

The *Journal of the American College of Cardiology* published a study in November 2012: “Staged Left Ventricular Recruitment after Single-Ventricle Palliation in Patients with Borderline Left Heart Hypoplasia.”

The abstract can be found at the following link: <http://www.ncbi.nlm.nih.gov/pubmed/23062531>

The NPC-QIC Research and Publication Committee reviewed this article, and a summary of the findings can be found below.

Main Finding from this Study:

Most children with hypoplastic left heart syndrome (HLHS) require three surgeries in the beginning of their lives for survival: the Norwood, Glenn and Fontan surgeries. This result of these surgeries is a heart that only has one pumping chamber (instead of the usual two chambers). For children with HLHS, the right ventricle pumps blood to the body. The process of these surgeries all together is called “single-ventricle palliation”, or SVP. (SVP is the term used in the article, but it might be more familiar to many readers as the “Fontan route”.)

SVP is recommended for most children with HLHS. This is because the left ventricle is too small to help pump blood to the body. In some children, however, the left ventricle is “borderline”, meaning that it *might*, with the help of multiple surgeries and other procedures designed to promote growth, be able to do the work of pumping blood to the body.

The authors of this paper describe an approach at Boston Children’s Hospital for children with borderline hearts. They show that for certain patients, it is possible to get the left ventricle to grow using a combination of different surgeries and other procedures. Some of the patients who undergo these surgeries and procedures are able to have a “biventricular” circulation, meaning the left ventricle pumps blood only to the body and the right ventricle pumps blood only to the lungs. This is more similar to a healthy normal heart. Perhaps more importantly, this demonstrates that there is the potential for growth of the left ventricle in patients with borderline left heart structures.

About this study:

- **Why is this study important?**

Many children with HLHS who undergo SVP do well. Unfortunately, however, many babies and children do not survive SVP and others have medical problems related to the heart and other organs. This has prompted investigators to explore the possibility of getting the left ventricle to grow in the hope of keeping the left ventricle as the chamber that pumps to the body (this is called a biventricular circulation). These children could (we don’t know yet) have better survival and fewer medical problems than those that undergo SVP.

The authors of this paper describe ways of getting the left ventricle to grow; they call this “staged left-ventricular recruitment”, or SLVR. SLVR procedures include surgeries and catheter-based procedures designed to promote blood flow through the left side of the heart. This includes procedures on the mitral valve, the aortic valve, and within the left ventricle itself. These are all components of the heart that are affected by HLHS, and patients must meet certain qualifications of these components to be a candidate for SLVR.

- **How was this study performed?**

The researchers looked back at patients diagnosed with borderline heart at Boston Children’s Hospital between 1995 and 2010. They compared 34 patients with borderline hearts who underwent traditional SVP, and 34 patients who underwent SLVR. They compared the sizes of different left-sided heart structures. Specifically, they compared the size of the mitral valve, aortic valve, and left ventricle.

- **What were the results of the research?**

- At birth, the sizes of left heart structures were similar in both groups of patients, although the patients in the SLVR group tended to have a slightly larger left ventricle, and patients in the SLVR group were more likely to have had a procedure as a fetus (during the pregnancy) or shortly after birth to open a blocked aortic valve.
- Patients in the SLVR group did demonstrate growth in their left-sided heart structures. Specifically, they had a larger mitral valve, aortic valve, and left ventricle than patients who had SVP. The most growth was seen after the Glenn surgery.
- 12 of the 34 patients in the SLVR group were able to achieve a biventricular circulation. 18 of the patients had either Glenn or Fontan type hearts, and 1 underwent transplant. Those with Glenn or Fontan circulation will either continue with SLVR or undergo SVP.
- Patients who had a traditional SVP typically had 3 surgeries, whereas patients who underwent SLVR typically had 4 surgeries.
- Patients who had a small hole created in the top heart chambers to direct blood flow into the left ventricle were more likely to have growth of left-sided heart structures.
- There were 3 deaths (8%) in the SLVR group and 7 deaths (20%) in the SVP group. Although this difference was noted, the number of patients in the study overall was too small to know if that difference was due to the way they were treated.

- **What are the limitations of this study?**

- The study looked at a small numbers of patients. This limits the ability to draw conclusions that can be applied to the general population. Borderline patients encompass a wide range of patients with many different sizes of the left side of the heart.
- There may be aspects of the hearts in the SLVR that are different from the SVP group. Patients in the SLVR group were more likely to have a procedure as a fetus or newborn to relieve blockage of the aortic valve. This may mean that there are differences between the two groups despite the size of their left hearts being similar.

- This study does not describe how the SVP and SLVR patients are doing clinically, either in the short or long term.
- Important questions that could be addressed in the future include:
 - What does the future hold for promoting the left side of the heart to grow?
 - How are SLVR patients doing clinically, and what are their long-term outcomes? How do they compare to the SVP patients?
 - When is the best time to undergo SLVR?
- **What are the takeaway messages considering the results and limitations of this study?**
 - This study shows promise for promoting growth of the left ventricle in patients with a borderline heart. Future study is required to explore the best way to achieve that growth, but this study offers hope.
 - In order to undergo SLVR, children must meet certain qualifications. It is important to realize that SVP is recommended for the most children with HLHS. In a select group of patients with HLHS—those with a borderline heart—surgeries and other procedures to encourage the left heart structures to grow are possible. Among such patients, some, but not all, will achieve a biventricular circulation.
 - SLVR includes a variety of surgeries and catheter-based procedures on different parts of the left heart (the mitral valve, aortic valve, and ventricle), and most patients who undergo SLVR will have a Norwood and Glenn procedure prior to achieving a biventricular circulation.
 - Compared with patients who undergo SVP, patients who undergo SLVR have larger left heart structures, and 12 out of 34 patients who underwent SLVR were able to achieve a biventricular circulation.
 - More studies are required to understand the best way to promote left heart growth and to understand the short and long-term quality of life for patients with biventricular circulation.

The Bottom Line:

Doctors at Boston Children’s Hospital are working to grow the left side of the heart for some children with HLHS. These procedures are still being studied, but they seem to help the left heart grow for some children. In order for your child to qualify, he or she must meet certain criteria. You may wish to talk to your cardiologist about SLVR, and whether your child may be right for it. Dr. Emani (the author of the article reviewed here) can also be contacted at sitaram.emani@cardio.chboston.org.