

Dynamic Contrast-Enhanced Magnetic Resonance Lymphangiography and Percutaneous Lymphatic Embolization for the Treatment of Plastic Bronchitis

► Interventional radiologists at Penn Medicine have developed a program to combine advanced lymphatic imaging and percutaneous interventions for the diagnosis and treatment of plastic bronchitis in adults.

Plastic bronchitis (PB) is an uncommon lung disease defined by the production of occlusive branching casts from the tracheobronchial tree (Figure 1). Casts are composed of lymphocytes or proteinaceous materials and vary in magnitude and branching extent from small bronchial segments (occurring in asthma or pulmonary infection) to the entire lung (>30.5 cm) in individuals with lymphatic disease.

PB is thought to arise when elevated central venous pressures and abnormal flow in the thoracic duct cause retrograde flow into the pulmonary lymphatic channels that seeps into the airways and engorges the lung parenchyma. Principally affecting individuals with surgically-repaired congenital heart disease, PB has been identified in patients with sickle cell disease, chronic pulmonary disease and the primary lymphatic disorders, as well.

Under the direction of **Drs. Maxim Itkin and Gregory Nadolski**, interventional radiologists at Penn IR have developed an evidence-based initiative to diagnose and treat PB that combines dynamic contrast-enhanced magnetic resonance lymphangiography (DCMRL) with percutaneous lymphatic embolization.

DCMRL was developed to image the central lymphatic system, and involves bilateral injection of gadolinium into the inguinal lymph nodes and image acquisition using time-resolved central k-space dynamic T1-weighted magnetic resonance imaging. Lymphatic embolization employs a transabdominal catheter to place coils coated with organic endovascular glue into affected lymphatic vessels to block leakage.

In a landmark 2016 study,¹ Dr Itkin and colleagues used DCMRL to demonstrate the long-suspected finding that abnormal pulmonary and mediastinal lymphatic flow from the thoracic duct is a cause of plastic bronchitis in adults. The same study established the efficacy of percutaneous transabdominal lymphatic embolization as a treatment modality for adults with plastic bronchitis.

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References

1. Itkin MG, McCormack FX, Dori Y. Diagnosis and Treatment of Lymphatic Plastic Bronchitis in Adults Using Advanced Lymphatic Imaging and Percutaneous Embolization. *Ann Am Thorac Soc* 2016;13:1689–1696.



► **Figure 1:** Plastic bronchitis is marked by the formation and expectoration of large gelatinous or rigid branching airway ‘casts.’ Abnormal pulmonary lymphatic flow resulting in molding of congealing lymphatic fluids in the airway has been documented as a cause of the disease.

CASE STUDY

Mr. R, a 67-year-old man, was referred to Penn Interventional Radiology for lymphangiogram and thoracic duct embolization following a 20-year history of pulmonary disease, including pulmonary alveolar proteinosis, and respiratory complications (e.g., chylothorax, chronic respiratory failure, plastic bronchitis). Despite obesity (BMI 30.78 kg/m²) and a decades-long daily requirement for oxygen, Mr. R was managing relatively well until several months prior to his arrival at Penn, when he began to experience progressive exertional dyspnea, orthopnea and cough productive of casts.

At presentation, Mr. R recounted a long series of procedures to clear his airways, including bronchoscopies, vest treatments, hypertonic saline nebulizer treatments, albuterol inhalation and guaifenesin therapy. His most recent procedure, an attempted thoracic duct embolization at a hospital in his home state, was abandoned after 12 hours.

Following this experience, Mr. R was treated for pulmonary nodular amyloidosis (PNA) and his airway clearance regimen increased. To address his persistent oxygen requirement, he had CT pulmonary angiography, which demonstrated acute bilateral pulmonary emboli and a large left-sided pneumothorax, and was discharged on room air at rest and 3L oxygen with ambulation. He continued to cough up small amounts of cast-like material and to experience orthopnea. At this point, arrangements were begun to list him for lung transplant.

Seeking an alternative, Mr. R was referred by his primary care provider to Penn Interventional Radiology and the Pulmonary Service at Penn Medicine for consideration of dynamic contrast MR lymphangiogram with possible thoracic duct embolization.

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CASE STUDY (Continued from front page)

The Procedure ~ Mr. R's procedure followed a day-long transport by vehicle from his home in the southern United States, and was preceded at Penn Medicine by admission to OBS, followed by admission to Pulmonary Services.

Under general anesthesia, Mr. R first had intranodal lymphangiography to identify the cisterna chyli and thoracic duct. The cisterna chyli, which sits near the base of the thoracic duct, was then accessed transabdominally via percutaneous means under fluoroscopic guidance with a 22g chiba needle, and a wire placed in the thoracic duct to guide a microcatheter.

Contrast dye was then injected to evaluate the pulmonary lymphatic system (Figure 2), followed by blue dye and bronchoscopy to confirm the leakage and cleanse the lungs of casts. Endovascular glue and coils were then introduced through the catheter to embolize the thoracic duct, and the catheter removed.

Mr. R remained in the hospital for three days and was discharged to home, where he began a full and dramatic recovery. Within weeks of his procedure, his respiration normalized to the point that home oxygen therapy was no longer necessary.

ACCESS

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► **Figure 2:** Thoracic duct injection with contrast demonstrating abnormal pulmonary lymphatic flow in a patient with plastic bronchitis.

FACULTY TEAM

A national leader in research, education and patient care, Penn Interventional Radiology is also one of the oldest and largest IR programs in the United States. In addition to performing more than 12,000 procedures annually, Penn IR has an inpatient admitting service, inpatient consult service and daily outpatient consultation and follow-up clinic.

Treating Plastic Bronchitis at Penn Medicine

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