HISTORY

45-year-old white man.

CHIEF COMPLAINT: Progressive dyspnea, orthopnea and ankle edema of six months duration.

PRESENT ILLNESS: He first noted the nearly simultaneous onset of dyspnea with exertion and peripheral edema two years ago. At that time, he was found to have cardiomegaly and an abnormal electrocardiogram. He was treated with digitalis, diuretics and salt restriction. There is no history of rheumatic fever, heart murmurs, hypertension, diabetes, or chest pain.

Question: What diagnosis is suggested by this history?
**Answer:** The history of simultaneous onset of biventricular failure, cardiomegaly and an abnormal electrocardiogram is consistent with a dilated cardiomyopathy. No specific etiology is suggested by history.

**PHYSICAL SIGNS**

**a. GENERAL APPEARANCE** – 45-year-old dyspneic man with peripheral edema.

**b. VENOUS PULSE** - The CVP is estimated to be 12 cm H₂O.

**Question:** How do you interpret the jugular venous pulse?
**Answer:** The mean central venous pressure is elevated. In the absence of tricuspid stenosis, this reflects an increased right ventricular filling pressure. There is a prominent or giant “a” wave (arrow), indicating enhanced right atrial contraction against increased resistance.

c. **ARTERIAL PULSE - (BP = 110/90 mm Hg)**

![Graph showing carotid and ECG waves]

**Question:** How do you interpret the carotid pulse?
**Answer:** The arterial pulse is small and the pulse pressure diminished, reflecting a low stroke volume. On another occasion when the patient was examined, the pulse had a different character, as shown below.

**Question:** What is the significance if this arterial pulse?
Answer: The pulse is dicrotic, a type of twice-beating pulse produced by a palpable dicrotic wave (broken arrow) in diastole. It is commonly found in younger patients with severe myocardial disease who have compliant vessels, decreased stroke volume and increased peripheral vascular resistance.

d. PRECORDIAL MOVEMENT

Question: How do you interpret this precordial movement?
**Answer:** The sustained systolic impulse reflects an enlarged right ventricle. The palpable presystolic impulse (arrow) is due to right atrial contraction. The palpable early diastolic impulse (broken arrow) is related to passive filling. They are enhanced due to reduced right ventricular compliance and systolic function.

d. PRECORDIAL MOVEMENT *(continued)*

**Question:** How do you interpret this apical precordial movement?
**Answer:** The location and size of the impulse indicates that the left ventricle is dilated. The systolic movement is sustained, reflecting left ventricular hypertrophy and/or dysfunction. The palpable presystolic impulse (arrow) indicates forceful left atrial contraction while the early diastolic impulse (broken arrow) reflects passive left ventricular filling. Both are due to a poorly compliant left ventricle.

**Proceed**
e. CARDIAC AUSCULTATION

**Question:** How do you interpret these auscultatory findings?
**Answer:** The second heart sound splits normally with inspiration. The pulmonic component is increased, reflecting increased pulmonary arterial pressure.

Prominent fourth (arrow) and third (broken arrow) heart sounds are present at the apex. These sounds are caused by the tensing and vibration of the mitral apparatus and associated structures due to the rapid deceleration of blood as it enters the poorly compliant left ventricle. There is a holosystolic murmur at the apex. All of these findings are also well heard posterolateral to the mitral area over the enlarged left ventricle.

**Questions:**
1. *What is the most likely cause of the murmur at the apex?*
2. *What is its clinical significance?*
Answers:

1. This murmur is most likely due to mitral regurgitation.

2. The recent onset of the murmur is against chronic mitral regurgitation. There is no history of fulminant left ventricular decompensation as may occur in acute mitral regurgitation due to ruptured chordae tendineae. Hence, this patient’s murmur is most likely due to dysfunction of the mitral apparatus associated with a dilated poorly contracting left ventricle. It is unlikely that the major hemodynamic problem in this patient involves a primary abnormality of the mitral valve.

Proceed
Question: How do you interpret the acoustic events at the lower left sternal edge?
Answer: At the lower left sternal edge the fourth heart sound (arrow) and the third heart sound (broken arrow) increase in intensity with inspiration. This indicates their right ventricular origin, as the drop in intrathoracic pressure which occurs during inspiration enhances right ventricular filling. These pathologic filling (or gallop) sounds correlate with the presystolic and early diastolic precordial movements previously described.

f. PULMONARY AUSCULTATION

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

Proceed
**Answer:** In the lower right posterior lung field, breath sounds are absent, reflecting a pleural effusion associated with chronic congestive failure. In the lower left posterior lung field, there are inspiratory crackles, reflecting chronic pulmonary congestion. In all other lung fields, there are normal vesicular breath sounds.

**ELECTROCARDIOGRAM**

**Question:** How do you interpret this ECG?
**Answer:** The ECG shows sinus rhythm, with a premature ventricular beat in lead III, left axis deviation, left ventricular hypertrophy and left ventricular conduction delay. Abnormal P waves suggest left atrial enlargement or intra-atrial block.

The patient has bedside findings of right ventricular enlargement. This is obscured electrocardiographically by the dominant left ventricular mass.

**Proceed**
Question: How do you interpret these chest X rays?
**Answer:** The PA chest X ray shows a large cardiac silhouette due to marked cardiomegaly, pericardial effusion, or both. There is a large pleural effusion in the lower right lung field. There is also venous congestion with loculated fluid in the right interlobar fissure (arrow). This type of fluid collection is called a “phantom tumor,” as it may disappear with therapy. The lateral X ray confirms the pleural effusion and shows posterior displacement of the barium filled esophagus (broken arrow), suggesting left atrial enlargement.

**Question:** Based on the history, physical examination, ECG and chest X rays, what is your diagnostic impression and plan to further evaluate this patient?
Answer: The most likely diagnosis is primary disease of the myocardium, that is, dilated (congestive) cardiomyopathy. An ischemic etiology is possible, but the absence of angina, myocardial infarction and diabetes makes this diagnosis unlikely.

Numerous other etiologies are possible, but most likely will not be identified. In these instances, it would be classified as idiopathic.

Echocardiography would resolve the question of a pericardial effusion, rule out occult valvular disease and confirm cardiac enlargement and poor ventricular function.
Question: How do you interpret this echocardiogram?

RV = Right Ventricle
LV = Left Ventricle

PARASTERNAL SHORT AXIS (SYSTOLE)
Answer: This view shows a markedly dilated left ventricle. In the real-time video, severely hypokinetic wall motion is seen with an estimated ejection fraction of 20%. The remainder of the echo study shows that all chambers are dilated. No pericardial effusion is present and the four valves all appear normal. Doppler study shows mild mitral regurgitation.

Question: Is cardiac catheterization necessary?
**Answer:** Cardiac catheterization is not necessary to confirm the clinical diagnosis of cardiomyopathy. It would determine if the etiology is ischemic, i.e., secondary to coronary artery disease. Specific etiologic information may be obtained by endomyocardial biopsy. Hemodynamic assessment of pulmonary vascular resistance is helpful in transplant evaluation.

The patient’s cardiac catheterization follows.

**Proceed**
ADDitional DATA: (mm Hg)

Right Atrial Mean = 12
Right Ventricle = 50/12
Pulmonary Artery = 50/30
Pulmonary Artery = 30
Wedge
Cardiac Index 2.0 L / Min / M²

Question: How do you interpret these data?
**Answer:** The elevated left ventricular end diastolic pressure (arrow) of 28 mm Hg (normal 5-12 mm Hg) reflects reduced left ventricular function. The elevated pulmonary artery wedge and pulmonary artery pressures mirror the increased left ventricular filling pressure. The cardiac index is low (normal 2.5 - 3.5 L/Min/M²). A left ventricular cineangiogram was performed (not shown) which demonstrated a dilated, poorly but symmetrically contracting left ventricle with mild mitral regurgitation. The coronary arteries were normal.

**Question:** How would you treat this patient?
**Answer:** His digoxin, diuretics and salt restriction were continued for symptom relief. (Goal digoxin levels are < 1 ng/mL).

The cornerstone of therapy involves administration of drugs that have been shown not only to improve symptoms but also to prolong survival. These include: 1) vasodilators, including ACE-inhibitors; 2) beta-adrenergic blocking agents; and 3) aldosterone blocking agents in patients with more severe heart failure.

He was strongly urged to discontinue all alcohol intake. Because of the absence of atrial fibrillation, prior thromboembolic events, or intracardiac thrombus, anticoagulation was not indicated in this patient.

Patients with cardiomyopathy may be sensitive to digitalis. His serum potassium was carefully monitored, as hypokalemia from diuretic therapy is another predisposing factor to digitalis intoxication.

**Question:** What are the common clinical and electrocardiographic manifestations of digitalis intoxication?
**Answer:** The common symptoms are anorexia, nausea, vomiting and visual disturbances.

Premature ventricular contractions are the most frequent nonspecific ECG finding. The most frequent specific ECG finding is accelerated junctional rhythm.

In addition to the clinical and ECG manifestations of digitalis intoxication, monitoring the serum drug level may be helpful in management.

**Proceed**
The therapeutic digoxin level is 1-2ng/ml. Serum digoxin levels should be obtained at least 6 hours after the last dose. Serum levels of any drug obtained too soon after administration, i.e., before equilibrium between blood and tissue, may give falsely high values. Digoxin is excreted by the kidney and, therefore, the dose should be reduced in patients with decreased renal function.

**Question:** What are the basic hemodynamic therapeutic actions of digitalis, diuretics and vasodilators?
**Answer:** Digitalis increases the force of myocardial contraction, improving myocardial performance.

Diuretics decrease fluid volume and can thus relieve symptoms of congestion in patients with excess salt and water accumulation.

Vasodilators are most helpful in patients with poor ventricular function and congestive failure, especially when there is associated mitral regurgitation. The decreased after-(pressure) load from arteriolar dilatation makes it easier for the weak ventricle to expel blood into the aorta, decreasing the amount of mitral regurgitation and increasing forward stroke volume and cardiac output. Some vasodilators reduce pre-(volume) load from venular dilatation, resulting in pooling of blood in the periphery.

These therapies may enhance survival.

**Question:** What is the natural history of this disease and what other therapeutic options are available?
**Answer:** The natural history is one of progressive deterioration with approximately 50% mortality in 2-3 years. The usual causes of demise include ventricular arrhythmias, systemic embolization and progressive heart failure.

Biventricular pacing may be helpful in patients with a QRS interval > 130 msecs to improve symptoms. In patients with dilated cardiomyopathy associated with a prior myocardial infarction and a left ventricular ejection fraction of less than or equal to 30% or patients with documented ventricular arrhythmias, placement of an automatic implantable defibrillator has been shown to improve survival. In patients with supraventricular tachydysrhythmias, amiodarone is the drug of choice.

In patients with severe mitral regurgitation, mitral valve annuloplasty may be indicated.

Progressive heart failure despite maximal medical therapy may be an indication for implantation of a ventricular assist device or for cardiac transplantation.
SUMMARY

The cardiomyopathies may be classified into three clinical types:

1. Dilated (Congestive)
2. Hypertrophic (with or without obstruction)
3. Restrictive (and/or obliterative)

Cardiomyopathy is primary myocardial disease, where the abnormality is in the myocardium, and does not involve the heart valves, pericardium, coronary arteries or lungs, nor is it secondary to a shunt lesion or hypertension.

Proceed
SUMMARY (continued)

Dilated cardiomyopathy is most often idiopathic. A significant percentage is hereditary. Identifiable causes include alcohol, viruses, toxic drugs and the peripartum state. The major differential diagnostic consideration is coronary artery disease. In the absence of clinical data to suggest a rare cause, routine extensive workup is not likely to be fruitful.

Patients with dilated cardiomyopathy may present with heart failure, cardiomegaly, emboli and arrhythmias. The mainstay of medical therapy currently consists of ACE-inhibitors, beta-adrenergic blockers, and aldosterone blocking agents. In addition, the traditional use of diuretics and digitalis continues. Anticoagulation is indicated for intracardiac thrombi, atrial fibrillation, or thromboembolic events.

The typical gross pathology follows.
The disease is diffuse and involves both ventricles, resulting in a flabby musculature. The left ventricle shown here is markedly dilated. The mitral valve is anatomically normal.

Proceed for Case Review
To Review This Case of Dilated Cardiomyopathy:

The **HISTORY** is typical, with nearly simultaneous onset of biventricular failure in the absence of identifiable causes.

**PHYSICAL SIGNS:**

a. The **GENERAL APPEARANCE** reveals a dyspneic 45-year-old man with peripheral edema.

b. The **JUGULAR VENOUS PULSE** mean pressure is elevated at 12 cm H$_2$O, reflecting the increased right atrial filling pressure caused by the poorly compliant right ventricle. The wave form shows a giant “a” wave from an increased right atrial contraction.

Proceed
c. The **CAROTID PULSE** is diminished due to the low stroke volume.

d. **PRECORDIAL MOVEMENTS** reveal sustained systolic impulses over both the right and the left ventricles with presystolic and early diastolic impulses that coincide with active and passive filling of the diseased ventricles.

Proceed
e. **CARDIAC AUSCULTATION** reveals normal splitting of the second heart sound with an increase in the pulmonic component reflecting pulmonary hypertension. In some patients, the second heart sound may be more widely split than normal with little respiratory motion. This occurs when the severely failing right ventricle cannot augment its stroke volume with inspiration. When left bundle branch block is present, paradoxical splitting of the second heart sound may occur.

Third and fourth heart sounds are present at the apex and lower left sternal edge. In the latter area they increase with inspiration, reflecting their right ventricular origin. The systolic murmur at the apex is due to mitral regurgitation secondary to abnormalities of the mitral apparatus. These findings are also well heard posterolateral to the mitral area over the enlarged left ventricle.

f. **PULMONARY AUSCULTATION** reveals absent breath sounds in the lower right posterior lung field and inspiratory crackles in the lower left posterior lung field. In all other lung fields, there are normal vesicular breath sounds.

**Proceed**
The **ELECTROCARDIOGRAM** shows an intraventricular conduction delay, left axis deviation and left atrial and ventricular hypertrophy.

The **CHEST X RAYS** show diffuse cardiomegaly with pulmonary vascular congestion, a “phantom tumor,” as well as a large pleural effusion in the lower right lung field.

**LABORATORY STUDIES** include the echocardiogram which shows a dilated poorly contracting left ventricle.

While invasive study is unnecessary in this patient (unless he becomes a heart transplant candidate), typical catheterization data show elevated filling pressures, a dilated poorly contracting left ventricle and mild mitral regurgitation with normal coronary arteries.

**TREATMENT** consists of restricted activity, low salt diet, vasodilators (especially ACE-inhibitors), beta-adrenergic blockers and aldosterone blocking agents, diuretics and digitalis.