

Table 1. ACC/AHA 2022 GL on Aortic Disease. TAA Syndromes and Conditions Attributable to a Heritable or Genetic Cause

Condition	Gene	Clinical Features
Syndromic HTAD*		
Marfan syndrome	<i>FBN1</i>	Aortic root aneurysm, aortic dissection, TAA, MVP, long bone overgrowth, arachnodactyly, dolichostenomelia, scoliosis, pectus deformities, ectopia lentis, myopia, tall stature, pneumothorax, dural ectasia
Loeys-Dietz syndrome	<i>TGFBR1, TGFBR2, SMAD3,† TGFBR2, TGFBR3</i>	TAA, branch vessel aneurysms, aortic dissection, arterial tortuosity, MVP, craniosynostosis, hypertelorism, bluish sclera, bifid/broad uvula, translucent skin, visible veins, club feet, dural ectasia, and premature osteoarthritis and peripheral neuropathy†
Vascular Ehlers-Danlos syndrome	<i>COL3A1</i>	TAA, AAA, arterial rupture, aortic dissection, MVP, bowel and uterine rupture, pneumothorax, translucent skin, atrophic scars, small joint hypermobility, easy bruising, carotid-cavernous fistula
Arterial tortuosity syndrome	<i>SLC2A10</i>	Tortuous large and medium sized arteries, aortic dilation, craniofacial, skin and skeletal features
Shprintzen-Goldberg syndrome	<i>SKI</i>	Craniosynostosis, skeletal features, aortic dilation
Ehlers-Danlos syndrome with periventricular nodular heterotopia	<i>FLNA</i>	X-linked, periventricular nodular heterotopia, TAA, BAV, MV disease, PDA, VSD, seizures, joint hypermobility
Meester-Loeys syndrome	<i>BGN</i>	X-linked, TAA, aortic dissection, MV disease
LOX-related TAA	<i>LOX</i>	TAA, BAV, aortic dissection, Marfanoid habitus in some
Smooth muscle dysfunction syndrome	<i>ACTA2</i>	TAA, moyamoya-like cerebrovascular disease, pulmonary hypertension, pulmonary disease, hypoperistalsis, hypotonic bladder, congenital mydriasis [‡]
Nonsyndromic HTAD (Familial TAA)		
FTAA	<i>ACTA2</i>	TAA, aortic dissection, premature CAD and moyamoya-like cerebrovascular disease, livedo reticularis, iris flocculi
FTAA	<i>MYH11</i>	TAA, aortic dissection, PDA
FTAA	<i>MYLK</i>	Aortic dissection at relatively small aortic size
FTAA	<i>PRKG1</i>	Aortic dissection at young ages at small aortic sizes
FTAA	<i>MAT2A</i>	TAA, aortic dissection, BAV
FTAA	<i>MFAP5</i>	TAA, aortic dissection, skeletal features may be present
FTAA	<i>FOXE3</i>	TAA, aortic dissection
FTAA	<i>THSD4</i>	TAA, aortic dissection
Bicuspid Aortic Valve-Associated Ascending Aortic Aneurysm		
Familial BAV/AS and TAA	<i>NOTCH1</i>	Aortic valve stenosis, TAA
BAV with TAA	<i>TGFBR2, MAT2A, GATA5, SMAD6, LOX, ROBO4, TBX20</i>	Syndromic and nonsyndromic HTAD and FTAA with an increased frequency of BAV
Turner syndrome	<i>XO, Xp</i>	BAV, CoA, TAA, aortic dissection, short stature, lymphedema, webbed neck, premature ovarian failure

*Some individuals with pathogenic variants in a gene that can lead to syndromic HTAD have very few or no syndromic features, and variants in some genes causing syndromic HTAD may also lead to nonsyndromic HTAD.

†SMAD3 premature osteoarthritis and peripheral neuropathy.

AAA indicates abdominal aortic aneurysm; AS, aortic stenosis; BAV, bicuspid aortic valve; CAD, coronary artery disease; CoA, coarctation of the aorta; EDS, Ehlers-Danlos syndrome; FTAA, familial thoracic aortic aneurysm (and dissection) syndrome; HTAD, heritable thoracic aortic disease; MV, mitral valve; MVP, mitral valve prolapse; PDA, patent ductus arteriosus; TAA, thoracic aortic aneurysm; and VSD, ventricular septal defect.

Table 2. ESC 2024 Guidelines on Aortic and Peripheral Diseases.**Recommendations for the management of patients with heritable thoracic aortic disease**

It is recommended that medical management of patients with HTAD is individualized and based on shared decision-making. ¹³⁸⁶	I	C
It is recommended that patients with known or suspected syndromic or non-syndromic HTAD are evaluated in a centre with experience in the care of this patient group. ⁸⁸⁸	I	C

HTAD, heritable thoracic aortic disease.

Recommendations for genetic testing and aortic screening in aortic disease

Genetic testing		
In patients with aortic root/ascending aneurysms or thoracic aortic dissection, gathering family history information for at least three generations about TAD, unexplained sudden deaths, and peripheral and intracranial aneurysms is recommended. ^{880,1396–1402}	I	B
In patients with aortic root/ascending aortic aneurysms or thoracic aortic dissection and risk factors for HTAD, ^c genetic counselling at an expert centre and subsequent testing, if indicated, is recommended. ^{1399,1403–1408}	I	B
In patients with HTAD who have a pathogenic/likely pathogenic variant, genetic testing of at-risk biological relatives (i.e. cascade testing) is recommended, irrespective of age. ^{70,1407,1409}	I	C
In patients with HTAD, guidance of clinical management by the underlying gene/variant, when known, should be considered. ^{70,1391,1410–1416}	IIa	B
Aortic imaging screening		
In patients with TAD with risk factors for HTAD, ^c with a negative family history of TAD and in whom no (likely) pathogenic variant is identified, TTE ^d screening aortic imaging of FDRs ^e is recommended. ^{1396,1402}	I	B
Imaging screening of family members of patients with TAD with risk factors for HTAD ^e in whom no (likely) pathogenic variant is identified should be considered starting at age 25, or 10 years below the youngest case, whichever is younger. If the initial screening is normal, continued screening every 5 years until the age of 60 should be considered. ²⁵	IIa	C

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; FDR, first-degree relative; HTAD, heritable thoracic aortic disease; TAD, thoracic aortic disease; TTE, transthoracic echocardiography.

d CMR/CCT may be indicated if the aortic root/ascending aorta cannot be visualized adequately.

e Parents, siblings, children.

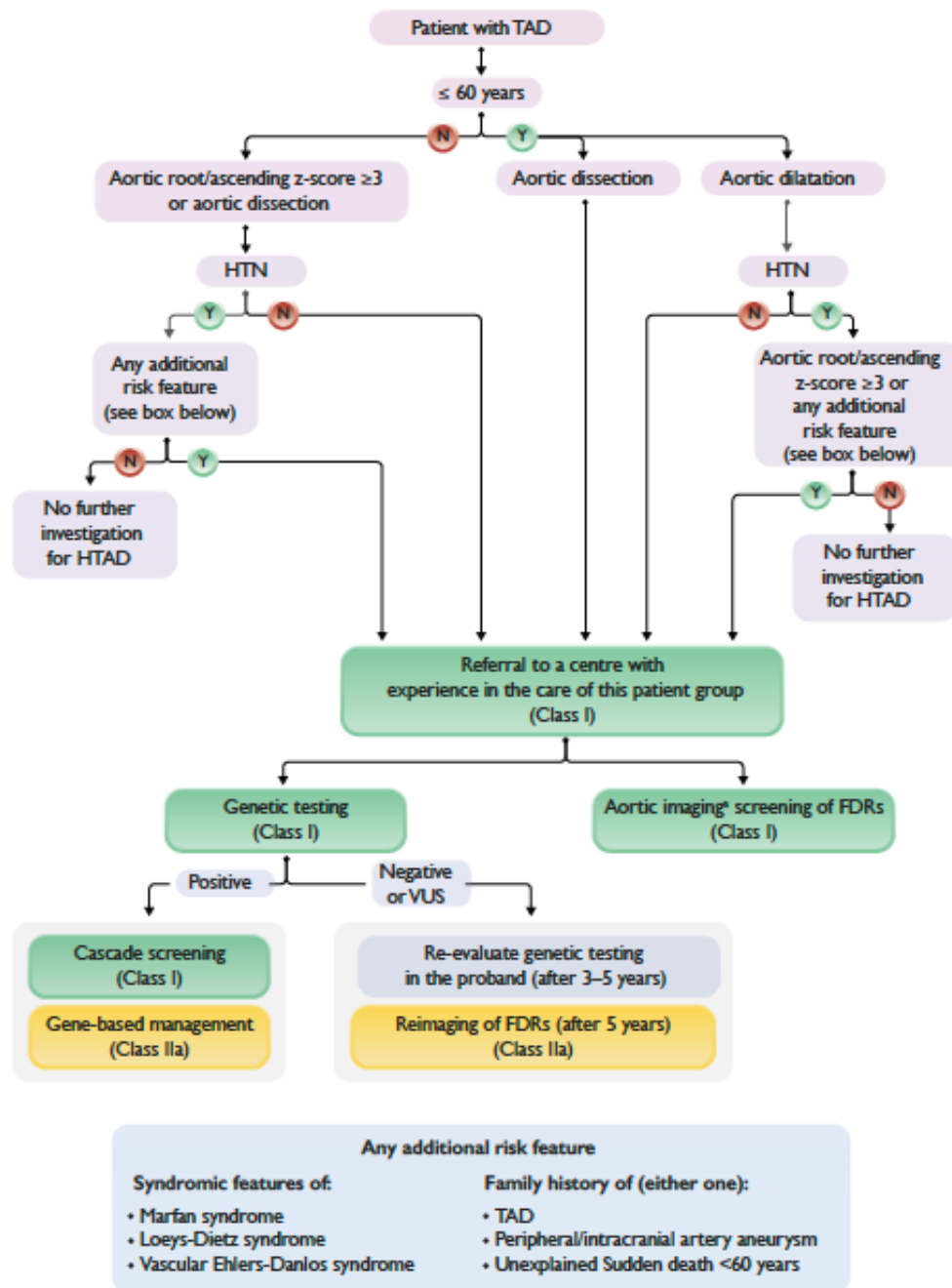


Figure 1. ESC 2024 Guidelines on Aortic and Peripheral Diseases. Algorithm for genetic and imaging screening in patients with thoracic aortic disease.

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; FDR, first-degree relative; HTAD, heritable thoracic aortic disease; HTN, arterial hypertension; TAD, thoracic aortic disease; TTE, transthoracic echocardiography; VUS, variant of uncertain significance. a mainly by TTE, but also consider CMR or CCT if the aortic root/ascending aorta are not properly visualized.

Table 3. Revised Ghent Criteria

In the absence of family history
1. Ao ($Z \geq 2$) and ectopia lentis
2. Ao ($Z \geq 2$) and FBN1 mutation
3. Ao ($Z \geq 2$) and systemic features (≥ 7 points), as presented below
4. Ectopia lentis and FBN1 mutation and aortic diameter at the sinuses of Valsalva above indicated Z-score or aortic root dissection
In the presence of family history
5. Ectopia lentis and family history of Marfan's
6. Systemic features (≥ 7 points) and family history of Marfan's
7. Ao ($Z \geq 2$ above 20 years old, ≥ 3 below 20 years) and family history of Marfan's
Scoring of systemic features
Wrist and thumb sign—3 (wrist or thumb sign—1)
Pectus carinatum deformity—2 (pectus excavatum or chest asymmetry—1)
Hindfoot deformity—2 (plain pes planus—1)
Pneumothorax—2
Dural ectasia—2
Protrusio acetabuli—2
Reduced US/LS and increased arm/height and no severe scoliosis—1
Scoliosis or thoracolumbar kyphosis—1
Reduced elbow extension—1
Facial features (3/5)—1 (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)
Skin striae—1
Myopia >3 dioptres—1
Mitral valve prolapse (all types)—1

US: upper segment, LS: lower segment.

Table 4. Imaging in Marfan Syndrome**ACC/AHA 2022 GL on Aortic Disease. Diagnostic and Surveillance Aortic Imaging in Marfan Syndrome****Initial Diagnosis and Surveillance Imaging**

In patients with Marfan syndrome, a TTE at the time of initial diagnosis, to determine the diameters of the aortic root and ascending aorta, and 6 months thereafter, to determine the rate of aortic growth; annual surveillance TTE if the aortic diameters are stable; CT or MRI of the thoracic aorta if the aortic root, ascending aorta, or both are not adequately visualized on TTE. **I C-EO**

In adults with Marfan syndrome, after the initial TTE, CT or MRI of the thoracic aorta to confirm the aortic diameters and assess the remainder of the thoracic aorta. **Ila C-EO**

Imaging After Aortic Root Replacement

In patients with Marfan syndrome who have undergone aortic root replacement, surveillance imaging of the thoracic aorta by MRI (or CT) to evaluate for distal TAD, initially annually and then, if normal in diameter and unchanged after 2 years, every other year. **I C-LD**

In patients with Marfan syndrome who have undergone aortic root replacement, surveillance imaging every 3 to 5 years for potential AAA. **Ila C-LD**

ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for vascular imaging in Marfan syndrome

In patients with MFS, TTE is recommended. ^{70,171,1458,1459}		
• At least annually in patients with an aortic root diameter <45 mm in the absence of additional risk factors ^c	I	C
• At least every 6 months in patients with an aortic root diameter <45 mm in the presence of additional risk factors ^c	I	C
• At least every 6–12 months in patients with an aortic root diameter ≥45 mm in the absence of additional risk factors ^c	I	C
In patients without previous aortic surgery, complete peripheral vascular and thoracoabdominal aorta imaging by CMR or CCT and DUS is recommended at the first evaluation, and subsequently every 3–5 years if stable. ^{70,1455,1459}	I	C
In patients with MFS who have undergone aortic root replacement, surveillance imaging of the thoracic aorta by CMR (or CCT) is recommended at least every 3 years. ^{70,1458}	I	C

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; DUS, duplex ultrasound; MFS, Marfan syndrome; TTE, transthoracic echocardiography.

c Risk factors: aortic root diameter > 40 to ≤45 mm and family history of aortic dissection at small aortic dimensions (i.e. < 50 mm); resistant hypertension (hypertension persisting notwithstanding three or more antihypertensive medications prescribed by a physician with experience in hypertension treatment); and rapid growth of the aorta (annualized growth rate ≥3 mm or more in adults).

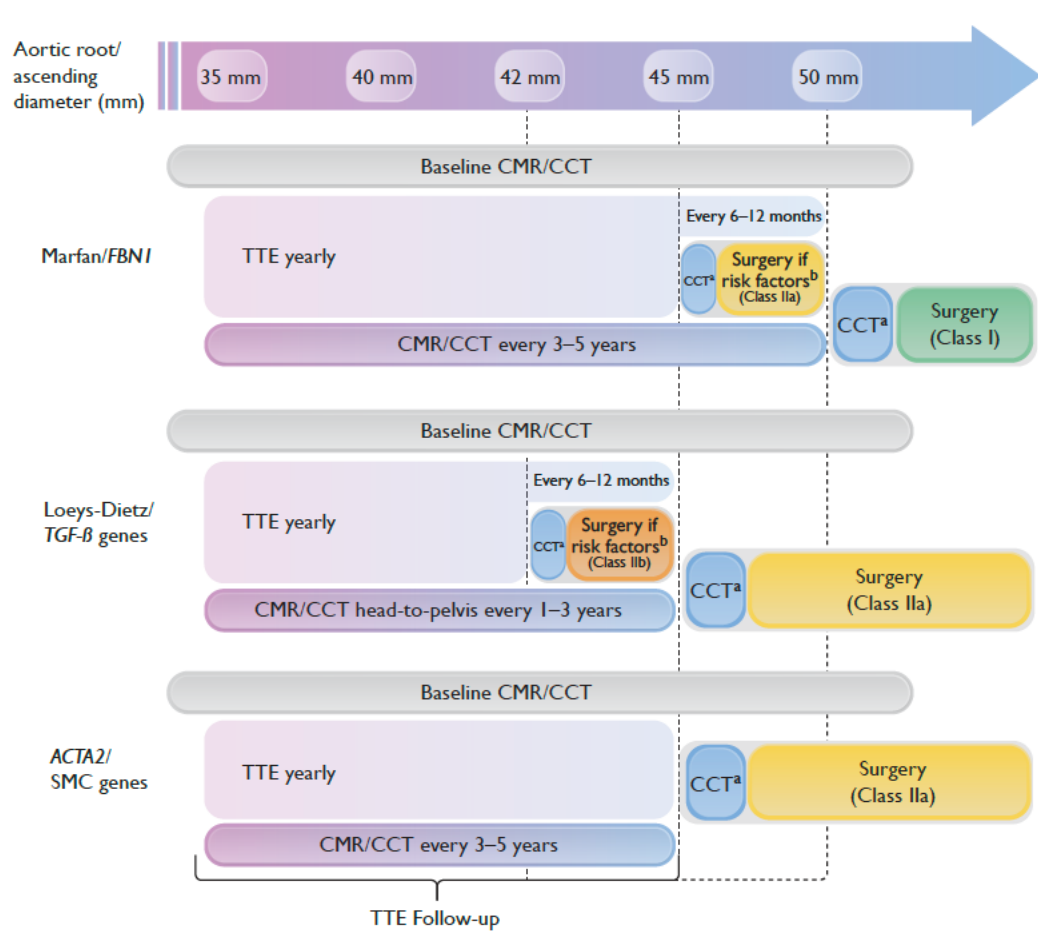


Figure 2. ESC 2024 Guidelines on Aortic and Peripheral Diseases. Algorithm for imaging surveillance in patients with syndromic and non-syndromic heritable thoracic aortic disease. a Pre-surgical CCT.

b See respective tables of recommendations for aortic surgery in Marfan and Loeys-Dietz syndrome

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; DUS, duplex ultrasound; HTAD, heritable thoracic aortic disease; SMC, smooth muscle cell; TTE, transthoracic echocardiography.

Table 5. Medical Therapy of Marfan Syndrome**ACC/AHA 2022 GL on Aortic Disease. Medical Therapy in Marfan Syndrome**

Treatment with either a beta blocker or an ARB, in maximally tolerated doses (unless contraindicated), to reduce the rate of aortic dilation.	I A
Use of both a beta blocker and an ARB, in maximally tolerated doses (unless contraindicated), to reduce the rate of aortic dilation	IIa C-LD

ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for medical treatment in Marfan syndrome

In patients with MFS, treatment with either a BB or an ARB, in maximally tolerated doses (unless contraindicated), is recommended to reduce the rate of aortic dilatation. ^{1461,1462}	I	A
In patients with MFS, the use of both a BB and an ARB, in maximally tolerated doses (unless contraindicated), should be considered to reduce the rate of aortic dilatation. ^{1463,1464}	IIa	A

ARB, angiotensin receptor blocker; BB, beta-blocker; MFS, Marfan syndrome

Table 6. Surgery in Marfan Syndrome**ACC/AHA 2022 GL on Aortic Disease. Marfan Syndrome Interventions: Replacement of the Aortic Root in Patients With Marfan Syndrome**

In aortic root diameter of ≥ 5.0 cm, surgery to replace the aortic root and ascending aorta	I B-NR
In aortic root diameter of ≥ 4.5 cm, and features associated with an increased risk of aortic dissection, surgery to replace the aortic root and ascending aorta, when performed by experienced surgeons in a Multidisciplinary Aortic Team	IIa B-NR
In maximal cross-sectional aortic root area (cm ²) to patient height (m) ratio of ≥ 10 , surgery to replace the aortic root and ascending aorta, when performed by experienced surgeons in a Multidisciplinary Aortic Team.	IIa C-LD
In aortic diameter approaching surgical threshold, who are candidates for valve-sparing root replacement (VSRR) and have a very low surgical risk, surgery to replace the aortic root and ascending aorta, when performed by experienced surgeons in a Multidisciplinary Aortic Team.	IIb C-LD

Features Associated With Increased Risk of Aortic Complications in Marfan Syndrome

Family history of aortic dissection
 Rapid aortic growth (≥ 0.3 cm/y)
 Diffuse aortic root and ascending aortic dilation
 Marked vertebral arterial tortuosity

Replacement of Primary (Nondissected) Aneurysms of the Aortic Arch, Descending, and Abdominal Aorta in Patients With Marfan Syndrome

In a nondissected aneurysm of the aortic arch, descending thoracic aorta, or abdominal aorta of ≥ 5.0 cm, surgical intervention to replace the aneurysmal segment. IIa C-LD

ESC 2024 Guidelines on Aortic and Peripheral Diseases Recommendations for aortic surgery in Marfan syndrome

Surgery is indicated in patients with MFS who have aortic root disease with a maximal aortic sinus diameter ≥ 50 mm. ^{70,172,1466–1468}	I	B
Surgery to replace the aortic root and ascending aorta, using the valve-sparing surgery technique, is recommended in patients with MFS or related HTAD with aortic root dilatation when anatomical features of the valve allow its preservation and the surgeon has specific expertise. ^{70,1466,1469}	I	B
Surgery should be considered in patients with MFS who have an aortic root aneurysm with a maximal aortic sinus diameter ≥ 45 mm and additional risk factors. ^{c,1467,1469}	IIa	C
In patients with MFS and an aneurysm of the ascending aorta, aortic arch, descending thoracic aorta, or abdominal aorta of ≥ 50 mm, surgical replacement of the aneurysmal segment by a surgeon with specific expertise should be considered. ^{1467,1469}	IIa	C

HTAD, heritable thoracic aortic disease; MFS, Marfan syndrome.

c Family history of aortic dissection at small aortic dimensions (i.e. < 50 mm); resistant hypertension (hypertension persisting notwithstanding three or more antihypertensive medications prescribed by a physician with experience in hypertension treatment); and rapid growth of the aorta (annualized growth rate ≥ 3 mm or more in adults).

Table 7. ESC 2024 Guidelines on Aortic and Peripheral Diseases Recommendations for pregnancy in women with Marfan syndrome

It is recommended that all women with MFS: • Have a pre-conception evaluation to address the risks of maternal CV and other complications • Have follow-up in a centre with access to a pregnancy heart and vessel team. ¹⁴⁷³	I	C
It is recommended that couples in which a partner has or is at risk for HTAD be offered pre-conception genetic counselling.	I	C
Imaging of the whole aorta (by CMR/CCT) is recommended prior to pregnancy.	I	C
Follow-up during pregnancy is recommended with a frequency determined by aortic diameter and growth. ^{1337,1474,1475}	I	C
Intake of BBs during pregnancy is recommended. ¹⁴⁷⁶	I	C
Prophylactic aortic root surgery is recommended in women desiring pregnancy with aortic diameters >45 mm. ^{1435,1472}	I	C
Prophylactic aortic root surgery may be considered in women desiring pregnancy with aortic diameters of 40–45 mm. ^{1472,1475,1477}	IIb	C
ARBs are not recommended during pregnancy. ^{1478–1480}	III	B

ARBs, angiotensin receptor blockers; BBs, beta-blockers; CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; CV, cardiovascular; HTAD, heritable thoracic aortic disease; MFS, Marfan syndrome.

Table 8. ESC 2024 Guidelines on Aortic and Peripheral Diseases Recommendations for physical exercise in patients with Marfan syndrome

It is recommended to individualize physical activity in patients with MFS based on aortic diameter, family history of aortic dissection, and pre-existing fitness.	I	C
Regular moderate aerobic exercise with a level of intensity informed by aortic diameter is recommended in most patients with MFS.	I	C
For patients who present with aortic dissection and/or have undergone aortic surgery, post-operative cardiac rehabilitation aiming at improving both physical and mental health should be considered. ^{73,1483,1484,1486}	IIa	B

Table 9. Imaging in Loeys-Dietz Syndrome**ACC/AHA 2022 GL on Aortic Disease. Imaging in Loeys-Dietz Syndrome**

A baseline TTE is recommended to determine the diameters of the aortic root and ascending aorta, and 6 months thereafter to determine the rate of aortic growth; if the aortic diameters are stable, annual surveillance TTE.	I C-EO
In a dilated or dissected aorta and/or arterial branches at baseline, annual surveillance imaging of the affected aorta and arteries with MRI or CT.	I C-EO
A baseline MRI or CT from head to pelvis to evaluate the entire aorta and its branches for aneurysm, dissection, and tortuosity. ¹	I C-EO
In patients without dilation of the aorta distal to the aortic root or ascending aorta and without dilated or dissected arterial branches, surveillance imaging from chest to pelvis with MRI (or CT) every 2 years, but imaging may be more frequent depending on family history.	IIa C-EO
In patients without dilation of the cerebral arteries on initial screening, periodic imaging surveillance for cerebral aneurysms with MRI or CT every 2 to 3 years.	IIa C-EO

ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for imaging follow-up in Loeys-Dietz syndrome

In patients with Loeys-Dietz syndrome, TTE at baseline and subsequently every 6–12 months, depending on aortic diameter and growth, ^c is recommended. ^{70,1390,1488}	I	C
In patients with Loeys-Dietz syndrome, a baseline arterial imaging study from head to pelvis with CMR or CCT and subsequent surveillance with CMR or CCT or DUS every 1–3 years is recommended. ^{70,1488}	I	C

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; DUS, duplex ultrasound; TTE, transthoracic echocardiography.

c More frequent imaging if aortic root/ascending diameter ≥ 42 mm and aortic growth rate ≥ 3 mm per year.

Table 10. Therapy of Loeys-Dietz Syndrome**ACC/AHA 2022 GL on Aortic Disease.****Medical Therapy in Loeys-Dietz Syndrome**

Treatment with a beta blocker or an ARB (unless contraindicated), or both, in maximally tolerated doses. IIa C-EO

Replacement of the Aorta in Patients With Loeys-Dietz Syndrome

In patients with aortic dilation, the surgical threshold for prophylactic aortic root and ascending aortic replacement should be informed by the specific genetic variant, aortic diameter, aortic growth rate, extra-aortic features, family history, patient age and sex, and physician and patient preferences. I C-LD

In Loeys-Dietz syndrome attributable to a pathogenic variant in *TGFBR1*, *TGFBR2*, or *SMAD3*, surgery to replace the intact aortic arch, descending aorta, or abdominal aorta at a diameter of ≥ 4.5 cm, with the specific genetic variant, patient age, aortic growth rate, family history, presence of high-risk features, and surgical risk informing the decision. IIb C-EO

Surgical Thresholds for Prophylactic Aortic Root and Ascending Aortic Replacement in Loeys-Dietz Syndrome Based on Genetic Variant

<i>Genetic Variant</i>	<i>Presence of High-Risk Features*</i>	<i>Aortic Diameter (cm)</i>	<i>COR</i>
<i>TGFBR1</i>	No	≥ 4.5	I C-LD
<i>TGFBR2</i>	No	≥ 4.5	I C-LD
<i>TGFBR1</i>	Yes	≥ 4.0	IIb C-EO
<i>TGFBR2</i>	Yes	≥ 4.0	IIa C-LD
<i>SMAD3</i>	—	$\geq 4.5^{\dagger}$	IIa C-EO
<i>TGFB2^{\ddagger}</i>	—	$\geq 4.5^{\dagger}$	IIb C-EO
<i>TGFB3</i>	—	$\geq 5.0^{\dagger}$	IIb C-EO

*Aortic surgery may be recommended at smaller aortic diameters in Loeys-Dietz syndrome attributable to *TGFBR1* and *TGFBR2* pathogenic variants when there are features that associate with a higher risk of aortic dissection, including: certain specific pathogenic variants; women with *TGFBR2* and small body size; severe extra-aortic features (ie, craniosynostosis, cleft palate, hypertelorism, bifid uvula, marked arterial tortuosity, widened scars, and translucent skin); family history of aortic dissection (especially at young age or relatively small aortic diameter); and aortic growth rate >0.3 cm/y.

† Family history, age, and aortic growth rate also inform surgical thresholds.

‡ Pathogenic variants in the *TGFB2* gene are different than variants in the *TGFBR2* gene.

ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for aortic root surgery in Loeys-Dietz syndrome

Aortic root replacement should be considered for patients with Loeys-Dietz syndrome if the aortic root diameter exceeds 45 mm. ^{1388,1390,1489–1492}	IIa	C
It may be considered to adjust the threshold for surgery according to the underlying gene, taking associated risk features ^c into account. ¹³⁹¹	IIb	C

cHigh-risk features include certain specific pathogenic variants; women with *TGFBR2* variants and small body size; severe extra-aortic features; family history of aortic dissection (especially at young age or relatively small aortic diameter); and aortic growth rate ≥ 3 mm per year.

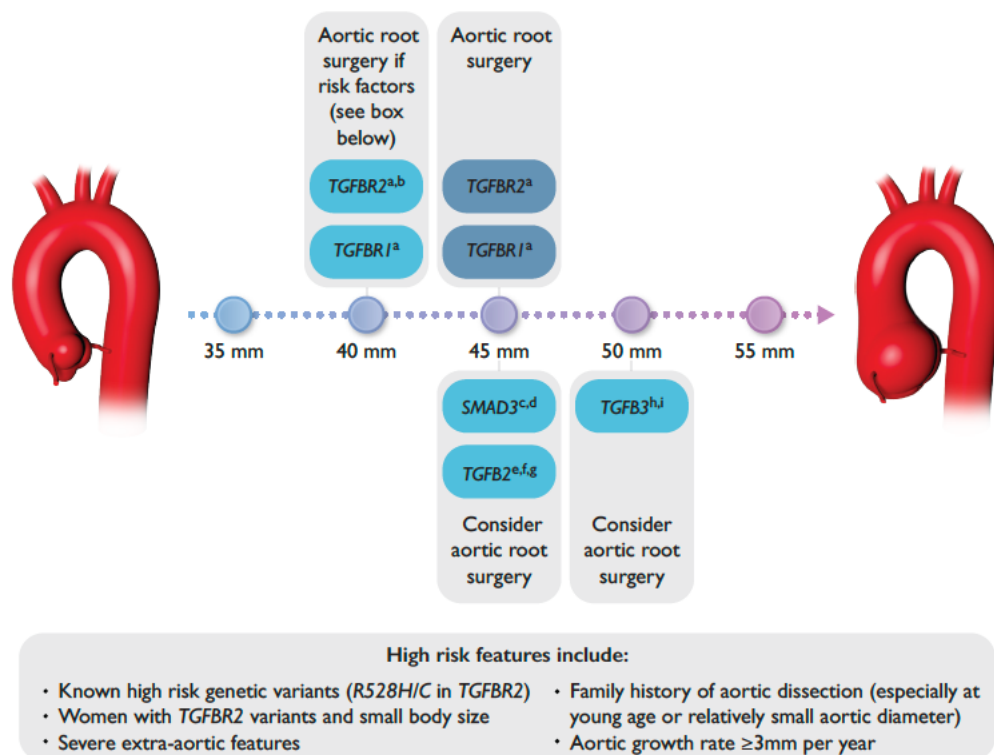


Figure 3. ESC 2024 Guidelines on Aortic and Peripheral Diseases. Suggested thresholds for prophylactic aortic root/ascending replacement in Loeys-Dietz syndrome.

Table 11. ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for medical treatment in patients with vascular Ehlers-Danlos syndrome

In patients with vEDS, regular vascular surveillance of the aorta and peripheral arteries by DUS, CCT, or CMR is recommended. ^{1439,1443}	I	C
Treatment with celiprolol should be considered in patients with vEDS. ^{1441,1444,1445}	IIa	B

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; DUS, duplex ultrasound; vEDS, vascular Ehlers-Danlos syndrome.

Table 12. ACC/AHA 2022 GL on Aortic Disease. Surgical Thresholds for Prophylactic Aortic Root and Ascending Aortic Replacement in Nonsyndromic Heritable Thoracic Aortic Disease Based on the Genetic Variant and Additional Risk Factors for Aortic Dissection

Genetic Variant	Risk Factors	Aortic Diameter (cm)	COR
ACTA2	No	≥ 4.5	IIa C-LD
ACTA2	Yes†	≥ 4.2	IIb C-EO
PRKG1	No	≥ 4.2	IIb C-LD
PRKG1	Yes†	≥ 4.0 †	IIb C-EO

*Patient has risk factors for aortic dissection (family history of type A aortic dissection with minimal aortic enlargement, aortic growth rate ≥ 0.3 cm/y) or significant valve disease requiring surgery.

†Earlier surgery may be considered in patients with a family history of type A aortic dissection in the setting of no or minimal aortic dilation, aortic growth rate ≥ 0.3 cm/y, or at the patient's request.

Table 13. ACC/AHA 2022 GL on Aortic Disease. Surgical Considerations for Nonsyndromic Heritable TAA and No Identified Genetic Cause

In asymptomatic patients with aneurysms of the aortic root or ascending aorta with nonsyndromic heritable thoracic aortic disease (nsHTAD) and no identified genetic cause, determining the timing of surgical repair requires shared decision-making and is informed by known aortic diameters at the time of aortic dissection, TAA repair, or both in affected family members. **I C-LD**

In asymptomatic patients with aneurysms of the aortic root or ascending aorta with nsHTAD and no identified genetic cause but no information on aortic diameters at the time of dissection or aneurysm repair in affected family members and who have no high-risk features for adverse aortic events, repair the aorta when the maximal diameter reaches ≥ 5.0 cm. **I C-LD**

In patients with aneurysms of the aortic root or ascending aorta with nsHTAD and no identified genetic cause and a maximal aortic diameter of ≥ 4.5 cm, who have high-risk features for adverse aortic events, or who are undergoing cardiac surgery for other indications, aortic repair when performed by experienced surgeons in a Multidisciplinary Aortic Team. **IIa C-LD**

Table 14. ESC 2024 Guidelines on Aortic and Peripheral Diseases. Recommendations for imaging and surgery in *ACTA2*-related heritable thoracic aortic disease

Annual monitoring of the aortic root/ascending aorta with TTE to evaluate aortic root/ascending aorta enlargement is recommended. ¹⁴⁹⁸	I	C
Imaging of the aorta with CMR/CCT every 3–5 years is recommended. ¹⁴⁹⁸	I	C
Prophylactic aortic root surgery should be considered with an aortic diameter ≥ 45 mm, or lower in cases with other risk factors. ^{c,1499}	IIa	C

CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; TTE, transthoracic echocardiography.

c Risk factors for aortic dissection: family history of dissection with no or minimal dilatation or young age; rapid growth ≥ 3 mm per year.