

Case Study: Autoimmune Hepatitis and Pancytopenia in Pregnancy

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Background

- Autoimmune hepatitis (AIH) is a chronic inflammatory disease of unknown etiology characterized by progressive hepatocellular inflammation and necrosis that may progress to cirrhosis.
- Treatment goals include improving symptoms, reducing liver inflammation, and preventing disease progression.^{1,2} This starts with glucocorticoid treatment, with azathioprine as maintenance.
- Overall prognosis is high, with 10-year survival rates of 90%,³ however cirrhosis is associated with higher mortality rates.⁴

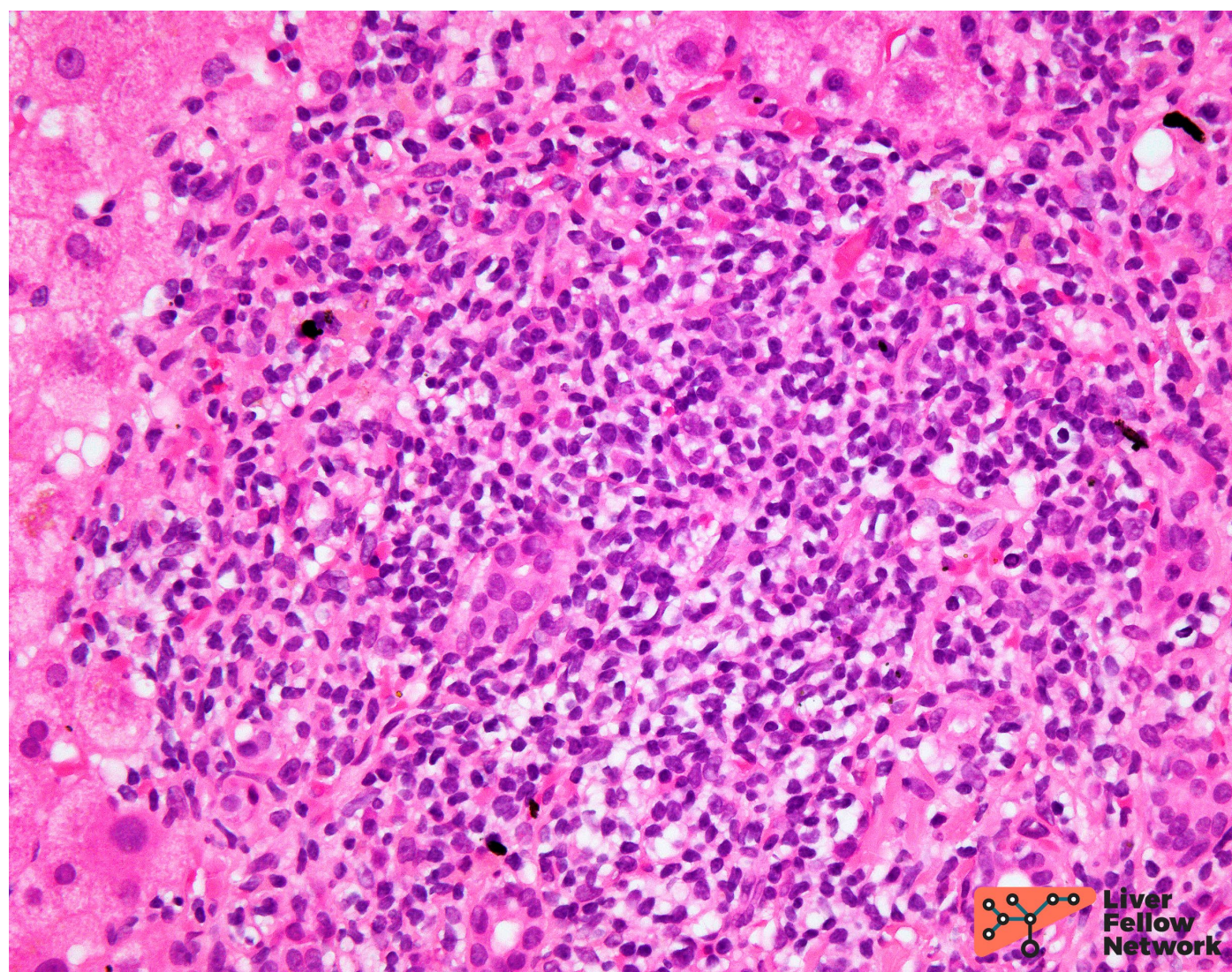


Figure 1: Liver biopsy of autoimmune hepatitis (AIH) showing portal inflammation, composed of lymphocytes, scattered plasma cells, and rare eosinophils. Pathology Pearls Post 6: Autoimmune Hepatitis. Bell, Phoenix. Figure on AASLD. Available from: <https://www.aasld.org/liver-fellow-network/core-series/pathology-pearls/pathology-pearls-post-6-autoimmune-hepatitis-aih> [Accessed 12 Oct, 2023]

- AIH disproportionately affects reproductive-aged females. AIH in pregnancy is associated with many adverse maternal and fetal outcomes.^{5, 6}

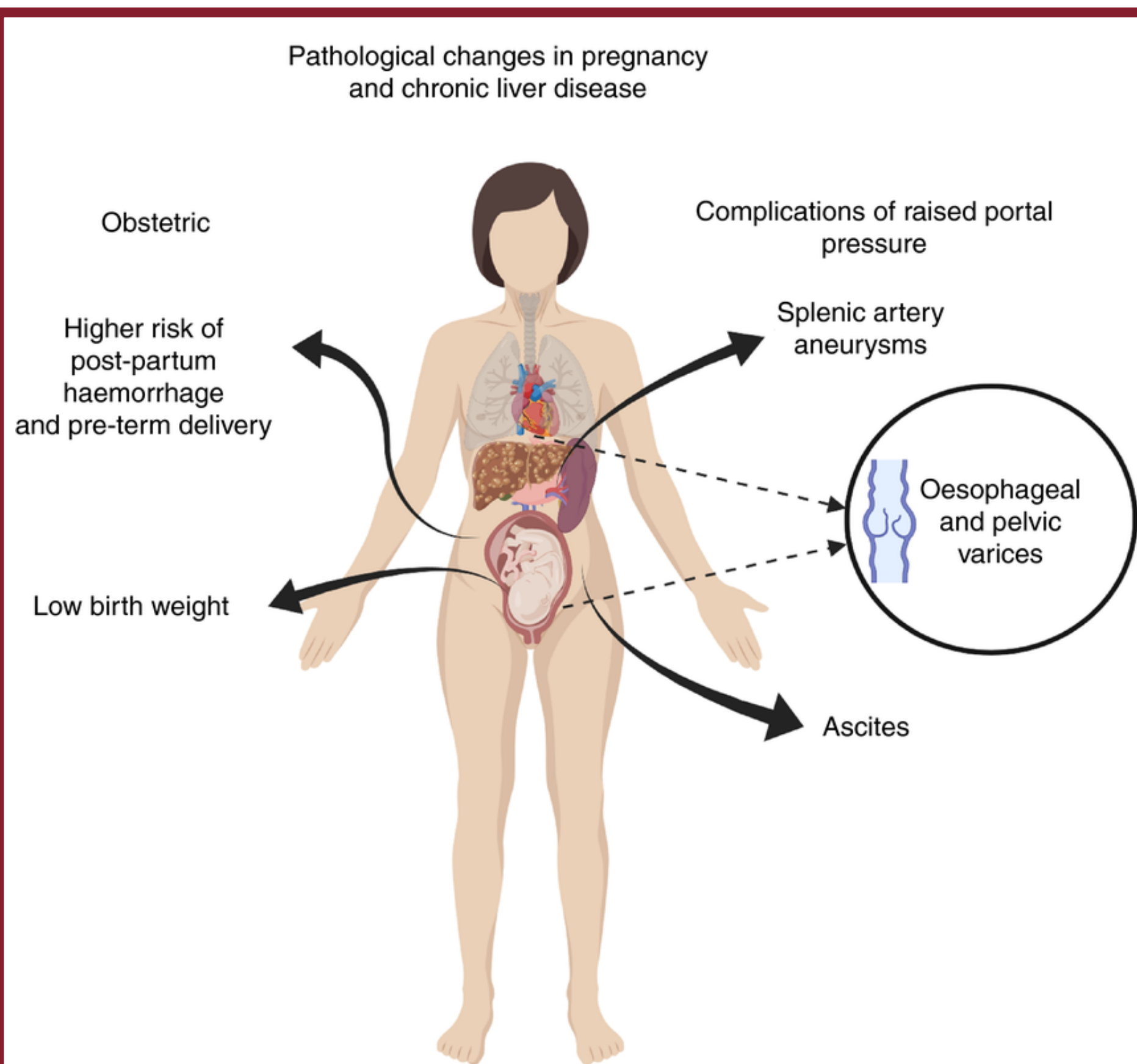


Figure 2: Review article: chronic liver disease and pregnancy - Scientific Figure on ResearchGate. Available from: https://www.researchgate.net/figure/Pathological-changes-in-pregnancy-and-chronic-liver-disease-Created-withBioRendercom_fig2_342534364 [Accessed 12 Oct, 2023]

Case Presentation

Presenting Illness and Past Medical History

- A 29-year-old G4P0121 female presented to prenatal care at 7 weeks gestation with diagnosis of AIH with cirrhosis. The patient was incidentally found to be pregnant during workup of new-onset pancytopenia.
- She had a history of spontaneous preterm delivery at 36 weeks complicated by cirrhosis, sepsis, thrombocytopenia, acute kidney injury, and spontaneous bacterial peritonitis (SBP).

Pregnancy Course

- The current pregnancy was co-managed by Maternal Fetal Medicine, Gastroenterology, and Hematology.
- The patient was maintained on azathioprine, prednisone, and trimethoprim/sulfamethoxazole throughout pregnancy.
- She developed mild pulmonary hypertension suspected due to right heart disease from AIH.
- Between 31 and 32 weeks, she was hospitalized multiple times for management of steroid-unresponsive pancytopenia of unclear etiology with severe Coombs-negative anemia unresponsive to packed red blood cell transfusion. She had received 19U PRBC in transfusions since her pregnancy started. (See admission labs on right.)

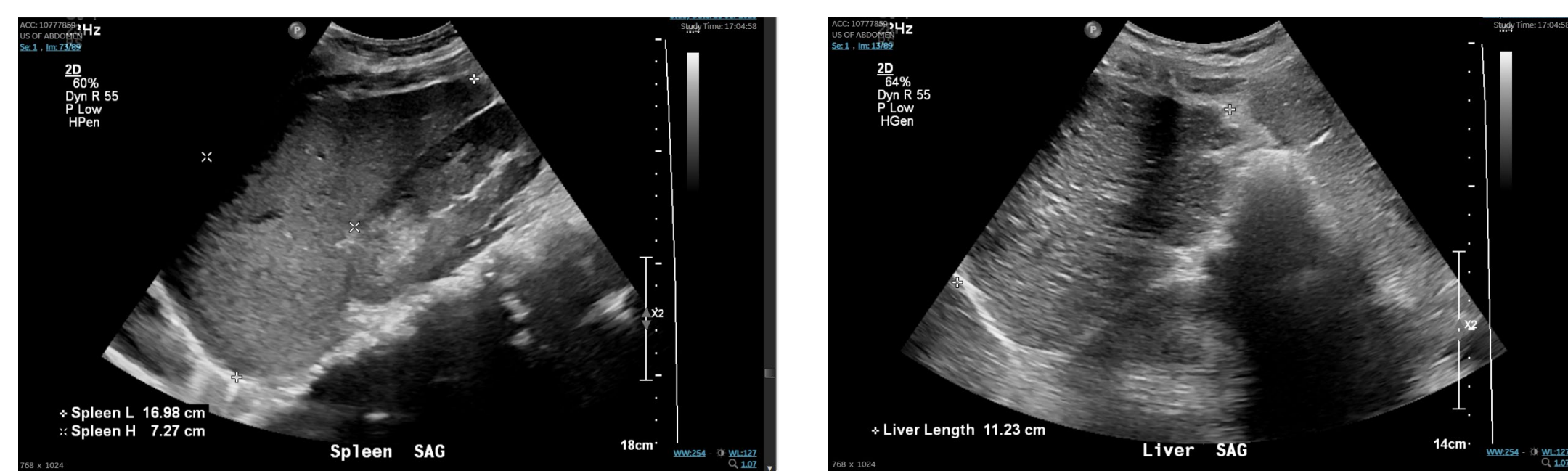
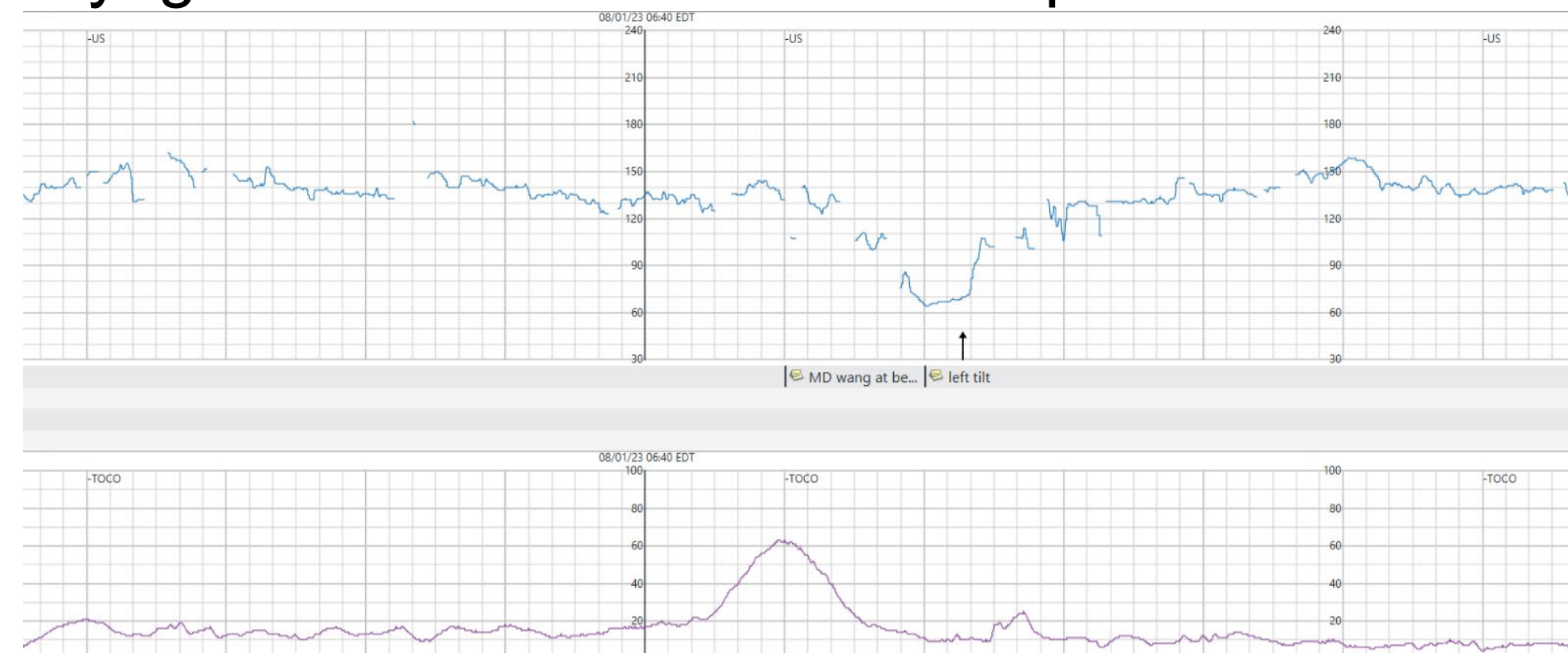


Figure 3: Abdominal ultrasound demonstrated known cirrhosis, splenomegaly

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|----------------------|--------|
| WBC | 1.60 |
| RBC | 2.05 |
| Hemoglobin | 6.4 |
| Hematocrit | 18.1 |
| MCV | 88.3 |
| MCH | 31.2 |
| RDW | 19.9 |
| Platelet | 56 |
| Anisocytosis | 2+ |
| Basophilic stippling | 1+ |
| Elliptocytes | 1+ |
| Microcytosis | 2+ |
| Ovalocytes | 1+ |
| Poikilocytosis | 2+ |
| Spherocytes | 1+ |
| Reticulocyte count | 3.19% |
| Fibrinogen | 216 |
| ALT/AST | 11/24 |
| BUN/Cr | 0/0.54 |
| Albumin | 2.4 |

Delivery Decision

- At 33 weeks, decision was made for delivery via cesarean section due to persistent Category II fetal heart tracing with fetal breech presentation.
- For preoperative optimization, the patient was transfused 1U platelet, 4U packed red blood cells, albumin, and cryoglobulin with additional blood products on call to the operating room.



Postpartum Complications

- Delivery was complicated by hemorrhage with estimated blood loss of 1L, requiring multiple blood products (3U PRBC, 4U FFP, 1U platelet) as well as tranexamic acid.
- Postpartum course was complicated by sepsis secondary to urinary tract infection, and decompensated cirrhosis with hepatic encephalopathy and coagulopathy.

Conclusion

- This case highlights the challenges in management of AIH in pregnancy, particularly the difficulty in balancing the need for immunosuppression with the increased risk of infection that may lead to sequelae such as spontaneous bacterial peritonitis and sepsis.
- The clinical presentation of AIH can be complex, and demonstrates the need for a multidisciplinary team in the care of patients who have complications.

Citations

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Acknowledgements

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