



Differential Diagnosis of Aortic Regurgitation (AR)

Disease	Notes
Bicuspid aortic valve	Bicuspid valve prevalence is 1%; less common congenital valve abnormalities include unicommissural and quadricuspid valves; may be familial in some cases. Incomplete valve closure or cusp prolapse; some degree of stenosis due to incomplete leaflet opening or superimposed calcific changes is common; associated with aortic root dilation.
Infective endocarditis	Leaflet perforation or destruction, vegetations may be present; commonly associated with fever, chills, systemic findings (embolic phenomena). Tissue destruction most often is the cause of AR, but vegetations can interfere with proper cusp coaptation; most common cause of acute AR.
Rheumatic fever	Fibrous, retracted cusps unable to oppose during diastole; common cause worldwide. The mitral valve is always affected, resulting in mitral stenosis.
Myxomatous valve disease	Diffusely thickened redundant leaflet with prolapse in diastole. Seen in approximately 5% of mitral valve prolapse patients. Usually mild AR.
Calcific degeneration	Focal cuspal fibrous thickening, calcified deposits restrict leaflet motion and coaptation; incidence increases with age. Mitral annular calcification is often present as well; usually causes only mild-moderate AR; rarely requires surgery.
Rheumatoid arthritis	Aortic valve nodules deform leaflet closure. Aortitis resulting in aortic dilation also is seen in patients with rheumatoid arthritis.
Aortic dissection	AR may be central due to aortic dilation or may be due to disruption of the attachment of the cusp to the aortic wall; Dissection of the aortic wall may occur secondary to root dilation associated with hypertension, degenerative changes (cystic medial degeneration), systemic connective tissue disorders (Marfan's and Ehlers-Danlos syndromes), pregnancy, and direct trauma.
Marfan's syndrome	Aortic dissection is the most common cause of death in Marfan's syndrome. Diagnosis should be suspected in young patients with musculoskeletal signs of Marfan's syndrome and chest pain. May be the initial manifestation of disease. Other cardiovascular features include mitral valve prolapse, mitral regurgitation, and dilation of the aortic root.
Hypertension	Dilated ascending aorta/root, aortic valve sclerosis, usually causes only mild to

moderate AR. Other characteristics of hypertensive heart disease are LV hypertrophy, aortic valve sclerosis, and mitral annular calcification.

Aortic aneurysm	Aortic aneurysm of any cause results in AR due to central noncoaptation of the leaflets. Aneurysms may be idiopathic or familial.
Ankylosing spondylitis	Ascending aortic dilation, aortic wall thickening, thickened cuspal margins; clinical signs of ankylosing spondylitis. Aortic thickening leads to leaflet distortion and prevents coaptation.
Aortitis	Takayasu's or giant cell aortitis may result in AR due to aortic root dilation.
Syphilis	Mild aortic dilation with aortic wall thickening due to inflammation; clinical history or serologic testing confirmatory. Aortic thickening leads to leaflet distortion and prevents coaptation.

Table adapted from Physicians Information and Education Resource (PIER), Aortic Regurgitation module.