

## Clinical Guideline

**MANAGEMENT OF AORTOPATHY IN PREGNANCY**

<b>SETTING</b>	University Hospitals Bristol
<b>GUIDELINE FOR</b>	The cardiac obstetric multidisciplinary team
<b>PATIENT GROUP</b>	Pregnant women with dilated aortas, including those with Marfan's Syndrome (MFS), Loeys-Dietz Syndrome (LDS), vascular Ehlers-Danlos Syndrome (vEDS), Turner's Syndrome (TS) and related connective tissue diseases (CTDs) causing aortopathy, familial thoracic aneurysm and dissection syndrome, bicuspid aortic valve (BAV) with associated aortopathy, post Ross operation, or congenital heart repair associated with enlargement of the ascending aorta.

**GUIDANCE**

Pregnant women with dilated aortas, particularly those with Marfan's Syndrome (MFS), Loeys-Dietz Syndrome (LDS), vascular Ehlers-Danlos Syndrome (vEDS), Turner's Syndrome (TS) and other inherited aortopathies, are at risk of aortic dissection in pregnancy and in the post-partum period due to haemodynamic changes and hormonal effects on the abnormal arterial wall. The highest risk period is in the third trimester, peripartum and the early post-partum period. For the purposes of this document, MFS refers to Marfan's Syndrome and those conditions with a high risk of aortic dissection as above. The risk has been studied most in patient with MFS and BAV:-

In MFS, dissection risk is increased in women with:

- a family history of dissection;
- previous aortic dissection or aortic surgery;
- aortic regurgitation; or
- a rapidly growing aorta.

Risk is thought to be reduced by beta-blockade.

Aside from patients with LDS and vEDS who are at high risk of dissection, regardless of aortic size, dissection is unlikely in pregnancy if the aorta measures <45mm\* and none of the above features are present. However, the clinician must be mindful that dissection can still occur and Type B dissection in particular, cannot be predicted by aortic root size.

All patients with Marfan's syndrome should be considered high-risk pregnancies and be managed in a cardiac surgical centre. Vaginal delivery with a passive second stage of labour is the aim, unless the aortic root measures >45mm, in which case elective Caesarean section for cardiac reasons is recommended. Trans-thoracic echocardiography should be undertaken at least 6 weekly until 6 weeks post-partum. All patients should have blood pressure carefully controlled, ideally with beta-blockers if tolerated and undergo 4 weekly growth scans from 26 weeks gestation (due to beta-blocker associated IUGR).

BAV aortopathy is relatively common and the risk of dissection is lower with few dissections reported in registries and one prospective longitudinal study. The ascending aorta is more often dilated, rather than the aortic root. Pregnancy should be avoided if the aorta measures >50mm. As with MFS blood pressure control and surveillance of the aorta by echocardiography is

recommended, though 6-8 weekly echoes are sufficient unless the aorta is >45mm. Vaginal delivery should be the aim and elective Caesarean section for cardiac reasons is not necessary.

Patients can be risk-stratified as follows,

**EXTREMELY HIGH RISK:** MFS with Sinus of Valsalva measuring >45mm  
 MFS with a growing sinus of Valsalva  
 MFS with previous surgery/dissection  
 All LDS and all vEDS

**VERY HIGH RISK:** MFS with Sinus of Valsalva measuring 40-45mm  
 MFS with aortic regurgitation  
 MFS with family history of dissection  
 All TS

**HIGH RISK:** MFS with Sinus of Valsalva measuring <40mm  
 BAV aortopathy with ascending aorta >45mm

**MODERATE RISK:** BAV aortopathy with ascending aorta 40-45mm

**LOW RISK:** BAV aortopathy with ascending aorta <40mm

**The following table is a guideline for the management of these patients.**

	Beta-block	Frequency of scan	Elective epidural	Second Stage
MFS, LDS, vEDS, TS	All	4 weekly	All	Elective Caesarean section if <b>EXTREMELY HIGH RISK</b> , consider in <b>VERY HIGH RISK</b> Passive for VERY HIGH RISK and HIGH RISK
BAV with aorta>50mm	Yes	4 weekly	All	Passive
BAV with aorta 45-50mm	Yes	4-6 weekly	All	Semi passive with passive descent and up to 30 minutes pushing
BAV with aorta 40-45mm	No	6-8 weekly	Low threshold	Management of second stage as per usual obstetric guidelines
BAV with aorta <40mm	No	8-12 weekly	No	Management of second stage as per usual obstetric guidelines

Patients who have had the **Ross operation** or other repaired congenital heart defects associated with an enlarged aorta should be treated as for BAV aortopathy. Other thoracic aortic aneurysm syndromes should be treated as for MFS by a specialist team.

BAV +/- BAV aortopathy is common in TS. Pregnancy can occur in mosaic TS and with assisted conception. Death from aortic dissection has been reported in up to 2% pregnancies, though there is little evidence to guide management. Dissection can occur in the absence of hypertension or congenital heart disease. All patients should be treated as BAV with aorta >50mm with delivery in a surgical centre. Those with CHD or hypertension should be treated as

MFS as above with beta-blockade and consideration of elective Caesarean section.

\*Aortic measurements should be indexed to body surface area. This is especially important in TS.

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**RELATED DOCUMENTS**      Regional Referral Pathway

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**AUTHORISING BODY**      Obstetric Cardiology MDT

**SAFETY**      Careful management with an experienced multi-disciplinary team is advised with an individual care plan for each woman

**QUERIES**      Contact any of the following via switchboard

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