Updates on Arterial Complications in Marfan Syndrome, Ehlers-Danlos Syndrome: Endografts Can be Valuable

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Disclosures

• Consultant: Cook, WL Gore.

Procedural Considerations in CTD

• Done in operating room (+/- GA)
• Strict BP control
  • Reduce catheter whip
• Induced hypotension
  - SBP < 90 mmHg for endo
  - SBP 70-80 mmHg for surgery
• Careful crossing-selection in dissected vessels of VEDS
  - Celiac
  - External iliac

Endovascular frontier.....

• Think creatively...

Endovascular frontier.....

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Endovascular frontier.....

• Operating room-catheter suite hybrid room..

Selection of VEDS Patients for Therapy:

*influence of specific genotypes*

• COL3A1 mutation is the etiology of VEDS; (Gly-X-Y).
  – “Frame shift” due to glycine substitution
  – Destroys the 3D architecture of the Collagen 3 trimer
  – Collagen 3 is degraded in the cell & not secreted, total mass <10% of nl.

• COL3A1 mutations can be haploinsufficient.
  – Early “stop” in transcription, thus some Type 3 collagen is created
  – Total mass ≈ 50% of nl → Better handling

Stent-graft therapy in CTD

• 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 Dissections in MFS

• The arch and ascending are at risk for rAAD. (Dong, 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 rAAAD cases, yet the overall rate of rAAD was only 1.3% of total.
  – “RTAD was the most common complication in Marfan patients after stent-graft therapy”

Pre-Procedure

Back Table Fenestration:
Cook TX2 34x152mm

Celiac 8mm
SMA 8mm
R Renal 6mm
L Renal 6mm
Follow-up 3D CTA: No Endoleak Seen up to 36mos

MFS TEVAR: Bridge to definitive surgery

1wk 1mo 6mo Referred

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

Open TAAA and TEVAR in CTD

21 yo MFS woman studying abroad in Barcelona: acute Type B with SMA malperfusion

18mos

• Future is likely for complementary roles of competing therapies

Conversion technique after TEVAR for TAAA.

Contemporary CTD Management

• Multidisciplinary evaluation.
• Liberal use of adjunctive techniques to reduce operative trauma in the endovascular & open setting.
• Procedures should be in the operating room setting versus “cath-lab.”
• Stent-graft therapy in CTD is defined in limited fashion.

Thank you