

**“A big gap in care.”** Not too long ago, kids born with heart concerns rarely lived into adulthood. Now as many as 15,000 Coloradoans do, but while pediatricians may be uncomfortable treating them as adults, adult cardiologists aren't always trained in the specialized care these patients continue to need. Children's and UCH are creating ways to spread the word about all kinds of no-longer-fatal chronic childhood diseases.



## *Congenital disease poses unique problems*

# As Heart Patients “Age Out” of Pediatric Care, Children's & UCH Forge a Transition to Adult Care

*By Tyler Smith*

Choose virtually any vantage point on the Anschutz Medical Campus, and one can see the imposing physical structures of both The Children's Hospital and University of Colorado Hospital. Avenues that offer easy connections for pedestrians and automobiles run between them.

But transitioning patients from the pediatric world of Children's to the adult medical care of UCH is not as simple as taking the road from east to west, as Joseph Kay, MD, and others can attest. As director of the Adult Congenital Heart Disease program at UCH, Kay, an assistant professor of Medicine and Pediatrics in the CU School of Medicine's Division of Cardiology, knows the problems firsthand.

He and colleague Elizabeth Yeung, MD, specialize in treating patients saddled from birth with heart defects that endanger their health and put them at even higher risk of complications than patients with acquired heart conditions. Managing their conditions closely is imperative, yet the care they receive as they move from the pediatric to the adult world too often lapses, Kay says.

**Rocky road.** *“We're doing a poor job of managing congenital heart disease,” he states. The reasons are varied. Young adults in general may push their health care to the side because of financial problems, lack of insurance or a perception it's not a priority.”*

*But the health care system itself frequently doesn't serve congenital heart patients in particular well, either, Kay adds.*



*Joseph Kay and Elizabeth Yeung are among the few physicians who treat both kids and adults with congenital heart disease.*

“There is often poor education from pediatric providers about these patients' long-term needs,” he asserts. Adult providers, moreover, often have little training in treating or even recognizing congenital heart problems.

“Neither is ideally set up to deal with these patients' conditions,” Kay says.

Medical procedures don't spell the end of congenital heart problems, adds George Warren, MD, a retired physician who is a patient of Kay's.

Warren suffers from tetralogy of Fallot, a cluster of four congenital defects that affect the septum, the pulmonary valve, the right

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ventricle and the aorta. At 17, Warren underwent surgery that included a patch to repair holes in the aorta and pulmonary artery. His surgeon also whittled away fibrous tissue from the ventricle.

"When I was discharged, the surgeon – a good friend – told me what he had done, but added he couldn't fix the problems entirely," Warren recalled. "With congenital heart problems, a repair is not a repair. You're not made normal. People have to follow the conditions for the rest of their lives."

**Broader problem.** Easing patients through uncertain times and monitoring their care is a challenge that extends beyond the field of cardiology, says Kay, who began thinking about the problem during his residency in internal medicine and pediatrics.

"During my rotation in pediatric cardiology," he remembers, "I learned that many pediatricians didn't feel qualified to deal with adult issues, and that many adults were not confident treating congenital defects. I saw it as a big gap in care.

"But it's not just cardiology," he adds. "The issue applies to all the chronic diseases of childhood, including cancer, neurology and hematology. There is a paucity of adult providers who understand the totality of the needs of these patients. They aren't like the 25-year-old with an acquired condition."

**Unique problems.** Patients with congenital heart problems can pay a high price for that knowledge gap. For example, those born with single ventricle defects – meaning only one chamber of the heart pumps blood normally – are at greater risk of cirrhosis of the liver, Kay notes.

Arrhythmias, or irregular heartbeats, plague disproportionate numbers of these patients. So does pulmonary hypertension, which results from septal defects that create excessive, damaging blood flow to the heart and lungs. Women with congenital defects are more likely to have high-risk pregnancies.

"Providers must approach these patients differently than they do others with acquired heart diseases," Kay says. "Without that, they won't get the appropriate transition of care."

Too often, Yeung adds, patients who leave Children's care go to adult-care cardiologists in the community whose experience extends only to those with acquired heart conditions.

"They don't know when to intervene," she says. "They may do an echocardiogram and look at the heart, but they don't know when to do something about what they see."

**Both sides of the street.** By contrast, Kay and Yeung not only specialize in congenital heart disease, they see patients at both UCH and Children's. The program works closely with other specialists at UCH and Children's, including perinatologists, pediatric interventionists, high-risk obstetricians, pulmonologists, geneticists and hepatologists. Key services stand nearby, including surgery, electrophysiology, heart failure and transplant.

The program, the only one of its kind on the eastern slope of the Rockies "between Montana and Mexico," sees 700 to 800 patients a year, Kay says, up from about 250 in 2003.

"But there are 9,000 to 15,000 people in Colorado with adult congenital heart disease we should be following or consulting with their community providers."

Kay and Yeung are intent on doing more than managing the disease, however. Both are keenly interested in finding ways to improve transitional care for congenital heart disease patients. Kay was principal site investigator for a National Institutes of Health study of 12 centers that treat adult congenital heart defects.

The two-year study, which concluded in April, examined how long it took patients to move from pediatric to adult care, the barriers they faced and whether or not follow-up and education would improve their care. Researchers have just begun analyzing the study data, Kay says.

Yeung analyzed lesions from some 400 adult congenital heart disease patients older than 18 years of age, looking for lapses in their care, the reasons they occurred and the consequences.

The research showed that patients who didn't come back for care as adults were more apt to have heart procedures, surgeries or changes in their medications, she says. "It also seemed that those with moderate disease had the highest incidence of lapses of care," she adds.

**Closing the gap.** The reasons she cites for the gaps in care echo many of the points Kay makes. Patients whose care lapsed were sometimes stymied by a lack of financial resources or insurance. Others didn't know where to go to get care, a problem that might be addressed by their pediatric providers, she adds.

*"We're not always empowering young adults to have the information they need to move forward," Yeung says, noting she sometimes sees kids who have no idea they have a heart condition.*

*"We have to make sure they have the information they need so that when they go into the real world, they will be ready to deal with [their health] issues."*

Yeung says Children's created a short survey for patients and their families that asks them to list their diagnoses and surgeries, their activities and any restrictions caused by their condition.

"If they don't know, we help them review and make sure they have their paperwork and imaging," she says.

As the time for the transition to adult care gets closer, Yeung says, she begins to "broach the topic" with discussions about patients' plans for pregnancy, jobs and college. "We try to unshelter them and help them to get started thinking" about the transition to adulthood. "Some are more ready than others," she adds.

It's also important for adult providers to see congenital heart disease patients at the pediatric institution, Kay says. "Adult providers can provide during adolescence education about their disease and the need for follow-up care," he explains. "That should start at age 12 to 14 so we're sure they get it."