

Doctor:

Hello, Umer. I see you are 2.5 years old, weighing 12 kg and measuring 87 cm in height. You are the product of a consanguineous marriage, and you have two siblings who are alive and healthy. However, you had two siblings who passed away due to polycystic kidney disease. How are you feeling today?

Patient's Caregiver (or Parent):

Umer seems fine today, but he has had some past medical issues. He was diagnosed with polycystic kidney disease at the age of 6 months after being investigated for a minor illness. His symptoms included abdominal distension at 6-7 months of age. He also experienced hematemesis and melena multiple times. He underwent endoscopy with band ligation seven times, the last one being in May 2022. His most recent episode of upper GI bleeding was in March 2022.

Doctor:

I understand. I can see you have a history of complications due to polycystic kidney disease. Let me review your previous lab results and ultrasound. In your last ultrasound from October 2020, it showed bilateral cystic kidney disease associated with hepatic fibrosis, though there was no evidence of progressive portal hypertension.

Can you share any new symptoms or concerns today?

Patient's Caregiver (or Parent):

No new complaints today. The only ongoing issue is his abdominal pain, which he has had for the last 15 days. He is on medications for his condition, including Inderal 5 mg twice daily and Dmax drops.

Doctor:

Thank you for the update. I will review his recent medical records and tests. His latest blood tests

show a bilirubin level of 0.4, ALT of 19, and ALP of 70. His INR was 1.07, which seems within normal limits. His hemoglobin in March 2022 was 10.0 g/dL, and his platelet count was 151. There is no jaundice or pallor observed in the examination. However, his liver size is noted to be slightly enlarged, with a 3 cm liver span, and his spleen is 2 cm palpable. His abdominal distension and the presence of ascites are concerning.

Doctor:

I will order repeat baseline labs, including CBC, LFTs, RFTs, S/E, INR, and ultrasound of the abdomen. We need to monitor his condition closely. Additionally, we may proceed with liver biopsy as part of his upcoming procedure at the Children's Hospital.

I recommend you follow up in the pediatric nephrology clinic after one month.

Doctor:

Hello, Umer. I see you are now 3 years old. Your weight is still 12 kg. You have been diagnosed with autosomal recessive polycystic kidney disease and congenital hepatic fibrosis. How have you been feeling recently?

Patient's Caregiver (or Parent):

Umer is doing well, but he still experiences some abdominal distension. We haven't had any active complaints recently, but his weight and height are still below the 5th percentile for his age. He continues with the same medications: Inderal 10 mg in the morning and 5 mg in the evening, along with Dmax drops once daily.

Doctor:

It's good to hear there are no active complaints. During today's examination, Umer has a distended abdomen with superficial veins and a spleen that is 6 cm palpable. His liver is 1 cm enlarged, and there is ascites present. His kidneys are not palpable on bimanual examination.

The liver biopsy from May 2023 confirmed congenital hepatic fibrosis, with 60-70% of the biopsy volume showing fibrous bands. There's no evidence of cirrhosis, which is reassuring.

Doctor:

I reviewed his endoscopy results, and the most recent one from May 2023 showed grade II varices at the 12 o'clock position. Previous endoscopies also showed grade 1 and grade 2 varices, with banding applied during several of them.

Doctor:

For today's follow-up, we will proceed with the necessary investigations including CBC, RFTs, LFTs, S/E, and INR. We will also repeat the urine C/S and urine R/E. Given his condition, we are considering liver transplantation as a definitive treatment.

You can continue his current medications, and we will schedule follow-up after the lab results.

Plan for Future:

Repeat CBC, LFT, RFT, S/E, INR

Ultrasound abdomen and pelvis

Liver biopsy scheduled

Continue current medications (Inderal, Dmax)

Follow-up visit in one month after lab tests