WINDOW TYPE OF DUCTUS ARTERIOSUS WITH SHUNT REVERSAL

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Patent ductus arteriosus has been classified by Edwards (1960) into three anatomical types: (1) cylindrical, (2) funnel, and (3) window types. The window type is least common, and in fact is rarely encountered (Taussig, 1960). It is seen more frequently in adults and Keys and Shapiro (1943) reported a frequency of 17 per cent among 60 persons of 17 years of age or older with patent ductus arteriosus. A case is described in an adult who also had severe pulmonary hypertension with right to left shunting thus satisfying the criteria for it to be called Eisenmenger Syndrome (Wood, 1958).

CASE REPORT

A 31-year old Chinese housewife was admitted into hospital with a history of palpitation for 3 years and breathlessness on exertion for one year. In the three months prior to admission her symptoms became worse and she noticed swelling of her abdomen and legs in addition to worsening of her effort dyspnoea. There were no other significant symptoms.

Physical examination revealed a thin wasted woman with mild central cyanosis. There was no clubbing of fingers or toes and no differential cyanosis. Jugular pressure was elevated to 8 cm. above the sternal angle with a dominant "a" wave. Pulse rate was regular at 96/min. and B.P. 140/80. The lungs were clear but the apex beat was displaced to the left fifth intercostal space just lateral to the mid-clavicular line. The cardiac impulse was that of right ventricular enlargement and a long diastolic thrill was palpable in the left parasternal region. A fourth heart sound was heard in addition to a long early diastolic murmur at the left sternal edge. A soft ejection systolic murmur was audible in the pulmonary area. Pulmonary second sound appeared normally split. The liver was enlarged to 3 cm. below the right costal margin but there was no peripheral oedema.

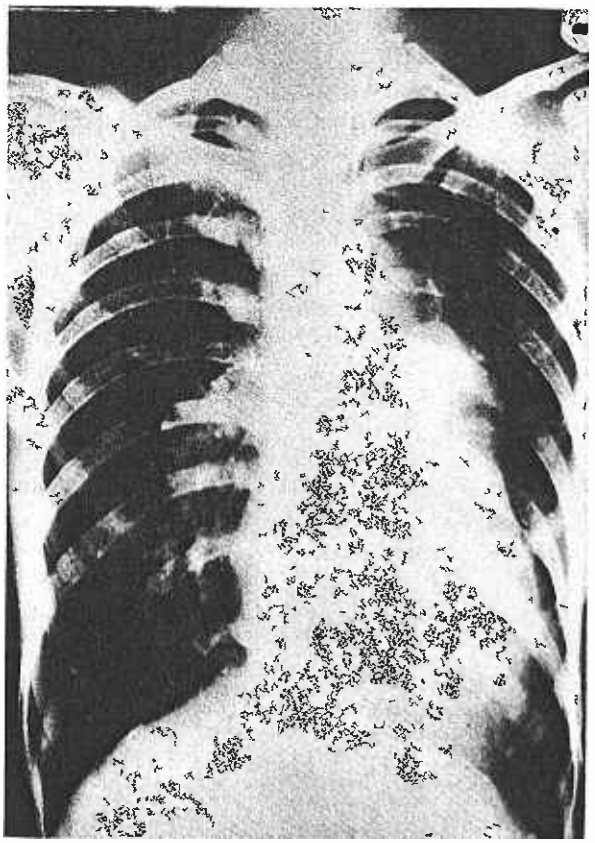

Fig. 1. Chest X-ray showing enlarged heart with dilated pulmonary vessels radiating from the hila. The pulmonary arteries are markedly enlarged with peripheral pruning of the vessels. Calcification of the aortic wall in the position of a ductus is seen.

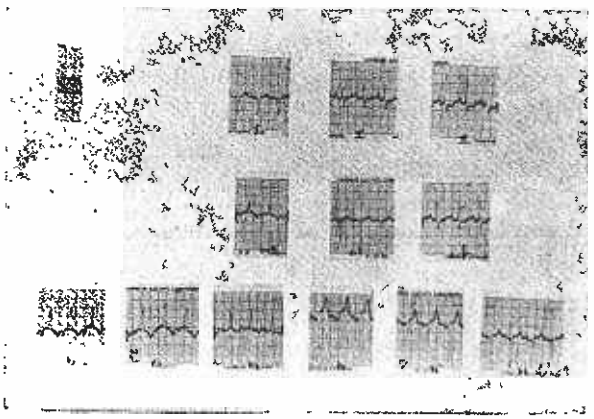

Fig. 2. E.C.G. showing marked right axis and right ventricular hypertrophy.

Chest X-ray (Fig. 1) showed enlarged cardiac silhouette with very dilated pulmonary vessels radiating from the hila and peripheral pruning of the vessels. Calcification of the aorta in the position of a ductus was also seen. Electrocardiography showed sinus rhythm with marked right axis and right ventricular hypertrophy (Fig. 2). Haemoglobin was 16.0 gm. %. Venous angiogram showed marked dilatation of right atrium, right ventricle and pulmonary artery but no evidence of right to left shunt was seen (Fig. 3).

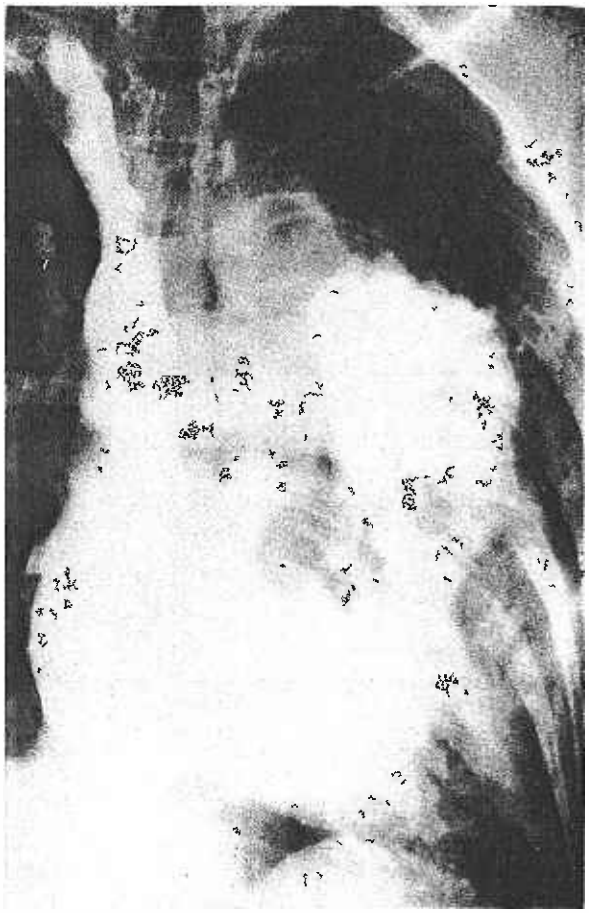

Fig. 3. Venous angiocardiogram showing marked dilatation of right atrium, right ventricle and pulmonary artery but no evidence of right to left shunt.

She was diagnosed as having severe pulmonary hypertension with pulmonary incompetence and a right to left shunt at either aorto-pulmonary, atrial or ventricular level. A formal cardiac catheterisation with oxygen saturation studies was not performed because of lack of facilities. The patient gradually deteriorated with intractable heart failure and increasing cyanosis until death supervened five weeks after admission.

POST-MORTEM FINDINGS

At autopsy, the main findings were in the cardiovascular system. The heart was moderately enlarged. The left ventricle was of normal size, its wall measuring 10 mm. in thickness. The right ventricle was dilated and its wall was markedly hypertrophied, measuring 12 mm. in thickness (Fig. 4). No valvular defects were detected. The pulmonary orifice, however, was increased in size, measuring 9.5 cm. in circumference.

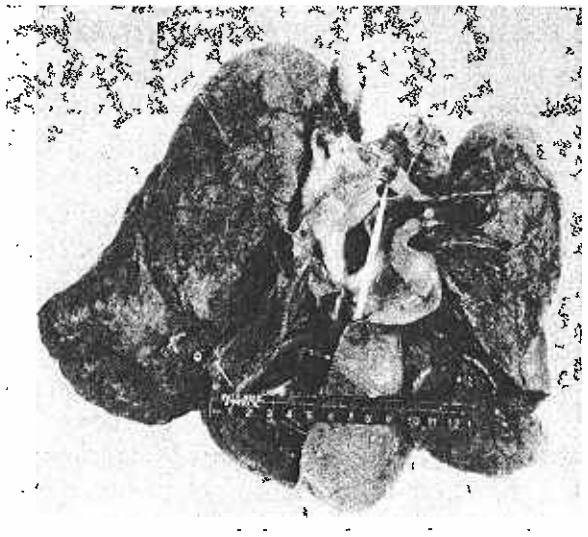

Fig. 4. The heart is enlarged. The right ventricle, which has been opened into, shows marked hypertrophy (thickness of ventricular wall = 12 mm.). The "window" ductus, situated at the bifurcation of the pulmonary trunk, is shown by the pointer.

The pulmonary trunk was dilated (Fig. 4). Small scattered patches of atheromatous thickening were seen on the endothelial surface of the pulmonary trunk and its branches. At its bifurcation, there was a large circular defect measuring 1 cm. in diameter, with a smooth, thickened, rounded, and partly calcified margin. It opened into the aorta just beyond the origin of the left subclavian artery. No length or neck of a ductus could be demonstrated.

The liver weighed 840 gm., was congested, and on cut section showed a "nut-meg" appearance. The spleen weighed 180 gm. and was congested and firm. No other abnormal findings were detected on gross examination.

Post-mortem angiographic studies of the pulmonary arterial system (after injection of "micropaque" into the main pulmonary arteries) revealed the changes expected to be found in long-standing pulmonary hypertension (Fig. 5). The large elastic arteries were seen to be dilated. The muscular arteries were markedly narrowed and the peripheral arterioles were not visualised.

Histological examination revealed the changes of secondary pulmonary hypertension in the

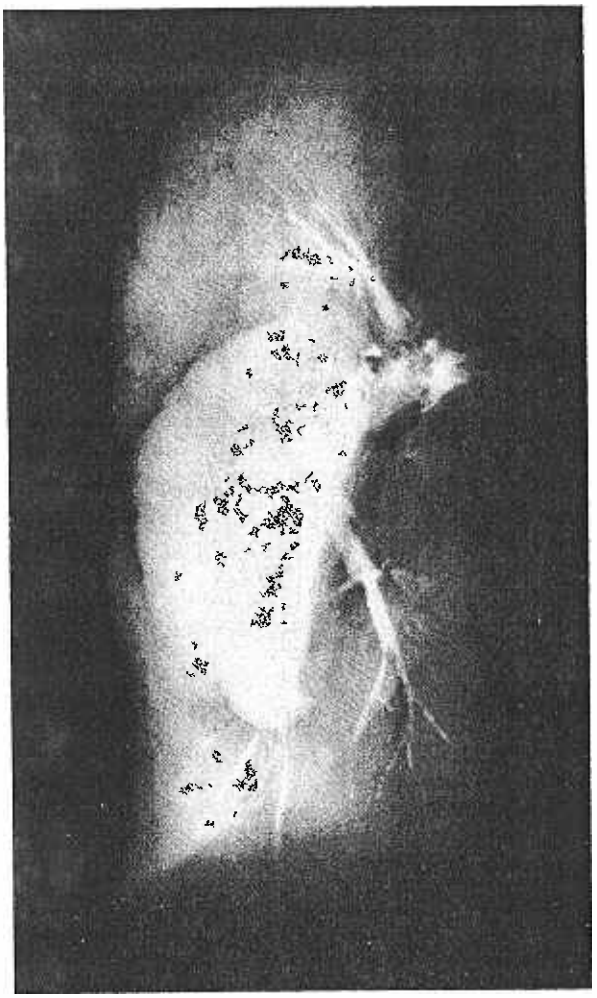

Fig. 5. Post-mortem angiogram of the right lung (antero-posterior view). The large elastic arteries are dilated. The smaller muscular arteries are seen to be narrowed, while the peripheral arterioles are not visualised.

lungs (Fig. 6) and of chronic venous congestion in the liver and spleen. There were no other significant histopathological findings.

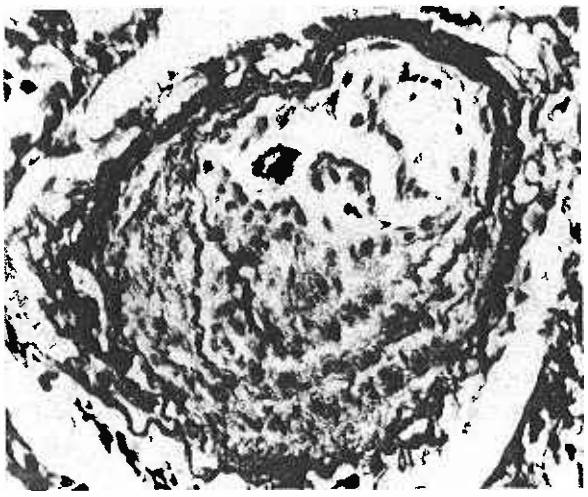

Fig. 6. A small pulmonary muscular artery showing wrinkling and reduplication of the internal elastic lamina and fibro-elastic thickening of the intima (Elastic stain X 460).

DISCUSSION

The diagnosis of Eisenmenger syndrome with severe pulmonary hypertension and right to left shunt was made ante-mortem but the actual site of the shunt reversal could not be determined despite angiography.

In the presence of severe pulmonary hypertension and early shunt reversal, the classical signs of patent ductus arteriosus would be absent (Evans and Short, 1958). In this patient, there was no collapsing pulse nor a continuous murmur. There was a diastolic murmur attributable to pulmonary incompetence caused by dilatation of the pulmonary artery. In these cases, differential cyanosis may be obvious if reverse shunting occurs through the ductus, since deoxygenated blood from the pulmonary trunk would be carried distally down the aorta causing cyanosis of the lower extremities and clubbing of the toes, while the upper limbs remain normal.

In this patient some radiological calcification at the site of a ductus suggested the level of the shunt.

The patient eventually succumbed from intractable heart failure. At autopsy, the presence of congestion of liver and spleen, right ventricular hypertrophy and atherosclerotic narrowing of the pulmonary arterial vasculature, indicated long-standing pulmonary hypertension. A large window defect between the main pulmonary artery and aorta at the usual site of a ductus was located. This appeared to be the site of a large left to right shunt throughout life until terminal stages when, because of the large volume of flow through the lungs, pulmonary arteriosclerosis and narrowing of muscular arteries developed. Thus as pulmonary arterial resistance started to exceed systemic resistance, reversal of shunt began to occur causing central cyanosis and ultimately right-sided heart failure.

In the window type of patent ductus, the defect is located at the usual site of a classical ductus, *i.e.*, at the bifurcation of the pulmonary trunk and opening into the aorta just beyond the origin of the left subclavian artery (Taussig, 1960). This entity is therefore distinct from the aorto-pulmonary window which is a communication between the two great vessels at their origin and which results from a different embryological anomaly. The window ductus differs from the usual type of ductus in that the lumen of the pulmonary artery communicates directly with that of the aorta, so that the ductus has no reco-

gnisable length (Edwards, 1960). In fact the ductus is absent and there is a window where the two great vessels lie in apposition at the distal end of the pulmonary trunk (Sherman, 1963). The window ductus is found more frequently in adults possibly because it develops gradually and results from the pulmonary side of the typical ductus being gradually effaced by the dilatation of the pulmonary artery (Edwards, 1960).

The window ductus is an important entity to the surgeon because of the obvious technical difficulty of trying to dissect, ligate and divide the communication which has virtually no neck. Calcification of the great vessels would make dissection even more hazardous. Operation, therefore, should always be performed with pump by-pass to allow better vision and access to the surgeon. Unfortunately window ductus cannot be diagnosed with any certainty preoperatively but one should be alert to its possibility in the presence of a large left to right shunt, and dilated or calcified aorta or pulmonary artery at the site of a ductus, occurring in an adult.

Surgery in this patient could not be offered because shunt reversal had already started to occur and the mortality would be forbidding. (Campbell, 1955., Reid et al., 1964., Ellis et al., 1956).

SUMMARY

A case of window type of patent ductus arteriosus with severe pulmonary hypertension and shunt reversal is described. The difficulties in diagnosis and management are discussed.

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