**Good Practice Paper No. XX**
**Peer Review Draft – June 2025**

# 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 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, 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.

## 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),[3] it carries a high maternal–fetal mortality rate and accounts for approximately 1 in 6 –10 of maternal cardiac deaths recorded in UK maternal mortality reports in the past 20 years. 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 informed that their pregnancy will be high risk by definition and that they will receive focused care by a pregnancy heart team (at a minimum comprised of an obstetrician, a cardiologist, a maternal medicine specialist, an obstetric anaesthetist, a neonatologist and a midwife) 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. During adolescence, girls should be made aware of pregnancy with aortopathy being high risk, and the safety of pregnancy (and



appropriate contraception) should be considered an essential part of routine cardiac consultations for every woman of childbearing age.

The most concerning pregnancies occur in women with hereditary thoracic aortic disease (HTAD), including syndromic conditions, such as Marfan syndrome (MFS), Loeys-Dietz syndrome (LDS) and vascular Ehlers-Danlos syndromes (vEDS). Women with bicuspid aortopathy are also at risk, although at larger aortic diameters.[6] Other rare causes of dissection include: women with hypertensive disorders of pregnancy; 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.[7]

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 high 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.[8] 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.[9,10] Guidance suggests using aortic height index (AHI), as with other patients at the extremes of body size.[11]

Women who have had previous aortic surgery were thought to be at higher risk, but this is not evidenced in 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 multidisciplinary team (MDT) comprised of cardiology, obstetric and assisted reproduction specialists, in line with the the recommendations.[12]

*3.2 Risk assessment*

The 2018 European Society of Cardiology (ESC) guideline was helpful in guiding risk assessment, but risk must be individualised (see Table 1).[6] 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.[13–17] 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 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.[6]

| | Low risk mWHO II | Intermediate risk mWHO II–III | High risk mWHO III | Extremely high risk mWHO IV |
|---|---|---|---|---|
| **HTAD** | | No aortic dilatation | 40–45 mm | > 45 mm, all vEDS |
| **BAV** | | < 45 mm | 45–50 mm | > 50 mm |
| **TS** | No aortic dilatation | | ASI 20–25 mm/$m^2$ | ASI > 25mm/$m^2$ |

ASI, aortic size index; HTAD, Heritable thoracic aortic disease; BAV, bicuspid aortic valve; TS, Turner Syndrome; mWHO, modified World Health Organization; 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. Similarly, although the ESC suggests that women with repaired Tetralogy of Fallot with dilated aortas are at high risk, more recent data suggests this is extremely rare and the risk has potentially been over-estimated.[18]

*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.[6] |
| **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). Refer to genetics if women wish to be considered for PGT-M. |
| **Medication adjustments** | Discuss the need to stop teratogenic medications, e.g. angiotensin receptor blockers/ACE inhibitors , continue beta-blockers, and use appropriate thromboprophylaxis regimens. |
| **Contraception** | Discuss the need for safe contraception while pending investigations and postpartum contraception options. |
| **General prenatal care** | Folic acid supplementation (3 months prior to conception), cervical smear up to date (within 3 years), 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 labour, premature 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 for a prolonged postpartum inpatient stay (up to 10 days). |
| **Contact details** | Provide contact details so that it is clear who to contact if they become pregnant. |

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. Obstetricians 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.

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 MFS, LDS or 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.

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 a 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.
- **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 2018 ESC guideline recommends beta-blockers in all women with MFS and HTAD during pregnancy (Class IIa C recommendation).[6]



- **Blood pressure monitoring**: implement strict blood pressure monitoring with clearly defined parameters for the antenatal team (typically 135/85 mmHg or less, but dependent on individual circumstances). Hypertension is an independent risk factor for aortic dissection. Consider home blood pressure surveillance if possible.
- **Aortic surveillance plan**: discuss the plan for aortic surveillance according to the World Health Organization (WHO) risk classification (Table 1) 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.
- **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) 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 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);
  - 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 contact details for the high risk pregnancy heart team.

All women with aortopathy or 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 perfomed without gadolinium contrast. If a 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.[19] 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, however there is a potential risk of fetal growth restriction, which



is dose dependent.[add ref back in] While the effect is thought to be small (i.e. around an average reduction in fetal weight of around 200 g), women taking these drugs require serial fetal growth scans at 28, 32 and 36 weeks of gestation. Increased surveillance may be needed if any fetal concerns are identified. All neonates will require glucose monitoring for 24 hours following birth because of the risk of neonatal hypoglycaemia with antenatal beta-blocker use.[20]

- **Optimise, diagnose and treat other comorbidities** (e.g. pre-eclampsia, pre-existing and gestational diabetes and anaemia).

*4.3 Fetal considerations and monitoring*

Referral of the woman to the fetal medicine unit and/or shared care with the pregnancy heart team may be required in the following cases. 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 in the first trimester by chorionic villus sampling and in the second trimester by amniocentesis.
- **Fetal assessment**: women who have not had PGT-M, and/or have chose not to have invasive fetal DNA testing, can be referred for assessment of phenotypic features in the fetus characteristic of the maternal condition, e.g talipes or amniotic bands in vEDS or LDS.
- **Fetal growth**: women can be referred to the fetal medicine team for fetal growth surveillance if they are taking beta-blockers.

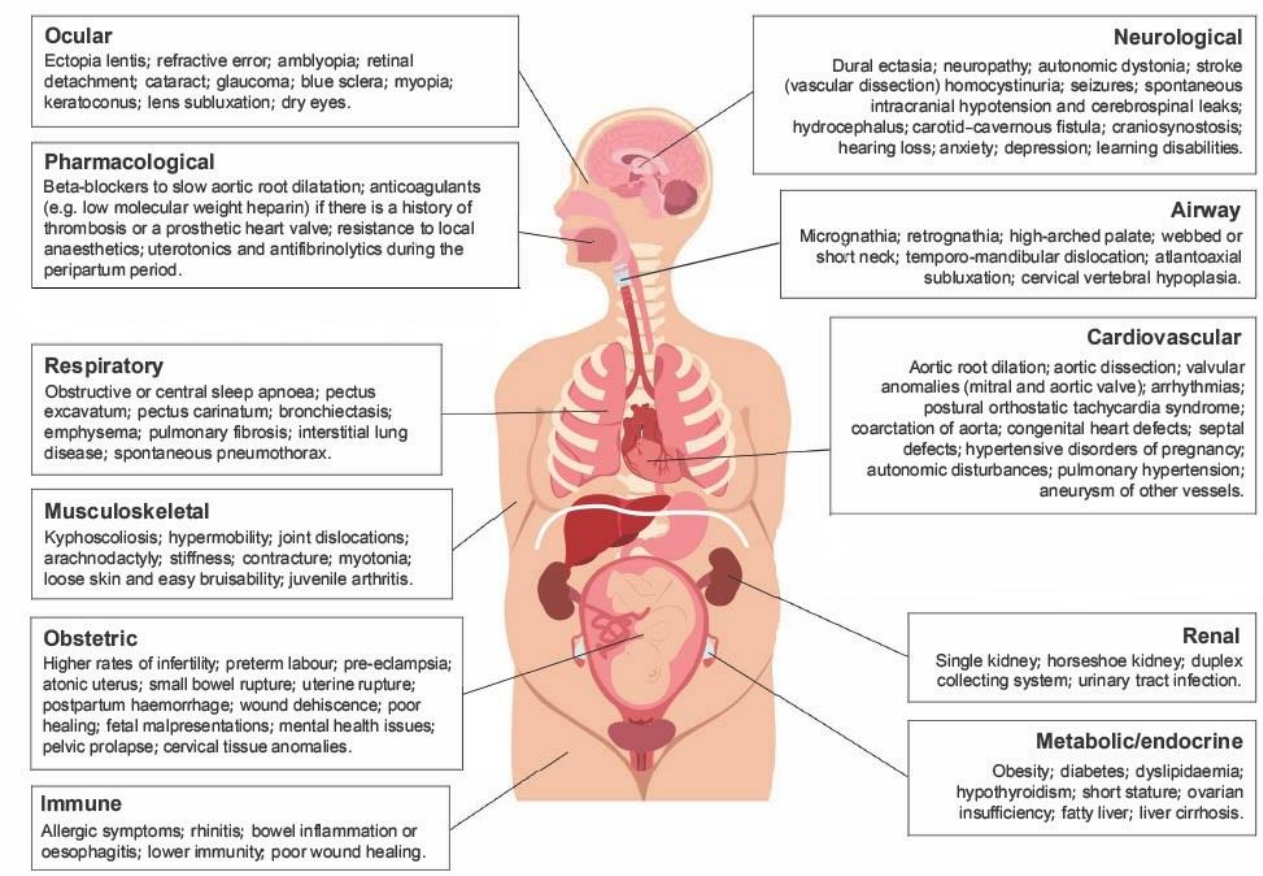
*4.4 Specific antepartum anaesthetic considerations*

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 aortic dissection, rapidly expanding aorta or aortopathy at surgical threshold) cardiothoracic anaesthetists should also be involved in birth planning. These women may, in addition to their aortopathy, have other cardiac and non-cardiac anomalies that may influence peripartum care.[21–25] 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.






Pregnant women with aortopathy should be offered review in an obstetric anaesthesia antenatal assessment clinic ideally before $28^{+0}$ weeks.[26]

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.[27] 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.

## 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. This may involve copies being held on the electronic record, in patient handheld notes and/or on the labour ward/birthing suite. The birth plan should be discussed and finalised with the woman; and owing to the risk of premature birth, iatrogenic or otherwise, this plan should be outlined and recorded early.

A copy of the care plan should be provided to the woman in line with MBRRACE-UK recommendations.[4]



The birth plan should document:

- The location of birth.
- Recommended mode of birth.
- Gestation at which birth is recommended.
- 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).
- Recommended 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 – 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 teams are being co-managed between local teams and the pregnancy heart team. As detailed in section 4.2.1, 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).[28]
- 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 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. In high risk cases, the location of birth may need to be in surgical theatres, adjacent to cardiac surgery. 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 genetic aortopathy. Vaginal birth is preferred for almost all women with cardiac disease for a number of reasons. 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 is however advised for the 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).[6,28]

- **Timing of birth**: while the evidence is limited, most teams would recommend birth before $39^{+0}$ weeks to avoid the development of the complications of hypertensive disease. With the highest risk women, where birth in a surgical centre is mandated, planned birth at 38 weeks (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, who are at increased risk of premature birth and uterine rupture.

**Table 3.** Recommended birth management (modified from AHA and 2018 ESC guidelines).[6,28]

| | Low risk mWHO II | Intermediate risk mWHO II–III | High risk mWHO III | Extremely high risk mWHO IV |
|---|---|---|---|---|
| **HTAD (including MFS and LDS)** | | < 40 mm | 40–45 mm<br>Family history of dissection | > 45 mm, all vEDS<br>Previous dissection<br>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$^2$ |
| **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.

*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 to counter the haemodynamic effects of Valsalva manoeuvre. These potentially 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.[6,28] 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 may be considered to decrease dural puncture risk.[29]
- Other neuraxial techniques described for MFS include intrathecal catheter following a dural puncture and combined spinal–epidural analgesia.
- For women taking anticoagulants, national guidelines[30] for the time intervals 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 in women who seem resistant to the effect of local anaesthetics or in whom neuraxial analgesia cannot be provided.[31]

*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), along with tachycardia.
- A combined spinal–epidural (CSE) anaesthesia technique is recommended over a single-shot spinal anaesthetic for a caesarean birth, although this decision needs to be individualised. The use of CSE provides greater block reliability and consistency of intrathecal local anaesthetic, with the more gradual introduction of epidural anaesthesia, which 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 use to facilitate caesarean birth is described in vEDS, some guidelines[32,33] recommend using general anaesthesia, especially in those who have a history of 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 of in vEDS.
- General anaesthesia for a caesarean birth can safely be utilised for obstetric (e.g. unplanned caesarean, significant postpartum haemorrhage [PPH], anaesthetic (e.g. ineffective neuraxial anaesthesia or anticoagulation or instrumentation of the spine), cardiac indications (e.g. heart failure) or maternal preference.
- Careful airway manipulation to avoid atlanto-axial subluxation and temporomandible joint dislocation is recommended especially in patients with hypermobility and EDS.
- 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 Appendix 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 needs to 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.[34]
- 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.
- Early tranexamic acid is recommended if there is a concern about blood loss.
- A massive obstetric haemorrhage call should be activated early (e.g. 20% loss of circulating blood volume taking into account woman's weight). Guidance from MBRRACE-UK[35] emphasises that arbitrary volumes for activation of haemorrhage protocols may be detrimental for women with a lower (or higher) body mass index.
- Appropriate preparation for transfusion including intra-operative 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 steroids should be offered to women as per RCOG Green-top Guideline No. 74 *Antenatal corticosteroids to reduce neonatal morbidity and mortality*.[36] If more than $37^{+0}$ weeks of gestation the evidence is less robust and steroids may not improve neonatal outcomes. Therefore, a joint discussion to assess the risks and benefits should take place between the obstetrician and the woman.
  - Magnesium sulphate should be considered for fetal neuroprotection in line with National Institute for Health and Care Excellence guideline [NG25] *Preterm labour and birth*.[37]
- **Preterm premature 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, local guidelines on the management of PPROM should be followed.
- **Wound healing**
  - Wound healing and scarring can be abnormal in women with connective tissues disorder. All members of the team and the woman should be informed and vigilant for the early signs of wound infection.

*5.8 Postpartum care*

Most aortic dissections occur postpartum, and most of these occur within the first week.[38] 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 educate women, community midwives, general practitioners/primary care providers about warning signs of dissection.



Emergency medicine clinicians also need to be vigilant and rule out dissection when a pregnant or recently pregnant woman presents with chest 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-blocker 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 lumborus block may be utilised for postoperative analgesia following caesarean birth.
- **Duration of inpatient stay** should be clearly defined.
- **Women at high risk** should have a repeat echocardiogram pre-discharge.
- **Contraception** plan should be discussed and documented. In the postpartum period these will be non-estrogen containing options. Long-term combined oral contraceptive pill should also be avoided in women with aortopathy who have mechanical valves, atrial arrhythmia or hypertension.
- A clear plan for further cardiology follow-up and imaging should be in place at the time of discharge.

## 6. Management of acute aortic dissection in pregnancy

### *6.1 Type of dissection*

Acute aortic dissection (AAD) is usually described by the Stanford classification:[39]

- **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 is essential for the formulation of management strategies and mobilisation of MDTs. 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). 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). 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 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 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 CT. The dissection flap may be visible.

*6.3 Acute general management*

Management principles for acute aortic dissection:

- **Vascular access and fluid resuscitation**: ensure adequate venous access early. Fluid resuscitation should occur and ensure blood products available.
- **Aggressive pain relief**: opiates plus antiemetics.
- **Hypertension management:** labetalol, hydralazine, intravenous glyceryl trinitrate can all be used.
- **Critical care:** ensure the women is in a critical care environment where she can be monitored closely with electrocardiogram, 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.
- **Neonatal team:** 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 maternal–fetal medicine team and neonatal team.

*6.4 Definitive management*

The Acute Aortic Dissection Pathway Toolkit[40] was developed to ensure 24-hour regional cover for AAD management in the UK, which involves a 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:

- Definitive management of type A dissection requires open heart aortic surgery using cardiopulmonary bypass.
- 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%).[41]
- For a viable fetus, between $22^{+0}$ and $26^{+6}$ weeks of gestation, a risk-based approach to decision-making should be considered where time permits. Maternal wellbeing takes precedence over the fetus, who would be considered viable before 26 weeks. The birth of a fetus in an emergency situation at less than $26^{+6}$ weeks of gestation may be associated with a unfavorable prognosis, particularly when there has not been adequate time to administer fetal steroids for the maturation of the lungs. Where possible, an informed decision should be made with the woman

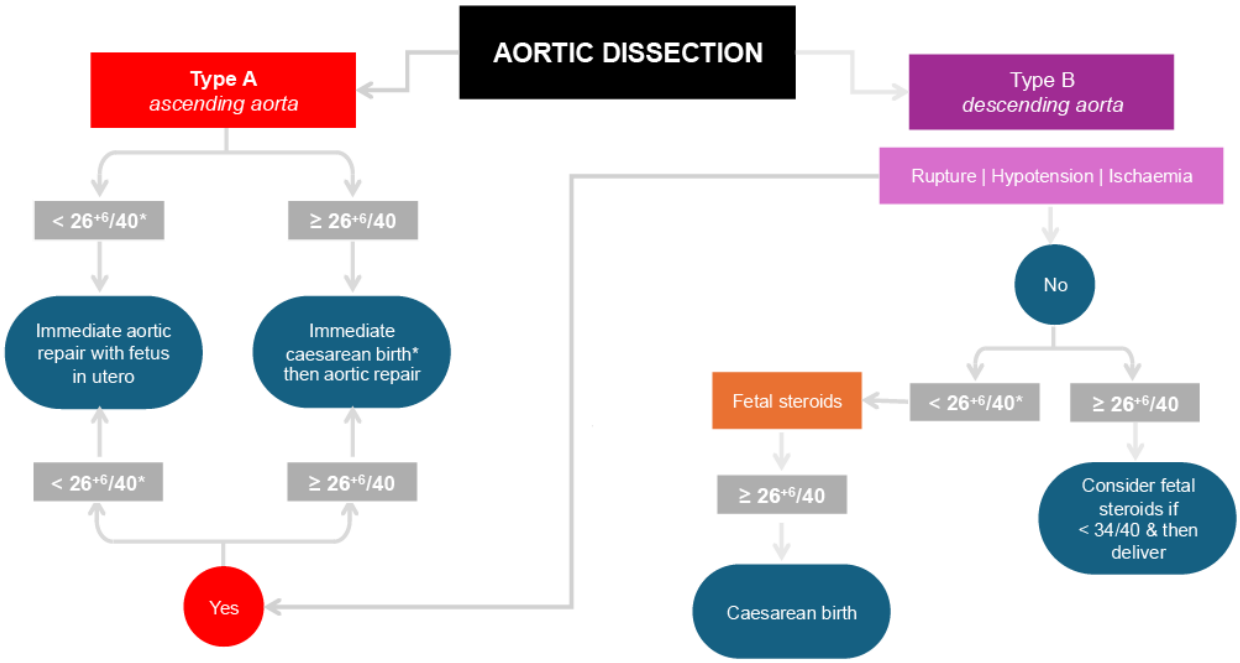


and the pregnancy heart team (including neonatal) regarding the risks of prematurity versus the risk to the fetus from cardiopulmonary bypass, using guidance such as the 2019 British Association of Perinatal Medicine Framework for Practice.[42]

- In the event that a 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.
- Type B aortic dissection in pregnancy is usually treated medically in the first instance with appropriate antihypertensive therapy. However, 20% will go on to develop complications that require intervention (either endovascular repair or open surgery).
- Good blood pressure and heart rate control with medical treatment in stable type B dissections may permit continuation of pregnancy. The timing, mode of 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.[43]
- Women who have had an aortic dissection early in pregnancy (before 20 weeks) should be counselled about the option of termination of pregnancy given the high risk nature of the pregnancy going forward.

**Figure 2.** Decision making relevant to aortic dissection type and weeks of gestation.




* For women 22–26+6/40 where time permits individualised decision making should take place regarding viability (see section 6.4).

*6.5 Cardiac arrest*

Cardiac arrest should be treated in line with the recommended advanced life support guidelines.[44,45] Manual displacement of the uterus (if more than 20+0 weeks of gestation) 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 pregnanices.

*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.[46] Healthcare professionals should continue to report adverse maternal and fetal outcomes to MBRRACE-UK and Maternity and Newborn Safety Investigations (MNSI).[44,46] Although AAD is a rare occurence, 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.[47]

## 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 dissect are unaware that they are 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.

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783 **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.<br>• Risk of airway trauma, atlanto-axial subluxation and temporomandibular joint subluxation during airway manipulation.<br>• 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.<br>• Monitoring for respiratory depression following use of intrathecal long-acting opioids.<br>• Monitor airway pressures during general anaesthesia as at risk of ventilator-associated pneumothorax.<br>• Avoid 50:50 mixture of oxygen:nitrous oxide in women with previous history of pneumothorax or pneumomediastinum.[48]<br>• Critical care support may be necessary. |
| **Cardiovascular** | • Aortic root dilation<br>• Aortic dissection<br>• Valvular abnormalities (mitral and aortic valve)<br>• Coarctation of aorta<br>• Congenital heart defects<br>• Hypertensive disorders of pregnancy<br>• Aneurysm of other vessels | • Physiological changes of pregnancy and labour causing tachycardia and hypertension.<br>• Serial echocardiography.<br>• Good labour analgesia to mitigate sympathomimetic effects of labour.<br>• ECG/NIBP/$SpO_2$ with arterial line for invasive blood pressure monitoring in women at high risk during labour and operative birth.<br>• Appropriate IV access and blood products including intra-operative cell salvage.<br>• Activation of major obstetric haemorrhage if necessary.<br>• Avoidance of swings in blood pressure (hypotension and hypertension) and tachycardia during operative birth or general anaesthesia.<br>• Maintenance of systolic arterial pressure at > 90% accurate baseline using vasopressors (e.g. phenylephrine infusion) during neuraxial anaesthesia.<br>• Suppress pressor response to laryngoscopy and extubation.<br>• Point-of-care ultrasound as necessary and if expertise available. |



 

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| **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.<br>• Ensure pressure areas are padded.<br>• 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.<br>• Consider pre-puncture ultrasound to facilitate neuraxial analgesia (epidural) during labour.<br>• If neuraxial analgesia not feasible, consider alternatives including remifentanil patient-controlled analgesia, intramuscular opioids or 50:50 mixture of oxygen : nitrous oxide (Entonox®).<br>• Combined-spinal epidural anaesthetic preferable for caesarean birth.<br>• 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.<br>• Monitoring blood sugar in line with national recommendations.[49] |
| **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.<br>• Anticoagulants (e.g. low molecular weight heparin) if there is a history of thrombosis or a prosthetic mechanical heart valve.<br>• Resistance to local anaesthetics.<br>• Uterotonics and antifibrinolytics during the peripartum period. | • Continue beta blockers during labour.<br>• Anticoagulants may need to be tailored prior to neuraxial analgesia and anaesthesia in line with national guidelines.<br>• Appropriate monitoring of anticoagulation as necessary.<br>• Need to use higher (but safe) doses of local anaesthetic or alternative forms of pain relief, or conversion to general anaesthesia if necessary.<br>• Avoid ergometrine and consider using tranexamic acid to decrease PPH.<br>• Administer anti-emetics to decrease the risk of nausea, vomiting and organ rupture.<br>• Multimodal analgesia following operative birth – avoid non-steroidal |



 

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| | | anti-inflammatory agents in patients with bleeding diathesis. |
| **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.<br>• Higher caesarean birth rates (63%).<br>• PPH rates and sequalae may be higher. |

784 ART, assisted reproductive techniques; ECG, electrocardiogram; NIBP, non-invasive blood pressure; PPH, postpartum haemorrhage; $SpO_2$, oxygen saturation.
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**Appendix II:** Dural ectasia and its relevance to anaesthesia.[27,50–53]

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

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

**Appendix III:** Glossary

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|---|---|
| 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 areas |
| CMR | cardiac magnetic resonance |
| CSE | combined spinal–epidural |
| CT | computerised 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] |
| PGT-M | preimplantation genetic testing for monogenic disorders |
| PPH | postpartum haemorrhage |
| PPROM | preterm premature rupture of the membranes |
| TS | Turner syndrome |
| vEDS | vascular Ehlers-Danlos syndromes |

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The Chair of the Patient Safety Committee was: Dr SL Cunningham MRCOG, Stoke-on-Trent[1]; Dr CJ Calderwood FRCOG, Clydebank[2]; and the Vice Chair was: Dr J Elson FRCOG, Nottingham.
[1]until May 2024; [2]from June 2024

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

The review process will commence in XXXX, unless otherwise indicated.