HISTORY

39-year-old woman.

CHIEF COMPLAINT: Mild dyspnea on exertion and recurrent palpitations, increasing over the past six months.

PRESENT ILLNESS: A heart murmur was first noted during childhood. She recalls joint pains without definite arthritis in her teens. There is no history of maternal illness during pregnancy. Her growth and development were normal and she never had cyanosis.

Question: What category of heart disease is suggested by this history?
**Answer:** The early appearance of a murmur suggests congenital heart disease. Many patients give a history of arthralgia, but unless true arthritis is present, the diagnosis of rheumatic fever is unlikely.

**PHYSICAL SIGNS**


b. **VENOUS PULSE** - The CVP is estimated to be 6 cm of H₂O.

**Question:** How do you interpret the jugular venous pulse?
**Answer:** The venous pulse pressure is normal, but the wave form is unusual in that the height of the “v” wave (broken arrow) is equal to or slightly greater than the “a” wave (arrow). This finding is highly suggestive of an atrial septal defect, wherein the normally dominant left atrial “v” wave is transmitted to the right atrium and, hence, into the jugular veins. Mild tricuspid regurgitation may also produce a prominent “v” wave.

**Proceed**
c. ARTERIAL PULSE - (BP = 120/80 mm Hg)

Question: How do you interpret the arterial pulse?
**Answer:** The arterial pulse is normal.

d. PRECORDIAL MOVEMENT

![Diagram showing precordial movements and ECG](image)

**Question:** How do you interpret the precordial movements?
**Answer:** There are brisk, nonsustained (hyperdynamic), early systolic impulses at the upper and lower left sternal edge, suggesting volume overload of the pulmonary artery and right ventricle.

e. **CARDIAC AUSCULTATION**

![Cardiac Auscultation Diagram]

**Question:** How do you interpret these auscultatory findings?
**Answer:** The second heart sound is widely split (0.07 sec.) and does not move with respiration. Before one can conclude that there is fixed and not just wide splitting of the second heart sound, auscultation should be performed with the patient standing and during the Valsalva maneuver. These maneuvers decrease venous return, and tend to bring the two components of the second heart sound closer together so that respiratory variation may become clearer.

Wide splitting is consistent with an atrial level shunt. Since the right ventricle receives both shunted blood and systemic venous return, ejection is prolonged, delaying the pulmonic valve closure. Alternatively, wide splitting may be explained by an increase in pulmonary vascular compliance that results in an inertial delay of P2 (“hang out”). Fixed splitting is due to variation in shunt volume thereby keeping right ventricular volume constant as respiration varies systemic venous return.

**Proceed**
Answer (continued): There is an early to midsystolic murmur at the upper left sternal edge. The early systolic timing of the murmur suggests that it is related to the turbulence of flow alone, as the majority of the blood is ejected from the ventricles in early systole. This murmur (flow across the pulmonic valve) and the associated fixed splitting of the second heart sound are consistent with a left to right shunt at the atrial level, such as an atrial septal defect. No audible murmur is generated at the site of an atrial septal defect, as blood flows through it without turbulence because there is no significant gradient between the right atrium and left atrium.

Proceed
Question: How do you interpret the auscultatory findings at the lower left sternal edge?
**Answer:** Single first and second sounds are present. In an atrial septal defect (ASD), the first sound at the lower left sternal edge may be split, with an increase in the tricuspid component. This may be seen with volume overload of the right ventricle associated with vigorous ventricular contraction.

There is a mid-diastolic murmur (arrow) due to increased flow into the right ventricle. This finding indicates that the size of any atrial level shunt is significant.

**f. PULMONARY AUSCULTATION**

**Question:** How do you interpret the acoustic events in the pulmonary lung fields?

**Proceed**
**Answer:** In all lung fields, there are normal vesicular breath sounds.

**ELECTROCARDIOGRAM**

**Question:** How do you interpret this ECG?
Answer: The ECG shows sinus rhythm. The classic rsR’ in lead V1 with a normal QRS duration is consistent with volume overload of the right ventricle. In some individuals, a small terminal r’ in lead V1 may be a normal finding.

CHEST X RAYS

Question: How do you interpret the patient’s chest X rays?
**Answer:** The cardiac silhouette is enlarged. The extremely prominent pulmonary artery (arrow), the inconspicuous aortic arch and the increased pulmonary arterial vascularity are typical of an adult atrial septal defect. In the lateral view, the cardiac silhouette fills in the retrosternal space (broken arrow), consistent with right heart enlargement. The left atrium is not enlarged.

The history, physical examination, ECG and chest X rays are all consistent with an ostium secundum atrial septal defect. The size of the right heart structures, the prominent pulmonary vascularity and the diastolic murmur suggest that the shunt is greater than 2.0 to 1.

**Question:** How would you evaluate the patient’s complaint of palpitations?
Answer: The patient’s palpitations are most likely caused by an atrial dysrhythmia. These occur in adults with atrial septal defects and become more frequent with increasing age. They are best documented by continuous ECG monitoring for a period of at least several hours. The strip below was obtained when the patient had symptoms.

Questions:
1. How do you interpret this rhythm strip?
2. Would an echocardiogram help to confirm your clinical diagnosis of atrial septal defect?
**Answers:**

1. The rhythm strip shows atrial fibrillation with a moderate ventricular response of approximately 100.

2. Two-dimensional echocardiography must be obtained in order to confirm the diagnosis, localize the defect in the atrial septum, determine its size, and evaluate other cardiac structures and function.

   An ostium secundum atrial septal defect is the most common type, and is usually well defined by the subcostal four chamber view. This is demonstrated on the patient’s study that follows.

**Proceed**
LABORATORY- ECHOCARDIOGRAM
TWO-DIMENSIONAL SUBCOSTAL FOUR CHAMBER VIEW

Proceed

RV = Right Ventricle
RA = Right Atrium
LV = Left Ventricle
LA = Left Atrium
ASD = Atrial Septal Defect
This study demonstrates a large secundum ASD (arrows) and right heart dilation.

Color Doppler or pulsed wave Doppler can also locate the shunt. Continuous wave Doppler can quantify it.

The patient’s study follows.
The arrow heads in the four-chamber apical view identify the atrial septal defect. The orange color (arrow head) identifies the left to right shunt flow.

**Question:** Is cardiac catheterization necessary?
Answer: Cardiac catheterization is not indicated in this condition. Data expected in an atrial septal defect would be as follows.

Proceed
**LABORATORY (continued) - CATHETERIZATION DATA**

<table>
<thead>
<tr>
<th>SITE</th>
<th>PRESSURE (mm Hg)</th>
<th>OXYGEN SATURATION(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior Vena Cava (SVC)</td>
<td>Mean = 4</td>
<td>72</td>
</tr>
<tr>
<td>Mid Right Atrium (MRA)</td>
<td>Mean = 4</td>
<td>86</td>
</tr>
<tr>
<td>Right Ventricle (RV)</td>
<td>31/3</td>
<td>88</td>
</tr>
<tr>
<td>Right Pulmonary Artery (RPA)</td>
<td>25/8</td>
<td>88</td>
</tr>
<tr>
<td>Left Ventricle (LV)</td>
<td>120/8</td>
<td>96</td>
</tr>
<tr>
<td>Femoral Artery (FA)</td>
<td>124/80</td>
<td>96</td>
</tr>
</tbody>
</table>

**Question:** How do you interpret these data?
**Answer:** A significant (14%) increase in oxygen saturation is present at the right atrial level, caused by shunting of highly oxygenated blood from the left atrium to the normally less oxygenated right atrium.

Based on the saturation data, the ratio of pulmonary blood flow to systemic blood flow in this example is approximately 3 to 1.

The slight systolic pressure gradient from the right ventricle to the right pulmonary artery is due to flow alone.

**Question:** *What therapy do you recommend?*
**Answer:** Many ostium secundum atrial septal defects, even if large, can be closed by transcatheter techniques. In some cases, however, surgery may be necessary. Decisions regarding closure technique are based upon the particular anatomic characteristics of the defect. Ostium primum and sinus venosus defects require surgery.

The recommended age for closure of atrial septal defects is after infancy and before school age. This patient’s defect was closed without complication and with relief of her symptoms.

Bacterial endocarditis prophylaxis is not required for an isolated atrial septal defect. Endocarditis prophylaxis is indicated for six months after closure.

**Proceed**
SUMMARY

Defects in the atrial septum are of three general types:

1. Sinus Venosus Defects - located high in the septum at the superior vena cava - right atrial junction;
2. Ostium Secundum Defects - located in the midseptum in the region of the fossa ovalis;
3. Ostium Primum Defects - located low in the septum at the atrioventricular valves.

Ostium secundum atrial septal defects are by far the most common and are occasionally familial. There is a higher incidence in females, and about 20 percent of patients have associated mitral valve prolapse. Anomalous return of a right upper lobe pulmonary vein to the superior vena cava occurs in most patients with a sinus venosus defect. Partial anomalous pulmonary venous return without an atrial septal defect occurs rarely but can present a similar clinical picture. Bony abnormalities of the forearm and the thumb (Holt-Oram syndrome) may be a clinical clue to the presence of an atrial septal defect.
SUMMARY (continued)

From a patient with an atrial septal defect associated with a “fingerized thumb” (Holt-Oram syndrome). In this case, severe pulmonary vascular resistance occurred, causing reversal of the shunt (right to left), and resulting in clubbing of the fingers.

Proceed
SUMMARY (continued)

Uncomplicated ostium secundum defects usually are asymptomatic in children and young adults, although palpitations, dyspnea and fatigue may occur. Atrial dysrhythmias and heart failure increase in frequency with age. The defect may not be recognized for years because of the lack of symptoms and the fact that the flow murmur across the pulmonary outflow tract may be interpreted as “innocent.”

With large atrial septal defects, there usually is no pressure gradient between the atria. The direction and magnitude of the shunt relate to the increased compliance of the right atrium and ventricle relative to the left.
SUMMARY (continued)

Pulmonary vascular obstructive disease due to an atrial septal defect occurs rarely in the adult. When pulmonary resistance increase is substantial, closing the atrial septal defect may not help the patient. In some patients, the left ventricle may become less distensible with age, resulting in an increase in left to right shunting.

The typical pathology follows.

Proceed
Specimen from patient who had unsuccessful surgery. A patch is sutured over the 2 cm defect. The marked right ventricular hypertrophy in this specimen is secondary to pulmonary vascular obstructive disease.

Proceed for Case Review
To Review This Case of Ostium Secundum Atrial Septal Defect in an Adult:

The patient’s **HISTORY** is typical, with a murmur heard in childhood and adult onset of dyspnea and dysrhythmia.

**PHYSICAL SIGNS:**

a. The **GENERAL APPEARANCE** is that of a normal woman.

b. The **JUGULAR VENOUS PULSE** mean pulse is normal and the “a” and “v” waves are approximately equal, as the dominant left atrial “v” is transmitted through the defect.

c. The **ARTERIAL PULSE** is normal.

d. The **PRECORDIAL MOVEMENT** reveals hyperdynamic pulmonary artery and right ventricular impulses due to volume overload of the right heart.

**Proceed**
e. **CARDIAC AUSCULTATION** reveals wide fixed splitting of the second heart sounds from the volume overload of the right ventricle which occurs without significant respiratory variation.

The early crescendo-decrescendo murmur at the upper left sternal edge is due to increased flow across the pulmonic valve.

The diastolic flow rumble across the tricuspid valve indicates that the shunt is significant in size.

f. **PULMONARY AUSCULTATION** reveals normal vesicular breath sounds in all lung fields.

Proceed
The **ELECTROCARDIOGRAM** shows the classic rsR’ in lead V1 associated with right ventricular volume overload.

The **CHEST X RAYS** show right heart and pulmonary artery enlargement with increased pulmonary vascularity.

**LABORATORY STUDIES** reveal right ventricular volume overload and a large secundum atrial septal defect on two-dimensional echocardiography with a left-to-right shunt shown on the color flow Doppler.

The **TREATMENT** is closure of the defect.