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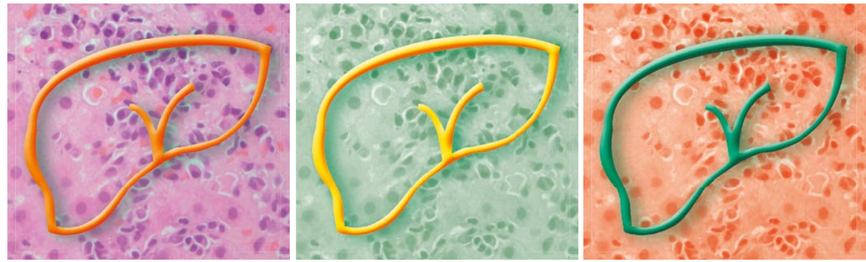
Treatment Algorithm Autoimmune Hepatitis



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Autoimmune Hepatitis: Treatment Algorithm Recommendations

Induction Therapy – Goal: Normalization of Transaminase and IgG Levels

Treatment with predniso(lo)ne

Start with **predniso(lo)ne** (0.5 – 1 mg/kg BW/day) followed by stepwise dose reduction. When transaminases decrease and bilirubin levels are below 6 mg/dl (100 µmol/L) after at least 2 weeks of **predniso(lo)ne** therapy

plus

Azathioprine 50 mg/day (patients without decompensated cirrhosis) followed by dose increase (up to 1 – 2 mg/kg BW/day) based on treatment response and potential side effects

Patients who are highly susceptible to the adverse effects of predniso(lo)ne, e.g. patients with diabetes, NAFLD/NASH, osteoporosis (e.g. post-menopausal), glaucoma, or patients who wish to avoid potential adverse cosmetic effects (acne, weight gain, Cushing syndrome, hirsutism)

Induction therapy with lower dose of **predniso(lo)ne** (30 mg/day)

plus

Azathioprine 50 mg/day for patients without decompensated cirrhosis and bilirubin < 6 mg/dl (100 µmol/L) followed by dose increase (up to 1 – 2 mg/kg BW/day) based on treatment response and potential side effects

Stepwise reduction of **predniso(lo)ne** according to the decrease of transaminases

For patients without cirrhosis:
induction therapy
with **budesonide** (3 x 3 mg/day)

plus

Azathioprine 50 mg/day followed by dose increase (up to 1 – 2 mg/kg BW/day) based on treatment response and potential side effects

Patients with azathioprine intolerance or contraindications (leukopenia, thrombocytopenia, cholestatic liver disease, gastrointestinal side effects, pancreatitis)

Induction therapy using **predniso(lo)ne** monotherapy (0.5 – 1 mg/kg BW/day) followed by stepwise dose reduction according to the decrease of transaminases

or

For patients without cirrhosis:
budesonide monotherapy (3 x 3 mg/day)

Maintenance Therapy

■ **Duration: usually lifelong**

■ **Treatment withdrawal: should only be attempted for patients with complete biochemical and immunological remission (normal transaminase and IgG levels) demonstrated multiple times within a period of at least 2 years who have also achieved histological remission (liver biopsy)**

Continue maintenance therapy
with **azathioprine** (1 – 2 mg/kg BW/day)

Possibly as a combination of
azathioprine (1 – 2 mg/kg BW/day)
with **predniso(lo)ne** (≤ 7.5 mg/day)

or

For patients without cirrhosis:
with **budesonide** (2 x 3 mg/day)

Continue maintenance therapy
with **azathioprine** (1 – 2 mg/kg BW/day)

Possibly as a combination of
azathioprine (1 – 2 mg/kg BW/day)
with **predniso(lo)ne** (≤ 7.5 mg/day)

or

For patients without cirrhosis:
with **budesonide** (2 x 3 mg/day)

Taper **budesonide** and
continue maintenance therapy
with **azathioprine** (1 – 2 mg/kg BW/day)

or

Continue combination therapy
with **budesonide** (2 x 3 mg/day)

plus

Azathioprine (1 – 2 mg/kg BW/day)

Continue maintenance therapy
with low-dose
predniso(lo)ne (≤ 7.5 mg/day)

or

For patients without cirrhosis:
with **budesonide** (2 x 3 mg/day)

or

Consider off-label treatment with
mycophenolate mofetil (2 x 1 g/day)

For patients with relapse on maintenance therapy

■ **Increase the dose or reinitiate predniso(lo)ne or budesonide therapy followed by stepwise dose reduction and possible increase of azathioprine dose (up to 2 mg/kg BW/day)**

For patients with inadequate response

■ **Check compliance, diagnosis, and for possible overlap with PBC or PSC**

■ **If inadequate compliance due to adverse effects of predniso(lo)ne therapy is suspected (patients without cirrhosis):**
Switch to budesonide (3 x 3 mg/day) and taper predniso(lo)ne

■ **For patients with inadequate response to budesonide-based therapy, switch to predniso(lo)ne (starting dose > 20 mg/day) and increase the azathioprine dose (up to 2 mg/kg BW/day). Stepwise reduction of predniso(lo)ne dose according to the decrease of transaminase levels**

■ **Second-line treatment options*:**
Mycophenolate mofetil, cyclosporine A or tacrolimus (off-label) as add on to or replacement of standard therapy in consultation with a hepatology center

■ **Third-line treatment options*:**
Rituximab or infliximab (off-label) as add on to or replacement of standard therapy in consultation with a hepatology center**

* Consideration of patient-specific factors such as comorbidities, tolerability, drug interactions/toxicity, and treatment response after initiation.

** Can induce AIH.

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