

Diagnosis and Treatment of Protein Losing Enteropathy using Liver Lymphangiography and Hepatoduodenal Lymphatic Embolization

► Penn interventional radiologists are performing embolization procedures in combination with novel lymphatic imaging modalities, such as liver lymphangiography and MR lymphangiography to ameliorate protein depletion in patients with protein losing enteropathy (PLE).

PLE is a condition defined by rapid loss of serum proteins into the intestinal lumen. The condition can be caused by many GI disorders, as well as by a variety of lymphatic congestion disorders. In the latter, PLE is the result of leakage from small gaps in the intestinal lymphatic system, particularly at retroperitoneal vessel-bound malformations in the region of the liver and duodenum. The highest concentration of the proteins in the lymph is in liver lymph (90% of the blood level) and for this reason, in most of cases of lymphatic PLE, the abnormal communications between the liver lymphatics and intestine play an important role.

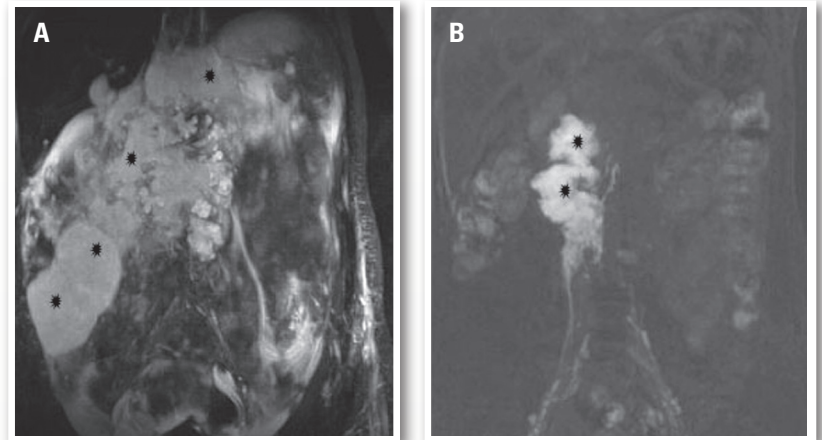
Reductions in albumin (hypoalbuminemia) result in reduced osmotic pressure of the plasma allowing escape of the fluid from the vessels into soft tissue, causing tissue swelling, and ascites. Loss of some some blood proteins can result in coagulopathy.

PLE is confirmed by a decrease in albumin and protein levels in the blood and plasma, and by assessing levels of a sensitive protein marker, alpha-1-antitrypsin, in the feces. At Penn Interventional Radiology, these procedures are complemented by dynamic contrast MR lymphangiography and liver lymphangiography in suspected PLE.

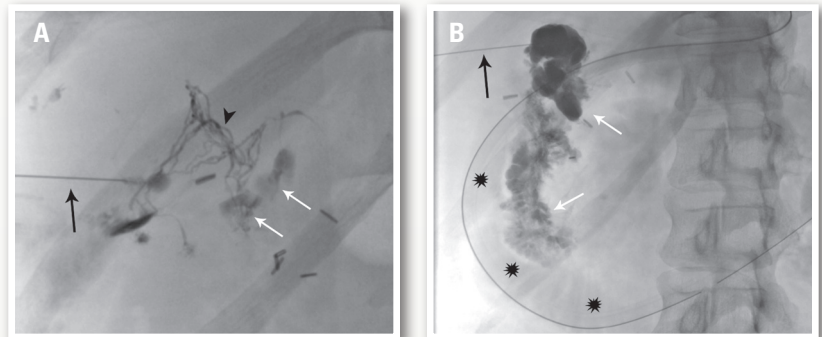
Treatment of PLE at Penn Medicine

The treatment of PLE has evolved dramatically at Penn Interventional Radiology with the development of a technique that employs a biological glue to seal leaky lymphatic vessels. Diluted with an oil based contrast agent to allow visualization under fluoroscopic guidance, the glue is injected through a small bore needle into the hepatic lymphatics, where it propagates, spreads and eventually blocks the holes from which protein is escaping into the gut.

Recent studies have confirmed that liver lymphatic embolization results in sustained improvement of serum albumin levels and the symptoms of PLE in patients with radiographic and endoscopic confirmation of lymphatic leakage into the duodenum.



► **Figure 1:** MRI lymphangiography. (A) Unenhanced coronal T2 MRI sequence of the abdomen demonstrates multiple large T2 hyperintense lymphatic masses (black stars). (B) Coronal MR sequence following injection of contrast through bilateral inguinal lymph nodes demonstrates only partial opacification of these masses (black stars).



► **Figure 2:** Technical steps of fluoroscopic lymphangiography. (A) 25 G needle (black arrow) positioned in the periportal space. The liver lymphatic ducts (black arrowhead) are connected to subhepatic lymphatic mass (white arrows). (B) Injection of the contrast into subhepatic mass through 25 G needle (black needle), demonstrates the progression of the contrast into the periduodenal mass (white arrows) and leakage of the contrast into duodenum (black stars). Note the oro-duodenal tube positioned to provide negative suction in the duodenum.

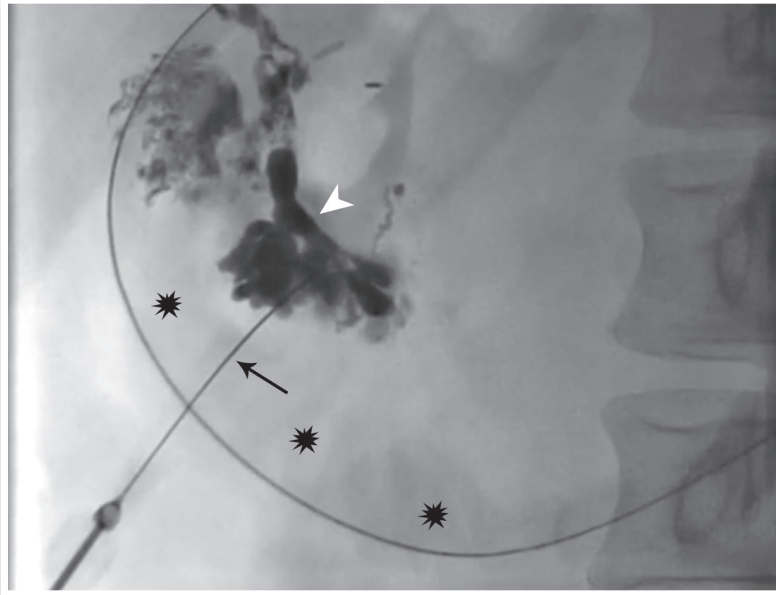
CASE STUDY

Mrs. M, a 47-year-old woman, presented to Penn Interventional Radiology for evaluation 11 years after the partial resection of several retroperitoneal cystic lymphangiomas. The surgery provided relief from her symptoms of abdominal bloating and pain, but at current presentation, she complained of worsening abdominal bloating, development of bilateral foot edema, postprandial abdominal cramping and significant diarrhea.

Mrs. M's serum albumin level was 2.3 g/dL (normal range 3.5-5.1 g/dL). Suspecting PLE, an alpha 1 anti-trypsin level assessment was performed, which measured 3.88 mg/g (0.032-0.344 mg/g), confirming this diagnosis. Given the presence of cystic lymphangiomas, and the absence of primary intestinal or hepatic disorders, Mrs. M was categorized as having primary intestinal lymphangiectasia.

(Continued on back)

CASE STUDY (Continued from cover)



► **Figure 3:** Fluoroscopic image of n-BCA glue injection into the periduodenal mass (white arrowhead) through 25 G needle (black arrow). Note the contrast in the duodenum (black stars).

After self-reported diet modifications, described by Mrs. M as a high protein “celiac” diet, her symptoms improved. However, one year later her diarrhea, edema and bloating worsened again, prompting her to seek treatment. At her second consultation, Mrs. M’s albumin level had fallen to 1.7 g/dL.

Dynamic contrast MR lymphangiography (DCMRL) at this time demonstrated small volume ascites and large lymphatic malformations in the peritoneum and retroperitoneum. Because these masses were only partially opacified following contrast injection into inguinal lymph nodes (Figure 1), and the retroperitoneal lymphatic masses were close to the duodenum and liver, the source of the leak was presumed to be a result of abnormal lymphatic communication between these organs.

A liver lymphangiography and hepatoduodenal lymphatic embolization procedure was planned. Under continuous ultrasound guidance, intrahepatic lymphatic channels were accessed with a 25 G spinal needle (BD, Franklin Lakes, NJ). Iodinated contrast was then injected into the liver lymphatics until multiple large periduodenal lymphatic masses opacified (Figure 2A).

An oroduodenal tube with added side holes was then placed with its tip in second and third portions of the duodenum and placed on suction (-20 mm Hg), to create a negative pressure gradient. This exposed the underlying leak, which appeared when contrast escaped into the duodenum from the periduodenal lymphatic masses (Figure 2B).

The opacified periduodenal masses were then accessed under fluoroscopic guidance with a 25 G spinal needle and embolized with a 3:1 mixture of Lipiodol and n-BCA glue (TRUFILL®, Codman Neuro, MA) (Figure 3).

The following day, Mrs. M reported cessation of her diarrhea, and two days after the procedure she was discharged to home. Two weeks later, she reported near complete resolution of soft tissue edema, relief of abdominal pressure and return of regular bowel movements. Her weight decreased from 135 lbs. to 116 lbs. and her albumin increased from 1.7 g/dL to 2.7 g/dL. At present (four months after the procedure) she continues to report stable relief of her symptoms.

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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 Interventional Radiology has an inpatient admitting service, in-patient consult service and daily outpatient consultation and follow-up clinic.

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