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Commentary: Skeleton in the closet: Toward durable repair of atrioventricular valve in univentricular circulation

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Children with unbalanced atrioventricular septal defects (uAVSD) are amongst the highest risk patients with single ventricle physiology.¹ Early reports showed extremely poor outcomes in these children,^{2,3} which has spurred interest in the pursuit of strategies to recruit or convert patients with uAVSD to a biventricular circulation.⁴ However, it has recently become clear that survival with single ventricle palliation is similar to that of complex biventricular strategies,^{5,6} and that atrioventricular valve (AVV) failure is the major factor contributing to morbidity and mortality in patients with univentricular circulation.⁷

Arrigoni and colleagues⁸ report the outcomes of 151 children with uAVSD from a binational cohort, including 6 centers across Belgium and the Netherlands. They demonstrated survival of 68% at 20 years, better survival than previously reported. However, they observed that more than half of the patients required reoperation on the AVV by 10 years, and that the presence of severe AVV regurgitation was the only independent risk factor for mortality. AVV surgery was not a risk factor for failing to achieve Fontan completion. They conclude that patients

CENTRAL MESSAGE

As survival of patients with unbalanced atrioventricular septal defect has improved, attention must shift to treating and preventing atrioventricular valve regurgitation.

with greater-than-moderate AVV regurgitation should undergo AVV repair.

These findings reiterate the importance of AVV function for the long-term outcomes of patients with uAVSD. Indeed, patients with uAVSD have the highest risk of AVV failure of all patients with single ventricle physiology.⁹ Yet, AVV repair in these patients is extremely challenging, and we have previously shown that failed repair is associated with a very high risk of mortality, especially in infants.¹⁰ Early experience suggests that even in patients with biventricular conversion, the AVV remains a major cause of morbidity.⁴ Thus, a durable repair of AVV is crucial in both univentricular and biventricular circulation. How can we achieve a durable repair? In nature, both exo- and endoskeleton provide crucial support for all tissues and permit lasting long-term durability of the entire organism. In the heart, such support to the valves is provided by the fibrous skeleton of the heart (Figure 1, A). The normal human heart has evolved to have a well-defined fibrous skeleton, particularly at the central fibrous body, the site of continuity of the aortic, mitral, and tricuspid annuli. On the other hand, the heart with a common AVV has a maldeveloped fibrous skeleton of the heart (Figure 1, B), which is particularly attenuated in those with hypoplastic left-sided heart structures.¹¹ Should we not aim to emulate nature? Should restoration of the fibrous skeleton of the heart become a routine part of our surgical armamentarium? Bridging annuloplasty aims to reconstruct the fibrous

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FIGURE 1. A, Fibrous skeleton in the normal heart. B, Maldeveloped fibrous skeleton in the heart with complete atrioventricular septal defect.

skeleton of the heart and prevent progressive annular dilatation in patients with a common AVV.¹² Although early results appear promising, long-term follow-up is required to see if these results are durable.

Given the high rate of AVV failure in patients with uAVSD, is there a role for prophylactically stabilizing the AVV annulus before the development of significant regurgitation? Arrigoni and colleagues⁸ have demonstrated that AVV surgery does not increase the risk of mortality or decrease the risk of achieving Fontan completion, demonstrating the safety of AVV repair. Future studies should aim to identify how best to prevent the progression of AVV in these patients, and which patients, if any, would benefit from prophylactic stabilization of the AVV annulus. Perhaps by intervening before significant AVV regurgitation develops we can break the cycle of progressive annular dilatation, regurgitation, and ventricular dysfunction and improve the long-term survival of this high-risk group of patients. When it appears that surgical armamentarium is exhausted and there is nothing left in the armory, we should look into the depths of the closet. We may find new ideas!

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