Aortic valve disease during pregnancy

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NYHA and other classifications of cardiovascular disability

Class	NYHA functional classification [1]	Canadian Cardiovascular Society functional classification [2]	Specific activity scale [3]
I	Patients with cardiac disease but without resulting limitations of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain.	Ordinary physical activity, such as walking and climbing stairs, does not cause angina. Angina with strenuous or rapid prolonged exertion at work or recreation.	Patients can perform to completion any activity requiring ≥7 metabolic equivalents (ie, can carry 24 lb up 8 steps; do outdoor work [shovel snow, spade soil]; do recreational activities [skiing, basketball, squash, handball, jog/walk 5 mph]).
II	Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain.	Slight limitation of ordinary activity. Walking or climbing stairs rapidly, walking uphill, walking or stair-climbing after meals, in cold, in wind, or when under emotional stress, or only during the few hours after awakening. Walking more than 2 blocks on the level and climbing more than 1 flight of ordinary stairs at a normal pace and in normal conditions.	Patients can perform to completion any activity requiring ≥5 metabolic equivalents (eg, have sexual intercourse without stopping, garden, rake, weed, roller skate, dance foxtrot, walk at 4 mph on level ground) but cannot and do not perform to completion activities requiring ≥7 metabolic equivalents.
III	Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less-than-ordinary physical activity causes fatigue, palpitation, dyspnea, or anginal pain.	Marked limitation of ordinary physical activity. Walking 1 to 2 blocks on the level and climbing 1 flight in normal conditions.	Patients can perform to completion any activity requiring ≥2 metabolic equivalents (eg, shower without stopping, strip and make bed, clean windows, walk 2.5 mph, bowl, play golf, dress without stopping) but cannot and do not perform to completion any activities requiring >5 metabolic equivalents.
IV	Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort is increased.	Inability to carry on any physical activity without discomfort. Anginal syndrome may be present at rest.	Patients cannot or do not perform to completion activities requiring >2 metabolic equivalents. Cannot carry out activities listed above (specific activity scale III).

Aortic stenosis —

The most common cause of aortic stenosis among women of childbearing age is congenital bicuspid aortic valve disease

Aortic stenosis due to rheumatic heart disease is an uncommon cause and is generally accompanied by mitral stenosis.

Pregnancy is usually well tolerated in patients with mild and moderate aortic stenosis, but patients with severe aortic stenosis tolerate the hemodynamic alterations of pregnancy poorly.

HF and atrial and ventricular arrhythmias are the common pregnancy complications reported in women with aortic stenosis and these occur most often in women with severe aortic stenosis

In women with fixed outflow tract obstruction, the inability to accommodate to the increased cardiac output and stroke volume can increase the LV end diastolic pressure, precipitating pulmonary edema or arrhythmias.

Maternal deaths and aortic dissection in women with bicuspid valve aortopathy are rare

In a review of publications between 1985 and 2019 on pregnancy outcomes in women with aortic stenosis in medium or higher Human Development Index countries,

rates of pulmonary edema and new or recurrent arrhythmias were 9 and 4 percent for women with severe aortic stenosis and 8 and 2 percent for women with moderate aortic stenosis.

Maternal mortality for women with severe aortic stenosis was 2 percent

High rates of obstetric and fetal/neonatal complications have been reported in women with significant aortic stenosis

Aortic stenosis is staged according to	valve anatomy,
	stenosis severity,
	LV geometry and function,
	and symptoms

Patients with aortic stenosis should receive preconception counseling by a cardiologist experienced in managing VHD during pregnancy.

Asymptomatic women with severe aortic stenosis require careful preconception assessment

blood pressure response to exercise can be helpful in risk stratification

All women with symptomatic aortic stenosis should have a valve intervention prior to pregnancy.

If symptoms develop during pregnancy, restricted activities are a reasonable first step.

Pulmonary edema should be treated with diuretics.

Rarely, women with persistent symptoms despite medical therapy may need to be considered for a valve intervention (either valvuloplasty or aortic valve replacement) during pregnancy

Stages of valvular aortic stenosis

Stage	Definition	Valve anatomy	Valve hemodynamics	Hemodynamic consequences	Symptoms
A	**At risk of AS**	▪ Bicuspid aortic valve (or other congenital valve anomaly) ▪ 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 ▪ Rheumatic valve changes with commissural fusion	▪ Mild AS: Aortic V_{max} 2.0 to 2.9 m/s or mean ΔP <20 mmHg ▪ Moderate AS: Aortic V_{max} 3.0 to 3.9 m/s or mean ΔP 20 to 39 mmHg	▪ Early LV diastolic dysfunction may be present ▪ 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 ΔP ≥40 mmHg ▪ AVA typically ≤1.0 cm^2 (or AVAi ≤0.6 cm^2/m^2) ▪ Very severe AS is an aortic V_{max} ≥5 m/s or mean ΔP ≥60 mmHg	▪ LV diastolic dysfunction ▪ Mild LV hypertrophy ▪ 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 ΔP ≥40 mmHg ▪ AVA typically ≤1.0 cm^2 (or AVAi ≤0.6 cm^2/m^2)	▪ LVEF <50%	▪ None

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 mmHg ▪ 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 ▪ LV hypertrophy ▪ Pulmonary hypertension may be present	▪ Exertional dyspnea or decreased exercise tolerance ▪ Exertional angina ▪ 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 mmHg ▪ Dobutamine stress echocardiography shows AVA ≤1.0 cm^2 with V_{max} ≥4 m/s at any flow rate	▪ LV diastolic dysfunction ▪ LV hypertrophy ▪ LVEF <50%	▪ HF ▪ Angina ▪ 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 mmHg ▪ Indexed AVA ≤0.6 cm^2/m^2 ▪ Stroke volume index <35 mL/m^2 ▪ Measured when patient is normotensive (systolic BP <140 mmHg)	▪ Increased LV relative wall thickness ▪ Small LV chamber with low stroke volume ▪ Restrictive diastolic filling ▪ LVEF ≥50%	▪ HF ▪ Angina ▪ Syncope or presyncope

Bicuspid aortic valve

- is the most prevalent congenital cardiac abnormality, affecting 1 percent of the population with a 2 to 3:1 male predominance
- A bicuspid aortic valve may be functionally normal or it may be stenotic and/or regurgitant.
- Bicuspid aortic valve is the most common cause of congenital aortic stenosis (AS) and the most common cause of isolated aortic regurgitation (AR).
- Dilation of the ascending aorta is likely to coexist independent of valvular function

DETECTION AND EVALUATION —

- typically detected by echocardiography performed to evaluate the murmur of AS and/or aortic regurgitation (AR) or an ejection click.
- An isolated, functionally normal bicuspid aortic valve is likely to go unrecognized in women of childbearing age because auscultatory signs are inconspicuous or unrecognized
- Rarely, a bicuspid aortic valve is first identified during pregnancy or postpartum after a patient presents with dissection of the thoracic aorta

- ✓ All patients with bicuspid aortic valve should undergo preconception transthoracic echocardiography,
  - ✓ including assessment for aortic valve dysfunction (AS and AR)
  - ✓ and ascending aortic dilation.
- ✓ Evaluation should also include cross-sectional imaging (eg, computed tomography or magnetic resonance imaging) to detect aortic coarctation and assessment for features of Turner syndrome

COUNSELING —

- Women with bicuspid aortic valve should be counseled prior to and during pregnancy regarding potential risks and their management.
- Potential risks that should be discussed include heritable congenital heart disease, aortic enlargement or dissection, complications of AS and/or aortic regurgitation (AR), and endocarditis.

HERITABILITY —

Bicuspid aortic valve occurs sporadically and as an autosomal dominant inherited disorder with variable penetrance.

an isolated lesion or associated with other congenital cardiovascular defects or aortopathy syndromes

Large family studies have found the prevalence of bicuspid aortic valve to be approximately 9 to 10 percent in first-degree relatives of the individual with bicuspid aortic valve

Fetal ultrasound is recommended when the mother has bicuspid aortic valve since fetal congenital heart disease has been reported in approximately **4 percent** of women with congenital AS

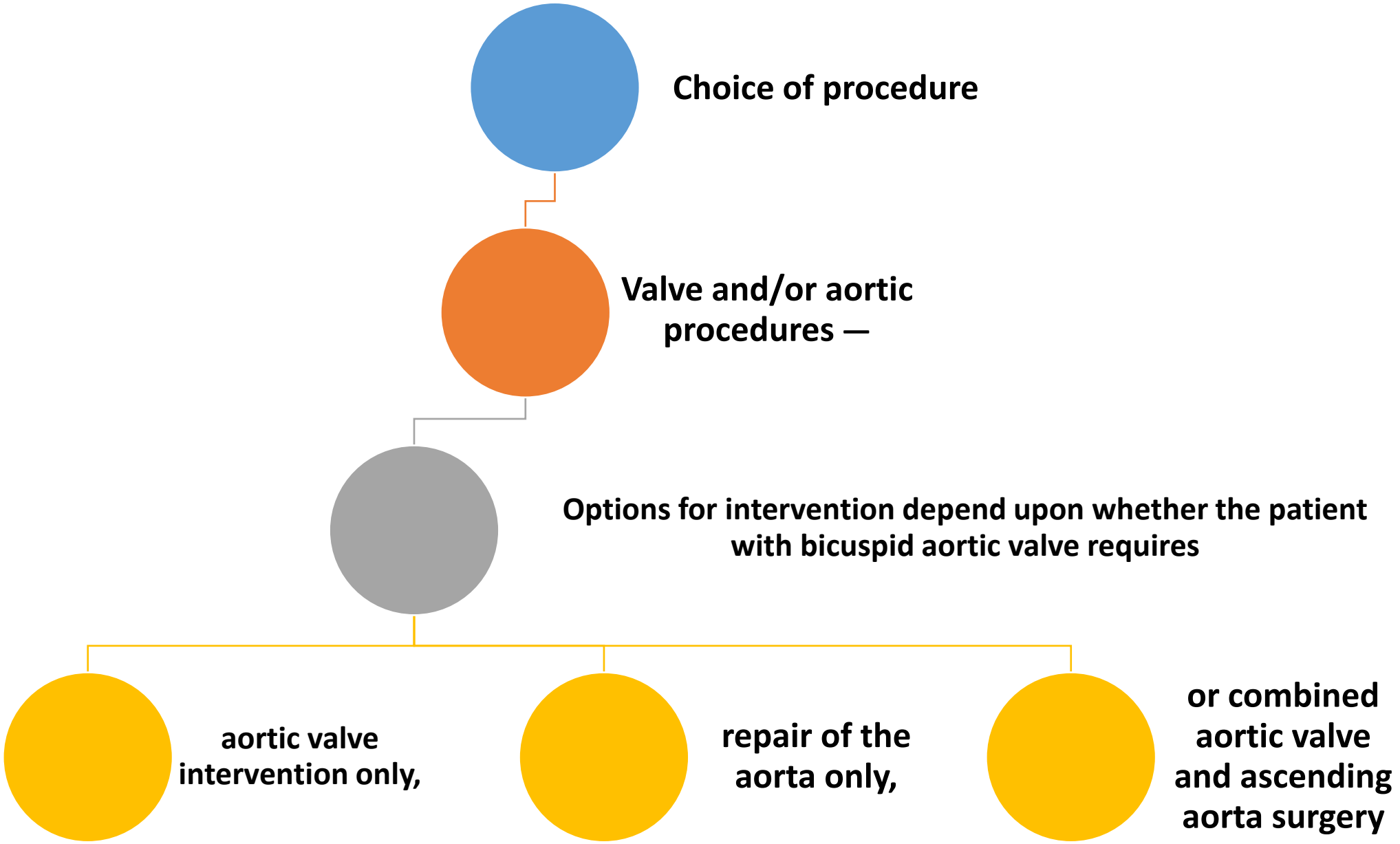
Bicuspid aortic valve is frequently associated with congenital abnormalities of the aorta

(eg, coarctation of the aorta and patent ductus arteriosus)

SURGERY PRIOR TO OR DURING PREGNANCY

Timing —

- **For patients with an indication for valve or aortic surgery, preconception (or postpartum) surgery is preferred to surgery during pregnancy because of the fetal risks of surgery.**
- **Valve or aortic surgery should be avoided during pregnancy if possible.**
- **The maternal risks are similar to those in nonpregnant women, but cardiopulmonary bypass during pregnancy incurs risks for the fetus.**
- **Indications for intervention for AS, aortic regurgitation (AR), and aortic dilation prior to pregnancy are aimed at reducing the risk of maternal and fetal complications during and following pregnancy, including the risk of requiring surgery during pregnancy.**


Choice of procedure
Valve and/or aortic procedures —
Options for intervention depend upon whether the patient with bicuspid aortic valve requires
aortic valve intervention only,
repair of the aorta only,
or combined aortic valve and ascending aorta surgery

Choice of valve procedures —

Most patients with bicuspid aortic valve with an indication for valve intervention require valve replacement	Alternatives to aortic valve replacement are available for only selected patients with severe bicuspid AS or AR	For AS, only selected patients are candidates for balloon or surgical aortic valvotomy	For AR, only selected patients are candidates for aortic valve repair

Aortic valve replacement for severe AS is performed by

- surgical aortic valve replacement (SAVR)
- or transcatheter aortic valve implantation (TAVI)
- although data on TAVI are more limited in patients with bicuspid aortic valve than in patients with tricuspid valve

Aortic valve replacement for severe AR associated with bicuspid aortic valve is generally performed by SAVR

- The choices for SAVR
  - are a bioprosthetic valve,
  - a homograft valve,
  - a pulmonary autograft (Ross procedure),
  - and a mechanical valve
  - TAVI involves implantation of a bioprosthetic valve

The long-term utility of implantation of a bioprosthetic aortic valve in women of childbearing age is not well established

Although anticoagulation is generally not required for bioprosthetic valves (after the first three to six months), there is a high rate of structural failure, which may exceed 60 percent at 15 years in younger patients.

The failure rate may not be as high for newer-generation bioprosthetic valves.

Another option that avoids a need for anticoagulation is a Ross procedure that replaces the aortic valve with the patient's own pulmonary valve (pulmonary autograft) and replaces the pulmonary valve with a homograft

advantage :
anticoagulation, with its attendant risks (including fetal risks during pregnancy from vitamin K antagonist is not required

cardiovascular complications are common after the Ross procedure,

reoperation/reintervention is often required involving one or both valves

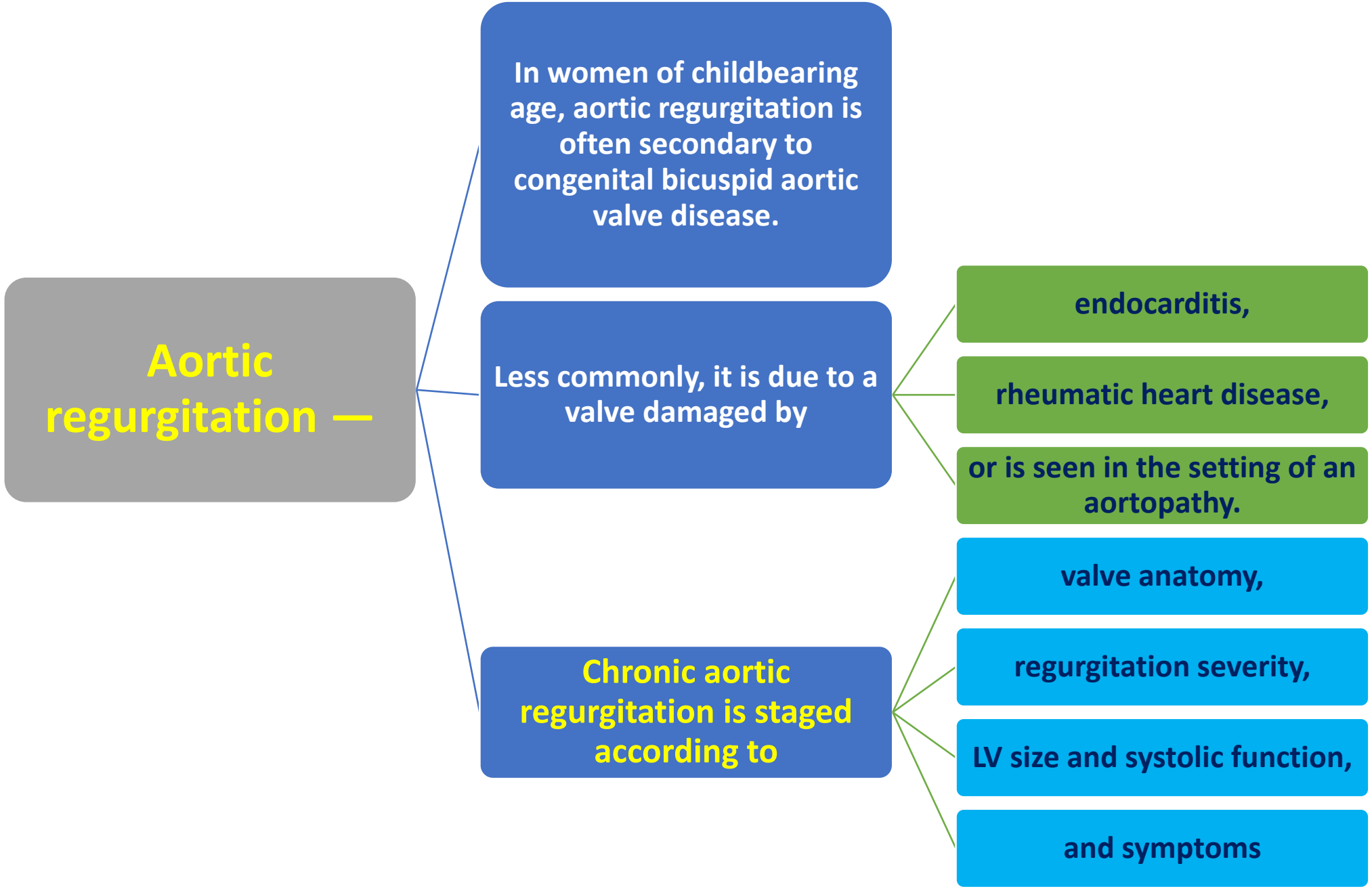
The least satisfactory option for valve replacement in anticipation of pregnancy is a mechanical prosthesis, which is generally durable but requires life-long anticoagulation.

VKA (eg, warfarin) is considered the safest form of anticoagulation for the mother with a mechanical heart valve.

However, warfarin crosses the placenta, and the fetus is anticoagulated.

Warfarin therapy is associated with a dose-dependent risk of embryopathy (particularly during the first trimester), and later in gestation it increases the risk of fetal bleeding and fetal loss.

Heparin does not cross the placenta, and low molecular weight heparin use requires anti-Xa dose adjustment during pregnancy for patients with mechanical valve prostheses and is associated with low fetal risk but higher risk of maternal thromboembolism and mortality than VKA


Aortic regurgitation —
In women of childbearing age, aortic regurgitation is often secondary to congenital bicuspid aortic valve disease.
Less commonly, it is due to a valve damaged by
endocarditis,
rheumatic heart disease,
or is seen in the setting of an aortopathy.
Chronic aortic regurgitation is staged according to
valve anatomy,
regurgitation severity,
LV size and systolic function,
and symptoms

A study of pregnancy outcomes in women with moderate or severe valve regurgitation reported on 40 pregnancies in women with aortic regurgitation

Most women with aortic regurgitation had bicuspid aortic valves.

Maternal cardiac events occurred in 5 percent of the pregnancies

Women with asymptomatic aortic regurgitation (stage B or C1) and preserved LV systolic function tolerate pregnancy well.

Women with severe aortic regurgitation with symptoms (stage D), LV systolic dysfunction, severe LV dilation (stage C2), or pulmonary hypertension may develop cardiac complications during pregnancy.

Women with severe aortic regurgitation with LV systolic dysfunction (LVEF <30 should be advised to avoid pregnancy

If a woman with severe aortic regurgitation and significant LV systolic dysfunction presents early in pregnancy, we suggest termination of pregnancy followed by corrective surgery before another attempt at pregnancy.

If the mother declines termination, we manage the patient medically and, as recommended by the American College of Cardiology/American Heart Association guidelines, operate only for refractory NYHA class III or IV symptoms

Symptomatic patients can be treated with

- diuretics
- and, if necessary, vasodilators (eg, nifedipine)
- Angiotensin converting enzyme inhibitors and angiotensin II receptor blockers should not be used during pregnancy because of potential harm to the fetus

Cardiac surgery should be avoided during pregnancy if possible.	• **The maternal risks are similar to those in nonpregnant women, but cardiopulmonary bypass during pregnancy poses risks for the fetus** • **It is preferable to delay surgery, if possible, until the fetus is viable; a cesarean delivery can then be performed as part of a combined procedure**
Pregnant women with aortic regurgitation due to Marfan syndrome are at risk for aortic dissection;	• **the highest risk is in those with aortic root dilatation** • **The prophylactic use of beta blockers may minimize aortic dilation during pregnancy**

Stages of chronic aortic regurgitation in adults

Stage	Definition	Valve anatomy	Valve hemodynamics	Hemodynamic consequences	Symptoms
A	At risk of AR	▪ Bicuspid aortic valve (or other congenital valve anomaly) ▪ Aortic valve sclerosis ▪ Diseases of the aortic sinuses or ascending aorta ▪ History of rheumatic fever or known rheumatic heart disease ▪ 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) ▪ Dilated aortic sinuses ▪ Rheumatic valve changes ▪ Previous IE	▪ Mild AR: • Jet width <25% of LVOT; • Vena contracta <0.3 cm; • RVol <30 mL/beat; • RF <30%; • ERO <0.10 cm^2; • Angiography grade 1+ ▪ Moderate AR: • Jet width 25 to 64% of LVOT; • Vena contracta 0.3 to 0.6 cm; • RVol 30 to 59 mL/beat; • RF 30 to 49%; • ERO 0.10 to 0.29 cm^2; • Angiography grade 2+	▪ Normal LV systolic function ▪ Normal LV volume or mild LV dilation	▪ None

C	**Asymptomatic severe AR**	▪ Calcific aortic valve disease ▪ Bicuspid valve (or other *congenital abnormality*) ▪ Dilated aortic sinuses or ascending aorta ▪ Rheumatic valve changes ▪ IE with abnormal leaflet closure or perforation	▪ Severe AR: • Jet width ≥65% of LVOT; • Vena contracta >0.6 cm; • Holodiastolic flow reversal in the proximal abdominal aorta • RVol ≥60 mL/beat; • RF ≥50%; • ERO ≥0.3 cm^2; • Angiography grade 3+ to 4+; • In addition, diagnosis of chronic severe AR requires evidence of LV dilation	▪ C1: Normal LVEF (≥50%) and LVESD ≤50 mm ▪ C2: Abnormal LV systolic function with depressed LVEF (<50%), 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 ▪ Bicuspid valve (or other congenital abnormality) ▪ Dilated aortic sinuses or ascending aorta ▪ Rheumatic valve changes ▪ Previous IE with abnormal leaflet closure or perforation	▪ Severe AR: • Jet width ≥65% of LVOT; • Vena contracta >0.6 cm; • Holodiastolic flow reversal in the proximal abdominal aorta; • RVol ≥60 mL/beat; • RF ≥50%; • ERO ≥0.3 cm^2; • Angiography grade 3+ to 4+; • 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 to 50%), or severe LV dysfunction (LVEF <40%) ▪ Moderate to severe LV dilation is present	▪ Exertional dyspnea or angina or more severe HF symptoms