Rebuilding A Whole Heart For Children With Only Half

Using a combination of surgical procedures developed over the last 11 years, surgeons at Boston Children's Hospital have established a new approach for rebuilding the heart in children born with a severe heart defect called hypoplastic left heart syndrome (HLHS). This "staged left ventricle recruitment" (SLVR) strategy uses the existing standard single-ventricle treatment for HLHS and additional procedures to spur the body's capacity for healing and growth and encourage the small left ventricle in these children to grow and function.

Members of Boston Children's Departments of Cardiac Surgery and Cardiology—including Sitaram M. Emani, MD; Wayne Tworetzky, MD; James E. Lock, MD; and Pedro J. del Nido, MD—reported their results to date with the SLVR strategy in the November 6 issue of the *Journal of the American College of Cardiology*.

HLHS is a rare but severe congenital heart defect in which the left ventricle, the chamber of the heart that pumps blood out into the body, does not develop properly. The majority of children born with HLHS undergo a series of three procedures, together known as single ventricle palliation (SVP), culminating in the Fontan procedure. Surgeons use the sequence to reconstruct a child's heart so that it can function with only a single working ventricle, but only about half of patients treated in this way survive to adulthood.

"Many of the children who reach adulthood eventually require a heart transplant because their Fontan circulation doesn't work effectively or causes additional complications," according to Emani, a pediatric cardiac surgeon who specializes in cardiovascular surgery for newborns and children with complex congenital heart disease. In 2001, del Nido, Emani and their colleagues started considering ways to help children with HLHS avoid SVP as an end goal and improve long-term outcomes by reconstructing the heart so that it has two working ventricles.

"Children have an enormous growth and healing potential," Emani explained. "We realized that rather than give up on the left ventricle and commit a child to single ventricle circulation for life, we could leverage that growth potential in a staged approach that would promote growth in the left ventricle and gradually recruit it back into operation."

With the SLVR approach, a child undergoes initial single ventricle palliation with the Norwood procedure in order to stabilize their circulation. Surgeons then perform additional procedures over time, such as mitral or aortic valve repair, removal of endocardial fibroelastic (EFE) tissue (tough tissue that can restrict the heart's pumping motion) and closure of atrial septal defects (gaps in the wall between the heart's left and right atriums). Between each procedure, surgeons allow the child's

heart to heal and strengthen, setting the stage for the next operation.

"The goal is to promote blood flow through the patient's left ventricle and thereby stimulate growth of the ventricle," Emani said. "If we can do that, we give the ventricle an opportunity to function." Emani noted that the SLVR approach is highly customizable based on each individual child's unique heart anatomy. It also requires a high degree of collaboration and an open approach to cardiac care involving multiple medical and surgical sub-specialties.

In their report, the authors document their experiences developing and refining the SLVR approach between 2001 and 2010. In that time, 34 children were treated using the approach, 12 of whom were successfully converted to complete, two-ventricle circulation. In the short term, survival rates among SLVR-treated patients have been slightly better than a comparable group of children who underwent SVP. In addition, patients who underwent SLVR also displayed significant growth in the valves and chambers on the left side of the heart, even if surgeons could not get the left ventricle to function completely, demonstrating that the strategy can effectively rehabilitate the heart and successfully support its growth and function.

Compared to SVP, the SLVR approach has had equal if not better outcomes over the last decade. The approach's overall benefits are geared toward improved long-term outcomes and survival into adulthood. The team notes that in the short term the SLVR approach is more invasive and intensive than the single ventricle approach. But they expect that the long-term benefits of SLVR—especially in terms of reduced risks of future hospitalization, surgery and heart transplantation—will far outweigh the initial costs.

"We believe the SLVR strategy could revolutionize the pediatric cardiology field's approach to treatment of HLHS," Emani says. "The general consensus has been that conversion to single ventricle circulation is the safest approach, but it has significant limitations when it comes to adult survival. While we are still refining the process, we think our two-ventricle approach will help children with HLHS not just survive but thrive into adulthood."

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