EM CASE OF THE WEEK.

BROWARD HEALTH MEDICAL CENTER
DEPARTMENT OF EMERGENCY MEDICINE

BROWARD HEALTH
Care Warriors

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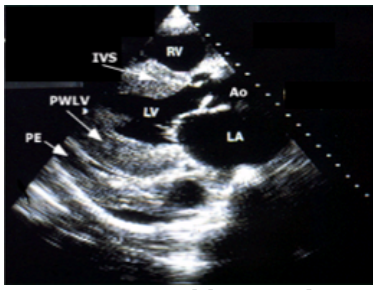
Hypertrophic Cardiomyopathy

A 19-year-old female with a past medical history of hypertrophic cardiomyopathy and Wolff-Parkinson-White syndrome presents to the ED with several episodes of palpitations, each about three seconds in duration, with initial onset approximately five hours prior. She has experienced these symptoms several times in the past, and had a pacemaker/AICD placed three years prior to this evaluation. She did not feel her defibrillator activate. She is otherwise asymptomatic, denying chest pain, light-headedness, weakness, or diaphoresis. Patient is afebrile and vitals are within normal limits. On physical exam, the patient is in no acute distress. Her heart is maintaining a regular rhythm and no murmurs are noted. The remainder of her physical examination is within normal limits. Her ECG is consistent with left ventricular hypertrophy and is unchanged from prior ECGs. Which of the following is the most appropriate initial step in the treatment of this patient's condition?

A. Immediately call interventional cardiology for cardiac catheterization.
B. Have the AICD interrogated and notify the patient's cardiologist.
C. Give the patient anti-platelet therapy and repeat the ECG in an hour.
D. Give the patient a one-liter bolus of IV fluids to maintain left ventricular diastolic filling, then re-evaluate.
E. Discharge the patient after explaining to her that symptoms are inevitable and are the reason why she has the implanted device.



Parasternal long axis echocardiogram in a patient with hypertrophic cardiomyopathy UpToDate, 2017.

Hypertrophic cardiomyopathy is a genetic heart muscle abnormality of varying severity that can lead to symptoms such as chest pain, palpitations, and can cause sudden cardiac death. Shown above is image of an echocardiogram demonstrating hypertrophy of the interventricular septum (IVS).

EM Case of the Week is a weekly "pop quiz" for ED staff.

The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

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The correct answer is B, have the AICD interrogated and notify her cardiologist.

Hypertrophic cardiomyopathy (HCM) is a genetic disease that is caused by mutations in the genes regulating cardiac muscle. There are over 1500 gene mutations associated with HCM. This heterogeneity leads to a wide variety of clinical manifestations and disease severity. It can lead to both systolic and diastolic dysfunction, ischemia, and valve disorder. Clinical presentations include but are not limited to chest pain, palpitations, shortness of breath, and syncope. HCM can cause sudden cardiac death if hemodynamic obstruction occurs.

Discussion

Symptoms can occur due to obstruction of cardiac output, arrhythmia, ischemia, or failure to achieve sufficient diastolic filling. Physical examination of patients with HCM may be completely normal if the patient is not experiencing cardiac output obstruction. If left ventricular output obstruction is present, a harsh systolic murmur heard best at the apex might be detectable. Physical exam maneuvers that increase preload, such as passive raising of the legs, increase diastolic filling and can reduce the murmur's intensity. Heart failure is the most common presentation of HCM. One-quarter to one-third of patients with HCM experience angina. Acute myocardial infarction in patients with HCM is more likely to present without ST segment elevation. Arrhythmias in HCM can originate from the atria or ventricles and present with palpitations, episodes of syncope, and shortness of breath. HCM should be considered in the diagnostic approach to patients presenting with syncope, particularly in young patients. Ventricular arrhythmias are a significant threat to patients with HCM, as they can lead to sudden cardiac death when they are sustained. Sudden cardiac death in HCM results from acutely decreased cardiac output as a result of either decreased preload or ineffective contractility.

Decreased preload can result from hypovolemia, such as in dehydration, hemorrhage, or mitral regurgitation, and ineffective contractility can result from arrhythmia. Diagnostic testing when HCM is considered include ECG and echocardiogram. If HCM is diagnosed, ambulatory ECG monitoring and stress testing for risk assessment may also be performed. ECG findings often include left ventricular hypertrophy with repolarization abnormalities and Q waves in the inferior and/or lateral leads. "Giant" inverted T waves are sometimes seen in the precordial leads. On echocardiogram, diagnosis of HCM is confirmed with measurement of left ventricular wall width >15mm, or 13mm in a patient with family history of HCM. Echocardiography can also accurately and noninvasively measure the degree, if any, of outflow obstruction.

Treatment

Treatment of HCM has not been evaluated in any large clinical trials. The clinical approach to symptomatic patients is based on reduction of outflow obstruction, decreasing myocardial oxygen demand, and increasing left ventricular diastolic filling. Beta blockers are a commonly preferred first choice for treatment. It is also important for patients to avoid volume depletion, which leads to impaired diastolic filling and reduced cardiac output. Vasodilators and diuretics, if necessary, should be used with caution. Arrhythmias render patients vulnerable to reductions in cardiac output, and therefore should be treated aggressively. While guidelines currently do not recommend routine antibacterial prophylaxis in patients with HCM, many experts do favor administering prophylaxis before dental procedures. If symptoms persist despite medical management, alternatives include alcohol ablation and surgical myectomy. Patients with HCM should undergo risk stratification to determine if AICD is indicated. AICD should be placed in patients with a history of SCD survival or sustained ventricular tachycardia. AICD should also be placed in patients with multiple risk factors for SCD.

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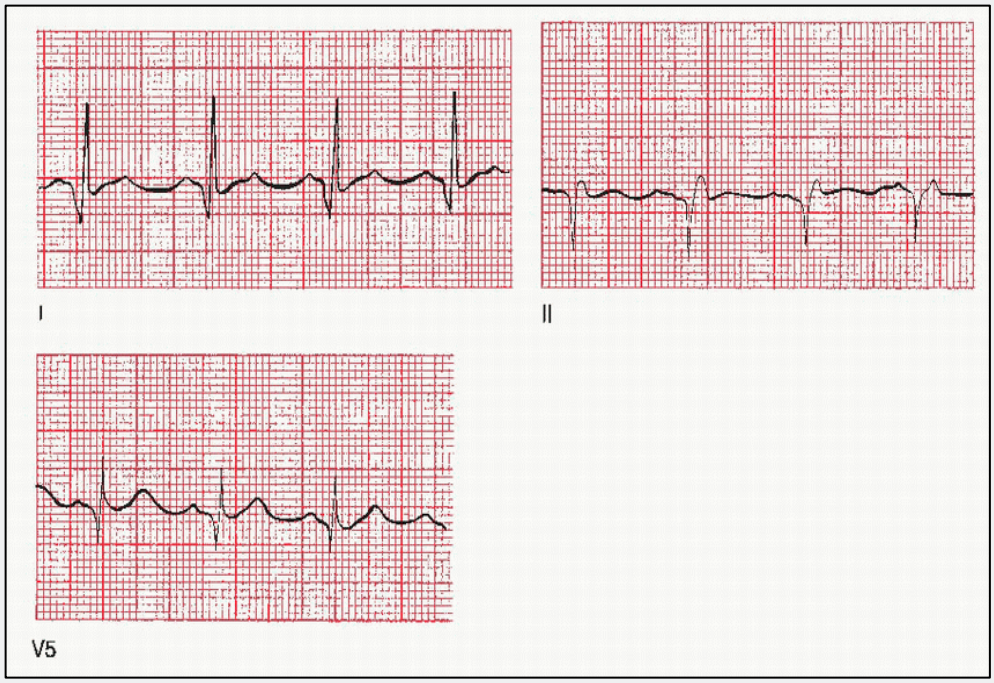
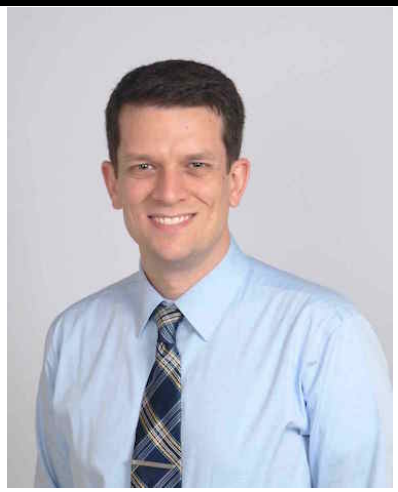


This diagram illustrates the prominent Q waves that may be found in the inferior and lateral leads of patients with HCM. ECGs in HCM are typically abnormal. They often show evidence of left axis deviation, left ventricular hypertrophy with repolarization abnormalities, prominent Q waves in the inferior and lateral leads, and occasionally large inverted T waves in the pre-cordial leads.

Take Home Points

- Hypertrophic cardiomyopathy is an autosomal dominant genetic disease occurring in approximately 1 in 500 adults.
- Patients' experiences can vary greatly, from asymptomatic to sudden cardiac death.
- HCM is an important consideration in the assessment of any patient with syncope, but particularly so in young patients.
- ECG and Echocardiogram are important diagnostic tests in the workup of the patient in whom HCM is being considered.
- While no specific ECG pattern is diagnostic, there are several abnormal findings associated with HCM. Echocardiogram is an excellent and inexpensive tool in the diagnosis of HCM.
- The first step in the treatment of HCM is medical management with beta blockers, which can effectively target the pathological physiology in HCM.
- If medical management is ineffective, invasive treatment with alcohol ablation or surgical myectomy can be considered.
- Because HCM can cause sudden cardiac death, AICD placement is indicated in some patients and therefore all patients should be risk stratified with a thorough medical history, physical examination, and pertinent diagnostic testing.



ABOUT THE AUTHOR

This month's case was written by Garrett Van Ostran. Garrett is a 4th year medical student from NSU-COM. He did his emergency medicine rotation at BHMC in February 2017. Garrett plans on pursuing a career in Internal Medicine after graduation.

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