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Partial Atrioventricular Canal: Pitfalls in Technique

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Partial atrioventricular (AV) canal represents approximately 25% of all AV septal defects. While often grouped with secundum ASD from the perspective of cardiopulmonary physiology, clinical presentation, and timing of surgical correction, their optimal management truly requires an understanding of their anatomic similarities to other forms of common AVC defects. By most measures, outcomes for surgical management of partial AV canal has improved over the last four decades, though some aspects of these defects continue to pose important challenges. Current experience has witnessed the reduction in early mortality and only rare complete heart block. Left AV valve dysfunction remains the most common indication for reoperation (10%) with LVOT stenosis the next most common reason (10% to 15% incidence, 5% to 10% reoperation rate). It is important to understand in this population that postoperative left AV valve problems and LVOT stenosis may be intimately linked, both from an etiologic standpoint, and with respect to their management. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 10:42-46 © 2007 Elsevier Inc. All rights reserved.

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Many view repair of partial atrioventricular (AV) canal in a similar level of complexity to that of secundum atrial septal defect (ASD), yet most certainly recognize the presence of subsets of these patients that can pose challenges during initial management or the need for challenging reintervention. Common atrioventricular canal defects (AVCs) were classified into three anatomic forms by Wakai and Edwards[1] in 1958. Partial AVC, also commonly referred to as primum ASD, is characterized by fibrous continuity of the superior and inferior leaflets of the common AV valve forming separate right and left sided valve orifices, a primum ASD, absence of a ventricular level communication, and typically a cleft in the anterior leaflet of the left AV valve. Partial AV canal represents approximately 25% of all AV septal defects. While often grouped with secundum ASD from the perspective of cardiopulmonary physiology, clinical presentation, and timing of surgical correction, their optimal management truly requires an understanding of their anatomic similarities to other forms of common AVC defects. Though lacking a ventricular level communication, partial AVC defects share the same anatomic features of the scooped-out ventricular septum, elongated left ventricular outflow tract (LVOT), and trifoliate left AV valve configuration characteristic of complete and transitional AVC defects.[2] The discrepancy in length of the ventricular septum, as measured in the left ventricle comparing the inlet to apex with the outlet to apex distance, has been well described.[2-4] Two factors contribute to this disparity: the "scooped-out" deficiency of the inlet septum resulting in a shorter inlet to apex dimension compared with normal (Fig. 1), and the rightward and anterior displacement of the aortic root (the so-called "unwedged position") resulting in a longer apex to outlet dimension compared with normal (Fig. 2). Understanding this anatomy may be critical in the surgical management of two of the most common reasons for reintervention in this population: left AV valve dysfunction and LVOT stenosis. In addition, partial AVC, by virtue of its firm fixation of the bridging leaflets of the common AV valve (particularly superiorly) to the scooped out septum is often reported to have a higher incidence of AV valve problems and LVOT stenosis compared with other forms of common AVC.

By most measures, outcomes for surgical management of partial AV canal have improved over the last four decades. Although some aspects of these defects continue to pose important challenges.[5] In older series on surgical management of partial AVC, the important outcome parameters reported were mortality (10% to 20%), left AV valve regurgitation

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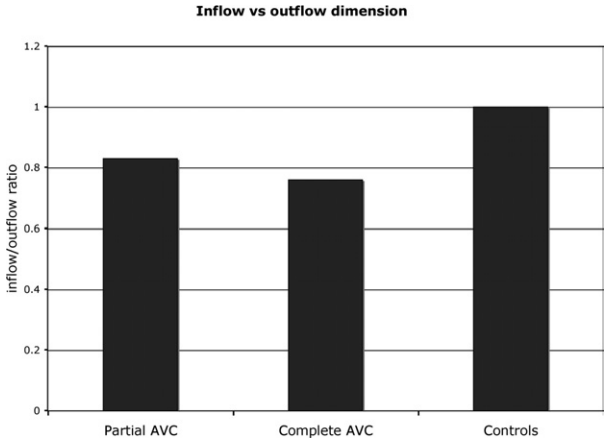

Figure 1 Comparison of left ventricular inflow versus outflow dimensions between AV canal defects and normal hearts. (Reprinted with permission.[4])

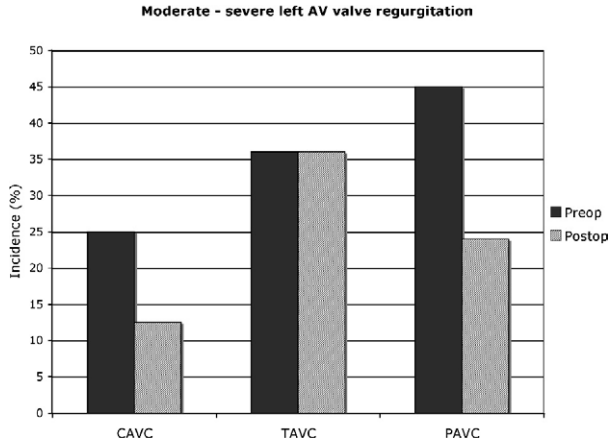

Figure 3 Incidence of preoperative and postoperative left AV valve regurgitation comparing different types of AV canal defect. CAVC, complete AV canal; TAVC, transitional AV canal; PAVC, partial AV canal. (Data from Michielon et al.[6])

(25% to 50% incidence; 10% to 20% reoperation rate), and arrhythmias (complete heart block in 1% to 10% of cases). Current experience has witnessed the reduction in early mortality to less than 3% and only rare complete heart block. Left AV valve dysfunction remains the most common indication for reoperation (10%), with LVOT stenosis the next most common reason (10% to 15% incidence; 5% to 10% reoperation rate). In this population it is important to understand that postoperative left AV valve problems and LVOT stenosis may be intimately linked, both from an etiologic standpoint and with respect to their management. While LVOT stenosis can certainly exacerbate the degree of left AV valve insufficiency even in the face of only mild valve deformity, the fixation of the superior portion of the anterior valve leaflet to the scooped-out septal crest, along with abnormalities in cordal and papillary muscle architecture, may be contributing causes of the LVOT obstruction itself.

In the current era, can we do better to prevent the need for reoperation in this patient population, and when reoperation is necessary, can we manage the reoperation better to avoid recurrence of the problem?

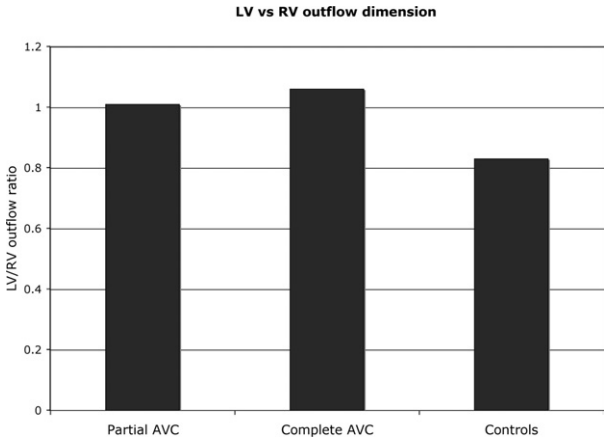

Figure 2 Comparison of left ventricular and right ventricular outflow lengths between AV canal defects and normal hearts. (Reprinted with permission.[4])

AV Valve Regurgitation

The majority of patients with partial AVC defects exhibit a trifoliate left AV valve with a characteristic cleft extending a variable proportion of the distance between the valve's free edge and its attachment to the septum. While most agree that the cleft is a "normal" part of the valve architecture in AVC defects and that reconstruction does not ever re-create a replica of the anterior leaflet of a normally formed mitral valve, closure of the cleft is performed routinely by most surgeons, certainly when any insufficiency exists on preoperative imaging or at the time of intraoperative passive testing. Patients with partial AVC have been observed to have a higher incidence of preoperative left AV valve insufficiency than other forms of AVC (Fig. 3),[6] and the incidence of significant postoperative insufficiency and the risk for reintervention to address insufficiency have been found to correlate both with the degree of preoperative regurgitation and the amount of early postoperative regurgitation.[7] Risk for reintervention is seen as both an early and late postoperative issue, with the incidence of reoperation for residual AV valve insufficiency leveling off as long as 8 to 10 years following initial repair.[8] Highlighting the challenges in dealing with managing AV valve regurgitation is the high incidence of second re-operation for regurgitation, reported as high as 25%.[9] Younger age at initial repair has been found by some to be a risk factor for late AV valve regurgitation,[8] while others have found this complication to be less frequent in children operated on earlier.[6] Because annular dilation and thickening and rolling in of the leaflet edges along the cleft are likely contributors to AV valve regurgitation in this anomaly, and these are both likely to worsen with time, some degree of preoperative insufficiency may be acquired and may be prevented by earlier surgical repair. In the series that found worse regurgitation (pre- and post-operative) in patients with partial AV canal, it

is of interest that the average age at repair for partial AV canal was 10.7 years compared with 10 months and 6 months for transitional and complete AV canals, respectively.[6] Unusual cordal attachments from the leaflet to the septum, which may tether leaflet mobility and contribute to valve function as well as to potential LVOT stenosis, may be resected as long as they lend no support to the free edge of the leaflet.

Two anatomic variations warrant specific comment. Double orifice left AV valve and a "potentially parachute" valve caused by the fusion of the papillary muscles into a single body are reported in approximately 10% of cases. The double orifice valve may be unrecognized on preoperative studies. Careful intraoperative inspection of the valve typically reveals a small, secondary orifice in the postero-medial aspect of the valve supported only by a single papillary muscle, thus resulting in limited excursion during diastole. The major orifice is often associated with a cleft and may be supported by both papillary muscles. Division of the bridge of leaflet tissue between the two orifices must be avoided to prevent a flail central segment of the valve and severe insufficiency. Limited closure of the cleft associated with the major orifice may be considered with careful attention not to excessively narrow the inflow dimensions of the valve. The presence of a single left ventricular papillary muscle and thus the potential for a stenotic, parachute valve is easily demonstrated on short-axis echocardiogram views. Careful intraoperative inspection is necessary because some patients reported to have a single papillary muscle by preoperative study will be found to simply have hypoplasia of one papillary muscle with true support of the valve segregated to two separate zones in the LV. In cases where all cordae insert to a single LV papillary muscle, the cleft of the left AV valve represents the major orifice for diastolic flow and complete closure risks the creation of important postoperative stenosis. Partial closure of the base of the cleft is often possible to address insufficiency without creating significant obstruction to flow.

LVOT Stenosis

Autopsy and echocardiographic studies have shown that the characteristic anatomy of the ventricular septum, common AV valve, and its papillary muscle configuration leads to an "at risk" configuration for either inlet or outflow stenosis in as many as three quarters of AVC patients.[4,10,11] The actual incidence of clinically significant stenoses is much less (5% to 10%), but has been noted up to three times more frequently in patients with partial AVC compared with other forms.[10] Similar to what has been observed with AV valve regurgitation, the risk for reintervention for LVOT stenosis does not appear to decline even many years following initial operation,[8] and even more concerning is that the need for second reoperation for LVOT stenosis has been observed to be 35% to 45%.[12,13]

The narrowing and elongation of the LVOT that is characterized as the "gooseneck" deformity of AVC defects may be more pronounced in partial AVC than in complete forms because of the firm fixation of the valve leaflets to the scooped-out septum resulting in this deformity being evident not only in diastole but also through systole. As mentioned previously, many studies (autopsy and clinical) have shown the substrate for potential LVOT stenosis in the majority of patients with AV septal defects. In addition to the configuration of the ventricular septum already mentioned, abnormal positioning of papillary muscles or accessory attachment of cordae to the septum may contribute to this potential. In studies predating the routine use of echocardiograms for preoperative evaluation, LVOT gradients were typically not investigated during diagnostic catheterization resulting in under-recognition of this problem.[14] Despite the anatomic predisposition for development of LVOT stenosis, important gradients are seen in as few as 1% of patients at the time of presurgical evaluation. Development of LVOT stenosis in the postoperative period is noted in 3% to 10% of patients and is recognized across a spectrum of time intervals from months to many years following original surgical correction. Most series report resection of subaortic fibrous tissue with septal myomectomy as the primary technique used in addressing the stenosis, but a disturbingly high rate of recurrent stenosis is noted when this technique alone is used in this population, underscoring the complexity of the underlying anatomy contributing to the development of this complication.

As emphasized by Van Arsdell et al,[12] management of this complication must often incorporate a number of techniques. Resection of fibrous tissue that may represent accessory endocardial cushion tissue, anomalous cordal attachments, or reaction because of chronically turbulent flow through the narrowed and malaligned outflow tract is virtually always a component of the repair. Septal myomectomy is also recommended. Some have reported[15] use of a modified Konno technique to create a VSD with patch closure from the right side similar to what has been successfully reported in other forms of tunnel-like subaortic stenosis. Two techniques have been described that specifically address the scooped-out ventricular septum characteristic of AVC defects as well as the attachment of the valve leaflets to the crest. One involves detaching all bridging leaflet components of both AV valves from the ventricular septal crest, essentially re-creating an inlet VSD analogous to a Rastelli type C complete AVC defect, then patching the VSD with reattachment of the valves to the top of the patch.[16] The anterior aspect of this patch directly augments the dimension of the septum in the subaortic region. The rationale for this stems from the observation that LVOT stenosis is rare after repair of Rastelli type C complete AVC compared with type A, transitional, or partial AVC; all of which share the distinction of having attachments of valve to the superior septal crest and may be repaired primarily without significant augmentation of the superior portion of the ventricular septum. The second technique (Fig. 4) involves mobilization of the left-sided valve components only, either the superior portion, or the entire reconstructed septal leaflet.[17,18] Pericardial patch augmentation of the base of the leaflet similarly moves the left AV valve farther into the left atrium during ventricular systole, thus augmenting the dimension of the LVOT. Such leaflet augmentation may also be beneficial in addressing residual left AV valve insufficiency[9] by advancing the septal leaflet's free edge farther posteriorly

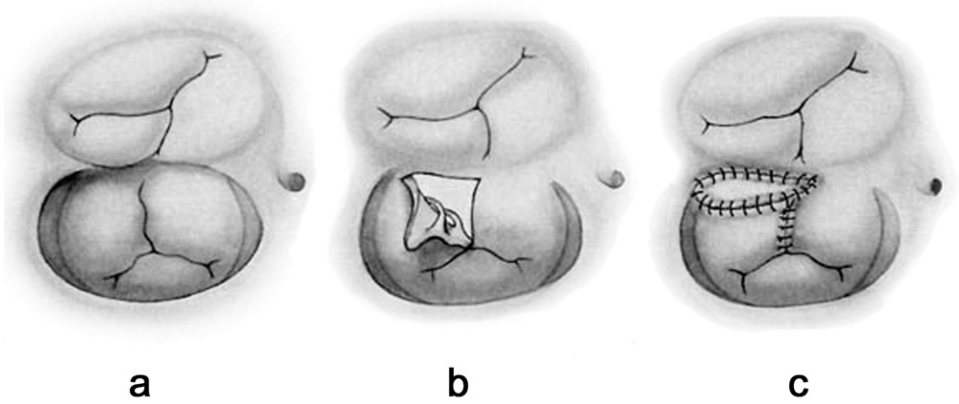

Figure 4 Technique of anterior leaflet augmentation for relief of LVOT stenosis in AV canal defects. a, partial AV canal defect; b, mobilization of the base of left superior bridging; c, pericardial patch augmentation of leaflet and closure of left AV valve cleft. (Reprinted with permission.[18])

allowing more surface for coaptation. Given that no alteration is made to the valve's free edge cordal support, central prolapse should not be seen with such a technique.

Multiple Left-Sided Obstructions

A subgroup of patients with partial AVC defects represents a particular challenge for management. Approximately 10% of children born with partial AVC defects will also exhibit hypoplasia of multiple left-sided cardiac structures including potentially obstructive anomalies of the left AV valve as described above, variable degrees of LV hypoplasia, LVOT stenosis, and aortic coarctation. Similar to patients with Shone's complex, they often require multiple reoperations, typically to address complex left AV valve or LVOT problems, and have been observed to have a dramatically different mortality risk compared with children without such associated findings, in the range of 20% to 30%.[19,20] While most children with uncomplicated partial AVC defects exhibit few symptoms early in life, following a course not unlike a child with a large secundum ASD, this subgroup exhibits the signature characteristic of significant symptoms of congestive heart failure (CHF) in the first months of life despite the complete absence of any ventricular level communication. Some have suggested following a single-ventricle reconstruction pathway in some of these patients, although this is unappealing in the face of AV valve insufficiency (which may commonly be present), and is difficult to justify in patient whose left side has been supporting adequate systemic circulation for a period of time and whose right ventricle may be deconditioned given the absence of a ventricular level communication.

Significant symptoms of CHF in an infant with partial AVC defect warrants careful evaluation for the presence of any left heart obstructive lesions, and management of these must be incorporated into the repair strategy in addition to closure of the septal defect. Utilization of some of the repair strategies described previously, including septal augmentation for LVOT stenosis, may be considered as part of the primary repair. When aortic coarctation is an associated lesion, consideration of single-stage complete repair of the arch and intracardiac anatomy should be given. Staged management with coarctation repair first in hopes of making a significant impact on the symptoms of CHF with arch repair alone is often unsuccessful. A higher incidence of aortic restenosis because of underlying arch hypoplasia has been reported, and significant symptoms of CHF will often persist in the face of other levels of left heart hypoplasia.

Summary

Patients with partial AV canal who present with symptoms very early in life should raise suspicion of complex anatomy, including multiple left-sided hypoplastic lesions, which may pose significant challenges to management and predict a worse prognosis.

Because some determinants of left AV valve regurgitation may be acquired rather than congenital, even in patients with AV canal defects (such as thickening and rolling of leaflet edges and annular dilatation), earlier repair of partial AV canal may result in a lower incidence of late insufficiency.

LVOT stenosis in this population is characterized by complex anatomy and simple membrane resection, and myomectomy alone should rarely be performed for its treatment. Superior valve leaflet augmentation may be a valuable adjunct in the surgical management of both LVOT stenosis and AV valve regurgitation.

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