

An uncorrected atrial septal defect presenting as a rare cause of cardiac cirrhosis

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INTRODUCTION

- The most common causes of liver cirrhosis include chronic viral hepatitis, alcohol abuse, cryptogenic, or medications however, cirrhosis due to cardiac causes (cardiac cirrhosis) is an uncommon etiology.
- The exact prevalence of cardiac cirrhosis has been undetermined as patients often have other comorbidities.
- This unique case demonstrates an uncorrected atrial septal defect (ASD) manifesting as cardiac cirrhosis.

THE CASE

History Present Illness:

66 YOM with non-ischemic cardiomyopathy, systolic heart failure with EF of 30%, atrial fibrillation and ESRD on HD who presented with hypoxia requiring 15L of supplemental oxygen.

Pertinent ROS:

- + Dyspnea, distended abdomen
- Fevers, n/v, diarrhea or other complaints

Pertinent Physical Exam:

Cardiac: RRR, S1/S2 present, no murmurs

Lungs: diminished lung sounds in RLL, transitioned to BiPAP for oxygen support

Abdomen: distended, decreased bowel sounds, non-tender

Pertinent Imaging:

CT abdomen/pelvis: large right pleural effusion with complete collapse of RLL, cardiomegaly and cirrhosis with ascites

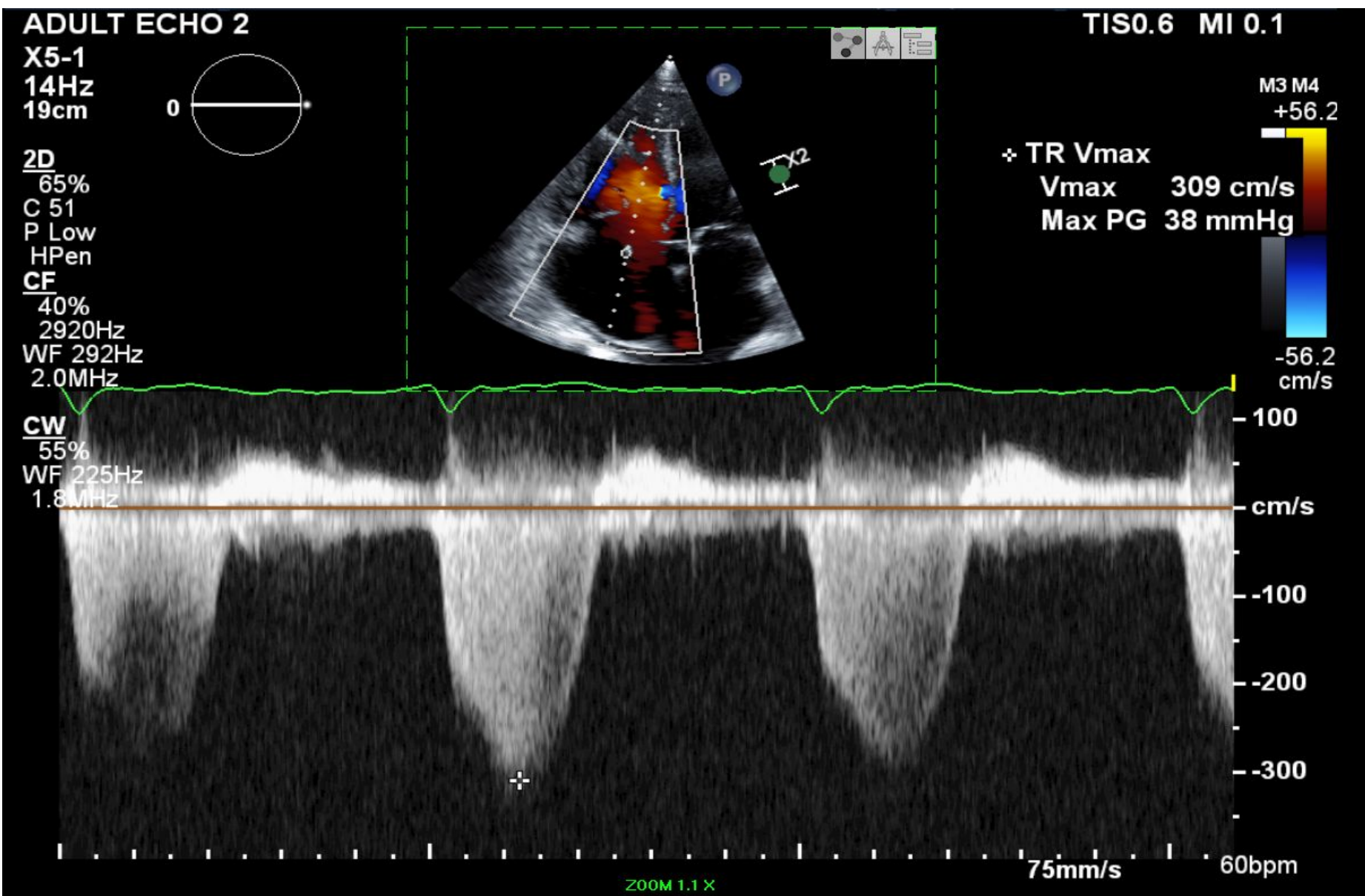


Image 1: Echocardiogram: apical four chamber view shows moderate tricuspid regurgitation (TR), TR peak gradient of 38 mmHg (reference range: <35 mmHg) and TR peak velocity of 309 cm/s (reference range: <280cm/s).

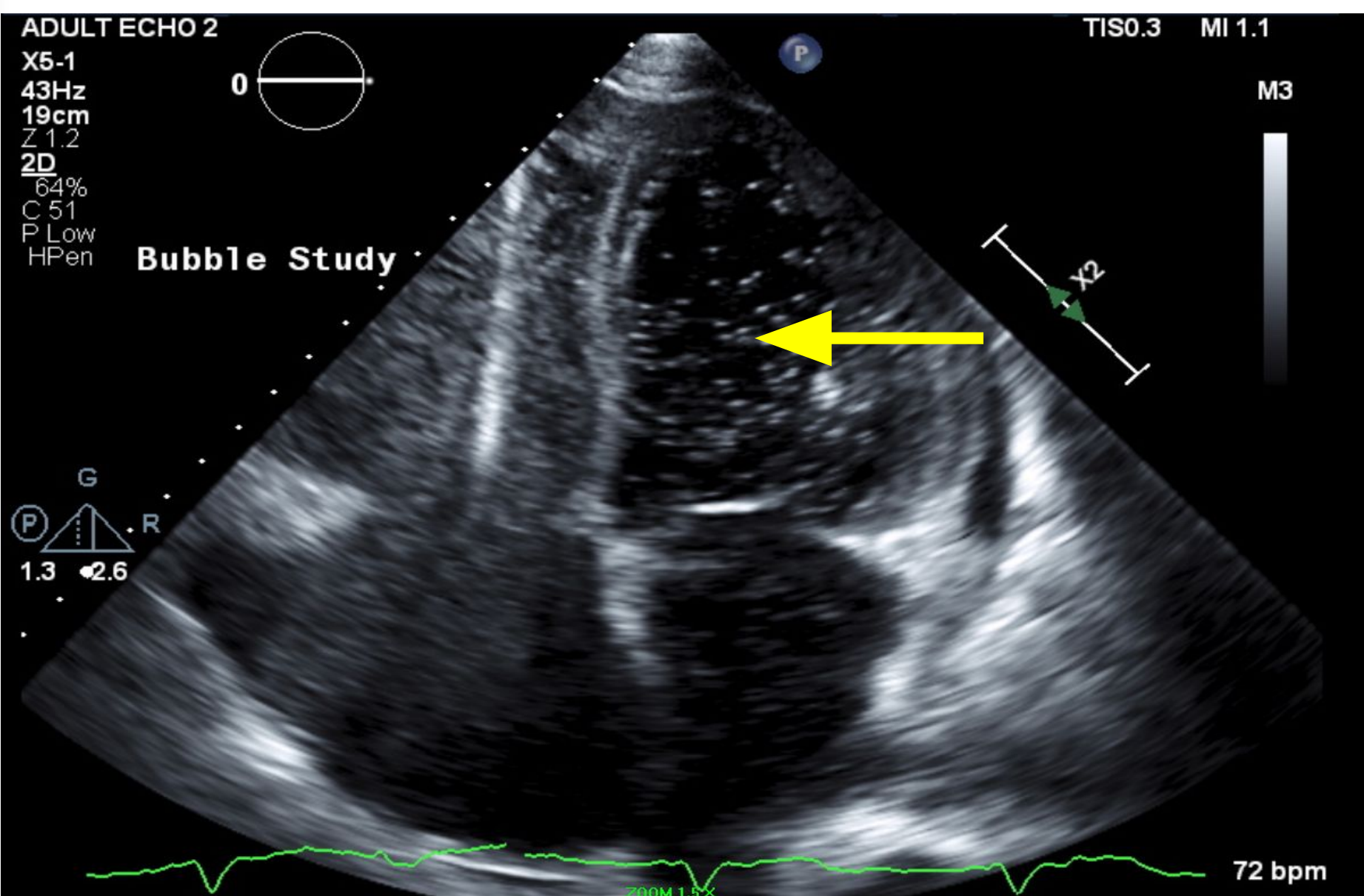


Image 2: Echocardiogram: apical four chamber view with bubble study shows bubbles in the second ventricular beat.

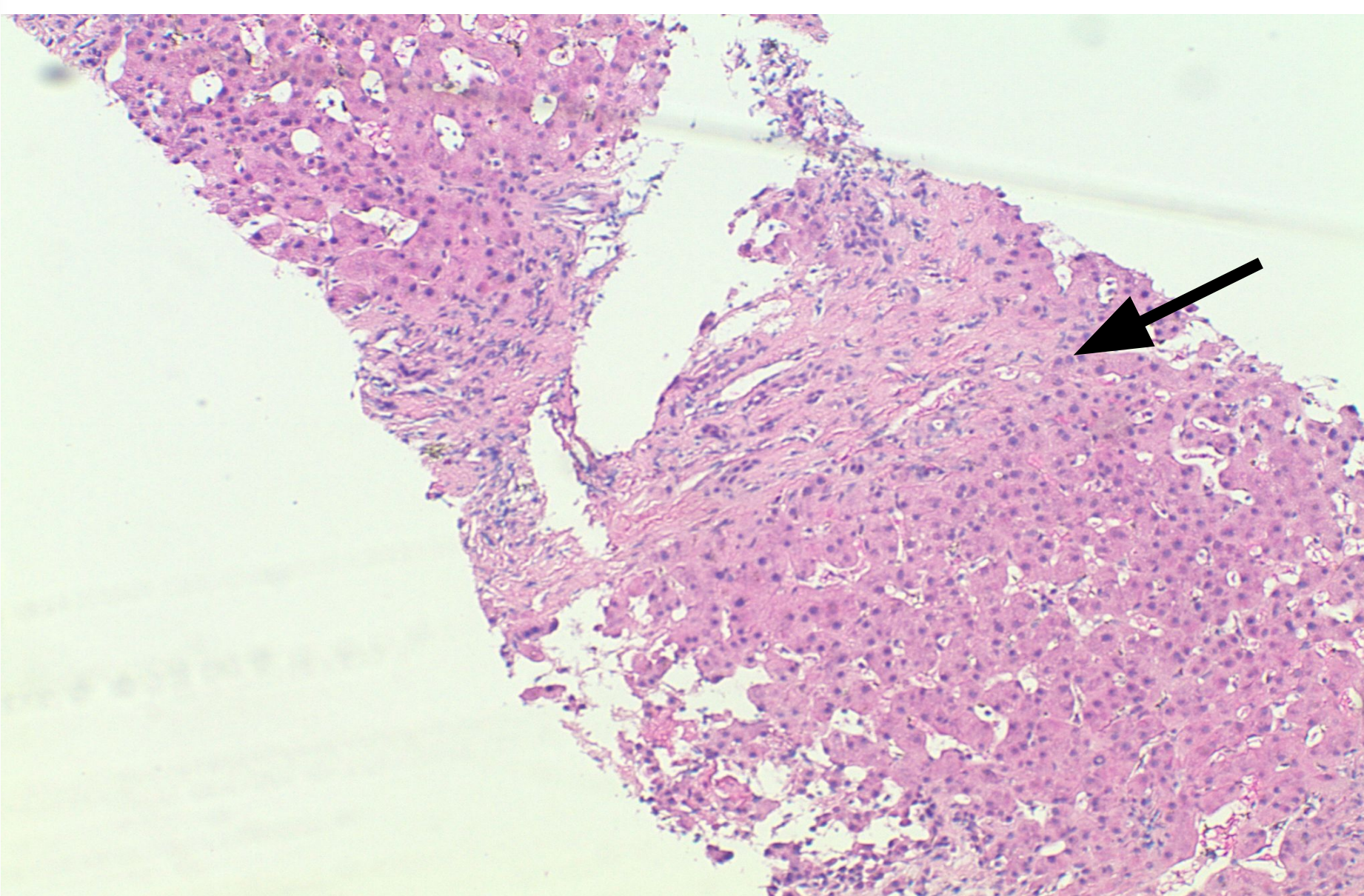


Image 3: Liver biopsy: Iron stain shows mesenchymal iron deposition consistent with history of dialysis.

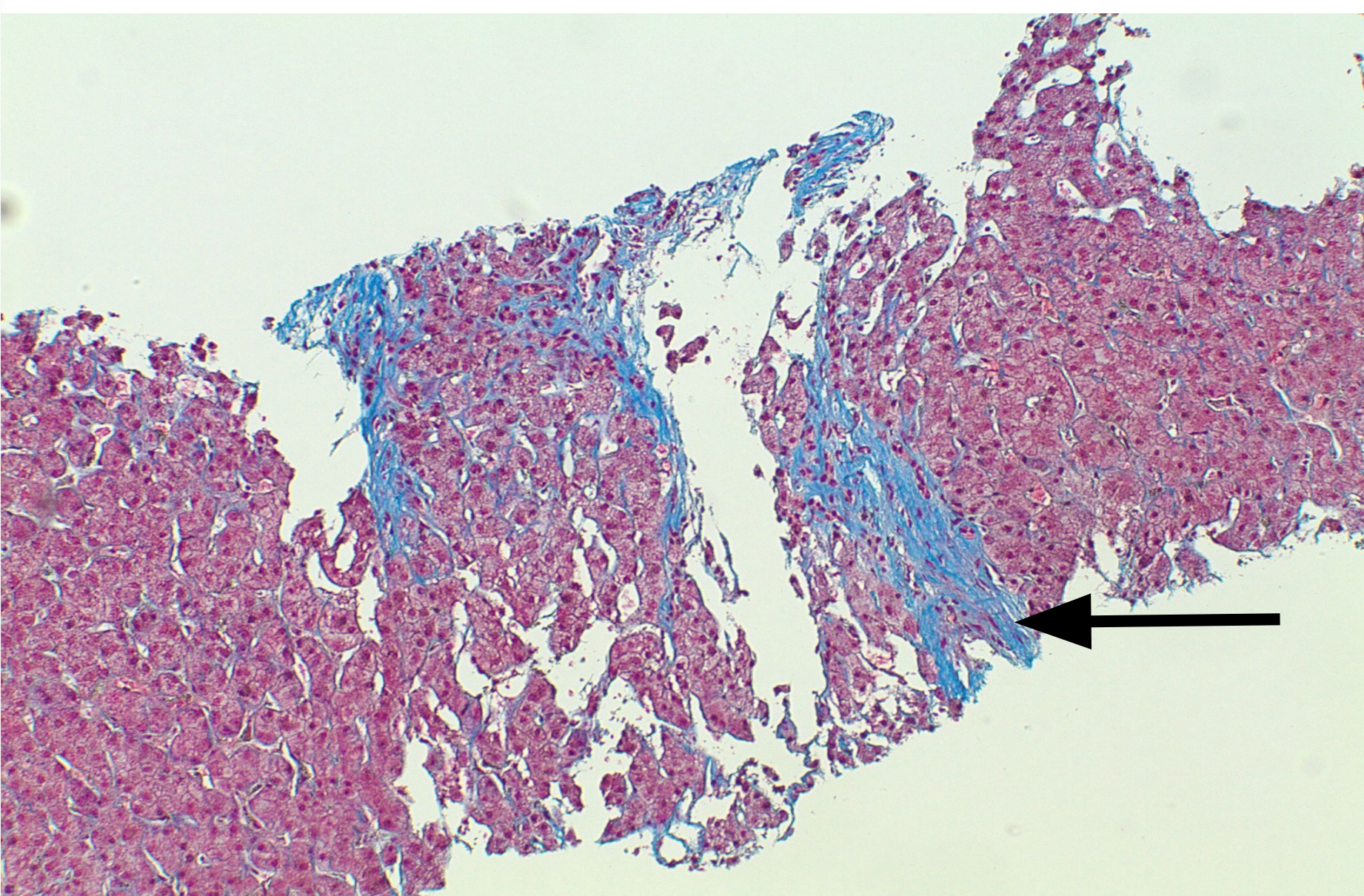


Image 4: Liver Biopsy: Trichrome stain highlights sinusoidal dilation, periportal and focal bridging and focal pericellular fibrosis.

COURSE

- **Admit for respiratory support and workup of new onset ascites**
- **Paracentesis:** showing high SAAG and an **ascites protein of 3.9**, concerning for cardiac etiology of ascites
- **Right thoracentesis:** transudative effusion 2/2 RHF vs hepatic hydrothorax
- **Echocardiogram:** EF 30-35%, S3DD, LAD, RAD, RV enlarged with reduced global systolic function, elevated systolic pressure with possible ASD
- **Portosystemic pressure:** right hepatic and portal vein pressures were normal (reference range: 3-7 mmHg and 5-10 mmHg) and hepatic venous pressure gradient of 0 mmHg (reference range: 1-5 mmHg)
- **Right Heart Catheterization:** confirmed sinus venosus ASD with left to right shunt at the SVC-RA junction
- **Liver biopsy:** pericellular fibrosis and sinusoidal dilation confirming cardiac cirrhosis

SIGNIFICANCE & CONCLUSION

- New onset ascites with an elevated ascites protein should prompt a workup for cardiac etiology.
- Cardiac cirrhosis is rare and if found, it is typically due to valvular diseases such as tricuspid regurgitation (TR).
- In our case, TR and RV dysfunction are complicated by the L to R intra-atrial ASD shunt rather than just the most common cause, left heart failure.
- The shunt added additional pre-load burden to the venous system, worsening hepatic congestion, leading to cirrhosis.
- Approx. 95% of cases with ascites fluid protein > 2.5 are of cardiac sources and it was the key clue in discovering the cause of cirrhosis.
- Using the ascitic fluid protein along with other diagnostic tests, a rare cause of cardiac cirrhosis, ASD with left to right shunt, was identified.

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