Valvular Heart Disease

Cardiac murmurs

• Cardiac murmurs are often the first sign of underlying valvular disease.
• May be systolic or diastolic, pathological or benign.
• Systolic murmurs may be due to physiological increases in blood velocity or might indicate as yet asymptomatic cardiac disease.
• Diastolic murmurs are almost always pathological and require further evaluation.
• An ECG and CXR, although readily available tests, provide limited diagnostic information.

Echocardiography

Echocardiography can evaluate valve function by the following imaging modalities:

• 2-D: Valve motion and morphology; LV size and function.
• Doppler: Blood flow velocity; valve gradients; haemodynamic data.
• Colour flow: Valvular regurgitation

Management of cardiac murmurs

• Murmur + cardiac symptoms: refer to cardiologist.
• In asymptomatic patients with cardiac murmurs, an echo is indicated in the following instances:
  - Murmur + abnormal cardiac signs
  - Murmur + abnormal ECG or CXR
  - Diastolic or continuous murmurs
  - Pansystolic or late systolic murmurs

Aortic stenosis

Aetiology

• Aortic stenosis may be congenital or acquired.
• Congenital malformations may be tricuspid, bicuspid or unicuspid.
• Acquired causes include the following:
  - Degenerative disease
  - Rheumatic disease
  - Calcific e.g. end-stage renal failure, Paget’s disease
  - Miscellaneous e.g. rheumatoid involvement

Aetiology of aortic stenosis

A: Normal aortic valve
B: Congenital aortic stenosis
C: Rheumatic aortic stenosis
D: Calcific aortic stenosis
E: Degenerative aortic stenosis
Aortic stenosis

Grading and severity

• Aortic valve area must be reduced to 25% of normal before significant circulatory changes occur.
• Grading of stenosis severity is as follows:
  - Normal valve area = 3-4cm²
  - Mild stenosis = 1.5-3cm²
  - Moderate stenosis = 1.0-1.5cm²
  - Severe stenosis ≤ 1.0cm²
• When stenosis is severe, the peak gradient across the aortic valve is usually > 60mmHg.

Pathophysiology of aortic stenosis

Aortic stenosis

LV outflow obstruction

LV systolic pressure

Aortic pressure

LV hypertrophy

LV dysfunction

Myocardial ischemia

LV failure

Natural history of aortic stenosis without operative treatment:

Aortic stenosis

Physical findings

• Slow rising pulse
• Reduced systolic and pulse pressure
• Systolic thrill over the aortic area
• Ejection systolic, crescendo-decrescendo murmur
• Soft or inaudible second heart sound
• ECG: LVH, AV node conduction defects

Aortic stenosis

Medical therapy

• Conservative treatment should be offered for mild to moderate aortic stenosis and to asymptomatic patients with severe aortic stenosis as follows:
  - Advise to report symptoms
  - Avoid vigorous exercise
  - Antibiotic prophylaxis for endocarditis
  - Regular follow-up ± echocardiography

Aortic stenosis

Surgical therapy

• Aortic valve replacement should be offered to the following:
  - Symptomatic pts with severe AS
  - Pts with severe AS undergoing CABG surgery
  - Pts with moderate AS undergoing CABG surgery
  - Asymptomatic pts with severe AS and LV dysfunction
• Balloon valvuloplasty can play a temporary role as a bridge to surgery in haemodynamically unstable patients, or as palliation for patients with serious comorbid conditions
Aortic stenosis

Aortic valve replacement

- In the absence of LV dysfunction, operative risk is 2-8%.
- Indicators of higher mortality are NYHA class, LV dysfunction, age, concomitant coronary artery disease, and aortic regurgitation.
- Valve replacement usually results in reduced LV volumes, improved LV performance and regression of LV hypertrophy.

Aortic regurgitation

Aetiology

- Either due to primary disease of the aortic valve or wall of the aortic root or both.
- Causes of primary aortic valve disease include:
  - Congenital eg. bicuspid aortic valve
  - Acquired: rheumatic valve disease, infective endocarditis, trauma, connective tissue disease.
- Causes of primary aortic root disease include:
  - Degenerative, cystic medial necrosis (eg. Marfan’s), aortic dissection, syphilis, connective tissue disease, hypertension.

Pathophysiology of aortic regurgitation

- LV volume (↑)
- Stroke volume (↑)
- Diastolic BP (↑)
- LV mass (↑)
- Systolic BP (↓)
- LV dysfunction
- Myocardial ischaemia
- LV failure

Clinical history

- In chronic severe AR, the left ventricle gradually enlarges while the patient remains asymptomatic. Symptoms usually develop after cardiomegaly and LV dysfunction have occurred. Dyspnoea is the principal complaint. Syncope is rare and angina is less frequent than in aortic stenosis.
- In acute severe AR, LV decompensation occurs readily with fatigue, severe dyspnoea and hypotension.

Physical findings

- Collapsing pulse.
- Wide pulse pressure due to both raised systolic blood pressure and reduced diastolic blood pressure.
- Displaced, diffuse and hyperdynamic apex beat.
- Early blowing diastolic murmur.
- ECG: Left axis deviation, LV hypertrophy.
- CXR: Cardiomegaly, aortic calcification, aortic root dilatation.

Management

- Medical treatment includes:
  - Diuretics, digoxin, salt restriction
  - Vasodilators
  - Endocarditis prophylaxis
- Indications for surgical treatment depend on symptoms, and LV size and function.
- Without surgery, death usually occurs within 4 years of developing angina and within 2 years after onset of heart failure.
Aortic regurgitation

Surgical therapy

- Severe acute AR requires prompt surgical intervention.
- Indications for valve replacement in pure, severe, chronic AR include:
  - Symptomatic patients with normal LV function
  - Symptomatic patients with LV dysfunction or dilatation
  - Asymptomatic patients with LV dysfunction or dilatation (EF < 50% or end-systolic diameter > 55mm)
- Aortic valve and root replacement are indicated in patients with disease of the proximal aorta and AR of any severity when the aortic root diameter is ≥ 50mm.

Mitral stenosis

Aetiology

- Rheumatic fever is the predominant cause.
- Rarely, mitral stenosis is congenital and observed almost exclusively in infants and young children.
- Miscellaneous rare causes include carcinoid, SLE, rheumatoid arthritis and mucopolysaccharidoses.
- Causes of left atrial outflow obstruction that may simulate mitral stenosis include left atrial myxoma, ball-valve thrombus, infective endocarditis with large vegetation and cor triatriatum.

Pathophysiology

- Normal mitral valve area = 4-6cm².
- Usually, a mitral valve area ≤ 2.5cm² must occur before the development of symptoms.
- A mitral valve area >1.5cm² usually does not produce symptoms at rest.
- The first symptoms in mild mitral stenosis are usually precipitated by exercise, emotional stress, infection, pregnancy or fast atrial fibrillation.
- A mitral valve area ≤ 1cm² equates to severe mitral stenosis.
- Pulmonary hypertension results from backward pressure, pulmonary arteriolar constriction and organic obliterative changes in the pulmonary vascular bed.

Rheumatic mitral stenosis

- Rheumatic mitral stenosis is due to four forms of fusion: commissural (30%), cuspal (15%), chordal (10%) or combined (45%).
- The stenotic mitral valve is typically funnel-shaped; the orifice is frequently shaped like a fish mouth.
- Symptoms usually occur in the 3rd or 4th decade, but mild MS in the aged is becoming more common.
- 25% of patients with rheumatic mitral valve disease have pure mitral stenosis and two-thirds are female.
- May be associated with an atrial septal defect – Lutembacher’s syndrome.

Natural history

- Long latent period of 20 to 40 years from the occurrence of rheumatic fever to onset of symptoms.
- Once symptoms develop, there is a further 10 years before symptoms become disabling.
- Once significant limiting symptoms occur, the 10-year survival rate is 5-15%.
- When there is severe pulmonary hypertension, mean survival falls to < 3 years.
- Mortality from untreated mitral stenosis is due to progressive heart failure (60-70%), systemic embolism (20-30%) and pulmonary embolism (10%).
Mitral stenosis

Clinical features

- The main symptom is dyspnoea due to reduced lung compliance.
- Haemoptysis may also occur.
- Approximately 15% of patients experience angina due to either coincidental coronary artery disease, right ventricular hypertension or coronary embolisation.
- Embolic events may occur and 80% of such patients are in atrial fibrillation.

Physical findings

- Mitral facies – pinkish-purple patches on the cheeks.
- Tapping apex beat – palpable first heart sound.
- Right ventricular heave, loud P2 indicating pulmonary hypertension.
- Loud first heart sound.
- Opening snap.
- Rumbling, mid-diastolic murmur with presystolic accentuation in sinus rhythm.

Echo evaluation

- Assessment of valve morphology: degree of leaflet thickness, mobility and calcification and extent of subvalvular fusion.
- Estimation of left atrial size.
- Doppler echo: estimation of mitral valve area, transvalvular gradient and PA pressure.

Medical treatment

- The asymptomatic patient with mild mitral stenosis should be managed medically. Medical therapy includes:
  - Avoidance of unusual physical stress.
  - Salt restriction.
  - Diuretics if needed.
  - Control of heart rate – β-blocker or digoxin.
  - Anticoagulation for AF or prior embolic event.
  - Annual follow-up.
  - Echocardiography if deterioration in clinical condition.

Management of symptomatic mitral stenosis

- Patients with symptoms should undergo clinical re-evaluation with echocardiography.
- NYHA class II symptoms and mild mitral stenosis may be managed medically.
- NYHA class II symptoms and at least moderate stenosis (MVA ≤ 1.5 cm² or mean gradient ≥ 5 mmHg) may be considered for balloon valvuloplasty.
- NYHA class III or IV symptoms and severe mitral stenosis should be considered for balloon valvuloplasty or surgery.

Balloon mitral valvuloplasty

- The technique involves passing a balloon flotation catheter across the interatrial septum after trans-septal puncture and dilating the balloon within the mitral valve orifice.
- Results of the procedure are highly dependent on the experience of the operator.
- 80-95% of patients have a successful procedure.
- Complications include severe MR, residual ASD, myocardial perforation, emboli, MI and death.
- Overall event-free survival is 50% to 65% over 3-7 years.
- The underlying mitral valve morphology is the most important factor in determining outcome.
- Relative contraindications include the presence of a left atrial thrombus and significant mitral regurgitation.
Mitral stenosis

**Mitral valve replacement**

- Mitral valve replacement is indicated in patients with severe mitral stenosis and contraindications to surgical commissurotomy or balloon valvuloplasty.
  - Restenosis following surgical commissurotomy or balloon valvuloplasty.
  - Significant mitral stenosis and regurgitation.
  - Extensive calcification of the subvalvular apparatus.
- Operative mortality ranges from 3-8% in most centres.
- Postponement of surgery until the patient reaches NYHA class IV symptoms should be avoided.

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Mitral regurgitation

**Aetiology**

Mitral regurgitation may be caused by abnormalities of the valve leaflets, chordae tendineae, papillary muscles or mitral annulus:

- Valve leaflets
  - myxomatous degeneration causing mitral valve prolapse
  - shortening, rigidity, deformity and retraction due to rheumatic heart disease
  - vegetations due to infective endocarditis
- Chordae tendineae
  - congenital, infective endocarditis, trauma, rheumatic fever, myxomatous
- Papillary muscles
  - myocardial ischaemia, congenital abnormalities, infiltrative disease
- Mitral annulus
  - dilatation eg. ischaemic or dilated cardiomyopathy
  - calcification due to degeneration, hypertension, aortic stenosis, diabetes, chronic renal failure

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Mitral regurgitation

**Clinical features**

- Symptoms usually occur with LV decompensation: dyspnoea and fatigue.
- Physical findings include:
  - Pulse: sharp upstroke
  - Apex: displaced, hyperdynamic
  - Heart sounds: pansystolic murmur loudest at the apex, radiating to the axilla and accentuated by expiration.

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Mitral regurgitation

**Natural history**

- The natural history of chronic MR depends on the volume of regurgitation, the state of the myocardium and the underlying cause.
- Preoperative LV end-systolic diameter is a useful predictor of postoperative survival in chronic MR.
- The preoperative LV end-systolic diameter should be < 45mm to ensure normal postoperative LV function.

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Mitral regurgitation

**Medical treatment**

- Symptomatic patients may benefit from the following drug therapy whilst awaiting surgery:
  - Vasodilator therapy
  - Diuretics
  - Digoxin / Beta-blockers in presence of atrial fibrillation.
  - Endocarditis prophylaxis

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Mitral regurgitation

**Surgical treatment**

- Surgery is indicated in the presence of symptoms or left ventricular end systolic diameter ≥45mm.
- The surgical procedure consists of either mitral valve repair or replacement.
- Mitral valve repair better preserves LV function and avoids the need for chronic anticoagulation.
- However, mitral valve repair is technically more demanding and often not possible to perform in severely deformed valves.
Mitral regurgitation

Acute mitral regurgitation

Aetiology

Important causes of acute mitral regurgitation include:
- Infective endocarditis causing disruption of valve leaflets or chordal rupture.
- Ischaemic dysfunction or rupture of papillary muscle.
- Malfunction of prosthetic valve.

Chronic versus Acute MR

<table>
<thead>
<tr>
<th>Finding</th>
<th>Chronic MR</th>
<th>Acute MR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms</td>
<td>subtle onset</td>
<td>obvious</td>
</tr>
<tr>
<td>Appearance</td>
<td>normal/mildly</td>
<td>severely ill</td>
</tr>
<tr>
<td>Tachycardia</td>
<td>not striking</td>
<td>always present</td>
</tr>
<tr>
<td>Apex beat</td>
<td>displaced</td>
<td>not displaced</td>
</tr>
<tr>
<td>Systolic thrill</td>
<td>common</td>
<td>absent</td>
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<tr>
<td>Murmur</td>
<td>harsh pansystolic</td>
<td>soft or absent</td>
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<tr>
<td></td>
<td></td>
<td>systolic component</td>
</tr>
<tr>
<td>ECG-LVH</td>
<td>usually present</td>
<td>absent</td>
</tr>
<tr>
<td>CXR</td>
<td>severe cardiomegaly</td>
<td>normal heart size</td>
</tr>
</tbody>
</table>

Medical therapy

The following therapies may be beneficial in reducing the severity of MR:
- Vasodilator therapy
- Inotropic therapy
- Intra-aortic balloon counterpulsation

Surgical therapy

- Indicated in patients with acute severe MR and heart failure.
- Higher mortality rates than for elective chronic MR, but unless treated aggressively, the outcome is usually fatal.

Mitral valve prolapse

General features

- Occurs in 2-6% of the general population
- Twice as common in women.
- Due to myxomatous proliferation of the mitral valve.
- Usually occurs as a primary condition, but may be a secondary finding in connective tissue diseases e.g. Marfan’s syndrome.
- Vast majority of patients are asymptomatic.
- Symptoms may include palpitations, dizziness, syncope, or chest discomfort.
- The principal physical finding is the mid-systolic click, followed by a late systolic murmur in the presence of regurgitation.

Echocardiographic criteria

- M-mode criterion: ≥ 2mm posterior displacement of one or both leaflets.
- 2-D echo findings: Systolic displacement of one or both leaflets within the left atrium in the parasternal long-axis view; leaflet thickening, redundancy, chordal elongation and annular dilatation.
- The diagnosis of mitral valve prolapse is even more certain when leaflet thickness is >5mm.
- Echocardiography is useful in the risk stratification of patients with mitral valve prolapse.
Mitral valve prolapse

Natural history

- In most patients, mitral valve prolapse is associated with a benign prognosis.
- Complications may occur in patients with a systolic murmur, thickened leaflets an increased LV or LA size, especially in men > 45 years old:
- Complications include progressive mitral regurgitation, infective endocarditis, cerebral emboli, arrhythmias and rarely sudden death.

Management

- Asymptomatic patients without MR or arrhythmias have an excellent prognosis – follow-up every 3-5 years.
- Patients with a typical systolic murmur should receive endocarditis prophylaxis.
- Patients with a long systolic murmur may show progression of MR and should be reviewed annually.
- Patients with previous embolic events should be given antiplatelet / anticoagulant therapy.
- Severe MR requires surgery, often mitral valve repair.

Prosthetic valves

Prosthetic valves may be divided into 2 broad categories:

Mechanical valves
- Very good durability.
- Require long-term anticoagulation.
- May cause mild haemolysis.

Bioprosthetic (tissue) valves
- Porcine variety most commonly used.
- Limited durability.
- Anticoagulation for first 3 months only.

Mechanical valves

Designs and flow patterns of different types of mechanical valves

Tricuspid stenosis

- Always rheumatic in origin and when present accompanies mitral valve involvement.
- The anatomical changes and physiological principles are similar to those of mitral stenosis.
- The low cardiac output state causes fatigue; abdominal discomfort may occur due to hepatomegaly and ascites.
- The diastolic murmur of tricuspid stenosis is augmented by inspiration.
- Medical management includes salt restriction and diuretics.
- Surgical treatment should be carried out in patients with a valve area <2.0cm² and a mean pressure gradient >5mmHg.
### Tricuspid regurgitation
- Most common cause is annular dilatation due to RV failure of any cause; may also be caused by intrinsic valve involvement.
- Well tolerated in the absence of pulmonary hypertension; in the presence of pulmonary hypertension, cardiac output declines and RV failure may worsen.
- Symptoms and signs result from a reduced cardiac output, ascites, painful congestive hepatomegaly and oedema.
- The pansystolic murmur of TR is usually loudest at the left sternal edge and augmented by deep inspiration.
- Severe functional TR may be treated by annuloplasty or valve replacement. Severe TR due to intrinsic tricuspid valve disease requires valve replacement.

### Pulmonary stenosis
- Most commonly due to congenital malformation and usually an isolated anomaly.
- Survival into adulthood is the rule, infective endocarditis is a risk and right ventricular failure is the most common cause of death.
- Rheumatic involvement of the pulmonary valve is very uncommon and rarely leads to serious deformity.
- Carcinoid plaques may lead to constriction of the pulmonary valve ring.

### Pulmonary regurgitation
- Most common cause is ring dilatation due to pulmonary hypertension, or dilatation of the pulmonary artery secondary to a connective tissue disorder.
- May be tolerated for many years unless complicated by pulmonary hypertension.
- The clinical manifestations of the primary disease tend to overshadow the pulmonary regurgitation.
- Physical examination reveals a right ventricular heave and a high-pitched, blowing, early diastolic decrescendo murmur over the left sternal edge, augmented by deep inspiration.
- Pulmonary regurgitation is seldom severe enough to require specific treatment. Surgery may be required because of intractable RV failure.