

# Chapter 8

## Aortic Regurgitation



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### Name and Synonyms

Aortic Regurgitation

- Aortic Insufficiency

### Incidence/Epidemiology

- The most common causes of aortic regurgitation (AR) in the developed world are congenital bicuspid aortic valve and aortic root enlargement from diseases such as Marfan's syndrome.
- In the developing world, the most common cause of aortic regurgitation is rheumatic heart disease.
- The prevalence of aortic regurgitation increases with age, with about a 2 % prevalence of moderate to severe AR in men over 70, and about a 2.5 % prevalence of moderate to severe AR in women over 70.

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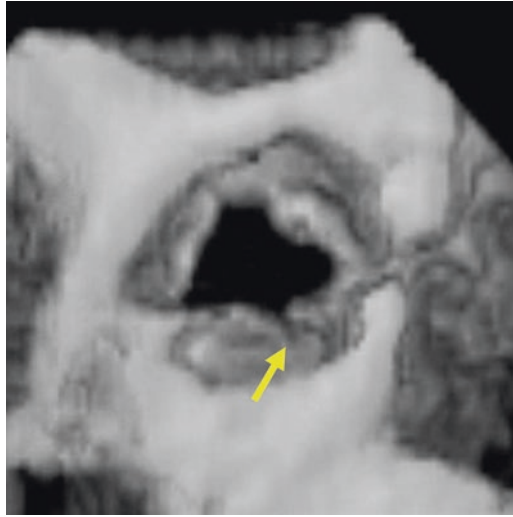
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C. V. Pollack, Jr. (ed.), *Differential Diagnosis of Cardiopulmonary Disease*,  
[https://doi.org/10.1007/978-3-319-63895-9\\_8](https://doi.org/10.1007/978-3-319-63895-9_8)

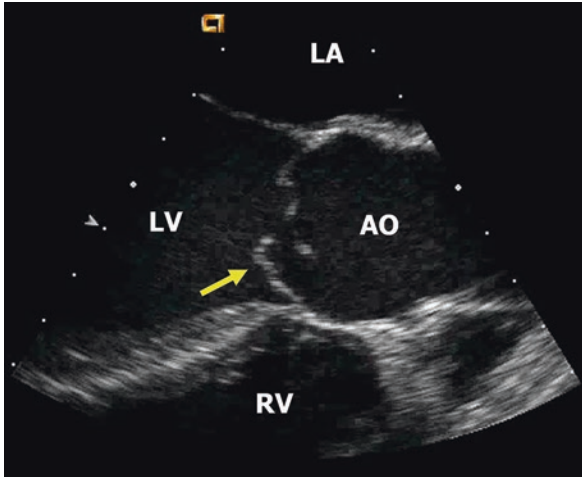
## Differential Diagnosis

- The differential diagnosis of AR is broad, and includes all the pulmonary and cardiac causes of exertional dyspnea, such as myocardial ischemia, pulmonary edema/CHF, pulmonary embolism, and pneumonia.

## Pathophysiology and Etiology

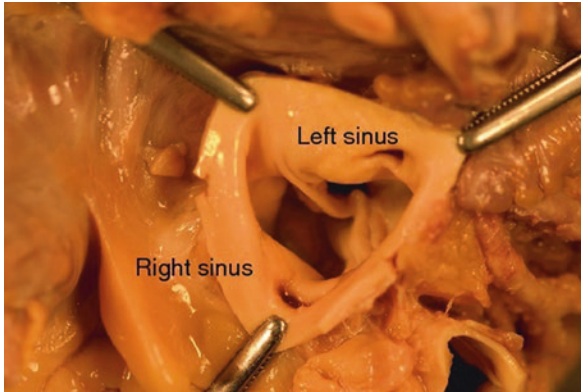


Aortic valve from a patient with aortic insufficiency. The three-dimensional transesophageal echocardiogram demonstrates a tear in the right cusp (arrow). [Garcia M, Liu Z, Rakowski H. Chapter 4. In: Vannan M, Lang RM, Rakowski H, Tajik AJ. Atlas of echocardiography. Philadelphia: Current Medicine; 2005. (Braunwald E, editor. Atlas of heart diseases; vol. 16). ISBN: 1-57340-217-6, 2005-12-05] *Caption from original*



Aortic valve from a patient with aortic insufficiency. This transesophageal echocardiography image demonstrates a bicuspid valve with severe anterior cusp prolapse (arrow). Ao—aorta; LA—left atrium; LV—left ventricle; RV—right ventricle. [Garcia M, Liu Z, Rakowski H. Chapter 4. In: Vannan M, Lang RM, Rakowski H, Tajik AJ. Atlas of echocardiography. Philadelphia: Current Medicine; 2005. (Braunwald E, editor. Atlas of heart diseases; vol. 16). ISBN: 1-57340-217-6, 2005-12-05] *Caption from original*

- Aortic regurgitation may result from either primary aortic valvular disease or aortic root disease.
- Primary valve disease. There are multiple causes of primary valve disease that may result in aortic regurgitation. The final common pathophysiologic pathway involves some deformity/disease of the valve leaflets that prevents normal systolic opening and/or diastolic closure. Usually these processes cause fibrosis, scarring, and hypertrophy of the valve leaflets. The causes of primary valve disease include:
  - Rheumatic valve disease, which is usually associated with concomitant mitral valve disease.
  - Congenital bicuspid aortic valve, which more commonly results in aortic stenosis, but may also develop into aortic regurgitation.



Bicuspid aortic valve. The origin of the coronary arteries can be appreciated in each of the sinuses. BAV was found during autopsy of a 41-year-old male who died from ischaemic heart disease [Suárez-Mier MP, Morentin B, Cobo M, Castedo E, García-Pavía P. Pathology of the Heart Valves. In: Lucena JS, García-Pavía P, Suarez-Mier MP, Alonso-Pulpon LA, editors. Clinico-Pathological Atlas of Cardiovascular Diseases [Internet]. Cham: Springer International Publishing; 2015 [cited 2016 Apr 4]. p. 171–200. Available from: [http://link.springer.com/10.1007/978-3-319-11146-9\\_7](http://link.springer.com/10.1007/978-3-319-11146-9_7)] *Caption from original*

- Membranous, subaortic stenosis
  - Infective endocarditis
  - Syphilis
  - Ankylosing spondylitis
  - Ventricular septal defect
  - Non-penetrating chest trauma
  - Prior use of fenfluramine-phentermine
- Primary aortic root disease. Aortic root disease occurs with any process that causes widening and dilatation of the aortic root. This causes separation of the aortic valve leaflets and prevents normal opening and closing of the valve. Aortic root disease may result from:
    - Marfan's disease
    - Idiopathic
    - Annuloaortic ectasia
    - Osteogenesis imperfecta
    - Severe hypertension
    - Retrograde dissection of the aorta to the aortic root
    - Syphilitic aortitis
    - Ankylosing spondylitis

- Hemodynamic consequences. When the aortic valve does not close completely during diastole, regurgitant blood will flow back from the aorta into the left ventricle. This increases the total stroke volume of the left ventricle because in addition to normal blood flow from the left atrium, regurgitant blood flow from the aorta also fills the left ventricle during diastole. This results in an increased left ventricular end diastolic volume (LVEDV), or increased preload. Initially, the heart compensates for the volume increase with left ventricular (LV) dilatation and subsequent hypertrophy, maintaining a near-normal cardiac output and blood pressure. Eventually, however, the left ventricle can no longer dilate or hypertrophy any further, and the end-diastolic volume continues to increase, leading then to a decrease in stroke volume and forward flow. Initially, these changes manifest as exertional dyspnea and fatigue, as the cardiac output can be maintained during rest but not with exertion. These processes may take many years to develop, and there is generally a long asymptomatic period. It may take 15–20 years for symptoms to develop. This is typical, chronic aortic regurgitation.
- In some cases (especially with endocarditis, dissection, trauma) significant aortic regurgitation may develop acutely. In this situation, the left ventricle cannot handle the massive increase in LVEDV to sufficiently maintain stroke volume, and pulmonary edema or cardiogenic shock develops quickly.

## Presentation

### *Typical/“Classic”*

- Aortic regurgitation is generally a chronic, slowly progressive disease and may be asymptomatic for many (typically 15–20) years.
- Early complaints are somewhat nonspecific but may include:
- An unusual awareness of the heart beating, especially when lying down and on the left side. In these positions, the left ventricle becomes closer to the chest wall.
  - Persistent sinus tachycardia.
  - Palpitations with frequent ectopy. Ectopic beats may become even more uncomfortable than usual, as there is even more LV filling during the compensatory pause, so the eventual beat feels even “stronger” than usual.
  - Noticing that the head “bobs” with the heart beat when sitting quietly.
  - Chest pain, either exertional or nocturnal, often accompanied by severe diaphoresis and not significantly alleviated by sublingual nitroglycerin.
- The typical presentation is one of slowly worsening exertional dyspnea, associated with the typical diastolic murmur of aortic regurgitation. Often, the diastolic murmur is the first manifestation appreciated on routine examination, which then leads to the diagnosis.

## *Atypical*

- The atypical presentation of aortic regurgitation is the acute presentation of pulmonary edema or cardiogenic shock.

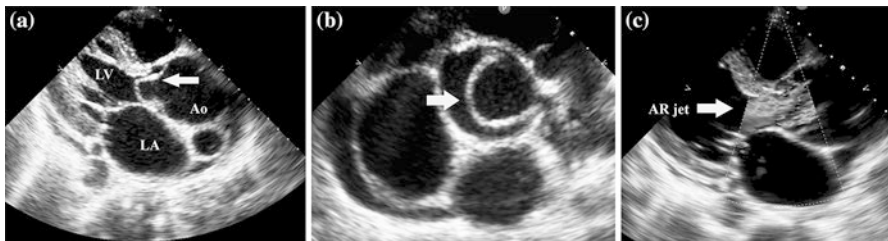
## *Primary Differential Considerations*

- The differential diagnosis of AR is basically limited to other valve disorders, acute coronary syndrome, very proximal aortic dissection, and infective endocarditis.

## History and Physical Exam

### *Findings That Confirm Diagnosis*

- A consistent history and physical examination should be confirmed with a transthoracic echocardiogram.



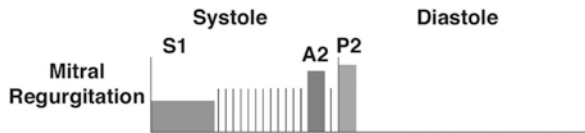
Transthoracic echocardiogram a parasternal long-axis view showing the dissection flap prolapsing into the LVOT (arrow), b parasternal short axis view showing a circular dissection flap (arrow) and c colour Doppler image showing severe aortic regurgitation (arrow). LVOT left ventricular outflow tract, LV left ventricle, LA left atria, Ao aorta, AR aortic regurgitation [Rajesh GN, Sajeer K, Anishkumar N, Sajeer CG, Krishnan MN. Intimo-intimal intussusception–circumferential aortic dissection: a rare mechanism of severe acute aortic regurgitation. *Journal of Echocardiography*. 2014 Sep;12(3):118–9.] *Caption from original*

## Factors That Suggest Diagnosis

- Auscultatory findings.
  - Typical auscultatory findings can strongly suggest the diagnosis of aortic regurgitation.
  - The aortic closure sound (A2) may be soft or absent.
  - The classic murmur of chronic aortic regurgitation is a blowing, high-pitched, decrescendo, diastolic murmur, heard best in the third intercostal space along the left sternal border with the patient sitting up, leaning forward, at end expiration. There is little correlation between the intensity of the murmur and the degree of regurgitation. However, as the degree of regurgitation becomes greater, the duration of the murmur will increase and eventually become holosystolic. The murmur is usually accentuated by maneuvers that increase systemic vascular resistance, such as sustained handgrip, and squatting. The murmur decreases with Valsalva maneuver.

<http://www.easyauscultation.com/cases-listing-details?caseID=124>

The classic murmur of chronic aortic regurgitation. [Aortic Regurgitation (Decrescendo Diastolic Murmur); Easy Auscultation; [www.easyauscultation.com](http://www.easyauscultation.com); copyright 2014, MedEdu LLC]



Caused by an insufficiency of the mitral valve, the mitral regurgitation murmur is a systolic murmur best heard over the mitral auscultation area. A2 = aortic valve closure; P2 = pulmonic valve closure; S1 = first heart sound [Bojanov G. Blood Pressure, Heart Tones, and Diagnoses. In: Iaizzo PA, editor. Handbook of Cardiac Anatomy, Physiology, and Devices [Internet]. Totowa, NJ: Humana Press; 2009 [cited 2016 Apr 4]. p. 243–55. Available from: [http://link.springer.com/10.1007/978-1-60327-372-5\\_16](http://link.springer.com/10.1007/978-1-60327-372-5_16)] *Caption from original*

- There is often an associated midsystolic ejection murmur, heard best at the base of the heart, that radiates into the carotids.
- The Austin Flint murmur is sometimes heard in severe, chronic AR. It is a soft-pitched, rumbling, mid- to late diastolic murmur, heard best at the apex. It is caused by turbulence from the interaction of the usual antegrade blood flow from the left atrium and the retrograde flow from the aorta. This turbulence may result in displacement of the mitral valve leaflets.

<https://www.youtube.com/watch?v=y5CncRHI38>

Video of the Austin Flint murmur.

- Acute AR is associated with a softening of S1 due to the early closure of the mitral valve from the acutely increased LV end-diastolic volume. Often a soft, short, early diastolic murmur also is present.
- Many physical examination findings are associated with chronic, severe AR:
  - Widened pulse pressure. AR is often associated with systolic hypertension and a low diastolic pressure. A clue to disease progression is diastolic blood pressure that seems to increase over time, a result of the worsening regurgitant flow leading to increased end-diastolic volume. As the diastolic pressure approaches the LVEDV, they will eventually become equal and then rise, as the diastolic pressure cannot be below the LVEDV.
  - de Musset's sign: head bobbing with the heartbeat.
  - Corrigan (water hammer) pulse. The pulse is palpated with a forceful, rapid rise, and then quickly dissipates.

<https://www.youtube.com/watch?v=OR19BOodAVI>

Video demonstrating Corrigan's pulse in aortic regurgitation.

- Quincke's pulse (capillary pulsations). While holding pressure at the tip of the nail, one can see pulsations of the skin at the nail bed; may also be seen at the lips.

[https://www.youtube.com/watch?v=9m\\_ORAQDFHM](https://www.youtube.com/watch?v=9m_ORAQDFHM)

Video demonstrating Quincke's pulse

- Traube's sign. A "pistol-shot" sound heard over the femoral artery.
- Duroziez's sign. A back-and-forth bruit heard over the femoral artery after light compression with the head of the stethoscope.

### ***Factors That Exclude Diagnosis***

- A normal echocardiogram excludes aortic regurgitation.

### **Ancillary Studies**

#### ***Laboratory***

- Lab tests for patients with these nonspecific but questionably cardiac complaints should include a CBC, complete metabolic panel, cardiac biomarkers, and a B-type natriuretic peptide.



## *Electrocardiography*

- The ECG in chronic, severe aortic regurgitation often shows LV hypertrophy with an associated strain pattern. This is usually seen as large-voltage QRS complexes with ST depression and T-wave inversions in the inferolateral leads.

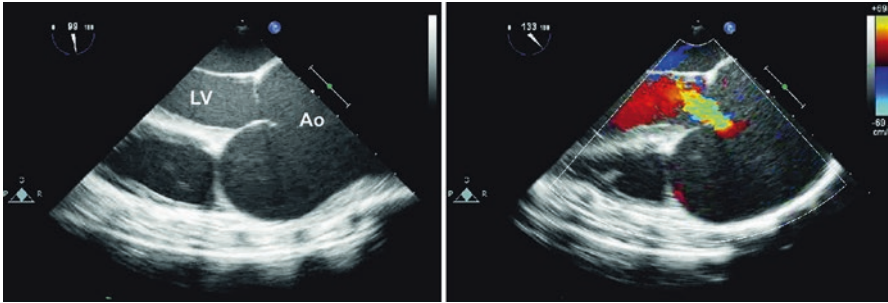


Electrocardiograph (ECG) in a patient with hypertrophic cardiomyopathy (HCM) showing left ventricular (LV) hypertrophy and T-wave inversion [Merlo M, Cocciolo A, Brun F, Sinagra G. Hypertrophic Cardiomyopathy: Clinical Assessment and Differential Diagnosis. In: Pinamonti B, Sinagra G, editors. Clinical Echocardiography and Other Imaging Techniques in Cardiomyopathies [Internet]. Cham: Springer International Publishing; 2014 [cited 2016 Apr 4]. p. 85–94. Available from: [http://link.springer.com/10.1007/978-3-319-06019-4\\_9](http://link.springer.com/10.1007/978-3-319-06019-4_9)] *Caption from original*

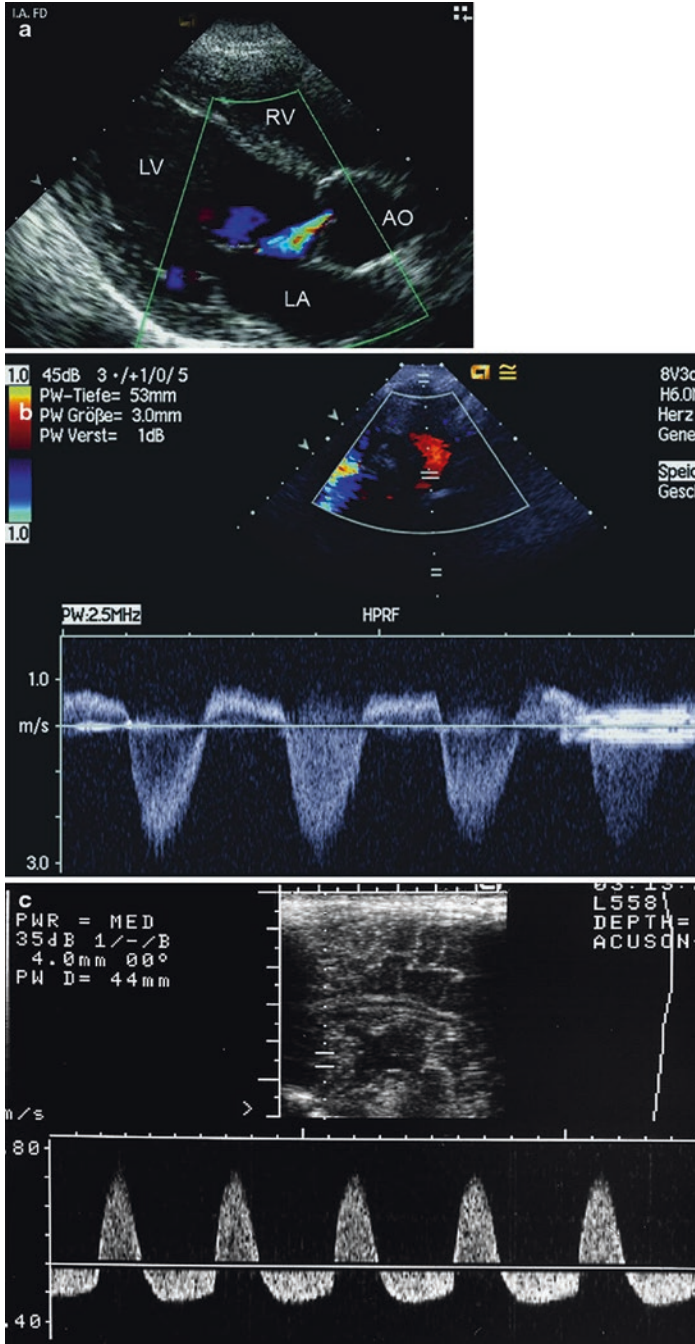
## **Imaging**

- A transthoracic echocardiogram is the diagnostic study of choice in the diagnosis and evaluation of aortic regurgitation. It can document the presence of aortic regurgitation, quantify the regurgitant flow, and define the cause of aortic regurgitation.
- A chest x-ray may be normal, but also may show cardiomegaly associated with LV hypertrophy. In acute aortic regurgitation, the chest x-ray may be consistent with pulmonary edema.

- Cardiac catheterization and angiography may be useful if all other imaging modalities (echo, cardiac MRI) are inconclusive, as well as for preoperative evaluation of aortic regurgitation. Before valve replacement surgery, coronary angiography is often performed to evaluate for concurrent coronary artery disease.



Markedly dilated aortic root as seen from the mid esophageal aortic valve long axis view without color mapping (left panel) and with color mapping, showing central aortic regurgitation (right panel). Ao aorta, LV left ventricle [ Lopez L, Ventura R, Choueiter NF. Outflow Tract Anomalies. In: Wong PC, Miller-Hance WC, editors. Transesophageal Echocardiography for Congenital Heart Disease. London: Springer-Verlag London; 2014. p. 283-305. [https://doi.org/10.1007/978-1-84800-064-3\\_11](https://doi.org/10.1007/978-1-84800-064-3_11)] *Caption from original*



(a) Colour flow imaging of aortic insufficiency. Diastolic flow in the left ventricular outflow tract shown in the parasternal long axis view. The picture shows mild

diastolic backflow from the aorta into the left ventricular outflow tract. AO aorta, LA left atrium, LV left ventricle, RV right ventricle. (b) Flow measurement in the aortic arch in severe aortic regurgitation. Colour Doppler shows retrograde flow in the aortic arch which is displayed red. Pulsed Doppler shows diastolic backflow displayed above the baseline. (c) Flow measurement in the anterior cerebral artery in a patient with severe aortic regurgitation. Diastolic backflow caused by severe aortic regurgitation [Deeg K-H. Cardiovascular Diseases Which Influence the Flow in the Extracardial Arteries. In: Deeg K-H, Rupprecht T, Hofbeck M, authors. Doppler Sonography in Infancy and Childhood [Internet]. Cham: Springer International Publishing; 2015 [cited 2016 Apr 4]. p. 679–730. Available from: [http://link.springer.com/10.1007/978-3-319-03506-2\\_15](http://link.springer.com/10.1007/978-3-319-03506-2_15)] *Caption from original*

## Special Populations

### *Age*

- Aortic regurgitation is more common in middle-aged and older adults, especially because the latent period before the appearance of symptoms may be two decades.
- Aortic regurgitation may be seen in children, in whom it is usually associated with either severe aortic stenosis or a ventricular septal defect. Most cases in children are mild and asymptomatic, and need only be followed with serial exams and echocardiography.

### *Co-morbidities*

- Aortic regurgitation may be diagnosed in relatively healthy patients but also may present in the setting of numerous other cardiovascular diseases and risks.

## Pitfalls in Diagnosis

### *Critical Steps Not to Miss*

- Consideration of the diagnosis is the first critical step. In patients with hemodynamic compromise, an echocardiogram should be performed early to assess all hemodynamic parameters and determine the need for urgent operative intervention.

## *Mimics*

- The entire constellation of diagnoses that underlies dyspnea, chest pain, and other valvular diseases can mimic the presentation of aortic regurgitation.

## **Time-Dependent Interventions**

- Time-dependent interventions in aortic regurgitation are necessary only when acute aortic regurgitation is considered and cardiac and pulmonary function is compromised. In such patients, urgent valve replacement may be a lifesaving procedure.
- For chronic aortic regurgitation, there must be a well-planned and considered approach to operative intervention.

## *Overall Principles of Treatment*

- Acute aortic regurgitation.
  - In acute aortic regurgitation, urgent valve replacement is indicated. Intravenous diuretics and vasodilators (such as sodium nitroprusside) may be helpful as a bridge to surgery.
  - In acute aortic regurgitation, intra-aortic balloon counterpulsation (IABP) is contraindicated, as it may worsen the regurgitation. Intravenous beta-blockers are relatively contraindicated, as they may decrease cardiac output even further.
- Chronic aortic regurgitation.
  - All patients diagnosed with aortic regurgitation, even those who are asymptomatic, must have their systolic blood pressure controlled. Aortic regurgitation is often associated with difficult-to-control systolic hypertension due to increased stroke volume. The goal should be less than 140 mm Hg. Most patients may benefit from vasodilator therapy with ACE inhibitors, dihydropyridine calcium channel blockers, or hydralazine.
  - Patients with early symptoms and mild dyspnea often also benefit from the addition of a diuretic.
  - Surgery with aortic valve replacement is indicated for all symptomatic patients with chronic aortic regurgitation.
  - The optimal time for surgery seems to be after the onset of LV dysfunction but before the development of symptoms. It is important to remember that patients with chronic AR do not become symptomatic until after LV dysfunction develops.

- Appropriately timed surgery often may restore normal LV function. However, surgery delayed for more than 1 year after the onset of LV dysfunction or symptoms often fails to restore normal LV function.
- If there are no indications for surgery, it is generally recommended that the patient be followed up with exams and echocardiograms every 3–6 months.
- Indications for operation in the asymptomatic patient include:
  - An LV ejection fraction <50 %
  - An LV end-systolic dimension < 55 mm or an end-diastolic dimension <75 mm
  - A regurgitant fraction  $\geq$ 50 %
  - A regurgitant volume  $\geq$  60 mL

### ***Disease Course***

- Patients with AR may remain asymptomatic for decades, but many patients eventually require valve replacement surgery.
- Overall operative mortality is 3 %, but in the presence of prolonged LV dysfunction, mortality approaches 10 % and is then also associated with a late mortality approaching 5 % per year from LV failure.

### ***Related Evidence***

Papers of particular interest have been highlighted as:

\*\* *Of key importance*

### ***Practice Guideline***

Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP 3rd, Guyton RA, O'Gara PT, Ruiz CE, Skubas NJ, Sorajja P, Sundt TM 3rd, Thomas JD; ACC/AHA Task Force Members. 2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2014 Jun 10;129(23):e521-643. <https://doi.org/10.1161/CIR.0000000000000031>. PMID: 24589853. <http://www.ncbi.nlm.nih.gov/pubmed/24589853> \*\*

Lancellotti P, Tribouilloy C, Hagendorff A, Moura L, Popescu BA, Agricola E, Monin JL, Pierard LA, Badano L, Zamorano JL; European Association of Echocardiography. European Association of Echocardiography recommendations for the assessment of valvular regurgitation. Part 1: aortic and pulmonary regurgitation (native valve disease). *Eur J Echocardiogr*. 2010 Apr;11(3):223-44. <https://doi.org/10.1093/ejechocard/jeq030>. PMID: 20375260. <http://www.ncbi.nlm.nih.gov/pubmed/20375260> \*\*

## ***Review***

Lung B, Vahanian A. Epidemiology of acquired valvular heart disease. *Can J Cardiol*. 2014 Sep;30(9):962-70. <https://doi.org/10.1016/j.cjca.2014.03.022>. PMID: 24986049. <http://www.ncbi.nlm.nih.gov/pubmed/24986049> \*\*

Prodromo J, D'Ancona G, Amaducci A, Pilato M. Aortic valve repair for aortic insufficiency: a review. *J Cardiothorac Vasc Anesth*. 2012 Oct;26(5):923-32. <https://doi.org/10.1053/j.jvca.2011.07.014>. PMID: 22703946. <http://www.ncbi.nlm.nih.gov/pubmed/22703946> \*\*

Hamirani YS, Dietl CA, Voyles W, Peralta M, Begay D, Raizada V. Acute aortic regurgitation. *Circulation*. 2012 Aug 28;126(9):1121-6. <https://doi.org/10.1161/CIRCULATIONAHA.112.113993>. PMID: 22927474. <http://www.ncbi.nlm.nih.gov/pubmed/22927474> \*\*

Goldbarg SH, Halperin JL. Aortic regurgitation: disease progression and management. *Nat Clin Pract Cardiovasc Med*. 2008 May;5(5):269-79. <https://doi.org/10.1038/ncpcardio1179>. PMID: 18364707 <http://www.ncbi.nlm.nih.gov/pubmed/18364707> \*\*

Bekeredjian R, Grayburn PA. Valvular heart disease: aortic regurgitation. *Circulation*. 2005 Jul 5;112(1):125-34. PMID: 15998697. <http://www.ncbi.nlm.nih.gov/pubmed/15998697> \*\*

Enriquez-Sarano M, Tajik AJ. Clinical practice. Aortic regurgitation. *N Engl J Med*. 2004 Oct 7;351(15):1539-46. PMID: 15470217. <http://www.ncbi.nlm.nih.gov/pubmed/15470217> \*\*

Use PubMed Clinical Queries to find the most recent evidence. Use this search strategy:

“Aortic Valve Insufficiency”[Mesh] OR “Aortic Regurgitation”