

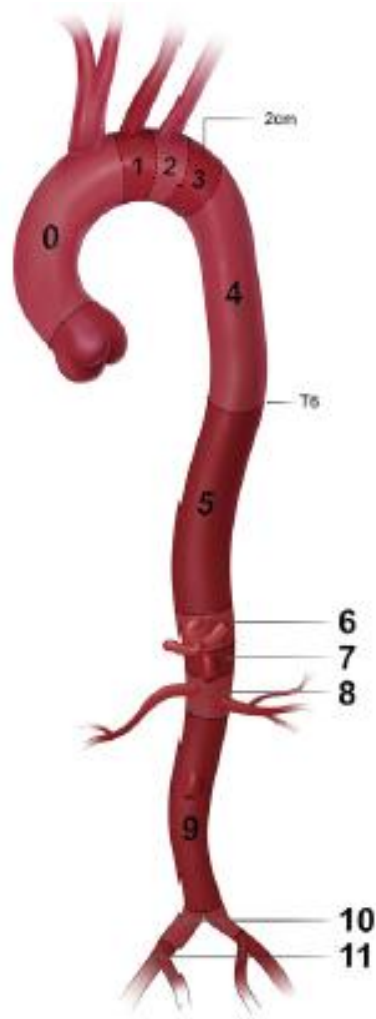


Figure 1. ACC/AHA 2022 GL on Aortic Disease. Classification of aortic anatomy segments by 11 landing zones.

Zone 0 (involves the ascending to distal end of the origin of the innominate artery); Zone 1 (involves the origin of the left common carotid; between the innominate and the left carotid); Zone 2 (involves the origin of the left subclavian; between the left carotid and the left subclavian); Zone 3 (involves the proximal descending thoracic aorta down to the T4 vertebral body; the first 2 cm distal to the left subclavian); Zone 4 (the end of zone 3 to the mid-descending aorta – T6); Zone 5 (the mid-descending aorta to the celiac); Zone 6 (involves the origin of the celiac; the celiac to the superior mesenteric); Zone 7 (involves the origin of the superior mesenteric artery; the superior mesenteric to the renals); Zone 8 (involves the origin of the renal arteries; the renal to the infrarenal abdominal aorta); Zone 9 (the infrarenal abdominal aorta to the level of aortic bifurcation); Zone 10 (the common iliac); Zone 11 (involves the origin of the external iliac arteries).

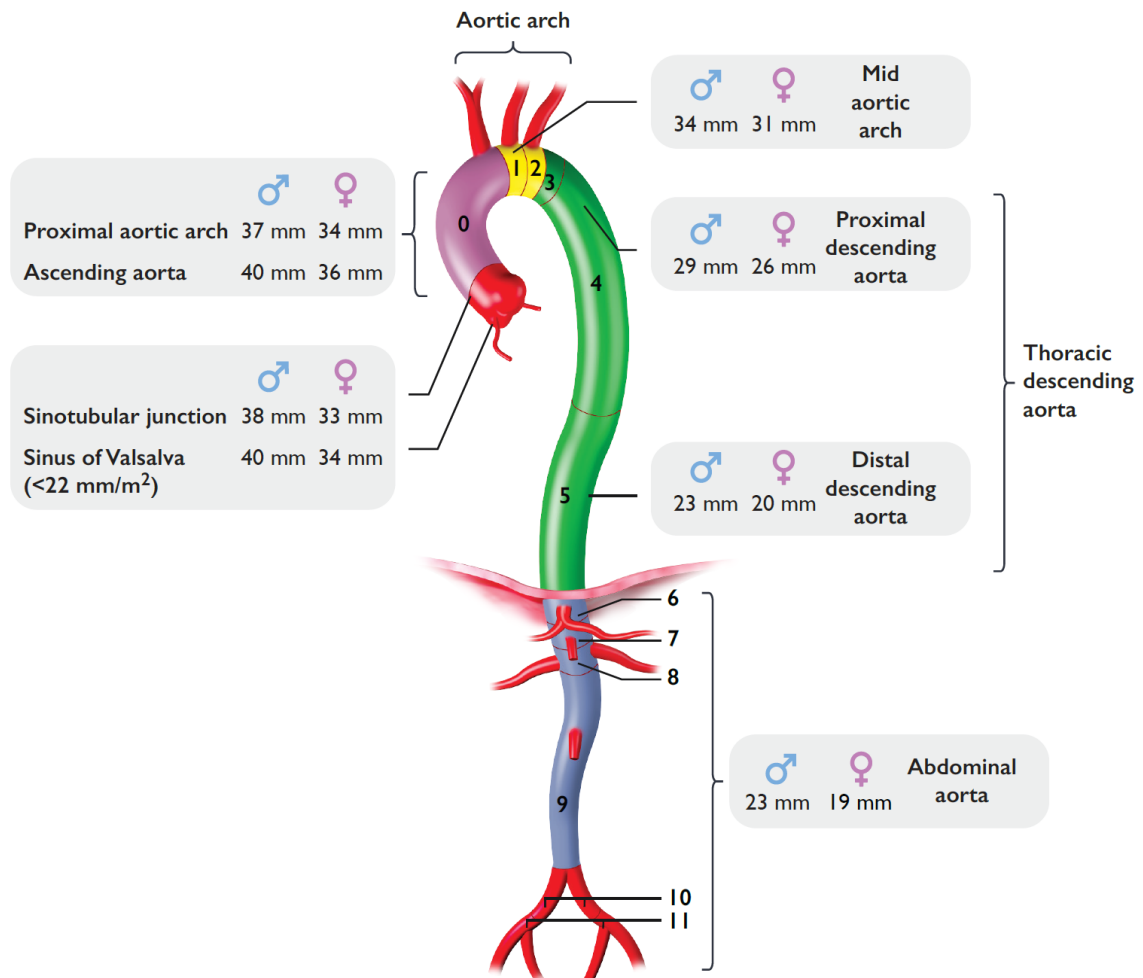


Figure 2. ESC 2024 GL on peripheral and aortic diseases. Anatomy and aortic segments and upper normal values for aortic dimensions. Numbers represent the 11 aortic segments based on the Society for Vascular Surgery/Society of Thoracic Surgeons (SVS/STS) classification for surgical and endovascular purposes.

Z-scores can be calculated for aortic root and ascending aorta. Calculation of z-scores can be performed following these links: <https://www.marfan.fr/accueil/z-score-calculus/> or <https://marfan.org/dx/z-score-adults>.

Table 1. ACC/AHA 2022 GL on Aortic Disease. Recommendations for Aortic Imaging Techniques to Determine Presence and Progression of Aortic Disease.

In patients with known or suspected aortic disease:

Aortic diameters measured at reproducible anatomic landmarks perpendicular to axis of blood flow, and these measurement methods reported in a clear and consistent manner. In cases of asymmetric or oval contour, report the longest diameter and its perpendicular diameter.	I B-NR
Keep episodic and cumulative ionizing radiation doses as low as feasible while maintaining diagnostic image quality.	I C-LD
When performing CT or MR imaging, the root and ascending aortic diameters should be measured from inner-edge to inner-edge, using an electrocardiographic-synchronized technique. If there are aortic wall abnormalities, such as atherosclerosis or discrete wall thickening (more common in the distal aorta), the outer-edge to outer-edge diameter should be reported.	I C-EO
The aortic root diameter should be recorded as maximum sinus to sinus measurement. In the setting of known asymmetry, report multiple measurements, and obtain both short- and long-axis images of the root to avoid underestimation of the diameter.	I C-EO
A dilated root or ascending aorta should be indexed to patient height or BSA in the report, to aid in clinical risk assessment.	IIa C-LD
When performing echocardiography, measure the aorta from leading-edge to leading-edge, perpendicular to the axis of blood flow.	IIa C-EO
Using inner-edge to inner-edge measurements, particularly on short-axis imaging.	IIb C-EO

Table 2. ESC 2024 GL on peripheral and aortic diseases. Recommendations for imaging of the aorta

It is recommended that aortic diameters are measured at pre-specified anatomical landmarks, and the largest diameter of the section be perpendicular to the longitudinal axis. ^{134,135}	I	C
It is recommended in cases of serial imaging of the aorta over time to use the same imaging modality with the same measurement method. ¹⁵⁹	I	C
It is recommended to consider renal function, pregnancy, age, and history of allergy to contrast media to select the optimal imaging modality with minimal radiation exposure and lowest iatrogenic risk, except for emergency cases. ^{159–161}	I	C
Indexing aortic diameters to BSA, along with the use of nomograms, z-scores, or other indexing methods, should be considered for more accurate assessment of aortic size, especially for body sizes at the lower end of the normal distribution. ^{156–158}	IIa	B

BSA body surface area

Table 3. ESC 2024 GL on peripheral and aortic diseases. Recommendations for thoracic aortic measurements

TTE is recommended as the first-line imaging technique in evaluating thoracic aortic diseases. ^{159,165}	I	B
It is recommended to report aortic diameters using the leading-to-leading edge convention in end-diastole by echocardiography. ^{137,139,140,159}	I	C
It is recommended to report aortic diameters using the inner-to-inner edge convention in end-diastole by CCT or CMR. ^{137,143,144,159}	I	C
It is recommended to report aortic diameters from images obtained with the double-oblique technique (not axial images) by CCT or CMR. ¹⁴⁸	I	C
ECG-triggered CCT is recommended for comprehensive diagnosis, follow-up, and pre-invasive treatment assessment of the entire aorta, particularly the root and ascending aorta. ¹⁵⁹	I	C
CMR is recommended for diagnosis and follow-up of thoracic aortic diseases, especially when chronic follow-up is required. ^{166–168}	I	C
The aortic root should be measured using the cusp-to-cusp distance. Also, the presence of asymmetry (>5 mm) among distances should be reported. ^{137,146}	IIa	C
If an increase of ≥ 3 mm per year in aortic diameters by TTE is observed, confirmation by CCT/CMR should be considered. ^{137,159}	IIa	C
Chest X-ray may be considered in cases of low clinical probability of AAS; however, a negative exploration should not delay dedicated aortic imaging in high-risk patients. ^{162–164}	IIb	C

AAS, acute aortic syndrome; CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; ECG, electrocardiogram; TTE, transthoracic echocardiography.

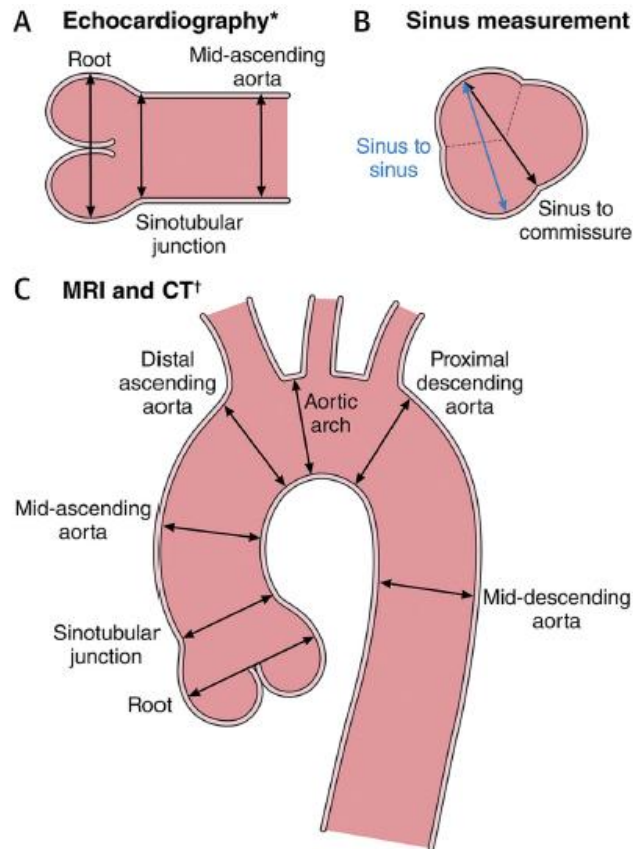


Figure 3. ACC/AHA 2022 GL on Aortic Disease. Aortic Imaging Techniques to Determine the Presence and Progression of Aortic Disease.

(A) Schematic shows the leading-edge to leading-edge measurement technique used in echocardiography, from left to right: measurement of the aortic root (sinuses of Valsalva), sinotubular junction, and proximal tubular ascending aorta. (B) Inner-wall to inner-wall measurements of the aortic root used in MRI and CT. In addition, a consistent approach to measuring all 3 sinuses with MRI and CT is necessary. The sinus-to-commissure and sinus-to-sinus measurements can both be used, but consistency is necessary for interval surveillance. (C) Standard measurement locations for MRI and CT with the inner-wall to inner-wall technique.

Table 4. ACC/AHA 2022 GL on Aortic Disease. Recommendations for Multidisciplinary Aortic Teams

For patients with acute aortic disease that requires urgent repair,
a multidisciplinary team should determine the most suitable intervention.

I C-EO

For patients who are asymptomatic with extensive aortic disease,
or who may benefit from complex open and endovascular aortic repairs,
or with multiple comorbidities for whom intervention is considered,
referral to a high-volume center (performing at least 30-40 aortic procedures annually)
with experienced surgeons in a Multidisciplinary Aortic Team to optimize treatment outcomes.

IIa C-LD

Table 5. ACC/AHA 2022 GL on Aortic Disease. Recommendations for Shared Decision Making.

In patients with aortic disease, shared decision-making when determining the appropriate thresholds for intervention, deciding on the type of surgical repair, choosing between open surgical versus endovascular approaches; and in medical management and surveillance.	I C-LD
In patients with aortic disease who are contemplating pregnancy or who are pregnant, shared decision making when considering the cardiovascular risks of pregnancy, the diameter thresholds for prophylactic aortic surgery, and the mode of delivery.	I C-EO

Table 6. ACC/AHA 2022 GL on Aortic Disease. Physical Activity and Quality of Life

For patients with significant aortic disease, education and guidance about avoiding intense isometric exercises (eg, heavy weightlifting or activities requiring the Valsalva maneuver), burst exertion and activities, and collision sports.	I C-EO
For patients who have undergone surgery for aortic aneurysm or dissection, postoperative cardiac rehabilitation.	I C-EO
In patients with thoracic or abdominal aortic aneurysms whose BP is adequately controlled, encourage 30 to 60 minutes of mild-to-moderate intensity aerobic activity at least 3 to 4 days per week.	IIa C-LD
In patients with clinically significant aortic disease, screen for anxiety, depression, and posttraumatic stress disorder and, when indicated, provide resources for support; provide education and resources to minimize patients' concerns, support optimal decision making, and enhance quality of life.	IIa C-LD

Table 7. ESC 2025 GL on pregnancy. Recommendations for aortopathies, cardiac surgery, and pregnancy

Counselling		
It is recommended that women with aortic disease have counselling about the risk of aortic dissection in pregnancy and the post-partum period. ^{404,423}	I	C
It is recommended that women with a history of aortic dissection or -surgery have pre-pregnancy counselling about the high risk by an extended Pregnancy Heart Team ^c considering the presence and type of genetic variant, aortic morphology, growth rate, and aetiology of aortic dissection. ^{7,381,424}	I	C
It is recommended that women with vascular Ehlers–Danlos syndrome wishing to become pregnant are counselled regarding the very high risk of pregnancy-related adverse events by a multidisciplinary team, considering family history, genetic variant, and previous vascular events. ^{7,396,397,425}	I	C
Imaging		
Imaging of the entire aorta ^d (CT or CMR) is recommended before pregnancy in women with known or suspected aortic disease. ^{390,424,426}	I	C
In women with aortic dilatation related to BAV, imaging (with TTE, and CMR/CT if needed) of the aortic root, ascending aorta, and descending aorta (to rule out coarctation) is recommended before pregnancy. ^{427,428}	I	C
In women with low-risk aortic disease (mWHO 2.0 classes II and II–III), one-time echocardiographic imaging between 20 and 30 weeks of gestation and imaging at 6 months post-partum is recommended. ^{393,429}	I	C
In women with moderate to high-risk aortic disease (mWHO 2.0 classes III and IV), repeated echocardiographic imaging every 4–12 weeks (depending on diagnosis and severity of dilatation) is recommended during pregnancy and until 6 months post-partum. ^{393,429,430}	I	C
CMR (without gadolinium) imaging of the entire aorta is recommended in pregnant women at risk of or with known aortic dilatation who have not had recent pre-pregnancy cross sectional imaging. ¹³⁵	I	C
Treatment—medical		
When a woman with known aortic dilatation, history of dissection, or P/LP variant associated with aortic disease becomes pregnant, strict and individualized BP control is recommended. ^{431,432}	I	C
Beta-blocker therapy ^e throughout pregnancy and in the post-partum period is recommended in women with MFS and other HTADs. ⁴³³	I	C
Celiprolol is recommended in women with vascular Ehlers–Danlos syndrome during pregnancy and lactation. ⁴³⁴	I	C
Treatment—intervention/surgical		
It is recommended that indications for pre-pregnancy aortic root and/or ascending aortic surgery are guided by aortic morphology, underlying pathology, family history, genetic variant, previous vascular events, and patient's preference. ^{62,383}	I	C
It is recommended that centres managing pregnancies in women with moderate to high-risk aortic disease (mWHO 2.0 class III/IV) can provide cardiovascular surgery in case of peripartum adverse events. ^{435,436}	I	C
Specific conditions		
In women with MFS and aortic root diameters >45 mm, surgery before pregnancy is recommended. ^{380,424}	I	C
In women with LDS with P/LP variants in <i>TGFBR1</i> , <i>TGFBR2</i> , and aortic root diameters ≥45 mm, surgery before pregnancy is recommended. ^{437–439}	I	C

In women with nsHTAD with P/LP variants in <i>MYH11</i> , <i>ACTA2</i> , <i>PRKG1</i> , or <i>MYLK</i> , and aortic root diameters ≥ 45 mm, surgery before pregnancy is recommended. ^{385,386,440}	I	C
In women with BAV and aortic root or ascending aortic diameter ≥ 50 mm, surgery before pregnancy is recommended. ^{441,442}	I	C
In women without an identifiable P/LP variant with aortic root or ascending aortic diameters ≥ 50 mm, surgery before pregnancy is recommended.	I	C
In women with HTAD and aortic arch, descending aortic, or abdominal aortic diameters ≥ 50 mm, surgery before pregnancy should be considered. ^{392,443}	IIa	C
In women with LDS with P/LP variants in <i>TGFβ2</i> , <i>TGFβ3</i> , <i>SMAD2</i> , and <i>SMAD3</i> , and aortic root diameters ≥ 45 mm, surgery before pregnancy should be considered. ^{437–439}	IIa	C
In women with BAV and root phenotype or family history of aortic aneurysm or dissection, surgery before pregnancy should be considered if the aorta is ≥ 45 mm.	IIa	C
In women without an identifiable P/LP variant with aortic root or ascending aortic aneurysm ≥ 45 mm, surgery before pregnancy should be considered in the presence of a family history of aortic aneurysm, aortic dissection, uncontrolled arterial hypertension, or on patient's preference.	IIa	C
In women with MFS and aortic root diameters between 40 and 45 mm, surgery before pregnancy may be considered if risk factors (growth > 3 mm/year, family history of aortic dissection) are present. ⁴⁴⁴	IIb	C
In women with LDS with P/LP variants in <i>TGFβR1</i> or <i>TGFβR2</i> and aortic root diameters ≥ 40 mm, surgery before pregnancy may be considered. ^{387,437,445,446}	IIb	C
In women with nsHTAD and aortic root diameters ≥ 40 –44 mm, surgery before pregnancy may be considered depending on the genetic variant, family history, and aortic growth rate. ^{385,386,440}	IIb	C
Delivery		
In women with an aorta < 40 mm, vaginal delivery is recommended. ¹⁶³	I	C
In women with vascular Ehlers–Danlos syndrome, caesarean section at 37 weeks is recommended for obstetrical reasons. ⁴⁰⁰	I	C
In women with an aorta 40–45 mm, vaginal delivery with epidural anaesthesia and an expedited second stage should be considered.	IIa	C
In women with an aorta ≥ 45 mm, caesarean section should be considered.	IIa	C
In women with acute, subacute, or chronic aortic dissection, caesarean section should be considered.	IIa	C
In women with an aorta 40–45 mm, caesarean section may be considered.	IIb	C
The use of ergometrine post-delivery is not recommended in women with aortopathy.	III	C
Cardiac surgery during pregnancy		
Delivery before cardiac surgery should be considered as soon as the foetus is viable, taking gestational age, comorbidities, and the available level of neonatal care into account. ^{163,421,435,447}	IIa	C
Cardiac surgery may be considered during pregnancy when conservative and medical therapy has failed, and in situations that threaten the mother's life or that are not amenable to percutaneous treatment.	IIb	C

BAV, bicuspid aortic valve; BP, blood pressure; CMR, cardiovascular magnetic resonance imaging; CT, computed tomography; HTAD, heritable thoracic aortic disease; LDS, Loeys–Dietz syndrome; MFS, Marfan syndrome; mWHO, modified World Health Organization; nsHTAD, non-syndromic heritable thoracic aortic disease; P/LP, pathogenic/likely pathogenic; TTE, transthoracic echocardiogram.

C Extended Pregnancy Heart Team: regular team + multidisciplinary aortic team.

d In women with vascular Ehlers–Danlos syndrome and LDS, imaging should encompass the entire aorta, including supra-aortic vessels as well as iliac and femoral arteries.

e See Chapter on Pregnancy for beta-blocker choice.

Table 8. ACC/AHA 2022 GL on Aortic Disease.

Counseling and Management of Aortic Disease in Pregnancy and Postpartum

Prepregnancy

In patients with genetic aortopathies attributable to syndromic (Marfan syndrome, Loeys–Dietz syndrome, vascular Ehlers–Danlos syndrome) and nsHTAD and who are contemplating pregnancy, genetic counseling before pregnancy to discuss the heritable nature of their condition. I C-LD

In patients with syndromic and nsHTAD, Turner syndrome, BAV with aortic dilation, and other aortopathy conditions, aortic imaging (with TTE, MRI or CT, or both as appropriate) before pregnancy to determine aortic diameters. I C-LD

In patients with syndromic and nsHTAD, Turner syndrome, BAV with aortic dilation, and other aortopathy conditions, who are contemplating pregnancy, counseling about the risks of aortic dissection related to pregnancy. I C-LD

During Pregnancy

- In patients with aortic aneurysms, or at increased risk of aortic dissection, or both, pregnancy to be managed by a multidisciplinary team including a maternal fetal medicine specialist and cardiologist, and, if logistically feasible, that delivery be planned in a hospital where the capability for emergency aortic repair is available. IIa C-EO
- Guideline-directed treatment of hypertension in pregnant patients with aortopathies I C-LD
- In patients with syndromic and nsHTAD, beta-blocker therapy during pregnancy and postpartum, unless contraindicated. I C-LD
- In pregnant patients with an aortopathic condition or a dilated aortic root or ascending aorta, surveillance TTE to monitor aortic diameters and aortic valve function each trimester and again several weeks postpartum, although imaging may be more frequent depending on aortic diameter, aortic growth rate, and underlying condition. I C-LD
- In pregnant patients with aortic disease who require surveillance imaging of the aortic arch, I C-LD descending, abdominal aorta, or all 3, MRI without gadolinium preferred to CT to avoid radiation exposure to the fetus.

Delivery in Pregnant Patients With Aortopathy

- Cesarean delivery in patients with a history of chronic aortic dissection. I C-EO
- Vaginal delivery (when otherwise appropriate), in an aortic diameter of <4.0 cm. I C-EO
- Cesarean deliver in a diameter of the aortic root, ascending aorta, or both, of ≥ 4.5 cm. IIa C-EO
- In a diameter of the aortic root, ascending aorta, or both, of 4.0 cm to 4.5 cm, vaginal delivery with regional anesthesia, expedited second stage, and assisted delivery. IIb C-EO
- In patients with syndromic and nsHTAD, and a diameter of the aortic root, ascending aorta, or both, of 4.0 cm to 4.5 cm, cesarean delivery. IIb C-EO

Surgery Before Pregnancy in Women With Aortic Disease

Aortic surgery before pregnancy:

- Marfan syndrome and an aortic root diameter of >4.5 cm I C-LD
- If the aortic root diameter is 4.0 cm to 4.5 cm, especially if there are risk factors for aortic dissection (ie, rapid aortic growth of ≥ 0.3 cm/y or a family history of aortic dissection). IIb C-LD
- In Loeys-Dietz syndrome attributable to pathogenic variants in TGFB2 or TGFB3 and an aortic diameter of ≥ 4.5 cm. IIa C-EO
- If the Loeys-Dietz syndrome is attributable to pathogenic variants in TGFBR1, IIb C-EO

TGFB2, or SMAD3, and the aortic diameter is ≥ 4.0 cm.

In nsHTAD and an aortic diameter of ≥ 4.5 cm.

I C-EO

If the aortic diameter is 4.0 cm to 4.4 cm, , depending on the molecular diagnosis, family history, and aortic growth rate.

IIb C-EO

In Turner syndrome and ASI of ≥ 2.5 cm/m².

I C-LD

In wih BAV (in the absence of Turner syndrome or an HTAD) and an aortic diameter of ≥ 5.0 cm.

I C-EO

In patients with sporadic aortic root aneurysms, ascending aortic aneurysms, or both and a diameter of ≥ 5.0 cm

I C-EO

Prophylactic Aortic Surgery Before Pregnancy in Women With Aortopathic Conditions

Condition	Surgical Threshold Before Pregnancy* by Aortic Diameter (cm) or Aortic Size Index (cm/m ²)
Marfan syndrome	>4.5 cm
Marfan syndrome with risk factors (rapid aortic growth of ≥ 0.3 cm/y; family history of aortic dissection)	4.0–4.5 cm
Loeys-Dietz syndrome (attributable to pathogenic variants in <i>TGFB1</i> , <i>TGFB2</i> , or <i>SMAD3</i>)	≥ 4.0 cm
Loeys-Dietz syndrome (attributable to pathogenic variants in <i>TGFB2</i> or <i>TGFB3</i>)	≥ 4.5 cm
Nonsyndromic heritable thoracic aortic disease	≥ 4.5 cm†
Turner syndrome	≥ 2.5 cm/m ²
Bicuspid aortic valve	≥ 5.0 cm‡

*Shared decision-making is required to determine the surgical threshold before elective aortic root, ascending aortic surgery, or both and is informed by the condition, specific pathogenic variant, age, body size, aortic growth rate, phenotype, and family history of aortic dissection, and surgery at smaller aortic diameters may be considered depending on individual circumstances.

†Aortic dissection related to pregnancy has occurred at small aortic diameters in women with *ACTA2* and *MYLK* pathogenic variants. Prophylactic aortic surgery before pregnancy at smaller aortic diameters may be reasonable in these conditions and other nonsyndromic heritable thoracic aortic disease and may be informed by the molecular diagnosis, family history, and aortic growth rate.

‡Prophylactic aortic surgery may be considered at smaller aortic diameters depending on body size, aortic growth rate, and family history.

Pregnancy in Patients With Aortopathy: Aortic Dissection and Aortic Surgery in Pregnancy

In acute type A aortic dissection during the first or second trimester of pregnancy, urgent aortic surgery with fetal monitoring.	I C-LD
In acute type A aortic dissection during the third trimester of pregnancy, urgent cesarean delivery immediately followed by aortic surgery.	I C-LD
In acute type B aortic dissection during pregnancy, medical therapy unless endovascular or surgical therapy is required to manage acute complications.	I C-EO
In progressive aortic dilation during pregnancy, prophylactic aortic surgery, depending on individual circumstances.	IIb C-EO