Ground-breaking study to improve quality of life and outcomes for kids born with heart defect

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A trial on shunts used to direct blood flow to the lungs, led by researchers at the University of Michigan's C.S. Mott Children's Hospital, will lead to better outcomes for kids worldwide born with hypoplastic left heart syndrome, the most common severe heart birth defect.

Under the leadership of the University of Michigan, the first multi-institutional randomized prospective trial ever conducted in congenital heart surgery was just completed by the Pediatric Heart Network with funding from the National Heart, Lung, Blood Institute.

Kids with the hypoplastic left heart syndrome have hearts that don't develop properly in the womb. Because the left side of the heart fails to develop, they are often referred to as being born with half a heart.

This groundbreaking study is published in the May 27, 2010 issue of the New England Journal of Medicine. Richard G. Ohye, M.D., Division Head of the Pediatric Cardiovascular Surgery at the University of Michigan's C.S. Mott Children's Hospital, is the study chair and lead author.

"Congenital heart disease is much more common than people realize," says Ohye. "Heart abnormalities are the most common birth defect, and it occurs in almost one out of every 100 live births."

Twenty years ago, doctors were unable to save these children with hypoplastic left heart syndrome. Today, because of efforts by the University of Michigan Congenital Heart Center and other centers like it around the world, most of these infants can be saved through a series of three operations that can be compared to re-plumbing their heart.

Research into this congenital defect has been rare because most centers do not see enough patients with any one diagnosis to be able to effectively study it. Hypoplastic left heart syndrome occurs in about 1,000 kids every year in the United States and it accounts for roughly about 8 percent of all different congenital heart defects.

"We can't really rebuild the heart so we have to make do with what there is -- the one pumping chamber," Ohye says. "So we re-plumb the heart so that the kids can get by with just the single ventricle or pumping chamber."

The treatment of hypoplastic left heart syndrome requires three operations and the first one is done around the time of birth, the second one at about four to six months of age and the last one at 18 to 24 months of age.

The University of Michigan has had a pioneering role in the treatment of hypoplastic left heart syndrome.

Prior to the early 1980s, there was no operation for it at all, so all the kids unfortunately died as newborns. Since that time, survivals have dramatically improved. Initially, they were quite low. During the 1990s, hospital survivals for the first operation were only about 40 percent. Now, at experienced centers, they're up above 90 percent.

In the first stage operation for hypoplastic left heart syndrome, there are two different versions. Researchers were interested in finding out which

had a better survival rate.

One version gets blood directly from the heart through a tube, or shunt, to the artery to the lungs. The other allows the blood first to exit the heart through the reconstructed aorta and then bring the shunt from that reconstructed aorta over to the artery to the lungs.

"What we found was that the shunt that went directly from the heart to the artery to the lungs did better over the course of the first year," Ohye says. "We also found that they had tended to have a few more complications though, and so I think the jury's still out over the long run which is going to be better."

"We're going to continue to follow these kids," says Ohye. "We're almost up to a four-year follow-up at this point and then we'll continue to follow them and plan on looking at them again when they're about 8 or 10 years old."

This research also is significant because it is the very first randomized prospective trial ever done in congenital heart surgery. The study enrolled 555 kids and will provide a tremendous opportunity to observe them all the way into adulthood and follow them and see how they do.

"It is important to test anything we do to manage our patients in a rigorous scientific way, and it's the first time that we've ever gotten together, many centers - 15 in this case - and agreed that this was really important to do," Ohye adds. "We showed that we can do it and that we believe that it's important to do for our kids."

"Not only did the study answer an important question about how we care for them at birth, but it's going to continue to help us understand how we take care of them throughout their lives," says Ohye.

More information: Journal reference: N Engl J Med 2010;362:1980-92.