The Use of Extracorporeal Circulation in the Treatment of Pulmonary Alveolar Proteinosis

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Keywords: bronchopulmonary lavage, pulmonary alveolar proteinosis, partial cardiopulmonary bypass, respiratory support

Abstract

(J. Extra-Corp. Technol. 18(4) p.225-227 Winter 1986) The use of a modified extracorporeal circuit has many uses in the treatment of several disease processes. Chief among these are partial and total support for cardiac and respiratory failure, isolated limb perfusion for the administration of chemical agents in the treatment of some forms of cancer, and in support of certain neurological procedures. Previously unreported in perfusion literature is the use of extracorporeal circulation in support of bronchopulmonary lavage as a treatment for severe pulmonary alveolar proteinosis. A 31-year-old male with this condition was successfully treated using bronchopulmonary lavage with a modified extracorporeal circuit. The technique is described.

Introduction

Over the past several years, there have been many attempts to utilize cardiopulmonary bypass for uses other than open heart surgery. Among these have been extracorporeal membrane oxygenation (ECMO) for long term cardiac and pulmonary support, isolated limb perfusion for hyperthermia and the delivery of chemotherapeutic drugs, and support for profound hypothermia and hypotension during certain types of neurosurgical procedures. During this time, the increasing sophistication of the oxygenating devices has allowed an improvement in the techniques utilized for these procedures.

One procedure not previously discussed in the perfusion literature is the use of cardiopulmonary bypass as an adjunct for the treatment of pulmonary alveolar proteinosis. Pulmonary alveolar proteinosis is a disease of unknown causes that is manifested by the accumulation of a granular, proteinaceous material, high in lipid content, in the alveolar spaces and the bronchioles. The disease is associated with pulmonary impairment, reductions in vital capacity, residual capacity, residual volume, and total lung capacity. The proteinaceous material interferes with gas exchange leading to an impairment of diffusion and arterial hypoxia.\(^1\)\(^2\)\(^3\)

The most effective and immediate treatment for pulmonary alveolar proteinosis has been whole lung bronchopulmonary lavage, which entails flooding the lungs continuously with large amounts of warmed saline.\(^4\)\(^5\)\(^6\) An improvement in symptoms and pulmonary function usually occurs within 24 to 48 hours. The technique of whole lung lavage was first described in 1965. However, it has met with varying degrees of success. There still appears to be some controversy over patient selection. This report will detail the elective application of a modified cardiopulmonary bypass circuit to provide temporary pulmonary support during a bronchopulmonary lavage for pulmonary alveolar proteinosis.
Case Report

A 31-year-old white male was admitted for an elective bronchopulmonary lavage of the left lung. The patient had a six month history of progressive shortness of breath. The patient had been previously admitted for a bronchoscopy and open lung biopsy which confirmed the diagnosis of pulmonary alveolar proteinosis. During that admission the patient underwent a bronchopulmonary lavage of the right lung with no problems. Arterial oxygen saturations during the procedure remained above 85%. The patient was admitted at this time for a lavage of the left lung. Significant laboratory data included a PaO$_2$ on nasal oxygen of 68 torr and a hemoglobin of 18.4% with a hematocrit of 54.9%. Physical findings were unremarkable with the exception of a pneumothorax on the right side and rales heard over the bases of both lungs. The patient weighed 80 kilograms and was 178 centimeters in height.

The patient was brought to the operating room with a chest tube in place on the right side. The initial plan was to attempt a lavage of the left lung. However, during the early test phase of the procedure, the patient's arterial saturation dropped to less than 70%. The testing phase consisted of clamping off the left lumen of the dual lumen endotracheal tube. To see if the right lung would support oxygenation of the patient. At this time it was decided to place the patient on cardiopulmonary bypass in order to support him during the lavage.

The cardiopulmonary bypass circuit consisted of a membrane oxygenator, a centrifugal pump, and polyvinyl chloride tubing. Because of the potential for long term pulmonary support following the procedure, there were no filters or reservoirs used. The heat exchanger portion of the oxygenator was not connected to a water bath because it was felt that, for the short term, the patient would maintain normothermia due to the lavage solution being at a temperature of 40°C. The patient was heparinized with 10,000 units of beef lung heparin. Cannulation was accomplished via the left femoral vein and artery. The circuit was primed with 1,000 cc of a balanced electrolyte solution and partial bypass was initiated. Bypass flows were calculated at 1.2 LPM/M$^2$ and averaged about 2 to 3 LPM during the bypass procedure. Activated clotting times were maintained at 300 seconds and were checked every 15 minutes. Arterial oxygen saturation remained above 95% during the lavage and the patient tolerated the procedure well. The FiO$_2$ of the oxygenator gas was maintained at 100% and the gas flow averaged 1 LPM. At the conclusion of the lavage, the patient was allowed to stabilize for 20 minutes and then was slowly weaned from bypass. Total pulmonary support time was 100 minutes. The heparin was reversed with protamine and the patient was decannulated. The patient was transferred to the recovery room with an arterial oxygen saturation of 96% and a PaO$_2$ of 120 torr on 100% oxygen via endotracheal tube. The patient was discharged from the hospital seven days later without the need for nasal oxygen and remains symptom-free six months later.

Discussion

While most reports in the medical literature describe the use of ECMO to support lavage of both lungs at one time, it was felt, where our patient was concerned, that a staged procedure would allow the opposing lung to support the body and would eliminate the need for cardiopulmonary support during the procedure. However, as indicated by the test phase described above, the patient was not able to tolerate the second procedure without support. This was probably due to the pneumothorax and perhaps an incomplete lavage during the first admission.

There is an indication in the literature of some patients undergoing a worsening pulmonary status after the lavage. Because of this possibility, the perfusion team needs to be prepared for the possible long term support of the pulmonary circulation. A modified cardiopulmonary bypass circuit, mounted on a small cart, with the oxygenator and centrifugal pump attached and facilities for oxygen and compressed air, allows the team to move the patient to the intensive care unit should it become necessary. While the heat exchanger portion of the oxygenator was not used during the short bypass procedure, its availability would be useful in the event of a longer procedure with its attendant heat loss. The Maxima oxygenator was chosen for this procedure because of its large fiber bundle and its ability to handle varying blood and gas flows throughout the procedure. The elimination of an arterial filter and other ancillary lines and reservoirs enables the team to decrease the amount of heparin used, resulting in fewer problems with bleeding and heparin reversal. However, the activated clotting time needs to be monitored closely as most of these patients will be maintained at normotherm-
mia and metabolize heparin faster than one is accustomed to during hypothermic open heart surgery.

The increasing sophistication of perfusion devices and techniques allows the perfusionist to assist in procedures previously not considered possible. While there have been reports of using cardiopulmonary bypass for bronchopulmonary lavage in neonates, the ease in which this system can be applied in adults may result in the perfusion team's future involvement with this procedure.

References