

## Value of Endovascular TAAA in Patients with Heritable Thoracic Aortic Diseases: Which Device(s) Are Best and Why

**Gustavo S. Oderich, MD, FACS**  
 John P. & Kathrine G. McGovern Distinguished Chair, Professor of Surgery, Chief of Vascular and Endovascular Surgery, Director of the Aortic Center

Department of Cardiothoracic & Vascular Surgery | UTHouston McGovern Medical School | VEITH INSTITUTE

## Faculty disclosures

*G.S. Oderich*

- Consulting, research grants, scientific advisory board  
 WL Gore, Cook Medical Inc., GE Healthcare and Centerline Biomedical
- Investigational use of devices  
 Cook Fenestrated and Branched Grafts
- Special thank you  
 Research fellows: Lucas R. Kanamori MD, Bruno Schmid MD and Dora Babocs MD  
 Research faculty: Ying Huang MD PhD  
 Medical illustrations and graphics: David Factor and Chris Akers  
 3D CTA Lab: Laura Ocasio MD and Thanila A. Macedo MD

### CLINICAL PRACTICE GUIDELINE

#### 2022 ACC / AHA Guideline for the Diagnosis And Management of Aortic Disease

**Intact Thoracoabdominal aortic aneurysm**

**Open Surgical Repair is recommended** In patients with genetically triggered aortic diseases (Class I)

26M with LDS      49M with MFS

Baselbacher E et al. Circulation 2022

## Open TAAA Repair for CTDs

**Results of Open Surgical Repair in Patients With Marfan Syndrome and Distal Aortic Dissection**  
 127 patients (43 year-old) Mortality: 4%

**72 patients (41 years-old) Mortality: 11%**

**Open Descending and Thoracoabdominal Aortic Repairs in Patients Younger Than 50 Years Old**  
 314 patients (<50 years-old) Mortality: 6%

## Aortic reoperations in HTAD patients

**Beyond the Aortic Root: Staged Open and Endovascular Repair of Arch and Descending in Patients With Connective Tissue Disorders**

At 10-years after index procedure:  
 - 98 reinterventions/100 patients

- At least one reintervention: **61%**
- Aortic-related death prior to reintervention: **15%**
- Alive without event: **30%**

Roselli J et al. Ann Thorac Surg 2016

## Aortic reinterventions in HTAD patients

**No FB-EVAR**

Roselli J et al. Ann Thorac Surg 2016

### Aortic wall fragility

- Patients with HTADs were excluded from stent-graft pivotal trials
- Fragility of the aortic wall
- Progression of disease:
  - Device radial force
  - Aortic enlargement
  - Stent-graft induced trauma

HTADs are associated with retrograde dissection and stent-induced new entry tear

*Dong Z et al. Circulation 2009*

### Genotype-surgical phenotype correlation

No abnormality

- FBN1
- FNA1 null
- TGFBR2
- TGFBR1
- SMAD3
- COL3A1 null
- COL3A1 large amino acid substitution

*WIP*

### Clinical series of TEVAR for MFS

Author	n	Prior Repair	PLZ Zone 0-2	Retrograde Dissection	30-day Mortality	Type IA endleak	Follow up (months)	Reintervention	Endo	Open
Akin et al (2008)	8	83%	ns	ns	0	0	63	0	38%	
Norden et al (2009)	7	100%	6 (86%)	ns	14%	0	16	17%	0	
Gesbusch et al (2008)	6	50%	2 (30%)	ns	0	17%	33	15%	0	
Botta et al (2009)	12	100%	ns	ns	0	8%	31	8%	8%	
Marcheix et al (2008)	15	73%	2(15%)	ns	0	27%	25	20%	30%	
Waterman et al (2012)	16	94%	4(25%)	ns	6%	19%	9	20%	45%	
Eid-Lidi et al (2015)	10	50%	ns	ns	10%	ns	60	30%	0	
Huu et al (2022)	37	13 (50%)	6 (15%)	ns	5.5%	ns	36	14%	45%	
<b>Total</b>	<b>69</b>				<b>5%</b>	<b>19%</b>	<b>32</b>	<b>20%</b>	<b>22%</b>	

*Adapted from Bakker G et al. Cardiovasc Med Biol 2017*

### FB-EVAR indications in patients with HTADs

**Indications**

- **FL enlargement** post TEVAR
- **Symptomatic/contained ruptured aneurysm**
- **Stent-induced new entry tear**
- **Reoperative repair** after open DTA/TAAA repair
- Hostile surgical anatomy
- High clinical risk or complicated course following prior repair

- Cook patient-specific TAAA platform
- Cook t-Branch® / TAAA+®
- Gore TAMBE®
- Artivion E-nside® TAAA
- Medtronic Valiant® TAAA

*WIP*

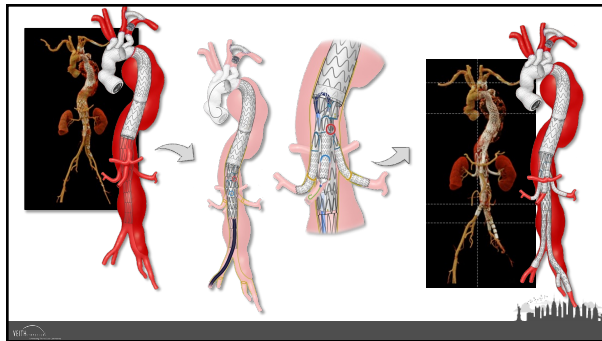
### FB-EVAR in patients with HTADs

- 857 patients treated by FB-EVAR (543 Extent I-IV TAAAs)
- **15 patients (1.7%)** had confirmed HTADs (8 male, median a 58 years-old (IQR, 49-70)
  - Marfan Syndrome, 9
  - Loays-Dietz Syndrome, 4
  - vascular Ehlers Danlos Syndrome, 1
  - Extent II TAAA in 12, Extent III TAAA in 2, Extent IV in 1
- No mortality
- Length of stay, 6±4 days
- Median follow up, 20 months
- Three patients (20%) required secondary interventions

*Tribe L (Dobersch G) et al. Ann Surg 2016 (under review)*

- 52-year-old man with LDS
- 8.5-cm Extent II TAAA
- Multiple complications from total arch repair: cardiac arrest (x2), respiratory failure (trach), temporary dialysis, L below knee amputation
- Stable CHF with EF 40%
- Moderate COPD

*WIP*



### TAAA/arch branch for HTADs

- 427 FB-EVAR patients
- 17 (4%) had HTADs
  - Mean age, 51±8 years-old
  - **1 arch, 16 TAAA**
  - Technical success 100%
  - No mortality
- Mean follow up, 3.4 years
- Reintervention in one patient

Endovascular Repair of Thoracoabdominal and Arch Aneurysms in Patients with Connective Tissue Disease Using Branched and Fenestrated Devices

Clough R et al. Ann Vasc Surg 2007.

### EVICTUS STUDY

EndoVascular Intervention in patients with Connective Tissue disease

- 18 clinical sites
- US, UK, European Union, China and New Zealand

LIPPSALA UNIVERSITY

### EVICTUS study

- 171 patients with underlying HTADs treated in 18 centers by endovascular repair of TAA/ TAAAs (2005-2020)
- 152 (89%) had chronic post-dissection and 19 (11%) had degenerative aneurysms
- **16% of all procedures** performed in patients with HTADs

83% Marfan Syndrome, 142  
10% Loays-Dietz Syndrome, 17  
7% Vascular Ehlers Danlos Syndrome, 12

### Patient characteristics

	MS n = 142	LDS n = 17	vEDS n = 12
Age (years, IQR)	49 (38-58)	53 (44-58)	40 (28-75)
Male sex	62%	53%	83%
Hypertension	56%	59%	58%
Renal impairment	11%	18%	0%
Prior open aortic repair	<b>81%</b>	<b>88%</b>	<b>50%</b>
Dissection	90%	88%	75%
Degenerative aneurysm	10%	12%	25%
Emergency indication	<b>40%</b>	<b>24%</b>	<b>75%</b>

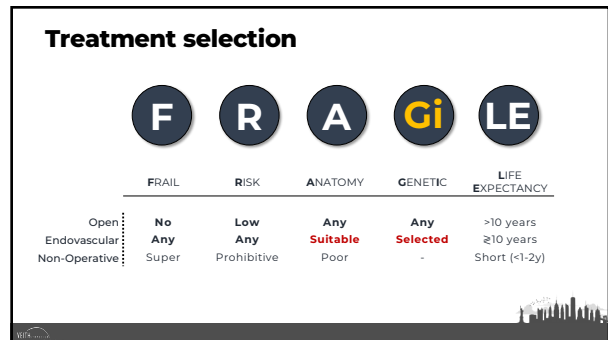
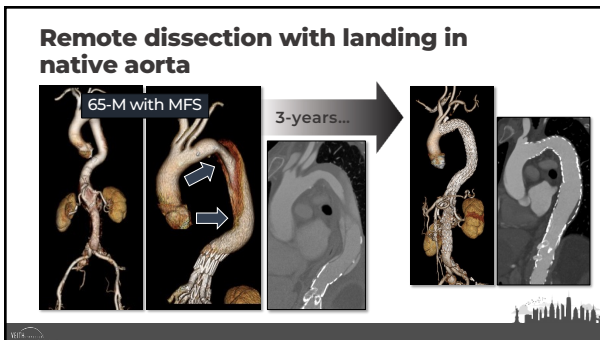
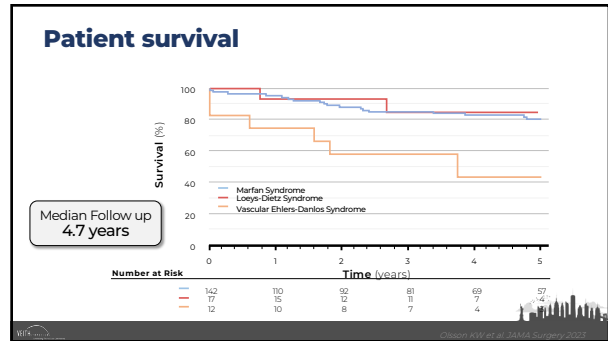
### Endovascular approach

	MS n = 142	LDS n = 17	vEDS n = 12
Proximal landing zone in surgical graft	<b>56%</b>	<b>59%</b>	42%
Distal landing zone in surgical graft	15%	6%	8%
FBEVAR	<b>17%</b>	<b>47%</b>	<b>34%</b>
Parallel grafts	3%	0%	0%
Debranching	31%	12%	17%

- 30-day or in-hospital mortality: **3%**
- Primary technical success: 98%
- Stroke: 6%
- Spinal cord injury: 0.5%
- Renal impairment: 1.5%

### Secondary procedures

	MS n = 142	LDS n = 17	VEDS n = 12
Any secondary procedure	54%	59%	42%
Proximal endovascular extension	5%	0%	0%
Distal endovascular extension	19%	18%	0%
Branch stenting	3%	12%	17%
Embolization	15%	24%	25%
Repair of different segment	8%	18%	0%
Conversion to open repair	9%	6%	0%
Repair of different segment	17%	18%	0%



- ### Conclusion
- Open surgical repair remains the gold standard for CTD patients
  - FB-EVAR experience is limited, but this should be considered in high risk patients and those who need redo TAAA repair
  - The ideal patient has sealing zones based on open surgical grafts
  - Preliminary experience shows high technical success and low mortality
  - Limitations remain access, cost, reinterventions and need for surveillance

### Thank You!

Department of Cardiothoracic & Vascular Surgery | UTHealth Houston | McGovern Medical School

@gustavooderich@uth.tmc.edu  
 go4aorta@gmail.com  
 @GustavoOderich  
 go4aorta