

Long-Term Transplant-Free Survival of Patients with Single Ventricle Congenital Heart Disease: Does Ventricular Morphology Matter?

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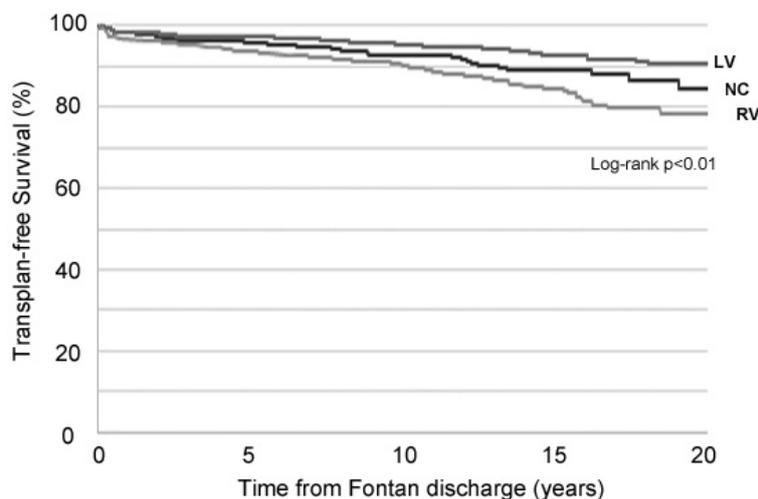
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Background: There have been conflicting studies regarding the association of ventricular morphology with long-term outcomes for children with single ventricle (SV). We used data from the Pediatric Cardiac Care Consortium (PCCC), a large multi-center US-based surgical registry, to examine the association of ventricular morphology with long-term transplant-free survival for children with SV.

Methods: This is a retrospective cohort study of individuals who had surgery for SV at <1 year of age at 45 centers in the PCCC from 1982-2003 and were linked with records from the US National Death Index (NDI) and Organ Procurement Transplantation Network (OPTN) registry through 2014. To examine overall survival and survival among those who had successful Fontan palliation, we constructed separate Kaplan-Meier transplant-free survival curves and mortality/transplant hazard ratios, adjusting for sex, syndrome, age at Fontan, type of Fontan (aortopulmonary, lateral tunnel or extracardiac), creation of fenestration at the time of Fontan, center, and birth era (early:1982-1992, middle: 1993-1997, and late: (1998-2003).

Results: Of the 3903 patients identified, the dominant single-ventricle morphology was right ventricle (RV) in 43.4%, left ventricle (LV) in 36.3%, and non-classifiable (NC) in 20.3%. Primary heart transplant or in-hospital death following first surgery occurred in 15.2% of LV, 17.6% of NC and 45.6% of RV. Among those who survived to discharge following the first surgery, overall 20-year transplant-free survival was 71.6% for LV, 63.2% for NC and 52.7% for RV. Compared to those with dominant LV, adjusted 20-year hazard ratio for mortality/transplant was 2.1 (95% CI 1.8-2.4) for RV and 1.4 (95% CI 1.2-1.7) for NC. Among the 1498 patients who survived Fontan hospitalization, 15-year transplant-free survival was 84.5% for RV, 92.8% for LV, and 88.8% for NC. Compared to those with dominant LV, adjusted 15-year hazard ratio for mortality/transplant following Fontan was 2.4 (95% CI 1.6-3.7) for RV and 1.6 (95% CI 1.2-3.0) for NC.

Conclusion: In this study, long-term transplant-free survival for patients with SV was greater for those with LV morphology. These findings have important prognostic value for providers, patients, and families.



	At risk				
LV	639	620	563	329	106
NC	326	311	250	111	27
RV	421	395	312	144	29