1 Patient Study 1

Sequence of data presentation

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1 History 1965

This patient is a chronic alcoholic. He was told of a heart murmur at age 20 in 1939 and has had dyspnea on exertion since that time. He has had two admissions for heart failure, in 1962 and 1964.

2 Chest X ray 1965

![Chest X ray 1965](image)

Figure 1.1 Chest X ray 1965.
3 Cardiac catheterization 1966

![Cardiac catheterization data 1966](image)

**Figure 1.2** Cardiac catheterization data 1966. Numbers in brackets are mean pressures (mmHg), numbers in the denominators are end-diastolic pressures in the ventricles and diastolic pressures in the pulmonary artery and aorta.

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<th>Table 1.1 Additional cardiac catheterization data 1966</th>
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<td>LVH, left atrial enlargement, subaortic chamber, MR</td>
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4 ECG 1967

![ECG 1967](image)

**Figure 1.3** ECG 1967.

5 History 1966–1968

The patient was offered surgery in 1966 for hypertrophic subaortic stenosis but refused. He was treated with diuretics warfarin and digoxin, and advised to stop drinking alcohol. He was diagnosed as having schizophrenia in 1967.

The patient has had intermittent atrial fibrillation since 1966. In 1968 he returned to hospital with exertional dyspnea.
6 Chest X ray 1968

Figure 1.4 Chest X ray 1968.

7 Phonocardiogram 1968

Figure 1.5 Phonocardiogram of carotid pulse and apexcardiogram 1968. Carotid pulse: D = dicrotic notch; P = percussion wave; T = tidal wave. Apex cardiogram (ACG): rfp = rapid filling phase; sfp = slow filling phase; E&O correspond to onset of ejection & mitral valve opening respectively.

8 Physical examination 1968

BP 160/90, HR 100/min.
No increase in jugular venous pressure. Prominent ‘a’ wave in neck veins.
Pulsus bisferiens.
Apex was in the 6th interspace in the anterior axillary line. RV lift 1+. 
S1–1 decreased. S2 paradoxically split. S3 and S4 at apex.
Grade 4/6 systolic murmur with ejection/pansystolic quality, best heard at LLSB, radiating to apex and axilla.
The murmur increased in intensity with a valsalva maneuver.
No edema or clubbing.
A few rhonchi were heard in the lung fields.

9 **ECG 1969**

![ECG 1969](image)

*Figure 1.6 ECG 1969.*

10 **Chest X rays 1965–1969**

![Chest X rays 1965–1969](image)

*Figure 1.7 Chest X rays 1965–1969.*
The patient had recurrent heart failure and atrial fibrillation, and died suddenly in 1969.  

*What would you expect to find at autopsy?*

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### Answers and commentary

1. **History 1965**  
The patient had a heart murmur and heart failure, which could be due to valvular heart disease. Alcoholic cardiomyopathy might also account for his symptoms of heart failure.

2. **Chest X ray 1965**  
The heart is probably enlarged and there are increased lung markings. The entire lung fields are not fully seen for technical reasons.

3. **Cardiac catheterization 1966**  
This showed moderately severe pulmonary hypertension (PA mean of 35 mmHg), a systolic gradient of 87 mmHg between LV inflow and LV outflow tracts, and no gradient in systolic across the aortic valve. The cardiac index was normal. There was LVH, LAE, and mitral regurgitation. Coronary angiography was not available at this time.  
Impression: hypertrophic subaortic stenosis.

4. **ECG 1967**  
The ECG shows sinus rhythm. Rate 78/min. PR 0.16 s. QRS 0.09 s. QRS axis +50°, LVH with repolarization abnormalities and left atrial enlargement.

5. **History 1966–1968**  
The patient continues to have evidence of heart failure. As he had refused surgery he was treated with anti-congestive measures and anticoagulants.

6. **Chest X ray 1968**  
There is now cardiomegaly with enlargement of the proximal PAs and some pulmonary congestion, which would account for his dyspnea.

7. **Phonocardiogram 1968**  
The carotid pulse shows a rapid upstroke, a bisferiens pulse with the percussion (P) wave being much taller than the tidal (T) wave. There is a diamond-shaped systolic murmur recorded at the apex and an apical S3.  
A bisferiens pulse with a taller percussion wave than the tidal wave is characteristic of HCM. The other cause of a bisferiens pulse is seen in aortic regurgitation, but in that case the P wave and T wave are about equal.  
The presence of a systolic murmur rather than a diastolic murmur (of aortic regurgitation) makes it even more likely that HCM is the correct diagnosis.  
The rapid carotid upstroke is also characteristic of LV outflow obstruction rather than aortic stenosis. In aortic stenosis the carotid upstroke is delayed and is of decreased amplitude (pulsus parvus et tardus).  
The presence of an S3 is unusual in HCM, but in this case is probably due to LV dysfunction.

8. **Physical examination 1968**  
The history of alcoholism and heart failure could represent an alcoholic cardiomyopathy. The physical examination, however, is more in favor of HCM in view of the apical systolic murmur that increases with stage 2 valsalva maneuver and the bisferiens carotid pulse detected on phonocardiography.  
The presence of moderate cardiomegaly is unusual in HCM but may be seen in advanced cases.

9. **ECG 1969**  
Ventricular rate: 70–80/min. QRS 0.08 s. QRS axis +50°. Further ST depression and sagging noted due to LVH and digoxin effect.  
Atrial fibrillation is now seen.
10 Chest X rays 1965–1969
There is a progressive increase in cardiac size and pulmonary congestion.

11 Course 1969
The loss of the atrial contribution to cardiac output in a patient with a stiff LV (due to LVH) led to progressive heart failure. His sudden death in 1969 was probably due to a ventricular arrhythmia. At autopsy the heart weighed 850 g. He had HCM with a dilated LA full of thrombi. There was evidence of peripheral emboli (kidneys, left temporal area), possibly because he was not taking anticoagulants on a regular basis (alcoholism, schizophrenia). There was pulmonary congestion but no pulmonary embolism. There was no significant coronary artery disease.

Key points
1. The presence of a rapid carotid upstroke and an aortic systolic murmur that increases during the valsalva maneuver is suggestive of HCM.
2. The loss of the atrial contribution to cardiac output (due to atrial fibrillation) may precipitate LV failure.
3. Patients with HCM may die suddenly from ventricular arrhythmias.
4. Anticoagulants are indicated in patients with atrial fibrillation, but may not be advisable if the patient is an alcoholic and has a mental disorder.

Further reading