

Time to Rethink Adult Congenital Heart Disease

BY JOYCE FRIEDEN

Associate Editor, Practice Trends

One morning a few months ago, Dr. Dan Murphy was awakened at 3 a.m. with news of a tragic case.

The patient, a 38-year-old man, had been operated on twice during childhood for a heart defect—once to get him stable and then again to fix the defect. Once he reached adolescence, however, he was lost to follow-up.

In the intervening years, he had developed a leaky valve, and his heart had gotten bigger and bigger. “His heart muscle had been damaged by being allowed to have that extra load all these years,” Dr. Murphy said. “He is probably going to either get a heart transplant or die.”

The real tragedy of the case, Dr. Murphy said, was that “I think a lot of the problem could have been prevented if he had been seen earlier.” It’s these kinds of cases that Dr. Murphy has in mind as he works to expand Stanford (Calif.) University’s Adult Congenital Heart Clinic.

Because of the success of pediatric cardiac care, many patients who would have died from their heart defects are now living well into adulthood. Long-term studies of these patients have shown that they often have few problems for the next 10 or 15 years. However, “We’ve found that longer term, we start to see things develop,” said Dr. Murphy, director of the clinic.

Part of that is simply a result of changes associated with aging, he continued. “We get older and middle age has its effects on all of us, including people with heart defects. We see rhythm changes, we see cardiac muscle failure in some populations, and we see changes in valves that were functioning before that begin to deteriorate.”

Some of these patients continue to see their pediatric cardiologists until they are well into their 30s, while others see general cardiologists, Dr. Murphy said. But some patients with milder defects may not end up seeing anyone at all.

The ideal background for physicians taking care of these patients is “they need to be trained in pediatric cardiology as well as adult medicine and adult cardiology,” said Dr. Mary Mullen, staff cardiologist at Boston Children’s Hospital as well as at the Boston Adult Congenital Heart Service. “Backgrounds tend to vary. I personally trained in medicine and pediatrics.”

To start figuring out a road map for the specialty, the American College of Cardiology sponsored a conference in Bethesda, Md., in October 2000, which resulted in an article addressing coordination of care, educational requirements, access to care, and special needs of adult congenital heart disease patients (J. Am. Coll. Cardiol. 2001;37:1161-98; www.acc.org). In 2004, the ACC formed the Congenital Heart

Disease and Pediatric Cardiology section, “To provide a resource for all cardiovascular specialists who are involved in the care of pediatric and adult patients with congenital heart disease.”

The case of Amy Verstappen, a patient with congenitally corrected transposition of the great arteries, is a perfect example of why treating physicians need sophisticated backgrounds. “Two years after the birth of my daughter, I started having some fatigue and was misdiagnosed as having dilated cardiomyopathy,” said Ms. Verstappen, who is president of the Adult Congenital Heart Association, a patient organization. “I kept saying I had this defect and they kept saying, ‘It doesn’t matter; your only option is a transplant.’”

The irony of that diagnosis, she continued, “was that I had a normal heart size—no one had done a chest x-ray. And the echocardiogram was getting really bad ejection fraction numbers, which were inaccurate.”

Once she got to a specialist, Ms. Verstappen found out that she never had dilated cardiomyopathy and that she could have tricuspid valve replacement instead of a transplant. “I had the valve replacement and I’ve been stable since.”

Several of the physicians Ms. Verstappen saw remarked that she had a “funny” mitral valve. But that was because they were actually looking at the tricuspid valve, she

noted. Reflecting on the treatment she got before she saw a specialist in this area, Ms. Verstappen said, “I’m not mad at [the doctors who misdiagnosed me], but I think people can do a better job of getting a referral when somebody with a complex disease comes along.”

Dr. Murphy agreed. “It’s a continuing education problem,” he said. “The patients have a right to see a specialist who stays up to date and who’s well trained and experienced. A specialized center like this, and the cardiologists who work in it, serve as education resources not just for the patient, but also for the primary care physician and for the local cardiologist, especially in the case of patients who come a long way. They can see their regular cardiologist for most things, and get a re-assessment from us” when needed.

But first, cardiologists need to learn which patients to refer. To help them figure that out, Dr. Roberta Williams, chairman of pediatrics at Children’s Hospital of Los Angeles, and Dr. Carol Warnes, professor of medicine at the Mayo Clinic in Rochester, Minn., are developing guidelines for cardiologists on how to manage adult congenital heart disease patients.

“It’s sort of a cookbook, to give them a handle on what they should be looking for, what should trigger a referral to a specialized center, and what are the long-term outcomes,” Dr. Williams said. The guidelines, which Dr. Williams said



Specialized adult congenital heart disease centers serve as educational resources not just for patients, but also for primary care physicians, Dr. Murphy says.

would be available in about a year, are being cosponsored by the ACC and the American Heart Association.

In some geographic areas, “the best expertise [in this area] is in the children’s hospital, because the anesthesiologists and the cardiologists there are best trained to manage these patients,” Dr. Mullen said.

Some cardiologists are starting to get the message. Dr. Murphy said his clinic sees about 10 patients a week, most of whom are complex and require long visits.

Dr. Joseph Perloff, founding director of the Adult Congenital Heart Disease Center at the University of California, Los Angeles, also has seen demand grow at his facility. “It was apparent decades ago that advances in surgical techniques in congenital heart disease would result in long-term survival, and there were no facilities or training programs” to help these patients, he said.

So Dr. Perloff started the center shortly after he arrived at UCLA in 1979; the center was initially open only 1 day a week. Today it is open 4 days a week and has a registry of almost 3,000 patients, most of them referred by cardiologists in the community. The Adult Congenital Heart Association, in conjunction with the International Society of Adult Congenital Heart Disease, is putting together a directory of self-described adult congenital heart clinics nationwide; so far, they have received 37 surveys from centers that describe themselves as such, according to Ms. Verstappen.

UCLA also has the first and largest training program for the specialty; the program admits one or two fellows per year, depending on the strength of the applicant pool, Dr. Perloff said. Physicians may apply for the fellowship after they finish residency, which must include 2 years in general cardiology and some pediatric cardiology as well. The fellowship lasts 1 year, “but usually the fellows stay on another year to complete a research study or develop a particular area of interest,” he noted.

Other training programs include those at the University of California, San Fran-

cisco, the Mayo Clinic, the University of Iowa, Iowa City, and the University of Pennsylvania, Philadelphia. But more need to be started, Dr. Perloff said.

He noted that currently there are only about 1,200 board-certified pediatric cardiologists, far too few to take care of the estimated 900,000 adults with congenital heart disease nationwide. In fact, “there are now more adults with congenital heart disease than infants and children,” he said.

One problem with recruiting people into training programs is that for cardiologists, “training is already very long” with 4 years of residency and 2 years of specialty training, Dr. Williams said. “To sustain this kind of [subspecialty] training, other aspects of training may need to be shortened, maybe by double-counting certain things, or having a loan forgiveness program or something to help them, because if you delay starting practice for 2 more years, the debt burden is so high a lot of them wouldn’t be able to afford to do it.”

This situation is not helped by the fact that many adult congenital heart disease patients have trouble getting health insurance. Coverage for these patients under their parents’ insurance policies typically stops when they turn 18 or 21. “How many 19-year-olds working at whatever job someone that age can have, will have health insurance?” Dr. Williams said. Treating uninsured or underinsured patients then adds to the physicians’ financial woes.

To bring more attention to the issues surrounding these patients, the Adult Congenital Heart Association and several other organizations sponsored a “lobby day” in Washington on Feb. 8, aimed in part at getting Congress to fund a registry of congenital heart disease patients. “We need basic data to help us plan for the needs of adults with congenital heart disease; we need to do evidence-based medicine,” Ms. Verstappen said. “We also want to help children with congenital heart disease. Right now, we are doing surgeries in children without the data we could have from adults.”