All about the coronaries

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Left to right: Syed Bukhari, MD, Manan Desai, MD, and Can Yerebakan, MD

Central Message

Coronary blood flow can successfully be sustained with an aorta–right ventricle shunt implantation in severe cases of pulmonary atresia with intact ventricular septum.

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Pulmonary atresia with intact ventricular septum is a rare congenital heart disorder with poor prognosis in severe cases. The fate of the disease is determined by such morphologic determinants as the degree of the dysplasia of right ventricular structures, such as the ventricular cavity, tricuspid valve, and pulmonary valve. The spectrum of treatment can extend from interventional or surgical right ventricular outflow procedures,[1] to systemic–pulmonary artery shunt procedures, and ultimately to inevitable cardiac transplantation in severe cases. The source of coronary artery supply affects the timing and the type of the required procedure. A right ventricle–dependent coronary circulation with bilateral coronary ostial atresia, for instance, would considerably limit the odds of survival without cardiac transplantation.[2,3] One important goal of surgical intervention in severe cases is to ensure an adequate coronary blood flow. Lower diastolic pressures after a systemic-pulmonary shunt procedure can jeopardize the coronary blood flow, which may already be critical because of abnormal coronary anatomy.[4] Any acute hemodynamic compromise from coronary ischemia is an extremely challenging problem in this entity. Even salvage extracorporeal membrane oxygenation therapy may decompress the right side of the heart, lower the right ventricular filling pressures, and worsen the right ventricle–dependent coronary circulation, leading to fatal ischemia.[5]

In this issue of the *Journal*, Sakurai and colleagues[6] describe a successful surgical technique in a patient with bilateral coronary atresia and thus a completely right ventricle–dependent coronary perfusion. Their surgical approach involved the addition of an aorta–right ventricle shunt combined with tricuspid valve closure and a right-modified Blalock-Taussig shunt for coronary perfusion. Although similar attempts have already been reported,[7,8] Sakurai et al[6] are the first to achieve success—survival of the patient without cardiac transplantation—in a case of bilateral coronary ostial atresia. Our colleagues from Japan are to be congratulated not only for successfully saving the patient but also for transitioning to the next stage of the single-ventricle pathway. The challenge in this particular strategy may be the high variability in anatomy and resultant unpredictable flow dynamics. In other case series, left ventricular volume overload was reported while some of the patients had precipitously low systemic diastolic pressure that required a snaring of the aorta–right ventricle shunt.[8] The selection of the right ventricular shunt size may not be straightforward, and the size may require secondary adjustments. In this case, a successful revision of the aorta–right ventricle shunt was performed when the patient was 7 months old to correct shunt stenosis. The described strategy is of importance in that it presents a feasible salvage option for patients in extremis, enabling a bridge to a single-ventricle pathway or to cardiac transplantation. Cardiac transplantation will still remain the therapy of choice in severe cases until novel options can be proved to produce comparable long-term outcomes.[3]

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