Royal College of Obstetricians & Gynaecologists

Prevention and Management of Aortic Dissection in Pregnancy

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Prevention and Management of Aortic Dissection in Pregnancy

This is the first edition of this guidance. This guidance is for healthcare professionals who care for women, non-binary and trans people who are at risk of aortic dissection in pregnancy, and those clinicians and non-clinicians tasked with commissioning and delivering care.

Within this document we use the terms woman and women's health. However, it is important to acknowledge that it is not only women for whom it is necessary to access women's health and reproductive services in order to maintain their maternal health and reproductive wellbeing. Services and delivery of care must therefore be appropriate, inclusive and sensitive to the needs of those individuals whose gender identity does not align with the sex they were assigned at birth.

1. Purpose

The purpose of this document is to ensure that women at risk of aortic dissection are identified, carefully monitored, counselled and effectively cared for throughout pregnancy, in order to reduce the risk of complications.

This guidance will:

- Define and increase awareness of women at risk for aortic dissection.
- Highlight the need for preconceptual assessment and counselling for women at risk.
- Recommend pathways for multidisciplinary peripartum care for women at risk.
- Recommend pathways for women who present with dissection during pregnancy and the postpartum period.

While this document sets out management and care pathways, care should always be guided by shared, informed decision making with the woman, respecting her preferences wherever it is clinically safe to do so.

2. Background

Pregnancy is known to be a risk factor for aortic dissection.[1,2] This is thought to be because of hormone-induced changes in the connective tissue, and the significant haemodynamic changes initiated by pregnancy and the postpartum period. Although aortic dissection in pregnancy is rare (0.5–1.1 per 100 000 pregnancy-related hospitalisations), it carries a high maternal–fetal mortality rate and accounts for approximately 1 in 6 of maternal cardiac deaths recorded in UK maternal mortality reports in the past 20 years.[3] Importantly, the majority of women who experience dissection during pregnancy are not known to have aortopathy prior to the event (Registry of Pregnancy and Cardiac Disease [ROPAC] 75%; Mothers and Babies: Reducing Risk through Audits and Confidential Enquiries across the UK [MBRRACE-UK, 90%]).[4,5] This highlights the need for increased awareness, counselling and close surveillance of those at risk.

3. Identification and pre-pregnancy counselling of women at high risk

3.1 Identification of women at high risk

Any woman with aortic disease should undergo detailed pre-pregnancy counselling with a consultant obstetrician and cardiologist with expertise in maternal cardiology.

Women should be referred early to the pregnancy heart team if they:

- Report a history at booking consistent with aortopathy or a condition associated with aortopathy (Table 1).
- Report a history at booking of aortopathy or aortic dissection (or sudden death) in a first-degree relative under the age of 60 and have not previously been investigated themselves.
- Report having a first-degree relative with Marfan syndrome (MFS), Loeys–Dietz syndrome (LDS) or vascular Ehlers–Danlos syndrome (vEDS) and have not undergone genetic testing/screening themselves.
- Report a history consistent with inflammatory vasculitis.
- Report a history of previous aortic surgery, intervention or aortic trauma.

During adolescence, girls with known aortopathy should be informed about the risks of pregnancy and discussions about contraception should be considered an essential part of routine cardiac consultations for every woman of childbearing age. Women should be informed that their pregnancy will be higher risk by definition and that they should receive focused care by a pregnancy heart team – at a minimum comprised of an obstetrician, a cardiologist, a maternal medicine specialist (an obstetric physician or an obstetrician with experience of high-risk cardiac pregnancy), maternal medicine midwife and an obstetric anaesthetist. Other members of the multidisciplinary team (MDT) may include (when necessary) a neonatologist, fetal medicine specialist, cardiac surgeon and cardiac anaesthetist in a tertiary or regional maternal medicine centre, referred to as an MMC (or sometimes a 'maternal medicine hub') in England, in line with the Maternal medicine networks: service specification.

Women at greatest risk are those with hereditary thoracic aortic disease (HTAD). HTAD is diagnosed when there is a presence of a highly penetrant pathogenic variant(s) in a known HTAD gene; thoracic aortic disease and one or more additional family members with thoracic aortic disease; or thoracic aortic disease with specific clinical features of a syndrome associated with thoracic aortic disease.[6] These include conditions such as MFS, LDS and vEDS. Not all HTAD is syndromic. Women with non-syndromic thoracic aortic disease (NsTAD) will typically have been identified through family screening, and some genotypes carry a particularly high risk. The absence of syndromic features means that the diagnosis and importance of a positive family history is often missed.[7]

Women with bicuspid aortopathy are also at risk, although at larger aortic diameters.[8] Other rare causes of dissection include: women with hypertensive disorders of pregnancy; coarctation of the aorta; substance abuse (e.g. cocaine); vasculitis (e.g. Takayasu arteritis); infection (e.g. syphilis); previous congenital cardiac surgery; and those who sustain trauma-related deceleration injuries.[9]

Genetic testing can aid in risk assessment, diagnosis, and family screening. However, a negative test result is only reassuring if the woman is confirmed negative for a known pathogenic variant present in her family, e.g. if a first-degree relative has a *FBN1* Marfan variant, but the woman has been screened and does not have the same gene. Women with aortopathy (e.g. a dilated aorta) should still be considered at risk even if a specific causative gene has not been identified and should receive appropriate care.

For women with a family history of aortic disease or dissection where no gene is available for predictive testing, they should be considered higher risk, even with a normal aorta. Since assessing genotype and phenotype can be time-consuming, early referral to cardiology or clinical genetics (depending on local services) is recommended.

Women with Turner syndrome (TS) may become pregnant if mosaic or if they have undergone assisted reproductive techniques (ART). TS is an independent risk factor for dissection and women who have co-existing hypertension and/or congenital heart disease are at higher risk than those who are normotensive and those with structurally normal hearts.[10] Women with TS have short stature and lower body surface areas (BSA), so aortic measurements need to be corrected for BSA and are usually quoted as aortic size index (ASI). However, using ASI may pseudo-normalise aortic size in women who are overweight, which is common in TS.[11,12] Guidance suggests using aortic height index (AHI). Height and weight should also be considered when assessing aortic measurements in any woman.[13]

Women who have had previous aortic surgery, typically aortic root replacement (with or without valve replacement) were previously thought to be at higher risk of aortic dissection, but this has not been strongly supported in the published literature. However, those with previous aortic dissection remain in the highest risk groups. Detailed counselling is required for these women, and it is important to ensure that they are fully informed of the risks. Those with known or at high risk of aortopathy who are seeking ART should have an up-to-date assessment and discussion with a MDT comprised of cardiology, obstetric and assisted reproduction specialists, in line with recommendations.[14]

3.2 Risk assessment

The 2025 European Society of Cardiology (ESC) guideline is helpful in guiding risk assessment, but risk must be individualised (see Table 1).[8] While absolute size of the aorta is an important issue in predicting risk, a history of dissection, evidence of growth, and hypertension must also be taken into account. In bicuspid aortopathy, root phenotype, associated coarctation and aortic length are also risk factors for dissection.[15–18] Increasingly, in MFS and vEDS, the genotype can be used to help guide risk, although no genotype is immune from dissection. Furthermore, type B dissection (dissection that starts in the descending aorta) can occur in women with an aortic root that is not dilated and cannot easily be predicted; and those who have required previous aortic root surgery also remain at increased risk of type B dissection.[1] For this reason, all women with HTAD (syndromic or non) are advised to be treated as being at risk.

Table 1. The European Society of Cardiology categorisation of risk based on aetiology and aortic size.[8]

	Low risk mWHO 2.0 II	**Intermediate risk mWHO 2.0 II–III**	**High risk mWHO 2.0 III**	**Extremely high risk mWHO 2.0 IV**
HTAD		No aortic dilatation	40–45 mm Marfan with previous aortic root replacement	> 45 mm, all vEDS
BAV		< 45 mm	45–50 mm	> 50 mm
TS	No aortic dilatation		ASI 20–25 mm/m^2	ASI > 25mm/m^2
Previous aortic dissection			Stable diameter	Increasing diameter

ASI, aortic size index; HTAD, heritable thoracic aortic disease; BAV, bicuspid aortic valve; TS, Turner Syndrome; mWHO, modified World Health Organization[19]; vEDS, vascular Ehlers–Danlos Syndrome.

Although the ESC states that women with TS with no aortic dilatation are low risk, it is advisable to treat all as being potentially at risk of dissection until further evidence is published. Current data[20] suggest that women with repaired Tetralogy of Fallot with dilated aortas are very unlikely to experience a dissection, while historical data has overestimated this risk.

3.3 Pre-pregnancy counselling

The following considerations should form part of any pre-pregnancy discussion, and counselling and should be offered by teams with appropriate maternal cardiac and aortopathy expertise to all women considered at risk of aortic dissection.

Table 2. Preconceptual assessment and counselling.

Up-to-date cross-sectional imaging	Ensure imaging is current (usually within 1–2 years).
Risk of aortic dissection	Assess the individual risk of aortic dissection.[8] Discuss alternatives to pregnancy (adoption/surrogacy) for those at high risk.
Prophylactic surgery consideration	Consider the need for preventative surgery before conception.
Assessment of comorbidities	Evaluate both cardiac and non-cardiac comorbidities.
Genetic risk for baby	Discuss genetic risks and options: PGT-M, CVS, amniocentesis, or postnatal testing (cord blood). Where relevant, refer to genetics for full diagnostic work-up and to discuss options regarding PGT-M.
Medication adjustments	Discuss the need to stop fetopathic medications during pregnancy, e.g. angiotensin receptor blockers/ACE inhibitors, continue beta-blockers, and use appropriate thromboprophylaxis regimens.
Contraception	Discuss the need for safe and effective contraception while pending investigations and postpartum contraception options.
General prenatal care	Folic acid supplementation (3 months prior to conception), cervical screening up-to-date, smoking cessation, lifestyle modifications (blood pressure control, healthy weight, diabetes management, etc.)
Antenatal visit burden	Discuss the frequency and need for antenatal visits to the tertiary centre.
Fetal growth and fetal echocardiogram	Discuss the potential need for additional fetal growth scans and fetal echocardiograms.
Blood pressure control	Ensure meticulous blood pressure control throughout.
Non-cardiac complications of connective tissue disorders	Discuss obstetric risks such as premature birth, preterm prelabour rupture of membranes, and postpartum haemorrhage.
Mode of birth	Assess likelihood of passive vaginal birth or planned caesarean birth for women at high risk (see section 5.2).
Postpartum stay duration	Prepare women at high risk for a prolonged postpartum inpatient stay (may be up to 7–10 days).
Contact details	Provide contact details so that it is clear who to contact if they become pregnant. Counsel on the symptoms and signs of dissection and emergency care plan.

ACE, angiotensin-converting enzyme; CVS, chorionic villous sampling; PGT-M, preimplantation genetic testing for monogenic disorders.

4. Antenatal care

4.1 Referral pathways

Referral pathways should be clearly defined for women who have, or are suspected to have, aortopathy during pregnancy. Obstetric teams at local hospital or primary care providers should refer all women with aortic dilatation, defined as high risk (Category C) in the Maternal medicine networks: service specification, for care by the pregnancy heart team at a tertiary or appropriate regional maternal medicine centre. Women who have been seen for preconceptual counselling should also have direct access to the pregnancy heart team.

4.1.1 First antenatal consult with women at risk

At the first meeting with the woman at an appropriate regional maternal medicine or tertiary centre, the following issues need to be considered:

- **History and imaging review**: review the woman's history and relevant imaging. Consider non-contrast cardiac magnetic resonance (CMR) imaging for women with high risk aortopathy who have not had cross-sectional imaging in the past 1–2 years.
- **Risk assessment and counselling**: gauge the woman's understanding of their condition and provide risk assessment and counselling for those not previously counselled. The conversation around risk should be tailored to their (and their partner's) understanding of the risks involved and their own assessment of it.
- **Medication review**: rationalise medications, e.g. stopping angiotensin receptor blockers, angiotensin-converting enzyme (ACE) inhibitors and any other teratogenic drugs.
- **Beta-blockers**: consider beta-blockers in women at risk and discuss the risks versus benefits. Guidelines recommend beta-blockers in the vEDS population (celiprolol is recommended – Class I C recommendation) and, despite a lack of robust evidence in pregnant women, the 2025 ESC guideline recommends beta-blockers in all women with MFS and HTAD during pregnancy (Class IIa C recommendation).[8] The use of beta-blockers in pregnancy has been shown to have an association with fetal growth restriction (FGR); however, the causal relationship is complex as hypertension itself is known to cause FGR. There have also been data demonstrating a potential risk of neonatal bradycardia, hypotension and hypoglycaemia at term; however, these risks have not been identified in more recent RCTs.[21–23] It is therefore recommended that these babies are reviewed postpartum according to local neonatal guidelines.
- **Blood pressure monitoring**: implement strict blood pressure monitoring with clearly defined parameters for the antenatal team (typically 120–129 mmHg systolic target, but dependent on individual circumstances).[8,13] Hypertension is an independent risk factor for aortic dissection. Consider home blood pressure surveillance if possible. Women with a history of coarctation of the aorta or previous type B dissection should have blood pressure monitoring in their right arm as well as left, owing to a possible discrepancy. If there is a discrepancy more than 15 mmHg, clinicians should review previous imaging to understand why. Typically, right arm blood pressure is recommended for longitudinal monitoring in coarctation when there is uncertainty about left subclavian involvement.[8] Prompt medical review should be sought if the discrepancy is new, larger than usual, or associated with chest or back pain.
- **Aortic surveillance plan**: discuss the plan for aortic surveillance according to the modified World Health Organization 2.0 risk classification (Table 1)[8] and the distribution of aortopathy, e.g. patients with aneurysms distal to the aortic root may need CMR surveillance if dimensions are concerning – more than 3 mm/year increase is considered rapid growth. Discuss the symptoms and signs of dissection and how to access emergency care.
- **Fetal plan and considerations**: establish a fetal plan and address any relevant considerations, e.g. the options of prenatal screening or fetal DNA testing for those with a predictive gene who have not had preimplantation genetic testing for monogenic disorders (PGT-M) and wish to have their baby tested after birth. Invasive testing may be an option (e.g. chorionic villus sampling [CVS]) and should be discussed both with the woman and a fetal medicine specialist. There is also the option for parents to arrange genetic testing after the baby is born – this should be done in consultation with specialists in paediatric cardiology and/or clinical genetics.
- **Discussion regarding likely place of birth ± mode of birth**: for women at high risk discuss the likelihood of needing to give birth in the tertiary centre/regional maternal medicine centre and whether a caesarean birth is likely to be indicated.
- **Liaison with local obstetric and primary care team**: liaise with the local obstetric team who conducted the initial booking (if not already involved) and primary care team so that they are aware of the plan made with the woman. Involving local obstetric and cardiology teams in MDT meetings is invaluable to ensure good communication between all healthcare professionals caring for the woman.

4.1.2 Counselling

- **Risk of aortic dissection**: counsel on the individual risk of aortic dissection.
- **Red flag symptoms**: educate on red flag symptoms during pregnancy and postpartum, including:
  - acute intense chest or back pain (particularly tearing subscapular pain); pain may emanate from, or radiate to, the neck or abdomen;
  - unexplained loss of consciousness; and/or
  - sudden, new neurological events.

Emergency plan: provide a clear plan for what women should do if they experience these symptoms, including to call 999 and give medical professionals assessing them the contact details for the pregnancy heart team.

Women known to be at increased risk should consider the following precautions:

- **Wear a MedicAlert bracelet or necklace** stating their heart condition and emergency contact details.
- **Carry an information card or wallet card** with details of their diagnosis, current medications, and specialist team contact information.
- **Enter their medical details** into their **phone's health or emergency information app** so that emergency services can access this even if their phone is locked.

All women with aortopathy who are considered at risk of aortic dissection should be cared for by the pregnancy heart team.

4.2 Specific antenatal obstetric considerations

- **Aortic imaging** in each trimester can be performed safely with echocardiography. If cross-sectional imaging is required, CMR can safely be performed without gadolinium contrast. If an elective computerised tomography (CT) scan is required, women should be counselled on the risk of radiation. The risk of an adverse effect is highest in the first trimester of pregnancy, during organogenesis, and decreases thereafter. The average radiation dose received from a chest CT is 5–12 milligray (mGy) for the mother, and 0.01–0.66 mGy for the fetus. The average exposure limit for the fetus should ideally be less than 1 mGy, although fetal risk is considered negligible under 10–50 mGy.[24] Although most aortic dissections occur in the first few days postpartum, those that occur antenatally most often occur in the third trimester.
- **Medication** needs to be considered to ensure that blood pressure is controlled. Women are at an increased risk of dissection with any hypertensive disease of pregnancy. Beta-blockers are considered safe in pregnancy. There have been data on the potential risk of FGR, however, as described above, the causal relationship is complex. While the overall effect on fetal size is thought to be small (approximately a 200 g reduction in birth weight), women taking these medications should have risk factors for fetal growth identified, including those determined by the Saving babies' lives care bundle (SBLCB) in England or trust/health board recommendations for the devolved nations of the UK.[25] These risk factors should be used to plan fetal growth surveillance. It is recognised that women with complex health conditions may have additional risk factors, outside the remit of the SBLCB, which necessitate increased surveillance above the parameters set in the care bundle. For example, a woman may be deemed at low risk using the SBLCB tool, but receiving a beta-blocker, as described above, and receiving care in a maternal medicine centre because of aortic root dilatation. In these rare situations, the specialist team may recommend serial growth scanning, rather than symphysis-fundal height measurement as the most appropriate tool to monitor the pregnancy. Increased surveillance is appropriate if any fetal concerns arise. Neonates may require glucose monitoring for 24 hours following birth and it is advised that clinicians follow local postpartum neonatal guidance.[21–23]
- **Optimise, diagnose and treat other comorbidities**, e.g. pre-eclampsia, pre-existing and gestational diabetes and anaemia.

4.3 Fetal considerations and monitoring

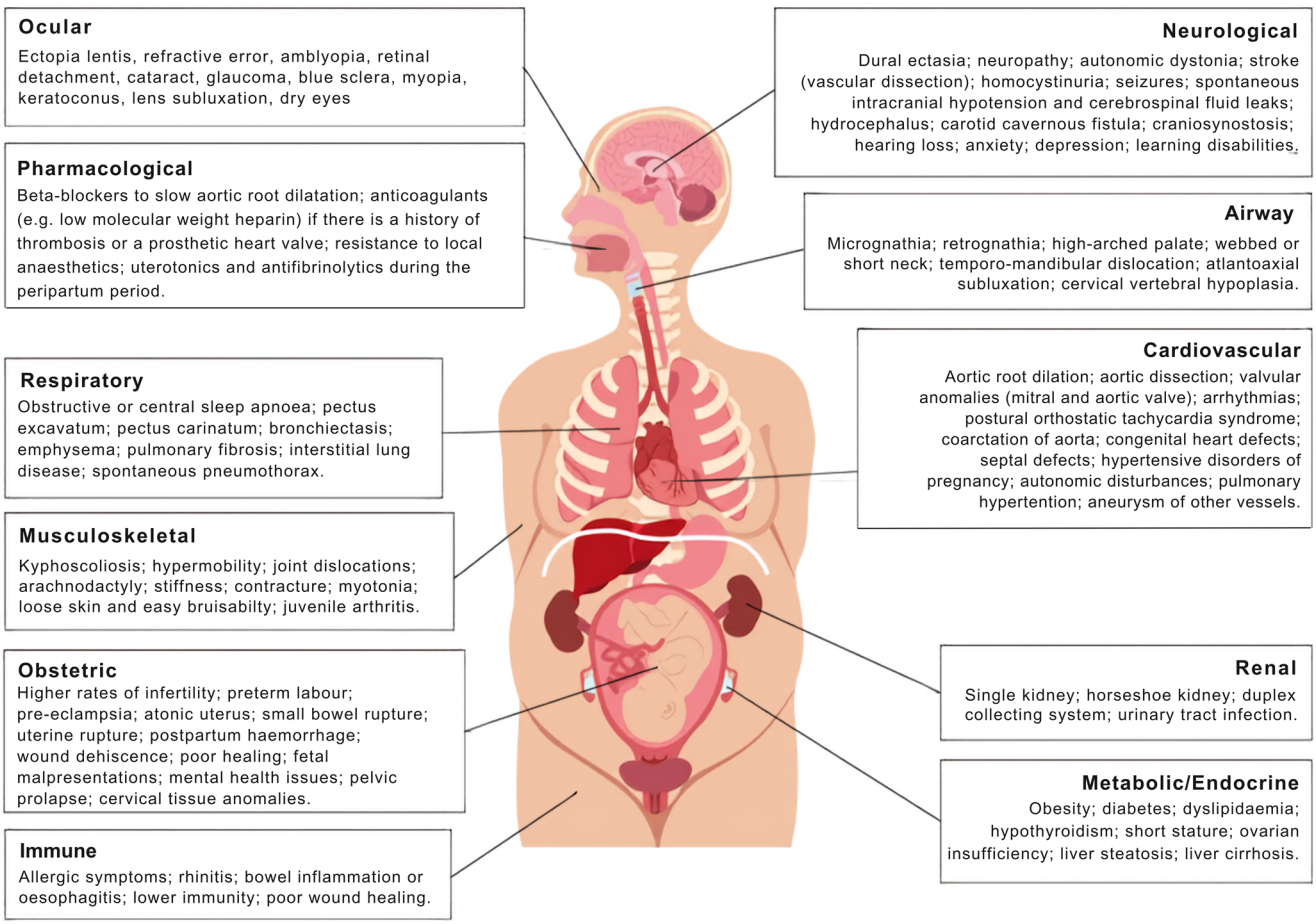
If, following individualised risk assessment, the woman is deemed to be at higher risk of having a pregnancy affected by FGR, ultrasound assessment of the fetus should be offered per local growth surveillance pathways. Tertiary level fetal medicine support is important for:

- **Genetic counselling and testing**: discussion regarding invasive fetal testing for inherited conditions can be undertaken early in the pregnancy, if no PGT-M has taken place. If, after expert counselling, the decision to test the fetus is made, this can be done by CVS and amniocentesis.
- **Fetal assessment**: women who have not had PGT-M, and/or have chosen not to have invasive fetal DNA testing, can be referred for assessment of phenotypic features in the fetus that are characteristic of the maternal condition, e.g. talipes or amniotic bands in vEDS or LDS.

4.4 Specific antepartum anaesthetic considerations

As key members of the pregnancy heart team, obstetric anaesthetists with interest/experience in maternal cardiology need to be involved at the start of planning for the birth in women with aortopathy. In the case of women at very high risk (i.e. those presenting with previous aortic dissection, a rapidly expanding aorta or aortopathy at surgical threshold), the aortic MDT should be involved in birth planning. This usually involves cardiothoracic as well as vascular surgeons, vascular imaging consultants, cardiologists, and specialist nurses with a specialist interest, alongside cardiothoracic anaesthetists. These women may, in addition to their aortopathy, have other cardiac and non-cardiac anomalies (depending on the aetiology of aortopathy) that may influence peripartum care.[26–30] These anomalies are shown in Figure 1, and the relevant anaesthetic considerations are summarised in Appendix I.

Figure 1. Cardiac and non-cardiac manifestations and anaesthetic considerations in women with aortopathy. Not all of these manifestations are associated with all aetiologies of aortopathy.



Pregnant women with aortopathy should be reviewed in an obstetric anaesthesia antenatal assessment clinic ideally before 24^{+0} weeks.[31]

The anaesthetic assessment in clinic should include:

- **A comprehensive assessment** of the primary condition along with any related medical, musculoskeletal (e.g. kyphoscoliosis/corrective spine surgery/hypermobility) and obstetric comorbidities.
- **Details of any previous anaesthetic interventions** including a difficult airway, any specific history suggestive of failed epidural analgesia/anaesthesia, and any observed resistance to local anaesthetics (e.g. dental procedures).
- **Any previous spinal imaging** (e.g. CMR). Dural ectasia is commonplace in MFS and is also seen in LDS and vEDS.[32] Its features and anaesthetic relevance are highlighted in Appendix II.
- Discussion about the **risks and benefits regarding the available analgesia and anaesthesia** techniques during labour or for any operative intervention.

5. Birth planning

An MDT birth plan should be in place with easy access for all staff caring for women at risk of aortic dissection. The birth plan should be discussed and finalised with the woman taking her own wishes for the birth into consideration wherever safely possible. A copy of the care plan should be provided to the woman in line with MBRRACE-UK recommendations.[4]

It may also be held on the electronic record, in patient handheld notes and/or on the labour ward/birthing suite. Given the risk of premature birth (iatrogenic or otherwise) this plan should be outlined and recorded early.

The birth plan should document:

- The location of birth.
- Recommended mode of birth.
- Gestation at which birth is recommended.
- Trial of vaginal birth – induction of labour method.
- Management of the second stage – oxytocin augmentation.
- Analgesic options during the first and second stages of labour.
- Appropriate haemodynamic monitoring.
- Recommended blood pressure target during labour.
- Considerations for caesarean birth, including anaesthetic options.
- Management of the third stage, including preferred uterotonic agents (e.g. avoidance of ergometrine).
- Cord blood instructions if postnatal genetic testing planned.
- Neonatal considerations might be included, e.g. where women have been on anticoagulants or other medications which require additional monitoring.
- Recommended maternal postpartum monitoring, including location, frequency of observations and blood pressure target.
- Advised duration of postpartum stay and location.
- Postpartum imaging required before discharge and who should review.
- Contraception advice on discharge.
- Who to contact in an emergency – the cardiology resident on-call will know the contacts for their cardiothoracic referral centre, if not on site. Contact list should also include the relevant contact details of the pregnancy heart team.

Communication between the members of the pregnancy heart team is key to good maternal–fetal outcomes, particularly if care is being co-managed by local teams and the pregnancy heart team. As detailed in section 3.1, involving local obstetric and cardiology teams in MDT meetings is advised.

5.1 Birth location

- For women at high risk (Table 1) a planned caesarean birth may be recommended to allow co-location with the cardiac surgical team (Table 3).[33]
- For women at very high risk who need cardiothoracic or vascular surgical backup in case of dissection or acute vascular event, it is important to involve the surgical team, including a cardiothoracic anaesthetist in the birth planning (including MDT meetings) and scheduling. In women with vEDS, premature birth is common and the plan must take into account that the birth may not happen at the scheduled time.

Co-location of birth (i.e. obstetrics and cardiac surgery) may not always be possible. For those at high risk, the location of birth may need to be in surgical theatres, either cardiac or general theatres, depending on availability and set up in individual centres. Each plan needs to be individualised to the woman, and expertise and availability of the pregnancy heart team to mitigate risk.

5.2 Mode of birth

- **Mode of birth**: there are no randomised controlled trials to determine the safest birth method for individuals with aortopathy. Vaginal birth is preferred for almost all women with cardiac disease for a number of reasons, albeit with MDT awareness during individualised birth planning that the Valsalva manoeuvre can induce variations in heart rate, blood pressure, intrathoracic pressure, and cardiac output during the second stage of labour.[34] Some of these effects may be mitigated by an effective epidural analgesia that may permit prolongation of the passive phase of the second stage allowing descent of fetal head. Expert opinion often recommends shortening the second stage of labour by an assisted vaginal birth in order to reduce adverse cardiac outcomes, although more evidence is needed to substantiate this viewpoint.[35]

  The haemodynamic changes that occur at the time of caesarean are more sudden and profound than those occurring at vaginal birth. There is an increased risk of infection, haemorrhage and venous thromboembolism. Furthermore, future pregnancies will incur the risk of uterine rupture during labour if the woman chooses to have a vaginal birth. Caesarean birth might be advised for women at highest risk, because of the need for planning of teams and coordination of birth in surgical/cardiac theatres with a cardiac surgeon on standby (Table 3), and specifically for women with vEDS because of a high risk of uterine rupture or third-degree tear.[8,33] While a discussion should be had with the woman about the safest mode of birth taking into account their individualised risks and benefits, this should consider all options, including an unplanned labour, so that they are able to make a fully informed decision. This discussion should be recorded in the medical records and birth plan.
- **Timing of birth**: while the evidence is limited, most teams would recommend birth before 39^{+0} weeks of gestation to avoid the development of the complications of hypertensive disease. For women at the highest risk, where birth in a surgical centre is mandated, planned birth at 38^{+0} weeks of gestation (whether by induction of labour or caesarean) for geographical reasons is practical. The timing of birth should be a shared decision between the woman and the pregnancy heart team once all maternal and fetal risks have been evaluated. Birth between 34^{+0} and 36^{+6} weeks of gestation may be considered for women with vEDS or LDS with a typically more aggressive genotype, who are at increased risk of premature birth and uterine rupture.

Table 3. Recommended birth management (modified from AHA and 2025 ESC guidelines).[8,33]

	Low risk mWHO 2.0 II	Intermediate risk mWHO 2.0 II–III	High risk mWHO 2.0 III	Extremely high risk mWHO 2.0 IV
HTAD (including MFS and LDS)		< 40 mm	40–45 mm Family history of dissection	> 45 mm, all vEDS Previous dissection Aortic growth in pregnancy
BAV	< 40 mm	40–45 mm	45–50 mm	> 50 mm
TS		No aortic dilatation	ASI 20–25 mm/m^2	ASI > 25 mm/m^2
Previous aortic dissection			Stable diameter	Increasing diameter
Neuraxial analgesia/ anaesthesia		Recommended	Recommended	Recommended
Birth	No specific precautions	Consider expedited active second stage* (IIa C)	Consider vaginal birth or planned caesarean* (ESC IIa C)	Caesarean birth recommended

AHA, American Heart Association; ASI, aortic size index; ESC, European Society of Cardiology; HTAD, Heritable thoracic aortic disease; BAV, bicuspid aortic valve; TS, Turner syndrome; mWHO, modified World Health Organization.

* There are no robust data on shortening the second stage of labour, however the benefits are widely recognised.

5.3 Labour analgesia

Goals of labour analgesia in women at risk of aortic dissection include provision of good pain relief during labour to mitigate the sympathomimetic effects, especially tachycardia, hypertension and increase in cardiac output associated with uterine contractions, and also to counter the haemodynamic effects of the Valsalva manoeuvre. These effects may increase the shear stress on the aorta and increase the risk of dissection.

- Epidural analgesia is the gold standard for women at high risk and is recommended for all women with aortopathy, where feasible.[8,33] It can also be used to extend the analgesia to provide surgical anaesthesia in case of an emergent caesarean birth.
- Dural ectasia is not a contraindication to performing a labour epidural. Pre-puncture neuraxial ultrasound aids in identification of the correct intervertebral level, increases first-pass success rate and may decrease dural puncture risk.[36,37]
- Other neuraxial techniques described for MFS include intrathecal catheter following a dural puncture and combined spinal–epidural analgesia.[38,39]
- For women taking anticoagulants, national guidelines[40] for the time intervals recommended for their cessation prior to performance of neuraxial analgesia (or anaesthesia) need to be adhered to.
- Reported non-neuraxial analgesia techniques for women that have given birth vaginally include a 50:50 mixture of nitrous oxide and oxygen, and intramuscular opioids.
- Remifentanil patient-controlled labour analgesia (with appropriate 1:1 midwifery and haemodynamic monitoring) may be a useful alternative for women who request remifentanil, those who seem resistant to the effect of local anaesthetic or for whom neuraxial analgesia cannot be provided.[41] In maternity units that do not offer remifentanil, fentanyl patient-controlled analgesia may provide a viable alternative.[42]

5.4 Anaesthetic for caesarean birth or operative interventions

- Key principles of anaesthesia during the perioperative period include avoiding hypertension, hypotension (swings in blood pressure), and tachycardia.
- A combined spinal–epidural (CSE) anaesthesia technique is recommended over a single-shot spinal or a de novo epidural anaesthetic for a caesarean birth, although this decision needs to be individualised.[36,43,44] The use of CSE provides greater block reliability and consistency of intrathecal local anaesthetic, with the more gradual introduction of epidural anaesthesia, and it can potentially be extended if necessary.
- For women having neuraxial (spinal, epidural or a CSE) anaesthesia, emphasis should also be on maintenance of blood pressure with a vasopressor infusion (e.g. phenylephrine, noradrenaline), although hypertensive surges should be avoided. These vasopressors have an advantage over ephedrine in avoiding tachycardia and maintaining better neonatal acid–base balance.
- Evidence on women with vEDS is sparse. Although spinal anaesthesia to facilitate caesarean birth is described in vEDS, some guidelines[45,46] recommend using general anaesthesia, especially in those who have a history of a bleeding diathesis, spinal pathology or spontaneous organ rupture. The benefits of neuraxial blockade should be balanced against the increased (but unquantified) risk of neuraxial haematoma in vEDS.
- General anaesthesia for a caesarean birth can safely be utilised for obstetric reasons (e.g. unplanned caesarean, significant postpartum haemorrhage [PPH]), anaesthetic reasons (e.g. ineffective neuraxial anaesthesia attributed to dural ectasia or anticoagulation or instrumentation of the spine), cardiac reasons (e.g. heart failure), or maternal preference.[43,47]
- Careful airway manipulation to avoid atlantoaxial subluxation and temporomandibular joint dislocation is recommended, especially in patients with hypermobility and EDS.[46]
- It is advisable to attenuate the pressor response associated with laryngoscopy and intubation, and to employ video laryngoscopy to reduce the likelihood of encountering a difficult intubation during general anaesthesia.
- Women with aortopathy may also present to the anaesthetist for a variety of other procedures varying from ART, termination of pregnancy, or following birth (e.g. repair of tear or removal of adhered placenta). For minor surgical procedures, sedation or a single-shot spinal anaesthetic technique may be safely utilised, and the decision should be individualised.
- Additional anaesthetic considerations during the perioperative period are highlighted in Appendices I and II.

5.5 Monitoring during birth

- Non-invasive blood pressure and pulse oximetry, along with fluid balance monitoring should be utilised during labour in women at low risk.
- Arterial line placement for invasive blood pressure monitoring is usually reserved for women with aortopathy at high risk, those with concomitant cardiac problems or those at risk of major PPH.

5.6 Uterotonics and postpartum haemorrhage management

Uterotonics can cause profound haemodynamic effects. Oxytocin causes dose-dependent hypotension and tachycardia with rebound increase in cardiac output. Ergometrine causes intense vasoconstriction and there is a risk of hypertension. Since some women with aortopathy are at increased risk of PPH, risk versus benefit of a more cautious uterotonic approach should be assessed:

- For women at high risk, where possible, avoid boluses and titrate oxytocin with a modified infusion in the third stage.
- Carbetocin may be utilised based on its haemodynamic profile, but there is scant literature of its reported use in pregnant women with heart disease.[48]
- Where possible, it is advisable to avoid ergometrine, which causes intense vasoconstriction.
- Carboprost (Hemabate®) can be administered, however this can cause mild hypertension in large doses.
- Tranexamic acid should be administered in the setting of PPH as there is evidence that it reduces life-threatening bleeding.[49] In this high risk patient group, tranexamic acid can also be considered prophylactically; despite sparse but growing evidence.

- A massive obstetric haemorrhage call should be activated early (e.g. 20% loss of circulating blood volume, taking into account the woman's body mass index [BMI]). Guidance from MBRRACE-UK[50] emphasises that arbitrary volumes for activation of haemorrhage protocols may be detrimental for women with a lower (or higher) BMI.
- Appropriate preparation for transfusion, including intraoperative cell salvage, and administration of desmopressin may be considered in vEDS as necessary.

5.7 Other obstetric considerations during birth

The following complications are more common in women with connective tissue disorders.

- **Prematurity** (preterm birth or extreme preterm birth)
  - Antenatal measures to improve fetal outcomes (e.g. steroids and magnesium sulphate) should be offered to women as per latest local or national guidance, e.g. RCOG Green-top Guideline No. 74 *Antenatal corticosteroids to reduce neonatal morbidity and mortality,*[51] the British Association of Perinatal Medicine Framework for Practice (BAPM) *Antenatal Optimisation Toolkit*[52] and National Institute for Health and Care Excellence (NICE) guideline [NG25] *Preterm labour and birth*.[53]
- **Preterm prelabour rupture of the membranes (PPROM)**
  - Some women, e.g. those with vEDS and TS, have been shown to be at increased risk of preterm birth. If labour does not occur spontaneously following PPROM, the RCOG Green-top Guideline No. 72 *Care of Women Presenting with Suspected Preterm Prelabour Rupture of Membranes from 24^{+0} Weeks of Gestation* should be followed.[54]
- **Wound infection**
  - Wound infection, healing and scarring can be atypical in women with connective tissue disorders. For further management refer to the NICE guideline [NG125] *Surgical site infections: prevention and treatment*.[55]

5.8 Postpartum care

Most aortic dissections occur postpartum, and most of these occur within the first week.[56] The pregnancy heart team must prioritise effective blood pressure management and conduct a postpartum echocardiogram. For women at high risk, the team may decide to extend their stay up to a week postpartum, following a discussion of the associated risks with the woman as part of shared and informed decision making.

It is also essential to emphasise the importance of medication adherence and to educate women, midwives, and general practitioners/primary care providers about the warning signs of aortic dissection.

Emergency medicine clinicians also need to be vigilant and rule out dissection when a pregnant or recently pregnant woman presents with chest/back pain in an emergency department.

The following are important considerations:

- **Location of postpartum care**: women at high risk should be monitored in an obstetric high-dependency unit setting for 12–24 hours post birth to ensure pain control and blood pressure are optimised.
- **Blood pressure management**: target blood pressure should be clear and drugs added as needed. Beta-blockers may be continued and ACE inhibitors (e.g. enalapril) can be recommenced safely, even for those who are breastfeeding. There is insufficient safety evidence for breastfeeding on angiotensin receptor blockers. A clear plan should be made to change any ACE inhibitor prescribed to an angiotensin receptor blocker once breastfeeding has ceased.
- **Multimodal analgesia**: using paracetamol, nonsteroidal anti-inflammatory agents (if not contraindicated), along with opioids and local infiltration of local anaesthetic or a transversus abdominal plane or a quadratus lumborum block may be utilised for postoperative analgesia following caesarean birth.

- **Duration of inpatient stay** should be clearly defined, agreed and documented in the woman's clinical record.
- **Women at high risk** should have a repeat echocardiogram pre-discharge.
- **Contraception** plan should be discussed and documented. Long-term use of the combined oral contraceptive pill should be avoided in women with an increased risk of venous thromboembolism, in particular women with aortopathy, mechanical valves, atrial arrhythmia or hypertension. For further advice, please see the College of Sexual and Reproductive Health guideline *Contraception After Pregnancy*.[57]
- A clear plan for further cardiology follow-up and imaging should be in place at the time of discharge. A discharge summary detailing the care should be provided to community midwifery teams and primary care providers (e.g. general practitioner) to ensure continuity of care.

6. Management of acute aortic dissection in pregnancy

6.1 Type of dissection

Acute aortic dissection (AAD) is usually described by the Stanford classification.[58]

- **Type A**: any involvement of the ascending aorta, regardless of intimal tear location or extent of dissection. Urgent open heart surgery in a cardiothoracic centre is required.
- **Type B**: involves the aorta distal to the origin of the left subclavian artery. These are generally medically managed in the first instance.

It is vital when reviewing a pregnant or a recently pregnant woman with chest pain that people '**Think Aorta**' and consider aortic dissection within the differential diagnosis. A dissection refers to a disruption of the intimal layer of the aorta, with bleeding into the space between the intima and media or the arterial wall. AAD is an emergency where rapid recognition and diagnosis is essential for the formulation of management strategies and mobilisation of MDTs. Untreated type A risk of mortality increases by 1% per hour from diagnosis.[59] This is particularly important where AAD type A has occurred when survival is dependent on early surgery.

6.2 Presentation and diagnosis

- **Symptoms**: acute sudden onset tearing pain radiating to the back (typically interscapular). The pain can also be transient (even when analgesia has not been administered) because the dissection temporarily stabilises. Women can also feel short of breath, have haemoptysis (coughing up blood), feel faint or collapse. Signs of visceral, neurological and or limb ischaemia may also be present.
- **Signs**: tachycardia, tachypnoea, differential blood pressure between the arms (more than 20 mmHg between systolic blood pressure readings) may be present, but only if dissection occurs distal to the right subclavian artery, and is specific, but not sensitive, for acute dissection. If type A involves the aortic valve there could be a new onset early diastolic murmur of aortic regurgitation. If the dissection has extended proximally into the coronary arteries or distally into the head and neck vessels there will be signs of an acute myocardial infarction (MI) or stroke respectively. There may also be hypotension, which could be because of myocardial ischaemia, tamponade, aortic valve incompetence or the extent of the dissection/aortic bleed into the vessel wall.
- **Investigations**: there is often an overcautious approach to the imaging required to make the diagnosis of aortic dissection, particularly in pregnancy.
  - A chest X-ray is only abnormal in 60–90% cases and classically shows a widened mediastinum.
  - For a definitive diagnosis, the woman should undergo an urgent CT scan with contrast, ensuring that the whole aorta is imaged.
  - Transthoracic echocardiography can identify pericardial effusions, aortic valve pathology and assess left ventricular function, but should not be allowed to delay the CT. The dissection flap may be visible.
  - Electrocardiogram (ECG) may show ST elevation if the dissection includes the coronary ostia, with right coronary (inferior MI) being more common than the left coronary artery (anterior MI), because of aortic anatomy.

6.3 Acute general management

Management principles for AAD:

- **Vascular access and fluid resuscitation**: ensure adequate venous access early. Fluid resuscitation should occur and ensure blood products are made available.
- **Aggressive pain relief**: opiates plus antiemetics.
- **Hypertension management**: labetalol, hydralazine, intravenous glyceryl trinitrate can all be used as necessary. Initially, intravenous therapy with arterial line blood pressure monitoring should be used, aiming for systolic blood pressure at < 120 mmHg and mean arterial pressure at 60 mmHg.
- **Critical care**: the women must be in a critical care environment, where she can be monitored closely with ECG, arterial line and urinary catheter.
- **Liaise with cardiothoracic team**: according to local referral pathway. Additional input will also be needed from the cardiothoracic surgical, vascular and critical care teams, and interventional radiology where available.
- **Fetal monitoring: maternal resuscitation is the priority. Fetal monitoring is ONLY performed once the patient has been stabilised**. Gestational relevant fetal monitoring using handheld Doppler fetal monitor and/or electronic fetal monitoring with cardiotocography (CTG) and/or ultrasound should be undertaken by the obstetric team.

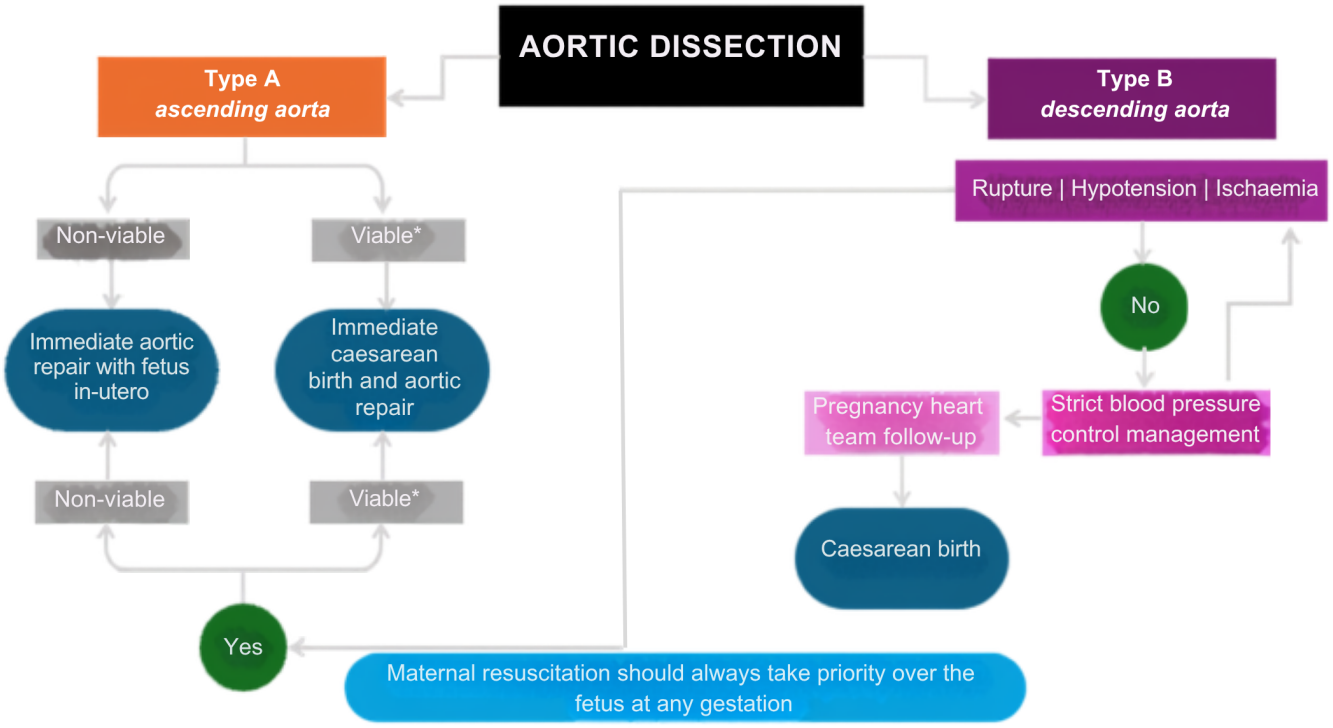
6.4 Definitive management

The Acute Aortic Dissection Pathway Toolkit[60] was developed to ensure 24-hour regional cover for AAD management in the UK, involving an MDT approach with rapid image transfer and review. Women with confirmed aortic dissection should be transferred immediately to a cardiothoracic centre (if not already in one) by the regional adult critical care transfer service. In addition to inputs from the pregnancy heart team, the following are required:

- Definitive management of type A dissection, which requires open heart aortic surgery using cardiopulmonary bypass.
- Maternal wellbeing must take precedence over the fetus regardless of fetal gestation/viability.
- If the fetus is considered pre-viable, then aortic surgery should take place without birth or evacuation of the uterus, accepting that there is a high risk of fetal mortality (20–40%).[61]
- For a viable fetus, a risk-based approach to decision making should be considered where time permits.
- The birth of an extremely preterm baby may be associated with immediate and long term morbidity, as well as mortality, particularly when there has been inadequate time for preterm optimisation. Where possible, an informed decision should be made with the woman and the pregnancy heart team regarding the risks of prematurity versus the risk to the fetus from cardiopulmonary bypass, using guidance such as the 2026 British Association of Perinatal Medicine Framework for Practice[62] and other relevant guidance as outlined in section 5.7. Importantly, the neonatal team should be involved early and provide antenatal counselling if a preterm birth is required for a maternal indication. They will also be involved in the preparation of neonatal resuscitation and transfer if the birth is performed in a non-obstetric environment (e.g. cardiac theatres).
- In the event that an MDT decision is made to first expedite birth, experienced teams need to act rapidly to perform a caesarean birth in cardiac theatres and proceed directly to dissection repair.
- Isolated type B aortic dissection in pregnancy is usually treated medically in the first instance with appropriate antihypertensive therapy. However up to 20% of cases will go on to develop complications that require intervention. Treatment of complicated type B during pregnancy is the same as for a non-pregnant person – either thoracic endovascular aortic repair [TEVAR] or open surgery.[63]
- Good blood pressure and heart rate control with medical treatment in stable type B dissections may permit continuation of pregnancy. The timing of the birth (e.g. caesarean), and the anaesthetic management in these stable patients with type B dissection needs to be individualised by the pregnancy heart, cardiothoracic, surgical and vascular teams, balancing the maternal comorbidities and the fetal prematurity risks.

- If endovascular repair is considered following a type B dissection, the team may need to decide whether it can be facilitated before, during, or after caesarean birth.[64]
- Women who have had an aortic dissection early in pregnancy should be counselled about the option of termination of pregnancy given the high risk nature of the pregnancy progressing. In the UK, termination of pregnancy is legal at any point during the pregnancy if maternal life is at risk and the termination of a pregnancy is considered lifesaving.

Figure 2. Decision making relevant to aortic dissection type.



Where time permits, individualised decision making should take place regarding viability – rather than based on a fixed gestational age

6.5 Cardiac arrest

Cardiac arrest should be treated in line with the recommended advanced life support guidelines.[65,66] Manual displacement of the uterus (if more than 20^{+0} weeks of gestation or if the uterus is above the level of umbilicus on palpation) and a perimortem resuscitative hysterotomy may be necessary.

6.6 Long-term care

Women presenting with aortic dissection in pregnancy should be assumed to have an inherited aortopathy unless there is another clear cause. Genetic testing should be performed while the woman is in hospital and screening of first-degree relatives arranged. Follow-up should be in an inherited cardiac conditions or specialist aortopathy clinic. Women should be advised regarding preconceptual counselling and assessment prior to considering a further pregnancy. This is particularly important if a mechanical valve is implanted at the time of their urgent surgery, as this carries an additional high risk in future pregnancies.

6.7 Incident review, debrief and simulation training

The occurrence of AAD and any adverse outcomes associated with the AAD during pregnancy should trigger an incident review as part of effective risk management processes. Appropriate debrief is recommended for the woman, family, and staff involved in AAD.[67] Healthcare professionals should continue to report adverse maternal and fetal outcomes according to their respective national frameworks (e.g. MBRRACE-UK/ Maternity and Newborn Safety Investigations/maternal medicine network where available), in line with national recommendations.[65,67] Although AAD is a rare occurrence, it is associated with significant maternal and fetal

morbidity and mortality. Therefore, simulation-based MDT training on AAD may improve team performance, emergency preparedness, and may contribute to improvements in specific maternal and perinatal outcomes.[68]

7. Conclusion

Pregnancy increases the risk of aortic dissection and is consistently one of the commonest causes of maternal death in registries. Most women who experience an aortic dissection are unaware that they were at risk. Identifying those women and offering appropriate pre-pregnancy counselling is key to improving maternal and fetal outcomes, both during pregnancy and long term. Women at risk of and presenting with aortic dissection associated with pregnancy require complex decision making and should be cared for by expert MDTs to ensure appropriate antenatal maternal and fetal surveillance and safe birth planning.

References

1. Roman MJ, Pugh NL, Hendershot TP, Devereux RB, Dietz H, Holmes K, et al. Aortic complications associated with pregnancy in Marfan syndrome: The NHLBI National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC). *J Am Heart Assoc*. 2016;5:e004052.
2. Nasiell J, Norman M, Lindqvist PG, Malmstedt J, Bottinga R, Blennow M. Aortic dissection in pregnancy: a life-threatening disease and a diagnosis worth considering. *Acta Obstet Gynecol Scand*. 2009;88:1167–70.
3. Wang Y, Yin K, Datar Y, Mohnot J, Nodoushani AY, et al. Aortic dissection during pregnancy and puerperium: contemporary incidence and outcomes in the United States. *J Am Heart Assoc*. 2023;12:e028436.
4. Knight M, Bunch K, Tuffnell D, Shakespeare J, Kotnis R, Kenyon S, Kurinczuk JJ, editors. *Saving Lives, Improving Mothers' Care – Lessons learned to inform maternity care from the UK and Ireland Confidential Enquiries into Maternal Deaths and Morbidity 2015–17*. Oxford: National Perinatal Epidemiology Unit, University of Oxford; 2019.
5. Campens L, Baris L, Scott NS, Broberg CS, Bondue A, et al.; ROPAC investigators group. Pregnancy outcome in thoracic aortic disease: data from the Registry of Pregnancy and Cardiac Disease. *Heart*. 2021;107:1704–9.
6. Milewicz DM, Cecchi AC. Heritable thoracic aortic disease overview. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. *GeneReviews®* [Internet]. Seattle (WA): University of Washington; 2003 (Updated 2023) [www.ncbi.nlm.nih.gov/books/NBK1120/]. Accessed 6 Aug 2026.
7. Holmes KW, Markwardt S, Eagle KA, Devereux RB, Weinsaft JW, Asch FM, et al.; GenTAC Investigators. Cardiovascular outcomes in aortopathy: GenTAC registry of genetically triggered aortic aneurysms and related conditions. *J Am Coll Cardiol*. 2022;79:2069–81.
8. De Backer J, Haugaa KH, Hasselberg NE, de Hosson M. 2025 ESC guidelines for the management of cardiovascular disease and pregnancy. *Eur Heart J*. 2025;46:4462-458.
9. Hebballi R, Swanevelder J. Diagnosis and management of aortic dissection. *Continuing Educ Anaesth Crit Care Pain*. 2009;9:14–8.
10. Carlson M, Silberbach M. Dissection of the aorta in Turner syndrome: two cases and review of 85 cases in the literature. *BMJ Case Rep*. 2009;2009:bcr0620091998.
11. Chevalier N, Letur H, Lelannou D, et al.; French Study Group for Oocyte Donation. Materno-fetal cardiovascular complications in Turner syndrome after oocyte donation: insufficient pre-pregnancy screening and pregnancy follow-up are associated with poor outcome. *J Clin Endocrinol Metab*. 2011;96:E260–7.
12. Silberbach M, Roos-Hesselink JW, Andersen NH, et al.; American Heart Association Council on Cardiovascular Disease in the Young; Council on Genomic and Precision Medicine; and Council on Peripheral Vascular Disease. Cardiovascular health in Turner syndrome: a scientific statement from the American Heart Association. *Circ Genom Precis Med*. 2018;11:e000048.
13. Mazzolai L, Teixido-Tura G, Lanzi S, Boc V, Bossone E, Brodmann M, et al.; ESC Scientific Document Group. 2024 ESC guidelines for the management of peripheral arterial and aortic diseases. *Eur Heart J*. 2024;45:3538–700.
14. English K, Frise C, Trinder J, Cauldwell M, Simpson M, Adamson D, et al. Best practice recommendations for medically assisted reproduction in patients with known cardiovascular disease or at high risk of cardiovascular disease: a consensus statement supported by multiple professional societies. *Hum Fertil*. 2024;27:2278295.

15. Smith K, Gros B. Pregnancy-related acute aortic dissection in Marfan syndrome: a review of the literature. *Congenit Heart Dis*. 2017;12:251–60.
16. Coady MA, Rizzo JA, Hammond GL, Mandapati D, Darr U, Kopf GS, et al. What is the appropriate size criterion for resection of thoracic aortic aneurysms? *J Thorac Cardiovasc Surg*. 1997;113:476–91.
17. Matura LA, Ho VB, Rosing DR, Bondy CA. Aortic dilatation and dissection in Turner syndrome. *Circulation*. 2007;116:1663–70.
18. Jondeau G, Detaint D, Tubach F, Arnoult F, Milleron O, Raoux F, et al. Aortic event rate in the Marfan population: a cohort study. *Circulation*. 2012;125:226–32.
19. Thorne S, MacGregor A, Nelson-Piercy C. Risks of contraception and pregnancy in heart disease. *Heart*. 2006;92:1520–5.
20. Shehata M, Bashir R, Al-Alao B, Salmasi MY, Asimakopoulos G, Hoschtitzky A. Type A aortic dissection following a tetralogy of Fallot repair. *JTCVS Tech*. 2023;22:145–6.
21. De Bruin R, van Dalen SL, Franx SJ, Ramaswamy VV, Simons SHP, Flint RB, et al. The risk for neonatal hypoglycemia and bradycardia after beta-blocker use during pregnancy or lactation: a systematic review and meta-analysis. *Int J Environ Res Public Health*. 2022;19:9616.
22. UK Teratology Information Service. Use of beta-adrenoceptor blocking drugs (beta-blockers) in pregnancy. May 2024, Version 4 [uktis.org/monographs/use-of-beta-adrenoceptor-blocking-drugs-beta-blockers-in-pregnancy/]. Accessed 6 Aug 2026.
23. Ramlakhan KP, Roos-Hesselink JW, Basso T, Greenslade J, Flint RB, Krieger EV, et al. Perinatal outcomes after in-utero exposure to beta-blockers in women with heart disease: data from the ESC EORP registry of pregnancy and cardiac disease (ROPAC). *Int J Cardiol*. 2024;410:132234.
24. Wang PI, Chong ST, Kielar AZ, Kelly AM, Knoepp UD, Mazza MB, et al. Imaging of pregnant and lactating patients: part 1, evidence-based review and recommendations. *AJR Am J Roentgenol*. 2012;198:778–84.
25. NHS England. *Saving babies' lives: version 3 – A care bundle for reducing perinatal mortality* [Internet]. 2023 (Updated 2025) [www.england.nhs.uk/long-read/saving-babies-lives-version-3/]. Accessed 6 Aug 2026.
26. Yagyu T, Noguchi T. Diagnosis and treatment of cardiovascular disease in patients with heritable connective tissue disorders or heritable thoracic aortic diseases. *Cardiovasc Interv Ther*. 2024;39:126–36.
27. Dulac Y, Pienkowski C, Abadir S, Tauber M, Acar P. Cardiovascular abnormalities in Turner's syndrome: what prevention? *Arch Cardiovasc Dis*. 2008;101:485–90.
28. Gouda P, Kay R, Habib M, Aziz A, Aziza E, Welsh R. Clinical features and complications of Loeys-Dietz syndrome: a systematic review. *Int J Cardiol*. 2022;362:158–67.
29. Malfait F, Castori M, Francomano CA, Giunta C, Kosho T, Byers PH. The Ehlers–Danlos syndromes. *Nat Rev Dis Primers*. 2020;6:64.
30. Pollock L, Ridout A, Teh J, Nnadi C, Stavroulias D, Pitcher A, et al. The musculoskeletal manifestations of Marfan syndrome: diagnosis, impact, and management. *Curr Rheumatol Rep*. 2021;23:81.
31. Jones B, Bhatia K. Cardiac disease in pregnancy. *Anaesth Intensive Care Med*. 2022;23:448–54.
32. Böker T, Vanem TT, Pripp AH, Rand-Hendriksen S, Paus B, Smith HJ, et al. Dural ectasia in Marfan syndrome and other hereditary connective tissue disorders: a 10-year follow-up study. *Spine J*. 2019;19:1412–21.
33. Isselbacher EM, Preventza O, Hamilton Black J 3rd, Augoustides JG, Beck AW, Bolen MA, et al. 2022 ACC/AHA guideline for the diagnosis and management of aortic disease: a report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. *Circulation*. 2022;146:24:e334–e482.
34. Keating N, Rooney G, McGuinness S, McAuliffe FM. A review of the effects of Valsalva manoeuvre in labour and birth. *Obstet Med*. 2025;18:4–8.
35. Cauldwell M, Von Klemperer K, Uebing A, et al. The management of the second stage of labour in women with cardiac: A mixed methods study. *Int J Cardiol*. 2016;222:732–736.
36. Meng ML, Arendt KW. Obstetric anesthesia and heart disease: practical clinical considerations. *Anesthesiology*. 2021;10.1097/ALN.0000000000003833.
37. Young B, Onwochei D, Desai N. Conventional landmark palpation vs. preprocedural ultrasound for neuraxial analgesia and anaesthesia in obstetrics – a systematic review and meta-analysis with trial sequential analyses. *Anaesthesia*. 2021;76:818–31.

38. Buser R, Mordecai M, Brull S. Combined spinal-epidural analgesia for labor in a patient with Marfan's syndrome. *Int J Obstet Anesth*. 2007;16:274–6.
39. Vacula BB, Gray C, Hofkamp MP, Noonan PT, McAllister RK, Pilkinton KA, et al. Epidural analgesia complicated by dural ectasia in the Marfan syndrome. *Proc (Bayl Univ Med Cent)*. 2016;29:385–6.
40. Harrop-Griffiths W, Cook T, Gill H, Hill D, Ingram M, Makris M, et al. Regional anaesthesia and patients with abnormalities of coagulation. *Anaesthesia*. 2013;68:966–72.
41. Knapp C, Bhatia K, Columb M, Elriedy M. Remifentanil patient-controlled analgesia for labour in pregnant patients with heart disease. *Int J Obstet Anesth*. 2023;55:103902.
42. Show KL, Ngamjarus C, Kongwattanakul K, et al. Fentanyl for labour pain management: a scoping review. *BMC Pregnancy Childbirth*. 2022;22:846.
43. Lacassie HJ, Millar S, Leithe LG, Muir HA, Montaña R, Poblete A, et al. Dural ectasia: a likely cause of inadequate spinal anaesthesia in two parturients with Marfan's syndrome. *Br J Anaesth*. 2005;94:500–4.
44. Heck A, Clayton R, Robinson C. Dural ectasia in Marfan syndrome. *Int J Obstet Anesth*. 2014;23:287–8.
45. Carness JM, Lenart MJ. Spinal anaesthesia for cesarean section in a patient with vascular type Ehlers-Danlos syndrome. *Case Rep Anesthesiol*. 2018;2018:1924725.
46. Wiesmann T, Castori M, Malfait F, Wulf H. Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s). *Orphanet J Rare Dis*. 2014;9:109.
47. Allyn J, Guglielminotti J, Omnes S, Guezouli L, Egan M, Jondeau G, et al. Marfan's syndrome during pregnancy: anesthetic management of delivery in 16 consecutive patients. *Anesth Analg*. 2013;116:392–8.
48. Saadat F, Dob DP, Cox ML, Johnson MR, Gatzoulis MA. Carbetocin as a uterotonic in a parturient with a Fontan circulation. *Anaesth Rep*. 2024;12:e12272.
49. Ker K, Sentilhes L, Shakur-Still H, et al. Tranexamic acid for postpartum bleeding: a systematic review and individual patient data meta-analysis of randomised controlled trials. *Lancet*. 2024;404:1657–67.
50. Knight M, Bunch K, Tuffnell D, Shakespeare J, Kotnis R, Kenyon S, Kurinczuk JJ, editors. *Saving Lives, Improving Mothers' Care – Lessons learned to inform maternity care from the UK and Ireland Confidential Enquiries into Maternal Deaths and Morbidity 2016–18*. Oxford: National Perinatal Epidemiology Unit, University of Oxford; 2020.
51. Stock SJ, Thomson AJ, Papworth S; on behalf of the RCOG. Antenatal corticosteroids to reduce neonatal morbidity and mortality. Green-top Guideline No. 74. *BJOG*. 2022;129:e35–e60.
52. British Association of Perinatal Medicine. *Antenatal Optimisation Toolkit*. 2020 (Updated 2021) [www.bapm.org/pages/194-antenatal-optimisation-toolkit]. Accessed 6 Aug 2026.
53. National Institute for Health and Care Excellence. *Preterm labour and birth*. NICE guideline [NG25]. 2015 (Updated 2022) [www.nice.org.uk/guidance/ng25]. Accessed 6 Aug 2026.
54. Thomson AJ, on behalf of the Royal College of Obstetricians and Gynaecologists. Care of Women Presenting with Suspected Preterm Prelabour Rupture of Membranes from 24^{+0} Weeks of Gestation. *BJOG* 2019;126:e152–166.
55. National Institute for Health and Care Excellence. *Surgical site infections: prevention and treatment*. NICE guideline [NG125]. 2019 (Updated 2020) [www.nice.org.uk/guidance/ng125/]. Accessed 6 Aug 2026.
56. Curtis SL, Swan L. Aortopathy in pregnancy. *Heart*. 2022;108:1851–7.
57. College of Sexual and Reproductive Health. FSRH Guideline: *Contraception After Pregnancy*. London: Faculty of Sexual & Reproductive Healthcare; 2017 (Amended 2020) [www.fsrh.org/Common/Uploaded%20files/documents/contraception-after-pregnancy-guideline-oct2020.pdf]. Accessed 6 Aug 2026.
58. Agarwal S, Kendall J, Quarterman C. Perioperative management of thoracic and thoracoabdominal aneurysms. *BJA Educ*. 2019;19:119–25.
59. Acharya M, Mariscalco G. Diagnosis and acute management of type A aortic dissection. *Br J Cardiol*. 2023;30:62–8.
60. NHS. *Acute aortic dissection pathway toolkit*. April 2022 [scts.org/news/463/acute_aortic_dissection_pathway_toolkit]. Accessed 6 Aug 2026.
61. Jha N, Jha AK, Chand Chauhan R, Chauhan NS. Maternal and fetal outcome after cardiac operations during pregnancy: a meta-analysis. *Ann Thorac Surg*. 2018;106:618–26.

62. British Association of Perinatal Medicine. *Perinatal management of extreme preterm birth before 27 weeks of gestation: a BAPM framework for practice* (2026) [www.bapm.org/resources/perinatal-management-of-extreme-preterm-birth-before-27-weeks-of-gestation]. Accessed 6 Aug 2026.
63. Mussa FF, Kougias P. Management of Acute Type B Aortic Dissection. *New Engl J Med*. 2025;393:895–905.
64. Bi T, Duan X, Yin Y. TEVAR for acute type B aortic dissection in pregnant women (35 weeks gestation) with Takayasu's arteritis after cesarean section: a rare case report and literature review. *Front Cardiovasc Med*. 2025;11:1498914.
65. Chu J, Johnston TA, Geoghegan J; on behalf of the Royal College of Obstetricians and Gynaecologists. Maternal collapse in pregnancy and the puerperium. Green-top Guideline No. 56. *BJOG*. 2020;127:e14–e52.
66. Deakin C, Clarke AB, Boulton A, DeBono J, Gosling A, Greig P, et al.; on behalf of the Resuscitation Council UK. *Special circumstances guidelines 2025* [www.resus.org.uk/professional-library/2025-resuscitation-guidelines/special-circumstances-guidelines]. Accessed 6 Aug 2026.
67. Knapp C, Bhatia K. Maternal collapse in pregnancy. *Br J Hosp Med (Lond)*. 2022;83:1–12.
68. Fransen AF, Banga FR, Van De Ven J, Mol BWJ, Oei SG. Multi-professional simulation-based team training in obstetric emergencies for improving patient outcomes and trainees' performance. *Cochrane Database Syst Rev*. 2020;12:CD011545.
69. Wilson-Baig N, Bhatia K, Kochhar P, Bonner S. Intrapartum spontaneous pneumomediastinum with subcutaneous emphysema – management and recommendations to prevent recurrence in future pregnancies. *Int J Obstet Anesth*. 2020;41:122–4.
70. National Institute for Health and Care Excellence. *Diabetes in pregnancy: management of diabetes and its complications from pre-conception to the postnatal period*. NICE guideline [NG3]. 2015 (Updated 2020) [www.nice.org.uk/guidance/ng3/]. Accessed 6 Aug 2026.
71. Skjellerup N. Successful spinal anaesthesia for caesarean section in a patient with Marfan syndrome complicated by dural ectasia. *Int J Obstet Anesth*. 2017;35:88–92.

Appendix I: Summary of anaesthetic considerations in women with aortopathy.

System	Features	Anaesthetic considerations
Airway	Micrognathia, retrognathia (atypical jaw position), high-arched palate, webbed or short neck, temporomandibular dislocation, atlantoaxial subluxation, and cervical vertebral hypoplasia.	• Prepare for difficult airway. • Risk of airway trauma, atlantoaxial subluxation and temporomandibular joint subluxation during airway manipulation. • Consider videolaryngoscopy to facilitate endotracheal intubation.
Respiratory	Obstructive or central sleep apnoea, pectus excavatum, pectus carinatum, spontaneous pneumothorax.	• Potential for respiratory deterioration as pregnancy progresses leading to preterm birth. • Monitoring for respiratory depression following use of intrathecal long-acting opioids. • Monitor airway pressures during general anaesthesia as at risk of ventilator-associated pneumothorax. • Avoid 50:50 mixture of oxygen:nitrous oxide in women with previous history of pneumothorax or pneumomediastinum.[69] • Critical care support may be necessary.
Cardiovascular	• Aortic root dilation. • Aortic dissection. • Valvular anomalies (mitral and aortic valve). • Coarctation of aorta. • Congenital heart defects. • Hypertensive disorders of pregnancy. • Aneurysm of other vessels.	• Physiological changes of pregnancy and labour causing tachycardia and hypertension. • Serial echocardiography. • Good labour analgesia to mitigate sympathomimetic effects of labour. • ECG/NIBP/SpO_2 with arterial line for invasive blood pressure monitoring in women at high risk during labour and operative birth. • Appropriate IV access and blood products including intraoperative cell salvage. • Activation of major obstetric haemorrhage if necessary. • Avoidance of swings in blood pressure (hypotension and hypertension) and tachycardia during operative birth or general anaesthesia. • Maintenance of systolic arterial pressure at > 90% accurate baseline using vasopressors (e.g. phenylephrine infusion) during neuraxial anaesthesia. • Suppress pressor response to laryngoscopy and extubation. • Point-of-care ultrasound as necessary and if expertise available.

Musculoskeletal	Kyphoscoliosis, hypermobility, joint dislocations, arachnodactyly, stiffness, contracture, myotonia. loose skin and easy bruisability.	• Careful positioning during labour and operative birth to avoid joint dislocations and injuries secondary to joint laxity. • Ensure pressure areas are padded. • Kyphoscoliosis or corrective surgery for scoliosis with instrumentation of the spine may cause difficulty in placement of neuraxial block and/or unpredictable neuraxial analgesia/anaesthesia.
Neurological	Dural ectasia, neuropathy, stroke (vascular dissection), cerebrospinal fluid leaks, craniosynostosis, anxiety, depression.	• Higher risk of dural puncture and inadequate spinal anaesthesia. • Consider pre-puncture ultrasound to facilitate neuraxial analgesia (epidural)/ anaesthesia • If neuraxial analgesia not feasible, consider alternatives including remifentanil patient-controlled analgesia, intramuscular opioids or 50:50 mixture of oxygen:nitrous oxide (Entonox®). • Combined spinal–epidural anaesthetic preferable for caesarean birth. • Monitoring of neuromuscular blockade.
Ocular	Ectopia lentis, retinal detachment, blue sclera, myopia, keratoconus, lens subluxation.	Protective tapes and use of ointments during general anaesthesia.
Immune	Allergic symptoms, rhinitis, bowel inflammation or oesophagitis, lower immunity, poor wound healing.	Asepsis during surgical/invasive procedures.
Metabolic/ endocrine	Obesity, diabetes, dyslipidaemia, hypothyroidism, short stature, ovarian insufficiency.	• Glycaemic control during the peripartum period. • Monitoring blood sugar in line with national recommendations.[70]
Renal	Single kidney, horseshoe kidney, duplex collecting system, urinary tract infection.	Need to monitor renal function as necessary.

Pharmacological	• Beta-blockers to slow aortic root dilatation. • Anticoagulants (e.g. low molecular weight heparin) if there is a history of thrombosis or a prosthetic mechanical heart valve. • Resistance to local anaesthetics. • Uterotonics and antifibrinolytics during the peripartum period.	• Continue beta-blockers during labour. • Anticoagulants may need to be tailored prior to neuraxial analgesia and anaesthesia in line with national guidelines. • Appropriate monitoring of anticoagulation as necessary. • Need to use higher (but safe) doses of local anaesthetic or alternative forms of pain relief, or conversion to general anaesthesia if necessary. • If neuraxial analgesia not feasible, consider alternatives, e.g. remifentanil/fentanyl patient-controlled analgesia. • Avoid ergometrine and consider using tranexamic acid in the setting of PPH. • Administer antiemetics to decrease the risk of nausea, vomiting and organ rupture. • Multimodal analgesia following operative birth – avoid non-steroidal anti-inflammatory agents in patients with bleeding diathesis or those with renal dysfunction.
Obstetric	Higher rates of infertility, preterm labour, pre-eclampsia, atonic uterus, small bowel rupture, uterine rupture, PPH, wound dehiscence, poor healing, fetal malpresentations, mental health issues, pelvic prolapse and cervical tissue anomalies.	• Women may present for ART or termination. • Higher caesarean birth rates (63%). • PPH rates and sequalae may be higher.

ART, assisted reproductive techniques; ECG, electrocardiogram; NIBP, non-invasive blood pressure; PPH, postpartum haemorrhage; SpO_2, oxygen saturation.

Appendix II: Dural ectasia and its relevance to anaesthesia.[32,39,43,44,71]

Definition	Widening of the dural sac or spinal nerve root sleeves; associated with bony erosions of the posterior vertebral body.
Prevalence	• MFS: 63–90% • LDS: 50–73% • Ehlers–Danlos syndrome: reported cases
Symptoms	• Asymptomatic • Back pain • Headaches • Weakness • Loss of sensation (above and below the affected limb)
Risks in anaesthesia	• Increased risk of: ➢ Dural puncture during epidural for vaginal birth. ➢ Inadequate spinal anaesthesia during caesarean. ➢ Intraoperative analgesic supplementation or conversion to general anaesthesia following neuraxial anaesthesia for operative intervention. • Most cases in anaesthetic literature relate to MFS.
Recommendations for imaging	**MRI of the lumbosacral spine:** • Recommended for MFS and LDS. • Ideally performed during: ➢ Preconception counselling. ➢ Antenatal period after the first trimester (if no prior imaging available).
Benefits of imaging	• Confirms presence of dural ectasia. • Provides information for optimal intervertebral level for neuraxial analgesia.

MFS, Marfan syndrome; LDS, Loeys–Dietz syndrome.

Appendix III: List of abbreviations.

AAD	acute aortic dissection
ACE	angiotensin-converting enzyme
AHA	American Heart Association
AHI	aortic height index
ART	assisted reproductive techniques
ASI	aortic size index
BAV	bicuspid aortic valve
BSA	body surface area
CMR	cardiac magnetic resonance
CSE	combined spinal–epidural
CT	computed tomography
CVS	chorionic villous sampling
ECG	electrocardiogram
ESC	European Society of Cardiology
GTN	glyceryl trinitrate
HTAD	hereditary thoracic aortic disease
LDS	Loeys–Dietz syndrome
MBRRACE-UK	Mothers and Babies: Reducing Risk through Audits and Confidential Enquiries across the UK
MDT	multidisciplinary team
MFS	Marfan syndrome
MNSI	Maternity and Newborn Safety Investigations
mWHO	modified World Health Organization [risk assessment]
NIBP	non-invasive blood pressure
NsTAD	non-syndromic thoracic aortic disease
PGT-M	preimplantation genetic testing for monogenic disorders
PPH	postpartum haemorrhage
PPROM	preterm prelabour rupture of the membranes
SpO_2	oxygen saturation
TS	Turner syndrome
vEDS	vascular Ehlers–Danlos syndrome

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The final version is the responsibility of the Patient Safety Committee of the RCOG.

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