Clinical Outcomes Of TBADs In Patients With Connective Tissue Diseases (Marfans, Loey-Dietz, etc) Are Worse Than In Those Without

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No relevant financial disclosures

Marfans Syndrome (MFS)
- Autosomal Dominant
- Fibrillin I TGFBR2
- Variable Expression – mild to severe
- 25% Patients give no family history
- CV, ocular, skeletal>Lung, skin, dura
- Key symptom= Thoracic aneurysm/dissection <50 yo

Loeys-Dietz (LDS)
- Autosomal Dominant
- TGFBR1 (1/3) TGFBR2(2/3)
- Similar to MFS – early onset aortic root dilatation, early onset dissection
- + generalized arterial tortuosity, aneurysms throughout arterial tree
- Dissections occur at a smaller aortic diameter than MFS
- Life expectancy reduced compared to MFS (mean age death 26 years)
- Increased risk pregnancy associated complications
LDS
• 54 yo Female
• Aortic root and valve replacement 2011 55mm ascending
• Peripheral aneurysms
• Aortic tortuosity
• 2012 JRAAA – operated 45mm
• Regular MRI/CT surveillance

Ehlers Danlos IV
• AD
• COL3A1 - Collagen
• 50% Thoracic/abdominal aorta
• 50% Peripheral
• 12% risk of death in pregnancy

34 yo Male
• 3 siblings – all with ascending aortic repair
• Rapidly expanding, dissected JRAAA
• Threshold Type I TAAA

Imperial Dissection Database 2005-2015

Imperial Type B: Intervention Group (37)

Mortality:
• In Hospital mortality = 3/37 (8%)
  - 2 retrograde dissections, 1 MOF
• Late mortality: 3 (8%)
  – paraplegia, multiple endoleaks, late rupture

Complications:
• Retrograde dissection 2 (5%)
• Transient paraparesis 2 (5%)
• Paraplegia 1 (2.7%)
• Respiratory major 5 (13.5%)
• Respiratory minor 10 (27%)
• Cardiac 6 (16%)
• Acute renal failure 10 (27%)
• GI 8 (21%)
Acute TBD: Results

Re-interventions (surgical and endovascular) : 21%

- 5 Early re-interventions (within 1 year): 2 acute Type A repairs (1 died), open Type IV (1), TEVAR relining (1), oesophagectomy (1), RSA aneurysm repair (1), JRAAA (1) – ALL MARFANS

- 3 Late re-interventions: 1 Type Ia endoleak at 10 yrs needing full arch hybrid, 1 Type III – relining, 1 fistula, 1 Type IV repair – 2/3 MARFANS

Acute Type B: Surveillance Group (33)

- 12 dilated >40mm

- 3 dilated >55mm

- 4 required intervention (2 Type As, 1 jRAAA, 1 renal stent

- 4 died in follow up – ALL connective tissue

Chronic TBD : Interventions

<table>
<thead>
<tr>
<th>Emergent Presentation (33)</th>
<th>Elective Presentation (137)</th>
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</thead>
<tbody>
<tr>
<td>TEVAR 16</td>
<td>TEVAR 16</td>
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<tr>
<td>Arch Hybrid 3</td>
<td>Arch Hybrid 15</td>
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<tr>
<td>Visceral Hybrid 3</td>
<td>Visceral Hybrid 20</td>
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<tr>
<td>Type III 1</td>
<td>Type III 1</td>
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<tr>
<td>Type IV 2</td>
<td>Type IV 2</td>
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<tr>
<td>JRAAA 4</td>
<td>JRAAA 2</td>
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<tr>
<td>FEVAR 2</td>
<td>FEVAR 2</td>
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<tr>
<td>EVAR 1</td>
<td>EVAR 1</td>
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<tr>
<td>Connective Tissue 21%</td>
<td>Connective Tissue 14%</td>
</tr>
</tbody>
</table>

Chronic TBD: Results

Mortality:

- In Hospital mortality = 7/94 (7.4%)

Complications:

- Retrograde dissection 1 (1%)

- transient paraparesis 5 (5.3%)

- paraplegia 6 (6.3%)

- Stroke 5 (5.3%)

- Respiratory major 12 (12.7%)

- Respiratory minor 25 (26.5%)

- Cardiac 10 (10.6%)

- Acute renal failure 18 (19.1%)

- GI 15 (16%)

Summary

- 18% of all TBD – connective tissue disorder

- Acute in-hospital mortality 33%

- Acute late deaths 33%

- Acute early re-interventions 100%

- Acute late re-interventions 66%

- Deaths in surveillance 75%

- Chronic in-hospital mortality 57%

- Chronic re-interventions 50%
Conclusions
Patients with connective tissue disease and TBD:

- Higher periprocedural and later mortality
- Higher early and late complication rate
- Higher re-intervention rate
- Higher risk in surveillance