CVID under treatment at a primary immunodeficiency clinic of a tertiary-level hospital.

Patients / Materials and Methods: A retrospective and descriptive study examined the medical records of patients with confirmed CVID.

Results and Discussion: out of the eleven patients with CVID, eight were women, and the median age was 34 years (range 23-72). PH was suspected in five (45.4%), with three patients experiencing clinically significant PH and one case complicated by variceal bleeding. Table 1 compares both groups (with and without PH). Thrombocytopenia was found in most patients, consistent with the higher incidence of splenomegaly. Liver biopsies performed only in two patients with suspected PH excluded cirrhosis but identified regenerative nodular hyperplasia in one case. Both cases had liver stiffness measurements by shear wave elastography, showing a median of 14.2 kPa. No association was identified with other non-infectious complications of CVID (gastrointestinal and pulmonary disease).

Conclusions: Liver disease is often underdiagnosed in patients with CVID, with portal hypertension appearing to be frequent. Early screening is essential to avoid severe complications.

	With portal hypertension (n=5)	Without portal hypertension (n=6)
Years from CVID diagnosis	11,4 (7-14)	7,6 (4-17)
Female	4	4
Age at PH diagnosis	45,6 (22-70)	-
Pulmonary disease	4	4
Gastrointestinal disease	1	1
Cholestasis	2	0
Splenomegaly	4	0
Thrombocytopenia	4	0
Gastro-esophageal varices/Upper bleeding	2 (3 no data)/1	0
Ascites	1	0

Table: comparison between patients with and without evidence of portal hypertension.

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P-77 TRIPLE THERAPY FOR DIFFICULT-TO-TREAT PRIMARY BILIARY CHOLANGITIS: A SYSTEMATIC REVIEW AND SINGLE-ARM META-ANALYSIS

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Conflict of interest: No

Introduction and Objectives: High-risk patients with primary biliary cholangitis (PBC) who respond incompletely to ursodeoxycholic acid (UDCA) require additional treatment with fibrates or obeticholic acid (OCA). Despite this, 30-50% of these patients continue to exhibit elevated alkaline phosphatase (ALP) and bilirubin levels, classifying them as difficult-to-treat PBC. This study aims to evaluate the effects of triple therapy (UDCA + OCA + fibrates) on liver biochemistry in patients with difficult-to-treat PBC.

Patients / Materials and Methods: We systematically reviewed EMBASE, PubMed, and Cochrane databases to identify eligible studies. Pooled analyses were performed for change-from-baseline data. We also conducted subgroup analyses based on the sequencing of the specific add-on drug used as third-line therapy. Statistical analyses were performed using RStudio (2023.12.1+402).

Results and Discussion: Two studies provided change-from-baseline data, encompassing 95 patients under triple therapy, of whom 68.4% (n=65) had fibrates added to UDCA+OCA dual therapy. Overall, patients under triple therapy presented with decreased ALP [-0.82 x upper limit of normal (ULN), 95%CI -0.96 to -0.68], bilirubin (-0.06 x ULN; 95%CI -0.11 to -0.01), and GGT (-3.18 x ULN; 95%CI -4.57 to -1.79) levels compared to the last available result on dual therapy. No significant change was noted for AST (-0.08 x ULN; 95%CI -0.44 to 0.28) and ALT (-0.21 x ULN; 95%CI -0.61 to 0.20) concentrations. However, the addition of OCA to UDCA+fibrates dual therapy significantly reduced AST (-0.53 x ULN; 95%CI -0.73 to -0.33; p-value for subgroup differences < 0.001) and ALT (-0.69 x ULN; -0.97 to -0.40; p<0.001) levels. On the other hand, adding fibrates to the UDCA+OCA scheme was superior in reducing ALP levels (p=0.049).

Conclusions: Triple therapy appears to reduce liver enzyme levels in patients with difficult-to-treat PBC. Further studies are warranted to clarify the optimal sequencing and to identify the subgroups that benefit the most from this combination therapy.

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P-78 DECOMPENSATED CIRRHOSIS IN A LARGE MULTINATIONAL COHORT IN LATIN AMERICA: MORTALITY IS TOO HIGH IN THE REGION REGARDLESS OF ETIOLOGY AND COUNTRY

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