



Mended  
Little  
Hearts

# Congenital Heart Defects

Not Your Average Heart Disease

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## **I**f you were asked to picture someone with heart disease, what would that person look like? For most

of us, the person we picture would be over 50 with several of the risk factors for heart disease. We typically do not picture, however, a young child or a young adult who was born with heart disease – someone with a congenital heart defect – yet, congenital heart defects are the most common birth defect in the U.S. and affect about 1.8 million Americans today.

### **What are congenital heart defects (CHD) and how do they differ from acquired heart disease?**

Congenital heart defects are conditions present at birth that affect the structure and function of the heart, according to the U.S. Center for Disease Control and Prevention. Simply put, they occur when the heart fails to form properly long before the baby is born. As a result, the heart has missing pieces, pieces in the wrong places, backwards pieces, and/or additional pieces that don't belong. Unlike acquired heart disease, there is no known way to prevent heart defects at this time, and doing heart-healthy things like eating a nutritious diet and exercising regularly cannot cure or change a congenital heart defect.

There are thirty-five recognized types of congenital heart defects, and the severity of the defects can vary greatly. About 30 percent of these defects are complex and may require surgery, or multiple surgeries, and/or other procedures in childhood. Congenital heart defects remain a leading cause of death for children, but the good news is that today, more than 85 percent of children born with congenital heart defects are living into adulthood.

As wonderful as it is, there is a challenge created by so many of these children living into adulthood: These adult hearts aren't the same hearts that adult cardiologists typically deal with – these hearts look weird, for lack of a better term, and have unique issues. In addition, because until fairly recently most people with CHD didn't live to adulthood, pediatric cardiologists, who are used to these weird hearts, are not used to dealing with these hearts in adults who have different issues from kids.

Dr. Michael Earing, Director of Adult Congenital Heart Disease at the Divisions of Adult Cardiovascular Medicine and Pediatric Cardiology at the Medical College of Wisconsin, explained this issue: "A pediatric cardiologist in training is required to take one month of

adult cardiology experience. An adult cardiology fellow, in his or her three years of fellowship, is required to take one month of congenital heart disease. Therefore, neither the adult cardiologist nor the pediatric cardiologist is a specialist in adult congenital heart issues."

Imagine that you have acquired heart disease that needs treatment. You have a choice between seeing your general practitioner, who has some cardiology training, or seeing an adult cardiologist for treatment. Which would you choose?

Most of us would choose to see the adult cardiologist because he or she specializes in our problem and has a better understanding of our needs. Similarly, adults with CHD need to seek specialized care for their special needs.

### **The Adult Congenital Heart Association (ACHA) is working with Mended Little Hearts to educate families about lifelong care.**

Families who have children with congenital heart defects want their children to grow up and become healthy adults. That's why Mended *Little Hearts* works with the Adult Congenital Heart Association (ACHA) to create opportunities for education about lifelong specialized care for people with congenital heart defects.



**Amy Verstappen, President and CEO of the ACHA, is a passionate advocate for specialized lifelong care for CHD adults**

Amy Verstappen, President and CEO of the ACHA, is a passionate advocate for specialized lifelong care for CHD adults, and she exemplifies the importance of understanding the special needs of adults with CHD. “I was extremely educated in my disease, I had excellent pediatric cardiology care, I had no insurance barriers – I had everything exactly right,” she said. “The missing piece was that I was told to see a cardiologist, but I wasn’t told to see a special cardiologist.”

“I have a tricuspid valve where a mitral valve should be in my heart,” she said, “so on my medical records, I have 15 years’ worth of adult trained cardiologists saying, ‘Boy, she has a really weird mitral valve.’ Nowhere did they recognize that the valve is not, in fact, a mitral valve at

specializes in adult congenital heart disease. “Around the year 2000, the adult population surpassed the pediatric population.” This means that Mended *Little* Hearts, in order to adequately support families with children who have CHD, needs to understand that these children are very likely to become adults with CHD and to prepare them for that.

Carol Raimondi, 35, gives another example of how congenital heart disease differs from acquired heart disease and why adults with CHD need specialized care. Carol was born with multiple congenital defects and has been seeing cardiologists all her life. She has had multiple surgeries.

When Carol turned 16, she decided to start looking for an adult cardiologist because she felt that her pediatric cardiologist would always talk to her through her mom. She was not told, however, to see an adult cardiologist who specialized in CHD. She said, “Even though the adult cardiologist that I found was very good, I felt a bit lost because it seemed like my heart, with its strange parts and configurations, was all new territory for him.”

When Raimondi had an open-heart surgery at age 20, she said that as soon as her cardiologist completed her surgery, he came out and told her mother that he would never operate on her again. “He said it was just too difficult because of my anatomy and because of the scar tissue there,” she said. “That was kind of a wake-up call for us. I thought, ‘Okay, this can’t be good.’ Who do you follow up with at that point? You want to have somebody who feels confident.”

Afterwards, Raimondi had multiple infections including an infection in the sternum. She was on IV antibiotics at home for a long time, but she was able to get well enough for her wedding. However, a week after her wedding, she returned to the hospital. She had developed endocarditis, an infection of the heart lining. “That was frustrating. I was still trying to get that normal feeling in life,” Carol said. “I wanted to be a regular newlywed.”

She was eventually referred to another hospital where there was an adult congenital heart specialist. She felt that she finally had a cardiologist who understood her heart and her unique needs. “I was just amazed at the ease with which he was able to do my echocardiograms (echos),” Carol said. “He was familiar with my defect and familiar with how things should look on my echo. Where other doctors would spend an hour searching trying to figure out what was what on my echo, he was able to find exactly what I needed right away.”



**Carol Raimondi was born with multiple congenital defects and has been seeing cardiologists all her life.**

all, because they didn’t have training in CHD. Not understanding their hearts can have severe consequences, and can even be fatal, for adults with CHD. CHD is a different disease, and there are guidelines that say it needs different care.”

Verstappen works with MLH to help parents understand that their child’s disease is different from acquired heart disease not because they are kids, but because they have a different disease called congenital heart disease. Because it is a different disease, they are going to need different care from adults with acquired heart disease.

Today, more adults than children are living with congenital heart defects. “The population of adults with congenital heart disease now living in the United States is believed to be one to one-and-a-half million,” said Dr. Mark Townsend, an internist and pediatrician who

Her specialist also helped her decide if she could get pregnant and worked in collaboration with her OB/GYN during her pregnancy. “I could be followed by both, and I felt very secure with my doctor,” she said. “I just felt that I was at the right place.”

Christine Yuhas, Education Coordinator for Mended Little Hearts of Lynchburg, Va., had a similar experience. Christine had her first surgery at five days old after being diagnosed with ventricular septal defect (a hole between the two bottom chambers of the heart) and a coarctation of the aorta (a pinch in the aorta that decreases blood flow through the vessel).

When Christine, now 25, moved to Virginia in 1997, she began seeing an adult cardiologist who did not specialize in congenital heart defects. However, in 2006 when she started having questions about pregnancy, her doctor couldn’t confidently answer her questions.

Eventually, Christine was referred to Dr. Mark Townsend who specializes in adult congenital cardiology. “Seeing someone who specializes in adult congenital cardiology has made such a difference,” she said. “It is so nice to have somebody who gets it. To be able to walk into the doctor’s office and sit down and ask him whether it is okay if I have a child and whether my heart can handle it, whether I can pass CHD on to my child, and other important questions, and for him to be able to answer those question with a true understanding of my heart, gives me the confidence to make good decisions about my health and the health of a baby that I might have.”

Because Christine understands that her heart disease is different from acquired heart disease, she understands the benefits of seeking specialized care. She said, “In a ‘normal’ heart, problems with the heart might look one way, but in a CHD heart, problems might look completely different. Also, things that are ‘normal’ in a CHD heart, may be problematic in the ‘normal’ adult heart. For example, the muscle of the CHD heart might be thickened in places, and that is normal. A cardiologist inexperienced in CHD hearts might see this as a problem needing treatment, while the adult CHD specialist will understand that no treatment is required. You need someone who is experienced with congenital patients and who understands the defects.”

### **Lifelong, specialized care for adults with CHD**

Dr. Townsend said part of the reason many adult CHD patients don’t seek follow-up care is that many who had surgical interventions in the ’70s and ’80s were told that they were cured. “If you were born with heart disease, chances are you were not cured despite the fact that many patients with congenital heart

disease were erroneously told they were cured and that they didn’t need further follow-up.” Even today, many parents of children with CHD who had surgeries in childhood believe their child is cured and doesn’t



**MLH of Tulsa, OK, putting CHD awareness on our nation’s highways.**

need further care.

However, we now know that many people with CHD who had surgery as a child are showing up with some changes in their hearts 20 years later, and some of those changes can cause problems.

Dr. Earing stated, “Most patients now survive into adulthood, and although most patients do well, multiple different problems can develop. Some of these are minor and some are major. But the key concept is that if you have regular follow-up by an adult CHD specialist, you can prevent a lot of those major problems before they occur.”

Dr. Earing explained that as adolescents with CHD become adults, some things can cause even more problems, particularly when it comes to pregnancy, birth control or even the type of occupation the patient chooses. “These are all special circumstances that an adult congenital heart specialist can help with compared to a regular cardiologist,” Dr. Earing said. “Pregnancy can be a huge issue in certain people. The changes that occur during pregnancy most of the time are well tolerated. But in some women, it can cause a lot of problems even if they are doing fairly well before they become pregnant.”

The ACHA’s website ([www.achaheart.org](http://www.achaheart.org)) provides a listing of clinics and programs across the country, and there is one in almost every state. If the program in your state is too far away, he recommends contacting those programs and asking them if they know of someone closer to you. The important thing is that all people with CHD, both children and adults, get the care they need so they can live longer, healthier lives. ❤️