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Commentary: Despite best intentions: Developing better strategies for patients with pulmonary atresia with intact ventricular septum

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Pulmonary atresia with intact ventricular septum (PA/IVS) encompasses a spectrum of morphologies with varying degrees of right heart hypoplasia and abnormalities in coronary perfusion. The latter creates a notoriously fragile circulation with high mortality. Defining categorical management strategies for single ventricle (SV) versus biventricular (BiV) physiology, particularly those with right ventricle-dependent coronary circulation (RVDCC), remains challenging and ill defined.

Sukhavasi and colleagues¹ report their large, single-center experience with PA/IVS, define patient characteristics, and report long-term outcomes. Their institutional approach was dichotomous in allocating patients into either a BiV or SV pathway based almost exclusively on the decision to decompress the RV, which in turn was mainly based on an absence of RVDCC. They did not have a protocolized approach but made team decisions that also took RV size into account. Of note, there was no institutional policy to pursue one-and-one-half repair as a definitive circulation. Of the intended BiV cases (54% of patients), only 56% achieved a BiV end state at a mean follow-up of 6 years. The data suggest that 6% converted to SV, 8% are likely to be one-and-one-half circulations, with 5% mortality/transplant. A further 11%

Angiogram showing extreme RV-dependent coronary circulation with coronary ostial atresia.

CENTRAL MESSAGE

Mortality remains high in patients with PA/IVS with right ventricle-dependent coronary circulation. Strategies tailored to morphologic and anatomic characteristics are required to improve outcomes.

are described as having moderate to severe RV hypoplasia potentially influencing their suitability for a BiV repair; therefore, approximately 70% to 75% of those with intended BiV ultimately achieved that end state.

Although decision making was not initially based on categorical *z* scores, it is interesting to note that the vast majority of BiV repair had tricuspid valve *z* score >-2.5 and that all patients who were likely to have one-and-one-half repair had *z* scores of -2.8 to -5.3 , which is very much in line with previously reported observations. As expected, those achieving BiV end state had significantly larger tricuspid valve *z* scores and almost zero incidence of RVDCC. The SV group had significantly higher mortality (34% vs 4.7%), the majority of which was in patients with RVDCC (despite not decompressing the RV) and the majority of deaths occurred before age 2 years, an important observation.

The study provides further unequivocal evidence that cases with RVDCC (particularly with coronary ostial atresia) (Figure 1) are the highest-risk patients and risk of myocardial ischemia remains a limiting factor in both short- and long-term survival.² This study reports a 26% incidence of RVDCC (defined as any stenosis or interruption that would place any size of myocardium at risk for ischemia with RV decompression) and was associated with an overall mortality of 42%—which begs

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FIGURE 1. Angiogram showing extreme RV-dependent coronary circulation with coronary ostial atresia.

the question of whether or not different strategies could improve the prognosis for this group. Primary transplantation should be considered, especially in the setting of ostial atresia,³ and use of patent ductus arteriosus stents (used in 5 SV patients) can facilitate waitlist logistics and avoid surgical morbidity. Patent ductus arteriosus stents may be underrepresented due to an era effect with the dramatic increase in cath lab interventions in recent years. For patients with ongoing risk of myocardial ischemia or infarction, the use of ventricular assist devices has also been described,^{4,5} as well as the use of aorto-RV shunts.⁶ Larger multi-institutional studies are required to delineate the influence of aortocoronary connections, coronary anatomy considerations, and thresholds of myocardium at risk to aid clinical decision making in this high-risk cohort.

It is an oversimplification to commit to SV versus BiV strategy based purely on whether or not the RV can be decompressed as a neonate because only 70% to 75% of cases will achieve BiV by this assumption. The well-recognized morphological characteristics of RV size

are still very important and tricuspid valve z scores remain strong predictors of the ultimate destination for the RV—as summarized in the authors' statement that "morphology at birth dictates outcome." Past lessons have taught us that it is difficult to demonstrate true growth of the RV despite early decompression and aggressive rehabilitation,^{7,8} and so use of a one-and-one-half ventricle intended strategy is a valid destination in certain borderline cases (probably 5%-10% of the population). By recognizing a spectrum of intended strategies, one can optimize the physiological end state by not forcing an inadequate BiV physiology or restricting one to an exclusive SV physiology. Only time will tell how expanded strategies for PA/IVS will influence long-term survival.

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