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# FRAMEWORK FOR GAUGING VALVE DISEASE SEVERITY

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**PRACTICE GUIDELINE**

# 2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines

*Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Anesthesiologists, and Society of Thoracic Surgeons*

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# CRITERIA OF THE SEVERITY OF VALVE DISEASE

1. Presence/absence of symptoms
2. Severity of VHD
3. Response of the left/right ventricle to the volume/pressure overload
4. Effects on the pulmonary circulation
5. Change in heart rhythm

# STAGES OF PROGRESSION OF VHD

| Stage | Definition | Description |
|---|---|---|
| A | At risk | Patients with risk factors for development of VHD |
| B | Progressive | Patients with progressive VHD (mild-to-moderate severity and asymptomatic) |
| C | Asymptomatic severe | Asymptomatic patients who have the criteria for severe VHD:<br>C1: Asymptomatic patients with severe VHD in whom the left or right ventricle remains compensated<br>C2: Asymptomatic patients with severe VHD, with decompensation of the left or right ventricle |
| D | Symptomatic severe | Patients who have developed symptoms as a result of VHD |

# STAGES OF VALVULAR AS

| Stage | Definition | Valve Anatomy | Valve Hemodynamics | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| A | At risk of AS | • Bicuspid aortic valve or other congenital valve anomaly)<br>• Aortic valve sclerosis | • Aortic $V_{max}$ <2 m/s | • None | • None |
| B | Progressive AS | • Mild-to-moderate leaflet calcification of a bicuspid or trileaflet valve with some reduction in systolic motion or<br>• Rheumatic valve changes with commissural fusion | • Mild AS: Aortic $V_{max}$ 2.0–2.9 m/s or mean $\Delta P$ <20 mm Hg<br>• Moderate AS: Aortic $V_{max}$ 3.0–3.9 m/s or mean $\Delta P$ 20–39 mm Hg | • Early LV diastolic dysfunction may be present<br>• Normal LVEF | • None |
| C | Asymptomatic severe AS | | | | |
| C1 | Asymptomatic severe AS | • Severe leaflet calcification or congenital stenosis with severely reduced leaflet opening | • Aortic $V_{max}$ ≥4 m/s or mean $\Delta P$ ≥40 mm Hg<br>• AVA typically is ≤1.0 $cm^2$ (or AVAi ≤0.6 $cm^2/m^2$)<br>• Very severe AS is an aortic $V_{max}$ ≥5 m/s or mean $\Delta P$ ≥60 mm Hg | • LV diastolic dysfunction<br>• Mild LV hypertrophy<br>• Normal LVEF | • None: Exercise testing is reasonable to confirm symptom status |
| C2 | Asymptomatic severe AS with LV dysfunction | • Severe leaflet calcification or congenital stenosis with severely reduced leaflet opening | • Aortic $V_{max}$ ≥4 m/s or mean $\Delta P$ ≥40 mm Hg<br>• AVA typically ≤1.0 $cm^2$ (or AVAi ≤0.6 $cm^2/m^2$) | • LVEF <50% | • None |

# STAGES OF VALVULAR AS

| Stage | Definition | Valve Anatomy | Valve Hemodynamics | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| D | Symptomatic severe AS | | | | |
| D1 | Symptomatic severe high-gradient AS | • Severe leaflet calcification or congenital stenosis with severely reduced leaflet opening | • Aortic $V_{max}$ >4 m/s or mean ΔP ≥40 mm Hg<br>• AVA typically ≤1.0 $cm^2$ (or AVAi ≤0.6 $cm^2/m^2$) but may be larger with mixed AS/AR | • LV diastolic dysfunction<br>• LV hypertrophy<br>• Pulmonary hypertension may be present | • Exertional dyspnea or decreased exercise tolerance<br>• Exertional angina<br>• Exertional syncope or presyncope |
| D2 | Symptomatic severe low-flow/low-gradient AS with reduced LVEF | • Severe leaflet calcification with severely reduced leaflet motion | • AVA ≤1.0 $cm^2$ with resting aortic $V_{max}$ <4 m/s or mean ΔP <40 mm Hg<br>• Dobutamine stress echocardiography shows AVA ≤1.0 $cm^2$ with $V_{max}$ ≥4 m/s at any flow rate | • LV diastolic dysfunction<br>• LV hypertrophy<br>• LVEF <50% | • HF<br>• Angina<br>• Syncope or presyncope |
| D3 | Symptomatic severe low-gradient AS with normal LVEF or paradoxical low-flow severe AS | • Severe leaflet calcification with severely reduced leaflet motion | • AVA ≤1.0 $cm^2$ with aortic $V_{max}$ <4 m/s or mean ΔP <40 mm Hg<br>• Indexed AVA <0.6 $cm^2/m^2$ and<br>• Stroke volume index <35 $mL/m^2$<br>• Measured when patient is normotensive (systolic BP <140 mm Hg) | • Increased LV relative wall thickness<br>• Small LV chamber with low stroke volume<br>• Restrictive diastolic filling<br>• LVEF ≥50% | • HF<br>• Angina<br>• Syncope or presyncope |

# STAGES OF CHRONIC AR

| Stage | Definition | Valve Anatomy | Valve Hemodynamics | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| A | At risk of AR | • Bicuspid aortic valve (or other congenital valve anomaly)<br>• Aortic valve sclerosis<br>• Diseases of the aortic sinuses or ascending aorta<br>• History of rheumatic fever or known rheumatic heart disease<br>• IE | • AR severity: none or trace | • None | • None |
| B | Progressive AR | • Mild-to-moderate calcification of a trileaflet valve bicuspid aortic valve (or other congenital valve anomaly)<br>• Dilated aortic sinuses<br>• Rheumatic valve changes<br>• Previous IE | • Mild AR:<br>◦ Jet width <25% of LVOT;<br>◦ Vena contracta <0.3 cm;<br>◦ RVol <30 mL/beat;<br>◦ RF <30%;<br>◦ ERO <0.10 $cm^2$;<br>◦ Angiography grade 1+<br>• Moderate AR:<br>◦ Jet width 25%–64% of LVOT;<br>◦ Vena contracta 0.3–0.6 cm;<br>◦ RVol 30–59 mL/beat;<br>◦ RF 30%–49%;<br>◦ ERO 0.10–0.29 $cm^2$;<br>◦ Angiography grade 2+ | • Normal LV systolic function<br>• Normal LV volume or mild LV dilation | • None |

# STAGES OF CHRONIC AR

| Stage | Definition | Valve Anatomy | Valve Hemodynamics | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| C | Asymptomatic severe AR | • Calcific aortic valve disease<br>• Bicuspid valve (or other congenital abnormality)<br>• Dilated aortic sinuses or ascending aorta<br>• Rheumatic valve changes<br>• IE with abnormal leaflet closure or perforation | • Severe AR:<br>○ Jet width ≥65% of LVOT;<br>○ Vena contracta >0.6 cm;<br>○ Holodiastolic flow reversal in the proximal abdominal aorta<br>○ RVol ≥60 mL/beat;<br>○ RF ≥50%;<br>○ ERO ≥0.3 $cm^2$;<br>○ Angiography grade 3+ to 4+;<br>○ In addition, diagnosis of chronic severe AR requires evidence of LV dilation | C1: Normal LVEF (≥50%) and mild-to-moderate LV dilation (LVESD ≤50 mm)<br>C2: Abnormal LV systolic function with depressed LVEF (<50%) or severe LV dilatation (LVESD >50 mm or indexed LVESD >25 mm/$m^2$) | • None; exercise testing is reasonable to confirm symptom status |
| D | Symptomatic severe AR | • Calcific valve disease<br>• Bicuspid valve (or other congenital abnormality)<br>• Dilated aortic sinuses or ascending aorta<br>• Rheumatic valve changes<br>• Previous IE with abnormal leaflet closure or perforation | • Severe AR:<br>○ Doppler jet width ≥65% of LVOT;<br>○ Vena contracta >0.6 cm;<br>○ Holodiastolic flow reversal in the proximal abdominal aorta;<br>○ RVol ≥60 mL/beat;<br>○ RF ≥50%;<br>○ ERO ≥0.3 $cm^2$;<br>○ Angiography grade 3+ to 4+;<br>○ In addition, diagnosis of chronic severe AR requires evidence of LV dilation | • Symptomatic severe AR may occur with normal systolic function (LVEF ≥50%), mild-to-moderate LV dysfunction (LVEF 40%–50%), or severe LV dysfunction (LVEF <40%);<br>• Moderate-to-severe LV dilation is present | • Exertional dyspnea or angina or more severe HF symptoms |

# STAGES OF MS

| Stage | Definition | Valve Anatomy | Valve Hemodynamics | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| A | At risk of MS | • Mild valve doming during diastole | • Normal transmitral flow velocity | None | • None |
| B | Progressive MS | • Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets<br>• Planimetered MVA >1.5 $cm^2$ | • Increased transmitral flow velocities<br>• MVA >1.5 $cm^2$<br>• Diastolic pressure half-time <150 ms | • Mild-to-moderate LA enlargement<br>• Normal pulmonary pressure at rest | • None |
| C | Asymptomatic severe MS | • Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets<br>• Planimetered MVA ≤1.5 $cm^2$<br>• (MVA ≤1.0 $cm^2$ with very severe MS) | • MVA ≤1.5 $cm^2$<br>• (MVA ≤1.0 $cm^2$ with very severe MS)<br>• Diastolic pressure half-time ≥150 ms<br>• (Diastolic pressure half-time ≥220 ms with very severe MS) | • Severe LA enlargement<br>• Elevated PASP >30 mm Hg | • None |
| D | Symptomatic severe MS | • Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets<br>• Planimetered MVA ≤1.5 $cm^2$ | • MVA ≤1.5 $cm^2$<br>• (MVA ≤1.0 $cm^2$ with very severe MS)<br>• Diastolic pressure half-time ≥150 ms<br>• (Diastolic pressure half-time ≥220 ms with very severe MS) | • Severe LA enlargement<br>• Elevated PASP >30 mm Hg | • Decreased exercise tolerance<br>• Exertional dyspnea |

# STAGES OF PRIMARY MR

| Grade | Definition | Valve Anatomy | Valve Hemodynamics* | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| A | At risk of MR | • Mild mitral valve prolapse with normal coaptation<br>• Mild valve thickening and leaflet restriction | • No MR jet or small central jet area <20% LA on Doppler<br>• Small vena contracta <0.3 cm | • None | • None |
| B | Progressive MR | • Severe mitral valve prolapse with normal coaptation<br>• Rheumatic valve changes with leaflet restriction and loss of central coaptation<br>• Prior IE | • Central jet MR 20%–40% LA or late systolic eccentric jet MR<br>• Vena contracta <0.7 cm<br>• Regurgitant volume <60 mL<br>• Regurgitant fraction <50%<br>• ERO <0.40 $cm^2$<br>• Angiographic grade 1–2+ | • Mild LA enlargement<br>• No LV enlargement<br>• Normal pulmonary pressure | • None |
| C | Asymptomatic severe MR | • Severe mitral valve prolapse with loss of coaptation or flail leaflet<br>• Rheumatic valve changes with leaflet restriction and loss of central coaptation<br>• Prior IE<br>• Thickening of leaflets with radiation heart disease | • Central jet MR >40% LA or holosystolic eccentric jet MR<br>• Vena contracta ≥0.7 cm<br>• Regurgitant volume ≥60 mL<br>• Regurgitant fraction ≥50%<br>• ERO ≥0.40 $cm^2$<br>• Angiographic grade 3–4+ | • Moderate or severe LA enlargement<br>• LV enlargement<br>• Pulmonary hypertension may be present at rest or with exercise<br>C1: LVEF >60% and LVESD <40 mm<br>C2: LVEF ≤60% and LVESD ≥40 mm | • None |
| D | Symptomatic severe MR | • Severe mitral valve prolapse with loss of coaptation or flail leaflet<br>• Rheumatic valve changes with leaflet restriction and loss of central coaptation<br>• Prior IE<br>• Thickening of leaflets with radiation heart disease | • Central jet MR >40% LA or holosystolic eccentric jet MR<br>• Vena contracta ≥0.7 cm<br>• Regurgitant volume ≥60 mL<br>• Regurgitant fraction ≥50%<br>• ERO ≥0.40 $cm^2$<br>• Angiographic grade 3–4+ | • Moderate or severe LA enlargement<br>• LV enlargement<br>• Pulmonary hypertension present | • Decreased exercise tolerance<br>• Exertional dyspnea |

# STAGES OF SECONDARY MR

| Grade | Definition | Valve Anatomy | Valve Hemodynamics* | Associated Cardiac Findings | Symptoms |
|---|---|---|---|---|---|
| A | At risk of MR | • Normal valve leaflets, chords, and annulus in a patient with coronary disease or cardiomyopathy | • No MR jet or small central jet area <20% LA on Doppler<br>• Small vena contracta <0.30 cm | • Normal or mildly dilated LV size with fixed (infarction) or inducible (ischemia) regional wall motion abnormalities<br>• Primary myocardial disease with LV dilation and systolic dysfunction | • Symptoms due to coronary ischemia or HF may be present that respond to revascularization and appropriate medical therapy |
| B | Progressive MR | • Regional wall motion abnormalities with mild tethering of mitral leaflet<br>• Annular dilation with mild loss of central coaptation of the mitral leaflets | • ERO <0.20 $cm^2$†<br>• Regurgitant volume <30 mL<br>• Regurgitant fraction <50% | • Regional wall motion abnormalities with reduced LV systolic function<br>• LV dilation and systolic dysfunction due to primary myocardial disease | • Symptoms due to coronary ischemia or HF may be present that respond to revascularization and appropriate medical therapy |
| C | Asymptomatic severe MR | • Regional wall motion abnormalities and/or LV dilation with severe tethering of mitral leaflet<br>• Annular dilation with severe loss of central coaptation of the mitral leaflets | • ERO ≥0.20 $cm^2$†<br>• Regurgitant volume ≥30 mL<br>• Regurgitant fraction ≥50% | • Regional wall motion abnormalities with reduced LV systolic function<br>• LV dilation and systolic dysfunction due to primary myocardial disease | • Symptoms due to coronary ischemia or HF may be present that respond to revascularization and appropriate medical therapy |
| D | Symptomatic severe MR | • Regional wall motion abnormalities and/or LV dilation with severe tethering of mitral leaflet<br>• Annular dilation with severe loss of central coaptation of the mitral leaflets | • ERO ≥0.20 $cm^2$†<br>• Regurgitant volume ≥30 mL<br>• Regurgitant fraction ≥50% | • Regional wall motion abnormalities with reduced LV systolic function<br>• LV dilation and systolic dysfunction due to primary myocardial disease | • HF symptoms due to MR persist even after revascularization and optimization of medical therapy<br>• Decreased exercise tolerance<br>• Exertional dyspnea |

# STAGES OF TR

| Stage | Definition | Valve Anatomy | Valve Hemodynamics* | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| A | At risk of TR | **Primary**<br>• Mild rheumatic change<br>• Mild prolapse<br>• Other (e.g., IE with vegetation, early carcinoid deposition, radiation)<br>• Intra-annular RV pacemaker or ICD lead<br>• Postcardiac transplant (biopsy related)<br>**Functional**<br>• Normal<br>• Early annular dilation | • No or trace TR | • None | • None or in relation to other left heart or pulmonary/pulmonary vascular disease |
| B | Progressive TR | **Primary**<br>• Progressive leaflet deterioration/destruction<br>• Moderate-to-severe prolapse, limited chordal rupture<br>**Functional**<br>• Early annular dilation<br>• Moderate leaflet tethering | **Mild TR**<br>• Central jet area $<5.0\ cm^2$<br>• Vena contracta width not defined<br>• CW jet density and contour: soft and parabolic<br>• Hepatic vein flow: systolic dominance<br>**Moderate TR**<br>• Central jet area $5–10\ cm^2$<br>• Vena contracta width not defined but $<0.70$ cm<br>• CW jet density and contour: dense, variable contour<br>• Hepatic vein flow: systolic blunting | **Mild TR**<br>• RV/RA/IVC size normal<br>**Moderate TR**<br>• No RV enlargement<br>• No or mild RA enlargement<br>• No or mild IVC enlargement with normal respirophasic variation<br>• Normal RA pressure | • None or in relation to other left heart or pulmonary/pulmonary vascular disease |

# STAGES OF TR

| Stage | Definition | Valve Anatomy | Valve Hemodynamics* | Hemodynamic Consequences | Symptoms |
|---|---|---|---|---|---|
| C | Asymptomatic severe TR | Primary<br>• Flail or grossly distorted leaflets<br>Functional<br>• Severe annular dilation (>40 mm or 21 mm/m$^2$)<br>• Marked leaflet tethering | • Central jet area >10.0 cm$^2$<br>• Vena contracta width >0.7 cm<br>• CW jet density and contour: dense, triangular with early peak<br>• Hepatic vein flow: systolic reversal | • RV/RA/IVC dilated with decreased IVC respirophasic variation<br>• Elevated RA pressure with "c-V" wave<br>• Diastolic interventricular septal flattening may be present | • None, or in relation to other left heart or pulmonary/pulmonary vascular disease |
| D | Symptomatic severe TR | Primary<br>• Flail or grossly distorted leaflets<br>Functional<br>• Severe annular dilation (>40 mm or >21 mm/m$^2$)<br>• Marked leaflet tethering | • Central jet area >10.0 cm$^2$<br>• Vena contracta width >0.70 cm<br>• CW jet density and contour: dense, triangular with early peak<br>• Hepatic vein flow: systolic reversal | • RV/RA/IVC dilated with decreased IVC respirophasic variation<br>• Elevated RA pressure with "c-V" wave<br>• Diastolic interventricular septal flattening<br>• Reduced RV systolic function in late phase | • Fatigue, palpitations, dyspnea, abdominal bloating, anorexia, edema |

# ASYMPTOMATIC VHD WITH NORMAL LVEF

## Frequency of echocardiogram

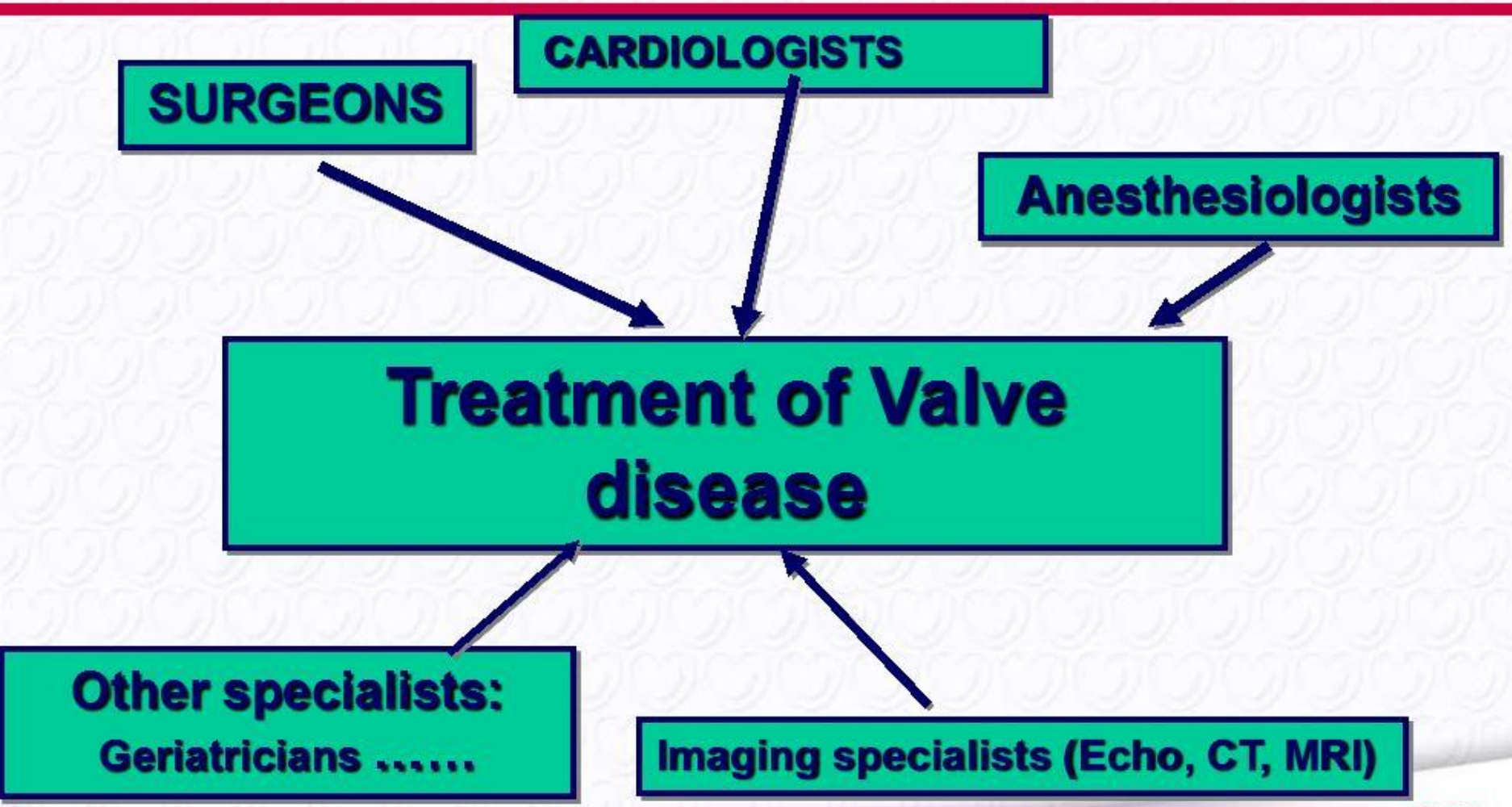
| Stage | Valve Lesion | | | |
|---|---|---|---|---|
| Stage | Aortic Stenosis* | Aortic Regurgitation | Mitral Stenosis | Mitral Regurgitation |
| Progressive (stage B) | Every 3–5 y (mild severity $V_{max}$ 2.0–2.9 m/s)<br>Every 1–2 y (moderate severity $V_{max}$ 3.0–3.9 m/s) | Every 3–5 y (mild severity)<br>Every 1–2 y (moderate severity) | Every 3–5 y (MVA >1.5 $cm^2$) | Every 3–5 y (mild severity)<br>Every 1–2 y (moderate severity) |
| Severe (stage C) | Every 6–12 mo ($V_{max}$ ≥4 m/s) | Every 6–12 mo<br>Dilating LV: more frequently | Every 1–2 y (MVA 1.0–1.5 $cm^2$)<br>Once every year (MVA <1.0 $cm^2$) | Every 6–12 mo<br>Dilating LV: more frequently |

# Essential questions in the evaluation of a patient for valvular intervention

- Is valvular heart disease severe?
- Does the patient have symptoms?
- Are symptoms related to valvular disease?
- What are patient life expectancy and expected quality of life?
- Do the expected benefits of intervention (versus spontaneous outcome) outweigh its risks?
- What are the patient's wishes?
- Are local resources optimal for planned intervention?

European Heart Journal 2012 - doi:10.1093/eurheartj/ehs109 & European Journal of Cardio-Thoracic Surgery 2012 - doi:10.1093/ejcts/ezs455).

# The « Heart Team »



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