

# Alveolar proteinosis lung lavage using partial cardiopulmonary bypass

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**ABSTRACT** An adult case of pulmonary alveolar proteinosis presented with an arterial oxygen tension of 27 mmHg (3.6 kPa) while breathing air. Dangerous hypoxaemia during lung lavage was avoided by using partial cardiopulmonary bypass.

Pulmonary alveolar proteinosis (PAP) is a relatively uncommon disease of unknown aetiology in which the alveoli are filled with an amorphous proteinaceous material, rich in phospholipid similar to pulmonary surfactant.<sup>1,2</sup> This alveolar filling can seriously compromise gas exchange. Although some cases are associated with only mild symptoms, in others cough, dyspnoea, and hypoxaemia can be disabling. Whole lung lavage with saline has been shown to result in substantial improvement in most patients.<sup>3-6</sup> However, lung lavage may cause transient worsening of hypoxaemia, making the procedure hazardous in those patients most needing lavage.<sup>7,8</sup> This report describes a patient with such severe arterial oxygen desaturation from PAP that cardiopulmonary bypass was used to maintain adequate arterial oxygenation during lung lavage.

## Case report

This 45-year-old black woman (ES) noted the onset of mild dyspnoea five years before admission to hospital. The dyspnoea became worse two and a half months before her first admission. She became dyspnoeic at rest and noted light-headedness and blurred vision during any exertion. She developed a cough which became productive of white sputum. She denied chills, fever, haemoptysis, rashes, or arthralgia. Weight loss of 70 pounds over one year was attributed to dieting. The initial admission was prompted by the onset of inspiratory left chest pain. The chest radiograph revealed a diffuse, bilateral

reticulonodular pulmonary infiltration. Arterial oxygen tension while breathing air was 46 mmHg (6.0 kPa) at that time. Open biopsy of the lingula demonstrated changes consistent with alveolar proteinosis. The alveoli were filled with granular PAS-positive, alcian blue-negative material. There was moderate interstitial inflammation. A trial of corticosteroid therapy for one month did not result in improvement and she was then admitted to our hospital for bronchopulmonary lavage.

In the past, she had experienced three episodes of pneumonitis, the last occurring eight years earlier. She had smoked half a pack of cigarettes a day for 10 years. For 10 years, she had worked as an assembler doing soldering in an electronics plant. Five years before admission, she left Philadelphia and moved to southern New Jersey where she lived in a rural area and worked as a nurses' aide in an institution for handicapped children.

Physical examination showed her to be in moderate respiratory distress. Blood pressure was 120/80 mmHg, pulse 100/min, temperature 37.1 °C, respiratory rate 34/min with mild use of accessory muscles. Neck vein distention, clubbing, and lymphadenopathy were absent. Crackles occurring late in inspiration were heard at both lung bases. The heart was not enlarged, and no murmurs were heard, but a soft fourth sound was present. The liver and spleen were not enlarged. There was no oedema. Routine laboratory examination was unremarkable except for elevation of the haemoglobin to 16.7 g/dl and a reduction of the albumin to 33 g/l. Serum LDH was 466 units (normal 100-225). Antinuclear antibody was negative, but rheumatoid factor was positive in a titre of 1:80. Arterial pH was 7.44 (36 nmol/l), P<sub>CO</sub><sub>2</sub> 31 mmHg (4.13 kPa), and P<sub>O</sub><sub>2</sub> 27 mmHg (3.60 kPa) breathing

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Table 1 Arterial oxygen pressure and percentage venous admixture on spontaneous and mechanical ventilation before, during, and after lung lavage

	Arterial $P_{O_2}$ (mmHg)			% Venous admixture
	Spontaneous ventilation 21% $O_2$	Spontaneous ventilation 100% $O_2$	Mechanical ventilation 100% $O_2$	Mechanical ventilation 100% $O_2$
Before lavage	27	59	126	32%
During lavage of left lung (cardiopulmonary bypass)			43	
One week after lavage of left lung During lavage of right lung	33		233 83	25%
Two weeks after lavage of right lung	48			

Conversion: mmHg  $\times$  0.133 = kPa.

Table 2 Results of pulmonary function tests

	$FEV_1$ (l)	FVC (l)	FRC (l)	TLC (l)	DLco (ml/min/mmHg)
Predicted	2.31	3.15	2.64	4.78	15.49
Before lavage	1.18	1.24	1.34	2.24	5.11
One week after lavage of left lung	1.24	1.40	1.14	1.85	7.02
Two weeks after lavage of right lung	1.55	1.76	1.34	2.41	11.52

room air (table 1). It was 59 mmHg (7.86 kPa) breathing 100% oxygen, giving an alveolar-arterial oxygen gradient of 654 mmHg (87.2 kPa) equivalent to a right-to-left shunt of 47% of the cardiac output. Pulmonary function tests were indicative of severe restrictive lung disease (table 2). Bronchoscopy revealed no endobronchial lesions and bronchial washings were negative for bacteria, mycobacteria, and fungi. The tuberculin test was negative and fungal antibodies were absent.

Because of the severity of the arterial hypoxaemia, bronchopulmonary lavage was performed while on partial cardiopulmonary bypass. A pulmonary artery catheter inserted at this time showed the pulmonary artery systolic pressure to be 41 mmHg, the mean pressure 17 mmHg, diastolic pressure 4 mmHg, and the pulmonary capillary wedge pressure 5 mmHg. Cardiac output was 5.26 l/min by thermodilution. A Robertshaw double-lumen tube was passed into the trachea under halothane anaesthesia. Mechanical ventilation of both lungs with 100% oxygen resulted in an arterial  $P_{O_2}$  of 126 mmHg (16.8 kPa) indicating a right-to-left shunt equal to 32% of the cardiac output. Because of the anticipated worsening during the lavage, partial venoarterial cardiopulmonary bypass was performed after full heparinisation using a Bentley BOS 10-bubble oxygenator. The left femoral artery was cannulated using a size 20 arterial cannula and the left common femoral vein was cannulated using a size 32 cannula. The venous cannula was advanced to the inferior

vena cava for optimal drainage. A bypass flow of 2.6 l/min (half of the cardiac output) was used. The left lung was then lavaged with 12.75 l of normal saline (23 cycles with an average volume/cycle of 554 ml), a total of 11.9 l being recovered. A protocol similar to that described by Rogers *et al* was used.<sup>8</sup> The effluent was initially turbid and full of debris, but gradually cleared. The lowest  $P_{O_2}$  of blood from the radial artery during the lavage was 43 mmHg (5.73 kPa) (at the end of a drainage phase) and the arterial  $P_{O_2}$  on 100% was 339 mmHg (45.2 kPa) after the lavage and cardiopulmonary bypass were terminated. Total bypass time was one and a half hours.

The right lung was lavaged seven days later without the aid of cardiopulmonary bypass, as arterial  $P_{O_2}$  had risen to 33 mmHg (4.4 kPa) on room air and 233 mmHg (29.7 kPa) during mechanical ventilation of both lungs with 100% oxygen (right to left shunt of 25%). The lowest  $P_{O_2}$  measured during the second lavage was 83 mmHg (11.1 kPa) at the end of a drainage phase, while ventilating the left lung with 100% oxygen. During the next five months, the arterial  $P_{O_2}$  rose to 69 mmHg (9.2 kPa) on room air and the pulmonary infiltrates slowly cleared. Subsequent recurrence necessitated a second bronchopulmonary lavage.

### Discussion

During the course of whole lung lavage for treatment of PAP, one lung is entirely filled and emptied repeatedly with a normal saline solution.<sup>3,4</sup> The lung

being lavaged is isolated from its mate by means of a double-lumen endotracheal tube. Since this lung has no ventilation, blood passing through it is not oxygenated. This mixed venous blood is added to oxygenated blood returning from the ventilated lung, causing lowering of arterial oxygen content. The amount of venous admixture is increased as pulmonary capillary blood flow increases in the lavaged lung.<sup>6,8</sup> The blood flow through the nonventilated, lavaged lung depends upon the relationship between pulmonary artery pressure and the airway pressure. When airway pressure exceeds pulmonary capillary pressure, the vessels collapse and blood flow ceases. If pulmonary capillary pressure exceeds airway pressure, blood flow will occur. Airway pressure in the lavaged lung varies greatly during the procedure. During drainage the pressure is atmospheric, while when the lung is filled with saline, the airway pressure depends upon the height of the saline reservoir above the lung—in this case 30 cm water above atmospheric. Blood flow occurs through the lavaged lung during drainage, but ceases as saline filling occurs and the airway pressure exceeds pulmonary capillary pressure.

If pulmonary oxygen transfer is sufficiently deranged before lavage, the arterial  $PO_2$  during lavage could fall transiently to a dangerously low level, causing tissue hypoxia. This could preclude use of this treatment in a patient needing it the most. Although most patients undergoing lavage are not severely hypoxaemic,<sup>5,6</sup> Ramirez-R<sup>3</sup> has reported lavage of a patient with PAP whose arterial  $PO_2$  was 28 mmHg (3.73 kPa) on room air and 124 mmHg (16.5 kPa) on 100% oxygen. In two other patients it was noted that arterial  $PO_2$  on 100% oxygen fell from 82 mmHg (10.9 kPa) to 38 mmHg (5.7 kPa) and from 71 mmHg (9.46 kPa) to 40 mmHg (5.3 kPa) respectively during lavage.<sup>4,9</sup>

Seard *et al*<sup>10,11</sup> reported simultaneous bilateral lung lavage with the aid of partial cardiopulmonary bypass in two children with PAP in whom tracheal size did not permit the use of a double-lumen tube. In these cases, one-third of the cardiac output was diverted through the oxygenator, resulting in an arterial  $PO_2$  of 25–30 mmHg (3.3–4.0 kPa) during lavage, a level we would not consider adequate for an adult.

Cooper *et al*<sup>12,13</sup> reported a critically ill 24-year-old woman with PAP in whom serial bilateral lung lavage was performed while on partial bypass. Her arterial  $PO_2$  before bypass was below 50 mmHg (6.7 kPa) while being mechanically ventilated with 90% oxygen and 10 cm PEEP. On partial bypass, her arterial  $PO_2$  fell to no lower than 77 mmHg (10.3 kPa) during lavage. Altose *et al*<sup>14</sup> describe the use of partial bypass during the initial unilateral lavage of a

32-year-old man with PAP whose arterial  $PO_2$  was 65 mmHg (8.7 kPa) breathing 100% oxygen by mouth-piece and 125 mmHg (16.7 kPa) when mechanically ventilated with 100% oxygen. His lowest arterial  $PO_2$  during lavage was 46 mmHg (6.1 kPa). Our patient is physiologically similar to this case.

Partial cardiopulmonary bypass was used during our patient's initial lung lavage because of her severe hypoxaemia and the degree of venous admixture on 100% oxygen. By its use, her arterial  $PO_2$  was maintained above 43 mmHg (5.7 kPa) even during cyclic drainage of the lavage fluid. This first lavage improved lung function to the point where lavage of the opposite lung was conducted seven days later without bypass.

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