PRACTICAL MANAGEMENT OF AORTIC VALVE DISEASE

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Diseases can rarely be eliminated through early diagnosis or good treatment, but prevention can eliminate disease.

Denis Parsons Burkitt

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VHD

- VHD in Africa affect mainly children and young adults and are a result of rheumatic fever.
- RF is a preventable disease, but in Africa the combination of a lack of resources, lack of infrastructure, political, social and economic instability, poverty, overcrowding, malnutrition and lack of political will contributes to the persistence of a high burden of RF, rheumatic valvular heart diseases and infective endocarditis.
A more recent review of the current evidence for the global burden of RF and RHD estimates that 15.6–19.6 million people have RHD (2.4 million children aged 5–14 years) causing 233 364–294 398 deaths from RHD each.

The highest prevalence of RHD is in SSA with a prevalence of 5.7 per 1000, compared with 1.8 per 1000 in NA, and 0.3 per 1000 in economically developed countries with established market economies.
Frequency of Affected Valves

- Mitral: 90%
- Tricuspid: 60%
- Aortic: 30%
## STAGES OF VHD PROGRESSION

<table>
<thead>
<tr>
<th>STAGE</th>
<th>DEFINITION</th>
<th>DESCRIPTION</th>
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</thead>
<tbody>
<tr>
<td>A</td>
<td>AT RISK</td>
<td>Patients with risk factors for development of VHD</td>
</tr>
<tr>
<td>B</td>
<td>PROGRESSIVE</td>
<td>Patients with progressive VHD (mild-to-moderate severity and asymptomatic)</td>
</tr>
<tr>
<td>C</td>
<td>ASYMPTOMATIC SEVERE</td>
<td>Asymptomatic patients who have the criteria for severe VHD:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>C1: Asymptomatic patients with severe VHD in whom the left or right ventricle remains compensated</td>
</tr>
<tr>
<td></td>
<td></td>
<td>C2: Asymptomatic patients with severe VHD, with decompensation of the left or right ventricle</td>
</tr>
<tr>
<td>D</td>
<td>SYMPTOMATIC SEVERE</td>
<td>Patients who have developed symptoms as a result of VHD</td>
</tr>
</tbody>
</table>
AORTIC VALVES

- Commonly
- AR
- AS

- (AR) is the diastolic flow of blood from the aorta into (LV). AR is due to incompetence of the aortic valve or any disturbance of the valvular apparatus (eg, leaflets, annulus of the aorta) resulting in the diastolic flow of blood into the left ventricular chamber.
- Three fourths of patients with significant aortic regurgitation survive 5 years after diagnosis.
- Half survive for 10 years.
- Patients with mild to moderate regurgitation survive 10 years in 80-95% of the cases.
- Average survival after Onset of (CHF) is less than 2 years.
Etiology of Aortic Regurgitation

- Valvular
- Subvalvular
- Supravalvular
Valvular Causes of AR

- Congenital Aortic Valve disease
  - Unicuspid
  - Bicuspid
  - Quadricuspid
- Rheumatic Aortic Valve disease
- Calcific Aortic Valve disease
- Aortic valve endocarditis
- Flail Aortic Valve
- Perforation of Aortic Valve
Bicuspid Aortic Valve
Calcific Aortic Valve Disease
Quadricuspid Aortic Valve
Aortic Valve Endocarditis
Etiology of Aortic Regurgitation Subvalvular level

- Fixed subaortic stenosis
  - Subaortic membrane
  - Fibromuscular type
  - Tunnel type
- Dynamic subaortic stenosis
Etiology of Aortic Regurgitation
Supravalvular level

- Annuloaortic ectasia
- Aneurysm of Sinus of Valsalva
- Supravalvular aortic stenosis
- Coarctation of aorta
- Paraaortic abscess
- Dissection of Aorta
- Aneurysm of Aorta
Congenital causes - Bicuspid aortic valve is the most common congenital cause

Acquired causes:
- Rheumatic fever
- Infective endocarditis
- Collagen vascular diseases
- Degenerative aortic valve disease
- Traumatic
- Postsurgical (including post-transcatheter aortic valve replacement)

Abnormalities of the ascending aorta, in the absence of valve pathology,
- Longstanding, uncontrolled hypertension
- Marfan syndrome
- Idiopathic aortic dilation
- Cystic medial necrosis
- Senile aortic ectasia and dilation
- Syphilitic aortitis
- Giant cell arteritis
- Takayasu arteritis
- Ankylosing spondylitis
- Other spondyloarthropathies
Acute AR

- Leads to increased blood volume in the LV during diastole. The LV does not have sufficient time to dilate in response to the sudden increase in volume.
- As a result, LV end-diastolic pressure increases rapidly, causing an increase in pulmonary venous pressure.
- As pressure increases throughout the pulmonary circuit, the patient develops dyspnea and pulmonary edema.
- In severe cases, heart failure may develop and potentially deteriorate to cardiogenic shock. Decreased myocardial perfusion may lead to myocardial ischemia.
- Early surgical intervention should be considered (particularly if AR is due to aortic dissection, in which case surgery should be performed immediately).
Chronic AR

- Gradual left ventricular volume overload that leads to a series of compensatory changes, including LV enlargement and eccentric hypertrophy.
- The resulting hypertrophy is necessary to accommodate the increased wall tension and stress that result from LV dilation (Laplace law).
- During the early phases of chronic AR, the LV ejection fraction (EF) is normal or even increased (due to the increased preload and the Frank-Starling mechanism). Patients may remain asymptomatic during this period.
- Eventually, the LV reaches its maximal diameter and diastolic pressure begins to rise, resulting in symptoms (dyspnea) that may worsen during exercise. Increasing LV end-diastolic pressure may also lower coronary perfusion gradients, causing subendocardial and myocardial ischemia, necrosis, and apoptosis.
- Grossly, the LV gradually transforms from an elliptical to a spherical configuration.
<table>
<thead>
<tr>
<th>Stage</th>
<th>Definition</th>
<th>Valve Anatomy</th>
<th>Valve Hemodynamics</th>
<th>Hemodynamic Consequences</th>
<th>Symptoms</th>
</tr>
</thead>
</table>
| 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  
       | |  
|       | | • Mild AR:  
       | |  
|       | | • Jet width <25% of LVOT;  
       | |  
|       | | • Vena contracta <0.3 cm;  
       | |  
|       | | • RVol <30 mL/beat;  
       | |  
|       | | • RF <30%;  
       | |  
|       | | • ERO <0.10 cm²;  
       | |  
|       | | • Angiography grade 1+  
       | |  
|       | | • Moderate AR:  
       | |  
|       | | • Jet width 25%–64% of LVOT;  
       | |  
|       | | • Vena contracta 0.3–0.6 cm;  
       | |  
|       | | • RVol 30–59 mL/beat;  
       | |  
|       | | • RF 30%–49%;  
       | |  
|       | | • ERO 0.10–0.29 cm²;  
       | |  
|       | | • Angiography grade 2+  
       | |  
|       | | • Normal LV systolic function  
       | |  
|       | | • Normal LV volume or mild LV dilation  
       | |  
|       | | • 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  
       | |  
| 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 cm²;  
       | |  
|       | | • Angiography grade 3+ to 4+;  
       | |  
|       | | 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)  
       | |  
|       | | C2: Abnormal LV systolic function with depressed LVEF (<50%) or severe LV dilatation (LVESD >50 mm or indexed LVESD >25 mm/m²)  
       | |  
|       | | 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:  
       | |  
|       | | • Doppler 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 cm²;  
       | |  
|       | | • 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  
       | |  

AR indicates aortic regurgitation; ERO, effective regurgitant orifice; HF, heart failure; IE, infective endocarditis; LV, left ventricular; LVEF, left ventricular ejection fraction; LVESD, left ventricular end-systolic dimension; LVOT, left ventricular outflow tract; RF, regurgitant fraction; and RVol, regurgitant volume.
Class 1 indication for echo in AR

1. Confirm presence and severity of acute AR
2. Diagnosis of chronic AR in patients with equivocal physical findings
3. Assessment of etiology of AR (assess valve morphology, and aortic root size and morphology)
4. Assessment of LV hypertrophy, volumes, and systolic function
5. Semi quantitative estimate of the severity of AR
Class 1 indication for echo in AR

6. Reevaluation of patients with mild, moderate, or severe AR and new or changing symptoms

7. Reevaluation of LV size and function in asymptomatic patients with severe AR

8. Reevaluation of asymptomatic patients with mild, moderate, or severe AR and enlarged aortic root
Case 1

Clinical Scenario

Asymptomatic patients with ≥ Moderate AR of Unknown duration

Clinical evaluation

Echocardiography

Within 3 mo

Within 3 mo
<table>
<thead>
<tr>
<th>Scenario</th>
<th>Clinical Evaluation</th>
<th>Echocardiography Evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asympatomatic patients with Severe AR, LVEDD &gt; 70 mm, or ESD &gt; 50 mm, but normal LVEF</td>
<td>Every 4-6 mo</td>
<td>Every 4-6 mo</td>
</tr>
</tbody>
</table>
Case 3

Clinical Scenario                  Echocardiography evaluation

Patients with known >mild AR with Progressive LV Every 6 mo Every 6 mo

Dilation or Declining LVEF
Case 4

Clinical Scenario                  Evaluation of Asymptomatic Patients with Severe AR, Normal LVEF, Every 6 mo And LVEDD >60 mm
Clinical Echocardiography
**Case 5**

<table>
<thead>
<tr>
<th>Clinical Scenario</th>
<th>Clinical Evaluation</th>
<th>Echocardiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymptomatic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patients with mild AR, normal or Midly dilated LV, Once a year</td>
<td>And normal ejection fraction</td>
<td>Once a year</td>
</tr>
</tbody>
</table>
Indications for AV replacement in symptomatic patients

1. Patients with normal resting systolic function (ejection fraction > 50%) and NYHA functional class III of IV symptoms

2. New onset of mild dyspnea and severe aortic regurgitation in patients with increasing LV chamber size or declining LVEF into the low-normal range

3. Patients with NYHA functional class II-IV symptoms and LVEF 25%-49%
Indications for AV replacement in symptomatic patients

4. Symptomatic patients with LVEF <25% and/or end-systolic dimension >60 mm

5. Patients with NYHA functional class II-III symptoms, if (I) symptoms and LV dysfunction are of recent onset and (II) short term vasodilator, diuretic, and/or intravenous positive inotropic therapy result in substantial improvement in hemodynamics or systolic function
Indications for AV replacement in symptomatic patients

6. Patients with NYHA functional class II symptoms and ejection fraction >50% at rest, but with progressive LV dilation or declining ejection fraction at rest on serial studies or declining effort tolerance on exercise testing; patients with Canadian Heart Association functional class II or greater angina with/without coronary artery disease

7. Patients with NYHA functional class IV symptoms and LVEF <25%
AV Replacement in asymptomatic patients

1. Patients with LVEF below the lower limit of normal at rest or 50% on two consecutive measurements

2. Patients with LVEDD >75 mm or ESD >55 mm, even if LVEF is normal

3. Patients with LVEDD of 70-75 mm or ESD or 50-55 mm with evidence of declining exercise tolerance or abnormal hemodynamic responses to exercise

4. Patients with disease of proximal aorta and aortic regurgitation of any degree if the aortic root dilation is >50 mm (valve replacement and aortic root reconstruction)

5. Asymptomatic patients with resting LVEF of 25%-49%
# 2014 AHA/ACC

## Summary of Recommendations for AR Intervention

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>COR</th>
<th>LOE</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVR is indicated for symptomatic patients with severe AR regardless of LV systolic function (stage D)</td>
<td>I</td>
<td>B</td>
<td>(31, 230, 231)</td>
</tr>
<tr>
<td>AVR is indicated for asymptomatic patients with chronic severe AR and LV systolic dysfunction (LVEF &lt;50%) (stage C2)</td>
<td>I</td>
<td>B</td>
<td>(212, 230, 241, 242)</td>
</tr>
<tr>
<td>AVR is indicated for patients with severe AR (stage C or D) while undergoing cardiac surgery for other indications</td>
<td>I</td>
<td>C</td>
<td>N/A</td>
</tr>
<tr>
<td>AVR is reasonable for asymptomatic patients with severe AR with normal LV systolic function (LVEF ≥50%) but with severe LV dilation (LVEDD &gt;50 mm, stage C2)</td>
<td>IIa</td>
<td>B</td>
<td>(226, 243, 244)</td>
</tr>
<tr>
<td>AVR is reasonable in patients with moderate AR (stage B) who are undergoing other cardiac surgery</td>
<td>IIa</td>
<td>C</td>
<td>N/A</td>
</tr>
<tr>
<td>AVR may be considered for asymptomatic patients with severe AR and normal LV systolic function (LVEF ≥50%, stage C1) but with progressive severe LV dilation (LVEDD &gt;65 mm) if surgical risk is low*</td>
<td>IIb</td>
<td>C</td>
<td>N/A</td>
</tr>
</tbody>
</table>

*Particularly in the setting of progressive LV enlargement.

AR indicates aortic regurgitation; AVR, aortic valve replacement; COR, Class of Recommendation; LOE, Level of Evidence; LV, left ventricular; LVEDD, left ventricular end-diastolic dimension; LVEF, left ventricular ejection fraction; LVESD, left ventricular end-systolic dimension; and N/A, not applicable.

*Nishimura, RA et al. 2014 AHA/ACC Valvular Heart Disease Guideline*
LOW FLOW LOW GRADIENT AS

- Severe AS has been defined as that associated with a valve area <1.0 cm$^2$,
- a mean transvalvular gradient >40 mm Hg,
- and a peak flow velocity >4.0 m/s$^1$.
- Patients with severe AS have a significant risk of cardiac morbidity and mortality
- and have improved symptoms and reduced mortality following aortic valve replacement (AVR).
as many as 30% of patients who have a calculated AVA in the severe range have other parameters suggesting mild or moderate disease (ie, mean gradient <30 mm Hg). These patients with low-flow/low-gradient AS (LF/LGAS) may truly have severe AS with resultant myocardial failure (true AS) or may have more moderate degrees of AS and unrelated myocardial dysfunction (pseudo-AS).
Surgical mortality from LF/LGAS has decreased significantly over the past several decades, these patients continue to have a high risk of adverse cardiac events and high mortality whether their disease is managed medically or surgically.

Determining which patients with LF/LGAS will benefit from AVR can be challenging. In this regard, the presence of contractile reserve is a powerful predictor both of perioperative mortality after AVR and of long-term survival.
In a multicenter study of dobutamine stress echocardiography for risk stratification prior to AVR, patients with contractile reserve had an operative mortality of 5% compared with 32% for patients without an augmented response to dobutamine. Furthermore, patients with contractile reserve had significantly improved long-term survival after AVR, whereas patients without contractile reserve had a dismal prognosis with or without valve replacement surgery, in large part related to the high perioperative mortality.
The purpose of valvular intervention

- is to improve symptoms and/or prolong survival
- minimize the risk of VHD-related complications such as asymptomatic irreversible ventricular dysfunction, pulmonary hypertension, stroke, and atrial fibrillation (AF).
Heart valve Team

- The management of patients with complex severe VHD is best achieved by a Heart Valve Team composed primarily of a cardiologist and surgeon.
- In selected cases, there may be a multidisciplinary, collaborative group of caregivers, including cardiologists, structural valve interventionalists, cardiovascular imaging specialists, cardiovascular surgeons, anesthesiologists, and nurses, all of whom have expertise in the management and outcomes of patients with complex VHD.
- The Heart Valve Team should optimize patient selection for available procedures through a comprehensive understanding of the risk–benefit ratio of different treatment strategies.
The greatest medicine of all is to teach people how not to need it.