

## Supplementary material

**Supplementary Table 1** Diagnostic criteria for AIH-PBC variant (adapted from [75])

PBC criteria*
ALP > 2×ULN or γ-GT > 5×ULN
AMA ≥ 1:40
Liver biopsy showing florid bile duct lesions
AIH criteria*
ALT > 5×ULN
IgG > 2×ULN or a positive test for SMA
Liver biopsy showing moderate or severe periportal or periseptal lymphocytic piecemeal necrosis

\*At least 2 of the 3 criteria should be satisfied for each disease  
*PBC, primary biliary cholangitis; AIH, autoimmune hepatitis; ALP, alkaline phosphatase; ULN, upper limit of normal; AMA, antimitochondrial antibodies; ALT, alanine aminotransferase; IgG, immunoglobulin G; SMA, smooth muscle antibodies*

**Supplementary Table 2** New diagnostic score for AIH-PBC variant diagnosis (adapted from [76])

Component	Result	Score
<b>Biochemical category</b>		
AST or ALT above ULN	>2	+3
	1.5-2	+2
	1-1.5	+1
	<1	0
ALP above ULN	>1	+2
	0.75-1	+1
	<0.75	0
Serum globulin above ULN	>1.5	+2
	1-1.5	+1
	<1	0
<b>Immunologic category</b>		
ANA, SMA, or LKM1	>1:80	+3
	1:80	+2
	1:40	+1
	<1:40	0
<b>Or</b>		
anti-SLA/LP, pANCA	Positive	+2
AMA	Positive	+3
<b>Histologic category</b>		
	Interface hepatitis	+3
	Lymphoplasmacytic	
	Hepatic rosettes	+1
	Granulomas	+3
	Florid ductal lesion	+1
	Ductular proliferation	+1
	Bile duct loss	+1
<b>Others category</b>		
Viral markers	Positive	-3
	Negative	+3
Drugs	Yes	-4
	No	+1
Alcohol	<25 g/day	+2
	>60 g/day	-2
Interpretation of score	Definitive	≥21
	Probable	19 or 20
	Rejected	<19

*AIH, autoimmune hepatitis; PBC, primary biliary cholangitis; AST, aspartate aminotransferase; ALT, alanine aminotransferase; ALP, alkaline phosphatase; ULN, upper limit of normal; ANA, antinuclear antibodies; SMA, smooth muscle antibodies; anti-LKM1, live kidney microsomal type 1 antibodies; anti-SLA/LP, antibodies against soluble liver antigens/liver pancreas; pANCA, perinuclear anti-neutrophil cytoplasmic antibodies; AMA, antimitochondrial antibodies*

**Supplementary Table 3** Simplified criteria for the diagnosis of autoimmune hepatitis (adapted from [70])

Parameter	Finding	Points
ANA or SMA positive	≥ 1:40	+1
ANA or SMA positive or anti-LKM positive or anti-SLA/LP positive	≥ 1:80 ≥ 1:40 Positive	+2*
Liver histology (presence of hepatitis is necessary)	Typical AIH** Compatible with AIH** Atypical**	+2 +1 0
Serum IgG levels	> ULN > 1.1 x ULN	+1 +2
Absence of viral hepatitis***	Yes No	+2 0
Sum		≥ 6: Probable AIH ≥ 7: Definite AIH

\*Addition of points achieved for all autoantibodies (maximum, 2 points).

\*\*Definition of typical lesions as in ref. 74; Compatible liver histology: chronic hepatitis with lymphocytic infiltration without all the features considered typical; Atypical: histological lesions supporting another diagnosis.

\*\*\*In chronic cases absence of hepatitis B and C viral markers; in acute cases absence of serological markers of acute hepatitis A, B, C, D and E is needed. ANA or SMA detection refers to the use of immunofluorescence assay on rodent tissues, not ELISA

ANA, antinuclear antibodies; SMA, smooth muscle antibodies; anti-LKM1, live kidney microsomal type 1 antibodies; anti-SLA/LP, antibodies against soluble liver antigens/liver pancreas; AIH, autoimmune hepatitis; IgG, immunoglobulin G; ULN, upper limit of normal; ELISA, enzyme-linked immunosorbent assay

**Supplementary Table 4** Proposed scoring criteria for the diagnosis of AIH and AIH-PSC variant in the pediatric population (adapted from [83])

Characteristic	Cutoff	AIH points	AIH-PSC points
ANA and/or SMA*	≥1:20**	1	1
	≥1:80	2	2
Anti-LKM1 <sup>+</sup> or	≥1:10**	1	1
	≥1:80	2	1
Anti-LC1	Positive**	2	1
Anti-SLA/LP	Positive**	2	2
pANNA	Positive	1	2
IgG	>ULN	1	1
	>1.2 ULN	2	2
Liver histology	Compatible	1	1
	Typical	2	2
Absence of viral hepatitis (A, B, E, EBV), MASH, Wilson, and drug exposure	Yes	2	2
Extrahepatic autoimmunity	Yes	1	1
Family history of autoimmune disease	Yes	1	1
Cholangiography	Normal	2	-2
	Abnormal	-2	2

Score ≥7 = Probable AIH; ≥8: Definite AIH. Score ≥7: Probable AIH-PSC; ≥8: Definite AIH-PSC.

\*Antibodies measured by immunofluorescence assay on a composite rodent substrate (kidney, liver, stomach).

\*\*Addition of points achieved for ANA, SMA, anti-LKM-1, anti-LC-1, and anti-SLA/LP autoantibodies cannot exceed a maximum of 2 points

*AIH, autoimmune hepatitis; AIH-PSC, autoimmune hepatitis – primary sclerosing cholangitis; ANA, antinuclear antibody; SMA, smooth muscle antibodies; anti-LKM-1, anti-liver kidney microsomal antibody type 1; anti-LC-1, anti-liver cytosol type 1; anti-SLA/LP, anti-soluble liver antigen/liver pancreas; pANNA, peripheral antinuclear neutrophil antibodies; IgG, immunoglobulin G; ULN, upper limit of normal, EBV, Epstein-Barr virus; MASH, metabolic-dysfunction associated steatohepatitis*