Valvular heart disease poses a common yet difficult problem in everyday clinical practice. A thorough clinical evaluation with basic common investigations such as an electrocardiogram (ECG) and a chest radiograph (CXR) remains the cornerstone of diagnosis. Echocardiography with colour flow and Doppler (not focus­
ated) plays a pivotal role in confirming the diagnosis, and assessing the severity of the valve lesions and concomitant pulmonary hypertension, other valve lesions and haemodynamic consequences.

Clinical evaluation (history and examination) remains the cornerstone of screening for valve pathology. An electrocardiogram (ECG) and a chest radiograph (CXR) are seen as important adjuncts to clinical evaluation and may provide important diagnostic clues to confirming pathology.

Clinical features and special investigation findings are described in Table 1.[2] Patients with mild disease and symptoms may require diuretic therapy and sodium restriction to reduce congestion. Beta-blockers are often prescribed, the rationale being that reducing the heart rate increases diastolic filling time, reduces the gradient and improves effort tolerance. It is generally used in patients with Class II - III symptoms. There is no evidence for its prognostic benefit or use in multivalve disease.[10]

Verapamil may also be used for heart rate control, but should not be administered concomitantly with beta-blockers. There is no benefit from the use of angiotensin-converting enzyme (ACE) inhibitors. Digoxin may be used for heart rate control in patients with AF, but is otherwise contraindicated.

Anticoagulation with vitamin K antagonists (warfarin) is advised in patients with previous embolic events, clot in the LA or AF (regardless of the presence of LA clot). Aspirin is not used, as its benefit in stroke prevention is low, with a similar bleeding risk to warfarin. The CHADS2­VASc score should not be applied to patients with MV disease and AF; as the stroke risk is already high, these patients should be anticoagulated. The new oral anticoagulants have not been tested in this setting and should not be used.

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Mitrval stenosis

Mitrval stenosis (MS) is almost exclusively caused by chronic rheumatic heart disease. The rheumatic process leads to inflam­
matin, resulting in commissural fusion, thickening and fibrosis of both the leaflets and subvalvalur apparatus. Fewer than half of patients with MS recollect an episode of acute rheumatic fever. Other rare causes of mitral valve (MV) obstruction include congenital MS, degenerative mitral annular calcification, atrial myxomas, large thrombus or vegetations, systemic lupus erythematosus (SLE) and carcinoid syndrome.

A normal MV area is 4 - 6 cm2. During diastole, the MV opens to allow the unobstructed flow of blood into the left ventricle. With MS, there is obstruction to ventricular inflow; the resultant pressure gradient causes an increase in left atrial (LA) pressure. Pulmonary oedema usually occurs secondary to this rise in pressure.

Any condition that increases heart rate, including sepsis, thyroid disease, anaemia, atrial fibrillation (AF) and pregnancy, may precipitate symptoms.

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For patients with moderate to severe disease or for those with an episode of acute pulmonary oedema, definitive surgical treatment should be considered and they should be referred for evaluation.

ARTICLE

An approach to the diagnosis and management of valvular heart disease

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The diagnosis of valvular heart disease is a difficult problem in everyday clinical practice. There is a wide spectrum of presentation – in some cases murmurs are found incidentally and in others, patients present very late with dire haemodynamic consequences of neglected valve lesions that may preclude them from definitive surgery. This review serves as a guide to the primary care clinician in the diagnosis and management of valve disease.

With the decline in rheumatic heart disease and the ageing population in the developed world, there has been a change in the disease patterns of valve lesions over the last few decades. Western populations are experiencing greater numbers of degenerative valve disease. In the developing world, however, rheumatic heart disease remains an important cause of valve pathology. Sliwa et al.[1] in the Heart of Soweto Study, showed an incidence of new cases of rheumatic heart disease of 23.5/100 000 cases per annum.

Clinical evaluation (history and examination) remains the corner­stone of screening for valve pathology. An electrocardiogram (ECG) and a chest radiograph (CXR) are seen as important adjuncts to clinical evaluation and may provide important diagnostic clues to confirming pathology.

Echocardiography with colour flow and Doppler (not focus­
ated) plays a pivotal role in confirming the diagnosis, and assessing the severity of the valve lesions and concomitant pulmonary hypertension, other valve lesions and haemodynamic consequences.

Invasive testing with cardiac catheterisation is reserved for patients in whom there is a discrepancy between clinical findings and echocardiography.

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matin, resulting in commissural fusion, thickening and fibrosis of both the leaflets and subvalvalur apparatus. Fewer than half of patients with MS recollect an episode of acute rheumatic fever. Other rare causes of mitral valve (MV) obstruction include congenital MS, degenerative mitral annular calcification, atrial myxomas, large thrombus or vegetations, systemic lupus erythematosus (SLE) and carcinoid syndrome.

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For patients with moderate to severe disease or for those with an episode of acute pulmonary oedema, definitive surgical treatment should be considered and they should be referred for evaluation.
Mitral stenosis

**History**
- Exertional dyspnoea
- Orthopnoea
- Paroxysmal nocturnal dyspnoea
- Acute frank pulmonary oedema
- Haemoptyysis
- Embolism: 20 - 25%, especially cerebral
- Chest discomfort
- Hoarseness: Ortner’s syndrome

**Physical findings**
- Normal character, small volume pulse in severe MS
- Irregularly irregular pulse: atrial fibrillation
- JVP may be elevated: prominent cv wave if concomitant significant TR
- Undisplaced, tapping apex beat
- Loud first heart sound
- Loud pulmonary component of S2 if pulmonary hypertension
- Opening snap after S2, if pliable valve
  - The closer to S2, the more severe the MS
- Low-pitched mid-diastolic murmur
  - Best heard at the apex
  - Longer murmur = more severe MS
  - If patient is in sinus rhythm, presystolic accentuation of the murmur may be audible
- Pulmonary crepitations at lung bases
- Other associated valve lesions: mitral regurgitation, tricuspid disease, aortic disease

**Special investigations**
- ECG
  - Usually unremarkable
  - In sinus rhythm, LA enlargement: broad P wave in lead II or a predominantly negative deflection in lead V1
  - In AF, no comment on LA size can be made
- CXR
  - Normal-sized heart (ventricle)
  - Upper-lobe blood diversion/interstitial oedema
  - Enlarged LA seen by:
    - Straight left heart border
    - Splaying on the coryna
    - Double shadow on the right heart silhouette

**Table 1. Clinical and special investigation features of mitral stenosis**

**Table 2. Clinical and special investigation features of mitral regurgitation**

Percutaneous balloon mitral valvuloplasty has superseded surgical valvotomy (closed and open) for patients with pliable valves.\(^1\) This is also the treatment of choice for pregnant MS patients with pliable valves and poor response to medical therapy. Those with calcified, non-pliable valves or significant concomitant mitral regurgitation (MR) are referred for valve replacement surgery.

Mitral regurgitation

MR may be classified depending on the clinical presentation (acute or chronic) or leaflet pathology (functional versus organic).

Acute MR is a medical emergency presenting with acute pulmonary oedema and hypotension and is usually caused by endocarditis, myocardial infarction with papillary muscle rupture or spontaneous rupture of the chordae. Afterload reduction with intravenous nitrates or nitroprusside and early surgical intervention is usually required.

The clinical and investigational features of chronic MR are summarised in Table 2. Signs of severity include clinical features of heart failure, pulmonary hypertension, a loud murmur grade ≥3/6 and presence of a third heart sound in the absence of heart failure.

Functional MR is characterised by normal leaflets and is secondary to a dilated and dysfunctional left ventricle. Echocardiography is required to accurately confirm this diagnosis and may differentiate ischaemic from non-ischaemic causes. Functional MR requires optimisation of heart failure therapy, whereas moderate or severe ischaemic MR may require surgery with possible coronary revascularisation.

Organic MR refers to primary leaflet abnormality resulting in MR. The cause is most often rheumatic in South African (SA) patients, whereas degenerative or myxomatous disease is most common in the developed world. Other causes include fibro-elastic disease, congenital (isolated cleft or atrioventricular (AV) canal defect), Marfan syndrome and endocarditis. Medical therapy has been shown not to be of benefit in MR. Indications for surgery include symptoms, development of AF, pulmonary hypertension, end-systolic diameter >40 mm and ejection fraction <60% on echocardiography. Surgical options include MV replacement or repair, which may mandate earlier surgery, provided a successful durable repair can be obtained.

Aortic stenosis

Aortic stenosis (AS) is the most common valve lesion in western countries and mainly a disease of the elderly. Common causes of AS include:
- degenerative trileaflet AS
- degenerative bicuspid AS
- rheumatic heart disease – there would usually be concomitant MV disease
- other (rare): congenital AS, Paget’s disease, end-stage kidney disease, chronic inflammatory diseases.

Haemodynamically significant obstruction usually occurs when the valve area is <1 cm². Left ventricular hypertrophy develops in response to the progressive obstruction. Risk factors for degenerative AS are similar to those for atherosclerosis, i.e. hypertension, diabetes, dyslipidaemia and smoking.
tricuspid regurgitation (TR) or a combination of TR and TS. Isolated patients with rheumatic tricuspid valve (TV) disease present with transcatheter aortic valve replacement is currently a possibility.[7] Tricuspid stenosis (TS) is rare and usually rheumatic in origin.[11] Most tricuspid valve disease being a bridge to surgery or in those too ill for surgery.[2,8­10]

Table 3. Clinical and special investigation features of aortic stenosis

Aortic stenosis

<table>
<thead>
<tr>
<th>History</th>
<th>Physical findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exertional dyspnoea</td>
<td>Small-volume, slow-rising pulses</td>
</tr>
<tr>
<td>Angina</td>
<td>Narrow pulse pressure</td>
</tr>
<tr>
<td>Syncope</td>
<td>JVP normal, unless heart failure or MV disease</td>
</tr>
<tr>
<td></td>
<td>Pressure-loaded undisplaced apex beat</td>
</tr>
<tr>
<td></td>
<td>Soft or single second heart sound</td>
</tr>
<tr>
<td></td>
<td>Crescendo-decrescendo ejection systolic murmur at base of the heart</td>
</tr>
<tr>
<td></td>
<td>Radiated to carotids</td>
</tr>
<tr>
<td></td>
<td>Longer murmur = more severe</td>
</tr>
<tr>
<td></td>
<td>High-pitched widely radiating murmur: Gallavardin effect – can be mistaken for MR</td>
</tr>
<tr>
<td></td>
<td>Systolic click in bicuspid valve may be heard</td>
</tr>
</tbody>
</table>

Special investigations

<table>
<thead>
<tr>
<th>ECG</th>
<th>Normal-sized heart (ventricle)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Aortic calcification</td>
</tr>
<tr>
<td></td>
<td>Post-stenotic dilatation: especially in bicuspid valves</td>
</tr>
</tbody>
</table>

Patients are asymptomatic for many years. Once symptoms occur, however, there is a rapid decline in life expectancy.[14]

Clinical features of AS are shown in Table 3.

There is no place for medical management of patients with symptomatic AS. It is a mechanical obstruction for which the definitive treatment is aortic valve replacement. These patients should therefore all be referred promptly for assessment for valve replacement surgery,[16] which prolongs and improves quality of life, even in octogenarians.[17] For those in whom the risk of surgery is too high, transcatheter aortic valve replacement is currently a possibility.[18]

Aortic regurgitation

Aortic regurgitation may occur as a result of leaflet pathology or secondary to aortic root pathology. Acute regurgitation is poorly tolerated and constitutes a medical and surgical emergency. It is commonly caused by infective endocarditis or aortic root dissection. Chronic regurgitation is well tolerated and patients are often asymptomatic for many years. Common causes of primary valve lesions are rheumatic heart disease, infective endocarditis, congenital bicuspid valves, and rheumatoid arthritis. Conditions primarily affecting the root, and hence causing regurgitation, are Marfan’s syndrome, syphilis, sero-negative spondyloarthritides, aortic dissection and osteogenesis imperfecta.

The clinical features are summarised in Table 4. Symptomatic aortic regurgitation requires referral for the assessment for aortic valve replacement. There is no place for medical therapy outside of it being a bridge to surgery or in those too ill for surgery.[24-34]

Table 4. Clinical and special investigation features of aortic regurgitation

Aortic regurgitation

<table>
<thead>
<tr>
<th>History</th>
<th>Physical findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long asymptomatic period</td>
<td>Collapsing pulses</td>
</tr>
<tr>
<td>Dyspnoea</td>
<td>Wide pulse pressure (difference between systolic and diastolic is &gt;50% of the systolic pressure)</td>
</tr>
<tr>
<td>Orthopnoea</td>
<td>Duroziez sign</td>
</tr>
<tr>
<td>Paroxysmal nocturnal dyspnoea</td>
<td>Heart failure signs: elevated JVP, leg swelling, crepitations at lung bases</td>
</tr>
<tr>
<td>Nocturnal angina</td>
<td>Early diastolic murmur at base of heart</td>
</tr>
<tr>
<td></td>
<td>Best heard with patient sitting forward, in end-expiration</td>
</tr>
<tr>
<td></td>
<td>Longer murmur = more severe</td>
</tr>
<tr>
<td></td>
<td>A systolic murmur (due to increased flow or concomitant AS)</td>
</tr>
<tr>
<td></td>
<td>An Austin-Flint murmur (late diastolic apical murmur)</td>
</tr>
</tbody>
</table>

Special investigations

<table>
<thead>
<tr>
<th>ECG</th>
<th>Cardiomegaly</th>
</tr>
</thead>
<tbody>
<tr>
<td>No specific diagnostic changes</td>
<td>Pulmonary congestion</td>
</tr>
<tr>
<td>Occasionally in severe AR-left ventricular hypertrophy, and left axis deviation may be seen</td>
<td></td>
</tr>
<tr>
<td>CXR</td>
<td></td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td></td>
</tr>
</tbody>
</table>

Prosthetic valves

There are two types of prosthetic valves: mechanical valves and bioprosthetic (tissue) valves. The major differences between the two relate to risk of thromboembolism (higher with mechanical valves) and structural deterioration (higher with bioprostheses).[34] Mechanical valves are classified into three groups: bileaflet, tilting disc and ball-cage. Bileaflet mechanical valves are most commonly implanted. Patients with mechanical valves require long-term anticoagulation.

The risk of thromboembolism is about 6 times higher without anticoagulants, and the risk of de novo valve thrombosis is also
higher. Warfarin is the anticoagulant of choice and the international normalised ratio should be between 2.5 and 3.5. Antiplatelet agents such as aspirin do not provide adequate protection and are not recommended without the use of anticoagulants.

Bioprostheses were developed to overcome the challenges of long-term anticoagulation and increased risk of thromboembolism associated with mechanical valves. A stented tissue valve consists of three tissue leaflets mounted on a ring with semi-rigid stents that facilitate implantation. Because stents add to obstruction and increase stress on the leaflets, stentless tissue valves were developed for the aortic position and are particularly useful for patients with small aortic roots. More recently, a transcatheter bioprosthesis has been developed, which can be implanted via a catheter at the aortic valve position. Homograft aortic valves are harvested from cadavers, sterilised with antibiotics and cryopreserved at −196°C for long periods before implantation. Pulmonary autografts (Ross procedure) involve removal of a patient’s native pulmonary valve and reimplantation to replace the diseased aortic valve. Bioprostheses were developed to overcome the challenges of long-term anticoagulation and increased risk of thromboembolism associated with mechanical valves. A stented tissue valve consists of three tissue leaflets mounted on a ring with semi-rigid stents that facilitate implantation. Because stents add to obstruction and increase stress on the leaflets, stentless tissue valves were developed for the aortic position and are particularly useful for patients with small aortic roots. More recently, a transcatheter bioprosthesis has been developed, which can be implanted via a catheter at the aortic valve position. Homograft aortic valves are harvested from cadavers, sterilised with antibiotics and cryopreserved at −196°C for long periods before implantation. Pulmonary autografts (Ross procedure) involve removal of a patient’s native pulmonary valve and reimplantation to replace the diseased aortic valve.[15] Homograft aortic valves are harvested from cadavers, sterilised with antibiotics and cryopreserved at −196°C for long periods before implantation. Pulmonary autografts (Ross procedure) involve removal of a patient’s native pulmonary valve and reimplantation to replace the diseased aortic valve.[15] Homograft aortic valves are harvested from cadavers, sterilised with antibiotics and cryopreserved at −196°C for long periods before implantation. Pulmonary autografts (Ross procedure) involve removal of a patient’s native pulmonary valve and reimplantation to replace the diseased aortic valve. Bioprostheses were developed to overcome the challenges of long-term anticoagulation and increased risk of thromboembolism associated with mechanical valves. A stented tissue valve consists of three tissue leaflets mounted on a ring with semi-rigid stents that facilitate implantation. Because stents add to obstruction and increase stress on the leaflets, stentless tissue valves were developed for the aortic position and are particularly useful for patients with small aortic roots. More recently, a transcatheter bioprosthesis has been developed, which can be implanted via a catheter at the aortic valve position. Homograft aortic valves are harvested from cadavers, sterilised with antibiotics and cryopreserved at −196°C for long periods before implantation. Pulmonary autografts (Ross procedure) involve removal of a patient’s native pulmonary valve and reimplantation to replace the diseased aortic valve.

**Conclusion**

The clinical evaluation remains essential in establishing a diagnosis in patients presenting with heart failure due to valvular heart disease. Awaiting results of special investigations should not hinder timeous referral. Those with symptomatic valvular heart disease should be referred promptly for specialist evaluation with a view to definitive surgical correction.

**Table 5. Clinical and special investigation features of tricuspid valve disease**

<table>
<thead>
<tr>
<th>Tricuspid stenosis</th>
<th>Tricuspid regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>History</strong></td>
<td><strong>History</strong></td>
</tr>
<tr>
<td>Progressive fatigue, oedema, anorexia</td>
<td>Well tolerated in absence of pulmonary hypertension, often asymptomatic</td>
</tr>
<tr>
<td>Minimal orthopnoea and paroxysmal nocturnal dyspnoea</td>
<td>Right heart failure (swollen abdomen, swelling of legs and painful, congestive enlargement of liver)</td>
</tr>
<tr>
<td>Pulmonary oedema and haemoptysis are rare</td>
<td>Throbbing pulsations in the neck (from elevated JVP) and eyeballs</td>
</tr>
<tr>
<td><strong>Physical findings</strong></td>
<td><strong>Physical findings</strong></td>
</tr>
<tr>
<td>Diastolic rumble at lower left sternal border, increasing in intensity with inspiration</td>
<td>Weight loss, cachexia, cyanosis and jaundice</td>
</tr>
<tr>
<td>Often confused with mitral stenosis</td>
<td>AF is common</td>
</tr>
<tr>
<td>Neck vein distention, with prominent a waves</td>
<td>Elevated JVP with prominent cv waves</td>
</tr>
<tr>
<td>Absent right ventricular lift/heave</td>
<td>Venous systolic thrill and murmur in neck</td>
</tr>
<tr>
<td>Hepatic pulsation</td>
<td>Tender hepatomegaly</td>
</tr>
<tr>
<td>Ascites, peripheral oedema</td>
<td>S3 gallop originating from RV</td>
</tr>
<tr>
<td>Associated murmurs of mitral and aortic valve disease</td>
<td>Loud P2 and parasternal heave if pulmonary hypertension present</td>
</tr>
<tr>
<td><strong>Special investigations</strong></td>
<td><strong>Special investigations</strong></td>
</tr>
<tr>
<td>ECG</td>
<td>ECG</td>
</tr>
<tr>
<td>Tall right atrial P waves and no RV hypertrophy</td>
<td>Usually nonspecific; incomplete RBBB, Q waves in V1, AF are common</td>
</tr>
<tr>
<td>CXR</td>
<td>CXR</td>
</tr>
<tr>
<td>Dilated RA without enlarged pulmonary artery segment</td>
<td>Marked cardiomegaly</td>
</tr>
</tbody>
</table>

**References**