Pediatric Cardiac Surgery Annual

Reoperation for Left Ventricular Outflow Tract Obstruction After Repair of Atrioventricular Septal

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Left ventricular outflow tract obstruction (LVOTO) is an important source of morbidity and mortality after repair of atrioventricular septal defect (AVSD). The intrinsic anatomy of the left ventricular outflow tract in AVSD is complex and predisposes to the development of LVOTO. LVOTO after repair of AVSD usually involves multiple levels and sources of obstruction, and surgical intervention must address each component of the obstruction. This includes fibromuscular obstruction, septal hypertrophy, and valve related sources of obstruction. Special attention is also directed to the anterolateral muscle bundle of the left ventricle, a well defined but under recognized feature of the left ventricular outflow tract in AVSD. It is present in all patients with AVSD, and resection of a hypertrophic anterolateral muscle bundle of the left ventricle should be incorporated in all operations for LVOTO after repair of AVSD. LVOTO after repair of AVSD has several unique features that must be taken into consideration to maximize outcome after surgical intervention. These include anatomic factors, technical aspects of surgical intervention, and proper selection of the operation used for relief of LVOTO.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 17:43-47

Introduction

Left ventricular outflow tract obstruction (LVOTO) is a well-described sequelae of repair of atrioventricular septal defect (AVSD). Indeed, a narrow left ventricular outflow tract (LVOT) has long been recognized as an intrinsic feature of hearts with AVSD.[1] LVOTO is second only to left atrioventricular (AV) valve regurgitation as an indication for reoperation after repair of AVSD. The reported incidence of LVOTO requiring reintervention after primary repair of AVSD varies from 0.5% to 4.5%.[2–7] Duration of follow-up is an important determinant of the observed incidence, as reoperation for LVOTO generally occurs at a mean interval of 5 to 7 years after primary repair.[2,8–10] The need for reoperation for LVOTO after AVSD repair has a significant impact on late survival (Fig. 1). LVOTO after repair of AVSD has several unique features that must be taken into consideration to maximize outcome after surgical intervention. These include anatomic factors, technical aspects of surgical intervention, and proper selection of the operation used for relief of LVOTO.

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Morphology of the LVOT in AVSD

The anatomy of the LVOT in AVSD has been extensively detailed.[11–14] Its main features are "unwedging" of the aortic root (anterior and rightward), disparate inlet and outlet septum lengths (short inlet, long outlet), and deficiency of the muscular interventricular septum (septal "scoop") Figs. 2 and 3).[15,16] The normal LVOT is quite short, measuring only a few millimeters. The aorta sits "wedged" between the mitral and tricuspid valves. In AVSD, the aortic valve is anterior and rightward, no longer positioned between the two normally formed AV valves. Vacating of the distal LVOT by the aortic root collapses the anteroposterior dimension of the LVOT. In addition, the unwedged position of the aortic root creates length at the distal end of the LVOT that is not present in normal hearts and results in discrepant inlet and outlet septal lengths. The muscular septal deficiency underlying the AV valves causes the leaflets to form a convexity toward the ventricular apex rather than toward the atria, as is normal, and this convexity measurably narrows the LVOT. These various intrinsic anatomic abnormalities combine to create a narrow and elongated LVOT with an abnormal outlet angle, classically described angiographically as the "goose neck" deformity.

LVOT geometry in AVSD is also significantly impacted by the anatomy of the AV valve and the subvalvar apparatus. Specifically, the superior bridging leaflet may be closely applied

http://dx.doi.org/10.1053/j.pcsu.2014.01.008

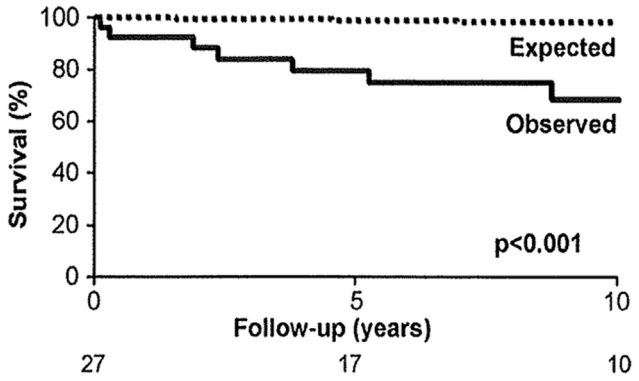

Figure 1 Actuarial survival after reoperation for LVOTO after repair of AVSD. A significant difference ($P < .001$) is observed as compare with age and gender matched population.

to the "scooped out" interventricular septum (as in some hearts with Rastelli Type A common AV valve configuration), or free floating (Rastelli Type C common AV valve). When closely adherent, the superior leaflet and associated primary and anomalous chordae may significantly crowd the distal LVOT. The papillary muscles of the left AV valve are rotated counterclockwise and are more closely spaced in AVSD compared with the normal heart. The anterolateral papillary muscle may have an abnormally high insertion, creating proximal LVOT narrowing. While closely spaced or parachute type arrangements of the papillary muscles are typically the substrate of left ventricular *inflow* obstruction, their abnormal positioning in AVSD brings the subvalvar and chordal apparatus toward the "center" of the proximal LVOT, where they may be a source of outflow tract turbulence. Accessory papillary muscles are frequently present in AVSD, as are anomalous chordal insertions and accessory fibrous bands. Thus, valvar and

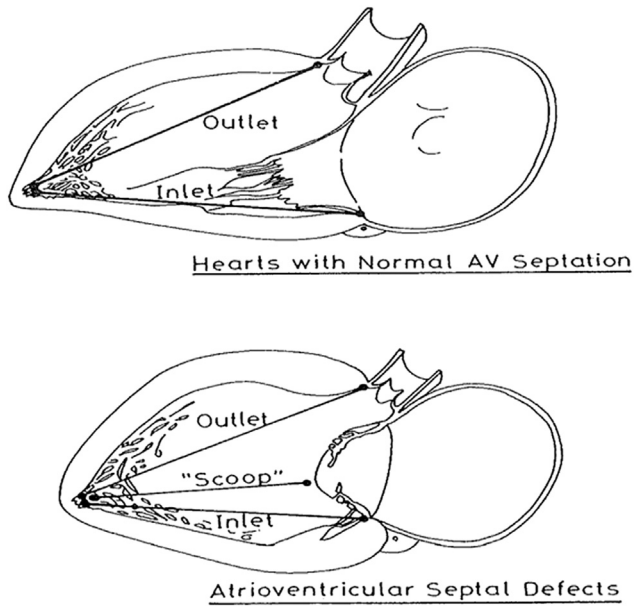

Figure 2 Illustration of measurements of the inlet and outlet septa from the left ventricular side in normal hearts (*top*) and hearts with atrioventricular septal defect (*bottom*).

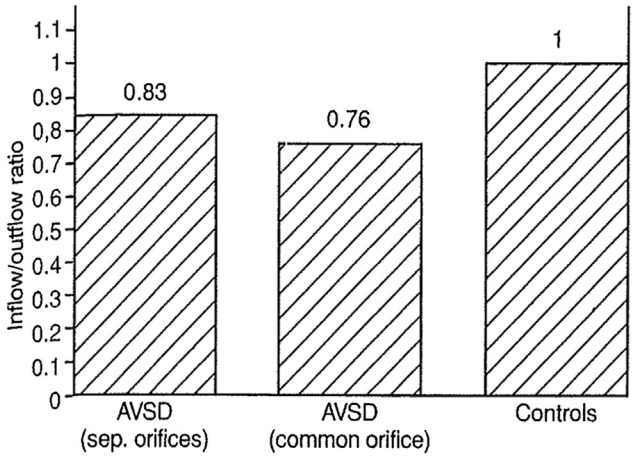

Figure 3 Ratios of the inlet and outlet septal dimensions in partial AVSD, complete AVSD, and in normal hearts. There is no significant difference in the ratios of partial and complete AVSD ($P > .05$). The difference is highly significant ($P < .01$) when hearts with AVSD (any form) are compared with normal hearts.

subvalvar abnormalities are critical features of LVOTO in AVSD (Fig. 5).[17]

Special Emphasis: Anterolateral Muscle Bundle of the Left Ventricle

This unfavorable geometry is further compromised by the known occurrence of encroachment on the LVOT by the anterolateral muscle bundle of the left ventricle (AML; Fig 4). In the surgical literature, the presence and importance of the AML in the setting of LVOTO after repair of AVSD has been largely unrecognized. The AML is a horizontal muscle bundle located between the left coronary cusp and the aortic leaflet of the mitral valve. Initially described by Moulaert and Oppenheimer-Decker,[18] it is present in roughly 40% of normal hearts. In a study of 77 hearts with AVSD, Draulans-Noe and Wenink[19] showed that the AML is present in 100% of AVSD specimens, and frequently bulged into the LVOT. In this morphometric analysis and in a corroborating echocardiographic and magnetic resonance imaging study, the authors noted that the AML alone did not create significant subaortic stenosis.[20] Nevertheless, operations for LVOTO that do not address encroachment by a hypertrophic AML almost certainly increase the risk of recurrence.

Predictors of LVOTO after AVSD Repair

Investigators have attempted to identify clinical and echocardiographic predictors of LVOTO after AVSD repair. Historically, patients without Trisomy 21 have been identified as being at increased risk for LVOTO, but more recent series refute this notion.[8,21] Likewise, evidence is mixed with regard to the impact of subtype of AVSD (partial, transitional, or complete) on the incidence of LVOTO after repair. Most recent series report similar reoperation rates amongst AVSD subtype.[3,6,8] Similarly, Rastelli Type A configuration of the common AV valve has been widely understood to predispose