Acquired Aortic Stenosis

Acquired aortic valve diseases which may cause AS include: (1) senile calcification of the aortic valve and (2) rheumatic valve disease.

Senile Calcification of the Aortic Valve

Senile degeneration of previously normal trileaflet aortic valves results from calcification of the valve leaflets. This is the most common cause of AS typically occurring in the 8th and 9th decades of life. The cusps are immobilised by a deposit of calcium along their flexion lines that begins at their bases and progresses toward the free edge (Fig. 7.11, left). These calcium deposits prevent the cusps from opening normally in systole. Commissural fusion with calcific AS is usually absent.

Degenerative "wear and tear", as a result of the normal mechanical stress on the aortic valve, is a proposed mechanism for this type of AS. Senile AS is commonly accompanied by mitral annular calcification and calcification of the coronary arteries.

Rheumatic Valve Disease

Rheumatic valve disease occurs secondarily to an episode or recurrent episodes of acute rheumatic fever (ARF). ARF occurs due to a group A beta-haemolytic Streptococcus infection and is most commonly encountered in developing nations and disadvantaged communities where there is overcrowding and malnutrition. ARF typically develops in children and adolescents between the ages of 5–14 years, however cases do occur in adults; the disease is rare in children under three years of age.

Valvular disease due to ARF occurs several years after the initial episode of ARF. An autoimmune response leads to chronic inflammation, commissural fusion, thickening, and calcification of cardiac valves. In rheumatic AS, this inflammatory process ultimately leads to retraction and stiffening of the free borders of the aortic cusps reducing the aortic orifice to a round or triangular opening (Fig. 7.11, right). Fusion usually affects all commissures equally but it may also be limited to only a single commissure; in this instance, the aortic valve becomes functionally bicuspid. Associated valvular calcification may or may not be present. The severity of the stenosis depends on the number of commissures that are adherent and the extent of commissural fusion. Importantly, rheumatic AS rarely occurs in isolation and is usually associated with mitral valve stenosis.

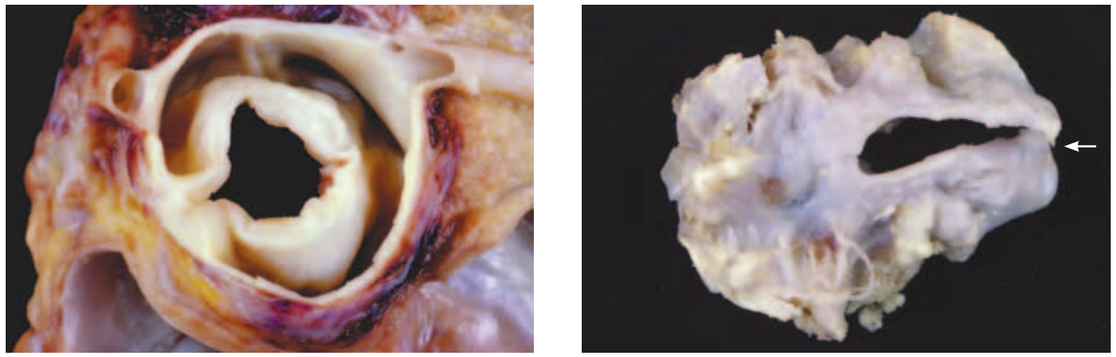
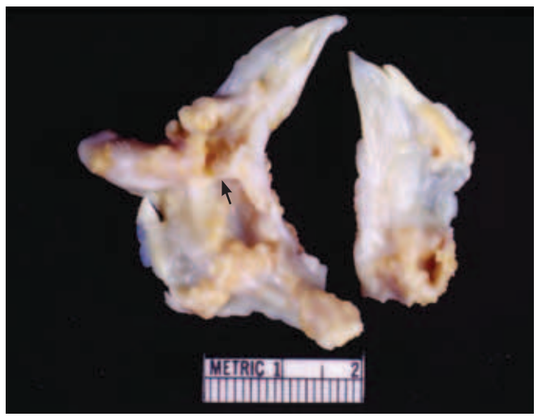


Figure 7.9 Pathological examples of unicuspid unicommissural aortic valves are shown. Observe that the unicommissural type of unicuspid valve has an eccentric opening ('keyhole" or "tear-drop" appearance) with a single commissural attachment to the aortic root (*arrow*). The non-calcified valve (*left*) had only mild stenosis, whereas the heavily calcified valve (*right*) was severely stenotic. Figure 9.40A shows an example of a unicuspid acommissural valve.



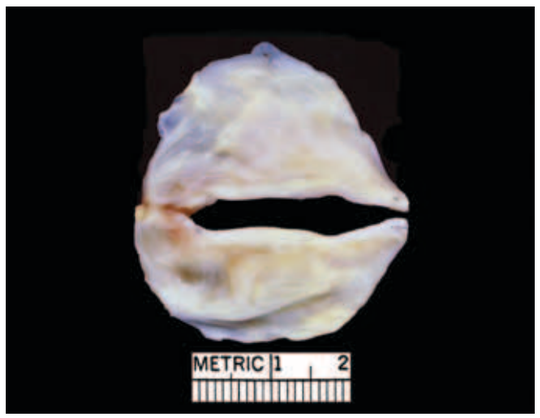


Figure 7.10 Pathological examples are shown of congenitally bicuspid aortic valves. Note that the bicuspid valve with a raphe (*left*) has one larger cusp to its left and one smaller cusp to the right; the raphe is seen in the centre of the larger cusp (*arrow*). Observe that the bicuspid valve without a raphe (*right*) has equal-size cusps.

Rheumatic Valve Disease
The valve most commonly affected by rheumatic heart disease is the mitral valve (65-70%) followed by the aortic valve (25%). The tricuspid valve is deformed in about 10% of patients; rheumatic tricuspid valve disease is almost always associated with mitral and aortic valve disease. The pulmonary valve is rarely affected.

Other Causes of LVOT Obstruction

Congenital obstruction to the outflow of blood from the LV to the aorta can also occur at the subvalvular and supravalvular levels. These lesions are discussed in further detail in Chapter 15.

Subvalvular LVOT obstruction may also occur due to dynamic obstruction. Dynamic obstruction may result from asymmetric septal hypertrophy which causes systolic anterior motion (SAM) of the anterior mitral valve into the LVOT; this is a feature of hypertrophic obstructive cardiomyopathy (see Chapter 6). Dynamic obstruction may also occur in elderly patients with concentric left ventricular hypertrophy (LVH) due to chronic hypertension; in these patients the aorta shifts to the right creating a basal interventricular septal bulge. This septal bulge can cause SAM of the anterior mitral valve into the LVOT resulting in dynamic LVOT obstruction.

Pathophysiology of Aortic Stenosis

Calcification, fibrosis and/or commissural fusion can hinder cusp mobility, cause thickening of the cusps or prevent complete valvular opening; all of which result in a decreased valve area and obstruction to LV emptying during ventricular systole.

In the adult patient, AS is a chronic lesion whereby the degree of LVOT obstruction gradually increases over time. Initially the LV end diastolic volume and systolic function remain normal and LV output is maintained by the development of LVH. LVH allows the LV to sustain large pressure gradients across the stenotic valve for many years without a reduction in the cardiac output, LV dilatation or the development of symptoms. However, as the LV becomes less compliant the LV end-diastolic pressure (LVEDP) increases and the LV dilates in order to maintain a normal cardiac output (see the Frank-Starling mechanism in Chapter 2). In addition, increased end-systolic wall stress due to the increased afterload placed on the LV ultimately results in a reduction in the LV stroke volume and contractility (see Laplace's law in Chapter 2).

Clinical Manifestations of Aortic Stenosis

The clinical manifestations of AS relate to the patho-physiological responses to chronic, severe AS. The primary manifestations of severe AS include angina pectoris, syncope, and congestive heart failure.

Angina Pectoris

Many patients with AS also have coexistent coronary artery disease (CAD). Therefore, these patients may also suffer angina. Furthermore, angina may be exacerbated by LVH. The mechanisms of myocardial ischaemia and subsequent angina in severe AS (unrelated to CAD) include:

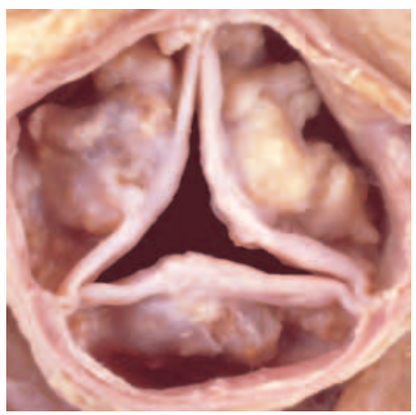
- compression of the intramyocardial coronary arteries due to elevated intracavity LV pressures, prolonged contraction and impaired myocardial relaxation,
- increased myocardial oxygen demand of the hypertrophied LV due to elevated LV systolic pressure and prolongation of the LV ejection time,
- impaired coronary blood flow due to elevation of LVEDP which lowers the diastolic aorta-to-LV pressure gradient and, therefore, the coronary artery perfusion pressure;
- decreased coronary perfusion time due to a decrease in the diastolic filling period,
- reduced coronary flow reserve.

Syncope or Presyncope

Patients with severe AS may experience syncope with exertion or rarely at rest. With exertion, syncope is caused by decreased cerebral perfusion due to peripheral vasodilatation and the inability of the heart to increase cardiac output across a fixed stenotic valve orifice. Syncope may also occur due to baroreceptor dysfunction and a vasodepressor response to increased LV systolic pressures during exercise. Syncope or presyncope may also be due to exercise-induced vasodilation which results in hypotension.

Congestive Heart Failure

Congestive heart failure (CHF) or congestive cardiac failure (CCF) refers to the inability of the heart to supply sufficient blood flow to meet the needs of the body. CHF can result from a number of cardiac causes including AS. The common symptoms of CHF include dyspnoea, peripheral oedema and decreased exercise tolerance.



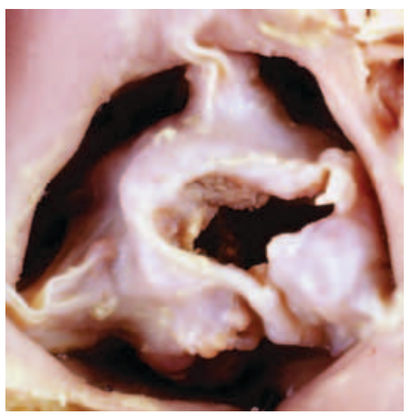


Figure 7.11 Pathological examples are shown of calcific aortic valve disease associated with severe stenosis.
The valve with degenerative disease (left) shows arch-like calcification of the cusps with no commissural fusion. In contrast, the valve with rheumatic disease shows marked commissural fusion, as well as further deformity by fibrosis and calcification.