PRIMARY PULMONARY HYPERTENSION

Report of one patient aged 23

By J. H. SWALLOW, M.B., B.CHIR., R.A.M.C.

This interesting though rare condition is now well recognized. Previous reports, with a clinical diagnosis confirmed by cardiac catheterization and oxygen studies are those of Nellen (one), Chapman (two), Johnson (one), Whittenburg (one), Dresdale (three), Wood (eight) and Cutler (three). The following case was diagnosed at an apparently much earlier stage, as judged by heart size and duration of symptoms, than has been previously recorded. However, the low arterial oxygen saturation and severe exertional dyspnoea are features of a more advanced stage of the disease.

Case Report

A 23-year-old man was admitted to hospital on February 24, 1955. There was nothing relevant in his past history or family history. During his National Service, from 1949-51, he spent some 18 months in Egypt, but there was nothing to suggest he had contracted schistosomiasis. He had never been exposed to irritant dusts.

His first symptom was recurrent small haemoptyses in March 1954, at which time a chest X-ray was normal. He remained fit until Christmas 1954, when he had further small haemoptyses, following which he became breathless on exertion. This gradually worsened and by the time of admission he could only walk about 50 yd. on the level at a moderate pace. When he walked he experienced a gripping umbilical pain which disappeared within a few seconds of stopping, but it was the shortness of breath rather than the pain that stopped him. He was not short of breath at rest and slept with only one pillow. There was no radiation of the pain and no history of chronic cough or ankle swelling.

He was rather thin. There was slight clubbing of the finger and toe nails. The hands were blue and cold. On exertion he assumed a ghastly pallor and his lips and tongue became markedly cyanosed. The radial pulses were equal, small and regular, and he had a resting tachycardia at 100/110. B.P., 125/85. Internal jugular venous pressure was raised 2 in. above the sternal angle; there was a large 'a' wave. Marked pulsation was seen in the third and fourth interspaces and on palpa-

tion there was a corresponding right ventricular heave. The apex beat was not displaced and the impulse right ventricular in type. The pulmonary artery could be felt pulsating in the second left interspace. The second sound in the pulmonary area was widely split with accentuation of the pulmonary component, and the splitting did not alter with the phases of respiration. There were no murmurs, no liver enlargement or pulsation and no other abnormal signs.

P/A chest X-ray showed a normal sized heart with a slight prominence of the pulmonary arteries at the hila, but in comparison with the film of March 1954 there had been a slight but definite increase in the transverse diameter of the heart. The peripheral lung fields looked somewhat oligæmic. There was no abnormal pulsation on screening and no evidence of right ventricular enlargement. E.C.G. showed only right ventricular preponderance. Haemoglobin, 107 per cent. (on two occasions). W.B.C.s, 9,000; normal differential; no eosinophilia. W.R., negative. Sputum, no A./A.F.B. on film or culture.

On March 21, 1955, he was transferred to the Middlesex Hospital for further investigation under the care of Dr. Evan Bedford. Cardiac catheterization gave the following results:

	Pressure (mm.Hg)	Oxygen Saturation (Per cent.)
Main pulmonary artery	86/40	53
Right ventricle	90/0	55 57
Right auricle	12/4	55 53
Superior vena cava	—	53
Brachial artery	120/84	82

Angiocardiography showed no evidence of shunt; the peripheral lung vessels filled poorly. Phonocardiograph recorded the wide splitting of the second heart sound with a delayed pulmonary component.

He was treated with 'Priscol' up to 50 mg. six-hourly and his exercise tolerance improved. However, recatheterization after two months showed the pulmonary artery pressure to be slightly higher at 100/46 mm. Hg.

Comment

Both central and peripheral factors here contribute to the cyanosis. The arterial oxygen saturation is remarkably low at 82 per cent., and the figures of this order have only previously been recorded in the presence of advanced heart failure and gross cardiac enlargement. It has been suggested that the cause of the central cyanosis is the presence of small pulmonary arteriovenous shunts such as have been demonstrated by Farber in Eisenmenger's complex. Ordway noted in two patients that the arterial oxygen saturation was not raised by breathing pure oxygen, but there are no other reports of respiratory function studies in cases of primary pulmonary hypertension. The arteriovenous oxygen difference of 5.7 ml. per cent. is raised and results from poor peripheral circulation and polycythaemia.

Haemoptysis, not a constant feature of this condition, was the presenting symptom here, and could well result from the rupture of small pulmonary arteriovenous shunts.

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