HISTORY

43-year-old woman.

CHIEF COMPLAINT: Progressive exertional dyspnea, orthopnea and nocturnal cough of two months duration.

PRESENT ILLNESS: At age four a murmur was first noted. While she was advised to limit her activities, she led an active life, including three uneventful pregnancies. There is no history of rheumatic fever, cyanosis, chest pain or neurological symptoms. Her mother was well during her pregnancy, and the patient’s birth, growth, and development were normal.

Question: What diagnosis is suggested by this history?
**Answer:** In the absence of rheumatic fever, the presence of a murmur early in life suggests congenital heart disease. Her symptoms indicate the recent onset of left heart failure.

Diagnostic considerations in this acyanotic woman include a ventricular septal defect (VSD) and patent ductus arteriosus (PDA). Symptoms of left heart failure are associated with these lesions as excess blood is recirculated to the left heart. Left ventricular outflow tract obstruction may also produce left heart failure, but the lack of exertional chest pain or syncope makes this diagnosis unlikely. Congenital aortic regurgitation (AR) or mitral regurgitation (MR), though less common, should be considered. Pulmonic stenosis is inconsistent with her history of left heart failure.

**Proceed**
PHYSICAL SIGNS

a. GENERAL APPEARANCE - Acyanotic woman appearing normal at rest.

b. VENOUS PULSE - The CVP is estimated to be 7 cm H₂O.

**Question:** How do you interpret the venous pulse?
**Answer:** The CVP and venous pulse contour are normal.

c. **ARTERIAL PULSE - (BP = 150/50 mm Hg)**

![](diagram.png)

**Question:** How do you interpret the arterial pulse?
**Answer:** The pulse pressure is wide with a low diastolic component. The carotid pulse is bounding with brisk upstroke. These changes reflect an increased stroke volume with rapid runoff as seen in PDA or AR.

The height of the systolic pressure is exaggerated by the artifact of pulse amplification. This artifact occurs because the arterial pulse wave is compressed as it travels distally into decreasing caliber arteries. Wide pulse pressure further magnifies this artifact.

The pulse is not typical of VSD alone, unless there is associated AR (as from prolapse of the right coronary cusp). The long history of murmur and the gradual onset of symptoms is inconsistent with acute rupture of a sinus of Valsalva aneurysm producing AR.

The only form of isolated left ventricular outflow obstruction associated with a brisk carotid upstroke is hypertrophic obstructive cardiomyopathy, but the wide pulse pressure with the low diastolic component makes this possibility unlikely. MR also produces a brisk upstroke, but with a normal pulse pressure and diastolic pressure.

**Proceed**
d. PRECORDIAL MOVEMENT

Question: How do you interpret the apical impulse?
**Answer:** The apical impulse is displaced laterally, enlarged and non-sustained, consistent with left ventricular volume overload as may be seen in PDA and AR.

e. **CARDIAC AUSCULTATION**

**Question:** How do you interpret the acoustic events at the upper left sternal edge?
Answer: The phonocardiogram shows a continuous murmur that peaks at the time of S2. The location of the murmur is consistent with PDA.

Continuous murmurs, by definition, continue through the second heart sound but not necessarily throughout the entire cardiac cycle. They result from a constant intravascular pressure difference that maintains turbulent blood flow throughout systole and all or part of diastole.

Question: What are some of the causes of continuous murmurs?
**Answer:** Specific causes of continuous murmurs, grouped according to mechanism, are given below:

<table>
<thead>
<tr>
<th>MECHANISM</th>
<th>EXAMPLES</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>I. RAPID BLOOD FLOW</strong></td>
<td></td>
</tr>
<tr>
<td>A. Venous</td>
<td>A. Venous hum</td>
</tr>
<tr>
<td>B. Arterial</td>
<td>B. Mammary souffle, bronchial collaterals</td>
</tr>
<tr>
<td><strong>II. HIGH TO LOW PRESSURE SHUNTS</strong></td>
<td></td>
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<tr>
<td>A. Systemic artery to pulmonary artery</td>
<td>A. PDA, surgical shunts</td>
</tr>
<tr>
<td>B. Systemic artery to right heart</td>
<td>B. Ruptured sinus of Valsalva aneurysm, coronary arteriovenous fistula</td>
</tr>
<tr>
<td>C. Arteriovenous fistula</td>
<td>C. Systemic, pulmonary</td>
</tr>
<tr>
<td><strong>III. LOCALIZED ARTERIAL OBSTRUCTION</strong></td>
<td>Pulmonary branch stenosis, systemic artery obstruction</td>
</tr>
</tbody>
</table>

**Question:** How does the contour of this patient’s continuous murmur help in the differential diagnosis?
**Answer:** This patient’s murmur peaks at the time of the second heart sound and is characteristic of a PDA. Coronary-cardiac fistulas and arteriovenous fistulas show a bimodal shape with peaks in both systole and diastole. A venous hum shows diastolic accentuation.

Mimics of a continuous murmur include the “to and fro” murmurs of either aortic stenosis with regurgitation or ruptured aortic sinus of Valsalva aneurysm and the combined murmurs of VSD and AR. Careful auscultation can differentiate these mimics from continuous murmurs.

Examples of these murmurs follow.

**Proceed**
Continuous murmurs and their mimics:

PDA (Peaks at S2)

ARTERIAL OBSTRUCTION (Peaks earlier in systole)

VENOUS HUM (Peaks in diastole)

AORTIC STENOSIS AND REGURGITATION (To and fro)

VSD WITH AORTIC REGURGITATION (To and fro)
e. CARDIAC AUSCULTATION (continued)

**Question:** How do you interpret the acoustic events at the apex?
**Answer:** A third heart sound (arrow) is followed by a short mid-diastolic rumble (broken arrow). These findings are also well heard posterolateral to the mitral area over the enlarged left ventricle.

The presence of a loud continuous murmur at the upper left sternal edge peaking at S2 is consistent with a large shunt due to a PDA (pulmonary to systemic flow ratio of 2.0 or more). The S3 and rumble are related to increased flow across the mitral valve as shunted blood recirculates from the aorta to the lungs and then to the left heart. Alternatively, the diastolic events could reflect some decrease in left ventricular compliance.

**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 electrocardiogram?
Answer: The ECG shows left ventricular hypertrophy associated with ST-T wave changes.

Question: How do you interpret this chest X ray?
Answer: The PA film shows an enlarged cardiac silhouette with a “left ventricular contour” (apex displaced inferolaterally). The aorta is slightly prominent (arrow) as is the pulmonary artery (broken arrow). Pulmonary vascularity is increased.

Question: Based on the history, physical exam, ECG and chest X ray, what is your diagnosis and plan for further evaluation?
**Answer:** The history, physical examination, ECG and X rays are all consistent with a large patent ductus arteriosus. Two-dimensional echocardiography provides information regarding chamber sizes and ventricular function. Doppler interrogation demonstrates flow from the aorta into the pulmonary arteries and can estimate shunt size.
LABORATORY

While not essential for diagnosis, catheterization defines the lesion, coronary anatomy and ventricular function. It also provides an opportunity to apply transcatheter device closure in selected cases. In this procedure, occlusion of the ductus may be accomplished by introducing a device, such as wire coils, that induces clotting. The patient’s study follows.
**LABORATORY - CATHETERIZATION DATA**

<table>
<thead>
<tr>
<th>SITE</th>
<th>PRESSURE (mm Hg)</th>
<th>OXYGEN SATURATION(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right Atrium</td>
<td>Mean = 3</td>
<td>65</td>
</tr>
<tr>
<td>Right Ventricle</td>
<td>30/3</td>
<td>65</td>
</tr>
<tr>
<td>Pulmonary Artery</td>
<td>31/13</td>
<td>83</td>
</tr>
<tr>
<td>Left Atrium</td>
<td>Mean = 16</td>
<td>96</td>
</tr>
<tr>
<td>Left Ventricle</td>
<td>150/16</td>
<td>96</td>
</tr>
</tbody>
</table>

Pulmonary vascular resistance is normal.

Angiography revealed normal coronary arteries.

**Question**: How do you interpret these data?
**Answer:** The right sided pressures are normal, while the left atrial and left ventricular pressures are increased, reflecting the patient’s left ventricular failure. There is a large increase in oxygen saturation in the pulmonary artery (normal = <3%), defining a large left-to-right shunt at the level of the great vessels.

**Proceed**
LABORATORY (continued)

X ray showing catheter introduced into the heart via the left brachial vein.

**Question:** Can you identify the route taken by this catheter?
**Answer:** The catheter course includes the left subclavian vein, superior vena cava, right atrium, right ventricle and pulmonary artery. The wide U loop (arrow) defines the catheter course in the right atrium, ventricle and pulmonary artery. The catheter then passes posteriorly through the patent ductus arteriosus and down the descending aorta (proven by visualizing the catheter extending well below the diaphragm).

**Proceed**
**Questions:**

1. How do you interpret this angiogram?
2. How would you treat this patient?
Answers:

1. The angiogram clearly outlines the patent ductus arteriosus (arrow) between the aorta (broken arrow right) and the enlarged pulmonary artery (broken arrow left). Its short, stout shape was considered unfavorable for coil closure.

2. After management of her congestive heart failure with digitalis, diuretics, salt restriction and an ACE-inhibitor, surgical closure was recommended. This procedure has a negligible risk in children, unless severe pulmonary vascular disease is present, and is usually carried out before age two. In older adults, bleeding complications associated with attempts to divide a calcified defect may occur. This patient’s surgery and postoperative course were uneventful.

Proceed for Summary
SUMMARY

Patent ductus arteriosus is the abnormal persistence of the fetal vascular structure that normally allows the majority of right ventricular output to bypass the lungs and enter the systemic circulation. It arises from the left pulmonary artery and enters the descending aorta just distal to the origin of the left subclavian artery.

The ductus closes functionally within a few hours postpartum, and closes anatomically within several weeks. The mechanism of this closure is related to a change in prostaglandins and to ductal smooth muscle contraction in response to increased systemic arterial oxygen. Delayed closure of the PDA is particularly common in premature infants.

Proceed
Patent ductus arteriosus is more common in females, in premature infants and following maternal rubella infection. It is also more common in infants born at high altitude. The mechanism responsible for its abnormal persistence at high altitudes may relate to associated hypoxemia. In contrast, the persistence of a PDA in premature infants may relate to the ductal musculature being less responsive to oxygen.

At birth, a murmur is typically absent because the initially high fetal pulmonary vascular resistance prevents significant left-to-right shunting. Subsequently, with the postpartum fall in fetal pulmonary vascular resistance, left-to-right shunting begins, producing a continuous murmur that peaks at the time of S2.
SUMMARY (continued)

Congestive heart failure and infective endocarditis are the most common complications in patients with PDA.

Congestive heart failure may occur in infants with large shunts. Patients with moderate shunts, however, are usually asymptomatic until after the third decade when the risk of cardiac failure increases. Patients with small shunts may remain asymptomatic throughout life.

When infective endocarditis occurs, it commonly begins at the site of impact of the ductus stream on the pulmonary artery wall.

Proceed
Pulmonary vascular disease is uncommon except in large shunts and in patients residing at high altitude. Patients with marked elevation of pulmonary vascular resistance are not suitable candidates for ductal closure.

This complication may result in reversal of the shunt, with flow from the pulmonary artery to the aorta (right to left), beyond the left subclavian artery. It may be recognized clinically by disproportionate cyanosis and clubbing in the lower extremities as compared to the upper extremities.

Proceed
Photograph of a patient with reversal of the shunt through a PDA, showing cyanosis and clubbing of the toes with normal fingers.

Proceed
A large patent ductus arteriosus extends from the left main pulmonary artery to the aorta just distal to the origin of the left subclavian artery.

Proceed for Case Review
To Review This Case of Patent Ductus Arteriosus in an Adult:

The **HISTORY** is typical, with a murmur heard in early childhood and the later onset of left ventricular failure.

**PHYSICAL SIGNS**

a. The **GENERAL APPEARANCE** is unremarkable with no cyanosis.

b. The **JUGULAR VENOUS PULSE** is normal in mean pressure and wave form.

c. The **ARTERIAL PULSE** is bounding with a brisk upstroke, and the pulse pressure is wide, reflecting an increased stroke volume with rapid runoff.

Proceed
d. **PRECORDIAL MOVEMENT** reveals an enlarged and inferolaterally displaced apical impulse due to left ventricular volume overload.

e. **CARDIAC AUSCULTATION** reveals the classic continuous murmur at the upper left sternal edge which peaks at the time of S2, and reflects the pressure gradient between the aorta and the pulmonary artery. The third heart sound and diastolic rumble at the apex are due to the increased flow across the mitral valve, and indicate that the shunt is large. These findings are also well heard posterolateral to the mitral area over the enlarged left ventricle.

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

The **ELECTROCARDIOGRAM** shows left ventricular hypertrophy.

Proceed
The **CHEST X RAY** shows increased pulmonary arterial vascularity, left ventricular enlargement, and some prominence of the aortic arch and pulmonary artery.

**LABORATORY STUDIES** include color flow Doppler echocardiography that demonstrates flow from the descending thoracic aorta into the pulmonic trunk.

Cardiac catheterization reveals a large left-to-right shunt at the level of the pulmonary artery by oxygen saturation data. Pulmonary vascular resistance is normal and there is evidence of mild left ventricular failure. The patent ductus arteriosus is confirmed both by catheter course and by contrast aortography.

**TREATMENT** consists of medical control of the congestive heart failure followed by closure of the ductus arteriosus.