

Valvular heart disease: recommendations for investigation and management

SUMMARY OF GUIDELINES PRODUCED BY A WORKING GROUP OF THE BRITISH CARDIAC SOCIETY AND THE RESEARCH UNIT OF THE ROYAL COLLEGE OF PHYSICIANS

Abstract—Despite the decline of rheumatic fever, valvular heart disease remains a common and important problem. The investigation and treatment of patients have been revolutionised by the widespread availability of echocardiography, which provides information about disease aetiology, accurate and non-invasive assessment of severity and a means of monitoring progression. These advances have facilitated progress in reconstructive valve surgery and the application of balloon valvuloplasty. Improved valve prostheses and reduction in peri-operative mortality have also broadened the indications for valve replacement surgery. A working party convened by the British Cardiac Society and the Royal College of Physicians of London have reviewed current practice and provided guidelines, summarised in this article, for the investigation and management of adults with valvular heart disease in primary care, district general hospitals and tertiary referral centres.

In 1995 the Joint Audit Committee of the British Cardiac Society and the Royal College of Physicians of London convened a working group to recommend guidelines for the investigation and management of adults with valvular heart disease. The group included representatives from general practice, the nursing profession, community medicine, district general hospital cardiology, tertiary centre cardiology and cardiac surgery, the legal profession and regional health authority purchasing bodies. This paper is a summary of the full guidelines [1], to which reference should be made for clarification or further information.

Initial assessment and referral: the role of the general practitioner

The general practitioner's priorities should be to identify and refer appropriately:

1. Patients in whom valvular heart disease is life-threatening.

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2. Patients with symptomatic valvular heart disease, who will benefit from exact diagnosis and planned management.
3. Patients in whom valvular heart disease may underlie the presenting condition, eg heart failure caused by aortic stenosis, atrial fibrillation caused by mitral valve disease.
4. Patients with valvular heart disease in special situations, eg pregnancy.

Assessment

A background of rheumatic fever, ischaemic heart disease, hypertension or intravenous drug abuse should be considered and potential symptoms of angina, syncope or dyspnoea evaluated. Examination should determine the heart rhythm and blood pressure and the presence of any cardiac murmurs or signs of heart failure.

The finding of a systolic murmur in conjunction with any of the above symptoms should not be dismissed as aortic 'sclerosis' (a defunct term). Given the poor prognosis of symptomatic aortic stenosis [2] and the difficulty of precise diagnosis, such patients should generally be referred urgently for a specialist opinion.

Investigation

Routine investigations are unnecessary. Simple blood tests, eg haemoglobin, thyroid function tests, should be requested according to clinical circumstances. An electrocardiogram or chest x-ray are unlikely to influence the referral decision and are unnecessary.

Referral

If a patient has suspected valvular heart disease it would be usual to refer unless the patient is unwilling or other conditions are judged to contra-indicate referral. The degree of urgency is indicated below: in uncertain cases telephone advice should be sought from an experienced specialist, ideally of registrar grade or above.

- *Emergency.* Valvular heart disease with uncontrolled symptoms or suspected infective endocarditis. Hospital admission will usually be required.

- *Urgent.* Symptomatic valvular heart disease. The patient should attend a specialist clinic as soon as possible.
- *Routine.* Diagnostic or long-term management issues. The patient should be seen in accordance with routine clinic procedure.

Hospital assessment: the role of the physician/cardiologist

Investigations

Electrocardiogram—All patients should have an electrocardiogram as part of the initial consultation.

Chest x-ray—A chest x-ray is not indicated routinely at the first assessment [3]. Baseline examination is useful if serial assessment is likely. The detection of coincident pulmonary disease may be important, particularly when invasive investigation is being considered.

Echocardiography—This is the non-invasive method of choice for the diagnosis and evaluation of valvular heart disease. Trans-oesophageal echocardiography is a specialised technique with specific indications in the assessment and management of valvular heart disease (Table 1). Referral to a centre with this facility may be necessary.

Assessment and advice—The general practitioner should be promptly informed of the specialist assessment, with emphasis on diagnosis and management, and any other important issues, eg endocarditis prophylaxis, anticoagulation. The patient should receive advice on the following issues, where appropriate:

- dental hygiene
- contraception or the possibility/advisability of pregnancy in women of childbearing age
- endocarditis awareness and prophylaxis
- anticoagulant control
- medication
- safety to drive.

Nurse specialists can play an important role in patient education and should be available if possible. Their advice can be supplemented by appropriate literature.

Follow-up arrangements should be made clear, with emphasis on any indications for early re-referral.

Follow-up

Many patients do not require hospital follow-up. Asymptomatic patients with a good long-term prognosis (eg 'innocent' murmurs, trivial mitral regurgitation, mild aortic regurgitation in a hypertensive patient) can be discharged after suitable advice and reassurance, with clear guidelines for re-referral.

The frequency of follow-up for patients with significant native valve disease should be determined according to the diagnosis and its severity. Most patients should attend a consultant-led clinic annually. Those

Table 1. Role of trans-oesophageal echocardiography in the assessment of valvular heart disease

- When trans-thoracic echocardiography fails to provide the necessary information
- Mitral valvuloplasty
- Valve repair
- Prosthetic valve assessment
- Infective endocarditis

with significant aortic stenosis, evidence of ventricular impairment/dilatation or symptoms suggestive of disease progression should be seen more often (Fig 1a, b).

The patient should be seen by an experienced cardiovascular physician, ie a consultant or staff grade cardiologist (or physician experienced in the care of cardiovascular disease), a senior registrar in cardiology or an experienced cardiology registrar or clinical assistant. Senior house officers should be supervised and have access to experienced colleagues for advice. Ideally, the patient should see a consultant every 2–3 years.

Facilities for an electrocardiogram, chest x-ray and echocardiogram should be available on the day of clinic attendance. Anticoagulant control should be reviewed. Patients with new or deteriorating symptoms, an embolic event or unexplained febrile illness require detailed investigation. If there is a definite suspicion of severe haemolysis or endocarditis the patient should be admitted to hospital.

Patients should be intermittently reviewed by the general practitioner, possibly in a formalised 'shared care' arrangement. Those with new signs or symptoms should be re-referred without delay. Any febrile illness of uncertain origin should be considered as infective endocarditis and referred urgently prior to any antibiotic administration.

Medical treatment

Vasodilators palliate valvular regurgitation [4] and left ventricular impairment and should be considered for all patients with significant aortic or mitral regurgitation. They are contra-indicated in aortic stenosis. Digoxin is the conventional treatment for atrial fibrillation and may have a role in patients with heart failure, even in sinus rhythm, in addition to diuretics and angiotensin converting enzyme inhibitors. Anticoagulant therapy will be discussed later.

Poor prognostic features and referral for intervention

Patients with the following features have a poor prognosis and should receive close attention: atrial fibrillation, left ventricular dysfunction or dilatation,

pulmonary hypertension, co-existent coronary artery disease and disease affecting more than one valve.

Patients with increasing symptoms, echocardiographic or clinical evidence of disease progression, or who develop new poor prognostic features should be considered for referral with a view to intervention. Ideally, elective investigations should be undertaken in the tertiary centre within three months of referral. In urgent circumstances the waiting time should not exceed one month; occasionally, immediate admission may be necessary (Table 2).

Interventional cardiology and cardiac surgery: the role of the tertiary centre

General indications for intervention

Mitral stenosis

All patients with symptomatic mitral stenosis should be actively considered for percutaneous balloon mitral valvuloplasty. Contra-indications include the presence of left atrial thrombus, severe subvalvar involvement or valve calcification, and significant mitral regurgitation. It is mandatory to detect these beforehand, using trans-oesophageal echocardiography [5].

The surgical alternatives are open mitral commissurotomy or mitral valve replacement. Recent studies comparing balloon valvuloplasty (in suitable patients) with surgery for mitral stenosis have shown no difference in outcome or complication rates [6,7].

Mitral regurgitation

Timing of intervention should be guided by the severity of symptoms and evidence of progressively increasing left ventricular size. Echocardiography is the investigation of choice; an end-systolic dimension greater than 5.5cm suggests the need for surgery [8].

The benefits of mitral valve repair [9] may favour earlier intervention if repair is thought feasible. Trans-oesophageal echocardiography is essential for pre- and peri-operative assessment [10].

Aortic stenosis

Symptomatic severe aortic stenosis has a very poor prognosis [2]. Unless there are exceptional circumstances, urgent surgery is always indicated. The presence of left ventricular impairment solely due to aortic stenosis is **not** a contra-indication to surgery. The management of patients with asymptomatic severe aortic stenosis is controversial: surgery is often indicated, especially if the trans-valve gradient exceeds 100mmHg.

Initial enthusiasm for balloon aortic valvuloplasty as an alternative to valve replacement in the elderly [11] has been tempered by poor long-term results [12]. The procedure should be restricted to occasional

patients with severe aortic stenosis in the following circumstances:

- pregnancy
- when other major surgery is proposed for underlying disease with an uncertain prognosis
- as a bridge to aortic valve replacement if immediate surgery is inadvisable.

Aortic regurgitation

Echocardiographic evidence of progressive left ventricular dilatation (end-systolic dimension >5.5cm), even in the absence of major symptoms, indicates the need for surgery [8]. Aortic valve replacement is usually necessary and some patients with dilatation of the ascending aorta (>5.5cm) may require root replacement.

Mixed valve disease

Assessment and treatment comprise a summation of the factors for each individual valve lesion. The mortality and morbidity of double valve replacement are significantly higher and this may influence the decision to proceed.

Special surgical considerations

The elderly

With appropriate patient selection, the results of valve replacement in the elderly can approach those in younger patients. Mortality after aortic valve replacement is consistently lower than after mitral valve replacement [13,14]. The risk of both procedures rises with advancing age or if there is co-incident coronary artery disease. A simple, quick procedure is important in the elderly; additional bypass grafting should only be undertaken for severe coronary artery disease.

Pre-operative coronary angiography

Echocardiographic techniques have negated the need for routine invasive assessment of valve disease. Most patients undergoing valve surgery require pre-operative coronary angiography to detect co-existent coronary artery disease. In some, eg the young with no history of chest pain or risk factors for coronary artery disease, this may be unnecessary. Equally, coronary angiography should not delay a potentially life-saving operation, eg emergency aortic valve replacement.

The choice of prosthesis

Mechanical prostheses—These offer durability and longevity. Despite the problems associated with anti-coagulation and small risks of thrombosis-in-situ they are the first choice for the majority of patients.

Tissue prostheses—*Heterografts (or xenografts)* offer a low

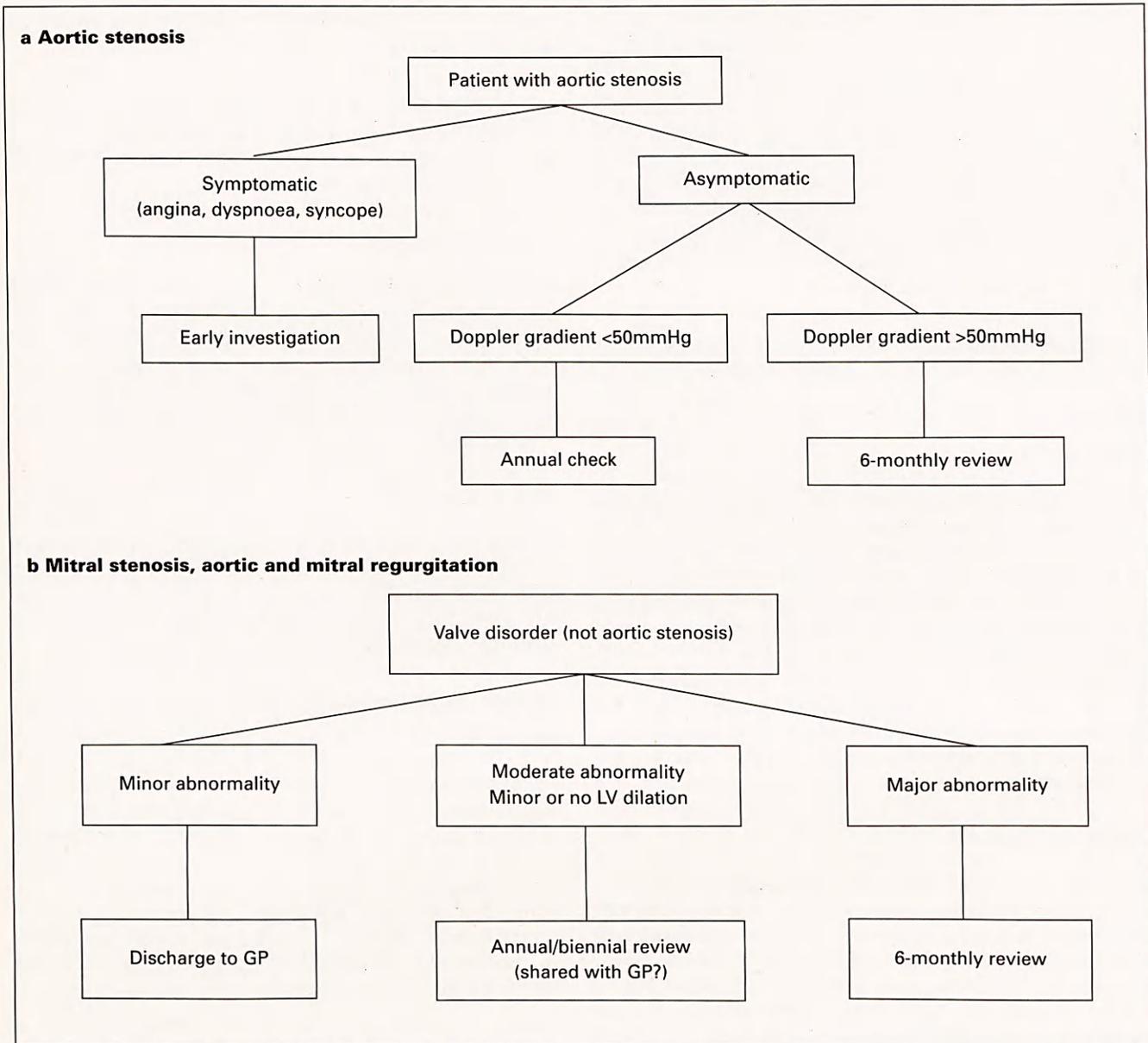


Fig 1. Suggested follow-up schemes in native valve disease

thrombo-embolic rate and avoid the need for anti-coagulation. The high incidence of long-term valve failure restricts their use to elderly patients with aortic valve disease whose life expectancy is less than that of the valve (10-12 years), situations where anticoagulants are contra-indicated and some patients with infective endocarditis. They are seldom used in the mitral position due to a high risk of complications [15,16] and also because anticoagulation is usually indicated for other reasons (left atrial enlargement and/or atrial fibrillation). *Homografts* are also associated with a low thrombo-embolic rate. Relative indications include aortic valve replacement in active endocarditis, aortic root replacement and aortic valve replacement in the young when a tissue valve is necessary.

Urgency of surgical treatment

The urgency should be defined for each patient and guided by clinical judgement. Circumstances will vary in individual centres; in particular, the quantity of emergency work undertaken can restrict elective waiting list admissions.

Follow-up post-procedure

Hospital discharge

The patient's condition and the presence of any cardiac murmurs at the time of discharge should be documented. The general practitioner should be

Table 2. Indications for expediting investigation and treatment

Urgent	Immediate
Aortic stenosis	Aortic stenosis
Angina	Syncope
Aortic regurgitation	Pulmonary oedema
Pulmonary oedema	Aortic regurgitation
Sudden deterioration	Dissection
Mitral stenosis	Mitral stenosis
Pulmonary oedema	Severe pulmonary oedema
Mitral regurgitation	Mitral regurgitation
Sudden deterioration	Intractable heart failure
	All
	Infective endocarditis

provided with essential information (Table 3), supplemented by a fuller summary in due course. Patients also need specific advice on recovery, exercise, sex, work, sport, driving, flying, dentistry, drug therapy (particularly anticoagulants) and the reporting of new symptoms, especially those suggesting valve failure or endocarditis. This should be accompanied by an information booklet.

Outpatient follow-up

The broad principles are the same as those in native valve disease. Intermittent follow-up by a physician with a cardiovascular interest or cardiac surgeon is satisfactory, providing there are no significant complications. Duplication of follow-up in the district general hospital and the interventional centre should be avoided.

The patient and general practitioner should be aware of the need for early review if problems arise. When medical advice is needed the general practitioner should contact a senior member of the team involved in the patient's hospital care.

Management in special circumstances

1. Antithrombotic management in valvular heart disease

This subject has been reviewed by a European working party which has produced comprehensive guidelines [17] including indications for anticoagulation and International Normalised Ratio (INR) recommendations for patients with prosthetic valves or native valve disease.

Background—In native valve disease the risk of thrombosis is related to the following factors: atrial fibrillation, left atrial enlargement, mitral obstruction and impaired left ventricular function [18].

General surgery in anticoagulated patients—The peri-

operative anticoagulation regime should be discussed in advance with a cardiovascular physician or cardiac surgeon. Patients with native valve disease can often stop or reduce their anticoagulants. Patients with prosthetic heart valves should *not* discontinue oral anticoagulation: if the patient cannot take oral medication, *intravenous* warfarin or heparin can be used. Heparin is less effective in preventing thrombosis: if it is used, then warfarin should be re-instituted as soon as possible. Most surgical procedures can be safely performed with an INR of around 2.0. In procedures resulting in large raw surfaces the risk of bleeding must be weighed against the risk of valve thrombosis if the INR falls below 2.0.

2. Infective endocarditis

Prevention—Working parties in the United Kingdom and America have produced guidelines [19,20] which are summarised in the full guidelines. Patient information cards are available from The British Heart Foundation.

Diagnosis—Diagnosis can be difficult. Minimum investigations include a full blood count, biochemical screen, erythrocyte sedimentation rate (and/or C-reactive protein) and at least three sets of blood cultures from two separate sites before starting antibiotic therapy. Trans-thoracic echocardiography should be performed though a negative study does not exclude the diagnosis. Trans-oesophageal imaging has a high negative predictive value [21] and is particularly sensitive in the detection of vegetations [22] or abscesses [23]. Echocardiographic features have been incorporated in the new 'Duke' diagnostic criteria [24] which are superior to the old von Reyn criteria.

Treatment—Optimal management requires close liaison between the cardiologist, cardiac surgeon and

Table 3. Minimum information for the general practitioner at the time of hospital discharge

- Consultant's name
- Junior doctor responsible for patient (legible)
- Direct line telephone number (if possible)
- Diagnosis
- Procedures performed (including dates and outcome)
- Complications (including any new arrhythmia)
- Drugs and dosage
- Anticoagulants (if relevant)
 - desired INR
 - therapeutic range
 - frequency of INR estimation
 - follow-up arrangements
- Clinic follow-up plans—who, where and when?
- Information given to the patient
- Indications for re-referral

microbiologist. Antibiotic therapy should be started promptly after blood cultures have been taken. Recommendations for the treatment of streptococcal and staphylococcal endocarditis have been published [25]. Indications for surgery include heart failure due to valvular regurgitation, abscess formation, failure to respond to medical therapy and an unstable infected prosthetic valve. Urgent surgery should not be delayed by an arbitrary period of pre-operative antibiotic therapy.

Prosthetic valve endocarditis—Rigorous prophylaxis is vital and intravenous cannulae, urinary catheters and indwelling lines should be avoided whenever possible. Prosthetic valve endocarditis has a mortality of approximately 50%, and further surgery is usually required, often as an emergency. It should be managed in a cardiothoracic surgical centre unless close communication and rapid transfer are possible. Trans-thoracic echocardiography is inadequate and prosthetic valve patients with unexplained pyrexia should be referred for trans-oesophageal imaging.

3. Valvular heart disease in pregnancy and young women

This group of patients should be managed by a specialist with experience in this area.

Planning of family—The oral contraceptive pill is safe in valvular heart disease, though anticoagulated patients should be aware of the potential drug interaction. Patients in New York Heart Association class I or II before conception will usually tolerate pregnancy without difficulty. If intervention seems likely this should be carried out before pregnancy if at all possible.

The choice of prosthetic valve for a young woman anticipating pregnancy is difficult. Bioprostheses do not require anticoagulation but degenerate more quickly in pregnancy [26]. Mechanical prostheses are durable but carry the need for anticoagulation. The choice should be made in consultation with the patient.

The pregnant patient with a murmur—These patients should be seen by an obstetrician and cardiologist as early as possible in case pregnancy is contra-indicated. Minimum investigations are a haemoglobin level and electrocardiogram; echocardiography should be performed if the diagnosis is uncertain.

Follow-up—The patient should be followed by an obstetrician and cardiologist. Deterioration is most common in mitral and aortic stenosis and these patients should receive particular attention.

Medical treatment—Beta-blockers may prevent deterioration in pregnant women with mitral stenosis [27]. Angiotensin converting enzyme inhibitors are contra-indicated in pregnancy.

Intervention—Balloon dilatation of mitral or aortic stenosis can be performed in early pregnancy. If valve replacement is required this should be deferred until pregnancy is sufficiently advanced to allow delivery by

Caesarean section immediately before cardiac surgery [28].

Endocarditis—Prophylaxis is not indicated to cover normal delivery in women with native valve disease. It should be used in patients with prosthetic valves or a history of previous endocarditis. Advice should be sought from a microbiologist concerning the choice of antibiotics.

Delivery—The mode of delivery and anaesthesia should be discussed in advance. Epidural anaesthesia should be avoided because of the risk of hypotension.

Anticoagulation & prosthetic valves in pregnancy—Patients with tissue valves who are not chronically anticoagulated generally do well but need careful follow-up because of the risk of valve degeneration. The risk of pregnancy for patients with mechanical prostheses is small. The main hazard is valve thrombosis and embolism, and meticulous anticoagulation is required. Most patients, particularly those with prosthetic valves, should continue oral anticoagulants with close INR monitoring. Warfarin carries a 3–5% risk of embryopathy [29] but the risk of fetal bleeding is very low at doses less than 5mg/day [30]. It is not significantly excreted in breast milk. Heparin is not teratogenic but has a narrow therapeutic index: its use should be confined to the first trimester of pregnancy in women whose warfarin requirement is high (ie greater than 5mg/day). The convention has been to admit patients with mechanical prostheses for peri-partum anticoagulant control with heparin. Anticoagulant control in this situation can be extremely difficult, and for this reason we recommend elective caesarean section at 38 weeks of pregnancy with only a brief 3-day interruption of warfarin therapy (two days preoperative and one day postoperative) [30].

Conclusions

These guidelines represent the consensus views of the working group. The aims have been to provide a balanced set of recommendations and to guide practice without substituting for individual clinical judgement. These standards should stimulate audit of the care of valvular heart disease and highlight areas where practice can be improved.

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A full version of these guidelines will be available from the College in September.