



LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

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1- VALVULAR AORTIC STENOSIS

The most common cause for congenital valvular aortic stenosis is BAV . Up to 80% of patients with a BAV will developed ascending aortic dilatation .

Patients frequently remain asymptomatic fo rmany years.Progression of stenosis varies and depends on initial severity, degree of calcification, age, and atherosclerotic risk factors.

Prognosis is good and sudden death is rare in asymptomatic patients with good exercise tolerance, even when stenosis is severe. Once symptoms (angina pectoris, dyspnoea, or syncope) occur, the prognosis deteriorates rapidly. Inpatients with BAV,cardiac mortality has been reported to be 0.3% per patient-year of follow-up , the frequency of aortic dissection 0.03%, and endocarditis 0.3%. Dilated aortic sinuses and/or ascending aorta have been found in 45% of patientsafter9yearsoffollow-up

Table 12 Diagnostic criteria for degree of aortic stenosis severity¹⁷³

| | 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.

Recommendations for intervention in valvular aortic stenosis

| Recommendations | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Symptomatic patients with aortic valve stenosis | | |
| In symptomatic patients with severe high-gradient AS (mean gradient ≥ 40 mmHg), intervention is recommended. ^{25,171,174,175} | I | B |
| Intervention is indicated in symptomatic patients with severe low-flow, low-gradient (mean gradient < 40 mmHg) AS with reduced EF and evidence of flow (contractile) reserve excluding pseudosevere AS. | I | C |
| Asymptomatic patients with severe aortic valve stenosis | | |
| Intervention is indicated in asymptomatic patients with severe AS and an abnormal exercise test showing symptoms on exercise clearly related to AS. | I | C |
| Intervention is indicated in asymptomatic patients with severe AS and systolic LV dysfunction (LVEF $< 50\%$) not due to another cause. | I | C |

| | | |
|--|-------------------|-----------------|
| <p>Intervention should be considered in asymptomatic patients with severe AS when they present with a fall in blood pressure below baseline during exercise testing.</p> | <p>Ila</p> | <p>C</p> |
| <p>Intervention should be considered 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:</p> <ul style="list-style-type: none"> ● Very severe AS defined by a $V_{max} > 5.5$ m/s. ● Severe valve calcification and a rate of V_{max} progression ≥ 0.3 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 mmHg confirmed by invasive measurement) without other explanation. | <p>Ila</p> | <p>C</p> |
| <p>Concomitant aortic valve surgery at the time of other cardiac/ascending aorta surgery</p> | | |
| <p>Surgery is recommended when patients with severe AS undergo surgery of the ascending aorta or of another valve, or CABG.</p> | <p>I</p> | <p>C</p> |
| <p>Patients with moderate AS undergoing CABG surgery or surgery of the ascending aorta or another valve should be considered for addi-</p> | <p>Ila</p> | <p>C</p> |

MEDICAL THERAPY :

Symptomatic patients require urgent surgery. Medical treatment for AS-related heart failure is reserved for non-operable patients. Neither statin treatment, nor any other medical treatment, has so far been shown to retard progression of AS.

SURGICAL/CATHETER INTERVENTIONAL TREATMENT:

In selected adolescents and young adults with non-calcified valves, balloon valvuloplasty may be considered. This may be the case in haemodynamically unstable patients as a bridge to surgery or to delay valve replacement in women with anatomically suitable valves and desire of pregnancy .

In patients with calcified valves, the treatment of choice is valve replacement. Mechanical valves are more durable than biological valves or homografts but require lifelong anticoagulation.

The Ross procedure (two-valve operation) has been suggested for patients of childbearing age and for those wanting to avoid anticoagulation.

Transcatheter aortic valve implantation currently has no place in the treatment of congenital AS, except in very rare cases with high surgical risk, when technically feasible.

Follow-up recommendations:

Lifelong and regular follow-up is required, and the intervals depend upon the degree of stenosis severity. It is also necessary after valve intervention at minimum yearly intervals.

Additional considerations :

- Pregnancy: contraindicated in severe symptomatic AS.
- Exercise/sports: patients with severe symptomatic and asymptomatic AS, and those with moderate stenosis due to BAV and dilated aorta, should avoid isometric exercise and high-intensity sports. In mild and moderate AS, more intensive physical activity is allowed.

Management of severe left ventricular outflow tract obstruction

Level of obstruction valvular?

No

Yes

See recommendation tables for Subaortic and supra-ventricular AS^a

Symptoms?

No

Yes

LVEF <50%?

Intervention

No

Yes

Exercise test unmask symptoms

Intervention

No

Yes

Blood pressure fall on exercise

Yes

No

Risk factors^b and low surgical risk

Intervention
IIa

No

Yes

Follow-up

Intervention
IIa

IIa

SUPRAVALVULAR AORTIC STENOSIS :

Supravalvular AS (SupraAS) may occur as a characteristic feature of Williams – Bueren Syndrome .

SupraAS may also be encountered in the setting of familial homozygous hypercholesterolaemia.

It may be associated with aortic valve abnormalities ,hypoplasia of the entire aorta, involvement of coronary ostia, or stenosis of major branches of the aorta or PAs.

The majority of patients present in childhood with symptoms of either outflow obstruction or myocardial ischaemia. While progression of SupraAS is rare in adulthood ,adults remain at risk for cardiac complication .

Sudden death occurs rarely, but it is more common in SupraAS with Williams– Bueren Syndrome with diffuse peripheral PA stenosis ,or with CAD, particularly related to an aesthetic procedures.

Surgical/catheter interventional treatment:

Surgery is the primary treatment: the operative mortality rate for fibrous diaphragm and hourglass deformity is <5%. Since the coronary arteries are under high pressure, surgery might be considered earlier than in patients with valvular AS, particularly when no valve substitute is required.

Following operative repair, the survival rate has been reported to be 80–85 % at 20 years . AR may be present in 25 % of patients , but usually it is not progressive after surgical relief of SupraAS .

Recommendations for intervention in supraaortic stenosis

| Recommendations | Class ^a | Level ^b |
|--|--------------------|--------------------|
| In patients with symptoms (spontaneous or on exercise test) and mean Doppler gradient ≥ 40 mmHg, surgery is recommended. | I | C |
| In patients with mean Doppler gradient < 40 mmHg, surgery is recommended when one or more of the following findings are present: <ul style="list-style-type: none">• 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 |
| Patients with mean Doppler gradient ≥ 40 mmHg ^c – but without symptoms, LV systolic dysfunction, LVH, or abnormal exercise test – may be considered for repair when the surgical risk is low. | IIb | C |

Subaortic stenosis :

Subaortic stenosis(SubAS) associated with aortic valve disease, VSD, AVSD, or Shone complex (multilevel left heart obstruction). It may also develop after repair of these lesions. It is caused by a fibrous ridge/ring in the LVOT proximal to the aortic valve or as a fibromuscular narrowing. SubAS has to be differentiated from obstructive hypertrophic cardiomyopathy .

The clinical course is highly variable. The presence of associated CHD, particularly a VSD, is related with SubAS progression; age seems not to play a role in disease.

Clinical findings mainly include a systolic ejection murmur at the left sternal border without systolic ejection click. A diastolic murmur refers to AR. • Echocardiography visualizes LVOT anatomy, associated aortic valve abnormality, degree of AR, LV function, LVH, and associated lesions. With Doppler echocardiography, the severity of the subvalvular obstruction is determined, but Doppler-derived gradients may overestimate the obstruction and may require confirmation by cardiac catheterization.

Surgical/catheter interventional treatment:

Surgical treatment is the only effective intervention and involves a complete resection of the fibrous ridge/ring and parts of the muscular base along the left septal surface.

Fibromuscular or tunnel-type SubAS requires more extensive resection or a Konno procedure. Surgical results are good, but restenosis may occur. In patients with low surgical risk and morphologically well suited to repair, the threshold for intervention is lower than in aortic valve stenosis since no valve implant is required. In the case of moderate or severe AR, the aortic valve must be repaired or replaced at the time of surgery.

Recommendations for intervention in subaortic stenosis

| Recommendations | Class ^a | Level ^b |
|--|--------------------|--------------------|
| <p>In symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥ 40 mmHg^c or severe AR, surgery is recommended.</p> | I | C |
| <p>Asymptomatic patients should be considered for surgery when one or more of the following findings are present:</p> <ul style="list-style-type: none"> ● Mean gradient < 40 mmHg but LVEF $< 50\%$. ● AR is severe and LVESD > 50 mm (or 25 mm/m² BSA) and/or EF $< 50\%$^d. ● 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. | IIa | C |
| <p>Asymptomatic patients may be considered for surgery when one or more of the following findings are present:</p> <ul style="list-style-type: none"> ● 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). | IIb | C |

THANK YOU