Managing the aortic root in pregnancy

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The aorta during pregnancy

- $\frac{1}{2}$ of all aortic dissections in women <40y occur during pregnancy (Hirst et al, Medicine (Baltimore) 1958)
- Pregnancy is associated with a 25-fold increased risk for dissection (Naziell et al, Eur J of Obst & Gyn 2010)

The putative association of pregnancy and acute dissection may be largely an artifact of selective reporting (Nienaber et al, Circulation 2004; Thalmann et al, EJCTS 2011)

- The causal relation between pregnancy and aortic dissection is largely overestimated (Oskui et al, Am J Cardiol, 1994)
- Pregnancy is a revealer of aortic fragility and not a factor increasing aortic deficiency (Pacini et al, IJC 2009)
The aorta during pregnancy

- Compression of aorta and iliac arteries → ↑ outflow resistance in lower arterial tree

- ↑ in CO → initiate intimal tear

- Hormonal factors (oestrogens) → fragmentation of reticulin fibers
The aorta during pregnancy

- Compression of aorta and iliac arteries → outflow resistance in lower arterial tree
- Hormonal factors (oestrogens) → initiate intimal tear
  - Non-genetic: hypertension, atherosclerosis
  - Inflammatory
  - Genetic
    - Syndromal: Marfan, FTAAD, Loeys Dietz, Bicuspid Aortic valve, Aneurysm-Osteoarthritis
    - Non-Syndromal: Turner Syndrome

+ underlying condition ↓
↑ risk for aortic dissection
The aorta during pregnancy

The IRAD registry

15 pregnant/3712 Aortic Dissections

- 5 type A – mean ARD 5.2cm
- 6 type B - mean Ao diam 3.3cm

11 complete data: age 33y

- 1 1st Trim
- 3 3rd Trim
- 7 postpartum

8 underlying CTD:
- 4MFS
- 1 BAV
- 1 LDS
- 1 EDS
- 1 FTAAD

Braverman et al, JACC 2012
The aorta during pregnancy

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# Pregnancy in Marfan syndrome

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<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>N (px)</th>
<th>Aortic event</th>
<th>Management</th>
<th>MFS dx</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Pyeritz 1981</td>
<td>Retrospective</td>
<td>26 (105)</td>
<td>0</td>
<td>?</td>
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<td>1% risk for serious cardiac complications if nl AO</td>
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<td>Rossiter 1995</td>
<td>Prospective</td>
<td>21 (45)</td>
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<td>Low complication risk if specialized care - ↑long term complic</td>
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<td>18 (22)</td>
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<tr>
<td>Mulder 2012</td>
<td>Meta-analysis</td>
<td>78 (145)</td>
<td>4 (type B)</td>
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# Pregnancy in Marfan syndrome

## Study Design

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## Specialized Care

- Diagnosis known/counseling prior to px
- Ao 40mm – Growth Rate
- β-Blockers
- Strict FU
- Tertiary Care Center
- Delivery Plan
- C- section when Ao >40-45mm
Pregnancy in Marfan syndrome

• Outcome after aortic root surgery

Pre-pregnancy aortic root replacement does not normalize the risk of pregnancy in a woman with Marfan syndrome and aortopathy.

- 3 elective
- 2 progressive AR
- 2 emergency for type A
- 2 type B dissection

Mulder and Meijboom, JACC, 60, July 2012
Rossiter et al, Am J Obstetr Gynec 1995

Donnelly et al, JACC July 2012
Meijboom et al, Eur Heart J 2005
Pregnancy in BAV

• No dissections in a series of 88 women (216 pregnancies) in Olmsted County
  – 6 had aorta >40mm; 1 >50mm

• Documented progression of aortic dilatation in 23 patients (4mm/y)

• No association between gravidity and AVR, development of aortic dilatation, or surgery of the ascending aorta

➢ risk of pregnancy-associated dissection in BAV is low.

= Consistent with previous population-based studies in pts with normally functioning BAVs in which no aortic dissections were observed.

Pregnancy in Turner Syndrome

• Aortic dissection in TS: 1.4% (Gravholt et al, Cardiol in the Young 2006)

• Retrospective series of 85 cases:
  – Age at dissection 30.4y
  – Often associated HTN/CHD
  – 7 dissection during pregnancy (after assisted reproductive technology)

• Diffuse and disorganized rapportation

ITSD registry (Carlsson and Silberbach, BMJ 2009)
Pregnancy in Turner syndrome

ITSAD registry

20 aortic dissections

Age 18-48y

17 type A

2 pregnancy related

Absolute ARD 4.1cm

Aortic index 2.8cm/m²

Silberbach M, Gentac meeting 2012
## Guidelines

<table>
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<td>Marfan</td>
<td><strong>β</strong>-blockers recommended&lt;br&gt;Surgery pre px if AR&gt;45 mm (IC)</td>
<td>Risk of major CV complications is low if AR&lt;40mm</td>
</tr>
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<td>BAV</td>
<td>Surgery pre px&lt;br&gt;If AR &gt;50mm (or &gt;27 mm/m^2 BSA) (IIC)</td>
<td>Women with AR&gt;45mm should be counseled about high risk</td>
</tr>
<tr>
<td>Turner</td>
<td>Surgery pre px if index &gt;27mm/m^2)</td>
<td></td>
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<td>General</td>
<td>• Counseling pre px (IC)&lt;br&gt;• Consider caesarean delivery If AR&gt;45mm (I C)&lt;br&gt;• If type B dissection: px contraindicated (III C)&lt;br&gt;• Strict BP control (IC)</td>
<td>• Counseling pre px (class I)&lt;br&gt;• Caesarian section if “enlargement” (class IIa)&lt;br&gt;• Prophylactic surgery if progressive dilatation or AR (class IIb)</td>
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Conclusions

• Pregnancy confers a risk for aortic dissection in the presence of an underlying condition
• Outcome is better in patients with
  • Known diagnosis
  • Special Care
• Pregnancy affects long term outcome in Marfan syndrome
• Pre-pregnancy aortic root replacement does not normalize the risk
• BAV may be less severe
• Turner may be more severe
• Need for prospective multicentre trials
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