SURGERY FOR CONGENITAL HEART DISEASE

EDITORIAL: PARTIAL LEFT VENTRICULECTOMY FOR DILATED CARDIOMYOPATHY IN CHILDREN

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Partial left ventriculectomy or ventricular volume reduction surgery to treat end-stage heart failure has gained much attention because of the potential for obviating or delaying the need for heart transplantation. This procedure has been advocated for patients who are not considered candidates for transplantation or in centers where transplantation is not readily available. Since its early description by Batista and associates[1] in 1996, what has now come to be called the "Batista" operation has been applied by many centers throughout the world and in patients with widely different causes for heart failure.[2,3] The common pathophysiology in these patients is a dilated cardiomyopathy, and the indications for the procedure are based on the concept that left ventricular (LV) dilatation without compensatory increase in LV wall thickness has led to a significant increase in LV wall stress. The elevated LV wall stress results in a mechanical load that the ventricle cannot overcome, leading to further dilatation. In other terms, the LV muscle mass/LV cavity volume relationship has been significantly reduced by LV dilatation. It is therefore reasonable to infer that a reduction in LV cavity volume will result in lower wall stress with decreased mechanical load and improved stroke work/end-diastolic volume (preload recruitable work).[4] Furthermore, if partial ventriculectomy is combined with another procedure to reduce end-diastolic pressure, such as mitral valve repair/replacement[2] or elimination of left-to-right shunts (personal experience), then a sustained improvement in LV dynamics may be achieved. Central to this hypothesis is the assumption that the LV muscle is intrinsically normal or has recoverable contractile function, such that the decrease in wall stress will result in improved contractility or a parallel leftward shift in the stroke work/end-diastolic volume relationship. Perhaps this is the one variable that is least controlled for in the numerous reports now in the literature describing results with adults receiving partial ventriculectomy for dilated cardiomyopathy (DCM).

In children, one additional variable needs to be considered: DCM does not progress in a significant proportion of infants and children, and some undergo spontaneous recovery. Indeed, spontaneous recovery of LV function in as high as 45% of cohorts has been noted.[5] Several reports have addressed the problem of predicting outcome in children with DCM. Most reports with larger series (>20 patients) have found that older age at presentation and the presence of fibroelastosis are strong predictors of poor outcome.[6,7] Other reported risk factors include the presence of a mural thrombus and a spherical shape to the LV.[5] Of the children who do recover, most have definite improvement in LV function within 3 months. However, those with progressive dilatation or ventricular arrhythmias are at high risk for early death.

Two case reports published in this Journal describe the results with partial left ventriculectomy in children with severe LV dysfunction and dilatation. Berger and associates[8] report a case of a 3-day-old infant with unexplained LV dysfunction and severe mitral regurgitation in whom systemic perfusion was maintained by the right ventricle via the patent arterial duct. Partial left ventriculectomy and mitral valve repair were associated with gradual recovery at 8 months' follow-up. Histologic examination of the excised tissue was reported as not showing a specific abnormality. In a previous report, Yoshii and colleagues[9] described a 5-month-old infant with LV dysfunction and dilatation progressing over a 2-month period, associated with severe mitral regurgitation, endocardial fibroelastosis, and endomyocardial biopsy with no abnormality. This infant also underwent partial ventriculectomy with mitral valve repair, and histologic examination showed endomyocardial fibrous thickening. Improved LV function was noted at 1 month's follow-up.

These reports illustrate some of the problems sur-

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geons face in deciding treatment of children with DCM. In the first case, although LV function was severely depressed, determining the prognosis was difficult because of the scarcity of experience in managing neonates with DCM. Whether continued inotropic support and prostaglandins to maintain ductal patency would have resulted in a similar outcome is not known. Mitral insufficiency, although severe, may also have recovered along with LV function, as is frequently seen in infants after repair of anomalous origin of the left coronary artery from the pulmonary artery. Unfortunately, because of the lack of experience with neonatal DCM, each surgeon is left to decide management on the basis of available resources and personal experience.

In the second case, the progression of LV dilatation, despite treatment with vasodilators and inotropic agents and evidence of endocardial fibroelastosis, predicted a very poor prognosis. The early favorable results with ventriculectomy and mitral valve repair are encouraging. However, early redilatation of the LV may still be seen in this patient, as has been reported in larger series of adult patients.

Because of the low incidence of DCM in children and the heterogeneous approach to their management among many centers, it is unlikely that any one pediatric center will accumulate sufficient experience with partial ventriculectomy to determine which patients are the best candidates. Unless a coordinated multicenter effort is made to study patients prospectively, we will need to rely on the growing experience with this procedure in adult patients to guide decisions about patient selection and operative technique. For now, each center will need to evaluate potential candidates individually on the basis of carefully assessed and known prognostic factors in children and the availability and results of alternative treatments such as transplantation.

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