VEITH SYMPOSIUM Connecting The Vascular Community 51st

Reasons to change from open to endovascular aortic aneurysm repair in patients with connective tissue disorders: When is open repair mandatory in this setting?

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No Disclosures

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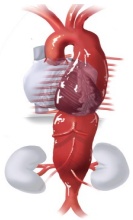
Complex aortic pathologies

- "Remaining" indications for open TAAA repair
  - Connective tissue disease
  - (relatively) young patients
  - Post dissection TAAA without endo options
  - Conversion after TEVAR
    - –persistent endoleak (1a/1b), no endo options
    - –infection
    - –aorto bronchial fistulae
    - –aorto esophageal fistula
    - –progression of aneurysmal disease without endo options



Treatment of thoracoabdominal aortic disease in patients with connective tissue disorders

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many device- and aorta-related complications both in the short term and in the long term. Despite the lack of level 1 evidence, open repair currently remains the standard approach to treatment of aortic disease due to CTDs. Open branched graft repair in particular is the preferred technique. Endovascular interventions may be cautiously used in patients with CTDs in selective circumstances. (J Vasc Surg 2018;68:1257-67.)

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Editor's Choice – Open Thoracic and Thoraco-abdominal Aortic Repair in Patients with Connective Tissue Disease

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WHAT THIS PAPER ADDS
This article confirms that open surgery is a durable treatment for patients with connective tissue disease but nevertheless entails relevant risks. The article emphasizes the benefit of staged operating strategies.

Outcomes After Endovascular Aortic Intervention in Patients With Connective Tissue Disease

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171 patients with CTD
89% post dissection aneurysms
11% degenerative aneurysms
80% previous aortic surgery

Indication	No. (%)		
	Marfan syndrome (n = 142)	Loeys-Dietz syndrome (n = 17)	Vascular Ehlers-Danlos syndrome (n = 12)
Dissection[a]	128 (90.1)	15 (88.2)	9 (75.0)
Aneurysm[b]	14 (9.9)	2 (11.8)	3 (25.0)
Emergent[c]	57 (40.1)	4 (23.5)	9 (75.0)
Malperfusion	12 (8.5)	2 (11.8)	1 (8.3)
Rupture	8 (5.6)	0	3 (25.0)

Procedure	No. (%)		
	Marfan syndrome (n = 142)	Loeys-Dietz syndrome (n = 17)	Vascular Ehlers-Danlos syndrome (n = 12)
Proximal landing zone in surgical graft	79 (55.6)	10 (58.8)	5 (41.7)
Distal landing zone in surgical graft	21 (14.8)	1 (5.9)	1 (8.3)
Branched	13 (9.2)	3 (17.6)	2 (16.7)
Fenestrated	11 (7.7)	5 (29.4)	2 (16.7)
CHIMPS	4 (2.8)	0	0
Debranching	44 (31)	2 (11.8)	2 (16.7)
Staged repair	24 (16.9)	4 (23.5)	1 (8.3)

Procedure	No. (%)		
	Marfan syndrome (n = 142)	Loeys-Dietz syndrome (n = 17)	Vascular Ehlers-Danlos syndrome (n = 12)
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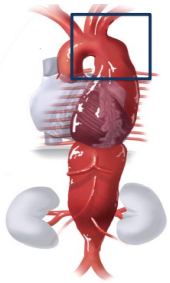
CHIMPS	4 (2.8)	0	0
Debranching	44 (31)	2 (11.8)	2 (16.7)
Staged repair	24 (16.9)	4 (23.5)	1 (8.3)

Outcome	No. (%)		
	Marfan syndrome (n = 142)	Loeys-Dietz syndrome (n = 17)	Vascular Ehlers-Danlos syndrome (n = 12)
Primary technical success	140 (98.6)	17 (100)	11 (91.7)
Mortality	3 (2.1)	0	2 (16.7)
Conversion	3 (2.1)	0	0
Acute coronary syndrome	0	0	0
Stroke	8 (5.6)[a]	0	2 (16.7)
Spinal cord ischemia	1 (0.7)[b]	0	0
Kidney impairment	3 (2.1)[c]	0	0

eTable 4. Secondary procedures

	Marfan syndrome (n=142)	Loeys-Dietz syndrome (n=17)	Vascular Ehlers-Danlos syndrome (n=12)
Secondary procedures, n (%)	76 (53.5%)	10 (58.8%)	5 (41.7%)
Secondary endovascular procedures, n (%)	57 (40.1%)	8 (47.1%)	3 (25.0%)
- Proximal extension, n (%)	7 (4.9%)	0 (0.0%)	0 (0.0%)
- Distal extension, n (%)	27 (19.0%)	3 (17.6%)	0 (0.0%)
- Relining (for endoleak type III), n (%)	1 (0.7%)	0 (0.0%)	0 (0.0%)
- Branch relining/extension, n (%)	4 (2.8%)	2 (11.8%)	2 (16.7%)
- Embolization, n (%)	21 (14.8%)	4 (23.5%)	3 (25.0%)
- Repair of different aortic segment, n (%)	11 (7.7%)	3 (17.6%)	0 (0.0%)
Secondary open procedures, n (%)	39 (27.5%)	4 (23.5%)	4 (33.3%)
- Conversion, n (%)	13 (9.2%)	1 (5.9%)	0 (0.0%)
- Bypass/deviation, n (%)	2 (1.4%)	0 (0.0%)	2 (16.7%)
- Other, n (%)	3 (2.1%)	0 (0.0%)	2 (16.7%)
- Repair of different aortic segment, n (%)	24 (16.9%)	3 (17.6%)	0 (0.0%)

Problem area for endografting in CTD patients



Gothic configuration
Involvement arch vessels
Dissection
Rigid septum
Short sealing zone
Vulnerable tissue

Inadequate apposition
Bird beaking
Type 1a endoleak
Malperfusion
Retrograde type A dissection
Aneurysm progression

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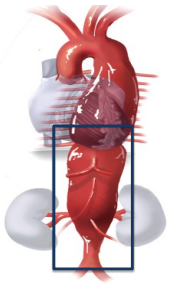
Distal aortic landing zone



Less vulnerable
Less complex morphology
Easier to handle
Type 1b easier to manage
Extension procedures

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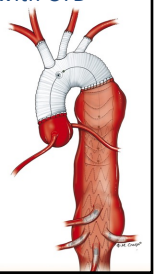
Main argument to change from open to endovascular TAAA repair in patients with CTD

Safe proximal landing zone
- Elephant trunk, frozen elephant trunk
- Surgical graft
- Well positioned endograft
- Adequate native aortic segment for proximal sealing

If not: open surgery still the gold standard



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