



## Operations for Patients With Vascular-Ehlers Danlos Syndrome: How to Make it Go Well!

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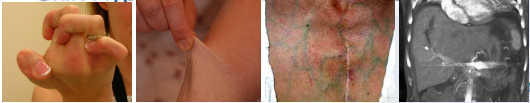
## Disclosures

- None



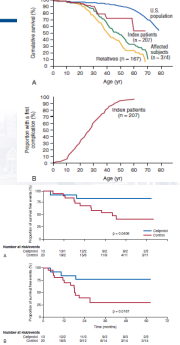
## Ehlers-Danlos Syndrome (EDS)

- 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

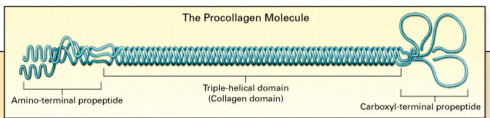


## Patient Management and Selection


- Vascular events increase with age.
  - 30% patients have family history of event
  - Over assignment of relatives w/ early demise to EDS
  - Malignant family history (esp<40)
- Medical management with beta blockade may reduce frequency of events (Lancet, cell/protol)



## Collagen structure

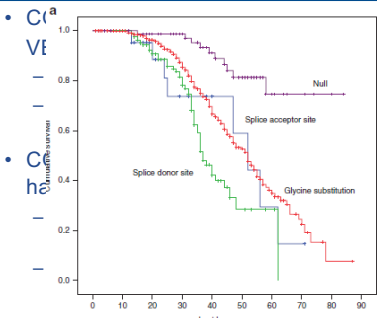
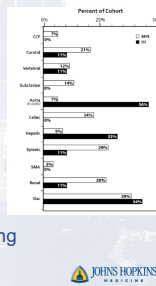



Gene	Polypeptide	Protein	Proportion of Total Protein	Function
	Normal polypeptide		1/8	Normal
COL3A1	Normal polypeptide		3/8	Abnormal
Mutated COL3A1	Mutated polypeptide		3/8	Abnormal
	Mutated polypeptide		1/8	Abnormal

From NEJM Pyeritz 342 (10): 2000 

## Selection of VEDS Patients for Therapy: influence of specific genotypes (Shalub, JVS, 2014)

- $Ct^a$  VE
- $Che$

### Truth, Lies, and Statistics.....

– Most common arterial pathology is pseudoaneurysm, and these are often Asx

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### Endovascular frontier.....

- Think creatively...

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### Endovascular frontier.....

- Cath lab...

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### Endovascular frontier.....

- Think creatively...

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### Endovascular frontier.....

- Operating room-catheter suite hybrid room..

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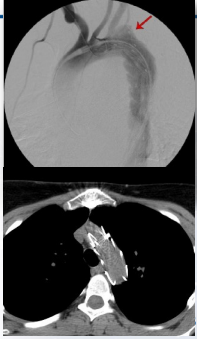
### Procedural Considerations in VEDS

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

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### 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.

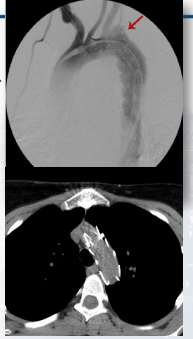


### VEDS: "Room in the Inn" for endografts?

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.

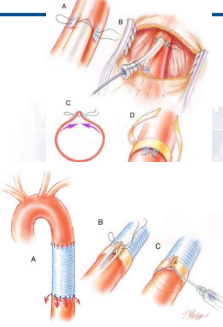
**AND,**

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



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