CHEST

Official publication of the American College of Chest Physicians

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Chest 2009;136;1678-1681
DOI 10.1378/chest.09-2295

ISSN:0012-3692

CHEST Postgraduate Education Corner

PROCEDURE VIDEOS ONLINE

Whole-Lung Lavage for Pulmonary Alveolar Proteinosis

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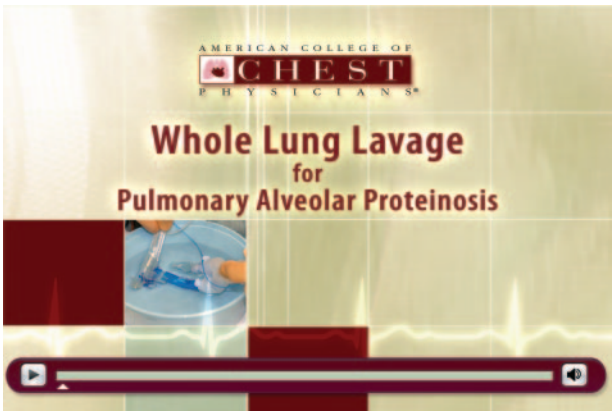
Pulmonary alveolar proteinosis (PAP) is a disease characterized by the deposition of amorphous lipoproteinaceous material in the alveoli secondary to abnormal processing of surfactant by macrophages. Whole-lung lavage often is performed as the first line of treatment for this disease because it is a means to wash out the proteinaceous material from the alveoli and reestablish effective oxygenation and ventilation. Whole-lung lavage is a large-volume BAL that is performed mainly in the treatment of PAP. In brief, it involves the induction of general anesthesia followed by isolation of the two lungs with a double-lumen endotracheal tube and performance of single-lung ventilation while large-volume lavages are performed on the nonventilated lung. Warmed normal saline solution in 1-L aliquots (total volumes up to 20 L) is instilled into the lung, chest physiotherapy is performed, then the proteinaceous effluent is drained with the aid of postural positioning. The sequence of events is repeated until such time as the effluent, which is initially milky and opaque, becomes clear. This procedure results in significant clinical and radiographic improvement secondary to the washing out of the proteinaceous material from the alveoli. The whole-lung lavage video details all aspects of the procedure, including case selection, patient preparation and equipment, a step-by-step review of the procedure, and postoperative considerations. ***(CHEST 2009; 136:1678–1681)***

Abbreviation: PAP = pulmonary alveolar proteinosis

Editor's Note: *In this series, the article is intended to complement a video, available at* [http://chestjournal.chestpubs.org/site/misc/videos/media1/index.html]. *The video contains footage and narration to accompany the text, from clinical background through postprocedural care. The first time you access the video, you may be asked to update your Web browser's Flash Player plugin. If you are not automatically prompted to update the plugin, you can download the latest version of the Flash Player for free by visiting http://www.adobe.com/go/getflash.*



Whole-lung lavage is a large-volume BAL that is performed mainly in the treatment of pulmonary alveolar proteinosis (PAP), a disease that is characterized by the deposition of amorphous lipoproteinaceous material in the alveoli. Patients with

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PAP have a primary or acquired form of macrophage dysfunction that results in abnormal processing of surfactant, and, over the course of time, phospholipids and surfactant apoproteins accumulate in the alveoli.[1–4] Figure 1 shows a representation of the

Manuscript received September 28, 2009; accepted September 29, 2009.

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DOI: 10.1378/chest.09-2295

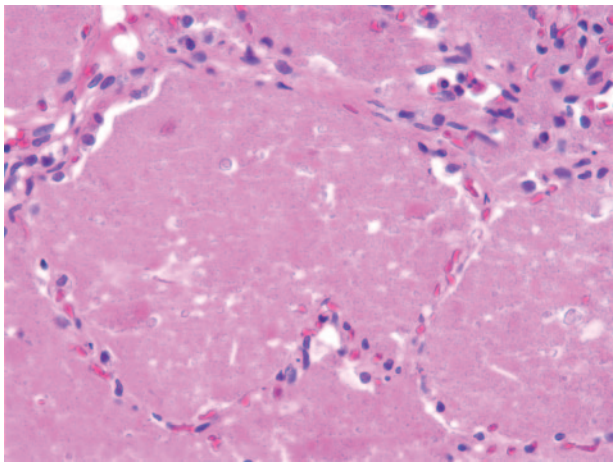

FIGURE 1. Histology of PAP. The classic histologic features of PAP include a well-preserved alveolar structure, the accumulation of amorphous lipoproteinaceous material within the alveoli that stains pink with periodic acid-Schiff stain, and a distinct absence of inflammatory cells.

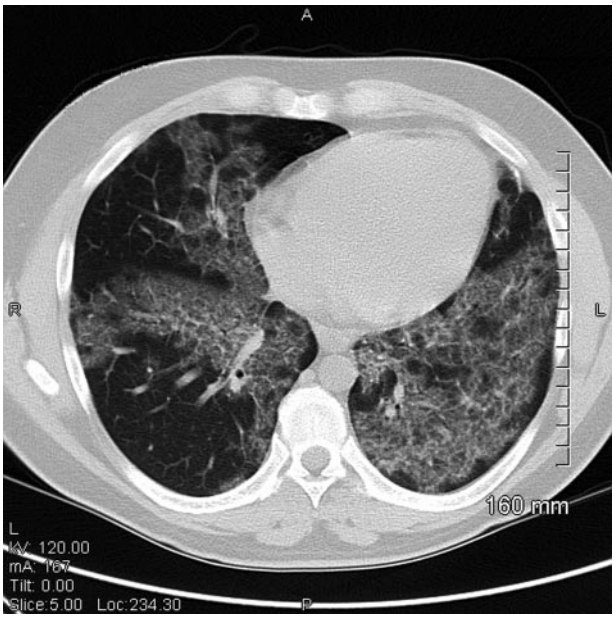

FIGURE 2. CT scan image of PAP. The classic CT scan features of PAP include patchy areas of ground glass with thickening of the interlobular septae. This combination of findings is commonly known as *crazy paving*.

pink amorphous material filling the alveoli as seen in the typical histology of PAP. PAP is divided into primary and secondary, or acquired, forms. The cause of primary PAP is as yet unknown. In the acquired type of PAP, usually there is a readily identifiable cause, such as occupational dust exposure, atypical infection, hematologic malignancy, or allogenic bone marrow transplantation.

PAP presents in a subacute fashion with a gradual onset of symptoms. Approximately 33% of patients are asymptomatic at presentation, whereas others present with a myriad of symptoms, including dyspnea, dry cough, fever, malaise, and respiratory failure.[5] The physical manifestations of PAP are quite nonspecific, and pulmonary function test results are consistent with a restrictive defect. The combination of septal thickening and ground-glass opacities have led to the name given to the overall radiographic appearance, *crazy paving*. A representative CT scan image is shown in Figure 2.[6]

INDICATIONS

Whole-lung lavage results in significant symptomatic and radiographic improvement in patients with PAP. The indications of whole-lung lavage include pathologic diagnosis of PAP obtained by either transbronchial lung biopsy or open lung biopsy, severe dyspnea or hypoxemia, Pao_2 at sea level of < 65 mm Hg, and alveolar-arterial oxygen tension gradient of ≥ 40 mm Hg or a shunt fraction exceeding 10% to 12%.

CONTRAINDICATIONS

There are few contraindications, the most common being uncorrectable blood dyscrasias, anesthetic risks, and cardiopulmonary instability. The cardiopulmonary instability in particular is a relative contraindication because the whole-lung lavage may lead to a fairly rapid improvement in oxygenation once the proteinaceous material is washed out of the alveoli.

PREPROCEDURAL CONSIDERATIONS

Informed Consent

Informed consent is obtained from the patient, after a discussion about the risks and benefits of the procedure. The risks associated with whole-lung lavage include those specifically related to the general anesthesia, double-lumen endotracheal intubation, and mechanical ventilation necessary for the procedure; those risks related to the lavage itself; and, finally, the need for continued postprocedure ventilatory support and monitoring in a critical care setting.

Equipment

Multiliter bags (up to 15 L to 20 L) of normal saline solution are used to lavage the lungs, and, in order to maintain the patient's core temperature, the solution is run through a blood warmer. In addition, a warming blanket (Bair Hugger; Arizant, Inc; Eden Prairie, MN) is placed over the patient during the procedure. IV tubing with a stopcock forms the

lavage and drainage limbs, and these are connected to the bronchial lumen of a dual-lumen endotracheal tube. Multiple drainage receptacles are necessary to collect the effluent. A representation of the circuit is shown in Figure 3.

We recommend that patients be placed on a procedure table that can be manipulated electrically to allow for Trendelenburg and reverse-Trendelenburg positioning in order to facilitate filling and drainage. A bronchoscope with an adequate suction channel is used to aspirate any residual lavage fluid as well as to verify endotracheal tube position.

Procedure

The procedure is performed under general anesthesia. After the patient is intubated with a double-lumen endotracheal tube, flexible bronchoscopy is performed to confirm the appropriate tube placement. Both the bronchial and the tracheal balloons are inflated to isolate the lungs, and mechanical ventilation is initiated. Lung isolation is reconfirmed by immersing the end of each lumen of the endotracheal tubes in water and observing for air bubbles while the other lung is being ventilated. The patient then is turned to a lateral decubitus position, with the lung being lavaged in the nondependent position (up). Because the procedure may last a few hours, meticulous care is taken to avoid ischemic complications to the extremities by placing supporting pillows in the dependent axilla, under the head, and between the thighs. Before initiating the procedure, the tubing limb to the treatment lung is opened to allow for the lung to "degas" and ensure appropriate oxygenation during single-lung ventilation.

With the patient's head end slightly elevated (*ie*, the reverse-Trendelenburg position), warm (37°C) normal saline solution is allowed to flow into the nondependent lung through the endotracheal tube limb. After 1 L of normal saline solution has flowed in, the inflow tubing is clamped, and chest percussion is performed for approximately 4 to 5 min. The foot of the bed then is elevated (Trendelenburg position), and the clamp on the outflow tube is opened to drain the effluent by gravity into a container. When the flow diminishes, the outflow tube is clamped, the head end is elevated, and the process is repeated. The initial effluent is milky in appearance and tends to settle on standing. The fluid becomes progressively less opaque, and after 10 to 15 lavages the effluent is clear. Ongoing charting of installed and drained volumes is important to minimize the risks related to overdistention of the alveoli from the residual lavage fluid, and pleural fluid samples are sent for cytologic and microbiological analysis. The procedure is terminated at this point.

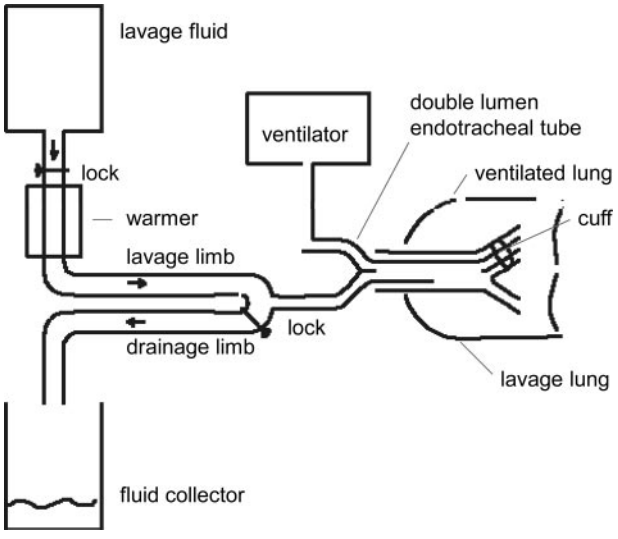

Figure 3. Whole-lung lavage equipment and setup. The whole-lung lavage circuit is as follows: the lavage fluid is hung in multi-liter bags from an IV pole and run through a warmer. A lock is located between the lavage fluid and warmer to control the volumes and timing of the lavage fluid being instilled. One lung is ventilated while the other is being lavaged. The lavage and drainage limbs are in continuity with the lavage lung only. There is a lock in the drainage limb to control the timing of drainage into the fluid collector.

Postprocedural Considerations

Once the residual saline solution solution is aspirated, ventilation of the lavaged lung is resumed. The patient is turned onto his or her back and, ideally, extubated in the operating room. Once stable, he or she is transferred to the recovery room and then back to the hospital ward or ICU for monitoring overnight. Disposition depends on the preoperative disease severity and patient comorbid illness and on the postoperative ventilatory status. A radiograph is performed to ensure that the patient did not sustain any complications from the procedure, such as pneumothorax or pleural effusion. In the case of bilateral disease, the contralateral lung may be lavaged in 24 to 48 h if the patient remains stable. If a pneumothorax is found on the side of the treated lung and a subsequent lavage is planned, a pleural drain may need to be placed to prevent intraprocedural tension pneumothorax.

Complications

Whole-lung lavage generally is well tolerated. The major potential complication is intraoperative refrac-

tory, which tends to be more common while the first lung is being lavaged. Low oxygen saturations (percentages in the high 70s to 80s) are not uncommon early in the procedure; however, they generally improve throughout the case without any other intervention. Hyperbaric oxygen, cardiopulmonary bypass, and temporary venovenous extracorporeal gas exchange all have been used in the past, but in more recent studies,[7] they have not been found to be necessary in most cases. Other more common and less dangerous complications include pneumothorax, pleural effusion, and hydropneumothorax, which can be avoided by meticulous charting of the infused saline solution and the output, and by taking care not to allow instilled fluid to exceed the fluid drained by more than a few hundred milliliters in consecutive lavages. Spillage of lavage fluid into the contralateral (ventilated) lung also may occur and should be considered if an imbalance is noted between the instilled and the drained volumes. If spillage is a concern, then any excess fluid should be aspirated from the ventilated lung and the dual-lumen endotracheal tube should be readjusted to ensure no further ongoing leak.

Outcomes and Follow-up

In the majority of reported cases, a significant clinical improvement was reported following whole-lung lavage for PAP. Approximately 15% of patients will have a relapsing course, requiring repeat procedures.[7] Replacement of granulocyte-macrophage colony-stimulating factor administered by daily subcutaneous injection has resulted in an increase in alveolar macrophage production, a reduction in the risk of opportunistic infections, and a reversal of the defect leading to surfactant accumulation within the alveoli and may be used in refractory cases.[1–4]

Acknowledgments

Financial/nonfinancial disclosures: The authors have reported to the ACCP that no significant conflicts of interest exist with any companies/organizations whose products or services may be discussed in this article.

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