Cardiomyopathies are primary diseases of the myocardium associated with cardiac dysfunction in the absence of causative ischemic or chronic valvular lesions. (1) (2) Physiologically, cardiomyopathies can be classified into 3 major groups: dilated (DCM), hypertrophic (HCM), and restrictive (infiltrative) cardiomyopathies (RICM). The incidence of cardiomyopathies is as follows: DCM > HCM/HOCM) > RICM, with DCM being the most common (60%).

**Dilated Cardiomyopathy:**

Dilated cardiomyopathies (DCM) are characterized by dilation of all 4 cardiac chambers with impaired biventricular systolic function, reduced cardiac output, impaired contractility, elevated left ventricular end diastolic pressures and to a less consistent extent diastolic dysfunction.(2) Potential causes include: idiopathic, toxins (alcohol, doxorubicin, cobalt, snake bites), metabolic (thiamine deficiency, acromegaly), peripartum, infections (postviral, Chagas’ disease), and genetic disorders (Duchenne’s muscular dystrophy, sickle cell anemia).(2)

With the exception of Chagas’ disease, echo features of DCM are fairly uniform despite the etiology. The principal findings of DCM include dilation of all four cardiac chambers and severely diminished systolic function.(2, 3) LV systolic dysfunction is global/diffuse and symmetric. In contrast, ischemia usually produces regional wall motion abnormalities, which are focal or segmental. Although wall thickness may be normal or even increased and overall cardiac mass is increased, end-diastolic wall thickness to end-diastolic cavity radius ratio is severely decreased.(3)

Progressive dilation causes apical displacement of the mitral valve leaflet coaptation point (restrictive Carpentier class III leaflet motion) and annular dilatation which consequently results in mitral (MR) and tricuspid (TR) regurgitation.(5) When appropriate, reductive annuloplasty of
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the mitral and tricuspid valves decreases regurgitation, improves LV morphology, reverses ventricular remodeling, and slows the progression of heart failure.(6) Elevated left atrial pressures from LV systolic failure and MR, along with primary involvement of the right ventricular myocardium contribute to RV systolic dysfunction.(7, 8)

Not surprisingly, severe chamber dilation (9,10), severe ventricular dysfunction(7), elevated right ventricular systolic pressure(11), severe MR(12), and irreversible diastolic dysfunction(13,14) are prognostic indicators associated with increased mortality. Interestingly, a favorable response to low-dose dobutamine indicates a better prognosis.(3, 4)

Diminished blood flow (stasis) secondary to dilated cardiac chambers and decreased systolic function increases the risk of intracavitary thrombosis, especially in the LV apex and the left atrial appendage (LAA). Two dimensional echo is 100% sensitive and 97% specific for diagnosing thrombus.(3)

Chagas’ disease is caused by Trypanosoma cruzi, a protozoan parasite endemic to South and Central America. Unlike other forms of DCM, patients with Chagas’ disease have focal and segmental disease. Patients typically have a left ventricular apical aneurysm with minimal involvement of the ventricular septum.(2, 15, 16)

Hypertrophic Cardiomyopathy:

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant (variable penetrance and variable expressivity) disease associated with asymmetric hypertrophy of the myocardium. Although systolic function is usually preserved, diastolic function is always abnormal and dynamic subaortic obstruction occurs in some individuals (25%).(17) HCM has many synonyms including: asymmetric septal hypertrophy, and idiopathic hypertrophic subaortic stenosis, (IHSS). Current classification reflects pathophysiology; if there is no
obstruction, the disease is referred to as hypertrophic cardiomyopathy, and if dynamic left ventricular outflow tract obstruction occurs (25%), the disease is referred to as hypertrophic obstructive cardiomyopathy (HOCM). (2, 17, 18) Clinically, HCM/HCOM is associated with a high incidence of sudden death, especially during exertion. Angina, exercise intolerance, arrhythmias, and syncope are frequent symptoms. (2, 18) Four types of HCM/HOCM are described based on location of hypertrophy.

Type I: isolated basal anteroseptal hypertrophy
Type II: hypertrophy of the basal anteroseptal and inferoseptal walls with sparing of the anterolateral, inferolateral (posterior), and inferior walls.
Type III: Extensive hypertrophy that spares only the basal inferolateral (posterior) wall
Type IV: Isolated apical hypertrophy

Note, in every type the basal inferolateral (posterior) wall is spared (normal thickness), and the ratio of the anteroseptal to inferolateral (posterior) basal wall is greater than or equal to 1.0 to 1.3. (2, 17) Echocardiographic features of HCM/HOCM during dynamic left ventricular outflow tract (LVOT) obstruction include the following:

- 2D echocardiography: normal systolic function, septal hypertrophy with narrowing of the LVOT and systolic anterior motion of the anterior mitral valve leaflet.
- Pulse wave Doppler: highest velocity noted at the site of dynamic outflow obstruction with sample gate placed below the aortic valve.
- Continuous wave Doppler: classic late-peaking “dagger shaped” velocity curve
- Color flow Doppler: turbulent flow in subaortic region with a posteriorly directed mitral regurgitant jet.
- M-mode through the aortic valve: midsystolic closure of the aortic valve with coarse fluttering of the leaflets
Diastolic dysfunction (universal finding).

Restrictive (Infiltrative) Cardiomyopathy:

Restrictive (infiltrative) cardiomyopathy (RICM) is characterized by severe diastolic dysfunction from a stiff hypertrophied left ventricle with (at least initially) normal systolic function. (2, 3) Diastolic dysfunction results from replacement or infiltration of normal myocardium with abnormal noncompliant substances. The etiology can be primary (Loeffler’s hypereosinophilic syndrome, idiopathic restrictive cardiomyopathy, endomyocardial fibrosis), or secondary (amyloidosis, hemochromatosis, glycogen storage diseases). In all cases, pulse wave Doppler shows a restrictive diastolic filling profile. Systolic function is initially preserved or mildly impaired. As the disease progresses systolic function decreases and chamber dilation occurs. As the patient deteriorates further, the clinical picture becomes indistinguishable from dilated cardiomyopathy. Although rare, amyloidosis is the most common etiology for RICM, and it results from abnormal layering of proteins within the heart. This abnormal protein creates a symmetric thickening or “psuedohypertrophy” of the myocardium, which takes on a distinctive 2D appearance often described as ‘granular, speckled or starry skied’ (19). Tissue Doppler analysis of diastolic function differentiates RICM (E_M < 8 cm/sec) from constrictive pericarditis (E_M > 8 cm/sec) which also has a restrictive transmitral pulsed wave Doppler diastolic filling profile. (20)

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