

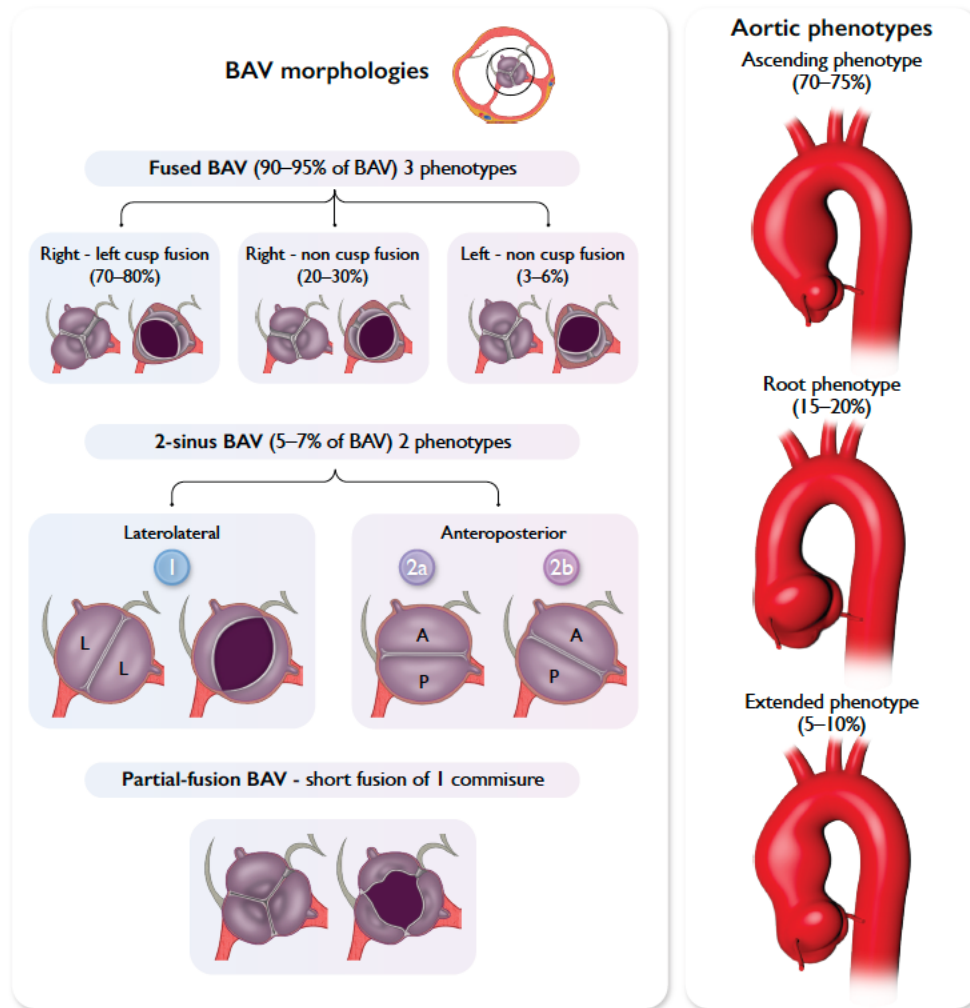


Figure 1. 2024 ESC Guidelines for the management of peripheral arterial and aortic diseases. Bicuspid aortic valve, valvulo-aortopathy nomenclature.

A, anterior; BAV, bicuspid aortic valve; L, lateral; P, posterior. Although preferential associations exist, each of the three valve types—‘fused BAV’, ‘2-sinus BAV’, and ‘partial-fusion BAV’—can be variably associated with dilatation predominantly located at the sinuses of Valsalva (‘root phenotype’, 15%–20%) or at the tubular (supra-coronary) tract (‘ascending phenotype’, 70%–75%). A minor proportion of patients present with equal dilatation of the sinusal and tubular segments or ascending dilatation extending into the proximal arch (‘extended phenotype’, 5%–10%)

Table 1. ESC 2020 GL on ACHD. Diagnostic criteria for degree of aortic stenosis severity

	Mild AS	Moderate AS	Severe AS
$V_{\max}$ (m/s) ^a	2.6–2.9	3.0–3.9	≥ 4.0
Mean gradient (mmHg) ^a	<20	20–39	≥ 40
AVA (cm ²)	>1.5	1.0–1.5	<1.0
AVAi (cm ² /m ² BSA)	>0.85	0.60–0.85	<0.60
LVOT velocity/aortic valve velocity	>0.50	0.25–0.50	<0.25

AS = aortic stenosis; AVA = aortic valve area; AVAi = indexed aortic valve area; BSA = body surface area; LVOT = left ventricular outflow tract; $V_{\max}$ = maximum Doppler velocity.

^aAt normal transvalvular flow.

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for Congenital Diseases of the Aortic Valve

Diagnostic	
Adults with a BAV should be screened for the presence of COA and aortic aneurysm to obtain early diagnosis and determine a management plan.	1 B-NR
Adults with a BAV who have aortic diameter >4 cm at the level of the sinuses or the ascending aorta, or who have risk factors for aortic dissection,* should undergo lifelong surveillance for progression of thoracic aortic disease to improve early detection of aortic growth.	1 B-NR
In first-degree relatives of adults with a BAV, 1-time echocardiographic screening for the presence of a congenitally abnormal aortic valve or aortic aneurysm is reasonable to obtain early diagnosis	2a B-NR

Congenital Diseases of the Aortic Valve: Imaging Testing Intervals for Asymptomatic Patients With Normal LV Function

Frequency of Echocardiography	
Progressive (Stage B)	Aortic stenosis ■ Every 3–5 years (mild severity, V_{max} 2.0–2.9 m/s or mean ΔP <20 mm Hg) ■ Every 1–2 years (moderate severity, V_{max} 3.0–3.9 m/s or mean ΔP 20–39 mm Hg) Aortic regurgitation ■ Every 3–5 years (mild severity) ■ Every 1–2 years (moderate severity)
Severe (Stage C)	Aortic stenosis ■ Every 6–12 months ($V_{max} \geq 4.0$ m/s or mean $\Delta P \geq 40$ mm Hg) Aortic regurgitation ■ Every 6–12 months ■ Dilating left ventricle: more frequently
Aortic dilation >4 cm at the level of the sinuses, the ascending aorta, or both	Periodic echocardiogram, CMR, or CT at intervals determined by: • Degree and rate of progression of aortic dilatation • Family history of aortic dissection • History of coarctation

CMR indicates cardiovascular magnetic resonance; CT, computed tomography; LV, left ventricular; V_{max} , maximum velocity; and ΔP , peak pressure gradient.

Table 3. ACC/AHA 2025 GL on ACHD. Recommendations for Congenital Diseases of the Aortic Valve

Therapeutic	
In adults with a congenitally abnormal aortic valve who meet indication for valve replacement, surgical and transcatheter options should be reviewed by heart valve and ACHD expert teams in a shared decision-making process to determine a management plan.	1 C-LD

In young adults with congenital aortic valve stenosis, a noncalcified valve with no more than mild aortic regurgitation, and an indication for intervention, balloon valvuloplasty may be reasonable to delay the need for aortic valve surgery.

*Risk factors: family history of aortic dissection, aortic growth rate ≥ 0.3 cm/year, COA, “root phenotype” aortopathy.

Table 4. 2024 ESC Guidelines for the management of peripheral arterial and aortic diseases. Recommendations for bicuspid aortic valve-associated aortopathy management

When a BAV is first diagnosed, initial TTE to assess diameters of the aorta at several levels is recommended. ^{1001,1510,1522}	I	B
Surgery for bicuspid aortopathy is recommended when the maximum aortic diameter is ≥ 55 mm. ^{70,172,899,969,1001}	I	B
Surgery for bicuspid aortopathy of the root phenotype ^c is recommended when the maximum aortic diameter is ≥ 50 mm. ^{70,893,981,986,1001,1519,1523}	I	B
CCT or CMR of the entire thoracic aorta is recommended at first diagnosis and when important discrepancies in measurements are found between subsequent TTE controls during surveillance, or when the diameter of the aorta exceeds 45 mm. ^{1001,1510}	I	C
Screening by TTE in FDRs of BAV patients with root phenotype ^c aortopathy and/or isolated aortic regurgitation is recommended. ^{1001,1510,1514}	I	C
Surveillance serial imaging by TTE is recommended in BAV patients with a maximum aortic diameter > 40 mm, either with no indication for surgery or after isolated aortic valve surgery, after 1 year, then if stability is observed, every 2–3 years. ^{70,1001}	I	C
Screening by TTE in FDRs of all BAV patients should be considered. ^{70,1001,1500,1510,1515}	IIa	B
In patients with low surgical risk, surgery for bicuspid aortopathy of ascending phenotype ^d should be considered when the maximum aortic diameter is > 52 mm. ^{153,172,981}	IIa	B
In patients with low surgical risk and ascending phenotype bicuspid aortopathy, surgery should be considered at a maximum diameter ≥ 50 mm if any of the following is the case: ^{70,153,155,981,1001} • Age < 50 years • Shorter stature^e • Ascending aortic length ≥ 11 cm^f • Aortic diameter growth rate ≥ 3 mm per year^g • Family history of acute aortic syndrome • Aortic coarctation • Resistant hypertension^h • Concomitant non-aortic-valve cardiac surgery • Desire for pregnancy	IIa	C
Surgery for bicuspid aortopathy in patients undergoing aortic valve surgery should be considered at a root or ascending diameter ≥ 45 mm. ^{70,172,969}	IIa	C

BAV, bicuspid aortic valve; BP, blood pressure; CCT, cardiovascular computed tomography; CMR, cardiovascular magnetic resonance; CSA/h, cross-sectional area-to-height ratio; FDRs, first-degree relatives; TTE, transthoracic echocardiography.

c Root phenotype = aortic dilatation with sinus diameter $>$ tubular diameter.

d Ascending phenotype = aortic dilatation with tubular diameter $>$ sinus diameter.

- e Patient height between 1.50 and 1.69 m (yielding a CSA/h ratio $>13 \text{ cm}^2/\text{m}$).
- f Curvilinear distance at aortic centreline between the ventriculo-aortic junction and the origin of the innominate artery.
- g In order to ascertain real rapid growth, side-by-side re-evaluation of images obtained with the same modality and technique should be performed.
- h Hypertension persisting notwithstanding three or more antihypertensive medications prescribed by a physician with experience in hypertension treatment, including diuretics.

Table 5. ESC 2020 GL on ACHD. Recommendations for intervention in valvular aortic stenosis

Symptomatic patients with aortic valve stenosis

Intervention in symptomatic patients with severe high-gradient AS (mean gradient $\geq 40 \text{ mmHg}$) I B

Intervention in symptomatic patients with severe low-flow, low-gradient (mean gradient $<40 \text{ mmHg}$) AS with reduced EF and evidence of flow (contractile) reserve excluding pseudosevere AS. I C

Asymptomatic patients with severe aortic valve stenosis

Intervention in asymptomatic patients with severe AS and an abnormal exercise test showing symptoms on exercise clearly related to AS. I C

Intervention in asymptomatic patients with severe AS and systolic LV dysfunction (LVEF $<50\%$) not due to another cause. I C

Intervention in asymptomatic patients with severe AS when they present with a fall in blood pressure below baseline during exercise testing. IIa C

Intervention in asymptomatic patients with normal EF and none of the above-mentioned exercise test abnormalities if the surgical risk is low and one of the following findings is present: IIa C

- Very severe AS defined by a $V_{\text{max}} > 5.5 \text{ m/s}$.
- Severe valve calcification and a rate of V_{max} progression $\geq 0.3 \text{ m/s/year}$.
- Markedly elevated BNP levels (>3 -fold age and sex-corrected normal range) confirmed by repeated measurements without other explanation.
- Severe PH (systolic PAP at rest $>60 \text{ mmHg}$ confirmed by invasive measurement) without other explanation.

Concomitant aortic valve surgery at the time of other cardiac/ascending aorta surgery

Surgery when patients with severe AS undergo surgery of the ascending aorta or of another valve, or CABG. I C

Additional valve replacement in patients with moderate AS undergoing CABG surgery or surgery of the ascending aorta or another valve IIa C

AS = aortic stenosis; BNP = B-type natriuretic peptide; CABG = coronary artery bypass graft; EF = ejection fraction; LV = left ventricle/ventricular; LVEF = left ventricular ejection fraction; PAP = pulmonary artery pressure; PH = pulmonary hypertension; V_{max} = maximum Doppler velocity.

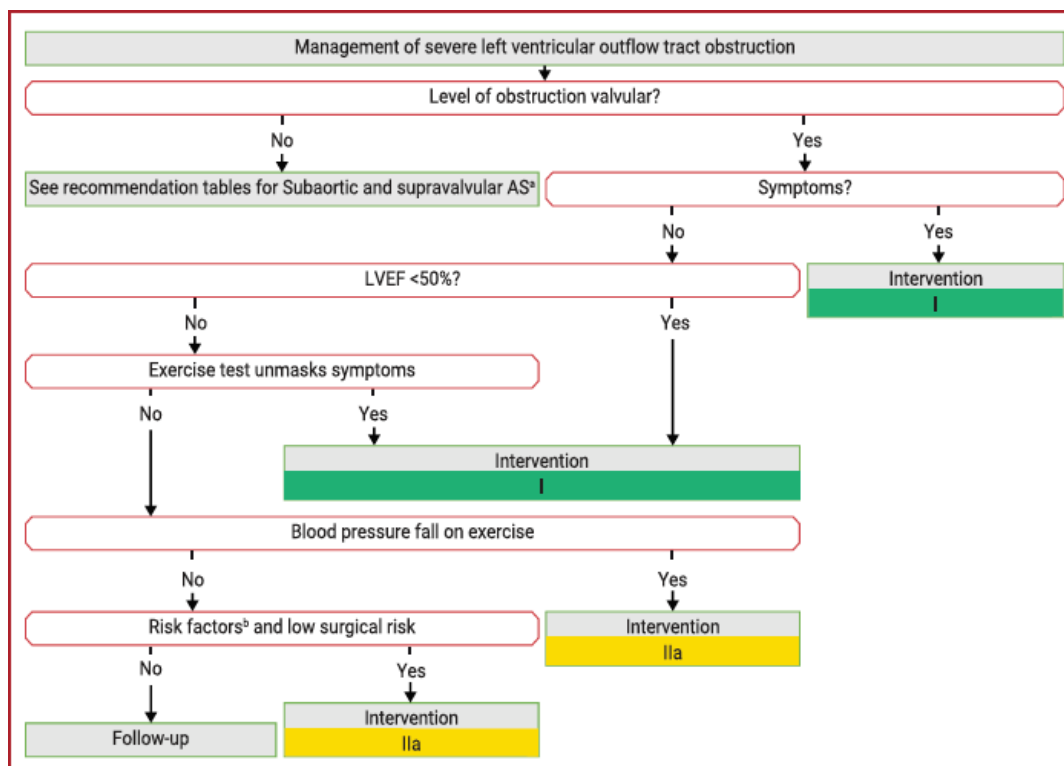


Figure 2. ESC 2020 GL on ACHD. Management of severe left ventricular outflow tract obstruction.

AS = aortic stenosis; LVEF = left ventricular ejection fraction; LVOTO = left ventricular outflow tract obstruction; PAP = pulmonary artery pressure; PH= pulmonary hypertension; SubAS = subaortic stenosis SupraAS = supraaortic stenosis.

a There are fundamental differences in management decisions compared to valvular AS, particularly because a valve substitute with its consequences is generally not required.

b Peak velocity >5.5m/s; severe calcification + peak velocity progression ≥ 0.3 m/s/y; markedly elevated neurohormones (>3-fold age- and sex-corrected normal range); severe PH (systolic PAP >60mmHg without other explanation).

Table 6. ACC/AHA 2025 GL on ACHD. Recommendations for subaortic stenosis**Diagnostic**

Stress testing for adults with LVOT obstruction to determine exercise capacity, symptoms, electrocardiographic changes, or arrhythmias, in the presence of otherwise equivocal indications for intervention IIb-C-LD

Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE	24	24	12	12
Exercise test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.

†6-minute walk test or CPET, depending on the clinical indication.

ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; SubAS, subaortic stenosis; and TTE, transthoracic echocardiogram.

Table 7. ESC 2020 GL on ACHD. Recommendations for intervention in subaortic stenosis

Surgery in symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient > 40 mmHg^a or severe AR, I C

Surgery for asymptomatic patients when one or more of the following findings are present: IIa C

- Mean gradient < 40 mmHg but LVEF $< 50\%$.
- AR is severe and LVESD > 50 mm (or 25 mm/m² BSA) and/or EF $< 50\%$ ^b.
- Mean Doppler gradient is > 40 mmHg^c and marked LVH present.
- Mean Doppler gradient is > 40 mmHg^c and there is a fall in blood pressure below baseline on exercise.

Surgery for asymptomatic patients when one or more of the following findings are present: IIb C

- Mean Doppler gradient is > 40 mmHg,^c LV is normal (EF $> 50\%$ and no LVH), exercise testing is normal, and surgical risk is low.
- Progression of AR is documented and AR becomes more than mild (to prevent further progression).

AR = aortic regurgitation; BSA = body surface area; EF = ejection fraction; ESC = European Society of Cardiology; LV = left ventricle/ventricular; LVEF = left ventricular ejection fraction; LVESD = left ventricular end systolic diameter; LVH = left ventricular hypertrophy.

^a Doppler-derived gradients may overestimate the obstruction and may need confirmation by cardiac catheterization.

^b See 2017 ESC Guidelines on the management of valvular heart disease.

Table 8. ACC/AHA 2025 GL on ACHD. Recommendations for Supravalvar Aortic Stenosis

Diagnostic

In adults with suspected supravalvar aortic stenosis or with Williams syndrome, baseline aortic imaging is recommended to define the anatomy and severity of supravalvar aortic stenosis. 1 C-LD

In adults with Williams syndrome and supravalvar aortic stenosis, baseline coronary imaging is recommended to evaluate for abnormalities such as coronary ostial stenosis. 1 C-LD

Routine Follow-Up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24	24	6-12	3-6
Electrocardiogram	24	24	12	12
Transthoracic echocardiogram†	24	24	12	12

Therapeutic

In adults with supravalvar aortic stenosis who are symptomatic and/or have diminished LV systolic function due to stenosis, surgical repair is recommended to alleviate symptoms and preserve ventricular function. 1 B-NR

Adults with supravalvar aortic stenosis and coronary ostial stenosis with symptoms of ischemia should undergo coronary artery revascularization to alleviate symptoms and preserve ventricular function. 1 C-LD

In adults with asymptomatic severe supravalvar aortic stenosis, normal ventricular size and function, and a normal exercise stress test, surgical repair may be considered to avoid worsening of cardiac function and symptoms. 2b C-EO

Table 9. ESC 2020 GL on ACHD. Recommendations for intervention in supralvalvular aortic stenosis

Surgery in patients with symptoms (spontaneous or on exercise test) and mean Doppler gradient ≥ 40 mmHg	I C
Surgery in patients with mean Doppler gradient <40 mmHg, when one or more of the following findings are present: - Symptoms attributable to obstruction (exertional dyspnoea, angina, syncope). - LV systolic dysfunction (EF $<50\%$ without other explanation). - Surgery required for significant CAD or valvular disease.	I C
Repair when the surgical risk is low for patients with mean Doppler gradient ≥ 40 mmHg ^a but without symptoms, LV systolic dysfunction, LVH, or abnormal exercise test.	I Ib C
CAD = coronary artery disease; EF = ejection fraction; LV = left ventricle/ventricular; LVH = left ventricular hypertrophy. a Doppler-derived gradients may overestimate the obstruction and may need confirmation by left heart catheterization.	

Table 10. ACC/AHA 2025 GL on ACHD. Recommendations for Cor Triatriatum Sinister

Diagnostic	
In adults with unrepaired cor triatriatum sinister, transesophageal echocardiography and/or crosssectional imaging can be useful to identify the size and location of the membrane orifice and to assess for the presence of associated congenital cardiac defects.	2a B-NR
In adults with repaired cor triatriatum sinister and symptoms compatible with recurrent membrane obstruction, it is reasonable to evaluate for the presence of residual congenital cardiac defects, including pulmonary vein stenosis	2a B-NR
Therapeutic	
In adults with unrepaired cor triatriatum sinister and symptoms attributable to membrane obstruction, surgical resection is recommended to reduce the consequences of left atrial hypertension.	1 B-NR
In adults with unrepaired cor triatriatum sinister and atrial fibrillation or atrial flutter, prior stroke, or left atrial thrombus, chronic anticoagulation is recommended to prevent embolic stroke, whether or not conventional thromboembolic risk factors are present.	1 C-LD
In asymptomatic adults with unrepaired cor triatriatum sinister and severe membrane obstruction, surgical resection can be useful to prevent the sequelae of left atrial hypertension	2a C-LD
In adults with unrepaired cor triatriatum sinister and atrial fibrillation or atrial flutter, a rhythm control strategy can be beneficial to avoid clinical decompensation.	2a C-LD
In highly symptomatic adults with unrepaired cor triatriatum sinister,	2b C-LD

significant membrane obstruction, and elevated operative risk,* catheter-based intervention may be considered as a bridge to surgical resection.

*For example, decompensated heart failure, severe pulmonary hypertension, or pregnancy (see supportive text for details).

Routine Follow-Up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36-60	12-24	6-12	3-6
Electrocardiogram	36-60	12-24	6-12	3-6
Transthoracic echocardiogram	36-60	12-24	12	12

Table 11. ACC/AHA 2025 GL on ACHD. Recommendations for Congenital Mitral Stenosis Including Shone Complex Routine Follow-Up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36-60	12-24	6-12	3-6
Electrocardiogram	36-60	12-24	12	12
Transthoracic echocardiogram	36-60	24	12	12

Table 12. ACC/AHA 2022 GL on Aortic Disease. Turner Syndrome.

Diagnostic Testing, Surveillance, and Surgical Intervention for Aortic Dilation in Turner Syndrome

TTE and cardiac MRI at the time of diagnosis to evaluate for BAV, aortic root and ascending aortic dilation, aortic coarctation, and other congenital heart defects. I B-NR

In patients ≥ 15 years old, use ASI (ratio of aortic diameter [cm] to BSA [m²]) to define the degree of aortic dilation and assess the risk of aortic dissection. I B-NR

In patients without risk factors for aortic dissection, surveillance imaging with TTE or MRI to evaluate the aorta every 5 years in children and every 10 years in adults, as well as before planning a pregnancy. I C-LD

In patients with an ASI > 2.3 cm/m², surveillance imaging of the aorta at least annually. I C-EO

In patients with risk factors for aortic dissection, surveillance aortic imaging at an interval depending on the aortic diameter, ASI, and aortic growth rate. I C-EO

In patients ≥ 15 years old and have an ASI of ≥ 2.5 cm/m² plus risk factors for aortic dissection, surgical intervention to replace the aortic root, ascending aorta, or both. IIa C-LD

In those without risk factors for aortic dissection, surgical intervention to replace the aortic root, ascending aorta, or both.

I Ib C-EO

Risk Factors for Aortic Dissection in Patients With Turner Syndrome

Aortic coarctation

Aortic dilation

Bicuspid aortic valve

Hypertension

Table 13. ESC 2024 Guidelines on Aortic and Peripheral Diseases

Recommendations for imaging in women with Turner syndrome

To take the smaller body size of women (≥ 15 years) with TS into account, the use of the ascending ASI (ratio of aortic diameter [mm] to BSA [m^2]), AHI (ratio of aortic diameter [mm] to height [m]), or aortic z-score is recommended to define the degree of aortic dilatation and assess the risk of aortic dissection. ^{153,1417,1421,1423,1428,1429}	I	C
It is recommended to define imaging and clinical surveillance intervals according to the estimated risk for dissection, based on the ascending ASI and concomitant lesions. ^{c,1420,1421}	I	C

AHI, aortic height index; ASI, aortic size index; BSA, body surface area; TS, Turner syndrome.

c Concomitant lesions: hypertension, aortic coarctation, bicuspid aortic valve

Recommendations for aortic surgery in women with Turner syndrome

Elective surgery for aneurysms of the aortic root and/or ascending aorta should be considered in women with TS who are ≥ 15 years of age, have an ascending ASI $> 23 \text{ mm/m}^2$, an AHI $> 23 \text{ mm/m}$, a z-score > 3.5 , and have associated risk factors for aortic dissection ^c or are planning pregnancy. ^{70,1417,1421}	IIa	C
Elective surgery for aneurysms of the aortic root and/or ascending aorta may be considered for women with TS who are ≥ 15 years of age, have an ascending ASI $> 25 \text{ mm/m}^2$, an AHI $> 25 \text{ mm/m}$, a z-score > 4 , and who do not have associated risk factors for aortic dissection. ^{c,70,1417,1421}	IIb	C

AHI, aortic height index (ratio of aortic diameter [mm] to height [m]); ASI, aortic size index (ratio of aortic diameter [mm] to BSA [m^2]); TS, Turner syndrome.

c Bicuspid aortic valve, elongation of the transverse aorta, coarctation of the aorta, and/or uncontrolled hypertension (despite more than three classes of antihypertensive drugs).

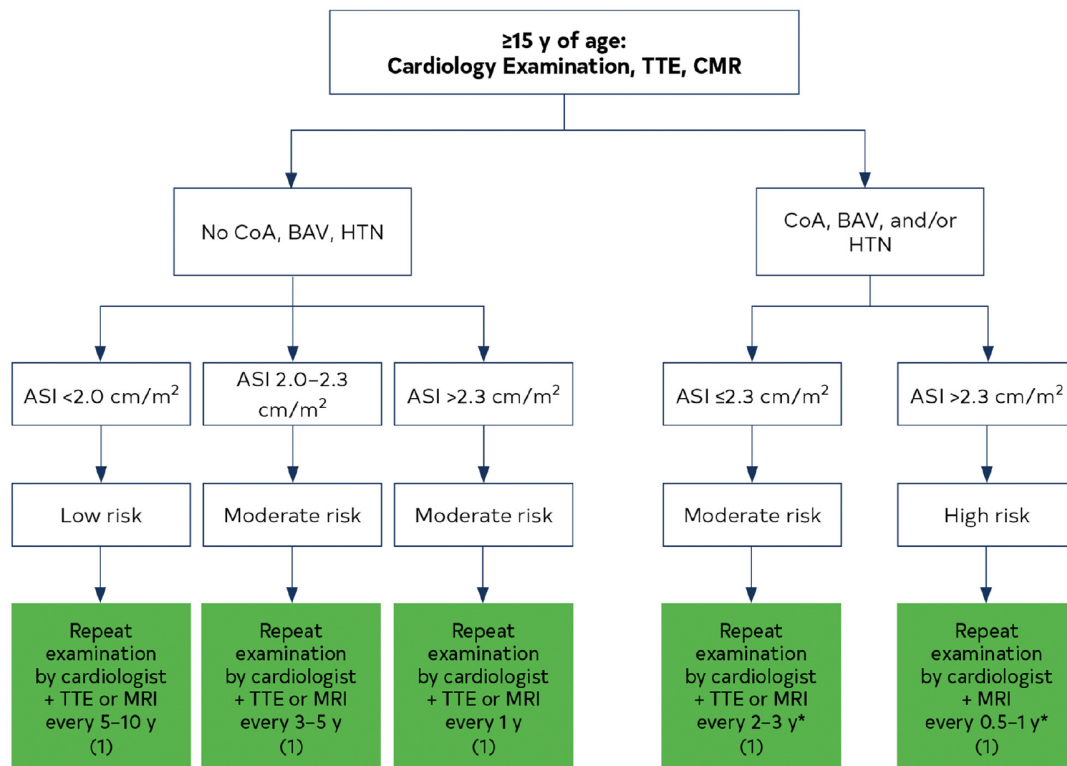


Figure 3. ACC/AHA 2022 GL on Aortic Disease. Suggested Aortic Monitoring Protocol for Girls and Women With Turner Syndrome Who Are ≥ 15 Years of Age.

*Surveillance frequency may vary depending on disease severity (ie, aortic valve dysfunction, severity of coarctation obstruction, hypertension, and left ventricular hypertrophy).

ASI indicates aortic size index; BAV, bicuspid aortic valve; CoA, coarctation of the aorta; HTN, hypertension; MRI, magnetic resonance imaging; and TTE, transthoracic echocardiography.

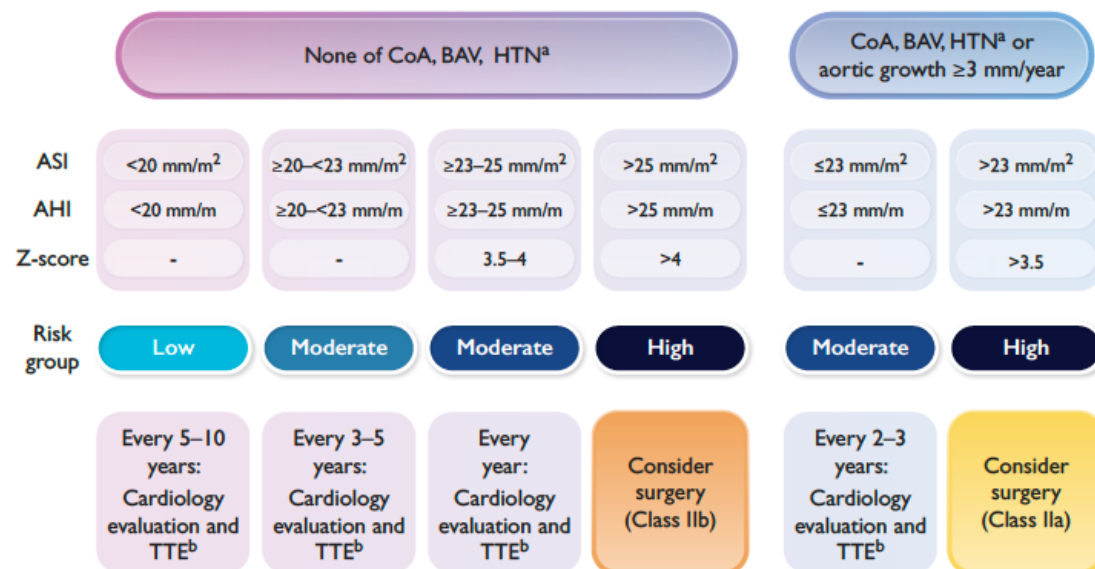


Figure 4. ESC 2024 Guidelines on Aortic and Peripheral Diseases Algorithm for surveillance in women (≥ 15 years) with Turner syndrome.

AHI, aortic height index (ratio of aortic diameter [mm] to height [m]); ASI, aortic size index (ratio of aortic diameter [mm] to BSA [m^2]); BAV, bicuspid aortic valve; BSA, body surface area; CCT, Cardiovascular Computed Tomography; CMR, cardiovascular magnetic resonance; CoA, coarctation of the aorta; HTN, arterial hypertension; TTE, transthoracic echocardiography.

^a HTN: arterial hypertension, not under control despite more than three classes of antihypertensive drugs.

^b CMR (preferably) or CCT if inadequate visualization of the ascending aorta.