VALVULAR HEART DISEASE

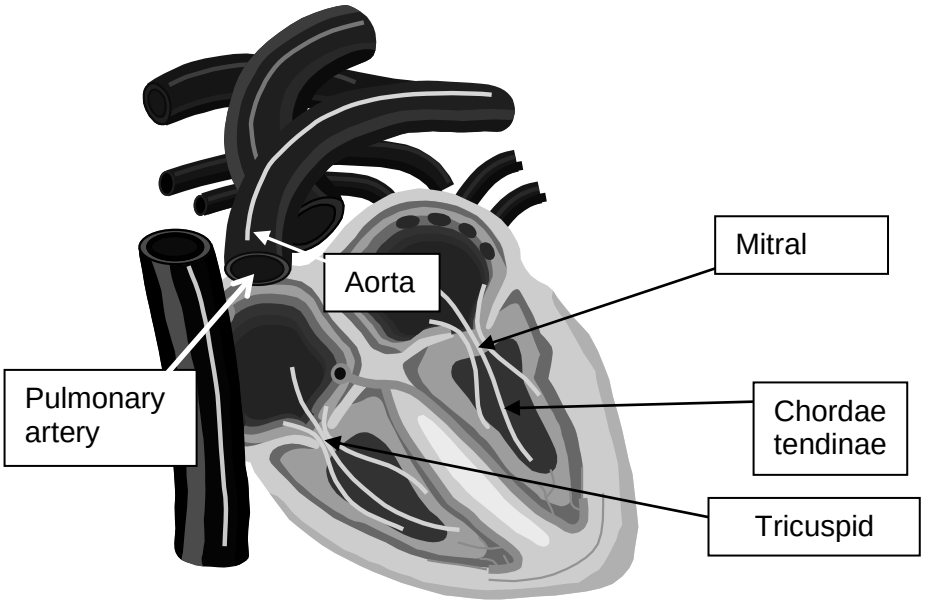
1 Introduction

1.1 Definition

Valvular Heart Disease is due to a structural abnormality in one or more of the four heart valves. (see fig. 1) This leads to valvular inefficiency in the cardiac pump cycle. This may result in disability due to cardiac failure and reduced cardiac output.

The inefficiency may be due to increased resistance to flow through a narrowed valve - stenosis, and/or leakage back through an inadequately closed valve - regurgitation.

Figure 1.



Roughened irregularities on the surface of the damaged or abnormal valves cause turbulent blood flow. This can be heard as a cardiac murmur.

The turbulent and slowed blood flow can lead to the formation of thromboses, which may embolise to the lung or the systemic circulation.

The roughened valve surface may become infected, leading to endocarditis and worsening of the condition due to further structural damage.

2 Description

2.1 Aetiology

Valvular heart disease may be congenital, acquired through infection, or degenerative.

Congenital:

As well as directly causing morbidity, congenital abnormalities may predispose the valves to acquired and degenerative changes.

Each year, the number of adults and adolescents with congenital heart disease increases by 7% as a result of successful cardiac surgery in early life. An estimated 80% of people with congenital heart disease will survive to at least adolescent age. Of these 75% will have had surgery.

The range of possible abnormalities is wide. Mild congenital cardiac valve abnormalities may produce no symptoms or disabling effects until one of the following occurs.

- Myocardial dysfunction
- Infective endocarditis
- Embolic event
- Arrhythmia

Some cases of congenital valve disease have complex and inoperable conditions. Post-natal adaptive changes may allow survival past childhood. This small group is likely to include individuals with severe cardiac disability and cyanosis.

More cases of congenital heart disease have acyanotic rather than cyanotic congenital heart disease. Many abnormalities are mild, with no disabling effects.

Of the general population, 1-2%, have a congenital anomaly of the aortic valve. A bicuspid aortic valve may cause only mild obstruction, and consequently will not present clinically until the 7th or 8th decade, when the valve has calcified.

In general, calcification occurs sooner in males, and where there is greater initial obstruction.

Acquired valve disease:

Acquired valve disease as a consequence of infection is most commonly due to rheumatic fever. This results from an immunological response to cell membrane antigens of the Lancefield Group A streptococcus. The immunological response also affects other tissues such as joints, lungs, kidneys and skin.

Virulent organisms such as Staphylococcus aureus can affect normal heart valves, whereas less virulent organisms normally only affect valves that have previously been damaged, or have a congenital abnormality.

Infective endocarditis is a major complication of valvular heart disease. The infection worsens the already damaged valve and causes systemic illness. Overall, mitral valve prolapse is the commonest underlying valve defect. In the elderly, the aortic valve is more usually affected. However, of those diagnosed with infective endocarditis, 40% have no history of underlying heart disease. Currently, chronic rheumatic heart disease accounts for less than 50% of cases. Intravenous drug users are estimated to have an annual risk of 2-5% of developing endocarditis. In more than 50% of cases the infecting organism will be Staphylococcus aureus. Fungal endocarditis and multiple infections are both more common in the addict population.

Other, rarer, infections can also cause acquired valve disease.

Degenerative valve disease:

This occurs later in life and is associated with calcification of the valve leading to stenosis and regurgitation.

There may be sudden onset of valve dysfunction following myocardial infarction due to papillary muscle rupture.

2.2 Prevalence

In North America and Western Europe the incidence of rheumatic fever has significantly reduced in recent years.

However, this is not true globally, and in developing countries rheumatic heart disease still accounts for a large proportion of cardiac morbidity.

It is common in the Indian subcontinent and other parts of Asia.

Acute rheumatic fever is a disease of childhood with a peak incidence between the ages of 5 and 15 years. The development of symptomatic valvular dysfunction tends to occur after a period of 20 to 40 years and so the typical patient presents in the 4th or 5th decade.

Degenerative valve disease is now more common in the developed world than disease acquired through rheumatic fever.

Degenerative valve disease with calcification presents in later life and typically causes problems in the 7th and 8th decades.

The incidence of infective endocarditis annually is 22 cases per million in the UK, and 49 cases per million in the USA.

3 Diagnosis

The diagnostic table (**3.8**) details the specific features for each valve in turn.

3.1 Left sided valve disease.

Mitral valve prolapse causing mitral regurgitation is the most common type of valvular disorder. In the majority of cases it is asymptomatic. It may cause transient symptoms.

In general, left sided valve disease - mitral and aortic - causes symptoms of left ventricular failure and poor cardiac output. When the disease is severe the right side of the heart can be affected as well, and congestive cardiac failure ensues.

A combination of aortic stenosis with regurgitation, and mitral stenosis with regurgitation gives a mixed picture of signs and symptoms that echocardiography and other tests can elucidate.

Three characteristic symptoms of poor cardiac output are breathlessness, anginal chest pain, and effort syncope. Orthopnoea is another feature characteristic of left ventricular failure (LVF). Paroxysmal nocturnal dyspneoa occurs in the later stages of the disease. Basal crepitations and other signs of LVF may be apparent in these cases. Breathlessness on exertion may be observed at examination.

In addition to effort syncope due to inadequate cardiac output, syncope in calcific degenerative aortic stenosis may occur due to transient dysrrhythmias as the atrioventricular node may be affected by calcification.

Atrial fibrillation is common in mitral valve disease.

3.2 Right sided valve disease.

Tricuspid stenosis with regurgitation, and pulmonary stenosis with regurgitation are rarely single valve abnormalities. They usually accompany left sided valve disease or cardiac abnormalities.

In right sided valve disease signs and symptoms of right ventricular failure predominate - raised JVP, oedema, and hepatomegaly with fatigue.

3.3 Special Investigations

The single best investigation for valvular heart disease is Echocardiography with Doppler. This test is both diagnostic and assesses suitability for surgery.

Patients who have concomitant coronary disease still need to have cardiac catheterisation performed to ascertain coronary anatomy and perfusion.[1]

3.4 Innocent Murmurs

It is estimated that 8% of the population have what is termed an innocent or physiological murmur. The mechanism for this is thought to be benign turbulent blood flow in the right ventricular outflow tract. The murmur is usually ejection systolic in type, and well localised to the left sternal edge. An echocardiogram and Doppler will clarify the presence or absence of any significant structural abnormality.

3.5 Mitral Valve Prolapse

2.4% of the population have this condition in which the mitral valve leaflet protrudes into the left atrium. Recent research shows that there is no difference in long-term complications in patients with simple mitral valve prolapse and those without.[2] Symptoms of anxiety and palpitations are therefore unlikely to be directly due to the prolapse. Occasionally, the condition progresses to 'floppy valve'. Antibiotic prophylaxis is needed if there is more than a click audible clinically, or if Doppler imaging demonstrates mitral regurgitation. Mitral valve prolapse with demonstrable regurgitation is a focus for endocarditis.

3.6 Infective Endocarditis

Infective endocarditis is due to micro-organisms growing on valves or in an intracardiac abscess. It can be diagnosed pathologically by blood culture or histology of an infective vegetation.

It commonly presents as a non-specific illness of fatigue, low-grade fever, and myalgia.

Clinically it is diagnosed using the specific definitions of:

- Two major criteria or
- One major and three minor criteria or
- Five minor criteria.

The criteria are listed in the table in Appendix A.

If the condition has vascular phenomena of arterial emboli, septic pulmonary emboli or intracranial haemorrhage there may be disabling peripheral, pulmonary, or neurological symptoms. The worsening of the valve function can lead to heart failure.

3.7 Atrial Fibrillation and Embolic Events

Atrial fibrillation is a common complication of mitral valve disease, especially mitral stenosis. The diagnosis may be suggested by an abnormal pulse ('irregularly irregular') and confirmed by the presence of characteristic ECG changes.

Emboli can occur from atrial thromboses and commonly affect the cerebral circulation – causing TIA or stroke. Aortic valve disease can also lead to emboli which may cause amaurosis fugax (ischaemia causing transient blindness), TIA and stroke.

Prosthetic valves can produce thromboses and emboli.

Because of the high incidence of embolic events, anti-coagulation is indicated in mitral valve disease and for prosthetic valves.

3.8 Diagnostic table – a basic guide

Term	**Diagnostic criteria**		
	History/Symptoms	**Clinical examination**	**Investigation**
Aortic Stenosis	Angina, Exertional syncope, Dyspnoea. (LVF) Infective endocarditis, TIAs (amaurosis fugax is typical), Stroke.	Reduced amplitude pulse. Ejection systolic murmur.	Echocardiography (Doppler assessment). ECG shows LV hypertrophy and strain.
Aortic Regurgitation	Late presentation of LVF symptoms: Fatigue, Dyspnoea, Orthopnoea, Paroxysmal Nocturnal Dyspnoea.	"Collapsing" hyperdynamic pulse. Early diastolic murmur. Pulmonary oedema with basal crepitations in LVF.	Echocardiography. CXR shows left ventricular dilatation and may show pulmonary oedema in LVF.
Mitral Stenosis	Late presentation of LVF as above. +/- Palpitations from AF.	Malar flush. Pulse frequently AF. Heart sounds - Loud S1, Opening snap. Mid diastolic murmur.	Echocardiography + Doppler. (CXR and ECG may be unhelpful)
Mitral Regurgitation	Late presentation of LVF as above. +/- Palpitations from AF.	Pulse "jerky" or AF. Apex beat hyperdynamic +/- systolic thrill. Pansystolic murmur, 3rd heart sound.	Echo-Doppler.

Pulmonary Stenosis	Most commonly congenital. Usually asymptomatic. May have features of low cardiac output – exertional dyspnoea and syncope	Prominent ejection systolic murmur at upper left sternal edge.	Echocardiography. ECG shows Right Ventricular hypertrophy.
Pulmonary Regurgitation	Most commonly congenital.	/	Echocardiography.
Tricuspid Stenosis	Commonly part of mixed valve disease and Left sided symptoms predominate.	Signs of RVF - JVP↑ Oedema. Hepatomegaly. Mid-diastolic murmur.	Echocardiography.
Tricuspid Regurgitation	Most commonly "functional" as a result of right ventricular dilatation.	Signs of RVF as above. Pansystolic murmur.	Echocardiography.

4 Treatment

The natural history of valvular heart disease has been changed by the advent of earlier surgical intervention.

Prior to this, outcome was adversely affected by myocardial dysfunction brought on by chronic disease.

An important aspect of management is antibiotic prophylaxis for at-risk individuals undergoing routine minor surgical procedures e.g. dental extraction.

The following groups should be offered prophylaxis:

Those with:

- Congenital heart defects
- Previous history of rheumatic fever
- Heart murmurs (unless 'innocent')
- Known valvular disease
- Prosthetic valves
- Previous history of endocarditis

Anticoagulation therapy reduces the risk of embolic phenomena in mitral valve disease with atrial fibrillation and following valve replacement with artificial prosthetic valves. All cases of mitral valve disease should be anti-coagulated, unless contraindicated, as should all cases with prosthetic valves.[5]

4.1 Aortic Stenosis

Conservative management has little to offer symptomatic patients. Medication, such as diuretics or vasodilators may be detrimental as they reduce left ventricular function.

All patients will need antibiotic prophylaxis for dental or minor surgical interventions.

In asymptomatic patients 6-12 month follow-up is necessary, to enable appropriate surgical referral should symptoms develop or, ideally, before symptoms develop.

In asymptomatic patients, the need for surgery will be indicated by diagnostic tests (i.e. Echo/Doppler) looked at together with ventricular function.

Surgical procedures include valve replacement with mechanical or biological valves (useful in patients where anticoagulation may be undesirable).

Coronary bypass grafting can be usefully carried out at the same time if required.

Balloon valvuloplasty may be a more appropriate procedure for patients with co-existing serious conditions or the very elderly. This procedure, though low risk, is really only palliative in terms of outcome, giving symptomatic relief for 6-18 months.

4.2 Aortic Regurgitation

Conservative management is favoured as many patients have mild, asymptomatic disease. After 10 years, more than half the patients with untreated aortic regurgitation are still alive.

Prophylactic antibiotic therapy is needed even in mild cases.

Surgery can be delayed for 3-4 years in more severely affected cases by the use of an angiotensin-converting enzyme inhibitor.

Indications for valve replacement include evidence of left ventricular dilatation or reduced left ventricular function.

Regular review of asymptomatic patients is important as left ventricular function can deteriorate over a relatively short period of time (1-2yrs), without a change in symptoms.

4.3 Mitral Stenosis

Medical management is aimed at controlling atrial fibrillation if present, reducing the possibility of systemic thromboembolism, and reducing left atrial pressure. Antibiotic prophylaxis for minor surgical procedures and anti-coagulation therapy are necessary.

Surgical intervention should be considered in patients who respond poorly to medical management. A number of procedures can be offered, choice being driven by factors such as the patient's age, the structure of the valve and surgical resources. Procedures include balloon valvuloplasty, open mitral valvotomy and mitral valve replacement.

In young patients whose valves are stenosed but still mobile and noncalcified, the procedure of choice is balloon valvuloplasty. This procedure can also offer useful palliation to older patients.[1]

4.4 Mitral Regurgitation

Mild cases may require no treatment. Atrial fibrillation may need the usual medication to control the ventricular rate. Anticoagulants are recommended in patients with atrial fibrillation.

Surgical intervention should be considered in patients before deteriorating symptoms of fatigue and breathlessness develop.[4] Left ventricular dysfunction is shown by echocardiogram in an asymptomatic patient and operation prevents irreversible heart muscle damage. Reconstruction of the valve as opposed to replacement of the valve gives good long-term results.

4.5 Mixed Mitral Valve Disease

Treatment is as for mitral stenosis. If surgery is indicated valve replacement rather than valvotomy/valvuloplasty is the procedure of choice.

4.6 Tricuspid Valve Disease

Treatment of associated problems is necessary. Replacement of the tricuspid valve carries a high risk of valve thrombosis. This procedure is therefore avoided if possible.

4.7 Antibiotic Prophylaxis

Some operative procedures can cause transient bacteraemia. This can result in infective endocarditis of prosthetic valves and valves with pre-existing lesions. Antibiotic prophylaxis is therefore given to at-risk individuals before:

- Dental procedures
- Upper respiratory tract procedures
- Genito-urinary procedures
- Obstetric, gynaecological and gastro-intestinal procedures

A large dose of amoxicillin is given preoperatively, except in patients who have had penicillin in the previous month and those who are penicillin-allergic. The British National Formulary contains the latest recommendations for prophylactic therapy.

4.8 Anticoagulant therapy

Anticoagulants are used to prevent thrombi forming on prosthetic heart valves and to prevent embolisation in rheumatic heart disease and atrial fibrillation. Warfarin is the drug of choice.

5 Prognosis

Rheumatic Fever: Appropriate treatment of this condition has reduced mortality from 25% to 1% with 90% of hearts normal after 10yrs.

Following valve replacement mortality is now much reduced.[3]

Aortic Stenosis: survival rates and symptomatic results are good overall with studies typically showing 5yr survival of 70-90% and 10yr survival of 70-75% following valve replacement. Once symptomatic, 3yr survival drops to 25% without surgical treatment.

Aortic Regurgitation: Mortality following valve replacement mirrors that for aortic stenosis.

Mitral Stenosis: the prognosis of this progressive disease has improved greatly with surgical treatment. After valvotomy a patient may maintain good valve function for several decades (though this procedure will not arrest rheumatic changes/susceptibility to infective endocarditis.)

The life span of biological replacement valves is limited to about 10yrs in patients over the age of 21yrs (and much less in younger patients). If these valves are used then patients are likely to need repeat surgery.

Mitral Regurgitation: long term, mitral valve reconstruction gives better results compared with replacement - the early mortality being the same - 0-2%. Patients who have mitral regurgitation secondary to infarction/ischaemia do less well, having an early mortality of 10-20% post operatively.

Late Complications: there are a number of potential late complications following valve replacement. These include:

1. Infection - infective endocarditis.
2. Prosthetic dysfunction - this can take the form of either an acute life threatening event such as acute clot formation, or more insidious changes to the structure of the valve due to chronic clot formation.
3. Left ventricular disease - this is a significant cause of mortality and morbidity. It often presents with features of right ventricular disease rather than left.

6 Main Disabling Effects

Ideally, surgical treatment will be offered and carried out before the onset of disabling symptoms. The aim is to prevent the development of irreversible damage to heart muscle, and long term disability due to cardiac failure. Such individuals may well report little in the way of disability day to day prior to surgery.

A good outcome following a surgical procedure can be expected.

Recovery after replacement of either the aortic or mitral valves is usually rapid, with patients generally resuming fulltime work in 2-3 months after surgery.[6]

Many cases will be on anti-coagulants. The risk from bleeding is only slightly increased in these individuals. The estimated rate of serious bleeding being only 2%.[6] Anti-coagulation does not usually give rise to any functional impairment. Atrial fibrillation may cause disabling symptoms of palpitations and breathlessness. Atrial fibrillation reduces exercise tolerance [4] and careful history taking will clarify whether the symptoms cause limitation in activities.

Patients who have had embolic events should be assessed for the possible effects of the emboli. These may be physical and psychological/cognitive.

The role of rehabilitation in valvular heart disease is largely focussed on individuals who have undergone heart surgery. Prior to surgery, patients may have become deconditioned physically due to a limited exercise capacity and fatigue. Disabling effects will be ameliorated by a suitable exercise programme. Where mechanical prosthetic valves have been used, very vigorous activities may not be tolerated, due to fixed valve orifices.

In symptomatic cases, functional disability may occur as the result of:

Reduced exercise tolerance due to:

- Breathlessness
- Angina
- Fatigue

The main cause of disablement is therefore reduced effort tolerance from whatever cause.

There are two scales used to measure cardiac disability due to dyspnoea and angina - the New York Heart Association Classification (NYHA) and the Canadian Cardiovascular Society Classification (CCS). These may be quoted in consultant or GP factual reports. They are detailed in Appendix A.

The assessment of claimants is made from:

1. The History of Activities of Daily Living (Typical Day) taking variability into account.
2. Informal Observation of the claimant's activities at examination. Breathlessness on minor effort.

3. Clinical examination findings may show signs of cardiac failure corroborating the report of disabling symptoms.
4. Drug therapy and frequency of hospital review will indicate the severity of the condition.

Some claimants may underreport the day-to-day effects of effort intolerance, failing to describe factors such as slow walking pace. A good history taking technique is essential to ascertain the reliable and repeatable functional ability.

In the IB-PCA the main functional areas initially affected by reduced exercise tolerance are Walking Up and Down Stairs, and Walking.

Exemption from the IB-PCA should be considered if the limitation of effort tolerance is severe. Such cases will usually show little day-to-day variability and are likely to be on maximal therapy. A further disabling feature in aortic stenosis is effort syncope. This is potentially fatal, and, exemption or application of a non-functional descriptor may be appropriate.

Appendix A - Disability Assessment Scales used in Cardiac Medicine and Surgery

New York Heart Association (NYHA) Classification

Disability from dyspnoea due to cardiac disease

- Class 1 - Patients with cardiac disease but without dyspneoa during normal activities.
- Class 2 - Cardiac disease resulting in mild/moderate dyspneoa on normal exertion.
- Class 3 - Marked dyspneoa on normal exertion.
- Class 4 - Any exertion causes dyspneoa, or symptoms at rest.

Canadian Cardiovascular Society (CCS) Classification

Disability due to angina

- Class 1 - Angina only on strenuous or prolonged exertion.
- Class 2 - Slight limitation due to angina with normal activities.
- Class 3 - Marked limitation due to angina with normal activities.
- Class 4 - Unable to undertake any physical activities. Angina at rest.

Diagnostic criteria for infective endocarditis	
MAJOR CRITERIA	
1)	*Positive blood culture for infective endocarditis*
i)	Typical micro-organism for infective endocarditis from two separate blood cultures.
	Viridans streptococci, *Streptococcus bovis*, HACEK group*,
	Or community acquired *Staphylococcus aureus* or enterococci in the absence of a primary focus, *or*
ii)	Persistently positive blood culture, defined as recovery of a micro-organism consistent with infective endocarditis from:
i)	Blood cultures drawn more than 12 hours apart, *or*
ii)	All of three or a majority of four or more separate blood cultures, with first and last drawn at least 1 hr. apart.
2)	*Evidence of endocardial involvement*
a)	Positive echocardiogram for infective endocarditis
i)	Oscillating intracardiac mass, on valve or supporting structures, *or* in the path of regurgitant jets, *or* on implanted material in the absence of an alternative anatomic explanation *or*
ii)	Abscess *or*
iii)	New partial dehiscence of prosthetic valve *or*

b) New valvular regurgitation (increase or change in pre-existing murmur not sufficient)
MINOR CRITERIA
Predisposition: predisposing heart condition *or* intravenous drug use.
Fever: ≥ 38.0° C (100.4 ° F)
Vascular phenomena: major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial haemorrhage, conjunctival haemorrhages, Janeway lesions.
Immunological phenomena: glomerulonephritis, Osler's nodes, Roth spots, rheumatoid factor.
Microbiological evidence: positive blood culture but not meeting major criterion as noted previously.
Echocardiogram: conistent with infective endocarditis but not meeting major criterion as noted previously.

*HACEK = *Haemophilus* spp., *Actinobacillus actinomycetemcomitans, Cardiobacterium hominis, Eikenella* spp., and *Kingella kingae*.

7 References

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