Valvular Heart Disease

Valvular disease can come to clinical attention due to stenosis, insufficiency (synonyms: regurgitation or incompetence), or both. *Stenosis is the failure of a valve to open completely, which impedes forward flow. Insufficiency results from failure of a valve to close completely, thereby allowing reversed flow.* These abnormalities can be present alone or coexist, and may involve only a single valve, or more than one valve. *Functional regurgitation* is used to describe the incompetence of a valve stemming from an abnormality in one of its support structures, as opposed to a primary valve defect. For example, dilation of the right or left ventricle can pull the ventricular papillary muscles down and outward, thereby preventing proper closure of otherwise normal mitral or tricuspid leaflets. Functional mitral valve regurgitation is particularly common and clinically important in IHD, as well as in dilated cardiomyopathy.

The clinical consequences of valve dysfunction vary depending on the valve involved, the degree of impairment, the tempo of disease onset, and the rate and quality of compensatory mechanisms. For example, sudden destruction of an aortic valve cusp by infection (infective endocarditis; see later) can cause acute, massive, and rapidly fatal regurgitation. In contrast, rheumatic mitral stenosis typically develops indolently over years, and its clinical effects can be well tolerated for extended periods. Certain conditions can complicate valvular heart disease by increasing the demands on the heart; for example, the increased output demands of pregnancy can exacerbate valve disease and lead to unfavorable maternal or fetal outcomes. Valvular stenosis or insufficiency often produces secondary changes, both proximal and distal to the affected valve, particularly in the myocardium. Generally, valvular stenosis leads to pressure overload cardiac hypertrophy, whereas mitral or aortic valvular insufficiency leads to volume overload; both situations can culminate in heart failure. In addition, the ejection of blood through narrowed stenotic valves can produce high speed "jets" of blood that injure the endocardium where they impact.

Valvular abnormalities can be congenital (discussed earlier) or acquired. *Acquired valvular stenosis* has relatively few causes; it is almost always a consequence of a remote or chronic injury of the valve cusps that declares itself clinically only after many years. In contrast, *acquired valvular insufficiency* can result from intrinsic disease of the valve cusps or damage to or distortion of the supporting structures (e.g., the aorta, mitral annulus, tendinous cords, papillary muscles, ventricular free wall). Thus, valvular insufficiency has many causes and may appear acutely, as with rupture of the cords, or chronically in disorders associated with leaflet scarring and retraction.

The causes of acquired heart valve diseases are summarized in Table 12-8. The most frequent causes of the major functional valvular lesions are:

- *Aortic stenosis:* calcification and sclerosis of anatomically normal or congenitally bicuspid aortic valves
- *Aortic insufficiency:* dilation of the ascending aorta, often secondary to hypertension and/or aging
- *Mitral stenosis:* rheumatic heart disease
- *Mitral insufficiency:* myxomatous degeneration (*mitral valve prolapse*)

Table 12-8 Major Etiologies of Acquired Heart Valve Disease

Mitral Valve Disease	Aortic Valve Disease
Mitral Stenosis	**Aortic Stenosis**
Postinflammatory scarring (rheumatic heart disease)	Postinflammatory scarring (rheumatic heart disease) Senile calcific aortic stenosis Calcification of congenitally deformed valve
Mitral Regurgitation	**Aortic Regurgitation**
Abnormalities of Leaflets and Commissures	
Postinflammatory scarring Infective endocarditis Mitral valve prolapse Drugs (e.g., fen-phen)	Postinflammatory scarring (rheumatic heart disease)
Abnormalities of Tensor Apparatus	
Rupture of papillary muscle Papillary muscle dysfunction (fibrosis) Rupture of chordae tendineae	Degenerative aortic dilatation Syphilitic aortitis Ankylosing spondylitis Rheumatoid arthritis Marfan syndrome
Abnormalities of Left Ventricle and/or Annulus	
LV enlargement (myocarditis, dilated cardiomyopathy) Calcification of mitral ring	

LV, Left ventricular.
Modified from Schoen FJ: Surgical pathology of removed natural and prosthetic valves. Hum Pathol 18:558, 1987.

Calcific Valvular Degeneration

Heart valves are subjected to high levels of repetitive mechanical stress, particularly at the hinge points of the cusps and leaflets; this is a consequence of (1) 30 to 40 million or more cardiac contractions per year, (2) substantial tissue deformations during each contraction, and (3) transvalvular pressure gradients in the closed phase of each contraction of approximately 120 mm Hg for the mitral and 80 mm Hg for the aortic valve. It is therefore not surprising that these delicate structures can suffer cumulative damage and calcification that lead to clinically important dysfunction.

Calcific Aortic Stenosis

The most common of all valvular abnormalities, calcific aortic stenosis is usually the consequence of age-associated "wear and tear" of either anatomically normal valves or congenitally bicuspid valves (in approximately 1% of the population). The prevalence of aortic stenosis is estimated at 2% and is increasing as the general population ages. Aortic stenosis of previously normal valves (termed senile calcific aortic stenosis) usually comes to clinical attention in the seventh to ninth decades of life, whereas stenotic bicuspid valves tend to become clinically significant 1 to 2 decades earlier.

Aortic valve calcification is likely a consequence of recurrent chronic injury due to hyperlipidemia, hypertension, inflammation, and other factors similar to those implicated in atherosclerosis. Bicuspid valves incur greater