

## **Second-line and Third-line therapy for Autoimmune Hepatitis**

*A position statement from the European Reference Network on Hepatological Diseases (ERN RARE-LIVER) and the International Autoimmune Hepatitis Group*

### **Lay summary for patients**

*Published July 2020 in Journal of Hepatology (doi: 10.1016/j.jhep.2020.07.023)*

Autoimmune hepatitis (AIH) is a chronic autoimmune disease of the liver. An autoimmune disease is a condition in which your immune system mistakenly attacks your body. The immune system of people with AIH mistakenly attacks the body's own liver cells causing damage and inflammation. To control inflammation in the liver and lower the immune system's activity, patients require immunosuppressive treatment.

#### 1<sup>st</sup> Line Treatment (Standard Treatment)

Standard treatment includes the use of steroids and azathioprine. At diagnosis, if the AIH is severe, steroids are needed because they control liver inflammation quickly. Azathioprine helps to reduce inflammation in the long term and taking it means that the steroid dosage can be reduced.

Most people with AIH take these medications for life and do very well. However, some patients need alternatives to 1<sup>st</sup> line treatment because they experience unwanted side effects (intolerance) or their liver blood test results do not improve enough (insufficient response). The European Reference Network on Hepatological Diseases (ERN RARE-LIVER) has now published recommendations on how to proceed in these situations (see Figure 1).

#### 2<sup>nd</sup> Line Treatment

For patients with side effects under standard treatment, mercaptopurine and mycophenolate mofetil are the drugs of choice for 2<sup>nd</sup> line treatment. For example, some patients taking azathioprine develop nausea, and they need to stop taking azathioprine. In this situation, most patients tolerate mercaptopurine or mycophenolate mofetil as an alternative to azathioprine and no longer experience nausea.

The aim of treatment in AIH is normalization of liver blood tests about 6 to 12 months after starting standard treatment. Sometimes this cannot be achieved and so doctors must first try to optimize the standard treatment. This means adjusting the dosage of azathioprine, and checking for other liver diseases that might also be causing the abnormal liver blood test results. A liver biopsy might be needed for this.

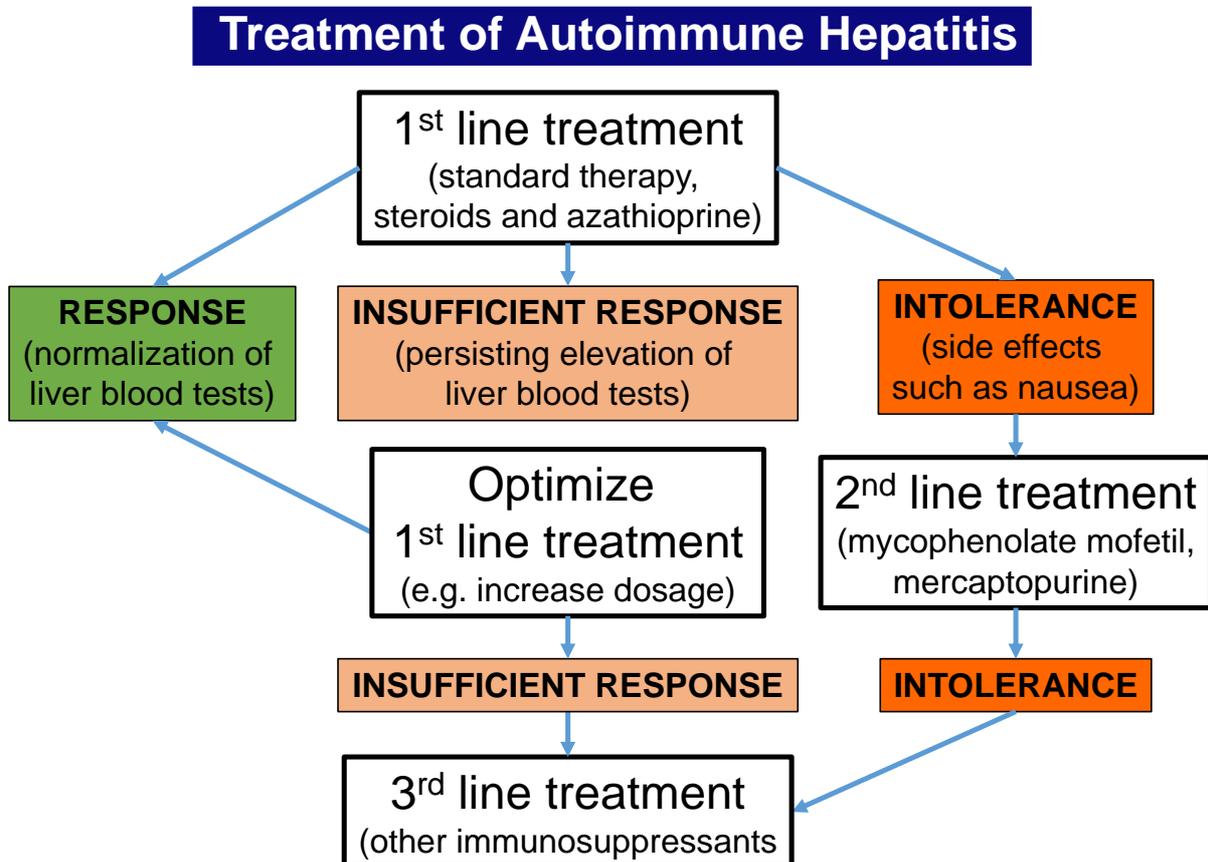
#### 3<sup>rd</sup> Line Treatment

If the liver blood tests still do not normalize, other immunosuppressants such as tacrolimus or infliximab are needed (3<sup>rd</sup> line treatment). The ERN RARE-LIVER has published expert guidance

on treatment protocols for these 3<sup>rd</sup> line treatments for AIH, because until now, there have not been any clear recommendations on the dosages required.

This publication on second-line and third-line AIH treatments will support doctors in managing their patients when standard treatments do not work. These new recommendations will help to standardize and improve the treatment of AIH patients across Europe. Standardizing treatment also allows doctors to start to understand which patients are likely to benefit most from the various treatment options available.

Figure 1: Treatment decisions for autoimmune hepatitis (AIH)



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