See Article page 1166.

Commentary: Atrioventricular valve insufficiency: Achilles' heel in single-ventricle physiology

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Atrioventricular (AV) valve insufficiency in patients with a functional single ventricle causes significant chronic volume overload to the systemic ventricle, which negatively influences all stages of single-ventricle physiology, resulting in high mortality and failing Fontan circulation.^{1,2} The multicenter study conducted by Arrigoni and colleagues³ sheds a light on the long-term influence of AV valve insufficiency on patients with unbalanced AV septal defect (AVSD). Patients with unbalanced AVSD and the common AV valve are known to have various degrees of AV valve structural abnormalities, which often hinders successful single-ventricle palliation and maintenance of the Fontan circulation in the long term.² The study provides unique new data on this matter. Firstly, the study includes very long-term follow-up of AV valve function in patients with unbalanced AVSD. Perhaps most important, variability in clinical practice among the participating institutions regarding threshold and timing of surgical intervention to a regurgitant AV valve gives a unique perspective for patients with significant untreated AV valve insufficiency acting as a control group. The study confirmed what we currently believe when treating patients with single-ventricle physiology with a significantly regurgitant AV valve: concomitant AV valve repair did not increase mortality nor decrease the rate of Fontan completion, and the patients who had significant AV valve insufficiency in the follow-up echocardiogram had a significantly lower survival rate.³ The findings from this study are consistent

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CENTRAL MESSAGE

The BeNe experience confirmed the importance of maintaining AV valve competency in patients with a single-ventricle physiology, and untreated AV valve insufficiency is related to lower survival rate.

with another recent, large study conducted by King and colleagues.⁴ The study reported the freedom from Fontan failure in patients with valvular failure at 10 and 20 years after Fontan surgery were 77% and 54%, respectively, and suggested Fontan circulation was likely to fail in patients with AV valve failure twice as high as those without AV valve failure.⁴

One of our studies, which investigated the mechanism of AV valve insufficiency in patients with single-ventricle physiology, demonstrated that approximately 85% of the AV valve with significant insufficiency had structural abnormalities such as leaflet dysplasia, prolapse, cleft, or double orifice.¹ Moreover, our follow-up study showed that leaflet dysplasia and residual AV valve insufficiency are identified as predictors for AV valve re-repair or replacement.⁵ There was a notion in the past that the volume unloading operation, such as bidirectional cavopulmonary shunt, might improve the degree of AV valve insufficiency. Although we sometimes see this phenomenon in patients with minor AV valve insufficiency without major AV valve structural abnormalities, the majority of the cases with significant AV valve insufficiency will not be improved only by the volume-unloading surgery. Lastly, the systemic single ventricle that is exposed to chronic volume overload caused by AV valve insufficiency may have significant myocardial dysfunction following successful AV valve repair, which adversely influences single-ventricle physiology.¹ This is additional rationale to intervene in significant AV valve

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insufficiency earlier than later before irreversible myocardial damage occurs. The investigators from the BeNe experience—the scientific cooperation of Belgium and Netherlands—should be congratulated on the well-conducted cohort study, which adds new knowledge in the management of patients with single-ventricle physiology.

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