Ostium Secundum Atrial Septal Defect discovered after 60 years of age.

Clinical Case Portal

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Case Report

The diagnosis of atrial septal defect should be considered when investigating patients with possible intra-cardiac shunts causing right heart failure. Imaging with cardiac transthoracic echocardiography with contrast made the pre-operative diagnosis possible, and clearly demonstrated the defect anatomy and blood flow through the defect on color Doppler confirmed with a positive bubble study. Appropriate management with surgical closure was then instituted.

Patient history prior to current observation :

A 61 year old man presented with an 18 month history of declining exercise capacity. In recent months, he noted onset of more profound exertional dyspnea and bilateral lower extremity edema.

Clinical findings on admission, evolution and outcome :

Clinical exam revealed an irregularly irregular rhythm at 70 beats/min, and blood pressure of 120/70 mm Hg. There was a fixed split S2, and a 2/6 holosystolic murmur, along the left sternal edge. There was trace pedal edema bilaterally. No other abnormalities were found on examination. The **ECG** showed atrial fibrillation at 70 beats/min, right axis deviation (QRS axis at + 127), incomplete right bundle branch block (fig. 1). On the **chest x-ray** the cardiac index was 0,6; the main pulmonary artery segment of left cardiac border was prominent suggesting pulmonary hypertension; clear expanded lungs and costophrenic angles prevail (fig. 2).

Stress Testing: on exercise electrocardiography, the patient exercised 4:30 minutes on a standard Bruce protocol (6.2 METS), stopping because of shortness of breath. Exercise ECG was negative for ischemic changes.

Initial **transthoracic echocardiography** showed severe biatrial enlargement, severe RV enlargement with mildly reduced RV systolic function (fig. 3). Mild pulmonary hypertension (estimated PASP 30mmHg + RA pressure). There was moderate tricuspid regurgitation and mild mitral regurgitation. There was interventricular septal flattening in diastole (“D”-shaped septum) consistent with RV volume overload (fig. 4), a dilated IVC with blunted respirophasic variation. There is a significant ostium secundum atrial septal defect (about 2.0 cm in diameter) (fig. 5) with evidence of bi-directional shunt noted by color Doppler interrogation (fig. 6) and injection of agitated saline contrast (fig. 7) via the left antecubital vein.

Cardiac catheterization revealed no significant epicardial coronary disease. In addition, the catheterization revealed the following : Mean right atrial pressure 10 mm Hg, right ventricular pressure 32/5 mm Hg, Pulmonary artery pressure 34/11 (mean 19), Mean pulmonary capillary wedge pressure 11 mm Hg. Superior vena cava O2 Sat: 63%, and Pulmonary artery 02 saturation 79%. The pulmonary artery blood flow relative to systemic blood flow was 2.0:1.0 (i.e. Qp/Qs > 1.5). Summary: No significant coronary disease. Right heart catheterization revealed normal pressure and a step-up in saturations consistent with the patient's known atrial septal defect.

Clinical evolution

Given the significant secundum atrial septal defect in a patient who was increasingly limited by exertional dyspnea, he was referred for open surgical closure (there was concern that moderate-severe tricuspid regurgitation may need to be repaired also).

Outcome

The patient underwent successful atrial septal defect closure with Dacron patch. Post-op echo bubble study shows excellent result with no residual shunt (fig. 8).

Discussion

Atrial septal defect is the most frequently encountered major congenital cardiac disorder in the adult population, with a prevalence of 0.2 to 0.7 per thousand (1). Most (75%) are ostium secundum defects; the remainder are ostium primum (20%), sinus venosus defects (<5%), and the rare coronary sinus (<1%). Patients often present late in life because symptoms are rare until later in life and physical findings may go undetected (2).

Patients with moderate sized atrial septal defect typically have a systolic ejection murmur (from increased flow across the pulmonary valve) best heard at the 2nd left intercostal space, a mid-diastolic rumble (from increased flow across the tricuspid valve), and a wide or fixed split S2 (from right ventricular conduction delay and increased pulmonary flow delaying pulmonary valve closure) (1).

The natural history includes development of progressive symptoms (dyspnea in our patient), cardiomegaly, atrial fibrillation and flutter, right ventricular hypertrophy, and pulmonary hypertension (may get a loud P2). Patients with atrial septal defect have a reduced life expectancy (2, 3). The typical natural history involves the onset of atrial fibrillation, with an incidence ranging from 13 to 52 percent among patients older than 40 years, as well as the progression of pulmonary arterial hypertension in up to 53 percent of patients, which results in congestive heart failure and functional limitation (3). Patients with hemodynamically significant shunts and/or symptoms who meet current criteria should be referred for closure (surgical or percutaneous) of their defect (4, 5, 6).

Hemodynamically insignificant atrial septal defects (Qp/Qs < 1.5) do not require closure; however, if the patient has had a stroke secondary to paradoxical emboli it can be closed to prevent another such stroke (4). Significant atrial septal defects i.e. Qp/Qs >= 1.5 or associated with right ventricular volume

overload, should be closed if closure criteria are met. There is an improvement in exercise capacity in asymptomatic and mildly symptomatic adults after atrial septal defect closure (7). Patients with fixed severe pulmonary hypertension that show no pulmonary artery reactivity when tested with oxygen, nitric oxide, or other pulmonary vasodilator, will not benefit from closure of their defect.

Conclusion

At present, appropriately selected ostium secundum atrial septal defects are indicated for closure devices. These devices include the Amplatzer Septal Occluder, the Atrial Septal Defect Occlusion System, the Buttoned Device, the Guardian Angel (Angel Wings), the Helex Septal Occluder, the StarFlex (Cardioseal, Clamshell), and transcatheter patch closure of atrial septal defects (8, 9). An adequate inferior and superior rim around the defect enables device closure without impinging on adjacent structures such as the superior or inferior vena cavae or atrioventricular valves (10).

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Fig. 1 :

Ostium Secundum ASD_Electrocardiogram

Fig. 2 :
Ostium Secundum ASD_Chest X-Ray (AP view)

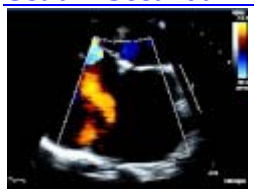
Video 1 :
Ostium Secundum ASD_Transthoracic Echocardiogram (PLAX view)

Video 2 :
Ostium Secundum ASD_Transthoracic Echo_PSAX View

Video 3 :
Ostium Secundum ASD_Transthoracic Echo_A4C View

Video 4 :

Ostium Secundum ASD_Transthoracic Echo_A4C Color Doppler



Video 5 :

Ostium Secundum ASD_Transthoracic Echo_A4C View - Bubble Study

Video 6 :

Ostium Secundum ASD_Transthoracic Echo Post-Surgery_A4C View Bubble Study