

Gender: Female

Age: 6

MRN: 745887

Diagnosis: Budd-Chiari syndrome

History and Physical:

Patient:

Hello, doctor. I am Dua Fatima, a 4-year-old girl. I weigh 16.20 kg, and I'm from Mianwali. I have five siblings, all of whom are healthy. My family history is unremarkable, and I was born via normal vaginal delivery at term with no complications. I'm up-to-date with all my vaccinations, and my development has been normal.

Doctor:

Hello, Dua. I see you are here today for an opinion. Can you tell me more about your health history?

Patient:

I had some issues about a year ago, when I was in the U.S. I developed abdominal pain and swelling. My doctors there referred me to Mianwali because my liver was enlarged. They did some tests, and then I was sent to the Childrens Hospital Lahore (CHL). I also have a history of fits, which I'm still dealing with.

Doctor:

I understand. How has your liver been since then? Have you noticed anything unusual like ascites (fluid buildup)?

Patient:

Yes, I do have ascites. The doctors have been monitoring it. They diagnosed me with cryptogenic liver cirrhosis and autoimmune hepatitis. My past surgeries have been unremarkable, and there's no

remarkable family history of similar conditions.

Doctor:

Thank you for sharing that with me. Let's take a look at your lab results from earlier tests. Your bilirubin levels are normal, but your ALT and AST are elevated, which indicates some liver stress. Your INR is 1.13, which is within the normal range. We'll need to monitor these closely.

Doctor:

Your abdominal ultrasound from June showed altered liver texture with gross ascites, and your CT scan results confirmed a congested liver. Your liver biopsy from July showed chronic hepatitis with minimal activity and cirrhosis. No malignancy was found, which is a good sign.

Doctor:

Your medications currently include Carvedilol, Spironolamide, and Vitamin K, which are helping manage your condition. You will also continue with these medications as we proceed with your treatment plan.

Doctor:

Based on your condition, we will need to conduct a few more tests. I'd recommend checking for ANA, IgG, ceruloplasmin, and 24-hour urinary copper levels. A liver transplant is likely to be the definitive treatment for you. We will also need to perform a follow-up abdominal ultrasound, CT scan, and EGD from CHL.

Doctor:

In the meantime, you will need to follow a salt-free diet. Your medications will continue as they are, but please bring a donor for your next visit. It will be important to have a donor who is 18 or older, as the prospective donor is currently 15 years old.

Follow-Up Visit on 29th August, 2023:

Patient:

Hello, doctor. Im Dua Fatima again. I have been feeling feverish with abdominal distension.

Doctor:

Lets check your condition. I notice grade 1 clubbing and some prominent superficial veins in your abdomen. Your liver is palpable, but theres no ascites at the moment. Your CT scan from 30th August confirms hepatosplenomegaly with a congested liver and ascites.

Doctor:

We also did an ultrasound, which showed that your liver is enlarged with a heterogeneous texture, and the caudate lobe is prominent. Your liver biopsy from July confirmed chronic hepatitis with cirrhosis, but no malignancy was detected. Your liver is still under significant stress, and the treatments will continue to focus on managing that.

Doctor:

We will continue with your salt-free diet, and Im prescribing Aldactone and Inderal to help manage your liver function. Please take the recommended medications for constipation and other supportive care. We will be referring you to the liver transplant team regarding the donor workup. If you experience any changes, please contact us.

Follow-Up Visit on 6th September, 2023:

Patient:

Doctor, Ive been having a fever and abdominal distension again. My condition seems a bit worse.

Doctor:

Lets check you over. You still have grade 1 clubbing, but your liver is now 3 cm below the costal margin, and ascites has returned. Well continue monitoring your liver function with Aldactone and Inderal, and Im adding Vitamin K as well.

Doctor:

I see that we need to proceed with more lab tests, including Protein C, Antithrombin III, CBC, LFT, and INR. Well follow up with you after 10 days and adjust treatments accordingly.

Follow-Up Visit on 18th September, 2023:

Patient:

Hello, doctor. Im feeling much better now. My condition has stabilized, and Im not having fever anymore.

Doctor:

Thats good to hear. You are still an active and alert child. We have reviewed your lab results, and I see youre doing better. The ultrasound from 22nd August confirmed an enlarged liver with nodular echotexture, but theres no ascites at the moment. The CT scan from 30th August showed signs of Budd Chiari Syndrome along with chronic liver disease changes.

Doctor:

Your Shearwave elastography from 18th September indicates F4 stage of hepatic fibrosis, so we need to continue careful monitoring of your liver. Youre doing well on your medications, and well proceed with your next steps.

Follow-Up Visit on 24th October, 2023:

Patient:

Doctor, Im back for my follow-up. Ive been feeling fine, except for a few issues with mouth ulcers.

Doctor:

Its good to hear that youre feeling well. Your condition still shows signs of Budd Chiari Syndrome, but no new issues have arisen. Well continue to monitor your liver function closely with ultrasound and regular checkups.

Doctor:

Your latest ultrasound shows an enlarged liver with a heterogeneous texture and ascites, but well monitor these developments. Well repeat the EGD after a year and continue managing your medications.

Overall Plan Moving Forward:

Continue with current medications: Aldactone, Inderal, and Vitamin K.

Maintain a salt-free diet.

Follow up with liver transplant team and donor workup.

Monitor liver function with regular tests and imaging.

Repeat EGD in a year.

End of Visit Summary:

We are working closely on your liver transplant evaluation, and we'll keep an eye on your condition with regular follow-ups. If you feel any changes or discomfort, please contact us right away.