Aortopathy Guidelines and Management

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Disclosures

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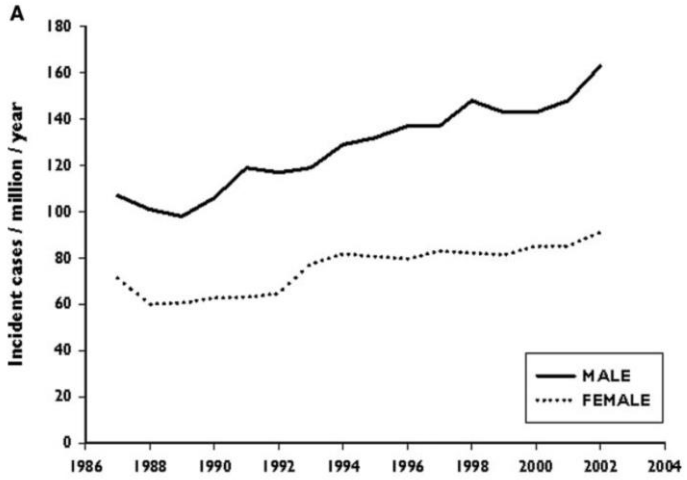
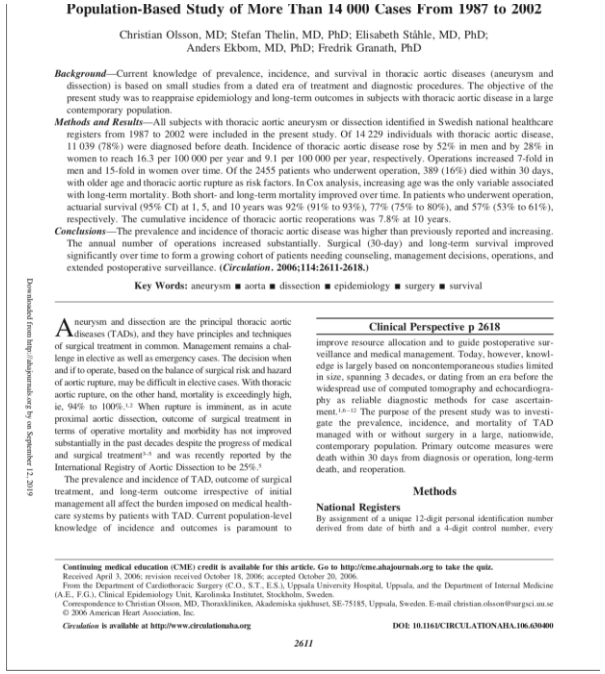
Epidemiology

Thoracic Aortic Aneurysm and Dissection

Increasing Prevalence and Improved Outcomes Reported in a Nationwide Population-Based Study of More Than 14 000 Cases From 1987 to 2002

Christian Olsson, MD; Stefan Thelin, MD, PhD; Elisabeth Ståhle, MD, PhD;
Anders Ekbom, MD, PhD; Fredrik Granath, PhD





Population-Based Study of More Than 14 000 Cases From 1987 to 2002

Christian Olsson, MD; Stefan Thelin, MD, PhD; Elisabeth Ståhle, MD, PhD;
Anders Ekbom, MD, PhD; Fredrik Granath, PhD

Background—Current knowledge of prevalence, incidence, and survival in thoracic aortic diseases (aneurysm and dissection) is based on small studies from a dated era of treatment and diagnostic procedures. The objective of the present study was to reappraise epidemiology and long-term outcomes in subjects with thoracic aortic disease in a large contemporary population.

Methods and Results—All subjects with thoracic aortic aneurysm or dissection identified in Swedish national healthcare registers from 1987 to 2002 were included in the present study. Of 14 229 individuals with thoracic aortic disease, 11 039 (78%) were diagnosed before death. Incidence of thoracic aortic disease rose by 52% in men and by 28% in women to reach 16.3 per 100 000 per year and 9.1 per 100 000 per year, respectively. Operations increased 7-fold in men and 15-fold in women over time. Of the 2455 patients who underwent operation, 389 (16%) died within 30 days, with older age and thoracic aortic rupture as risk factors. In Cox analysis, increasing age was the only variable associated with long-term mortality. Both short- and long-term mortality improved over time. In patients who underwent operation, actuarial survival (95% CI) at 1, 5, and 10 years was 92% (91% to 93%), 77% (75% to 80%), and 57% (53% to 61%), respectively. The cumulative incidence of thoracic aortic reoperations was 7.8% at 10 years.

Conclusions—The prevalence and incidence of thoracic aortic disease was higher than previously reported and increasing. The annual number of operations increased substantially. Surgical (30-day) and long-term survival improved significantly over time to form a growing cohort of patients needing counseling, management decisions, operations, and extended postoperative surveillance. (***Circulation***. **2006;114:2611-2618.**)

Key Words: aneurysm ■ aorta ■ dissection ■ epidemiology ■ surgery ■ survival

Aneurysm and dissection are the principal thoracic aortic diseases (TADs), and they have principles and techniques of surgical treatment in common. Management remains a challenge in elective as well as emergency cases. The decision when and if to operate, based on the balance of surgical risk and hazard of aortic rupture, may be difficult in elective cases. With thoracic aortic rupture, on the other hand, mortality is exceedingly high, ie, 94% to 100%.[1,2] When rupture is imminent, as in acute proximal aortic dissection, outcome of surgical treatment in terms of operative mortality and morbidity has not improved substantially in the past decades despite the progress of medical and surgical treatment[3–5] and was recently reported by the International Registry of Aortic Dissection to be 25%.[5]

The prevalence and incidence of TAD, outcome of surgical treatment, and long-term outcome irrespective of initial management all affect the burden imposed on medical healthcare systems by patients with TAD. Current population-level knowledge of incidence and outcomes is paramount to

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improve resource allocation and to guide postoperative surveillance and medical management. Today, however, knowledge is largely based on noncontemporaneous studies limited in size, spanning 3 decades, or dating from an era before the widespread use of computed tomography and echocardiography as reliable diagnostic methods for case ascertainment.[1,6–12] The purpose of the present study was to investigate the prevalence, incidence, and mortality of TAD managed with or without surgery in a large, nationwide, contemporary population. Primary outcome measures were death within 30 days from diagnosis or operation, long-term death, and reoperation.

Methods

National Registers

By assignment of a unique 12-digit personal identification number derived from date of birth and a 4-digit control number, every

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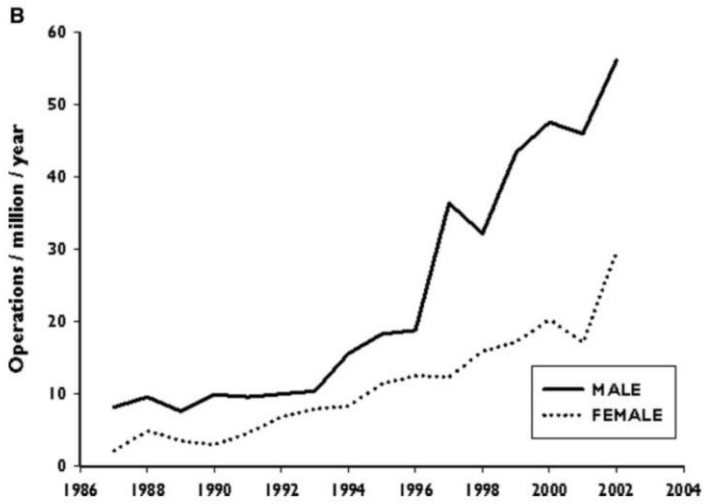
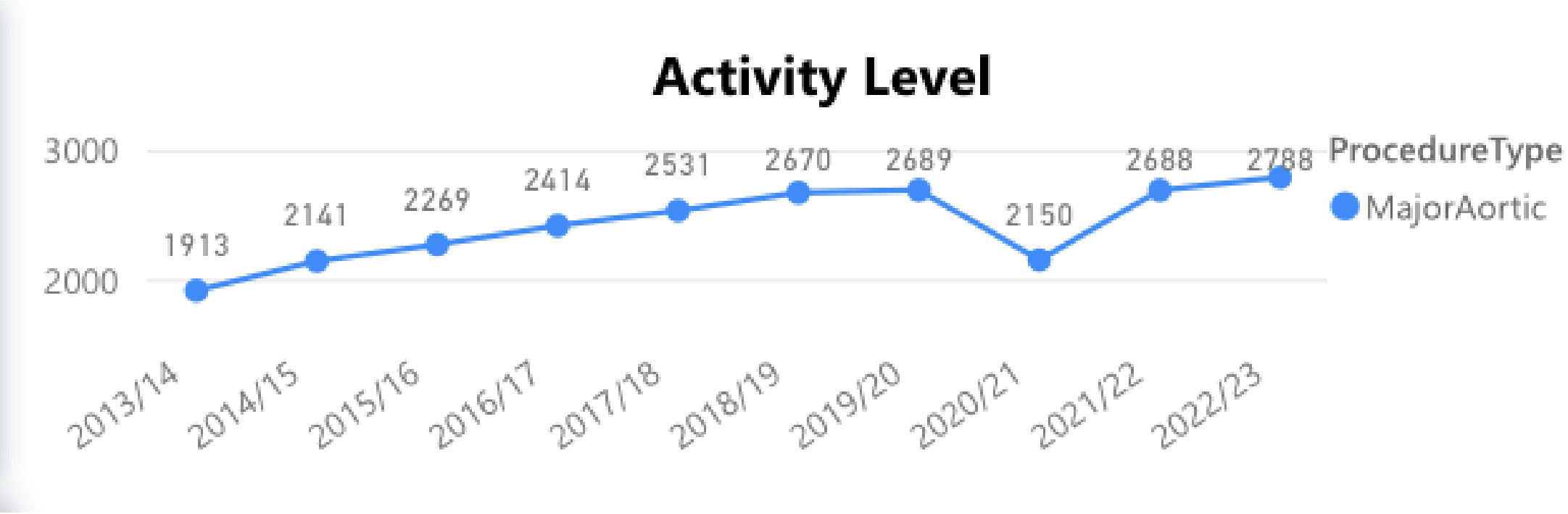


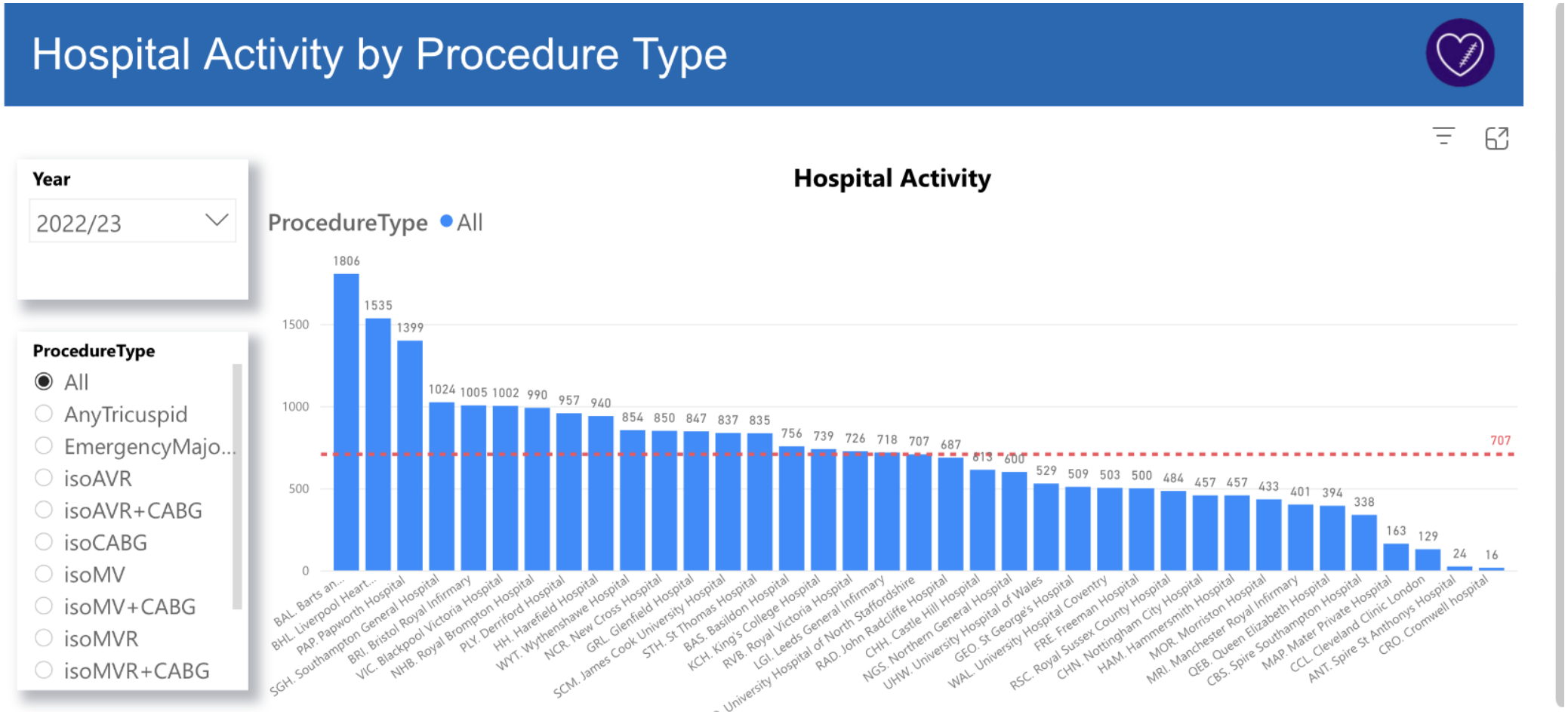
Thoracic Aortic Disease is increasing

Trend of UK Elective Aortic Surgery 2013 - 2023

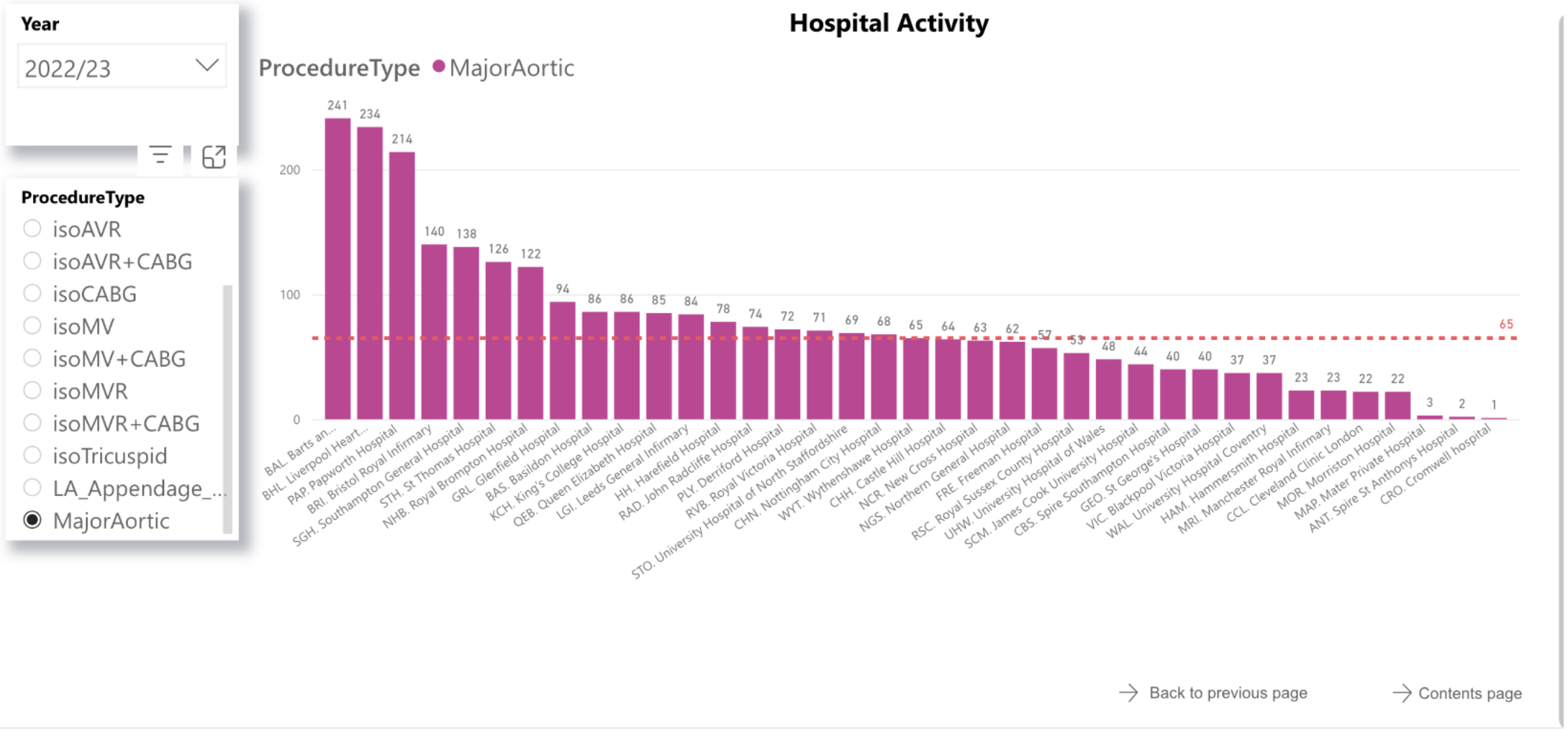


Trend of UK Emergency Aortic Activity 2013 - 2023

UK Cardiac Surgery Activity 2022/23



UK Elective Aortic Surgery 2022/23



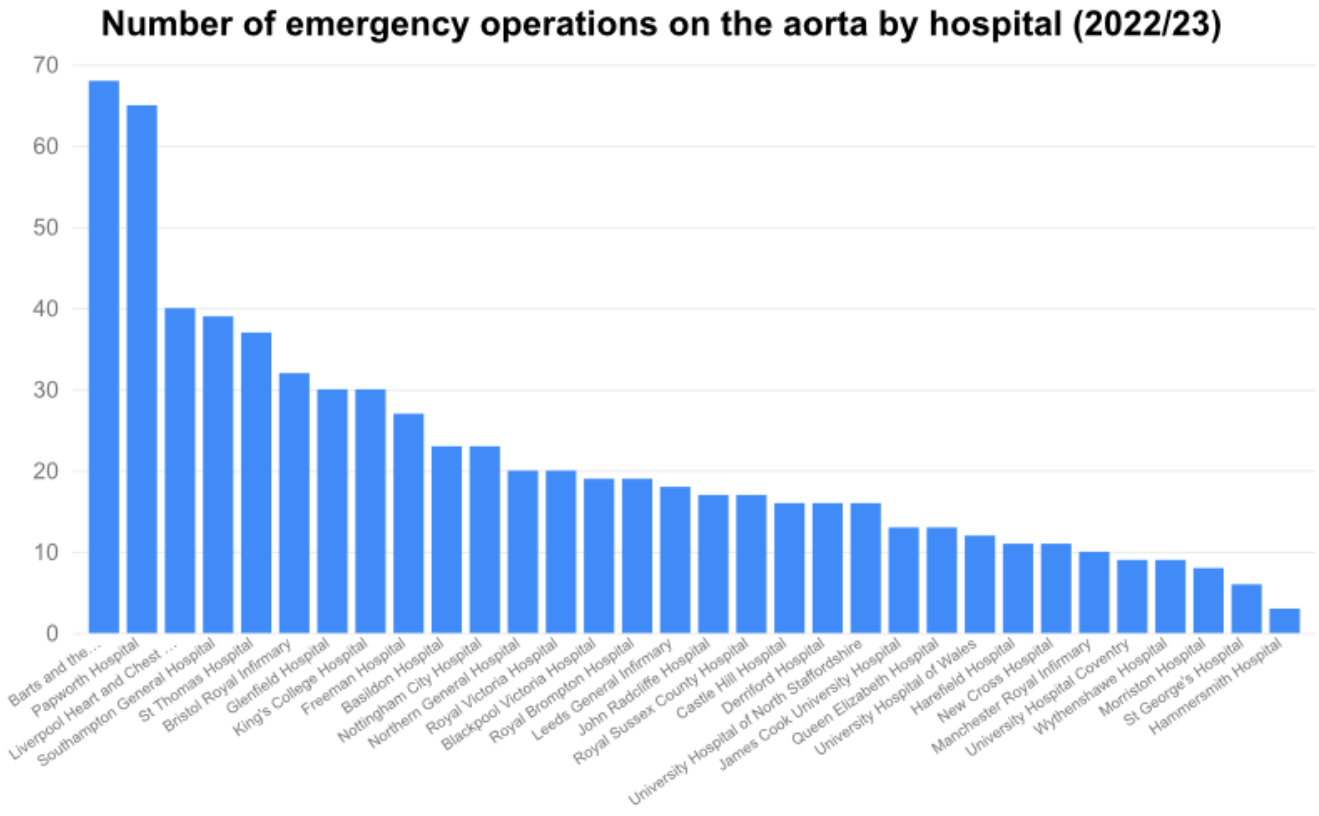
UK Emergency Aortic Surgery 2022/23

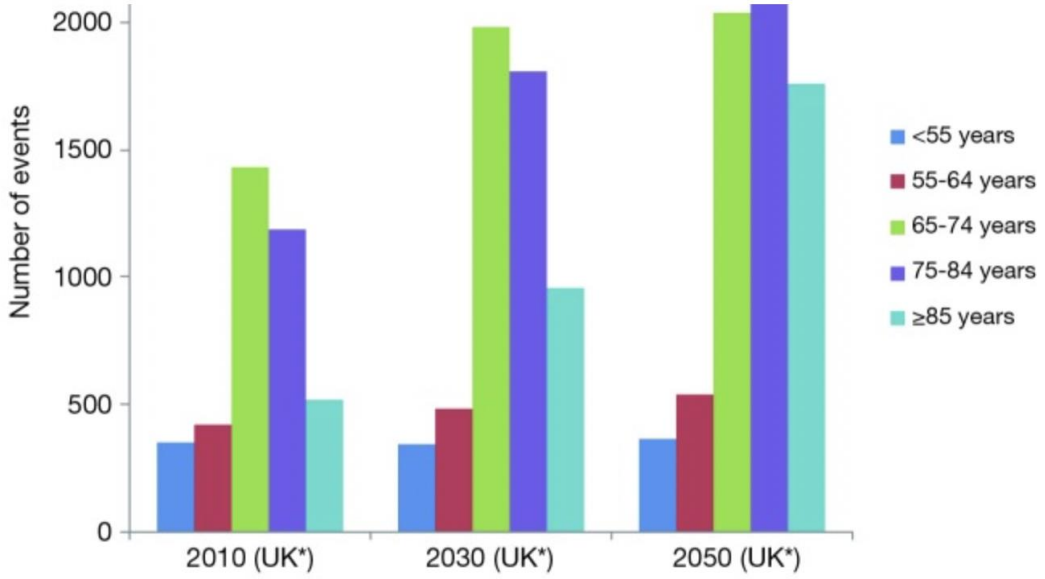
In 2022/23, the majority of hospitals performing emergency operations on the aorta carried out fewer than 24 operations

A total of 697 emergency operations were performed on the thoracic aorta in 2022/23. Most (but not all) of these were for acute aortic dissection.

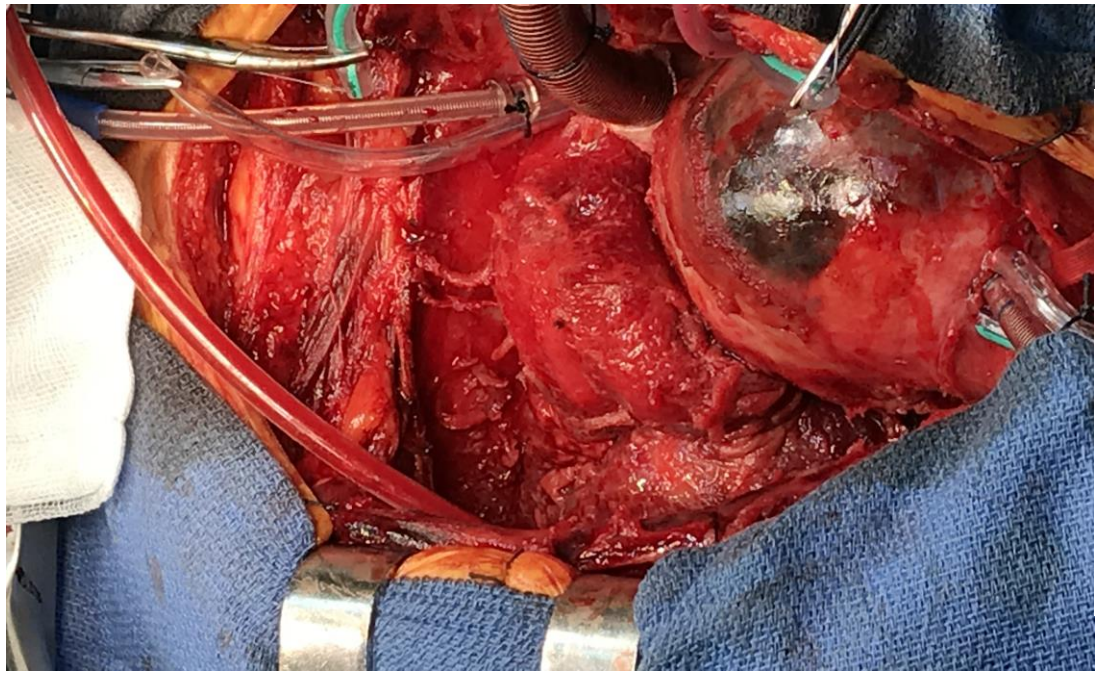
The 2023 NACSA audit report suggested possibly improved mortality outcomes in centres undertaking 24 or more operations per year (based on UK results from the last decade).

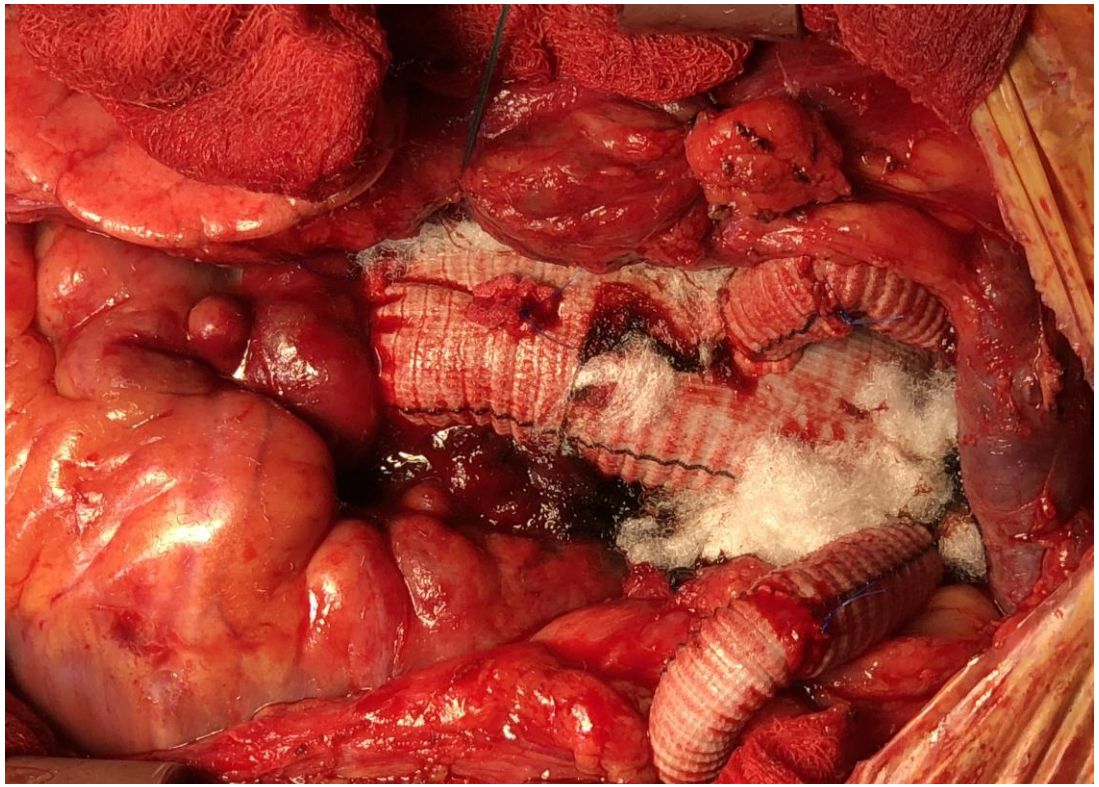
In 2022/23, out of 32 hospitals performing emergency operations on the aorta, 23 did not reach the minimum recommended number of procedures, with the lowest carrying out just three cases.





*Estimated rates based on Office of National Statistics 2010 UK census population and population projections by age and sex for 2030 and 2050





Projected number of incident dissection events occurring in the UK population in 2010 – 2050 stratified by sex and age

2010				2030			2050		
Years	Men	Women	Total	Men	Women	Total	Men	Women	Total
<55	348	0	348	344	0	344	368	0	368
55-64	346	75	421	394	86	481	451	91	542
65-74	888	542	1430	1235	745	1980	1299	738	2036
75-84	585	602	1187	947	863	1811	1158	1035	2193
≥85	89	431	520	219	738	957	423	1336	1759
Total	2256	1650	3906	3140	2433	5573	3698	3200	6898

Global Trend of Aortic Diseases

- Increase population in older age group
- Increasing number of GUCH patients transition with improving clinical outcomes of early stage interventions
- Awareness of aortic diseases

Circulation

Check for updates

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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AIM: The "2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease" provides recommendations to guide clinicians in the diagnosis, genetic evaluation and family screening, medical therapy, endovascular and surgical treatment, and long-term surveillance of patients with aortic disease across its multiple clinical presentation subsets (ie, asymptomatic, stable symptomatic, and acute aortic syndromes).

METHODS: A comprehensive literature search was conducted from January 2021 to April 2021, encompassing studies, reviews, and other evidence conducted on human subjects that were published in English from PubMed, EMBASE, the Cochrane Library, CINHL Complete, and other selected databases relevant to this guideline. Additional relevant studies, published through June 2022 during the guideline writing process, were also considered by the writing committee, where appropriate.

STRUCTURE: Recommendations from previously published AHA/ACC guidelines on thoracic aortic disease, peripheral artery disease, and bicuspid aortic valve disease have been updated with new evidence to guide clinicians. In addition, new

*Writing committee members are required to recuse themselves from voting on sections to which their specific relationships with industry may apply; see Appendix 1 for detailed information. †SCA representative. ‡ACR representative. §AHA/ACC Joint Committee on Clinical Data Standards liaison. ‖Lay stakeholder representative. ¶SCAI representative. #AATS representative. **ACC/AHA Joint Committee on Performance Measures liaison. ††AHA/ACC Joint Committee on Clinical Practice Guidelines liaison. ‡‡STS representative. §§SVS representative. ‖‖AHA/ACC staff representative.

ACC/AHA Joint Committee on Clinical Practice Guidelines Members, see page e445.

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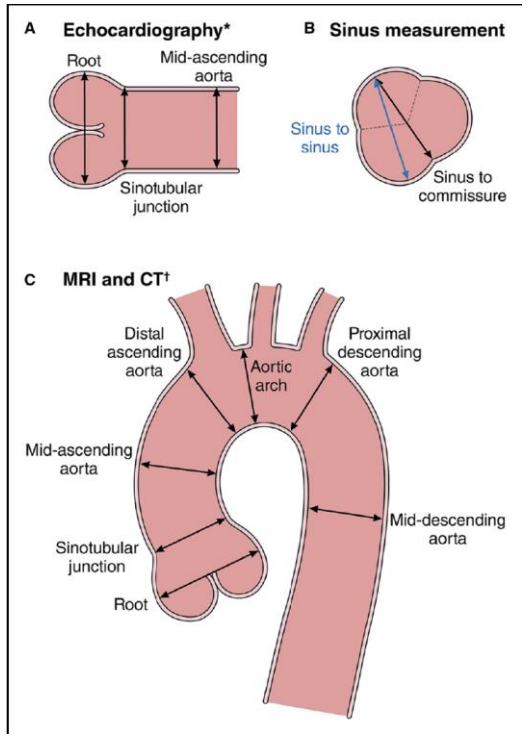

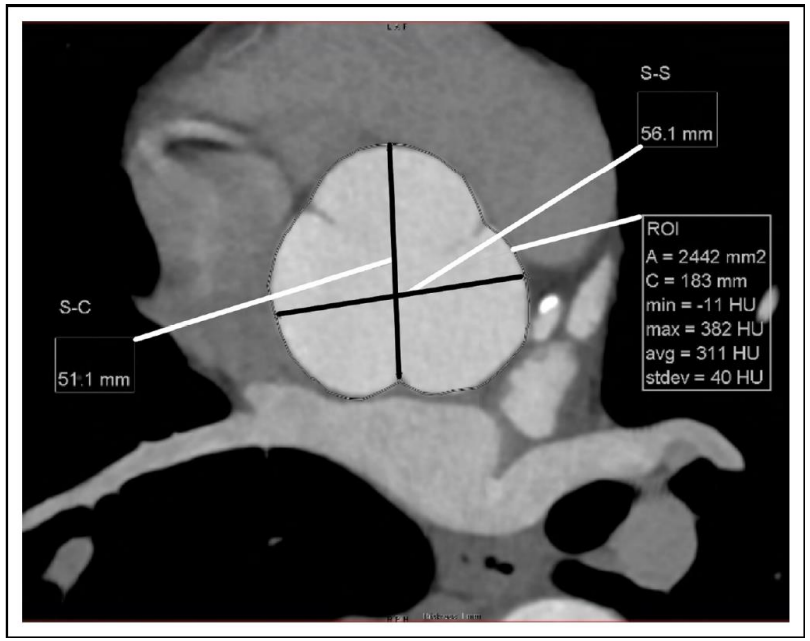

Recommendations for Marfan Syndrome Interventions: Replacement of the Aortic Root in Patients With Marfan Syndrome Referenced studies that support the recommendations are summarized in the Online Data Supplement.		
COR	**LOE**	**Recommendations**
1	**B-NR**	1. In patients with Marfan syndrome and an aortic root diameter of ≥5.0 cm, surgery to replace the aortic root and ascending aorta is recommended.[1-4]
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 (see Table 10), surgery to replace the aortic root and ascending aorta is reasonable, when performed by experienced surgeons in a Multidisciplinary Aortic Team.[1,3,4]
2a	**C-LD**	3. In patients with Marfan syndrome and a maximal cross-sectional aortic root area (cm2) 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.[5]
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.[2-4]

5. SHARED DECISION-MAKING

Recommendations for Shared Decision-Making		
COR	**LOE**	**Recommendations**
1	**C-LD**	1. In patients with aortic disease, shared decision-making is recommended when determining the appropriate thresholds for intervention, deciding on the type of surgical repair, choosing between open surgical versus endovascular approaches; and in medical management and surveillance.[1-6]
1	**C-EO**	2. In patients with aortic disease who are contemplating pregnancy or who are pregnant, shared decision-making is recommended when considering the cardiovascular risks of pregnancy, the diameter thresholds for prophylactic aortic surgery, and the mode of delivery.

ESC European Society of Cardiology
European Heart Journal (2024) **45**, 3538–3700
https://doi.org/10.1093/eurheartj/ehae179

ESC GUIDELINES

2024 ESC Guidelines for the management of peripheral arterial and aortic diseases

Developed by the task force on the management of peripheral arterial and aortic diseases of the European Society of Cardiology (ESC)

Endorsed by the European Association for Cardio-Thoracic Surgery (EACTS), the European Reference Network on Rare Multisystemic Vascular Diseases (VASCERN), and the European Society of Vascular Medicine (ESVM)

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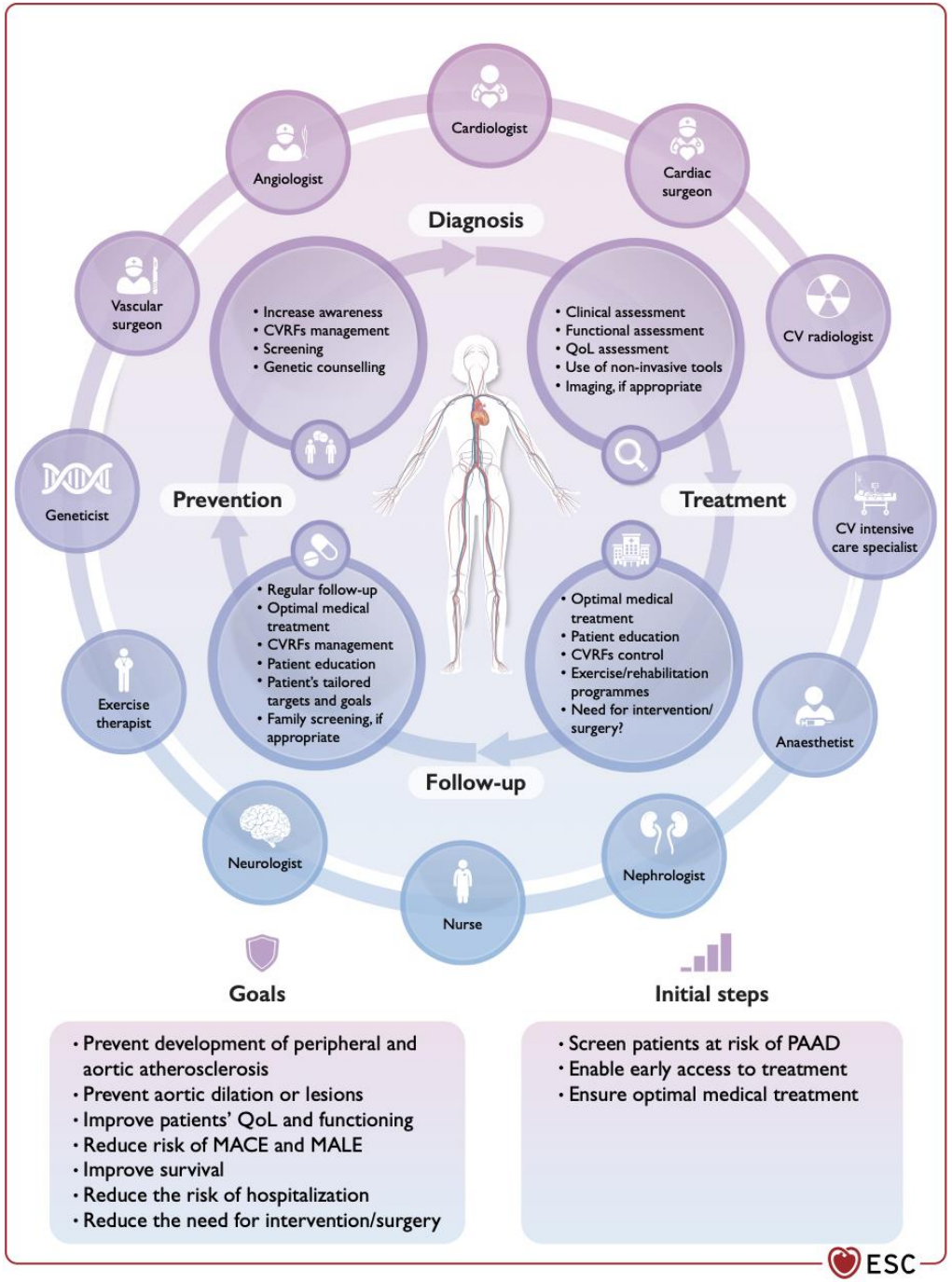
[2] Representing the European Reference Network on Rare Multisystemic Vascular Diseases (VASCERN)

ESC Clinical Practice Guidelines (CPG) Committee: listed in the Appendix.

ESC subspecialty communities having participated in the development of this document:

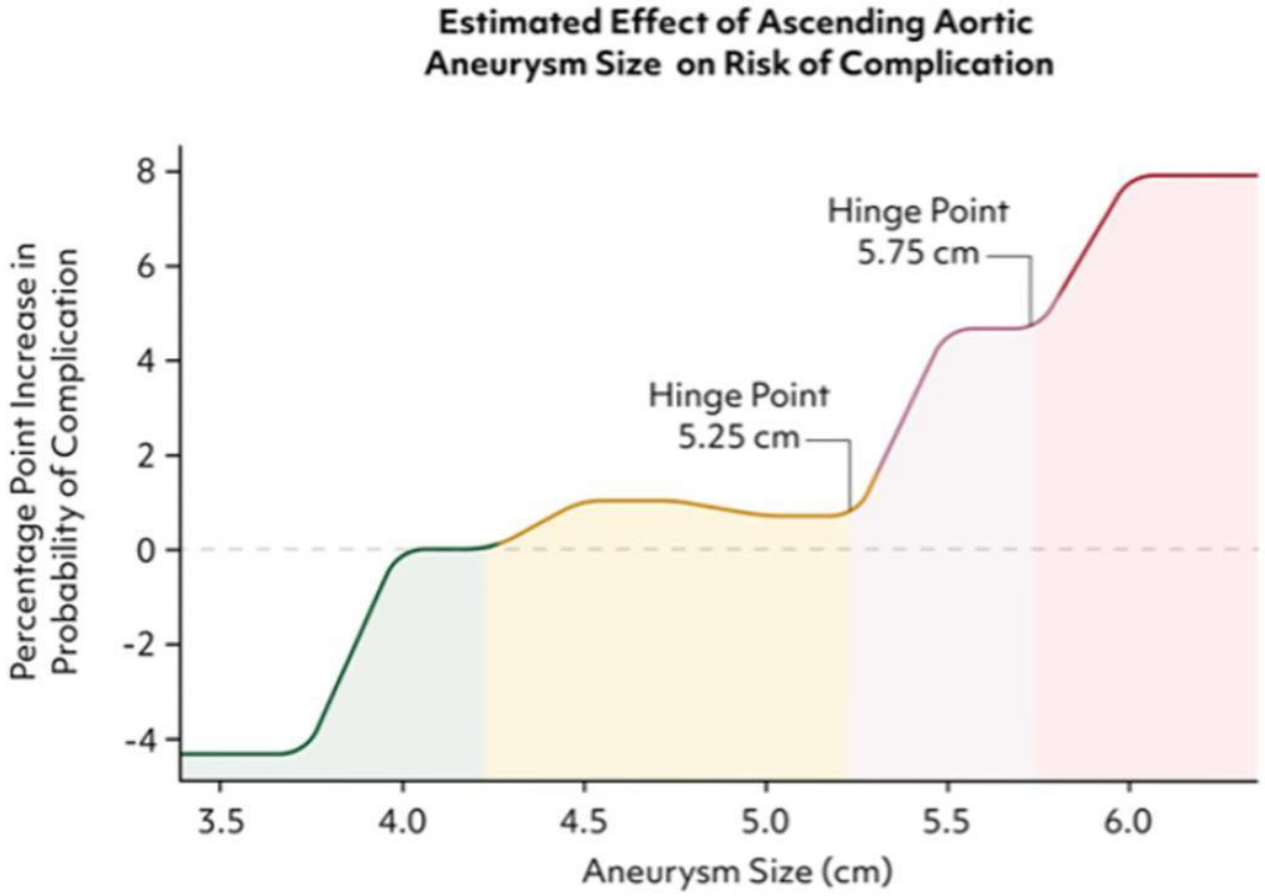
Associations: Association for Acute CardioVascular Care (ACVC), Association of Cardiovascular Nursing & Allied Professions (ACNAP), European Association of Cardiovascular Imaging (EACVI), European Association of Preventive Cardiology (EAPC), European Association of Percutaneous Cardiovascular Interventions (EAPCI), Heart Failure Association (HFA).

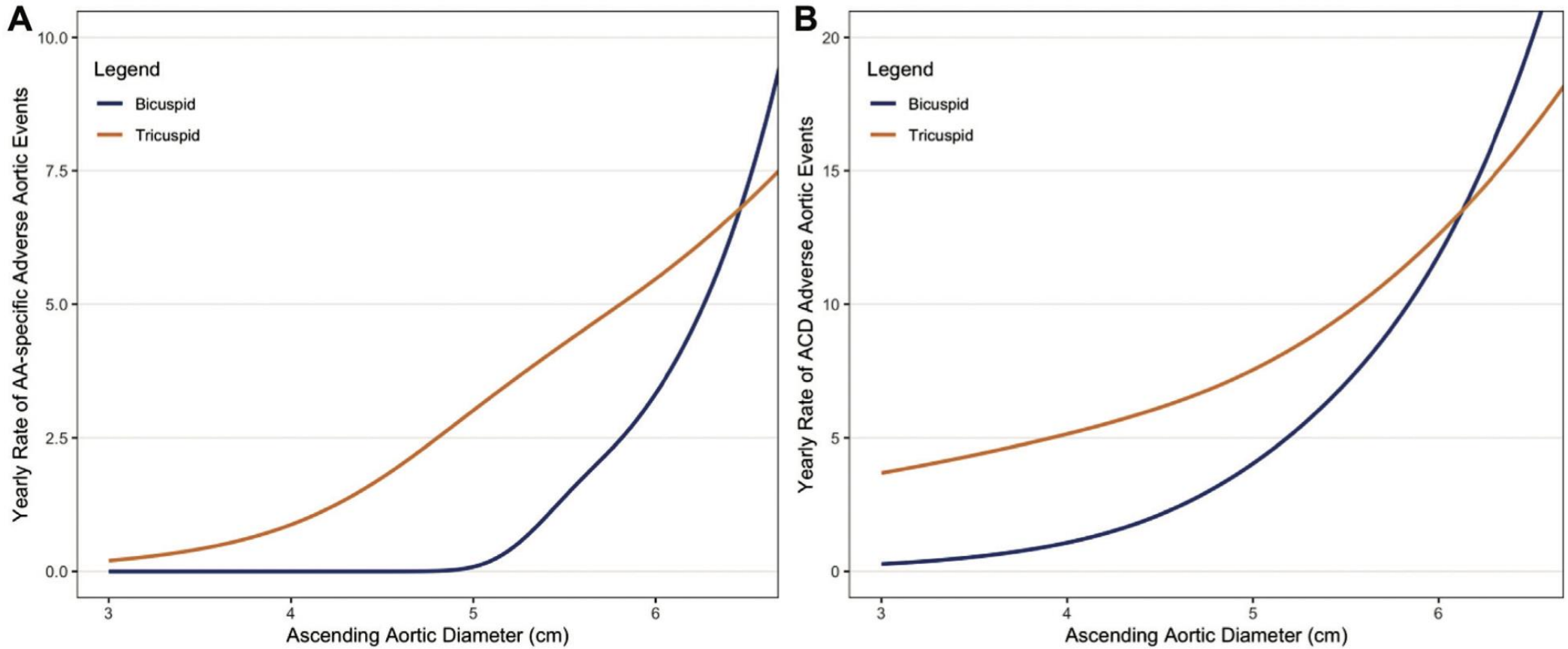


Recommendations for surgery in aortic root and ascending aorta dilatation associated with tricuspid aortic valve		
In patients with dilatation of the tubular ascending aorta who can be offered surgery with low predicted risk, ascending aortic replacement should be considered at a maximum diameter >52 mm.	IIa	B
In patients undergoing surgery for tricuspid aortic valve disease who have concomitant dilatation of the aortic root or ascending tubular aorta, and low predicted surgical risk, ascending aorta or root replacement should be considered at a maximum diameter ≥45 mm, otherwise ≥50 mm.	IIa	B
SAPT with low-dose aspirin (75–100 mg/day) should be considered for the first 3 months after valve-sparing aortic surgery when there are no other baseline indications for OAC.	IIa	C
In patients undergoing non-aortic-valve cardiac surgery who have concomitant dilatation of the ascending aorta or aortic root with a maximum diameter ≥50 mm, concomitant aortic surgery should be considered.	IIa	C
Recommendations for surgery in aortic arch aneurysms		
In patients with low or intermediate operative risk with an aortic arch aneurysm and recurrent episodes of chest pain not attributable to non-aortic causes, open surgical replacement of the arch is recommended.	I	C
In patients undergoing open surgical repair of an aortic arch aneurysm, an elephant trunk or frozen elephant trunk procedure should be considered if the aneurysmal disease extends into the proximal descending thoracic aorta.	IIa	C

Recommendations for follow-up after treatment of aortic aneurysms		
After open repair of TAA, an early CCT is recommended within 1 month, and then yearly CCT follow-up for the first 2 post-operative years and every 5 years thereafter is recommended if findings are stable.	I	B
After 5 post-operative years without complications, continuing long-term follow-up of TEVAR by CCT every 5 years should be considered.	IIa	B
If growth of the excluded aneurysm is observed, without evidence of type I or III endoleak, repeating CCT every 6–12 months, depending on the growth rate observed, should be considered.	IIa	C
In low-risk patients, from 1 year post-operatively after EVAR, repeating DUS/CEUS every 2 years should be considered.	IIa	B
If any abnormality during DUS/CEUS is found, confirmation should be considered using additional CCT or CMR (based on potential artefacts).	IIa	B

Change in Threshold



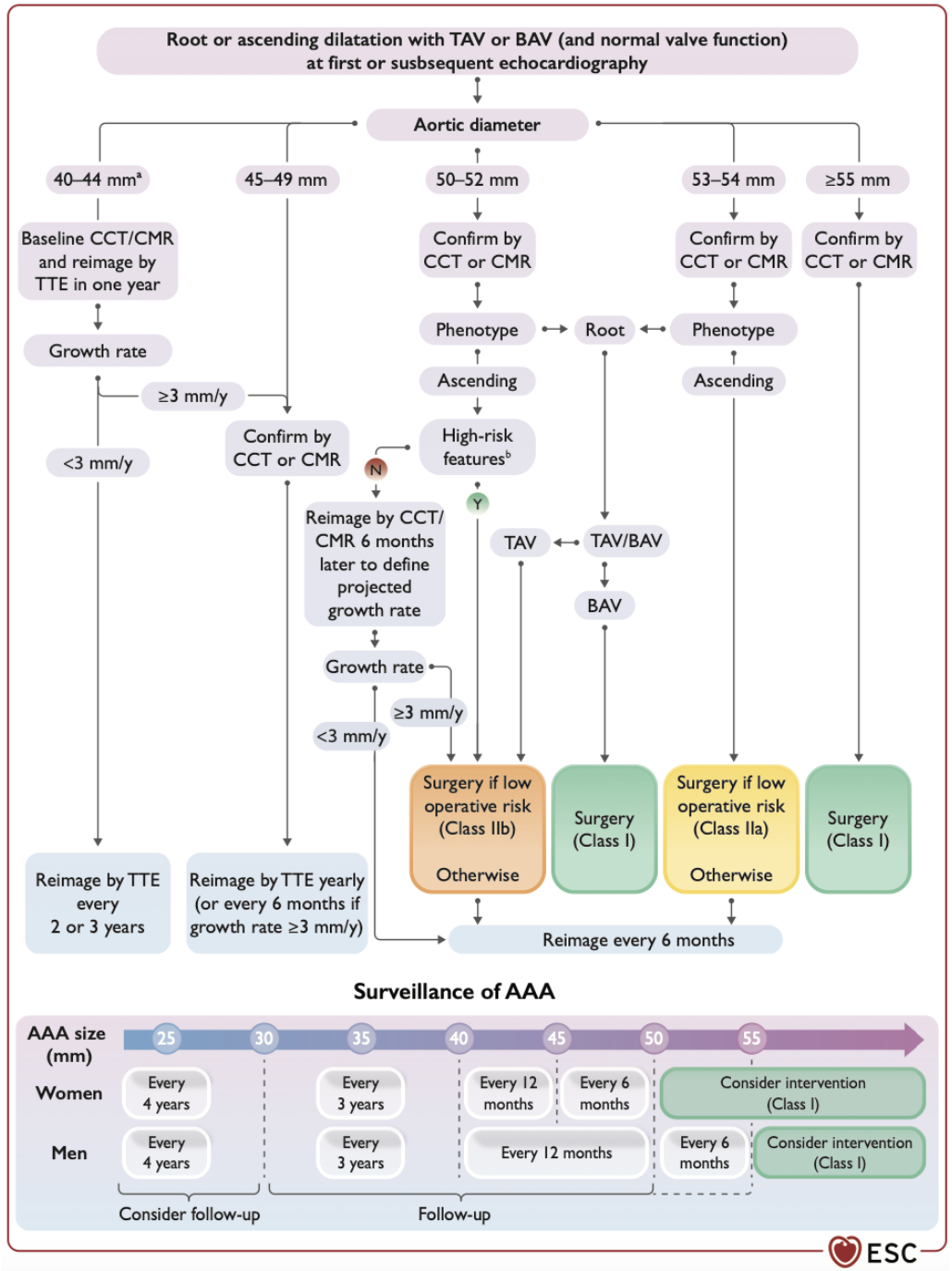


Recommendations for bicuspid aortic valve-associated aortopathy management		
Surgery for bicuspid aortopathy of the root phenotype is recommended when the maximum aortic diameter is ≥50 mm.	I	B
Screening by TTE in FDRs of BAV patients with root phenotype aortopathy and/or isolated aortic regurgitation is recommended.	I	C
In patients with low surgical risk, surgery for bicuspid aortopathy of ascending phenotype should be considered when the maximum aortic diameter is >52 mm.	IIa	B

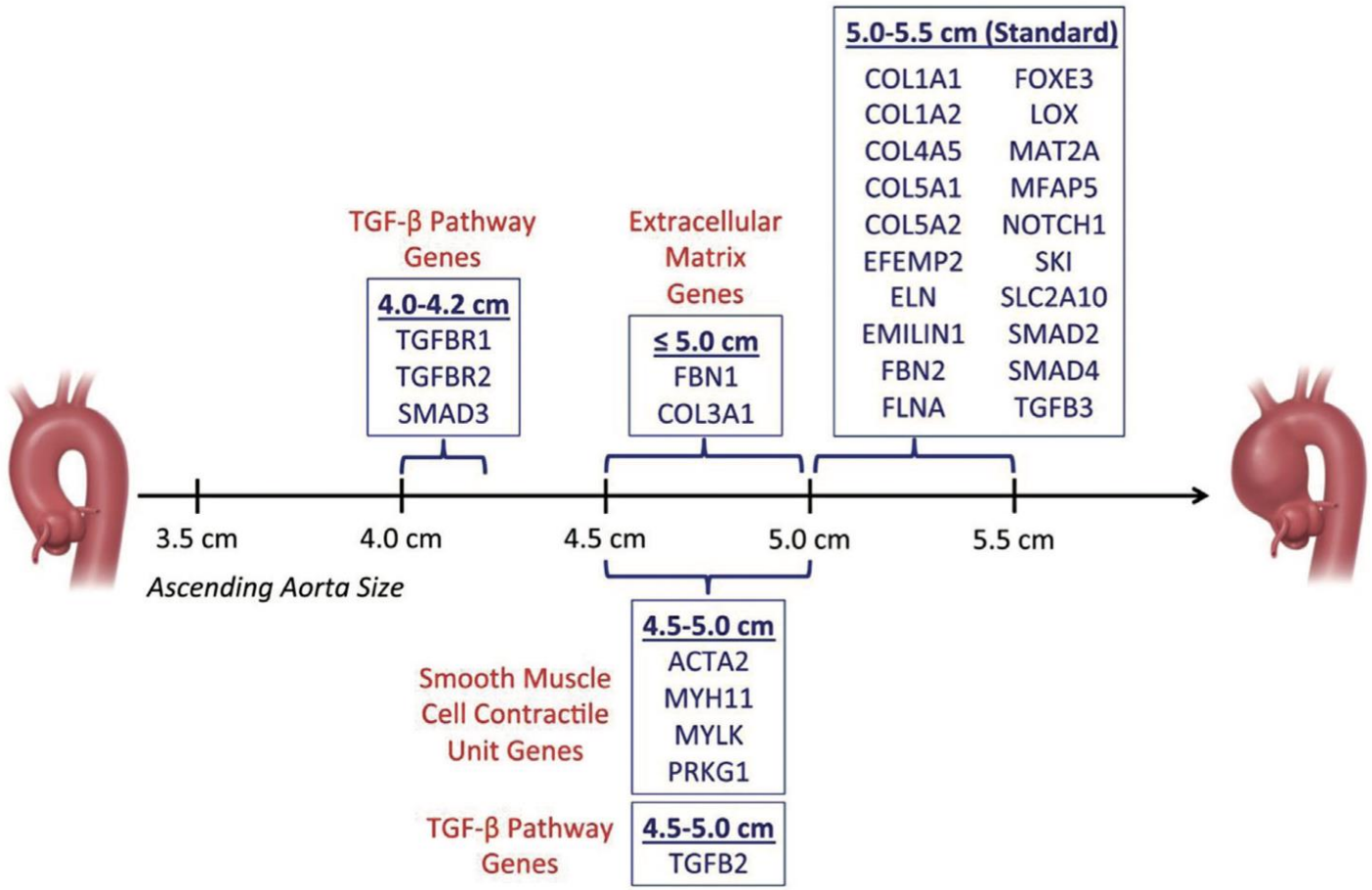
Recommendations for bicuspid aortic valve-associated aortopathy management					
Cardiac MRI or CT is indicated in patients with BAV when the morphology of the aortic root and the ascending aorta cannot be accurately assessed by TTE.	I	C	CCT or CMR of the entire thoracic aorta is recommended at first diagnosis and when important discrepancies in measurements are found between subsequent TTE controls during surveillance, or when the diameter of the aorta exceeds 45 mm.	I	C
In the case of aortic diameter >50 mm or an increase of >3 mm per year measured by echocardiography, confirmation of the measurement is indicated, using another imaging modality (CT or MRI).	I	C			
In the case of a diameter of the aortic root or the ascending aorta >45 mm or an increase of >3 mm per year measured by echocardiography, annual measurement of aortic diameter is indicated.	I	C	Surveillance serial imaging by TTE is recommended in BAV patients with a maximum aortic diameter >40 mm, either with no indication for surgery or after isolated aortic valve surgery, after 1 year, then if stability is observed, every 2–3 years.	I	C
In cases of BAV, surgery of the ascending aorta is indicated in the case of: • Aortic root or ascending aortic diameter >50 mm in the presence of other risk factors (coarctation of the aorta, systemic hypertension, family history of dissection, or increase in aortic diameter of >3 mm per year).	I	C	In patients with low surgical risk and ascending phenotype bicuspid aortopathy, surgery should be considered at a maximum diameter ≥50 mm if any of the following is the case: • Age <50 years • Short stature • Ascending aortic length ≥11 cm • Aortic diameter growth rate >3 mm per year • Family history of acute aortic syndrome • Aortic coarctation • Resistant hypertension • Concomitant non-aortic-valve cardiac surgery • Desire for pregnancy	IIa	C

Recommendations for imaging and surgery in *ACTA2*-related heritable thoracic aortic disease		
Annual monitoring of the aortic root/ascending aorta with TTE to evaluate for aortic root/ascending aorta enlargement is recommended.	I	C
Imaging of the aorta with CMR/CCT every 3–5 years is recommended.	I	C
Prophylactic aortic root surgery should be considered with a diameter ≥45 mm, or lower in cases with other risk factors.	IIa	C

Recommendations for aortic surgery in women with Turner syndrome		
Elective surgery for aneurysms of the aortic root and/or ascending aorta should be considered in women with TS who are ≥15 years of age, have an ascending ASI >23 mm/m^2, an AHI >23 mm/m, a z-score >3.5, and have associated risk factors for aortic dissection or are planning pregnancy.	IIa	C
Elective surgery for aneurysms of the aortic root and/or ascending aorta may be considered for women with TS who are ≥15 years of age, have an ascending ASI >25 mm/m^2, an AHI >25 mm/m, a z-score >4, and who do not have associated risk factors for aortic dissection.	IIb	C

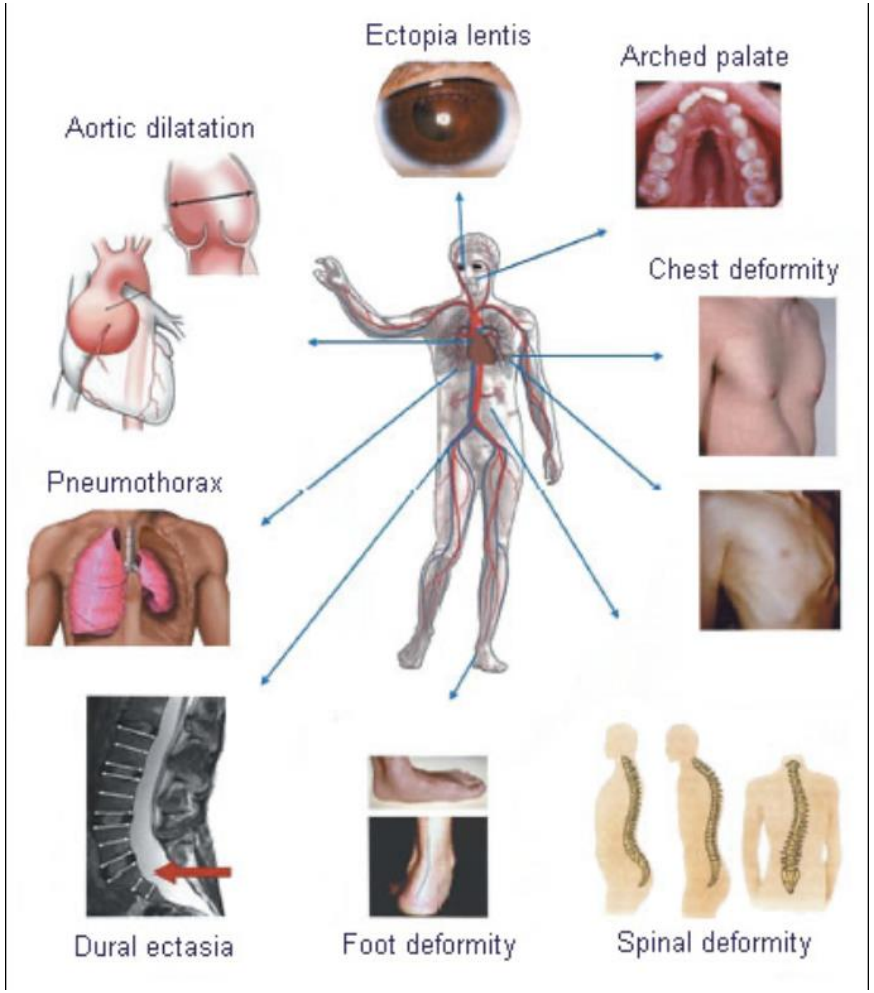

Root or ascending dilatation with TAV or BAV (and normal valve function) at first or susbsequent echocardiography
Aortic diameter
40–44 mm[a]
45–49 mm
50–52 mm
53–54 mm
≥55 mm
Baseline CCT/CMR and reimage by TTE in one year
Confirm by CCT or CMR
Confirm by CCT or CMR
Confirm by CCT or CMR
Growth rate
Phenotype
Root
Phenotype
≥3 mm/y
Ascending
Ascending
<3 mm/y
Confirm by CCT or CMR
High-risk features[b]
N
Y
Reimage by CCT/ CMR 6 months later to define projected growth rate
TAV
TAV/BAV
BAV
Growth rate
≥3 mm/y
<3 mm/y
Surgery if low operative risk (Class IIb) Otherwise
Surgery (Class I)
Surgery if low operative risk (Class IIa) Otherwise
Surgery (Class I)
Reimage by TTE every 2 or 3 years
Reimage by TTE yearly (or every 6 months if growth rate ≥3 mm/y)
Reimage every 6 months
Surveillance of AAA
AAA size (mm)
25
30
35
40
45
50
55
Women
Every 4 years
Every 3 years
Every 12 months
Every 6 months
Consider intervention (Class I)
Men
Every 4 years
Every 3 years
Every 12 months
Every 6 months
Consider intervention (Class I)
Consider follow-up
Follow-up
ESC

HTAD


5.0-5.5 cm (Standard)
COL1A1
COL1A2
COL4A5
COL5A1
COL5A2
EFEMP2
ELN
EMILIN1
FBN2
FLNA
FOXE3
LOX
MAT2A
MFAP5
NOTCH1
SKI
SLC2A10
SMAD2
SMAD4
TGFB3
TGF-β Pathway Genes
4.0-4.2 cm
TGFBR1
TGFBR2
SMAD3
Extracellular Matrix Genes
≤ 5.0 cm
FBN1
COL3A1
3.5 cm
4.0 cm
4.5 cm
5.0 cm
5.5 cm
Ascending Aorta Size
Smooth Muscle Cell Contractile Unit Genes
4.5-5.0 cm
ACTA2
MYH11
MYLK
PRKG1
TGF-β Pathway Genes
4.5-5.0 cm
TGFB2

Marfan Syndrome

- Incidence 1 in 10,000
- FBN1 gene defect
- Autosomal dominant
- Affect fibrillin and elastic fibers of connective tissue
- Aortic aneurysm 60-80%
- Aortic dissection
- Mitral valve prolapse
- Life expectancy increased to 70

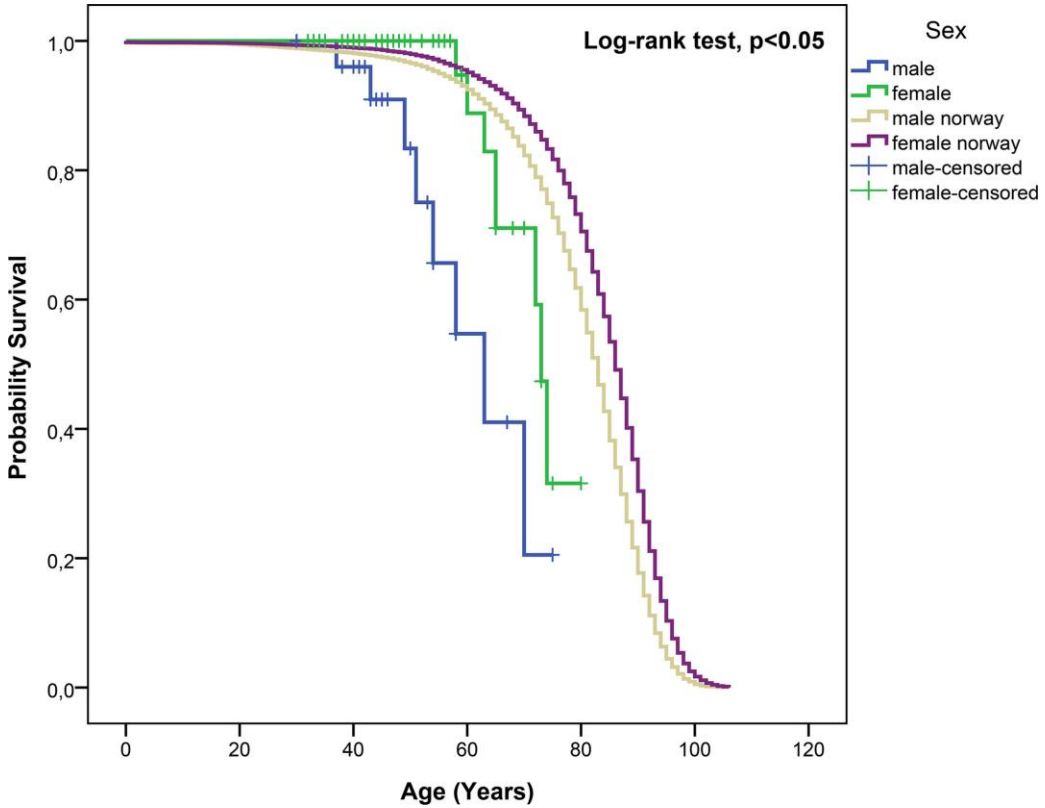


Marfan Syndrome and Thoracic Aneurysm

- 60 - 80 % of adult develop thoracic aneurysm
- 50 % of aortic dissection patients under 40 years of age

26. Januzzi JL, Isselbacher EM, Fattori R, et al. Characterizing the young patient with aortic dissection: results from the International Registry of Aortic Dissection (IRAD). *J Am Coll Cardiol* 2004;43:665-9. 10.1016/j.jacc.2003.08.054

Survival, causes of death, and cardiovascular events in patients with Marfan syndrome

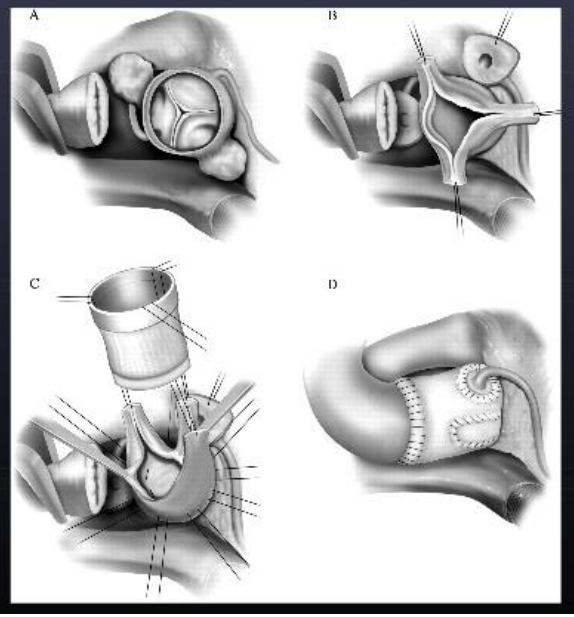


Molec Gen & Gen Med, Volume: 6, Issue: 6, Pages: 1114-1123, First published: 04 November 2018, DOI: (10.1002/mgg3.489)

Native Valve Sparing Root Replacement

Valve-sparing aortic root replacement: the inclusion (David) technique

Operative Techniques in Thoracic and Cardiovascular Surgery 2005;10(4):246–258
David TE, Feindel CM: An aortic valve-sparing operati(in for patients with aortic incompetence and aneurysm of the asrending aorta. J Thorac Cardiovasc Surg 103:617-622, 1992



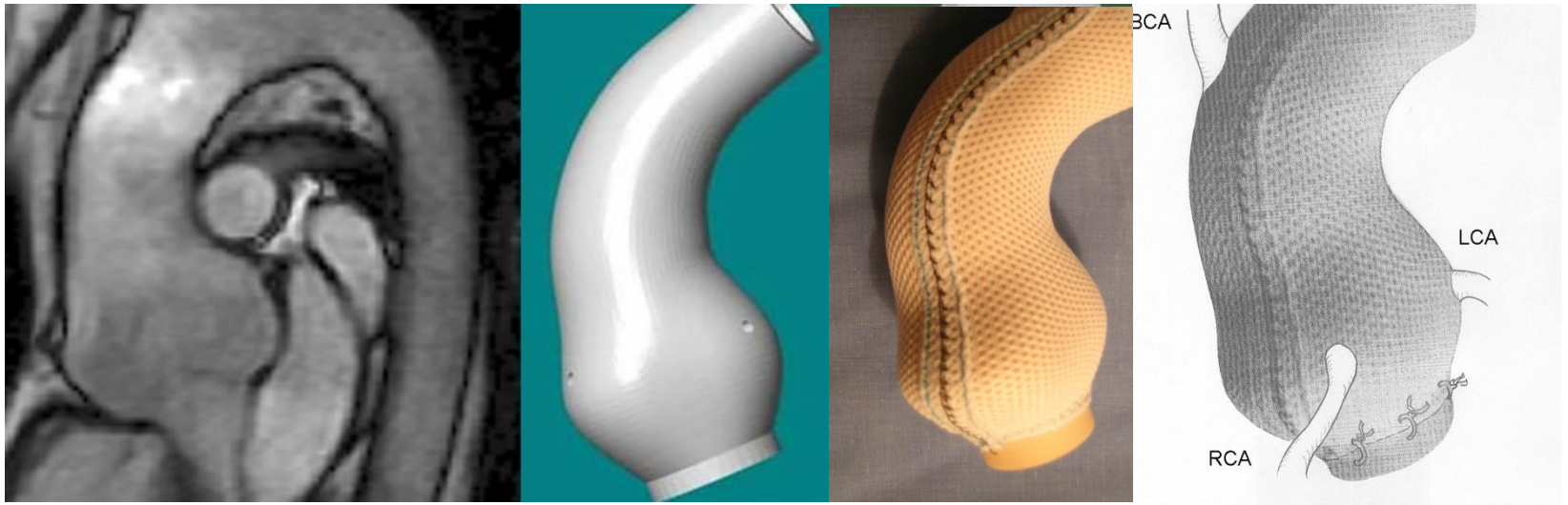
Minimally Invasive Valve Sparing Root Replacement

Professor A. Oo
Barts Heart Centre

Barts Heart Centre

NHS Barts Health

Aortic root support



Personalised External Aortic Root Support

Patient-led clinical research: CAD and RP

PEARS: challenges

Meticulous dissection is helped by the absence of heparin.

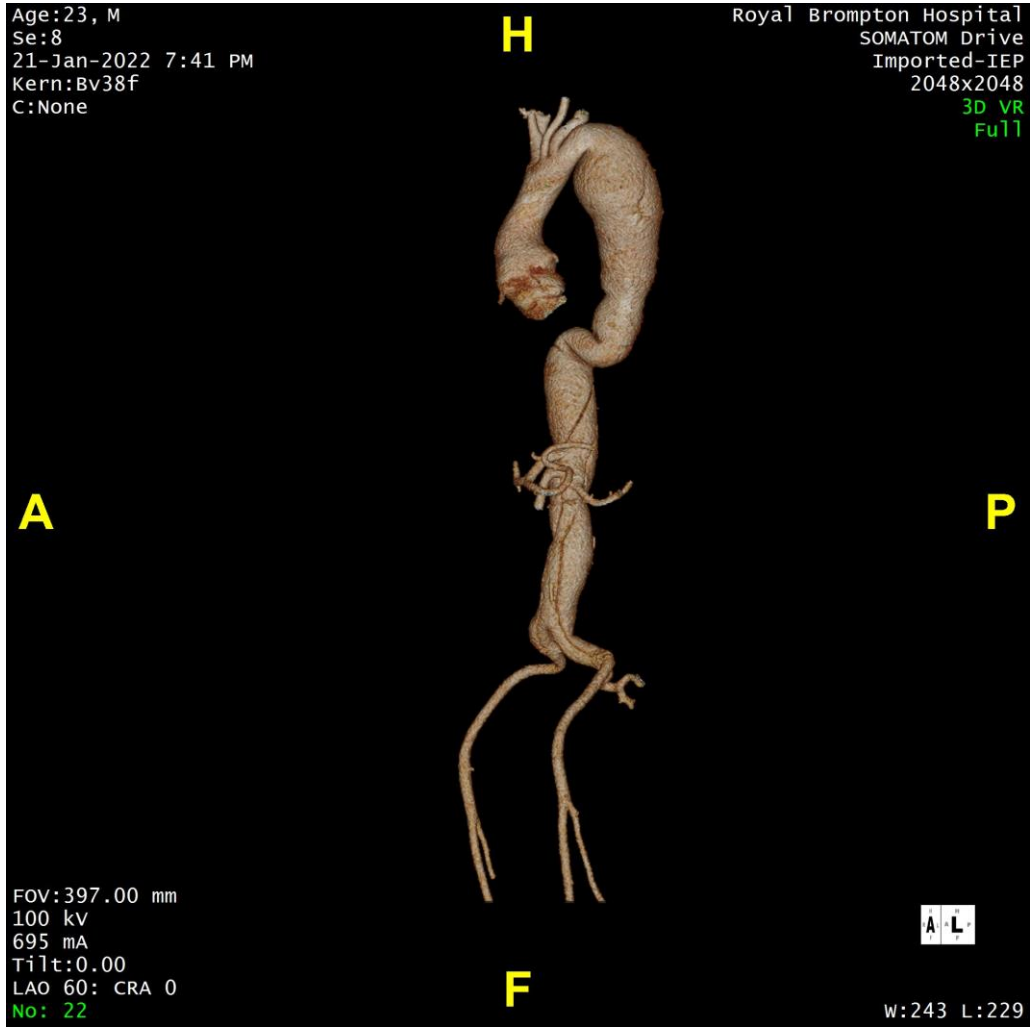
It is important that the hem of the sleeve is secured at the ventriculo-aortic junction.

Injury to the coronary ostia can be part of the learning curve.

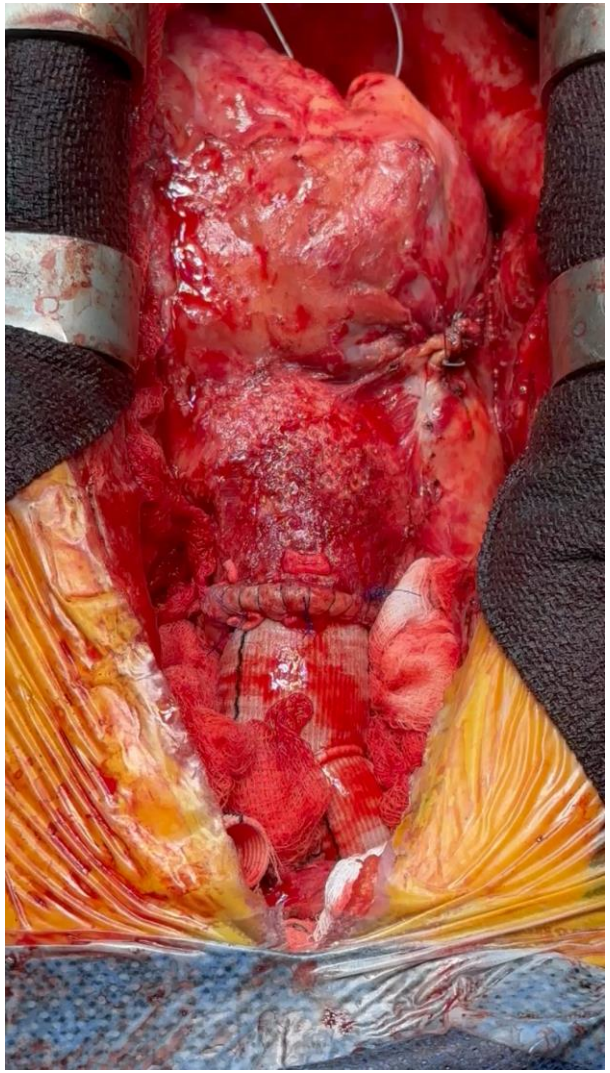
The arch appears less likely to dilate:

NVB:

The need for continued surveillance in Marfan and other genetically-driven aortopathies

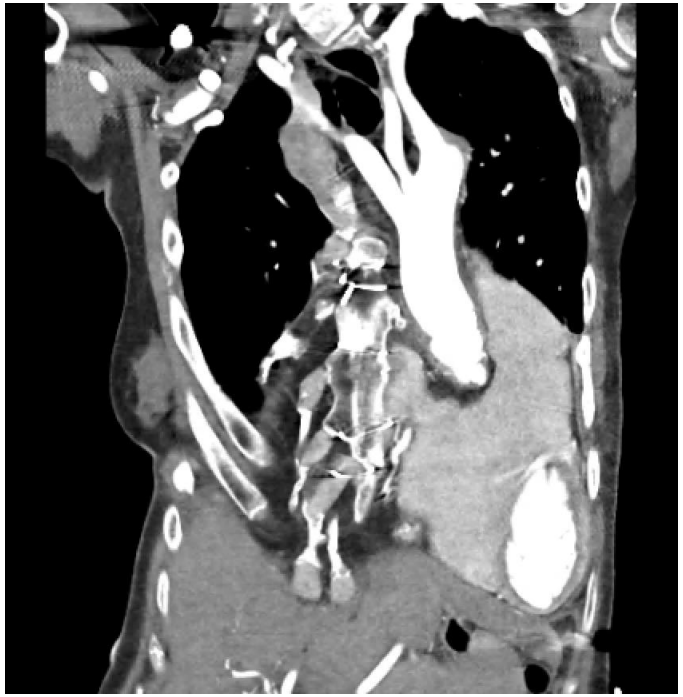


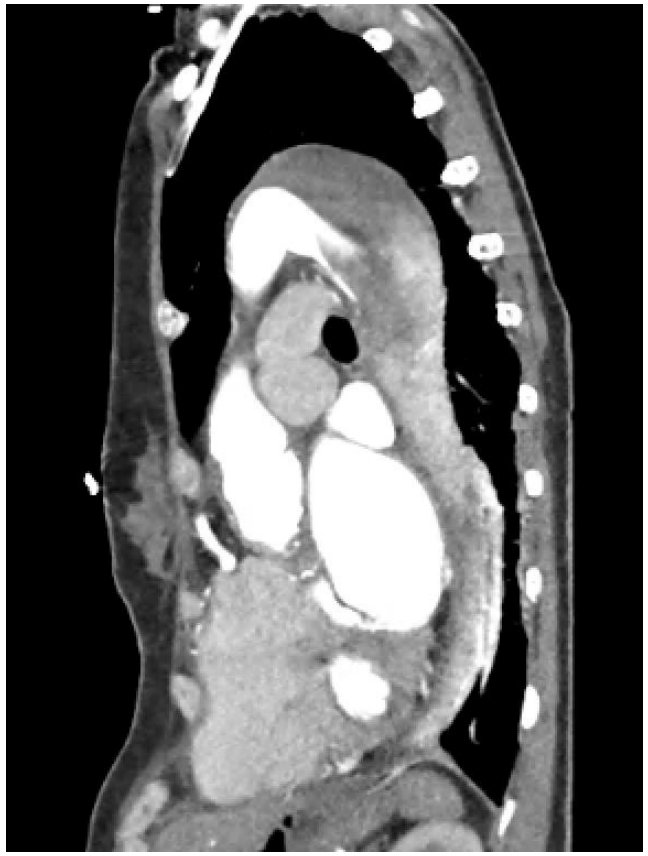
Redo Ascending and Arch + FET for PEARS

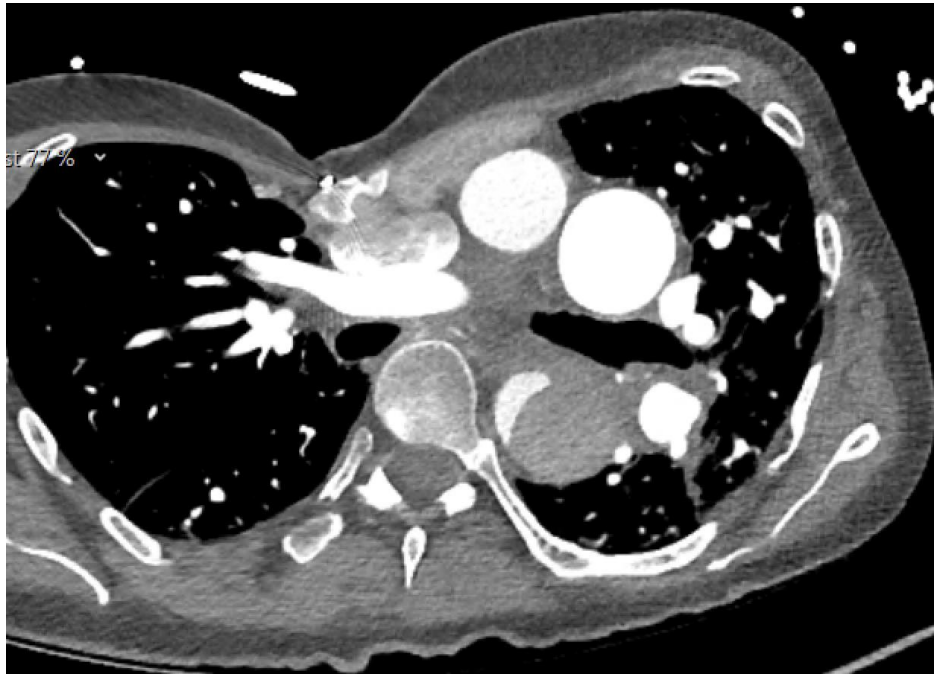


Case Study

- 51 F
- Marfan’s Syndrome
- Failed Pectus repair 1984
- Bilateral Pleurectomy for pneumothorax
- Bio-Root replacement 2002
- Severe AR due to tissue valve degeneration
- Type B Dissection Nov 2019
- Rapid expansion of DTA 6 mm in 2 months



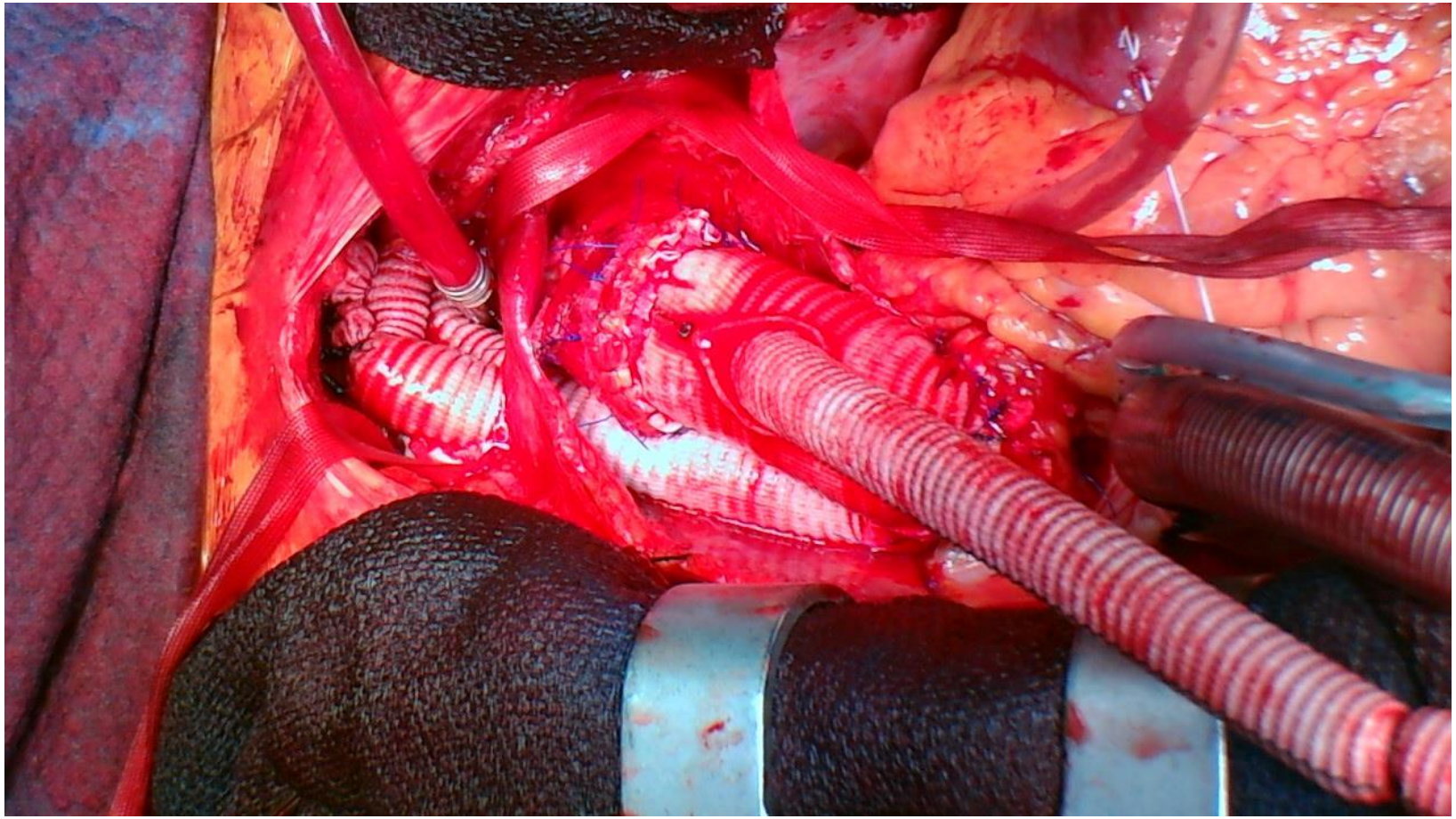




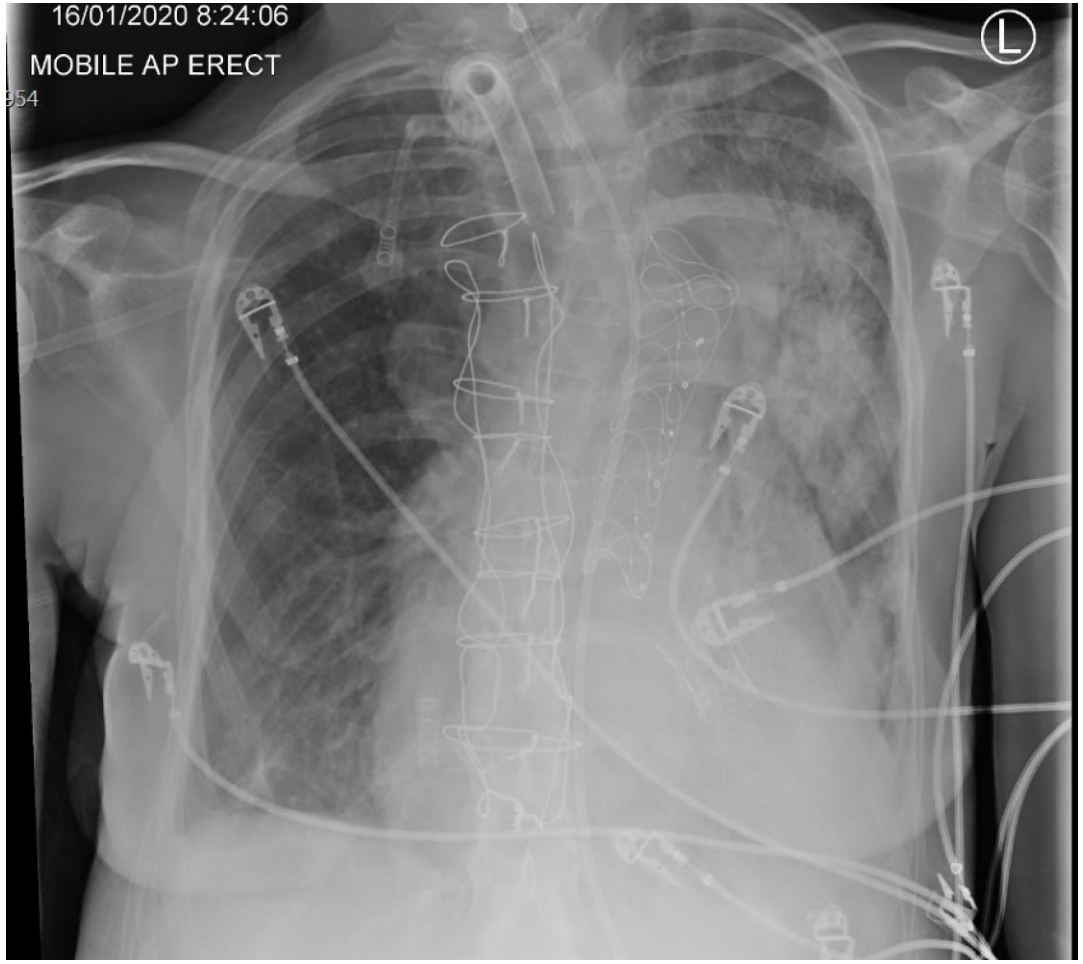
Procedure

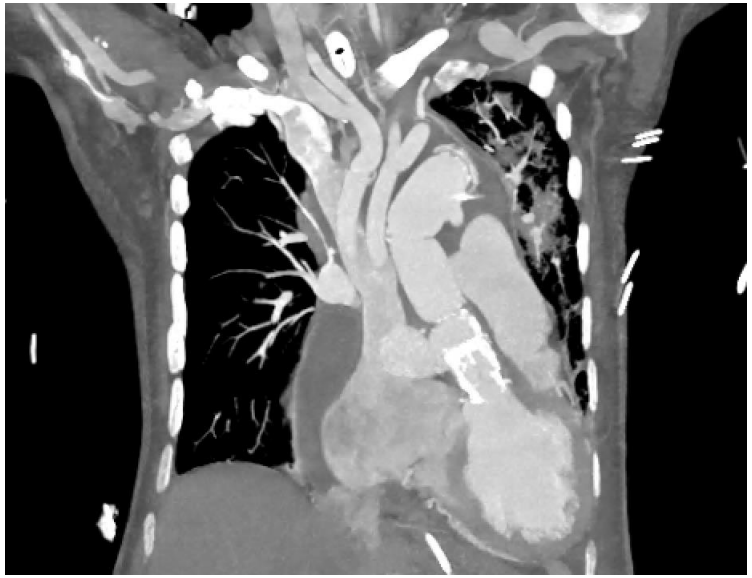
- Redo-sternotomy
- CPB – Arch and RA
- Cooled to 22^{o}C (Bilateral SACP)
- AV excised – Percival S Sutureless valve
- Debranched arch with Trifurcated graft
- Terumo Aortic Thoraflex 30mm 15 cm FET
- Zone 2 distal anastomosis

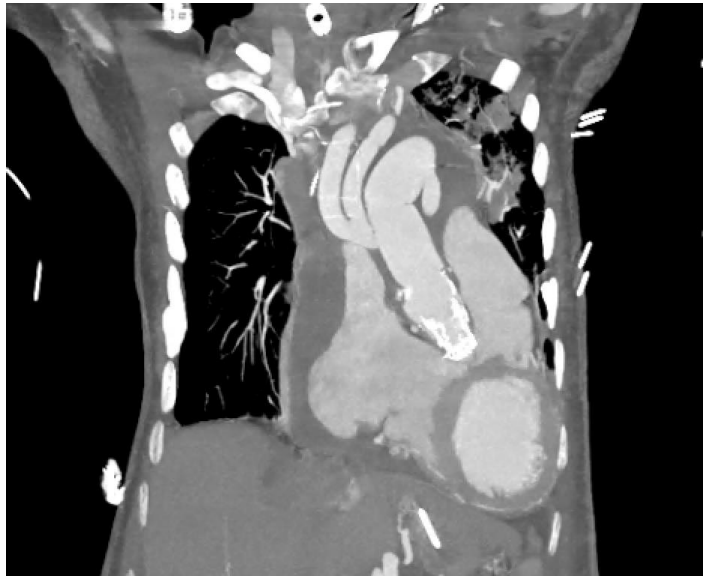
Anteflow FET + Trifurcated Graft

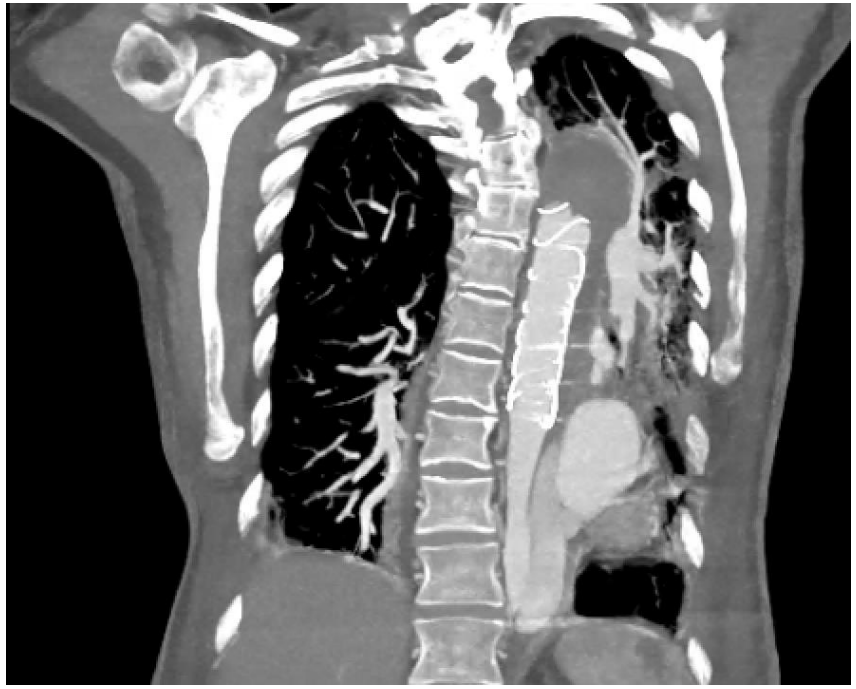


Postop









Guidelines

Recommendation for Replacement of Primary (Nondissected) Aneurysms of the Aortic Arch, Descending, and Abdominal Aorta in Patients With Marfan Syndrome		
COR	**LOE**	**Recommendation**
2a	**C-EO**	1. In patients with Marfan syndrome and a nondissected aneurysm of the aortic arch, descending thoracic aorta, or abdominal aorta of ≥5.0 cm, surgical intervention to replace the aneurysmal segment is reasonable.

Guidelines

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]

Guidelines

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.

Evidence of Endovascular Treatment of Acute and Chronic Aortic Pathology in Marfan Syndrome

From the Southern Association for Vascular Surgery

Endovascular treatment of acute and chronic aortic pathology in patients with Marfan syndrome

Alyson Lee Waterman, MD, MPH,[a] Robert Joseph Feezor, MD,[a] W. Anthony Lee, MD,[c] Philip J. Hess, MD,[b] Thomas M. Beaver, MD, MPH,[b] Tomas D. Martin, MD,[b] Thomas Stuart Huber, MD, PhD,[a] and **Adam Wayne Beck, MD,[a]** *Gainesville and Boca Raton, Fla*

Background: **In patients with Marfan syndrome, the complications of aortic degeneration, including dissection, aneurysm, and rupture represent the main cause of mortality. Although contemporary management of ascending aortic disease requires open surgical reconstruction, endovascular repair is now available for management of descending thoracic and abdominal aortic pathology (ie, thoracic endovascular aortic repair [TEVAR], endovascular aneurysm repair [EVAR]). The short- and long-term benefit of endovascular repair in Marfan patients remains largely unproven. We examine our outcomes after EVAR in this patient population.**

Methods: **All patients with a diagnosis of Marfan syndrome who were treated with TEVAR/EVAR were evaluated in a retrospective review. Perioperative, procedure-specific and patient covariate data were aggregated. Primary endpoints were overall mortality and procedural success as divided into three categories: (1) successful therapy, (2) primary failure, or (3) secondary failure.**

Results: **Between 2000 and June 2010, 16 patients were identified as having undergone 19 TEVAR/EVAR procedures. These included three emergent operations (two for acute dissection/malperfusion and one for anastomotic disruption early after open repair). All 16 patients had previously undergone at least one (range, 1-5) open operation of the ascending aorta or arch at a time interval from 33 years to 1 week prior to the index endovascular repair. During a median follow-up of 9.3 months (range, 0-46 months), there were four deaths (25%). Six patients (38%) had successful endovascular interventions. Despite early success, there was one death in this group at 1 month postintervention. Seven patients (44%) experienced primary treatment failure with five undergoing open conversion and one undergoing left subclavian coil embolization (the seventh was lost to follow-up and presented 4 months later in cardiac arrest and expired without repair). There were three deaths in the primary treatment failure group. Two patients experienced secondary treatment failure. One underwent the index TEVAR for acute dissection with malperfusion and required a subsequent TEVAR for more distal aortic pathology. He is stable without disease progression. The other patient underwent open conversion after a second EVAR with four-vessel "chimney" stent grafts and is stable with his entire native aorta having been replaced.**

Conclusions: **Aortic disease associated with Marfan syndrome is a complex clinical problem and many patients require remedial procedures. Endovascular therapy can provide a useful adjunct or bridge to open surgical treatment in selected patients. However, failure of endovascular therapy is common, and its use should be judicious with close follow-up to avoid delay if open surgical repair is required. (J Vasc Surg 2012;55:1234-41.)**

Marfan syndrome is a connective tissue disease in which the vascular manifestations of the syndrome, specifically dissection/aneurysm and subsequent rupture, are the largest source of morbidity and mortality for affected individuals. Since the late 1960s, patients with Marfan syndrome have undergone aortic root and valve replacement as described by Bentall[1] to treat ascending aortic aneurysms of 5 cm or greater. Aortic surgery for individuals with Marfan syndrome brought a much improved life expectancy and quickly became the standard of care for treating these patients.[2]

Despite operative success and improved survival following ascending aortic repair, it is well known that patients with Marfan syndrome continue to experience aortic degeneration throughout their lives, leading to remedial aortic interventions. In a study, including 675 Marfan patients undergoing aortic root replacement, Gott et al[2] reported that 14% of patients had a history of previous aortic surgery. Those patients who had undergone a previous aortic operation had a 60-day mortality that was fivefold higher than those who had not.[3] Gott's study also showed that subsequent dissection or rupture of the residual aorta was the leading cause of late death (defined as greater than 30 days). Additionally, other authors have reported that reoperation rates for Marfan patients is as high as 27%,[4] with a mortality rate of reoperation up to 31%.[5]

From the Division of Vascular Surgery, Department of Surgery,[a] and Division of Thoracic and Cardiovascular Surgery, Department of Surgery,[b] University of Florida, Gainesville; and the Christine E. Lynn Heart Institute, Boca Raton Regional Hospital, Boca Raton.[c]

Author conflict of interest: Drs Feezor, Huber, and Beck have received grant support from Cook, Medtronic, and Gore. Dr Lee is a consultant for Cook.

Presented at the Thirty-fifth Annual Meeting of the Southern Association for Vascular Surgery, January 19-22, 2011.

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doi:10.1016/j.jvs.2011.11.089

- 16 patients underwent 19 TEVAR/EVARs
- Endovascular treatment can be used as adjunct or bridge to open surgery

Marfan Syndrome

Endovascular Treatment for Type B Dissection in Marfan Syndrome: Is It Worthwhile?

Davide Pacini, MD,* Alessandro Parolari, MD, PhD,* Paolo Berretta, MD, Roberto Di Bartolomeo, MD, Francesco Alamanni, MD, and Joseph Bavaria, MD

Department of Cardiac Surgery, S. Orsola-Malpighi Hospital, University of Bologna, Bologna; Dipartimento di Scienze Cardiovascolari, Università degli Studi di Milano, Milan; Unit for Clinical Research in Atherothrombosis, Centro Cardiologico Monzino Istituto Di Ricovero e Cura a Carattere Scientifico, Milan, Italy; and Department of Cardiovascular Surgery, Hospital of the University of Pennsylvania, Philadelphia, Pennsylvania

Marfan syndrome is the most frequently inherited disorder of connective tissue and is strongly associated with aortic dilatation, dissection, and rupture; in these patients, type B dissection occurs substantially. It is not known whether stent grafting, which is now frequently used in type B aortic dissection and descending thoracic aneurysms in non-Marfan patients, is a valuable option in Marfan patients, and reports from the literature are sparse and sporadic. We performed a systematic review of studies reporting the early and late results of endovascular stent grafting in Marfan patients with type B dissection in the attempt to quantify possible benefits or potential drawbacks of this approach in these usually very sick patients. Although associated with a low operative risk (1.9%), endovascular stent grafting in patients with Marfan syndrome carries a substantial risk of early and late complications, mainly endoleaks and surgical conversions, and of death at midterm follow-up. Because these complications are relatively more frequent in patients undergoing endovascular stent grafting for chronic dissections, these data suggest caution against the routine use of endovascular stent grafting in Marfan patients.

(Ann Thorac Surg 2013;95:737–49)

REVIEW

Marfan syndrome (MFS) is the most common inherited disorder of the connective tissue. It involves multiple organ systems, and the incidence of this autosomal-dominant condition is 2 to 3 per 10,000 individuals. The diagnosis of this syndrome, even if genetic testing is available, is still made using the Ghent criteria. MFS carries an increased risk of aortic dilatation, dissection, and rupture, which are responsible for the increased mortality rate. The success of current medical and surgical treatment of MFS patients has substantially improved the life expectancy of affected patients. Pathologic dilatation of the aortic root, namely annuloaortic ectasia, is the typical vascular lesion existing in approximately 75% to 85% of patients with MFS [1]. Open surgical treatment for this proximal aortic disease is well established, with excellent long-term results [2]. However, the whole aorta in MFS is diseased, and patients can experience complications everywhere in the aorta, even beyond the primary surgical repair.

Endovascular treatment, which has been demonstrated to be effective in type B aortic dissection and descending thoracic aneurysms in non-Marfan patients, is still under scrutiny in MFS patients [3]. In these patients, such as in patients with other connective tissue disorders, the aorta is prone to dilate, and theoretically, an endovascular solution may have limited durability. In addition, the results of stent grafting in these patients are somewhat sporadic and not conclusive. The aim of this study is to perform a systematic review of studies reporting endovascular repairs in MFS patients with type B dissection to describe the outcomes and evaluate the practice of endovascular stent grafting in these delicate patients.

Material and Methods

Search Strategy

A systematic search was performed using the PubMed database to identify all studies reporting the results and evaluating the outcome of endovascular treatment of descending acute and chronic thoracic aortic dissections in patients with Marfan disease. Original articles, case series, and individual reports published in English from January 2000 to November 2011 were considered, and we looked for all the studies reporting the outcome of patients who underwent endografting for descending thoracic aorta dissection in MFS. The language of the articles was defined as reported in PubMed. Unpublished data or data reported only in an abstract were not included.

Three separate Boolean search strategies were used:

- The first search, using the search string "Marfan **and** dissection **and** endovascular or endografting **or** stent **or** stenting **or** graft **or** grafting **or** TEVAR repair" (thoracic endovascular aneurysm repair),

*These authors contributed equally to this paper.

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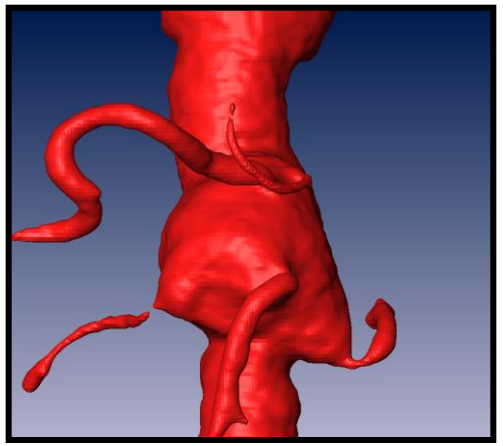
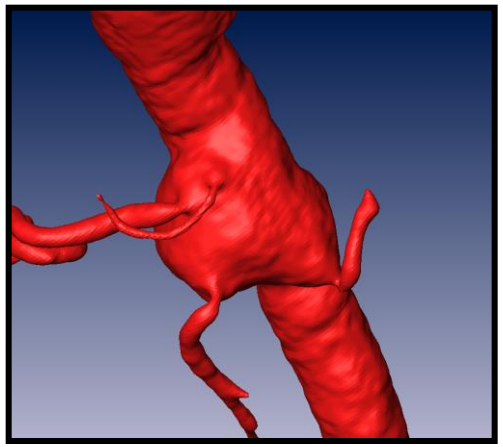
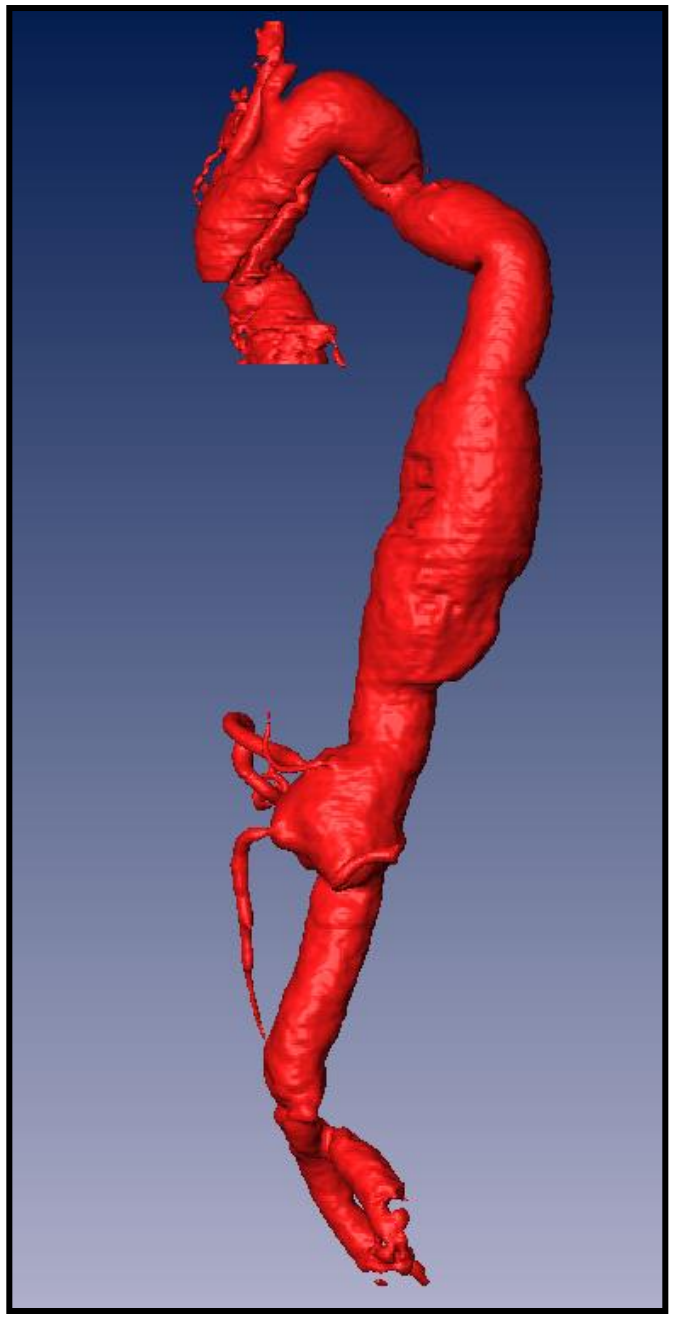
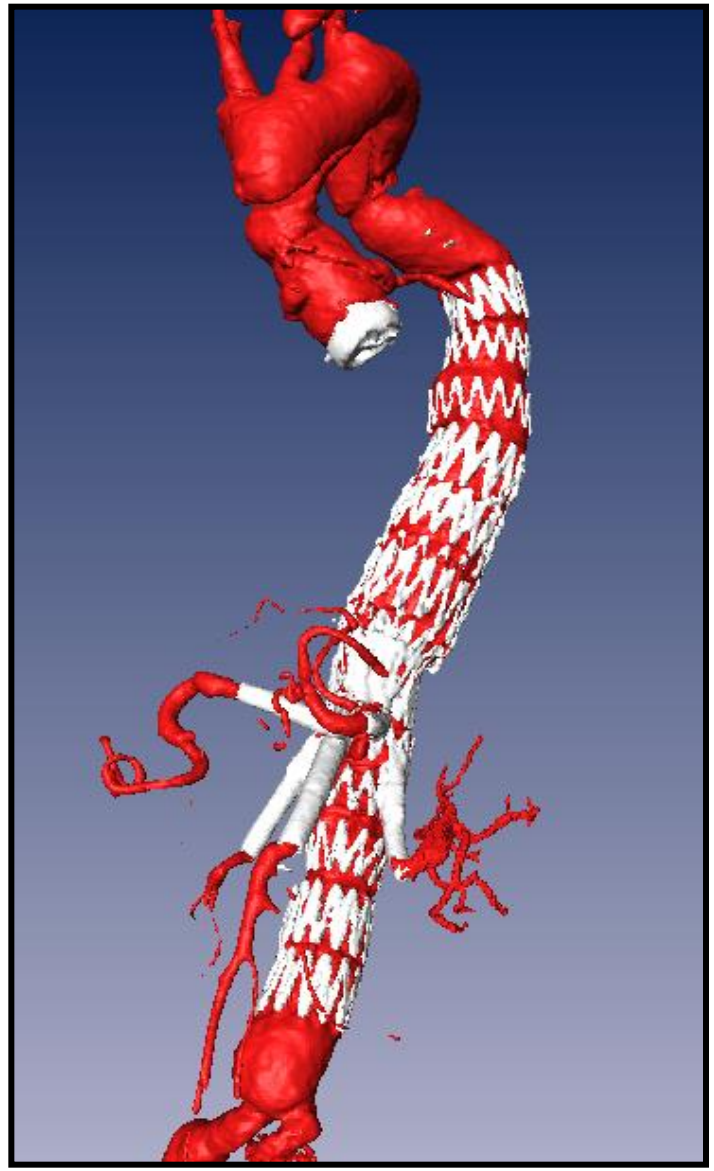
- Systematic Review
- 54 patients
- 12 papers
- 81% landing to native aorta in proximal LZ
- Death 1.9%
- CVA 1.9%
- Conversion to surgery 3.7%
- Endoleak 22%

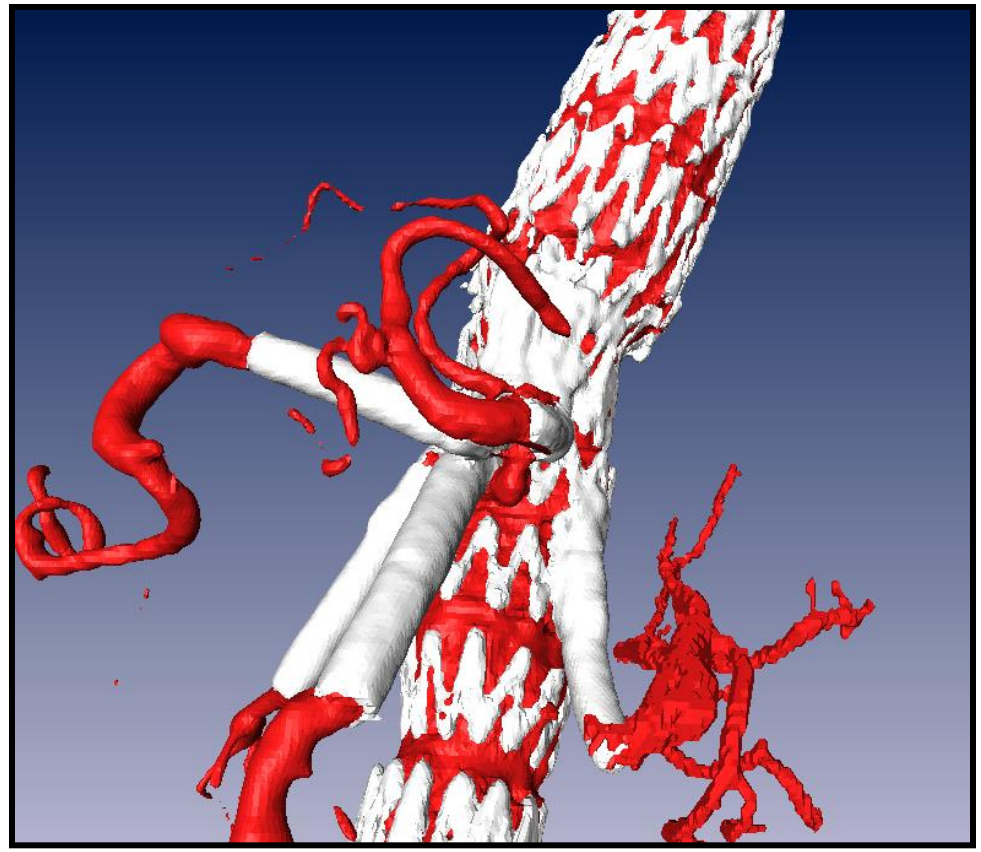
Lawyer
Marfanoid
Previous:
- Aortic root replacement
- Open repair AAA
- Open repair TAA
- Patch rupture











Ehlers-Danlos Syndrome

Endovascular Procedures in Patients With Ehlers–Danlos Syndrome: A Review of Clinical Outcomes and Iatrogenic Complications

Presented at the 21st Annual Winter Meeting of the Peripheral Vascular Surgery Society, Steamboat Springs, CO, January 28-30, 2011.

Ying Wei Lum, Benjamin S. Brooke, George J. Arnaoutakis, Timothy K. Williams, James H. Black III
Division of Vascular Surgery and Endovascular Therapy, Department of Surgery, The Johns Hopkins Hospital, Baltimore, MD

- 26 patients over 16 yrs
- 48 procedures
- Low late complications

Role of Endovascular Therapy

Expedient repair to safe life than gold standard treatment

- Rupture/ Trauma
- Exclusion of patch aneurysms
- Treatment of anastomosis failure

Key Principle of Endovascular Therapy

Graft to Graft Stenting

Problems with Connective Tissue Disorders

- Young patients
- Weak aortic wall
- Complications occur at smaller sizes
- Previous multiple interventions

Problems with Stent-Grafting in Connective Tissue Disorders

Retrograde Ascending Aortic Dissection During or After Thoracic Aortic Stent Graft Placement

Insight From the European Registry on Endovascular Aortic Repair Complications

Holger Eggebrecht, MD; Matt Thompson, MD; Hervé Rousseau, MD; Martin Czerny, MD; Lars Lönn, MD; Rajendra H. Mehta, MD, MS; Raimund Erbel, MD; on behalf of the European Registry on Endovascular Aortic Repair Complications

Background—Single-center reports have identified retrograde ascending aortic dissection (rAAD) as a potentially lethal complication of thoracic endovascular aortic repair (TEVAR).

Methods and Results—Between 1995 and 2008, 28 centers participating in the European Registry on Endovascular Aortic Repair Complications reported a total of 63 rAAD cases (incidence, 1.33%; 95% CI, 0.75 to 2.40). Eighty-one percent of patients underwent TEVAR for acute (n=26, 54%) or chronic type B dissection (n=13, 27%). Stent grafts with proximal bare springs were used in majority of patients (83%). Only 7 (15%) patients had intraoperative rAAD, with the remaining occurring during the index hospitalization (n=10, 21%) and during follow-up (n=31, 64%). Presenting symptoms included acute chest pain (n=16, 33%), syncope (n=12, 25%), and sudden death (n=9, 19%) whereas one fourth of patients were asymptomatic (n=12, 25%). Most patients underwent emergency (n=25) or elective (n=5) surgical repair. Outcome was fatal in 20 of 48 patients (42%). Causes of rAAD included the stent graft itself (60%), manipulation of guide wires/sheaths (15%), and progression of underlying aortic disease (15%).

Conclusions—The incidence of rAAD was low (1.33%) in the present analysis with high mortality (42%). Patients undergoing TEVAR for type B dissection appeared to be most prone for the occurrence of rAAD. This complication occurred not only during the index hospitalization but after discharge up to 1050 days after TEVAR. Importantly, the majority of rAAD cases were associated with the use of proximal bare spring stent grafts with direct evidence of stent graft–induced injury at surgery or necropsy in half of the patients. (***Circulation***. **2009;120[suppl 1]:S276–S281.**)

Key Words: aorta ■ TEVAR ■ stent graft ■ complications ■ dissection

Thoracic endovascular aortic repair (TEVAR) continues to be increasingly used as a less invasive treatment option for patients with thoracic aortic aneurysms and dissections, particularly in those deemed at high risk for conventional open surgical repair.[1,2] Growing technical experience and improving stent graft devices have resulted in better patient outcomes and expanded clinical indications. Available observational nonrandomized data suggest that the risk of acute complications of TEVAR, most notably paraplegia and stroke, appears to compare favorably with open surgery.[3] However, as with any new technology, TEVAR bears the risk of unusual, previously unanticipated, severe complications. One of these complications is retrograde ascending aortic dissection (rAAD), which has been highlighted as a potentially lethal complication of TEVAR in previous case series, raising significant concern about the safety of this relatively minimally invasive procedure.[4–11] However, the small number of patients in these case series (maximum, n=7 in a single series[11]) precludes reliable insight into the true incidence and patient- and procedure-related factors associated with this event and its outcomes in large number of patients undergoing TEVAR. This information may provide the opportunity for designing appropriate strategies not only to minimize this complication but also to diagnose and treat this complication early and effectively once it occurs, in the hope of improving future procedural safety and outcomes.

We analyzed data from the European Registry on Endovascular Aortic Repair Complications (EuREC, www.tevarcomplications.

From the Department of Cardiology (H.E., R.E.), West-German Heart Center Essen, University of Duisburg-Essen, Essen, Germany; the Department of Vascular Surgery (M.T.), St George's Vascular Institute, St George's Hospital NHS Trust, London, United Kingdom; the Department of Radiology (H.R.), Centre Hospitalier Universitaire, Hopital de Rangueil, Toulouse, France; the Department of Cardiothoracic Surgery (M.C.), University of Vienna Medical School, Vienna, Austria; the Department of Interventional Radiology (L.L.), Faculty of Health Sciences, Rigshospitalet University Hospital, University of Copenhagen, Copenhagen, Denmark; and Duke Clinical Research Institute (R.H.M.), Durham, NC.

Presented in part at American Heart Association Scientific Sessions 2008, November 8–12, 2008, New Orleans, La.

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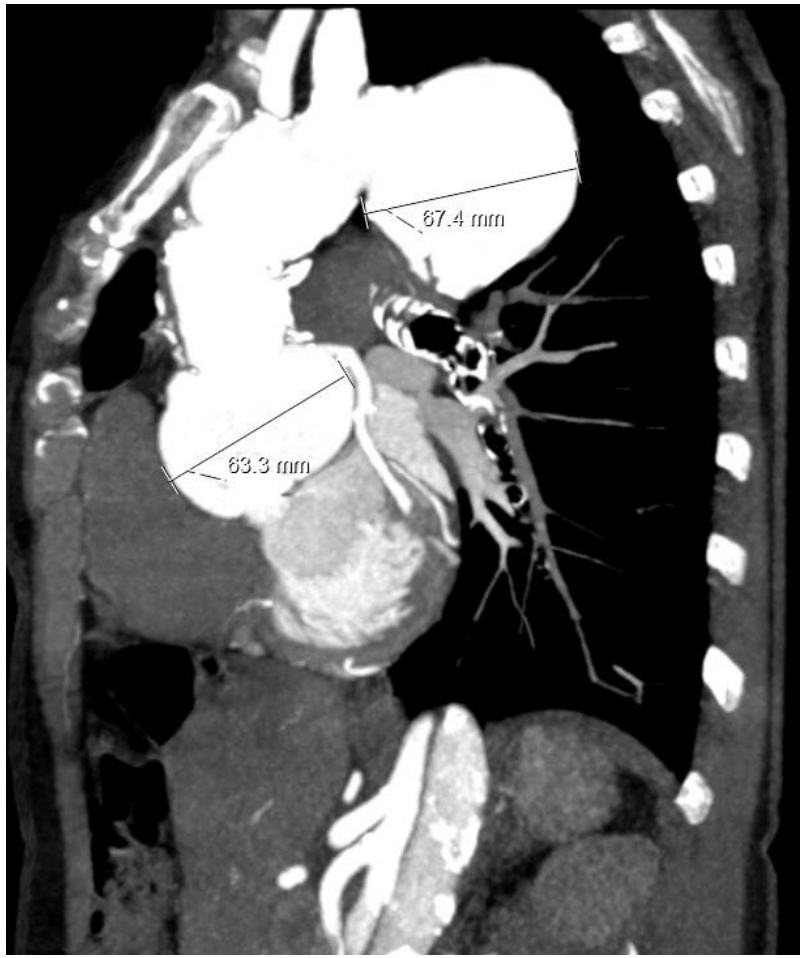
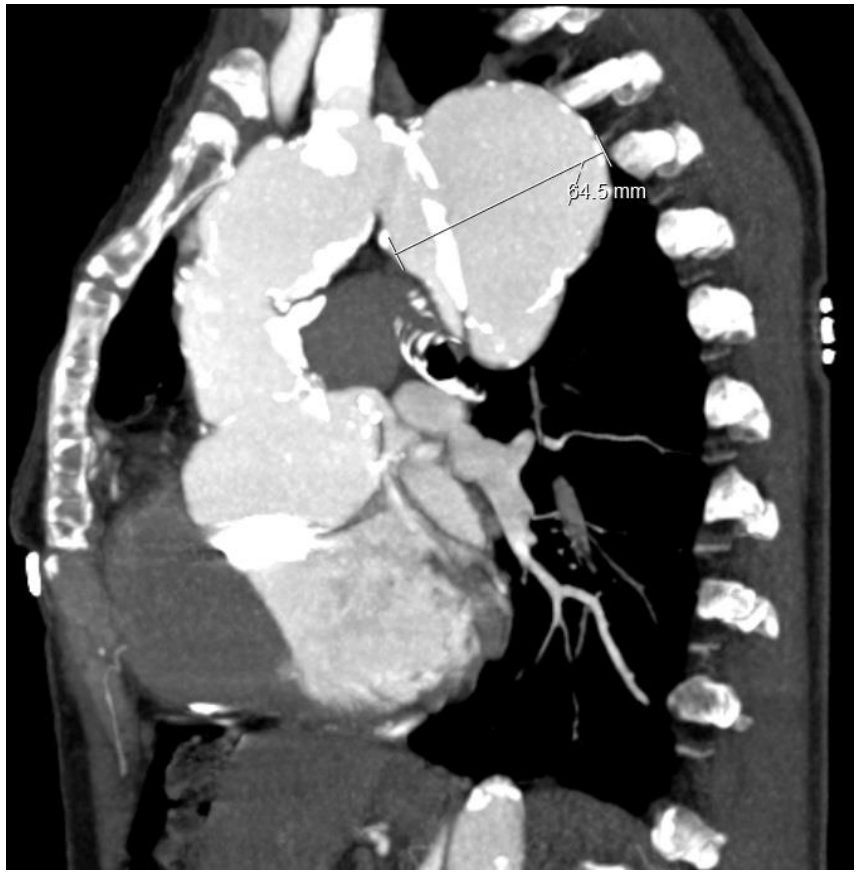
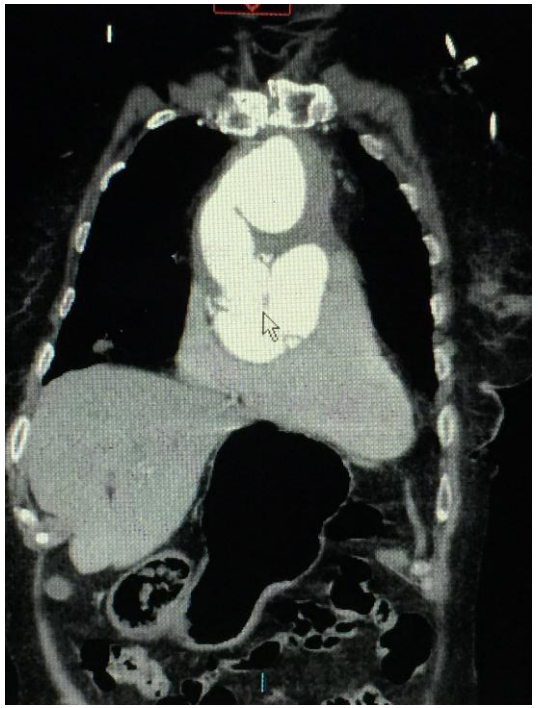
Circulation **is available at http://circ.ahajournals.org** **DOI: 10.1161/CIRCULATIONAHA.108.835926**

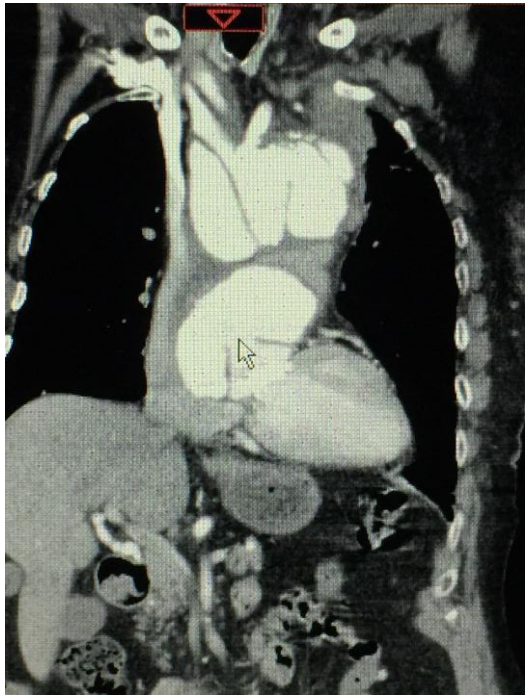
Previous Type A Repair

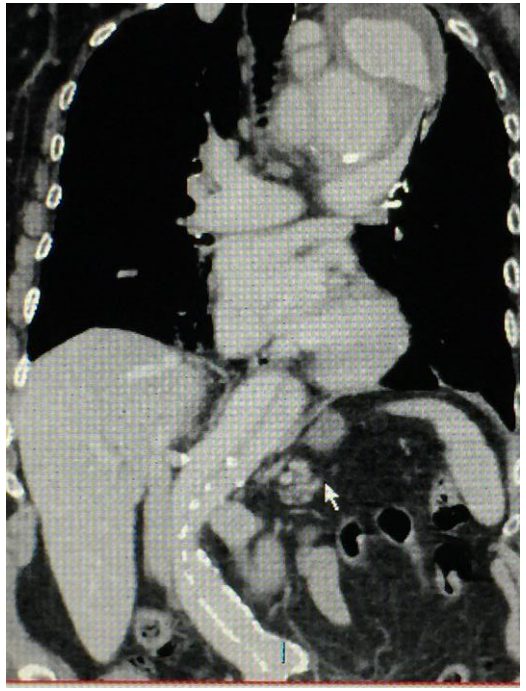


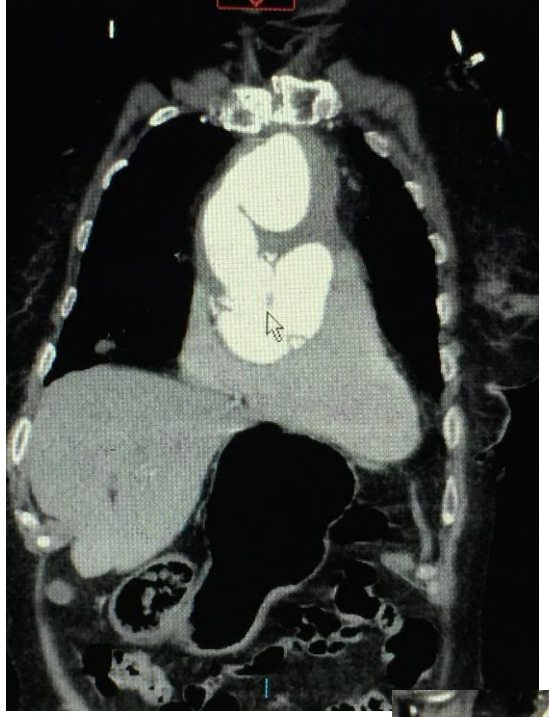
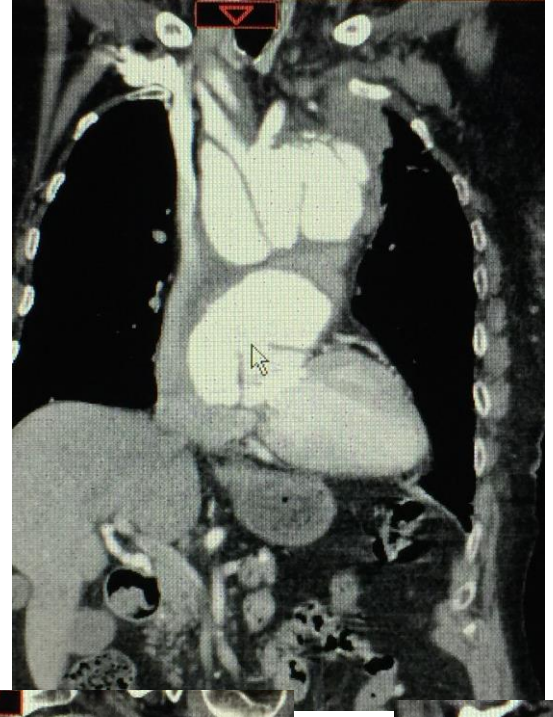
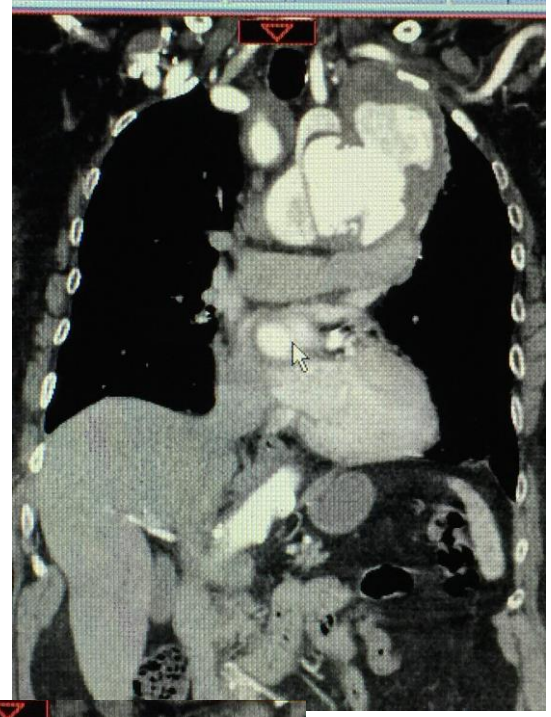
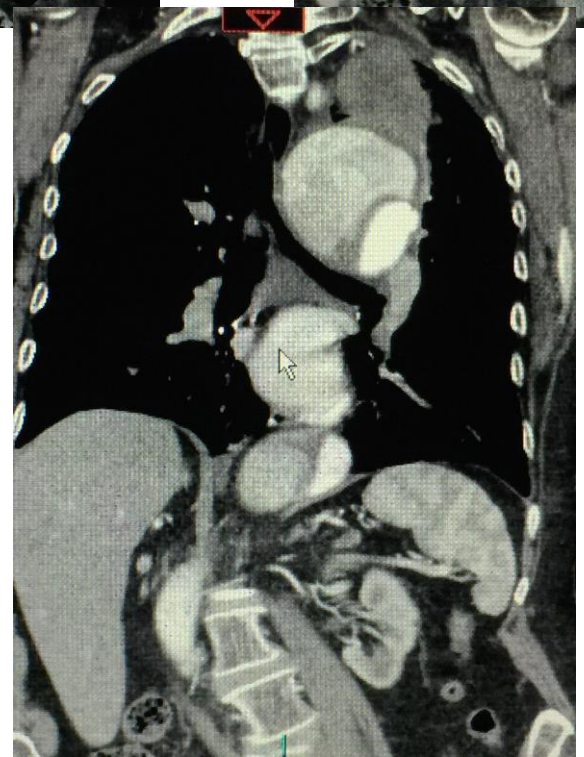
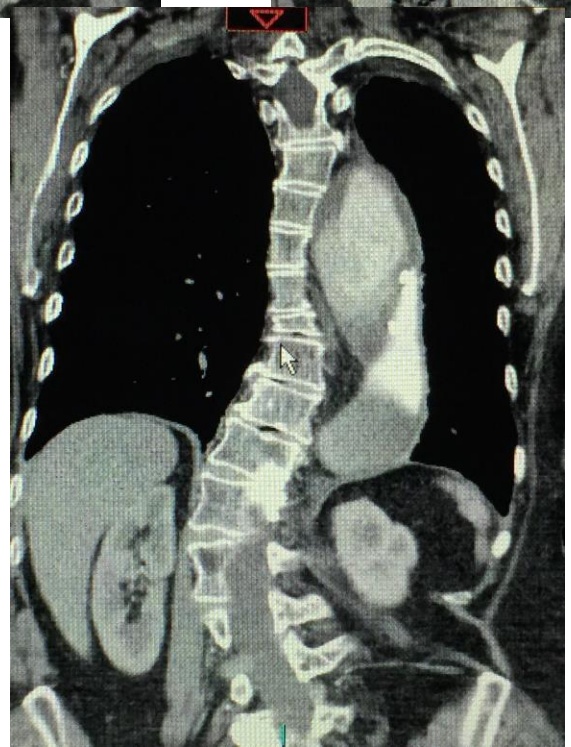


Chronic Dissection

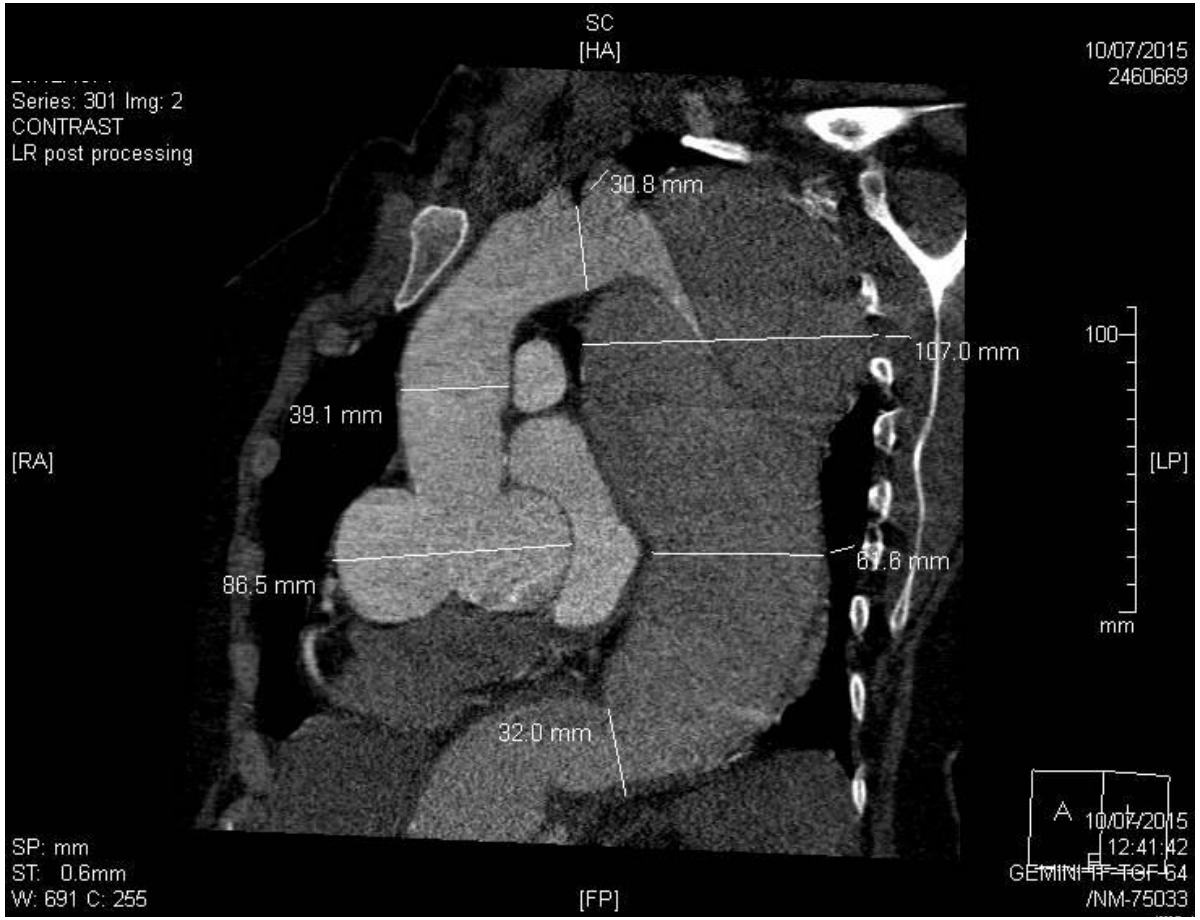






Previous AVR + AsA
Chronic Type B Dissection

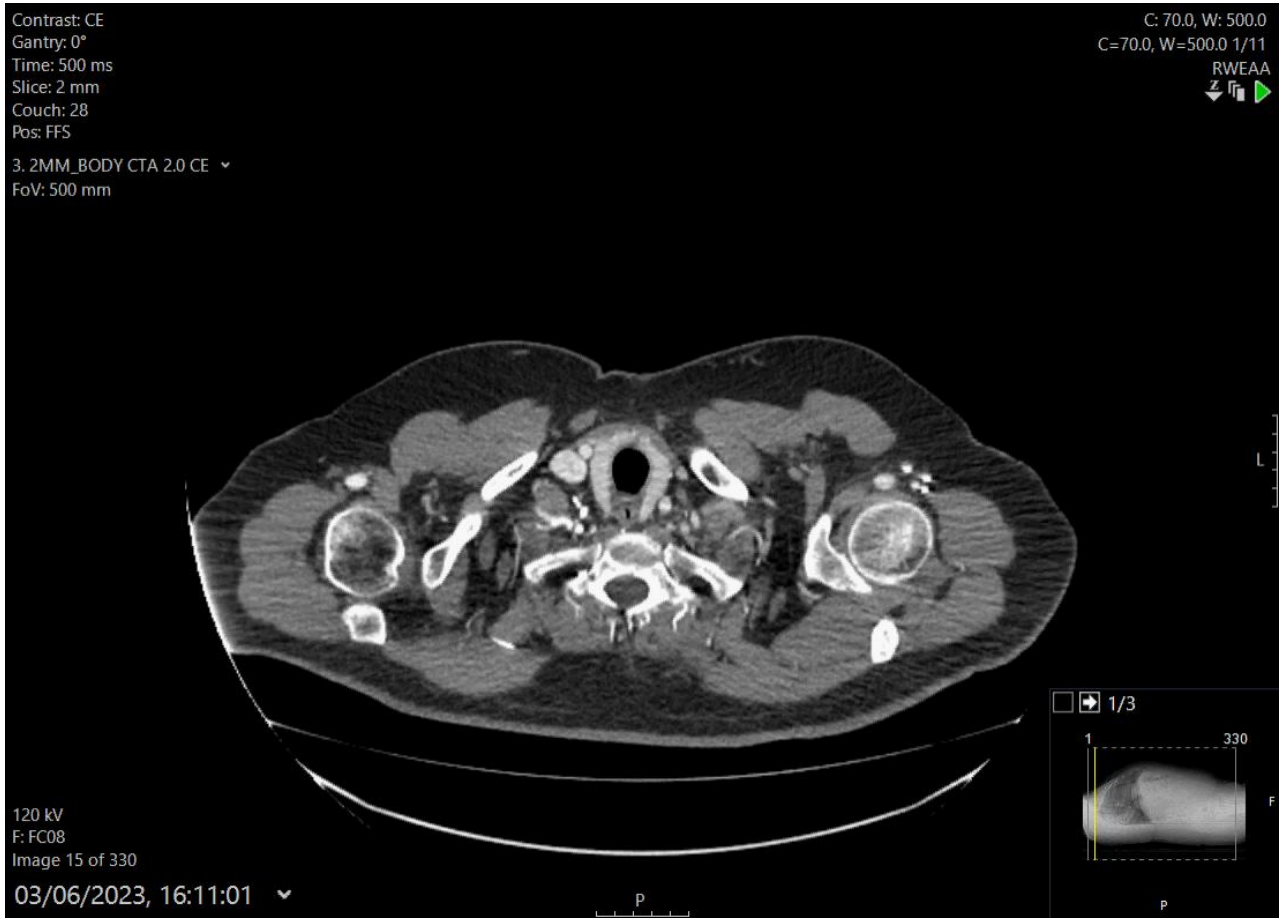


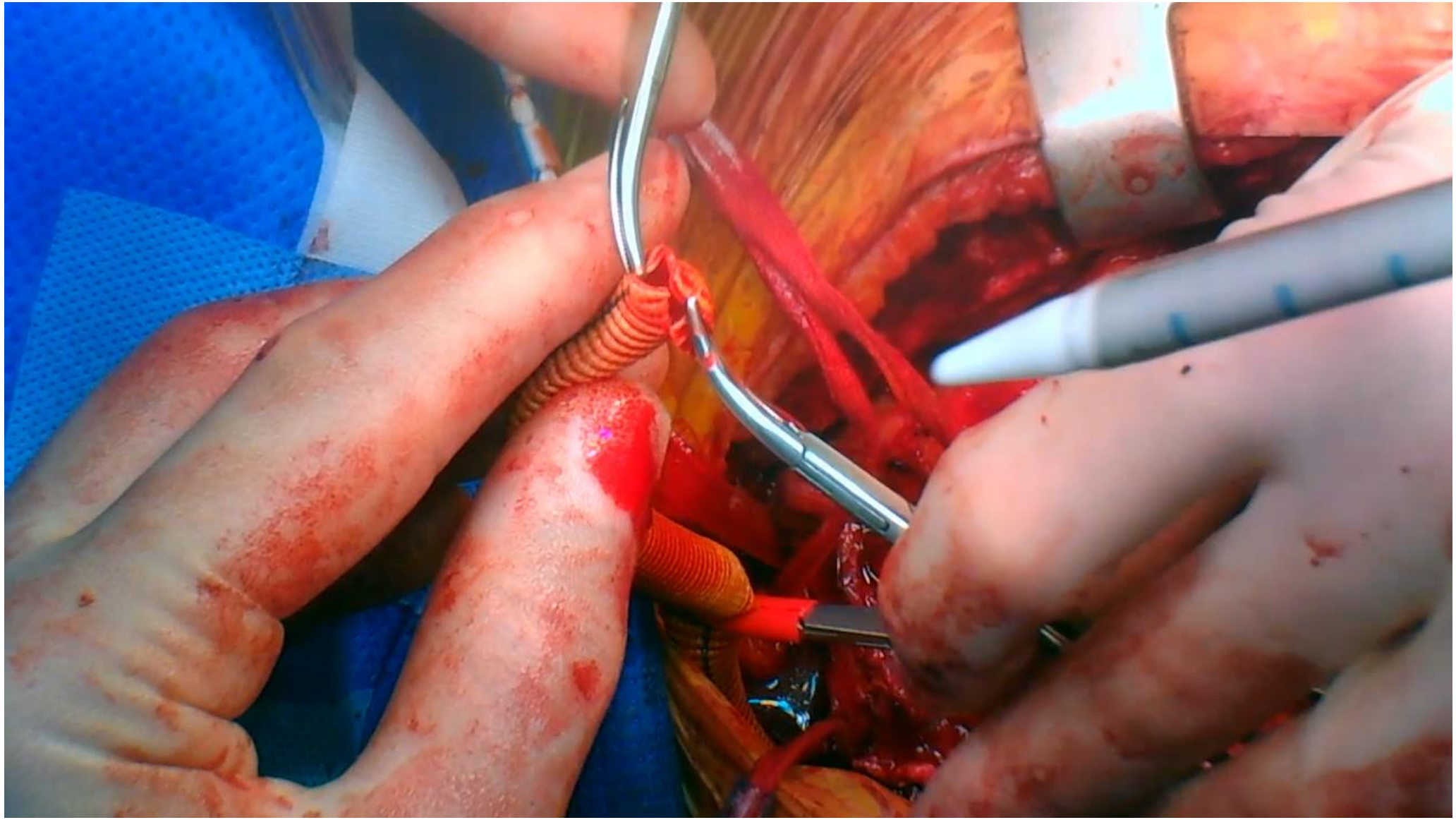
Syndromic - HTAD

Preoperative CT

56 yr old
Previous Acute Type A Dissection Repair
Arch+DTA expansion 85 mm
Normal AV function

Operative Strategy
Redo sternotomy
Innominate cannulation
HCA @ 24^0 C
SACP
Custodial





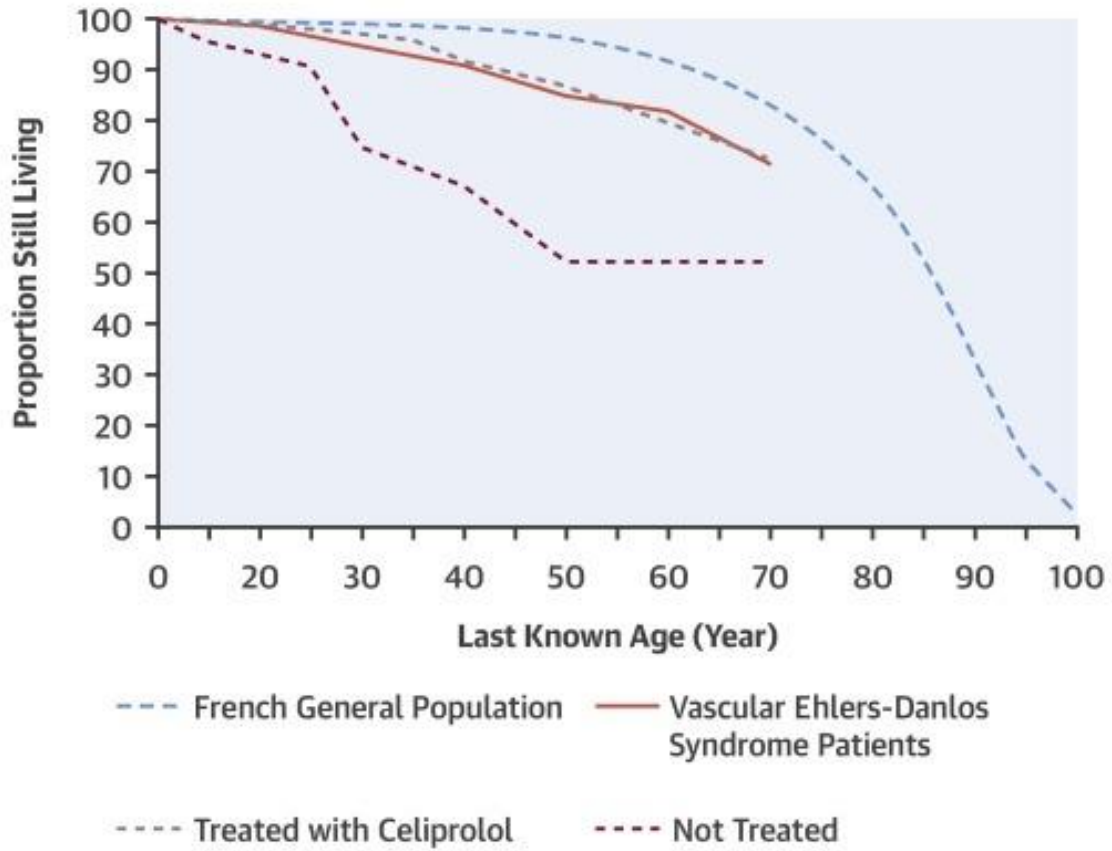
Bicuspid Aortopathy

- Incidence
- Guidelines
- Management
- Outcomes

Case Study 1

- Phil
- Redo root, arch and FET
- Turner – TAAA and root+arch

CENTRAL ILLUSTRATION: Comparing Survival of Patients With Vascular Ehlers-Danlos Syndrome in the 2015 French Population



Frank, M. et al. J Am Coll Cardiol. 2019;73(15):1948-57.

Case Presentation

- 32 yr. Male
- +ve TGF-β2 mutation
- +ve Family history of Aortic Aneurysm
- Aortic Root 5.3 cm
- AV valve – Trileaflet, No regurgitation

Barts Heart Centre

NHS
Barts Health
NHS Trust

Featured Article

Outcomes of aortic root replacement in patients with Marfan syndrome: the role of valve-sparing and valve-replacing approaches

Joseph S. Coselli[1,2,3]^, Irina V. Volguina[1,4]^, Lynna Nguyen[1,4]^, Susan Y. Green[1,4]^, Scott A. LeMaire[1,2,3,4]^, Marc R. Moon[1,2,3]^

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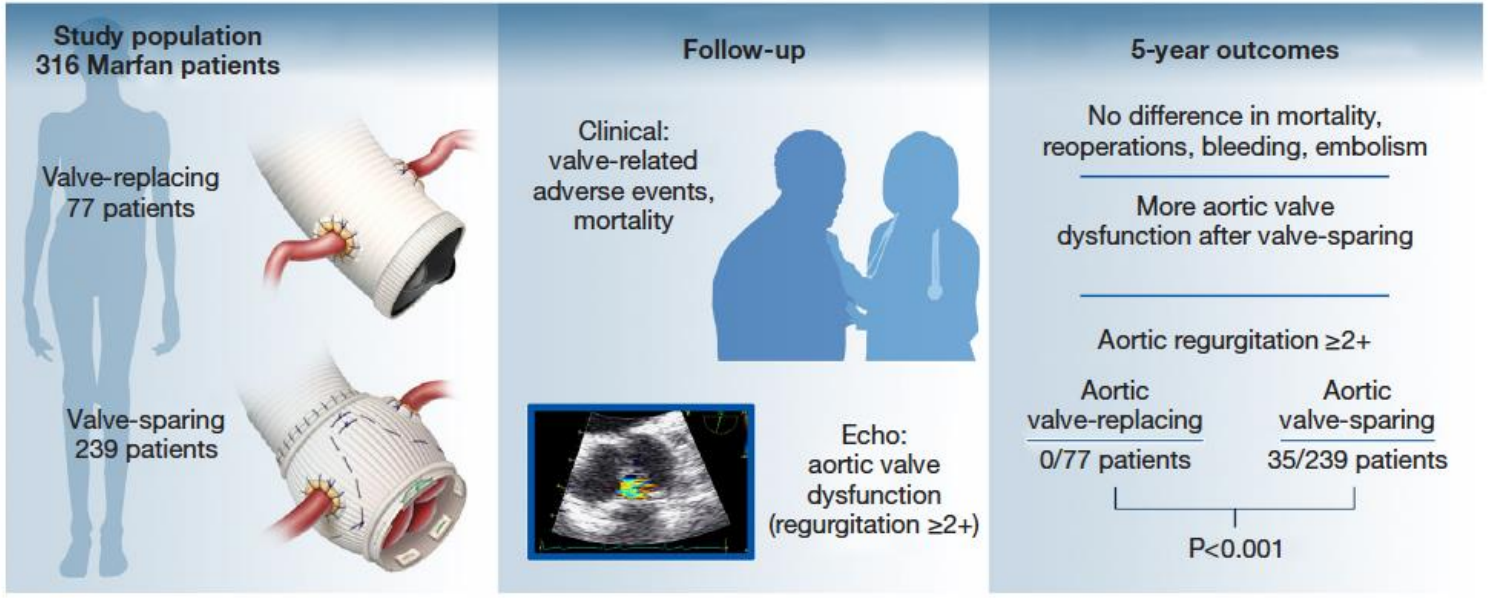
Aortic root replacement in patients with Marfan syndrome valve-replacing versus valve-sparing



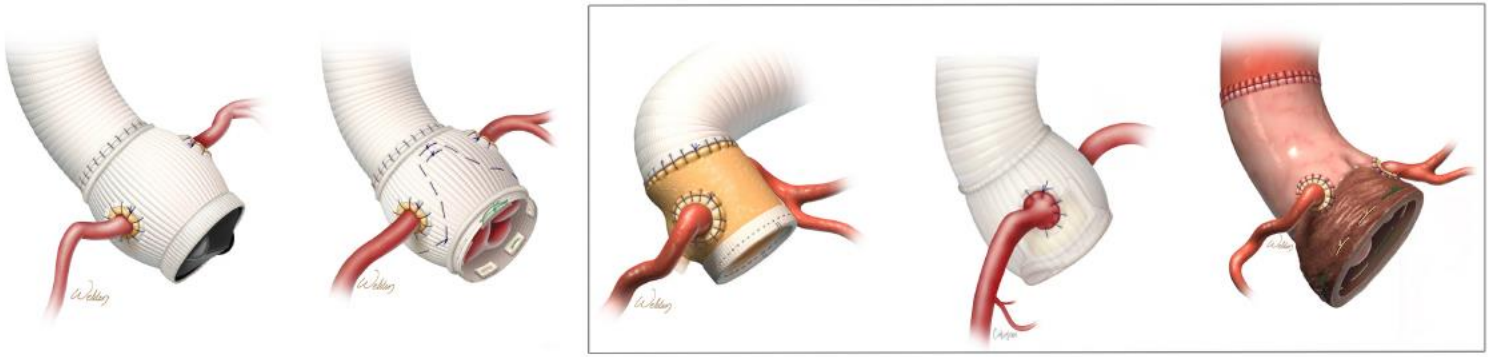


Table 1 Preoperative characteristics stratified by type of aortic root replacement

Variable	Mechanical CVG (n=113)	Valve sparing (n=62)	Bioprosthetic root (n=48)	P value
Age, years	36 [30–47]	38 [28–51]	43 [33–57]	0.3
Male	72 (64%)	38 (61%)	27 (56%)	0.7
Proximal aneurysm without dissection	83 (73%)	62 (100%)	39 (81%)	<0.001
Any aortic dissection	37 (33%)	10 (16%)	17 (35%)	0.046
Aortic dissection (proximal aortal)	30 (27%)	0	9 (19%)	<0.001
Chronic DeBakey type I	25 (22%)	0	9 (19%)	<0.001
Prior DeBakey type II	5 (4%)	0	0	0.08
Chronic DeBakey type III (distal aorta)	7 (6%)	10 (16%)	8 (17%)	0.08
Aortic root diameter, mm	55 [50–60]	50 [47–53]	52 [50–56]	<0.001
Coronary artery disease	12 (11%)	4 (7%)	9 (19%)	0.1
Cerebrovascular disease	14 (12%)	2 (3%)	4 (8%)	0.1

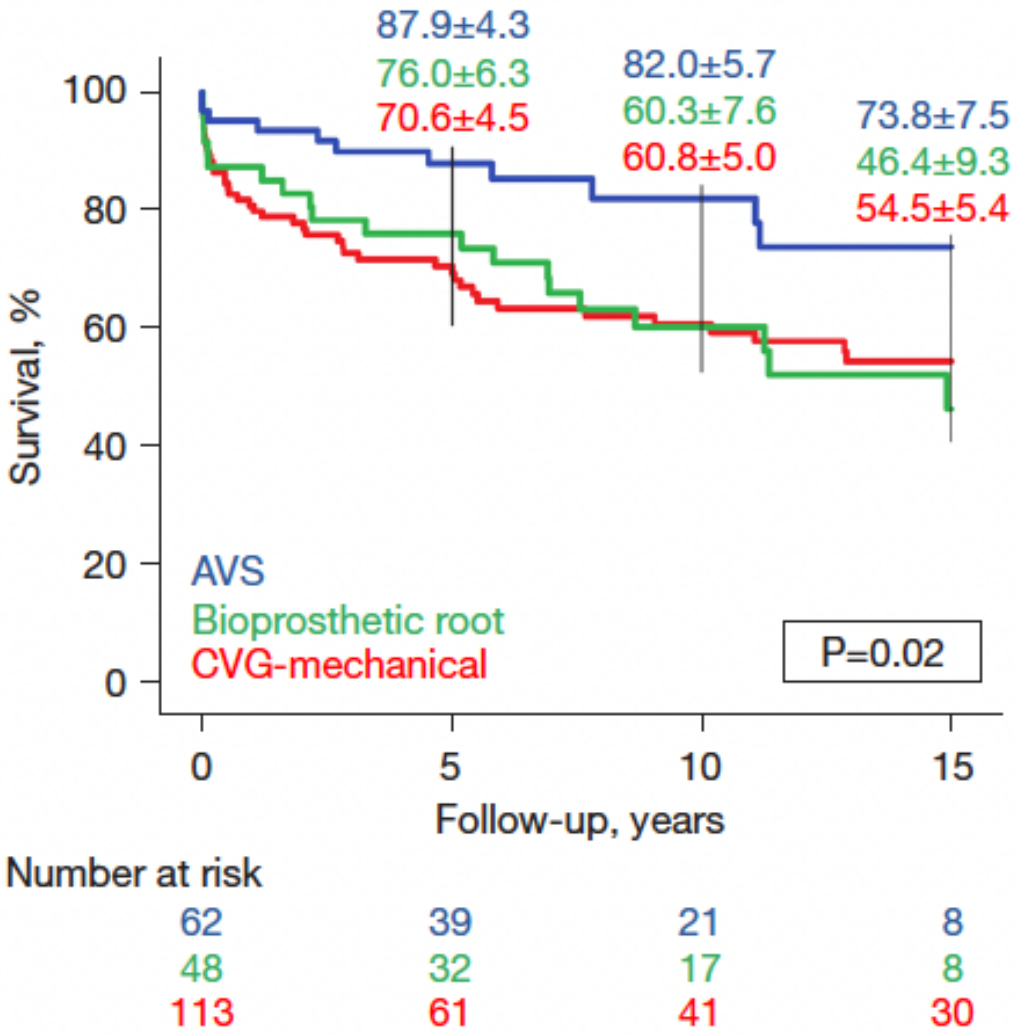
Table 2 Operative details stratified by type of aortic root replacement

Variable	Mechanical CVG (n=113)	Valve sparing (n=62)	Bioprosthetic root (n=48)	P value
Characteristics of repair				
1st aortic intervention[1]	69 (61%)	55 (89%)	29 (60%)	<0.001
2nd aortic intervention	27 (24%)	6 (10%)	13 (27%)	0.04
3rd or greater aortic intervention	17 (15%)	1 (2%)	6 (13%)	0.002
Repair before 2005	73 (65%)	14 (23%)	16 (33%)	<0.001
Reoperation (redo sternotomy)	42 (37%)	1 (2%)	11 (23%)	<0.001
Perfusion and ischemia				
CPB time, min	154 [132–186]	164 [146–213]	162 [139–193]	0.5
Hypothermic circulatory arrest	39 (35%)	22 (36%)	26 (54%)	0.052
HCA time, min	28 [20–42]	19 [15–22]	24 [17–32]	0.02
Aortic clamp time, min	89 [80–111]	112 [102–143]	99 [78–124]	<0.001
Cardiac ischemic time, min	104 [83–126]	120 [105–143]	114 [94–140]	0.01

Table 3 Early outcomes stratified by type of aortic root replacement

Variable	Mechanical CVG (n=113)	Valve sparing (n=62)	Bioprosthetic root (n=48)	P value
Operative death	5 (4%)	1 (2%)	4 (8%)	0.2
Persistent stroke	1 (1%)	0	0	0.6
Persistent renal failure‡	2 (2%)	1 (2%)	2 (4%)	0.6
Bleeding requiring reoperation	4 (4%)	3 (5%)	3 (6%)	0.7
Cardiac complications	50 (44%)	23 (37%)	22 (46%)	0.6
Arrhythmia	37 (33%)	15 (24%)	15 (31%)	0.5
Cardiac failure	10 (9%)	3 (5%)	4 (8%)	0.6
Pericardial effusion requiring drainage	10 (9%)	3 (5%)	2 (4%)	0.4
Respiratory failure	22 (20%)	6 (10%)	10 (21%)	0.2
Necessitating tracheostomy	8 (7%)	0	3 (6%)	0.1
Survivor ICU LOS, days	3 [2–5]	2 [2–4]	3 [2–5]	0.3
Survivor overall LOS, days	10 [8–13]	7 [6–10]	8 [7–13]	<0.001

Long term outcomes



Management of the aortic arch in patients with Loeys–Dietz syndrome

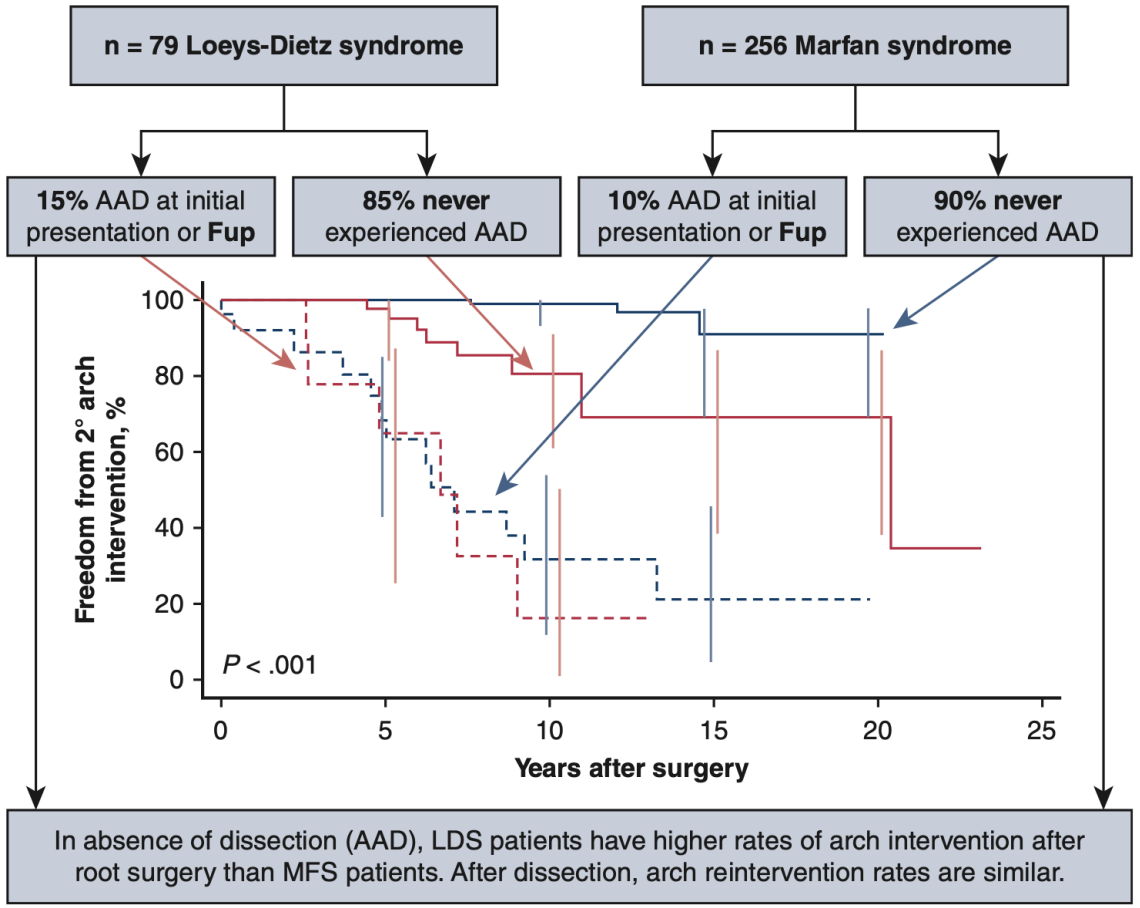
Florian S. Schoenhoff, MD,[a,b] Diane E. Alejo, BA,[a] James H. Black, MD,[c] Todd C. Crawford, MD,[a] Harry C. Dietz, MD,[d] Joshua C. Grimm, MD,[a] Jonathan T. Magruder, MD,[a] Nishant D. Patel, MD,[a] Luca A. Vricella, MD,[a] Allen Young, MD,[a] Thierry P. Carrel, MD,[b] and Duke E. Cameron, MD[a]

Central Message

In the absence of dissection, patients with LDS have a greater rate of arch intervention after root surgery than patients with MFS. After dissection, arch reintervention rates are similar in the 2 groups.

Perspective

In patients with LDS with acute type A aortic dissection treated with a tear-oriented approach, the need for subsequent arch interventions is not greater than in patients with MFS as long as hemiarch replacement is performed. Patients with LDS undergoing elective root repair have an increased risk for subsequent arch interventions, so consideration should be given to complete removal of the distal ascending aorta.



General Considerations

- High index of suspicion for diagnosis as well as associated issues
- Genetic testing and counselling
- Life time management
- Education and training for the patients, family and healthcare workers

Results of Open Surgical Repair in Patients With Marfan Syndrome and Distal Aortic Dissection

Joseph S. Coselli, MD, Susan Y. Green, MPH, Matt D. Price, MS, Jonathan A. Hash, BS, Yafei Ouyang, BS, Irina V. Volguina, PhD, Ourania Preventza, MD, Kim I. de la Cruz, MD, and Scott A. LeMaire, MD

Division of Cardiothoracic Surgery, Michael E. DeBakey Department of Surgery, Baylor College of Medicine, Houston; Cardiovascular Research Institute, Baylor College of Medicine, Houston; and Department of Cardiovascular Surgery, Texas Heart Institute, Houston, Texas

Jan 2004 – Jan 2014

127 patients

DeBakey I	73
DeBakey III	54

Ann Thorac Surg
2016;101:2193–201

Table 1. Preoperative Characteristics Stratified by DeBakey Classification

Characteristics	All (n = 127)	DeBakey I (n = 73)	DeBakey III (n = 54)	*p* Value
Age at onset of dissection, years, mean ± SD	36.3 ± 10.7	36.2 ± 10.2	36.3 ± 11.5	0.9
Age at TAAA repair, years, mean ± SD	43.4 ± 12.5	45.1 ± 11.1	41.1 ± 14.0	0.08
Time from dissection to distal aortic repair, years, no. [IQR] (n = 126)	5.2 [2.1–9.8]	6.5 [3.5–13.9]	2.9 [0.6–6.0]	<0.001
Men, n (%)	72 (57)	43 (59)	29 (54)	0.6
Confirmed Marfan syndrome (Ghent criteria), n (%)	67 (53)	34 (47)	33 (61)	0.1
Acute or subacute aortic dissection, n (%)	11 (9)	4 (6)	7 (13)	0.2
Additional distal aortic dissection, n (%)	11 (9)	8 (11)	3 (6)	0.4
Additional proximal aortic dissection,[a] n (%)	10 (8)	0	10 (19)	0.0001

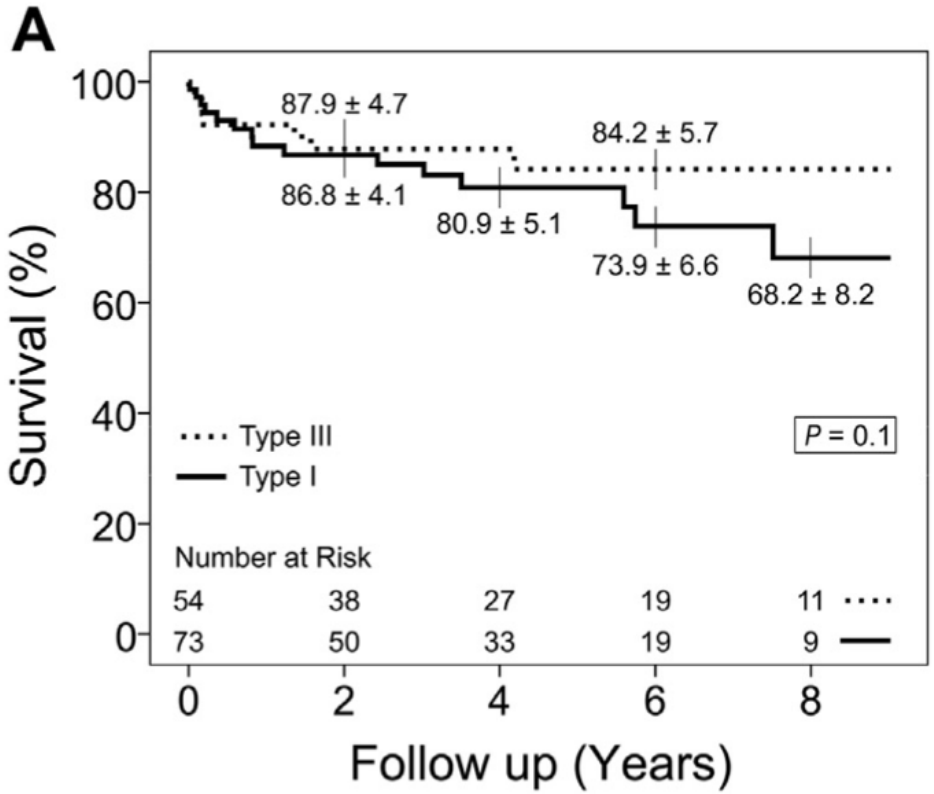
Ann Thorac Surg
2016;101:2193–201

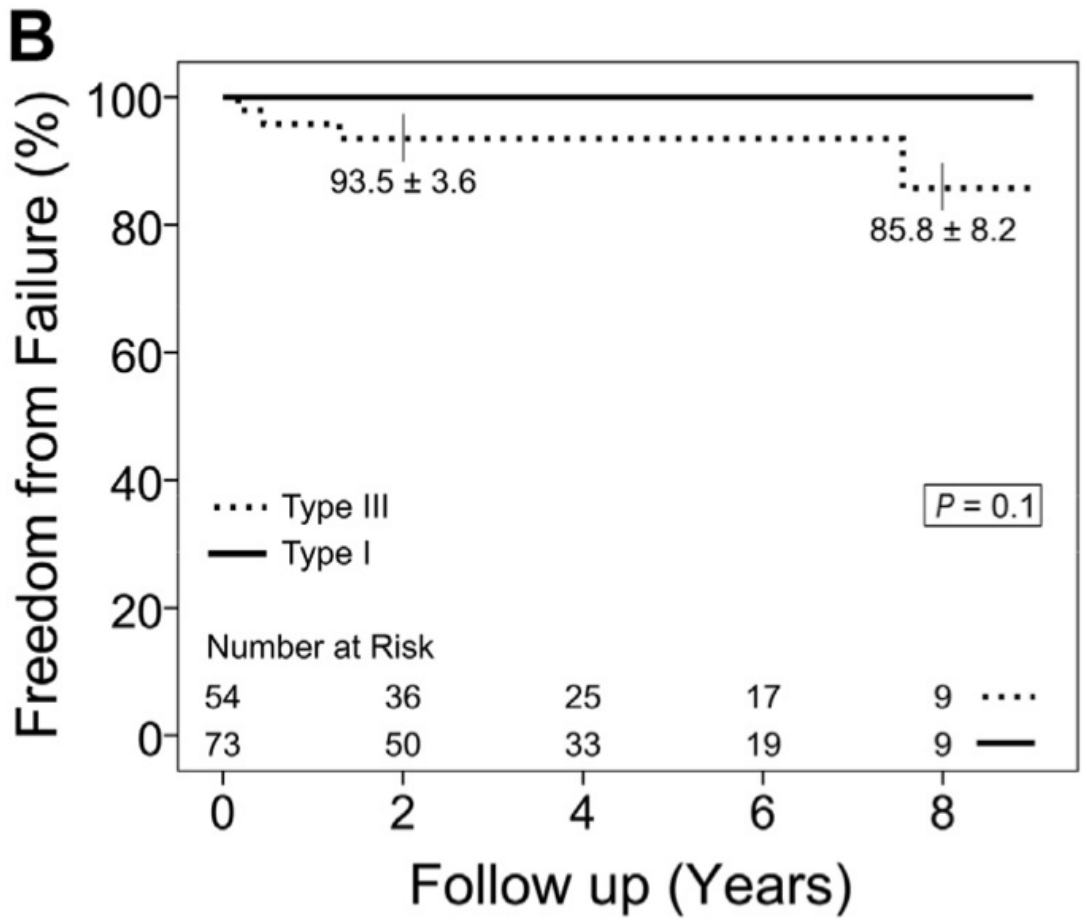
Table 2. Operative Details Stratified by DeBakey Classification

Operative Details	All (n = 127)	DeBakey I (n = 73)	DeBakey III (n = 54)	*p* Value
Urgency of operation				
Elective	98 (77)	59 (81)	39 (72)	0.3
Urgent	22 (17)	10 (14)	12 (22)	0.2
Emergent	7 (6)	4 (6)	3 (6)	1.0
Aortic repair details				
Extent of thoracoabdominal aortic aneurysm repair				
Extent I	27 (21)	12 (16)	15 (28)	0.1
Extent II	66 (52)	44 (60)	22 (41)	0.03
Extent III	26 (21)	13 (18)	13 (24)	0.4
Extent IV	8 (6)	4 (6)	4 (7)	0.7
Redo thoracotomy	25 (20)	14 (19)	11 (20)	0.9
Extraction of endograft	7 (6)	3 (4)	4 (7)	0.5
Reverse elephant trunk	14 (11)	10 (14)	4 (7)	0.4
Elephant trunk completion repair	12 (9)	12 (16)	0	0.01

Table 3. Early Outcomes Stratified by DeBakey Classification

Outcomes	All (n = 127)	DeBakey I (n = 73)	DeBakey III (n = 54)	*p* Value
Adverse event[a]	8 (6)	6 (8)	2 (4)	0.5
Operative death	5 (4)	3 (4)	2 (4)	1.0
In-hospital	5 (4)	3 (4)	2 (4)	1.0
30-day	2 (2)	1 (1)	1 (2)	1.0
Stroke	1 (1)	1 (1)	0	1.0
Permanent[b]	0	0	0	…
Spinal cord deficits	5 (4)	2 (3)	3 (6)	0.7
Permanent paraplegia[b]	1 (1)	1 (1)	0	1.0
Permanent paraparesis[b]	1 (1)	1 (1)	0	1.0
Acute renal dysfunction	12 (9)	9 (12)	3 (6)	0.2
Permanent renal failure necessitating dialysis[b]	6 (5)	5 (7)	1 (2)	0.2
Survival with life-altering complication[c]	3 (2)	3 (2)	0	0.3





Ann Thorac Surg
2016;101:2193–201

Special Considerations

- Timing for treatment
- Surgery vs Intervention
- Surgical technical considerations
- Management of expectation

Life Time Management

- Diagnosis and education
- Family screening and counselling
- Surveillance – Imaging and symptoms
- Lifestyle modification
- Family planning
- Multiprofessional team management

Conclusions

- Aortopathy patient population is increasing over last 3 decades
- Life expectancy of this group of patients is improving due to advancement in diagnosis and timely management with multimodality patient tailored approach in treatments
- The good quality service with specialization in aortopathy, dedicated clinical pathway, MDT based management and comprehensive life time management will improve survival and quality of life of this patient group