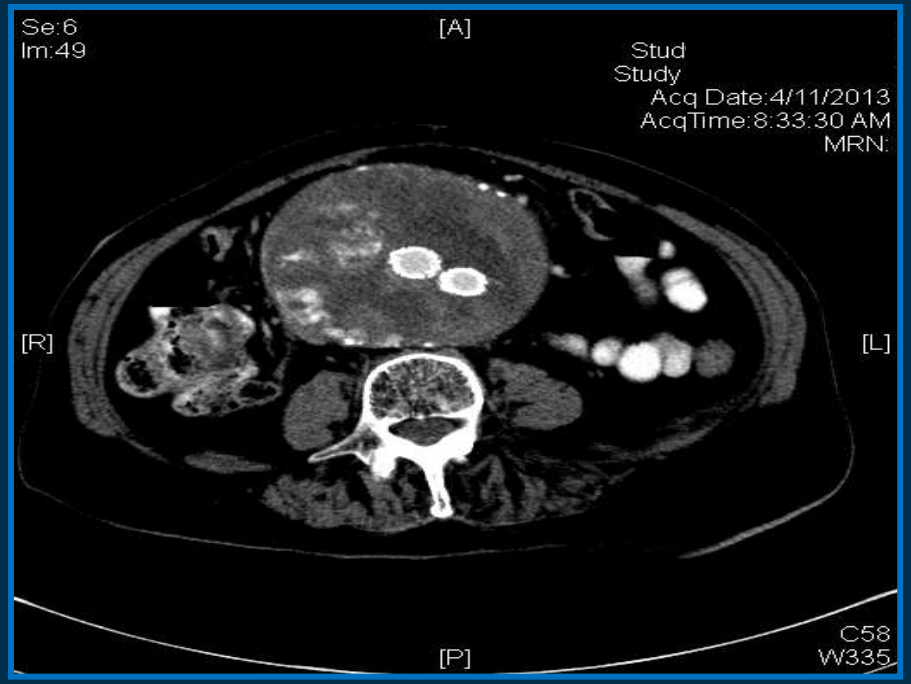
Open Versus Endovascular Repair of AAA

Recommendations for Open Versus Endovascular Repair of AAA		
COR	**LOE**	**Recommendations**
1	A	1. 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 recommended.
1	B-NR	2. In patients undergoing elective endovascular repair for nonruptured AAA, adherence to manufacturer's instructions for use is recommended.

Open Versus Endovascular Repair of AAA (con't.)

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.
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.

Endoleak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MRN:
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C58
W335


Shift Overlay from 60ox to 7FE0
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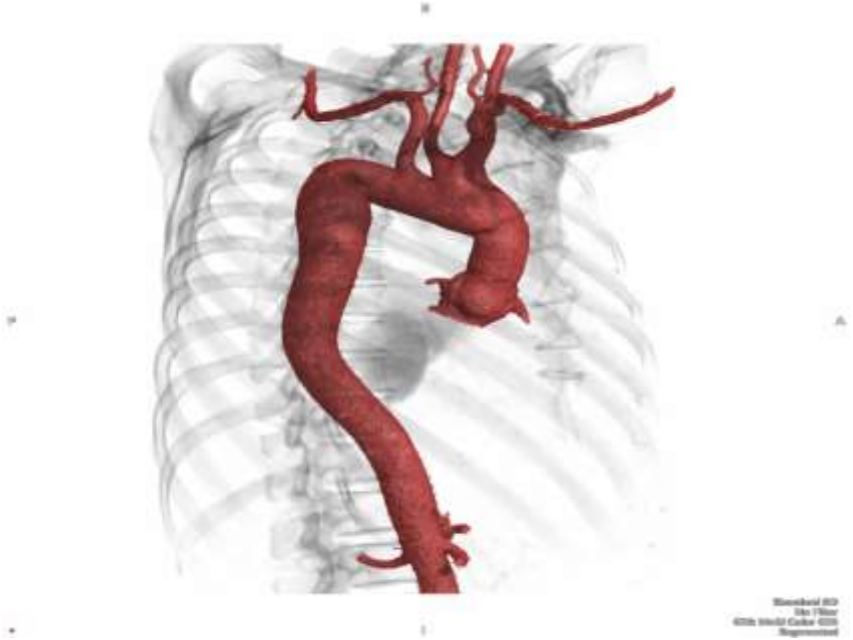
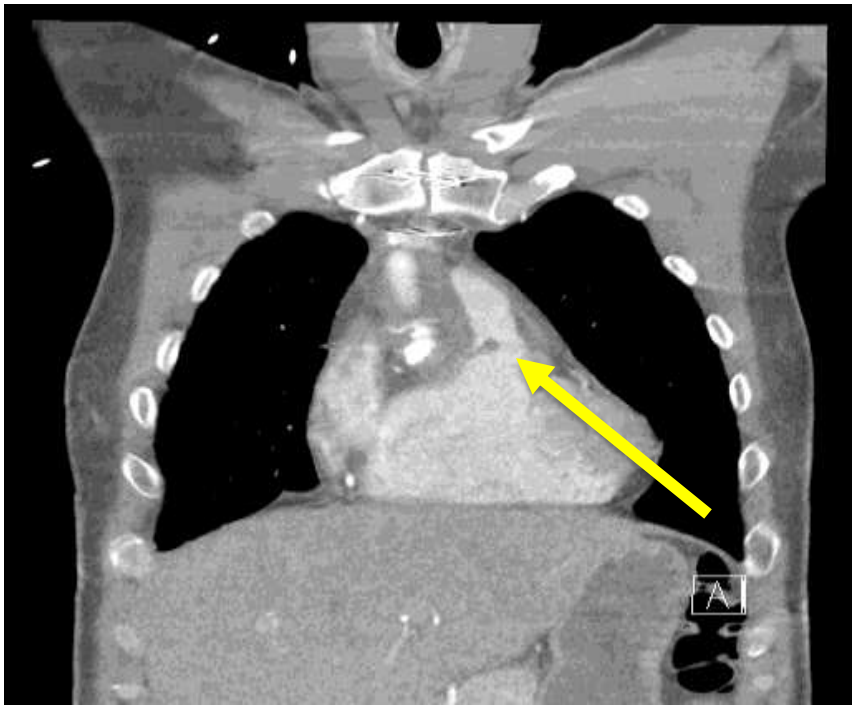
C40
W295

Aortic Aneurysm Disease

04/2022: Ross procedure with root and ascending aortic replacement. Post-op PE and atrial flutter.

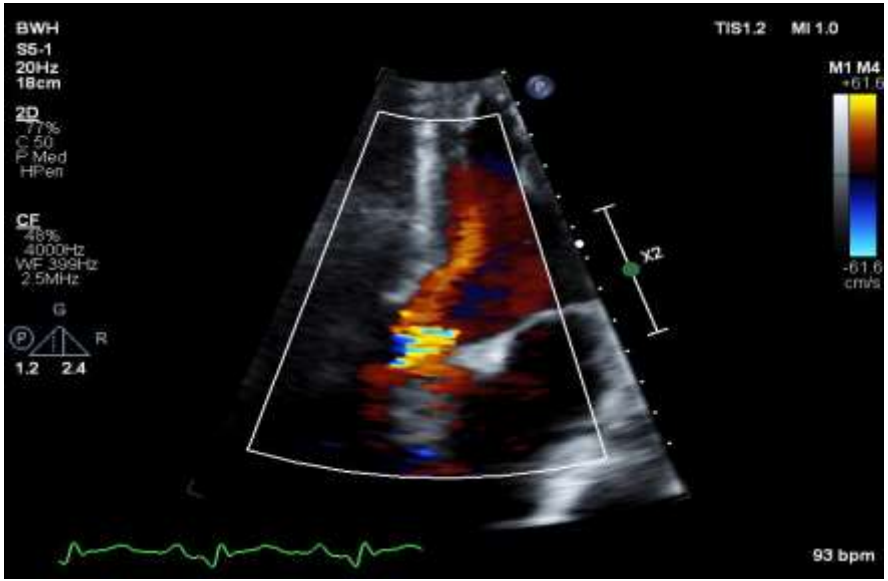
Aortic Aneurysm Disease





3 months post-op

Aortic Aneurysm Disease



8 months post-op