The left atrioventricular valve: The Achilles' heel of incomplete endocardial cushion defects

Meena Nathan, MD, MPH

From the Department of Cardiac Surgery, Boston Children's Hospital, Boston, Mass; and Department of Surgery, Harvard Medical School, Boston, Mass.

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Address for reprints: Meena Nathan, MD, MPH, Department of Cardiac Surgery, Bader 273, 300 Longwood Ave, Boston, MA 02115 (E-mail: meena.nathan@cardio.chboston.org).

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Residual cleft: a common indication for reoperation.

Central Message

Despite excellent early outcomes, repair of incomplete endocardial cushion defects in infancy is associated with higher LAVV and LVOT reintervention.

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Incomplete forms of endocardial cushion defects include partial and transitional atrioventricular septal defects (AVSDs). These defects are characterized by 2 distinct atrioventricular valve orifices, unlike the complete atrioventricular canal defect that is characterized by a single/common atrioventricular valve orifice.^{1,2} In the current era, mortality after repair of partial and transitional AVSD is low (0.3% and 0.9%, respectively)²; however, despite modern advances in imaging and operative techniques, approximately 10% to 15% of these patients undergo unplanned reinterventions, particularly for left atrioventricular valve (LAVV) regurgitation or left ventricular outflow tract (LVOT) obstruction.^{1,3-10}

In this issue of the *Journal*, Mery and colleagues¹¹ report on outcomes of 265 patients with partial (67%) or transitional AVSDs over a 22-year period. In one of the largest single-center reviews to date, they report low mortality (98% survival at 10 years) and 15% and 19% reintervention rates at 5 years and 10 years, respectively. Although this was a retrospective review, they have reasonable follow-up having reached out to families and care providers for information. They have demonstrated that younger age (repair in the first year of life) and more complex anatomy are associated with reoperations, with LAVV reinterventions and LVOT reinterventions being the most common. As expected from a retrospective review, data on preoperative anatomy and physiology were based on reports and not confirmed by review of echocardiograms in a core laboratory; thus, the authors were unable to create predictive models for reinterventions.

There is conflicting information about the association between age of repair and interventions on the LAVV or LVOT.³⁻⁷ In several studies, younger age is an important risk factor in reintervention.³⁻⁵ In contrast, the Pediatric Heart Network Investigators^{6,7} demonstrated a prevalence of moderate or greater LAVV regurgitation more common in children who were aged more than 4 years at partial/transitional AVSD repair. Despite such conflicting data, it

makes intuitive sense that repair in infancy preselects a higher-risk cohort who require more urgent repair (of what is essentially an atrial level shunt and is usually well tolerated) and are more likely to have significant abnormalities not only of the LAVV but also of other left-sided structures. This increases their risk for reoperations on the LAVV or LVOT.

Not surprisingly, preoperative LAVV regurgitation was the single most important predictor for LAVV reinterventions in partial and transitional AVSD.^{4,5,8,9} As expected, severity of LAVV regurgitation at discharge predicted reintervention on the LAVV.¹⁰ The cause of LAVV regurgitation can be attributed to abnormal morphology of the LAVV encompassing abnormal leaflets, atypical clefts, and abnormal subvalvular apparatus that results in leaflet prolapse or restriction. At primary repair, every effort should be made to address these abnormalities and ensure adequate tension-free coaptation of the LAVV leaflets.¹²

As nicely illustrated by Mery and colleagues,¹¹ younger age at repair is associated with higher rates of left-sided reintervention. Preoperative LAVV morphology and function remain the most important markers for successful repair and thus predictors of postdischarge reintervention on the atrioventricular valve.

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