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A case of the Eisenmenger Syndrome

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INTRODUCTION

In 1897 Eisenmenger described a case of a 32 year old man with cyanosis and exercise intolerance. Congestive heart failure was a preterminal event and death followed an episode of haemoptysis.

Post-mortem examination showed a large defect in the membranous septum, right ventricular enlargement and overriding oarta. More recently it has been appreciated that the position of the aorta is irrelevant to the diagnosis of the Eisenmenger complex.

Wood pointed out that development of pulmonary hypertension to systemic levels with high pulmonary vascular resistance could result from a variety of intracardiac defects and suggested that the term "Eisenmenger Syndrome" be used to include these entities.

The definition of Eisenmenger complex is defined by Wood (1968) as "Pulmonary hypertension at systemic level, due to a high pulmonary vascular resistance (over 800 dynes Sec/Cm) with reversed or bidirectional shunt through a large ventricular septal defect.

The phrase "Eisenmenger reaction" is used synonymously with Eisenmenger Syndrome.

A 58 year old Zambian male patient was admitted to the acute medical admission ward of the University Teaching Hospital, Lusaka, Zambia on 15th March, 1975. He gave a two week history of palpitations, breathlessness on mild exertion and pain in the abdomen.

Also he gave a one week's history of fullness of the abdomen and loss of appetite.

He had a mild non-productive cough. He had been well until two weeks before admission.

There was no history of similar episodes before and also no history of recurrent sore throat or joint pains. There was no history of haemoptysis, chest pains or syncope.

On examination his general condition was fair though he looked ill. The jugular venous pressure was elevated to about 8cm but had a normal pulse contour. It was 84/min and fibrillating and the B.P. 110/50. Apex beat was in the 7th intercostal space just outside the midclavicular line. There was a systolic thrill best felt at the 4th left sternal border. No heaves were palpable.

Heart sounds were normal except the pulmonary second sound which was loud. There was a grade IV pansystolic murmur best heard at the 4th left sternal border. The same murmur was also heard at the apex and was conducted well to the axilla. A grade II mid-diastolic murmur was heard in the mitral area.

He also had bilateral basal crepitations. Breath sounds were vesicular.

The liver was palpable 5cm below the right costal margin. It was smooth and tender. The spleen was not palpable. Other systems were normal.

The E.C.G. showed atrial fibrillation, a right bundle branch block and right ventricular hypertrophy and strain (Fig I).

Chest X-ray (Fig.II) showed very marked dilation of the main pulmonary artery and of the left and right main pulmonary arteries with considerable periphral pulmonary oligaemia. No evidence of left atrial or left ventricular enlargement was seen. A conclusion of severe Pulmonary Hypertension was made.

Angiocardiography was not very successful due to technical difficulties, though a catheter was passed into the right atrium and pressure injection of contrast media into right atrium done. This showed a jet of contrast flowing from right atrium into left atrium demonstrating the presence of an atrial septal defect.

DISCUSSION

The clinical presentation, electro-cardiogram and chest radiograph offer little help in determining the site of the shunt. Cardiac catheterisation assists in localising the lesion though a second defect may be present and remain undemonstrated. The late onset of symptoms suggests that shunt is truly at the atrial level.

MANAGEMENT

Surgical correction of the underlying defect is the treatment of choice provided the pulmonary vascular bed is reactive.

Surgery is by correction of the defect or pulmonary arterial banding in the patient whose level of total pulmonary resistance falls to normal or near normal with totazoline. Unfortunately we were unable to do any of these procedures. Congestive cardiac failure was controlled with digitalis and diuretics.

FIG. I

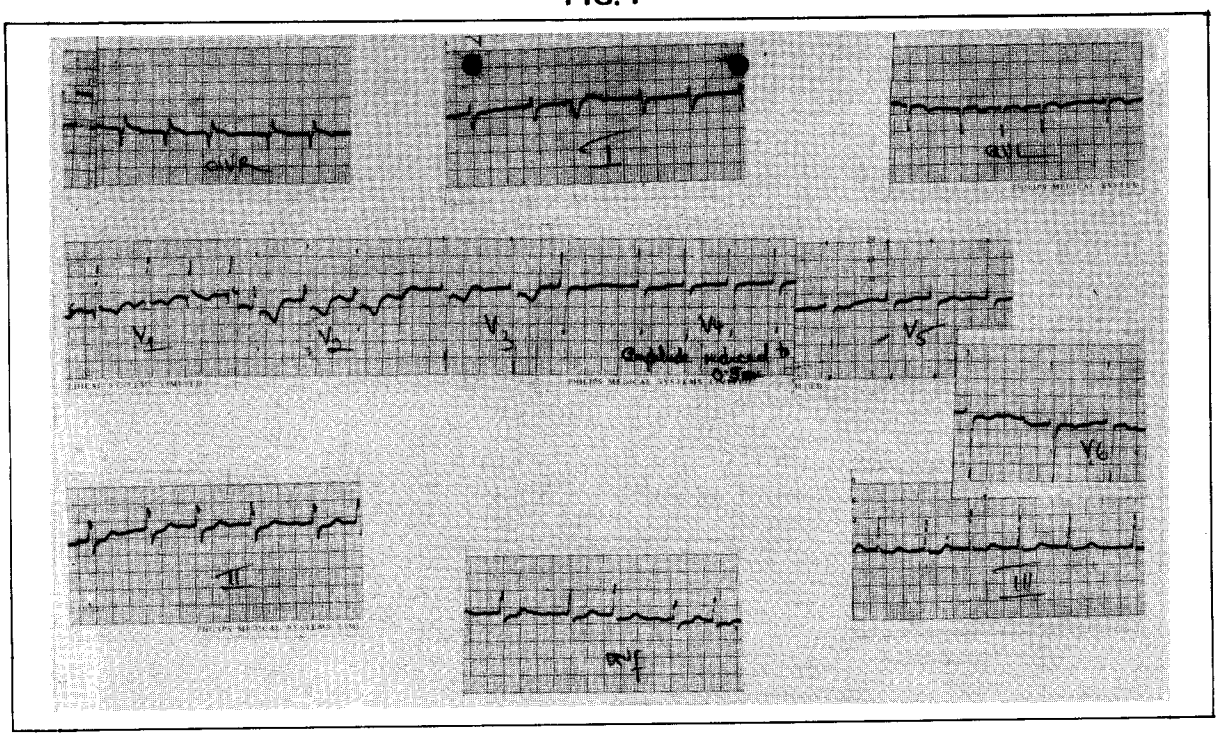
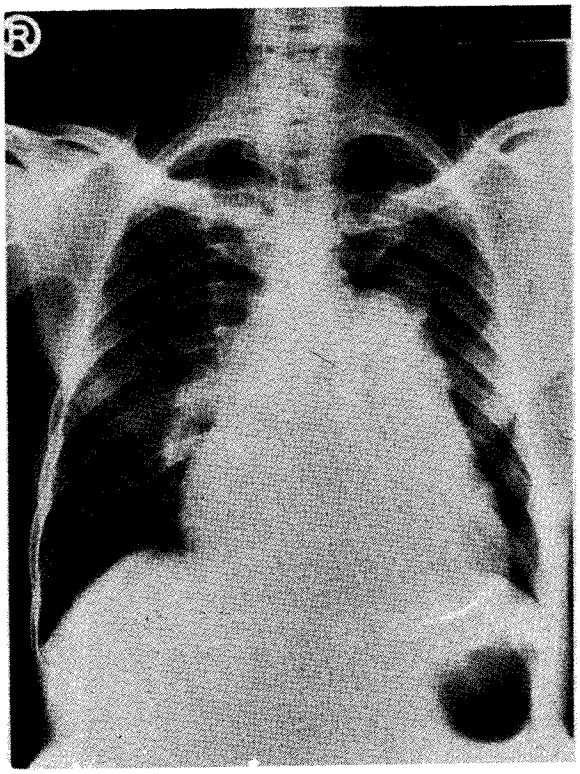


FIG. II



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REFERENCES

Eisenmenger, V. (1897) "Die angeborener Defecte der kammerscheidewand des Herzens." Z. Klin. Med. 32, supp: 1.

Wood, P. (1968) Diseases of the Heart and circulation Eyre & Spottiswoode, London.