# Lymphatic Disorders of the Abdomen



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# **Disorders of Lymph Flow in the Abdomen**













#### Disorders of Lymph Flow in the Abdomen: Protein Losing Enteropathy











### Disorders of Lymph Flow in the Abdomen: Protein Losing Enteropathy

14 year old male with Tricuspid stenosis, pulmonary atresia, intact ventricular septum s/p staged palliation to lateral tunnel Fontan

- Unremarkable post procedure courses until he was 11 years old when he presented with diarrhea, edema, and hypoalbuminemia and diagnosed with PLE

- Medically managed with budesonide but after minimal to no significant improvement it was stopped after ~18 months. Continued with diuresis and symptom management.

- Cardiac catheterization: SVC m11, Bilateral PCWP m5, Fenestration dilated
- Struggled with ongoing edema and diarrhea with worsening electrolyte abnormalities. Ultimately started on IV albumin replacement.

Given medically refractory symptoms, he was referred for lymphatic evaluation and possible intervention.





#### **IN-DCMRL** in **PLE**







## **PLE Treatment: Intrahepatic Embolization**







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# **PLE Treatment: Intrahepatic Embolization**







### Disorders of Lymph Flow in the Abdomen: Protein Losing Enteropathy

14 year old male with Tricuspid stenosis, pulmonary atresia, intact ventricular septum s/p staged palliation to lateral tunnel Fontan

- Post procedure developed SIRS response, Dopamine for <24h
- Anticoagulation initiated on post procedure day #1
  - Developed melena and downtrending hgb c/f GI bleed
  - Transfused PRBCs, anticoagulation held and monitored without further bleeding

Discharged on post procedure day #6

- Albumin levels: Pre 1.9 and max post procedure 3.5g/dl ~3 weeks post procedure, but abruptly decreased to 1.8 – 2.5 g/dL within the following month





#### **PLE Treatment: Intrahepatic Embolization Outcomes**

Very limited follow up data published

- Initial description by Itkin et al (2017)
  - 8 Patients treated with IH only
    - 3 patients with sustained improvement (longest f/u 140d)
    - 3 patients with early recurrence (<4 months)
    - 2 patients with no response
- Gewillig et al. (2019)
  - 7 patients included
    - 5 patients had second procedure
    - 6/7 patients with improvement (Longest f/u 17 mo, range 4-17mo))



Smith et al (Manuscript in preparation)





### Disorders of Lymph Flow in the Abdomen: Protein Losing Enteropathy

5 year old with Tricuspid stenosis/VSD, L-TGA s/p staged palliation to 20mm Extracardiac fenestrated fontan

- Diagnosed 2 years post Fontan with PLE when presenting with hypoalbuminemia and edema after an RSV illness
- Initiated on diuretics, budesonide, and received IV albumin infusions
- 9 months after diagnosis with continued hypoalbuminemia, edema, diarrhea, and development of ascites. Admitted for milrinone infusion and aggressive diuresis and albumin supplementation
- Cardiac catheterization with SVC pressures of 16mmHg and EDP of 8mmHg. Systemic saturation of 87% with minimal to no fenestration present.

Given Refractory medical therapy was referred for lymphatic evaluation and intervention.





#### **IH-DCMRL** in **PLE**







#### **PLE Treatment: Intramesenteric DCMRL**











#### Intrahepatic vs. Intramesenteric DCMRL: Extrahepatic sources of PLE







## IH and Periduodenal embolization Strategy







#### **IH and Periduodenal embolization Strategy**





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### **PLE – Leak with Blue Dye Injection**



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#### **PLE – Embolization**







# IH and Periduodenal embolization Strategy







## Disorders of Lymph Flow in the Abdomen: Protein Losing Enteropathy

5 year old with Tricuspid stenosis/VSD, L-TGA s/p staged palliation to 20mm Extracardiac fenestrated fontan

Post procedure course:

Developed pancreatitis that resolved by post procedure day 3 (resolution of lipase)
Day 10 had recurrent pancreatitis with hyperbilirubinemia, Gallstone identified (resolved with continued medical management)

- Albumin levels increased: 2.1 g/dL (pre) -> day 5 (3.8) and remained >3g/dL now for ~5 years





# **Outcome: Albumin levels**

- Medications reduced or discontinued; No further IV Albumin supplementation in complete responders
- Albumin levels increased in all patients after one procedure
  - Albumin median increased from 2.3 -> 4.3 g/dL
  - Median time to achieve albumin >3: 15d (IQR 6-20d)





Gartenberg E, et al. Intrahepatic and periduodenal embolization for PLE patients with congenital heart disease JACC 2023



# IH and Periduodenal embolization: Outcomes

- Albumin levels increased in all 10 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)





## **Outcome: Albumin levels**



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Smith et. al (manuscript in preparation)

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### **PLE Outcome: TD Obstruction**





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# Summary

- PLE pathogenesis is complex and there are multiple contributing sources/organs that ultimately converge on the duodenum
- Multidisciplinary approach is necessary
  - Gastroenterology and Interventional Radiology
- IH and Periduodenal embolization can lead to improved PLE symptoms after one procedure
  - All 10 patients responded, with 50% having sustained response
  - Reduction or stop medical therapies (i.e. budesonide, albumin)
- Further investigation of larger cohorts are needed to determine if early outcome is maintained and to identify those patients who are more likely to respond





# **8 y/o with Severe Ascites**

Tricuspid atresia with dTGA, VSD s/p staged palliation (Norwood BTS, hemifontan, 20mm Extracardiac nonfenestrated Fontan with MV repair and pacemaker placement)

Post Fontan course complicated by:

- Sternal abscess (s/p I&D), diaphragm paralysis (plication x2), chylothorax (s/p VATS x2),
- (1yr post) Endocarditis with infection of Fontan conduit
  - Fontan revision with 18mm Contegra (nonfenestrated) removal of PM
    - Multiple CVAs and neurologic injury with seizures
    - SVT on atenolol





# **8 y/o with Severe Ascites**

7 years of age developed recurrent Ascites with multiple paracentesis (1.5 - 4L)

- Cloudy yellow fluid: Lymphocytes 20% 55%, TG 118 413
- Cardiac Cath: Cavopulmonary 16-17mmHg, wedge 12, arterial saturation 85%
  - Junctional bradycardia with sinus node dysfunction







Repeat cardiac catheterization:

- Cavopulmonary 17mmHg, PCWP 10mmHg
  - Notable junctional rhythm throughout case
  - Retrograde TD access during case revealed PLPS, no obvious leak into peritoneum
  - Repeat Paracentesis
    - Lymphocyte 19%, Triglycerides 133
    - TD lymphocyte 99%
- Recovery in CICU with frequent junctional bradycardia
  - Discussion on repeat pacemaker placement
  - Timing of MRL? (Also still with abandoned epicardial leads)





#### **8 y/o with Severe Ascites: CHOP Evaluation** MRL approved by MR safety council with abandoned epicardial leads























MRL showed:

- leak from intrahepatic injection,
- PLE signature with ?intralumenal leak
- Pulmonary lymphatic perfusion

Timing of lymphatic intervention and pacemaker?

- Underwent epicardial pacer placement (AAI rate 70bpm)





## 8 y/o with Severe Ascites: Lymphatic Intervention







# IH US Contrast Lymphangiogram







# 8 y/o with Severe Ascites: Lymphatic Intervention







Post-intervention:

- Unremarkable immediate post procedure course, transferred to CCU on PPD #2
- Fluid volume decreased from ~1.5L/day to <200cc/day
- Drain removed on Post procedure day #20
- Continued low fat diet and discharged home on daily Lasix
- No further ascites since procedure (~5 years ago)



