Penn Medicine

CLINICAL BRIEFING™

Penn Cardiovascular Surgery • Penn Center for Inherited Cardiovascular Disease

Surgical Myectomy for Obstructive Hypertrophic Cardiomyopathy

Cardiac surgeons at Penn Medicine are performing septal myectomy for obstructive hypertrophic cardiomyopathy (HCM), a heritable condition characterized by thickening of the myocardium in the absence of abnormal loading conditions. Hypertrophy of the ventricular septum in conjunction with systolic anterior motion of the mitral valve can lead to obstruction of blood flow from the left ventricle to the aorta. While obstruction is not a universal finding in patients with HCM, approximately 70% of patients with the condition will have outflow obstruction at rest or with exercise.

At Penn Heart and Vascular, the diagnosis and treatment of HCM involves the coordinated care of cardiologists, genetic specialists and surgeons. Together, these specialists develop treatment strategies for the various presentations of HCM. Thus, patients with atrial fibrillation may receive anti-arrhythmic or rate-controlling agents, or invasive treatment including radiofrequency ablation. Patients deemed to be at high risk for sudden cardiac death or life threatening ventricular arrhythmias may be candidates for an implantable defibrillator.

First line therapy for patients with symptomatic outflow tract obstruction (gradient >30 mm Hg) at rest or after exercise provocation includes beta blockers, calcium channel blockers and disopyramide. If medical therapy is not effective and severe outflow tract obstruction remains Penn Heart and Vascular offers septal myectomy and alcohol septal ablation (ASA).

Septal myectomy removes a portion of the obstructing septal tissue to improve blood flow. The surgery is performed in severely symptomatic patients with proven obstructive disease who meet selection criteria. ASA is a percutaneous procedure in which alcohol is selectively delivered into a coronary artery to cause necrosis of the septum in the area of greatest obstruction. Generally, the procedures are considered comparable in safety and efficacy.

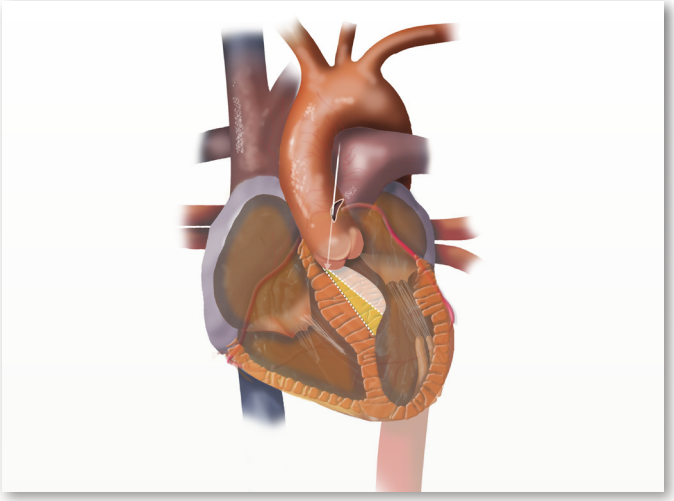

Figure 1: Septal myectomy to treat hypertrophic cardiomyopathy.

CASE STUDY

Mr. G was referred to Penn Heart and Vascular for an evaluation after an episode of severe chest pressure and dyspnea accompanied by a spike in blood pressure sent him to a local ER. At age 60, he had a 25-year history of obstructive hypertropic cardiomyopathy and a more recent history of coronary artery disease, for which he'd received a total of 5 drug-eluding stents. His HCM had progressed since his 30s, when his maximal septal thickness was measured at 1.5 cm. An echocardiogram 3 months before his arrival at Penn found a hyperdynamic left ventricle with severe asymmetric septal hypertrophy (septal wall thickness 3.0 cm) and systolic anterior motion of the mitral valve. His presentation at Penn Medicine was precipitated by the need for additional HCM management and a personal wish to discuss possible surgical interventions for his condition.

At Penn, Mr. G reported dyspnea during mild activity and a steady and limiting decline in stamina, but denied having palpitations, dizziness or syncope. Outside of a harsh systolic murmur, his physical examination and vitals were unremarkable. His EKG revealed sinus bradycardia, biatrial enlargement and left ventricular hypertrophy with repolarization abnormality. Upon completing a 6-minute walk test, he was estimated to have NYHA functional class III heart failure.

After achieving maximally tolerated doses of medical therapy, an echocardiogram revealed a peak left ventricular outflow tract gradient of 193 mmHg with valsalva. Following a lengthy surgical consultation, Mr. G decided to proceed with septal myectomy surgery.

During the four-hour surgery, approximately 3 grams of muscle were removed from the proximal ventricular septum of Mr. G's heart to widen the outflow tract (Figures 1 & 2), resulting in an immediate gradient reduction. From the operating room, he was taken to the surgical intensive care unit, where in the next hours he was weaned off all pressors and inotropic support and extubated without difficulty.

(Continued on the back)

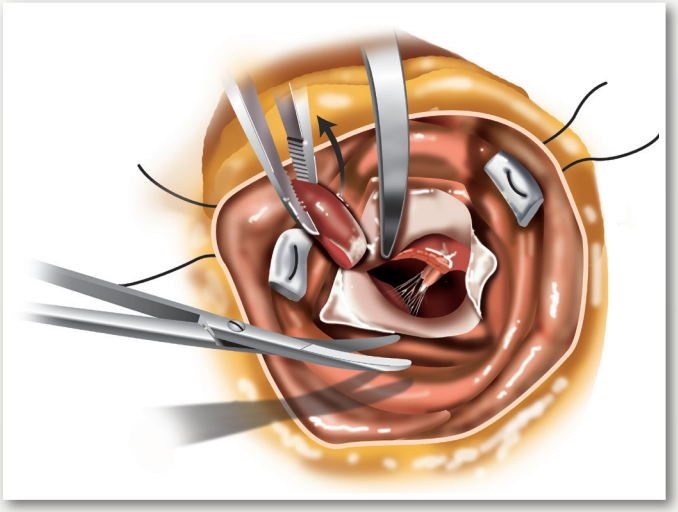

Figure 2: Retraction of right coronary aortic cusp with excision of hypertrophic tissue (arrow) during septal myectomy.

CASE STUDY *(Continued from cover)*

With the establishment of hemodynamic stability, he was transferred to the step-down unit. There, he was diuresed until euvolemic and had his chest tubes and wires removed.

Mr. G remained in-hospital for 7 days, after which he was discharged to home. At his one-month follow-up visit, he reported feeling no dyspnea, orthopnea or dizziness. His energy had improved substantially, and he was walking 2300 steps a day, and noted a marked improvement in breathlessness. His blood pressure was well controlled, and overall, he felt well. It was determined at this visit to have him proceed with cardiac rehabilitation.

FACULTY TEAM

At Penn Heart and Vascular, our specialists are among the select number of surgeons in the nation with the experience and skill necessary to perform septal myectomy. Among the nation's most experienced in all forms cardiovascular surgery, including transplantation, mitral and aortic valve surgery and robotic surgery, Penn cardiovascular surgeons have consistently acquired or developed innovative technologies to complement their skills in the operating room. This combination of technical and clinical expertise, experience and vision drives the constant improvement that defines the division of cardiovascular surgery.

▸ Treating Hypertrophic Cardiomyopathy at Penn Medicine

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