Transplantation in children with failed palliation of hypoplastic left heart syndrome: A good solution?

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Transplantation in patients with single-ventricle circulation after initial palliation is typically associated with worse outcomes. Previous studies have shown 1-year survival after transplant for this group to be as low as 70%.^{1,2} In this issue of *The Journal of Thoracic and Cardiovascular Surgery*, Alsoufi and colleagues³ analyze data from the Pediatric Heart Transplant Study to report outcomes for transplantation in children who previously had undergone palliation for hypoplastic left heart syndrome. The Pediatric Heart Transplant Study database is a multicenter database of currently more than 50 centers that report cardiac transplants in patients younger than 18 years. Alsoufi and colleagues' current study of 253 patients is the largest study to date looking at this specific subset of patients.³

Findings in most cases were not surprising, such as the fact that United Network for Organ Sharing status 1 and need for mechanical circulatory support were associated with death before transplant. The 1-, 5-, and 10-year survivals after transplant were 82%, 68%, and 59%, respectively. One of the more interesting findings from the study was that transplant after single-ventricle stage (ie, Norwood vs Glenn vs Fontan) palliation did not affect outcome. This finding is in contrast to previous single-center studies, which have demonstrated worse outcomes with heart transplants after the Fontan procedure.^{4,5} One reason for this may be that all patients in this study were younger than 18 years. This may be indicative of the more complex nature of adults with long-standing Fontan physiology and the usually more complicated nature of their cases from a surgical and medical perspective.

The limitations of the article are clearly mentioned by Alsoufi and colleagues³ and have to do mainly with the limitations inherent in working with a multi-institutional retrospective registry. There is likely significant sampling error introduced into the study, which is clearly indicated by the fact that "donor cause of death other than anoxic brain injury" was found to be a risk factor for mortality.

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Central Message

Cardiac transplantation in children with failed palliation for hypoplastic left heart syndrome can have excellent results.

See Article page 167.

See Editorial page 1404 in the December issue.

As Alsoufi and colleagues³ mention in their article, a larger report from the same Pediatric Heart Transplant Study database showed that survival after transplant was not affected by cause of donor death, which seems to indicate that sample size played a significant role in the statistical analysis.

This article should serve as an excellent reference for transplantation in children with failed palliation for hypoplastic left heart syndrome. Outcomes continue to improve for children with this condition, and should transplant become a necessity, the results can be encouraging.

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