When are Endografts the Right Option for Treatment of Marfan Syndrome Patients?

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Disclosures:

None

Historical Perspective

“There is no disease more conducive to clinical humility than aneurysm of the aorta.”

“The tragedies of life are largely arterial.”

Concerns about TEVAR in MFS

1. CTD exclusion of all devices to date
   - Device radial force.
   - Tendency of devices to straighten.
   - Bare metal stents?

2. Fragility of the aortic wall
   - Stent graft induced trauma.
   - Retrograde dissection.
   - Failure to control aorta remote to stent.

“Retrograde aortic dissection was the most common complication for MFS.” (Circ 2009)

- The arch and ascending are at risk for rAAD. (Circulation, 2009)
  - Distal ascending and proximal arch are usually guidewire related.
  - Whole arch dissection is usually stent-graft induced (60%+).
- MFS pts accounted for 12% of rAAD, yet were <1% of population.
- 50% necropsy showed rAAD from bare springs, all MFS.

- 12 year old Loeys-Dietz Syndrome....
**Stent-Graft Induced New Entry Tears**

- 33% incidence in MFS patients vs 3% in non-MFS patients. (JVS, 2010)
- No difference in mortality of proximal vs. distal SINE (25% vs 28%).
- Authors did not impugn bare stents.
- 2.5% w/ Medtronic device, 2.4% report from AZ Heart w/ Gore TG.
- No relationship of new entry tear to oversizing.
  - 15% oversizing in SINE vs 16% in non-SINE.

**Fate of the Aorta after Aortic Dissection in Marfan Syndrome (IRAD)**

- IRAD (1996-2017)
  - 258 MFS/6,424 AD pts
  - MFS: younger (38 v 63), known aneurysms, previous surgery.
  - MFS AD: Association with aortic annulus diameter but not distal aorta diameter
  - 55% re-intervention rate
  - 100% survival after open surgery for TBAD (N=27/258).
  - vs 17.6% mortality for non-MFS open surgery TBAD.

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**Fate of Aorta after TEVAR in MFS**

- Geisbusch, 8pts (JEVT, 2008):
  - Perioperative complications in 25%,
  - 38% reintervention rate
  - 50% patients developed de-novo aneurysms.
- Nordon, 7pts (JVS, 2009):
  - 14% mortality
  - 33% reintervention rate for endoleak.
  - 83% false lumens thrombosed.
  - Yet all DTAs continued to dilate (7mm/yr on average).

**Where does TEVAR fit into TBAD for Marfan Syndrome?**

- Evidence suggests TEVAR may be safe in short term, but device issues are central in local aortic complications, especially in acute Type B.
- There is potential benefit of TEVAR to stabilize acute DTA emergencies:
  - “Bridge” to definitive therapy in rupture
  - Surgical graft- surgical graft sealing zones
  - Allow referral to center where open TAAA surgery has matured excellent results.
- STS/AATS Consensus, 2012
MFS TEVAR:  Bridge to definitive surgery

- Allowed referral to our center where open TAAA surgery has matured excellent results.

Conversion technique after TEVAR for TAAA.

Endovascular Therapy in MFS Patients

- Multidisciplinary evaluation by geneticist, physician, and surgeon.
- Drive BP down (SBP <90) during endovascular intervention.
- Liberal use of techniques to reduce operative trauma.
- Stent-graft therapy in CTD is defined in limited fashion.
  - Graft-to-graft sealing zones.
  - Revision procedures, reoperative exposures
  - Emergent “Bridge” to referral

Thank you