Coarctation of the Aorta and Ventricular Septal Defect

Stephen N. Morris, M.D.*

CASE SUMMARY

The patient was a 27-year-old Negro woman who was admitted to the hospital for cardiac evaluation. A cardiac murmur had been discovered at the time of her first pregnancy four and one-half years earlier, and it was thought to represent a ventricular septal defect. The patient had not had orthopnea, paroxysmal nocturnal dyspnea, cyanosis or chest pain. She was able to do her own housework and was not limited in her physical activities. It is of note, however, that she had also been hypertensive since her first pregnancy, and had transient ankle edema at that time and again with her second pregnancy three and one-half years ago. Since then, she had received medication with adequate control of her blood pressure. There was no history of rheumatic fever.

Physical examination revealed a well-developed, slender Negro woman who was comfortable at rest. Pulse rate was 90 per minute; blood pressure was 162/90 in the left arm and 160/84 in the right arm. Funduscopic examination disclosed slight silver wiring and arteriolar narrowing. There was no jugular venous distention. Her chest was clear to percussion and auscultation. Her PMI was in the fifth intercostal space in the midclavicular line. There was a systolic thrill along the left sternal border at the third and fourth intercostal space. A grade IV/V holosystolic murmur was heard in this area. It radiated widely and was heard in the back as well. The second heart sound split widely but closed with expiration. The liver was not palpable. There was no cyanosis, clubbing or edema of her extremities. Femoral, popliteal, dorsalis pedis, and posterior tibial pulses could not be palpated. Clinical impression was that of congenital heart disease with a ventricular septal defect and possibly with associated coarctation of the aorta.

Results of laboratory tests included normal CBC, urinalysis, VDRL, prothrombin time, platelet count, BUN, serum creatinine and electrolytes. Her electrocardiogram also was normal. Cardiac fluoroscopy revealed some left ventricular enlargement. Chest roentgenogram showed some pulmonary venous distention, particularly in the upper lung fields, but no findings suggestive of coarctation of the aorta could be identified.

Right and left heart catheterization revealed normal right atrial, right ventricular and pulmonary artery pressures. Mean pulmonary capillary wedge pressure was 8 mm Hg. There was no pressure gradient found across the aortic valve. During simultaneous pressure recordings in the ascending and descending aorta, however, a pressure gradient of 64 to 72 mm Hg was noted across a coarctation of the aorta which was at the level of the take-off of the left subclavian artery. Left ventricular cineangiograms also revealed a ventricular septal defect with an associated aneurysm of the ventricular septum. Pulmonary vascular resistance was normal. The patient's cardiac output was 5.8 L/min with pulmonary flow of 9.4 L/min, giving a left-to-right ventricular shunt of 3.6 L/min.

How would you manage this patient and why?

Comments by

Robert C. Schlant, M.D.*

This patient presents the problem of an asymptomatic 27-year-old black mother of two who has a coarctation of the aorta with a 64-72 mm Hg pressure gradient in association with a ventricular septal defect producing a pulmonary-to-systemic flow ratio of 1.62 with normal pulmonary vascular resistances. The coarctation of the aorta, which is relatively common in association with ventricular septal defect in infancy1 produces an increased left ventricular pressure and a greater left-to-right shunt than would otherwise be present. The aneurysm of the ventricular septum described in this patient was most probably an aneurysm of the pars membranacea2 which is occasionally found in association with a ventricular septal defect. Interestingly, it has been suggested that such aneurysms can occasionally result from closure of a ventricular septal defect in that location.

In view of the patient's age and the degree of severity of the coarctation, I would recommend that the coarctation be repaired to avoid the later complications of coarctation including heart failure, aortic rupture, or infection. I would not recommend repair of the ventricular septal defect at the time of the coarctation repair, but rather would follow the patient medically. I would repeat the right heart catheterization approximately one year after repair of the coarctation. I would anticipate that the left-to-right shunt might decrease due to an anticipated decrease in left ventricular systolic pressure. Whether or not to repair the ventricular septal defect would depend upon the hemodynamic findings at that catheterization. If the left-to-right shunt decreases significantly with the pulmonary/systemic flow ratio less than 1.5, I would probably not recommend surgery since the probability of her developing significant pulmonary vascular resistance is quite low in view of her normal pulmonary artery pressure and pulmonary vascular resistances.

*Professor of Medicine and Director, Division of Cardiology, Department of Medicine, Emory University, Atlanta. Reprint requests: Dr. Schlant, 69 Butler Street, SE, Atlanta 30303

*Trainee in Cardiology, Department of Medicine, Indiana University School of Medicine, Indianapolis, Indiana.

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at age 27 during her first catheterization. I would not expect the ventricular defect to close spontaneously at her age after coarctation repair, though this could conceivably occur. On each visit I would emphasize to the patient the importance of taking prophylactic antibiotics prior to any dental or surgical procedure likely to be associated with transient bacteremia. Following the repair of her coarctation, she should be followed carefully especially for persistence of some degree of systemic hypertension despite relief of the coarctation and for recurrence of hypertension, which may recur several years later.

References

Comments by
Dwight C. McGoon, M.D.

This young mother presents the classic findings of coarctation of the aorta and small ventricular septal defect, and she has been entirely asymptomatic except for transient ankle edema during pregnancy. The surgical management of these two cardiovascular anomalies should be considered separately since, from a technical standpoint, a different surgical approach for the correction of each is advantageous. The ventricular septal defect is best approached through a median sternotomy, and the coarctation of the aorta through a left posterolateral thoracotomy.

Although some surgeons have reported combining correction of both defects at a single operation, the disadvantages of so doing outweigh the advantages except in unusual circumstances.

The coarctation of the aorta has resulted in significant hypertension and represents a distinct potential threat to her health and well being. Clearly, surgical correction of the coarctation on an elective basis is indicated. This would be approached through a left posterolateral thoracotomy in the bed of the nonresected fifth rib. We are not informed of the appearance of the coarctate segment on the angiocardiogram, but we may assume that it is not atypical and that, therefore, resection of the coarctate segment and end-to-end anastomosis can be anticipated. Similarly, reduction of her arterial pressure to or nearly to normal levels would be anticipated in the several days or first few weeks following operation.

The ventricular septal defect is small or "restrictive," causing no elevation in pulmonary arterial pressure, and no electrocardiographic abnormalities. However, the pulmonary flow is approximately 1.6 times systemic flow. One might anticipate that as left ventricular systolic pressure falls as a result of the repair of the coarctation of the aorta, the pulmonary-to-systemic flow ratio might also fall modestly. The decision whether or not to recommend operation for closure of the ventricular septal defect is admittedly borderline. If these relationships were encountered in childhood, my colleagues and I would definitely not recommend operation in view of a significant chance for spontaneous closure. In a patient of this age, this is probably less likely, yet the chief threat of this defect to the patient's health is the development of bacterial endocarditis, which carries a risk in a low order of magnitude. I would defer the decision about the ventricular septal defect until two years after repair of the coarctation, and perhaps would repeat catheterization at that time. Unless new evidence appeared, I would probably not advise operation for closure of the small ventricular septal defect.

*Surgical Service, Mayo Clinic, Rochester, Minnesota.
Reprint requests: Section of Publications, Mayo Clinic, Rochester, Minnesota 55901