Diagnostic (and Therapeutic) innovations to Accurately Map the Lymphatic System

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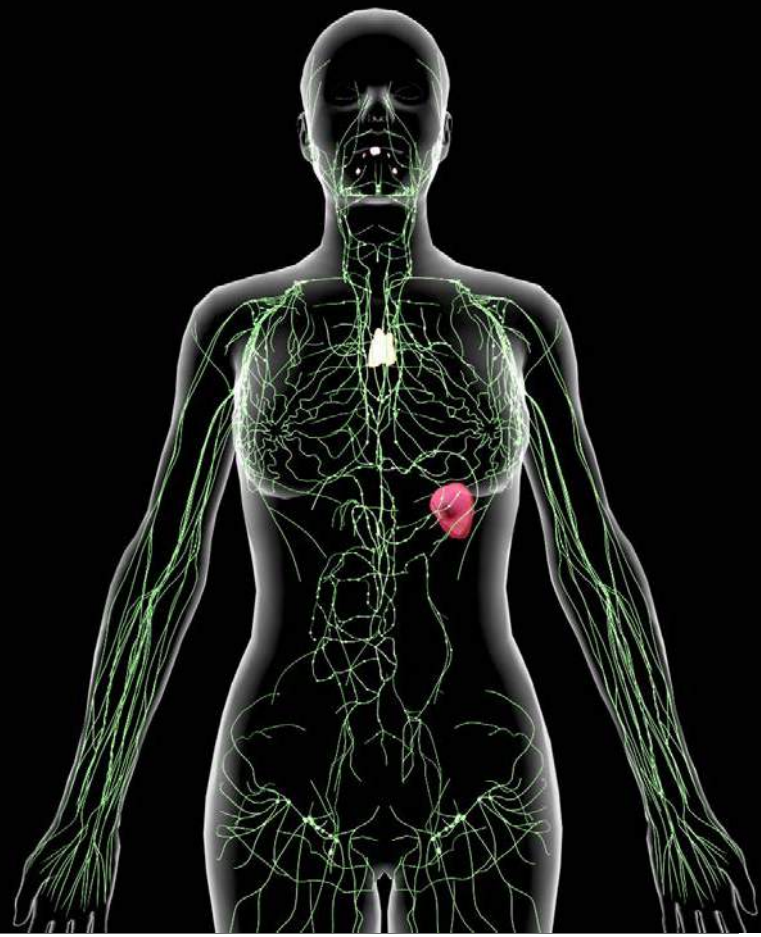

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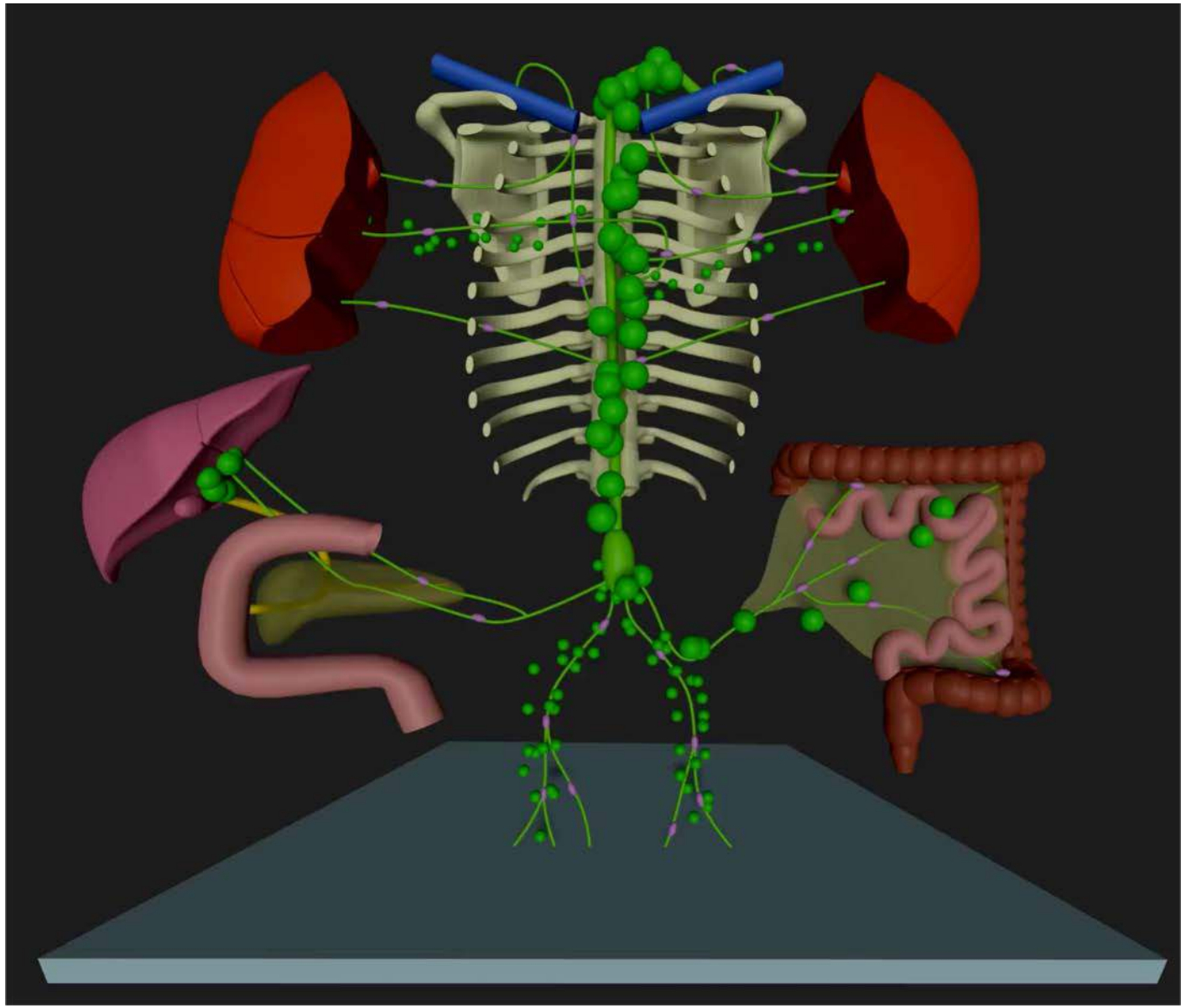
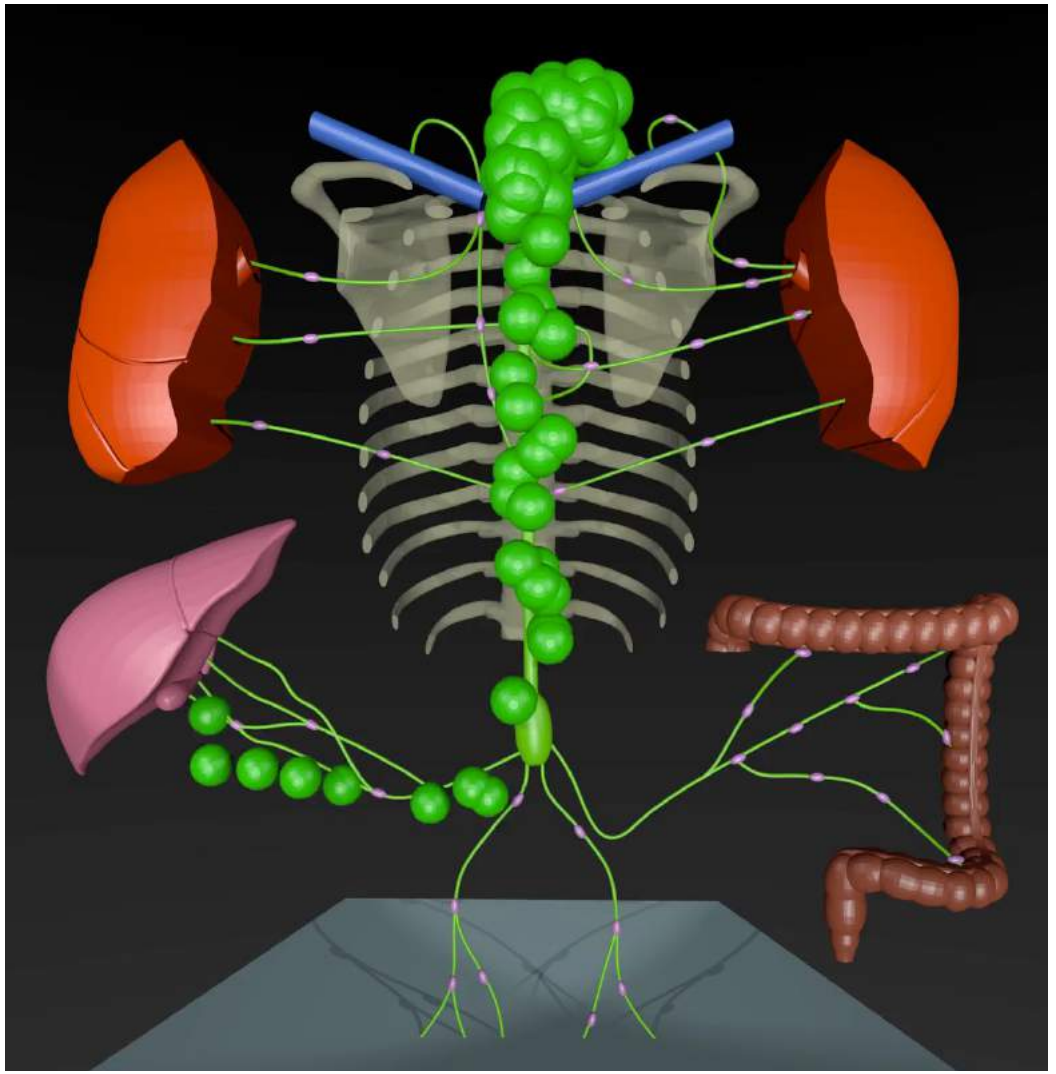
Disclosure

- None

Lymphatic Anatomy and Flow



Elevated Thoracic Duct Pressure in Fontan Patients

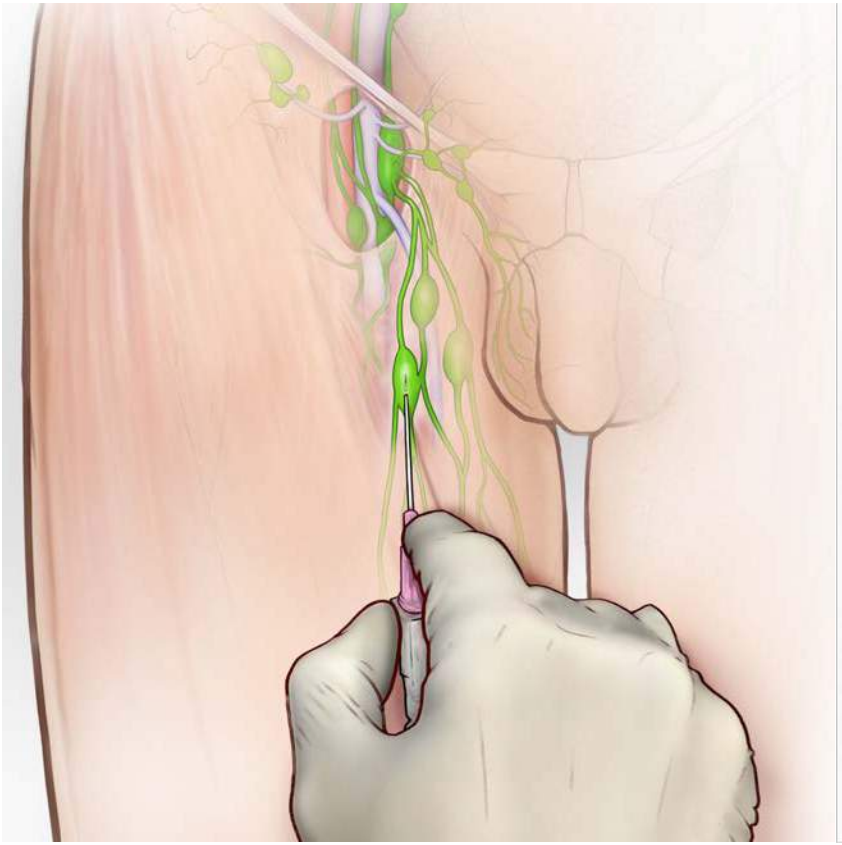


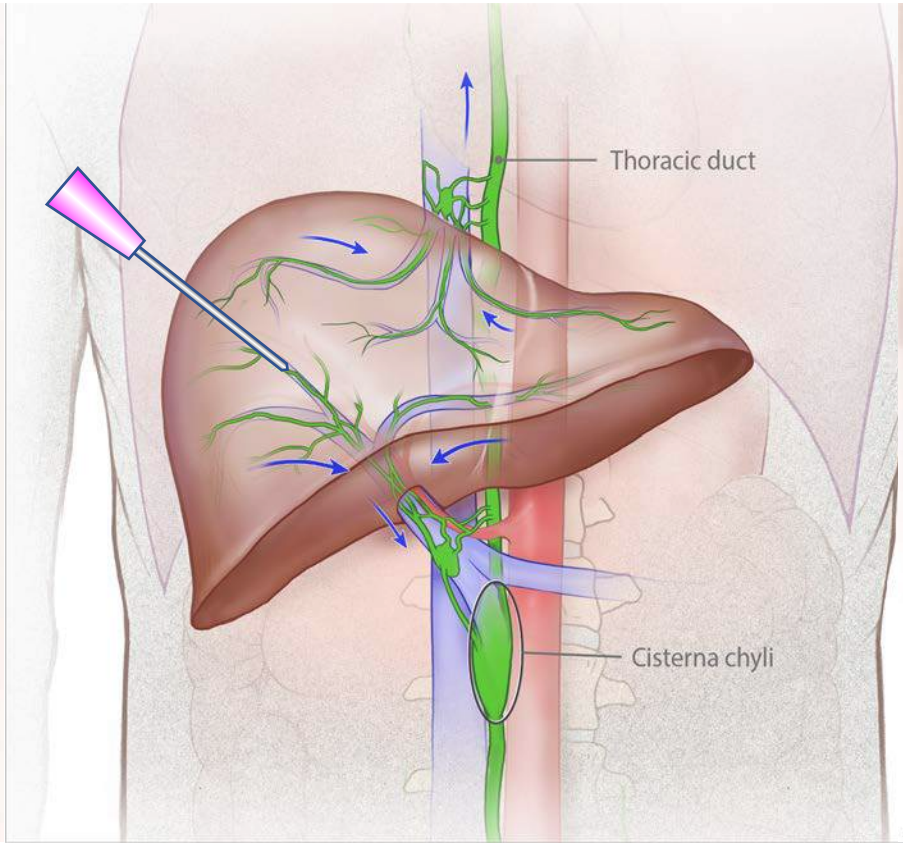
Manifestations of lymphatic dysfunction

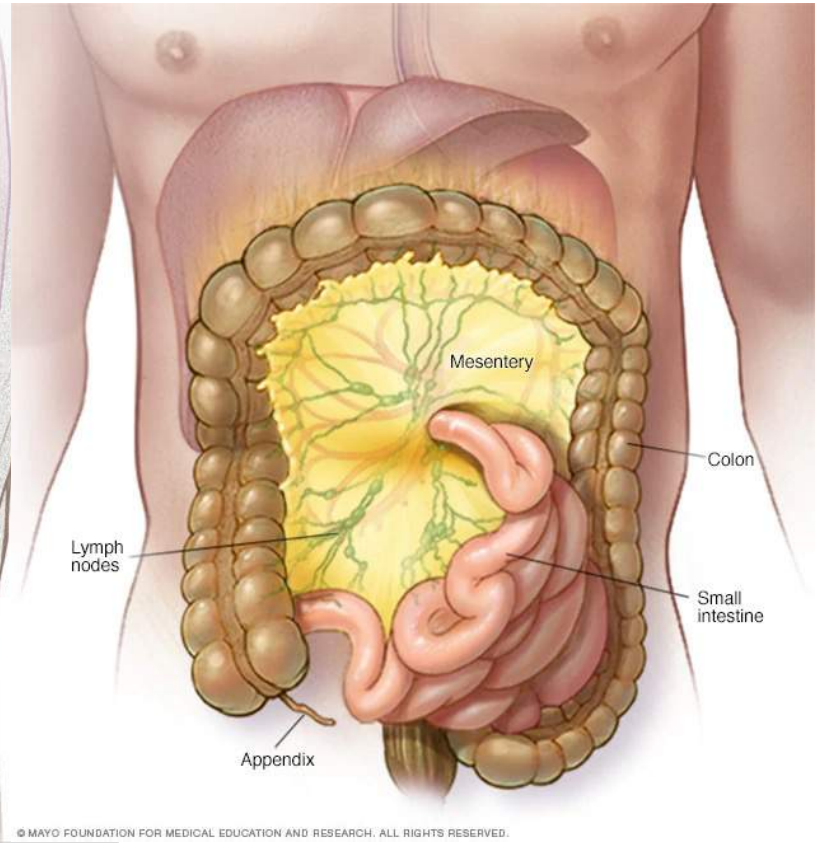
Malfunction in the ability of the lymphatic system to properly remove interstitial fluid

- Lymphedema
- Ascites
- Pleural effusions (chylothorax)
- Chylopericardium
- Plastic Bronchitis
- Interstitial lung disease
- Protein losing enteropathy

Intranodal, Intrahepatic, intramesenteric Lymphangiography







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(Dori et al. Radiology 2014, Biko et al. Eur J. Rad 2019)

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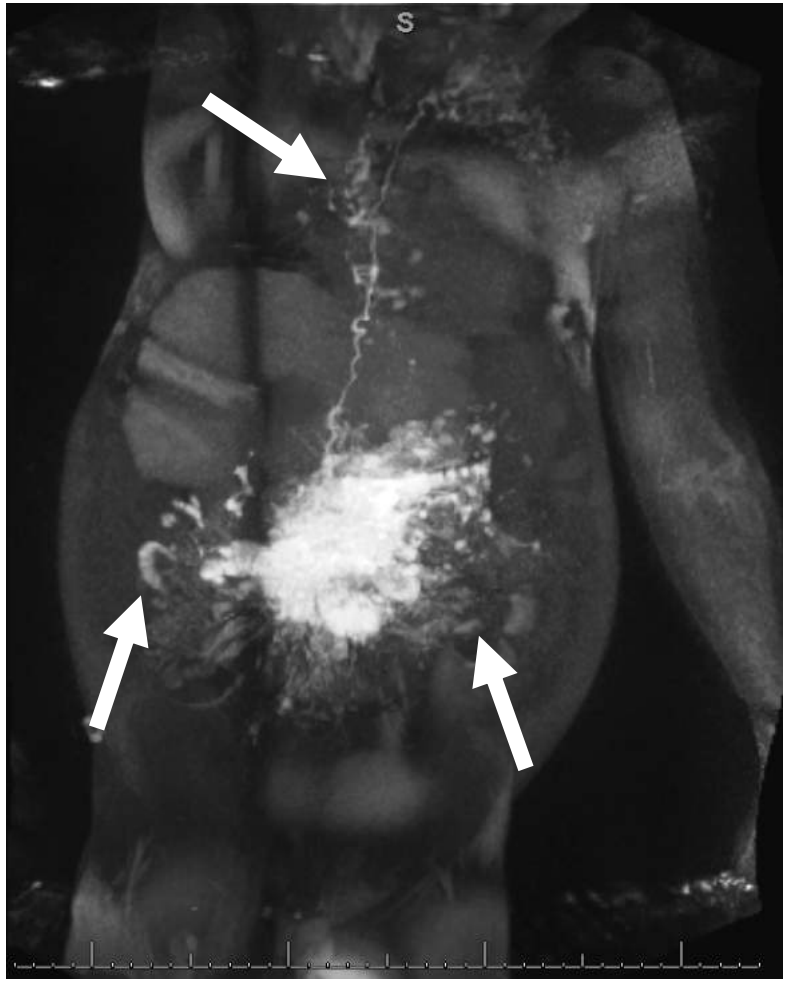
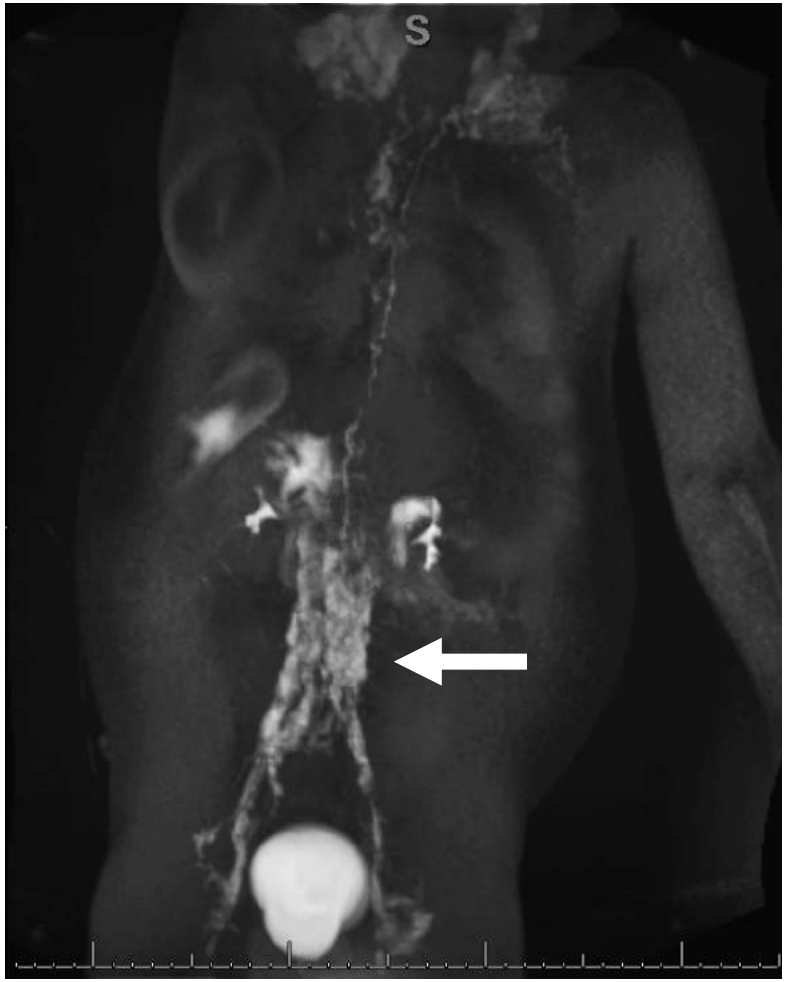
Multicompartment Imaging

IM-DCMRL

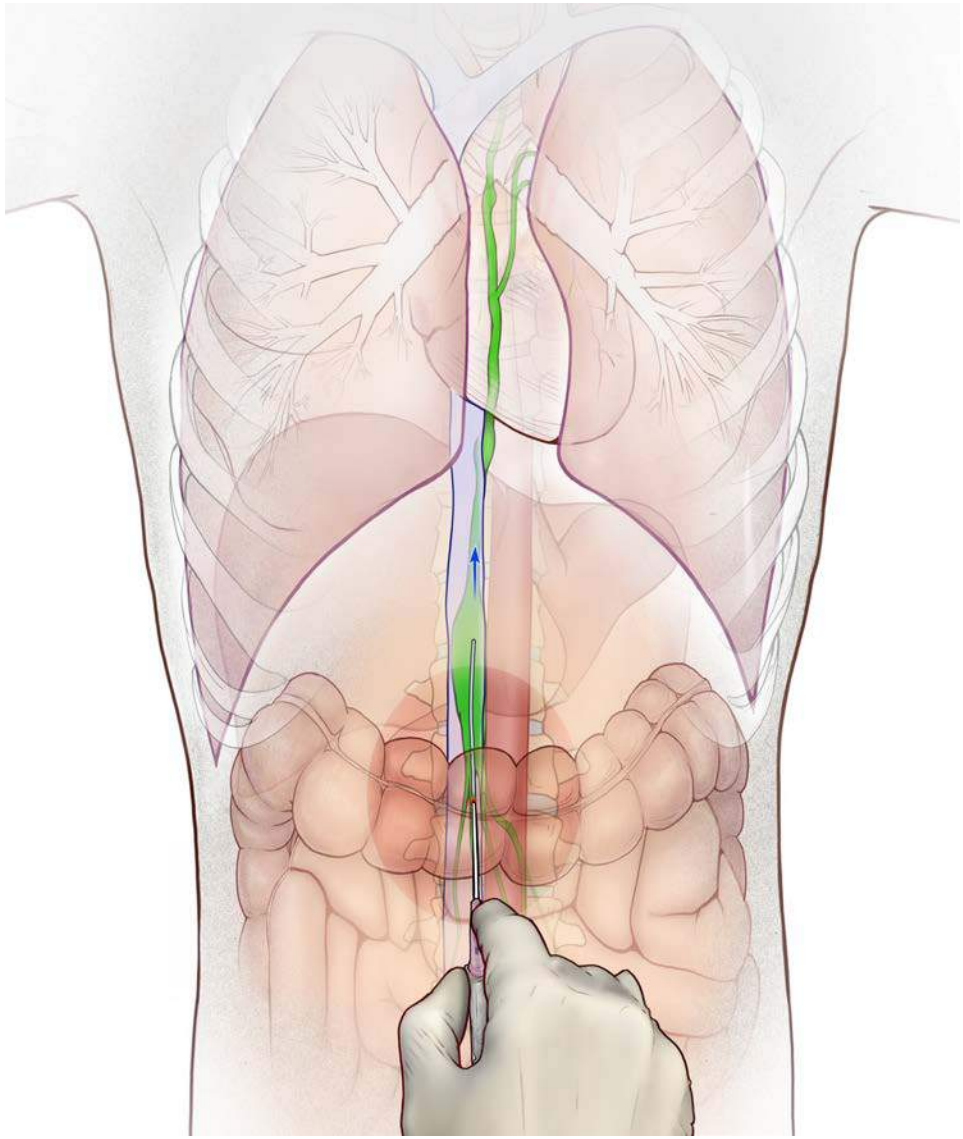


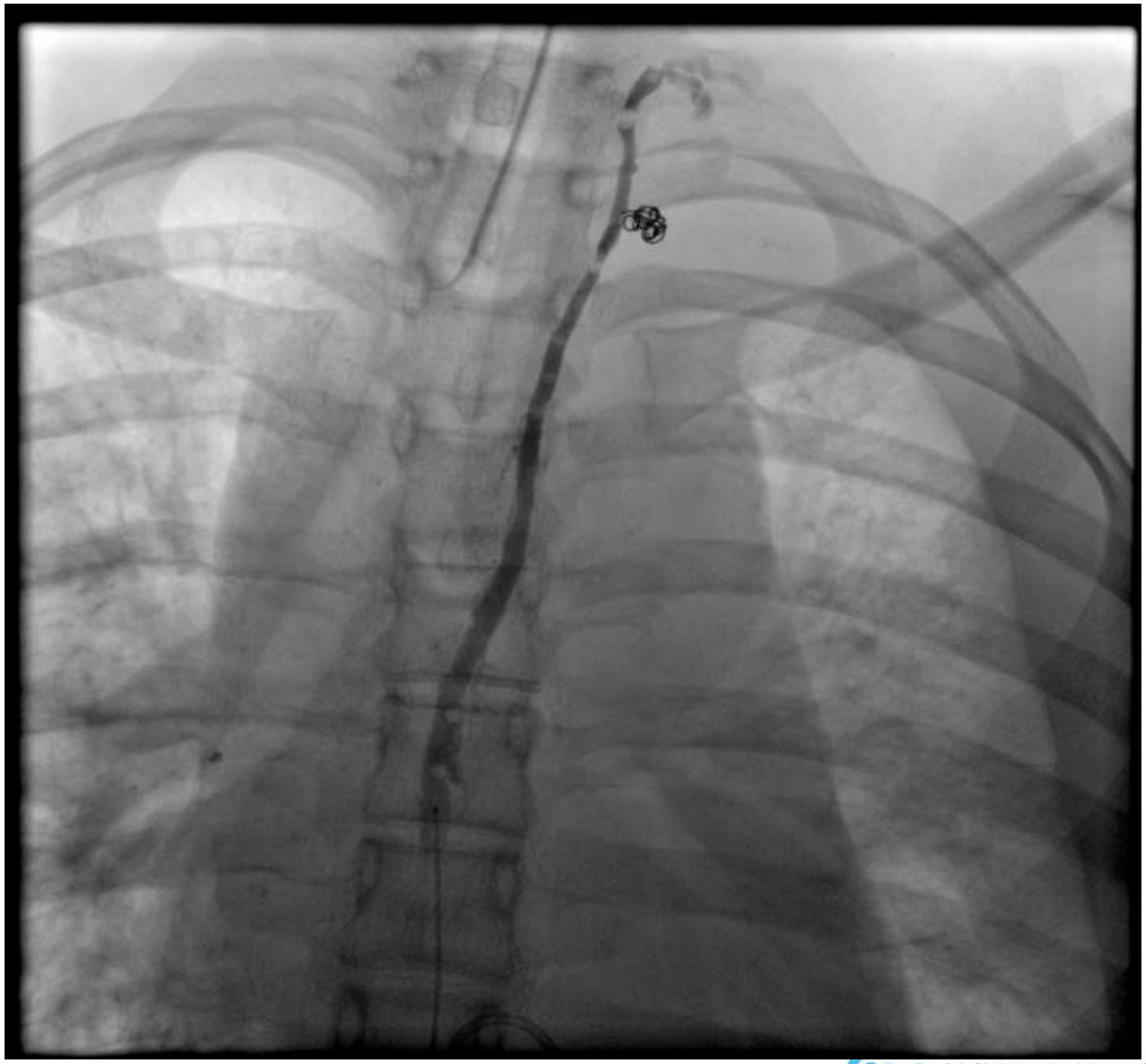
IH-DCMRL

IN-DCMRL



Percutaneous Transabdominal Lymphatic Access





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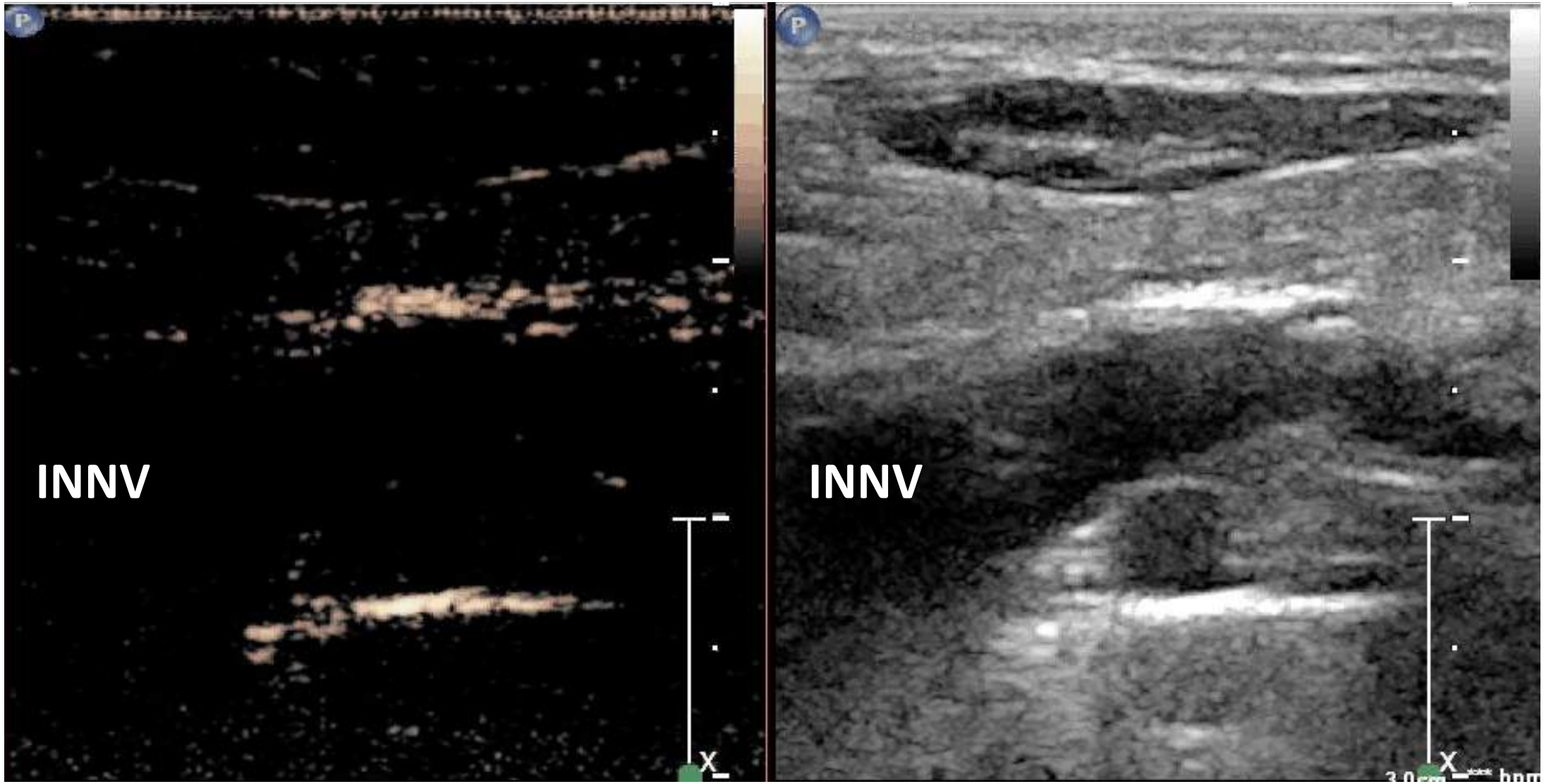
TD Outlet Patency with US Contrast IN Lymphangiogram



(Mejia et al. JVIR 2020)

Lymphatic Flow Disorders: Treatment

- Interventional
  - Thoracic Duct Embolization (TDE)
  - Selective lymphatic duct embolization (SLDE)
  - Lymphatic decompression
  - Balloon occlusion or dilation
  - Stent dilation or exclusion

- Surgical
  - Lymphovenous anastomosis (LVA)
  - Pleurectomy (with directed embolization)
  - Pleurodesis
  - INNV rerouting

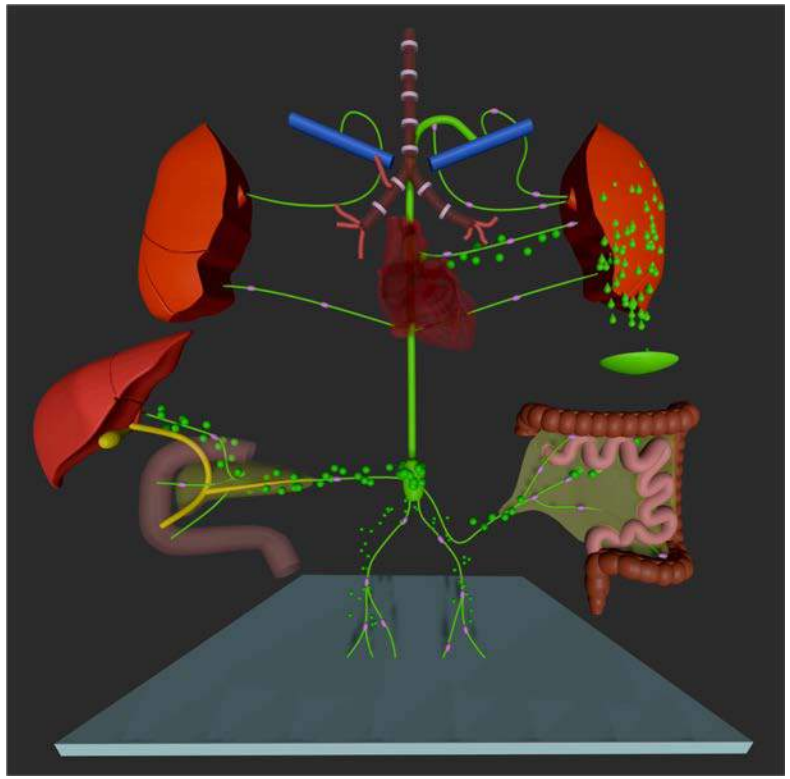
- Medical
  - Aimed at treating underlying secondary causes of lymphatic dysfunction
  - Lymphatic specific therapies

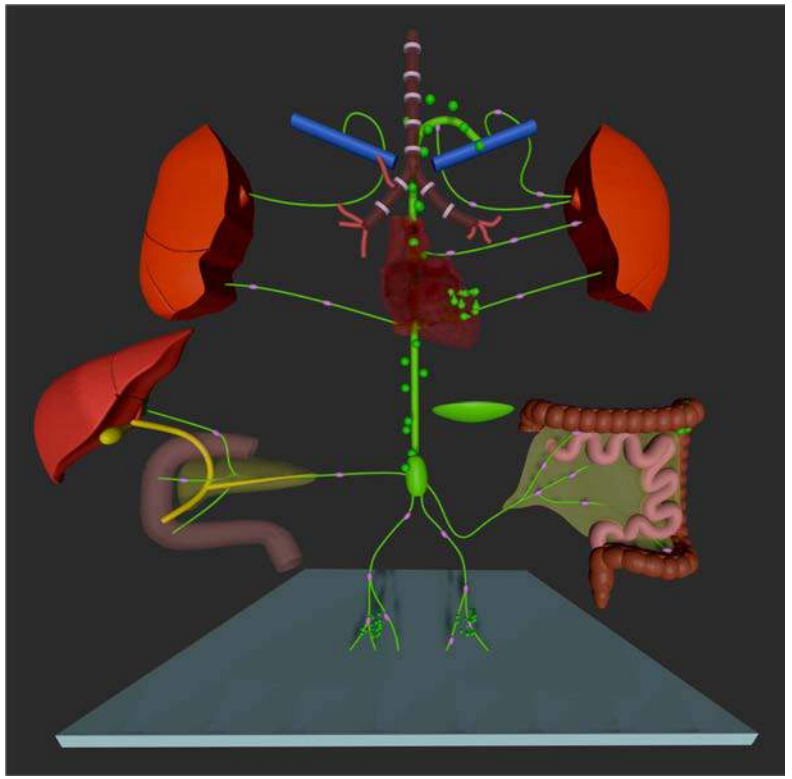
Disorders of Lymph Flow in the Thorax

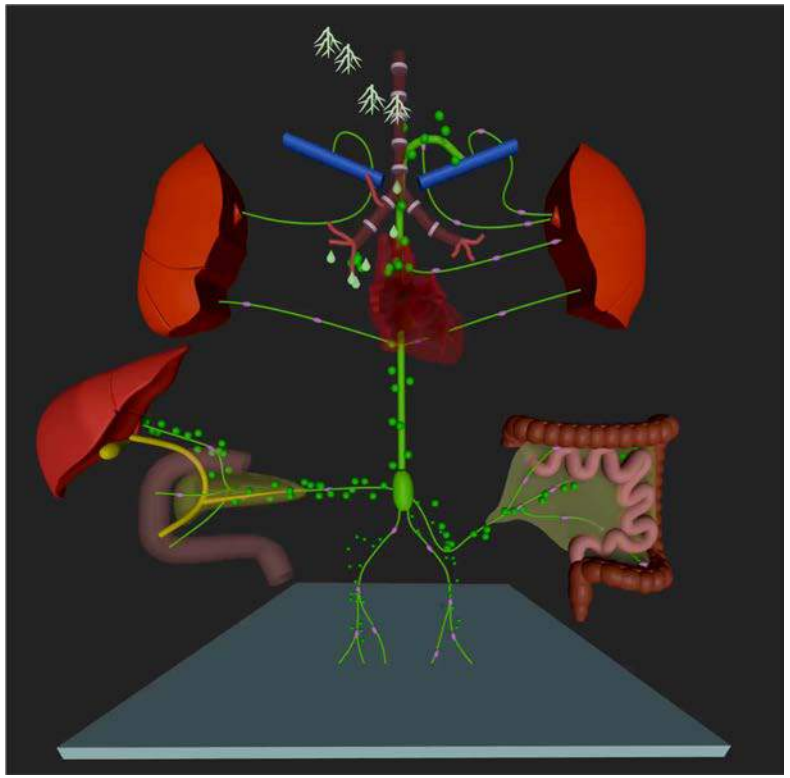
CTx

Chylopericardium

PB







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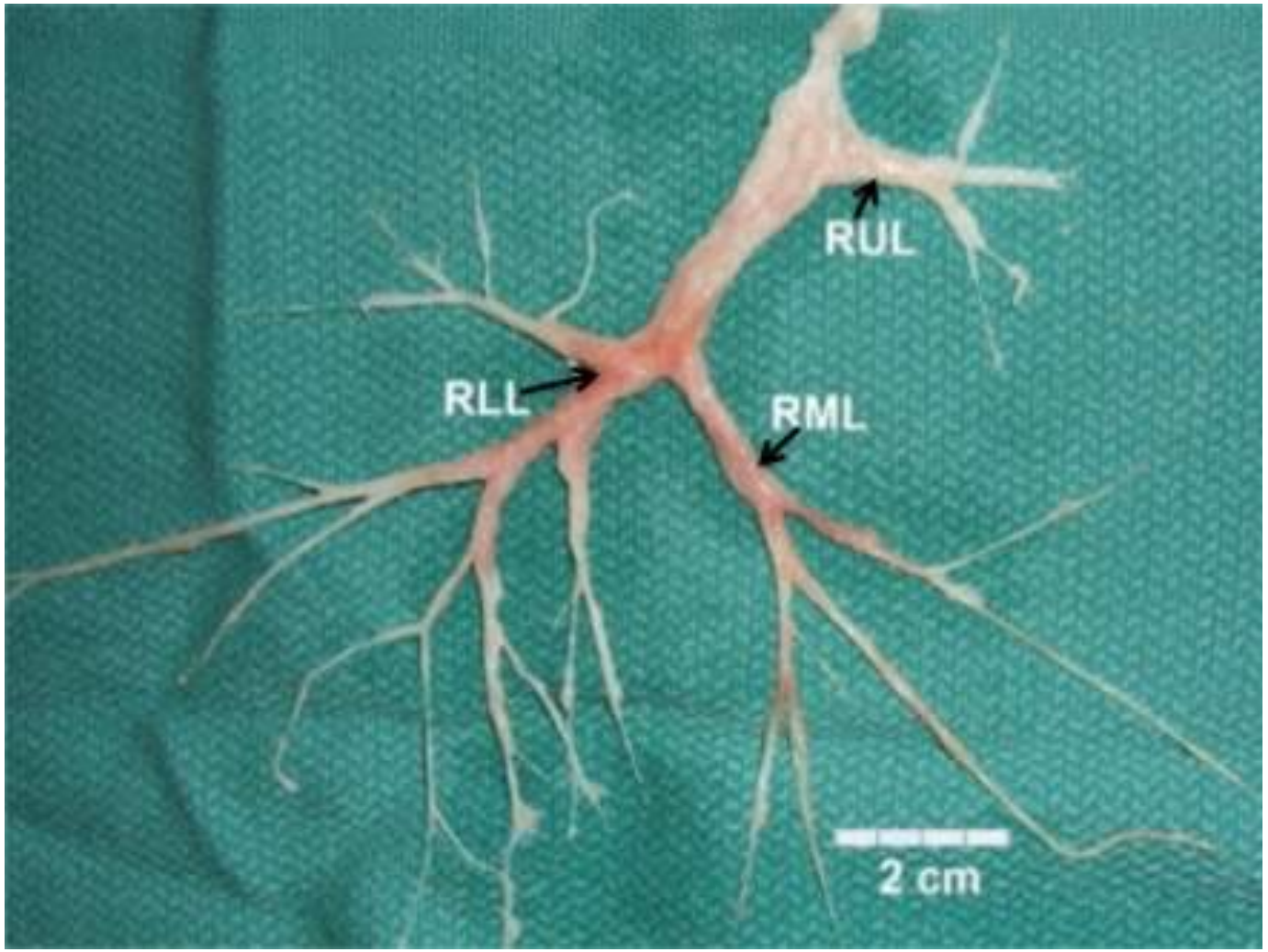
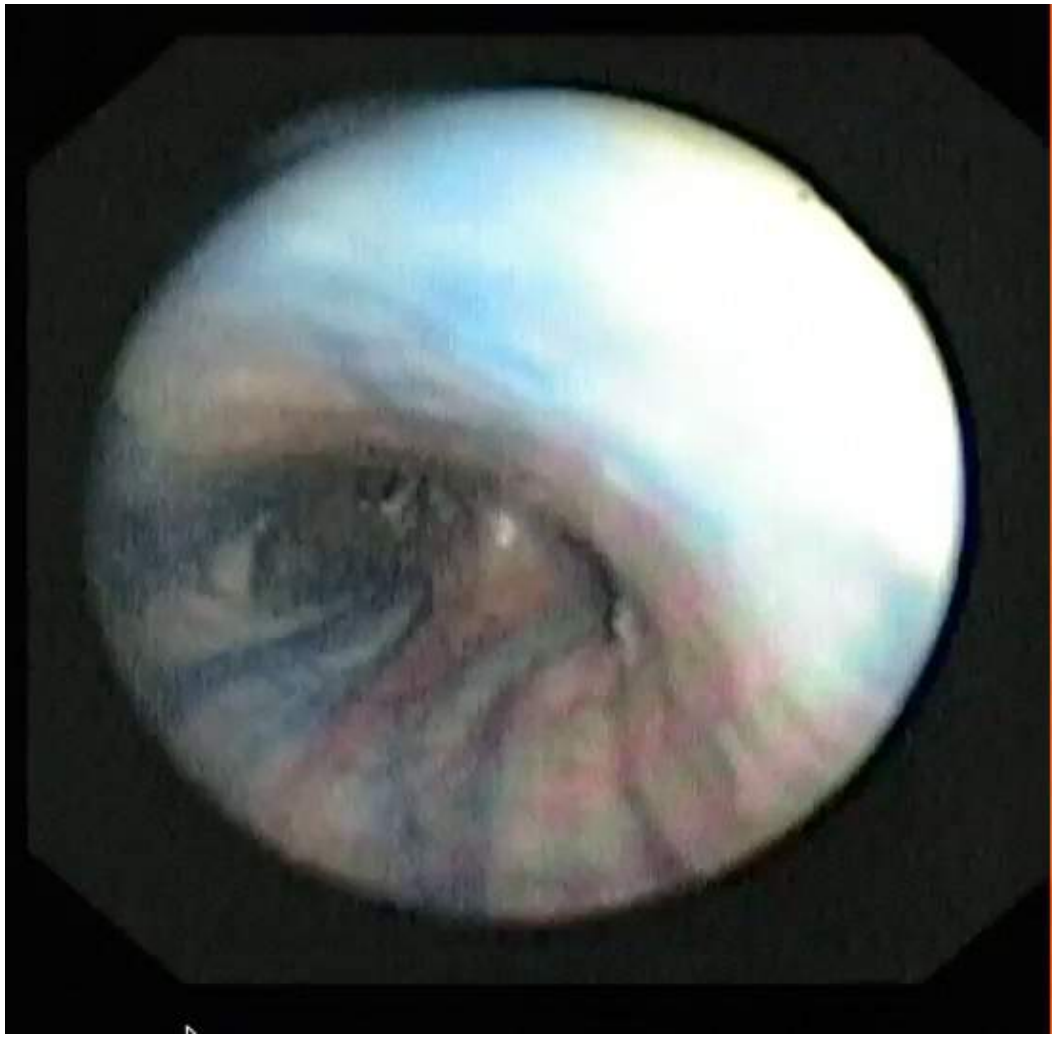
Plastic Bronchitis





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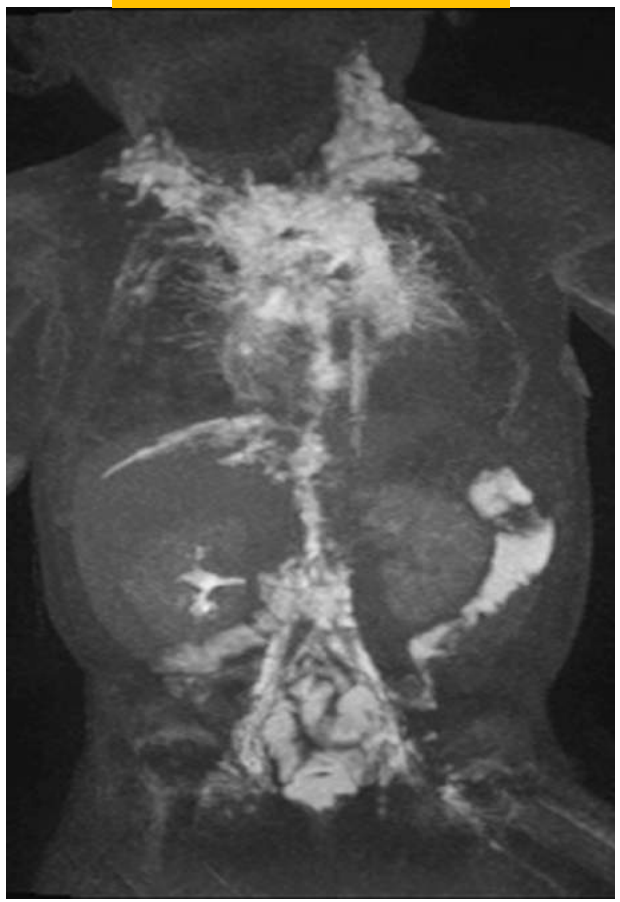
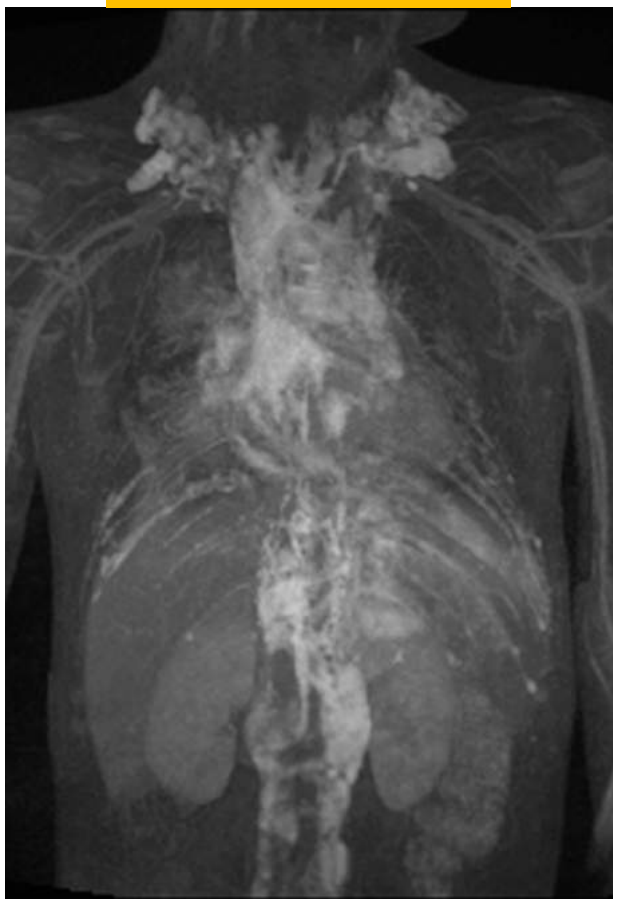
Pulmonary Lymphatic Perfusion In Patients with SV

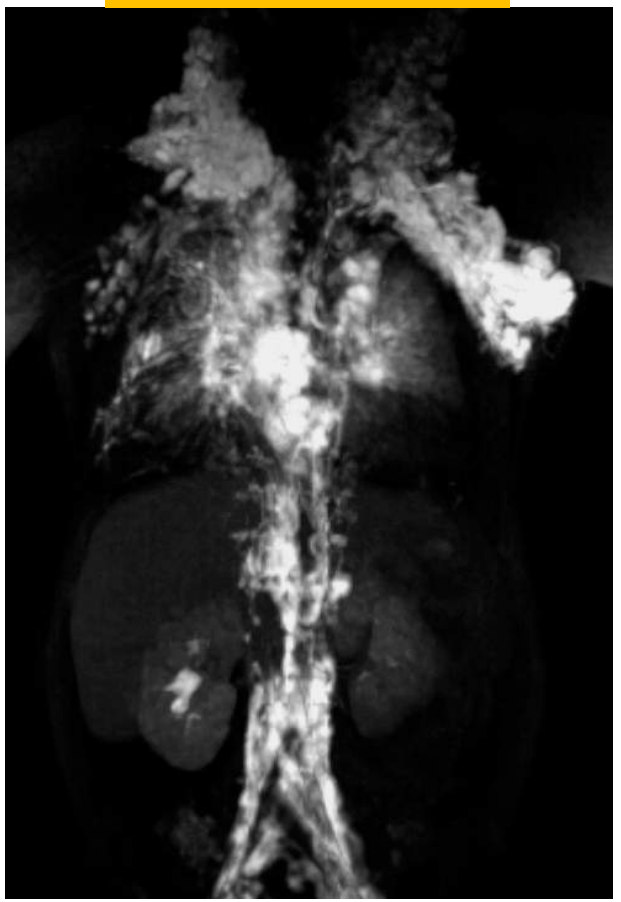
Glenn with chylothorax



Fontan with chylothorax



Fontan with PB



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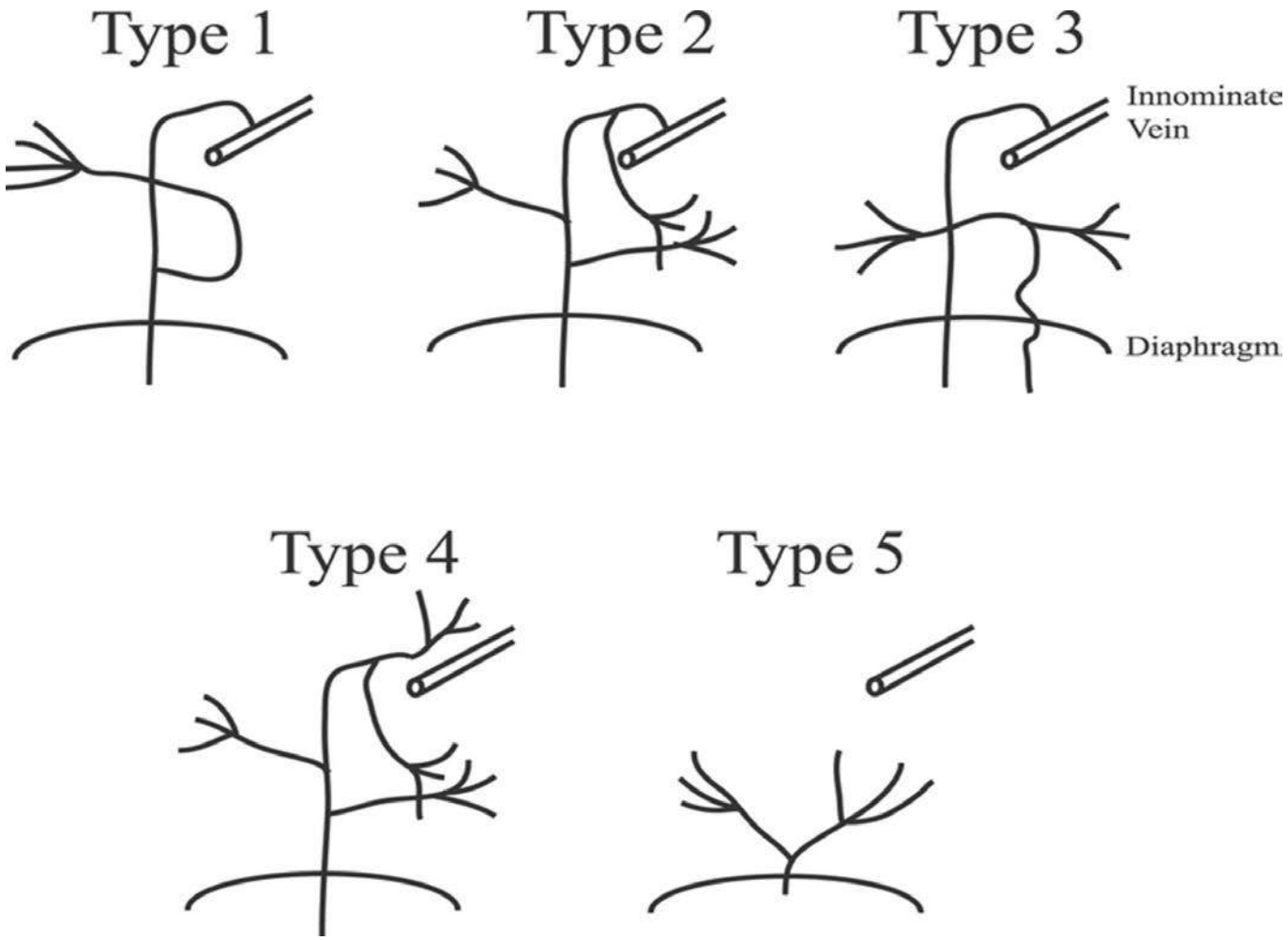
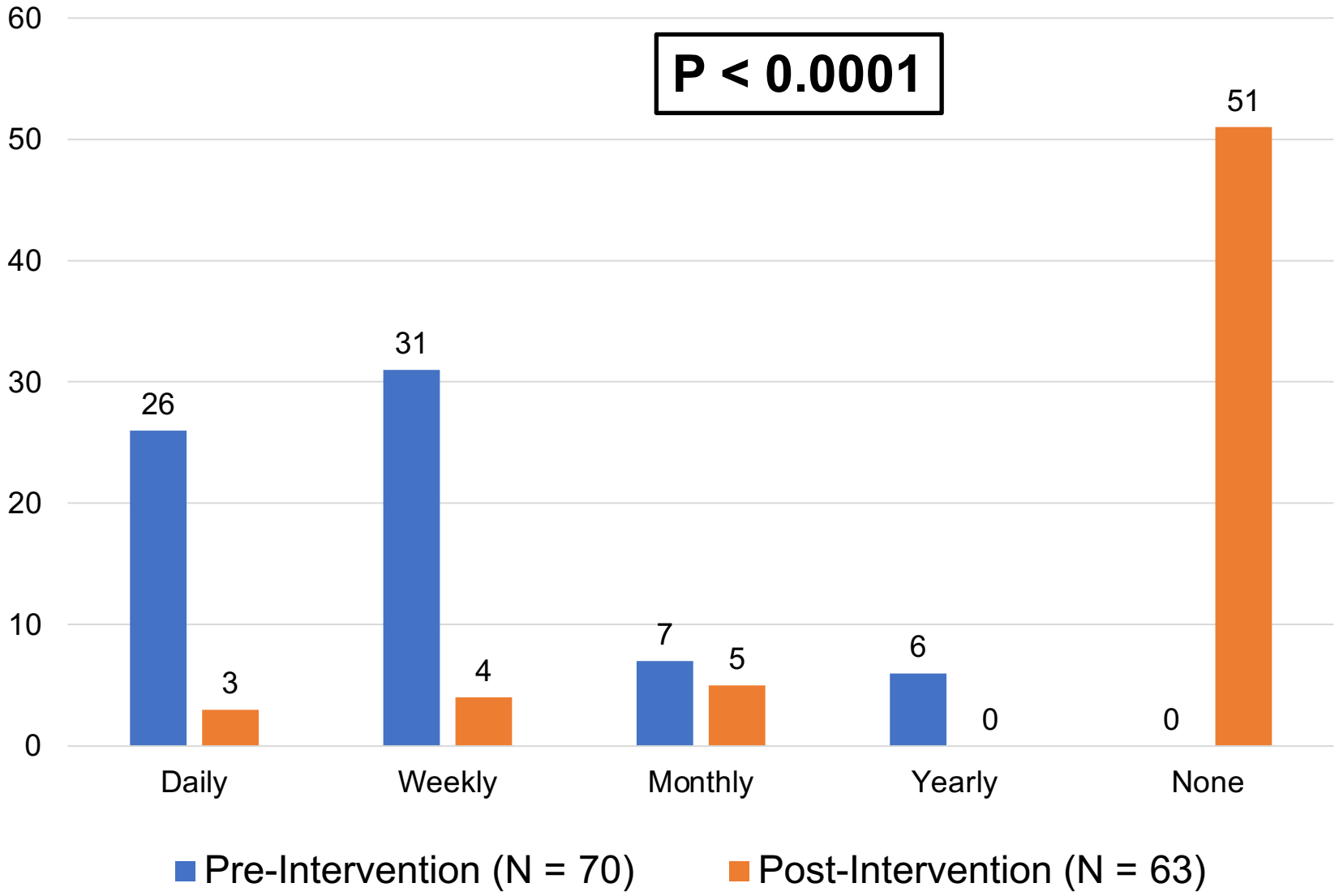
Pulmonary Lymphatic Perfusion In Patients with SV



Dori et al (Circulation, 2016)

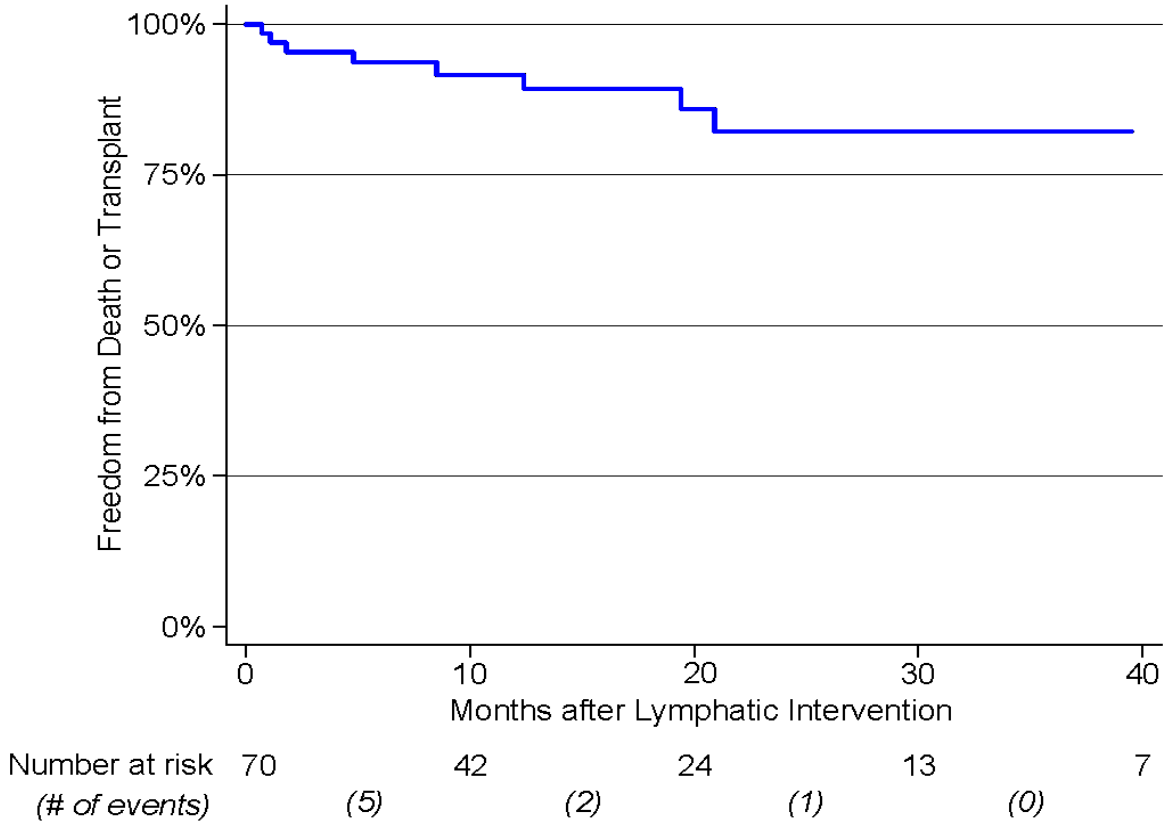
PB treatment: Lymphatic Intervention (SLDE)

Casting Frequency

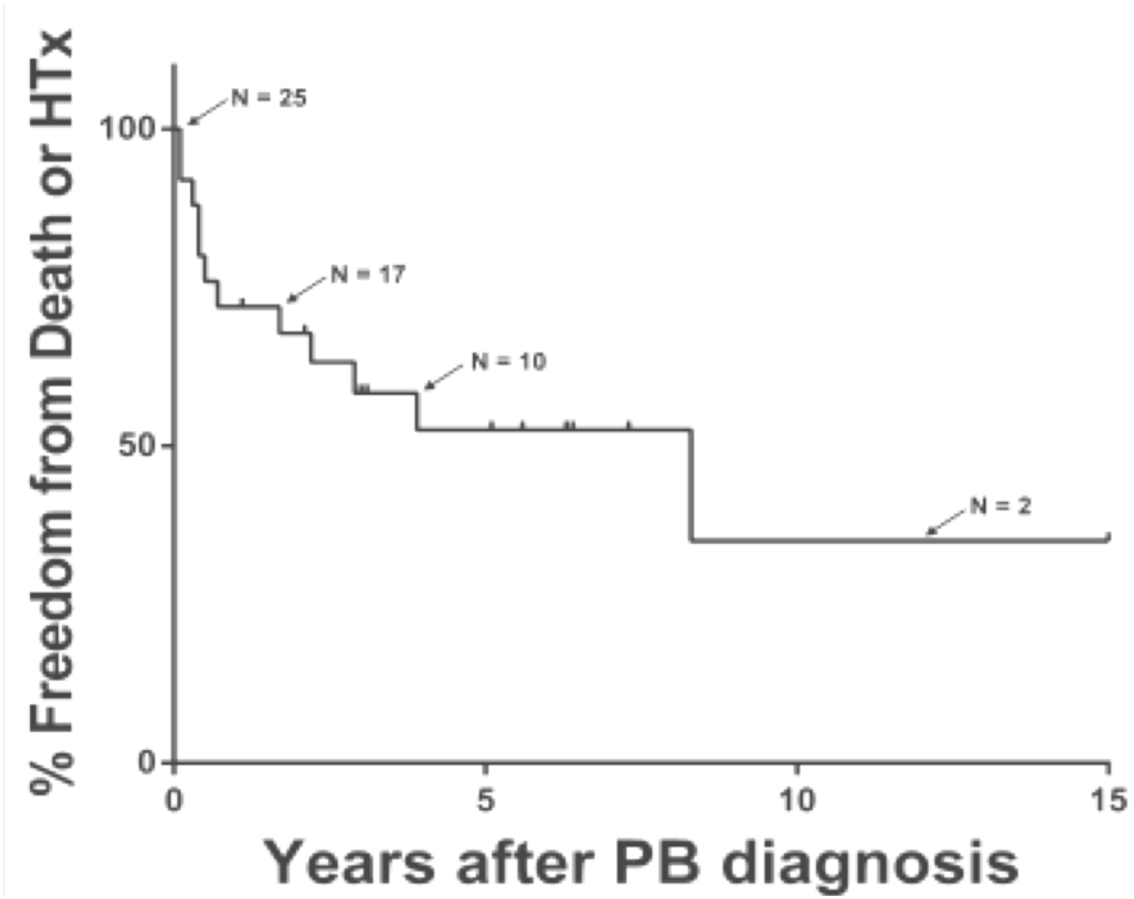


- Median follow up 19 months
- 51/63 (**81%**) with no casting
- Of those, 54% had immediate cessation

Transplant-Free Survival



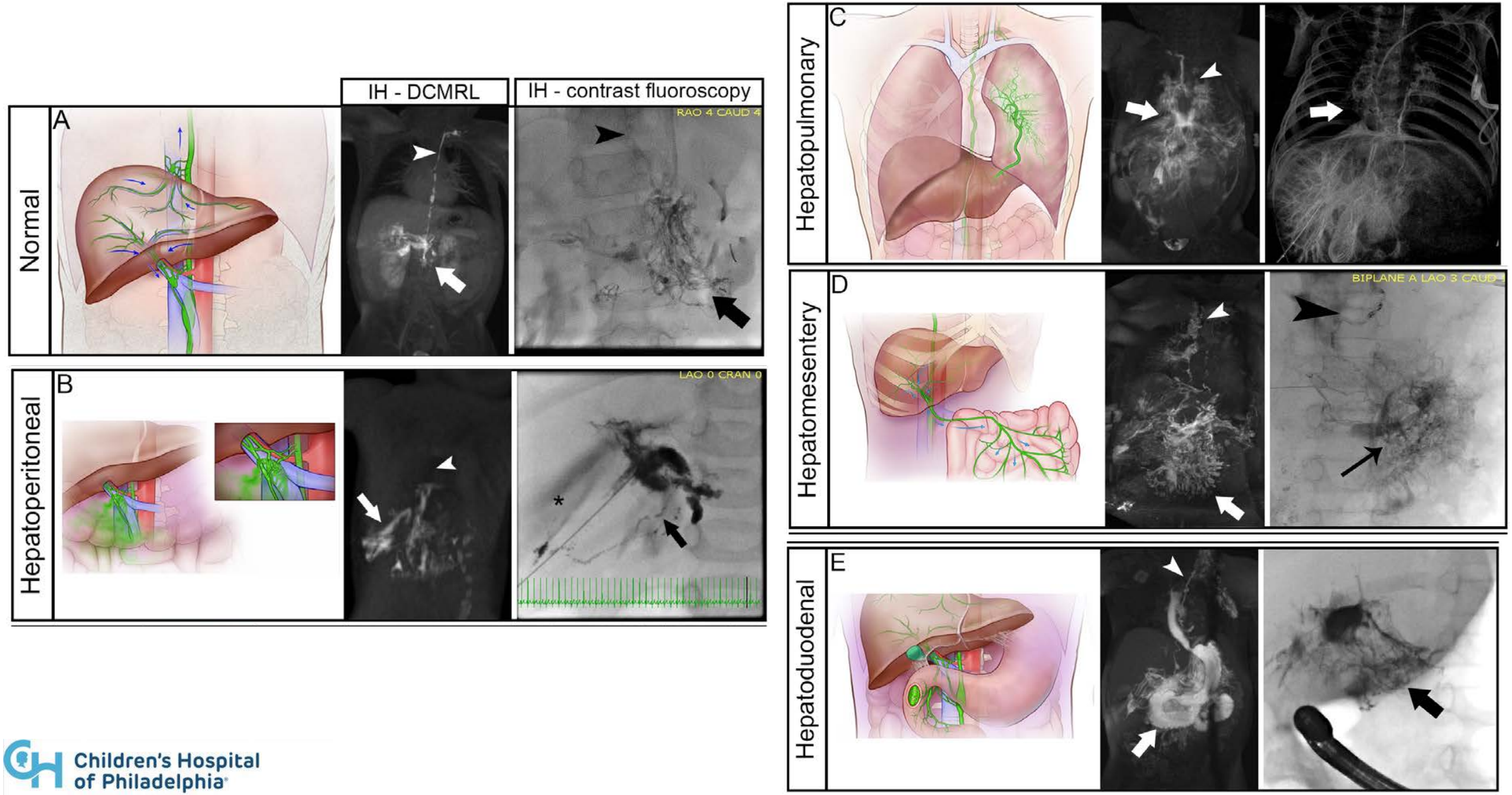
Median follow up 19 months (IQR 8 - 30)
→ 8/70 (11%) with death or transplant



Median follow up 30 months (IQR 8 - 67)
→ 12/25 (48%) with death or transplant

Schumacher KR et al. J Am Heart Assoc. 2014

Intrahepatic Lymphatic Imaging



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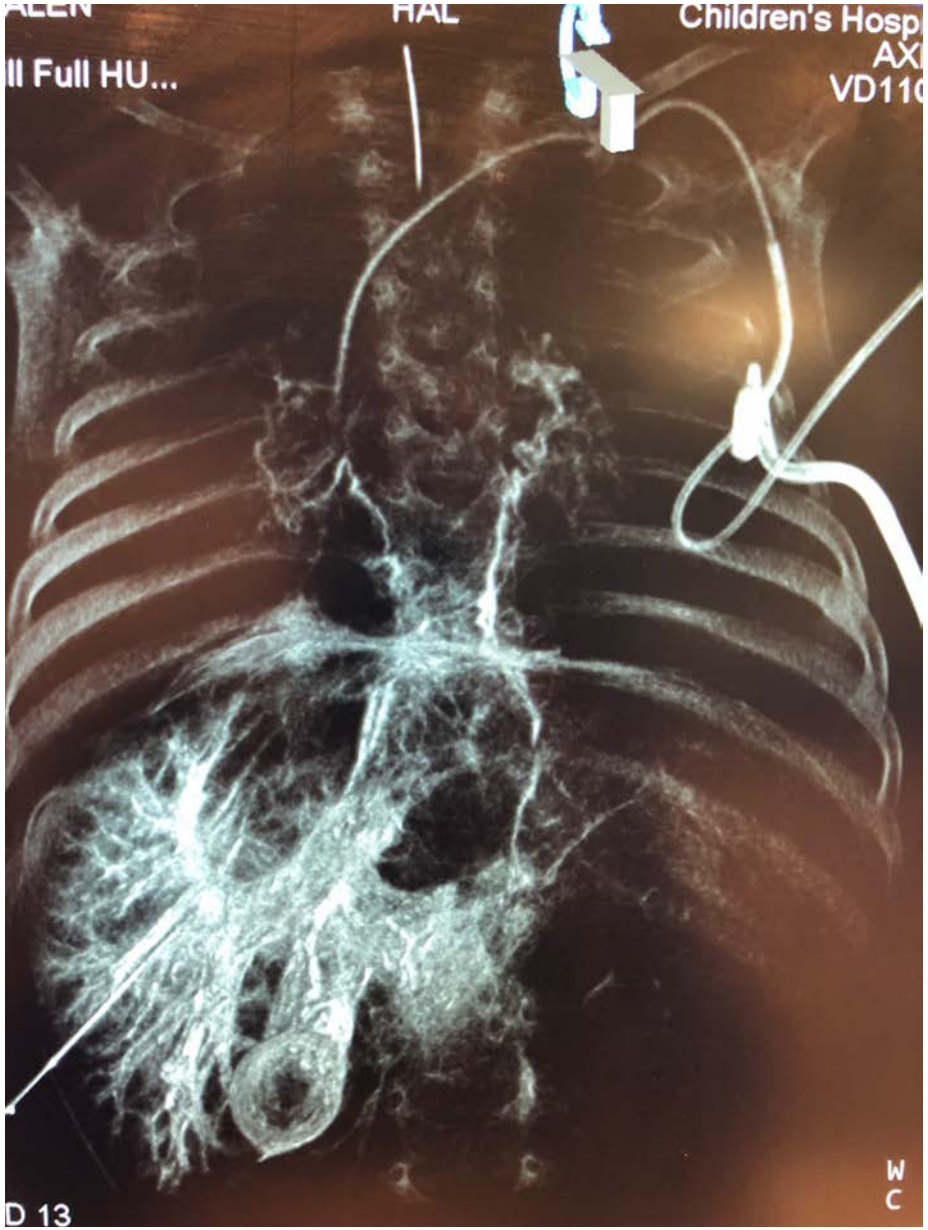
Smith et al. (European Radiology, 2021)

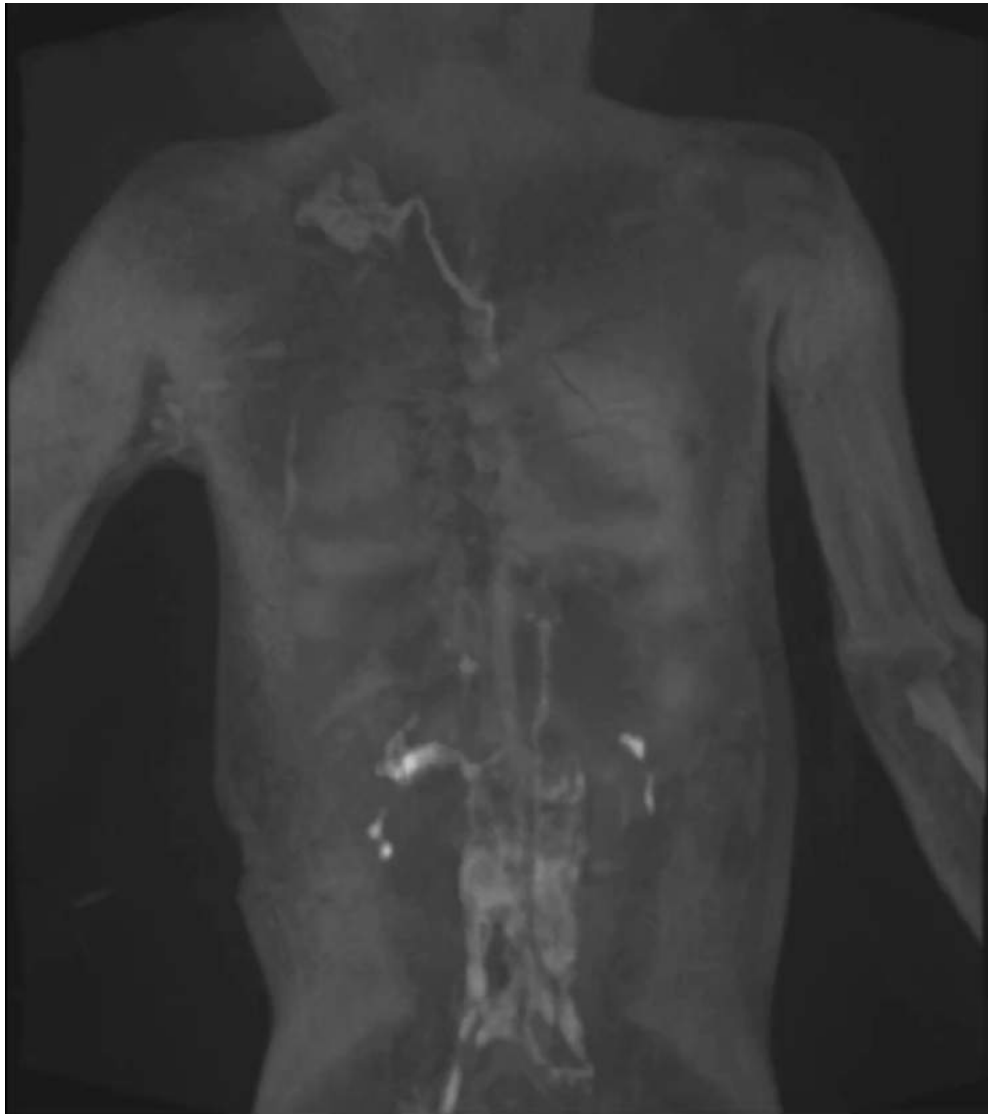
Hepatopulmonary Connections

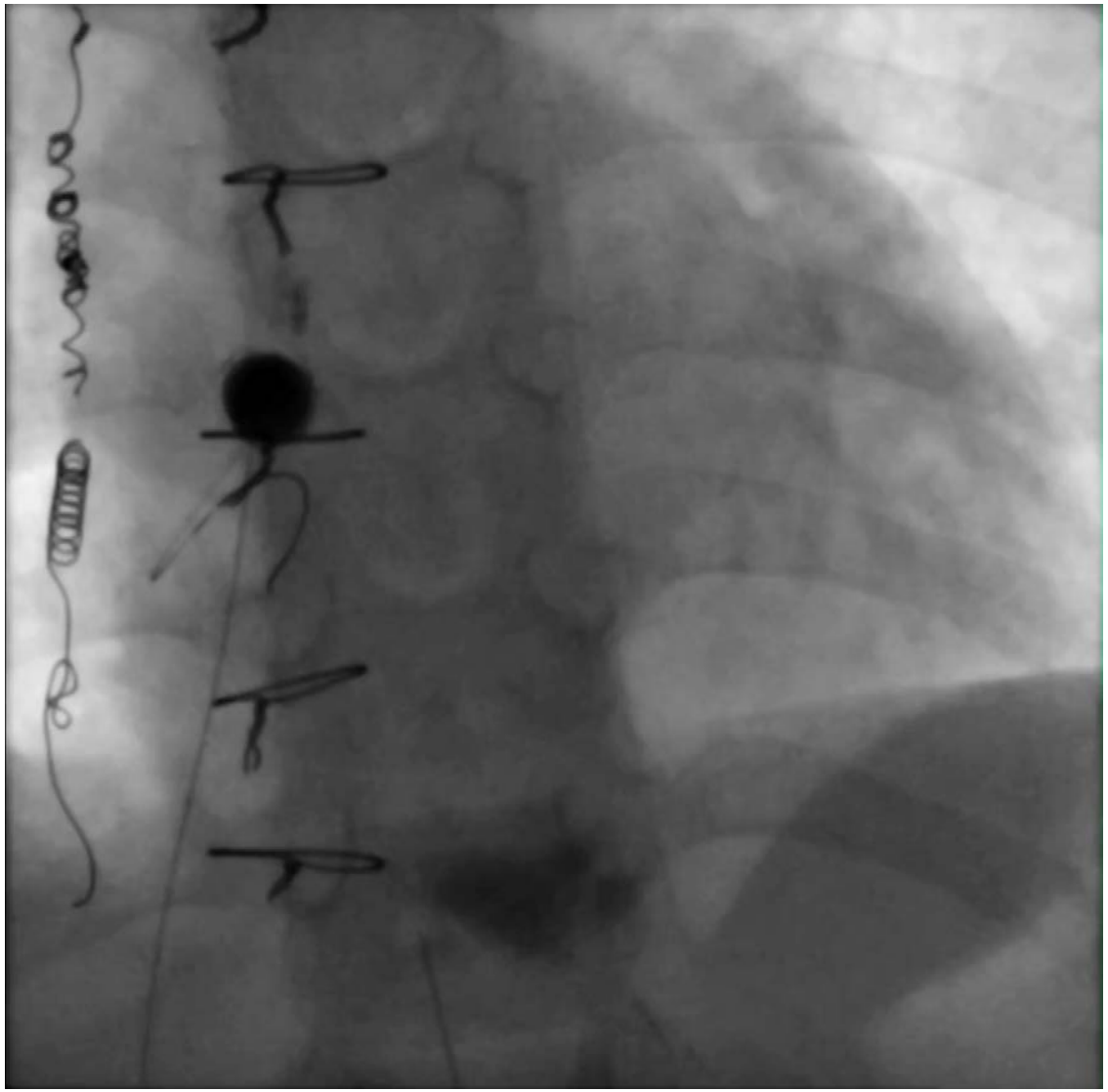
- 157 patients with chylothorax or PB with intrahepatic imaging

- 46 Patients (28%) had hepatopulmonary connections
  - (31 CTx, 19 PB)
  - 13 patients had prior intervention

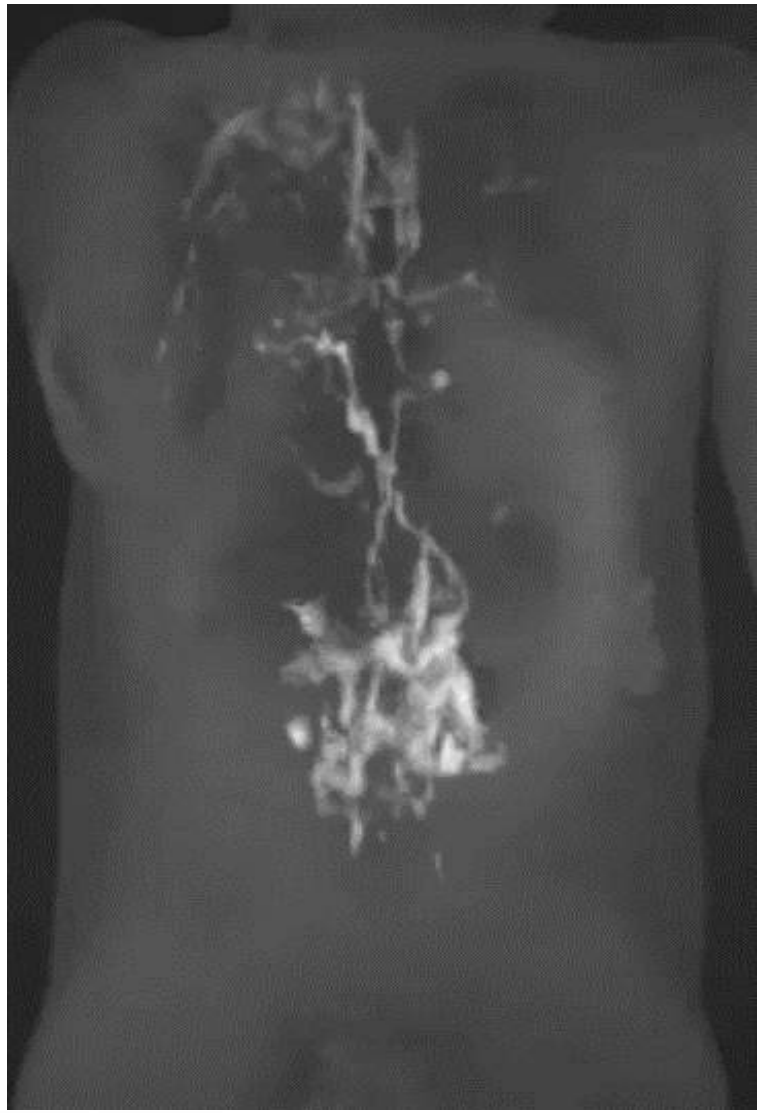


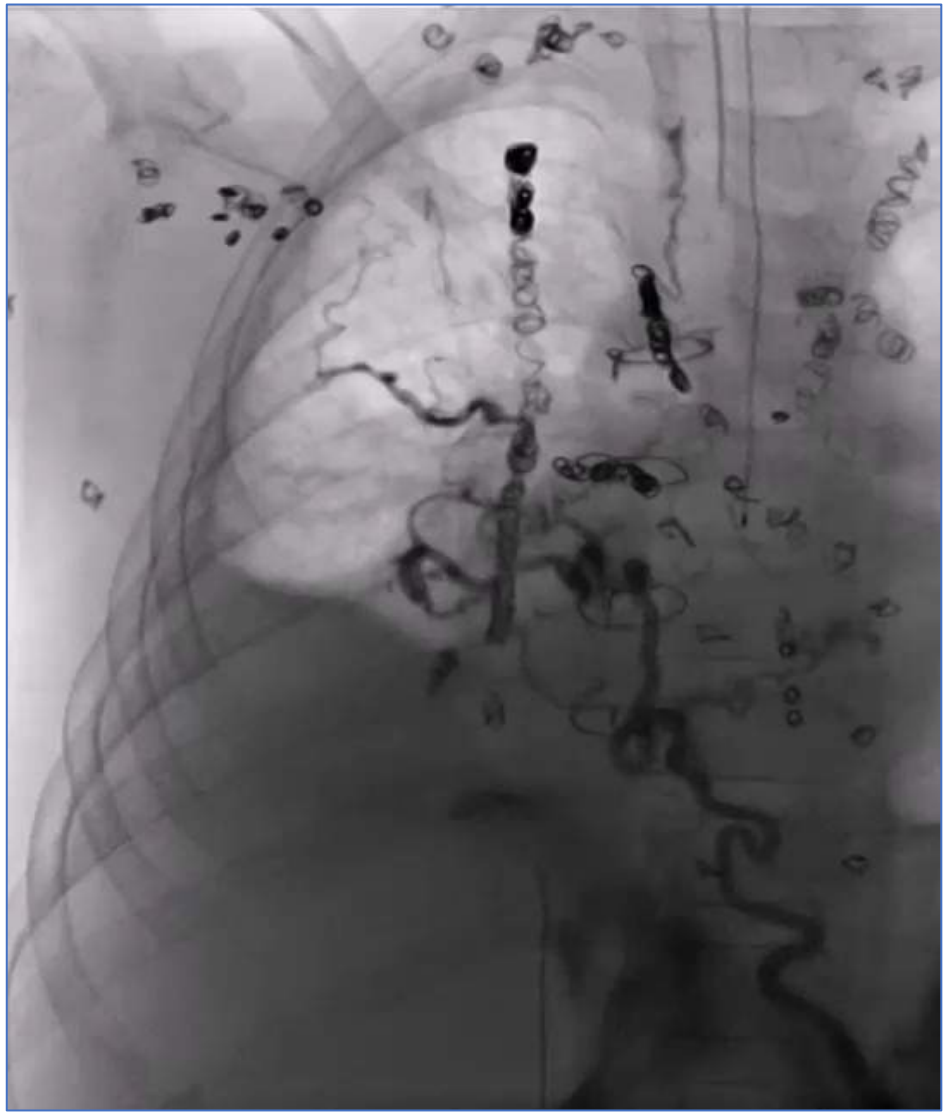
Hepatopulmonary Connections in Patient with PB





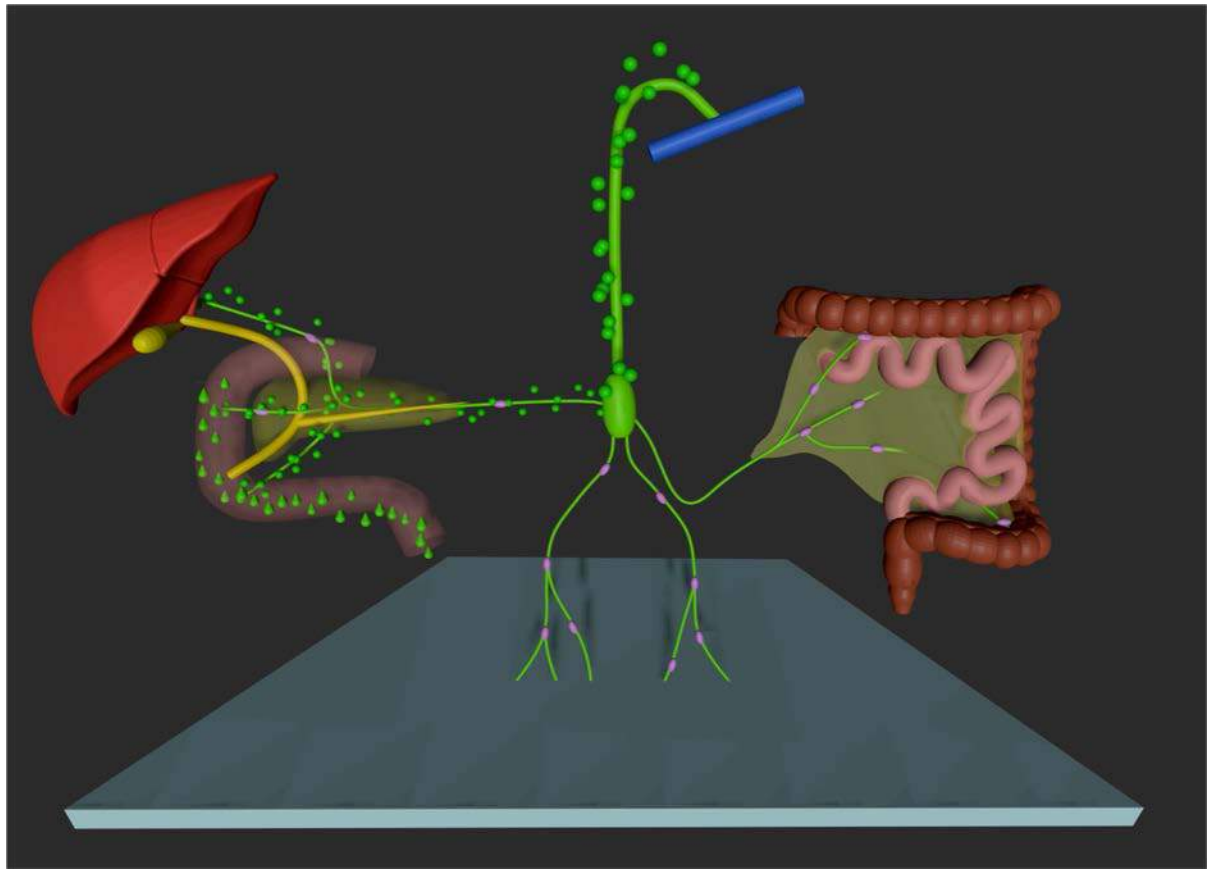
Hepatopulmonary Connections in Patient with PB



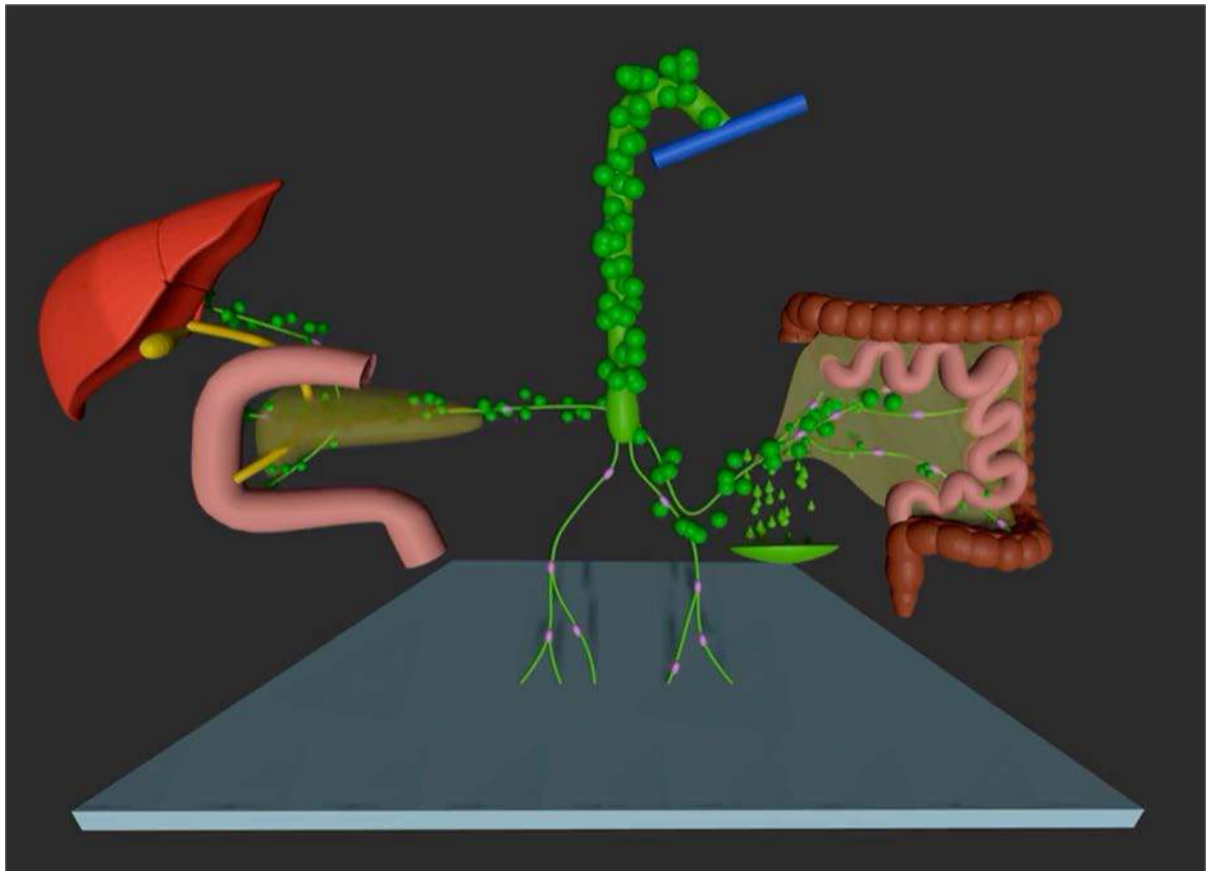


Disorders of Lymph Flow in the Abdomen

PLE



Ascites



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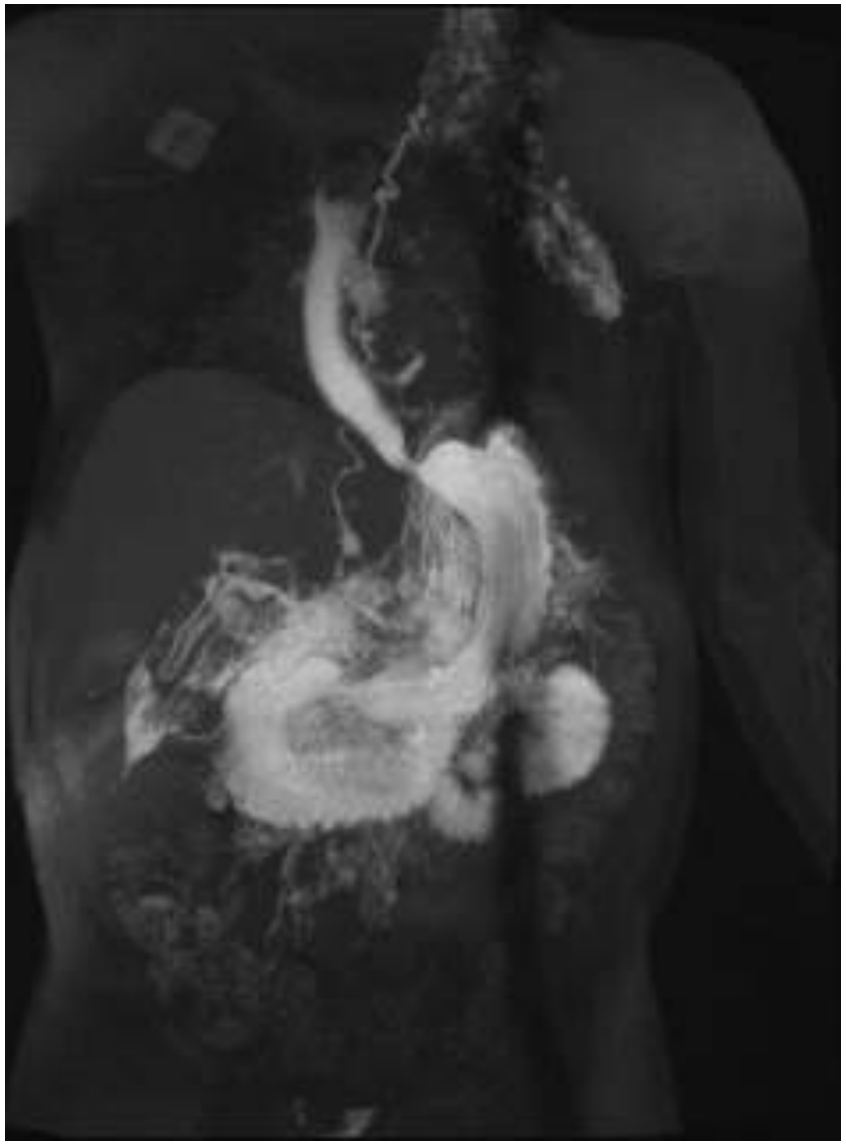
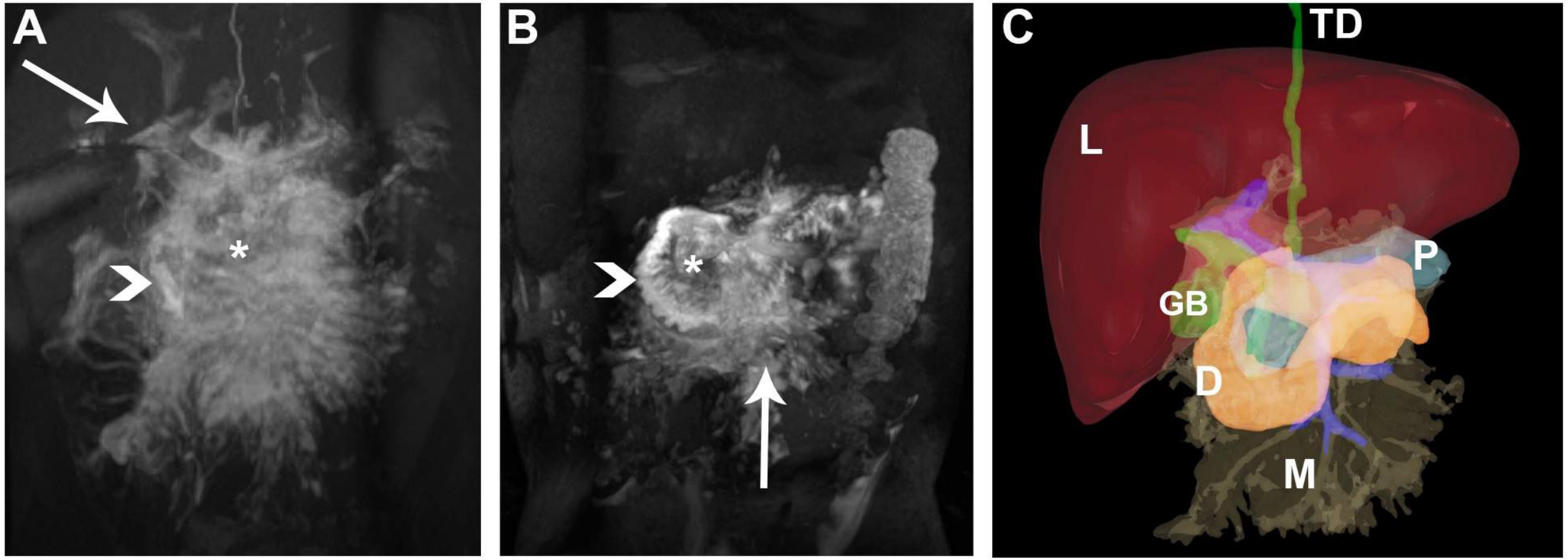
IH-DCMRL in PLE



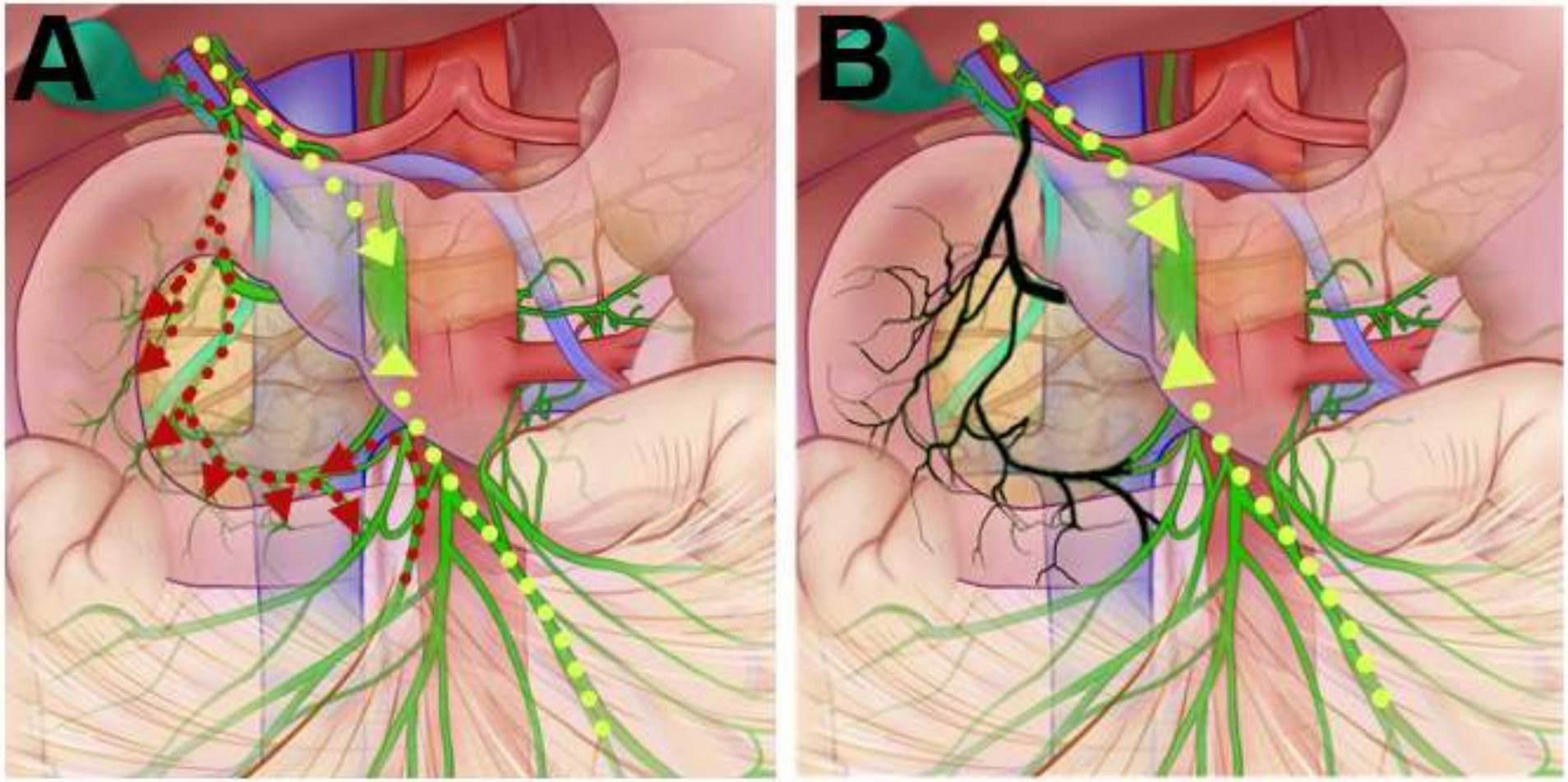
Intrahepatic vs. Intramesenteric DCMRL: Extrahepatic sources of PLE

IH-DCMRL

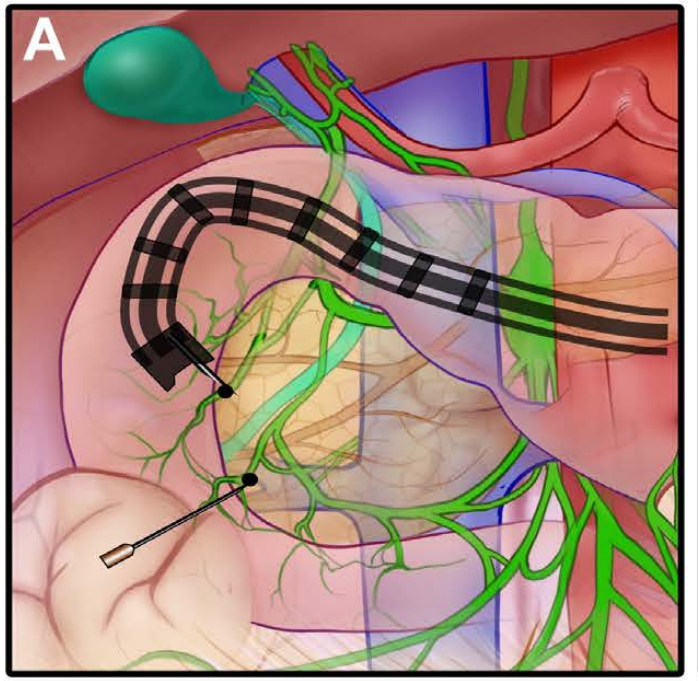
IM-DCMRL

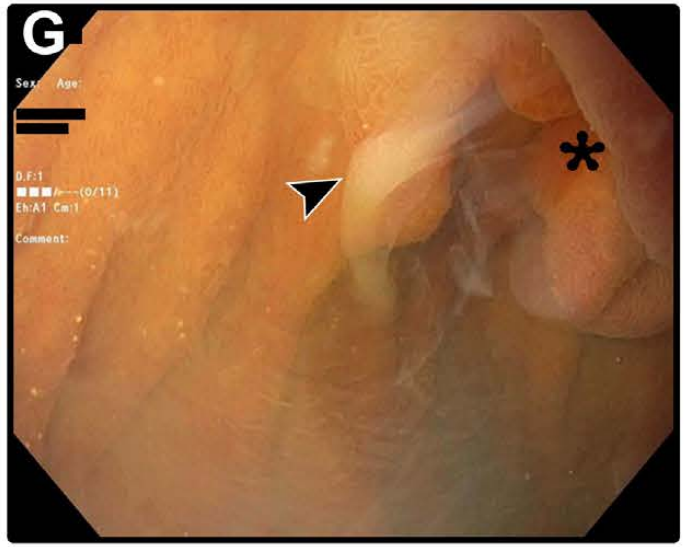


IH and Periduodenal embolization Strategy

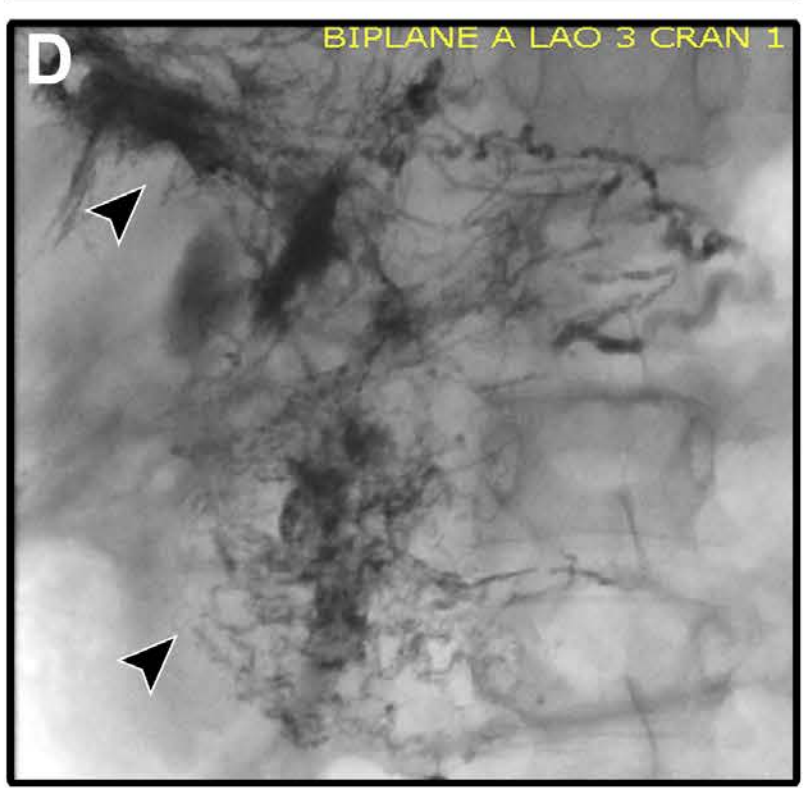


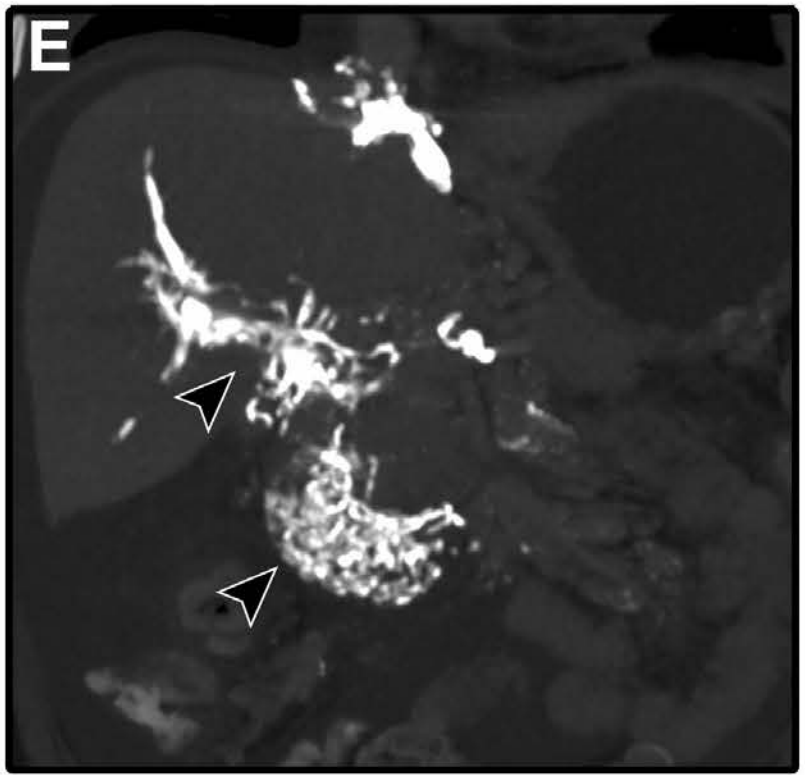
IH and Periduodenal embolization Strategy

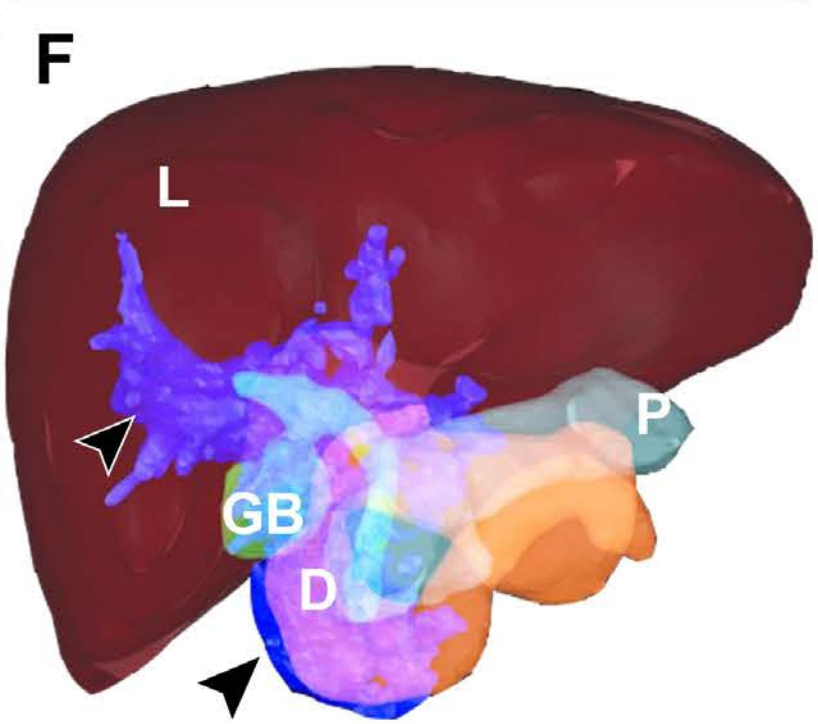




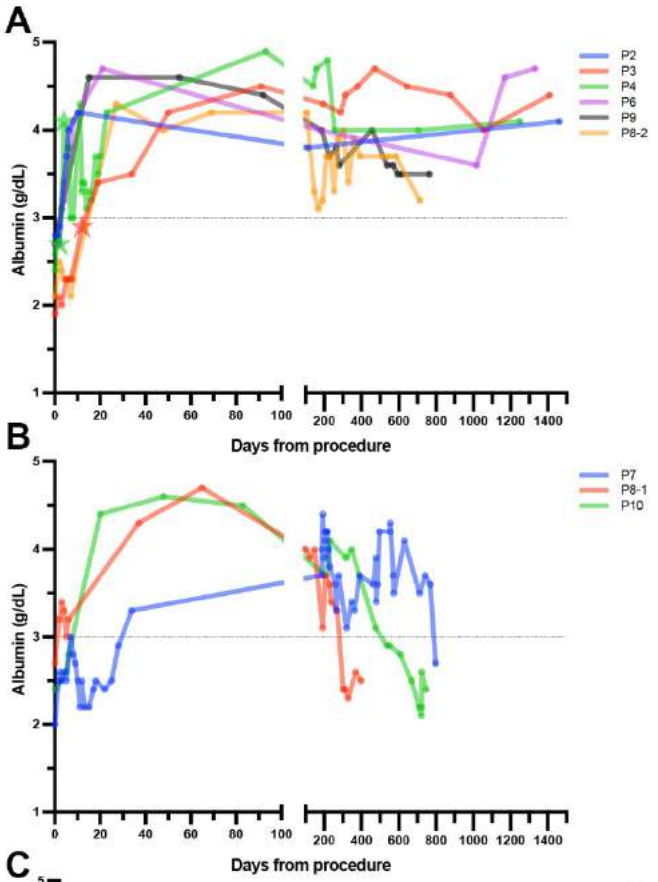
IH and Periduodenal embolization Strategy

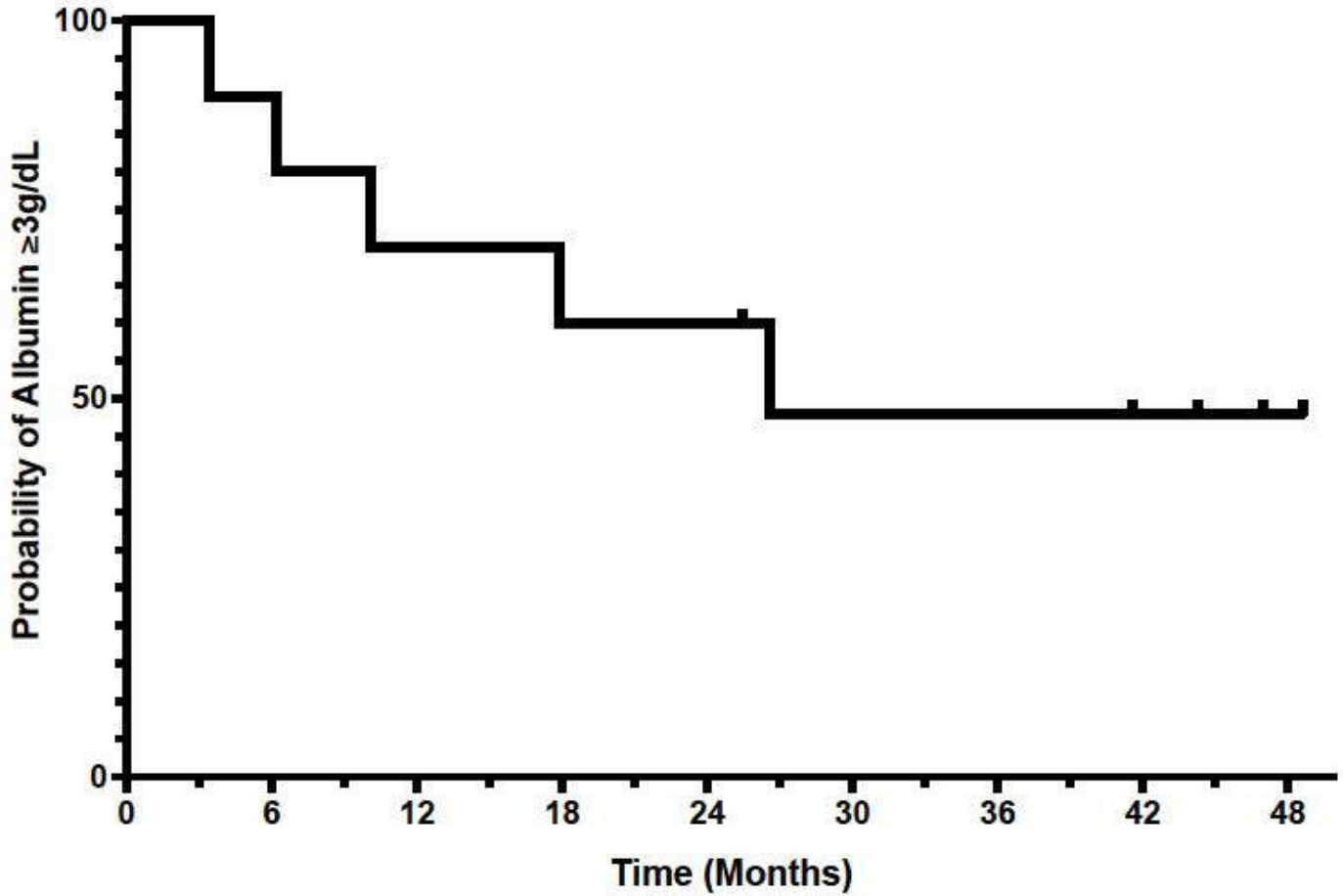






Outcome: Albumin levels





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IH and Periduodenal embolization: Outcomes

- Albumin levels increased in all patients after one procedure
  - Pre procedure: median 2.3 to max post procedure: median 4.3
  - Median time to achieve albumin >3: 15d (IQR 6-20d)
- Median length of stay was 6.5d (range 4-50d)
- Medications reduced or discontinued; No further IV Albumin supplementation in complete responders

- Complications:
  - Transient Pancreatitis (9/12 procedures, 75%)
  - Hyerbilirubinemia (4/12 procedures, 33%)
  - Two patients with non-occlusive thrombus (Portal vein or mesenteric vein)
  - One patient with late hemoperitoneum (PP Day 8, upon anticoagulation restart)

Summary

Keys to diagnosis and treatment of Lymphatic failure

- Multicompartment imaging is necessary
  - Non “traditional” sources of lymphatic perfusion are more common than previously recognized
- New therapeutic strategies for PLE treatment are available
- Focus on selective embolization, but recognize other compartments may worsen
- Be aware of multicompartment failure (more to follow)
- **Do not ligate the thoracic duct** in patients with multicompartment failure
  - Advanced therapies to modify underlying lymphodynamics

Thank You!