When And How To Treat Patients With Genetic Aortic Disorders By Endovascular Aortic Techniques?

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Disclosures

- Consultant: Cook Medical, Philips, Getinge, Terumo Aortic, Arterica, Medyria
- Research-grants: Cook Medical, Philips, Terumo Aortic, Medtronic
- Travel-grants: Cook Medical, Getinge proctoring speaking-fees,
- Speaking fees: Cook Medical, Philips, Getinge
- Shares: Mokita-Medical, Arterica, Medyria, Siemens, Philips
- IP: Cook Medical, Terumo Aortic, Mokita Medical
- Royalties: Cook Medical, Terumo Aortic
Open TAAA-Repair
Open TAAA Repair
Endovascular TAAA Repair
Genetisches Aorten Syndrom (GAS)
The Dogma in GAS-Patients

- GAS-patients are young and fit
- Open TAAA-repair is well tolerated by GAS-patients
- GAS-patients do not tolerate well stent-grafts in native landing zones.
- Endovascular therapy cannot offer good long-term results
GAS: Genetisches Aorten Syndrom
The Reality in GAS-patients:

* GAS-patients frequently are old, have multiple prev. operations and are cardio-pulmonarily compromised.

* Open TAAA-repair is not tolerated very well in many GAS-patients and they are frequently turned down for OR.

* GAS-patients tolerate stentgrafts in native landing zones under certain circumstances well.

* Endovascular Therapy can offer good long-term results in GAS-patients.
**ESVS-Guidelines**


Anders Wanhainen, Fabio Verzini, Isabelle Van Herzeel, Eric Allaire, Matthew Bown, Tina Cohnert, Florian Dick, Joost van Herwaarden, Christos Karkos, Mark Koelemay, Tilo Köhler, Ian Loftus, Kevin Mani, Germano Melissano, Janet Powell, Zoltán Széderin

**Recommendation 123**

In young patients with suspected connective tissue disorders and abdominal aortic aneurysms, open surgical repair is recommended as first option

<table>
<thead>
<tr>
<th>Class</th>
<th>Level</th>
<th>References</th>
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<tr>
<td>I</td>
<td>C</td>
<td>[785, 782]</td>
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**Editor’s Choice — Management of Descending Thoracic Aorta Diseases**

Clinical Practice Guidelines of the European Society for Vascular Surgery (ESVS)


**Recommendation 72**

In patients with genetic syndromes associated with thoracic aortic aneurysm dilatation ≥50 mm, surgery should be considered

<table>
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**Recommendation 73**

In patients with a genetic syndrome and enlarged aortic diameter <50 mm, surgery may be considered according to body surface area in patients of small stature or for rapid progression or more aggressive diseases or with a family history of dissection

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**Recommendation 74**

In patients with connective tissue disorder, endovascular repair may be considered in redo operations or in emergencies as bridging procedures

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Wanhainen et al. 2019; Eur J Vasc Endovasc Surg 57:8-93

Riambau et al. 2017; Eur J Vasc Endovasc Surg 53:4-52
Why Are GAS-patients Treated Endovascular?

- Diagnosis unknown
- Not fit for OR
- Bridging between 2 replaced aortic segments
Early TEVAR-Experience in GAS

<table>
<thead>
<tr>
<th>Author</th>
<th>Number of patients</th>
<th>Mean (min)</th>
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<tbody>
<tr>
<td>Ince et al. [16]</td>
<td>6</td>
<td>51(1)</td>
</tr>
<tr>
<td>Nordon et al. [17]</td>
<td>6</td>
<td>16(3)</td>
</tr>
<tr>
<td>Geisbüsch et al. [18]</td>
<td>6</td>
<td>32.8(5)</td>
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<td>Botta et al. [19]</td>
<td>12</td>
<td>31(3)</td>
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<tr>
<td>Marcheix et al. [20]</td>
<td>15</td>
<td>25(1)</td>
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<tr>
<td>Waterman et al. [21]</td>
<td>15</td>
<td>9(0-31)</td>
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<tr>
<td>Eid-Lidt et al. [22]</td>
<td>9</td>
<td>59.6 (3-5)</td>
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<tr>
<td>Total</td>
<td>69</td>
<td>32</td>
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</tbody>
</table>
F/B EVAR in CTD

Endovascular Repair of Thoracoabdominal and Arch Aneurysms in Patients with Connective Tissue Disease Using Branched and Fenestrated Devices

Rachel E. Clough, Teresa Martin-Gonzalez, Katrien Van Calster, Adrien Hertault, Rafaëlle Spear, Richard Azzouzi, Jonathan Sobocinski, and Stéphan Haulon, Lille, France

- 2004-2015, 17 patients with GAS
- FU 3.4 years
- 16 TAAA, 1 arch aneurysm
- Technical success 100%
- No early mortality/stroke
- 1 Reintervention, 1 death, 1EL III

Cook Branched Arch Endograft

Hamburg Experience 2012-2017:

- **Cases:** 54
  - **Aneurysm:** 28 (52%)
  - **Residual dissection:** 24 (44%)
  - **Acute Type A:** 2 (4%)
  - **Elective:** 43 (80%)
  - **Urgent:** 11 (20%)
  - **CTD:** 5 (9%)

- **30d-Mortality:** 3 (6%)
  - **in CTD:** 0 (0%)

- **Stroke:** 6 (11%)
  - **in CTD:** 1 (20%)

Tsilimparis et al. 2018; submitted
Land in Replaced Vessels
EVAR/TEVAR in Patients with GAS

- Hamburg 2010-2018
- Age: 55 years
- 30 patients:
  - 23 Marfan
  - 5 Loeys-Dietz syndrome
  - 2 vasc. Ehlers-Danlos syndrome
- 20 TEVAR
- 10 EVAR
- 4 Hybrid-EVAR
- 6 F/B EVAR
EVAR/TEVAR in Patients with Marfan Syndrome

- 23 patients: 55 Jahre
- Aneurysm: 11
- Dissection: 12
  - TAAD: 4
  - TBAD: 8
- Elective: 18
- Urgent: 5
  - Rupture: 2
  - Symptoms: 3
EVAR/TEVAR in Patients with Marfan Syndrome

* Technical success 100%

* Proximal Landing:
  * Graft: 7
  * Native segment: 16

* Distal Landung:
  * Graft: 6
  * Native segment: 17

* ICU: 2 Tage

* 30d Mortalität 4 (17%)
  * Hybrid EVAR 2/4 (50%)
  * F/BEVAR 1/5 (20%)
6B-EVAR in GAS
4y post Zone 2 TEVAR in Marfan
5y post Zone 2 TEVAR und FEVAR in Marfan
8y post Zone 2 TEVAR in EDS IV
There is no place for dogmas in aortic surgery

Patients with GAS expect to be offered modern contemporary minimally invasive techniques when justifiable

Endovascular Strategies should be considered in GAS-patients and preferred in high-risk patients

Treatment of GAS-patients should be centralised to aortic-centers that can offer all treatment options.
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University Heart & Vascular Center Hamburg

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• Aortic arch
• Thoracoabdominal aorta
• Aortoiliac disease

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