CLINICAL PROBLEMS IN CARDIOPULMONARY DISEASE

Management of Ventricular Septal Defects in Adults

Clinical Evaluations by Alexander S. Nadas, M.D., and S. Gilbert Blount, Jr., M.D.

Case Summary

Michael J. Mirro, M.D.*

The patient is a 20-year-old white man who was admitted to the hospital on June 21, 1977, for evaluation of a cardiac murmur. The patient has had a history of a cardiac murmur since the age of 11 weeks. He was evaluated at that time and was thought to have a ventricular septal defect; however, cardiac catheterization was not performed.

The patient's condition first became symptomatic during adolescence, when he noted dyspnea and easy fatigability during gym classes in high school. At present, the patient states that he experiences dyspnea with minimal exertion and fatigue. He has also noted the onset of a diffuse substernal "aching" pain in the chest that occurs with exertion, is nonradiating, and is associated with moderate dyspnea.

Physical examination revealed a gracile man with a blood pressure of 140/80 mm Hg; his pulse rate was 90 beats per minute and regular. There was no jugular venous distention. The carotid upstroke was brisk and full. The chest was clear to auscultation and percussion.

The cardiovascular examination disclosed an apical impulse in the sixth intercostal space 9 cm from the midsternal line. A palpable lift was felt along the left sternal border and at the apex. There was also a palpable systolic thrill along the left sternal border. The first heart sound (S₁) was normal, and the second heart sound (S₂) was widely split; however, it narrowed with expiration. There was a grade 4/6 holosystolic murmur that was heard best at the left sternal border and apex. A soft, grade 2/6 apical diastolic rumble was audible at the apex. There was a gallop rhythm with a third heart sound that was audible at the apex. The findings from the remainder of the examination were unremarkable.

The electrocardiogram revealed sinus rhythm, incomplete right bundle-branch block, an axis of −30°, and inversion of the T wave in leads V₁ to V₃. The posteroanterior and lateral chest x-ray films disclosed minimal cardiomegaly, with right ventricular prominence (Fig 1). The echocardiogram showed a dilated left ventricle and left atrium. There was exaggerated motion of the interventricular septum and posterior wall, compatible with left ventricular volume overload. The phonocardiogram revealed diminished intensity of S₁, a widely split S₂, and an intense pansystolic murmur that was recorded best at the left sternal border.

Right and left cardiac catheterization (transseptal) revealed a mean right atrial pressure of 7 mm Hg, with an "a" wave of 10 mm Hg and a "v" wave of 8 mm Hg. Right ventricular pressure was 41/8 mm Hg, with a simultaneous pulmonary arterial pressure of 30/14 mm Hg (mean, 22 mm Hg). The mean left atrial pressure was 13 mm Hg, and the left ventricular pressure was 143/16 mm Hg. The pulmonary vascular resistance was 80 dynes-sec/cm². The oxygen content was 12.7 volumes percent in the superior vena cava, 14.8 volumes percent in the right atrium, 15.2 volumes percent in the inferior vena cava, 16.2 volumes percent in the pulmonary artery, and 17.5 volumes percent in the left ventricle. The oxygen consumption at rest was 270 ml/min, yielding a systemic flow of 8.7 L/min, with an index of 5.65 L/min/m² m. The pulmonary flow was 20.8 L/min, and the shunt flow was 12.1 L/min. The ratio of the pulmonary blood flow to the systemic blood flow was 2.4:1. Curves obtained using the technique of indicator dye dilution revealed the presence of a left-to-right shunt at the ventricular level. Left ventricular cineangiograms showed the presence of a high membranous ventricular septal defect. The findings from left atrial and supravalvular aortic injections were unremarkable.

In summary, this patient is a mildly symptomatic 20-year-old man with a membranous ventricular septal defect and a left-to-right shunt (2.4:1).

*Cardiology Fellow, Indiana University School of Medicine, Indianapolis.

FIGURE 1. Posteroanterior chest x-ray film showing minimal cardiomegaly, with right ventricular prominence.
QUESTIONS

1. How would you manage this patient?
2. If surgery is recommended, what are the criteria for closure of a ventricular septal defect in an adult, an adolescent, and a child?
3. What is the possibility, if any, of spontaneous closure at this age?
4. What is the possibility that, if left untreated, this patient would develop Eisenmenger’s complex?

Comments by
Alexander S. Nadas, M.D.**

1. I do not have much doubt that I would recommend surgical closure of this defect in this patient with a moderate left-to-right shunt and mild pulmonary arterial hypertension, particularly in view of the fact that there was no evidence of a right-to-left shunt.

2. I believe that a net left-to-right shunt with a ratio of pulmonary-to-systemic flow of 2:1 or more (in childhood and beyond) is adequate indication for surgical closure. The presence of any degree of pulmonary arterial hypertension (defined as a mean pulmonary arterial pressure of 20 mm Hg or more) makes this recommendation almost mandatory.

3. Most of the spontaneous closures occur in patients under two years of age. Although defects continue to close or diminish in size beyond this age, the rate of closure is rather low. One cannot really count on it in patients in the third decade and beyond.

4. The possibility that this patient would develop Eisenmenger’s complex if surgery is not performed is reasonably high, perhaps 15 to 25 percent. Surely (and this may be the most important point), the possibility of developing pulmonary vascular obstruction is appreciably higher than that of spontaneous closure.

Comments by
S. Gilbert Blount, Jr., M.D.†

Although the ECG was not shown, it is stated that the axis in the frontal plane was −30°, and while about 10 to 15 percent of the patients with the usual ventricular septal defect will show such an axis, one would, of course, also think of the possibility of an endocardial cushion defect, if indeed the ECG showed true left anterior fascicular block.

Concerning the answers to the specific questions, the following points should be made:

1. Certainly, in a patient of this age who is symptomatic and has a ratio of pulmonary-to-systemic blood flow of 2.4:1, I would recommend surgical closure of the ventricular septal defect as soon as it could be conveniently arranged.

2. Concerning the question about the criteria for closure of a ventricular septal defect in an adult, adolescent, and child, there are no brief and simple answers to this question, as each patient would have to be considered individually; however, in general, the following might be said:

Let us consider the adult first. I would advise surgery enthusiastically in any adult with a normal pulmonary vascular resistance and a ratio of pulmonary-to-systemic blood flow of two or greater. I would, under no circumstances, of course, advise surgery in an adult with a ventricular septal defect that would fall into the category of so-called Eisenmenger’s complex. Now, in between, there are many circumstances that would govern the enthusiasm with which one would advise surgery. Thus, age would be a consideration, as would the level of the pulmonary vascular resistance. The higher the pulmonary vascular resistance, the greater the risk of the surgery and, frequently, the less to be gained.

Regarding the child, I would not advise surgery in any child with normal pulmonary vascular resistance and a ratio of pulmonary-to-systemic blood flow of less than 1.5:1. The possibility of spontaneous closure is good; and, of course, the younger the child when first seen, the higher the probability of spontaneous closure. There have been patients with much larger pulmonary blood flows in whom the ventricular septal defects have closed spontaneously. Closure usually occurs during the first four or five years of life. Thus, if the child was in the first year or two of life and had no evidence of failure and minimal cardiac enlargement, even if the ratio of pulmonary-to-systemic blood flow was somewhat larger than 1.5:1, I would still follow the patient to the age of about five years. Then, if closure had not occurred or if evidence of decreasing size of the defect was not suggested, I would advise surgical closure. Although there is a distinct possibility of spontaneous closure after five years of age, I would, in general, advise surgical closure in children five years of age or older with a ratio of pulmonary-to-systemic blood flow of greater than 2:1.

I believe that the question relative to surgical closure in adolescents has been largely answered in the foregoing; however, in any adolescent with a ratio of pulmonary-to-systemic blood flow of 1.8:1 or greater, I would advise surgical closure, although, admittedly, there is an outside possibility that some defects might close spontaneously. Nevertheless, when one considers the very minimal risk of surgical

**Chief, Department of Cardiology, Children’s Hospital Medical Center, Boston.
†Head, Division of Cardiology, University of Colorado, Denver.
closure today and the probable complications that might develop without surgery, I believe it best to advise surgery. For the adolescent with a very small ventricular septal defect (that is, with a ratio of pulmonary-to-systemic blood flow of less than 1.5), one might wish to follow the patient, particularly if there have been signs suggesting a decrease in size of the defect. The decision and the timing of surgical closure, if decided upon, would vary from patient to patient.

3. In answer to question 3, the majority of spontaneous closures occur during the first few years of life, and, thus, at 20 years of age, the chances for spontaneous closure are greatly reduced; however, it is certainly possible that this defect might still close spontaneously, although the older the patient, the smaller the possibility. We have documented spontaneous closure in patients in their 20s, and I believe that spontaneous closure can occur at almost any age; however, in this particular case of a 20-year-old individual with a ratio of pulmonary-to-systemic blood flow of 2.4:1, I would think that the chances of spontaneous closure were somewhat remote.

4. This brings us to the question about Eisenmenger's complex. Given a patient 20 years of age with normal pulmonary vascular resistance, the probability of the development of Eisenmenger's complex in the usually accepted sense is most remote; however, the development of increasing pulmonary vascular resistance, secondary to possible embolization or thrombosis in situ, is a definite possibility and not infrequently happens toward the end of the natural history of this defect. This could, of course, result in the development of severe pulmonary hypertension and reversal of the shunt; however, I do not consider this as the development of Eisenmenger's complex in the usual sense.

DISCUSSION

The patient underwent surgical closure of his ventricular septal defect on June 30, 1977, without complication. The patient was seen four months after surgery, and he had returned to work. At present, he is asymptomatic and denies pain in the chest or dyspnea.

As Nadas and Blount have already stated, the reasons for recommending surgical closure depend heavily upon knowledge of the natural history of this disorder.1-3 The probability of spontaneous closure of the defect in an adult is very small, and, if untreated, the possibility of development of serious complications (infective endocarditis, congestive heart failure, Eisenmenger's complex, and, possibly, aortic regurgitation) would still remain; however, the data available in the literature on the probability of developing progressive pulmonary vascular obstruction are not clear.4 Studies of the natural evolution of this disorder contain, of necessity, populations of patients in whom surgical closure of the defect often interrupted the natural history.4 Therefore, we are not certain what natural course would be taken in those individuals with large left-to-right shunts and normal pulmonary vascular resistance (ie, those individuals who are most likely to undergo surgery). Despite this, the data suggest that patients with ventricular septal defects with normal or mildly elevated pulmonary arterial pressures and normal pulmonary vascular resistance did not develop increasing pulmonary vascular obstruction with the evolution of a balanced or reversed shunt (Eisenmenger's complex).

REFERENCES