Case 1:

22 year old male with HLHS (MS/AA) s/p staged palliation to a lateral tunnel fenestrated fontan with recurrent PLE

- Prenatally diagnosed with HLHS and underwent staged palliation with course complicated by
  - Recurrent coarctation s/p angioplasty
  - Progressive Tricuspid regurgitation after Hemifontan to severe at time of fontan s/p valve repair
  - Post fontan with prolonged pleural effusions and significant sinus node dysfunction s/p atrial epicardial placement

- At 4 years old had fenestration closure secondary to cyanosis

- Over next decade had intermittent cyanosis with exercise
  - Several catheterizations with:
    - LPA and lateral tunnel stent angioplasty
    - Multiple venous collateral embolizations
    - Each evaluation noted increasing Fontan and RV end diastolic pressures

- 17 years old developed PLE with peripheral edema, diarrhea, and hypoalbuminemia

Case 1:

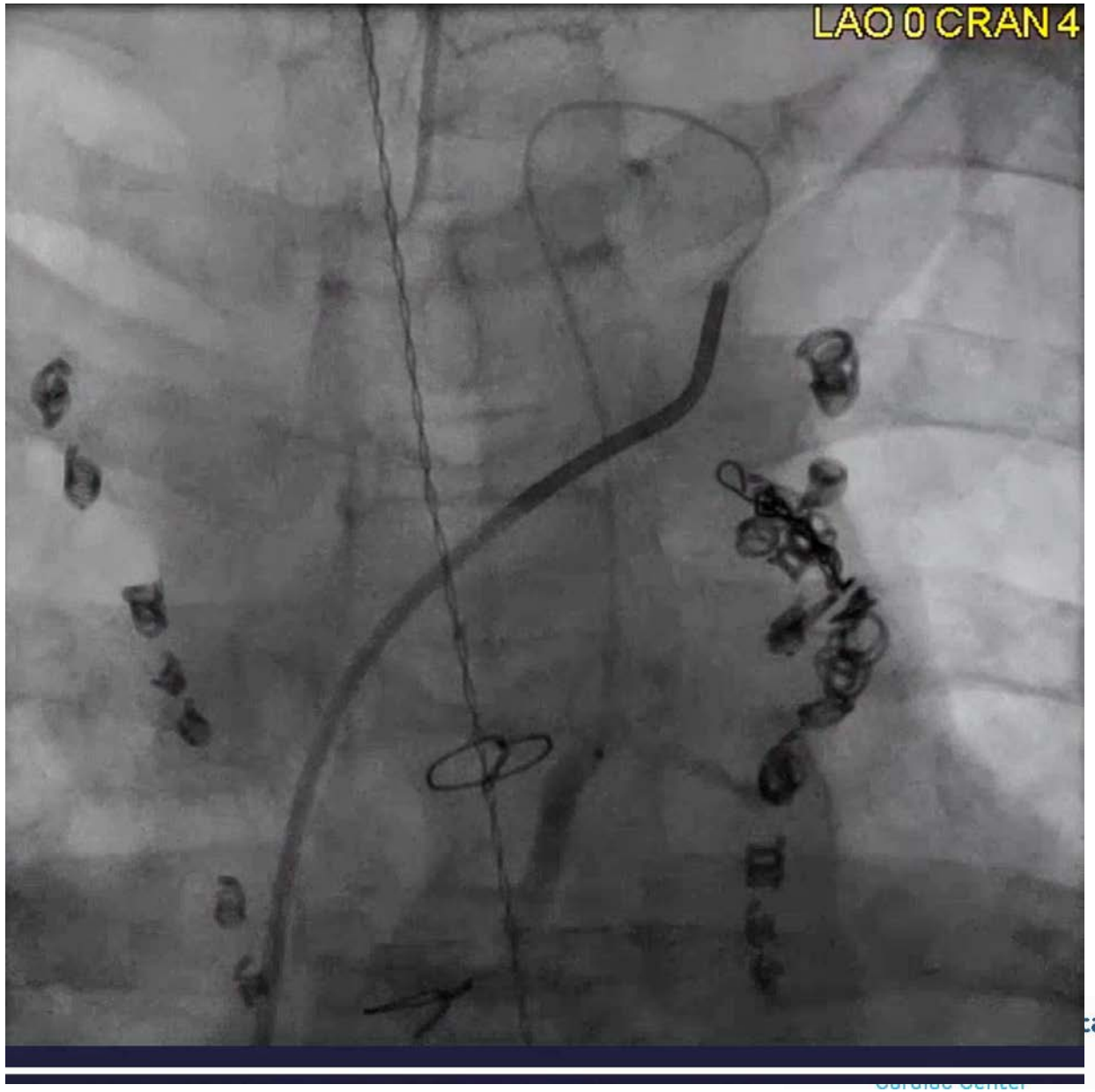
- 17 years old developed PLE with peripheral edema, diarrhea, and hypoalbuminemia
  - Catheterization noted Fontan 18mmHg, EDP 14, angioplasty of LPA stent
  - Trivial TR and normal systolic ventricular function

- Initiated HF regimen with carvedilol and Started on Budesonide
  - Improvement in edema and normalized albumin level for ~2 years

- At 19 years of age developed significant cushingnoid features, weight gain, and stress fractures and PLE was no longer controlled on high dose steroid therapy

Case 1:

- At 21 years of age referred to CHOP lymphatics for evaluation and treatment:
  - No MRI at initial presentation given pacemaker, no ascites or pleural effusions on US/CXR imaging
  - Hemodynamic catheterization noted
    - Fontan of 15mmHg, Wedge pressure of 8mmHg
    - Angiography with no Fontan obstructions
    - TD confirmed patent with lumason and accessed retrograde with dilated and tortuous course and no PLPS
  - Had multiple hepatic and periduodenal lymphatics embolized



Case 1:

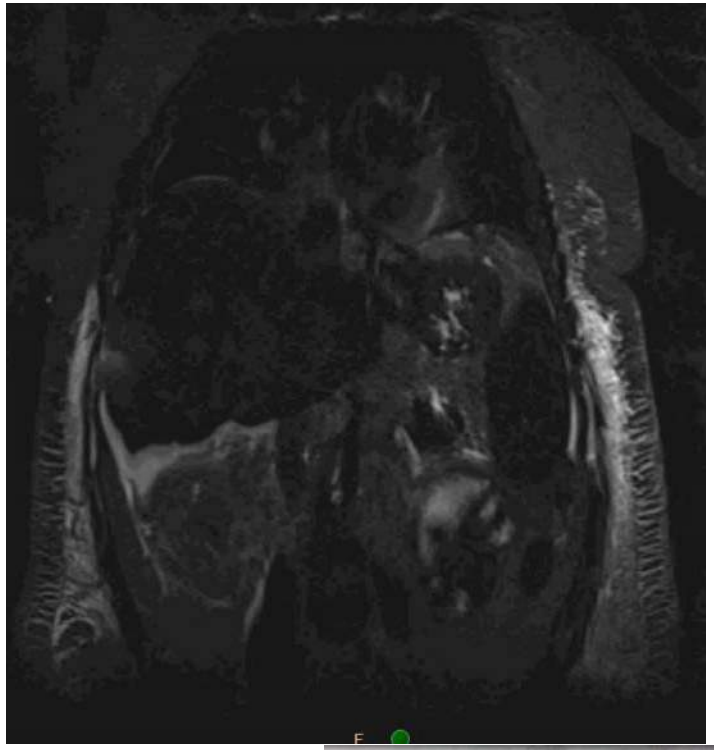
- Unremarkable recovery with no complications and discharged home on Day 4.
- Albumin normalized on post procedure day 4 and swelling significantly improved. Subjectively felt more energy
- For 8 months he had normal albumin and no diarrhea/swelling, but unfortunately symptoms returned…
- And now developed significant ascites…

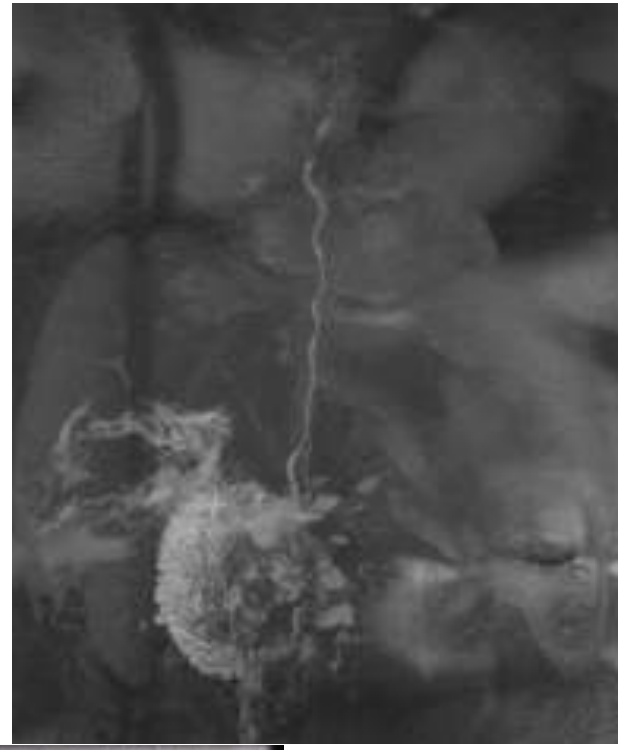
Case 1:

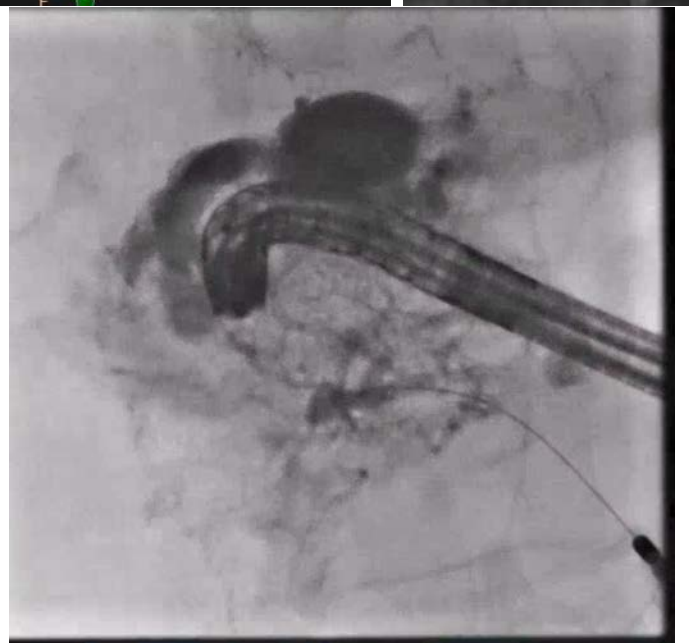
MR lymphangiogram: T2 space imaging: small volume ascites and bilateral pleural effusions. Mesenteric and supraclavicular edema and significant subcutaneous edema. DCMRL with copious intraduodenal leak with TD coursing to left venous angle.

Cath/Lymphatic intervention:
Innominate vein 20 mmHg, TD 21mmHg, Bilateral PCWP 11mmHg. TD patent to left venous angle.

- Repeat lymphatic embolization







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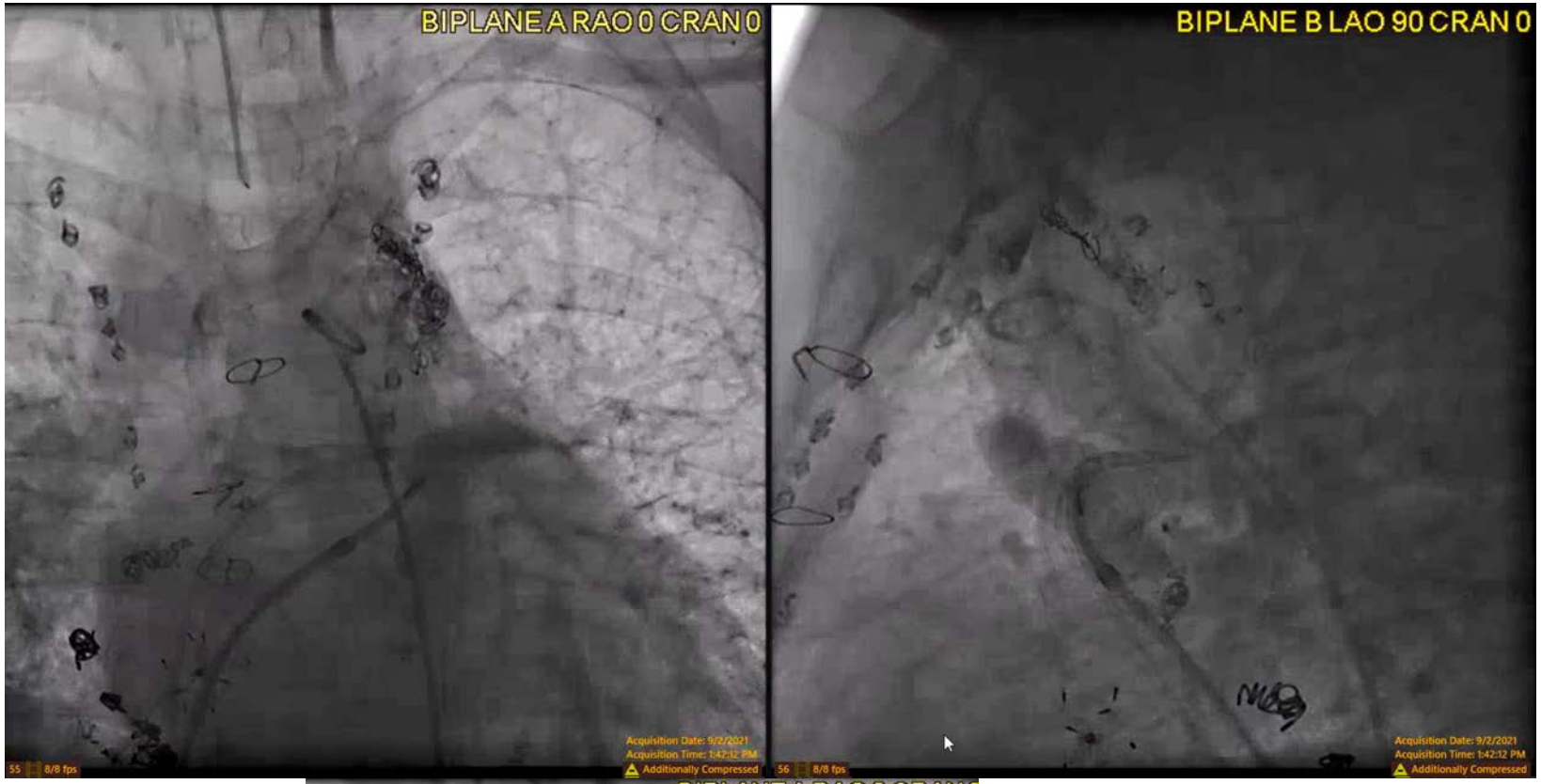
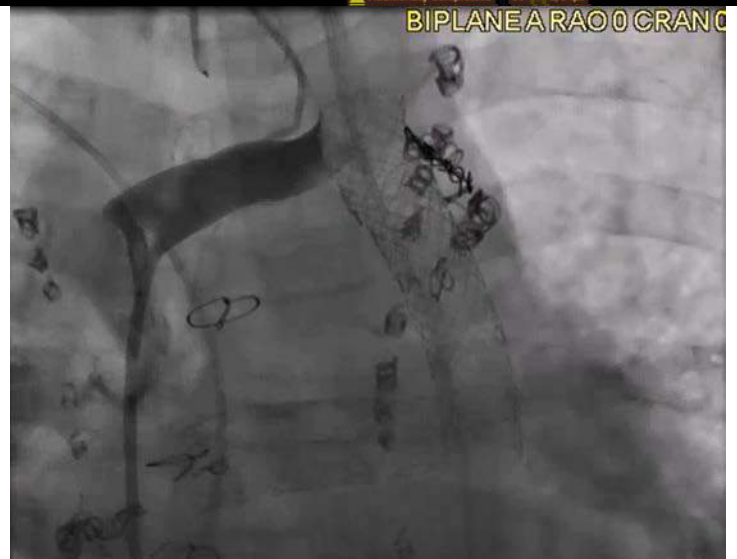
Jill & Mark Fishman Center for Lymphatic Disorders

Children's Hospital of Philadelphia
Cardiac Center

Case 1:

- Underwent TD Decompression 2 weeks after PLE embolization
- Extravascular covered stent pathway from base of innominate vein to LA Appendage
- Pre
  - Fontan m17
  - LIJ m17
  - LA m10
  - DAo 94%
- Post
  - Fontan m20
  - LIJ m15
  - LA m15
  - DAo 89%





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Case 1:

- Significant improvement with resolution of symptoms and albumin >3 g/dL
- 3 months later
  - Diarrhea, swelling, and hypoalbuminemia
  - Significantly desaturated to high 70s

- Found to have endovascular leak that was sealed with Covered CP stent
  - Pre
    - Fontan m17, LIJ m16, LA m12
    - DAo 70%
  - Post
    - Fontan m18, LIJ m13, LA m13
    - DAo 88%

- Symptoms again resolved and continues to have albumin >3 ~18 months after decompression with no ascites