Management of a Mildly Symptomatic 65-Year-Old Woman with an Atrial Septal Defect in the Setting of Normal Pulmonary Artery Pressure

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CASE PRESENTATION

A 65-year-old woman was referred to our institution for evaluation of a heart murmur. She had learned of a heart murmur eight years ago. Three to four years prior to referral, she began therapy with furosemide and digoxin; however, she did not recall her symptoms at that time. She felt well until two months prior to admission when she was admitted to her local hospital for treatment of pneumonia. Since that time she had been bothered by a persistent nonproductive cough. The patient and her family noted a gradual increase in dyspnea on exertion in the last month, but she denied orthopnea, nocturnal dyspnea, chest pain and weight gain. She has had mild ankle edema.

There was a history of hypertension, but she denied having had rheumatic fever, myocardial infarction or other cardiovascular disease. She had had two uncomplicated pregnancies. Medications on admission included furosemide, 40 mg twice daily; digoxin, .25 mg daily; nifedipine, 10 mg three times daily and a potassium supplement. She had had no surgical procedures and family history was negative for heart disease.

Physical examination disclosed a well-developed, thin woman in no apparent distress. Blood pressure was 140/70 mm Hg without orthostatic changes. Heart rate was 90 with occasional extrasystolic beats. Respiratory rate was normal, and she was afebrile. The jugular veins were not distended. The lungs contained fine bibasilar rales.
Figure 2. Standard 12-lead electrocardiogram.

which tended to clear with coughing. Cardiac examination disclosed a harsh grade 3/6 mid-systolic ejection murmur best heard at the left lower sternal border, but which radiated diffusely across the precordium. The second heart sound was not clearly split. A right ventricular heave was palpable in the left parasternal region. The apical impulse was located 2 cm to the left of the midclavicular line. Extremities were without clubbing, cyanosis or edema, and the remainder of findings were normal.

Admission data included a complete blood count, urea nitrogen, creatinine and electrolytes which were normal. A Chem-24 panel was within normal limits except for mildly elevated triglycerides. Chest roentgenogram (Fig 1a and b) revealed cardiomegaly with a cardiothoracic ratio of 15:25. The pulmonary arteries were massively dilated. The pulmonary veins were also enlarged. The aorta was normal in size and configuration. The lung fields suggested fibrous scarring in both bases. The electrocardiogram (Fig 2) demonstrated right axis deviation and nonspecific intraventricular conduction delay. An echocardiogram demonstrated mild-to-moderate right ventricular enlargement with paradoxic septal motion consistent with right ventricular volume overload. Left ventricular motion was normal, and no valvular abnormality was detected. Mild left atrial enlargement was present. Phonocardiogram (Fig 3) demonstrated an early systolic ejection murmur with no clear splitting of the second heart sound. Prolongation of the pre-ejection period to 0.16 second was also found.

Hospital Course

The patient was admitted for elective cardiac catheterization. She was found to have a secundum atrial septal defect with a left-to-right shunt and a pulmonary to systemic flow ratio of 3.2 to 1. Pulmonary arterial systolic pressure was 23 with a mean pressure of 15 and the pulmonary resistance was 1.1 Wood units, both within normal limits. Coronary arteriogram revealed normal arteries. During the procedure the patient developed bradycardia requiring administration of atropine. This was followed by the onset of asymptomatic atrial fibrillation with a ventricular response rate of 110 and stable blood pressure. Shortly after return to her hospital room this converted to normal sinus rhythm. That night she experienced transient hypotension that responded to administration of fluids.

Questions

1. How should this patient be managed? Do you recommend surgical closure of the septal defect, and why?
2. Would there be a significant difference in surgical outcome if the patient underwent surgical closure while stable versus waiting until evidence of hemodynamic compromise?
3. If you do not recommend surgical repair of the defect, are there any findings which would cause you to modify this approach later?
This patient has an atrial septal defect with a large left-to-right atrial shunt and normal pulmonary pressure. Atrial arrhythmias, at first intermittent, are common. She has pulmonary symptoms, cough and dyspnea on exertion, and peripheral edema. Upon clinical examination of the patient, the heart is enlarged with a right ventricular heave. Cardiac enlargement is also evident on X-ray examination of the chest. Radiographic evidence of enlarged pulmonary arteries, and cardiographic evidence of an enlarged right ventricle, with paradoxic motion of the ventricular septum, are consistent with findings of a large left-to-right interatrial shunt.

Closure of the atrial septal defect in this patient is recommended. Surgical repair of atrial septal defects in 16 patients over 60 years of age, six of whom had pulmonary artery systolic pressures of 50 mm Hg or greater, with no operative mortality and with low morbidity, was reported from our institution. The condition of most of these patients improved significantly. Sutton et al reported closure of atrial septal defects in 66 patients who were 60 years or older. Among them, four patients died (6 percent), all of whom had undergone concomitant procedures. Decreased symptoms were noted in all groups of survivors, regardless of the patient's preoperative pulmonary artery pressure or functional classification.

The natural history of patients with an atrial septal defect has been reported by Markman, et al and Craig and Selzer and their colleagues. All three reports reflect a rapid increase in disability and death after the age of 40 years. The mortality in patients between ages 40 to 50 approximates 5 percent per annum and increases thereafter. The incidence of symptoms similarly increases in the older patient, with heart failure being related more to the occurrence of atrial arrhythmias than to the presence of pulmonary hypertension. The occurrence of elevated pulmonary pressure, rare before 20 years of age, rises significantly after age 30. About 30 percent of patients in their fifth and sixth decades have appreciable pulmonary hypertension. After an atrial septal defect is closed successfully, atrial fibrillation appears to persist more often than paroxysmal supraventricular tachycardia.

Closure of atrial septal defects in patients 60 years and older can be accomplished with low risk, and the outcome, at least in patients with symptoms and a large left-to-right shunt, appears preferable to continued medical management.

**References**


**Comments**

**Richard W. Campbell, M.D.†**

The patient is a 65-year-old woman who has an atrial septal defect with a left-to-right shunt and a pulmonary to systemic blood flow ratio of greater than 2, but no pulmonary hypertension. In general, uncomplicated atrial septal defects should be closed in all adults under 45 years of age with left-to-right shunts and a Qp/Qs greater than 1.5. Prior to 40 years of age, patients usually have only mild symptoms, either class 1 or 2. Despite no or only mild symptoms, prophylactic surgery is recommended because spontaneous closure in adults is rare and the operative mortality rate is less than 1 percent in young-to-middle aged patients with secundum or sinus venous defects.

The natural history of atrial septal defect is marked by increased mortality in the fourth decade in the unoperated patient; however, it is difficult to predict the clinical course in an individual patient. The single most important risk is the development of pulmonary hypertension. This risk is relatively high in patients between 18 and 40 years of age (14 percent). In this age group, the pulmonary hypertension can progress rapidly and become irreversible, converting an operable condition to an inoperable one. Therefore, prophylactic closure of atrial septal defects in patients under 45 years of age is indicated despite the absence of significant symptoms.

Even in the older patient who has chronic right heart failure but no pulmonary hypertension, life expectancy is improved and patients are symptomatically improved after surgery. The operative risk is increased to 6 percent in this age group, but still is acceptable. Because the patient had symptoms, supraventricular dysrhythmias, a history of pulmonary infection, and a significant left-to-right shunt with a

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pulmonary to systemic blood flow ratio of greater than 2 without pulmonary hypertension, surgery was proposed to her. She refused the surgery, stating that she was satisfied with her lifestyle and that her symptoms were not disabling.

Do we need to be concerned that her refusal of surgery at this time will necessarily result in increased mortality and conversion of an operable to an inoperable lesion, or can surgery be delayed until symptoms increase and the patient is dissatisfied with her lifestyle? Unlike the group younger than 45 years of age, the risk of developing severe obstructive pulmonary hypertension does not seem to be very high in middle-aged or older patients. Prophylactic closure is recommended to prevent the development of pulmonary hypertension. Since this is uncommon in the older age group, then prophylactic surgery in the older patient with only mild symptoms may not be necessary, especially in view of the increased surgical mortality. In the series of Sutton et al.1 of patients greater than 60 years of age, surgery improved the actuarial survival; however, this improvement did not become statistically significant until four-five years after surgery. Forfang2 followed ten middle-aged patients with atrial septal defect who were not operated on. Five died in the follow-up period. There were five survivors who were followed-up for 6½ to 15 years. All of the survivors were still acceptable candidates for surgery. Unlike the younger patients, the older patients are symptomatic from congestive heart failure and supraventricular arrhythmias, but not pulmonary hypertension.

The symptoms related with increasing age result from left ventricular dysfunction, pulmonary emboli, and bronchial-pulmonary infections. Our patient did demonstrate intermittent atrial dysrhythmias, but remains in sinus rhythm on anti-arrhythmic therapy. She also has had an episode of pneumonia. Her left ventricular function remains intact with a left ventricular ejection fraction of 57 percent and left ventricular end diastolic pressure of 3 mm Hg.

Even though older patients with atrial septal defect who are significantly symptomatic should be offered surgery because of possible improved survival and the expectation for symptomatic improvement, I am not sure we can justify recommending prophylactic surgery for asymptomatic or only mildly symptomatic older patients, since the risk of developing irreversible pulmonary hypertension and conversion from an operable to an inoperable condition does not seem to be high. Prophylactic surgery in the older symptomatic or mildly symptomatic patient with an atrial septal defect with a left-to-right shunt and pulmonary blood flow to systemic blood flow ratio greater than 1.5 without pulmonary hypertension probably does not need to be recommended.

Recommendation for surgery in the older patients should possibly be based upon presence of the symptoms rather than simply the presence of the lesion. It is entirely possible that surgery can be delayed to this point without paying the penalty of increased mortality or rendering the patient inoperable; however, when symptoms are significant from CHP and supraventricular dysrhythmias, surgery would not be withheld from a patient simply on the basis of advanced age.

REFERENCES

4 St John Sutton MG, Tajik AJ, McGoon DC. Atrial septal defect in patients ages 60 years or older: operative results and long-term postoperative follow-up. Circulation 1981; 64:402-09
5 Forfang K. Natural history of atrial septal defect of secundum type in the middle-aged. Cardiology 1975; 63:73-8

EDITOR’S NOTE

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We note with interest that the second heart sound is not split in this individual with atrial septal defect, a lesion which regularly produces wide and fixed splitting of this sound. Delay of electrical activation of the left ventricle probably explains the peculiarity of the second sound in this case, for the electrocardiogram (Fig 2) is compatible with left bundle branch block and the phonocardiogram (Fig 3) lends further support by demonstrating a delay of onset of the carotid pulse rise (pre-ejection period prolonged to 0.16 second, about 0.05 second beyond the longest normal value). Thus, delay of ejection of the left ventricle would delay aortic valvular closure sufficiently enough to coincide with that of the pulmonic valve and thereby account for a single second sound.

A point of clinical importance, however, is that relatively narrow or even normal splitting of this sound may be found in older individuals with atrial septal defects—even with normal electrical activation of the left ventricle.1 This unexpected finding is probably attributable to a loss of pulmonary compliance with age and a more abrupt pulmonary valve closure as the right ventricular pressure falls in early diastole.

After extended discussions with the patient and her family, the patient elected not to undergo surgical repair of the septal defect at the time of its discovery. She desired to be followed-up in the clinic with the option of accepting this treatment later if it were warranted by a change in her clinical condition. She has been followed-up for approximately 1½ years, with no change in her symptoms and signs.

REFERENCE

1 Tavel ME. Clinical phonocardiography and external pulse recording, 4th ed. Chicago: Year Book, 1985; 100