Clinical Guideline

MANAGEMENT OF AORTOPATHY IN PREGNANCY

SETTING	**University Hospitals Bristol**
GUIDELINE FOR	The cardiac obstetric multidisciplinary team
PATIENT GROUP	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 (FTAAD), 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), LoeysDietz 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.

Women should be counselled pre-pregnancy by experts in obstetric cardiology. Pregnancy should be avoided in women with vEDS, previous dissection, MFS with aorta >45mm, BAV with aorta >50mm and TS with aortic size index (ASI) >25mm/m^2.

In MFS, dissection risk is approx. 3-4%; Type A dissection risk is increased in women with:
- sinuses >45mm;
- a family history of dissection;
- previous aortic dissection or aortic surgery;
- aortic regurgitation; or
- a rapidly growing aorta.

Type B dissection is less predictable. Risk is thought to be reduced by beta-blockade, but not proven in pregnancy.

The risk of dissection in LDS and vEDS is not known but thought to be as high/if not higher than MFS. vEDS also carries a risk of uterine rupture.

BAV aortopathy is relatively common and the risk of dissection is lower with few dissections reported. The ascending aorta is more often dilated than the aortic root. Pregnancy should be avoided if the aorta measures >50mm.

Pregnancy can occur in mosaic TS and with assisted conception. Death from aortic dissection has been reported in up to 2% pregnancies. BAV +/- aortopathy is common in TS and increases dissection risk, as does hypertension, though dissection can occur in women with no cardiovascular disease. Aortic measurements should be indexed to body surface area/height in TS.

Mainstays of management are strict blood pressure control and surveillance by trans-thoracic echocardiography 4-12 weekly and up to 6 months post-partum, depending on risk. Surveillance by MRI may be required if the dilated portion is not visible on echo (e.g. distal ascending, arch or descending).

In patients with MFS and other heritable aortopathies beta-blockers (celiprolol in vEDS) should be considered. Women should undergo 4 weekly growth scans from 26 weeks gestation (due to beta-blocker associated IUGR).

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
BAV aortopathy with ascending aorta >50mm
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.

All high risk patients should be delivered in a surgical centre.

	Beta-block	**Frequency of scan**	**Elective epidural**	**Second Stage**
MFS, LDS, vEDS, TS	All	4 weekly	All	Elective Caesarean section if EXTREMELY HIGH RISK, consider in VERY HIGH RISK 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

Ergometrine should be avoided for all in the 3rd stage.

RELATED DOCUMENTS Regional Referral Pathway

REFERENCES

Regitz-Zagrosek V et al. Eur Heart J. 2018 Sep 7;39(34):3165-3241.

Silberbach M et al. Cardiovascular Health in Turner Syndrome: A Scientific Statement From the American Heart Association. Circulation: Genomic and Precision Medicine. 2018;11 (10).

AUTHORISING BODY Maternal Heart Team 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

Dr S Curtis, Consultant Cardiologist
Miss J Trinder, Consultant Obstetrician
Miss A Mohan, Consultant Obstetrician
Miss L Ashelby, Consultant Obstetrician