Bilateral partial lung lavage in an infant with pulmonary alveolar proteinosis

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Pulmonary alveolar proteinosis (PAP) is a rare disease in infancy, resulting from abnormalities of surfactant production or decreased catabolism of surfactant. The only effective treatment of the congenital form of PAP is bronchoalveolar lavage. A 4-month-old boy with severe PAP received bilateral partial lung lavage on two occasions resulting in clinical improvement. We performed partial lung lavage using a 3.1 mm flexible fibreoptic bronchoscope introduced through a 4.0 mm tracheal tube under general anaesthesia. The infant did not require extra-corporeal oxygenation during the procedure or postoperative ventilation. This method may offer a feasible option for performing lavage in a resource constrained environment.

Keywords: anaesthetic techniques, bronchoscopy; lung, lavage

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We present a case of congenital pulmonary alveolar proteinosis (PAP) in which bilateral partial pulmonary lavage utilizing a flexible fibreoptic bronchoscope (FB) through a tracheal tube (TT), without the use of extra-corporeal oxygenation, was performed. The infant did not require postoperative high care unit admission or ventilation. This technique has advantages over previously described techniques, especially in a resource constrained environment.

Case report

A 4-month-old boy was referred to the paediatric pulmonology service at our tertiary level hospital in South Africa. The infant presented with a 4 week history of dyspnoea and oxygen dependence. The infant had a male sibling who had died 2 yr previously at the age of 9 months from respiratory failure due to PAP. Physical examination revealed that he weighed 5.3 kg and on nasal prong O2 had moderate respiratory distress, with a ventilatory frequency of 60 bpm and peripheral cyanosis with saturation of 88%. Fine inspiratory crackles could be heard throughout the chest on auscultation. Cardiovascular examination was normal. The liver and spleen were enlarged. Investigations showed low total protein 55 g litre\(^{-1}\) and albumin 18 g litre\(^{-1}\), liver enzymes and full blood count were within normal range. LDH was elevated at 312 U litre\(^{-1}\). Urinary lysine was not increased, 432 nmol per mg Cr. Arterial blood gases on oxygen 2 litre min\(^{-1}\) were pH 7.37, \(P_{CO2}\) 5.03 kPa, and \(P_{O2}\) 5.74 kPa. The chest X-ray showed extensive airspace opacification, predominantly of the lower lobes. A CT of the chest showed bilateral nodular interlobular and peri-bronchovascular interstitial thickening. Microbiological investigations for respiratory fungal, viral, and bacterial pathogens were all negative. HIV testing was negative.

The infant was anaesthetized using propofol total i.v. anaesthesia. Cisatracurium 0.1 mg kg\(^{-1}\) was given to facilitate intubation with a size 4.0 mm uncuffed TT. The lungs were ventilated by hand with an \(F_{I/O2}\) of 0.8–0.9. A mixture of 1% lidocaine and epinephrine was sprayed into the TT for bronchial analgesia. I.V. acetaminophen at 15 mg kg\(^{-1}\) was given.

A well-lubricated 3.1 mm flexible FB was passed through a catheter mount into the bronchial tree; 20 ml aliquots of pre-warmed normal saline 0.9% were lavaged. A total of 160 ml were lavaged into the left lower lobe and 140 ml into the right lower lobe, until the turbid fluid obtained had cleared. The infant remained haemodynamically stable. The emptying phase of lavage was
undergone repeated lung lavage. Survival has been described in a few infants who have anticipated future lavage procedures. In our setting, lung separation sites have to be carefully planned and repaired in anticipation of future lavage procedures. We opted to perform the procedure, he was discharged to the ward on nasal prong O2 and had an uneventful overnight course. Over the following week, oxygenation improved. The procedure was repeated 1 week later and the intraoperative course was similar.

**Discussion**

PAP is a rare condition due to accumulation of surfactant in the alveoli resulting in poor gas exchange and hypoxia. PAP occurs in three forms: congenital, secondary, and acquired. The congenital form comprises a heterogeneous group caused by mutations in genes encoding surfactant proteins for the β chain of the receptor of granulocyte-macrophage colony stimulating factor. Abnormal surfactant protein B gene is the most commonly recognized genetic defect. Early presentation, especially in the neonatal period, is associated with a worse prognosis and high mortality. The only effective treatment of the congenital form is lung lavage; clearance of surfactant is not permanent but may produce clinical improvement. Long-term survival has been described in a few infants who have undergone repeated lung lavage.

Hypoxia and haemodynamic instability occur in patients with PAP undergoing pulmonary lavage. In children, strategies to minimize the risk of hypoxia during lavage can be broadly classified into extra-corporeal oxygenation techniques or pulmonary ventilation with modified lung separation techniques.

The extra-corporeal techniques involve either cardiopulmonary bypass or extra-corporeal membrane oxygenation, either with full bypass or partial bypass. The cannulation sites have to be carefully planned and repaired in anticipation of future lavage procedures. In our setting, the costs and unpredictability of postoperative ICU bed availability precluded the use of extra-corporeal techniques.

Lung separation in children and infants cannot be achieved with a double-lumen tube (DLT) because the paediatric airways are too small to accommodate even the smallest DLT. Modified ventilation techniques used include: bilateral lavage of the lungs through a single-lumen TT during periods of apnoea in a hyperbaric chamber; unilateral lavage with a Swan–Ganz catheter, through a rigid bronchoscope, in a 2.2 kg baby; and selective intubation of a main bronchus with a cuffed TT with unilateral lavage through an FB passed parallel to a TT in children over 5 kg. We opted to perform the partial lung lavage of the most affected lobes with an FB inserted through a TT, using the FB as a bronchial obturator and continuing to ventilate the other lung lobes.

Concern has been raised about normal saline spillage during lavage resulting in desaturation. However, one case report in an adult describes using ventilation of a partially filled lung to optimize surfactant removal without significant desaturation. As significant desaturation only occurred during the emptying phase of lavage but corrected with application of PEEP, an observation previously described, we do not believe that spillage contributed significantly to the intraoperative or postoperative hypoxia.

The prognosis for an infant with congenital PAP is poor. Our aim in performing the procedure was to confirm the diagnosis, obtain respiratory specimens for culture, and produce symptomatic improvement. Using the technique as described, we achieved these without exposing the infant to undue risk. The use of extra-corporeal techniques or intensive care facilities in a patient with such a poor prognosis would have been hard to justify in our resource limited setting. To our knowledge, this is the first description of both this anaesthetic technique and the lavage technique through a TT using the FB as an obturator in an infant with PAP. These techniques were found to be safe and effective, resulting in minimal postoperative respiratory compromise, and may therefore be especially useful in resource constrained settings.

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**References**