New source of lymphatic system “leak” discovered in children with rare complication from open heart surgery

Liver lymphatic system connection with chest opens treatment options for certain plastic bronchitis cases.

WILMINGTON, Del. (November 11, 2020)– Interventional radiologists with Nemours Children’s Health System have identified a new source of abnormal lymphatic flow between the liver and the lungs that may be responsible for some cases of plastic bronchitis. Plastic bronchitis is a rare but serious late complication in patients with congenital heart disease who had Fontan surgery. A report detailing the discovery of this fluid leak, and successful treatment of two cases was published in *European Heart Journal.*

In plastic bronchitis, lymph fluid drains into the lungs, blocking patients’ airways and causing difficulty breathing. Recent advances in the use of dynamic contrast-enhanced magnetic resonance lymphangiogram have identified that in 94% of patients, the cause is identified as lymph fluid draining from the thoracic duct that can be successfully treated by blocking or embolizing this duct. However, in a subset of patients, the leak cannot be identified and treated.

> *“Through sophisticated lymphatic imaging techniques, we have identified a new source of lymph fluid leak from the liver to the chest in patients with plastic bronchitis, opening up treatment options where there previously were none,” said Deborah A. Rabinowitz, MD, an interventional radiologist with Nemours Children’s Health System in Wilmington, Del.*

“Through sophisticated lymphatic imaging techniques, we have identified a new source of lymph fluid leak from the liver to the chest in patients with plastic bronchitis, opening up treatment options where there previously were none,” said Deborah A. Rabinowitz, MD, an interventional radiologist with Nemours Children’s Health System in Wilmington, Del. “This discovery signals a connection between the liver’s lymph system and chest structures that could explain pulmonary symptoms in patients with other conditions, such as congestive heart failure and liver cirrhosis.”

The case report details the care of two teen boys who had previously undergone Fontan surgery, an open heart surgery which directs blood flow to the lungs. The first patient was transferred to Nemours/Alfred I. duPont Hospital for Children after unsuccessful identification of lymph fluid leak. Further lymphatic imaging identified the leaking duct from the liver. Despite the complexity of the liver’s lymphatic anatomy, the abnormal pathway was identified and embolized to treat this patient’s condition.

The second patient was transferred to Nemours for emergent care for a persistent cough and fatigue. Once diagnosed with plastic bronchitis, the team did not identify the more common leaks in the thoracic ducts. But based on the experience with the first patient, the second patient underwent liver lymphangiogram, which identified a similar leak from the liver to the airway. This discovery enabled the team to embolize the leaking duct with immediate relief of his symmptoms.

This discovery improves our understanding of the role of the liver as a source of plastic bronchitis in select patients, and advances an innovative and effective treatment approach.

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About Nemours Children’s Health System. Nemours is an internationally recognized children's health system that owns and operates the Nemours/Alfred I. duPont Hospital for Children in Wilmington, Del., and Nemours Children's Hospital in Orlando, Fla., along with outpatient facilities in five states, delivering pediatric primary, specialty, and urgent care. Nemours also powers the world’s most-visited website for information on the health of children and teens, KidsHealth.org and offers on-demand, online video patient visits through Nemours CareConnect.

Established as The Nemours Foundation through the legacy and philanthropy of Alfred I. duPont, Nemours provides pediatric clinical care, research, education, advocacy, and prevention programs to families in the communities it serves.

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