reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Swisher M, Jonas R, Tian X, Lee ES, Lo CW, Leatherbury L. Increased postoperative and respiratory complications in patients with congenital heart disease associated with heterotaxy. *J Thorac Cardiovasc Surg*. 2011;141:637-44. 644.e1-3.
2. Alsoufi B, McCracken C, Schlosser B, Sachdeva R, Well A, Kogon B, et al. Outcomes of multistage palliation of infants with functional single ventricle and heterotaxy syndrome. *J Thorac Cardiovasc Surg*. 2016;151:1369-77.e2.
3. Jacobs JP, Pasquali SK, Morales DL, Jacobs ML, Mavroudis C, Chai PJ, et al. Heterotaxy: lessons learned about patterns of practice and outcomes from the congenital heart surgery database of the society of thoracic surgeons. *World J Pediatr Congenit Heart Surg*. 2011;2:278-86.
4. Loomba RS, Nijhawan K, Anderson R. Impact of era, type of isomerism, and ventricular morphology on survival in heterotaxy: implications for therapeutic management. *World J Pediatr Congenit Heart Surg*. 2016;7:54-62.
5. Jacobs JP, Anderson RH, Weinberg PM, Walters HL III, Tchervenkov CI, Del Duca D, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. *Cardiol Young*. 2007;17:1-28.
6. Van Praagh S, Kreutzer J, Alday L, Van Praagh R. Systemic and pulmonary venous connections in visceral heterotaxy, with emphasis on the diagnosis of the atrial situs: a study of 109 postmortem cases. In: Clark EB, Takao A, eds. *Developmental Cardiology: Morphogenesis and Function*. Futura Publishing Co; 1990:671-727.
7. Overman DM, Jacobs JP, Prager RL, Wright CD, Clarke DR, Pasquali SK, et al. Report from the Society of Thoracic Surgeons National Database Workforce: clarifying the definition of operative mortality. *World J Pediatr Congenit Heart Surg*. 2013;4:10-2.
8. Collett D. *Modelling Survival Data in Medical Research Second Edition*. Chapman & Hall/CRC; 2003:15-53.
9. Schoenfeld D. Partial residuals for the proportional hazards regression model. *Biometrika*. 1982;69:239-41.
10. Banka P, Adar A, Schaetzle B, Sleeper LA, Emani S, Geva T. Changes in prognosis of heterotaxy syndrome over time. *Pediatrics*. 2020;146:e20193345.
11. Lim H, Bacha E, Marx G, Marshall A, Fynn-Thompson F, Mayer JE, et al. Biventricular repair in patients with heterotaxy syndrome. *J Thorac Cardiovasc Surg*. 2009;137:371-9.e3.
12. Alongi AM, Kirklin JK, Deng L, Padilla L, Pavnica J, Romp RL, et al. Surgical management of heterotaxy syndrome: current challenges and opportunities. *World J Pediatr Congenit Heart Surg*. 2020;11:166-76.
13. Vodiskar J, Kido T, Strbad M, Cleuziou J, Hager A, Ewert P, et al. Outcomes of single ventricle palliation in infants with heterotaxy syndrome. *Eur J Cardio Thorac Surg*. 2021;60:554-61.
14. Marathe SP, Zannino D, Cao JY, du Plessis K, Marathe SS, Ayer J, et al. Heterotaxy is not a risk factor for adverse long-term outcomes after Fontan completion. *Ann Thorac Surg*. 2020;110:646-53.
15. Spigel ZA, Edmunds EE, Caldarone CA, Hickey EJ, Binsalamah ZM, Heinle JS. Total anomalous pulmonary venous connection: influence of heterotaxy and venous obstruction on outcomes. *J Thorac Cardiovasc Surg*. 2022;163:387-95.e3.
16. Foerster SR, Gauvreau K, McElhinney DB, Geva T. Importance of totally anomalous pulmonary venous connection and postoperative pulmonary vein stenosis in outcomes of heterotaxy syndrome. *Pediatr Cardiol*. 2008;29:536-44.
17. Jonas RA. Surgical management of the neonate with heterotaxy and long-term outcomes of heterotaxy. *World J Pediatr Congenit Heart Surg*. 2011;2:264-74.
18. Oshima Y, Yoshida M, Maruo A, Shimazu C, Higuma T, Inoue T. Modified primary sutureless repair of total anomalous pulmonary venous connection in heterotaxy. *Ann Thorac Surg*. 2009;88:1348-50.
19. McLennan D, Ruiz Solano EC, Handler SS, Lincoln J, Mitchell ME, Kirkpatrick E. Pulmonary vein stenosis: moving from past pessimism to future optimism. *Front Pediatr*. 2021;9:747812.
20. Shao P-L, Chen M-Y, Wu M-H, Wang J-K, Huang L-M, Chiu S-N. Nosocomial severe bacterial infection after cardiac surgery for complex congenital heart disease in heterotaxy syndrome. *Pediatr Infect Dis J*. 2020;39:e163-8.
21. Tan SY, Rosenthal J, Zhao X-Q, Francis RJ, Chatterjee B, Sabol SL, et al. Heterotaxy and complex structural heart defects in a mutant mouse model of primary ciliary dyskinesia. *J Clin Invest*. 2007;117:3742-52.
22. Banka P, Schaetzle B, Komarlu R, Emani S, Geva T, Powell AJ. Cardiovascular magnetic resonance parameters associated with early transplant-free survival in children with small left hearts following conversion from a univentricular to biventricular circulation. *J Cardiovasc Magn Reson*. 2014;16:73.
23. Prendiville TW, Barton LL, Thompson WR, Fink DL, Holmes KW. Heterotaxy syndrome: defining contemporary disease trends. *Pediatr Cardiol*. 2010;31:1052-8.

Key Words: biventricular, heterotaxy, single ventricle

Discussion

Presenter: Manan Desai

Dr Joseph Dearani *(Rochester, Minn)*. Many patients with heterotaxy syndrome require SV palliation and are at a higher risk for worse outcomes compared with patients without heterotaxy. However, some patients with heterotaxy are managed with a BiV strategy. The long-term outcomes of patients with heterotaxy managed with these 2 strategies is not completely understood, with little evidence in the literature. So you are addressing an important topic in an area for which little information exists. In general, we would expect patients with heterotaxy who are able to undergo a BiV repair to have superior outcomes compared with those who underwent SV palliation. I have 3 questions for you. Number one: Taking into consideration presumed superiority of normal physiology with a BiV circulation over Fontan palliation, why do you think this was not the case in your series? Was it related to atrioventricular valve issues, pulmonary vein issues, or to directing patients toward a BiV repair who were borderline candidates to begin with?

Dr Manan Desai *(Washington DC)*. I think it is a bit of everything you mentioned. Abnormal valves, a little bit of borderline ventricle of one or the other, a small right ventricle or a small left ventricle, and pulmonary vein issues. As my analysis for organ system failure preceding death shows, less than half of patients had a cardiac system involvement. So we are looking at a patient who has a multisystem involvement. So it's not only that something wrong with the heart resulted in the death of the patient. I emphasize the need for a multidisciplinary team required to take care of such patients because they have, as has been shown by

previous studies by our institution, abnormal ciliary clearance and other issues, which result in a higher incidence of tracheostomy and duration of ventilation. So it's a bit of both, and we should look at these patients as having a multisystem disorder rather than just a cardiac abnormality.

Dr Dearani. Okay. Total anomalous pulmonary venous connection was found to be a risk factor for death in your study. Have you looked at patients who underwent surgical intervention for the total anomalous veins compared with those who had the total veins present but did not have surgical intervention to see if the risk was related to technical issues related to those who actually underwent surgical repair?

Dr Desai. Unfortunately, we don't have that comparison. A patient with BiV repair usually needs a correction for a TAPVR because of the physiology. In the SV, however, you can avoid a total pulmonary venous return repair if the confluence is unobstructed.

Dr Dearani. Exactly.

Dr Desai. Only need for the repair would be pulmonary venous stenosis, which was also a common risk factor in our study. So those patients did end up getting sutureless repair, but the outcomes, as we see, were not as good.

Dr Dearani. Okay. Finally, many studies have demonstrated significantly worse outcomes in the asplenia cases compared with the polysplenia cases. Did you find a difference in outcomes between asplenia and polysplenia in your heterotaxy series?

Dr Desai. We had a high prevalence of splenic abnormalities. Up to 60%. As I have shown you, one of the slides had one of the splenic abnormalities, either asplenia or polysplenia. But when we did our univariate and multivariable analyses, it was not found to be a significant risk factor.