

MANAGEMENT OF AORTOPATHY IN PREGNANCY

SETTING	University Hospitals Bristol and Weston
GUIDELINE FOR	The cardiac obstetric multidisciplinary team
PATIENT GROUP	Pregnant women with dilated aortas, including those with Marfan Syndrome (MFS), Loeys-Dietz Syndrome (LDS), vascular Ehlers-Danlos Syndrome (vEDS), Turner Syndrome (TS) and related connective tissue diseases (CTDs) causing aortopathy, Hereditary Thoracic Aortopathy (HTA), 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 Syndrome (MFS), Loeys Dietz Syndrome (LDS), vascular Ehlers-Danlos Syndrome (vEDS), Turner 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. It is strongly recommended that pregnancy is avoided in women with vEDS, previous dissection, MFS with aorta >45mm, BAV with aorta >50mm and TS with aortic size index (ASI) >25mm/m².

In MFS, overall 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. Dissection 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. It is strongly

recommended that pregnancy is avoided if the aorta measures >50mm.

Pregnancy can occur in TS with assisted conception and mosaic TS. 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 well seen 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 regular growth scans (2-4 weekly dependent on obstetric factors and previous USS findings) from 26-28 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
Growing aorta (>3mm in previous year)
Previous HTA surgery or dissection
All LDS and all vEDS
TS with aorta >25mm/m²

VERY HIGH RISK: MFS with Sinus of Valsalva measuring 40-45mm
MFS with aortic regurgitation
Family history of dissection
HTA with dilated aorta
TS with dilated aorta <25mm/m²
BAV aortopathy with aorta >50mm

HIGH RISK: MFS with Sinus of Valsalva measuring <40mm
BAV aortopathy with ascending aorta 45-50mm
HTA or TS with normal aorta

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 echo	Elective epidural	Second Stage
Extremely high risk	All	4 weekly	All	Elective Caesarean section (CS)
Very high risk	Yes	4 weekly	All	Passive vaginal delivery or elective CS
High risk	Yes	4-8 weekly	All	Passive vaginal delivery
Moderate risk	No	8 weekly	Low threshold	Management of second stage as per usual obstetric guidelines
Low risk	No	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