Current Management Strategies for Ehlers-Danlos Syndrome and the Role of Endografts in Arteriopathy

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Ehlers-Danlos Syndrome (EDS)

- Hereditary Connective Tissue Disorder
  - Mutations in genes regulating collagen matrix

- Six Different EDS Subtypes
  - Classical, Hypermobility, Vascular, Kyphoscoliotic, Arthrochalasic, & Dermatosparactic.

- Characterized by Joint Hypermobility, Skin Hyperextensibility, & Tissue Fragility

Procedural Considerations in VEDS

- 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

Disclosures

- Consultant: Cook, WL Gore, Medtronic.

Truth, Lies, and Statistics.....

- Truth:
  - Surgery, even in the best of hands, is likely still a risky proposition.

- Lies, or at least urban legend:
  - There is no evidence to suggest a VEDS artery with normal diameter and no pathology ruptures

- Statistics:
  - Most common arterial pathology is pseudoaneurysm, and these are often Ax

Endovascular frontier.....

- Think creatively...
Endovascular frontier.....
- Cath lab...

Endovascular frontier.....
- Think creatively...

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.

VEDS: “Room in the Inn” for endografts?
1. Rupture of Type B dissections.
2. Proximal fixation in surgical graft.
3. Distal stent graft could be managed with conversion.
4. Compliant with follow-up.

AND,
1. Device can be placed without conduit (iliac fragility).
2. Minimal oversizing (<10%).
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 VEDS is defined in limited fashion.

Thank you