

# Pathophysiology of Cardiomyopathy: Its Diagnosis and Treatment

### Sock Hwee Tan<sup>\*</sup>

Department of Medicine, National University of Singapore, Lower Kent Ridge, Singapore

# DESCRIPTION

Cardiomyopathy is a heterogenous group of diseases of myocardium, associated with mechanical or electrical dysfunction which is usually but not invariably exhibits inappropriate ventricular hypertrophy or dilation. Restrictive Cardiomyopathy (RCM) is either an idiopathic or a systemic myocardial disorder. It involves the restriction of ventricular filling. This leads to reduced ventricular volumes or nearly normal systolic function. This is the least common type of cardiomyopathy. Poor ventricular compliance is the major abnormality in restrictive cardiomyopathies and inadequate filling of the ventricular cavities occurs during diastole and results in clinical manifestations.

Restrictive cardiomyopathy is classified as primary and secondary. Primary cardiomyopathy is resulting from genetic abnormalities of cardiac muscle which includes dilated, hypertrophic and restrictive types. Secondary cardiomyopathy is resulting from infections, metabolic and nutritional diseases, endocrine disorders, neuromuscular diseases, blood diseases and tumors. Dilated cardiomyopathy involves the dilation and impaired contraction of the left or both ventricles. In hypertrophic cardiomyopathy, left or right ventricular hypertrophy is often symmetrical, which usually involves the interventricular septum.

Restrictive cardiomyopathy is defined as the restricted filling and reduced diastolic size of either or both ventricles with normal or near-normal systolic function. Arrhythmogenic right ventricular cardiomyopathy is a progressive fibro-fatty replacement of the right ventricle.

Signs and symptoms are volume overload causes fatigue, dyspnea, orthopnea and nocturnal dyspnea, arrhythmia causes palpitations, syncope and exercise intolerance, reduced cardiac output leads to cognitive difficulties, angina, sudden cardiac death, tachycardia, bradycardia, raised jugular venous pressure.

#### Diagnosis

Some of the diagnostic studies performed are Chest X-ray (CXR), Electrocardiogram (ECG), Magnetic Resonance Imaging (MRI), cardiac catheterization, laboratory diagnosis of amyloid protein and endomyocardial biopsy. **Chest x-ray:** Normal sized ventricles, atrial enlargement, enlarged pericardial silhouette is observed due to pericardial effusion and also other signs of pulmonary congestion.

**Electrocardiogram:** Low voltage QRS complexes, atrial fibrillation, atrial and ventricular dysrhythmias, bundle branch blocks, non-specific ST segment/T wave changes and pathological Q waves are the changes observed.

**Echocardiogram:** It shows impaired filling, RV/LV free wall and septal thickening, thickened valves, presence of pericardial effusion and reduced mitral annular doppler tissue velocities.

**Cardiac magnetic resonance:** Late Gadolinium Enlargement (LGE) is observed. Amyloidosis diffuse the left ventricular subendocardial or transmural LGE and may also cause LGE involving the right ventricle and atrial walls. Myocardial iron overload can be assessed by cardiac T2 measurements. In addition, similar to echocardiography, CMR-based tissue tracking can be used to differentiate restrictive cardiomyopathy from constrictive pericarditis.

**Cardiac catheterization:** It is the square root sign/dip and plateau configuration. It is observed in both constrictive pericarditis and filling pressure of left ventricle larger than the right ventricle favors the diagnosis if restrictive cardiomyopathy rather constrictive pericarditis.

#### Treatment

Amiodarone is used to maintain the sinus rhythm and the atrial fibrillation. Diuretics and ACE inhibitors are used to treat heart failure symptoms. Most are irreversible and require cardiac transplantation, regardless poor prognosis pacemaker for conduction system disease. Autologous hematopoietic cell transplantation in conjunction with melphalan therapy is used. Heart transplantation is used only if the patient has isolated cardiac amyloid. Glucocorticoids are thought to halt or slow process of inflammation and fibrosis.

## CONCLUSION

RCM is a relatively rare form of cardiomyopathy with diverse inherited and acquired causes and manifestations. A high index of suspicion is essential to recognize early-stage RCM so that

Correspondence to: Sock Hwee Tan, Department of Medicine, National University of Singapore, Lower Kent Ridge, Singapore, E-mail: Sockhweetan9666@hotmail.com

Received: 01-Feb-2023; Manuscript No. AOA-23-21966; Editor assigned: 03-Feb-2023; PreQC. No. AOA-23-21966 (PQ); Reviewed: 17-Feb-2023; QC. No. AOA-23-21966; Revised: 24-Feb-2023; Manuscript No. AOA-23-21966 (R); Published: 03-Mar-2023, DOI: 10.35248/2329-9495.23.11.324

Citation: Tan SH (2023) Pathophyiology of Cardiomyopathy: Its Diagnosis and Treatment. Angiol Open Access. 11:324.

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effective treatment can be initiated and prognosis potentially improved. Amyloid cardiomyopathy presents as a hypertrophic and restrictive cardiomyopathy, angina, and is associated with a high mortality. Hemochromotosis and alcohol toxicity are reversible forms of secondary cardiomyopathies. The anthracyclines doxorubicin and daunorubicin are associated with cardiac toxicity.