Partial Left Ventriculectomy for End-Stage Dilated Cardiomyopathy in Small Children

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Dilated cardiomyopathy (DCM) in the pediatric population is a rare myocardial disorder that commonly results in congestive heart failure. The majority of patients are idiopathic. Freedom from death or transplantation is disastrous for all categories, particularly those with idiopathic DCM. Heart transplantation is required for these cases of DCM when medical treatment, such as angiotensin-converting enzyme inhibitors or β-blockade, or mechanical ventilation and circulatory support do not sufficiently improve the left ventricular (LV) dysfunction. When heart transplantation is performed, it provides superior survival rates: 83% and 63% for 1 year and 10 years, respectively, compared with non-transplant therapy. Unfortunately, however, in Japan small children are not legally provided with any chance for heart transplantation. In addition, the handful of opportunities for transplant tourism to foreign countries will soon be prohibited according to the agreement created in Istanbul in 2008. For the moment, especially in Japan, surgical alternatives for diluted heart failure have gained particular importance in the management of end-stage DCM.

In this issue of the Journal, Sugiyama et al describe partial left ventriculectomy (PLV) with an edge-to-edge procedure (Alfieri’s repair) and/or mitral valve replacement for idiopathic DCM. There are few serial studies of the PLV procedure used for DCM in children, so we congratulate the authors on their achievement of satisfactory outcomes with their surgical strategy for end-stage DCM in small children. Although mortality rates for children with DCM have been reported as high, 16–25% cases of idiopathic DCM exhibit improvement in LV function5,6. It is noteworthy that recovery of LV function to within the normal range has been observed after 3 years from initial examination in 69% of patients whose LV performance recovered. In this situation, both the indication and the decision of timing the surgical procedure in children with DCM are matters of difficulty just several months after onset.

The advantage of PLV over transplantation includes freedom from immunosuppressant therapy and from risk of infection. PLV appears to gain much predominance over heart transplantation for refractory small children. However, in adults the PLV procedure is recognized as unsuitable for nonischemic end-stage DCM according to the AHH/ACC guideline for chronic heart failure. A report from the Cleveland Clinic presented a significant early failure rate and even free survival at 3 years of only 26% in adults. Attempting PLV for pediatric DCM might be justified if the long-term durability of this procedure can be assessed by relatively large studies. However, even the use of LV assist devices in pediatrics is uncommon in Japan, so durability of more than 5 years in selected cases that the authors present is good news for these children.

Functional mitral regurgitation (FMR) in the setting of DCM, which occurs as a consequence of global LV dilatation, results in progressive exacerbation of LV function and clinical symptoms. The authors performed mitral leaflet approximation, so called edge-to-edge mitral valve repair (Alfieri’s repair), accompanied by PLV. The choice of this procedure for repairing MR makes sense from the aspect of the patient’s future growth because there are no prosthetics, such as an artificial ring. Regrettably, De Bonis et al reported that the edge-to-edge procedure could not prevent recurrence of MR, which occurred with progression of LV redilatation. It is now commonly considered that both procedures, PLV and edge-to-edge mitral repair, should be avoided in adults, from the perspective of poor durability. Alternative surgical procedures might be introduced into the pediatric clinical arena just as was done for adults, including overlapping ventriculoplasty (OLVP), which restores the left ventricle without any patches, and papillary muscle approximation (PMA), which gathers both papillary muscles together, correcting the tethering of the mitral leaflets.

After OLVP was introduced for the treatment of idiopathic DCM in adults, we integrated PMA and papillary muscle suspension (PMS) incrementally into the procedure. These additional techniques are attempted for reducing the mitral valve tethering and perpetuating the correct geometry of the valve and apparatus. The tethering phenomenon, which occurs as a result of displacement of the papillary muscles, plays the main role in FMR, whereas annular dilatation plays only a minor part. Hence, we think that OLVP with simple PMA not accompanied by PMS or ring annuloplasty would do well for geometric reconstruction in small children with growth potential.

The opinions expressed in this article are not necessarily those of the editors or of the Japanese Circulation Society.

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References

3. The Declaration of Istanbul on Organ Trafficking and Transplant Tourism. Participants in the International Summit on Transplant

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