

EDITORIAL COMMENT

Biventricular Repair or Single-Ventricle Palliation

Can Cardiovascular Magnetic Resonance Flow Imaging Help in Decision-Making?*

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It continues to be a challenge deciding whether patients with borderline hypoplastic left ventricles (b-HLHS) will be suitable for a biventricular repair (BVR) or if (long-/mid-term) survival can only be reached with single-ventricle (SV) palliation. Until recently, echocardiography has been the main imaging modality used to address this issue in clinical practice but has limitations as highlighted in a recent review of older and newer echocardiographic prediction models for BVR.¹ Cardiovascular magnetic resonance (CMR) imaging has gained some acceptance as a tool for answering the question “BVR or SV palliation?”, but the number of CMR or multimodality studies (echocardiography and CMR) are relatively small.²⁻⁴ In this issue of *JACC: Advances*, Kang et al⁵ provide new insights in the use of CMR flow imaging in b-HLHS patients. In this retrospective study, the authors demonstrate that CMR phase contrast flow measurements in the ascending aorta may be helpful for triaging patients to either BVR or SV palliation.⁵ In addition, the presence of aortic coarctation with the need for repair was associated with a better survival and those patients without endocardial fibroelastosis

were deemed more suitable for BVR.⁵ In the multivariable Cox regression analysis, ascending aortic flow of <1.5 L/min/m² and not requiring aortic coarctation repair were associated with a worse transplant-free and SV pathway conversion-free survival.

The rather positive impact of aortic coarctation on outcome can be explained by studies showing different effects of coarctation and critical aortic stenosis on the left ventricular myocardium.^{6,7} Whereas the hearts with aortic coarctation show normal myocardial organization, the hearts affected by congenital aortic stenosis are affected by variable amount of fibrosis.^{6,7}

Although this is a single-center retrospective study with a relatively small number of patients, the results contribute to the already existing literature and suggest that CMR flow data should be considered in decision-making in b-HLHS patients, and moreover, they appear to be superior when compared with volumetric analysis. Interestingly, no or only minor correlation was found between ascending aortic flow and end-diastolic left ventricular volume and mitral and aortic valve Z-scores. This is not surprising as the forward flow in the ascending aorta allows for assessment of the true hemodynamics, including the impact of multilevel left heart disease in the individual patient.

Kang et al⁵ also confirm another important issue, namely, that left ventricular volumes derived from 2-dimensional echocardiography are not well suited for decision-making in b-HLHS patients.⁴ CMR is the gold standard for ventricular volume measurements using the Simpson’s method.⁸ Echocardiography typically underestimates ventricular volumes, which can result in triaging b-HLHS patients to SV palliation who would otherwise be candidates for BVR.^{5,9} In fact,

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it is striking that based on the echocardiographic criterion of indexed left ventricular volume ≥ 20 mL/m², 83% of patients would have precluded from BVR in a study by Kang et al. Three-dimensional echocardiography and a thorough analysis of potential causes for differences between CMR and echocardiography might be an option to overcome the volumetric differences.^{10,11} However, CMR still has the advantage to provide unlimited planes not affected by imaging windows, making it a very attractive and more comprehensive imaging modality.

Nevertheless, studies have shown that other imaging and hemodynamic parameters can also improve the prediction of long-/mid-term biventricular survival. Banka et al³ investigated b-HLHS patients who underwent conversion from an SV to a biventricular circulation and showed that CMR together with echocardiographic parameters were helpful in predicting biventricular survival. The same group demonstrated that elevated left ventricular end-diastolic pressure was a risk factor for adverse outcome and suboptimal hemodynamics after biventricular conversion.¹² Thus, a multimodality approach in this complex and heterogeneous group of patients might be best for triaging children with b-HLHS to either BVR or SV palliation. Finally, intracardiac exploration should not be forgotten, as this can be essential for making the “final” decision for the most appropriate treatment.¹³

In summary, the study by Kang et al showed that CMR flow imaging in the ascending aorta can make a difference when dealing with the complex group of

patients with b-HLHS. Performing CMR scans in all newborns and young infants with b-HLHS, as part of an institutional protocol, appears to be a reasonable approach. A combination of imaging modalities including cardiac catheterization data is also advised and should be considered in future scoring systems. Furthermore, fetal CMR can provide information and should be taken into account.¹⁴ There is also a growing body of literature suggesting a positive impact of fetal interventions on postnatal BVR in select cases. Emerging technologies in fetal CMR might be helpful, together with echocardiography, in recognizing the most suitable patients and optimal timing for this alternative approach, enabling growth of the left heart myocardium.¹⁵ Finally, do not forget, although not scientifically supported, sometimes it is useful to also incorporate your gut feeling. Despite the difficulty treating b-HLHS patients, Kang et al have provided additional meaningful data that support the use of CMR in decision-making.

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