The Role of Stent-Grafts in Marfan Syndrome

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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.”
12 year old Connective Tissue Disorder….
Aortic biology is a now realized as marriage of homeostatic mechanisms and structural stressors.

Lindsey, Dietz, Nature 2013
Humphrey, Science, 2015
Diagnosis of MFS

• Revised in 2012
  – More emphasis on cardinal features of root aneurysm and lens dislocation
  – FBN1 testing weighted with a score of other systemic findings

• Differential diagnosis is MASS, LDS, CCA

• β-blocker ± ARB (Lacro, NEJM)
The Modern Problem: Aging of the Aorta in MFS

- 60 yo MFS, 17 yrs post CVG, nonsmoker, **no proximal graft**
Aging of the Aorta in MFS

• 52 yo MFS, 15 yrs post Bentall, 5yrs post TAAA, has proximal graft

How should we handle these aging CTD patients?
Dissection Changes the Natural History of Aortic Disease

• Biologic basis for Type B dissection has been postulated. (Development, 2000)
  - VSMCs from cardiac neural crest are also present at the isthmus

• MFS: time onset for Type B dissection after elective root aneurysm surgery is 14 yrs.

• DTA intervention after Type A dissection is 2.5 yrs.

• 50% MFS pts will require DTA surgery over a mean of 26yrs. (JTCVS, 2009)
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. (Song, Circulation, 2009)
  • Distal ascending and proximal arch are usually guidewire related.
  • Whole arch dissection is usually stent-graft induced (80%+).
• MFS pts accounted for 12% of rAAD, yet were only 1% of the whole study population.
• 50% necropsy showed rAAD from bare springs, all MFS.
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).
Fate of Aorta after TEVAR in MFS

• Beck, 16 pts (JVS, 2012):
  • 12% perioperative mortality.
  • 44% required surgical conversion in 32 mos follow-up.
  • All pts who had a 2nd TEVAR progressed to surgery.
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, European Guidelines
Graft to Graft Seal Zones…..

6 cm longitudinal distance to place 4 fenestrations and all anterior on hemicircumference

Branched v. Fenestrated?
Back Table Fenestration: Cook TX2 34x152mm

Celiac 8mm
SMA 8mm
L Renal 6mm
R Renal 6mm
Wire Access to Target Vessels
Follow-up 3D CTA:
No Endoleak Seen up to 48 mos
MFS TEVAR: *Bridge to definitive surgery*

- **preop**
- **1mo**
- **6mo**
- **Referred**

*TEVAR*(chronic)  New SINE  
New AD  TAAA

- Allowed referral to our center where open TAAA surgery has matured excellent results.
Conversion technique after TEVAR for TAAA.
Endovascular Therapy in Marfan Syndrome 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
  - “Bridge” to referral
Thank you

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