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Fall 2014

### Hypertrophic Obstructive Cardiomyopathy

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# Hypertrophic Obstructive Cardiomyopathy

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## What is Hypertrophic obstructive cardiomyopathy?

Hypertrophic obstructive cardiomyopathy or HOCM, is a genetic disease that is described as hypertrophy or enlargement of the left ventricle which obstructs the blood flow to the body (Yue-Cheng et al., 2012). HOCM is a disease that cannot be cured, but can be treated. One of the main goals of treatment is to focus on treating the symptoms. HOCM can be asymptomatic or symptomatic and can cause some major complications. Some of the complications of HOCM are arrhythmias and sudden cardiac death, "which is defined as natural death due to cardiac causes, heralded by abrupt loss of consciousness within one hour of the onset of acute symptoms" (Spirito et al, 2009, p. 1704).

Normal Heart



(Lookfordiagnosis.com, 2014)

HCM/Obstruction



(Lookfordiagnosis.com, 2014)

## Signs and Symptoms

Patients with HOCM can range from having no symptoms at all, asymptomatic, to being very symptomatic becoming restricted. Some of the common symptoms of HOCM are:

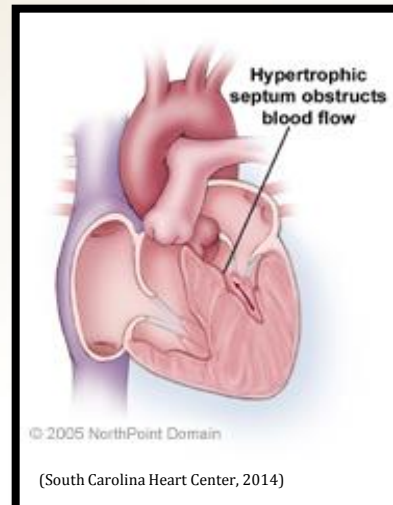
- Shortness of breath. "Most of the time shortness of breath correlates to the heart not being able to increase cardiac output upon exertion (Whitten, 2008, p. 47-48).
- Chest pain. Decrease perfusion in the microvascular circulation plays a role in the cause of chest pain (Whitten, 2008, p. 47-48).
- Syncope. Syncope occurs from decreased cerebral perfusion (Whitten, 2008, p. 47-48).
- Arrhythmias. Patients with HOCM will sometimes have arrhythmias due to ventricular remodeling, decrease cardiac output, microvascular ischemia, and hypotension. "The most common types of arrhythmias seen with HOCM patients are Atrial Fibrillation (A fib), Atrial Flutter (A flutter), supraventricular tachycardia (SVT), ventricular tachycardia (VT), and heart blocks (Whitten, 2008, p. 47-48).
- The most drastic symptom of HOCM is sudden cardiac death (SCD), which is common in young patients and previously asymptomatic patients (Prinz, Farr, Hering, Horstkotte, & Faber, 2011)."

## Pathophysiology and Diagnosis of HOCM

"HOCM is an autosomal dominant disease characterized by left ventricular (LV) hypertrophy and left ventricular outflow tract obstruction" (Yue-Cheng et al., 2012, p. 86) in the absence of an underlying condition causing hypertrophy (Ommen, 2011). In HOCM the myocardial sarcomere proteins result in muscle disarray and fibrosis, ultimately causing inappropriate left ventricular hypertrophy (Ommen, 2011). This abnormal gene is causing the muscle of the heart to thicken. The wall dividing the right and left ventricle becomes thick and gets in the way or obstructs the flow of the blood to the rest of the body. (Hypertrophic Cardiomyopathy, 2014). Because of the hypertrophy LV during the systolic period the hypertrophic interventricular septum protrudes into the left ventricular outflow tract (LVOT) and the anterior mitral valve leaflet moves forward, causing the LVOT to become narrowed or obstructed (Yue-Cheng et al, 2012 p. 88). The obstruction decreases coronary perfusion pressure and causes decreased in cardiac output (Whitten, 2008,).

Hearing a systolic murmur or holding an electrocardiograph (ECG) that looks abnormal could possibly indicate that the patient has HOCM. Confirmation of HOCM is made with an echocardiography. The echocardiography shows LV hypertrophy. Once the diagnosis of HOCM is made screening should be done for the immediate family for the autosomal dominant disorder (Nishimura & Holmes, 2004).

"HOCM is an autosomal dominant mutation of genes" (Gersh et al, 2011 p. 2710) and genetic testing is done to identify any relatives that may be affected with this disorder. Genetic testing is used to determine which first-degree family members are at risk for developing HOCM (Gersh et al, 2011). Genetic counseling can be done prior to the testing and will help the family members better understand the reasoning for the test, risk and benefits for taking the test, and can help the response of learning that an individual in the family does have a mutation of the gene (Gersh et al, 2011).



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(South Carolina Heart Center, 2014)

## Medical Therapy

Treatment is done to relieve the symptoms. In HOCM the goal is to improve the signs and symptoms by "decreasing the heart rate, decreasing outflow obstruction, decreasing the oxygen demand, improving the LV relaxation, and improve filling parameters and preventing major complications" (Whittwen, 2008, p49). Initially the first medication used will be a beta-blocker to help slow the heart rate to enhance diastolic filling (Nishimura & Holmes, 2004). If the patient cannot tolerate beta-blockers, a calcium channel blocker like Verapamil will be used to help improve the symptoms of HOCM (Nishimura & Holmes, 2004), and Verapamil can also improve the function of the LV (Hamada, Ikeda, & Shigematsu, 2014). "Beta-blockers are effective in relieving clinical symptoms, and calcium antagonists, such as Verapamil, are effective in attenuating LV diastolic dysfunction" (Hamada, Ikeda, & Shigematsu, 2014, p. 3). Antiarrhythmic medications such as Amiodarone, can be used in HOCM patients who have developed arrhythmias like A fib (Maron & Maron, 2013).



## Surgical Treatment

When pharmacological treatment is not enough, other options need to be considered. Two invasive procedures, surgical myectomy and percutaneous transluminal septal myocardial ablation (PT SMA), can be done to patients where medication is not working. "Surgical myectomy involves excision of a rectangular part of the thickened subaortic septum. Surgical myectomy ameliorates signs and symptoms in about 70% of patients" (Whitten, 2008, p. 50). PT SMA was introduced in 1995 as an alternative to myectomy and has been shown to reduce the LVOT obstruction and associated symptoms" (Jensen et al, 2011 p256). "PT SMA involves injecting ethanol into one or more the septal perforator arteries, producing a controlled infarction of the myocardial septum. A successful PT SMA results in septal thinning with reduction in the LVOT obstruction" (Whitten, 2008, p. 50). Between the two procedures surgical myectomy is the treatment of choice with HOCM patients. PT SMA is recommended for older patients or patients that absolutely do not want to have surgery (Maron & Maron, 2013).

## Implications for Nurses

"Nurses need to understand the pathophysiology, management, features, and complications of HOCM" (Whitten, 2008, p. 50). Nurses should talk with the patient regarding their medications and why they are on them. Nurses should also be aware of what HOCM is and the symptoms of it and what some of the major complications of HOCM are. When patients arrive to units post procedure, nurses should be monitoring vital signs, assessing the access site for signs and symptoms of bleeding, swelling, firmness and infection.

## Conclusion

HOCM is a disease that will require lifetime treatment and management. HOCM can be defined as thickening of the left ventricle with obstruction of blood flow. Chest pain, syncope, and shortness of breath are common symptom to see in patients with HOCM. HOCM can be treated with medications and surgical procedures for advanced cases where medications no longer work. "Nurses play a role in the knowledge of HOCM and how to detect problems early on" (Whitten, 2008 p. 52)

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