Don't Blame the Butter for Something the Bread Did

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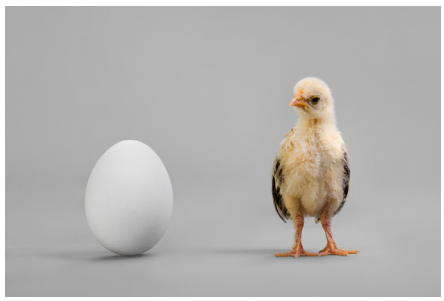
Despite the application of even the most advanced repair techniques, the systemic atrioventricular valves (SAVV) of some single ventricle patients ultimately require valve replacement. The timing of this intervention represents a careful balance of clinical necessity, likelihood of annular growth, patient size, and chronological proximity to the next-staged procedure. Indeed, the temptation is strong to restore valve competence as early as possible to enhance overall cardiac output in particular for those patients "stalled" in their clinical progress. Yet, the known complications of valve replacement in this smaller sized patient cohort are significant—heart block, stroke, valve thrombosis, low cardiac output, and mortality. Nakata et al in this issue of *Seminars* present their single-center experience with SAVV replacement (SAVVR) in single ventricle patients across the entire timeline of staged palliation in an effort to better elucidate optimal timing and candidates for this approach, given the poor results traditionally reported for this population.[1–3]



Clarifying which came first can be both fundamental and challenging.

Central Message

Systemic atrioventricular valve replacement for those with single ventricle disease demonstrates highest mortality in the smallest patients.

Although tempting to extrapolate from the adult literature the negative impact of patient-prosthesis mismatch as the fundamental culprit in these morbidities, in the pediatric literature this causality may be obscured by the fact that the smaller size of the patient who requires valve replacement may simply be a surrogate of disease severity (and degree of abnormality ["irreparability"] of the valve). The authors intriguingly utilized a new criterion, the geometric orifice area index (GOAI) to more accurately assess the degree of valve "oversizing" (determined by receiver operating characteristic curve analysis to have a threshold of 6). Yet, almost half of their patients utilized the 2 smallest commercially available valves in Japan, suggesting that even in the smallest patients the degree of oversizing was unavoidable.

Unfortunately, we cannot surmise from Nakata's data that implantation of smaller valves (eg, 15 mm) would have improved outcomes had they been available. Recent reports of "repurposed" transcatheter balloon expandable valves for AVVR offer the possibility of further upsizing with growth, but still confer a considerable morbidity and mortality even when including results of those implanted who had 2 ventricles.[4] An inherent appeal to using such tissue valves is the lack of necessity for anticoagulation, which is not a trivial advantage when considering Nakata's series reported an early thrombosis rate of 11% with associated 50% mortality.

The authors suggest that the therapeutic effect of SAVVR before Fontan completion could be limited, but that earlier SAVVR after Fontan (before the onset of more significant ventricular dysfunction) may be advantageous due to improvements in ventricular performance not unlike what has been demonstrated in teenagers and adults with rheumatic valve disease.[5] Indeed, coupled with a predicted sobering rate of progressive AVVR following Fontan in a considerable proportion of patients, this observation may serve to endorse a surgical strategy of earlier intervention despite perceived operative risk.[6]

But what, then, to make of SAVVR in the smaller patient? The scatterplot data would suggest that while there were indeed survivors with a GOAI >6, there were no survivors with a BSA <0.25 m^2; moreover, one of the strongest hazard ratio for death in the multivariable domain was patient size. A small, critically ill single ventricle infant develops severe AVVR, and that begets more illness—but which came first? Are we now vilifying the prosthetic valve as an accomplice to a feedback loop of disease severity and valve pathology—a Plutarchian play of chicken and egg?[7]

The complexity in managing this difficult patient population is no better illustrated than in figure 5, and the authors are to be commended for their assiduous pursuit of improved outcomes in this high-risk cohort. The inference from Nakata's

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series is that bigger may not always be better, and the need for SAVVR (in particular as a primary procedure) in single ventricle infants may, by its necessity, presage a poor outcome. Whether incremental improvements in the outcomes of these smaller patients can be achieved, either by reducing the GOAI (potentially with smaller transcatheter biological valves), or by refining anticoagulation remain to be seen; without a doubt, though, we can only do better.

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