The *Heart* of the Matter

Cardiology Associates, P.C.

Thursday, October 21

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A Message from Cardiology Associates, P.C.

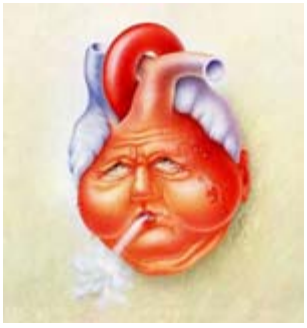
Dear Colleagues,

Our October, 2010 Referring Physician newsletter focuses on the diagnosis and treatment of hypertrophic cardiomyopathy (HCM). This condition causes a thickening of the myocardium and is prevalent in patients of all ages. Hypertrophic cardiomyopathy may cause severe and life-threatening health complications, including sudden heart failure. Dr. William Maxted discusses a case of a recent patient with HCM, and presents diagnosis and treatment options.

About the Author

Dr. William C. Maxted sees patients in our Annapolis and Bowie locations. He is a board-certified cardiologist with a special interest in consultative cardiology, echocardiography, and cardiac pacemakers. Dr. Maxted is a proud member of the American College of Cardiology and the American Society of Echocardiography.

The Inconspicuous Onset of HCM



Presentation of Case

A 45 year old man is referred for an echocardiogram because of an asymptomatic murmur. It shows evidence of a hypertrophic cardiomyopathy. The interventricular septum measures 1.6 cm, the posterior wall 1.1 cm. There is no significant dynamic gradient across the left ventricular outflow tract. How do you evaluate him? What do you tell him about his prognosis?

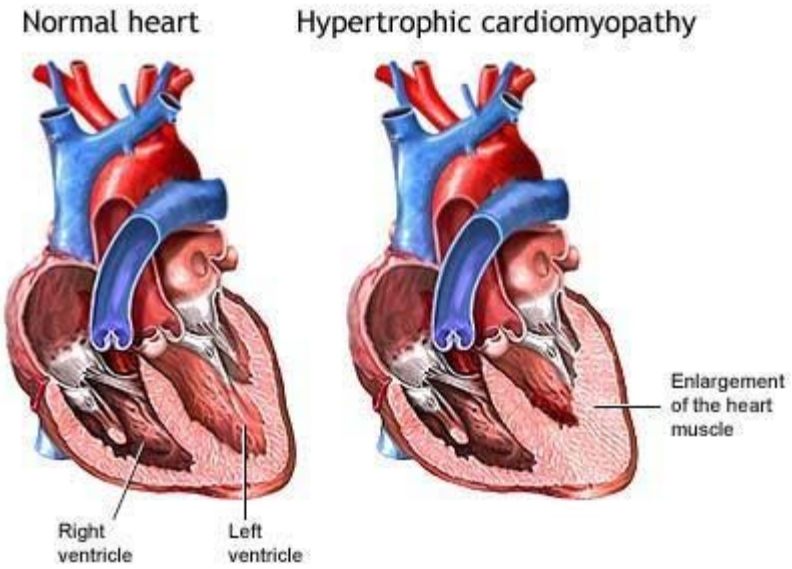
Incidence and Pathology

As the result of hypertrophic cardiomyopathy (HCM), the sarcomeres replicate causing myocardial cells to increase in size and the heart muscle to thicken. In addition, the normal alignment of muscle cells is disrupted, a phenomenon known as myocardial disarray - actually resembling fibroids. HCM also causes disruptions of the electrical conduction and can be associated with sudden death.

HCM is most commonly due to a mutation in one of nine sarcomeric genes that results in a mutated protein in the sarcomere.

Although assymetric septal hypertrophy is the most common manifestation of this entity, any portion of ventricular myocardium can be involved.

While most literature, so far, focuses on European, American, and Japanese populations, HCM appears in all racial groups. The incidence of HCM is about 0.2% to 0.5% of the general population.



Prognosis

The total mortality in all patients with HCM is 1.2 percent/year and does not differ significantly from that expected in the general U.S. population after adjustment for age, gender, and race. There are, however, a collection of risk factors that do increase the risk of sudden death.

Risk Factors

The strongest risk factors for sudden death include:

- A history of cardiac arrest of sustained ventricular tachycardia
- A family history of sudden cardiac death
- Syncope
- Multiple repetitive episodes of NSVT by holter monitor
- Massive LVH (septum >1/9 cm)
- Hypotension with exercise
- End-stage phase
- LV apical aneurysm
- A resting gradient > 30 mm Hg

When these risk factors are absent, the prognosis is generally good. In fact, the older the patient is at the time of their diagnosis, the better their prognosis.

Evaluation

In addition to a history designed to answer questions regarding those issues above, a typical approach for evaluation should include:

- An ECG,
- An echocardiogram to evaluate LV wall thickness as well as for dynamic outflow gradients,
- A holter monitor to evaluate for non-sustained ventricular tachycardia,
- A treadmill test to evaluate for an exercise induced drop in blood pressure.

Treatment

For those patients that fall into the high risk categories outlined above, an ICD is generally indicated. Treatment with beta blockers, calcium channel blockers, and disopyrimide can alleviate symptoms.

There are some patients that continue to be symptomatic despite medical therapy. Those patients can be treated with a surgical myomectomy or alcohol ablation of the septum. Dual chamber pacing has been tried in the past as well, but its success is debatable.

The patient described above had no history of syncope or arrhythmia. He had no family history of sudden cardiac death. He had no drop in blood pressure with exercise and no non-sustained ventricular tachycardia on his holter. Based on all of that, his prognosis was felt to be good. He will be followed clinically annually and by echocardiograms every five years.

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We are offering you this monthly newsletter as a way to provide cardiovascular news and update you on developments within our field. For your convenience, we are distributing our newsletter via e-mail. Visit our site at (www.heartcapc.com) and click the Referring Physician Newsletter link at the upper left corner of our home page. You will receive an e-Newsletter every month featuring an article or a case report from one of our physicians and links to other sources featuring new trends in the field of cardiology. Our focus will be on real questions and issues that we encounter in our day-to-day medical practice. In fact, if there is a topic that is of particular interest to you (or a question that is related to any of our articles) please e-mail your inquiries to our Project Manager, Nazar Snihur at nsnihur@heartcapc.com. (Of course, we will not share your e-mail address outside of our offices.)

Our Locations

Annapolis Cardiology Office
2002 Medical Parkway, Suite 500
Annapolis, MD 21401
Phone: 410-573-6480
Fax: 410-573-9413

Annapolis Vascular Office
2002 Medical Parkway, Suite 520
Annapolis, MD 21401
Phone: 410-571-8430
Fax: 410-573-5981

Bowie Office
4175 N. Hanson Court
Suite 100
Bowie, MD 20716
Phone: 301-809-6880
Fax: 301-805-4233

Irving Street 4800N
106 Irving Street, NW
Suite 4800N
Washington, DC 20010
Phone: 202-726-5484
Fax: 202-726-4587

Kent Island Office
1630 Main Street
Suite 208
Chester, MD 21619
Phone: 410-643-3186
Fax: 410-643-4098

K Street Office
2131 K Street, NW
Suite 800
Washington, DC 20037
Phone: 202-822-9356
Fax: 202-331-0451

Olney Office
18109 Prince Philip Drive
Suite 225
Olney, MD 20832
Phone: 301-774-5810
Fax: 301-774-0188