Commentary: A difficult group of patients

Emile Bacha, MD, FACS

The group from Boston Children’s has taken on to characterize and study and analyze a very difficult group of patients, namely those with residual or recurrent left atrioventricular valve (LAVV) lesions after atrioventricular canal defect (AVCD) repairs.1 These patients can often present a formidable surgical challenge, as evidenced by a rate of reintervention of one-third at a fairly short median follow-up time of 18 months. What other congenital heart operations have a one-third reintervention rate at early follow-up? None that I can think of. And what would the reintervention rate be at 5 years? Of course, in young infants and toddlers, a mitral valve replacement is equally fraught with problems and implies a certain reintervention to upsize the prosthesis. Therefore, a good repair is truly something for which to aim.

The anatomy of the LAVV in the setting of complex AVCD (often non-Downs, often partial AVCD) can be bewildering sometimes. The LAVV’s only opening can be the cleft itself, with a valve devoid of any commissural opening. Tissues can be thin and poorly developed. The annulus is often hypoplastic, and the left ventricular inflow can be crowded by a large coronary sinus connected to a left superior vena cava. The subvalvar apparatus often is characterized by a single papillary muscle, or short and dysplastic chordae that are disordered and misaligned. Thus, the findings are not surprising, but it is good to have a clear idea of current outcomes obtained under best conditions. Clearly, as a community, we still have a lot of work to do to serve these patients better. Compounding this sobering finding is the fact that these outcomes were obtained in a setting with deep surgical expertise and a large number of patients to study, implying experience with this particular lesion. A positive volume/outcomes relationship in pediatric cardiac surgery is especially strong for rare and difficult lesions, and this is definitely one. Therefore, to me, the principal value of the study is to “set the standard” for outcomes, so to speak, for this particular group of patients. The one exception is, as documented in this study, a patient whose cleft repair sutures have torn and requires “just” a cleft reclosure. Indeed, a residual cleft as the primary mechanism of LAVV regurgitation was protective in univariate analysis.

This study also suggests that those patients with a post-cardiopulmonary bypass transesophageal echocardiogram gradient of $\geq 5$ mm Hg with mild or great LAVV regurgitation represents a particularly high-risk group, with approximately 75% of patients needing redo LAVV intervention (meaning a third surgery). While I can’t argue with these findings, I have to caution that the gradient of 5 mm Hg does not jive with my clinical experience. We do aim for as little regurgitation as possible, and any more than mild $+$ regurgitation is certainly seen as a risk factor. However, we also aim for a “tight” valve,2 and it would be difficult to act upon a gradient of 5 or slightly greater.

References