UNIT 3 VALVULAR HEART DISEASES

Structure

3.0 OBJECTIVES

After going through this unit, you should be able to:

- diagnosis the valvular heart diseases, i.e., mitral valve, aortic valve and tricuspid valve;
- describe their pathophysiology;
- interpret and correlate various investigations related to the above mentioned valvular heart disease; and
- manage medically and advise surgery at appropriate time.

3.1 INTRODUCTION

Valvular heart disease is a common clinical problem. While rheumatic mitral valve disease seen more often in the underprivileged segments of society continues to be rampant, ischaemic mitral regurgitation is also seen with increasing frequency. Aortic stenosis is commonly associated with patients with chromic heart diseases and male are more susceptible to this diseases. While aortic regurgitation may be cause of primary aortic root disease or primary value disease. Tricuspid valve disease is relatively uncommon. Most of the time it is generally rheumatic in origin and is more common in female. The key principle in management is to understand the stage of the disease.

3.2 MITRAL STENOSIS

Most common cause of mitral stenosis is rheumatic fever. Nearly 30 per cent of patients with rheumatic fever may go on to develop pure mitral valve disease while 50 per cent of mitral stenosis patients may not be able to recall any history of rheumatic fever. All other etiologies of mitral stenosis are very rare and include congenital mitral stenosis, mitral annular calcification, rheumatoid arthritis, and infective endocarditis.

Mitral Valve

To understand mitral valve pathology better, it is important to have a clear idea of mitral valve structure. This may be understood from observations in surgical theater or from pathology specimens. The normal mitral valve is funnel shaped and has orifice area of about 4.0 sq.cm. It has following components:

Fig. 3.1: Components of mitral valve apparatus

1) **Annulus:** Annulus is a saddle shaped structure with medial and lateral portions forming the basal points and anterior and posterior aspects forming the apical points of the saddle. Anteriorly it merges with aortic annulus while posteriorly there is a C shaped discrete fibrous annulus. The normal diameter is about 2.5 – 3.5 cm. and circumference is about 8 – 9 cm. Normal motion and contraction contribute to the normal mitral valve function.

2) **Posterior Left Atrial Wall:** Change in the posterior left atrial wall distort mitral annulus and its contraction. Also sequential atrial and ventricular contraction that occurs in sinus rhythm is important for normal mitral valve closure as loss of sinus rhythm can cause mitral regurgitation.

3) **Leaflets:** Anterior leaflet is longer than wider and posterior leaflet is wider but shorter. Anterior leaflet occupies about one third and posterior about two thirds of annulus. Total combined area of the leaflet is more than double that of annulus. The posterior leaflet has three scallops–medial, central and lateral. Redundancy, loss or restricted motion of leaflets results in mitral regurgitation.

4) **Chordae:** There are 12 primary chordae that subdivide in to secondary and tertiary chordae attaching leaflets to the papillary muscles. Anterior papillary muscle supplies chordae to lateral aspects while posteromedial papillary muscle supplies to medial aspects of the leaflets. Rupture, fusion or redundancy of chordae can lead to regurgitation.

5) **Papillary Muscle:** Two papillary muscles are situated at two thirds of the long axis from the base. Their systolic contraction helps leaflets to overcome the systolic pressure and remain closed during systole without prolapse. Impaired contraction of these due to any cause like ischemia, infarction, necrosis, fibrosis or altered geometry will lead to mitral regurgitation.

6) **Left Ventricle:** Global as well as regional contraction of ventricular muscle is important for normal mitral valve closure. Impaired contractility leads to mitral regurgitation by one or more mechanisms.

Pathology

Following rheumatic fever, over next few years to decades, the typical funnel shaped mitral valve assumes a fish mouth appearance. During rheumatic fever endocardium of the leaflets gets inflamed, edematous and develops pin head vegetations at the tips or along the line of closure. These heal with fibrosis resulting in thickening of the leaflets with limited mobility. Some times there may be calcific deposits over the leaflets. The commissures get fused and in diastole the larger anterior leaflet pulls the posterior leaflet anteriorly. This decreases the valve opening in diastole. Thickened and fused chordae further limit the opening of the valve. Subvalvular apparatus, i.e., the chordae may fuse to such an extent that a secondary orifice may develop below the mitral valve limiting the diastolic blood flow. The abnormal valvular flows due to initial valvulitis may further worsen the thickening and fibrosis of the leaflets progressively. The progression may be rapid in some patients due to repeated attacks of rheumatic carditis, genetic or hormonal factors or due to increased hemodynamic load as seen in manual laborers.

Pathophysiology

Normally there is no pressure gradient between left atrium and the left ventricle during diastole. However, as the valve orifice size decreases pressure gradient develops across mitral valve in diastole and the mean left atrial pressure increases. Increase in left atrial pressure would cause increase in the pulmonary artery wedge pressure. Increase in pulmonary artery wedge pressure would cause interstitial congestion by way of Starling's forces and this presents as symptoms of dyspnoea. Further rise in pulmonary artery wedge pressure causes pulmonary edema and pulmonary arterial hypertension. In some patients there will be disproportionate rise in pulmonary artery pressure due to vasoreactivity of pulmonary arteries. As the pulmonary artery systolic pressure increases further the right ventricle fails and features of right heart failure would ensue.

Fig. 3.2: Mitral stenosis mechanism of diastolic murmur

Symptoms

The cardinal symptom of mitral stenosis is dyspnoea on exertion. Typically it progresses over a period of years. As the severity increases patient will have orthopnoea and paroxysmal nocturnal dyspnoea and class IV dyspnoea. The proximate reason for dyspnoea in mitral stenosis is increased pulmonary artery wedge pressure, which in turn depends upon the left atrial pressure. The pressure gradient across the mitral valve depends upon the size of the valve orifice, quantum of blood flow, i.e., cardiac output and the diastolic time period during which the blood flow

occurs. As the valve size decreases the gradient increases. Increase in cardiac output either due to exercise, pregnancy, anemia, fever or any other condition would increase the pressure gradient and there by the left atrial pressure. During sinus tachycardia or atrial fibrillation with fast ventricular rate, the diastolic time decreases and the pressure gradient and left atrial pressure increases. In all these situations in which the left atrial pressure increases patient will have increasing dyspnoea.

Patients with mitral stenosis sometimes report chest pain. This can be secondary to pulmonary artery hypertension and right ventricular ischaemia. Sometimes pulmonary infarction due to pulmonary embolism can cause pleuritic type of chest pain in these patients.

Palpitations in a patient with mitral stenosis usually herald the onset of atrial fibrillation. They may be paroxysmal or occur continuously at rest with increase on exertion. Some times they may precipitate pulmonary edema and heart failure. However, over a period of time symptoms tend to become quiescent.

Giddiness and syncope are unusual. They suggest severe mitral stenosis with limited cardiac reserve or fixed cardiac output. They should also raise a suspicion of ball valve thrombus in left atrium or left atrial myxoma.

Pedal edema is a late manifestation in the natural history of mitral stenosis. It suggests right heart failure due to severe pulmonary hypertension or associated organic tricuspid valve disease or combination of both.

Physical Signs

A typical malar flush is described in mitral stenosis. It is difficult to appreciate in Indians. Pulse is normal or low volume. Atrial fibrillation is a common arrhythmia in patients with mitral stenosis. Presence of ventricular ectopics should raise a suspicion of digitalis toxicity. In a patient with pulmonary hypertension '*a*' wave may be prominent in jugular venous pulse. With increasing severity of tricuspid regurgitation '*v*' wave becomes prominent. With onset of atrial fibrillation '*a*' *waves* disappear. While left ventricular enlargement is absent, with severe pulmonary artery hypertension, right ventricle may form the apex. First heart sound is loud, but with a calcific and non pliable valve it may be muffled. Intensity of pulmonary closure sound depends upon the severity of pulmonary artery hypertension. A loud opening snap (OS) indicates a pliable valve. Length of diastolic rumble at apex depends upon the severity of mitral stenosis. It may be mid diastolic or presystolic with mild mitral stenosis. Distance between aortic closure sound and opening snap indicates severity of mitral stenosis. Shorter A2–OS distance and longer murmur indicate severe mitral stenosis. Typically in patients with sinus rhythm mid diastolic murmur has presystolic accentuation. However, its presence in atrial fibrillation depends upon the cycle length. With severe pulmonary hypertension murmur of tricuspid regurgitation is present. Signs of right heart failure are often seen in patients with tight mitral stenosis and severe pulmonary hypertension.

Fig. 3.3: Timing of opening snap

Investigations

Electrocardiogram

Electrocardiographic changes are not specific for mitral stenosis and are never diagnostic for any valvular heart disease. They essentially reflect pathophysiological changes. Left atrial enlargement seen in more than 90 per cent of patients is reflected as increase in p-wave duration (>120 m sec.) in lead II or P mitrale. The p-wave axis is between + 45 to – 30 degrees. QRS axis correlates well with the severity of the mitral stenosis and degree of pulmonary hypertension. In pure or predominant mitral stenosis, QRS axis less than 60 degrees suggests a valve area more than 1.3 sq. cm., while a an axis more than that would indicate a valve area less than 1.3 sq. cm., right axis deviation also correlates with the degree of pulmonary hypertension. Absence of right axis deviation in the presence of features of pulmonary hypertension should suggest other associated valvular lesions causing left ventricular hypertrophy. Evidence of right ventricular hypertrophy in the form of R/S ratio more than 1 in lead V1 and poor progression of R-wave height in precordial leads also correlate well with the degree of right ventricular systolic pressure. Atrial fibrillation is common rhythm in patients with mitral stenosis and indicates a large left atrium.

Echocardiography

Echocardiography is diagnostic in mitral stenosis. There is varying degrees of thickening and calcification of leaflets with subvalvular fusion. Commissural fusion results in typical fish mouth appearance of mitral orifice as seen in para sternal short axis view. The same is responsible for anterior motion of posterior leaflet in diastole. Excursion of leaflets and E-F slope on M mode are decreased. Anterior mitral leaflet has a typical hockey stick appearance. Left atrium is enlarged and it becomes a source of clots. Morphological features of mitral valve are of importance in assessing the suitability of mitral valve for balloon valvuloplasty. These features are given in the table. A valve score less than 8 predicts a favourable outcome with balloon valvuloplasty, while a score more than 8 does not preclude valvuloplasty.

Fig. 3.4: Four chamber view showing turbulent flow in MV inflow area suggesting mitral stenosis continuous wave doppler is applied to calculate mitral valve gradient

Measurement of mitral valve and gradients across mitral valve are of importance in clinical decision making. Mitral valve area can be measured by planimetry from para sternal short axis view. Alternatively it can also be measured by different Doppler methods including continuity equation, pressure half time (PHT), and proximal isovelocity surface area methods. Mitral stenosis is considered mild if mitral valve area is >1.5 cm^2, moderate if it is between 1.1 to 1.5 cm^2 and severe if it is equal to or less than 1.0 cm^2. In patients with atrial fibrillation valve area as well as trans valvular gradients are better measured as an average of 5–10 cycles. In the presence of aortic regurgitation, ASD and non compliant left ventricle mitral valve area calculation by PHT method is fallacious. Similarly continuity equation is not applicable in the presence of mitral regurgitation. Errors in measurement are decreased if the color flow mapping is used to direct the continuous wave Doppler beam. While calculated valve area is independent of cardiac output, gradients can be erroneously low with bradycardia and low cardiac output state even in the presence of tight mitral stenosis.

Table 3.1: Echocardiographic Score Used to Predict Outcome of Balloon Valvulopasty

Grade	Mobility	Sub Valvular thickening	Leaflet thickening	Clacification
1	Highly mobile valve with only leaflet tips restricted	Minimal thickening just below the mitral leaflets	Leaflets near normal in thickness. (4-5 mm)	A single area increased echo brightness
2	Leaflet mid and base portions have normal mobility	Thickening of chordal structures extending upto one third of the chordal length	Mid leaflets normal, considerable thickening of margins (5-8 mm)	Scattered areas of brightness confined to leaflet margins
3	Valve continues to move forward in diastole mainly from base	Thickening extending to to distal third of chords	Thickening extending to entire leaflet (5-8 mm)	Brightness extending into the mid portion of the leaflets
4	No or minimal forward move-ment of the leaflets in diastole	Extensive thickening and shortening of all chordal structures extending down to the papillary muscles.	Considerable thickening of leaflet tissue (>8-10 mm)	Extensive brightness throughout much of the leaflet tissue.

Tran esophageal echocardiogram is helpful in detecting left atrial clots prior to planned balloon valvuloplasty or after an embolic event. Valve morphology can also be better assessed.

Cardiac Catheterization

Cardiac catheterization is rarely needed to diagnose mitral stenosis. An end diastolic gradient more than 5 mm Hg. across mitral valve as measured by the gradient between pulmonary artery wedge pressure and left ventricular diastolic pressure diagnoses mitral stenosis. Pressure measurements are mandatory prior to balloon mitral valvuloplasty. Heart rate and cardiac output have to be considered while evaluating pressure gradients. Mean pressure gradient as measured by cardiac catheterization closely approximate to that measured by Doppler examination. Mitral valve area is calculated using Gorlin's formula from the cardiac catheterization tracings.

Treatment

Presence of symptoms or any complications is an indication for treatment. Apart from penicillin prophylaxis no other treatment is warranted for an asymptomatic patient. Asymptomatic patients even with mild to moderate stenosis should avoid heavy unaccustomed exertion as it may precipitate pulmonary edema. Periodic evaluation including echocardiographic examination should be advised. Young women with mitral stenosis should plan pregnancy as even those with mild to moderate stenosis may become symptomatic during pregnancy.

Medical Treatment

Scope of medical treatment is limited and it is only palliative and not specific. Patients with mild to moderate mitral stenosis with minimal symptoms may be managed on medical treatment. Similarly patients who are not candidates for any intervention may be managed on medication. The principles are to relieve symptoms by diuretics, decrease heart rate and prevent thrombo embolic complications in patients with atrial fibrillation or in those with large left atrial chambers. In patients with sinus tachycardia without severe pulmonary artery hypertension beta blockers are the drug of choice. Beta blockers have to be given cautiously in those with severe pulmonary hypertension as sudden death may be precipitated. Beta blockers, calcium channel blockers or digoxine may be given to control ventricular rate in those with atrial fibrillation. In the absence of pulmonary hypertension and atrial fibrillation digoxine has limited role. Rheumatic fever and infective endocarditis prophylaxis needs to be advised.

Balloon Valvuloplasty

Whenever feasible valvuloplasty is the treatment modality of choice. One has to make sure that the valve is suitable and expertise to do the procedure is available. Surgical stand by is desirable. Immediate and long term results are directly related to pliability of the valve. With a pliable valve better valve area can be achieved and chance of immediate severe mitral regurgitation is less. A thorough trans thoracic echocardiography is mandatory prior to performing the procedure. Similarly in those with atrial fibrillation and in those with left atrial diameter more than 4 cm. trans esophageal examination (TEE) should be done to rule out the presence of any left atrial thrombi. TEE is also helpful in assessing the degree of associated mitral regurgitation if any. Patient with left atrial appendage clot may be given oral anticoagulants and followed closely over next few months till the clot dissolves and then go for balloon valvuloplasty. Such a strategy may or may not be useful for left atrial clots as they may or may not dissolve with oral anti coagulants. Higher valve scores and presence of commissural fibrosis and calcification may lead to sub optimal result and acute complications during the procedure. Mitral restenosis is a possibility after balloon valvuloplasty and the time taken for restenosis is dependent upon the initial pliability of the valve and the valve area achieved. Calcific valves tend to restenose early. Patients should continue with penicillin prophylaxis and have a periodic echocardiographic evaluation after balloon valvuloplasty. If symptoms recur repeat balloon valvuloplasty or valve replacement may be done. Complications of the procedure include local vascular complications,

vasovagal episodes, cardiac perforation and tamponade, acute mitral regurgitation, embolic complication due to missed left atrial thrombi and very rarely infective endocarditis. In terms of simplicity and costs balloon valvuloplasty is superior to surgical commissurotomy and all patients should be first considered for balloon valvuloplasty.

Surgical Treatment

Available surgical treatment modalities include closed mitral valvotomy, open mitral valvotomy, and mitral valve replacement.

With the advent of balloon valvuloplasty, closed mitral valvotomy is fast becoming extinct. Selection of patients is like that of balloon valvuloplasty. However, surgical morbidity is more. It may be considered in a rare situation, when balloon valvuloplasty cannot be done expeditiously in a very sick patient but surgical therapy is immediately available. Barring such a situation, there is hardly any scope for closed mitral valvotomy in modern day cardiology practice.

Open mitral valvotomy is an attractive procedure for the surgeon but neither to the patient nor to the cardiologist. It has been proved in a randomized trial that open mitral valvotomy is not superior to balloon valvuloplasty both in immediate and long term results. It is much more costly procedure with higher risk of morbidity and mortality compared to balloon valvuloplasty. It may be considered in a rare patient who has tight mitral stenosis, sinus rhythm and a large left atrial clot with a pliable valve or along with other open cardiac surgical procedures like aortic valve replacement and coronary artery by pass surgery.

Mitral valve replacement is the procedure of choice in a symptomatic patient with tight mitral stenosis and a non pliable valve. Any valve replacement is like buying a new disease and patient should be economically and logistically prepared for it. Patient will be committed for life long anticoagulation and possibility of infective endocarditis. Mechanical prosthetic valves are prone for valve thrombosis, and embolic complications. Inadequately monitored anticoagulant treatment could lead to valve thrombosis or hemorrhagic complications. Presence of atrial fibrillation by itself is not an indication for vale replacement in a patient with pliable valve and tight mitral stenosis, as the long term complications are more with prosthetic valve. Surgery should not be denied to a patient with significant symptoms as the delay in surgery will expose the patient to the complications of mitral stenosis and will adversely affect the long term outcome of the surgery. These include severe pulmonary hypertension, which some times may not come down following mitral valve replacement. A patient with advanced right heart failure and severe cardiac cachexia and hepatic and renal failure also is a poor candidate for surgery and patient should not be allowed to reach that stage before performing mitral valve replacement.

Complications of Mitral Stenosis

Hemoptysis

It is a common complication in patients with mitral stenosis and is related to the severity of mitral stenosis. Pulmonary edema and PND can cause hemoptysis as pulmonary capillaries may rupture due to sudden rise in pulmonary capillary pressure. Pulmonary embolism is common in patients with atrial fibrillation and heart failure and can lead to pulmonary embolism and hemoptysis. These patients with chronic pulmonary congestion are predisposed to bronchitis and winter bronchitis can be another cause of hemoptysis in these patients.

Atrial Fibrillation

As the left atrial size increase and atrial wall gets fibrosed, depolarization wave fronts get fragmented and atrial fibrillation sets in. Atrial fibrillation eventually develops in majority of patients with tight mitral stenosis. Initially it may be paroxysmal but later it becomes persistent and then chronic. Fast ventricular rate decreases diastolic filling time that would increase the mean left atrial pressure. Loss of atrial contraction also contributes to increase in mean left atrial pressure. This would lead to worsening of clinical picture due to increased pulmonary wedge pressure and development of pulmonary edema. Very often patient who has been stable

deteriorates following the onset of AF and becomes symptomatic necessitating surgery or balloon valvuloplasty.

Left atrial dilatation and stasis due to fibrillation causes development of atrial and atrial appendage thrombus. Incidence of thrombo embolic complications in AF due to rheumatic mitral stenosis is 17 times more than that seen in lone atrial fibrillation. Not uncommonly systemic embolization is a presenting symptom in patients with atrial fibrillation. All patients with atrial fibrillation with or without presence of documented left atrial clot or systemic embolization should receive oral anticoagulants. Even when the atrial fibrillation is intermittent, oral anti coagulants should be given. Also in patients with dilated left atrium more than 5 cm oral anti coagulation is considered even when there is no documented atrial fibrillation. Onset of atrial fibrillation is not related to the severity of mitral stenosis since many other co morbid conditions can generate atrial fibrillation. Presence of atrial fibrillation but not the severity of mitral stenosis determines the indication for oral anticoagulants.

Attempts to regain sinus rhythm either by pharmacological means or by electric cardio version often fails if the underlying disease is not tackled. Any attempt to regain sinus rhythm should be made only after excluding left atrial appendage clots. When amiodarone is given to a patient with atrial fibrillation and undiagnosed left atrial clot, sinus rhythm may suddenly be regained and embolization may occur with disastrous consequences. Hence one should be careful in starting amiodarone in a patient with mitral stenosis and atrial fibrillation. It is better to control ventricular rate by other means before starting amiodarone even when the patient is very sick. Trans esophageal echocardiogram is mandatory before starting amiodarone to a patient with mitral stenosis and AF. As far as possible, in patients with mitral stenosis sinus rhythm should be maintained. The best way of achieving this is by relief of mitral stenosis itself. After relieving mitral stenosis, if the patient has atrial fibrillation attempts should be made to regain sinus rhythm. Usually such attempts made before relieving mitral stenosis are futile or will not be long lasting. At the time of surgical correction either by way of open mitral valvotomy or mitral valve replacement surgeon should do maze procedure to regain sinus rhythm. If sinus rhythm is not regained after surgery or balloon mitral valvotomy, patient may be stared on amiodarone after excluding clots in left atrial appendage or left atrium. If after two weeks of treatment with amiodarone sinus rhythm is not regained one should attempt synchronized DC version after excluding left atrial thrombus. DC version may be ineffective when given without pretreatment with amiodarone. DC version should be given cautiously in sick patients with tight mitral stenosis as sometimes it may worsen patient's condition and precipitate pulmonary edema. It has been shown that long term out come with mitral valve replacement is better in those in whom sinus rhythm has been restored than when mere rate control was achieved.

Other Rare Complications

Infective endocarditis is a relatively rare complication of mitral stenosis and in the present era one may unexpectedly find it following balloon intervention. Cardiac cirrhosis, and cardiac cachexia due to long standing heart failure, hoarseness of voice due to long standing pulmonary hypertension are other described complications due to mitral stenosis that are seldom seen today.

Mitral Stenosis and Pregnancy

Since mitral stenosis is often seen in young women, it is not uncommon to see young women with pregnancy complicated by mitral stenosis. Unlike mitral regurgitation, it is poorly tolerated. During pregnancy cardiac output, blood volume and heart rate–all increase. This leads to increased flow during shortened diastolic period and pressure gradients increase across mitral valve for any mitral valve area. Hence patients with even moderate mitral stenosis become quite symptomatic during pregnancy. These young women require proper counselling before marriage and conception but unfortunately due to social conditions this is never effective. Mild to moderate mitral stenosis (MVA < 1.3sq, cm) may safely sail through pregnancy, but often more sicker patients will end up with problems. It is not uncommon to see a pregnant woman with tight mitral

stenosis presenting with pulmonary edema late in pregnancy. If seen in first trimester and the patient has more than mild mitral stenosis it is advisable to terminate the pregnancy and correct valvular lesion. If mitral stenosis at that time is mild to moderate and transvalvular gradients across mitral valve are low, pregnancy may be continued but close medical supervision is needed. Whenever symptomatic, such patient may be started on beta blockers and small dose of diuretics. These drugs can cause fetal growth retardation and should be given in the smallest doses required. If the patient with tight mitral stenosis is seen later in the pregnancy and is symptomatic, she should be offered balloon valvuloplasty if the valve is suitable. Closed mitral valvotomy and mitral valve replacement carry high risk of fetal wastage and maternal morbidity. However, during balloon valvotomy fetus may be exposed to radiation and proper lead shield has to be used and procedure completed by an expert in shortest possible time avoiding unnecessary fluoroscopic time. Pregnant patients with prosthetic mitral valve or with atrial fibrillation who require oral anticoagulants are usually managed with heparin during 6-12 weeks of pregnancy and again later during the last two weeks. Fetal teratogenecity and maternal valve related complications are least with this approach.

Juvenile Mitral Stenosis

Peculiar to developing countries is the problem of juvenile mitral stenosis. Patients with rheumatic fever develop tight mitral stenosis in their adolescence. In developed countries disease tends to present much later. This may be due to initial severe rheumatic carditis or recurrent rheumatic fever. Principles of management are similar to that in adults. Balloon valvuloplasty may be done as indicated and expertise is needed to do the same in young children safely.

Natural History of Mitral Stenosis

Untreated, mitral stenosis is a serious disease with observed 10-year mortality of 70–80 per cent in symptomatic patients. By 20 years virtually nobody survives. About 60 per cent die of progressive heart failure and about 20 per cent die of thrombo embolic complications. Mean survival drops to less than three years in patients with severe pulmonary hypertension. Younger asymptomatic patients or those with minimal symptoms, sinus rhythm and mild mitral stenosis have a better prognosis. However, even in these patients symptoms deteriorate in more than 50 per cent of patients over next 10 years. Though no formal studies are available treatment has significantly altered this natural history.

Check Your Progress 1

1) What happens to the length of the diastolic murmur and the A2-OS interval as the severity of mitral stenosis increases?

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2) What are the changes that may be seen on the Chest X-ray in mitral stenosis?

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3.3 MITRAL REGURGITATION

Mitral regurgitation is the most common valvular abnormality seen in clinical practice. Different disease processes leading to mitral regurgitation may affect different components of the mitral valve. Prevalence of the etiology depends upon surgical or medical series reporting the causes. Table 1 gives a list of various disease processes that cause mitral regurgitation.

Table 3.2: Causes of Mitral Regurgitation

1) **Primary Abnormalities of the Valve Apparatus**

Abnormalities of the Valve

Congenital

Cleft leaflet

Fenestrations

Post Inflammatory

Acute rheumatic fever

Chronic rheumatic Heart disease

Post infective

Infective endocarditis

Degenerative

Myxomatous Mitral valve

Involvement in systemic disease

Systemic lupus erythematosus

Rheumatoid Arthritis

Hypereosinophilic syndrome

Amyloidosis

Scleroderma

Abnormalities of the Annulus

Mitral Annular Calcification

Abnormalities of the Chordae

Idiopathic Chordal rupture

2) **Secondary to Cardiac Disease**

Dilated cardiomyopathy

Hypertrophic cardiomyopathy

Endomyocardial fibrosis

Marfan's syndrome

Coronary artery disease

Papillary muscle rupture

Papillary muscle dysfunction.

Remodeled Left ventricle after Infarction

Dilated left ventricle

Global LV dysfunction

3) **External Causes**

Radiation therapy

Methysergide

Post surgical

Post Valvuloplasty

Pathology

Mitral valve has a complex anatomy and involvement of any of the segments of mitral valve apparatus *viz.* leaflets, chordae, annulus, papillary muscle or underlying left ventricular muscle may cause mitral regurgitation. Congenital mitral regurgitation may be due to cleft mitral leaflet and is often associated with ostium primum type of ASD. Ostium secundum type is associated with mitral valve prolapse. Rheumatic involvement is the most important cause of mitral regurgitation in our country. Commissural fusion, thickening, calcification, shortening and retraction of leaflets and incomplete coaptation of leaflets characterize the pathology of valve. Sub valvular pathology with thickened and fused chordae contributes to mitral regurgitation. Only in about 10 per cent of the cases the lesion is a pure regurgitation while in majority of the cases it is a combined mitral stenosis and regurgitation. Myxomatous mitral valve is characterized by deposition of mucopolysaccahrides in the spongiosa layer. This leads to increase in valve area and length causing hooding of the leaflets. Chordal lengthening and tortuosity may be seen and rupture may occur. Chordae may be thick with mucopolysaccahrides deposition but lack strength. Rupture of chordae results in flail segment of the leaflet. In idiopathic degeneration of chordae usually posterior leaflets are involved and their rupture leads to mitral regurgitation. Sclerosis and calcification of mitral annulus is seen in elderly women who also may have associated hypertension and more often coronary artery disease. Calcific annulus has impaired contraction and also interferes with posterior leaflet coaptation.

Mitral regurgitation is considered secondary if the leaflets and chordae are not directly involved but mitral regurgitation results from changes in annular size or left ventricular geometry as seen in dilated and ischaemic heart disease. In ischaemic heart disease papillary muscle malfunction may be secondary to ischaemia, necrosis or fibrosis of the papillary muscle. Altered geometry with distal displacement of papillary muscle secondary to remodeling also leads to impaired

mechanics of mitral valve closure and thereby regurgitation. Venturi effect on anterior leaflet and also certain secondary structural changes of anterior leaflet cause mitral regurgitation in hypertrophic obstructive cardiomyopathy. Chordal and even leaflet encasement in endomyocardial fibrosis causes mitral regurgitation.

Pathophysiology

During left ventricular systole as the pressure rises in left ventricle, blood is pumped simultaneously both into aorta and left atrium. The effective orifice that allows regurgitation is called effective regurgitant orifice. The fraction of total left ventricular stroke volume that regurgitates in to left atrium is regurgitant fraction. In chronic mitral regurgitation compliant left atrium dilates and accommodates the regurgitant blood thus not allowing the mean left atrial pressure to rise. As the blood flow across mitral valve increases during next diastole, end diastolic volume of left ventricle increases. With increased pre load, left ventricular ejection fraction increases and maintains forward stroke volume in spite of regurgitation. During systole as the left ventricle is open to low pressure left atrium, afterload decreases. Thus in chronic mitral regurgitation left ventricular ejection fraction is more than normal due to increased preload and decreased afterload. As the mitral regurgitation progresses, left ventricle and left atrium dilate. Dilated large left atrium will have areas of fibrosis leading to generation of atrial fibrillation. Dilated chambers with dilated mitral annulus and altered geometry of papillary muscles and posterior left atrial wall lead to aggravation of mitral regurgitation. Hence mitral regurgitation begets mitral regurgitation. Eventually changes take place in left ventricular myocardium causing myocardial dysfunction. As stated earlier, increased preload and decreased afterload of left ventricle allow maintained left ventricular ejection fraction even in the presence of myocardial dysfunction. Left ventricular ejection fraction is not a load independent index of left ventricular systolic function and is not suitable to assess left ventricular systolic function in patients with chronic mitral regurgitation. Removal of the regurgitant orifice through which left ventricle has a low pressure outlet by mitral valve replacement or repair will effectively increase the afterload. Failing myocardium will now face afterload mismatch and left ventricular ejection fraction actually decreases after surgery. Understanding this concept is of fundamental importance in timing of mitral valve surgery.

Clinical Features

Symptoms

Symptoms depend upon underlying etiology of mitral regurgitation. Patients with mild mitral regurgitation and most of those with even severe mitral regurgitation are asymptomatic. However, patients with secondary mitral regurgitation due to ischaemic heart disease or dilated cardiomyopathy will usually be symptomatic of their underlying myocardial failure. Patients usually have dyspnoea on effort, palpitations and fatigue. Worsening symptoms usually denote worsening of underlying left ventricular dysfunction. Conversely left ventricular dysfunction may progress without any symptoms. Symptoms correlate more with hemodynamic status than severity of mitral regurgitation. Patients may also be asymptomatic as they gradually limit their physical activity. Patients with mitral valve prolapse may complain of chest pain. Sudden worsening of symptoms in a stable patient usually indicates chordal rupture or endocarditis. Decompensation due to superimposed hemodynamic burden or onset of atrial fibrillation may sometimes bring chronic mitral regurgitation to the clinical notice. Sometimes patients who present with severe symptoms may slowly settle down due to dilatation of left atrium with increased compliance. Symptoms of edema feet denoting right heart failure are usually a late feature.

Physical Signs

Pulse is of normal character but carotid upstroke may be brisk. Atrial fibrillation is often present in a patient with advanced disease. Blood pressure is normal. Jugular venous pressure is normal in compensated phase. Left ventricle is often dilated with a downward and laterally displaced forcible apex. A systolic left para sternal lift may be palpable as the regurgitant blood enters the left atrium and this is different from para sternal lift due to prominent right ventricle. Occasionally systolic thrill of mitral regurgitation is palpable. First heart sound (S1) is usually soft in rheumatic mitral regurgitation but it is normal in mitral valve prolapse. Second heart sound (S2) may be widely split. A third heart sound (S3) may be palpable at the apex. A fourth heart sound (S4) may be seen with recent onset severe mitral regurgitations and sinus rhythm. A holosystolic murmur starting with S1 and ending with S2 due to mitral regurgitation is audible at apex. In mitral valve prolapse it is a mid systolic murmur starting after a mid systolic click. Murmur radiates to axilla and back with a posteriorly directed jet as seen with anterior leaflet abnormalities, ischaemic and dilated cardiomyopathies. It radiates superiorly and medially towards base with posterior leaftlet abnormalities. Patients with severe mitral regurgitation due to valve pathology have loud and long murmurs while soft, short, barely audible early murmurs are present in patients with functional mitral regurgitation. Murmur is often not audible in patients with acute mitral regurgitation. Physical maneuvers like valsalva, squatting and respiration will help in differentiating it form other systolic murmurs. Mid diastolic murmur may follow an S3 especially in rheumatic mitral regurgitation and is unusual in mitral regurgitations of other etiologies.

Fig. 3.5: Mitral Regurgitation — Haemodynamics

Investigation

Electrocardiogram

Patients with severe mitral regurgitation often have atrial fibrillation. Left atrial enlargement is a common finding in those with sinus rhythm. Left ventricular hypertrophy with ST segment changes may be seen. Signs of pulmonary hypertensions are seen less often and are usually seen in patients with rheumatic etiology. Non specific ST–T changes may be seen in patient with mitral valve prolapse. Q-waves in Inferior leads and LBBB pattern may be seen in patients with functional mitral regurgitation due to ischaemic and dilated cardiomyopathy respectively.

Chest Radiogram

Enlarged left atrium is obvious on chest X-ray and it may occupy most of the cardiac silhouette in patients with aneurysmal left atrium. Left ventricular enlargement may also be seen. Mitral annular calcification may be seen left to the vertebral column in elderly women. Signs of left ventricular failure are seen some times but pulmonary artery dilatation and right ventricular enlargement are uncommon.

Echo and Doppler Evaluation

2 D echocardiography will help determine the morphology and etiology of mitral regurgitation. Rheumatic mitral regurgitation is characterized by thickened leaflets and chordae, commissural fusion and limited mobility of posterior mitral leaflet. Leaflets may contract and may not coapt with each other. In patients with SLE small vegetation may be found. In mitral valve prolapse either of the leaflets crosses the plane of mitral annulus in to left atrium during systole. Since the mitral annulus is of saddle shape an assessment should be made in para sternal long axis view than from an apical view. With myxomatous degeneration, mitral leaflets are thickened. Mitral valve prolapse may be present with no or any degree of regurgitation. A flail leaflet is diagnosed when there is complete loss of cooptation and tip of one of the leaflet prolapses in to leaflet atrium during systole. Ruptured chordae are better observed on trans esophageal echo. Mitral annular calcification with varying degrees of involvement of posterior leaflet, vegetations, perforations and cleft leaflets are obvious on 2 D echocardiographic imaging.

Fig. 3.6: Colour doppler shows central MR jet

In secondary or functional mitral regurgitation, global or regional left ventricular dysfunction is present with normal or mildly thickened mitral leaflets. Characteristic apical displacement of anterior mitral leaflet with tenting due to abnormal tension on the principal chordae results in incomplete leaflet closure with central jet of mitral regurgitation.

2 D guided M-mode measurements of left atrium, left ventricle dimensions and ejection fraction should be obtained and recorded at baseline with which future measurements can be compared. With Color flow imaging one can assess the relative sizes of left atrium and mitral regurgitant jet to estimate the severity of mitral regurgitation and this correlates well with the angiographic severity. However, one should keep in mind that color flow imaging depends upon the gain settings, pulsed repetition frequency, field depth, direction of jet and loading conditions. An eccentric MR jet is often underestimated. Doppler echo indices of severe MR include increased

antegrade flow velocities with a large E-wave, dense continuous Doppler wave spectrum of regurgitant jet and systolic flow reversal in pulmonary veins. Vena contracta is the narrowest portion of the mitral regurgitation jet down stream from the orifice and a biplane vena contracta more than or equal to 0.5 cm indicates severe mitral regurgitation. Using volumetric and PISA method, severity of regurgitation, regurgitant volume and fraction and regurgitant orifice area can be measured. Also from the velocity of mitral regurgitation left ventricle to left atrial pressure difference can be obtained which correlates with the left atrial mean pressure.

Severe mitral regurgitation is diagnosed when:

1) There is echocardiographic evidence of disruption of mitral valve apparatus like papillary muscle rupture, flail mitral leaflet etc.,
2) The effective regurgitant orifice area is $\geq$ 0.40 $cm^{2,}$
3) Mitral regurgitation volume > 60 cc,
4) Regurgitant fraction $\geq$ 55 per cent,
5) Pulmonary vein systolic flow reversal and
6) Mitral regurgitation jet reaches posterior wall of the left atrium.

Severe mitral regurgitation is suggested when:

1) The color flow area is $\geq$ 40 per cent of LA size,
2) Eccentric mitral regurgitation jet reaches the posterior wall,
3) Dense continuous wave Doppler signal is present,
4) E-wave velocity is $\geq$ 1.5 m/s for native valves and $\geq$ 2.0 m/s for prosthetic valves,
5) LV dimension is $\geq$ 7 cm,
6) LA size is $\geq$ 5.5 cm

TEE examination gives a superior window and is indicated whenever the etiology and severity can not be assessed accurately with trans thoracic echocardiography. TEE is also helpful in per operative assessment of mitral valve repair.

Cardiac Catheterization

Routine cardiac catheterization is not necessary in patients with mitral regurgitation. It may be done when there is discrepancy between clinical findings and non invasive tests or preoperatively to do coronary angiography before mitral valve replacement in a patient in whom associated coronary artery disease is a possibility. Pressure tracings may show a prominent '*v*' wave in pulmonary capillary wedge tracings but this in neither a sensitive nor specific finding even in a patient with severe mitral regurgitation as height of '*v*' wave depends upon left atrial compliance. Exercise hemodynamics may provide additional information. Left ventricular angiography may give a qualitative assessment of mitral regurgitation but this is not an objective method. Quantification of mitral regurgitation has been attempted by measuring forward cardiac output by Fick's method and stroke volume by left ventricular angiogram, but this is highly erroneous. However, left ventricular angiogram gives information about left ventricular size, function, severity of mitral regurgitation and regional wall motion abnormalities.

Management

Medical Management

Vasodilator therapy to reduce afterload may be beneficial in patients with chronic mitral regurgitation, but there is no evidence to suggest the same in an asymptomatic patient. In a symptomatic patient with primary mitral regurgitation, surgery should be done. Vasodilators should be used in a symptomatic patient with primary or secondary mitral regurgitation not undergoing surgery. Beta blockers, digoxine, calcium channel blockers and even amiodarone may have to be used for rate control in a patient with atrial fibrillation. Diuretics need to be given as required. Oral anticoagulants are indicated in a patient with atrial fibrillation to prevent embolic episodes though the rate of embolic episodes is low and INR in the range of 2.0–2.5 may be sufficient.

Serial Follow Up

Serial follow up will help to identify patients who develop symptoms and left ventricular dysfunction. It is important to identify early signs of left ventricular dysfunction since left ventricular dysfunction may precede symptoms and post-operative outcome depends upon the left ventricular size and function. Patients with mild mitral regurgitation with no left ventricular dilatation need to be followed once in a year. Annual echocardiograms are not indicated unless they develop new symptoms. Patients with moderate mitral regurgitation need annual evaluation including echocardiography. Patients with severe mitral regurgitation should be followed up once in six months to assess symptoms and left ventricular size and function. Even in an asymptomatic patient left ventricular end systolic dimension more than 45 mm and left ventricular ejection fraction less than 60 per cent are indications for mitral valve repair or replacement. When history is not obvious, exercise testing may be done to assess functional capacity objectively.

Indications for Surgery

Surgery is indicated in all symptomatic patients (class II and above) with severe mitral regurgitation and normal or decreased left ventricular function. Surgery is also indicated in an asymptomatic patient with left ventricular ejection fraction less than 60 per cent and or left ventricular end systolic dimension more than 45 mm. In patients with severe mitral regurgitation, even when the patient is asymptomatic, one should not allow left ventricular ejection fraction to fall less then 55 per cent as post operative recovery will not be good. Though left ventricular ejection fraction is a better index to follow than left ventricular end systolic dimension, surgery may be considered even if one of the parameters is satisfied. When left ventricular ejection fraction is less than 30 per cent or end systolic dimension more than 55 mm, mitral valve surgery carries high risk and may not be beneficial. Surgery is indicated in symptomatic or asymptomatic patient with recent onset severe mitral regurgitation and also in patients with atrial fibrillation. With onset of atrial fibrillation patients tend to become symptomatic and left ventricular dysfunction sets in. Atrial fibrillation tends to persistent even after mitral valve surgery if its duration is more than three months. Similarly asymptomatic severe mitral regurgitation patients with pulmonary artery systolic pressure of equal to or more than 50 mm Hg. at rest or equal to or more than 60 mm Hg. with exercise are also candidates for mitral valve surgery. The outcome in patients with ischaemic mitral regurgitation is worse than those with non ischaemic etiology as they have underlying left ventricular dysfunction. In patients with papillary muscle dysfunction due to reversible ischaemia, simple revascularization may be sufficient. Many patients with severe ischaemic mitral regurgitation need mitral valve replacement or repair.

Types of Surgery

Mitral valve repair, mitral valve replacement with preservation of part of or all of the valve apparatus and mitral valve replacement with removal of mitral valve apparatus are three different surgeries that can be performed. When the valve is suitable and surgical expertise is available

mitral valve repair should be the treatment of choice since it avoids the need for chronic anticoagulation, late prosthetic valve failure and preserves post operative left ventricular function. Preservation of mitral valve apparatus is essential to maintain normal shape, volume and function of mitral valve and it should be preserved as far as possible. Valve or annular calcification, rheumatic and ischaemic involvement, anterior leaflet involvement decrease the chances of repair. Mitral regurgitation due to uncalcified posterior leaflet prolapse due to degenerated mitral leaflets or ruptured chordae tendineae is often repairable. Mitral valve repair rather than mitral valve replacement should be considered seriously in patients with severe left ventricular dysfunction and in asymptomatic patients with good left ventricular function. In the former because any replacement rather then repair will only worsen the dysfunction while in the latter replacement will mandate chronic anticoagulation in otherwise asymptomatic patient.

Natural History

Patients with mild mitral regurgitation and normal left ventricular size and function do well long term, However, those with severe mitral regurgitation with or without left ventricular dysfunction do not de well and their 10 year survival is less than expected to a tune of 50 – 60 per cent in different series. Higher NYHA class, low LVEF, presence of pulmonary hypertension and AF are adverse prognostic factors. Patients who undergo surgery do better and those who undergo surgery at lower NYHA class and those with preserved left ventricular function do better post operatively.

Acute Severe Mitral Regurgitation

Acute severe mitral regurgitation is a medical emergency. Patient presents with breathlessness and low output state. He may be is shock with extreme dyspnoea. There may be features of background disease that led to mitral regurgitation like coronary artery disease usually an inferior wall myocardial infarction, infective endocarditis, or mitral valve prolapse with chordal rupture. In the absence of time for compensatory left ventricular and left atrial dilatation patient will have an acute decrease in forward output and acute increase in pulmonary wedge pressures leading to pulmonary edema. There will be tachycardia, tachypnoea and extremities may be cold with signs of left ventricular failure. One should not be looking for signs of chronic mitral regurgitation. Cardiomegaly is absent and systolic murmur is often short and may even be inaudible. An S3 and often an S4 are audible. Echocardiography is diagnostic and trans thoracic echo may not completely reveal the extent of mitral regurgitation. Trans esophageal echocardiography is required. This will not only show the severity of mitral regurgitation but also uncover the pathology and anatomy of the mitral valve. This is essential to plan type of surgery. Surgery is often mandatory. Medical management includes use of nitroprusside alone or with inotropic agents like dobutamine. Intra aortic balloon pump preeminently decreases the afterload and improves forward output as well as degree of mitral regurgitation. Whenever possible this should be used prior to planned surgery.

Check Your Progress 2

1) What are three common causes of mitral regurgitation?

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2) What are the signs on cardiac examination in mitral regurgitation?

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3) What are some of the echocardiographic features of severe mitral regurgitation?

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3.4 AORTIC STENOSIS

Left Ventricular outflow tract obstruction can occur due to various aortic valvular, subvalvular and supra valvular lesions (Table 3.3).

Valvular Aortic Stenosis

Etiology

Table 3.3: Etiology of Aortic Stenosis

Congenital	Unicuspid
	Bicuspid
	Tricuspid valve with fused commissures
Acquired	Rheumatic
	Degenerated Tricuspid valve
	Degenerated bicuspid valve
Unusual acquired	Hypercholesterolemia
	Obstructive vegetation
	Obstructive tumor
	Rheumatic arthritis
	Alkaptaneurea

Rheumatic aortic stenosis is often seen in young and middle aged individuals and is almost always associated with rheumatic involvement of mitral valve; Fibrocalcific degenerative changes of aortic valve is another important cause of aortic stenosis often seen in the elderly. Such changes occur more rapidly if the underlying valve is bicuspid or if atherosclerotic risk factors are present. Hypercholesterolemia typically hastens the degenerative changes of aortic valve and homozygous patients may present with aortic stenosis in younger age. Similarly hypertension, smoking and male sex also predispose to aortic valve degeneration leading to aortic stenosis.

Pathology

Congenital abnormalities of aortic valve may lead to unicuspid aortic valve. This may be dome shaped with central stenotic orifice. Unicuspid appearance may also be due to fusion of two commissures with an eccentric opening. Usually these patients become symptomatic in childhood or by early adulthood.

Congenital bicuspid aortic valve is one of the common congenital cardiac abnormalities (about 2 per cent) and it has higher prevalence in males. There seems to be certain genetic transmission and it may be associated with other left sided obstructive lesions like coarctation of aorta seen in about 10 per cent of patients. In some patients this may be part of generalized connective tissue disorder associated with cystic medial necrosis and focal apoptosis in aortic media leading to aortic root dilatation. Ascending aortic dissection occurs nine times more frequently in patients with bicuspid aortic valve compared to those with tricuspid valve. Due to altered flow pattern across bicuspid aortic valve, turbulence is generated leading to abnormal hemodynamic stress on the cusps. This results in micro thrombi formation, fibrosis and calcification resulting in aortic stenosis in majority of patients.

Rheumatic aortic valve stenosis is characterized by fusion of one or more commissures with variable cusp fibrosis and calcification. Mitral valve involvement is almost always present. Commissural fusion and edge calcification distinguish rheumatic etiology from degenerative etiology.

Degenerative aortic valve disease that is seen increasingly in elderly shares common pathogenesis with atherosclerosis. Primarily there is lipid accumulation, migration of inflammatory cells and dystrophic calcification in valve cusps. Same atherosclerotic risk factors like age, male sex, diabetes, hypertension and smoking and hyperlipidemia hasten valve degeneration and calcification. Early stages of fibrosis and mild calcification not leading to a significant hemodynamic abnormality are called aortic sclerosis. Extensive distortion due to fibrosis and calcification results in significant hemodynamic abnormality that results in aortic stenosis. While atherosclerosis vascular disease and valvular aortic stenosis seem to have similar etiopathogenesis — it should be noted that majority of patients with extensive coronary artery disease have normal aortic valves and only 50 per cent of patients with degenerative aortic valve disease have significant associated coronary artery disease.

Certain metabolic conditions like hyper cholesterolemia, Fabry's disease and alkaptaneuria lead to valvular stenosis due to metabolite deposition. SLE with associated anticardiolipin antibodies and radiation may also lead to aortic valve stenosis.

Pathophysiology

Patient is often asymptomatic till the orifice size decreases from normal 3 – 4 cm^2 to 1.5 cm^2. Usually valve area < 1.0 cm^2 is considered severe, 1 – 1.5 cm^2 moderate and >1.5 cm^2 is considered mild aortic stenosis. As the area decreases left ventricle to aorta gradient increases. With a severe AS, in the presence of normal cardiac output, mean gradient across aortic valve increases to more than 50 mm Hg. To generate more intracavitary pressure left ventricle undergoes concentric type hypertrophy, i.e., chamber size remains same or decreased but wall thickness increases. As per Laplace law increased wall thickness in the presence of normal or decreased cavity size maintains wall stress. As the severity of LVOT obstruction increases, afterload mismatch sets in, i.e., wall stress cannot keep pace with increasing left ventricular pressure. Left ventricular end systolic volume increases and left ventricular failure occurs. In addition to afterload mismatch some patients have decreased contractile function due to patchy fibrosis due to chronic myocardial ischaemia. While failure due to afterload mismatch responds well to relief of obstruction, failure due to decreased contractile function may not.

The degree of left ventricular hypertrophy varies between individuals. Elderly women tend to have small reduced left ventricular chambers with very thick left ventricular wall.

While left ventricular systolic function is maintained till late in the natural history of disease diastolic dysfunction sets in early. Patients have elevated left ventricular end diastolic pressure due to altered chamber compliance and this leads to prominent atrial contraction (atrial kick). Loss of atrial boost to diastolic filling due to any cause like in atrial fibrillation or complete heart block may suddenly precipitate heart failure.

Compensatory hypertrophy increases the capillary myocardial distance. Left ventricular hypertrophy that would squeeze coronary arteries during prolonged systole and elevated left ventricular diastolic pressure – both lead to demand supply mismatch and ischaemia. Prolonged and repeated ischaemic episodes cause angina, arrhythmias and myocardial necrosis with fibrosis and there by loss of systolic function.

Natural History

There is a prolonged latent period of few decades followed by rapid deterioration. Average survival is 2 – 3 years after the onset of symptom of angina, syncope and heart failure. The rate of progression of stenosis is variable but is consistent for an individual. On an average, in a patient with degenerative aortic valve stenosis valve area decreases by 0.12 cm^2 per year. Rise in trans valvular gradient varies between 10 – 15 mmHg. and in some patients it may increase in the range of 15 – 20 mm Hg. Rate of progression in an individual patient may not be accurate and close clinical follow-up is indicated. Sudden death rarely occurs in asymptomatic patients at an estimated frequency of 0.4 per cent per annum.

Clinical Features

Patients with congenital aortic stenosis usually present in childhood or adolescence. Those with rheumatic heart disease present during 30–50 years and with bicuspid valve between 40–60 years. Degenerative aortic valve disease patients present late in elderly age group. There is no direct correlationship with severity of disease and presence or absence of symptoms. However, those with mean transaortic gradient of more than 60 mmHg are often symptomatic. Symptoms include dizziness or syncope, angina and those of heart failure. Dizziness or syncope may be due to arrhythmias or LV dysfunction but more often due to inappropriate baroreceptor function that results in vasodilatation in the presence of fixed cardiac output.

Angina is noted in approximately 50–60 per cent of patients and in about 50 per cent of these patients it is due to associated CAD. Onset of heart failure symptoms usually portends poor prognosis. The life span is usually considered to be 5, 3 and 2 years after the onset of symptoms of angina, heart failure and syncope respectively.

While infective endocarditis may affect non calcified valves, it is uncommon to affect calcific valves. However, micro emboli from calcific valves may occur. Lower G.I. bleeding due to angiodysplasia of ascending colon is a known association of severe valvular aortic stenosis.

Physical Signs

Typically the carotid pulse in severe aortic stenosis is delayed (pulsus tardus). It is also decreased in amplitude (pulses parvus) when cardiac decompensation occurs. This pulse is fairly specific for severe aortic stenosis. However, patients with low cardiac output and mild stenosis will have a low volume pulse that is normal in timing.

In patients with atherosclerotic vessels carotid upstroke may be brisk even in severe aortic stenosis. A detectable time gap between the apical pulse and carotid pulse suggests prolonged ejection time of severe aortic stenosis. Wide pulse pressure in the presence of other findings of severe aortic stenosis suggests associated aortic regurgitation. Also wide pulse pressure with normal diastolic pressure is fairly common in elderly patients with concomitant hypertensive vascular disease.

Typically apical impulse is forcible and sustained and with decompensation of left ventricle it is displaced. A palpable S_4 is common in severe aortic stenosis. Absence of a valvular ejection

click in children should make one suspect the diagnosis of non valvular aortic stenosis. Usually S_2 is single due to muffled A_2. If the valve is pliable with less extent of calcification, it may produce paradoxical split in a patient with severe valvular aortic stenosis.

Typically the murmur of valvular aortic stenosis starts after S_1, is crescendo decrescendo ejection systolic murmur with maximum intensity at second right inter costal space and radiates to carotids. A grade I murmur in the absence of decompensation suggests mild lesion while the same in a patient with severe decompensation should not make one eliminate aortic stenosis as a cause of decompensation.

A grade IV murmur almost always suggests severe aortic stenosis. In children and elderly selective conduction of high frequency sounds to apical area may give risk to cooing murmur at apex (Gallavardin phenomenon). However, the intensity of murmur may vary with cardiac output, chest valve thickness, emphysema and direction of turbulent jet. A late peaking loud grade IV murmur is very specific to severe aortic stenosis.

Fig. 3.7: Ejection systolic murmur of aortic stenosis

Electrocardiogram

Typically left ventricular hypertrophy with strain pattern is seen in severe aortic stenosis. Total QRS amplitude in mm. tends to correlate and equals trans aortic gradient in pure aortic stenosis. A normal ECG though suggestive of mild aortic stenosis does not totally eliminate severe aortic stenosis. Left atrial enlargement, left axis deviation and various kinds of conduction abnormalities including left bundle branch block, 1° heart block or even complete heart block may occur. Though atrial fibrillation may occur in an advanced aortic stenosis with decompensation–one should eliminate concomitant mitral valve disease.

Roentgenogram

Cardiac enlargement does not occur in pure compensated aortic stenosis. Post stenotic dilatation of aorta may be seen. Fluoroscopy may reveal calcified aortic valve and often concomitant mitral annular calcium. Decompensated aortic stenosis may present with signs of left ventricular dilatation and pulmonary venous hypertension. Gross left atrial dilatation on roentgenogram should suggest associated mitral valve disease. Absence of ascending aortic dilatation gives a clue that valve may not be the site of stenosis.

Echo Cardiography

2D and M-mode Echo cardiography visualize thickened, often calcific aortic cusps with doming motion. In M-mode Echo cardiography relative sizes of aorta, left atrium, left ventricular dimensions and extent of left ventricular hypertrophy and function can be measured. 2D Echo cardiogram shows the number of cusps. But presence of raphae and distorted anatomy due to calcification may make if difficult to assess. Localization of stenosis to sub-valvular or supra

valvular or valvular location can easily be done. Ascending aorta may be disproportionately dilated in some patients with bicuspid aortic valve with a tendency for aortic dissection.

Fig. 3.8: Two dimensional echocardiography plax view showing thickened calcific aortic valve in a patient with severe AS

Hemodynamic severity of aortic stenosis can be measured by Doppler echocardiography. Peak and mean trans valvular aortic gradients, aortic valve area and ratio of LVOT and aortic valve time velocity integral (VTI) are important measurements. Meticulous search using all transducer positions should be made to obtain maximal aortic velocity. In patients with normal LV function and cardiac output, aortic stenosis is considered severe when peak aortic velocity is more than or equal to 4.5 m/sec; mean pressure gradient equal to or more than 50 mmHg, aortic valve area less than or equal to 0.75 cm^2 or LVOT/Aortic valve VTI ratio less than or equal to 0.25. It should be noted that in-patients with left ventricular dysfunction and/or low cardiac output, transvalvular pressure gradients will be under estimated while in situations with increased flow across aortic valve like aortic regurgitation and/or anemia transvalvular gradient will be over estimated. Since in both these situation LVOT and Aortic valve VTI move in the same direction and value area as measured by continuity equation makes use of ratio of these two, valve area measurements tends to be more accurate. Also valve resistance which takes into account valve area and flow across the valve is a more reliable index. Further while following a patient serially with echocardiography combined use of transvalvular gradients, left ventricular function and valve area calculation are more useful. Otto and his colleagues have shown that in asymptomatic patients, peak aortic jet velocities increase by 0.32 ± 0.34 m/sec per year, mean gradient by 7± 7 mmHg and valve area decrease by 0.12 ± 0.19 cm per year. Beyond a peak velocity of 4.0 m/sec most of the patients become symptomatic within 2 years. Aortic valve area is d" 0.75 cm^2 when peak aortic velocity is > 4.5 m/sec and mean gradient is e" 50 mm Hg. However, half of patients with aortic valve area d" 0.75 cm^2 have lower peak velocities and mean gradients due to lower cardiac output.

Ratio of LVOT/Aortic valve VTI is another good index in measuring aortic valve area. A ratio of LVOT/AV VTI is < 0.25 correlates well with a aortic valve area of < 0.75 m^2.

In a patient with low out put and low aortic valve gradients dobutamine echo (upto dose of 20 mcg/KG/mt) is useful in differentiating between true aortic stenosis and pseudo or functional aortic stenosis. With dobutamine infusion, in a patient with true aortic stenosis, LVOT velocity and aortic velocity increase proportionately but the ratio between both remains same or less than 0.25. Conversely in a patient with functional or pseudo aortic stenosis with severe left ventricular dysfunction–aortic velocity does not increase in proportion to LVOT velocity because aortic valve area increases with increased stroke value. This results in higher LVOT to aortic valve VTI ratio. It becomes more than 0.25 ruling out any significant aortic stenosis.

During echocardiography we should be careful in measuring trans aortic flow than mitral or tricuspid regurgitation jets. Aortic annulus measurement is important for the surgeon to plan aortic valve replacement.

Trans esophageal echocardiography (TEE) is an important complement to transthoracic window. It helps in assessing the severity of mitral regurgitation and pathology of mitral valve if any. Number of cusps can be confidently measured. Planimetry measurement of aortic valve area can also be done in TEE short axis view. Pre-operative TEE is helpful in assessing associated mitral regurgitation.

A well done echocardiographic assessment precludes the need for invasive hemodynamic measurement.

Exercise Testing

Exercise testing is relatively contraindicated in patients with aortic stenosis. Its diagnostic accuracy to pick up additional coronary artery disease is limited in view of baseline ECG changes and left ventricular hypertrophy. Exercise testing should not be performed in symptomatic patients. It is safe to do in asymptomatic patients provided it is supervised by an experienced physician and ECG and blood pressure are closely monitored. While prognostic information of exercise induced ECG changes is uncertain—it can provide data about patient's exercise capacity and exercise induced hypotension. Limited exercise tolerance and/or exercise induced hypotension is an indication for aortic valve replacement in a patient with moderate to severe aortic stenosis.

Cardiac Catheterization

The indications are to confirm hemodynamics when the non-invasive tests are not conclusive or there is a discrepancy between clinical evaluation and echocardiographic findings. It is also done pre-operatively to obtain coronary angiography if associated CAD needs to be ruled out. During cardiac catheterization right and left heart pressures and all necessary gradients should be measured in a meticulous manner. Transvalvular gradients, cardiac out put should be measured to calculate aortic valve area. Crossing a calcified degenerated aortic valve should be avoided unless it is clearly indicated. If entering left ventricle is not possible by retrograde approach transeptal puncture may be done to assess left ventricle to aorta gradient. When clinical impression correlates well with non-invasive evaluation–invasive hemodynamic evaluation with all its complications can be avoided.

Management of Asymptomatic Patient

The most common cause of death in a truly asymptomatic patient with severe aortic stenosis is aortic valve replacement (AVR). While expected incidence of sudden death in those patients is 0.4 per cent per annum, direct surgical mortality and post-operative complications are more in operated patients. However, patients with severe aortic stenosis may undergo aortic valve replacement if they are undergoing coronary artery bypass surgery, other valvular or aortic root surgery. Similarly, AVR may be done even for moderate aortic stenosis in some situations. AVR may also be considered in patients with asymptomatic severe "AS" if it is documented that they have left ventricular dysfunction or hypotensive response to exercise. It may also be considered for patients with very severe AS or excessive hypertrophy.

Follow-up

Asymptomatic severe aortic stenosis patient needs to be followed once in a year or even more frequently. Patients with moderate aortic stenosis need follow up once in two years and those with mild aortic stenosis over a five years provided they do not have any new symptoms or clinical worsening. Echo cardiogram should also be performed whenever there is change in symptoms on signs. In obese individuals or patients with poor echo window serial follow-up with

magnetic resonance imaging (MRI) is a good alternative. Patients should be advised to come for prompt follow-up, if they developed any exercise related symptoms.

Management of Symptomatic Patients

All symptomatic patients with severe aortic stenosis or even moderately severe aortic stenosis should undergo aortic valve replacement and they do well after surgery. Even patients with severe LV dysfunction should be offered aortic valve replacement, if they have severe aortic stenosis. However, if the cause for patient's left ventricular dysfunction is decreased myocardial function rather than afterload mismatch or if they do not have severe aortic stenosis they may not do well after aortic valve replacement.

Aortic valve replacement may not be indicated if the patient is very elderly, debilitated and has associated severe co morbid conditions like cancer, stroke etc. Patients with extreme degree of left ventricular hypertrophy also have increased surgical complications. In elderly patients age alone should not influence the decision regarding surgery.

Balloon Aortic Valvuloplasty

This is the procedure of choice in children and young individuals where the valve is not calcified. In elderly patients with calcified valves results are suboptimal. Peri procedural complications may be seen in upto 10 per cent of patients. Restenosis rates requiring repeat intervention are very high. In elderly patients this should be done only as a bridge to surgery as in the patients with cardiogenic shock or pulmonary edema. This may also be considered as an emergency procedure in patients who require non cardiac surgery or have serious comorbid conditions. Percutaneous aortic valve replacement is newer modality of treatment that has to be established.

Medical Therapy

Definitive therapy for aortic stenosis is surgical and medical therapy is only palliative. Infective endocarditis and rheumatic fever proplylaxis should be given as indicated. In patients who are not being considered for aortic valve replacement digoxine and diuretics can give symptom relief. In decompensated heart failure ACE inhibitors may be tried cautiously. Sinus rhythm should be maintained with amiodarone. Beta blockers or other drugs with negative inotropic effect should be used very cautiously or not at all. Nitrates may be given cautiously. Patients with moderate to severe aortic stenosis, even when asymptomatic, should be advised against any kind of vigorous exercise or physical activity. In degenerative valvular AS statin therapy retards the progression of the disease.

Low Gradient Aortic Stenosis

One may come across patients who have severe left ventricular dysfunction and modest gradients across aortic valve. This situation may be due to afterload mismatch in a patient with severe valvular aortic stenosis and left ventricular dysfunction or this situation can also be due to moderate aortic stenosis in a patient with intrinsically severe left ventricular muscle dysfunction. Valve area calculation or valve resistance calculations are not particularly useful in this situation with low cardiac output. Dobutamine stress echo cardiography is helpful in this situation. In a patient with intrinsic myocardial dysfunction, cardiac output, trans aortic gradient and calculated valve area — all increase. In a patient with severe aortic stenosis with afterload mismatch also dobutamine infusion would increase calculated valve area but it will still be in the range severe aortic stenosis. The latter patient may benefit from aortic replacement while the former would not.

Check Your Progress 3

1) What are the symptoms with aortic stenosis?

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2) What are the clinical features of valvular aortic stenosis?

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3) How do you assess the severity of aortic stenosis by echocardiography?

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3.5 AORTIC REGURGITATION

Aortic regurgitation is a common valvular disease that may present chronically or acutely. Normally the integrity of valve closure depends upon the anatomy of valve leaflets and on three dimensional geometry of aortic root and sinuses of Valsalva. Wide variety of causes may lead to aortic regurgitation. Usually 50 per cent of valve replacements are due to diseases of valve and 50 per cent due to root disease. However, in India predominant cause of aortic valve replacement is still rheumatic.

Table 3.4: Causes of Aortic Regurgitation

Acute	–	Aortic dissection
	–	Trauma
	–	Leaflet perforation due to endocarditis
Chronic		
Valve leaflet disease	–	Congenital
		Bicuspid
		Unicuspid
		Quadricuspid
		Fenestrations
	–	Rheumatic
	–	Infective endocarditis
	–	Myxomatous degeneration
	–	Fibro calcific degeneration
	–	Systemic Lupus Erythematosus

	–	Rheumatoid
	–	Pharmacological agents
Aortic Root Disease	–	Hypertension
	–	Idiopathic root dilatation
	–	Marfan's Syndrome
	–	Aortic dissection
	–	Annuloaortic ecstasia
	–	Cystic medial necrosis
	–	Syphilis
	–	Ankylosing spondylosis
	–	Takayasu arteritis
	–	Psoriatic arthritis
	–	Reiter's Syndrome
	–	Relapsing Polychondritis
	–	Kawasaki disease
	–	Osteogeneses imperfecta
	–	Giant cell Arteritis
	–	Ehlers – Danlos Syndrome
Sub Valvular	–	Discrete sub aortic stenosis
	–	VSD with aortic regurgitation due to prolapse

Majority of elderly hypertensive patients have trivial to mild aortic regurgitation due to varying degrees of root dilatation due to hypertension and degenerative changes in the valve. However, they are rarely of clinical importance and may not progress to severe aortic regurgitation.

Pathology

In Post Inflammatory situations scarring and retraction of valve cusps lead to aortic regurgitation. Varying degrees of commissural fusion leads to incomplete opening and closing in rheumatic heart disease.

15 – 20 per cent of bicuspid valve patients will have significant aortic regurgitation. In Marfan's Syndrome due to abnormalities in fibrillin and in cystic medial necrosis due to abnormalities in elastin aortic wall becomes weak, stretched out and does not support commissures leading to incomplete central closure. In inflammatory aortitis situations like syphilis and ankylosing spondylitis, medial inflammation and scarring cause root dilatation. In SLE with anti cardiolipin antibodies, Libman-Sacks endocarditis and in rheumatoid arthritis, rheumatoid nodules cause scarring of valve cusps.

Pathophysiology

Left ventricle responds to chronic aortic regurgitation by chamber dilatation and an increase in its compliance so that end diastolic pressure does not increase. This is accompanied by rearrangement of muscle fibers and addition of new sarcomeres leading to eccentric hypertrophy. As the chamber dilates—with preserved systolic function – stroke volume increases compensating the regurgitant volume. However, dilated chamber increases wall stress and afterload and to compensate for the increased afterload concentric hypertrophy ensues. Thus, chronic aortic regurgitation represents combined volume and pressure overload. During this compensatory phase involving preload reserve and concentric and eccentric hypertrophy, patient

remains asymptomatic with preserved left ventricular systolic function but with dilated left ventricle. This can go on for many years. Patient becomes symptomatic as the preload reserve gets exhausted and end diastolic pressure increases. Further increase in afterload leads to afterload mismatch and left ventricular systolic function declines. The changes occur very insidiously and patient may remain asymptomative till severe LV dysfunction sets in. As the chamber enlargement proceeds and geometry alters depressed myocardial function occurs and predominates over afterload mismatch.

Though both mitral and aortic regurgitation cause volume overload to left ventricle–aortic regurgitation has additional pressure overload as the increased stroke volume has to be ejected into high impedance aorta. This is also borne out by the increased left ventricular end systolic wall stress. In mitral regurgitation, the initial compensatory mechanism is increased ejection fraction with little ventricular dilatation but in aortic regurgitation it is ventricular dilatation with preserved ejection fraction.

In aortic regurgitation, coronary perfusion is impaired due to decreased aortic diastolic pressure and increased oxygen demands. This in severe cases leads to sub endocardial ischaemia.

Clinical Features

Symptoms

Patients with chronic severe AR may be asymptomatic for many years and it may be picked up on routine examination. Some patients may remain asymptomatic even with severe left ventricular systolic dysfunction. Excertional dyspnoea occurs as the left ventricle end diastolic pressure increases. Impaired coronary blood flow results in angina, sometimes occurring in the nights and awakening the patient from sleep (nocturnal angina). As the systolic dysfunction worsens, patients may have dyspnoea at rest, paroxysmal nocturnal dyspnoea, and even frank heart failure. Angina is more pronounced in patients with Leutic or Takayasu aortitis with coronary ostial disease. Patients may also complain of palpitation due to forcible ventricular contraction. Rarely sudden death may occur in patients with severe aortic regurgitation.

Physical Signs

Chronic severe aortic regurgitation is characterized by wide pulse pressure and multiple peripheral signs it produces. Sharp rapid upstroke of radial pulse followed by rapid down stroke is called Corrigan's pulse or water hammer pulse and is exaggerated by elevating the wrist. Carotids have two prominent systolic impulses called bisferiens pulse. There may be palpable thrill in carotids due to rapid ejection of increased stroke volume.

Due to rapid run off of blood from aorta in diastole and increased stroke volume, varieties of physician signs are described. Movement of head synchronously with arterial pulsation causes head bob. Auscultation over femoral arteries with gentle compression produces to and fro bruit called Duroziez's sign and it corresponds flow reversal in aorta. The prominent pulse waves may be audible over brachial and other peripheral arteries and are called pistol shot sounds. These interfere with measurement of diastolic blood pressure. In significant aortic regurgitation diastolic blood pressure is usually less than 70 mmHg and systolic blood pressure is elevated causing wide pulse pressure. Normal systolic blood pressure difference between lower and upper limbs of 10–20 mmHg is exaggerated in aortic regurgitation. The difference of more than 60 mmHg suggests severe aortic regurgitation, 40–60 mmHg difference suggest moderate aortic regurgitation and 20–40 mmHg difference mild.

Usually cardiac enlargement is present and left ventricular hyper dynamic apex is palpable outside mid clavicular line. S_1 is normal or slightly reduced in intensity. A_2 is louder in aortic regurgitation due to aortic root disease while it is muffled in valvular aortic regurgitation. Left

ventricular S_3 suggest left ventricular dysfunction. Constant ejection click may be audible in bicuspid aortic valve or with root dilatation.

Characteristic murmur is early diastolic murmur which is high pitched and blowing. It is best heard in left parasternal border third left intercostals space at end expiration with patient sitting and bending forward. A cooing or musical murmur suggests everted or perforated cusp (Seagull murmur). A murmur that is better heard on the right side of the sternum suggests aortic root disease. Ejection systolic murmur due to increased systolic flow across aortic valve is common and its presence does not necessarily mean aortic stenosis. Some patients have their cooing early diastolic murmur referred to apex. At apex one may additionally hear Austin Flint (mid diastolic) murmur. This may be due to aortic regurgitation jet pushing the anterior mitral leaflet up and causing relative mitral stenosis or just audibility of low pitched vibrations of aortic regurgitation itself. This needs to be differentiated from associated mitral stenosis.

Physical signs of aortic regurgitation are unmistakable, but with cursory and casual examination – one may miss them.

Investigation

Chest X–Ray

Left ventricular dilatation produces cardiomegaly on chest radiograph. Ascending aorta is often prominent. Egg shell calcification of ascending aorta suggests Leutic etiology while widened mediastinum makes one suspect aortic dissection. Valve calcification is better seen on fluoroscopy. With left ventricular failure obvious radiological signs may emerge. Mild or even moderate aortic regurgitation may produce no radiological signs.

Electrocardiogram

Electrocardiogram shows left ventricular dominance with voltage criteria for left ventricular hypertrophy. In moderate aortic regurgitation prominent q-waves are seen in lateral leads with iso-electric ST-segment. As the severity of AR increases and LV dilates, LVH with strain pattern appears with ST-segment depression and T-wave inversion in lateral leads. Conduction blocks may be seen in patients with post inflammatory AR or due to severe calcific aortic valve disease.

Echocardiogram

Echocardiography is done to confirm the diagnosis of aortic regurgitation, evaluate the cause of AR and assess the aortic valve and severity of aortic regurgitation. It is also used to assess the left ventricular size and function.

Diastolic flutter of anterior mitral leaflet and premature closure of mitral valve in severe, often acute, aortic regurgitation are noted. Dilated chamber with increased contractility suggesting volume overload pattern of left ventricle is seen. 2 D guided M-mode measurements should be made to record LV dimensions, mass and ejection fraction. It should be noted that dimension measurements can vary due to intra and inter observer variability, interim changes in loading conditions, instrumentation factors and physiological variability. Reliability is increased by comparing with previous recordings. Whenever changes are noted, it is prudent to repeat examination after a shorter interval. On the basis of 2 D Doppler and color flow imaging, severe aortic regurgitation can be differentiated from mild aortic regurgitation by following parameters:

Table 3.5: Comparision between severe AR and mild AR

Parameter	Severe AR	Mild AR
Regurgitant Jet width/LVOT diameter	$\geq$ 60 per cent	$\leq$ 30 per cent
Regurgitant Jet area/LVOT area	$\geq$ 60 per cent	$\leq$ 30 per cemt

Aortic regurgitant pressure half time	≤ 250 msec	≥ 400 msec
Mitral filling pattern	Restrictive	Normal
Flow reversal in descending aorta	Holodiastole	Early diastolic
Continuous Wave Doppler signal	Dense	Faint
Regurgitant Fraction	≥ 50 per cent	≤ 30 per cent
LV End Diastolic diameter	≥ 75 mm	≤ 60 mm
Effective regurgitant orifice	≥ 0.30 cm^2	≤ 0.1 cm^2

When expertise is available MRI gives equal information as obtained with echocardiographic examination. This is specially suitable in patients with poor echo windows.

Fig. 3.9: Appearance of valve and aortic root for deciding etiology of AR

Exercise Testing

It is helpful in assessing functional capacity, symptom status and hemodynamic effects in patients with severe AR with good LV function and equivocal symptoms or when patient leads sedentary life. Exercise ejection fraction, when abnormal may predict, poor long term out come. However, it is not clear if it has any independent additional predictive valve over and above LV size and function. It is not particularly indicated in patients with normal LV systolic function without severe LV dilatation.

Radionuclide ventriculography may be used in place of echocardiography where echo window is not suitable to assess LV size and function satisfactorily or when the echocardiographic data is inconclusive. It is also useful in serial follow up of patients.

Cardiac Catheterization

Often cardiac catheterization is not indicated as reliable information for management decision making can be obtained through non-invasive methods. Before Aortic valve replacement (AVR)

patient may have coronary angiography if indicated to rule out coronary artery disease. Similarly, cardiac catheterization or angiography may be done if there is any discrepancy between clinical findings and echocardiographic data. Hemodynamic measurements during exercise are occasionally helpful for determining the effect of aortic regurgitation on left ventricular function. In borderline cases right heart pressures on exercise may clarify the indications for AVR.

Natural History

In asymptomatic patients with normal LV function, the rate of progression to symptoms and/or LV Systolic dysfunction is about 4 per cent per year. Average mortality rate is 0.2 per cent per year. 1.3 per cent patients per year developed LV Systolic dysfunction without symptoms.

Systolic dysfunction precedes onset of symptoms about 25 per cent of patients. Serial follow up based only on symptom status evaluation is not reliable and quantitative evaluation of left ventricle is needed. Age, end systolic dimension or volume, end diastolic volume or dimensions are good indicators of onset of LV dysfunction, symptoms and/or death. Patients with end systolic dimension of > 50 mm have a likelihood of death, symptom onset and/or LV dysfunction of about 20 per cent year. In those with end systolic dimension between 40-50 mm the likelihood is about 6 per cent per year and in those with less than 40 mm it is zero. Patients with asymptomatic LV systolic dysfunction develop symptoms within 2-3 years at a rate of about 25 per cent per year. In the presurgical era, data has shown annual mortality rate of > 10 per cent in patient with angina and > 20 per cent in patients with heart failure. Symptomatic patient have poorer prognosis even when they have preserved LV function while mild AR in an elderly hypertensive may not need any serious follow up. Same is not true in an young patient with mild AR due to bicuspid aortic valve or aortic root dilatation.

Management

Vasodilators improve stroke volume, and reduce degree of regurgitation. This results from decrease in systemic vascular resistance and leads to reduction in left ventricular end diastolic volume, after load, wall stress and LV mass. They also preserve LV systolic function. Hydralazine and nefedipine when given at appropriate doses decrease left ventricular size and improve LV function. The dose should be titrated to bring down systolic blood pressure as much as possible without getting side effects. Less consistent results were obtained with ACE inhibitors and increased plasma renin activity is not an issue in these patients. Vasodilators may be given in any patient with severe AR with LV dysfunction and symptoms of heart failure when patient is a poor candidate for surgery. They may also be given to a patient with severe AR and LV dysfunction with or without symptoms to improve hemodynamic profile before surgery. They are also indicated in a patient with severe AR and normal LV function. Here they delay the onset of LV dysfunction, symptom onset and postpone the time for AVR and also improve operative outcomes. In this situation there is no proven role of any other medication. Vasodilators should not be used as a substitute for aortic valve replacement in a patient with severe AR who needs surgery either due to LV dysfunction or symptoms. They are also not indicated in patients with mild AR and normal sized left ventricle with good LV function. Following AVR, ACE inhibitors may be better choice in patients with persistent myocardial systolic dysfunction.

Serial Testing

Serial testing is indicated in asymptomatic patients with severe AR and preserved LV function since LV dysfunction may precede the onset of symptoms. The rationale is to identify those who develop symptoms, LV systolic dysfunction and rapid and progressive increase in LV size, when AVR becomes indicated. Patients with mild AR and normal LV size and systolic function need annual follow up or whenever signs and symptoms of worsening occur. Patients with severe AR

and LV dilatation (LVEDD > 60 mm) require follow up once in 6 – 12 months while those with LVEDD > 70 mm or LVESD > 50 mm need more frequent follow up. Also patients with aortic root dilatation need more frequent follow up. Patients with change in effort intolerance should have prompt evaluation. When the symptoms change in equivocal or uncertain, exercise testing may be done. Otherwise it is not indicated. Radio nuclide or MRI may be done when the echo window is sub-optimal and there is discrepancy between clinical and echocardiographic findings. Chest X-ray and ECG may have additional value to echo in some patients.

Surgical Therapy

The recommendation for Aortic valve replacement is only for those patients with Severe AR. If patients with mild AR have LV dilatation, dysfunction and symptoms–other causes need to be evaluated and ruled out. If there is uncertainty about the severity of AR–it needs to confirmed with angiographic or any other modality.

In patients with Severe AR with good LV function and symptoms of NYHA Class III–IV or angina Canadian class II or more, surgery is indicated. In patients with NYHA Class II dyspnoea exercise testing is useful. If onset of dyspnoea is recent or patient's LV size and LV function reach threshold values, i.e., LVEF < 50 per cent or LVEDD about 75 mm or LVESD about 55 mm – surgery is indicated.

AVR is indicated in all symptomatic patients NYHA Class II, III, IV and systolic dysfunction with EF 25 – 49 per cent. In patients with EF < 25 per cent and LVESD more than 60 mm surgery carries high risk. LV dysfunction may be irreversible and post operative morbidity is high. Patients with LV dysfunction do well if the duration of LV dysfunction is short or show improvement with intense vasodilatation, inotropic or diuretic therapy. Clinical judgement should be based on the fact that medical management alone carries very high mortality.

In patients with severe AR and no symptoms, LVEF < 50 per cent, LVEDD > 75 mm or LVESD > 55 mm are indications for surgery. Two consecutive measurements may be obtained in short duration before proceeding with surgery or findings may be confirmed in these asymptomatic patients, by 2 different modalities. AVR is also indicated in patients approaching these LV dimension if they have progressive increase in LV size or any evidence of decreasing exercise tolerance. These dimensions should be tailored to patient's body size but no guidelines are available. Patients with concomitant CAD, Hypertension or other valve disease are also likely to be more symptomatic even with smaller LV size.

Indications for surgery should not be based on the operative techniques used.

When aortic root dilatation is equal to or more than 50 mm root reconstruction should accompany aortic valve replacement.

Severity of pre-operative symptoms or reduced exercise tolerance, severity of left ventricular systolic dysfunction and duration of LV dysfunction–all predict post-operative survival and recovery of LV systolic function.

Acute Aortic Regurgitation

Infective endocarditis, aortic dissection and trauma often produce severe AR. Acute increase in left ventricular end diastolic volume especially in a small pressure loaded hypertrophied heart increases left ventricular diastolic pressure and causes pulmonary edema. In the absence of compensatory mechanisms forward stroke volume is markedly decreased and patient develops cardiogenic shock. Tachycardia ensues and pulse pressure is low due to low cardiac output. In such a sick patient physical findings are difficult to appreciate. First sound is muffled. Early diastolic murmur is short and soft. Apical diastolic rumble may be present. There are no

peripheral signs of chronic severe AR. Physical examination and Chest X–ray do not show any cardiomegaly.

Echocardiogram clinches the diagnosis. Severity and mechanism of AR and associated lesions will be known. Typically acute Severe AR causes short pressure half time of aortic regurgitant jet < 300 m sec., short deceleration time of early mitral filling wave < 150 m sec. and premature closure of mitral valve.

Acute severe AR is a surgical emergency. Pre-operatively patient may be stabilized with inotropes and vasodilatation like nitroprusside. IABP and beta blockers are contraindicated. Disease as well as treatment is associated with high mortality.

Check Your Progress 4

1) Name two causes of acute onset aortic regurgitation.

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2) Name four common causes of chronic aortic regurgitation.

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3) Name three common causes of aortic regurgitation caused by diseases of the aorta.

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4) What are the symptoms of aortic regurgitation?

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5) What are the clinical signs of chronic severe aortic regurgitation?

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6) What are some of the drugs used in moderate aortic regurgitation?

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7) What are the indications for surgery in asymptomatic severe aortic regurgitation?

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Tricuspid Valve Disease

Tricuspid valve disease is relatively uncommon compared to other valvular lesions. Tricuspid Regurgitation is more common than tricuspid stenosis and is often secondary rather than primary.

3.6 TRICUSPID STENOSIS

It is rare disease, with rheumatic etiology seen in 90 per cent of cases. In patients with rheumatic mitral stenosis only 3-5 per cent have concomitant tricuspid stenosis. Milder degrees of organic tricuspid valve involvement, up to 15-30 per cent are noted in autopsy series and in echocardiogram. Also tricuspid stenosis progresses much more slowly and develops clinical features on an average a decade later than mitral stenosis.

Unusual causes of tricuspid stenosis include carcinoid disease, congenital anomalies, infective endocarditis, Whipple's Disease and right atrial myxoma.

Pathophysiology

Obstruction to the tricuspid valve results in increased right atrial pressures, systemic venous congestion with right heart failure and low cardiac output state.

Clinical Findings

Since tricuspid stenosis, which is most often rheumatic, coexist with mitral stenosis, it is difficult to differentiate symptoms of one disease from other. In general, patient will have complaints of dyspnoea, fatigue and peripheral edema. Jugular venous pressure is characterized by prominent "a" wave and slow "y" descent in sinus rhythm. Absence of 'y' trough is noted in atrial fibrillation. Diastolic murmur generated across tricuspid valve has a distinct crescendo-decrescendo shape–a finding accentuated in 1^{o} heart block. It is typically located at lower left sternal border and increases with inspiration. Tricuspid opening snap is difficult to appreciate.

Fig. 3.10: Mid-diastolic murmur-tricuspid stenosis

ECG

AF is seen 50 per cent of cases. Right Atrial enlargement is obvious in those with sinus rhythm. Conduction abnormalities like 1^{0} heart block may be obvious.

Chest X-ray

Chest X-ray shows prominent right atrium and inconspicuous pulmonary artery.

Echocardiography

Findings are subtle. One should carefully search for organic tricuspid valve involvement in all rheumatic heart disease patients. Findings include thickened tricuspid valve leaflets with doming motion, commissural fusion and chordal thickening.

2D echo helps in determining the etiology and nature of tricuspid valve involvement. Doppler findings are very helpful. Normal velocities across tricuspid valve are less than 1m/sec with a mean gradient of less than 2 mmHg. Tricuspid stenosis is considered significant when mean gradient is $\geq$ 7 mmHg or PHT is $\geq$ 190 ms. It is better to measure at held expiration to avoid respiratory variation. Accurate Doppler evaluation excludes the need for cardiac catheterization.

Hemodynamic Measurements

Unless one suspects it clinically and echocardiographically, and plans the hemodynamic study—diagnosis of tricuspid stenosis is easily missed at routine catheterization. Prominent 'a' wave in right atrial pressure trace in a patient with sinus rhythm should make one suspect tricuspid stenosis. Pull back gradient especially in a patient with atrial fibrillation can easily miss the gradient. One should place two catheters simultaneously in RA and RV and measure the diastolic gradient. These small gradients can be exaggerated by exercise.

Management

Medical therapy is ineffective. With diuretics, symptoms of congestion are replaced by that of low cardiac output state. Occasional cases of balloon tricuspid valvuloplasty are reported. Since tricuspid stenosis is often accompanied by regurgitation and mitral valve disease simultaneous surgical correction with comissurotomy and tricuspid annuloplasty is performed. When tricuspid valve is destroyed badly, tricuspid valve replacement may be needed. In general it is avoided in view of poor long term results. Unrecognized and uncorrected rheumatic tricuspid stenosis can have deleterious effects on the results of mitral and aortic surgery. It is worth repeating that 10 per cent of patients with mitral valve disease and up to 50 per cent of patients with combined mitral and aortic valve disease have associated organic tricuspid valve disease. A diligent echocardiography examination avoids major post operative embarrassment.

3.7 TRICUSPID REGURGITATION

Though tricuspid regurgitation is a common valvular abnormality on echocardiography rarely is it due to primary organic disease.

Secondary or functional tricuspid regurgitation due to pulmonary hypertension of any cause, is much more common than primary tricuspid regurgitation. In these instances tricuspid valve structure is normal except for dilated tricuspid annulus. Right ventricular and right atrial dilatation further exaggerates this abnormality.

Primary causes or organic involvement of tricuspid regurgitation as opposed to its functional involvement are much less common. The common etiologies include rheumatic, traumatic, endocarditis, carcinoid, endomyocardial fibrosis and myxomatous valve prolapse. Rheumatic etiology is often accompanied by involvement of mitral valve. Similarly myxomatous degeneration is associated with mitral valve prolapse. Infective endocarditis is typically seen in drug addicts, but it is more commonly described with septic abortions and contaminated intra venous infusions in India. Endomyocardial fibrosis affecting right ventricular apex as well as mitral valve is confined to certain geographical regions. Tricuspid Regurgitation due to blunt injury chest can have a delayed presentation. Conduction abnormalities are often associated with traumatic tricuspid regurgitation. Ebstein's anomaly, a congenital heart disease is often associated with varying degrees of tricuspid regurgitation. Carcinoid valve disease is rare, and tricuspid

valvular involvement is associated with hepatic metastasis only. Pulmonary valve also is often involved in this disease.

Pathophysiology

Tricuspid regurgitation is associated with prominent venous filling waves and elevated right atrial venous pressures. Hepatic and systemic venous congestion and low output state would result as a consequence. In the primary tricuspid regurgitation, right ventricular pressure is normal and the diseased tricuspid valve or right ventricle is the cause for tricuspid regurgitation. Since right ventricle can tolerate high volumes, it enlarges and symptoms would be manifested at an advanced stage of the disease. Secondary tricuspid regurgitation is due to elevated pressures in right ventricle, which is more commonly due to elevated pulmonary artery pressures. Clinical features are predominantly due to disease processes causing elevation of pulmonary artery pressures.

Fig. 3.11: Systolic murmur of tricuspid regurgitation

Clinical Features

Patients with severe primary tricuspid regurgitation will have symptoms of fatigue, dyspnoea and effort intolerance, abdominal fullness and distension. In the absence of left heart diseases leading to tricuspid regurgitation, orthopnoea and PND do not occur. Physical examination will show elevated mean jugular venous pressure with prominent ‘v’ waves and sharp v-y collapse in the absence of tricuspid stenosis. Pulsatile hepatomegaly is often noted. Pan systolic murmur of tricuspid regurgitation is heard at lower left sternal border and it typically increases with inspiration due to increased venous return. In about 20 per cent of patients the murmur may not be audible. Lateral head bobbing is sometimes obvious at bedside. This is in contrast to vertical head bobbing that may be seen in aortic regurgitation. The height of ‘v’ waves depends upon the compliance and size of right atrium and may not exactly reflect severity of regurgitation. Systolic murmurs of low pressure TR tend to be early systolic in contrast to those of high pressure TR. This is because in the former, right atrial and right ventricular systolic pressure tend to equalize in early systole and regurgitation stops by mid systole. Intensity of murmur does not correlate with severity of regurgitation. Musical murmurs of TR may produce loud murmurs. In inspiration TR murmur typically increase due to increased venous return and this is called Carvello’s sign. This may not be seen in patients with right heart failure since failing right ventricle’s output cannot be increased further with inspiration.

Investigations

ECG in secondary TR shows evidence of right atrial overload and right ventricular hypertrophy with right axis QRS deviation. Chest X-ray shows evidence of right atrial enlargement and right ventricular type of cardiomegaly.

Echo Doppler Evaluation

Echocardiographic examination clarifies the diagnosis, etiology and severity of tricuspid regurgitation. Right atrial and ventricular dilatation depend upon severity of TR. High pressure TR may be associated with near normal size of RA and RV. Severe organic TR may be present with normal RV systolic pressure. In volume overload of RV, IVS shows a paradoxical motion in late systole while in pressure overload of RV, IVS motion is paradoxical in early systole. Inter atrial septum may be pushed to left side. There is no simple and reliable formula to calculate right ventricular ejection fraction and one has to depend upon suggestive visual impression. Color flow imaging shows the severity of TR and continuous wave Doppler imaging measures the TR jet velocity from which RV-RA systolic pressure gradient is calculated. Addition of estimated RA pressure to this gradient gives right ventricular systolic pressure. The following criteria suggest severe tricuspid regurgitation: A color flow regurgitant jet area > 30 per cent RA area, dense continuous wave Doppler signal annulus dilatation with incomplete leaflet coaptation, increased tricuspid inflow velocity [E-wave >1.0 m/sec], systolic flow reversal in the hepatic vein. Trivial to mild TR may be seen in echocardiogram of up to 65 per cent of normal subjects and these are of no clinical significance.

Fig. 3.12: Colour Doppler across tricuspid valve demonstrating severe tricuspid regurgitation

The etiology of TR is often clarified with echocardiogram. In Tricuspid Valve Prolapse it is often the myxomatous leaflets that prolapse, and is often present along with mitral valve prolapse. Ebsteins Anomaly has a characteristic appearance of attachment of septal leaflets. Carcinoid valve disease has typical appearance with shortened and thickened leaflets resulting in large area of incomplete coapatation. Rheumatic TR may be primary or secondary to thickened shortened leaflets with annular dilatation and associated mitral valve involvement is obvious. In the absence of pulmonary metastasis or PFO-left sided valves are not involved in carcinoid. With rheumatic etiology these are always involved.

Haemodynamics

Right Atrial pressure tracing shows a prominent 'v' wave and these cannot be appreciated in a patient with atrial fibrillation. RV angiography is never done to diagnose or assess severity of tricuspid regurgitation.

Natural History

Prognosis of the tricuspid regurgitation depends upon underlying cause, as most of the time tricuspid regurgitation is functional or associated with other valvular lesions. In-patients with severe mitral stenosis it is an independent predictor of poor late outcome. The 4 year survival rates for mild TR is 99 per cent, 90 per cent for moderate and 69 per cent for sever tricuspid regurgitation. In long term patients with severe tricuspid regurgitation develop signs and symptoms of systemic venous congestion and low output state.

Management

It depends upon the cause of tricuspid regurgitation. Specific therapy is directed towards the particular cause of pulmonary artery hypertension, which results in tricuspid regurgitation. Diuretics should be used with caution to relieve symptoms of venous congestion. Mean jugular venous pressure rather than top of 'v' wave should direct diuretic usage since inappropriate high dosage would result in low output state.

Tricuspid valve annuloplasty is often considered to decrease the severity of tricuspid regurgitation if the patient is undergoing surgery for associated left heart disease. This tends to reduce the severity of regurgitation but does not eliminate it. In patients with severe deformation replacement is needed. Complete heart block is an important potential complication. 5 and 10 year survival rates range from 55-80 per cent and 36-50 per cent respectively. Since mechanical valve have high risk of thrombogenicity, tissue prosthesis are preferred. However, in young patients and those who need anticoagulants for other reasons, mechanical valve may be considered.

Check Your Progress 5

1) Name three causes of primary tricuspid regurgitation.

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2) What are the clinical signs of tricuspid regurgitation?

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3.8 LET US SUM UP

Rheumatic fever is one of the important cause of mitral stenosis. It clinically present with exertional dysnea and having mid diastolic rumbling murmur. Ballon valvuplasty or close mitral valvulomy, open mitral valvotomy, mitral volume replace ment are the choice of treatment depending on the condition of disease. Mitral regurgitation is the most commom valvular disease observe in clinical practice. Mitral valve repair or mitral valve replacement is the treatment of

choice in all symptomatic patients (class 11 and above) with severe mitral regurgitation and normal or decreased left nontricular function. Angina occur 50-60 per cent of the aostic stenosis patient and crescendo decrescendo ejection systolic murmur can best heard in the second right intercostal space. Primary aortic valve disease or primary aortic root disease are main cause of aortic regurgitation. Patient will present dyspnea, followed by arthopnea, paronymal nocturnal dyspnear and excessive deaphoresis, water-hammer pulse, palpitation and decrecendo diastolic murmur. Tricuspid valve disease are relatively uncommon.

3.9 ANSWERS TO CHECK YOUR PROGRESS

Check Your Progress 1

1) The longer the diastolic murmur, the more severe the mitral stenosis.

   The shorter the A2-OS interval, the more severe the mitral stenosis.

2) The structures that enlarge in mitral stenosis are:

   The left atrium and appendage

   The pulmonary veins resulting in signs of pulmonary venous hypertension with perihilar venous congestion and Kerley lines.

   Enlargement of the pulmonary artery and main branches with peripheral narrowing due to pulmonary artery hypertension.

   Enlargement of the right ventricle (best seen in the lateral view) due to pulmonary hypertension and later tricuspid regurgitation.

   Enlargement of the right atrium from tricuspid valve disease.

   There can be marked enlargement of the left atrium and the right atrium.

Check Your Progress 2

1) Congenital

   Cleft leaflet

   Post Inflammatory

   Acute rheumatic fever

   Chronic rheumatic Heart disease.

   Post infective

   Infective endocarditis

   Degenerative

   Myxomatous Mitral valve

   Dilated cardiomyopathy

   Coronary artery disease

   Papillary muscle dysfunction.

   Remodeled Left ventricle after Infarction

2) Left ventricle is often dilated with a downward and laterally displaced forcible apex. Occasionally systolic thrill of mitral regurgitation is palpable. First heart sound (S1) is usually soft in rheumatic mitral regurgitation but it is normal in mitral valve prolapse. Second heart sound (S2) may be widely split. A third heart sound (S3) may be audible at the apex. A fourth heart sound (S4) may be seen with recent onset severe mitral regurgitations and sinus rhythm. A holosystolic murmur starting with S1 and ending with S2 due to mitral regurgitation is audible at apex. In mitral valve prolapse it is a mid systolic murmur starting after a mid systolic click. Murmur radiates to axilla and back with a posteriorly directed jet as seen with anterior leaflet abnormalities, ischaemic and dilated cardiomyopathies. It radiates superiorly and medially towards base with posterior leaftlet abnormalities. Patients with severe mitral regurgitation due to valve pathology have loud and long murmurs while soft, short, barely audible early murmurs are present in patients with functional mitral regurgitation. Murmur is often not audible in patients with acute mitral regurgitation. Physical maneuvers like valsalva, squatting and respiration will help in differentiating it form other systolic murmurs. Mid diastolic murmur may follow an S3 especially in rheumatic mitral regurgitation and is unusual in mitral regurgitations of other etiologies.

3) Severe mitral regurgitation is diagnosed when

   1) There is echocardiographic evidence of disruption of mitral valve apparatus like papillary muscle rupture, flail mitral leaflet etc.,
   2) The effective regurgitant orifice area is $\geq$ 0.40 $cm^{2,}$
   3) Mitral regurgitation volume > 60 cc,
   4) Regurgitant fraction $\geq$ 55 per cent,
   5) Pulmonary vein systolic flow reversal and
   6) Mitral regurgitation jet reaches posterior wall of the left atrium.

   Severe mitral regurgitation is suggested when

   1) The color flow area is $\geq$ 40 per cent of LA size,
   2) Eccentric mitral regurgitation jet reaches the posterior wall,
   3) Dense continuous wave Doppler signal is present,
   4) E-wave velocity is $\geq$ 1.5 m/s for native valves and $\geq$ 2.0 m/s for prosthetic valves,
   5) LV dimension is $\geq$ 7 cm,
   6) LA size is $\geq$ 5.5 cm

Check Your Progress 3

1) Symptoms include dizziness or syncope, angina and those of heart failure. Dizziness or syncope may be due to arrhythmias or LV dysfunction but more often due to inappropriate baroreceptor function that results in vasodilatation in the presence of fixed cardiac output.

   Angina is noted in approximately 50 – 60 per cent of patients and in about 50 per cent of these patients it is due to associated CAD. Onset of heart failure symptoms usually portends poor prognosis.

2) Typically the carotid pulse in severe aortic stenosis is delayed in upstroke and of small volume. The apical impulse is forcible and sustained and with decompensation of left

ventricle it is displaced. A palpable S_4 is common in severe aortic stenosis. Absence of a valvular ejection click in children should make one suspect the diagnosis of non valvular aortic stenosis. Usually S_2 is single due to muffled A_2. If the valve is pliable with less extent of calcification, it may produce paradoxical split in a patient with severe valvular aortic stenosis.

Typically the murmur of valvular aortic stenosis starts after S_1, is crescendo decrescendo ejection systolic murmur with maximum intensity at second right inter costal space and radiates to carotids.

In children and elderly selective conduction of high frequency sounds to apical area may give risk to cooing murmur at apex (Gallavardin phenomenon).

3) In patients with normal LV function and cardiac output, aortic stenosis is considered severe when peak aortic velocity is more than or equal to 4.5 m/sec; mean pressure gradient equal to or more than 50 mmHg, aortic valve area less than or equal to 0.75 cm^2 or LVOT/Aortic valve VTI ratio less than or equal to 0.25.

Check Your Progress 4

1) Aortic dissection

   Trauma

   Leaflet perforation due to endocarditis

2) Congenital - bicuspid

      Unicuspid

      Quadricuspid

      Fenestrations

   Rheumatic

   Infective endocarditis

   Myxomatous degeneration

   Fibro calcific degeneration

   Systemic Lupus Erythematosus

3) Hypertension

   Idiopathic root dilatation

   Marfan’s Syndrome

   Aortic dissection

   Annuloaortic ecstasia

   Cystic medial necrosis

   Syphilis

   Ankylosing spondylosis

4) Patients with chronic severe AR may be asymptomatic for many years and it may be picked up on routine examination.

   Excertional dyspnoea occurs as the left ventricle end diastolic pressure increases. Impaired coronary blood flow results in angina, sometimes occurring in the nights and awakening the patient from sleep (nocturnal angina). As the systolic dysfunction worsens, patients may have dyspnoea at rest, paroxysmal nocturnal dyspnoea, and even frank heart failure. Angina is more pronounced in patients with Leutic or Takayasu aortitis with coronary ostial disease. Patients may also complain of palpitation due to forcible ventricular contraction. Rarely sudden death may occur in patients with severe aortic regurgitation.

5) Chronic severe aortic regurgitation is characterized by wide pulse pressure and multiple peripheral signs it produces.

   Sharp rapid upstroke of radial pulse followed by rapid down stroke is called Corrigan's pulse or water hammer pulse and is exaggerated by elevating the wrist. Carotids have two prominent systolic impulses called bisferiens pulse. There may be palpable thrill in carotids due to rapid ejection of increased stroke volume.

   Due to rapid run off of blood from aorta in diastole and increased stroke volume, varieties of physicial signs are described. The prominent pulse waves may be audible over brachial and other peripheral arteries and are called pistol shot sounds. These interfere with measurement of diastolic blood pressure. In significant aortic regurgitation diastolic blood pressure is usually less than 70 mmHg and systolic blood pressure is elevated causing wide pulse pressure.

   Usually cardiac enlargement is present and left ventricular hyper dynamic apex is palpable outside mid clavicular line. S_1 is normal or slightly reduced in intensity. A_2 is louder in aortic regurgitation due to aortic root disease while it is muffled in valvular aortic regurgitation. Left ventricular S_3 suggest left ventricular dysfunction.

   Characteristic murmur is early diastolic murmur which is high pitched and blowing. It is best heard in left parasternal border third left intercostals space at end expiration with patient sitting and bending forward. A murmur that is better heard on the right side of the sternum suggests aortic root disease. Ejection systolic murmur due to increased systolic flow across aortic valve is common and its presence does not necessarily mean aortic stenosis. At apex one may additionally hear Austin Flint (mid diastolic) murmur. This may be due to aortic regurgitation jet pushing the anterior mitral leaflet up and causing relative mitral stenosis or just audibility of low pitched vibrations of aortic regurgitation itself. This needs to be differentiated from associated mitral stenosis.

6) Vasodilators improve stroke volume, and reduce degree of regurgitation. This results from decrease in systemic vascular resistance and leads to reduction in left ventricular end diastolic volume, after load, wall stress and LV mass. They also preserve LV systolic function. Hydralazine and nefedipine when given at appropriate doses decrease left ventricular size and improve LV function. The dose should be titrated to bring down systolic blood pressure as much as possible without getting side effects.

   Less consistent results were obtained with ACE inhibitors and increased plasma renin activity is not an issue in these patients. Vasodilators may be given in any patient with severe AR with LV dysfunction and symptoms of heart failure when patient is a poor candidate for surgery. They may also be given to a patient with severe AR and LV dysfunction with or without symptoms to improve hemodynamic profile before surgery. They are also indicated in a patient with severe AR and normal LV function. Here they

delay the onset of LV dysfunction, symptom onset and postpone the time for AVR and also improve operative outcomes. In this situation there is no proven role of any other medication. Vasodilators should not be used as a substitute for aortic valve replacement in a patient with severe AR who needs surgery either due to LV dysfunction or symptoms. They are also not indicated in patients with mild AR and normal sized left ventricle with good LV function. Following AVR, ACE inhibitors may be better choice in patients with persistent myocardial systolic dysfunction.

7) In patients with severe AR and no symptoms, LVEF < 50 per cent, LVEDD > 75 mm or LVESD > 55 mm are indications for surgery.

Check Your Progress 5

1) The common etiologies include rheumatic, traumatic, endocarditis, carcinoid, endomyocardial fibrosis and myxomatous valve prolapse.

2) Physical examination will show elevated mean jugular venous pressure with prominent ‘v’ waves and sharp v-y collapse in the absence of tricuspid stenosis. Pulsatile hepatomegaly is often noted.

Pan systolic murmur of tricuspid regurgitation is heard at lower left sternal border and it typically increases with inspiration due to increased venous return.