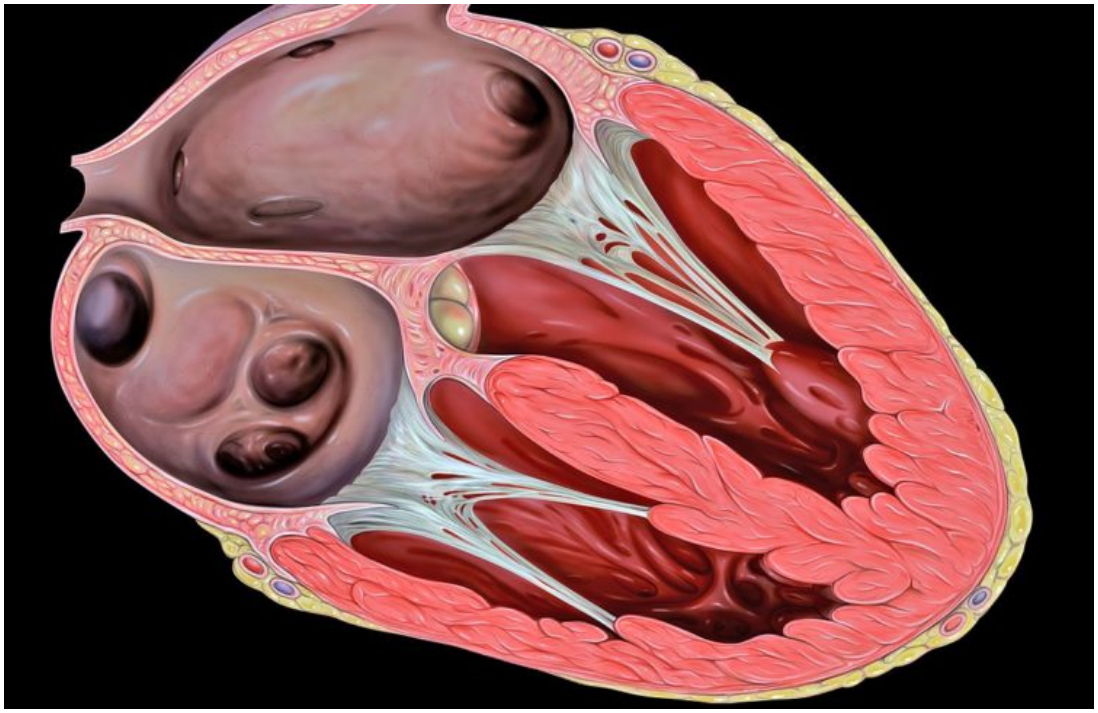
Cardiomyopathy (Heart Muscle Disease) — Symptoms and Causes

See online here

Though cardiomyopathies are not necessarily among the most studied topics, they appear quite often on exams and are actually one of the most common causes of heart insufficiency. Here, you get an overview of the important types of cardiomyopathies usually covered in exams–easy to understand and a huge time-saver!



Definition and Classification of Cardiomyopathy

Cardio = heart

Myo = muscle

Pathy = disease

As the name indicates, cardiomyopathy refers to a group of myocardial diseases associated with impaired systolic and diastolic function.

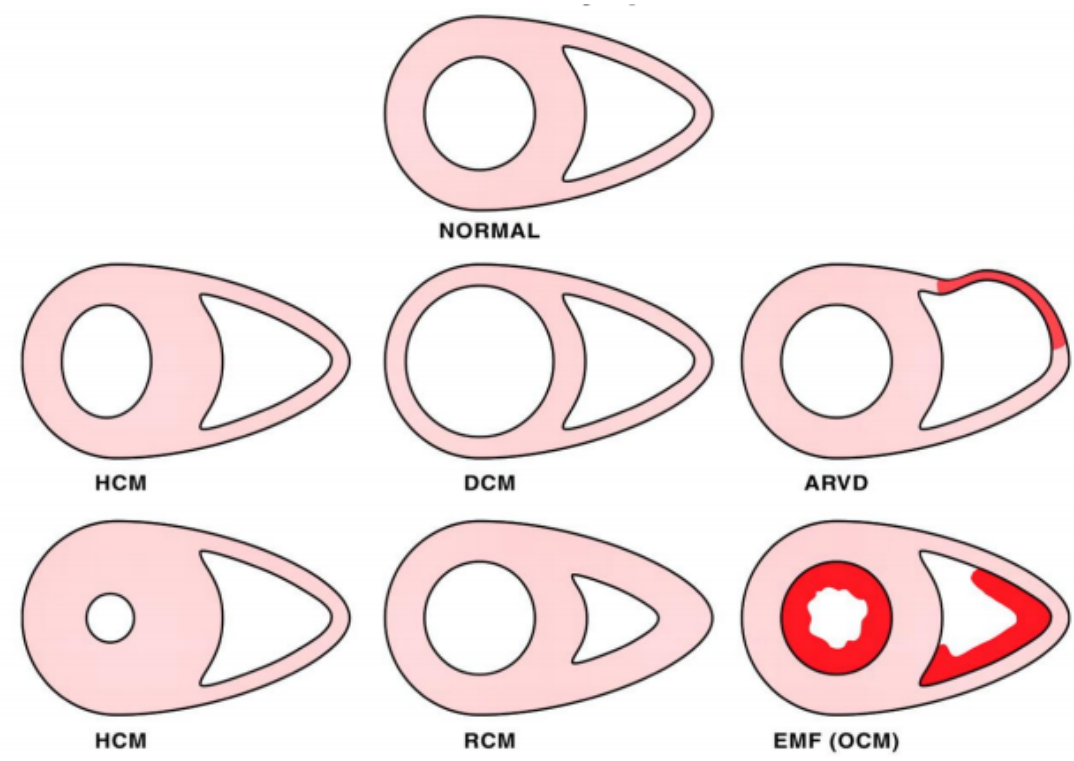
Two types of cardiomyopathy:

1. Primary type: heart muscle diseases predominantly involving the myocardium and of unknown etiology
    - Genetic: hypertrophic myopathies, arrhythmogenic right ventricular

dysplasia, glycogen storage disorders, mitochondrial myopathies
        - Acquired: inflammatory cardiomyopathies (myocarditis), peripartum cardiomyopathies, tachycardia-induced cardiomyopathies
        - Mixed: idiopathic dilated cardiomyopathies, restrictive cardiomyopathies
2. Secondary type: myocardial disease of known cause/associated with a systemic disease, for example, amyloidosis, chronic alcoholic use, etc.
        - Infiltrative cardiomyopathies including amyloidosis
        - Storage cardiomyopathies including hemochromatosis
        - Toxicity cardiomyopathies induced by drugs, e.g., chemotherapeutic agents
        - Inflammatory cardiomyopathies including sarcoidosis
        - Endocrine cardiomyopathies including hypo- or hyperthyroidism, and diabetes
        - Neuromuscular/neurological myopathies
        - Autoimmune myopathies including systemic lupus erythematosus, rheumatoid arthritis, and scleroderma

The WHO classifies 5 types based on cardiac changes:

- Dilated cardiomyopathies (DCM)
- Hypertrophic non-obstructive or hypertrophic obstructive cardiomyopathies (HNCM or HOCM)
- Restrictive cardiomyopathies (RCM)
- Arrhythmogenic right ventricular cardiomyopathies (ARVC)
- Unclassified cardiomyopathies

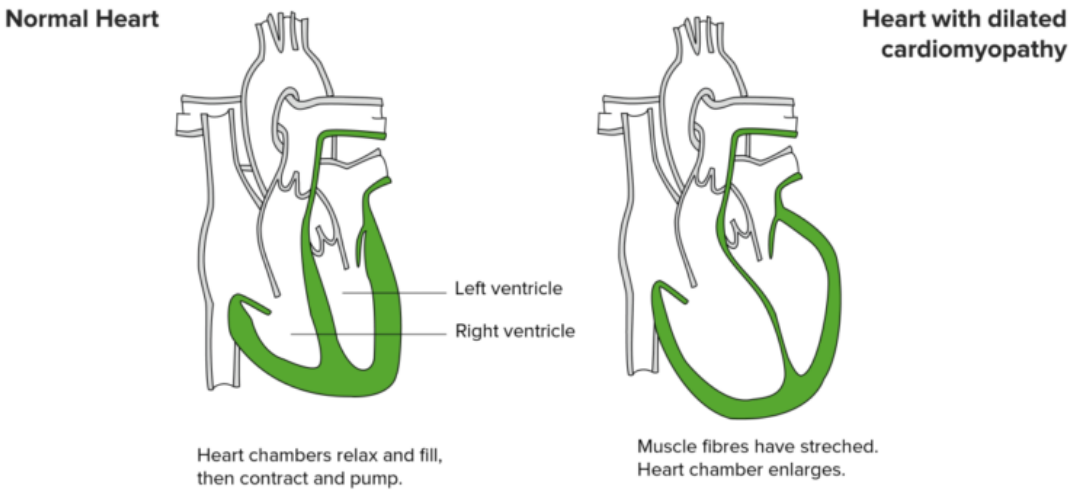


Different cardiomyopathies. Image by Lecturio.

Potential complications include heart failure, arrhythmias, and sudden death. Cardiomyopathy is the second most common cause of sudden death (ischemic heart disease is #1). In the past, few cardiomyopathies were thought to be genetic in origin;

currently, at least 1/3 of cardiomyopathies are known to be genetic. Prognosis for dilated cardiomyopathy is poor, especially when diagnosed late in the course when heart failure signs and symptoms are present.

Dilated Cardiomyopathy



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The most common type of non-ischemic cardiomyopathy is DCM, which has an incidence of 6/100,000 and affects more men than women. Causal processes are usually unknown; however, it is easily identified on the basis of an enlarged left-ventricular diameter observed via echocardiography and rarefication of cardiomyocytes observed via microscopic examination. DCM can be inherited or be a result of underlying conditions. Diseases such as diabetes, viral heart infection, thyroid disease, or abnormalities in the valves of the heart may lead to DCM. Postpartum cardiomyopathy occurs in women after giving birth and is associated with a sudden change in cardiovascular function and blood volume, though the exact cause is not known.

Patients having symptoms of a heart insufficiency–i.e. dyspnea, lung edema (left ventricular insufficiency), or blood stasis in the peripheral body (right ventricular insufficiency)–should also be observed for the likelihood of DCM. Other symptoms include pitting edema, weight gain, dizziness, fainting, and shortness of breath.

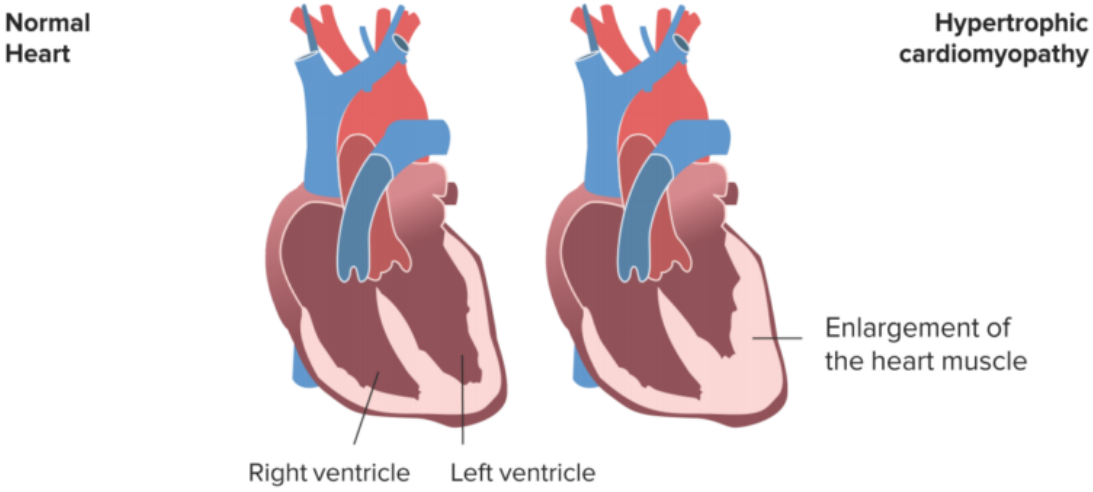
ECG, blood tests, X-rays, magnetic resonance imaging (MRI), myocardial biopsy, CT scans, and cardiac catheterization are some of the tests that may help diagnose DCM cardiomyopathy.

Treatment

Consequently, it is treated (correspondent to heart insufficiency) using diuretics, vasodilators, ACE-inhibitors, and beta-blockers which delays the progression of the disease and lowers the risk of sudden cardiac death. Such treatment also aims at improving cardiac function as well as alleviating most of the symptoms. A lifestyle change is usually necessary, such as including exercise as well as changes in diet, to improve treatment outcomes. Additionally, if arrhythmic side effects occur, the implantation of cardiac resynchronization systems is indicated. If the left ventricular ejection fraction (LVEF) falls below 35% during the course, oral anticoagulation using vitamin K

antagonists such as warfarin and an ICD is necessary. However, there are asymptomatic patients as well.

Hypertrophic Cardiomyopathy



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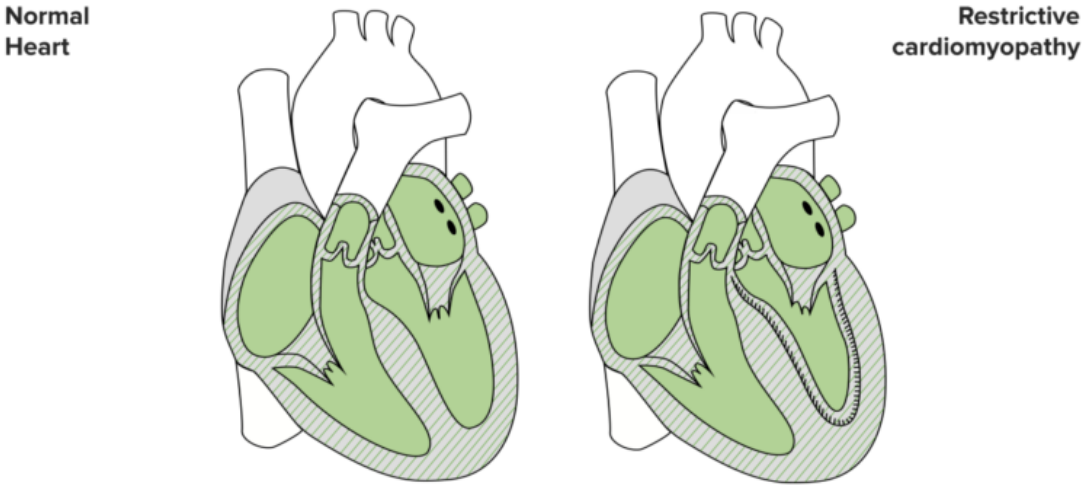
Hypertrophic cardiomyopathy can be diagnosed by 2-D echocardiography, chest radiography, radionuclide imaging, or cardiac MRI.

Treatment of hypertrophic cardiomyopathy

- B-blockers, verapamil (calcium channel blocker), disopyramide, amiodarone have been found useful
- Beta blockers in high dose are the #1 therapy to be tried
- No pharmacological treatment is known to improve prognosis
- Atrial and ventricular arrhythmias are common and often respond to anti-arrhythmic drug therapy including beta-blockade
- Outflow tract obstruction can be improved by:
    - Partial surgical resection (myectomy)
    - Iatrogenic infarction of basal septum (septal ablation with alcohol)
- Implantable cardiac defibrillation (ICD) for patients with risk of sudden death

Among the named therapy approaches for DCM, at this a Morrow myectomy or an interventional TASH is expedient. The latter describes the identification of an arterial branch that feeds the bulge and subsequent injection of alcohol to devitalize the muscular mass.

Restrictive Cardiomyopathy



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Though RCM is fairly uncommon in western countries, it is commonly observed in tropical regions. Contrary to DCM, RCM is a diastolic malfunction caused by an increased rigidity (i.e. reduced compliance) of the heart's wall due to increased connective tissue, leading to increased pressures in atria and ventricles. RCM can also be caused genetically and has been shown to be curable using molecules that silence genes responsible for abnormal protein production.

RCM is difficult to recognize by echocardiography, though there are clinical symptoms of heart insufficiency which serves as the basis for diagnosis.

Common causes of RCM are infiltrations of the myocardium, storage diseases, and endo-/myocarditis. Here, the Loeffler endocarditis is to be named to a special degree, which leaves scars and that way causes the rigidity. In this case, cortisone must be added to conventional medicinal therapy. Moreover, excision of the endocardium belongs to the surgical treatments. Corticosteroids are used to treat sarcoidosis and Loeffler endocarditis. Chemotherapy is used to treat amyloidosis. Endarterectomy treats endomyocardial fibrosis as well as Loeffler endocarditis.

Diagnosis is often difficult	Therapy
• Difficult to distinguish from constrictive pericarditis • Echo/doppler and MRI can make the diagnosis many times, BUT endomyocardial biopsy may need to be done to confirm the diagnosis	• Diuretics to combat pulmonary and systemic congestion • Usually, the prognosis is poor • Cardiac transplantation is indicated for patients with severe heart failure secondary to restrictive cardiomyopathy

Arrhythmogenic Right Ventricular Cardiomyopathy

The most uncommon type is the ARVC which is an inheritable type of cardiomyopathy associated with ventricular arrhythmia and tachycardia. ARVC can lead to sudden cardiac death.

Therapy includes antiarrhythmic drugs, ICD-implantation, and catheter ablation. Physical stress must be avoided. Patients with ARVC and a family history of the disease require changes in lifestyle as well as regular testing.

It is important to understand the fundamental pathomechanisms and types of therapy associated with cardiomyopathies. Indeed, details and numbers can leave behind a good impression; however, don’t lose yourself in details. Aside from that, the morphological differences between the different types can be memorized quite well by visualization.

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Notes