Gender: Male

Age: 7

MRN: 753394

Diagnosis: Budd-Chiari syndrome

History and Physical:

Doctor:

Hello, how are you today?

Patient's Mother:

Hello, doctor. We're here for a follow-up regarding my son, Jawad Hussain. Hes 7 years old and weighs 16 kg, standing at 105 cm. He is from Gujrat, and he has two siblings, one of whom is healthy. His birth was by C-section, and theres no known family history of any medical conditions.

Doctor:

Alright, I see. Could you please tell me more about his condition and any symptoms hes been experiencing?

Patient's Mother:

Yes, Jawad has been suffering from abdominal distension for the last six months. About six months ago, he developed this distension along with some mild pain. We initially went to a nearby clinic, where an ultrasound showed hepatomegaly and altered liver echotexture. He was referred to Childrens Hospital, where they did some lab tests and performed an endoscopy. A liver biopsy was attempted but couldnt be completed due to bleeding concerns. Recently, I noticed two episodes of blood in his stool, about 2-3 ml of fresh blood after he passed stool, but it was painless, and there hasnt been any other bleeding elsewhere. He also had high-grade fever with variations throughout the day, especially at night, and a dry cough. The symptoms improved after we started some medications prescribed by a local doctor.

## Doctor:

Thank you for explaining. I see that Jawads condition is quite serious. Do you have any history of complications or hospitalizations?

# Patient's Mother:

Yes, he was admitted to Childrens Hospital one month ago and three months ago as well. He has been diagnosed with hepatic fibrosis based on an ultrasound performed in July 2023, and there have been some decompensations like abdominal distension and mild pelvic ascites back in July.

## Doctor:

Understood. Lets go ahead and check his current status. During the examination, I observed that his weight and height are below the 25th and 5th percentiles, respectively. There was no jaundice, pallor, or edema. His liver is four fingers below the costal margin, and its firm in consistency. The spleen is also palpable, about three fingers below the subcostal margin. His abdomen is distended, with a prominent abdominal vein. I did not find any tenderness, but his liver and spleen could not be precisely measured due to the tense abdomen.

# Patient's Mother:

Thats correct. He seems to be sleeping a lot lately and doesnt respond well to pain or stimuli. Hes not very oriented either.

#### Doctor:

Thank you for that. We will continue monitoring him closely. Ive gone through his recent lab results and previous tests. His bilirubin is at 1.10, ALT is 16, ALP is 432, albumin is 3.4, INR is 1.17, WBC count is 8.14, hemoglobin is 11.44, and platelets are 244. His Hepatitis B surface antigen and anti-HCV tests were negative as of July 2023.

Patient's Mother:

Yes, we had those tests done, and they came back negative.

Doctor:

Great. As for the endoscopy, the last procedure on 27th July 2023 showed grade 2 varices with no gastroenteropathy, and one band was applied. The follow-up endoscopy on 12th October 2023 showed grade 1 varices with moderate to severe gastroenteropathy. Based on the ultrasound performed on 15th July 2023, it was noted that his liver was enlarged, measuring 13 cm, with altered echogenicity. The spleen size was 9 cm. His diagnosis of congenital hepatic fibrosis is confirmed.

Patient's Mother:

Yes, the doctors mentioned congenital hepatic fibrosis as the diagnosis.

Doctor:

For treatment, were currently prescribing the following medications: Inderal 10 mg twice a day, Oss-Pro syrup 5 ml once daily, Cefspan DS syrup 5 ml once daily, A Dew drops two drops daily, and Indrop D once every 15 days. I have also advised a liver biopsy and shearwave elastography in the plan. Additionally, well continue surveillance endoscopy and follow-up after three months.

Patient's Mother:

Thank you, doctor. We will follow the plan as suggested. Is there anything else we should be concerned about?

Doctor:

Currently, the focus is on managing his symptoms, preventing further complications like variceal bleeding, and closely monitoring liver function. Well continue to adjust his treatment based on his

response. Please also be mindful of his diet and the medications. Lets plan for a follow-up visit in three months. If his condition worsens before then, dont hesitate to contact us sooner.

Patient's Mother:

Thank you so much for your care, doctor. Well make sure to follow up regularly and keep you updated.

Follow-Up Visit on 26th February 2024

Doctor:

Hello, how has Jawad been doing since we last saw him?

Patient's Mother:

Hes been having some issues with black, tarry stools again, and weve noticed that his abdominal distention persists. He has also been sleeping a lot and is still quite irritable.

Doctor:

Lets review his recent tests. His latest endoscopy on 10th February 2024 showed grade 1 varices with gastroenteropathy. He had a follow-up ultrasound on 17th March 2024, which showed mild hepatomegaly with homogeneous texture. His liver enzymes are still elevated, and his albumin levels remain low.

Patient's Mother:

Yes, weve been following the treatment regimen, and his symptoms have been somewhat manageable, but theres still a lot of concern.

Doctor:

Well continue with the treatment plan as advised. Theres no change in his medications: Inderal, Oss-Pro, and other treatments. I would also recommend that you follow up with a specialist for further optimization of his medical care. Well keep a close eye on his liver function and general well-being.

Patient's Mother:

Understood, doctor. Well keep following up as instructed. Thank you for your ongoing support.

Doctor:

Youre welcome. Keep a close watch on his condition and reach out if you notice any significant changes. See you in three months for the next follow-up.

Final Follow-Up on 12th June 2024

Doctor:

Hello, how is Jawad doing today?

Patient's Mother:

Hes been struggling with his symptoms, but weve kept up with his medications. Hes been having more frequent black stools, and his distended abdomen has caused more discomfort.

Doctor:

Based on his latest examination and lab results, weve confirmed that his liver is still showing signs of significant strain. The next step is to consider liver transplantation. Well need to work closely with specialists for counseling and further evaluations. Lets continue the current medications and keep managing his symptoms as best as we can.

Patient's Mother:
Were ready to take any necessary steps. Thank you for your continued care.
Doctor:
Well continue to support you and Jawad through this. Lets keep a strong line of communication and

address any issues that arise.