Echocardiographic Evaluation of Cardiomyopathies
Andrew Maslow MD
Department of Anesthesiology
Rhode Island Hospital, Providence Rhode Island

Introduction
Cardiomyopathy is generally defined as a “disease of the myocardium associated with cardiac dysfunction”. Primary cardiomyopathies are divided into three major classifications: a) dilated cardiomyopathy (DCM), b) hypertrophic cardiomyopathy (HCM), and c) restrictive (or infiltrative) cardiomyopathy (RICM). Although there are functional and clinical overlaps, each of the primary cardiomyopathies has distinctive morphological and functional characteristics.

The incidence of cardiomyopathy is less than 1% in the general population, with DCM representing the vast majority of cases. Hypertrophic cardiomyopathy is less prevalent while RICM is the least common. Accurate diagnostic assessment of patients suspected of having a cardiomyopathy is important to establish prognosis and to institute appropriate treatment. This talk will focus on salient echocardiographic features of primary cardiomyopathies, but may include discussion and description of none-primary cardiomyopathies for completeness.

Dilated Cardiomyopathy
Dilated cardiomyopathy (DCM) accounts for 60% of all cardiomyopathies and is characterized by progressive myocyte hypertrophy, dilation and contractile dysfunction of one or both ventricles. Although ventricular wall thickness can be increased, the degree of hypertrophy is proportionally less compared to the amount of dilatation. The development of left ventricular (LV) hypertrophy is initially beneficial in reducing systolic wall stress, a major determinant of myocardial oxygen consumption. However, wall stress is never fully normalized and eventually stimulates LV remodeling, resulting in a reduced ejection fraction (EF) as the ventricle continues to dilate and assume a spherical shape. DCM is defined by increased end-systolic and end-diastolic volumes and a reduced LV EF (< 45%). Furthermore, mitral (MV) and tricuspid (TV) regurgitation may be present in association with increased ventricular volume and/or
pressure load. Cardiac thrombi may also develop in associated with reduced cardiac function, and are most commonly found in the LV apex or left atrial appendage (LAA).

Many patients with DCM may be asymptomatic, however, most patients range in age between 18 and 50 years, with progressive deteriorating DCM include clinical signs and symptoms all of which are consistent with LV heart failure. The right ventricle (RV) may be independently involved in rare cases of a familial form of DCM, however RV failure is usually a later and more ominous consequence of primary LV failure, and is usually associated with a particularly poor prognosis.

As many as fifty percent of patients die within 2 years. Five-year survival following the initial diagnosis has been reported in the 50-75% range depending upon the initiation of therapy, extent of cardiac remodeling and dysfunction, and advanced age. About 25% of patients with recent onset DCM improve spontaneously. Patients with DCM and advanced CHF with LV end-diastolic diameters > 4 cm/m² body surface area have twice the 1-year mortality rate compared to those patients with less significant ventricular dilatation.

**Echocardiographic Evaluation**

Classic 2-D echocardiographic features of DCM include the presence of increased systolic and diastolic (>7-8 cm) LV dimensions, reduction of LVEF (<45%), and development of a spherical configuration. Although DCM is defined by the presence of significant systolic dysfunction, concurrent diastolic dysfunction is common and may manifest anywhere within the full spectrum of severity from impaired relaxation to restriction. Symptoms of CHF in patients with DCM appear to be related to the severity of diastolic dysfunction.

The LV wall can vary from normal to increased thickness; however, relative wall thickness (i.e. the ratio of end-diastolic wall thickness to end-diastolic cavity radius) is severely diminished. Although not pathognomonic, LV and/or RV wall motion tend to be symmetrically and globally reduced in patients with DCM compared to the typical segmental and focal wall motion abnormalities more commonly associated with ischemic heart disease and coronary artery narrowing. Dobutamine stress echocardiography may RWMA associated with CAD, differentiating it from patients with idiopathic DCM.
is important to make this distinction since patients with ischemic cardiomyopathy may experience significant improvement in functional capacity with coronary revascularization.

Ventricular dilatation associated with DCM may produce functional atrial-ventricular valve incompetence. Incomplete closure of the MV and TV may develop due to annular dilatation, abnormal alignment of the papillary muscles related to the development of ventricular sphericity, and apical displacement of the coaptation point which increases tension on the leaflets (i.e. “apical tenting”).16,17

The risk of intracavitary thrombi, due to flow stasis, are higher for patients with DCM. Thrombi develop more commonly in the LV compared to the LA.18

**Hypertrophic Cardiomyopathy**

The classification of hypertrophic cardiomyopathy (HCM) has been simplified to reflect the pathophysiology and to accommodate the varied patterns of hypertrophy. While diastolic dysfunction occurs in almost all individuals with HCM, only 25% experience a dynamic, intermittent or episodic obstruction to ventricular systolic outflow.19,20 Patients are therefore subdivided into two related but distinct groups: 1) hypertrophic cardiomyopathy (HCM); and 2) hypertrophic obstructive cardiomyopathy (HOCM), which includes those with obstruction to ventricular systolic outflow.

Hypertrophic cardiomyopathy (HCM/HOCM) is defined as an abnormal thickening of the myocardium without chamber dilation, and in the absence of a demonstrable cause (e.g. aortic stenosis [AS], systemic hypertension). The incidence is approximately 0.2% in the general population, but reporting may vary depending on the referral patterns of the institution, and the diagnostic criteria.19,20

The age of onset, morphology, and pathophysiology of HCM/HOCM varies greatly. More commonly, HCM/HOCM presents in young adulthood from the second to fifth decades, and is characterized by a diffuse or asymmetric ventricular hypertrophy. Presentation of HCM/HOCM in older patients (≥ 65 years old) is becoming increasingly more recognized.21,22 Although commonly ascribed to HCM/HOCM, asymmetric hypertrophy has also been reported as an adaptive response to AS, systemic hypertension, and in certain congenital cardiac anomalies.
Hypertrophic cardiomyopathy is defined by the presence of LV hypertrophy (> 11 mm thickness), which occurs disproportionately in the ventricular septum by a ratio of > 1.3:1.0 relative to the measured free wall thickness. However, different patterns of hypertrophy have been reported including isolated proximal basal septal hypertrophy (“septal bulge”), posterior wall hypertrophy, concentric or diffuse hypertrophy, and RV hypertrophy in a small number of cases. Four types of hypertrophic cardiomyopathy have been described: Type I – hypertrophy limited to the anterior septum; Type II – hypertrophy of anterior and posterior septum; Type III – diffuse hypertrophy sparing only the basal posterior wall; Type IV – apical hypertrophy.

The mechanism of LVOT obstruction (LVOTO) is complex, involving ventricular hypertrophy and abnormalities of the MV apparatus. The common pathway is represented by a narrower ventricular outflow cavity with or without distortion of MV leaflet coaptation and subsequent MR. Scenarios more likely to result in LVOTO include asymmetric hypertrophy, a prominent basal septum, a narrowed outflow cavity (< 20-25 mm), and structural abnormalities of the MV apparatus. The latter includes anteriorly positioned papillary muscles, abnormal insertion of the papillary muscles into the mitral leaflet, elongated mitral leaflets, and disturbances in MV annular function (e.g. posterior annular calcification). Echocardiographic delineation of the mechanism of LVOTO is important for planning therapeutic interventions especially in regards to the requirement for, and timing of surgery.

**Echocardiographic Evaluation**

In patients with HCM/HOCM, systolic function is usually preserved (LVEF ≥ 55%) or hyperdynamic (LVEF ≥ 65%) and the ventricular dimensions are normal or reduced in size. A smaller percentage (< 5-10%) of patients may have reductions in systolic function with or without chamber dilation, suggestive of late- or end-stage cardiomyopathy. Focal and global systolic dysfunctions are due to disarray of the myocardial fibers with or without fibrosis. In addition, decreases in myocardial performance may be due to atheromatous CAD, hypertrophic involvement of the coronary arteries, reduction of coronary perfusion in a hypertrophied ventricle, or decreased coronary artery vasodilatory reserve.
Wall thickness ranges from normal (≤ 11mm) to greater than 30 mm with varying patterns from apex to base, concentric or asymmetric. The absence of hypertrophy does not rule out HCM/HOCM since development of myocardial thickening may be delayed until the second or third decade, emphasizing the need for routine follow-up of patients with a family history of HCM/HCOM.

Diastolic dysfunction, which may associated with ventricular hypertrophy, myocardial disarray, fibrosis, reduced systolic emptying, and/or coronary artery insufficiency occurs in almost all patients. Relief of LVOTO reduces LV pressures and improves coronary blood flow, myocardial oxygen balance, and ventricular filling.\textsuperscript{33} Severity of diastolic dysfunction ranges from normal to restrictive filling patterns, the latter reflecting greater degrees of hypertrophy and fibrosis.

Systolic outflow obstruction whether at the level of the LVOT or at the middle portion of the ventricular cavity results in high systolic flow velocities, incomplete systolic emptying, and increased ventricular cavity pressures. The Doppler profile is described as late peaking (> 1.4 m/s) and “dagger-shaped” in appearance reflective of a dynamic process vs. a fixed one. LVOTO can also be demonstrated using M-mode AoV leaflets, which will open normally but close prematurely in mid-systole due to dynamic obstruction of systolic outflow. Two-dimensional echocardiographic assessment may display the narrowing of the LVOT with or without systolic anterior motion (SAM) of the mitral leaflets and chordae. Specific features of the MV and mitral apparatus associated with SAM and LVOTO include redundant or elongated mitral leaflets, lax chordae, abnormally positioned papillary muscles, and mitral annular dysfunction.

Color Doppler analysis of the LVOT and MV demonstrates aliasing of the CFD jet in LVOT at the level of narrowing consistent with high velocity flow, and across the MV indicating significant MR. This ‘Y’ shaped CFD is consistent with LVOTO/SAM and MR.

The mechanism of SAM/LVOTO and MR is likely to involve some combination of the Venturi effect and abnormalities of the mitral apparatus. These variables may contribute differently from one patient to another. Nevertheless, delineating the mechanisms of SAM/LVOTO/MR with echocardiography provides useful information
when considering the best mode of therapy for patients who are symptomatic from outflow tract obstruction and MR.

**Restrictive and Infiltrative Cardiomyopathy (RICM)**

Restrictive cardiomyopathy (RCM) is a pathologic process in which diastolic function of one or both ventricles is severely impaired in the absence of a definitive systemic disease. While several diseases are associated with restrictive filling, only Loeffler’s hypereosinophilic endocarditis, endomyocardial fibrosis, and idiopathic restrictive cardiomyopathy qualify as primary restrictive cardiomyopathies. Secondary causes of RICM include infiltrative diseases such as amyloidosis, sarcoidosis, and storage diseases (e.g., glycogen storage disease, hemochromotosis), certain drugs (anthracyclines, ergotamine, methysergide, serotonin), and a number of miscellaneous causes (transplant rejection, radiation, cancers, toxins). Collectively, these diseases have significant overlap and have been categorized as restrictive/constrictive, infiltrative, congestive or obliterative cardiomyopathies reflecting the range of clinical and morphological presentations. Congestive cardiomyopathy more accurately reflects this group of disorders due to the presence of either pulmonary venous or vena caval congestion, which results in signs of left and right heart failure, respectively. However, since this term is vague and could include all cardiomyopathies, the diseases in this section will be referred to as “restrictive/infiltrative cardiomyopathies”: RICM).

RICM should be suspected when a patient presents with CHF, non-dilated ventricles, dilated atria, diastolic dysfunction, and poor response to medical therapy. While chest X-ray reveals cardiomegaly and a thickened heart, ECG demonstrates low QRS voltage consistent with replacement of the normal myocardium with non- or poorly conducting tissue. Tissue biopsy frequently confirms the diagnosis, however it may also show non-specific and non-diagnostic fibrotic changes of the endomyocardium and myocardium.

**Echocardiographic Evaluation**

Establishing the diagnosis and determining the severity of ventricular dysfunction is important to develop establish a treatment plan and prognosis. Differentiation from
more treatable causes of heart dysfunction (e.g. CAD, valvular heart disease, pericardial disease) is necessary to allow prompt performance of therapeutic procedures (e.g. pericardiectomy for pericarditis) when indicated.

**Amyloidosis** is the most commonly reported etiology of RICM and is caused by an abnormal layering of protein within the myocardial tissues including all cardiac chambers, the coronary arteries, cardiac conduction system, and heart valves. The heart has been described ‘rubbery’ and non-compliant. Although all patients have bi-atrial enlargement, only 20% had ventricular dilation, which was associated with other pathologies (e.g. CAD, primary pulmonary disease). Focal wall motion abnormalities suggest either amyloid or atheromatous involvement of the coronary arteries. A number of echocardiographic features differentiate the amyloid heart from other causes of hypertrophy and/or diastolic dysfunction. The echocardiographic appearance of the amyloid heart is classically described as ‘speckled’, granular, or ‘starry skied’. All cardiac tissues may be symmetrically or, infrequently asymmetrically thickened and ‘speckled’. Similar to other etiologies of RICM, the atria are enlarged while the ventricular cavity size is often normal or reduced. **However, infiltration of the atrial walls, especially the interatrial septum, differentiates amyloidosis from other causes of CHF.** In addition, RV thickening and speckling are more common with amyloidosis than other diseases. Intracardiac thrombi were found in 26% of patients, with a greater incidence in the atria.

Mortality for patients with cardiac amyloidosis is high, and survival beyond 2-3 years for patients presenting with CHF is less than 50%. Prognosis in patients with amyloidosis is based on myocardial wall thickness, severity of diastolic dysfunction, and systolic function. For patients with wall thickness \( \leq 12 \) mm the median survival is about 2.5 years, while survival is reduced for those with wall thickness between 12 and 15 mm (1.3 years), and least for those with wall thickness \( \geq 15 \) mm (0.4 years). The incidence of systolic dysfunction is 0%, 35%, and 70% in these 3 groups respectively. Prognosis is also related to the severity of diastolic dysfunction, which in turn is correlated with ventricular wall thickening. For patients with milder cardiac involvement, wall thickness is \( < 15 \) mm, and Doppler profiles suggest a pattern consistent with abnormal relaxation. In contrast, patients with wall thickness \( \geq 15 \) mm have flow
profiles suggestive of a restrictive filling defect. Normal RV free wall thickness is < 7 mm. Patients with RV wall thickness ≥ 7 mm the right sided Doppler patterns reveal a restrictive filling defect.

*Idiopathic restrictive cardiomyopathy* is an autosomal dominant disease associated with heart block and skeletal myopathy, which presents in the third and fourth decades. Histologic examination reveals variable degrees of interstitial fibrosis throughout the heart. For children under 10 years of age, the survival is less than 2 years, while more than 60% of adults survive beyond 4 years. Echocardiographic evaluation reveals diastolic dysfunction, relatively normal systolic function, variable myocardial thickening, atrial enlargement with or without thrombi, and pulmonary hypertension.

*Endocardial fibroelastosis* is found in children and characterized by a thick endocardium. Histology demonstrates infiltration of the endocardium with collagen and elastic tissue causing LV endocardial thickening with or without MV involvement. While the primary form is not associated with other congenital cardiac abnormalities, a secondary form may be found with LVOTO, aortic coarctation, coronary artery abnormalities, or hypoplastic left heart.

*Hypereosinophilic syndrome (Loffler’s endocarditis)* and *endomyocardial fibrosis* are a continuum of the same disease differing only by their presenting pathology. Although these diseases are uncommon, their occurrence is greater in parts of Africa and Asia, accounting for as much as 25% of cardiac deaths. While both are the result of cardiac eosinophilia, Loffler’s endocarditis presents with significant cardiac hypereosinophilia, while endomyocardial fibrosis represents a later stage characterized mainly by fibrosis. As both diseases progress, endocardial thickening and fibrosis develop with greater involvement of the MV and TV subvalvular apparatus producing valve insufficiency and/or stenosis. Involvement of the ventricular apex results in obliteration of the cavity, which may be further complicated by thrombus formation. Echocardiography demonstrates bi-atrial enlargement, endocardial thickening or deposits along the MV and TV associated papillary muscles, and along the apices of both ventricles. The involved tissues appear bright and echodense consistent with calcium deposition. While impairment to ventricular filling is present, systolic motion of the ventricular walls is usually preserved.
Hemochromatosis and Sarcoidosis are infiltrative processes due to iron deposition and non-caseating granulomas respectively. For both diseases, cardiac involvement is rarely seen in the absence of non-cardiac organ involvement. Cardiac involvement is seen in as many as 20-40% of patients.\(^{42,43}\) In sharp contrast to primary RCMs, systolic dysfunction is typical with varying degrees of diastolic dysfunction. A restrictive filling defect is rare.

A number of miscellaneous etiologies of RICM have also been described including carcinoid, radiation therapy, and various metabolic storage diseases.

**Left Ventricular NonCompaction**

Left ventricular non-compaction (LVNC) or hypertrabeculation results from an arrest of the normal fetal development of the myocardium. During its initial stages of development, the heart muscle has a spongy feel and appearance i.e. non-compacted. At this time (up until the 18\(^{th}\) week of gestation) the developing myocardium receives its nutrition via diffusion across cell membranes since the coronary arteries are not yet developed. This loose network of muscle trabeculations and bands maximizes the amount of surface area thereby facilitating the diffusion of blood into the tissue beds. At around the time (18\(^{th}\) week gestation) when the coronary arteries are developed the spongy trabeculated myocardium becomes more compacted so that it might begin to contract and pump, and take on the normal functions of the ventricle. Around this time, the delivery of nutrition to the myocardium has switched to the developing coronary circulatory system. Beyond the 18\(^{th}\) week the prominent trabeculations reduce in size and the spongy myocardium becomes more compacted. In time the trabeculations appear smaller and close to the surface.

Although the incidence of LVNC is difficult to ascertain, it is estimated that 0.015% to 0.05% of echocardiograms display images consistent with LVNC \(^{46}\). Among patients presenting with heart failure, this incidence may be greater \(^{47}\). The timing of arrested development dictates the degree of myocardial dysfunction. In its most severe form the pumping function of the myocardium is severely impaired. The genetic inheritance varies significantly, and mutations generally include genes coding for the myocardial filaments, cytoskeleton, and possibly coronary vasculature.
The clinical presentation of LVNC varies from asymptomatic to severe heart failure, the latter being characterized by a number of non-specific signs and symptoms of heart failure such as peripheral edema, dyspnea, fatigue, and reduced functional capacity. A host of arrhythmias (atrial and ventricular), conduction delays and blocks can be recorded as well. Tachycardias are not well tolerated. There is an increased incidence of intracavitary thrombi located within the trabecular mesh, but not likely to occur in the absence of systolic dysfunction.

During autopsy, noncompacted broad and coarse trabeculae, resembling multiple papillary muscles, and sponge-like interlacing smaller muscle bundles are found. There is an absence of well-formed papillary muscles. (48)

**Echocardiographic Evaluation**

The diagnosis of LVNC is based on echocardiography and cardiac magnetic resonance imaging (CMRI). Jenni et al proposed the following echocardiographic criteria for the diagnosis of LVNC:

1. Thickened LV wall consisting of two layers; a thin compacted epicardial layer and a markedly thickened endocardial layer with numerous prominent trabeculations and deep recesses with a maximum ratio of noncompacted to compacted myocardium > 2:1 at end-systole in the parasternal short-axis view during transthoracic echocardiography (49,50).

2. Color Doppler to highlight the flow within the recesses created by the deep trabeculations.

3. Involvement of the mid to apical inferior and lateral wall segments.

All three of these findings needed to make the diagnosis of LVNC. Apical and mid ventricular inferior and lateral walls may be affected most often. Although hypokinesis of the affected walls, diastolic dysfunction and thrombi may be recorded, these are not part of the criteria to establish the diagnosis.

Other proposed criteria included that by Chin et al:
1. The distance from the epicardial surface to the trough of the trabecular recess is \( \leq 0.5 \) of the distance from the epicardial surface to the peak of the trabeculations. Measurements were obtained from the apical four chamber or sub-xiphoid view at end-diastole during TTE (51).

Although both of these criteria highlight the presence of prominent trabeculations, the Jenni criteria has been shown to be more sensitive (52).

Cardiac magnetic resonance imaging (CMRI) displays a maximum ratio, during diastole, of noncompacted to compacted myocardial thickness of > 2.3 as assessed in three long-axis views (sens 86%; spec 99%). This finding distinguishes LVNC from other cardiovascular causes of prominent trabeculations (AS; HTN) (53).

Treatment is supportive, including standard therapies for heart failure and arrhythmia prevention. Oechslin et al described the outcome for 34 adults with LVNC over 44\(+\) 40 months (46). The mean age at diagnosis was 42\(+\) 17 years with 12 patients experiencing significant heart failure at the time of diagnosis. The mean LVEDD was 65\(+\)12 mm and an LVEF of 33\(+\) 13%. Apical and mid ventricular segments of the inferior and lateral walls were involved in > 80% of cases. Complications included heart failure (18; 53%), thromboembolic events (8;24%) and ventricular tachycardias (14; 41%). Sudden death occurred in 12, death related to heart failure in 4, and two others died of noncardiac causes. Four patients underwent heart transplantation and four received automated cardioverter/defibrillators. Presentation in the neonatal period carries a 14% morality at three years. For unclear reasons, there may be a period of recovery followed by significant deterioration (54).

**Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C)**

Arrhythmogenic right ventricular cardiomyopathy (ARVC) or arrhythmogenic right ventricular dysplasia (ARVD) is characterized, histologically, by fatty and/or fibrous infiltrate of the right ventricle. The severity varies from a functionally normal patient with mild structural changes to complete replacement of the RV myocardium with severe ventricular dysfunction and arrhythmias. The ventricular septum is typically
spared. Less commonly the left ventricle is involved.

Patients present between the ages of 10 and 50 years (mean age of 30 years). It is rarely diagnosed in infancy and uncommonly before age 10. The incidence is regionally different (rare in the United States) with a maximum occurrence of approximately 1:1000. However, ARVC may account for 11% of sudden cardiac death (SCD) in younger patients, and 22% of athletes suffering from SCD (55,56). The diagnosis is suspected for sudden cardiac death brought on by exercise.

The presentation of patients with ARVC varies depending the extent of infiltration. Symptoms include palpitations, syncope, atypical chest pain, and dyspnea (57). Symptomatic atrial (atrial fibrillation) and ventricular arrhythmias, the latter ranging from frequent premature ventricular contractions to ventricular tachycardia/fibrillation are typical (58).

The diagnosis is suspected for young patients with ventricular tachycardia with a left bundle branch block (LBBB) configuration or multiple morphologies. The QRS morphology is more likely that of a LBBB configuration since the arrhythmia is more likely to originate from the right ventricle. The diagnosis, however, depends on histologic demonstration of fibrofatty replacement of the right ventricle. However, since the sensitivity of tissue biopsy may be as low as 67%, other criteria have been established to determine the diagnosis of ARVC (59):

1. Global and/or Regional dysfunction and structural alterations
2. Fatty or fibrofatty replacement of the RV free wall
3. Repolarization or depolarization and conduction abnormalities on the ECG
4. Arrhythmias
5. Family history

The evaluation of suspected patients includes ECG, echocardiography, RNV, and MRI studies. Forty to fifty 40-50% have normal ECG at presentation, however within 6 years, almost all of one of the following (60):

1. Prolonged QRS
2. Incomplete or Complete bundle branch block
3. 30% have an epsilon wave
4. T wave inversion which correlates with degree of RV enlargement
5. QT dispersion
6. Prolonged S wave upstroke

Electrophysiologic testing demonstrates inducible ventricular arrhythmias and typically localizes the foci to the right ventricle.

**Echocardiographic Evaluation**

Echocardiographic evidence of ARVC includes right ventricular enlargement +/- regional wall motion abnormalities (61). Right heart enlargement characteristically includes the right ventricular outflow tract (> 30 mm diameter) (62). Right ventricular enlargement and dysfunction is common, however, RV failure is present in only 6% of patients. A later stage, the right ventricular becomes increasing dilated and dysfunctional.

Disease severity can be classified echocardiographically as below (61):

1. Mild: RVEDV < 75 ml/m² with localized hypokinesis or akinesis
2. Moderate: RVEDV 75-120 ml/m² with localized hypokinesis or akinesis
3. Severe: RVEDV ≥ 120 ml/m² with widespread akinesis/dyskinesis and diastolic bulging

Qualitative echocardiographic findings include trabecular derangement, and a hyperreflective moderator band (62).

Treatment is directed toward preventing sudden cardiac death. Although, not well defined, placement of implantable cardiac defibrillator follows similar guidelines for the general population for both primary and secondary therapies (63). Sotalol (63) may be the best pharmacologic therapy especially when ‘electrical storm’ occurs with ICD therapies. Amiodarone can also be effective (63). Treatment of atrial fibrillation also follows similar principles as the general population including cardioversion, rate control and anticoagulation. Since exercise is known to precipitate tachyarrhythmias, avoidance of exercise is advised.

More invasive therapies include radiofrequency ablation of a documented arrhythmogenic foci, and surgical resection of the right ventricular free wall to decrease the ventricular mass available to initiate ventricular tachycardias. This may also prevent the spread of VF/VT to the left ventricle.
High risk patients (64) include those with symptoms (syncope, hemodynamic instability, and/or VT/VF), evidence of RV failure, evidence of LV involvement, and increase in QRS duration of > 40 msec.

**Takotsubo’s (Stress) Cardiomyopathy**

Takotsubo (‘pot with narrow neck and round bottom used to catch octopi) Cardiomyopathy (TCM), or Stress Cardiomyopathy, was initially described in 1990 in the Japanese population, and named it such based on the end-systolic shape of the LV during ventriculography, which more commonly appears like ‘apical ballooning’ (64). TCM presents as an acute coronary ischemic event characterized by ECG changes and left ventricular dysfunction, however, cardiac catheterization does not reveal significant coronary artery pathology to explain the dysfunction.

The mechanism of dysfunction is not fully known, however, it appears to be related to surges in catecholamines, perhaps related to transient coronary artery vasospasm (65,66), or surges of calcium influx into the myocardial tissues affecting levels of oxygen free radicals and other ionic influxes (67). Catecholamine sensitivity may be more prominent in the ventricular apex explaining the more common pathophysiology.

Demographics show that it is more common in post-menopausal females (95%) experiencing some kind of stressful circumstance whether it be psychological or physical (68,69). The presentation ranges from asymptomatic ECG changes to cardiopulmonary instability (< 10-15%) and death (< 2-3%). More commonly patients present with ECG changes, chest pain, and/or dyspnea. In a review of 70 cases 13% of patients presented perioperatively. Cardiac enzymes may be mild to moderately elevated indicative of infarct, which may persist long-term.

**Echocardiographic Evaluation**

ECG and imaging analyses typically show apical (> 85%) dysfunction often described as systolic apical ballooning, and may be associated with more proximal (mid cavity or basilar) hyperkinesis with or without evidence of LVOTO (68,69). Ten to 15% of the cases may be variant described by basilar hypokinesis/akinesia with normal apical
function. These variant cases are associated with greater hemodynamic dysfunction and mortality.

**Diagnostic criteria include (70)**

1. Reversible LV dysfunction involving apex, midventricular, or basilar segments.
2. Absence of significant coronary artery disease to explain dysfunction
3. Acute ECG changes +/- transient elevations of cardiac enzymes

Depending on the severity of the initial presentation, ECG changes and LV dysfunction (i.e. wall motion abnormalities) may persist indefinitely similar to an infarct due to coronary artery obstruction.

Management is supportive and removal from the stressful situation. Administration of sympatholytics medications is suggested but not of proven value (71,72,73). For the large majority of cases, ECG and imaging changes are transient and resolve within 1-2 weeks. Elevated cardiac enzymes normalize within a few days. Long-term outcome is not well studied however, **Elesber et al** reported a recurrence of 11.4% for 100 patients followed for, on average, four years (74). The rate of recurrence was 2.9%/year for the first four years and then < 2%/year thereafter. Mortality at 4 years was no different than that expected for the general population of the same age.
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