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LIVER FUNCTION IN A NATIONAL COHORT OF BILIARY ATRESIA PATIENTS SURVIVING WITH NATIVE LIVER

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Aims: We assessed liver function and portal hypertension in a national cohort of biliary atresia (BA) patients surviving with native liver to guide follow-up and transition of care.

Methods: All BA patients surviving with native liver, who were born in the era of pediatric liver transplantation (LTX) since 1987 in one country, where BA treatment was nationally centralized in 2005 were included. Liver biochemistries and abdominal ultrasound (US) were performed in all, and gastroscopy, elastography and liver biopsy in patients aged ≥1 year (n=25). Histological liver fibrosis was graded according to Metavir staging (F0-F4).

Results: The 28 native liver survivors represented 37% of all BA patients treated during the study period, and 63% after centralization. 86% had type 3 BA, while median PE age was 58 (IQR, 32-82) days and mean follow-up age 8.4 (range, 0.5-25) years. Median bilirubin, alanine aminotransferase, prealbumin, prothrombin ratio and platelets were in reference range (**table**), and all of them normal in 32%. In US, fourteen patients (50%) had splenomegaly, whereas none had ascites. Eleven (39%) had received prophylactic sclerotherapy for esophageal varices. Mesenterico-systemic shunt was performed in one case for massive bleeding after band ligation of gastric varices. In liver biopsies at 7.3±4.6 years 92% had fibrosis, while 56% had only mild or moderate fibrosis (Metavir ≤2).Liver stiffness was increased in 78%. Follow-up age was unrelated to liver fibrosis stage (R=-0.19, P=0.35) or stiffness (r=-0.09, P=0.67), but correlated inversely with platelets (R=-0.40, P=0.039). Although patients with splenomegaly tended to be older (10.2±6.2 vs 6.6±5.2 years, p=0.098), cumulative occurrence of varices was not related to follow-up age (8.8±4.1 vs 9.7±6.5, P=0.96).

Conclusions: Liver fibrosis and complications of portal hypertension including varices, splenomegaly and hypersplenism represented the major manifestation of liver disease in native liver survivors, while biochemistries were well preserved, being normal in 32%.

Table. Liver biochemistries, stiffness and histological fibrosis in native liver survivors

	Bilirubin (µmol/l)	ALT (U/l)	Prealbumin mg/l	Prothrombin (%)	Platelets E9/l	Stiffness (kPa)	Metavir (F0-F4)
Mean	14	48	147	97	159	17.7	2.4
Median	9.5	38	147	93	133	12.7	2.0
IQR	4-16	23-51	110-174	76-117	86-235	6.5-26.6	2.0-4.0
Range	2-56	10-141	70-230	55-169	44-357	3.3-60.9	0-4.0
Reference	<20	<40	>130	>70	>150	≤6	0
Normal (%)	86%	61%	71%	82%	46%	22%	8%