



Children's Hospital
of Philadelphia®

Jill & Mark Fishman
Center for Lymphatic Disorders

CARDIOLOGY
2023

Lymphatics Case #3

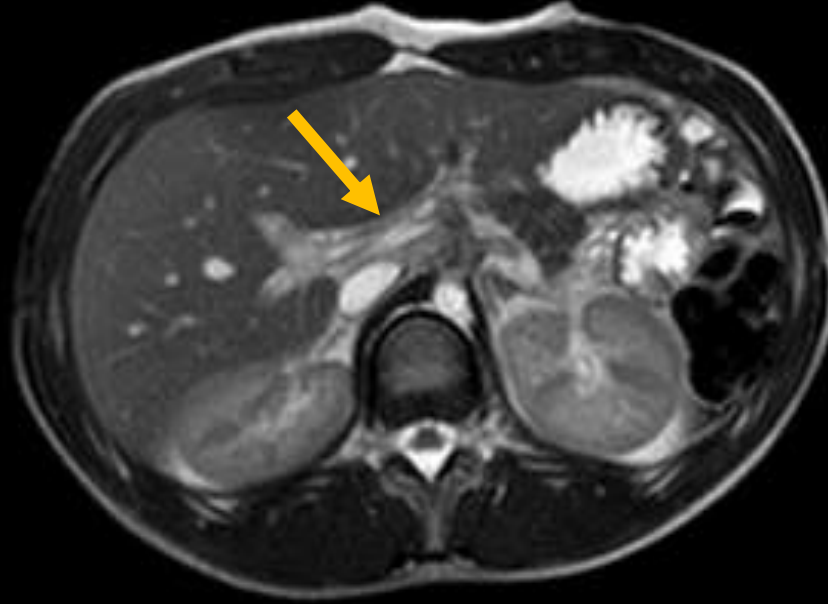
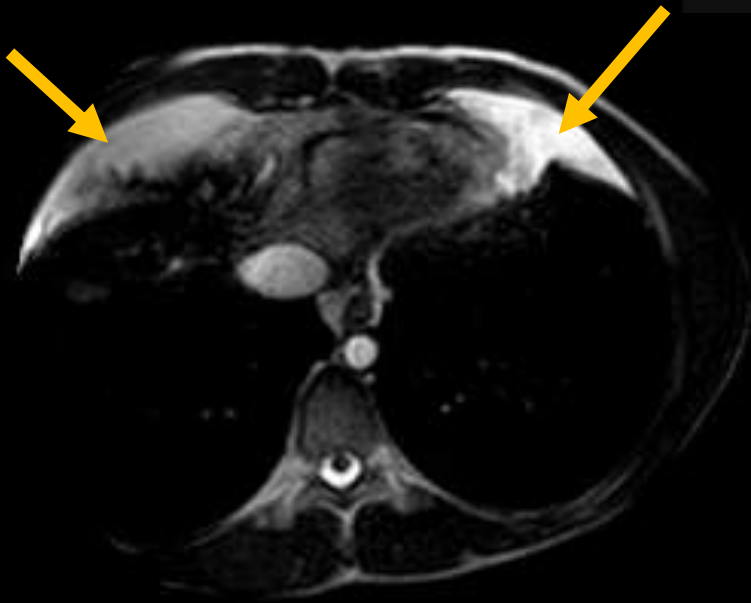


Case Presentation

- 16 yo previously healthy male with recent diagnosis of PLE
 - Elevated alpha 1 anti-trypsin levels in stool
 - Normal colonoscopy/normal intestinal biopsy
- Symptoms
 - Shortness of breath
 - 1-2 episodes of non-bloody loose stool per day
 - No abdominal pain, emesis, or nausea
- No recent travel history
- No pertinent family medical history

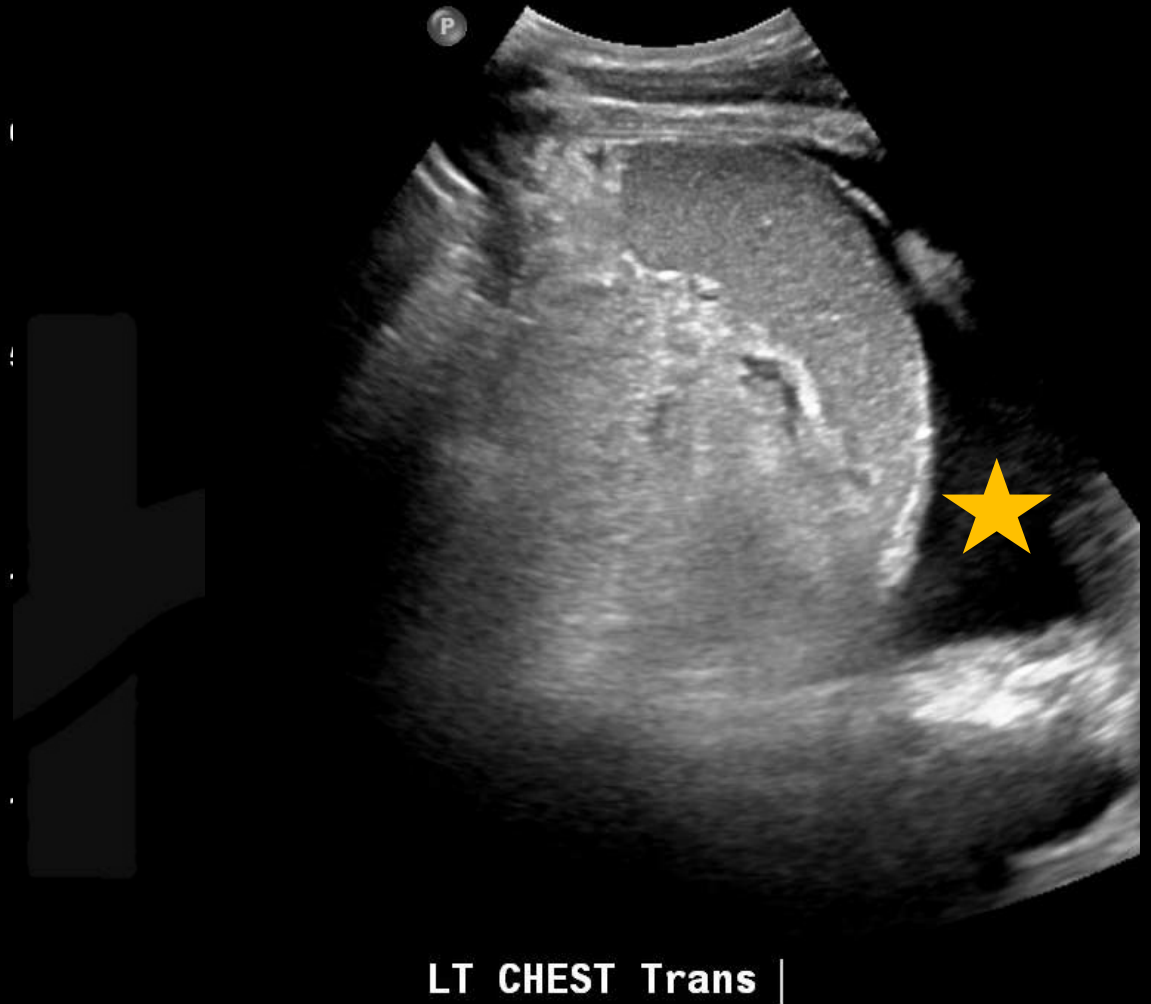
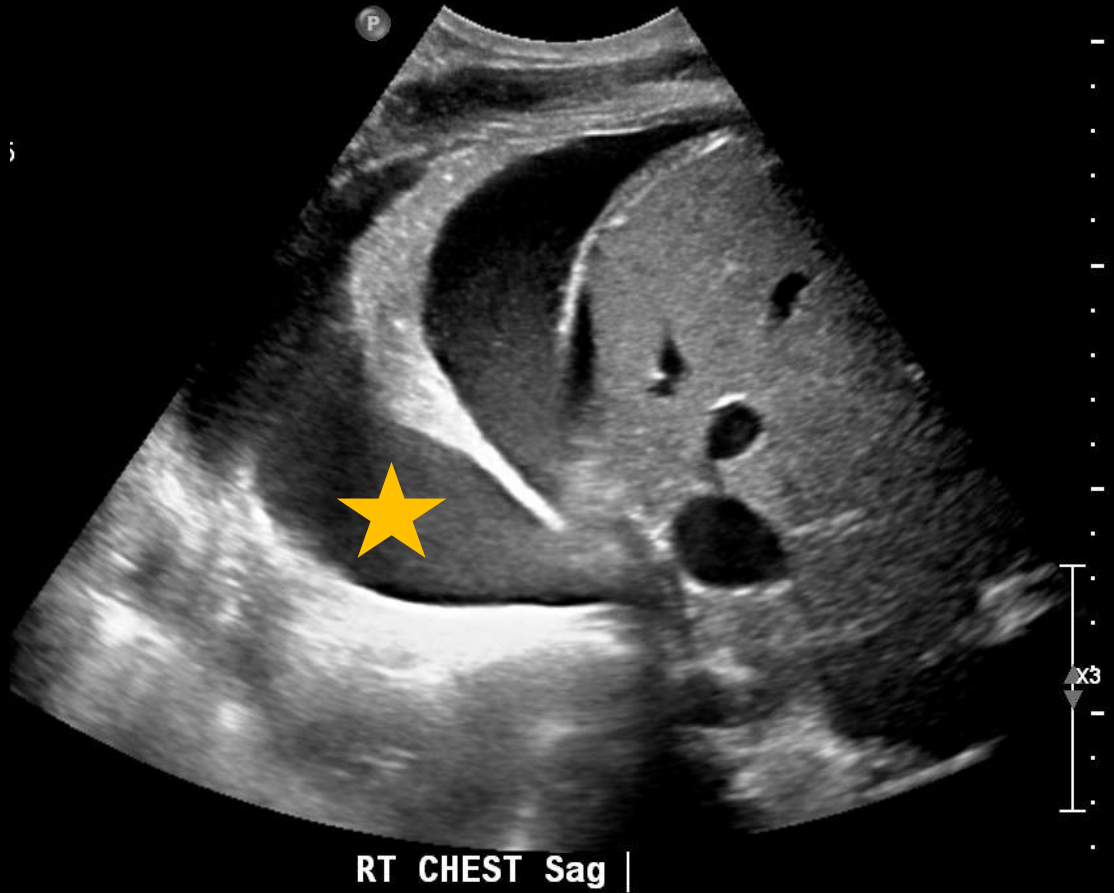
Outside Imaging

- Periportal edema
- Mesenteric/retroperitoneal edema
- Bilateral effusions

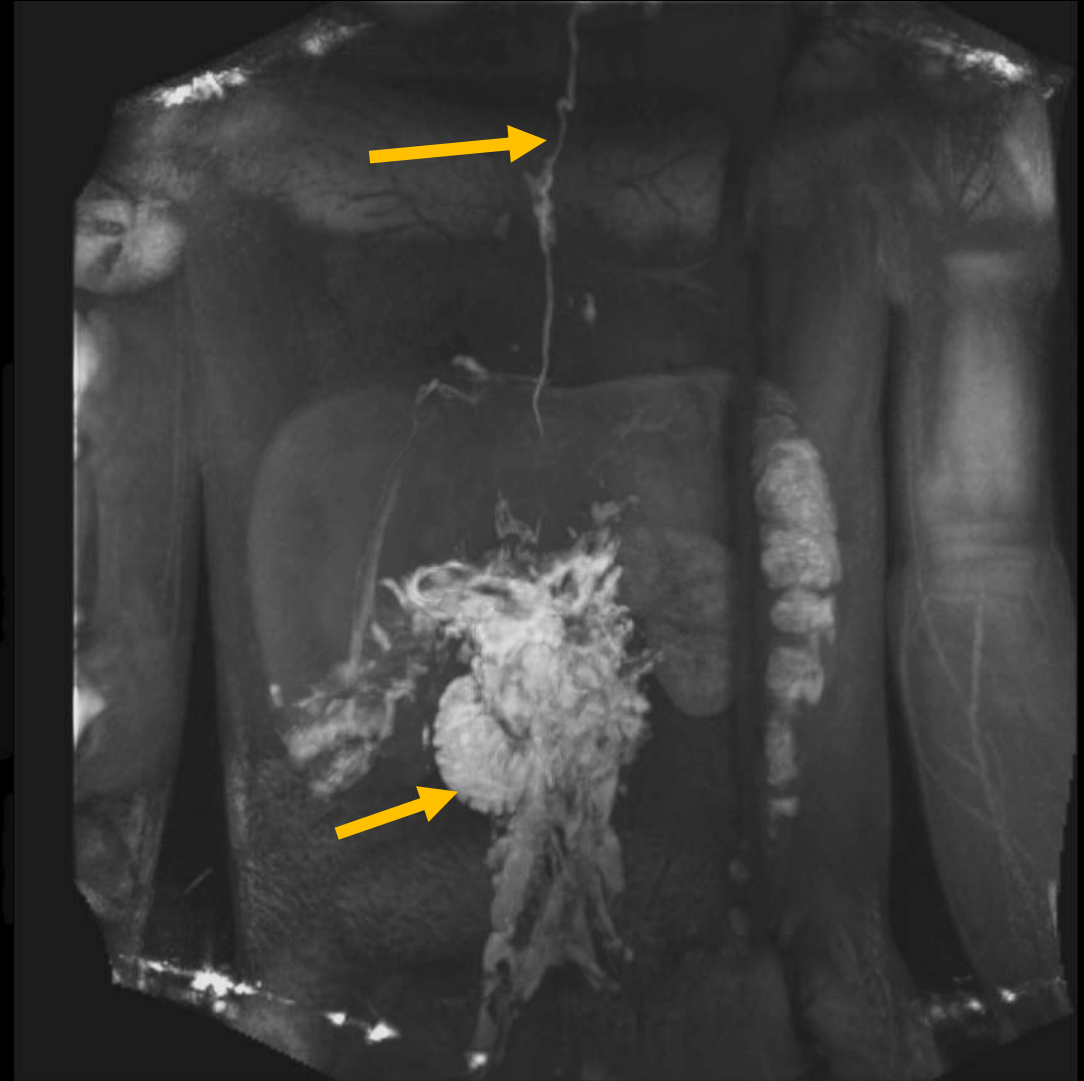
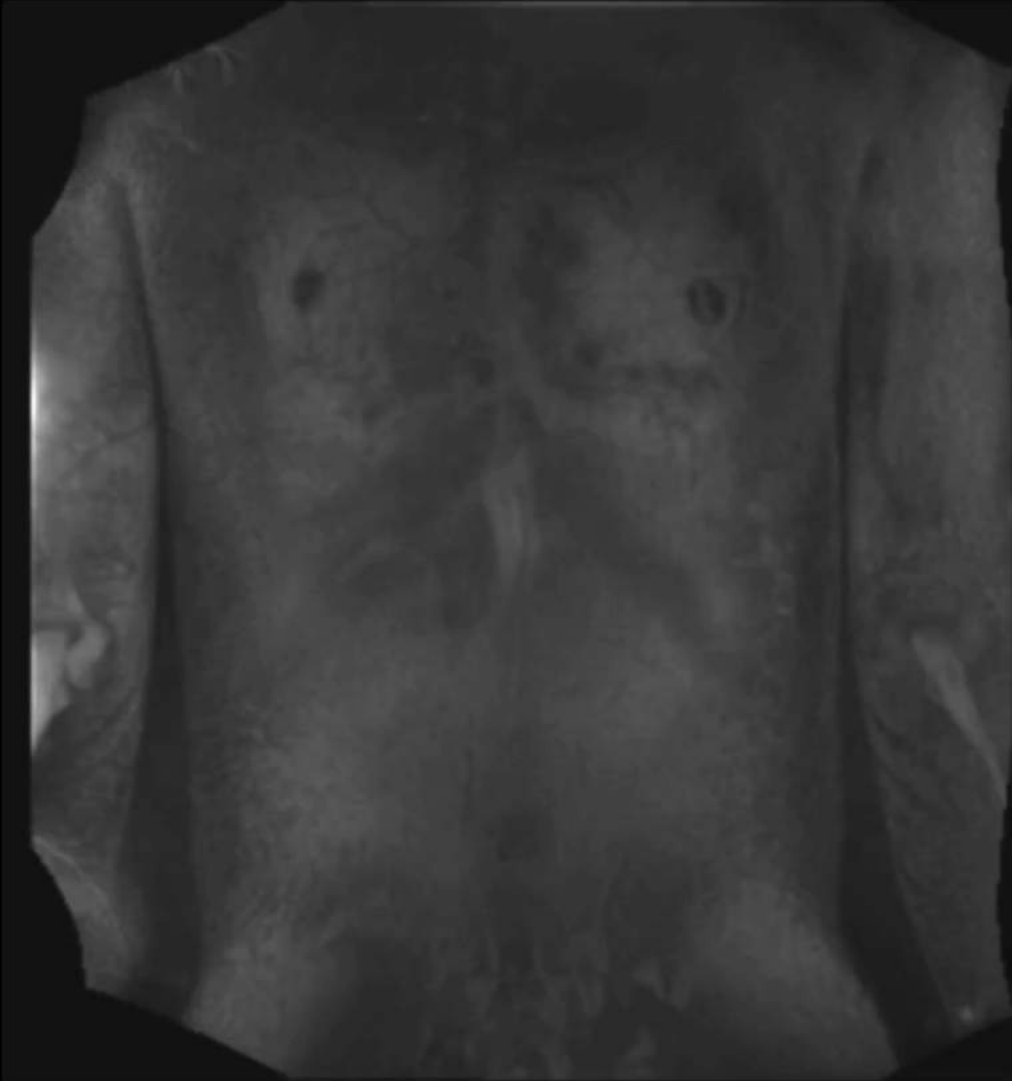


Outside Imaging

- Bilateral pleural effusions



MR Lymphangiogram



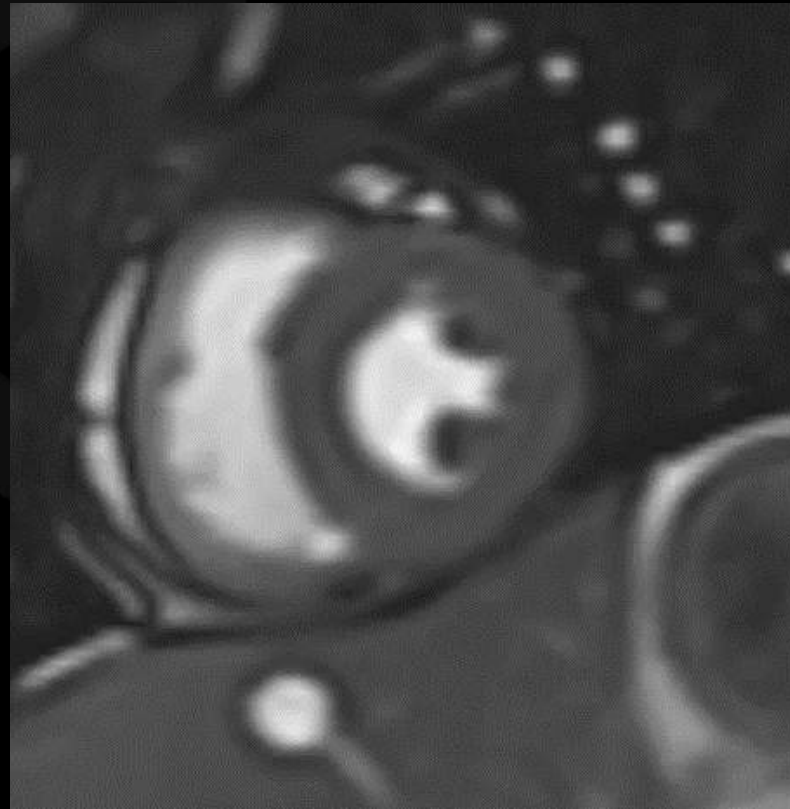
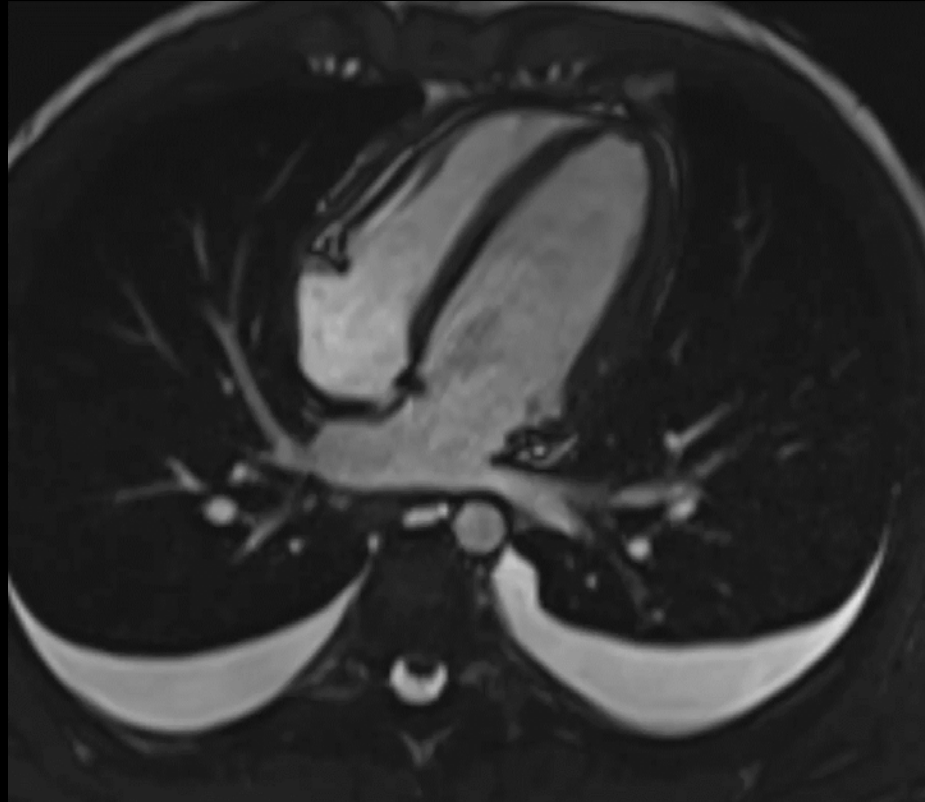
Cardiac Catheterization

Oximetry: Condition	Site	O2-Sat (%)	Sample Date/Time	PO2 (mmHg)
Rest	Superior Vena Cava	73.0	8/22/2018 12:09:31 PM	
Rest	Right Atrium	73.0	8/22/2018 12:09:51 PM	
Rest	Right Ventricle	71.0	8/22/2018 12:12:50 PM	
Rest	Pulmonary Artery Right	72.0	8/22/2018 12:24:39 PM	
Rest	Pulmonary Artery Left	71.0	8/22/2018 12:14:41 PM	
Rest	Artery Radial	96.0	8/22/2018 1:36:35 PM	124.0

Cardiac Catheterization

Pressures: Condition	Site	S/A (mmHg)	D/V (mmHg)	M/ED(mmHg)
Rest	Vein Right Femoral	14	13	13
Rest	Right Atrium	17	16	14
Rest	Right Ventricle	31	9	20
Rest	Pulmonary Artery Right	23	17	19
Rest	Pulmonary Artery Left	27	18	20
Rest	Pulmonary Capillary Wedge Right	16	16	14
Rest	Pulmonary Capillary Wedge Left	17	16	15
Rest	Left Ventricle	80	11	24
Rest	Ductus	15		14
Shunt Occlusion	Right Ventricle			
Shunt Occlusion	Left Ventricle	82	17	26
Shunt Occlusion	Aorta Ascending	74	49	59

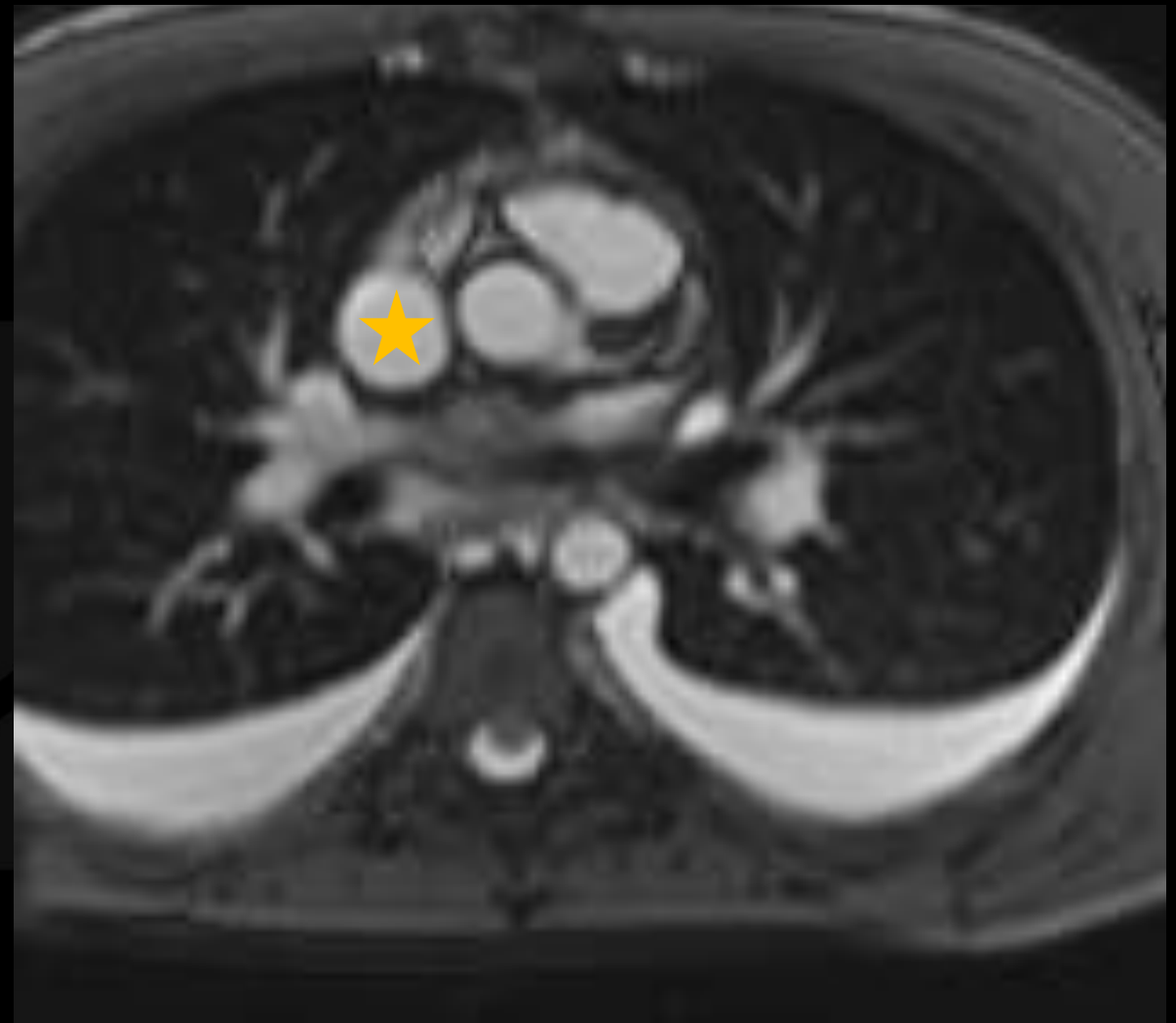
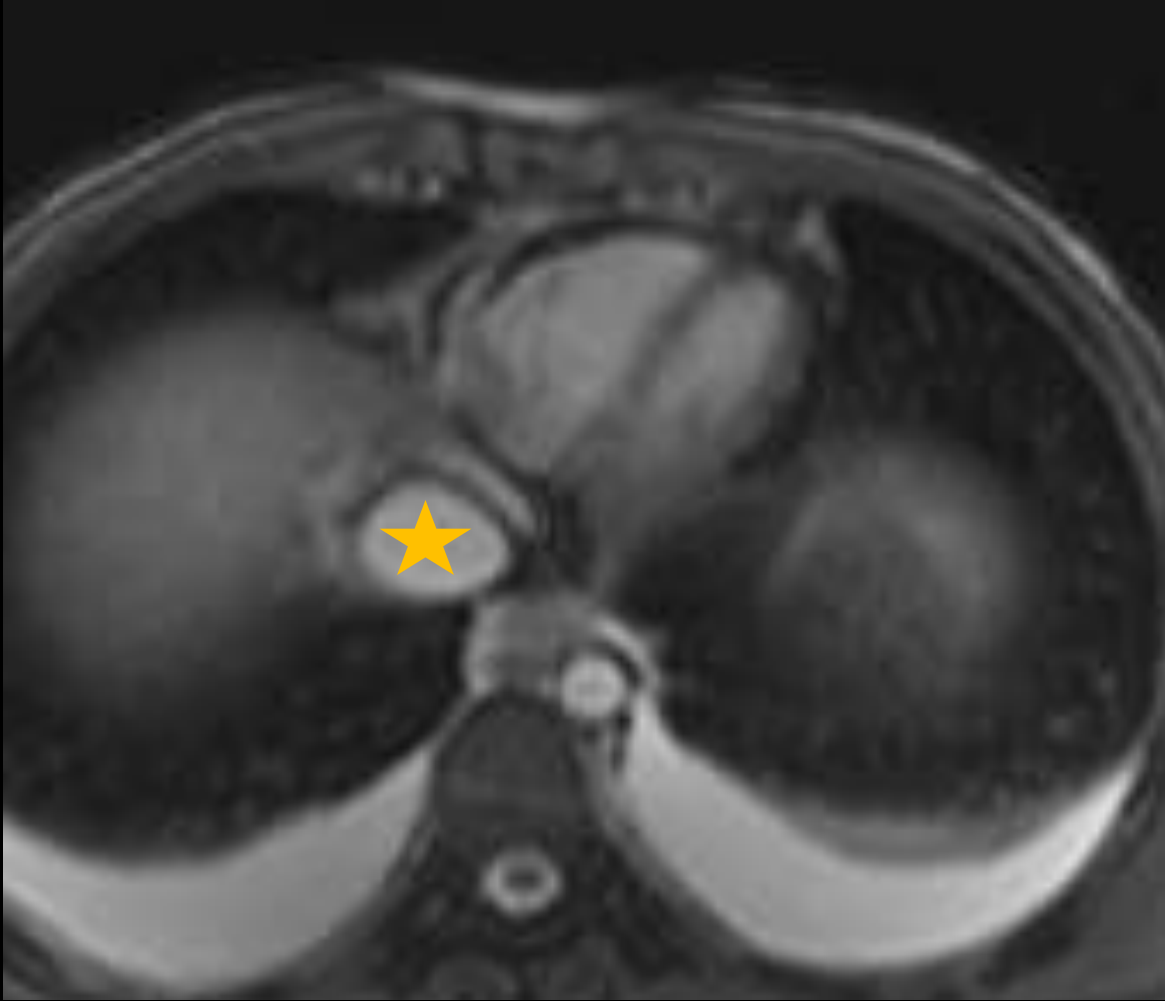
Cardiac MRI



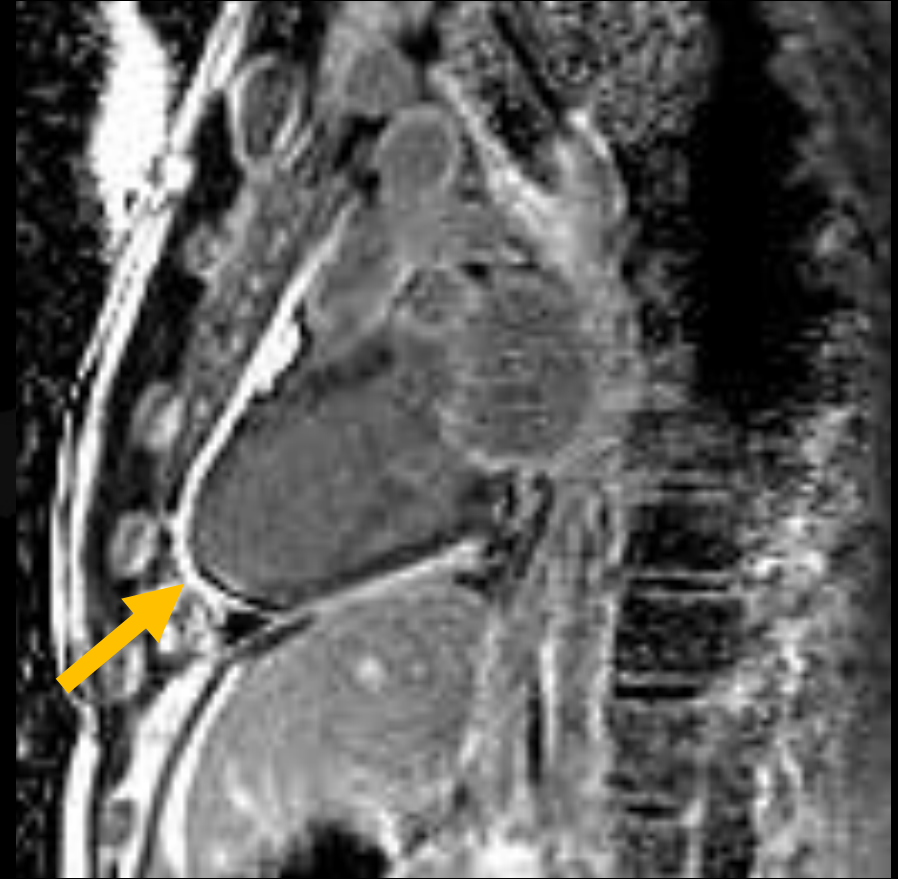
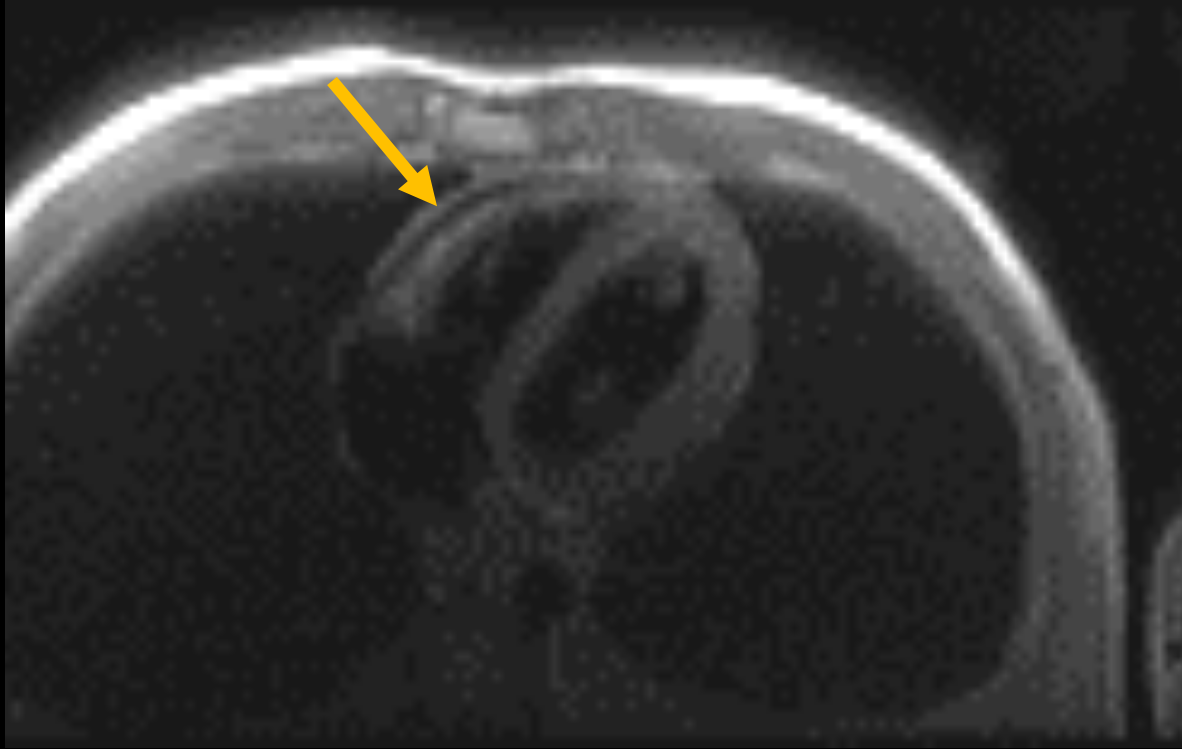
LVEF 75%
LVEDV 43 mL/m²

RVEF 70%
RVEDV 41 mL/m²

Cardiac MRI



Cardiac MRI



Status post pericardectomy

