The Hypertrophic Cardiomyopathy Program

A Family Affair
Introducing the Hypertrophic Cardiomyopathy Program

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The NewYork-Presbyterian Hospital/Columbia HCM Program provides seamless care for you and any family members who have HCM, at any age, and at every stage of this disease.

We help you to alleviate your symptoms and live full and active lives while managing this chronic condition. Our collaborative, cross-specialty program brings together leading specialists in pediatric and adult HCM, and we are known for providing exceptional continuity of care.

Nationally known for our expertise in genetics, noninvasive imaging, interventional techniques and cardiac surgery, the HCM team will carefully guide you through your treatment options. Our multidisciplinary team provides genetic testing and counseling to help you better understand how this condition may be passed from generation to generation and why some family members may show no symptoms, yet still need careful monitoring or preventive measures in the years ahead.

Our cardiologists manage this condition with the latest and most effective drugs, tailoring a personalized program that combines lifestyle choices, exercise, diet and nutrition. When necessary, we offer implantable cardioverter defibrillators (ICD) to prevent or...
control arrhythmias, allowing you to feel secure as you pursue your daily tasks.

If you have obstructive HCM—meaning a blockage in the flow of blood from one of the chambers of the heart, we offer comprehensive evaluation and offer the most advanced surgical procedures and minimally invasive options when appropriate. Our interventional and surgical teams are among the most experienced in the country, so you are assured of the best possible outcomes.

At present, there are only a few centers specializing in HCM in this country. Our goal is to provide you with comprehensive and compassionate treatment and the most innovative medical and surgical care. Our physician-scientists are also pursuing leading-edge research into treatments that will help future generations with HCM.

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Our multidisciplinary HCM team works to provide our patients with the highest standard of integrated care. Our collaboration brings together the following experts:

- Advanced imaging specialists who use echocardiography, CT scans and MRIs to refine the diagnosis

- Cardiologists who provide medical management and comprehensive treatment plans for adults with HCM

- Pediatric cardiologists who assess, monitor and provide medical management for HCM, from the pre-natal stage through adolescence

- Geneticists who test patients and their families for genes related to HCM

- Electrophysiologists who implant pacemakers and defibrillators both to prevent and to treat symptoms caused by HCM

- Interventional cardiologists who perform minimally invasive procedures to control symptoms of HCM in those with obstruction

- Cardiac surgeons who perform a "gold standard" operation called a myectomy, removing the overgrown heart muscle to alleviate symptoms and restore blood flow. Our world-renowned surgeons can also correct valve and other heart defects

- NYPH/Columbia has one of the premier heart transplantation programs in the world, and so in the rare situation, we are able to treat the very few patients who progress to end-stage HCM, offering them a new heart, and a new lease on life.
When patients come to our program, we take a complete family and personal medical history, and evaluate the risks for developing symptoms and complications associated with HCM.

The HCM Program offers advanced medical imaging to refine this diagnosis and includes the following services for adults and children:

- Advanced cardiac imaging, including 3D-echocardiography and MRI
- Echocardiography
- Cardiopulmonary exercise studies
- Arrhythmia management and electrophysiology
- Cardiac surgery
- Interventional cardiology
- Heart failure management
- Heart transplantation
- Pulmonary hypertension management

Once we confirm or refine the diagnosis, we educate patients and family members about the nature and course of this chronic illness.

While HCM can involve both lower chambers of the heart, it usually affects the left ventricle (the main pumping chamber). The muscle fibers of the heart thicken, making it harder for the heart to relax and let new blood into the chambers. Because the heart can’t fill completely, it cannot send enough blood to the rest of the body during exercise.
Hypertrophic Cardiomyopathy is an extremely common condition affecting at least one in 500 Americans. This condition can be diagnosed at any time between infancy and seniors.

HCM may be silent and symptoms may vary widely, from mild chest pain and difficulty breathing to serious arrhythmias, conduction problems, and heart failure.

Some patients with HCM may report shortness of breath, chest pain or discomfort, fainting or dizziness especially upon exertion, and palpitations—a rapid or irregular heartbeat. Because none of these symptoms are unique to HCM, the condition is often misdiagnosed as exercise-induced asthma, mitral valve prolapse, or anxiety/depression.

Atrial fibrillation, an irregular rhythm that affects the normal pumping of the heart, is a common complication associated with HCM, as is ventricular tachycardia, and other serious arrhythmias. Blood clots may form in the hypertrophic heart, and if undetected, may cause a stroke, heart attack, or reduced blood flow to the arms and legs.
Once your family is aware of the HCM diagnosis, it is important for everyone to learn about the disease. Our genetic specialists are like detectives who help you understand how the body produces this condition. Their job is to find the exact mutation in your DNA that gives you a propensity to develop HCM, and to express it in a certain clinical way.

HCM is the most common genetic abnormality of the heart. It is an autosomal dominant condition, meaning that parents have a 50% chance of inheriting, or passing on the predisposition to this disorder to their children.

Until recently, the only way to screen family members for HCM was through yearly evaluations with a cardiologist, echocardiogram and ECG testing.

Today, genetic testing is available as an option to determine who among your relatives needs to be followed medically, and who no longer has to be concerned about HCM. Genetic testing can also help with family planning. We offer the option to have an entire family evaluated for HCM, including a genetic profile and clinical evaluation on one day.

The way HCM manifests can vary from one family member to another. The same gene mutation can also manifest in different people at different ages. Further, not everyone with a genetic predisposition will develop HCM. We are presently engaged in clinical research to find out why this is the case, and to help future generations who are genetically at risk.

The advantage of coming to the HCM Program at NewYork-Presbyterian Hospital/Columbia is that we’re not satisfied with today’s answers. We are looking for tomorrow’s answers as well.
If a child has HCM, we will teach parents about the lifestyle restrictions they’ll need to observe to keep the young patient safe, yet still allow participation in normal school activities and gentle, non-competitive sports.

HCM is the leading cause of sudden cardiac arrest in student athletes and all people under the age of 40; however, sudden cardiac death risks are detectable and treatable for the majority of patients.

The HCM team works closely with the Division of Pediatric Cardiology at NewYork-Presbyterian Morgan Stanley Children’s Hospital to assess, follow and treat children with known or suspected HCM. This division is known for its tradition of ground-breaking state-of-the-art dedicated pediatric diagnostic and therapeutic interventions as listed above, as well as fetal echocardiography.
Medical Management

After a comprehensive evaluation, a cardiologist determines what medications will be most effective in addressing your individual symptoms.

Beta-blockers and calcium channel blockers can relieve certain symptoms, as can diuretics and anti-arrhythmia drugs. We choose a regimen that is tailor-made for you, along with the highly individualized treatment programs for any of your family members who are affected by HCM.

We also counsel your entire family about preventive measures including what kind of activities to avoid and how not to overstress your heart. In addition, we address diet and nutrition and help you determine the right level of exercise to support your general health and well-being.

Defibrillators and Pacemakers

Using risk stratification we are able to identity high-risk features for sudden cardiac arrest that may warrant an implantable cardioverter defibrillator to prevent or control arrhythmias.

A small group of HCM patients may require a pacemaker if there is damage to the electrical wiring of the heart.

We are likely to recommend an implantable cardioverter defibrillator if a patient has one or more of the following risk factors:

- Personal or family history of sudden cardiac arrest
- History of fainting or losing consciousness
- Heart wall that is more than 3 cm thick (1 cm is normal)
- Non-sustained ventricular tachycardia appearing on a heart monitor
- Decline or blunting in blood pressure during exercise
Through advanced testing, we can identify the 70% of patients who have left ventricular outflow tract obstruction at rest or with exertion.

Patients who fail medical therapy and remain symptomatic may benefit from surgical myectomy. If they are not a candidate for surgery, they may benefit from alcohol septal ablation, a catheter-based procedure.

Obstructions in the HCM heart can be addressed by septal ablation.

Our physicians and scientists are not just helping HCM patients control their symptoms and live with this condition now—they’re looking into the future to see what new approaches might improve quality of life and help us understand how HCM develops in children and in the elderly, and why this gene may express itself differently in family members. Here is a brief summary of the exciting work we are engaged in.

**Cardiomyopathy: Genetic Traits in Infants**

In 2011, Dr. Wendy Chung, MD received a $100,000 grant from the Children’s Cardiomyopathy Foundation (CCF) to identify new genes for cardiomyopathy in infants.

We know far less about the cause of cardiomyopathy in infants and their prognosis is generally worse, so this is an important area of inquiry. This CCF sponsored multi-center study will use the most advanced genetic methods to analyze all 20,000 human genes, looking for new genes...
related to infantile cardiomyopathy. This research should eventually help us to establish new targets for treatment and provide reproductive options for families who hope to have healthy children in the future.

The Relation of Cardiovascular Disease and HCM in the Aging Process

With funding from the National Institute on Aging, Dr. Mathew Maurer is exploring the higher prevalence of cardiovascular disease in older individuals. His studies may help us understand the mechanism that leads to manifestations of HCM in the elderly.

Stem-Cell Research: Understanding the Evolution of HCM

While we know a good deal about the genes that are defective in HCM patients, we know very little about the evolution of the hypertrophic heart muscle and what leads to a predisposition for sudden cardiac death. A recent seminal discovery has made it possible to reprogram the skin cells into stem cells. In the laboratory, Dr. Jonathan Lu has reprogrammed HCM cells and is watching their development. In doing so, he hopes to determine how they grow into a hypertrophic heart. This may lead to new insights into the origins of HCM, and to new treatments for this chronic disease.
"At 42, I got a second chance, and I feel like I’m just getting better and better every day."

John Cruz

In his late 20's, John Cruz worked in a Brooklyn warehouse, lifting heavy boxes and enjoyed playing football with his nephews in the local park.

Over the next few years he was hampered by asthma-like symptoms and chest pain, often symptoms of HCM. Treated at another hospital he ultimately required an implantable defibrillator to avoid sudden cardiac death.

In 2011, John’s cardiologist sent him to the HCM Program at NewYork-Presbyterian Hospital/Columbia to consult with Drs. Mathew Maurer and Hiroo Takayama. "They immediately made me feel comfortable. They said I probably needed a myectomy—surgery to cut away the damaged muscle in my heart—but they wanted to be sure."

John’s first echocardiogram indicated that he needed surgery, but a second test indicated that his heart was still functioning well enough.

To find out why John had these two different readings, Dr. Maurer gave him a modified stress test. "He told me to do as much as I comfortably could on the treadmill—I didn’t even last 10 minutes—then just sat down," John recalls.
This solved the medical mystery. When John was inactive, the gradient between his left ventricular outflow tract was between 15 and 20 mmHg. But, once he became active his gradient increased to greater than 80 mmHg.

"Dr. Maurer explained that my left ventricle closed at the top because of the thickness of the heart muscle. That meant I needed the surgery."

Dr. Takayama explained the procedure and told John what to expect post-op. "It all went according to plan," John said, "when it was over, I was so relieved and so happy to be alive. I wasn’t in as much pain as I expected to be."

John’s "asthma" symptoms also began to subside. "This turned out to be heart related—as Dr. Maurer predicted. I got a bonus from this surgery," John says, "it sure feels good to breath!"

John has high praise for the HCM team at NewYork-Presbyterian Hospital/Columbia. "Everyone was so caring, from the doctors and the nurses to the man who cleaned my room. Dr. Takayama and Dr. Maurer came to see me everyday and kept reminding me to move around. Their advice: take little five minute walks, then gradually go up to 10 and then 15 minutes. You’ve got to get your heart used to pumping."

Since his discharge, John walks his dog everyday, plays catch with his nephews and pushes his nieces on a swing. He also has the energy to paint and draw.

"At 42, I got a second chance," he says, "And I feel like I’m just getting better and better every day."
The Fleischaker Family:

"With HCM, it isn’t easy finding the balance between what you can do, and what might set you back", says Pam Fleischaker. "You have to keep on going and use the energy you have, but you’ve always got to be careful not to overdo it."

With this philosophy, Pam has succeeded in balancing a demanding career with raising a family, and her son Joey, who also has HCM, has blazed a trail in the world of sports.

Pam, 65, has worked as a newspaper editor and columnist, press secretary for Geraldine Ferraro, and as adviser on two national presidential campaigns. For more than two decades, she has played a major role in women’s politics.

At 34, Joey has been a sportscaster and a producer for both the NBC Olympics and the National Hockey League. As an extreme skier he has also conquered some of the toughest slopes in Europe, South American, Central Europe and Australia.

The Fleischakers have achieved these things with a combination of optimism and perseverance, and with the help of caring doctors who specialize in HCM. Pam suffered with atrial fibrillation for nearly twenty years, and when her symptoms worsened in 2008, she came to Dr. Donna Mancini at NYPH/Columbia to explore the option of a heart transplant. "I did my research," she says, "and learned that this hospital had more transplant experience than any other in the country."
After receiving a new heart, Pam found she had to work very hard on her rehabilitation. "I wish I had more encouragement to exercise in my 30’s and 40’s," she says, noting that it was key to bouncing back. Today Pam can walk a mile in under 30 minutes, practice yoga, and travel to her favorite spots in Santa Fe and Mexico. "My son," she adds, "has been my inspiration."

Dr. Mathew Maurer, the medical director of The HCM Program at NewYork-Presbyterian Hospital/Columbia is now monitoring Joey’s progress and helping him to maintain his high fitness level.

As a teenager, Joey played basketball and baseball, though most HCM patients refrain from competitive sports. He also added mountain biking and rock climbing to his repertoire, surprising his first team of cardiologists. In 1992, Joey received an implantable defibrillator as a preventative measure. Yet so far it hasn’t activated, and he’s shown no signs of atrial fibrillation. He now runs, goes to the gym or rides his bike for a minimum of two hours everyday. "I’ve finally discovered the level of exercise that will keep my symptoms at bay," he reports. "If I don’t work out, I’ll have trouble climbing stairs. If I do, my energy level is much better."

Warming up slowly is important, too. At night, Joey’s heart rate can drop as low as 30, so he has to wait for his body to rev up before he starts his routine: "I don’t just jump on the treadmill. I work up to it, and then I ease up to my standard pace." In addition, Joey follows a heart-healthy diet, focusing on natural foods, with plenty of vegetables and protein. These are good healthy habits for anyone but they are even more critical for those with HCM.

"Anything that makes my body work better, makes me feel better," Joey adds. "I may have some challenges ahead. But for now, no one would guess that I have HCM."
The HCMA is pleased to see NYPH/Columbia formalize its efforts in the HCMA Center of Excellence model that has served the HCM community so well. We look forward to partnering in efforts to educate, support and improve the lives of all families with HCM.
To learn more about our HCM program contact

The HCM Program and The Center for Advanced Cardiac Care

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