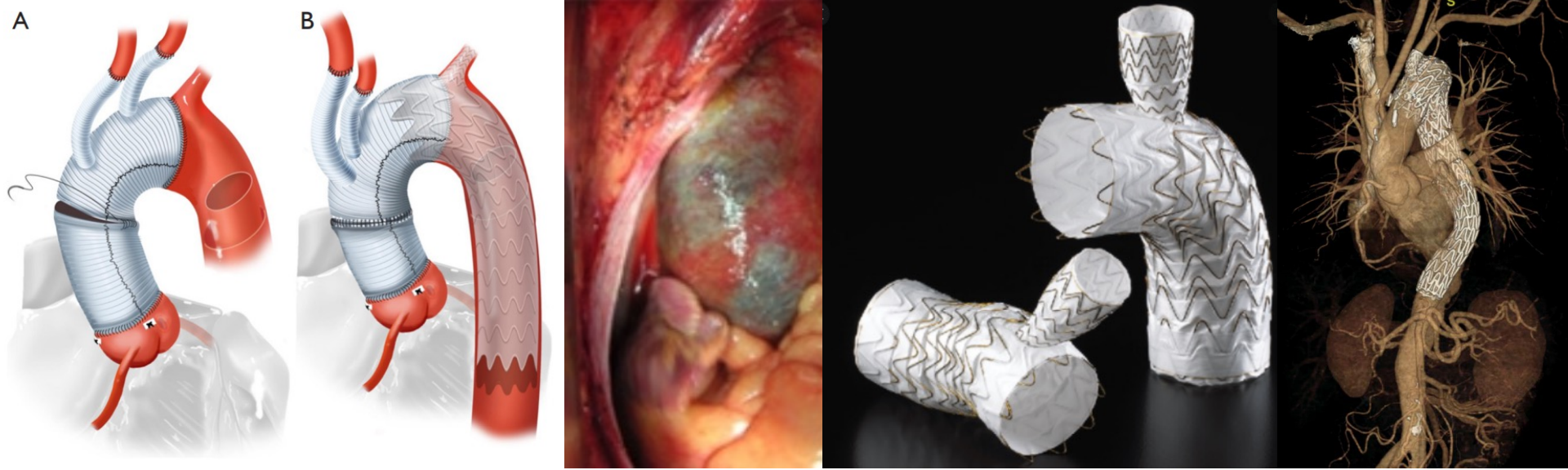




Surgical Strategies for Arch Repair in Genetic Aortopathies

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Disclosures

- ▶ None



Aortopathy watershed aortic events



Root aneurysm

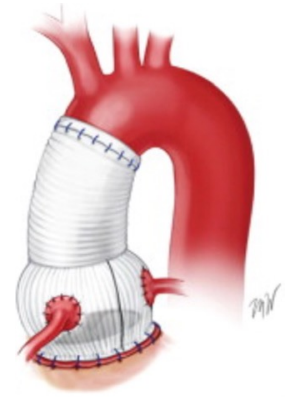
- Drives decision for elective aortic root replacement (ARR)
- TAAD can be minimized with medical therapy and ARR



Type B Dissection

- Occurs at normal diameters
- Risk factors: pregnancy, previous ARR, tortuosity
- Often drives need for TAAA repair

- Our understanding of genetic aortopathy has increased dramatically
- Heterogenous group of conditions and size thresholds for surgery



MYH11, MYLK, PRKG1, LOX

- Cause aggressive aortopathy
- Aortic events can happen at normal diameters
- Individualized approach

ACTA2

- No high-risk features

Loeys Dietz Syndrome

- TGFBR1, TGFBR2 no high-risk features^b
- SMAD3 variants

Marfan Syndrome

- High-risk features^a

Marfan Syndrome

- FBN1 mutations
- No high-risk features^a

Loeys Dietz Syndrome

- TGFBR1, TGFBR2 with high-risk features^b

4.0 cm

4.2 cm

4.5 cm

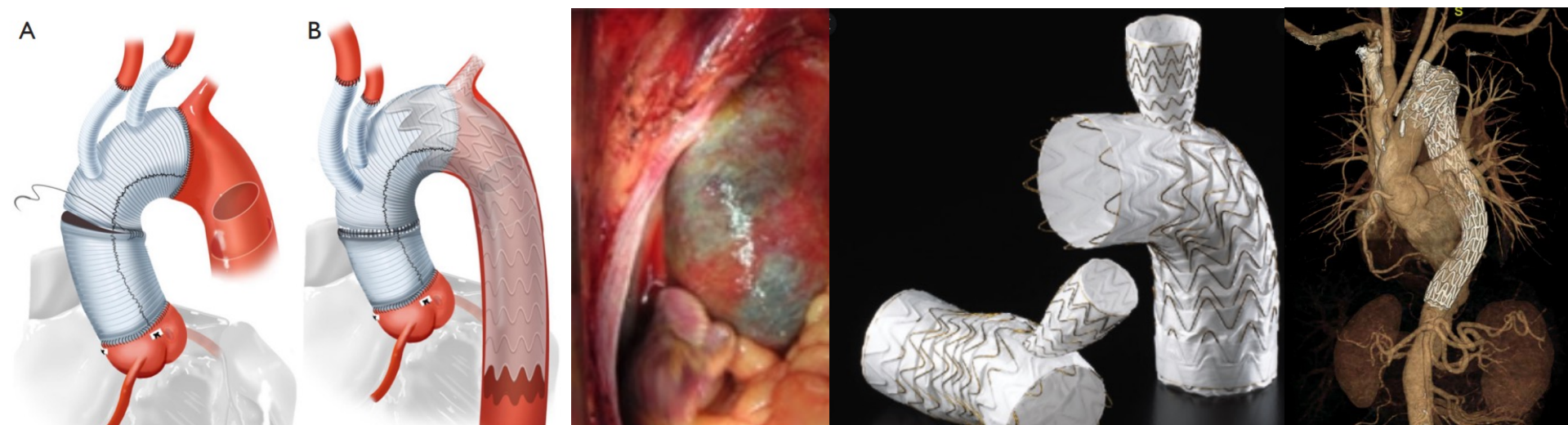
5.0 cm

a: Family history of aortic dissection, growth rate > 0.3 cm/y, severe vertebral artery tortuosity, combined root/ascending phenotype

b: Family history of aortic dissection, growth rate > 0.3 cm/y, severe extra-aortic features

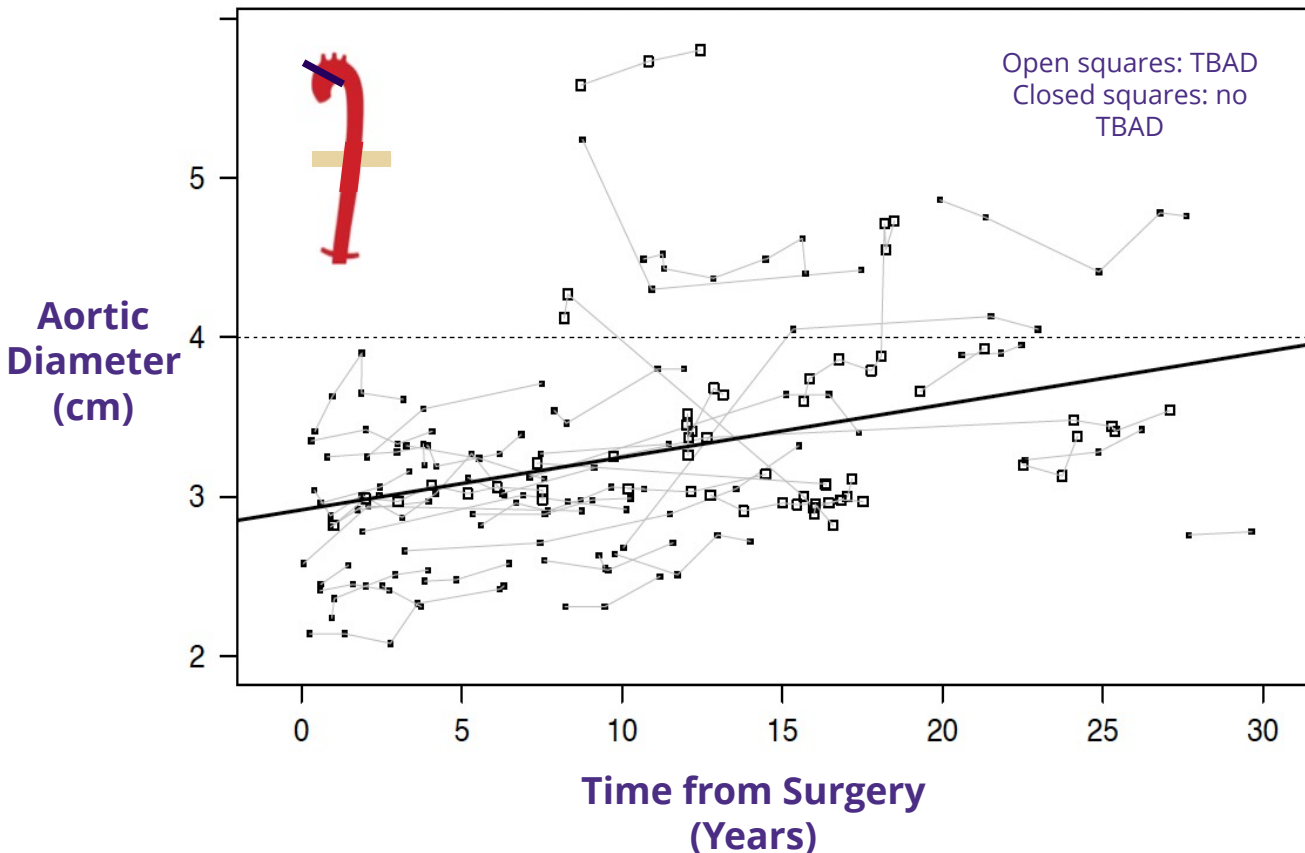
Key Arch Concepts for Aortopathy Patients

1. Management of arch during elective root surgery
2. Management of arch during acute type A aortic dissection
3. Management of arch during chronic residual arch dissection
4. Management of type B dissection



Management of arch during root surgery

Is a prophylactic arch operation necessary during elective root replacement in MFS?

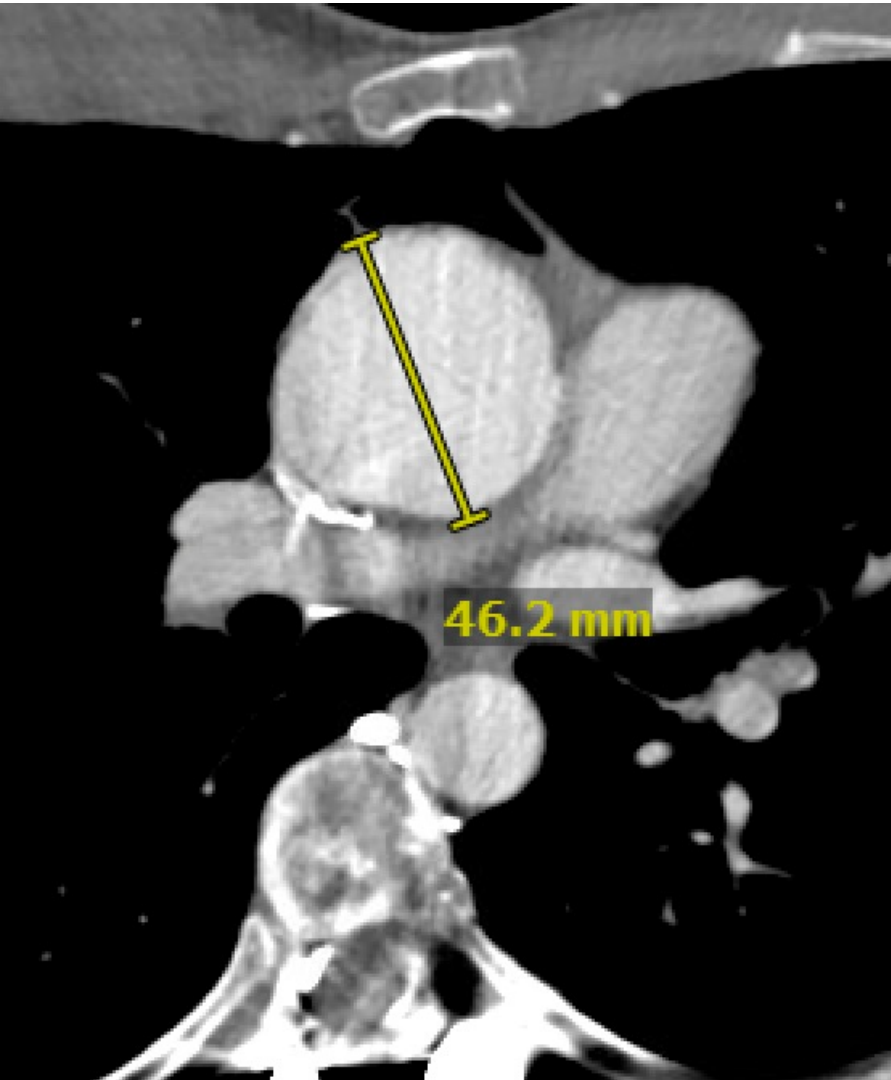


Rate of growth: 0.33 mm/yr

Rate of re-intervention on clamp/cannulation site: 1.6% (2 patients)

Rate of growth in standard MFS population: 0.3-0.6 mm/yr

This is not the case with LDS!!



27 yo F with LDS

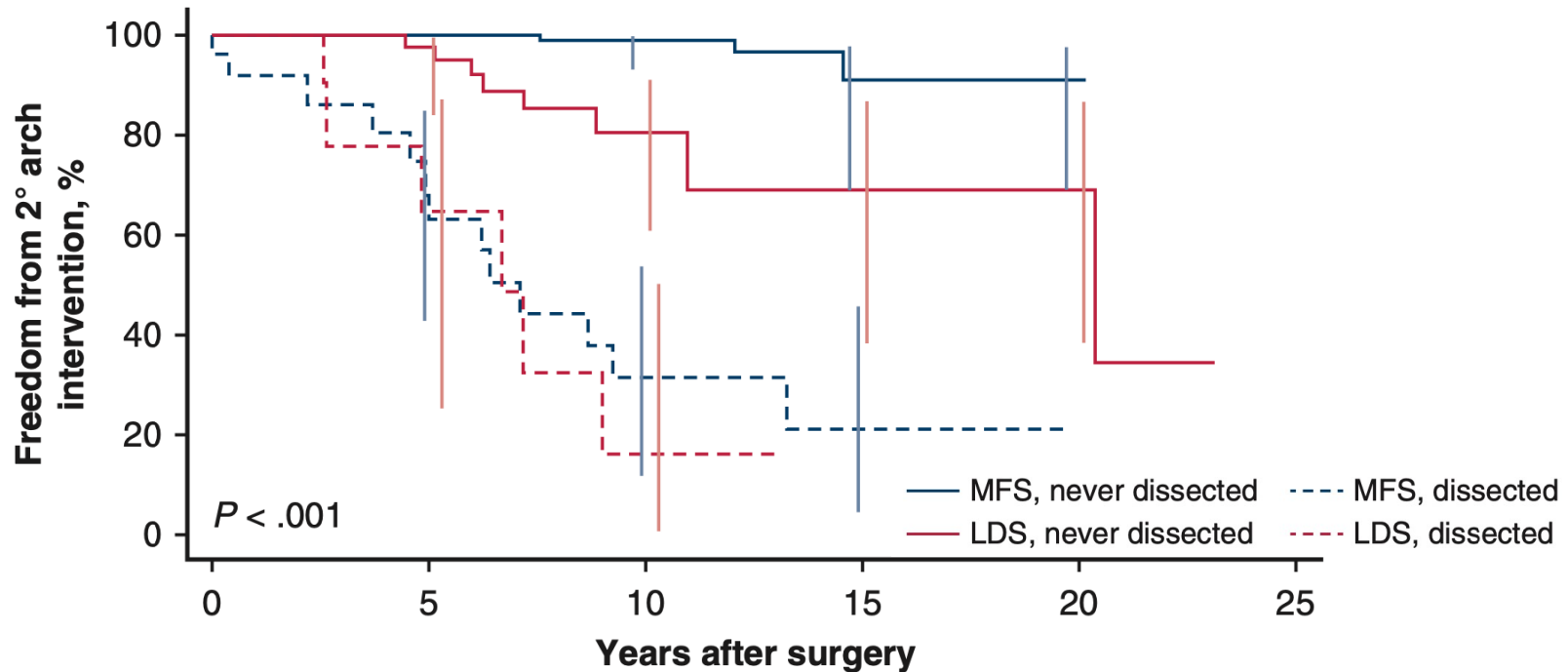
History of VSRR 10 years prior

Aneurysmal degeneration of
cross clamp site

Plan: Zone 2 arch



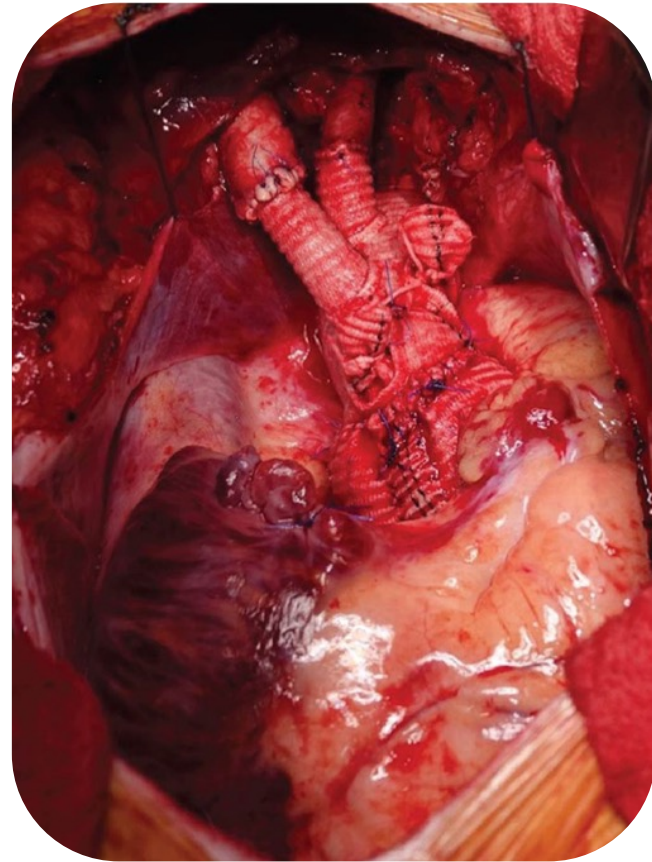
LDS has a unique aortic natural history compared to MFS



This has led many to advocate for performing an arch operation (hemiarch/total arch) at time of VSRR

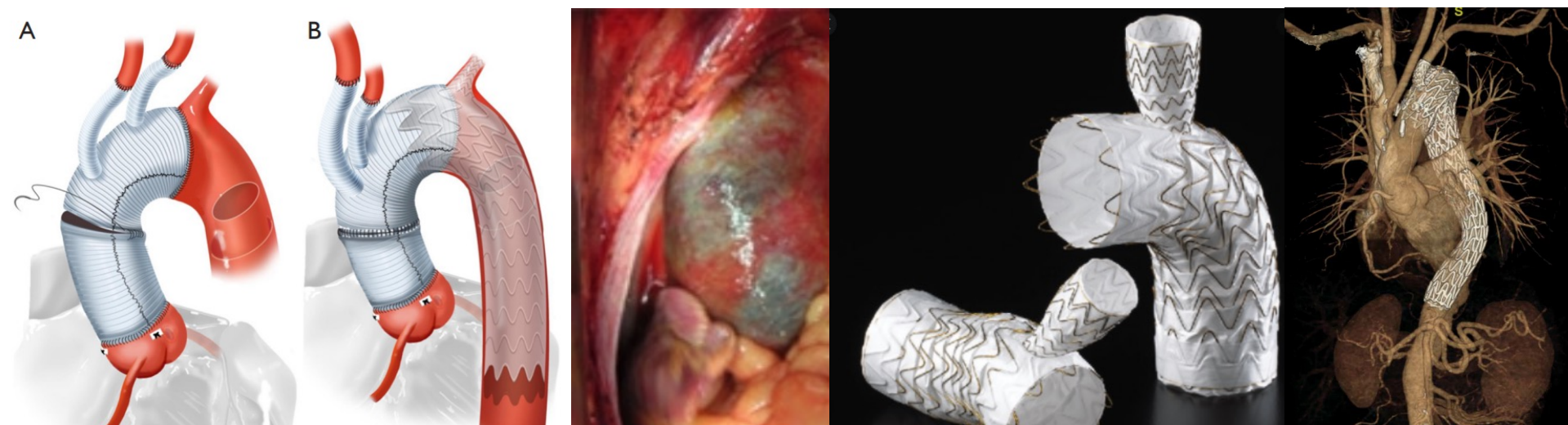
Management of arch during root surgery

- **MFS:** Arch can be left alone
- **LDS:** Arch operation typically required
- Others: Patient dependent but strong consideration for arch operation should be given.



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Genetic testing in TAAD

Characteristic	Pre-MTAP (n = 13)	MTAP (n = 43)
Pathogenic Mutation/VUS Positive	7 (58%)	18 (42%)
Mutation:		
- FBN1	2	5
- TGFBR2	1	2
- SMAD3	1	0
- MYH11	2	3
- MYLK	1	2
- ACTA2	0	1
- PRKG1	0	1
- LOX	0	1
- LZTR1	0	1
- SMAD6	0	1

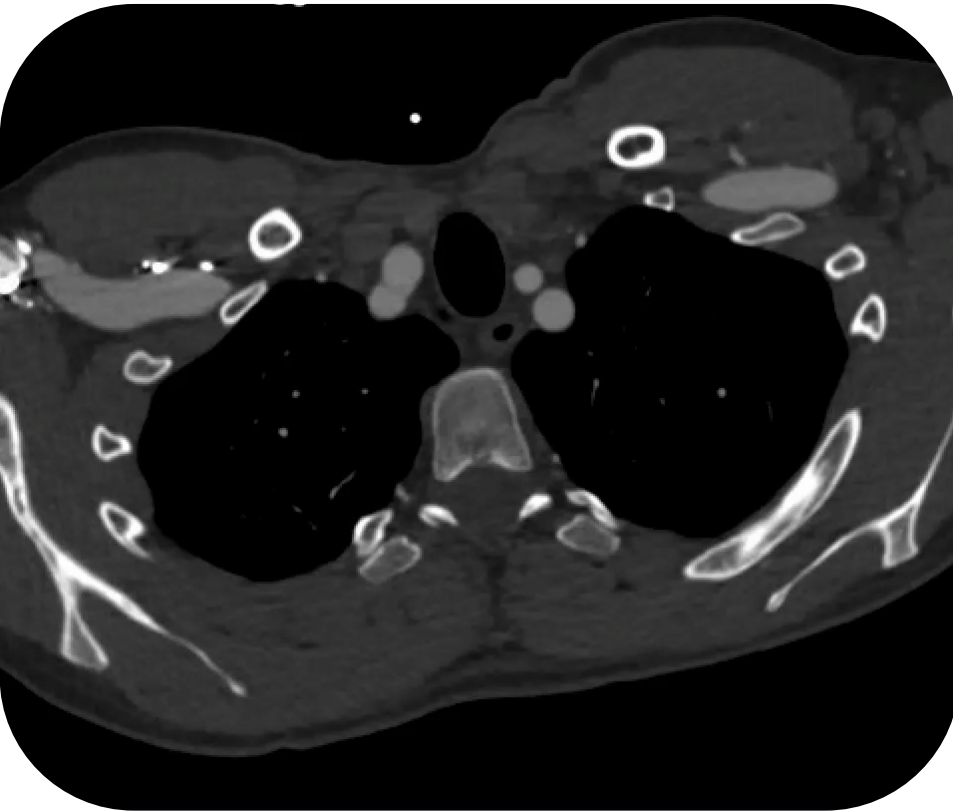
- ~50% VUS/pathogenic positivity rate!!
- Not “just” MFS- many variants identified
- Young patients, clinical features, family history, etc. need to be assumed to have aortopathy

Management of arch during acute type A dissection

- Type A dissection in genetic aortopathy patients is a total calamity
- Assumption must be made that the patient will ultimately require total aortic replacement
- **Root:** Typically replaced (VSRR vs Bentall)
- **Arch:** Zone 2 arch with interval TEVAR



Management of arch during acute type A dissection



23 yo M with LDS

History of VSRR at age 15

Acute chest pain radiating to back

Plan: Right axillary cannulation, Zone 2 arch

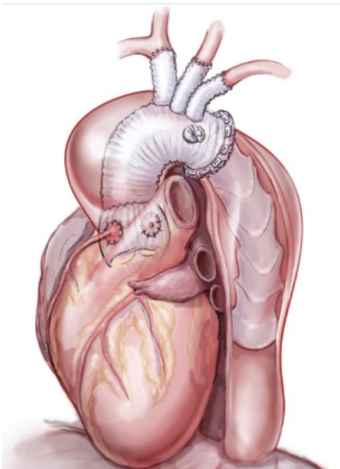
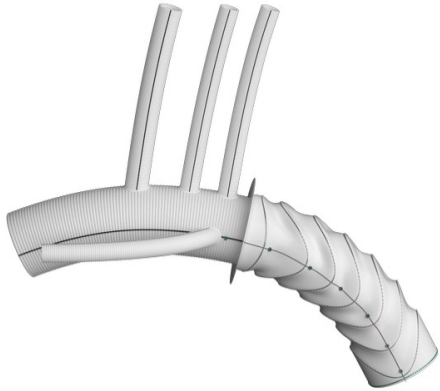
TBE/TEVAR performed 1-week post-op

Management of arch during acute type A dissection



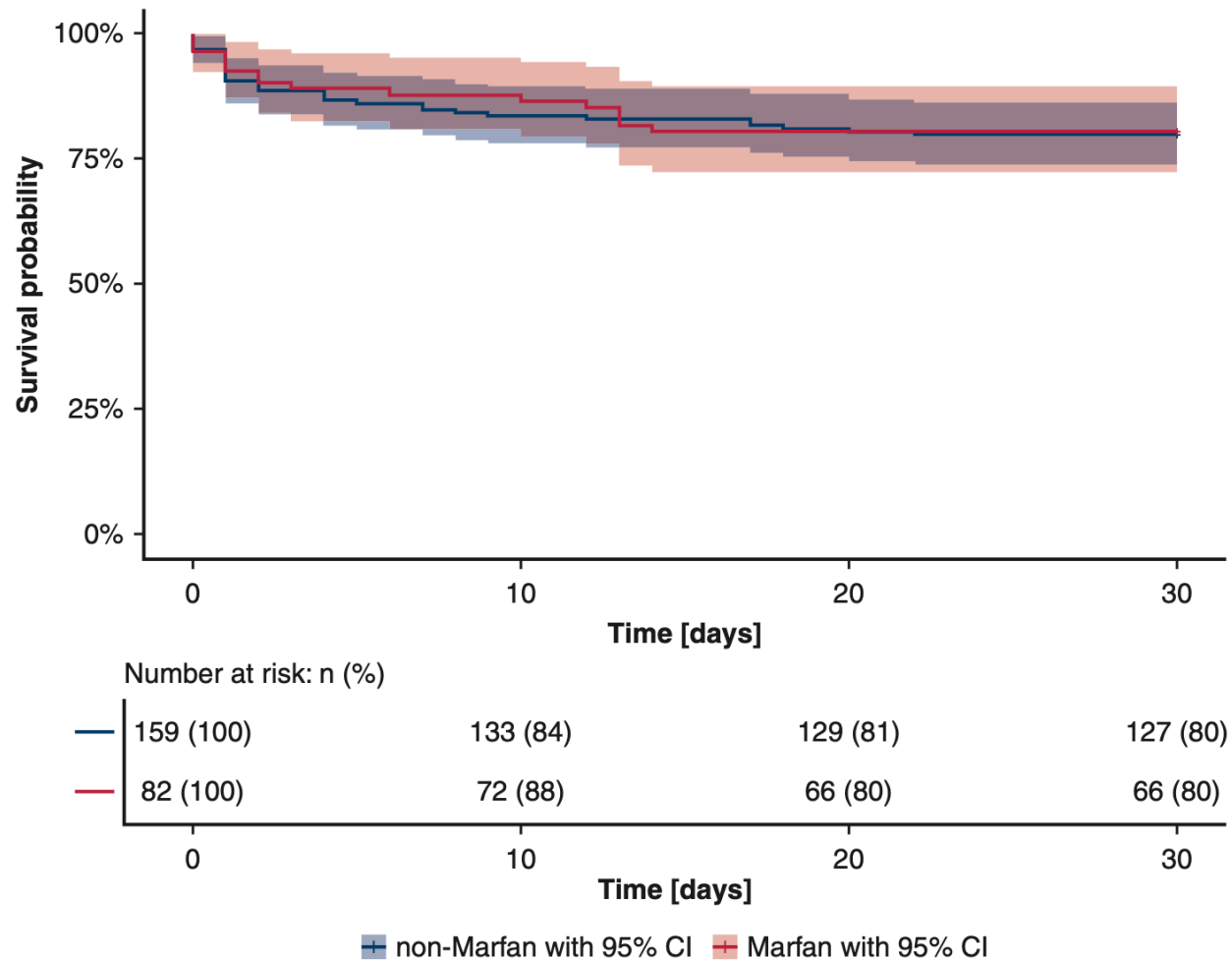
- Use of TBE controversial
- SINE very common- close monitoring
- Will require open TAAA repair; matter of timing

What about FET?

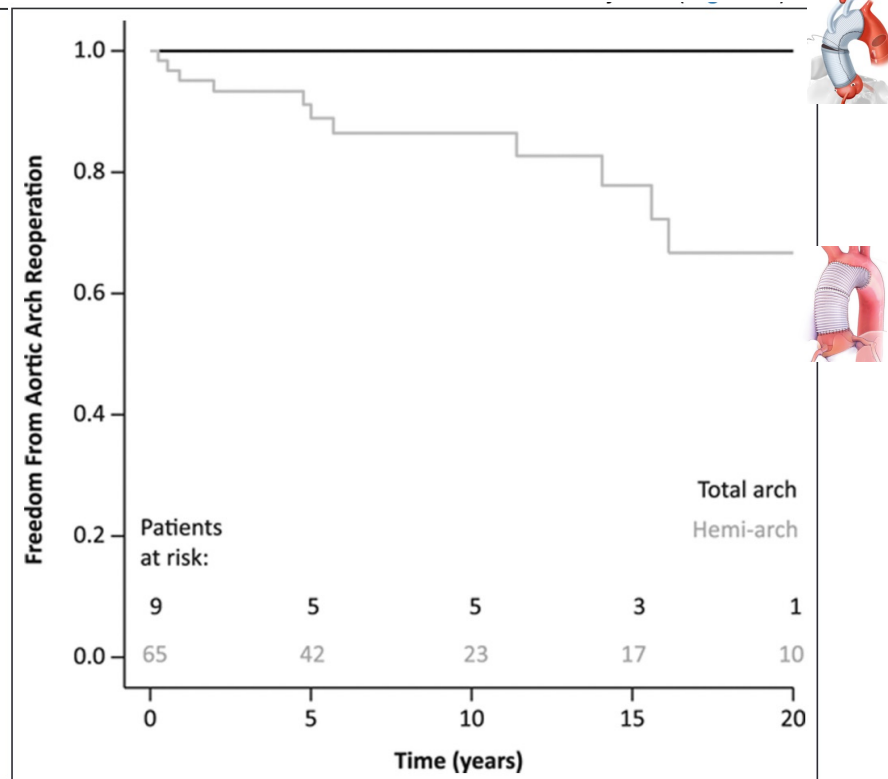
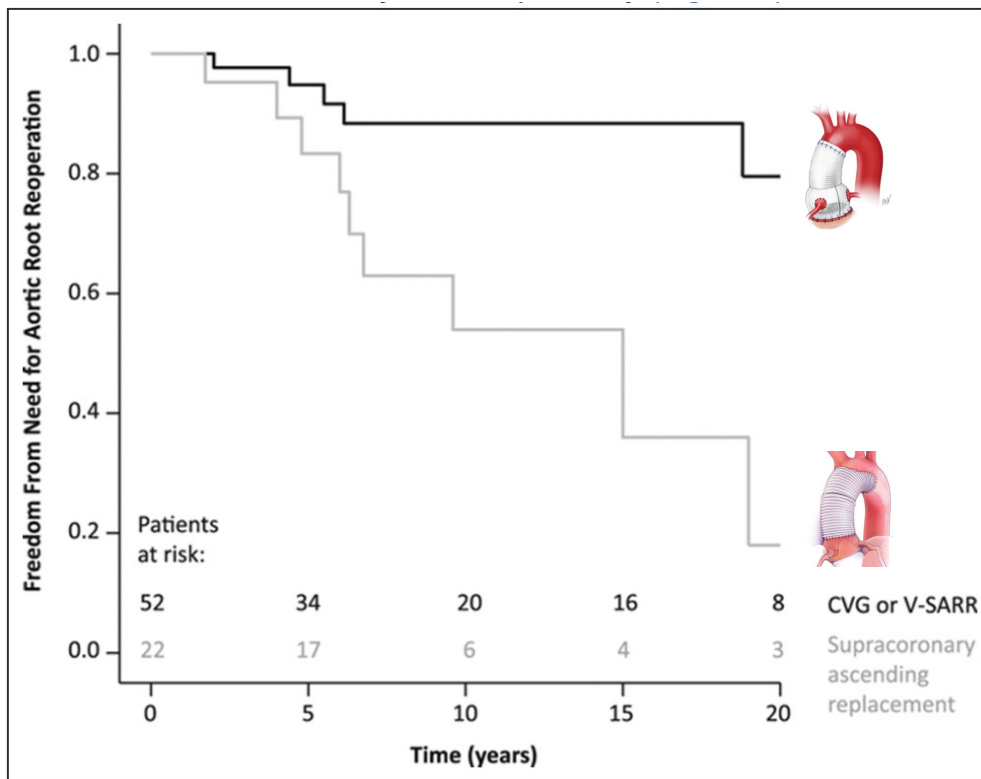


- Possible benefit in cases of severe abdominal malperfusion
- **Our preference is zone 2 arch followed by TEVAR**
 - Eliminates need to revascularize LSA during TAAD repair
 - Lower SCI risk by staging?
- All TAAD that receive zone 2 arch are offered TEVAR (within 6 weeks)

TAAD survival in MFS



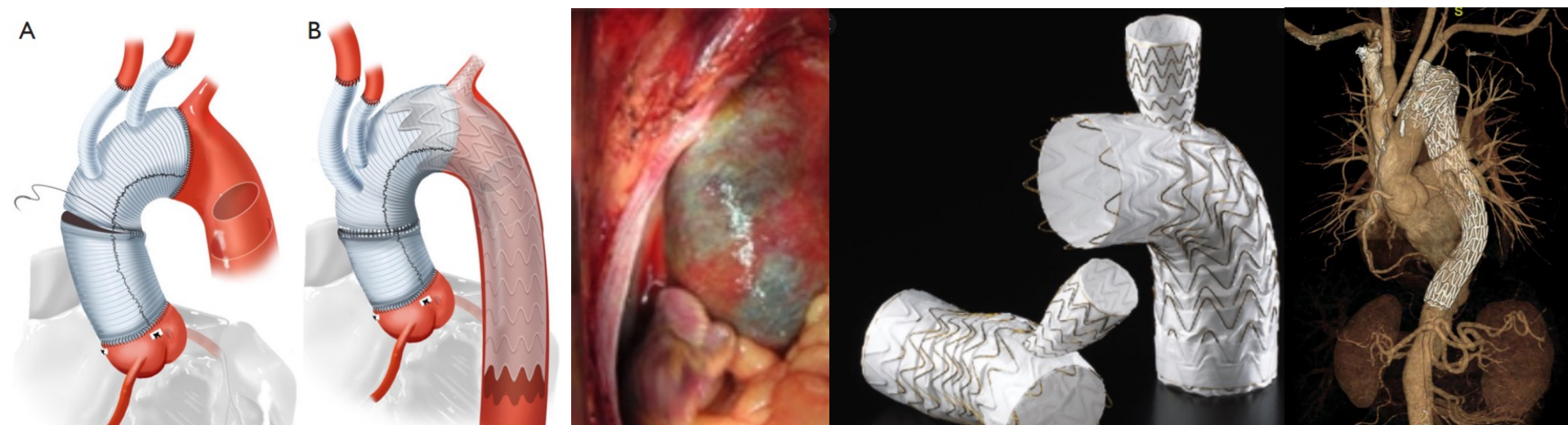
MFS patients have similar 30-day survival after TAAD repair



Failure to replace the aortic root and arch is associated with high reoperation rate.

Key Arch Concepts for Aortopathy Patients

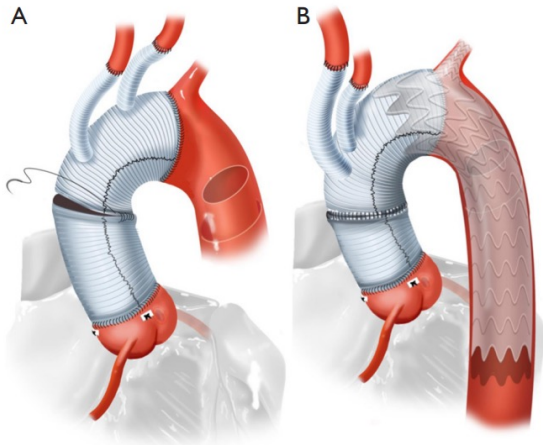
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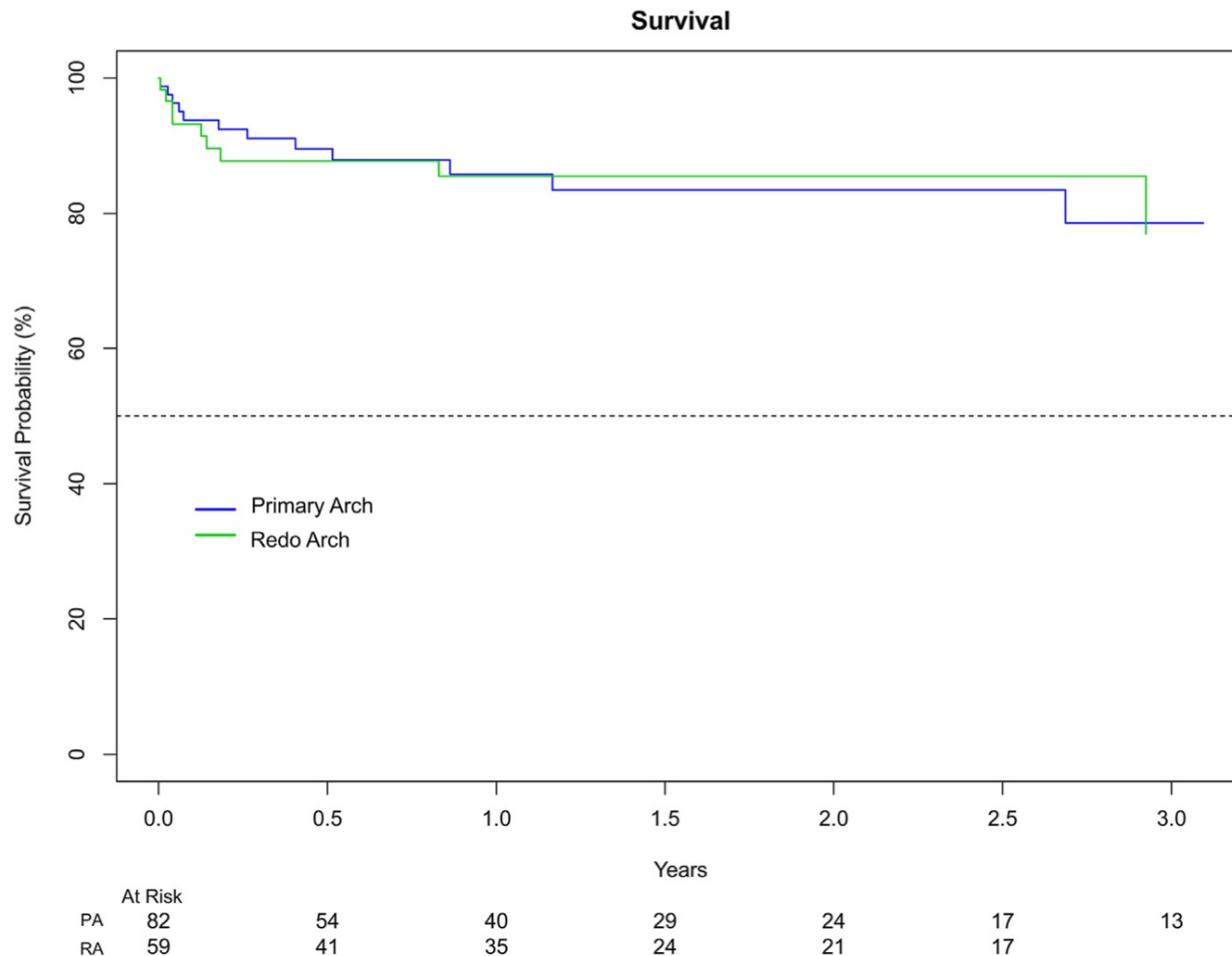
Management of arch during chronic residual dissection



- Most of these patients will have had previous ascending repair
- Planning is essential (re-entry, LZ creation, assessing valve/root pathology)
- Goal is to accommodate distal aortic repair as well as address any root pathology



Redo arch has similar outcomes

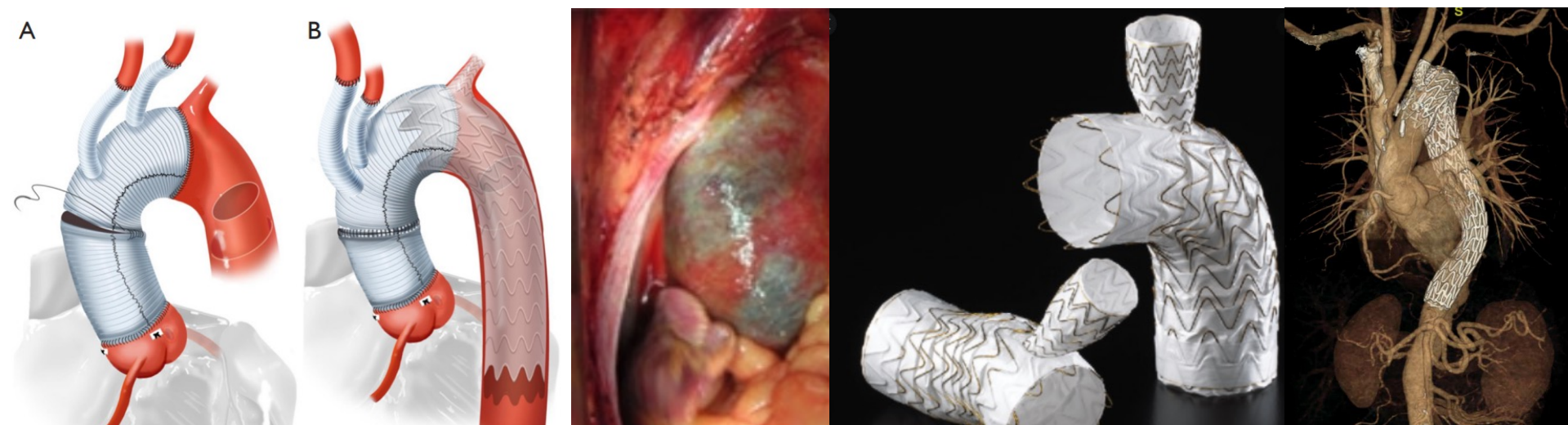


Stroke: 9.2%

In-hospital
mortality: 7.0%

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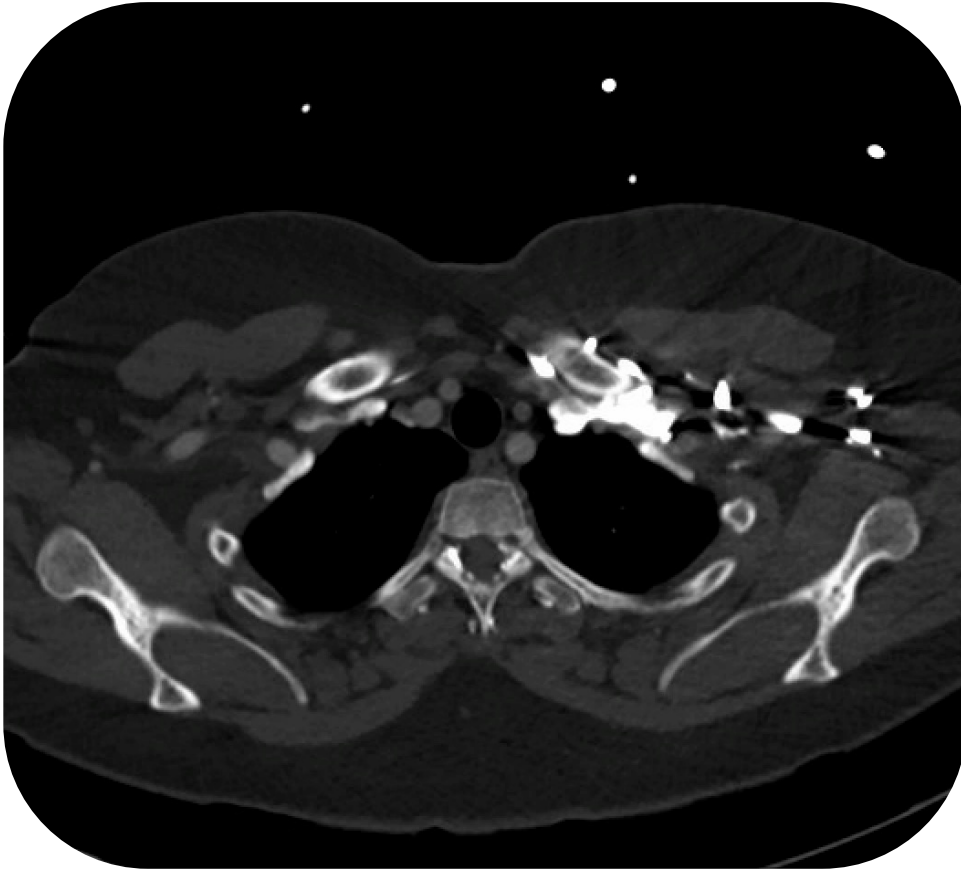


Management of type B dissection



- Occur more frequently than TAAD and typically with normal aortic dimensions
- Unique challenge given limitations of TEVAR in aortopathy patients
- Options include medical management, TEVAR, or open DTA/arch replacement +/- TEVAR
- Clinical status and anatomy dictate approach

Management of type B dissection



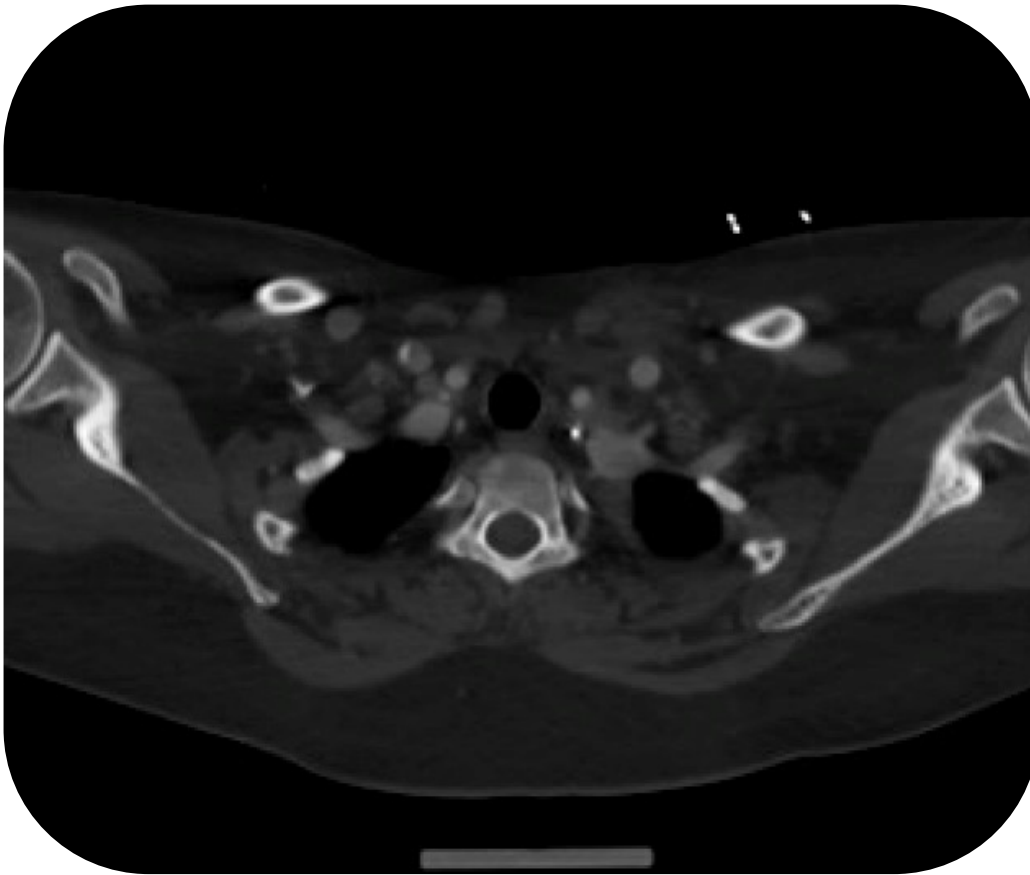
27 yo F with MFS

10/2020: TBAD during week 28 of second pregnancy

Plan: Medical management →
c/b pre-eclampsia

C-section at 30 weeks

Management of type B dissection



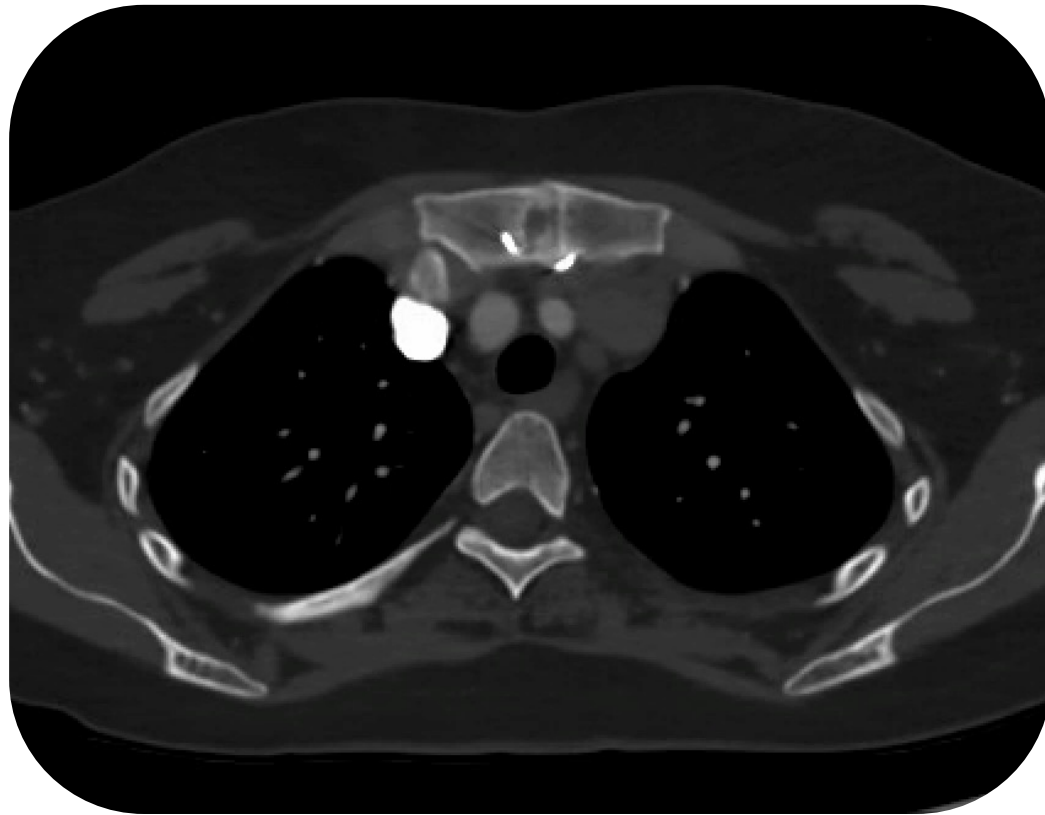
27 yo F with MFS

10/2020: TBAD during week 28 of second pregnancy

7/2021: DTA increased from 3.4 cm to 4.0 cm
- Intermittent chest pain

Plan: David V with zone 2 arch/FET

Management of type B dissection



27 yo F with MFS

10/2020: TBAD during week 28 of second pregnancy

7/2021: David V with zone 2 arch/FET

2/2022: Continued expansion of TAAA

Plan: Open TAAA repair

Management of type B dissection

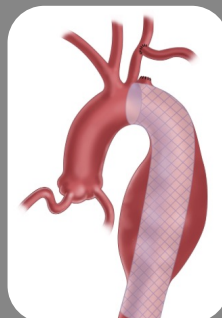
Medical Therapy

- Poor endo anatomy
- Lack of high-risk aortic features
- Allows time to recover from acute event



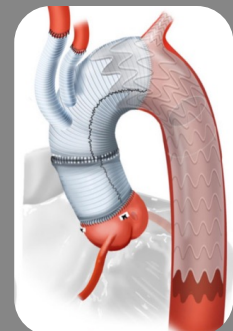
TEVAR

- Complicated dissection with malperfusion
- LZs vulnerable and need close f/u
- Risk of retrograde TAAD



Surgery

- Creates proximal LZ
- Can address proximal aortic pathology
- What to do with “normal” root?



Take Home Message

- Arch not required during MFS elective root
- Aortic dissection (of any type) is a watershed event
 - Typically begins path for complete aortic replacement
- These patients benefit from extensive operation during TAAD
- Role of endovascular therapy is still evolving

Thank you!

