Aortic valve stenosis - Congenital Heart Defects

Aortic stenosis is the most common form of left ventricular outflow tract obstruction that accounts for about 3 to 6% of all the congenital heart defects.

Congenital aortic stenosis is divided into three types:

1. Valvular,
2. Subvalvular (subaortic), and
3. Supravalvular.

This classification is based on the site of the obstruction.

Of these types, valvular stenosis is the most common and accounts for 75% of the cases of aortic stenosis.

Subvalvular(subaortic) stenosis is also categorized to three types:

1. Discrete membranous,
2. Fibromuscular and
3. Idiopathic hypertrophic.

Aortic stenosis can be a result of:

- Bicuspid aortic valve with fused commisures that does not open completely or
- Hypoplastic aortic valve annulus or
- Stenosis above or below the aortic valve (sub valvular or supravulvular stenosis).

Pathophysiology of aortic stenosis

An obstruction to the left ventricular outflow due to aortic stenosis increases the load of left ventricle.

Blood flows at an increased velocity across the obstructive valve or stenotic area and into the aorta.

During systole, left ventricular pressure rises dramatically to overcome the increased resistance at the aortic valve. This eventually leades to hypetrophy of the left ventricle.

An aortic valve of less than 0.5 sq cm/sq m body surface area or a pressure gradiant of more than 70 mmHg across aortic value is regarded as severe obstruction

An imbalance between oxygen demand and supply may lead to myocardial infarction in the left ventricle.

left sided heart failure occurs

Clinical signs and symptoms

Most patients with aortic stenosis are assymptomatic or show no signs, except easy fatigability and exercise intolerance, dizziness and syncope.

The most common clinical manifestations are;

- Severe congestive heart failure
- Metabolic acidosis
- Tachypnea
- Faint peripheral pulses, poor perfusion, poor capillary refill, cold skin.
- Poor feeding and feeding intolerance
- Yound child and adolescents
- Chest pains on exertion, decreased exercise tolerance
- Dyspnea, fatigue, shortness of breath
- Syncope, light headedness
- Palpitations
- Sudden death

In ***valvular aortic stenosis,*** pulses are normal but may be small with a slow upstroke, if the pressure gradiant exceeds 80 mm Hg.

Apex shows left ventricular thrust and a systolic thrill at right base, suprasternal notch and both carotid arteries (in mild disease only right carotid artery) may be found.

Auscultatory findings include a prominent ejection click that does not vary with respiration at the aortic area and lower left sternal border, P2 which is physiologically split and a grade 3 to 4/6 rough, medium to high-pitched ejection systolic mumur which is best heard at the first and second spaces and is radiated to suprasternal notch and the carotids, as also down the left sternal border and the apex.

In ***discrete membranous subvalvular aortic stenosis***, the clinical findings are essentially the same but there is no ejection click and a diastolic murmur of aortic regurgitation is usually present after the age of 5 years.

Fibromuscular subvalvular aortic stenosis is clinically almost impossible to differentiate from the discrete membranous type.

Idiopathic hypertrophic aortic stenosis (IHAS) too does not show any ejection click. The ejection systolic murmur (grades 2 to 3/6) is heard over the left sternal border and the apex.

A murmur of mitral regurgitation usually accompanies it.

In ***supravalvular aortic stenosis***, the patient has characteristic “elfin facies” with prominent forehead, epicanthal fold, depressed bridge of nose, overhanging lip, deformity of teeth and strabismus, and mental retardation.

It may coexist with metabolic disorders like idiopathic hypercalcemia and hypervitaminosis D.

The condition is often familial.

The cardiac findings include the thrill and, murmur which are best found in the suprasternal notch and along the carotids. The pulse and systolic pressure in the right arm is higher than in left arm.

Diagnosis of aortic valve stenosis

Auscultation- murmur heard best at right upper sternal border and an injection click

X-ray chest shows somewhat prominent left ventricle though heart size is usually within normal limits.

Dilatation of aorta suggests valvular and, sometimes, discrete membranous subvalvular stenosis.

An electrocardiogram(ECG) is normal in mild disease. In severe obstruction, the changes include left ventricular hypertrophy and strain which may be progressive and left ventricular strain

which warns that operative intervention is warranted.

Echocardiography is of value in the diagnosis and follow-up.

Serial cardiac catheterization is a dependable guide to the progression of the condition.

Cineangiography is important in determining the exact site of the stenotic lesion.

Treatment of aortic stenosis

Treatment in neonates

Stabilization with prostaglandin E1 (PGE1) infusion to maintain cardiac output through the PDA and a close follow up to monitor increassing gradient across the aortic valve or through the aorta

Use of inotropic support as needed.

Intubation and mechanical ventilation may be required.

Administration of a lifelong prophylactic medication against infective endocarditis.

Specialized invasive procedures such as; cardiac catheterization, aortic balloon valvuloplasty or aortic baloon angioplasty.

The patient who gets valve replacement has got to be on anticoagulants.

Surgical valvotomy, commisurotomy or myectomy/myotomy is indicated in the presence of significant manifestations or a large resting gradiant of 60 to 80 mm Hg.

Unfortunately, surgery in the form of valvotomy may be complicated by aortic regurgitation which is worse than the stenosis.

Management of child/adolescent

Medical management with close follow up to monitor increassing gradient across the aortic valve or through the aorta

Restrict strenious exercise and anaerobic exercise eg weight lifting

Restric participation in competitive sports

Aortic balloon valvuloplasty or aortic balloon angioplasty.

The patient who gets valve replacement has got to be on anticoagulants.

Administration of a lifelong prophylactic medication against infective endocarditis.

Surgical intervention valvotomy, myotomy or aortic valve replacement.

Unfortunately, surgery in the form of valvotomy may be complicated by aortic regurgitation which is worse than the stenosis.

Complications

- Congestive heart failure and pulmonary edema
- Dizziness, light headedness and syncope
- Palpitations, arrhythmias
- Infective endocarditis
- Sudden death