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Congenital arteriovenous fistula between the internal mammary artery and the pulmonary artery

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This is the fourth reported case of congenital arteriovenous fistula between the internal mammary artery and pulmonary artery. Precise and complete diagnostic evaluation is required to localize, delineate and appreciate the haemodynamic significance of this type of arteriovenous shunt. A brief review of the literature is given with suggestions for diagnosis.

Introduction

Congenital arteriovenous fistulas of the internal mammary artery with the pulmonary artery are very rare. Only three cases have been fully described in a recent review of the literature by Senno and coworkers[1]. A fourth patient whose clinical signs simulated a persistent ductus arteriosus prompted us to report the findings and the diagnostic procedures which were useful in this particular case.

Case report

A 28-year old woman was admitted to the Thoraxcenter for study of a continuous murmur which was found on routine physical examination in 1965. The patient had no family history of heart disease, gave no history of thoracic trauma and had no knowledge of a murmur having been noted on previous physical examinations. In January 1977, while under treatment for a left upper lobe bronchopneumonia the continuous murmur was again noted, a persistent ductus arteriosus suspected, and cardiac studies were recommended. At the time of admission, she was asymptomatic and afebrile. Height was 174 cm and weight 78 kg. On physical examination, there was no dyspnoea, cyanosis, clubbing of the fingers or oedema. Pulse and blood pressure were normal. A grade 4/6 high-pitched continuous machinery murmur was audible in the second intercostal space at the left sternal border and was confined to an area about two inches in diameter. The murmur was superficial and 'close to the ear' and was associated with a slight thrill. The heart sounds were normal. Routine laboratory tests and the electrocardiogram were normal. The phonocardiogram showed a continuous high frequency murmur overriding the second heart sound. The M-mode echocardiogram revealed slight enlargement of the right ventricle with an end-diastolic diameter of 36 mm (normal: 30 mm) but the interventricular septum moved normally. On a frontal view chestroentgenogram, the left hilum appeared dilated and nodular (Fig. 1); the lateral view confirmed the presence of a nodular lesion, just behind the sternum.

Percutaneous femoral right and left heart catheterization was performed which revealed normal pressures. However an increase in oxygen saturation ($\pm 3\%$) was repeatedly found in the peripheral left pulmonary artery beyond the main trunk. These results were consistent with a shunt of arterialized blood to the left pulmonary artery. This was not unexpected, because of the suspected persistent ductus arteriosus. The left ventriculogram was normal, with an end-diastolic volume at the upper limit of normal (91 ml m^{-2}). Biplane pulmonary angiography and supravalvular aortography failed to demonstrate a persistent ductus. However, after the venous phase of the pulmonary angiogram as well as during the aortography, a serpiginous collection of enlarged vessels was observed to the left and to the rear of the sternum in the third intercostal space.

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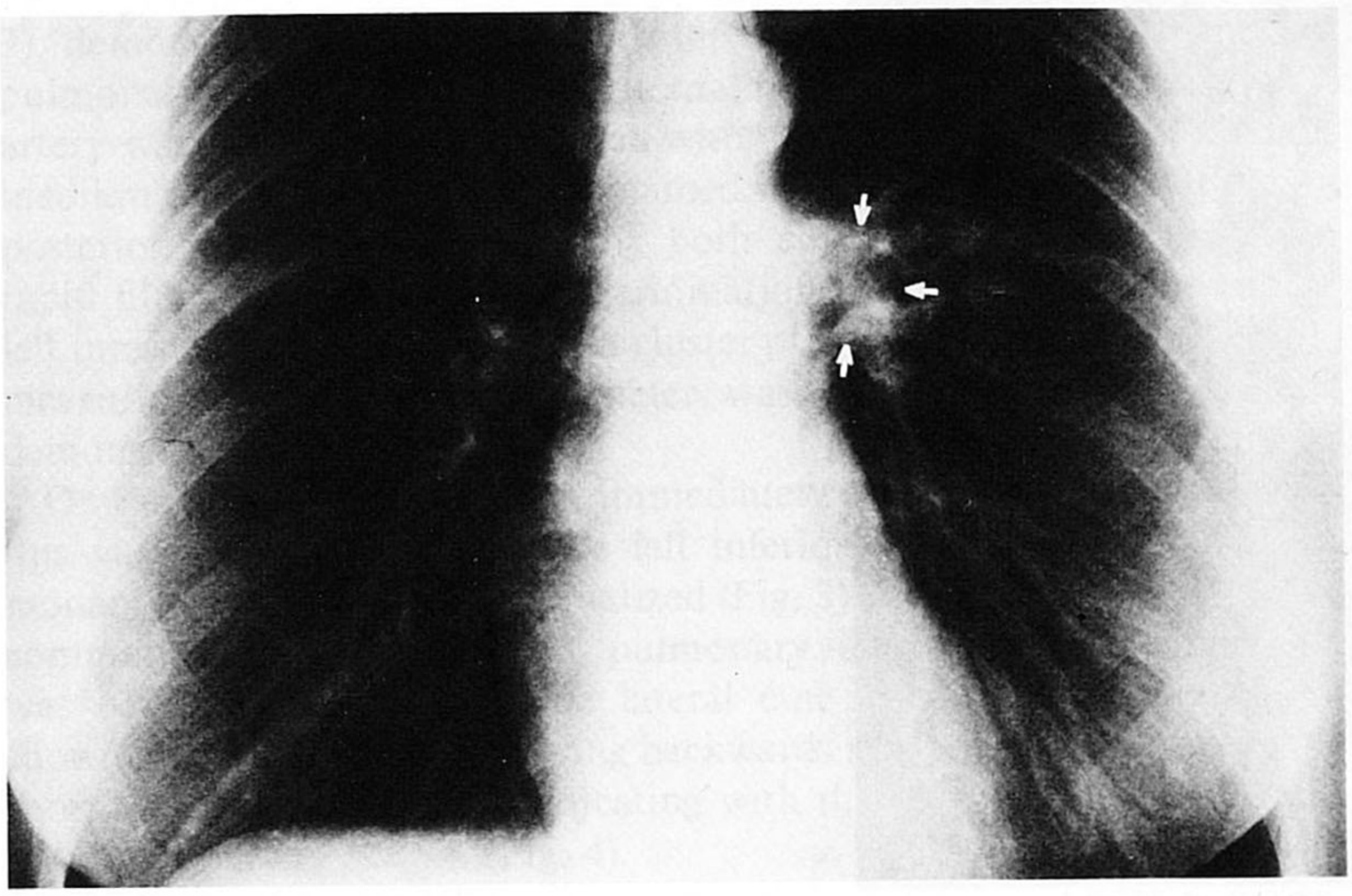

Figure 1 X-ray film of chest, antero-posterior view with dilated and nodular left hilum.

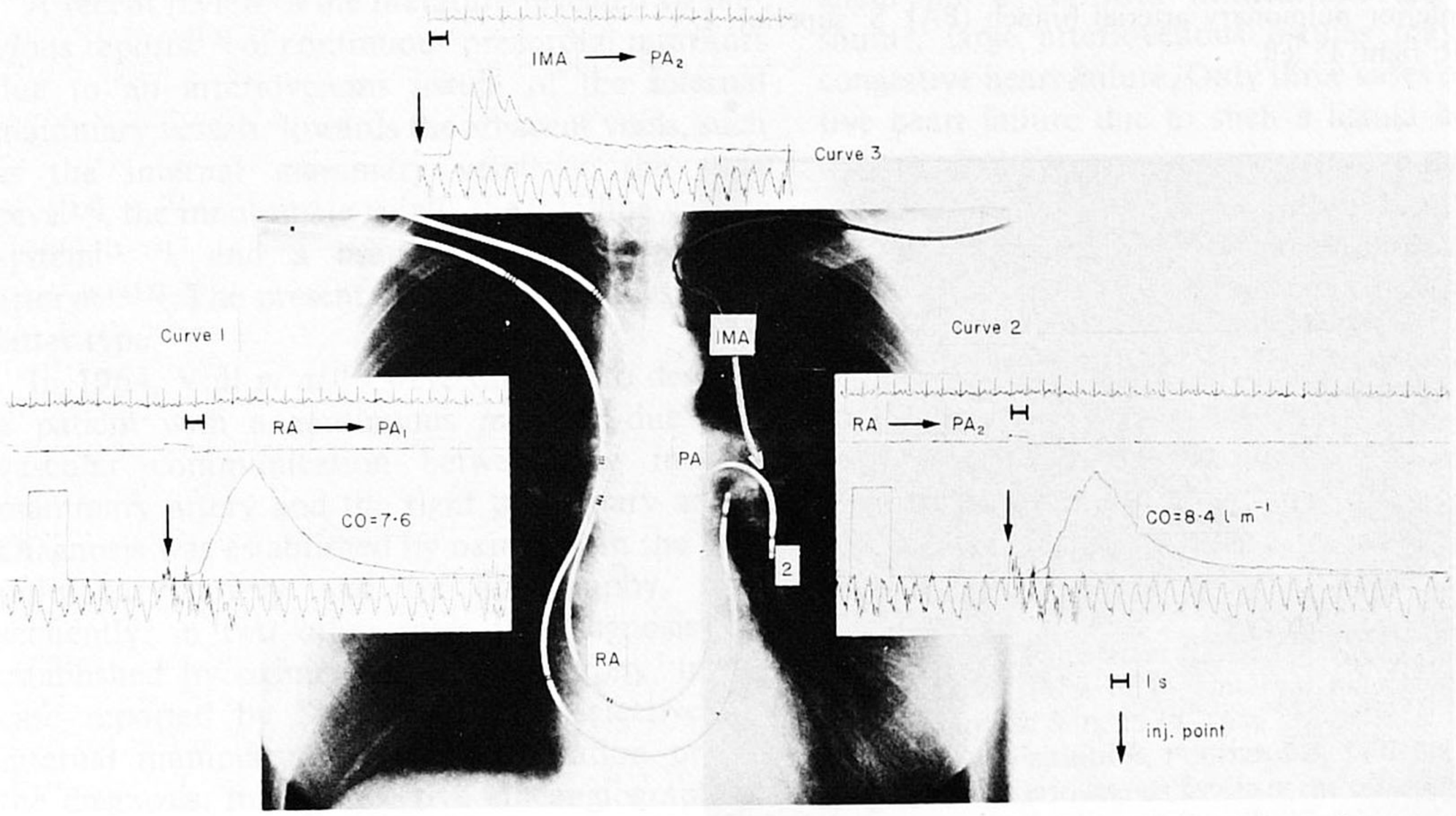

Figure 2 Curve 1: the tip of the thermodilution catheter is positioned in the main stem of the pulmonary artery (PA); the cold indicator for measurements of the cardiac output (CO) is injected into the right atrium (RA) CO 7·6 l m^{-1} (average of duplicated measurements). Curve 2: tip of the catheter, in the proximal left pulmonary branch (2); cold indicator for measurements of CO injected into RA CO 8·4 l min^{-1} (average of duplicated measurements). The difference in CO suggests that the catheter in the latter position is distal to the shunt. Curve 3: tip of the thermistance is positioned in the proximal left pulmonary branch (2). Selective injection in the internal mammary artery (IMA). The sudden pulsatile change of the intravascular temperature demonstrates a left to right shunt at the left pulmonary artery level.

On a subsequent examination a selective left internal mammary artery catheterization was done by a left brachial artery approach, using a Judkins left coronary artery catheter.

Just before the cineangiogram was made, several thermodilution curves were recorded with injection of 11 ml of cold water into the right atrium. The tip of the thermo-dilution catheter was first positioned

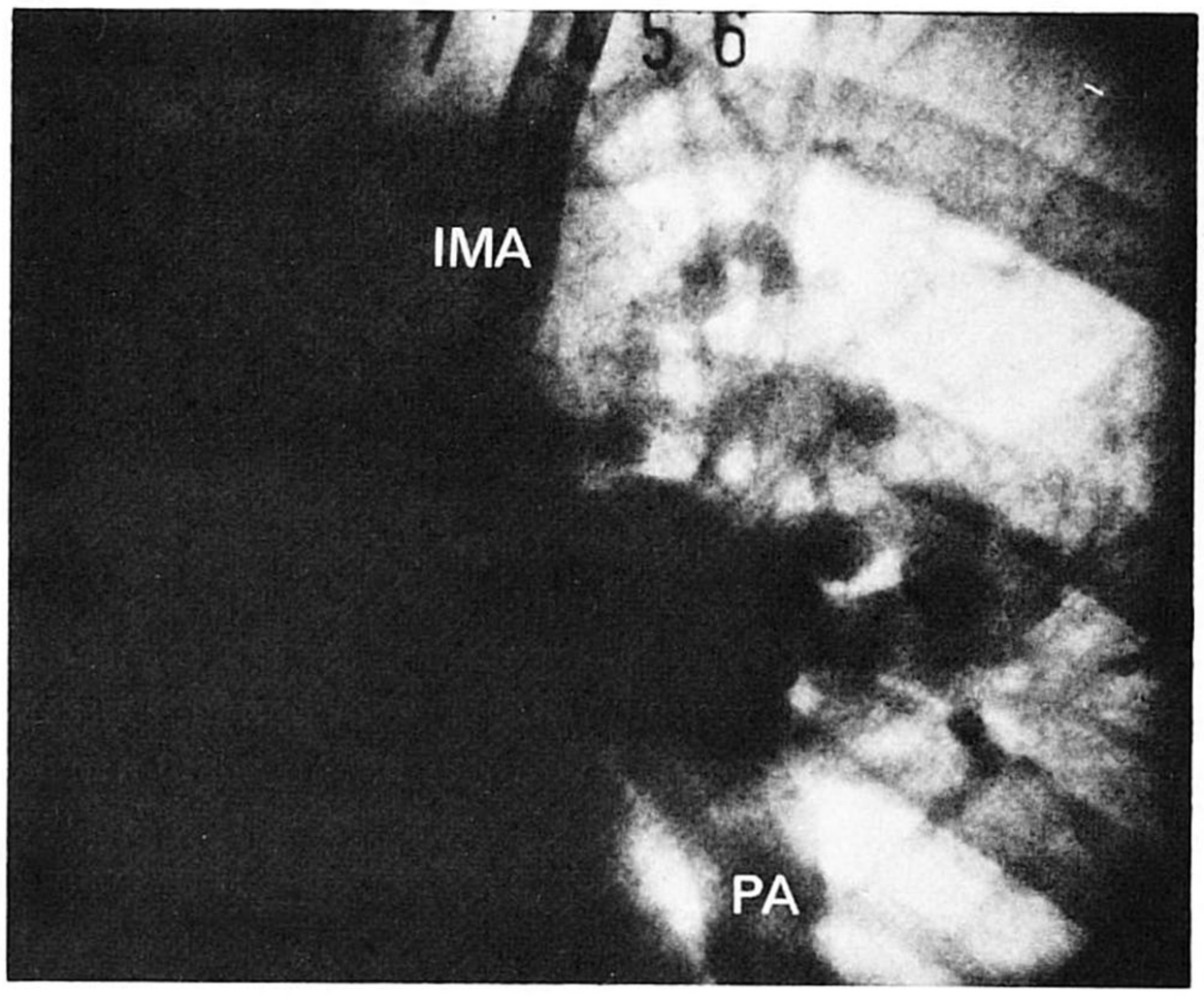

Figure 3 Selective left internal mammary arteriography, antero-posterior view; opacification of the internal mammary artery (IMA), filling of several dilated and tortous vessels and filling of the left inferior pulmonary arterial branch (PA). S: superior; I: inferior; R: right; L: left.

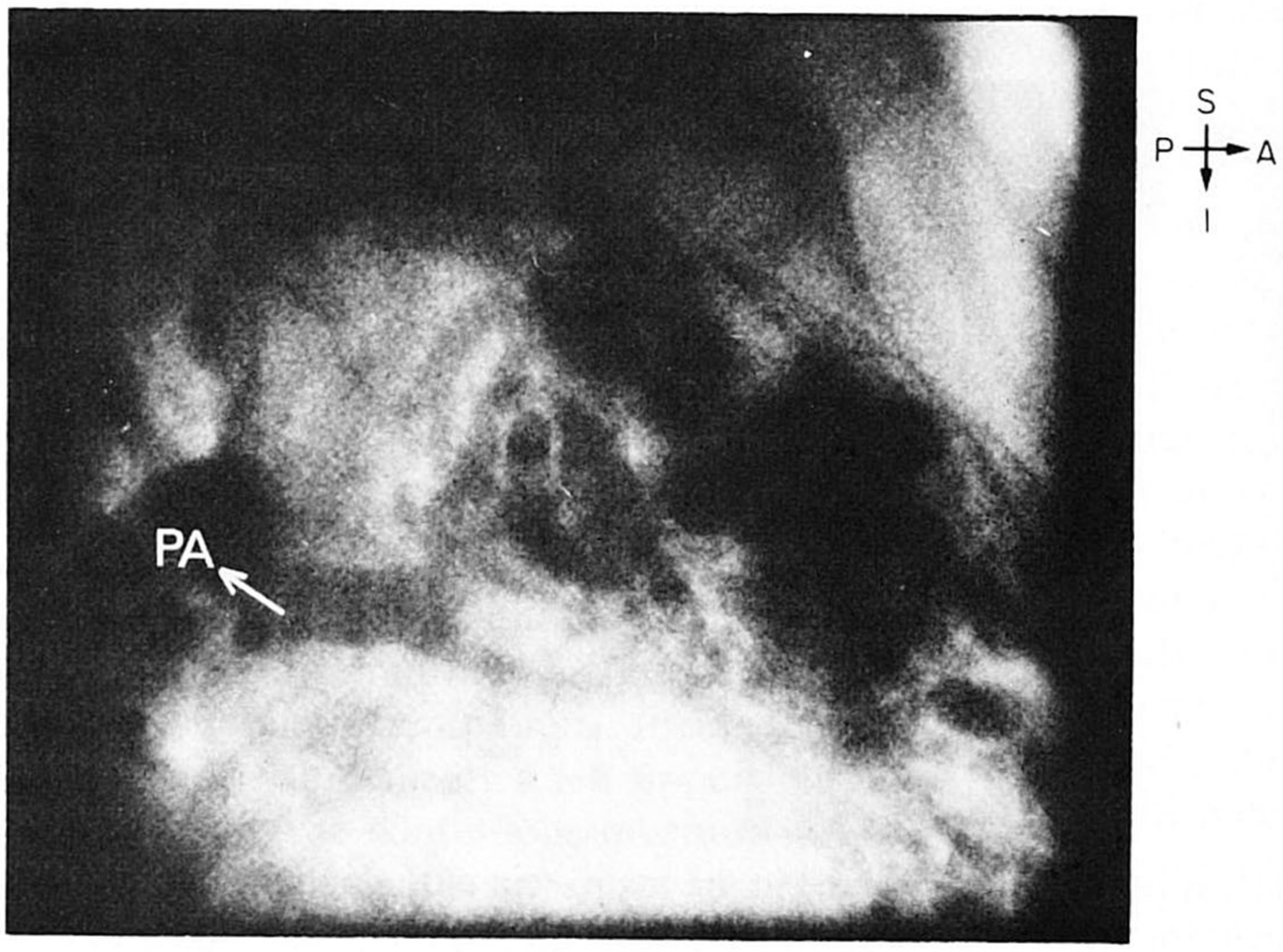

Figure 4 Selective left internal mammary artery (IMA) arteriography, lateral view: contrast medium flows backward into a short common trunk communicating with the left pulmonary artery (P.A.). S: superior; I: inferior; A: anterior; P: posterior.

in the main pulmonary artery (Fig. 2, curve 1) and subsequently in the proximal left pulmonary artery (Fig. 2, curve 2). The repeated difference in cardiac output suggested that the tip thermistor in the latter position was distal to the shunt. This could be demonstrated as follows: a selective injection of the cold indicator into the left internal mammary artery resulted in a sudden pulsatile change of the intravascular temperature measured downstream in the proximal left pulmonary branch (Fig. 2, curve

3), demonstrating a left to right shunt at the left pulmonary artery level. The left internal mammary artery was then selectively injected with a contrast medium and angiograms were obtained in antero-posterior and lateral views using both cine and rapid filmchanging technique. Termination of the left internal mammary artery in a cluster of vessels, measuring about 4 cm in diameter was clearly demonstrated (Fig. 3).

On the antero-posterior view, immediately under this vascular malformation, the left inferior pulmonary arterial branch was visualized (Fig. 3). The communication with the left pulmonary artery was best demonstrated on the lateral cine films showing contrast medium flowing backwards into a short common trunk communicating with the left pulmonary artery branch (Fig. 4).

Discussion

A recent review of the literature revealed 20 previous reports[1,2] of continuous precordial murmurs due to an arteriovenous fistula of the internal mammary vessels, towards the adjacent veins, such as the internal mammary vein[3–10], the vena cava[2,6], the innominate vein[2], the vitelline venous system[11–13], and a branch of the pulmonary artery[9,14,15]. The present case clearly belongs to the latter type.

In 1964, Voll *et al.*[14] were the first to describe a patient with a continuous murmur due to a vascular communication between the internal mammary artery and the right pulmonary artery. Diagnosis was established by oximetry in the right pulmonary artery and by aortography. Subsequently, in two other cases the diagnosis was established by oximetry and aortography. In the case reported by Stafford *et al.*[9] selective left internal mammary artery catheterization proved the diagnosis. Indeed selective cineangiography is the most useful and precise technique for delineating the exact anatomic malformation. Oximentry, indicator dilution curves, hydrogen inhalation and intracardiac phonocardiography may be helpful to detect a shunt in the pulmonary trunk but they do not identify the internal mammary fistula as its origin. Since in our patient, there was no past history of chest trauma or surgery, the abnormality is presumably congenital in origin. The early embryonic vascular system is plexiform. Initially, the arteries and veins consist simply of endothelial tubes and cannot be distinguished from each other histologically[16].

In a 10 mm embryo, the saccus aorticus divides so that the sixth aortic arch is continuous with the pulmonary trunk and the primitive pulmonary arteries. Of the intersegmental arteries, the seventh plays at this stage an important role in the formation of the subclavian arteries and their branches. They are located at about the level where the two dorsal aortas join. Any communication present at this early embryologic stage between the future pulmonary and systemic circulation may result in an arteriovenous fistula. The term congenital arteriovenous angioma seems acceptable as it takes into account the following factors: the congenital nature and multiplicity of the lesions, and the large dilated-appearing vascular sinuses as a part of the congenital defect being not entirely the result of secondary dilitation.

There is a risk of infection or rupture of these anomalous vessels because their walls are often thin and may give rise to the formation of an aneurysm[17]. As with intracardiac left to right shunts, large arteriovenous fistulas may lead to congestive heart failure. Only three cases of congestive heart failure due to such a fistula have been described[1,12,19].

For all these reasons surgical repair is indicated. However, the latter is not without risk as two deaths have been reported due to haemorrhage during the operation while another patient scarcely recovered after a severe bleeding[18]. However, our patient refused any surgical treatment since she was completely asymptomatic.

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