Stent-Grafts and Endovascular Repair can be a Reasonable Option for Treating Aneurysm Patients with Connective Tissue Disorders and Ehlers-Danlos Syndrome?

Vincent Riambau, MD, PhD
Professor and Chief of Vascular Division
Hospital Clinic, University of Barcelona

Yes, in selected cases

The wide spectrum of Connective Tissue Disorders related to Aortic Aneurysms

- Marfan syndrome
- Loeys-Dietz syndrome
- Ehlers-Danlos Syndrome
- Marfan Thoracic Aortic Aneurysm Family
- Aneurysms associated with osteoarthritis
- Arterial tortuosity syndrome
- Turner syndrome
- Aortic dissection in non-dissecting Aortic Aneurysms
- Neurofibromatosis Type I
- Autosomal Dominant Polycystic Kidney Disease

CTD are not candidates for endovascular repair

MFS patients

Acceptable early outcomes

High re-intervention rates

Aortic Endovascular repair (TEVAR/EVAR) in MFS

<table>
<thead>
<tr>
<th>Year</th>
<th>Patients</th>
<th>Survival</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>2003</td>
<td>1</td>
<td>100%</td>
<td>Hybrid approach</td>
</tr>
<tr>
<td>2005</td>
<td>6</td>
<td>0%</td>
<td>100% at 4 yrs</td>
</tr>
<tr>
<td>2006</td>
<td>15</td>
<td>9%</td>
<td>1.7% redo</td>
</tr>
<tr>
<td>2006</td>
<td>2</td>
<td>0%</td>
<td>2 redo</td>
</tr>
<tr>
<td>2007</td>
<td>2</td>
<td>0%</td>
<td>Talent</td>
</tr>
<tr>
<td>2008</td>
<td>2</td>
<td>0%</td>
<td>Talent</td>
</tr>
<tr>
<td>2012</td>
<td>16</td>
<td>6.23%</td>
<td>0% redo</td>
</tr>
<tr>
<td>2014</td>
<td>10</td>
<td>3.35%</td>
<td>1.6% TEVAR</td>
</tr>
</tbody>
</table>
Stent-graft for failed open surgery
Endovascular repair of thoracic aortic pseudoaneurysms and patch aneurysms

This important series is not included in Bergqvist’s review


Vascular Repair in EDS
Treatment of Vascular Ehlers-Danlos Syndrome
A Systematic Review

David Bengtsson, MD, PhD, Anu I. Lepanto, MD, PhD, and Angela Wedel, MD, PhD

Background: Vascular Ehlers-Danlos syndrome (EDS) is a rare connective tissue disorder with serious hemodynamic consequences. Most experience on treatment is based on case reports and small case series.

Methods: A systematic literature review was performed. PubMed and other relevant databases were searched.

Results: A total of 28 patients were identified with no gender predominance. Hemorrhages were present in 8%, whereas rupture in 17%. There was an aortic rupture without an underlying aneurysm. Cumulative rupture incidence was 10%. After surgery, the mortality was 20%, after endovascular procedures, 22%, in a group of miscellaneous cases, it was 33%

Conclusion: Vascular EDS is a serious disease with high mortality, which does not seem to have been influenced by new treatment methods. Problems in diagnosis should be sought only when necessary, primarily to identify the problematic patient. Other causes, the genetic molecular defect should be identified. The results of this review may be affected by publication bias. Finally, a case report should be included.


Vascular Repair in EDS
Contemporary management of vascular complications associated with Ehlers-Danlos syndrome

Bengtsson D, Bengtsson L, Halk H. Male factor for EDS? A systematic review

Case report
34 yo female, few weeks after the birth of her daughter...
We address consultations...

The John Hopkins answer

The Cleveland Clinic Response

Stanford University response

Rostock response
Endovascular tools and techniques are needed to manage vascular complications in connective tissue disorders.

- Stent grafts could be useful as a primary intervention,
  - Alone (high risk patients) or in combination with open surgery
  - As a bridge to open repair (emergencies)
  - After failed OR (redo)

No consensus is achieved nowadays about therapeutical strategies.

Connective tissue diseased patients should be addressed to aortic centers with large experience in every single aortic repair technique.

Close surveillance is recommended in any case.