

# Cardiomyopathy and anaesthesia

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Cardiomyopathies are diseases of the heart muscle and may present with cardiac dysfunction. Originally defined as conditions with unknown aetiology, the World Health Organization reclassification in 1995 was expanded to include all known causes and is based upon anatomical and physiological features. Within this classification, the three main identified types of cardiomyopathy are dilated, hypertrophic, and restrictive.

Patients with these conditions present to anaesthetists in elective and emergency situations, but may be diagnosed at anaesthetic pre-assessment.

## Pathophysiology

The major pathophysiological features of the cardiomyopathies are related to ventricular dysfunction; however, the nature of this dysfunction varies between the different types.

### Systolic dysfunction

This is the predominant form of dysfunction seen in dilated cardiomyopathy (DCM). There is a decrease in myocardial contractility that is usually global, reducing left ventricular ejection fraction. Initially compensation occurs with enlargement of the left ventricular cavity allowing an increase in stroke volume with an associated increase in force of contraction as described by the Frank–Starling relationship. Eventually, these compensatory mechanisms fail, cardiac output decreases, and left ventricular failure ensues. Up to 14% of the elderly population has echogenic evidence of systolic dysfunction.

### Diastolic dysfunction

This is the major feature of hypertrophic and restrictive cardiomyopathy, but some diastolic dysfunction can occur in the dilated form. It is characterized by impaired left ventricular filling with consequential raised left ventricular filling pressures. During diastole, a healthy left

ventricle undergoes active relaxation (an ATP-dependent process) after closure of the aortic valve. This relaxation continues during early left ventricular filling after mitral valve opening. In late diastole, left ventricular filling is passive and dependent on the compliance of the ventricle. Diastolic dysfunction can be caused by either impairment of active relaxation or a reduction in left ventricular compliance or a combination of both. Active relaxation is most commonly affected by ischaemia, while a reduction in ventricular compliance may be due to intrinsic myocardial changes including fibrosis or external restriction due to pericardial disease.

### Dilated cardiomyopathy

DCM is characterized by progressive cardiac dilation and results in impaired ventricular function. It has a prevalence of 36 per 100 000 population.<sup>1</sup> A large number of cases are idiopathic but within these there is a familial association. There is an increased incidence in males and in patients of Afro-Caribbean origin. The known causes are ischaemic, valve dysfunction and post-viral infection. DCM can also be found in association with sickle cell disease, muscular dystrophy, excess alcohol, hypothyroidism, and some chemotherapy agents. Identifying the cause is important as directed treatment may improve ventricular function. Peripartum cardiomyopathy (PPCM) has an incidence of 1 in 4000 live births, and has a quoted mortality of between 20% and 50%.<sup>2</sup> PPCM usually presents post 36 weeks but may develop for up to 5 months postpartum. It may recur in subsequent pregnancies; therefore, a cardiology assessment should be performed prior to further conceptions.

### Symptoms and signs

The low cardiac output state presents with tachycardia and as the condition progresses dyspnoea, ascites, and peripheral oedema can develop. As the atrio-ventricular ring dilates,

### Key points

There are three main types of cardiomyopathy: hypertrophic, dilated, and restrictive.

Patients with cardiomyopathy may be asymptomatic, but they still require careful anaesthetic management as symptoms are not predictive of risk.

Many cases may be diagnosed at anaesthetic pre-assessment.

Perioperative arrhythmias requiring intervention are common.

Anaesthetic perioperative management is dictated by the presenting pathophysiology.

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valvular regurgitation and arrhythmias can occur. Intracardiac thrombus, leading to embolic events may also be a feature. There is combined systolic and diastolic dysfunction in the most severe cases with impaired relaxation and elevated left ventricular end-diastolic pressures (LVEDP).

### Medical management

Systolic dysfunction is the major pathological component of DCM; medical therapy is as for chronic heart failure. A preoperative history of heart failure is an important risk factor for postoperative cardiac complications,<sup>3</sup> and risk increases when clinical signs are present before surgery. Optimal management of this heart failure should occur before an operation.

The effect of medical therapy including angiotensin-converting enzyme inhibitors (ACEIs), angiotensin II inhibitors, and diuretics should be determined. Other therapies include  $\beta$ -blockers, spironolactone, digoxin, biventricular pacing, and anticoagulants.

### Angiotensin inhibitors

ACEIs have been shown to improve survival in all grades of left ventricular failure<sup>4</sup> with angiotensin II inhibitors indicated in those who do not tolerate ACEI.<sup>5</sup> Whether these drugs should be given on the day of surgery is contentious. ACEIs are known to cause intraoperative hypotension, but this is often short-lived and generally responds to i.v. fluids and conventional treatment.<sup>6</sup> If the decision is taken to omit ACEIs, then they should be recommenced as soon as possible after surgery. Angiotensin II inhibitors cause more severe and prolonged episodes of intraoperative hypotension and are best omitted on the day of surgery.

### $\beta$ -Blockade

The use of  $\beta$ -blockers in the treatment of chronic heart failure has been shown to improve mortality, and is well established. In patients undergoing surgery and considered at high risk of coronary events,  $\beta$ -blockers have been shown to improve outcomes.<sup>7</sup> There is, however, no evidence that they improve perioperative outcome in patients with acute heart failure.

### Diuretics

Spironolactone is an aldosterone antagonist that has been shown to decrease mortality by 30% in those with severe (New York Heart Association class III or IV) chronic heart failure already on ACEIs and loop diuretics.<sup>8</sup> Loop diuretics are useful in relieving symptoms, despite being unproven in prolonging survival.

### Biventricular pacing

In addition to medication, some patients gain benefit from biventricular pacing. These patients lack coordination of ventricular contraction, due to delayed pathways of conduction through damaged myocardium. Re-synchronization of left and right ventricular contraction by coordinated stimulation from leads in both ventricles

(that improves stroke volume) is supported by recommendations from NICE (May 2007).

### Perioperative management

The optimal time for surgery is problematic, but medical control of heart failure for >1 week is desirable. Any arrhythmia should be appropriately treated, particularly atrial fibrillation (AF) where rate, rhythm control, or preferably both are important. Preoperative echocardiography is indicated to assess the degree of biventricular impairment and valvular dysfunction. The correction of any significant electrolyte abnormalities should be made perioperatively. This is especially important in relation to potassium and magnesium ions, both of which are likely to be depleted in patients receiving chronic diuretic therapy, and derangement may predispose to cardiac arrhythmias. Consideration should be given to the use of regional or local anaesthesia. Regional anaesthesia used alone or in combination with general anaesthesia has the advantage of reducing after load which can improve cardiac output. However, hypotension must be prevented to avoid myocardial hypoperfusion. Treatment of arterial pressure changes should be considered if a >10% decrease in systolic pressures occurs.

The overall aims of anaesthesia are to:

- avoid tachycardia;
- avoid/minimize the effects of negative inotropic agents, in particular anaesthetic drugs;
- prevent increases in afterload;
- maintain adequate preload in the presence of elevated LVEDP.

For patients undergoing higher risk procedures under general anaesthesia or those in whom heart failure management is not optimized, invasive direct arterial pressure monitoring is indicated. Central venous pressure monitoring offers some additional information on right ventricular preload but does not provide information on left heart pressures. Information from a pulmonary artery catheter can be useful, although may not improve outcome. Intraoperative transoesophageal echocardiography may also be useful for examining dynamic changes in cardiac performance and the response to inotropes and fluid loading. Where such technology is unavailable, then the use of oesophageal Doppler monitoring of aortic root velocities can provide information on cardiac performance. During anaesthesia, the maintenance of sinus rhythm and avoidance of tachycardia are additional goals. Inotropic support if required during and after surgery can be provided by the use of a variety of agents, including dobutamine, dopamine, phosphodiesterase inhibitors, and levosimendan. It may be necessary to counteract some of the peripheral vasodilator effects of general anaesthetic agents to assist coronary perfusion. This is achieved with the cautious use of norepinephrine which increases systemic vascular resistance and maintains mean arterial pressure. Postoperative regional anaesthesia can be beneficial and good quality pain management avoids increases in systemic vascular

resistance and heart rate. Cautious fluid management is important, and is best undertaken in an intensive care area.

## Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant condition. This important cause of sudden death in young adults affects significant numbers of the population who are unaware that they have the condition. The genetic associations are numerous and involve a number of different chromosomes, so screening is difficult to perform as not all mutations have been described.<sup>9</sup> It is also true that progression of an individual's disease cannot be predicted, with many patients having a normal life span being relatively unaffected. Diagnosis is based upon unexplained left ventricular hypertrophy which may be concentric or asymmetrical, diffuse or focal.

### Symptoms and signs

Although often asymptomatic, the presenting features are as for heart failure and include: shortness of breath, chest pain on exertion, syncope or pre-syncope, arrhythmias, and sudden cardiac death. Signs include: hypotension, a low volume pulse, left ventricular heave, ejection systolic murmur, and a mitral regurgitant murmur.

The major risk factors for sudden cardiac death are:

- a family history of sudden death;
- extreme hypertrophy of the left ventricular wall (>30 mm);
- unexplained syncope;
- non-sustained ventricular tachycardia.<sup>9</sup>

### Cardiac failure and ischaemia in HCM

In most patients, the left ventricular size is normal or reduced. A large number of HCM patients have diastolic dysfunction due to a failure of the left ventricle to undergo active relaxation. This leads to a compensatory increase in the contribution to LV filling from atrial contraction and explains why AF is poorly tolerated. An increase in end-diastolic pressure leads to a decrease in coronary perfusion pressure. In addition, the hypertrophied muscle, with a higher oxygen demand, makes the ventricle prone to ischaemia. Any decrease in systemic vascular resistance can lead to a further reduction in coronary blood flow.

### Arrhythmia

Both atrial and ventricular arrhythmias are common. When these occur, they can lead to rapid decompensation.

### Left ventricular outflow tract obstruction

In a subset of high-risk patients, hypertrophy is most prominent in the interventricular septum. This leads to dynamic left ventricular outflow obstruction in the sub-aortic region. This is exacerbated in situations where there is under filling of the left ventricle with forceful sub-aortic narrowing. The velocity of blood in the outflow

tract draws the anterior mitral valve leaflet towards the intraventricular septum. The result can be complete obstruction of the outflow tract coupled with mitral regurgitation as the anterior mitral valve leaflet is pulled away from the posterior leaflet.

### Medical therapy

Medical therapy is used for patients with symptoms of heart failure and includes  $\beta$ -blockade combined with or without disopyramide or verapamil. These should be continued in the perioperative period as they are thought to improve symptoms by reducing heart rate and by exerting a negative inotropic effect leading to improved coronary perfusion pressures. Patients thought to be at risk of sudden death may benefit from an internal cardio-defibrillator; the only intervention which may decrease this risk.<sup>10</sup> Surgical cardiac myomectomy and more recently non-surgical ablation have been used to treat septal hypertrophy and reduce left ventricular outflow tract (LVOT) obstruction.

### Perioperative management

Before operation, it is important to establish if the patient is symptomatic, as those with symptoms are more likely to suffer intraoperative problems. Any device present should be checked by the pacing clinic and clear instructions for its intraoperative management documented. The use of a magnet to deactivate any pacemaker device is to be discouraged and any changes to device management should be made using the appropriate manufacturer's software.

ECG abnormalities are present in 75–90% of cases, including high-voltage complexes, ST abnormalities, T wave inversion, and evidence of left atrial enlargement. However, these changes do not predict the severity of the condition. Echocardiography can demonstrate the degree of hypertrophy and may show systolic anterior movement of the mitral valve, but this is often a dynamic situation and its absence before operation does not mean that it will not occur intraoperatively.

In the patient with known HCM, the overall aim is to prevent or treat LVOT obstruction, arrhythmia, and ischaemia should they occur.

The strategy involves:

- maintenance of sinus rhythm;
- reduction in sympathetic activity to reduce chronotropy and inotropy;
- maintenance of left ventricular filling;
- the maintenance of systemic vascular resistance.

Premedication may assist in reducing sympathetic activity and therefore reducing cardiac workload. Adequate hydration is essential to maintain ventricular preload, particularly left-sided pressures. Invasive arterial monitoring placed before induction of anaesthesia facilitates a rapid response to changes in arterial pressure. Agents used for induction and maintenance of

anaesthesia should be selected to minimize decreases in systemic vascular resistance and prevent tachycardia or sympathetic surges. Ventilation should aim to minimize any reduction in venous return that may occur, and this can be achieved by small tidal volumes and rapid ventilatory frequencies.

The use of transoesophageal echocardiography during surgery is an ideal continuous monitor of the adequacy of left ventricular filling and the development of potential LVOT obstruction. Pulmonary artery wedge pressure measurements can be misleading as they may overestimate left ventricular filling, but are useful for monitoring trends. Hypotension should be treated with judicious volume resuscitation and  $\alpha$ -agonists. The use of agents with inotropic or chronotropic actions can increase myocardial oxygen demand. Hypertension should be treated with  $\beta$ -blockade after ensuring adequate analgesia and anaesthesia. Vasodilator agents, such as GTN, should be avoided. Postoperative management aims to avoid sympathetic stimulation by providing good pain control and avoidance of hypothermia. Regional anaesthesia is relatively contraindicated as it can cause a decrease in systemic vascular resistance and potentially lead to outflow obstruction, although it is worth noting that there are a number of case reports, particularly in obstetric anaesthesia, documenting its safe use.

Cardiac arrest in HCM is a special circumstance. The use of inotropic agents is contraindicated if the arrest is thought to be due to LVOT obstruction as this will only increase the obstruction.<sup>9</sup>  $\alpha$ -Agonists, i.v. fluids, and rapid correction of arrhythmias are more appropriate measures. The application of external defibrillator pads is recommended before induction of anaesthesia.

## Restrictive cardiomyopathy

Restrictive cardiomyopathy is rare outside tropical regions of the world, where endomyocardial fibrosis is found. In non-tropical regions, idiopathic fibrosis, the most common cause, is associated only with increasing age. Other rare causes include amyloidosis, sarcoidosis, haemochromatosis, and eosinophilic endocarditis.

Diastolic dysfunction is the major clinical feature producing reductions in ventricular compliance. Biventricular dysfunction may occur, with the right ventricle being more commonly affected. Hypertrophy of the ventricles is absent and systolic function remains normal until late in the disease. Diagnosis is based upon myocardial biopsy, which may show endocardial or myocardial disease.

## Symptoms and signs

In addition to the signs of left and right heart failure, patients often have a low volume pulse, regurgitant murmurs, a third heart sound, and a raised JVP with rapid X and Y descent that increases or fails to decrease on inspiration.

## Medical therapy

The aim is to lower the elevated filling pressures caused by low ventricular compliance without reducing cardiac output.

The medical therapy includes the use of ACEI or  $\beta$ -blockade, which can lead to improvement in some patients and symptomatic treatment with diuretics. None of these medications improves long-term outcome in restrictive cardiomyopathy. Arrhythmias are common and include AF, bradyarrhythmias secondary to SA or AV node involvement, and a variety of ventricular tachyarrhythmias. Primary treatment is with amiodarone or digoxin, with pacemakers and ICDs reserved for more malignant arrhythmias. Other medications include anticoagulation for those at higher risk of embolic disease and disease-modifying agents specific to the underlying condition.

Of specific note:

- Use of digoxin is relatively contraindicated in patients with amyloidosis as it is likely to precipitate arrhythmias.
- Cardioversion of patients in AF may result in complete heart block due to existing intrinsic pacemaker damage.

## Perioperative management

Echocardiographic assessment of the right and left ventricles is important. If right ventricular dysfunction predominates, then it is important to maintain a high right ventricular filling pressure.

The overall aims of anaesthesia are:

- to maintain adequate filling pressures;
- sinus rhythm to be maintained if possible;
- to manage electrolyte disturbances;
- to maintain SVR in the presence of relatively fixed cardiac output.

With right ventricular dysfunction as the predominant feature, the importance of preload manipulation should not be underestimated. Under general anaesthesia, the combination of vasodilatation, myocardial depression, and reduced venous return can compromise cardiac output. The addition of intermittent positive pressure ventilation may even lead to circulatory arrest, and for this reason, spontaneous ventilation is theoretically beneficial, but often impractical.

There is a risk of complete heart block and ventricular tachycardia during anaesthesia in patients who have damage to the intrinsic pacemaker system.

Regional anaesthesia in addition to general anaesthesia is avoided due to the decrease in systemic vascular resistance.

Postoperative management should include cardiac monitoring and maintenance of right ventricular filling pressures and also correction of electrolyte disturbances.

## Summary

Cardiomyopathies are more common than is widely appreciated. Whilst their impact upon the conduct of anaesthesia can often remain low, it is essential that clinical anaesthetists understand the

**Table 1** Pathological features and anaesthetic management aims of DCM, HCM, and restrictive cardiomyopathy

	Dilated	Hypertrophic	Restrictive
Cardiac output	Low	Low	HR dependent
Stroke volume	Low	Low	Fixed
Contractility	Reduced	High	Normal or reduced
<i>Anaesthetic aims</i>			
Preload	Maintain in the presence of elevated LVEDP	Maintain/high	High
Heart rate	Avoid tachycardia	60–80	High
Rhythm	Sinus preferred	Maintain sinus rhythm	Sinus preferred
Contractility	Inotropic support often required	Slight reduction beneficial	Promote with inotropes if required
Afterload	Reduce or maintain, avoid increases	Increase slightly or maintain	Maintain as cardiac output is fixed

‘ideal’ perioperative management aims, so that individualized directed care may be delivered (Table 1).

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Please see multiple choice questions 10–12