Case Discussion

Primary Sclerosing Cholangitis

Dr. Manzoor Ahmad
Ultrasound Trainer PAFP (Regd)

History

Patients name is Erum, aged 20 yrs
Complains of
1. Pale eyes since 2 yrs
2. Pain right upper abdomen since 2y
3. Fatigue 2y
4. Dark urine 2y
5. Intense itching 1y

Investigations

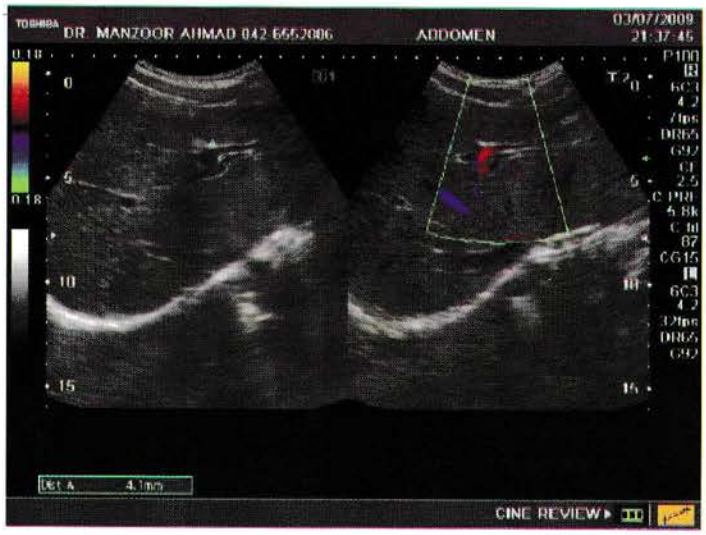
- Blood CBC
- Haemoglobin 9.8G/dl
- ESR 87mm 1st hour

Liver function tests

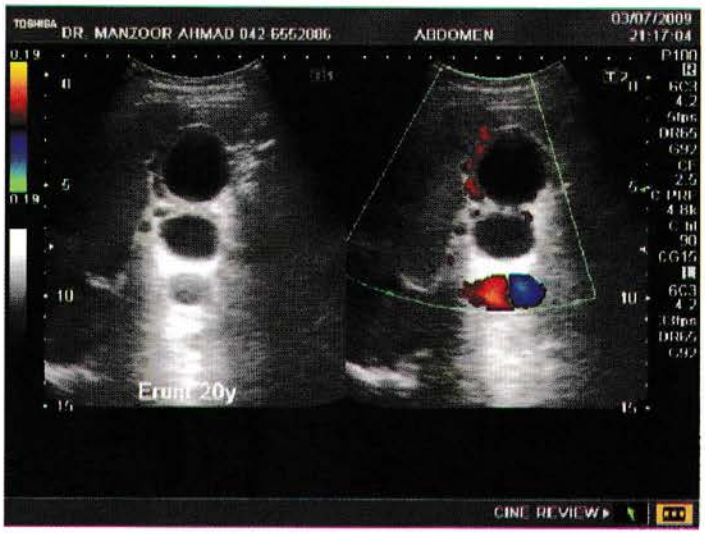
- Biluribin total 12mg/dl
- Cojugated biluribin 9.6 mg/dl
- ALT 54 U/L
- AST 74 U/L
- Alkaline phosphatase 220 U/L
- Malarial parasites negative
- HBSag negative
- Anti HCV negative

Ultrasound Examination

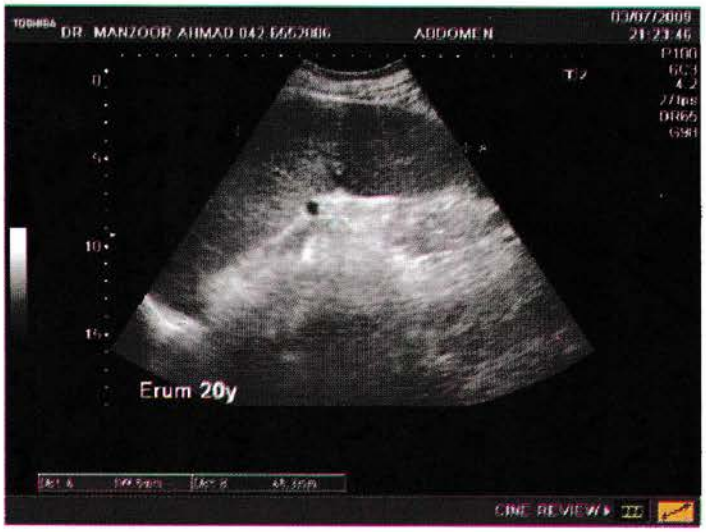
- Liver is normal in size and texture with no focal defect.
- Focally dilated IHB Channels are visible.
- IHBC show smooth narrowing toward porta hepatis. CHD is extermely narrow.
- CBD is 8mm in diameter.
- Extensive collaterization around thick walled gall bladder is visible
- Spleen is enlarged and its length is 20cm and splenic index is 140.
- RPV is 13mm. Portal vein division at porta hepatis appears to be narrowed resulting in opening of collateral channels
- Most likely some gradual fibrosing pocess involving porta hepatis which is producing narrowing of CHD and PV bifurcation thus resulting in obstructive jaundice and portal hypertension.
- Dilated hepatic ducts, which is confirmed by its avascularity on Doppler, and it narrows towards porta hepatis.



Dilated intrahepatic billiary channels

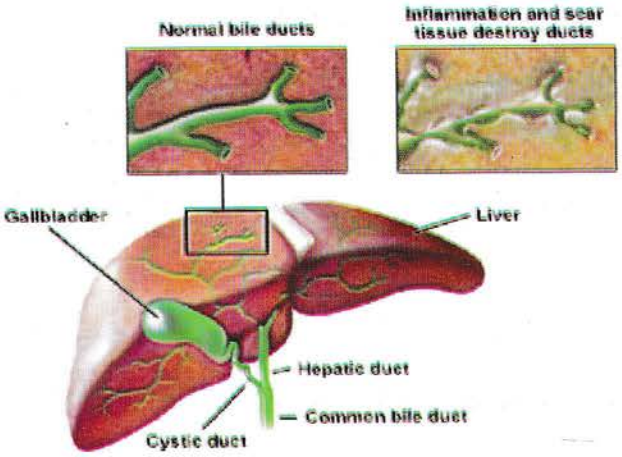


Thick walled gall bladder with vascular collaterals i.e varices of GB walls, due to portal hypertension.



Splenomegally, another sonographic evidence of portal hypertension. Length of spleen is 20 cm (NV 12 cm).

Primary sclerosing cholangitis (PSC)



- **Primary sclerosing cholangitis (PSC)** is a chronic liver disease caused by progressive inflammation and scarring of the bile ducts of the liver.
- The inflammation impedes the flow of bile to the gut, which can ultimately lead to liver cirrhosis and liver failure.
- The underlying cause of the inflammation is believed to be autoimmunity.
- PSC is often complicated by recurrent episodes of bacterial cholangitis.
- Patients with PSC also have an increased risk of cholangiocarcinoma.
- The definitive treatment is liver transplantation.
- Diagrammatic representation of normal hepatobilliary system, along with the abnormal appearance of bile ducts in PSC, where these are inflamed and scarred.

Pathophysiology

- Inflammation damages bile ducts, both intra and extra hepatic. The resulting scarring of the bile ducts blocks the flow of bile, causing cholestasis.
- Bile stasis and back-pressure induces proliferation of epithelial cells and focal destruction of the liver parenchyma, forming bile lakes.
- Chronic biliary obstruction causes portal tract fibrosis and ultimately biliary cirrhosis and liver failure.
- Bile assists in the enteric breakdown and absorption of fat; the absence of bile leads to fat malabsorption and deficiencies of fat-soluble vitamins (A, D, E, K).

Management

- Standard treatment includes ursodiol, a bile acid naturally produced by the liver, which has been shown to lower elevated liver enzyme numbers in people with PSC, but has not yet been proven effective at prolonging the life of the liver.
- Treatment also includes medication to relieve itching (antipruritics) and bile acid sequestrants (cholestyramine), antibiotics to treat infections, and vitamin supplements, as people with PSC are often deficient in vitamin A, vitamin D, vitamin E and vitamin K.
- In some cases, ERCP, which may involve stenting of the common bile duct, may be necessary in order to open major blockages (dominant strictures).
- Liver transplantation is the only proven long-term treatment of PSC. Indications for transplantation include recurrent bacterial cholangitis, jaundice refractory to medical and endoscopic treatment, decompensated cirrhosis and complications of portal hypertension.