Improved gas exchange after pneumonectomy in an adult with incomplete pulmonary vein atresia

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Abstract
Atresia of the pulmonary veins of the left lung and the right upper lobe with moderate pulmonary hypertension was diagnosed in a 20 year old man presenting with exertional dyspnoea and haemoptysis. After left pneumonectomy gas exchange appeared to have improved. This was shown by improved arterial oxygen pressures during all steps of exercise in a cycle ergometer test in comparison with values obtained before surgery. This improvement could be entirely attributed to a decrease in physiological dead space ventilation.

(Thorax 1994;49:723–725)

In contrast to complete atresia of the pulmonary veins, which is almost invariably rapidly fatal in early infancy, incomplete pulmonary venous atresia may present in childhood with failure to thrive, dyspnoea, recurrent respiratory infections, and haemoptysis. Many patients with pulmonary venous atresia have been reported in the literature, but we are aware of only one (female) patient who was followed with this condition until the age of 20. The mechanism of impaired gas exchange in this condition, which is associated with pulmonary artery hypertension, has not been studied but is believed to be due to gross inequality of ventilation and perfusion.

We have studied a young adult patient with pulmonary hypertension due to an atresia of the pulmonary veins of the left whole lung, and presumably also of the right upper lobe, who underwent extensive lung function testing including an exercise test with arterial blood gas analysis during each exercise increment, before and after left sided pneumonectomy.

Case report
A 20 year old man was referred for further analysis of his dyspnoea on exertion and small recurrent haemoptyses.

He had had surgery for an omphalocele during infancy. In childhood he experienced several episodes of bronchitis. A sweat test and a bronchogram were normal, as were findings during bronchoscopy. An iron deficiency anaemia was found with a prolonged bleeding time (15 minutes) due to platelet malfunction; clotting factor VIII was normal. The anaemia was believed to be due to recurrent episodes of epistaxis.

At the time of first presentation in our hospital his exercise tolerance was such that he was able to go to school by bicycle if weather conditions were favourable. Epistaxis had not occurred for some time, but haemoptysis had been more frequent.

On examination he appeared lean, body weight being 55 kg, height 1·67 m (body mass index 17·8 kg/m²). There was no dyspnoea, cyanosis, or oedema. Blood pressure was 100/60 mm Hg, the pulse was normal. The jugular venous pressure was normal. Breath sounds were diminished over the left lung. Auscultation of the heart revealed a loud, split second sound at the left sternal border; no murmurs were heard. Palpation revealed a left parasternal heave, otherwise physical examination was unremarkable. Laboratory tests were in the normal range except for a haemoglobin level of 12·8 g/dl.

The electrocardiogram revealed an incomplete right bundle branch block only. Lung volumes were near normal with mild, partly reversible bronchial obstruction and a small increase in residual volume. Diffusing capacity (single breath test for carbon monoxide: TLCO) was 50% of predicted; capillary volume appeared almost normal: 65·2 ml (85% pred), the membrane factor (DM) was 43% predicted; there was no evidence of an increased right to left shunt neither at rest (3%) nor at maximal exercise of 75 W breathing 100% oxygen (7%). Exercise testing was performed with a computerised cycle ergometer (Jaeger, Germany) in a 45% semirecumbent position at 60 rpm with an arterial indwelling catheter for serial blood gas analysis taken at 135 seconds after each increment in exercise load, in a three minute incremental exercise test schedule. Blood gas analysis was performed immediately after obtaining each specimen with an automated blood gas analyser (Corning, UK). Hypoxaemia was found during maximal exercise of 65 W, with an alveolar-arterial oxygen gradient increasing from 1·8 kPa to 7·7 kPa.

The chest radiograph showed enlarged pulmonary vessels in the right hilum with moderate displacement of the heart to the left; fluoroscopy revealed a normal pattern of breathing.

Echocardiography showed normal valves, a normal left atrium and left ventricle; the right atrium and right ventricle were dilated, and there was no evidence of intracardiac shunt. Right heart catheterisation revealed considerable pulmonary hypertension: pulmonary arterial pressures were 40/28 mm Hg (mean 32 mm Hg), with normal pulmonary capillary wedge pressure (3·8 mm Hg) and right atrial pressures (4·7 mm Hg). No intracardiac shunts were found.

A technetium-99m labelled albumin lung perfusion scan showed a perfusion defect to
the right upper lobe and to the left not matched by a krypton-81m labelled ventilation scan. Pulmonary angiography showed no blood flow to the right upper lobe and to the whole of the left lung, while the right lower and middle lobe artery branches were considerably dilated; the pulmonary veins were not seen on the left side. Computed tomography showed a poorly perfused but otherwise normal left pulmonary artery and right upper lobe pulmonary artery branch without evidence of intraluminal thrombus. Arteriography of the bronchial arteries did not reveal any abnormalities that were deemed appropriate for embolisation. Contrast injection in the aortic arch did not show an aberrant aortic supply to the pulmonary artery system. Coronary arteriography revealed an anomalous connection from the right coronary artery to the pulmonary artery system.

In the following two years the condition of the patient remained stable until he reported back to the hospital with massive haemoptysis requiring transfusion of six units of packed red blood cells, in addition to platelet transfusion. Fibrebronchoscopy showed bleeding from the left lower lobe. A second episode of massive bleeding within 24 hours urged us to perform a left lateral emergency thoracotomy after an attempt to embolise the bronchial artery system failed. At thoracotomy the pulmonary artery tree was normal, with no evidence of thrombi, but the left pulmonary veins were completely absent at the site of expected entrance into the left atrium. A left pneumonectomy was performed. Pathological examination revealed features consistent with secondary pulmonary hypertension, tortuous abnormal pulmonary veins, with intimal proliferation, but without signs of vasculitis, thromboembolic disease, or inflammatory or neoplastic changes. The postoperative course was uneventful. Four months after pneumonectomy the patient reported an improved exercise tolerance. Physical examination did not reveal any new abnormalities. Postoperative lung function tests showed the expected restrictive ventilatory defect, with a small further reduction in TLCO. When corrected for the alveolar volume, however, the diffusing capacity (KCO) had considerably improved.

On exercise VO2max had decreased from 24.4 to 21.4 ml/kg/min in the postoperative test, and the PaO2 at rest remained normal. Gas exchange and exercise tolerance were clearly improved, however (figure). The alveolar-arterial gradient for oxygen was lower and PaO2 was higher during all steps of the exercise test. The physiological dead space (Vd/Vt) was smaller during all steps of the postoperative test.

**Discussion**

These findings provide evidence that gas exchange can improve after pneumonectomy in patients with predominantly one sided pulmonary vascular disease. In our patient the improvement was even more striking as the right upper lobe, with a supposed similar condition to the left lung, was an additional disadvantage.

One of the three patients with unilateral pulmonary vein atresia reported by Swischuk et al. had improved exercise tolerance after pneumonectomy, which was also performed because of haemoptysis; however, in this 2.5 year old child there were no objective data to substantiate this clinical observation. The young adult female (patient no. 3 reported by Belcourt et al.) had normal pulmonary artery pressures when aged 15, and presented at 20 with similar complaints to our patient, including exertional dyspnoea and haemoptysis; she became well after left pneumonectomy.

The result of the TLCO is affected by the presence of the volume of blood (or haemoglobin) in the lung, and may have been increased preoperatively by the alveolar haemorrhage. Alternatively, carbon monoxide retention in the left lung may have been the result of diffusion to the bronchial artery system preoperatively. The KCO, which clearly improved postoperatively, was not, however, the limiting factor for exercise tolerance.

In the postoperative test Vd/Vt increased immediately at the first increment in exercise load, probably because of preferential ventilation of the more compliant left lung which was hardly perfused. It is important to note that dead space calculations were based on arterial blood gas samples for determination of Paco2:

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V_D, \text{PHYSIOL} / V_T = \frac{P_{aco2} - P_{etco2}}{Paco2}
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If the anatomical dead space using end tidal values (PETCO2) instead of arterial values (Paco2) had been used for analysis, an initial reduction in VD,ANAT/Vt in the preoperative test would have emerged with dead space ventilation remaining constant in the postoperative test, and the improved gas exchange would not have been diagnosed. The lower VO2max postoperatively may reflect the increase in efficiency of breathing: less oxygen is required by the respiratory muscles after the reduction in dead space ventilation.

We conclude that the impaired gas exchange in pulmonary hypertension with incomplete pulmonary vascular disease may improve after resection of affected lung tissue through reduction in physiological dead space, and that blood gas analysis during exercise testing in
Improved gas exchange after pneumonectomy

725


Thorax 1994;49:725-727

Spontaneous improvement in a patient with the hepatopulmonary syndrome assessed by serial exercise tests

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Abstract

A 37 year old patient with chronic active hepatitis progressing to cirrhosis presented with increasing breathlessness and was found to be hypoxic with finger clubbing. A progressive exercise study with measurement of oxygen saturation (SaO₂) showed abnormally high ventilation and desaturation to 81% at 100 W. Serial studies over nearly two years showed, first, deterioration, then improvement with lower ventilation and higher saturation levels at all work loads. This could not be correlated with any change in treatment with azathioprine, prednisolone, or propranolol.

(Thorax 1994;49:725-727)

About one third of patients with hepatic cirrhosis are found to have arterial hypoxemia without any of the usual cardiorespiratory causes. A few have obvious cyanosis with clubbing. The hypoxia is due either to right to left shunts through large arteriovenous connections within the lungs, or to peripheral vaso-dilatation of the fine branches of the pulmonary artery at both precapillary and capillary levels leading to incomplete diffusion equilibration between alveolar and end capillary blood. The pathophysiology has recently been fully reviewed.³

Case report

The patient presented at the age of 27 with nose bleeds, jaundice, anaemia, and swelling of legs and abdomen. Chronic active hepatitis was diagnosed by liver biopsy and, over subsequent years, further biopsies showed progression to cirrhosis despite treatment with prednisolone and azathioprine which had been prescribed for about five years, in varying dosage, before the present study. The cause of the chronic active hepatitis was never established. An autoantibody screen, including antibodies to smooth muscle and mitochondria, was negative. Hepatitis B serology was negative, and hepatitis C serology was not available at that time. The concentration of plasma α₁-antitrypsin was not directly measured, but the level of α₁-globulin was increased.

Ten years after his first attendance he complained of breathlessness and was referred for respiratory assessment. He was clubbed but not cyanosed at rest. He said he was unusually short of breath on exertion, but there was no serious limit to his exercise tolerance and throughout the period of the study he worked full time as a welder on a British Rail track gang, and could play 36 holes of golf in a day without discomfort. During the period of study standard liver function tests were abnormal, and varied without any systematic trend with ranges as follows: bilirubin 55-90 μmol/l, albumin 30-32 g/l, alkaline phosphatase 191-344 IU/l, gamma-GT 93-134 IU/l, and alanine transaminase 28-87 IU/l. He continued to drink alcohol, but it was difficult to assess in what quantity.

Blood gas measurements were obtained sitting (PaO₂ 9.2 kPa, PaCO₂ 4.3 kPa) and lying (PaO₂ 9.3 kPa, PaCO₂ 3.8 kPa). Pure oxygen was given from a balloon reservoir with careful exclusion of leaks from the breathing circuit whereupon PaO₂ rose to 60-6 kPa. A pulmonary perfusion scan with technetium-99 m labelled albumin microspheres, analysed according to the method of Chilvers et al, showed that 42% of the tracer passed through the lungs. The chest radiograph was normal. Lung volumes, both static and dynamic, were normal, but transfer factor for carbon monoxide (TLCO) and transfer coefficient (KCO) were...