

## Hypoplasia of the right hepatic lobe and portal hypertension A forty years follow-up

R. Willocx, B. Van Beers

Departments of Gastro-Enterology and Radiology, Cliniques Universitaires St Luc, Brussels, Belgium.

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In 1960, hepatosplenomegaly was discovered in five-year old girl. Diagnosis of juvenile cirrhosis was put forward by an eminent french consultant. To confirm the diagnosis, laparoscopy and splenoportography were done in 1962 ; normal bulging liver without edge, non-visualised gallbladder and enlarged spleen were noticed. The splenoportography showed a portal vein rising to the upper right diaphragm and irrigating the liver down below. Splenic pressure was 17.6 mm Hg and hepatic veins gradient was 4 mm Hg. Oral cholecystography revealed a supra-hepatic gallbladder. The only biological abnormality was a low platelet count.

In 1964, while in Brussels for a G.I. meeting, Prof. Dame Sheila Sherlock kindly agreed to examine the child and, thereafter, discussed with Dr Hans Popper on an eventual preventive spleno-renal shunt. Dr Popper requested the opinion of Dr Michael De Bakey who confirmed the general opinion to delay surgery until further sympatomatology.

In november 1964, the child was admitted at the Royal Free Hospital (London, U.K.) under the care of

Prof. Dame Sheila Sherlock : liver biopsy showed a normal histology and selective celiac arteriography a hepatic artery arising from the celiac axis and, almost immediately, dividing in the right and the left hepatic arteries, a patent portal vein, a patent splenic vein and an enlarged spleen. Platelet count was 57.000.

Years went on : liver became less and less palpable, splenomegaly did not change, platelet count oscillated between 110.000 and 140.000/mm<sup>3</sup>. Growth and menarche were without problem. The patient became more and more reluctant for further investigation. In 1991, oesogastrosocopy did not show any sign of portal hypertension.

At the end, in 2001, she underwent a magnetic resonance imaging examination, which confirmed the presumed diagnosis. Right liver hypoplasia and splenomegaly were seen on the transverse T1-weighted gadolinium enhanced gradient – echo image (Fig. 1a). Sagittal gradient – echo image showed subdiaphragmatic gallbladder and hypoplasia of the right liver lobe located above the right kidney (Fig. 1b).

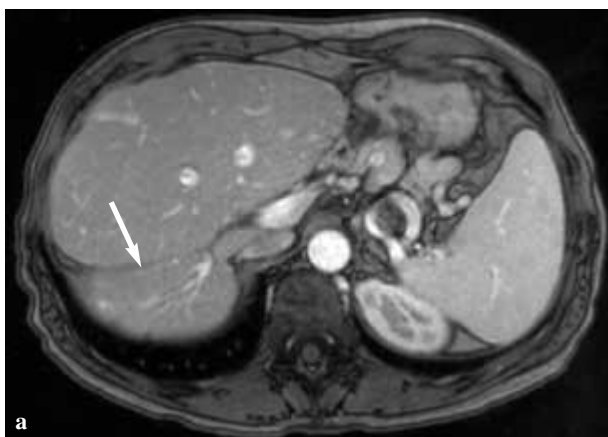


Fig. 1. — Transverse T1-weighted, gadolinium-enhanced, gradient-echo image (Fig. 1a) shows hypoplasia of right liver (arrow) and splenomegaly. Sagittal gradient-echo image (Fig. 1b) shows subdiaphragmatic gallbladder (long arrow) and hypoplasia of right hepatic lobe (short arrow) located above right kidney.

