

AIH Case Definitions*

AIH Case Definition

Variable	Points
ANA or SMA \geq 1:40	1
ANA or SMA \geq 1:80 OR LKM \geq 1:40 OR SLA Positive	2**
IgG > Upper normal limit	1
IgG > 1.10 times upper normal limit	2
Liver histology shows evidence of hepatitis AND COMPATIBLE with AIH	1
Liver histology shows evidence of hepatitis AND is TYPICAL AIH	2
Absence of viral markers	2

** Maximum addition of points for autoimmune markers ANA, SMA, LKM, or SLA is 2.

Definite AIH if score \geq 7
Probable AIH if score \geq 6

Possible AIH is defined as the following:

1. Elevated ALT or AST or histopathology indicating hepatitis,

ND

2. Absence of clearly defined and confirmed alternative diagnosis to AIH, such as viral hepatitis, alcoholic cirrhosis, etc., AND/OR absence of histology atypical for AIH.

(Note that AIH may occasionally occur in the presence of other hepatic disorders, such as

with overlapping syndromes. If AIH does occur with other liver diseases or as an overlap syndrome, then the other liver disease(s) should not be noted as definite/confirmed alternative to AIH. A defined/confirmed alternative diagnosis is one in which there is no evidence for AIH and clear evidence for another diagnosis).

Not AIH is defined as the presence of histology that is atypical for AIH (or inconsistent with AIH or consistent with another diagnosis) **OR** there is a clearly defined and confirmed alternative diagnosis.

Histology **consistent with** AIH if **ALL** of the following are found:

- Interface hepatitis, described as being periportal or periseptal hepatitis.
- Lymphoplasmacytic/lymphocytic infiltrate in portal tracts and extending into the lobule.
- Emperipolesis (active penetration by one cell into and through a larger cell).
- Formation of liver cell rosettes.

Histology **compatible with** AIH is a picture of chronic hepatitis with lymphocytic infiltration without all the features considered typical.

Histology **atypical** for AIH if showing signs of another diagnosis, such as steatohepatitis.

*Extracted from: EM Hennes, Zeniya M, Czaja AJ, et al. Simplified criteria for the diagnosis of autoimmune hepatitis. *Hepatology* 2008; 48: 169-176.

