A CASE OF PULMONARY ARTERIO-VENOUS FISTULAE.

By R. J. M. FRY, T. T. CHAPMAN, K. SHAW and R. WILSON.

This uncommon condition is now being recognised more frequently but, so far as we know, the case here recorded is the first treated in this country.

The condition has been reported more frequently in males than in females and shows a familial tendency in the cases in which telangiectases are found (over 50 per cent. cases). The lesion most frequently occurs in the lower lobes, the right lower lobe being the commonest site in the reported cases. Diagnosis of the condition has been made in a number of children under the age of 5 years, though in most of the reported cases the diagnosis was made in adults. In a number of the reported cases the condition existed from childhood (as in the case below). An early diagnosis is of importance, as the incidence of serious complications, haemoptysis, thrombosis, brain abscess etc. is high.

The clinical picture and the morphology of the disease will depend on the presence or absence of a right-to-left shunt and the size of the shunt. Obviously, if there is a right-to-left shunt the condition will have some of the features of cyanotic congenital heart disease, viz., cyanosis, polycythæmia, dyspnoea and clubbing of the fingers. Dizziness, faintness, numbness, or weakness of one side, recurrent epistaxis and haemoptysis are other commonly reported features. In those cases in which there is no right-to-left shunt the presenting symptom may be haemoptysis or haemothorax, and these may be fatal. On examination a murmur can sometimes be heard (in about half the reported cases) and may be accompanied by a thrill. X-ray examination in most cases shows an abnormal shadow, and the vascular shadows in the region are broader than normal. If the Valsalva manoeuvre is done while screening the shadow may increase in size, whereas if expiration against a closed glottis (Muller’s test) is carried out the shadow will increase in size.

The R.B.C., Hb level, P.C.V. and the O₂ saturation will depend on the degree of shunt present. The polycythæmia may be marked with a P.C.V. as high as 80 per cent. and the Hb level over 20 gms/100

<table>
<thead>
<tr>
<th>R.B.C. millions /cu.mm.</th>
<th>Hb. g/100 mls.</th>
<th>Art. O₂ content vols %</th>
<th>Ven. O₂ content vols %</th>
<th>O₂ Cap. vols %</th>
<th>O₂ Sat.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-operative</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6.7</td>
<td>20.0</td>
<td>19.1</td>
<td>14.1</td>
<td>26.8</td>
<td>70%</td>
</tr>
<tr>
<td>Post-operative</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5.5</td>
<td>15.6</td>
<td>19.4</td>
<td>14.5</td>
<td>20.9</td>
<td>93%</td>
</tr>
</tbody>
</table>
If there has been a severe haemorrhage or repeated bleeding the true picture may be masked. The cell volume is increased without a concomitant increase of the plasma volume. The cardiac output has been found to be within normal limits; whether this is so in all cases is not certain. The shunt may be over 50 per cent. of the total pulmonary flow. If the shunt is of a high order there may be an increase in the arterial $\text{CO}_2$ content. These findings differ markedly from those in systemic arterio-venous fistulae.

**Case Report**

P.F. aged 17 years. At birth (in the Rotunda Hospital) no abnormality was noted. During the first year the mother noticed that his left arm was weak. She also noticed that he developed slight cyanosis when he was about 9 years old. At this time (1948) he had an attack during which he could not talk or speak properly; there are no records of any doctor having seen this, and he slowly recovered.

The boy was first admitted to the Royal City of Dublin Hospital in 1949. At that time he was complaining of pain in the left loin, of about 6 months’ duration, occurring in paroxysms. There was a slight swelling in the area with tenderness, which was indefinite and diffuse. There was a left hemiparesis with marked wasting of the arm. In appearance the boy was well nourished, but looked pale and delicate. He was apathetic, spoke little and it was noted “that the child appears to sweat profusely.” He was discharged after fifteen weeks, when he had improved without any diagnosis of the cause of the pain.

He attended the out-patient department occasionally and also had physiotherapy for his arm. In February, 1956, he was again admitted to the Royal City of Dublin Hospital complaining of breathlessness and tiredness. He had been having severe nose bleeds once a week for three months. He had anorexia, melaena and symptoms of angina. On examination he was pale, he had facial acne with an oily, sweaty skin, he had marked clubbing of the fingers, and the blood pressure was 134/70 and the pulse 120. There was a mitral systolic murmur and a split second sound in the pulmonary area. He had a loud systolic murmur, maximal at the base of the right lung posteriorly. There was weakness of the left leg, with accompanying signs of an upper motor neurone lesion and a spastic paresis of the left arm with wasting. The stools contained occult blood. ECG showed a normal vertical heart. Blood examination. Hb. 8.2 gms/100 mls. The red blood cells were microcytic and hypochromic. W.B.C. 11,600/c.mm. (neutrophils, 55%; eosinophils, 3%; lymphocytes, 35%; monocytes, 7%). The patient was given two pints of blood, following which cyanosis became apparent. X-ray examination suggested the presence of a vascular anomaly in the lower lobe of the right lung. Barium swallow showed slight auricular enlargement of the heart. The blood picture changed during the following four months to that shown in Table 1. The patient was transferred to St. Mary’s Chest Hospital in June.

**Pulmonary Arterio-Venous Fistula**

\[
\text{Anoxaemia Increased} \rightarrow \text{Marrow Stimulated} \quad \text{Right to Left Shunt}
\]

\[
\text{Anoxaemia} \rightarrow \text{Polycythaemia and increase in Hb.}
\]

\[
\text{Anoxaemia compensated} \quad \text{CO}_2 \text{ Content normal}
\]

\[
\text{Increased Peripheral Resistance}
\]

\[
\begin{align*}
\text{Danger of Thrombosis} & \quad \text{reduced} \\
\text{Haemorrhage} \quad & \\
\text{Possibility of Thrombosis and Haemorrhage}
\end{align*}
\]