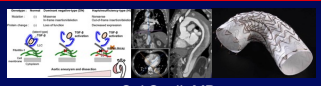
Results of Endovascular Graft Aortic Repair in Mixed Connective Tissue Disease (Heritable Aortic Disease): Is It Time To Change The Guidelines: In What Circumstances is Endovascular Repair Indicated?



Sal Scali, MD
Division of Vascular Surgery & Endovascular Therapy
University of Florida, Gainesville

Wednesday, November 20th, 2024 2:18-2:23PM
Session 44: More New Developments in AAAs

Disclosures

- None

Background

- Open aortic repair (OAR) is the 'gold standard' for connective tissue disease patients (CTD) with aortic pathology.
- CTD patients undergoing OAR have operative mortality ranging from 3-12%, SCI 4-7%, and 5-year survival of 50-60%.
- Clinical guidelines recommend against endovascular aortic repair in CTD patients.

Coselli et al. Ann Thorac Surg. 2016
Hicks et al. J Vasc Surg. 2017
Frankel et al. Ann Thorac Surg. 2020

Guidelines, CTD & Endo

Guideline	General Stance on EVAR/TEVAR	Specific Scenario for Endovascular Use	Focus
Chaikof et al. (2018)	Avoid if possible; open preferred	Select emergency (rupture/dissection)	Fragility of CTD tissue
Isselbacher et al. (2022)	Cautious consideration, selected scenarios	Acute TBAD or life-threatening complication	Tailored, multi-disciplinary approach
Wanhainen et al. (2024)	Strongly discouraged, except emergency	Salvage/emergency cases	Durability, long-term complications
Czerny et al. (2024)	Rarely recommended, only multidisciplinary consensus	Hemodynamic instability or anatomic complexity	Centralized care & risk mitigation

*Significant variation in CPG endo enthusiasm

Chaikof et al. J Vasc Surg. 2018
Isselbacher et al. Circulation. 2022
Wanhainen et al. Eur J Vasc Endovasc Surg. 2024
Czerny et al. Ann Thorac Surg. 2024

CTD & Endo Dilemma

ENDO

- Device-related Complications

-SINE – 10x higher (~3% vs. 30%)
-RTAD – 1-3x higher (~2% vs. 5%)
-Conversion to Open (0-33%)

- Reintervention Rates

- 14-60% [index & remote sites]

- Pediatrics

-Anatomic suitability, durability

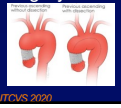
OPEN

- Open repair complications
-Dissection/malperfusion
15-30% mortality

Gambert et al. Eur J Vasc Endovasc Surg. 2022

- Rupture

- Redo surgery



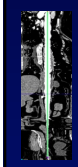
Glebova et al. J Vasc Surg 2018
Sandhu et al. JTCVS 2020

CTD & B/FEVAR

Annals of Vascular Surgery

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

- Single center, N = 17 {16 TAAAs, 1 arch} [2004-2015]
'unfit for open repair'
Mean follow-up = 3.4 years (0.3-7.4)



30-day Mortality: 0

SCI: 0

AKI: 19%

Reintervention 6% (N = 1)

Late death 6% (N = 1 @ 2yrs)

No conversion to open repair

Clough et al. Ann Vasc Surg 2017

Challenging Dogma - EVICTUS

JAMA Surgery | Original Investigation
Outcomes After Endovascular Aortic Intervention in Patients With Connective Tissue Disease

- 18 centers: Europe, Asia, US, New Zealand [2005-2020]
- All endovascular repairs/indications

N = 171
- 83% Marfan syndrome
-17% LDs/VEDs

Median age: ~50
Sex: ~63% male

Indications: 89% dissection, 11% aneurysm

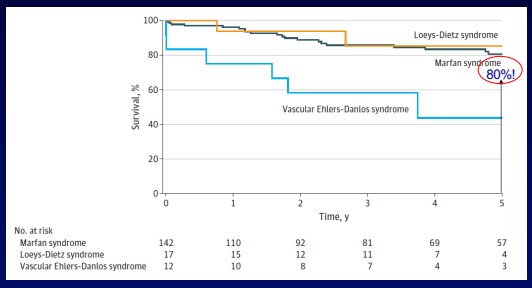
Aortic surgery: 79.5% prior open

30-day mortality: 2.9%

-RTAD: 1.2%
-Conversion: 2.3%

Olsson et al. JAMA Surgery 2024

CTD, Endo & Mid-Term Survival



Olsson et al. JAMA Surgery 2024

UF Experience

Variable, N = 43	No. (%)
Age, years (SD)	50 (16)
Sex, female	14 (33%)
- Marfan syndrome	34 (79%)
- Ehlers-Danlos syndrome	4 (9%)
- 'Other'/undefined	3 (7%)
- Loeys-Dietz syndrome	2 (5%)

- 80% prior open aortic operation
- **74% emergent/non-elective**

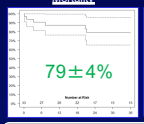
Types of Repair
TEVAR EVAR Fen/branchedEVA R Hybrid
11%
11%
4%
74%

*Mean Follow-up Time = 48±46 months
Median 29 [IQR 14, 76]

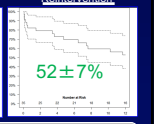
30-day mortality 5% (n=2)

30-day morbidity 29% (n=12)

Freedom from Aorta-related Mortality



Freedom from Aorta-related Reintervention



OR 3.1 95%CI 1.1-4.5, p = .03
≥ 2 Previous Open Aortic Operations

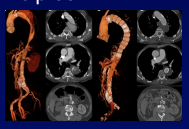
*Dacron LZ OR 0.8 95%CI 0.6-1.0, p = .06

Stinson G et al., JTCVS, In Revision

Optimizing Endo for CTD

Aortic Center Referral	Multi-disciplinary Team	Pediatric Case	Elective Cases (not routinely)
✓	✓	X	X

- Stent graft selection, oversizing principles
- Access vessel considerations
- Balloon molding (avoid)
- Staged/Bridging strategies
- 'Graft in Graft'



Pellenc et al. Eur J Vasc Endovasc Surg 2019
Stinson G et al., JTCVS, In Revision

Conclusions & Recommendations

- Historical norms being challenged for CTD + Endo.
- Endovascular repair likely reasonable option:
  - non-elective scenarios
  - anatomic high-risk [reduce complexity]
  - physiologic high-risk
  - adjunct/bridge to open aortic repair
- *High rate of Reintervention* – mandates differential surveillance intensity relative to non-CTD patients and needs to be disclosed.
- Device evolution may extend eligibility criteria.

Thank You

UF UNIVERSITY of FLORIDA