**ETIOLOGY**

Aortic Stenosis: Congenital, Rheumatic and Degenerative

Aortic Regurgitation: Valvular: Congenital, Rheumatic, and Degenerative

Root: Marfan syndrome, Aortic Aneurysm

**AORTIC STENOSIS**

Pathophysiology

Under normal conditions, the left ventricle pumps blood generating pressure to overcome peripheral resistance in the arterial system. This force is produced by contraction of the left ventricle and generates the necessary flow and perfusion pressure needed to supply the body and its organs. In a normal physiologic state, the pressure generated by the left ventricle is entirely transmitted across the aortic valve to the arterial system. However, in the setting of aortic stenosis, a fraction of the pressure generated by the left ventricle is spent overcoming the resistance of a narrowed valve. The subsequent loss in pressure results in less pressure being transmitted to the arterial system and the formation of a pressure difference (or gradient) between left ventricular and aortic pressures.

However, in the setting of aortic stenosis, a fraction of the pressure generated by the left ventricle is spent overcoming the resistance of a narrowed valve. The subsequent loss in pressure results in less pressure being transmitted to the arterial system and the formation of a pressure difference (or gradient) between left ventricular and aortic pressures. The severity of aortic stenosis and the hemodynamic burden it imposes is measured as a pressure gradient across the stenotic valve. The gradient is expressed as a peak gradient (peak LV systolic pressure - peak aortic systolic pressure) or mean gradient (LV systolic mean pressure - aortic systolic mean pressure). The peak pressure gradient is usually much higher than the mean gradient. A mean pressure gradient of >60 mmHg is an indicator of severe disease (mild disease <30 mmHg, moderate between 30 and 60 mmHg). As a result of generating higher systolic pressure (similar to hypertension), the left ventricle will develop left ventricular hypertrophy. The hypertrophied ventricle is less compliant (or stiffer) and requires higher filling pressure necessitating higher pulmonary venous pressure. This is known as diastolic dysfunction. Pulmonary venous hypertension results in pulmonary congestion which may manifest to varying degrees, as shortness of breath. As a result of the chronic increase in left ventricular pressure (as in the setting of untreated or uncontrolled hypertension), left ventricular systolic dysfunction may ensue. Left ventricular systolic dysfunction is evident with diminished ejection fraction (normal EF +/− 70%). This pathologic state thus results in LV diastolic dysfunction with superimposed LV systolic dysfunction. The resultant low ejection fraction may lead to lower stroke volume and diminished cardiac output. The body may compensate by generating higher pulmonary venous pressure thereby increasing left ventricular filling (increased ventricular volume). As a consequence of this compensatory mechanism, pulmonary venous hypertension and pulmonary congestion are aggravated with associated symptoms of shortness of breath.

In severe aortic stenosis (with high gradient), cerebral perfusion can be maintained as long as adequate pulmonary venous pressure is generated to fill the hypertrophied ventricle thereby maintaining stroke volume and cardiac output. However, in the setting of diminished filling pressures or low peripheral resistance, the left ventricle working at its optimum capacity may not be able to sustain sufficient flow and the necessary pressure to maintain cerebral perfusion. In this instance, the patient may develop symptoms of presyncope or syncope depending on the degree and duration of hypotension.

Left ventricular oxygen consumption is directly proportional to left ventricular pressure and myocardial perfusion is directly related to perfusion pressure. In the setting of aortic stenosis, oxygen demand may exceed oxygen supply (i.e. aortic diastolic pressure - elevated left ventricular diastolic pressure). This imbalance may precipitate the symptom angina.

Right ventricular dysfunction may result from long standing left ventricular diastolic and/or systolic dysfunction requiring higher pulmonary arterial pressure. This may result in systemic venous hypertension and manifest as peripheral edema.

**Management**

There is no indication for medical treatment in asymptomatic patients. However, these patients should be monitored for the development of symptoms or signs of left ventricular systolic dysfunction. 2D ECHO and nuclear scan are valuable tools and may show evidence of diminishing ejection fraction and increased end-systolic volume. Decreased ejection fraction should be associated with an increase in end systolic dimension. Subsequent imaging studies should be completed at intervals to evaluate for progression of disease. Patients with symptomatic disease and significant radiographic changes may benefit from surgical intervention with aortic valve replacement. Serial studies may
be necessary to confirm diagnosis and may be prudent before proceeding to valve replacement surgery.

Negative inotropes i.e. beta blockers and calcium channel blockers should be avoided.

Patients with aortic stenosis and hypertension should be treated with vasodilators as they have no inotrope effect.

Symptomatic patients may require aortic valve replacement. Those patients who opt to defer surgery and present with shortness of breath may be treated with judicious use of diuretics.

*Indications for Surgery*

1. Symptoms
2. Development of systolic dysfunction

**AORTIC REGURGITATION**

**Pathophysiology**

Aortic regurgitation results in increased end diastolic volume. Increased end diastolic volume is associated with increased end diastolic pressure. However, in chronic aortic regurgitation, left ventricular compliance is increased. Therefore, a large volume can be accommodated without requiring increased filling pressure i.e. pulmonary venous pressure. As a result, patients may not present with symptoms of shortness of breath. Chronic volume overload results in progressive left ventricular systolic dysfunction with diminished ejection fraction and increased end systolic dimension. Subsequently, the body compensates by further increasing end-diastolic volume with increased pulmonary venous pressure. This results in pulmonary congestion and shortness of breath. If stroke volume is not restored, low cardiac output ensues. Chronic pulmonary venous hypertension with associated pulmonary arterial hypertension may result in right ventricular systolic dysfunction. This may cause systemic venous hypertension with resultant peripheral edema.

**Management**

Asymptomatic patients should be treated with vasodilators i.e. ACE inhibitors in order to decrease aortic regurgitation.

Patients should be monitored for the development of symptoms and LV systolic dysfunction evidenced by decreased ejection fraction and increased end systolic dimension. 2D ECHO and nuclear studies provide good estimation of disease progression. Decreased EF should be associated with an increase in end systolic dimension. Inconsistencies with respect to ejection fraction and ventricular dimensions should be further evaluated with serial imaging. Diagnosis should be confirmed prior to proceeding to surgery.

*Indications for surgery*

1. Symptoms
2. Progressive LV systolic dysfunction.