

BRIEF COMMUNICATION

## Bilateral whole lung lavage in pulmonary alveolar proteinosis – A retrospective study



A. Silva<sup>a,\*</sup>, A. Moreto<sup>a</sup>, C. Pinho<sup>a</sup>, A. Magalhães<sup>b</sup>, A. Morais<sup>c</sup>, C. Fiuza<sup>d,1</sup>

<sup>a</sup> Medical Resident in Anesthesiology, Centro Hospitalar São João, Porto, Portugal

<sup>b</sup> Consultant in Pneumology, Centro Hospitalar São João, Porto, Portugal

<sup>c</sup> Medical Assistant in Pneumology, Centro Hospitalar São João, Porto, Portugal

<sup>d</sup> Consultant in Anesthesiology, Centro Hospitalar São João, Porto, Portugal

Received 31 January 2014 ; received in revised form 11 April 2014; accepted 17 April 2014

Available online 18 July 2014

### KEYWORDS

Pulmonary alveolar proteinosis;  
Whole lung lavage;  
Bilateral

**Abstract** Whole lung lavage (WLL) is the gold standard technique for the treatment of Pulmonary Alveolar Proteinosis (PAP). In this paper we evaluated and discuss bilateral WLL, namely the procedure work-up and the therapeutic efficacy.

Six bilateral WLL performed through a careful adherence to a modified *Royal Brompton Hospital (London) technique* were carried out without major complications and were associated with clinical and functional improvement of the PAP patients submitted to this procedure.

As there are benefits in terms of time, patient comfort and cost effectiveness compared to unilateral WLL, associated with the efficacy and safety observed, bilateral WLL seems to be a suitable first choice for therapeutic lavage in PAP patients.

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### PALAVRAS-CHAVE

Proteinose Alveolar Pulmonar;  
Lavagem Pulmonar Total;  
Bilateral

### Lavagem pulmonar total bilateral na proteinose alveolar pulmonar - um estudo retrospectivo

**Resumo** A lavagem pulmonar total (LPT) é a técnica de referência para o tratamento da Proteinose Alveolar Pulmonar (PAP). Neste documento avaliamos e discutimos a LPT bilateral, nomeadamente o procedimento levado a cabo e a eficácia terapêutica.

Foram executadas seis LPTs bilaterais, através da adesão cuidada a uma técnica modificada do *Royal Brompton Hospital (Londres)*, executadas sem complicações de maior e foram associadas à melhoria clínica e funcional dos pacientes com PAP submetidos a este procedimento.

\* Corresponding author.

E-mail addresses: [acaciomsilva@gmail.com](mailto:acaciomsilva@gmail.com), [fiuza.carlos@gmail.com](mailto:fiuza.carlos@gmail.com) (A. Silva), [anamoreto@gmail.com](mailto:anamoreto@gmail.com) (A. Moreto), [xanavp@gmail.com](mailto:xanavp@gmail.com) (C. Pinho), [adrimagalhaes08@gmail.com](mailto:adrimagalhaes08@gmail.com) (A. Magalhães), [antonio.moraisrpp@gmail.com](mailto:antonio.moraisrpp@gmail.com) (A. Morais), [fiuza.carlos@gmail.com](mailto:fiuza.carlos@gmail.com) (C. Fiuza).

<sup>1</sup> Rua da Madeira 57, 4520-035 Escapães.

Como existem benefícios em termos de tempo, conforto para o paciente e eficiência em termos de custos, comparando com uma LPT unilateral, associada à eficácia e segurança observadas, a LPT bilateral parece ser uma primeira escolha adequada para uma lavagem terapêutica em pacientes com PAP.

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## Introduction

Pulmonary Alveolar Proteinosis (PAP) is a rare diffuse lung disease with three clinical forms, congenital, acquired (idiopathic) and secondary, characterized by an accumulation of large amounts of a phospholipoproteinaceous material in the alveoli due to a deficiency in granulocyte-macrophage colony-stimulating factor (GM-CSF) activity<sup>1-14</sup> (Fig. 1). This is associated with an abnormal macrophage function and an impaired clearance of surfactant from the lungs. PAP has a prevalence of 3.7 cases per million, a male preponderance (4:1 male/female ratio) and 80% of the cases are reported during the third and fourth decade of life.<sup>6</sup>

Whole Lung Lavage (WLL), introduced in 1960s,<sup>3,4,7,10,15</sup> is still the gold standard treatment.<sup>6</sup> Unilateral WLL, with the lavage performed in each lung in different sessions separated by days/weeks, is the most frequent procedure. However, bilateral sequential WLL in the same treatment session is an attractive alternative, since it is significantly less time consuming, with a reduced amount of patient discomfort and is more cost effective.

Its efficacy has been attributed not only to the removal of lipoproteinaceous material from alveolar spaces, as well as the removal of anti GM-CSF antibodies, alveolar macrophages and type II epithelial cells. This therapeutic procedure is considered when a significant limitation in daily activities is reported by the patient and/or hypoxemia with

a  $pO_2 < 60$  mmHg, a  $P(A-a) O_2 \geq 40$  mmHg and a shunt fraction  $\geq 10\%$  is detected.<sup>12</sup>

In our Hospital, we performed the first WWL in 2010 and after five unilateral WLL, we moved on to a sequential bilateral WWL program.

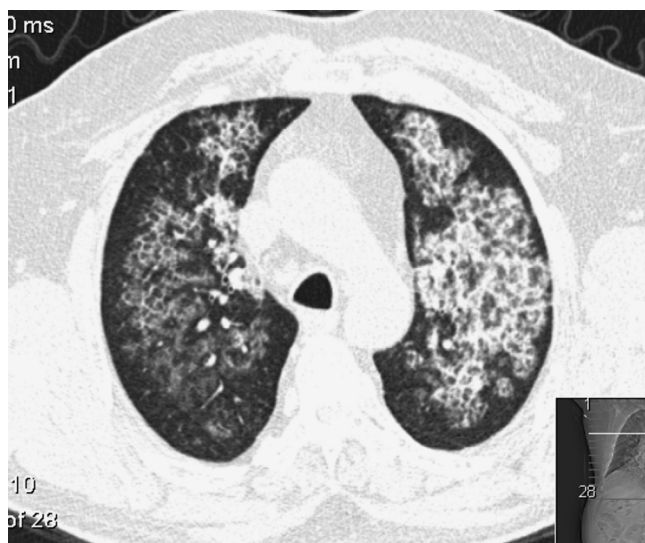
The aim of this report is to describe the bilateral WLL technique and to discuss its safety and effectiveness.

## Methods

In this retrospective study, we collected demographic and clinical data from the medical reports of three adult patients. The standard technique applied was a modified version of the *Royal Brompton Hospital (London) technique* protocol.<sup>6,10</sup>

Throughout the procedure, electrocardiography, pulse oximetry (SatO<sub>2</sub>), invasive blood pressure, central venous pressure (CVP), urine output, capnography, tidal volumes bispectral index (BIS), and central temperature were continuously monitored and arterial blood gases (ABGs) were done hourly. A total intravenous anesthesia (TIVA) was performed to allow the depth of the anesthesia to be managed, independently of ventilatory variations inherent to the procedure. Curarization was maintained throughout the entire procedure. A left double lumen tube (DLT) was introduced, selecting the biggest size possible, to ensure lung isolation and promote ventilation and WLL efficacy. Its correct position was confirmed by fiberoptic bronchoscopy and cuff pressure insufflation measured in order to prevent contralateral leakage from the lavage fluid.<sup>11</sup>

A pre-oxygenation with a FiO<sub>2</sub> of 100% was carried out during 5 min to ensure a correct alveolar denitrogenation and the occurrence of air bubbles, which could impair the removal of the lipoproteinaceous material from the alveoli. One-lung ventilation was started and lung isolation verified. Patients were ventilated by pressure-controlled ventilation, with pressure value under 30 cmH<sub>2</sub>O. The volume of saline to be instilled was calculated by the preoperative measurements of functional residual capacity (FRC). The right lung volume was calculated by 3/5 of the FRC and the left lung by 2/5 FRC (in the first cycle a smaller volume was instilled). The saline should be instilled under gravitational effect from a height not exceeding 40 cm above mid-axillary line, in order to prevent barotrauma and leakage to the ventilated lung.<sup>10</sup> Although some reports describe a 30° lateral decubitus positioning in order to preserve the ratio of ventilation/perfusion of the dependent ventilated lung, this increases the probability of contralateral lung inundation, so we preferred a dorsal decubitus positioning, with a reverse trendelenburg as well as trendelenburg



**Figure 1** CT Scan of a patient with Pulmonary Alveolar Proteinosis. Note the thickened interlobar septa within the opacified parenchyma producing a “crazy paving” pattern.

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