South West Maternal Medicine Network

NHS University Hospitals Bristol and Weston NHS Foundation Trust

Clinical Guideline

MANAGEMENT OF AORTOPATHY IN PREGNANCY

SETTING	South West Maternal Medicine Network
GUIDELINE FOR	Maternal heart teams in the South West of England
PATIENT GROUP	Pregnant women with dilated aortas, including those with hereditary thoracic aortic disease (HTAD), non syndromic and syndromic (Marfan's Syndrome [MFS], Loeys-Dietz Syndrome [LDS], vascular Ehlers-Danlos Syndrome [vEDS] and related conditions; Turner's Syndrome (TS); bicuspid aortic valve (BAV) associated aortopathy, and congenital heart conditions resulting in aortic dilatation (post Ross procedure, post arterial switch, Tetralogy of Fallot).

GUIDANCE

Pregnant women with dilated aortas, particularly those with hereditary thoracic aortic disease (HTAD) non-syndromic and syndromic (Marfan's Syndrome [MFS], Loeys-Dietz Syndrome [LDS], vascular Ehlers-Danlos Syndrome [vEDS] and related conditions; Turner's Syndrome (TS) and bicuspid aortic valve (BAV) related aortopathy, 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 the early post-partum period.

Pre-pregnancy counselling

All women should be counselled prior to conception by an expert in obstetric cardiology. Counselling will cover the following: inheritance, which is often autosomal dominant - genetic testing should be offered where appropriate, including the option of preimplantation genetic testing if a pathogenic variant is known; up-to-date cross-sectional imaging; risks to mother and foetus; and the likely management antenatally, during delivery and postnatally. Medication should be reviewed.

Surgery may be considered prior to pregnancy in high-risk women, depending on risk, aortic size, family history, and genotype (see https://uhbw.mystaffapp.org/17332/document_view.pdf)

Risk of dissection

In **MFS**, dissection risk is widely reported as approx. 3-4%. Most cases occur in women who are not aware they have the condition. Dissection is relatively rare in well managed cases, unless the aorta is very dilated. Risk is increased with a family history of dissection, history of aortic surgery, aortic regurgitation, or rapidly growing aorta.Type B dissection is less predictable and may occur in women with no aortic dilatation. Dissection risk is thought to be reduced by beta-blockade, but this is not proven in pregnancy.

There is limited data in pregnancy in women with **LDS** but increasing evidence to suggest that some patients behave in a similar way to MFS. Women with pathogenic variants inTGFBR1/2 are likely to be at higher risk. Expert counselling is recommended.

Women with **non-syndromic HTAD** maybe treated in a similar way to MFS. Variants in PRKG1 and ACTA2 may confer higher risk of dissection at smaller diameters.

BAV aortopathy is relatively common but the risk of dissection is much lower. The ascending aorta is more often dilated than the aortic root. Root pathology is more concerning. Women should undergo fetal echocardiography due to the increased risk of congenital heart disease in the fetus.

vEDS women typically dissect without initial dilatation. It is advised that pregnancy is avoided in this group of women due to the high-risk of adverse events - pregnancy-related mortality 5%, life-threatening vascular events up to 10%, including vascular dissection or rupture, and uterine rupture. Perineal tears, haemorrhage and prematurity common. Risk may be lower with normal cross-sectional imaging and specific genetic variants (e.g. null mutations).

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, and coarctation, though dissection has been reported in women with no cardiovascular disease. Aortic measurements should be indexed to body surface area/height in TS. Aortic height index is increasingly preferred over aortic size index due to the high prevalence of obesity in these women. Increased risk of hypertension, gestational diabetes, haemorrhage and small for gestational age babies. Caesarean rate often high.

Aortic dilatation can be seen after repair of some congenital heart conditions, for example after the Ross operation (where the aorta is referred to as the neo-aorta [the original pulmonary valve and root] and after the arterial switch repair for transposition of the great arteries (again the aorta is referred to as the neo aorta for the same reason). In both cases there is a suture line between the neo-aortic root and the native aorta. Dissection is rare post Ross and has not been reported post arterial switch in large series. Surgery should be considered when the neo-aorta exceeds 50-55 mm in Ross patients. Mild aortic dilatation is commonly seen after repair of Tetralogy of Fallot and is rarely of any consequence.

Risk can be stratified according to the condition, genotype, size of the aorta, and family history of aortic dissection. The European Society of Cardiology (ESC) recommends the following modified World Health Organisation risk categorisation, updated in August 2025. This has been modified.

Risk	**Low mWHO 2.0 II**	**Intermediate mWHO 2.0 II-III**	**High mWHO 2.0 III**	**Very high mWHO 2.0 IV**
Maternal cardiac event rate (%)	8-22	13-18	21-29	36-50
HTAD (including MFS and LDS)		Without aortic dilatation**	• 40-45 mm • previous aortic root replacement	> 45 mm*
Previous aortic dissection			stable diameter	increasing diameter*
BAV aortopathy	<40mm	40-45mm	45-50 mm	>50 mm*
TS		No cardiac disease (BAV, coarctation, ARD,	ASI 20-25 mm/m^2	ASI >25 mm/m^2*

		HT)		
Vascular EDS				all

* These patients would typically be offered surgery prior to conception.
** note unpredictability of type B aortic dissection in Marfans Syndrome, thus reasonable to consider all as high-risk.

Recommendations for antenatal management, delivery and postnatal management depending on risk.

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 part of the aorta is not well seen on echo (e.g. distal ascending, arch or descending aorta). Whole aorta MRI should be considered if not done within two years prior to pregnancy.

Beta-blockers are advised for all women with syndromic and non-syndromic HTAD. Specifically, in vEDS, celiprolol (up to 200 mg BD) is recommended for all. In those cases, women should undergo 4 weekly growth scans from 26 weeks gestation (due to beta-blocker associated IUGR). Neonatal hypoglycaemia may occur.

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

Risk	**Low mWHO 2.0 II**	**Moderate mWHO 2.0 II-III**	**High mWHO 2.0 III**	**Very high mWHO 2.0 IV**
Echo	Once at 20-30 weeks and 6 months postpartum	Once at 20-30 weeks and 6 months postpartum	4-12 weekly and up to 6 months postpartum	4 weekly
Epidural	As per obstetric requirement	Recommended	Recommended	Recommended
Location of delivery	Local hospital	Cardiac surgical centre	Cardiac surgical centre	Cardiac surgical centre
Second-stage	As per usual obstetric guidelines	Consider shortened active second stage	Passive vaginal delivery or ELCS†	ELCS† (37 weeks for vEDS)
Recommended postpartum stay (days)	Up to 3 and/or when BP controlled	Up to 3 and/or when BP controlled**	7	7

† location of Caesarean section to be discussed with MDT. Maybe appropriate to be in cardiac theatres for some women.

N.B. Non cardiac risks in women with connective tissue disease include increased miscarriage postpartum haemorrhage, preterm birth, cervical incompetence, premature rupture of membranes and increased tearing/poor wound healing.

Anaesthetic considerations

Women with Marfans Syndrome and Loeys-Dietz syndrome may have dural ectasia. This can affect the effectiveness of both spinal and epidural anaesthesia. MRI of the lumbosacral spine is advised prior to pregnancy, though can be done safely during pregnancy to help with delivery planning. Women should be advised of the unpredictable nature of the anaesthesia. Previous surgery for scoliosis may also affect the ability to achieve good analgesia in these women.

Appendix 1 – Evidence of Learning from Incidents

The following table sets out any incidents/ cases which informed either the creation of this document or from which changes to the existing version have been made.

Incidents	Summary of Learning
n/a	

Table A

REFERENCES	De Backer J et al. 2025 ESC Guidelines for the management of cardiovascular disease and pregnancy. Eur Heart J. 2025 Aug 29:ehaf193. Mazzolai L et al. 2024 ESC Guidelines for the management of peripheral arterial and aortic diseases. Eur Heart J. 2024 Sep 29;45(36):3538-3700. Royal college of obstetricians and gynaecologists good practice paper on the prevention and management of aortic dissection in pregnancy (currently open for consultation) aortic-dissection-pr-draft.pdf
RELATED DOCUMENTS AND PAGES	Regional Referral Pathway for Cardiac Disease in Pregnancy https://uhbw.mystaffapp.org/16416/document_view.pdf Management Chronic Thoracic Aortic Disease https://uhbw.mystaffapp.org/17332/document_view.pdf
AUTHORISING BODY	Pregnancy heart team at University Hospitals Bristol and Weston (maternal medicine obstetricians, obstetric cardiologist, obstetric anaesthetists, specialist midwives)
SAFETY	Careful management with an experienced multi-disciplinary team (the pregnancy heart team) is advised with an individual care plan for each woman
QUERIES AND CONTACT	Contact any of the following via switchboard Dr S Curtis, Consultant Cardiologist Dr V North, Consultant Cardiologist Miss J Trinder, Consultant Obstetrician Miss Aamna Ali, Consultant Obstetrician Miss A Mohan, Consultant Obstetrician Miss L Ashelby, Consultant Obstetrician
AUDIT REQUIREMENTS	Adherence to guideline will be audited periodically as part of maternal medicine departmental audit

Plan Elements	Plan Details

The Dissemination Lead is:	Dr Stephanie Curtis
Is this document: A – replacing the same titled, expired SOP, B – replacing an alternative SOP, C – a new SOP:	A
If answer above is B: Alternative documentation this SOP will replace (if applicable):	
This document is to be disseminated to:	South West Maternal Medicine Network South West and South Wales Congenital Heart Network
Method of dissemination:	Email
Is Training required:	No

Document Change Control

Date of Version	Version Number	Lead for Revisions	Type of Revision	Description of Revision
Oct 2018	1	Dr Stephanie Curtis		
Nov 2020	2	Dr Stephanie Curtis	Minor	
Sep 2022	3	Dr Stephanie Curtis	Minor	
June 2024	4	Dr Stephanie Curtis	Minor	New Trust Format
Nov 2025	5	Dr Stephanie Curtis	Major	Major revision required in line with new 2025 ESC guidelines, as well as scientific progression and published evidence in the field.