Case No. 3 (January 2004)

Male – age: 56

Medical History:

- Chronic ulcerative colitis (CUC) for years, managed by medical therapy
- Long enduring, frequent episodes of moderate hyperbilirubinemia prompting preliminary diagnosis of Gilbert's disease
- Hospitalization for worsening of jaundice and fever

Diagnostic procedures:

- Ultrasonography
- ERCP
- CT

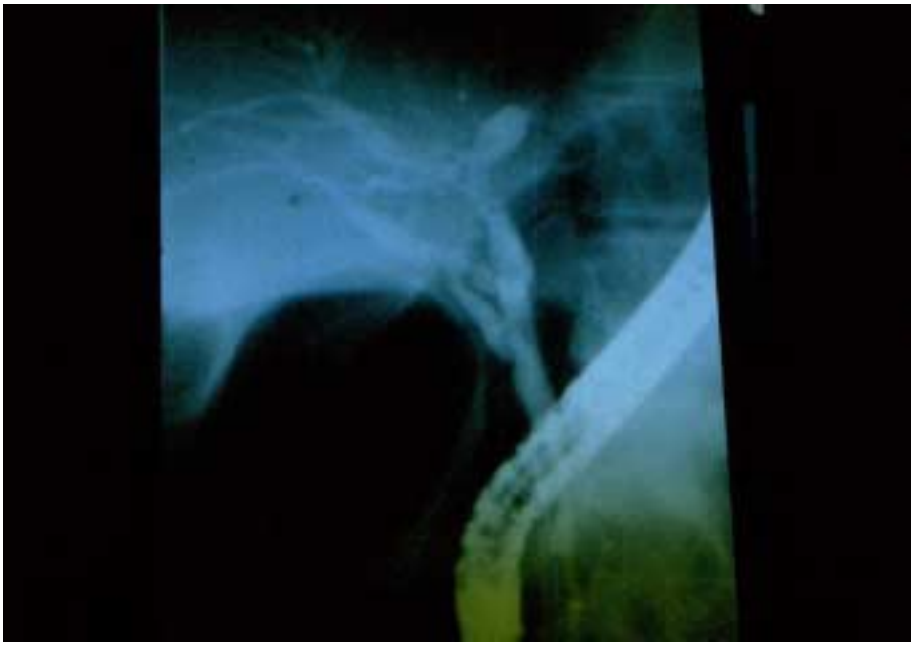


Fig. 1 – ERCP

Stones in the common bile duct. Dilatation of the left biliary system. Triple biliary confluence (2 ducts on the right; 1 on the left)

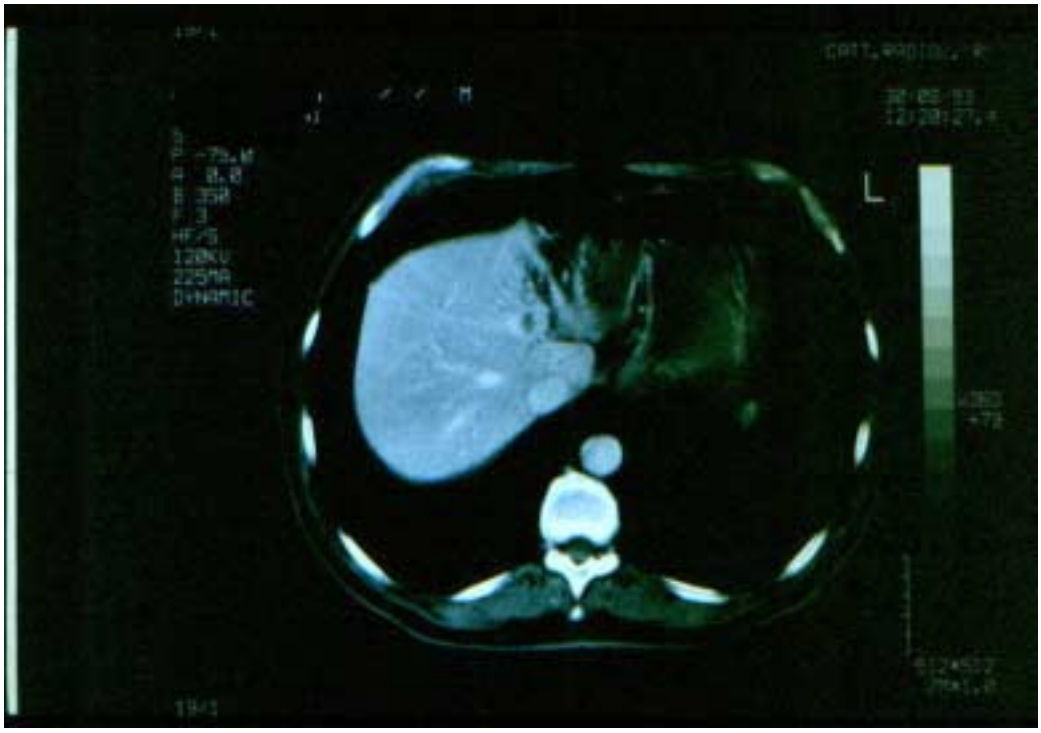


Fig. 2 – CT

Atrophy of the left liver (S2,S3,S4). The vasculo-biliary skeleton without the parenchymal component is visible.

Diagnosis:

- Lithiasis of the common bile duct
- Primitive dilatation of the left biliary intrahepatic system (localized Caroli's disease)
- Atrophy of the left liver (S2,S3,S4)

Therapeutic treatment:

- Endoscopic clearance (ERCP) of the common bile duct
- left hepatectomy (trisegmentectomy - S2,S3,S4)

Surgery:

Right S-shaped, prolonged to the left, subcostal laparotomy. The left liver (S2,S3,S4) is atrophic, i.e., smaller than normal, pink and pale. Dilated biliary ducts are visible through Glisson's capsule (Fig. 3).

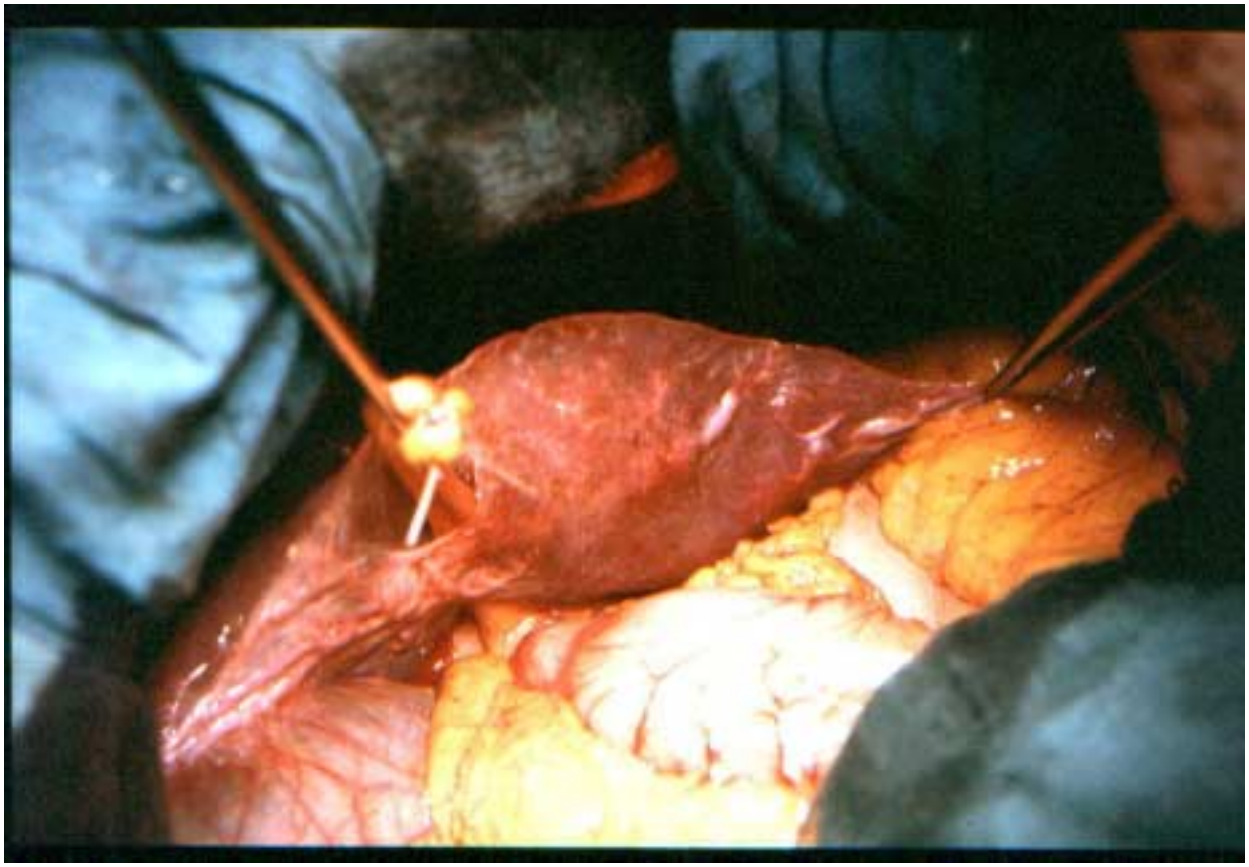


Fig. 3 – Left hepatic lobe – Dilated biliary ducts are visible through Glisson's capsule. Compared to the right lobe, the left lobe is pale and pink.

Cholangiography confirms ERCP data. The gallbladder is removed. Mobilization of the left lobe, dividing the appropriate ligamentous attachments, i.e., Teres, falciform and left triangular ligaments.

Dissection of the triangle of attack and preparation of the caval plane and hepatic veins.

Ultrasonography to control the median hepatic vein.

Dissection of the porta hepatis and identification of the hilar elements. Triple biliary confluence is confirmed. The left branches of the hepatic artery and the portal vein are ligated and transected, respectively.

Pringle's maneuver.

Incision of Glisson's capsule along the surgical plane, about 1 cm to the left of Castle's great scissure to safeguard the median hepatic vein.

Parenchymal incision by Kellyclasia. Exposure of ducts and vessels, which are individually ligated and transected. Finally, double ligation and transection of the left hepatic vein.

Left hepatectomy

Control of the surface of section (Fig. 4).

Suture without drainage.

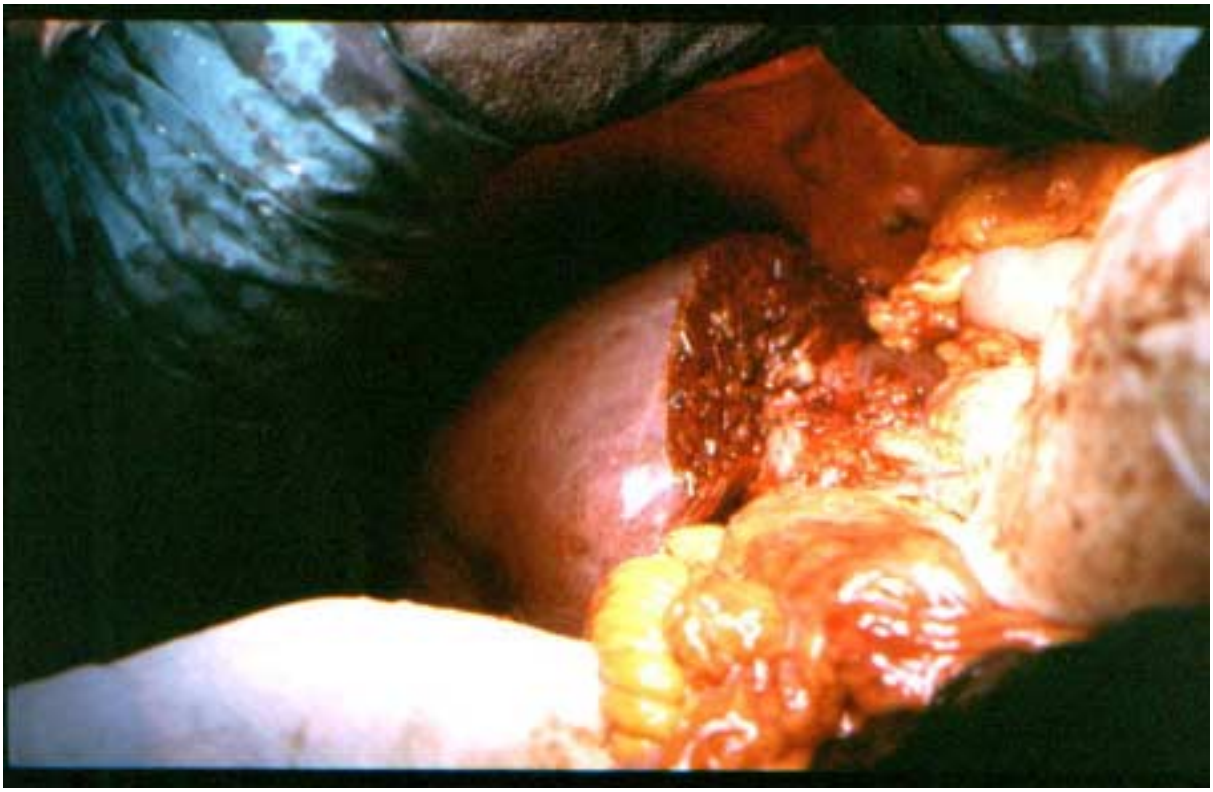


Fig. 4 –Residual right liver - Surface of section

Pathology

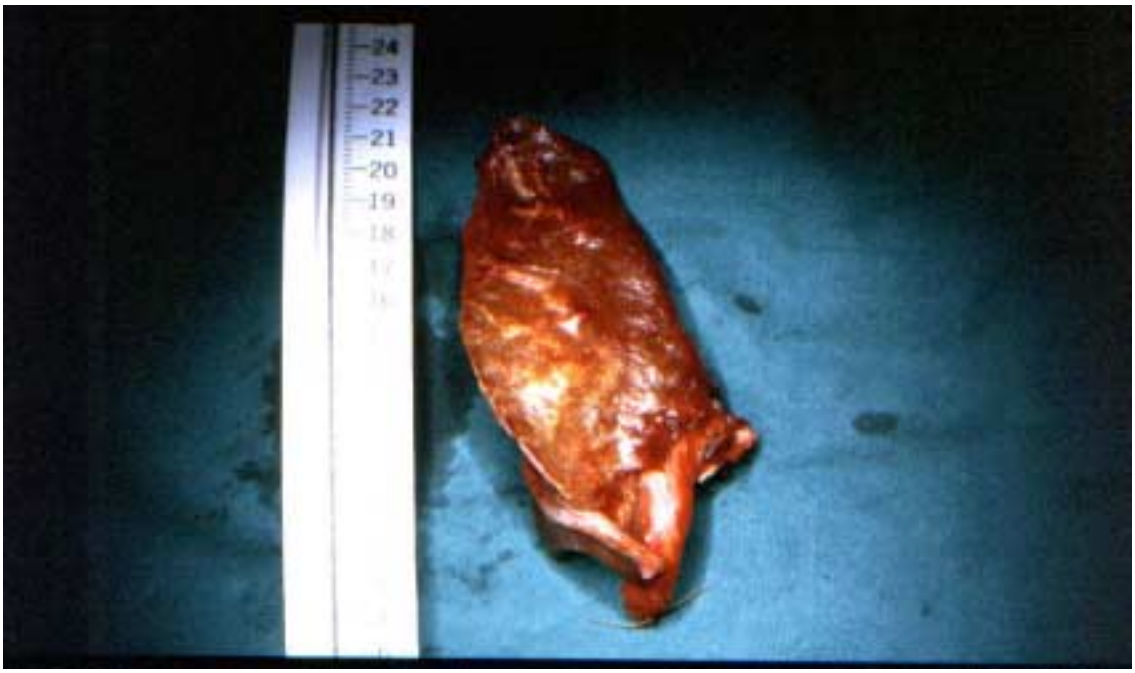
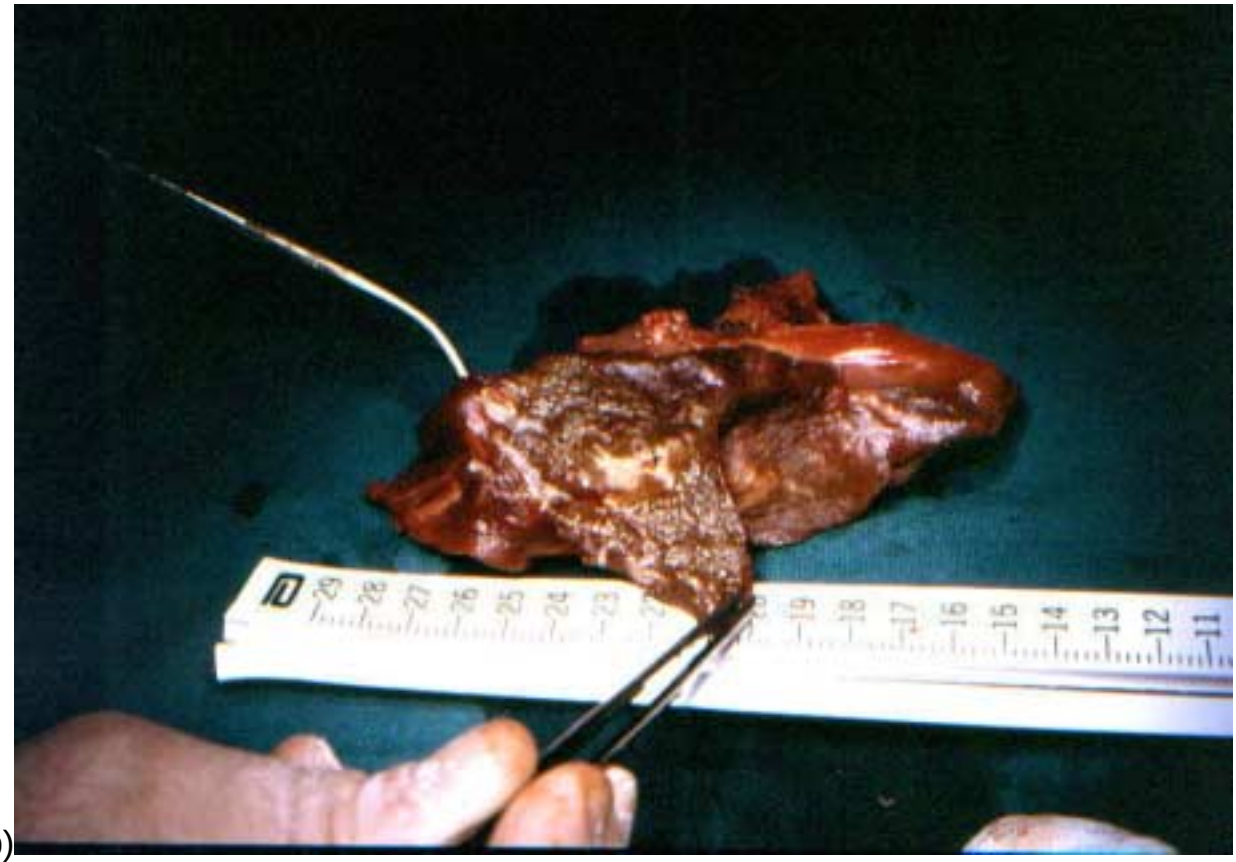
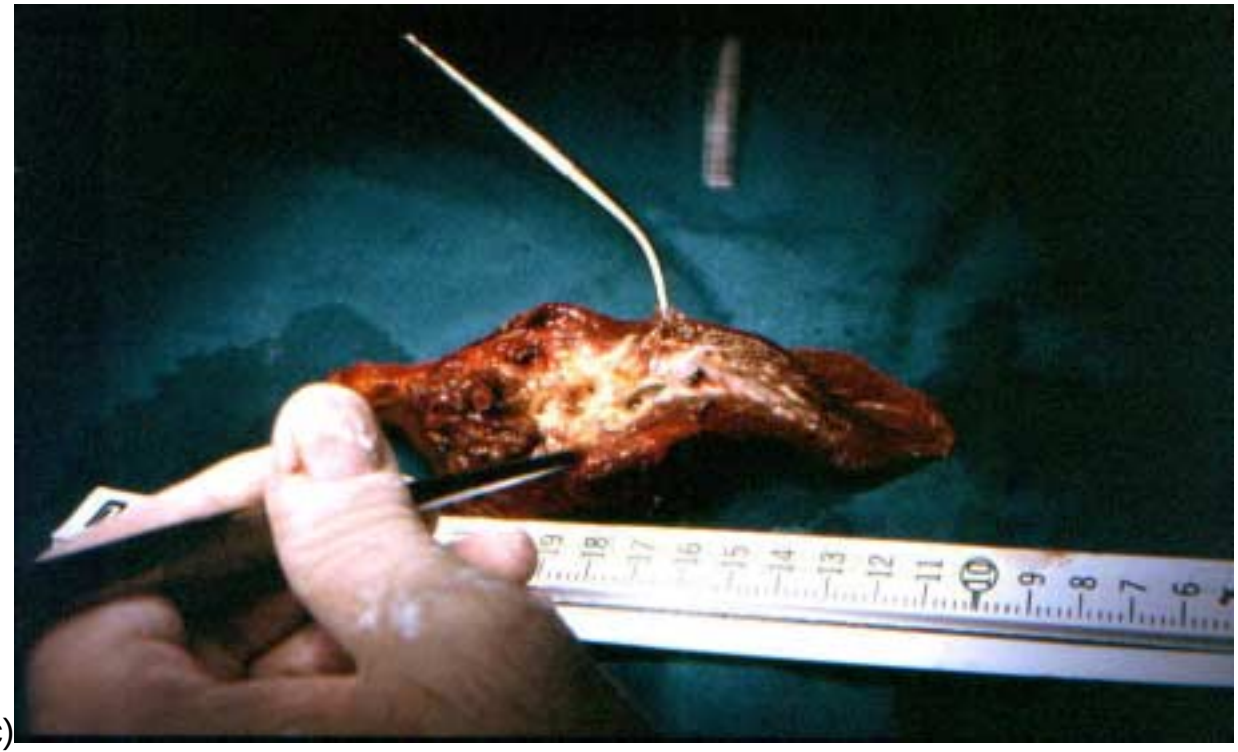
a) 

Fig. 5 – a) specimen of left liver

b) 



c)

Fig. 5 – a) specimen of left liver; b-c) atrophic parenchyma, ductal cystic dilatation and microlithiasis.

Histopathological findings:

- parenchymal atrophy
- the duct wall is fibrotic with varying degrees of acute and chronic inflammation. Epithelium is focally hyperplastic with ulcerated areas.

Follow-up

After seven years of relatively good health:

- obstructive jaundice (total bilirubinemia mg 12.4 – direct 10.7);
- Ultrasonography: moderate dilatation of intrahepatic biliary tract – stones in S8 and S6;
- ERCP: inserting of transhepatoduodenal drainage into the confluence of S7-S8;
- Cholangiography via PTC: intrahepatic lithiasis with stenosis of the confluence of the draining ducts S6,S7,S8;
- cleaning b.i.d. via PTC by physiologic solution;
- removal of stones via PTC.

Diagnosis

- Sclerosing cholangitis?
- Intrahepatic lithiasis
- Previous left hepatectomy
- Ulcerative colitis

Outcome

- Post-operative control: negative for both stenosis and intraductal lithiasis
- Bilirubinemia: progressive decrease to normality
- Home therapy: PPI, steroids, 5-ASA, tauroursodeoxycholic acid

Follow-up

After three years (ten years after the onset of intrahepatic biliary disease): no residual or subsequent pathological evidence.

Remarks

- The finding in an adult of dilatation of the left biliary system with cystic configuration and micro-lithiasis suggests the diagnosis of localized Caroli's disease-*simple type*, to be distinguished from the syndrome of the same name, which is characterized by congenital malformations, e.g., hepatic fibrosis.
- The condition of parenchymal atrophy is not often discussed in the literature.
- The association with CUC is interesting for pathogenetic reasons.
- The subsequent possible sclerosing cholangitis (primary? secondary?), for its reported association with CUC and for its putative common immunological bases, raises the suspicion of a pathogenetic correlation between Caroli's disease and these two disorders. The literature, however, does not seem to have paid much attention to this supposition.
- The seemingly propitious and enduring outcome appears, nevertheless, to contradict the above-mentioned hypothesis.

This case-report is being published to pique comparison and discussion. Peers and experts are invited to share their views.