EDITORIAL COMMENTARY

Kicking the can down the road … gently

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Hypoplasia of left heart structures presents along a spectrum, ranging from the most severe, mitral and aortic atresia, to the least severe, coarctation of the aorta with bicuspid aortic valve. Two alternative treatment strategies now exist, with assignment of a patient either to a biventricular circulation or to a univentricular arrangement, depending on the perceived functional adequacy of the left side of the heart. Thus a “continuous” disease must be...
managed with "categorical" therapies. For many patients presenting in the middle of the spectrum, it is difficult to discern which strategy is most appropriate, although several predictive scoring systems to guide such decisions have been proposed. Retrospective analysis suggests that inappropriate assignment may result in substantial morbidity, or even death. The report in this issue of the Journal by Yerebakan and colleagues from Giessen describes the outcome for a cohort of patients with varying degrees of left heart underdevelopment in whom the treatment assignment decision was delayed, with the patients initially undergoing a temporizing hybrid palliation in the neonatal period. No patients died in the interstage interval. Ultimately, 40 patients underwent biventricular reconstruction at a median age of nearly 7 months, with 5 deaths, 1 cardiac transplant, and 1 takedown to univentricular circulation, at a median follow-up of 7.9 years.

In an alternative approach suggested by the Boston group, initial palliation is a conventional first-stage Norwood-type operation, with either modified Blalock-Taussig shunt or right ventricle–to–pulmonary artery shunt, deferring the biventricular conversion until after the second or even third stage of palliation. Advantages of this approach include the opportunities to perform complex left-sided valve reconstruction, to resect endocardial fibroelastosis, and to enhance pulmonary blood flow, all before biventricular reconstruction. The Giessen approach does not permit these options, although approximately 10% of their patients had mitral or aortic reconstruction at the time of biventricular repair.

Not surprisingly, the Giessen patients were substantially younger at biventricular conversion (median age of 6.7 months vs 42 months in the Boston experience). The Giessen group had also not previously undergone heart operations at the time of biventricular conversion, very different from the Boston patients, who had undergone a median of 3 heart operations before definitive repair. An area of commonality between the 2 approaches is the focus on the atrial septum at initial palliation. The aim for both is to leave a restrictive interatrial communication after initial palliation. Indeed, no patient in the Giessen cohort underwent atrial septal intervention before biventricular conversion, guided by a threshold left atrial pressure of 15 mm Hg. The underlying hypothesis is that “forcing” blood to cross the left side of the heart, rather than the atrial septum, promotes growth of the mitral valve, left ventricle, and aortic valve. Comparison of echocardiographic measurements before the hybrid procedure with the same assessments before the biventricular conversion demonstrated a significant increase in aortic valve z scores (−2.13 to −1.21). The change in mitral valve z scores (0.52 to 0.98) was less impressive, however, and there was no significant change in the ratio of left to right ventricular length (0.90 to 0.92). Nonetheless, although the relative sizes of various left heart structures may have changed only slightly, if at all, the impact on the discriminant score (predicting suitability for biventricular repair) was a change from below the discriminant threshold of −0.65 to substantially above it by the time of biventricular conversion (−0.72 to 1.47).

With continued reference to the atrial septum, it is notable that at biventricular repair Yerebakan and colleagues chose to leave an atrial septal defect in 25% of the patients. Although it is not explicitly stated, here the concept was apparently away from forcing transmural flow to encourage growth and toward prevention of early left atrial hypertension–induced pulmonary edema. This principle is not new, first appearing in the prebypass era in the management of patients with mitral stenosis and much later in the neonatal biventricular repair of left heart complex. The Boston group also reports leaving an atrial septal defect in most of their patients at the time of biventricular repair.

For a neonate who is does not obviously fit into either the biventricular or univentricular paradigm, this report suggests that assignment can be safely and successfully delayed for several months. The approach that Yerebakan and colleagues have described is simple, avoids exposure of a fragile neonate to cardiopulmonary bypass with or without circulatory arrest, does not require multiple operations before definitive repair, and seems to permit correct patient management strategy assignment in the vast majority of cases. For centers wishing to duplicate this experience, it must be remembered that the Giessen group has among the longest and largest experience in the world with the hybrid approach, and subtle insights and judgmental nuances derived from such an extensive experience are often difficult to detect and convey in retrospective analyses.

References