ANESTHETIC CONSIDERATIONS IN PATIENTS WITH CARDIOMYOPATHIES

- A Review -

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Introduction

Cardiomyopathy literally means “heart muscle disease”, and refers to the deterioration of the function of the myocardium for any reason. Patients with cardiomyopathy are often at risk of dysrhythmias or sudden cardiac death.

Cardiomyopathies can generally be categorized into two groups, based on World Health Organization guidelines: extrinsic and intrinsic. In extrinsic cardiomyopathies the primary pathology is outside the myocardium. Most cardiomyopathies are extrinsic, because the most common cause is ischemia. Intrinsic cardiomyopathies is weakness in the muscle of the heart that is not due to an identifiable external cause. To make a diagnosis of an intrinsic cardiomyopathy, significant coronary artery disease should be ruled out. The term intrinsic cardiomyopathy does not describe the specific etiology of weakened heart muscle.

Anesthetic Implications

Anesthetic management, of patients with cardiomyopathy with reduced systolic function, is challenging and may be associated with high mortality1. Tabib and his group presented a retrospective analysis of 1500 autopsies following unexpected deaths and identified 43 deaths possibly related to anesthesia and surgery1. Pathological examination revealed cardiac lesions in 40 cases and 20% were due to cardiomyopathy (Table-1).

Table-1 Cardiac causes of death (Tabib et al)1

<table>
<thead>
<tr>
<th>Condition</th>
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<tr>
<td>arrhythmogenic right ventricular cardiomyopathy (14 cases)</td>
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<tr>
<td>coronary artery disease (9 cases)</td>
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<tr>
<td>cardiomyopathy (8 cases)</td>
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<tr>
<td>structural abnormalities of the His bundle (7 cases)</td>
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<tr>
<td>mitral valve prolapse (1 case)</td>
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<td>acute myocarditis (1 case)</td>
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Of note, arrhythmogenic right ventricular cardiomyopathy (ARVC) was identified in 35% in this subgroup series. ARVC is an inherited disease with fatty fibrotic tissue infiltration of the right ventricle which causes ventricular arrhythmias and sudden death. EKG of these patients presents with T wave inversion in the anterior leads2.

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Cardiomyopathies

Cardiomyopathy can be broadly classified as heart muscle disease which decreases cardiac function. It can be classified into four groups: dilated, hypertrophic, and restrictive or Takotsubo type (Table 2).

<table>
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<th>Table 2</th>
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<td><strong>Types of cardiomyopathies</strong></td>
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</table>

- **Dilated:**
  - Ischemic
  - Non-ischemic infections, chemotherapeutic agents, drug abuse, alcohol, and peripartum.
- **Hypertrophic:**
  - Septal hypertrophy-idiopathic hypertrophic, Secondary to Hypertension
- **Restrictive** (sarcoid)
- Takotsubo

Dilated cardiomyopathy (DCM) is defined by a large heart cavity with impaired systolic function of one or both ventricles (Fig. 1). It is characterized by ventricular dilatation and impaired systolic cardiac function. It is defined by the presence of (a) fractional myocardial shortening < 25% and/or ejection fraction < 45%; and (b) left ventricular end diastolic diameter > 117% excluding any known cause of myocardial disease. Familiar dilated cardiomyopathy accounts for 20-48% of all DCM and is defined by the presence of two or more affected relatives with DCM meeting the above criteria or a relative of a DCM patient with unexplained sudden death before the age of 35. The prevalence is 920/100,000. It occurs more frequently in males (3:1) and in African Americans (2.5:1) compared to Caucasians. It may be ischemic or non-ischemic. The ischemic type is related to atherosclerosis and ischemic heart disease. The non-ischemic type may be secondary to infections (HIV, Coxsackie virus, cytomegalovirus, toxoplasmosis, Chagas’ disease, trichinosis, leptospirosis, Lyme disease), chemotherapeutic agents (adriamycin, doxorubicin), drug abuse (alcohol, cocaine, methamphetamines and heroin) or during the peripartum period.

The clinical presentation of dilated cardiomyopathy includes symptoms such as dyspnea, orthopnea, weakness, fatigue and leg edema. Physical findings are similar to those seen in congestive heart failure. Patients may have increased jugular venous distention, rales and pulmonary edema, resting tachycardia, s3 and s4 heart sounds and cardiomegaly.

Hypertrophic cardiomyopathy may occur either related to increased hemodynamic workload or without provocation. The latter is known as hypertrophic obstructive cardiomyopathy (HOCM) and idiopathic hypertrophic subaortic stenosis (IHSS). The former is termed hypertensive hypertrophic cardiomyopathy. IHSS is transmitted in an autosomal dominant pattern with variable penetrance. Echocardiography (ECHO) shows disease in about one fourth of first degree relatives.

Restrictive cardiomyopathy is the least common cause of cardiomyopathy in western countries. It is most frequently due to sarcoïd disease.

Recently, Takotsubo cardiomyopathy has been described. It is a transient, reversible, left ventricular dysfunction causing severe hypotension and can mimic an acute coronary event. However, cardiac catheterization often reveals normal coronary arteries. It is rare, usually occurs in postmenopausal women associated with stress and chest pain. EKG may show ST elevation and ECHO and ventriculogram studies demonstrate left ventricular mid and apical ballooning with hypokinesia. The basal segment of the left ventricle may be hyperkinetic. It has been related to anaphylaxis after succinylcholine. Stress induced cardiomyopathy may also follow cephalosporin induced anaphylaxis. Sympathetic discharge can trigger transient cardiomyopathy. In one case report, although vital signs responded favorably to resuscitative efforts after an anaphylactic reaction during general anesthesia, cardiovascular collapse reappeared with transient ventricular tachycardia shortly after transfer to the intensive care unit. There was diffuse regional wall motion abnormalities in the mid ventricular region. Increased MB fractions of creatinine kinase and troponin T levels indicated myocardial necrosis but coronary catheterization indicated normal arteries.
Management

Two key factors exist in the management of patients with cardiomyopathies; one is to improve systolic function and the other is to prevent sudden death due to ventricular arrhythmias.

Several types of treatment for dilated cardiomyopathy aim at improving systolic function. Patients should initially be managed medically. Biventricular pacing, cardioplasty or cardiac transplant may also be required to improve cardiac function. Arrhythmias are managed with amiodarone and/or an automatic implantable cardioverter defibrillator (ICD). Amiodarone prevents life threatening arrhythmias and an ICD promptly treats the arrhythmias with an electrical shock impulse.

Medical management to improve systolic function includes administration of diuretics, beta-blockers, angiotensin converting enzyme (ACE) inhibitors, or angiotensin receptor blockers (ARB).

In the renin-angiotensin system, angiotensin II causes vasoconstriction, and release of aldosterone, vasopressin and anti diuretic hormone (ADH). All these factors increase the blood volume and cause hypertension. ACE inhibitors prevent angiotensin I from converting into angiotensin II and thus prevent the subsequent hypertensive effects. The net effect is a reduced afterload. In patients with cardiac disease, ACE inhibitors reduce the risk of death. They also slow heart emodling, prevent the heart from becoming less efficient over time and improve ventricular function.

Biventricular pacing devices are often used in patients with cardiomyopathies to improve systolic function (Fig. 2). Biventricular pacing (BVP) is beneficial for patients with severe cardiomyopathy in moderate to severe congestive heart failure with an EF <30% and in ventricular asynchrony. An interventricular conduction defect with a wide QRS complex is indicative of asynchrony of the two ventricles. Biventricular leads are programmed to synchronize the contraction of the right and left ventricles thereby improving the ejection fraction. BVP improves left ventricular (LV) systolic function, decreases LV size, decreases mitral regurgitation, and shortens the prolonged QRS interval. Quality of life is markedly improved.

Right ventricular pacing is achieved by the lead placed in the apex of the right ventricle. This lead in addition to pacing function has arrhythmia detection and defibrillation function. Two shock coils are incorporated in the right ventricular lead for defibrillation. Left ventricular pacing is obtained by a lead placed into the obtuse marginal branch of the coronary sinus (Fig. 2). The pacing of the two ventricles can then be synchronized.

Cardioplasty has been developed in several ways to improve systolic function. In the common type, the latissimus dorsi muscle is placed around the heart as a free flap, and its contraction synchronized to augment
ventricular systolic function. Mitral valve repair will also improve cardiac function.

Cardiac transplant may be recommended for patients with end stage dilated cardiomyopathy, not amenable to other therapies. Left ventricular assist devices may be used as a “bridge” prior to transplant when a patient is awaiting a donor.

**Antiarrhythmic drugs and ICDs**

In addition to low ejection fraction, patients with cardiomyopathy tend to develop ventricular arrhythmias and sudden death. Therefore, oral administration of amiodarone an antiarrhythmic medication is prescribed, or a cardioverter defibrillator is implanted to treat ventricular tachycardia.

An implantable cardioverter-defibrillator is more effective than amiodarone in reducing mortality in high-risk patients with previous myocardial infarction and is usually the primary treatment. Amiodarone may be used as an adjunct to reduce the frequency of ICD shocks.

A single chamber ICD consists of a generator which contains a battery and a small computer. A ventricular lead with two shock coils is attached to the generator (Fig. 3). The battery life usually ranges from 4 to 6 years. At the tip of the lead are a sensor and a pacer. These devices are able to distinguish between ventricular fibrillation and ventricular tachycardia (VT). If ventricular fibrillation (VF) occurs then a 25 Joules shock is delivered from the ICD right ventricular lead shock coil. If it is VT, then it may pace the heart faster than the rate of VT to override it and break the VT. This is referred to as antitachycardia pacing. When bradycardia occurs pacing function is initiated.

Other indications for ICDs are in patients likely to develop ventricular arrhythmias. These include IHSS, prolonged QT syndrome or Brugada syndrome. Brugada syndrome is an inherited disease with an increased risk of sudden cardiac death due to ventricular fibrillation. The EKG may manifest a right bundle branch block pattern.

**Preoperative preparation (Table 3)**

The preoperative preparation of these patients must be meticulous as they have minimal or no cardiac function.

**Preoperative assessment:**

<table>
<thead>
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<th>Volume status</th>
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<tr>
<td><strong>Continue Antiarrhythmic drugs</strong></td>
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<tr>
<td><strong>Drug interactions with ACEs inhibitors. Electrolytes-Potassium/Magnesium correction</strong></td>
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<tr>
<td><strong>Hemoglobin optimized</strong></td>
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<tr>
<td><strong>ICD-deactivation</strong></td>
</tr>
<tr>
<td><strong>Inotropes (Resistance to usual dose)</strong></td>
</tr>
<tr>
<td><strong>Intraaortic balloon pump if necessary</strong></td>
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*Fig. 3*  
*Chest X-Ray demonstrating the leads of ICD with two translucent segments-shock coils*
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reserve. Any decrease in myocardial contractility, heart rate, or vasodilatation can cause profound hypotension. Preoperatively, patients tend to be dehydrated, as most have been diuresed, a further cause for hypotension during anesthetic care. However this dehydration is generally beneficial for these patients as it improves limited cardiac function. But they easily can become hypotensive with anesthetics which cause vasodilatation. Preoperative hydration may not be desirable as it may lead to congestive heart failure. Fluid management is critical. To err on the hypovolemic state is prudent. Therefore a vasopressor to mitigate against the vasodilating effect of the anesthetic is a rational approach.

As patients may develop ventricular arrhythmias in the perioperative period, antiarrhythmic medications should be continued. Some patients may already have had an ICD implanted. During the anesthetic care drug interaction resulting in hypotensive episodes requiring vasopressor therapy has been reported when ACE inhibitors are combined with diuretic therapy10.

Arrhythmias occur when potassium or magnesium levels are decreased. These electrolytes should be measured preoperatively and corrected as necessary11.

Oxygen carrying capacity should be adequate. The main determinants of oxygen carrying capacity are cardiac output and hemoglobin. Therefore, hemoglobin should be maintained at a higher level and 13-14gms/100ml has been recommended12. To improve cardiac output, inotropes, biventricular synchronized pacing or an intraaortic balloon pump may be required.

Management of Patients with ICD prior to surgery13,14

Many patients with cardiomyopathies have an ICD for defibrillating function. Some patients may not be aware of the type, whether it is a simple pacemaker or an ICD. ICDs have larger generators than pacemakers (Fig. 4). The right ventricular lead of an ICD, unlike a pacemaker lead, has two sets of spiral segments which can be identified by chest X-Ray (Figs 3, 5).

The difficulty encountered with ICDs during surgery is that the energy discharged during the use of electrocautery can be mistaken by the sensor in the ICD as a tachyarrhythmia. The antiarrhythmic function of the ICD is activated and the patient receives an inappropriate shock. This shock, if discharged at the vulnerable phase of the cardiac electrical cycle, may induce ventricular arrhythmia. Also multiple shocks with repeated uses of the cautery, can damage the heart, decrease cardiac function, damage the generator or deplete the battery of the ICD. Therefore, all ICDs should be deactivated prior to surgery if use of cautery is planned. Magnet placement on the generator of some ICDs may deactivate its shock therapy function but not the pacing or sensing capability.

With other ICDs7 (Angeion, Guidant, St. Jude, Ventritex) placement of a magnet will not have any effect on function. Ideally, in the management of these patients, a cardiologist or the manufacturer representative should be consulted preoperatively.

Once the ICD is deactivated, defibrillator pads must be placed on the patient’s chest and connected to an external defibrillator as a stand-by to treat any ventricular arrhythmia that may occur.

Fig. 4
Comparison of size of ICD generator (left), with pacemaker generator

Fig. 5
ICD shock-coils are seen in this chest-X-Ray

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In a critically ill patient with cardiomyopathy, if the planned surgery is complex and absolutely necessary, an intraaortic balloon pump may be placed preoperatively\textsuperscript{15}.

**Anesthetic management**

Anesthetic management of patients with severe cardiomyopathies is associated with a high morbidity and mortality\textsuperscript{1} and therefore requires careful planning, preparation and monitoring (Table 4). Many of these patients present for ICD or biventricular pacemaker placement. However, they may present emergently or for any type of surgery.

Preoperatively the diagnosis of cardiomyopathy may be lacking or not easily forthcoming\textsuperscript{16}. A patient with a history of coronary artery disease with a low EF is usually treated with furosemide, ACE inhibitors and has an implanted ICD. Therefore, in the preoperative evaluation, these four signs (Quadratic sign; CHF, cardiomegaly, ICD, EKG changes) if present, should alert the anesthesiologist to a low EF. If the preoperative diagnosis of cardiomyopathy is missed, routine anesthetic management carries considerable risk. Patients will likely become profoundly hypotensive\textsuperscript{17,18}. Prompt recognition and immediate intervention of hemodynamic instability with appropriate vasoactive or inotropic medications is required to prevent deterioration. Anesthesiologists initially use neosynephrine in incremental doses of 100 \( \mu \)g every 30-40 seconds and initiate a norepinephrine infusion at 4-8\( \mu \)gs/min or dopamine at 5\( \mu \)gs/kg/min and adjust dose range to titrate the systolic blood pressure above 90-100 mmHg.

Patients can also manifest life threatening ventricular arrhythmias. Anesthesiologists should be prepared to use lidocaine, amiodarone or defibrillation to treat the arrhythmia.

<table>
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<th>Table 4</th>
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<td><strong>Presentation of cardiomyopathy patients</strong></td>
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In addition to the basic monitoring (BP, pulse oximeter, EKG, end-tidal CO\textsubscript{2}), direct arterial blood pressure monitoring is required to identify abrupt hemodynamic changes\textsuperscript{18}. In situations where the surgery is complex or of long duration, transesophageal echocardiography (TEE) monitoring is also appropriate\textsuperscript{19}. When a patient becomes hypotensive during anesthetic management, TEE differentiates the cause of hypotension as due to global hypokinesia, regional ischemic ventricular dysfunction or hypovolemia. These changes may be treated with inotropes, coronary vasodilators or fluids, as indicated. Recently, intraoperative Doppler tissue imaging (DTI) has been added as a valuable diagnostic tool\textsuperscript{20}. DTI displays and measures systolic and diastolic velocity from a myocardial region. It is simple to perform and independent of endocardial imaging. Assessment of systolic and diastolic function on regional (detection of ischemia) as well as global levels (ejection fraction, grading of diastolic function) and evaluation of filling pressure can all be derived from DTI signals.

The cardinal feature of dilated cardiomyopathy is the reduction in the systolic function or the left ventricular ejection fraction (LVEF). Patients with LVEF over 45% usually do not require any change in anesthetic technique.

Anesthetics not only depress central nervous system function but also cardiac function. They tend to depress the myocardium, slow the heart rate and dilate the blood vessels. Anesthetic management needs to be customized for those with LVEF below 45\%. Therefore, selecting the type and dose of anesthetics with minimal vasodilatation and myocardial depressing effect is prudent. Drugs like ketamine, etomidate and narcotics have minimal depressing effect on cardiac function and are used frequently\textsuperscript{18,19,21}. Conventional anesthetics like propofol, sodium thiopental or isoflurane in recommended doses depress cardiac function. However, recent laboratory studies indicated that sevoflurane in a porcine model decreased myocardial infarct size after prolonged coronary occlusion\textsuperscript{22}. Sevoflurane cardioprotection was substantiated in the juvenile intact organism.

Nerve blocks are a rational approach for appropriate surgery as they have minimal hemodynamic abrasion.
Anesthetic management of the parturient with cardiomyopathy is challenging, as the baby’s welfare must be considered in the management of the critically ill mother. Usually regional techniques are preferred for cesarean section in a normal parturient but may exaggerate the hypotension in a cardiomyopathic parturient. If general anesthetics are indicated in an emergency for a mother who is decompensating, drugs with minimal effect on the baby need to be selected. Etomidate with remifentanil has been successfully used without respiratory depression of the baby. Remifentanil crosses the placenta but is quickly metabolized by the baby’s liver. It must also be borne in mind that most of the cardiac drugs will also cross the placenta and/or be excreted in the mother’s milk and can affect the baby.

Use of vasoactive or inotropic drugs may be required frequently to counteract the negative effects of the anesthetics on cardiac function. Inotropic drugs such as dopamine, epinephrine, dobutamine and milrinone increase the ejection fraction significantly. In clinical reports dopamine is frequently used during the anesthetic care of these patients. Dopamine in the appropriate dose range has positive inotropic, chronotropic and vasoconstrictive effects making it an ideal agent to negate adverse cardiovascular effects of anesthetics.

The beta receptors in the myocardium, B₁ and B₂ receptors, control contractility and B₃ influence relaxation. With cardiomyopathy there is decreased myocardial beta-adrenergic receptor density or sensitivity of these receptors. Therefore conventional doses of beta stimulants may be inadequate and larger doses may be required.

**Conclusion**

Cardiomyopathies as a class are identified increasingly as a result of improved means of detection and an aging population. Also, presentation may be sudden or already well known. Consequently, patients with this underlying condition may present at any time for anesthesia or intraoperatively. It is essential that anesthesiologists understand the underlying pathology to better manage these patients.
References


