



National Pediatric Cardiology  
Quality Improvement Collaborative

## Research Explained

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### **Attrition between the superior cavopulmonary connection and the Fontan procedure in hypoplastic left heart syndrome.**

Lawrence KM, Ittenbach RF, Hunt ML, Kaplinski M, Ravishankar C, Rychik J, Steven JM, Fuller SM, Nicolson SC, Gaynor JW, Spray TL, Mascio CE.

*J Thorac Cardiovasc Surg.* 2020 Oct 22:S0022-5223(20)32871-3.  
doi: 10.1016/j.jtcvs.2020.10.053. Online ahead of print

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## ABOUT THIS STUDY

### **Why is this study important?**

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- The risk for death between the initial Norwood procedure to the superior cavopulmonary connection (SCPC, i.e. Glenn or hemi-Fontan) procedure for patients with HLHS has improved due to advancements in operative, perioperative, and interstage monitoring.
- The risk for death from the SCPC to the Fontan has been described, but previous data were limited due to relatively small number of patients studied, incomplete data, and minimal long-term data.
- Better understanding of the risk factors contributing to death between the period from the SCPC to the Fontan surgery may be helpful in decreasing deaths during this time interval.

### **What was the goal of the study?**

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- The main goal of this study was to document the incidence of failure to undergo a Fontan procedure in patients discharged after the SCPC procedure.
- A secondary goal was to identify any variables associated with failure to undergo a Fontan procedure.

## How was this study performed?

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- Data from all patients with HLHS or HLHS variants that underwent a Norwood and SCPC procedure between 1/1/1998 – 12/31/2017 were reviewed.
- Patients were excluded from the study if they died before hospital discharge after the SCPC, were suitable candidates awaiting their Fontan procedure, lost to follow-up, or underwent a two ventricular repair.
- Baseline information, operative data, hospital data, and interstage data were collected.
- Time period was divided over four eras (era 1: 1988-1994, era 2:1995-2001, era 3: 2002-2010, and era 4:2011-2017)
- Attrition was defined as death, non-suitability for Fontan procedure, or cardiac transplantation before the Fontan procedure.

## What were the results of the study?

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- 919 patients were initially reviewed. 63 patients were excluded (19 awaiting Fontan, 43 lost to follow up, and 1 underwent two ventricle repair), therefore the study consisted of 856 patients.
- 705 patients (82.4%) underwent a Norwood procedure with a modified Blalock-Taussig (BT) shunt whereas 146 patients (17.1%) had a right ventricle to pulmonary artery conduit (RVPA)
- Use of BT decreased from era 1 (99.2%) to era 4 (54.8%).
- There was a trend for performing the SCPC earlier over eras, 210 days old to 136 days old.
- The attrition rate was 8.3% (71 patients total: 52 patients died, 10 patients underwent heart transplantation due to poor heart function, and 9 patients were deemed unsuitable for SCPC).
  - Median age of death was 412 days old and median time of death from SCPC was 245 days old.
- This rate of attrition did not change over the four eras.
- After analysis, the values that were significantly associated with attrition were:
  - Atrioventricular valve (AVV) repair at the time of SCPC
  - Other procedures being performed at the time of SCPC
  - Total support time at SCPC
  - Length of hospital stay after the SCPC

- Use of a RVPA conduit
  - In the two most recent eras, RVPA conduit attrition was 14.3% (21/146) versus BT shunt attrition of 6.4% (18/283).

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

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- This was a retrospective study so some missing data could not always be found and studies such as echocardiograms were not standardized.
- 5% of SCPC survivors were lost to follow up, so some important factors may be missing from analysis.
- Cause of death was not always available, so some deaths may not necessarily have been due to heart issues.

### **What it all means?**

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- This is the first time, to the authors' knowledge, that there was a significant association between RVPA shunt and attrition between SCPC and Fontan time period compared to the BT shunt.
  - The single ventricle trial noted trends with late deaths in the RVPA group, but it was not significant.
  - The larger number of patients in this study, 856 vs 356 in the single ventricle trial, may explain why this difference was statistically seen.
- Previous risk factors for death after Norwood procedure such as gestational age, birth weight, genetic abnormality, and pulmonary vein abnormalities were not significant in this study.
  - This may be due to survival effect in which patients that had these risk factors had already died prior to SCPC discharge.
- Even though AVV repair at time of SCPC was associated with attrition, the decision to repair the AVV at the time of SCPC is complicated. Further studies are needed to determine the best option when there are significant AVV issues.
- Other procedures performed at time of SCPC, longer SCPC surgery time, and longer SCPC hospital stay are likely secondary markers of more complex patients, hence the higher attrition.
- Attrition did not change over four eras studied. This may be due to higher risk patients surviving to SCPC procedure due to improvements in surgical and medical management.
- Strategies to improve survival between the SCPC to Fontan are needed as more complex patients are surviving to this time-period.
- Due to attrition in most patients occurring 13.7 months after SCPC, increased monitoring and vigilance in this interstage period may aid in mitigating attrition.