

HOCM:

A Classic Hemodynamic Tracing

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CardioCase presentation

John's Sudden Pains



John, 42, is an active man who plays hockey regularly. One evening, he is brought to the emergency department after complaining of first-time chest pains and shortness of breath during a particularly competitive game.

His physical exam on arrival reveals a 3/6, late peaking, systolic murmur along the left sternal border. The murmur intensifies with the Valsalva manoeuver.

His chest pain resolves spontaneously. His ECG is shown below.

For more about John, see page 20.



Figure 1. John's ECG, showing normal sinus rate and rhythm, prominent voltages compatible with left ventricular hypertrophy and anterolateral T wave inversion.

About the authors...

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CardioCase presentation

John's Sudden Pains continued...

John's cardiac enzymes are normal. His ECG reveals asymmetric left ventricular hypertrophy. A subsequent angiogram reveals normal coronary arteries and the following hemodynamic tracing (Figure 2).

For John's followup, see page 21.

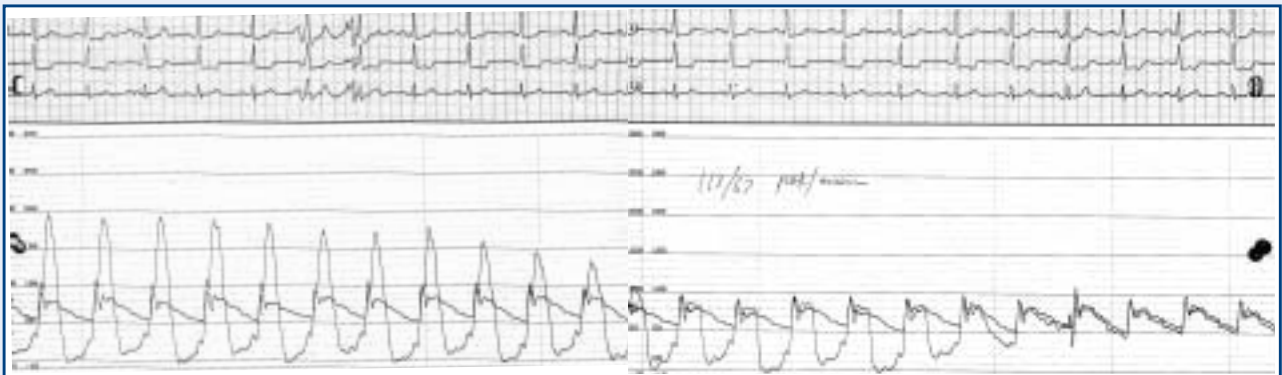


Figure 2. John's hemodynamic tracing, showing a very large gradient between the left ventricular (LV) apex and aortic pressures (approximately 90 mmHg), a reduction of pressure gradient as the catheter is withdrawn to the LV outflow tract and equalization of pressures as the catheter is pulled back across the aortic valve.

What's your CardioCase diagnosis?

CardioCase discussion

What's wrong with John?

Hypertrophic cardiomyopathy (HOCM) is inappropriate myocardial hypertrophy in the absence of an obvious cause. Hyperdynamic systolic function is common and a dynamic outflow tract gradient is seen in 25% of patients. These gradients are provoked by Valsalva and vasodilators. Manifestations include asymmetric hypertrophy, most commonly of the interventricular septum. Asymmetric hypertrophy of any of the left ventricle (LV) segments may be seen, including an apical hypertrophic variant commonly seen among Japanese HOCM patients.

Signs and symptoms

The majority of patients are asymptomatic and are diagnosed on screening evaluations. Reported symptoms are due to myocardial strain, like:

- dyspnea (90%),
- angina pectoris (75%) and
- fatigue.

Syncope and pre-syncope may be the result of the outflow tract gradient or due to arrhythmias (both ventricular and supraventricular). Sudden cardiac death (SCD) is the

John's Followup

John was initially treated with beta-blockers but continued to complain of shortness of breath. He subsequently underwent alcohol septal ablation with marked improvement in his outflow gradient and symptoms.

most catastrophic event and tends to occur in younger patients, typically during extreme exercise or competitive sports.

The hallmark physical finding is a harsh systolic murmur that varies in intensity, increases with standing and diminishes with squatting. There may be a prominent S₄ due to ventricular non-compliance. In the absence of an outflow tract gradient, the physical exam may be normal.

Etiology and course

Autosomal genetic mutations in many locations have been identified and these tend to occur in family clusters. It is important to screen family members of patients who have been diagnosed with HOCM. It has an approximate 0.2% prevalence, and most patients run a benign course with a 1% yearly mortality. Patients at high risk for SCD tend to be young, and have:

- a family history of SCD,
- a detectable genetic abnormality,
- a markedly elevated outflow tract gradient and
- asymptomatic complex ventricular arrhythmias.

Diagnosis

The diagnosis of HOCM may be suspected by findings of left ventricular hypertrophy on the ECG and confirmed by echocardiography. Asymmetric hypertrophy of an LV segment is the typical finding, though marked concentric hypertrophy (without an identifiable reason) can also be seen. This

may be associated with a dynamic pressure gradient across the outflow tract. Systolic anterior motion of the mitral valves during systole, leading to mitral regurgitation, may also be seen.

Management

Beta-blockers are the mainstay of treatment and are used to improve symptoms of chest pain and shortness of breath. Calcium channel antagonists may also be used. Disopyramide is a potent negative inotrope and is also used for symptom control. Amiodarone may be used for complex ventricular and supra ventricular arrhythmias, though these patients should be evaluated by an electrophysiologist. Implantable cardioverter defibrillator implantation is carried out for survivors of SCD or those at high risk for SCD.

The definitive management for symptoms is traditionally through cardiac myomectomy, but alcohol septal ablation via cardiac catheterization has shown very favourable results. 