Aortic Aneurysm and Dissection in Pregnancy - a Rare Case in a Marfan Woman with BAV: Echocardiographic Findings and Review of the Literature

T. Costagliola, R. Leone, P. Masiello, G. Mastrogiovanni, F. D’Auria, A. Longobardi and S. Iesu
Aortic dissection in pregnancy is one of the most life-threatening occurrences, but fortunately it is rare. There is an important association between this event and bicuspid aortic valve, as well as Marfan Syndrome, due to the involvement of connective tissue.

We describe a rare case of undiagnosed ascending aortic dissection occurred in a 22-year-old pregnant woman, affected by Marfan Syndrome and bicuspid aortic valve.
Our Case (1)

- 22 years old woman with Marfan syndrome
- Ascending aortic aneurysm
- Bicuspid aortic valve
- Familiarity for aortic dilatation
- Normal blood pressure; Atenolol 25mg/die
- No other risk factors
- Routinely trans-thoracic echocardiography to measure the aortic root and the growth rate of the AAA.
- Pregnancy! According to the present guidelines it was considered safe to continue the pregnancy.

Echocardiographic assessment of the aortic root should include—in addition to determining the maximum diameter—measurements at the ring, sinus, sinotubular junction, and distal ascending aortic levels.

Pregestational TTE parasternal long axis view of the aortic root and AAA.
Our Case (2)

During labor:
- stabbing chest pain, irradiated to the neck and the epigastrium spontaneously solved.
- ECG and blood tests came back normal, but no TTE was done.
- A delayed Caesarian section was performed without any known complications.

Five months later without symptoms:
- TTE check-up pointed out a chronic Stanford Type A ascending aortic dissection involving the aortic arch
- treated by the replacement of the ascending aorta sparing the BAV (Tirone David operation) combined with the replacement of the aortic arch and the supra aortic vessels.

A: Pregestational TTE parasternal long axis view of the aortic root and AAA. B: five months after delivery, parasternal short axis view of the BAV. C: ascending aortic dissection. D: 5 chamber view of the aortic dissection.
The changes in diameters:

<table>
<thead>
<tr>
<th></th>
<th>Pre gestational</th>
<th>28th week of pregnancy</th>
<th>5 months after delivery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Annulus</td>
<td>23 mm</td>
<td>23 mm</td>
<td>24 mm</td>
</tr>
<tr>
<td>Sinus of Valsalva</td>
<td>34 mm</td>
<td>34 mm</td>
<td>42 mm</td>
</tr>
<tr>
<td>Sino Tubular Junction</td>
<td>30 mm</td>
<td>36 mm</td>
<td>39 mm</td>
</tr>
<tr>
<td>Ascending Aorta Diameter</td>
<td>38 mm</td>
<td>38 mm</td>
<td>52 mm</td>
</tr>
<tr>
<td>Z-score</td>
<td>-1.89</td>
<td>0.217</td>
<td>0.871</td>
</tr>
</tbody>
</table>

Postoperative ETT: parasternal long axis (A-B) and short axis (C-D) view of the aorta, showing aortic reconstruction and minimal aortic insufficiency, due to an imperfect adhesion of the cusps.

WHO risk classification

- **WHO II-III** (depending on individual)
  - Mild left ventricular impairment
  - Hypertrophic cardiomyopathy
  - Native or tissue valvular heart disease not considered WHO I or IV
  - Marfan syndrome without aortic dilatation
  - Aorta <45 mm in aortic disease associated with bicuspid aortic valve

**Conditions in which pregnancy risk is WHO IV**

- Marfan syndrome with aorta dilated >45 mm
- Aortic dilatation >50 mm in aortic disease associated with bicuspid aortic valve
How About Guidelines?

Moreover:

Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults

2. All patients with a bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation. (LOE: B)
**ESC Guidelines on the management of cardiovascular diseases during pregnancy**

**Table 11: Recommendations for the management of aortic disease**

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Women with Marfan syndrome or other known aortic diseases should be counselled about the risk of aortic dissection during pregnancy and the recurrence risk for the offspring</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Imaging of the entire aorta (CT/MRI) should be performed before pregnancy in patients with Marfan syndrome or other known aortic diseases</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Women with Marfan syndrome and an ascending aorta &gt;45 mm should be treated surgically pre-pregnancy</td>
<td>I</td>
<td>Ila</td>
</tr>
</tbody>
</table>

**Recommendations on interventions on ascending aortic aneurysms**

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter ≥50 mm for patients with Marfan syndrome</td>
<td>I</td>
<td>C</td>
</tr>
</tbody>
</table>
| Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameters:  
  • ≥45 mm for patients with Marfan syndrome with risk factors  
  • ≥50 mm for patients with bicuspid valve with risk factors  
  • ≥55 mm for other patients with no elastopathy | Ila | C |
| Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid progression, aortic valve regurgitation, planned pregnancy, and patient’s preference | Iib | C |

**2014 ESC Guidelines on the diagnosis and treatment of aortic diseases**

**Recommendations for the management of aortic root dilation in patients with bicuspid aortic valve**

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
</table>
| In cases of BAV, surgery of the ascending aorta is indicated in case of:  
  • aortic root or ascending aortic diameter >55 mm  
  • aortic root or ascending aortic diameter >50 mm in the presence of other risk factors  
  • aortic root or ascending aortic diameter >45 mm when surgical aortic valve replacement is scheduled | I | Ila |
| Beta-blockers may be considered in patients with BAV and dilated aortic root ≥40 mm.  
Because of familial occurrence, screening of first-degree relatives should be considered | Iib | C |

**Cardiac Surgery Division**

Salerno, Italy
the lack of detailed clinical data precluded an assessment of the risk of aortic complications in relation to patients’ aortic root diameter before pregnancy. Therefore, we cannot comment on whether pregnancy increases the risk of aortic complications in women with an aortic root diameter <40 mm or <45 mm, which are the thresholds currently recommended for deciding on the safety of pregnancy in women with Marfan syndrome or other connective tissue disorders.
Pregnancy-related acute aortic dissection in Marfan syndrome: A review of the literature

April 2017
Congenital Heart Disease 12(4)
DOI 10.1111/chd.12465
Katherine Smith; Bernard Gros

Current guidelines rely on aortic root diameter for stratification of Marfan women into risk categories, but we identified several cases that would be missed by these guidelines. Specifically, the existing literature suggest that women with Marfan syndrome should take precautions throughout pregnancy, rather than the third trimester.

Conclusions

Patients with MFS, especially those whose initial aortic diameters ≥40 mm, planning a pregnancy or currently pregnant should be carefully counseled about the maternal and fetal risks throughout pregnancy. MFS patients whose aortic diameters ≥40 mm should be advised to ideally await pregnancy until prophylactic aortic surgery. As MFS varies in its phenotypic expression, each patient's risk of adverse cardiac events should be assessed individually through a joint Maternal Fetal Medicine and Cardiology Center.
Take home message

• Because of its rarity, pregnancy in patients with MFS remains a debated topic.
• Women with Marfan syndrome seem to well tolerate the pregnancy, up to an aortic root diameter of 45 mm, with good clinical care before, during, and after pregnancy.
• When possible surgical repair of enlarged aortic root in risk patients should be done before starting pregnancy.
• The 2010 AHA guidelines recommend the prophylactic aortic root replacement if the aortic root diameter exceeds 40 mm. However, this data is not in line with the European guidelines, which report that an aortic root diameter of 45 mm is actually safe. After our experience, we think that the strategy should be consistent with the stricter guidelines.
• When pregnancy starts, a strict prevention of systemic hypertension is mandatory and beta blocker use is recommended, as well as close echocardiographic surveillance.
• Dissection should be immediately treated by surgery and, after 30 weeks, Cesarean section and immediate surgery is the best treatment.
Thank you for your attention!