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Technique of Single-Stage Repair of Coarctation of the Aorta With Ventricular Septal Defect

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The results of single-stage and two-stage repair of coarctation of the aorta (CoA) with ventricular septal defect (VSD) have improved, but the optimal treatment strategy remains controversial. This article emphasizes the technical details for performing the single-stage repair of CoA with VSD and compares the results of this technique with the two-stage approach. A retrospective analysis of 46 patients who underwent completed surgical repair of CoA with VSD at Children's Hospital of Michigan, either using the single-stage (N = 23) or the two-stage (N = 23) techniques, was performed. The postoperative complications, hospital mortality, freedom from cardiac re-interventions, and actuarial survival were the same in both groups. The advantages of single-stage over two-stage repair include an earlier age at completion of repair, fewer operations, and fewer incisions. The one disadvantage of a single-stage repair was the increased need for delayed sternal closure compared with the two-stage approach, but this disadvantage has been neutralized in the recent era.

KEYWORDS Single-stage repair, Coarctation, Ventricular septal defect, Aorta, Two-stage Repair

Anatomy

The decision-making process for determining the optimal surgical strategy to address the neonate and infant with coarctation of the aorta-ventricular septal defect (CoA-VSD) includes a detailed knowledge of the patient's clinical condition, the anatomy of the entire aortic arch, and the identification of all important associated intracardiac defects. This pertinent anatomic information can usually be provided by echocardiography alone. Rarely, however, it is necessary to supplement echocardiographic data with cardiac catheterization/cineangiocardiography and/or computerized cross-sectional imaging. Despite the availability of this sophisticated diagnostic data, the management of CoA-VSD remains controversial[1] because 1) the ability to predict spontaneous closure or significant regression of the VSD is imperfect, 2) the determination of clinically significant proximal aortic arch hypoplasia is often subjective, and 3) because little is known about the comparative advantages of the single versus the two-stage approaches.

Spontaneous VSD Closure

Critical to surgical decision-making is the observation that when all patients, regardless of the size of the VSD, who present during a given time interval with CoA-VSD are analyzed, a significant percentage will experience spontaneous closure of perimembranous and muscular VSDs after repair of the CoA alone with or without pulmonary artery (PA) banding. Posterior malalignment and subarterial VSDs will never close spontaneously and large perimembranous VSDs, especially those with inlet and/or outlet extension, will rarely close spontaneously. In Brouwer's series,[1] for example, 58% of all patients who underwent CoA repair ± PA banding experienced spontaneous closure of the VSD. Although he-

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modynamic[1] and morphologic[1,2] predictors of which VSDs are likely to close spontaneously have been proposed, our knowledge of this aspect of the natural history of CoA-VSD remains imperfect. Furthermore, accurate estimates of VSD size by measuring VSD gradients and shunts are confounded by the frequent presence of a large patent ductus arteriosus (PDA) and by the aortic arch obstruction itself.

Aortic Arch Anatomy

Important transverse aortic arch hypoplasia coexists in as many as 65% of neonatal patients with CoA in recent series.[1,3,4] Associated intracardiac defects, such as VSD and various forms of left ventricular outflow tract obstruction, probably result in an even higher incidence of associated aortic arch hypoplasia. This assertion is supported by the hemodynamic flow theory that postulates a paucity of aortic blood flow during fetal development as an etiologic agent for the development of the CoA and the frequently associated aortic arch hypoplasia.[4–11] The proximal extent of clinically significant aortic arch hypoplasia will also have an important impact on the choice of the surgical strategy because resection with extended end-to-end anastomosis repair of CoA through a left thoracotomy incision cannot always address arch hypoplasia that is proximal to the left common carotid artery. Similarly, the presence of proximal arch hypoplasia in conjunction with a common trunk for the right-sided innominate artery and the left common carotid artery can present technical difficulties for coarctation repair through a left thoracotomy incision (Fig. 1).

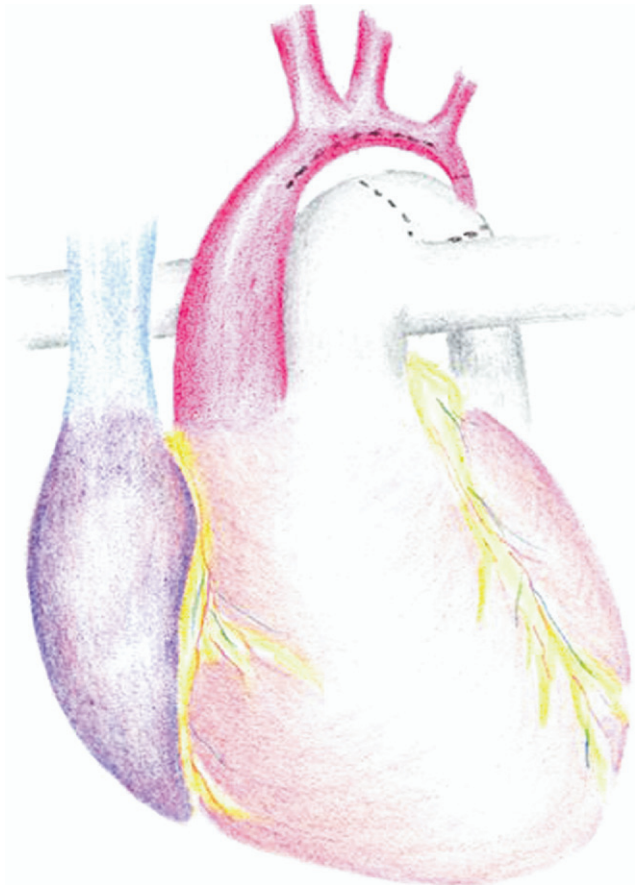

Figure 1 CoA with VSD is frequently associated with significant hypoplasia of the entire transverse aortic arch, which makes complete aortic reconstruction through a lateral thoracotomy incision difficult or impossible. This common situation is depicted in the illustration along with the proposed sites for ductal division, for distal transverse aortic arch ligation/division, and for the distal ascending/transverse aortotomy incision.

Categorization of Surgical Strategies

Three broad surgical strategies for the repair of patients who present with CoA-VSD are described in the literature. These strategies can be categorized as 1) complete single-stage repair, 2) committed two-stage repair, and 3) uncommitted two-stage repair.

The complete single-stage strategy is the most complex and comprehensive. It involves repairing both the coarctation and the VSD in one operation through a single median sternotomy incision always using cardiopulmonary bypass (CPB) ± deep hypothermic circulatory arrest (DHCA) ± isolated cerebral perfusion (ICP) ± isolated myocardial perfusion.[1,12–17] The committed two-stage approach is "committed" to the use of at least two operations (and usually two incisions) and is somewhat less complex than the single-stage approach. It consists of coarctation repair with PA banding (usually through a left thoracotomy incision without CPB) followed by interval PA debanding ± VSD closure through a median sternotomy incision using CPB.[18] The uncommitted two-stage strategy is the most optimistic of these three surgical categories for the repair of CoA-VSD in that it relies heavily, for its optimal outcome, on the tendency for some VSDs to close spontaneously. This approach is, therefore, not always "committed" to the performance of two surgical procedures but involves cooperation with nature to accomplish the complete (or near-complete) restoration of normal anatomy and physiology. The uncommitted two-stage strategy can be divided into two types. Type I consists of coarctation repair alone (usually through a left thoracotomy incision without CPB) and expectant management of the VSD. Interval closure of the VSD, through a median sternotomy incision with CPB, is later performed only if the patient still has a clinically significant VSD.[19] Type II has only been reported once and consists of coarctation repair with PA banding using an absorbable banding material followed by interval VSD closure, only if necessary.[20]

CoA-VSD With a Restrictive VSD

With this background, a reasonable and relatively non-controversial approach in well-compensated patients with CoA and an unequivocally small, restrictive, and clinically insignificant perimembranous or muscular VSD is a Type I uncommitted two-stage strategy consisting of repair of the CoA alone, through a left thoracotomy incision, followed by expectant management of the VSD.[1,19] In this situation there is usually no compelling reason to consider simultaneous repair of the VSD or placement of a PA band. One exception to this generalization is the coexistence of severe, proximal aor-