Pulmonary arteriovenous fistula with bilharzial pulmonary hypertension

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Congenital pulmonary arteriovenous fistulae, provided that they are not present in very large numbers, are best managed surgically, and there is no effective alternative management. Where pulmonary arteriovenous fistulae develop as a consequence of pulmonary hypertension it is rational to resect the fistulae if the cause of pulmonary hypertension—for example, mitral stenosis—is correctable. Pulmonary arteriovenous fistula in the presence of unexplained or uncorrectable pulmonary hypertension may be the safety valve on which life depends and should, therefore, not be resected. An example is reported of pulmonary arteriovenous fistula associated with bilharzial pulmonary hypertension in which resection of the fistula resulted in death.

Fistulous communications between branches of pulmonary arteries and veins within the lung may be congenital, when they represent vascular hamartomata, or acquired, when there is no certain evidence that fistulae develop on the basis of septal hypoplasia.

Congenital pulmonary arteriovenous fistula is a haemangiomatous malformation of the pulmonary vascular bed, and is commonly a manifestation of multiple hereditary haemorrhagic telangiectasia or Rendu-Osler-Weber disease. While occasionally found in the newborn, first recognition is usually after puberty; the lesion is usually more than a distended sack fed by an end-artery and drained by a vein, and is often part of an intricate vascular anomaly which affects neighbouring vessels and sometimes even the chest wall. Fistulae usually abut on the pleura; adjacent parenchyma is little disturbed.

The extent of dislocation of circulatory physiology depends on the size of the right-to-left shunt. One large, several small, and many minute fistulae may produce the same physiological disturbance. The percentage of right ventricular output shunted through the fistula varies widely—from 18 to 90 per cent in reported cases. In otherwise normal subjects probably 30 per cent of blood must be shunted past the pulmonary capillary bed before clinically detectable cyanosis will develop. Arterial oxygen unsaturation, within certain limits of anoxia, stimulates the erythро-
that, in circumstances of polycythaemia, as in
tetralogy of Fallot, pulmonary vascular resis-
tance may be increased from small multiple
thrombi which reduce effective pulmonary
blood flow. Therefore, though the heart is
usually normal in patients with congenital
pulmonary arteriovenous fistulae, it is im-
portant to recognize that it may enlarge when
the resistance offered to flow through the
fistula is unusually low or where over-all
pulmonary vascular resistance is significantly
increased.

Pulmonary arteriovenous fistula is re-
corded in association with mitral stenosis.
Lindgren (1946) and Steinberg and McClena-
han (1955) quoted examples of pulmonary
arteriovenous fistula in patients with mitral
stenosis, and three of six previously reported
elements of pulmonary arteriovenous fistula
(Le Roux, 1959) had mitral stenosis. In two
of these patients the mitral stenosis was diag-
osed clinically and shown surgically to be
severe; in the third it was estimated clinically
to be mild. It has been maintained that
the development of pulmonary arteriovenous
fistula in the presence of pulmonary hyper-
tension, the consequence, for example, of
mitral stenosis, may result from the rupture
of hypoplastic vascular septa in patients with
a generalized vascular dysplasia; may repre-
sent increase in the size of shunt through an
already established small pulmonary arterio-
venous fistula; or may represent enlargement
of anatomically normal arteriovenous shunts
known to exist in most organs at precapillary
level. In the lung these shunts are most
numerous at the apices of the segmental
subdivisions, in the visceral pleura, and at the
level of the respiratory bronchioles. By
means of in vivo perfusion experiments using
glass spheres 20 to 40 times the diameter of
the lumen of capillaries, Prinzmetal et al.
(1948) have shown arteriovenous shunts in
the lungs, and in most other organs in many
mammals. Gas analysis studies in man have
confirmed these findings, and the passage
through the lungs of large particles such as
clumps of tumour cells and certain parasites,
particularly Schistosoma cercariae, has com-
monly been cited as evidence of their exist-
ence.

While, therefore, it remains debatable
whether pulmonary arteriovenous fistula can
develop in the presence of pulmonary hyper-
tension without the pre-existence of hypo-
plastic septa between arteries and veins, it is
clinically well established that the lesion is
found in two separate circumstances: one in
which pulmonary arteriovenous fistula is
associated with other manifestations of a
congenital vascular hypoplasia, and the
other in which the pulmonary arteriovenous
fistula develops in the presence of pulmonary
hypertension.

Pulmonary hypertension in relation to
bilharziasis is well recorded (Shaw and Ghareeb,
1938; Cavalcanti et al., 1962; Turner, 1964;
Chaves, 1966; Winship, Kallichurum, and
Lapinsky, 1969); the pulmonary lesions are
found in association with Schistosoma mansoni
and Schistosoma haematobium; the histo-
logical features of pulmonary bilharzial
hypertension are parenchymatous pseudo-
tubercles which lie in close proximity to
small pulmonary arteries in which there are
bilharzial ova, which are either calcified or
ingested by foreign body giant cells. It is
often not possible to identify the ova in re-
spect of species and some of the pseudo-
tubercles are necrotic. In addition to the
granulomatous lesions, there is diffuse in-
timal hyperplasia, medial thickening, and
thrombosis, with organization in the arteri-
oles and muscular arteries. Diffuse intimal
thickening of arterioles not directly related to
ova are also seen; larger arteries show medial
and adventitial thickening. Bilharzial pseudo-
tubercles, with or without ova, need not be
numerous, and obliterative changes which
involve arteries not directly related to pseudo-
tubercles may be particularly striking. In
many of the reported examples 'angiomatoid'
lesions typical of chronic pulmonary hyper-
tension are found in relation to the tubercles,
and in these the vessels are dilated and thin
walled. It is probable that pulmonary hyper-
tension, when it develops in relation to bil-
harziasis, is a manifestation of individual
hypersensitivity rather than a result of
mechanical obstruction of the pulmonary
vascular bed. The angiomatoid lesions were
once thought to be a distinctive feature of
bilharzial arteriolitis but probably result from
recanalization of obliterated vessels and are a
non-specific result of severe pulmonary
hypertension from any cause (Heath and

An association between bilharzial pul-
monary hypertension and pulmonary arterio-
venous fistula has not been found on scrutiny
of available published papers.

**Case report**

An 11-year-old African girl was admitted to
hospital in April 1967 with a history of progres-
sive dyspnoea on effort and cough for a year. Six
months earlier she had had fever and joint pain;
she denied haematuria or previous gastro-intes-
tinal upsets. Radiographic features were those of
cardiomegaly, prominent proximal pulmonary
arterial segments, and a left peripheral, lobulated,
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**FIG. 1a** The pulmonary arteriovenous fistula has been shown at angiocardiology.

**FIG. 1b** Arteriovenous fistula in which the artery (A) is shown opening into a grossly dilated vein (V).

**FIG. 2a** Granulomatous focus around two bilharzial ova, adjacent to a small pulmonary artery. (H. and E. x 60.)

**FIG. 2b** Late angiomatous lesion adjacent to pulmonary artery. (H. and E. x 80.)
pulmonary opacity suggestive of a pulmonary arterio-fistula. The child was thin and undernourished; her fingers were normal; she was not cyanosed and did not have angiomatous malformations of skin or mucosae. There was a prominent 'A' wave in the neck; the systemic blood pressure was normal; there was a moderate right ventricular lift and a palpable second heart sound. The first heart sound in the mitral area was loud, with an ejection systolic click. There was an ejection systolic murmur, grade 3-6, maximal at the second left interspace but also heard along the left sternal border. At the apex this murmur was replaced by a continuous murmur which increased in intensity with deep inspiration, and was diminished by the Valsalva manoeuvre. The features of the electrocardiogram were those of right atrial and right ventricular hypertrophy. Cardiac catheterization showed moderate pulmonary arterial hypertension (mean pressure 50 mm. Hg), with a pulmonary vascular resistance of 25 units, a right-to-left shunt of 0.8 litres per minute, and moderately severe arterial desaturatation (72%); a large left-sided arteriovenous fistula was shown by cine-angiography (Fig. 1); it lay in the lingular segment of the left upper lobe.

The child was observed for two months in hospital and during this time the heart size was thought to have diminished. There was radiographic evidence of increase in size of the opacity shown to be that of an arteriovenous fistula. Resection of the fistula was, therefore, undertaken.

Through a standard left thoracotomy, through the bed of the 6th rib without rib resection, the vascular anomaly was seen on the lateral surface of the lingular segment. The fistula was deflated by ligation of the lingular artery. Formal lingulectomy was undertaken by division of the bronchus and vein. The bronchus was closed proximally with interrupted silk and the closure buried in pleura. The vein was divided between ligatures and the segments stripped out cleanly without bleeding. At this stage the heart began to fall; the pericardium was opened and an isoprenaline drip started; the heart recovered quickly, without the need for cardiac massage. The left atrium was seen to be normal; thrills were not appreciated on the surface of the heart; the left ventricle was very much smaller than the right. As a manifestation of sublime hind-sight the operation note ends: 'There can be no doubt that there is very much more wrong with this child than pulmonary arterio-venous fistula, and this is either incidental or a safety valve in a patient with obstructive pulmonary vascular disease'. Though the child recovered consciousness, left ventricular function remained unsatisfactory in early convalescence, and isoprenaline was required to maintain a reasonable cardiac output. The child deteriorated and died 12 hours after operation.

The resected specimen, which contained the arteriovenous fistula (Fig. 1b), was shown histologically to bear the features of bilharzial granulomata (Fig. 2a), angiomatous lesions (Fig. 2b), and vascular sclerosis.

References
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