CASE REPORTS

DIRECT COMMUNICATION OF A PULMONARY ARTERY WITH THE LEFT ATRIUM

BY

D. VEREL, R. G. GRAINGER, AND D. G. TAYLOR

From The Regional Cardiovascular Centre, City General Hospital, Sheffield 5

Pulmonary arteriovenous fistulae are not uncommon. They are usually congenital anomalies in which the pulmonary artery is connected to the left atrium through a leash of arteriovenous channels enmeshed in functionless lung tissue. Rarely a large communication connecting the pulmonary artery directly to the left atrium is found. Kroeker *et al.* reviewing earlier publications in 1963 reported one case and could find reference to only four other similar anomalies. A sixth case was recorded by Taussig (1960). Three of those reported were found at necropsy and three were operated on, the cases reported by Friedlich, Bing, and Blount (1950) and Taussig (1960) being those successfully treated. We report here a further patient with this condition in whom surgical treatment successfully relieved the patient's cyanosis.

Case Report

The patient was referred to the department in 1959 at the age of 5 years. He was the second of four children, the sibs being healthy, and his parents were aged 27 and 24 at the time he came under observation. He was born at home and admitted to hospital elsewhere, aged 12 hours, with cyanosis, congenital heart disease being diagnosed. He had grown normally and compared with his sibs was not stunted. He had always been cyanosed, however, and soon became breathless on exertion, having to rest while the others continued to play.

On examination there was obvious central cyanosis with moderate clubbing. The cyanosis did not lessen at rest. The apex beat was normal and there were no abnormal heart sounds. The electrocardiogram was within normal limits and chest radiograph was reported as normal. Fluoroscopy was not undertaken at this attendance. He was reviewed as an out-patient until December 1962, and during this time remained cyanosed with moderate dyspnoea; no syncopal attacks or squatting were reported. He had not been subject to inter-current infections. On admission in 1963 he was found to be moderately cyanosed with clubbing of the fingers and toes, and a normal venous pressure. The femoral pulses were easily felt. The apex beat was in the fifth space of the mid-clavicular line. There was a moderate left parasternal lift. The first sound was normal. The second sound was closely split and a third sound was audible at the sternal border in the fourth left space. A soft systolic murmur was heard in the aortic area and some observers could hear a soft continuous hum in the interscapular region. Investigations showed a haemoglobin of 18.6 g. per 100 ml. The electrocardiogram was normal. Right-sided cardiac catheterization showed no evidence of a left-to-right shunt. The pulmonary artery pressure was 20/10 mm. Hg; right ventricular pressure 20/0. Chest radiograph showed a rounded opacity overlying the right hilum (Fig. 1). Angiocardiogram showed that there was a large vessel in the posterior part of the right chest originating in the pulmonary artery and entering a circular aneurysm which connected directly to the left atrium, thereby bypassing the lungs (Fig. 2A and B). The arterial saturation at the time of catheterization was 81 per cent, indicating that approximately one-third of the cardiac output was bypassing the lung and passing directly through the shunt to the left atrium. In view of the extremely poor prognosis indicated by the reported cases and the boy's obvious disability, surgical treatment was advised.

An incision was made through the bed of the right sixth rib. A large aneurysm at the posterior aspect of the right lower lobe and apparently entirely separate from it was exposed. When it was mobilized, it became clear that it consisted of a diverticulum of the left atrium which opened into a single large branch of the pul-

FIG. 1.—Postero-anterior chest film showing rounded opacity overlying the right hilum. Film taken in 1963.

(A)

(B)

FIG. 2.—(A) Angiocardiogram illustrating filling of the large sac directly from the right pulmonary artery. On the lateral film. (B) The sac is seen to empty directly into the posterior aspect of the left atrium.

monary artery. The aneurysm was invaginated and the site of the opening into the pulmonary artery confirmed. The feeding branch was clamped and the inside of the aneurysm digitally examined, the finger being enclosed in a purse-string. It was confirmed that there was a very large opening into the left atrium, apparently distinct from both inferior and superior pulmonary veins. Accordingly clamps were applied and the diverticulum and feeding vessel were separately sutured. A systolic thrill, which had been palpable over the feeding branch of the pulmonary artery, was completely abolished.

After operation the child was no longer cyanosed and he made an uneventful recovery. His exercise tolerance over the ensuing months increased and his performance now exceeds that of which he was capable before the operation. The clubbing of the nails has disappeared. The hæmoglobin is now 13.8 g./100 ml.

Discussion

The patients previously described with this condition have fared badly in the absence of treatment. Two have been successfully operated on and the remainder have either died at attempted surgery or been found at necropsy to have the anomaly. The risks of leaving arteriovenous aneurysms untreated are discussed by Lucas, Lund, and Edwards (1961) who described a patient in whom fatal cerebral abscess occurred. Despite the difficulty encountered in the surgical correction in the patient we describe there seems no reasonable doubt that such treatment should be attempted in these patients.

The embryological origin of this anomaly has not been established with certainty. The anatomical features found in our case agree precisely with those described by others; the anomaly seems to be a clear-cut developmental derangement which is not similar to that in the commonly occurring arteriovenous aneurysm in which a multiple leash of vessels is found in the pulmonary tissue. Lucas *et al.* found that the pulmonary lobes were abnormal in their case and suggested that the vessel represented a normal pulmonary artery to left atrial connexion in which the lung tissue had not formed. In our patient, however, the remaining pulmonary veins were present and normal and the right lung had a normal division into three lobes.

Summary

A child with a direct communication between the pulmonary artery and left atrium is described. He was successfully treated surgically. In view of the poor prognosis of such cases it is desirable that they should be recognized and treated.

References

- Friedlich, A., Bing, R. J., and Blount, S. G., Jr. (1950). Physiological studies in congenital heart disease. IX. Circulatory dynamics in the anomalies of venous return to the heart including pulmonary arteriovenous fistula. *Bull. Johns Hopk. Hosp.*, **86**, 20.
- Kroeker, E. J., Adams, H. D., Leon, A. S., and Pouget, J. M. (1963). Congenital communication between a pulmonary artery and the left atrium. Physiologic observations and review of the literature. *Amer. J. Med.*, **34**, 721.
- Lucas, R. V., Jr., Lund, G. W., and Edwards, J. E. (1961). Direct communication of a pulmonary artery with the left atrium. *Circulation*, **24**, 1409.
- Taussig, H. B. (1960). *Congenital Malformations of the Heart*, 2nd ed., Vol. 2, p. 376. Harvard University Press Cambridge, Massachusetts.