Physiologic Studies in Congenital Absence of the Left Main Pulmonary Artery

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Congenital absence of a main pulmonary artery branch is now a well-recognized clinical entity, more than 60 cases having been reported in the literature. Diagnosis has heretofore been based primarily on radiologic findings in both routine x-rays and angiocardiography. Physiologic studies of pulmonary function, including differential bronchopulmonary function studies, provide valuable diagnostic information in addition to demonstrating the unusual behavior of a lung possessing normal ventilatory capacity despite grossly abnormal circulation. Few studies of this nature have been published. The present report of 2 such patients with congenital absence of the left pulmonary artery provides new physiologic data in these cases of such extreme imbalance of ventilation and perfusion.

Case Reports

Case 1

G. P., a 27-year-old white man was first seen in 1950 at the age of 22 years for evaluation of hemoptysis and weight loss of 14 pounds. Decreased breath sounds and rales were heard over the left upper lobe, patchy densities in the left lung field were seen radiologically, and a diagnosis of resolving pneumonitis was made. Bronchoscopy showed bronchial inflammation and bronchography was normal.

Since 1950 several episodes of hemoptysis had occurred. Extensive studies had been performed to rule out pulmonary tuberculosis and bronchiectasis. Pulmonary function studies in 1954 are presented in table 1. Maximum breathing capacity and lung volume studies were entirely normal. Exercise studies on a treadmill were also within normal limits, alveolar-arterial oxygen difference being 23 mm. Hg. Bronchopulmonary findings (table 2) were grossly abnormal, showing only 7 per cent oxygen uptake on the left side. Carbon dioxide production was 37 per cent of the total, and ventilation was approximately normal for both right and left lungs. During bronchopulmonary peripheral arterial blood samples showed 94 per cent oxyhemoglobin saturation when room air was breathed and 74 per cent when the right catheter lumen was occluded for 1 minute. With the left lumen occluded the patient was perfectly comfortable for several minutes. When he breathed pure nitrogen through the right lumen and room air through the left, oxyhemoglobin saturation dropped to 54 per cent in slightly more than 1 minute. During breathing of nitrogen through the left lumen of the catheter, however, the saturation dropped to 87 per cent after 3½ minutes.

On the basis of these findings a diagnosis of absence of the left main pulmonary artery was made, which was confirmed by angiocardiography (fig. 1). On bronchoscopic examination prominent vascular pulsations were noted in the submucosal region on the left, but no definite bleeding point was seen.

Because of repeated severe hemoptyses the left lung was removed in 1957. At operation the degree of bronchial arterial circulation to the left lung was very impressive (fig. 2). These vessels originated from both posterior and anterior aspects of the mediastinum and reached the lung through adhesive bands located at the apex of the left upper lobe, tip of the lingula, and inferior pulmonary ligament. There was no aortic arch on the left side. The pulmonary veins were considered normal.

A radiograph of the resected lung after injection of methylglucamine diatrizoate (Renografin) into the systemic bronchial circulation demonstrated these numerous dilated vessels (fig. 3).

Case 2

J. S., a 28-year-old white man was first seen in 1953 because of unusual fatigue of 8 months' duration and left-sided chest pain, increasing on deep breathing and particularly after hard work.

In 1946 x-rays had shown blunting of the left costophrenic angle and subsequently a left-sided fibrinous pleurisy developed, but no evidence of active tuberculous infection had been found.

Physical examination on admission to the hospital revealed only decreased breath sounds and

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Supported in part by funds made available by the Vermont Heart Association.
increased tactile fremitus in the left mid-lung field posteriorly. Pulmonary function studies revealed maximum breathing capacity, timed vital capacity, and lung volume determinations to be normal (table 1). Exercise tolerance test on a treadmill was normal. Alveolar-arterial difference was only 6 mm. Hg. Bronchospirometry (table 2) demonstrated that 35 per cent of the total ventilation was being accomplished on the left side. On the left oxygen consumption was 5 per cent of the total, carbon dioxide production was 26 per cent, and 29 per cent of the total vital capacity was produced on this side. Arterial oxyhemoglobin saturation was 96 per cent, both at rest and on exercise. While breathing pure nitrogen on the left side for 5 minutes the patient maintained oxyhemoglobin saturation at 95 per cent. When the nitrogen was introduced on the right side, however, saturation fell to 80 per cent after only 30 seconds.

Bronchospirometry showed crowding of the bronchial subdivisions of the left lung and markedly hyperemic bronchial mucosa. Diagnosis of absence of the left main pulmonary artery branch was made and was confirmed by angiocardiography (fig. 4).

Bronchospirometry in 1958 again showed 35 per cent of total ventilation, 5 per cent of oxygen uptake, and 26 per cent of carbon dioxide production being performed on the left side. Differential diffusing capacity for carbon monoxide was tested and 30 per cent uptake of carbon monoxide by the left lung was demonstrated. Diffusing capacity during exercise at a steady state was normal, 24 ml. per minute per mm. Hg. 3

**Discussion**

Congenital absence of a main branch of a pulmonary artery without associated intracardiac defects has been reported less than 20 times, the majority of cases having involvement of the right pulmonary artery. Aortic anomalies are very frequently associated with the absence of the pulmonary artery. An attractive recent embryologic explanation for the combination of anomalies is that of Anderson et al., 4 who suggested that proximal interruption of the pulmonary arch with persistence of a primitive pulmonary artery persists for the final picture. This theory is consistent with, and readily explains all the variations of absent pulmonary artery that have been seen.

Diagnosis in the past has usually rested on a combination of radiographic and clinical findings, angiocardiography providing definite confirmation in several instances prior to thoracotomy. 1, 5-7 Surprisingly enough, pulmonary function studies, and in particular bronchospirometric investigation, have very seldom been utilized to their full extent. These procedures are not only valuable diagnostically, but provide important physiologic data regarding ventilation and accessory perfusion of the lung deprived of its normal circulation.

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**Table 1**

<table>
<thead>
<tr>
<th>Case no.</th>
<th>MBC 1./min.</th>
<th>TVC % 3 min.</th>
<th>TLC 1. VC</th>
<th>FRC 1.</th>
<th>ERV 1.</th>
<th>RV 1.</th>
<th>Mixing Index</th>
<th>Resting Exercise</th>
<th>Arterial Dco</th>
<th>Art Sat, % N₂ to left lung</th>
<th>Art Sat, % N₂ to right lung</th>
<th>Minim. Calc., ml./min.</th>
<th>CO uptake ml./min.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>141</td>
<td>—</td>
<td>5.2 4.2</td>
<td>1.8</td>
<td>0.85</td>
<td>0.96</td>
<td>1.2</td>
<td>94</td>
<td>91</td>
<td>23</td>
<td>—</td>
<td>1838</td>
<td>—</td>
</tr>
<tr>
<td>2.</td>
<td>122</td>
<td>100</td>
<td>6.1 4.4</td>
<td>2.7</td>
<td>1.08</td>
<td>1.64</td>
<td>1.0</td>
<td>96</td>
<td>96</td>
<td>6</td>
<td>24</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

MBC, maximum breathing capacity; TVC, timed vital capacity; TLC, total lung capacity; VC, vital capacity; FRC, functional residual capacity; ERV, Expiratory reserve volume; RV, residual volume, Dco, diffusing capacity of carbon monoxide.

**Table 2**

<table>
<thead>
<tr>
<th>Ventilation,</th>
<th>O₂ uptake,</th>
<th>CO₂ prod.,</th>
<th>Vital cap.,</th>
<th>Art, Sat, % N₂ to left lung</th>
<th>Art, Sat, % N₂ to right lung</th>
<th>Minim. Calc., ml./min.</th>
<th>CO uptake, ml./min.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case no.</td>
<td>% total</td>
<td>% total</td>
<td>% total</td>
<td>% total</td>
<td>% total</td>
<td>ml./min.</td>
<td>ml./min.</td>
</tr>
<tr>
<td>1.</td>
<td>48 52</td>
<td>93 7</td>
<td>63 37</td>
<td>65 35</td>
<td>87</td>
<td>54</td>
<td>1838</td>
</tr>
<tr>
<td>2.</td>
<td>65 35</td>
<td>95 5</td>
<td>74 26</td>
<td>71 29</td>
<td>95</td>
<td>80</td>
<td>1810</td>
</tr>
</tbody>
</table>

(30 sec.)
ABSENT LEFT PULMONARY ARTERY

Figure 1
Venous angiocardiogram in case 1. Filling of right pulmonary artery is seen with absence of filling on the left.

As has been previously demonstrated the ventilation of a lung without a pulmonary artery may be considerable and even normal. This is true in the present 2 cases, in which the affected lungs accounted for 52 and 35 per cent, respectively, of the total ventilation.

To our knowledge studies of carbon dioxide production in such lungs have not been previously reported. In the present examinations this information was obtained by collecting expired air from each lumen of the Carlen’s bronchospirometric catheter and analyzing it in the Scholander microgas apparatus. The results, showing carbon dioxide productions by the affected lungs of 37 and 26 per cent, suggest a considerable degree of collateral circulation to these ventilated lungs. The presence of such a circulation has been discussed by others4, 6, 8 and is convincingly demonstrated in figures 2 and 3. That such large amounts of carbon dioxide are capable of being excreted from an arterial circulation is easily explained by the shape of the carbon dioxide dissociation curve.

Further evidence for a richly developed accessory circulation to these lungs is presented by the differential bronchospirometric oxygen uptake studies. Although it has generally been reported2, 5, 9 or assumed that little or no oxygen uptake occurs, in our cases the values were unequivocally significant even during the breathing of room air and with normal arterial oxyhemoglobin saturation. Some earlier studies in this regard2, 10 are qualified by the use of anesthesia and the presence of hypoventilation and arterial desaturation. Arterial blood normally but not 100 per cent saturated, will obviously absorb additional oxygen when perfused through an adequately ventilated lung with a higher alveolar oxygen tension.

The observation of a drop in oxyhemoglobin saturation from normal to 87 per cent in one of our patients during the breathing of pure nitrogen on the affected side (table 2) is in effect an expression of oxygen excretion and once again shows that oxygen exchange occurs in this lung. We are unable to explain the absence of desaturation in the second patient under similar circumstances; we suspect that his effective bronchial blood flow is not so large as the first patient’s. This appears rea-
sonable in view of the absence of hemoptysis.

As reasoned by Bloomer,\textsuperscript{10} the Fick principle may be utilized for calculation of bronchial blood flow. By use of the absolute uptake of oxygen by the affected lung, the simultaneously determined arterial oxyhemoglobin content and capacities, and on the assumption that the bronchial blood reaching the lung has the same saturation as brachial artery blood, minimal bronchial blood flow may be calculated. These minimal flows in our patients were calculated as 1,838 and 1,810 ml. Were blood leaving the affected lung less than 100 per cent saturated, as is probably the case, the calculated flow would be significantly higher. Despite such an increased left ventricular work load, our patients exhibited neither radiologic or electrocardiographic evidence of left ventricular hypertrophy nor clinical signs of cardiac decompensation.

With normal or nearly normal ventilatory dynamics in the absence of alveolar-capillary membrane block, diffusing capacity for carbon monoxide would be expected to be normal. This was indeed the case in our one patient so tested who showed a steady state $D_{O_2}$ of 24 ml per minute per mm Hg, which is within normal limits for his age.\textsuperscript{3} Although it is not feasible to determine the diffusing capacity of each lung separately, carbon monoxide uptake on each side (table 2) is in good agreement with the values of differential ventilation. This lends further credence to recently published work\textsuperscript{11} that diffusing capacity is closely related to ventilation.

**Summary**

Physiologic studies in 2 cases of congenital absence of the left pulmonary artery are presented. The significance of carbon dioxide production and oxygen uptake in the affected lung and their relationship to ventilation and perfusion are discussed. All functional and anatomic evidence points to a highly significant bronchial artery circulation substituting in part for the absent pulmonary artery.

**Summario in Interlingua**

Studios physiologic in 2 casos de absentia congenite del arteria sinistro-pulmonar es presentate. Es discutite le significacion del production de biaxydo de carbon e del captation de oxygeo in le affeite pulmon e le relation de illos al funcionamento del ventilacion e del perfusion. Omne le disponibile datos functional e anatomic suggere le existentia de un significativissime circulation de arteria bronchial que age in vieirato partial del absente arteria pulmonar.
References

William Beaumont recognized, grasped, and improved the opportunity which fell in his path, with a zeal and an unselfishness not excelled in the annals of medical science... His work remains a model of patient, persevering investigation, experiment, and research, and the highest praise we can give him is to say that he lived up to and fulfilled the ideals with which he set out and which he expressed when he said: "Truth, like beauty, when unadorned, is adorned the most, and, in prosecuting these experiments and inquiries, I believe I have been guided by its light."—Sir William Osler. Aphorisms from His Bedside Teachings and Writing. Edited by William Bennett Bean, M.D. New York, Henry Schuman, Inc., 1950, p. 110.