A Family Transforms Tragedy into Hope for the Future

There was no reason for Vivian and Paul Schafer to think that their son PJ had coronary artery disease. A sophomore at the Community College of Baltimore County in Catonsville, Maryland, 20-year-old PJ was captain of the men’s lacrosse team, loved spending time outdoors with family and friends, and completed a 12-day, 80-mile hike at the Boy Scouts’ Philmont Scout Ranch in New Mexico as an Eagle Scout.

Watching one of his son’s lacrosse games on March 23, 2003, Paul knew something was wrong. A few minutes into the fourth period, PJ came out of the game complaining of chest discomfort, and put ice on his shoulder and chest. By the time Paul reached him, PJ was lying motionless on his side and could not be revived by CPR. Tragically, he died from a sudden cardiac event.

“He was a healthy, college-level athlete in great shape,” Paul says. “Nobody would have ever guessed he had something like this.”

“It was just a total, total shock,” adds Vivian.

Searching for a meaningful way to memorialize their son, the Schafers started an annual charity golf tournament, dinner and silent auction to raise money for scholarships in PJ’s name for students at his high school and college alma maters. But after a couple of years, they wanted to do more. One of their physicians, Johns Hopkins preventive cardiologist Wendy Post, suggested funding research projects for young faculty members studying how to prevent sudden cardiac death and premature heart disease. The Schafers loved the idea.

Over the past 10 years, the Schafers have raised over $285,000 for heart disease research at the Johns Hopkins Ciccarone Center for the Prevention of Heart Disease through the PJ Schafer Cardiovascular Research Fund. It’s been a family affair, with help from PJ’s siblings Chelsea, a trauma nurse, and Bryan, a real estate agent. Paul’s mother, Louise Schafer, PJ’s cousin, Scott Musser, and PJ’s best friend, Matt Hook, also lend a hand.

“We have been blessed to have a large group of dedicated volunteers and friends that help make our event a huge success. Many have been with us from the start,” says Vivian, who works in food service for Howard County Public Schools.

The fund has supported at least 10 faculty members, including Chiadi Ndumele, for a project to better understand the link between obesity and heart disease; Erin Michos, for looking at vitamin D deficiency as a risk factor for heart disease and the early signs of heart disease in women; Saman Nazarian, for a project using endoscopy to visualize the outside surface of the heart during procedures in the treatment of ventricular arrhythmias; and Seth Martin and Bill McEvoy, for work on the detection of undiagnosed coronary artery disease.

“We’re very pleased with the results so far,” says Paul, a project manager for a consulting company. “We honestly believe that in the past 10 years, they have made some inroads into identifying potential risk factors for heart disease.”

Support from the family “has been extraordinarily helpful,” says Post, noting that researchers can take results from the pilot studies to support requests for larger federal research grants. “It’s been a win-win for everybody. The Schafers work really hard on the tournament to raise this money, and we’re incredibly grateful for their support. This is a wonderful tribute to PJ’s memory.”

The 13th annual PJ Schafer Memorial Golf Tournament will be held June 23 at the Compass Pointe Golf Club in Pasadena, Maryland. For more information, see pjschafer.com.

“I’m very grateful for the hard work of the Schafers and the other participants in the tournament. It’s been really good for us.”

—WENDY POST

Explore our new online resource to improve your heart health. Visit bit.ly/jh_heart.
When Mom Has Congenital Heart Disease

Esther Martin, 35, of Harrisburg, Pennsylvania, was born with Shone’s anomaly, the constellation of left-heart defects. By age 10, she had had four repairs, followed by aortic valve replacement as an adult, along with a pacemaker implantation for heart block. So when newlywed Martin and her husband wanted to start a family, the decision was anything but casual. “I’ve always wanted to have kids but wasn’t really sure it would be possible,” she says.

Martin’s uncertainty was amplified when a regularly scheduled follow-up only a few months before her wedding revealed a leaky mitral valve. The unexpected twist sent the couple to the altar months earlier than planned. Within a few weeks of her wedding, Martin was undergoing mitral valve replacement by Duke Cameron, director of cardiac surgery. Mere weeks post-surgery, she was talking baby with pediatric cardiologist Jane Crosson, co-director of the adult congenital heart disease program at Johns Hopkins.

Martin is among the growing number of women born with cardiac defects who attempt pregnancy and successfully carry to term—a testament to parallel successes in pediatric and adult cardiology, cardiac surgery, diagnostic imaging and maternal-fetal medicine. Because congenital heart defects are spectrum disorders and no two pregnancies are alike, the need for a tailored approach is that much more vital in women with heart defects.

“Most can have successful pregnancies provided they have careful preconception counseling and assessment, their condition is well-controlled, they are in good overall health and they are followed carefully,” Crosson says.

To ensure that the heart is coping with the substantially higher cardiac output of pregnancy, exercise stress testing is essential, Crosson says, yet Martin’s pacemaker would have rendered such testing less useful. Instead, Crosson says, yet Martin’s pacemaker would have rendered such testing less useful. Instead, they have periodic echocardiograms.

Women with congenital heart disease have a small but real risk of giving birth to a baby with a heart defect—about 5 to 7 percent, compared with 1 percent for the general population. In September 2014, Martin gave birth to a healthy boy.

Toward Better Treatment for Advanced Heart Failure

It was a scenario cardiologist Kavita Sharma encountered time and again during her residency at Johns Hopkins: A patient would present with signs of heart failure—shortness of breath, edema and fatigue—but a robust ejection fraction, or the amount of blood leaving the heart when it contracted.

Heart failure with preserved ejection fraction, or HFpEF, accounts for nearly one-half of the 6.6 million cases of heart failure in the U.S. and kills nearly 50 percent of people diagnosed with it within five years. Although there are at least nine well-studied treatments for heart failure with reduced ejection fraction, HFpEF has poorly understood pathophysiology and virtually no proven therapies.

The mysteries of the condition intrigued Sharma, and within months of finishing her cardiology fellowship in 2014, she launched the Johns Hopkins HFpEF program—one of only a handful of such programs in the country. The centerpiece of the program is a clinic dedicated to diagnosis and treatment. Sharma sees about four to five new patients a week.

Clinical management focuses on reducing recurrent edema and treating comorbidities, including secondary hypertension, atrial fibrillation, diabetes and chronic kidney disease—all of which are commonly seen in patients with HFpEF. These therapies are also the cornerstones of treating classic heart failure, but HFpEF may require modifications that address its idiosyncrasies.

For example, a recent study led by Sharma showed that patients with HFpEF are more vulnerable to acute kidney injury as a result of standard fluid-removal therapies when admitted to the hospital and require more delicate approaches to fluid removal. The Johns Hopkins group is using low-dose intravenous dopamine experimentally. Some evidence suggests that dopamine promotes blood flow to the kidneys, and the strategy, Sharma says, may help mitigate injury.

The Johns Hopkins program is also a main site in several ongoing federal studies to assess treatment strategies and disease progression. Sharma and colleagues are collecting demographic, lifestyle, risk factor and laboratory data to help clarify the pathophysiology of HFpEF and understand the mechanisms that fuel the racial and gender disparities observed in the condition.

“Our program goals are ambitious, but how could they not be?” says Stuart Russell, co-director of the HFpEF program and director of heart failure and transplantation at Johns Hopkins. “The challenges posed by this disease demand nothing less.”

The blue stain shows increased fibrosis in the heart muscle of someone with HFpEF.

Following Cardiac Cells in HFpEF

Johns Hopkins is the only site in the United States performing cardiac bioptries to better define the morphology of cardiac cells in HFpEF and glean insights about any molecular aberrations that account for the signature myocardial stiffness seen in HFpEF. The investigators are also performing genetic analyses seeking to pinpoint any DNA alterations that might illuminate heritable patterns of disease behavior.
What Will Your Legacy Be?
A single gift in 1873 from our founding benefactor, Johns Hopkins, inspired a revolution in American medicine. The Johns Hopkins Legacy Society honors Mr. Hopkins and welcomes those who make their own legacy gifts to secure the financial future of Johns Hopkins Medicine. There are many ways to become a member: Include Johns Hopkins in your estate plan; designate us as beneficiary of a retirement plan or life insurance policy; or give in a way that also provides income to you. To learn more about these and other creative ways to give, visit rising.jhu.edu/giving, or contact the Johns Hopkins Office of Gift Planning at 410-516-7954/800-548-1268 or giftplanning@jhu.edu.

Celebrating the Cardiac Surgery Critical Care Lecture

Lynne and Alan Van Praag visit the home of Glenn Whitman, director of the Johns Hopkins cardiovascular surgical intensive care unit, and his wife, Anna. The celebration took place prior to the annual Cardiac Surgery Critical Care Lecture, which was established with generous donations from the Van Praag family. From left are Glenn Whitman, Lynne Van Praag, Alan Van Praag and Anna Whitman.

A Bequest for the Ride of His Life

When John Sauter’s doctor told him that his heart could burst on a long-distance bike ride, he knew it was time to get help.

An active cyclist for most of his life, Sauter had started getting lightheaded and feeling dizzy on some of his rides. When he mentioned it during a routine physical, his primary care physician recommended that he be evaluated for aortic stenosis, a narrowing of the valve in the large blood vessel branching off the heart.

“He nailed it,” says Sauter.

As it turned out, Sauter’s aortic stenosis was a symptom of a larger problem. He was born with a bicuspid aorta, or two-valve, aorta. Normally, an aortic valve has three small flaps that regulate blood flow from the heart to the aorta. Those with a bicuspid aorta have only two flaps. The valve doesn’t function perfectly, but someone could go years without symptoms. “Bicuspid aortas are almost destined to fail eventually,” he says. “And mine did fail.”

The most pressing need Sauter faced was that his ascending aorta was dilating—meaning it was going to burst. If he wanted to keep up his active lifestyle, he’d need surgery.

An old college classmate of Sauter’s was also born with a bicuspid aorta, and he underwent a complicated surgery. “My friend said, ‘John, you have to get the best care you can, especially with that dilated ascending aorta.’” Sauter recalls.

Sauter checked out some of the hospitals near his home in Reston, Virginia, and thought they looked good. But when he came to Johns Hopkins in summer 2012 for a consultation, a young cardiologist told Sauter, “Dr. Cameron can do these complicated surgeries in his sleep.” That sealed the deal for him.

It took two years and four surgeries to get Sauter fully back on his feet. His daughter, Dawn Regan, was by his side the entire time. “She was my No. 1 angel throughout the whole thing,” he says.

To thank his care team, which included Duke Cameron, chief of cardiac surgery, heart surgeon Christopher Sciortino and several nurses, Sauter returned to Baltimore with Dawn over a prearranged celebratory lunch. “I owe my life to them,” he says. “I don’t have confidence I could have survived hardly any place else. That’s why I feel obligated to support them.”

Sauter started out making a small donation. Now, he sends in contributions several times a year and named the Heart and Vascular Institute as a beneficiary in his will. Dawn made a donation as well. With his fully healed heart, Sauter recently took his granddaughter on a 62-mile bike ride on an old tandem bike, and he completed another 62-mile ride by himself. And not once since the surgeries, he notes, has he experienced shortness of breath or heart symptoms.

Sauter also spends his time volunteering for his alma mater and former employer, Knox College, in Illinois, through alumni events in the Washington metro area. “Knox College has my heart, but Hopkins saved it,” he says.

Girl Scout Gives Her Hearts Away

Erika Kosar, right, and nurse Jackie Bradstock deliver handmade pillows to cardiovascular surgery patients at The Johns Hopkins Hospital. A member of Girl Scout Troop 81201 from Frederick, Maryland, Kosar continues a Girl Scout tradition that has provided more than 100 pillows to heart patients over the last four years.

Department Chairs Convene at the Achuff Lectureship

The second annual Stephen C. Achuff, M.D., Lectureship, established through patient philanthropy, welcomed guest speaker Vincent Gott, former director of cardiac surgery at Johns Hopkins. From left are former director Bill Baumgartner, Gott, Stephen Achuff and current director Duke Cameron.

23rd Annual Dean’s Symposium Stands Strong

Roger Blumenthal, director of the Ciccarone Center for the Prevention of Heart Disease, was a featured speaker at the 23rd annual Dean’s Symposium in Palm Beach, Florida. The event brings together generous supporters and highlights medical updates and research advances currently in progress at Johns Hopkins Medicine.

Giving Back

7954/800-548-1268 or giftplanning@jhu.edu.
The Curious Case of the ‘Nervous Heart’

It started with mild palpitations—sporadic at first, then increasingly frequent and bothersome. Next came chest discomfort, spikes in blood pressure, tremors and a jittery, something-is-not-right sensation described as a “nervous heart.”

For three years, the mystifying spells—lasting from a few minutes to an hour—plagued the 42-year-old man, sending him to various specialists across three countries and two continents, sometimes with blood pressure readings as high as 200/110 millimeters of mercury.

The patient ended up in the care of Miami cardiologist and former Johns Hopkins fellow Juan Rivera, who knew it was time to call on Oscar Cingolani, a Johns Hopkins cardiologist with expertise in treatment-resistant hypertension and a knack for cracking hard-to-solve cases.

Three years’ worth of clinical notes and medical tests gave Cingolani no clues. But the physical exam pointed to a possible culprit when Cingolani noticed the man’s fingers turned blue and his heart rate shot up right after gently pressing on his abdomen. The patient sweated profusely and experienced a flurry of premature atrial and ventricular contractions. In a matter of minutes, the symptoms subsided.

Even though a previous CT scan had shown normal adrenal glands, Cingolani immediately suspected trouble. A blood test after the patient’s spell showed mildly elevated levels of metanephrine, a finding sometimes suggestive of an adrenal gland tumor. A radioisotopic scan revealed that the left adrenal gland was overworking. Following a laparoscopic adrenalectomy, the patient’s symptoms vanished rapidly. He had a rare condition known as adrenal medullary hyperplasia. Cingolani says his improvement was dramatic.

Oscar Cingolani