

EDITORIAL COMMENT

## More Evidence for Regionalization\*



Carl L. Backer, MD,<sup>a</sup> Tara Karamlou, MD,<sup>b</sup> Karl F. Welke, MD<sup>c</sup>

**H**ear transplant in adults with congenital heart disease presents multiple challenges. These patients often present with complex anatomy, significant comorbidities, and have usually undergone numerous previous operations. Strategies that improve the survival of these complex patients should be carefully examined. In their report in this issue of the *Journal*, Nguyen et al. (1) from Seattle found that, for adults with congenital heart disease, post-transplant survival was best at the highest-volume regional centers. In addition, wait-list mortality and graft failure were reduced in centers that were Adult Congenital Heart Association (ACHA) Accredited Transplant Centers. An interesting finding that needs further analysis is that although wait-list mortality was lower at pediatric heart transplant centers caring for these patients compared to adult heart transplant centers, post-transplant graft failure at the pediatric heart transplant centers was higher. The authors postulate that regionalizing care of these complex patients would increase the volume of patients treated at each center and therefore improve overall results.

SEE PAGE 2908

These findings regarding adults with congenital heart disease and the impetus to regionalize care are consistent with studies evaluating the outcome of

regionalization for children undergoing congenital heart surgery. There is a positive correlation between surgical volume and survival for children undergoing congenital heart surgery. There appears to be an inflection point whereby survival is optimized at an annual case volume of >300 index cases (2). Regionalization of congenital cardiac care in countries other than the United States has been associated with a profound improvement in patient survival (3). We recently addressed hospital case volume distribution and patient travel patterns for congenital cardiac surgery in the United States (4). Our first analysis of state inpatient databases demonstrated that there are more U.S. hospitals performing congenital heart surgery than previously described. Many of these hospitals are low-volume centers in close proximity to one another. A critical finding of this study was that in the current system, patients with congenital heart disease are often bypassing their closest hospital and traveling long distances, mostly to the perceived high-volume centers of excellence. Our subsequent theoretical simulation model described the resulting landscape and survival of congenital heart surgery care in the U.S. if patients were sequentially relocated to higher-volume centers (5). With all hospitals performing >300 operations, between 12.5% to 17.4% fewer deaths were predicted to occur, roughly 1,000 lives saved over 10 years. At the same time, median patient travel distance increased from 38.5 miles to 69.6 miles. The conclusion of this simulation was that regionalization of congenital heart surgery in the United States to higher-volume centers may reduce mortality with a reasonable increase in patient travel distance.

These same findings regarding regionalization of care appear to apply to adults with congenital heart disease who require heart transplantation (1). It was interesting to note in the current report by Nguyen et al. (1) that nearly one-half of the centers transplanting adults with congenital heart disease transplanted <5 patients over an 8-year period. That is <1 patient per year! In fact, only 13 centers

\*Editorials published in the *Journal of the American College of Cardiology* reflect the views of the authors and do not necessarily represent the views of JACC or the American College of Cardiology.

From the <sup>a</sup>Pediatric Heart Transplant Program, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, Illinois; <sup>b</sup>Division of Pediatric Cardiac Surgery and the Heart and Vascular Institute, The Cleveland Clinic, Cleveland, Ohio; and the <sup>c</sup>Division of Pediatric Cardiothoracic Surgery, Levine Children's Hospital/Atrium Health, Charlotte, North Carolina. Disclosures: Dr. Backer is a consultant to W.L. Gore & Associates. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

transplanted >20 adults with congenital heart disease over the 8-year time period. The documented wait-list mortality and post-transplant graft failure curves from this paper show distinct improvements both in the higher-volume centers and in those centers that are ACHA accredited. These centers are required to have a multidisciplinary team with multiple board certified ACHA accredited cardiologists, and must include dedicated ACHA accredited heart failure specialists, congenital heart surgeons, and comprehensive diagnostic and interventional services by specialists in congenital heart disease. Adult congenital heart disease patients carry significant surgical risk and often require aorta and pulmonary artery reconstruction as well as venous or arterial rerouting techniques. These are operative strategies that may be more familiar to congenital heart surgeons than surgeons who specialize in acquired heart disease.

It is a slightly more complex issue to address the somewhat discordant findings of wait-list versus post-transplant outcomes in adults with congenital heart disease being treated at pediatric care centers. The wait-list mortality was lower among this group, but the risk of graft failure was higher. These transplant recipients were on average 14 years younger than recipients at adult hospitals, which may have contributed to retention at a pediatric care center. Ultimately, the data in this paper may not account for the combined patient (or hospital) factors that compel the care team to retain the patient at a pediatric institution. In addition, there are only a small number of pediatric hospitals transplanting adults with congenital heart disease. Out of 1,006 patients transplanted in this study, only 110 were transplanted at pediatric institutions.

It would be an interesting follow-up investigation to characterize the center and patient characteristics

further among these hospitals; in particular, knowledge about geographically proximate adult hospital “partners” may be important. The authors did not include hospitals’ experiences with heart transplants in adults with acquired heart disease or children who underwent heart transplant. It is likely that the care of these patients overlaps that of ACHD patients undergoing transplantation and should be added to the institutional experience. That said, it appears to be in patients’ best interests to be transplanted at an integrated ACHA accredited center that has the full capabilities to deal with adults with congenital heart disease. The findings of the current investigation corroborate previous investigations that have shown that patients with ACHD are best served by teams experienced with congenital rather than acquired heart disease (6,7).

In summary, post-transplant survival was better at the highest-volume regional centers when compared to other centers in a United Network Organ Sharing region. This finding is evidence that regionalized care of these complex adult patients may be associated with improved heart transplantation outcomes. Expertise in congenital heart anatomy and management appears to be important for the care of patients with complex congenital disease who are now adults and have advanced heart failure. This paper adds to the growing body of evidence that care for all patients with congenital heart disease (children and adults) should be regionalized.

---

**ADDRESS FOR CORRESPONDENCE:** Dr. Carl L. Backer, Division of Thoracic and Cardiovascular Surgery, Ann & Robert H. Lurie Children’s Hospital of Chicago, 225 E. Chicago Avenue, mc 22, Chicago, Illinois 60611. E-mail: [cbacker@luriechildrens.org](mailto:cbacker@luriechildrens.org). Twitter: [@karamlou](https://twitter.com/karamlou).

---

## REFERENCES

1. Nguyen VP, Dolgner SJ, Dardas TF, Verrier ED, McMullan DM, Krieger EV, et al. Improved outcomes of heart transplantation in adults with congenital heart disease receiving regionalized care. *J Am Coll Cardiol* 2019;74:2908-18.
2. Welke KF, O’Brien SM, Peterson ED, Ungerleider RM, Jacobs ML, Jacobs JP. The complex relationship between pediatric cardiac surgical case volumes and mortality rates in a national clinical database. *J Thorac Cardiovasc Surg* 2009;137:1133-40.
3. Lundström NR, Berggren H, Björkhem G, Jögi P, Sunnegårdh J. Centralization of pediatric heart surgery in Sweden. *Pediatr Cardiol* 2000;21:353-7.
4. Welke KF, Pasquali SK, Lin P, et al. Hospital distribution and patient travel patterns for congenital cardiac surgery in the United States. *Ann Thorac Surg* 2019;107:574-81.
5. Welke KF, Pasquali SK, Lin P, et al. Regionalization of congenital heart surgery in the United States. *Semin Thorac Cardiovasc Surg* 2019 Sep 10 [E-pub ahead of print].
6. Karamlou T, Diggs BS, Person T, Ungerleider RM, Welke KF. National practice patterns for management of adult congenital heart disease: operation by pediatric heart surgeons decreases in-hospital death. *Circulation* 2008;118:2345-52.
7. Karamlou T, Diggs BS, Ungerleider RM, Welke KF. Adults or big kids: what is the ideal clinical environment for management of grown-up patients with congenital heart disease? *Ann Thorac Surg* 2010;90:573-9.

---

**KEY WORDS** Adult Congenital Heart Association, adult congenital heart disease, heart transplantation, regionalization, United Network of Organ Sharing