

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

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Key Words: biventricular, heterotaxy, single ventricle

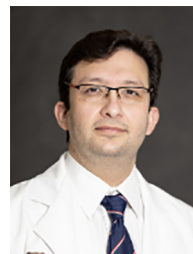
Discussion

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.