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PULMONARY VALVULAR STENOSIS WITH INTACT VENTRICULAR SEPTUM AND RIGHT AORTIC ARCH

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Pulmonary valvular stenosis with right-sided aortic arch and an intact ventricular septum is a rare anomaly. Brock and Campbell (1950) and Abrahams and Wood (1951) stated that the aortic arch was left-sided in simple pulmonary valvular stenosis. Three years later, Campbell (1954) reported one patient with a right aortic arch from a series of 75 cases of pulmonary valvular stenosis with an intact ventricular septum: details of this patient's history and the methods of exclusion of a ventricular septal defect were not specified. A search has failed to reveal any other reported cases of pulmonary stenosis and right aortic arch without an associated ventricular septal defect. This paper reports a second instance of this particular combination of defects.

Case Report

The patient, aged 46, was a white truck driver who came to the out-patient department with a complaint of mild shortness of breath most of his life. He was rejected for the Army in 1942 because of a cardiac murmur. During the past two years he had had præcordial pain, which had lasted 10 to 30 minutes; it occurred during exertion or rest, and was not relieved by nitroglycerine. He could walk only about two blocks. There was no history of orthopnœa, œdema of the ankles, or palpitation. Two years before, following an attack of præcordial pain, he had been told by his family physician that he had had a myocardial infarction and had been advised to limit his activity: he had not worked since that time. The past history was not significant.

Physical Examination. Blood pressure was 140/80 mm. Hg. A jugular 'a' wave was present. Examination of the lungs was normal. There was normal sinus rhythm with a rate of 80 a minute. A left parasternal tap was palpated in the third to fifth intercostal spaces. A grade 3/6 ejection murmur was audible over the præcordium and was loudest at the third intercostal space at the left parasternal border and radiated to the base of the neck and high posterior thoracic region. No ejection sound or diastolic murmur was audible, and no thrill could be felt. The second sound was moderately split and the pulmonic component was diminished in intensity. The liver was palpated 2 cm. below the right costal margin and was not pulsatile. No cyanosis or clubbing was present.

The cardiogram showed incomplete right bundle-branch block, right ventricular hypertrophy, and non-specific primary T wave changes.

The hæmoglobin was 16·5 g. per 100 ml. with a hæmatocrit of 49 per cent and a normal white cell count. Other laboratory tests were normal.

Right cardiac catheterization was performed. A pressure gradient of 99 mm. Hg across the pulmonary valve was observed. The right ventricular pressure was 114/0–12 and the pulmonary artery pressure was 15/6 mm. Hg. The arteriovenous oxygen saturation difference was 5·25 volumes per cent. The cardiac output was 5·7 l./min. with a cardiac index of 2·9 l./min./m.^{2} The calculated pulmonary valve area was 1·1 cm.^{2} and the diameter 1·2 cm. The mean right atrial oxygen saturation was 73, and the brachial artery saturation was 98 per cent.

The catheter slipped from the pulmonary trunk into the right atrium before blood samples could be collected from the pulmonary trunk, and this position was not reached again.

An angiocardiogram injected from the right atrium demonstrated pulmonary valvular stenosis and post-stenotic dilatation of the pulmonary trunk. Also the right-sided aortic arch and the descending aorta crossing to the left side of the vertebral column at the T10–11 level, shown in the chest radiogram (Fig. 1), were confirmed.

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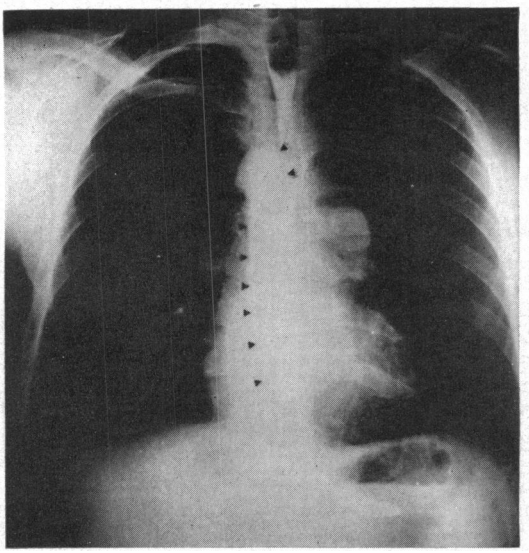

FIG. 1.—An antero-posterior chest radiogram with barium in the œsophagus demonstrates the right aortic arch with the descending aorta in the right thorax. The arrows demonstrate the aortic arch indenting the right side of the œsophagus. The lower arrows outline the lateral border of the descending aorta. This position of the aorta was verified by an angiocardiogram that is not reproduced. Post-stenotic dilatation of the pulmonary trunk was present.

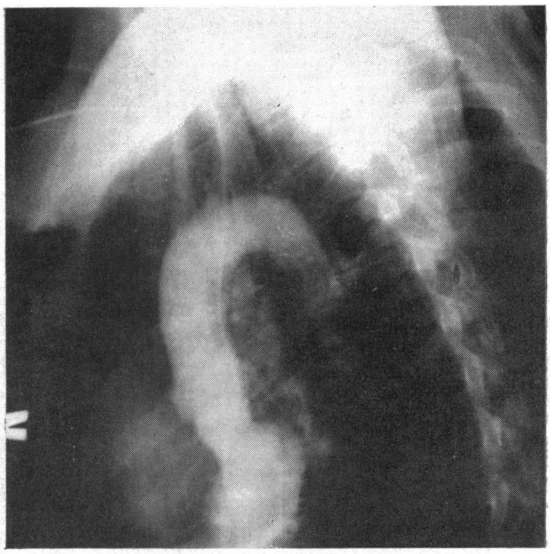

FIG. 2.—A left ventricular injected angiocardiogram demonstrates no left-to-right shunt at the ventricular or aortic level.

To determine whether a left-to-right shunt was present at the ventricular level, an angiocardiogram was obtained following injection of the contrast medium into the left ventricular cavity, using a retrograde aortic technique (Honick *et al.*, 1962): no left-to-right shunt was demonstrated (Fig. 2).

The patient was operated on for correction of the pulmonary valvular stenosis in April 1962 by David D. Snyder and Gilbert M. Campbell of the Surgical Department, using total cardiopulmonary bypass. Examination of the heart confirmed the enlarged right ventricle and post-stenotic dilatation of the pulmonary trunk. The anterior commissure was completely fused and the right postero-lateral commissure was partially fused while only the left postero-lateral was open. There was minimal infundibular stenosis. The pulmonary commissures were freed to the annulus. A diligent search revealed no ventricular septal defect. The patient has had a satisfactory convalescence.

Discussion

A right-sided aortic arch is the result of persistence of the right fourth aortic arch with the normally persisting left arch diminishing or disappearing entirely (Schnitker, 1952). This persisting arch may occur as the sole abnormality. A right aortic arch is associated also with about 60 per cent of patients with truncus arteriosus and 25 per cent with tetralogy of Fallot. Several cases with associated atrial or ventricular septal defects have been reported.

During the past 15 years the detection of pulmonary valvular stenosis has become relatively frequent. Brock and Campbell (1950) and Abrahams and Wood (1951) were among the first to emphasize that right aortic arch was frequently present with Fallot's tetralogy but absent with pulmonary stenosis when the ventricular septum was intact. As late as 1958 the same conclusion was reiterated by Nadas (1957) and Keith, Rowe, and Vlad (1958).

The exclusion of ventricular or atrial septal defect was thought to be established beyond reasonable doubt in this patient by the results of the right heart catheterization, the right and left ventricular angiocardiograms, and the diligent search for such a lesion at the time of the operation.

This patient and the one reported by Campbell (1954) are the only known instances of this rare combination of pulmonary valvular stenosis and an intact ventricular septum with a right aortic arch.

Summary

A second instance of pulmonary valvular stenosis, right aortic arch, and right-sided thoracic aorta is reported.

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