



National Pediatric Cardiology  
Quality Improvement Collaborative

## Research Explained

---

# Cumulative Comorbid Conditions Influence Mortality Risk after Staged Palliation for Hypoplastic Left Heart Syndrome and Variants

Emily Backes, Natasha Afonso, Danieele Guffey, James Tweddell, Sarah Tabbutt, Nancy Rudd, Ginny O'Harrow, Silvana Molossi, George Hoffman, Garick Hill, Jeffrey Heinle, Priya Bhat, Jeffrey Anderson, Nancy Ghanayem

J Thorac Cardiovasc Surg. 2023 Jan;165(1):287-298.e4. doi: 10.1016/j.jtcvs.2022.01.056. Epub 2022 Apr 2.

Steven Matthies (parent) and Matthew Moehlmann DO (Physician)

---

### ABOUT THIS STUDY

- The goal of this study was to learn if the presence of one high-risk feature was associated with reduced survival at one year of age compared to those without any high-risk features.
- Another aim was to learn if multiple high-risk features were associated with reduced survival at one year of age compared to those with one high-risk feature.

### Why is this study important?

---

- Patients with hypoplastic left heart syndrome (HLHS) (and other similar diagnoses) often have high-risk features (40% in this paper) which has traditionally been associated with reduced survival.
- High-risk features have usually been lumped together when comparing outcomes instead of looking at individual features themselves.
- This study looked at the outcome of survival to one year of age for patients with no high-risk features compared to patients with only one or multiple high-risk features.

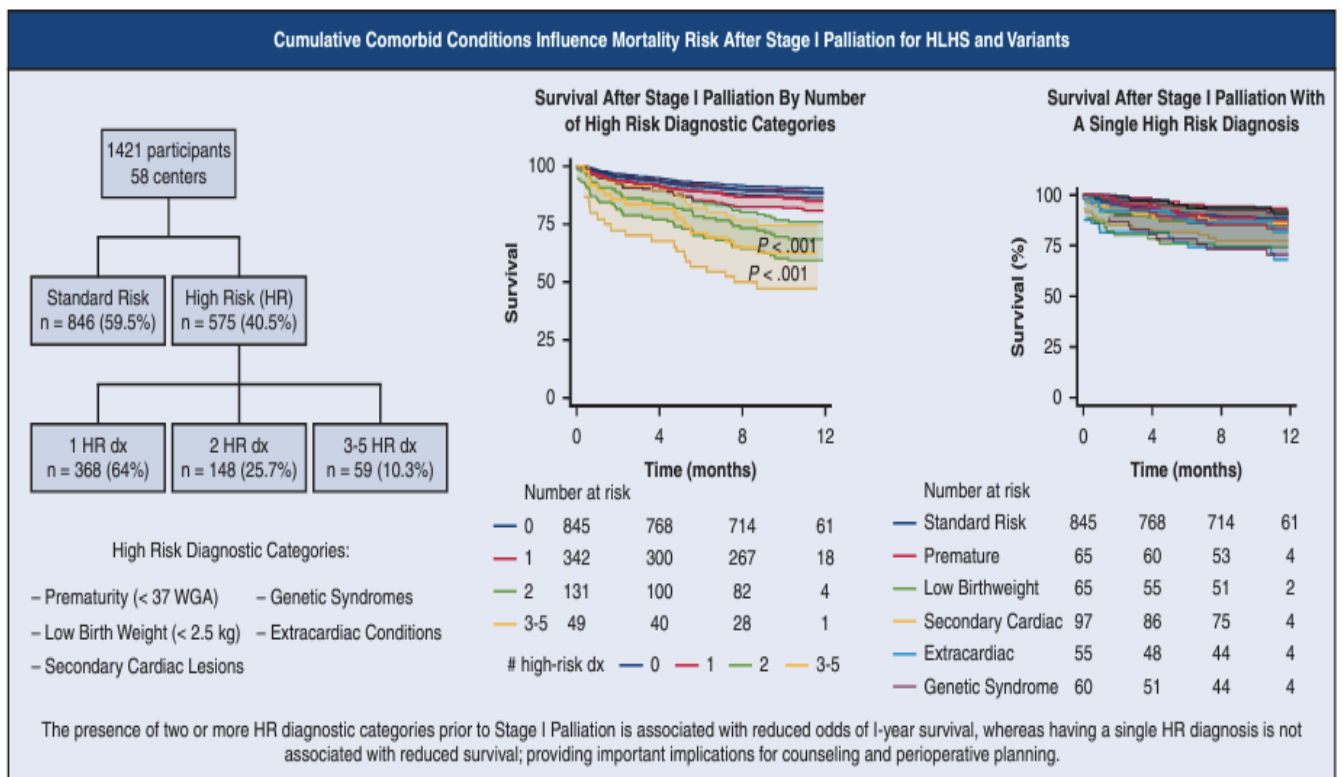
- High-risk features included secondary cardiac diagnosis, prematurity (<37 weeks gestation), low-birth weight (<2.5 kg), genetic syndrome, or other organ system anomalies

## How was this study performed?

- This study used information from a national database (phase 2 data from National Pediatric Cardiology Quality Improvement Collaborative registry).
- Survival to one year of age was compared between patients without high-risk features to those with one or more high-risk features.

## What were the results of the study?

- In the national collaborative, 40% had at least one high-risk feature.
- Patients without any high-risk features had survival to the first birthday of 88%. Those with one or more high-risk features (as a group) had survival of 76%.
- However, those with only one high-risk feature had similar survival to first birthday (84%) compared to those without high-risk features. Thus, no statistically significant difference was observed.
- Patients with two or more high-risk features had survival to one year of 56-64%.



### **What were the limitations of the study?**

---

- Outcomes of some patients that exited the study because of transplantation or different surgical plans were difficult to determine.
- The initial surgical strategy was often influenced by high-risk features. This makes it difficult to analyze these high-risk features without considering the surgical approach.

### **What it all means**

---

- A single high-risk diagnosis was not associated with reduced survival following surgery for HLHS and similar single ventricle diagnoses.