Editorial Comment

Coarctation of the Aorta and Ventricular Septal Defect in Infancy: Left Ventricular Volume and Management Issues*

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Combined coarctation of the aorta and ventricular septal defect. The association of coarctation of the aorta and ventricular septal defect resulting in congestive heart failure in early infancy is not uncommon. The underlying hemodynamic derangements resulting from this combination of lesions depend on a multitude of factors: 1) extent and severity of obstruction of the coarcted segment; 2) size and anatomy of the ventricular septal defect or defects; 3) presence of other left-sided obstructive disease (aortic valve, subaortic, mitral valve); and 4) patency of fetal channels (foramen ovale, ductus arteriosus).

Coarctation of the aorta with ventricular septal defect associated with a patent ductus has been described as "preductal" or "postductal." These terms cannot be taken literally in an anatomic sense because the coarctation is almost invariably in a "juxtaductal" position. However, they can be useful in describing the physiologic extremes of this combination of lesions (1). When severe isthmal aortic hypoplasia is part of the coarctation complex, the ductus arteriosus is oriented toward shunting blood from the pulmonary artery to the descending aorta. Under these circumstances of a physiologic "preductal coarctation" of the aorta, the right ventricle supplies blood to the lower body through the patent ductus arteriosus while also pumping a large volume of blood to the lungs at systemic pressure. In order for adequate blood to reach the lower body, the pulmonary circulation must reach maximal capacitance, and some degree of pulmonary vasconstriction also is present. The right ventricle must cope with a large preload and afterload at a systemic level. The left ventricle is often small; indeed, the preductal coarctation syndrome has been classified under the broad category of hypoplastic left heart syndrome.

In striking contrast, the infant with a "postductal coarctation" has a left ventricle that is responsible for pumping blood to the lower body across the coarctation at an increased afterload, and may also have the burden of increased preload because of the ventricular septal defect. A patent ductus arteriosus may add to the left to right shunt. Many infants with coarctation of the aorta and ventricular septal defect have hemodynamic characteristics that cannot be classified at either extreme, so that left ventricular chamber size and functional requirements may vary a great deal within the spectrum of this combination of lesions. However, because the left ventricle must supply at least the cardiac output required for the upper body proximal to the coarctation, it cannot be truly hypoplastic.

The present study. The study by Graham et al. (2) in this issue of the Journal has documented the absence of left ventricular volume loading in most infants with coarctation of the aorta and large ventricular septal defect apparently regardless of the patency or physiologic flow pattern of the ductus arteriosus. Preoperative left ventricular volume was not different in the 25 infants with the combined lesions as compared with that in 19 babies with isolated coarctation of the aorta. However, despite the absence of increased left ventricular volume in these patients with a large ventricular septal defect, the pulmonary to systemic blood flow ratio was extremely high. The authors could not demonstrate a statistically significant atrial or ductal left to right shunt to account for this. However, the large right ventricular volume in the eight patients studied, as well as increased pulmonary blood flow, can only be explained by the presence of a large interatrial left to right shunt.

Graham et al. (2) conclude that decreased left ventricular distensibility accounts for the absence of left ventricular volume loading. Left ventricular dysfunction would not be surprising among such infants, who have severe hemodynamic aberrations with possible superimposition of left ventricular myocardial hypoxemia during neonatal distress. Decreased diastolic compliance could explain the decreased left ventricular volume found in patients without a patent ductus or those with postductal coarctation. Otherwise, left to right shunts across the ventricular septal defect might be expected to be significant. Of course, decreased left ventricular volume would be the usual finding in the "preductal coarctation" syndrome.

It is well documented that left ventricular dysfunction following repair of coarctation of the aorta in infancy is reversible. In this study (2) it was demonstrated that in patients with a prior low left ventricular ejection fraction this variable normalized at a second study and, for the most part,
left ventricular end-diastolic volume increased. The latter finding cannot be clearly interpreted because most of these patients presumably had pulmonary artery banding at the time of coarctation repair (3). Because one purpose of the banding procedure is to reduce left ventricular preload, the increase that occurred is difficult to interpret. It may mean that pulmonary artery banding simply did not have significant effects on patients in whom left ventricular preload was not increased. An alternative explanation might be that after coarctation repair and normalization of left ventricular function, left ventricular volume would have increased still further without banding, preventing eventual increased left to right shunting at the ventricular level. The matter is further complicated by variability in the degree of constriction at the banding site, anywhere along a spectrum from inadequate to severely obstructive. Volume relations between the ventricles would depend on the initial or progressive rate, or both, of right ventricular outflow obstruction.

Surgical management of the associated lesions in infancy. Graham et al. (2) have added to our knowledge of the hemodynamics of coarctation of the aorta and ventricular septal defect. They have documented on a quantitative basis, using accepted methodology, the lack of volume loading of the left ventricle for the great majority of patients with this association of lesions even in the absence of the “preductal coarctation” syndrome. Left ventricular distensibility is decreased. Left ventricular contractile function may be depressed, and one might postulate that there may also be effects of mild (perhaps later to be significant) associated left ventricular abnormalities (e.g., aortic stenosis, mitral valve abnormality, subaortic obstruction). In some cases, the “large” ventricular septal defect might nevertheless be restrictive.

The findings in this study support an established clinical principle in the management of the vast majority of infants with coarctation of the aorta and ventricular septal defect: coarctation of the aorta is the dominant lesion and must be repaired. Whether also to surgically palliate the effects of the ventricular septal defect remains controversial. If pulmonary pressure remains elevated at the completion of the anastomosis, many surgeons will proceed with pulmonary artery banding (3,4). Other centers (5,6) advocate repair of the coarctation of the aorta without banding, even in the presence of a significant ventricular septal defect. Subsequently, if the patient cannot be managed medically because of a large left to right shunt, open heart repair of the septal defect is carried out. However, more often the septal defect may become smaller or close spontaneously and a second operation may never be needed; in some cases the defect is repaired electively later in childhood. Banding the pulmonary artery at the initial operation is reserved for infants with multiple defects, large apical muscular communications, a single ventricle or other complex lesions. In such instances early surgical elimination of the intraventricular left to right shunt would not be feasible and a persistent large left to right shunt would be expected. The long-term palliated status is preferable; more definitive surgery may be done later.

Implications. The findings from this study, indicating the absence of increased left ventricular volume in infants with coarctation and isolated ventricular septal defect, suggest that the ventricular left to right shunt is not an important cause of the congestive heart failure among the great majority of these patients. This may be why the pulmonary artery banding procedure most often is not necessary; dramatic improvement is noted after repair of the coarctation alone. Nevertheless, some groups continue to advocate pulmonary artery banding. On the basis of this report, preoperative and postoperative hemodynamic data collection in infants undergoing and in those not undergoing pulmonary artery banding should include ventricular volume studies. As a result, it may be possible to formulate specific preoperative criteria that will predict, prospectively, which if any infant with coarctation and isolated ventricular septal defect might benefit from pulmonary artery banding at the time of coarctation repair.

References


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