4.2.3

Cardiomyopathies

4.2.3 – Cardiomyopathies

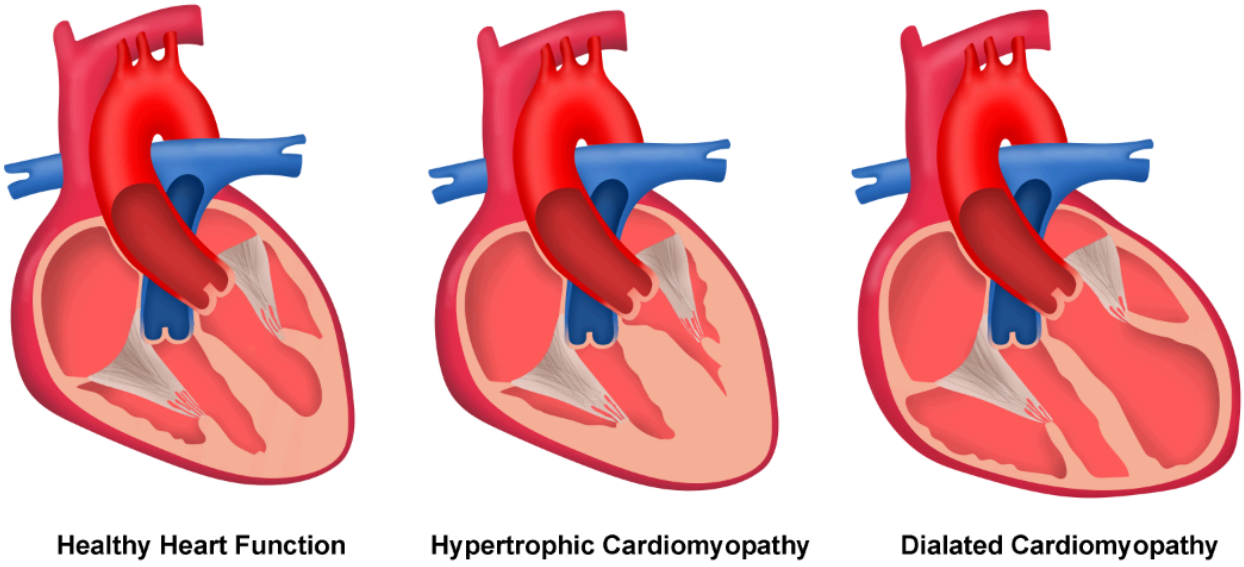

Image by Becky T F19

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is unexplained left ventricular hypertrophy. HCM appears to be a genetic disorder caused by mutations in genes that code for proteins of cardiac sarcomeres. Histologically, those with HCM show hypertrophied cardiac myocytes with disorganized myofibrils. This hypertrophy of the cardiomyocytes can cause left ventricular outflow obstruction because as the walls of the ventricle thicken, the chamber size of the ventricle shrinks. The decreased size of the left ventricle cavity prohibits an adequate amount of blood from filling the left ventricle, and as a result the heart is unable to pump the necessary stroke volume to maintain adequate cardiac output. As the medial wall of the ventricle (the septum) gets excessively large, it can cause additional blockage where outflow from the ventricles should occur (at semilunar area). This also decreases stroke volume.

Common symptoms of HCM include dyspnea (difficulty breathing), chest pain, syncope, and cardiac arrhythmias. HCM is the number one cause of sudden cardiac death in young athletes. Septal myectomy surgery to decrease the muscle thickening in the septum and widen the outflow tract from the left ventricle to the aorta can be useful in the treatment of

HCM. Note that as you read on, you should see that HCM is most likely to be a diastolic/preserved ejection fraction type of heart failure.

Dilated Cardiomyopathy

Dilated cardiomyopathy (DCM) is another cause for heart failure. With DCM, the cardiomyocytes have become damaged and died off. Some common causes for DCM that lead to damage and destroyed cardiomyocytes include genetic mutations, infections, toxins, alcoholism, chemotherapeutic drugs, and heavy metals. As the remaining heart cells spread out and become thinner to fill the void left by the destroyed cells, the ventricular wall thins as well. Over time, a stretched out and larger ventricular cavity volume can arise as ventricular pressure pushes out on the ventricular wall (especially during contraction). This dilated state leads to a reduced stroke volume because even though the ventricular chamber size is increased, the thin walls of the heart are not strong enough to maintain adequate pumping strength. The decreased stroke volume caused by contraction weakness is called systolic heart failure.

Common clinical manifestations of DCM include dyspnea, orthopnea (difficulty breathing when lying down) and reduced exercise capacity. Patients with DCM often have an ejection fraction of less than 25% (normal is 50-70%). They have increased risk for developing a thrombi because of stasis of blood in the heart chambers. When the heart chambers enlarge, they tend to separate out the valves that divide the atria from the ventricles. The stretched valves may not close all the way, and thus blood regurgitates back into the atria. This regurgitation manifests as AV (atrioventricular) valve murmurs. Arrhythmias can also be a problem because stretching the heart wall can influence conduction cells and action potential propagation.

Treatment for both HCM and DCM include diuretics that reduce cardiac preload, beta blockers that reduce cardiomyocyte oxygen consumption and can help with reflex tachycardia, and ACE inhibitors to help prevent afterload by reducing vasoconstriction and decreasing blood volume. Anticoagulants are used as well to prevent thrombus formation, while antiarrhythmic drugs are used to prevent arrhythmias. A left ventricular assist device (LVAD) can be implanted in severe cases to assist in the left ventricle pumping function. Cardiac transplantation is required for patients who do not respond to these treatments.

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