CASE REPORTS

PATENT DUCTUS ARTERIOSUS WITH PROGRESSIVE DILATATION OF THE PULMONARY ARTERY

BY

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A patient with patent ductus arteriosus was observed over a period of years to have progressive increase in size of the pulmonary artery. This was accompanied by the development of a pulmonary diastolic murmur and thrill, and of electrocardiographic changes of right ventricular hypertrophy with bundle-branch block. She died, aged 39 years, of pulmonary embolism.

Case History

The patient was born in 1913. A heart murmur was noticed in childhood, but she apparently led a normally active life and had no symptoms as a child.

At age 23 she had pneumonia, and after recovery became aware for the first time of dyspnoea on effort. A year later she was admitted to the Bristol General Hospital under Professor C. B. Perry as a possible case of bacterial endocarditis. No proof of this was found. She was then of average physique and normal colour, with no finger clubbing and no venous congestion or oedema. The apex beat was in the fifth left intercostal space beyond the mid-clavicular line, and the impulse was forceful in character. A systolic thrill was felt at the pulmonary area, where there was a continuous murmur. B.P. 140/90. A diagnosis of uninfected patent ductus arteriosus was made. The electrocardiogram (Fig. 1) showed evidence of left ventricular hypertrophy. In the following year she bore a child without difficulty; the child's heart was normal.

When the patient was re-examined at the age of 28 years the only change noted was that a diastolic murmur was now heard to the left of the sternum. X-ray showed enlargement of the heart and great prominence of the pulmonary artery, which was seen to pulsate vigorously on fluoroscopy. Two years later a second pregnancy was terminated at the fourth month by hysterotomy. In the next three years her condition remained unchanged. Her weight was 102 lb. and height 62 inches. The X-ray appearances at age 32 are shown in Fig. 2.

The diastolic murmur to the left of the sternum was noted to be louder, and accompanied by a thrill, when the patient was seen again at the age of 35. X-ray showed the main pulmonary artery to be larger than it was three years previously. B.P. 140/60. The electrocardiogram now showed right axis deviation (Fig. 1).

In 1952, when she was 39 years of age, the patient attended hospital for the last time. Dyspnoea on effort and fatigue towards evening were troublesome but not incapacitating. Her colour was normal, with no cyanosis or finger clubbing, and there were no signs of congestive failure. The apex beat was in the fifth interspace, 2.5 cm. beyond the mid-clavicular line, and the impulse was abrupt and forcible. A coarse diastolic thrill was felt to the left of the sternum, most obviously in the third left interspace. The first sound was loud at the apex, and the second sound loud at the pulmonary area. The continuous murmur was still present at the pulmonary area, and the diastolic murmur to the left of the sternum was rasping. The blood pressure was 140/70. X-ray and fluoroscopy showed that the heart was considerably enlarged; the aorta and main pulmonary artery were dilated and exhibited vigorous pulsation, which was also obvious in the chief branches of the pulmonary arteries. The main pulmonary artery was even more prominent than before (Fig. 2). The electrocardiogram showed right bundle-branch block and right ventricular hypertrophy (Fig. 1).

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FIG. 1.—Electrocardiograms showing change from left ventricular hypertrophy at age 24 to right bundle-branch block and right ventricular hypertrophy at age 39. The unmarked lead in the upper right corner is V1.

FIG. 2.—Teleradiograms showing the increase in prominence in the main pulmonary artery and aorta between the ages of 32 years (A) and 39 years (B). There is little accompanying change in the heart size or the heaviness of the pulmonary vascular shadows.

Three months later the patient was found dead in bed at home, having apparently been in her usual health the night before.

Necropsy Findings

Examination was limited to the thorax and abdominal cavity. The external appearances were normal. *Heart.* Pericardium normal, no effusion. Heart weight 570 g. There was well-marked hypertrophy of the right (1.1 cm.) and left (2.0 cm.) ventricular walls, and both chambers were dilated. A well-defined

area of endocardial thickening was seen below the cusps of the pulmonary valve (Fig. 3). There was a little generalized thickening of the endocardium of the left atrium, and the cusps of the mitral, aortic and pulmonary valves were a little thickened, but with no naked-eye evidence of rheumatism. The auricles contained no clot. The circumference of the aorta was 6.3 cm. at 3 cm. above the valve, and 7 cm. at the valve orifice (normal 7.7 to 8.0 cm.). There was a localized dilatation of the aorta at the site of the *patent ductus*. The ductus orifice measured 0.8 × 0.5 cm.; the aorta and pulmonary artery were in close contact

FIG. 3.—Pulmonary valve cusp and the thickened patch of endocardium immediately below, caused by regurgitation.

at this point, and the ductus had no appreciable length. The main pulmonary artery was much dilated, and its circumference at the valve was 10.3 cm. compared with the normal 8.5 to 9.0 cm. (Ross, 1948). There was no endocardial thickening either on the aortic or pulmonary arterial walls opposite the opening of the ductus. The right pulmonary artery was blocked by a large embolus, the origin of which was not found. The larger branches of the pulmonary artery were dilated, and their walls thickened, showing small scattered patches of atheroma.

Lungs. Pleural cavities normal. There was some brown induration at the bases of the lungs, but no other abnormality.

Abdomen. The spleen was nearly twice the normal size, and other organs were normal.

Histology. Left auricle: hypertrophy and endocardial fibrosis. The mitral valve showed irregular

fibrous thickening. The cusps of the pulmonary valve were covered with a thin layer of organizing fibrin and irregularly scarred. The left and right ventricles were hypertrophied, with no evidence of rheumatism.

In the lungs there were well-marked changes of pulmonary hypertension; the small arteries showed medial hypertrophy and fibro-elastic intimal thickening (Fig. 4).

FIG. 4.—Histology of the small arteries of the lung, showing (A) medial hypertrophy and intimal thickening, (B) increase in the elastic fibres in the intima and media.

Interpretation

The finding of a characteristic Gibson murmur on every examination between the ages of 23 and 39 years is evidence that the shunt was in the usual direction, i.e. from the aorta into the pulmonary artery. It appears to have been a large shunt in view of the wide systemic pulse pressure, the hypertrophy of the left ventricle, the dilatation and pulsation of the pulmonary artery, and the size of the ductus at necropsy. The patch of endocardial thickening below the pulmonary valve cusps is evidence of gross regurgitation through the valve. The histology of the pulmonary arteries indicates the presence of pulmonary hypertension; right ventricular hypertrophy would well be accounted for by the combination of pulmonary hypertension and pulmonary regurgitation.

The unusual features of this case are the development of pulmonary regurgitation, becoming more obvious as the pulmonary artery increased in size, and the electrocardiographic change from the pattern of left ventricular hypertrophy to that of right ventricular hypertrophy with right bundle-branch block.

Discussion

The association of pulmonary hypertension with patent ductus is well known (Cosh, 1953; Dammann *et al.*, 1953; Lukas *et al.*, 1954; Whitaker *et al.*, 1955). In the most severe type the pulmonary vascular resistance is so great that the pulmonary arterial pressure may roughly equal the aortic, so that a continuous murmur is not heard. In the mildest type the pulmonary resistance is of normal degree, and the pressure is raised merely passively by the presence of an increased pulmonary blood flow. This patient appears to occupy an intermediate position between these types. The histology of the pulmonary vessels suggests a considerable increase in resistance, so

that, in the presence of a large left-to-right shunt the pulmonary arterial pressure would be high; yet the presence of a continuous murmur indicates that the pulmonary arterial pressure remained lower than the aortic.

It is not known whether the pulmonary arterial pressure remained at the same level over the last decade of life, or whether it progressively increased, as appears to have happened in a similar case described by Harris (1955). Although the electrocardiographic changes suggest the latter, they may only reflect the development of pulmonary regurgitation and the resulting increased strain on the right ventricle. Presumably after many years the pulmonary artery became sufficiently dilated to give rise to valvular incompetence. The resulting big pulse pressure in the pulmonary artery may have been a factor in aggravating the dilatation and incompetence.

Dilatation of the pulmonary artery is a common finding in patients with patent ductus arteriosus. Progressive dilatation is unusual, and was not found in a review of 73 cases, many of whom were followed into adult life (Cosh, 1956). One of the cases of patent ductus reviewed by Campbell (1955, Case 19) had an increase in the size of the heart and main pulmonary artery over the course of ten years, when pulmonary hypertension had caused reversal of the flow through the ductus; she died after ligation of the ductus. Great dilatation, with rupture of the pulmonary artery into the pericardium, has been reported by Durno and Langdon Brown (1908) and by Moench (1924). Saccular aneurysms of the pulmonary artery accompanying patent ductus have been described by d'Aunoy and von Haam (1934).

Operative closure of the ductus in this patient would clearly have been hazardous, although it might well have been easier earlier in life before such great dilatation of the pulmonary artery occurred.

I wish to thank Professor C. Bruce Perry for permission to report this case, and Dr. O. C. Lloyd, Lecturer in Pathology, University of Bristol, for his report of the necropsy.

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