Acute liver failure and experience with therapy using the molecular absorbent recirculation system (MARS)

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Introduction and Objective: Acute liver failure (ALF) is a cause of urgent liver transplantation (LT). The molecular absorbent recirculation system (MARS) is an extracorporeal liver replacement device, considered bridging therapy for LT.

Case report: 24-year-old man with no relevant history was admitted due to asthenia, adynamic, hyporexia, jaundice, oral intolerance and transaminasemia. His laboratory studies are shown in Table 1 and include positivity for IgM hepatitis A (HAV). Other causes are excluded. Development of ALF with two minor criteria (King's College), received N-acetylcysteine, without response, management with MARS/PRISMA is started, one session (initial dialysate 1300 mL, increasing to 1800 mL, maintaining a flow of 150 mL/L, and eight bottles of 25% albumin (400 mL)). His evolution towards neurological, hepatic, and renal improvement. Discharged for improvement.

Discussion: MARS therapy is based on removing molecules, including medium-sized ones, especially those that are binding by albumin and, therefore, cannot be purified conventionally. The relative simplicity, the good tolerance and the results obtained so far make MARS the most promising alternative. There is some experience with the use of MARS in ALF due to HAV.

Conclusion: MARS therapy is useful in the management of patients with ALF due to HAV; its use has shown positive results impacting patient survival and even, in some cases, avoids liver transplantation. The number of sessions will depend on the clinical response.

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Table 1 Laboratory values before and after the use of MARS therapy.

	Before	After
Hemoglobin (g/L)	17.90	13
Leukocytes (cell/10^3)	10.4	6.10
Platelets (cell/10^3)	213	207
Glucose (mg/dL)	86	71
Creatinine (mg/dL)	3.2	1.93
Total bilirrubin (mg/dL)	16.1	10
Direct bilirrubin (mg/dL)	14	5.8
ALT (U/L)	2409	824
AST (U/L)	772	257
ALP (U/L)	110.5	77
GGT (U/L)	501	136
ALB (g/dL)	3.90	2.8
INR	3.4	1.1
PT (Sec)	36.9	21.8
aPPT (Sec)	45	31
	Hepatitis A virus IgM	

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Congenital hepatic fibrosis as a rare cause of non-cirrhotic portal hypertension

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Introduction and Objective: a case of congenital hepatic fibrosis is presented

Case report: A 19-year-old female with a medical story relevant only to epistaxis of 3 years long was referred to the liver clinic because of thrombocytopenia and transaminasemia. She denied data of decompensated advanced chronic liver disease and hepatic transition elastography was performed: S0 224, F4 (19.9 kPa). Portal Doppler ultrasound was performed: a diffuse liver disease with hepatomegaly, signs of portal hypertension, splenomegaly and ascites, and probable thrombosis of the distal splenic vein. Given the suspicion of hereditary thrombophilia, a genetic profile was requested (negative Leiden Factor V PCR, negative JAK 2 PCR, negative lupus anticoagulant, normal antithrombin III, normal protein C, normal protein S). Abdominal-pelvic angiotomography was performed: enlarged liver with no focal lesions, no dilatation of the bile duct, adequate permeability of the portal venous system, and enlarged spleen. The rest of the antibodies and tests for congenital metabolic disorders were requested (normal ANAS, normal ASMAs, normal Anti LKM1, normal AMA, normal IgG, normal ceruloplasmin, normal urine copper, low ferritin, normal transferrin). Active infection by hepatitis B, C and HIV viruses was ruled out. During follow-up, the patient developed variceal gastrointestinal bleeding, endoscopic variceal ligation was performed and management with a non-selective beta-blocker was initiated. A percutaneous liver biopsy was performed, reporting in histopathology: morphological changes consistent with malformation of the ductal plate of a congenital hepatic fibrosis type.

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Sclerosing cholangitis associated with IgG4 disease. Case Report

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Introduction and Objectives: This study aimed to present the case of a patient with sclerosing cholangitis and autoimmune pancreatitis associated with IgG4 disease.

Materials and Methods: A 44-year-old male with a history of allergic rhinitis, diabetes, and dyslipidemia. His clinical picture began with diffuse abdominal pain and jaundice; on physical examination, jaundice, a painful abdomen in the right upper quadrant and hepatomegaly. Liver biochemistry with transaminasemia and cholestasis. In auxiliary studies US with bile duct dilatation of 12.4 mm, ColangioMR was performed, reporting concentric wall thickening in T2 of the intra and extrahepatic bile duct that condition focal stenoses of the right hepatic duct and intrahepatic bile duct approximately 16 mm from the ampulla, conditioning segmental dilatations of the common hepatic and extrapancreatic bile duct up to 8.9mm. According to these findings, IgG4 serum levels were requested, reporting 1200mg/dl. Steroid treatment was started, presenting a favorable response.

Discussion: IG-G4-related disease is an autoimmune relapsing chronic multiorgan fibroinflammatory syndrome. Its maximum incidence is in Japan, with 336 to 1300 patients diagnosed/year; the estimated prevalence is 62/million subjects between 50 and 70 years of age. The main and most commonly affected organs are the pancreas, bile duct, salivary and lacrimal glands, retroperitoneum and lymph nodes.