# Autoimmune pancreatitis: Management

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## INTRODUCTION

Autoimmune pancreatitis (AIP) is an uncommon but well-established form of pancreatic inflammation. Unlike other pancreatic diseases, it typically has a dramatic response to glucocorticoid therapy. AIP can appear radiographically as a focal mass that is indistinguishable from pancreatic cancer or as a diffuse pancreatic process that can be pathognomonic for AIP. If untreated, AIP can lead to pancreatic insufficiency, fibrosis, and other complications. The initial discovery of elevated immunoglobulin G4 (IgG4) as a biomarker of AIP helped establish the disorder as distinct from other forms of chronic pancreatitis. However, it is now known that IgG4 levels are often normal in AIP and that other diseases such as pancreatic cancer may also have moderately elevated IgG4 levels.

This topic will review the management of AIP type 1 and type 2. The clinical manifestations and diagnosis of AIP and the management of IgG4-related disease are discussed in detail separately. (See "Autoimmune pancreatitis: Clinical manifestations and diagnosis" and "Treatment and prognosis of IgG4-related disease".)

## INDUCTION OF REMISSION

The approach to management of autoimmune pancreatitis (AIP) is based upon the manifestations of AIP and the presence of immunoglobulin G4-related disease (IgG4-RD). The goal of management is to alleviate the immediate symptoms of AIP and to prevent irreversible

liver fibrosis and pancreatic exocrine and endocrine failure. Treatment may prevent progression to chronic pancreatitis [1]. However, the fibroatrophic changes that accompany the initial presentation of the disease are typically permanent [1,2]. Management of AIP is largely based upon observational studies since there have been few randomized controlled trials ( algorithm 1). Our recommendations are largely consistent with the guidelines of the International Association of Pancreatology consensus conference for the treatment of AIP [3-5] and the Japanese consensus guidelines for treatment of AIP [6]. The management of IgG4-RD is discussed in detail separately. (See "Treatment and prognosis of IgG4-related disease".)

**Indications for therapy**

**Symptomatic patients** — Treatment for AIP is indicated in patients with any one of the following [3]:

- Pancreatic symptoms including obstructive jaundice, abdominal pain, or back pain.
- Symptomatic "other organ involvement" (OOI) associated with IgG4-RD (eg, jaundice from bile duct strictures in patients with overlapping IgG4-related sclerosing cholangitis [IgG4-SC]). (See "Pathogenesis and clinical manifestations of IgG4-related disease", section on 'Clinical manifestations'.)

**Asymptomatic patients** — Indications for treatment in asymptomatic patients with AIP include one of the following:

- Persistent pancreatic mass on imaging (eg, three to four weeks).
- Persistent liver test abnormalities (eg, two to three weeks) in a patient with bile duct obstruction or IgG4-SC, as this can lead to bacterial cholangitis, irreversible liver fibrosis, or cirrhosis.
- IgG4-RD with progressive subclinical lesions, potentially leading to severe, irreversible sequelae in vital organs (eg, biliary tree, kidney, aorta, mediastinum, retroperitoneum, mesentery). (See "Treatment and prognosis of IgG4-related disease", section on 'Treatment approach'.)
- Irreversible pancreatic exocrine and endocrine failure due to AIP.

Asymptomatic patients with AIP without these features require watchful waiting. About 10 to 25 percent of AIP patients improve spontaneously [7].

**Glucocorticoid therapy** — For most patients requiring treatment, we recommend initial treatment with glucocorticoid monotherapy.

**Dose and duration** — Our approach is to initiate treatment with prednisone 0.6 mg/kg (typically 40 mg) per day for four to six weeks [3]. Most patients demonstrate clinical and/or radiologic improvement within the first four to six weeks; many patients respond even earlier. Administration of high-dose glucocorticoids results in more rapid and successful induction of remission than conservative management [3]. Treatment may prevent progression to chronic pancreatitis [1].

Induction therapy with prednisone/prednisolone 20 to 30 mg/day with a minimum of 12 weeks of treatment may also achieve a response. Although lower doses of prednisolone appear to have comparable efficacy in retrospective studies, randomized trials are lacking [8]. In one retrospective, multicenter study of 65 AIP patients who were treated with one of three different prednisone regimens at the discretion of the treating clinician (low dose [10 to 20 mg daily]; medium dose [30 mg daily], and high dose [40 to 60 mg daily]), at six-month follow up, nearly all patients had complete resolution of symptoms [8]. In patients treated with 10 or 15 mg/day, this dose was maintained for at least six months. Patients treated with higher initial daily doses were continued for two to four weeks, after which the dose was tapered, usually by 5 mg per one to two weeks.

**Efficacy** — The majority of patients with AIP respond to glucocorticoids (80 to 99 percent), but patients may require a second course of treatment due to disease relapse [9-12]. (See 'Incidence and risk factors for relapse' below.)

In a retrospective study in Japan that included 563 patients with AIP, of whom 459 (82 percent) received glucocorticoids, remission rates were significantly higher in patients treated with glucocorticoids as compared with patients who did not receive glucocorticoid therapy (98 versus 74 percent) [5]. Indications for the use of glucocorticoids included obstructive jaundice (60 percent), abdominal pain (11 percent), associated extrapancreatic lesions except the biliary duct (11 percent), and diffuse enlargement of the pancreas (10 percent). In another retrospective, international multicenter study of 1064 patients (684 with type 1 AIP and 52 with type 2 AIP), remission rates in patients with type 1 and type 2 AIP who were treated with glucocorticoids were 99 and 82 percent, respectively [4]. Rates of relapse were significantly higher in patients with type 1 AIP as compared with type 2 AIP (31 versus 9 percent), and in those with associated IgG4-SC (56 versus 26 percent).

### Assessment of response to therapy

**Evaluation** — Most patients typically demonstrate a clinical and/or radiologic improvement within the first four to six weeks. Patients with AIP usually have a rapid and complete resolution of clinical symptoms such as abdominal pain or jaundice after initiation of glucocorticoids. To

assess response to treatment, we perform imaging tests (typically computed tomography or magnetic resonance imaging, whichever was obtained initially) to evaluate for radiologic improvement (eg, a decrease in focal mass, diffuse swelling, narrowed pancreatic duct or biliary strictures) at the end of the four- to six-week induction period.

Failure to respond to a trial of four to six weeks of glucocorticoids should raise the possibility of an alternative diagnosis. (See 'Steroid-refractory disease' below.)

Normalization of serum IgG4 levels alone is not used to assess response to therapy as serologic activity correlates poorly with clinical or radiologic remission [13]. Confirmation of histologic remission in patients with clinical and radiologic remission with treatment is not required.

**Patients who respond**

**Glucocorticoid taper** — In patients with a clinical and radiologic response to prednisone/prednisolone 40 mg/day for four weeks, we gradually taper the dose by 5 mg/week over a two-month period, with the goal of discontinuing glucocorticoids entirely unless patients have a high risk of relapse. (See 'Maintenance treatment for high relapse risk' below.)

There are other alternative approaches to glucocorticoid tapers [3]. The international consensus for the treatment of AIP recommends a taper by 5 to 10 mg/day every one to two weeks until a daily dose of 20 mg is reached, followed by tapering by 5 mg every two weeks. The duration of total induction treatment should generally last 12 weeks.

**Inability to tolerate taper** — Patients may initially improve but have worsening of disease or "flare" before AIP is in remission. This typically occurs during glucocorticoid dose reduction or withdrawal.

Treatment options for steroid-dependent AIP include continued low-dose glucocorticoids (prednisone 2.5 to 10 mg daily), thiopurines (azathioprine, 6-mercaptopurine), and rituximab.

**Steroid-refractory disease** — Failure to respond to a trial of glucocorticoids for four to six weeks should raise the possibility of an alternative diagnosis. The lack of response to a glucocorticoid trial warrants referral for surgical exploration for pancreatic cancer. If the diagnosis of AIP is re-established, steroid-refractory patients can be treated with rituximab. (See "Autoimmune pancreatitis: Clinical manifestations and diagnosis", section on 'Differential diagnosis'.)

**Special populations**

**Patients with contraindications to steroids** — Rituximab (anti-CD20 antibody) can be used as initial treatment in patients in whom glucocorticoids are contraindicated [14,15]. Other steroid-sparing agents such as thiopurines are poorly effective as single agents for induction of remission.

**Patients with obstructive jaundice** — Obstructive jaundice is a common complication of AIP. Patients with mild jaundice (incomplete obstruction) without cholangitis can be treated with glucocorticoids alone without biliary stenting [4,16]. However, in patients with AIP and co-existing IgG4-SC as part of IgG4-RD, biliary drainage is useful to prevent biliary infection, and use of brushing and cytology can differentiate IgG4-SC from biliary malignancy [3].

The management of biliary strictures in patients with AIP is controversial. Our recommendations are based on consensus opinion and observational studies because randomized trials are lacking. The clinical considerations and risks vary markedly among patients, as well as with the location of the strictures (including intra- and extra-hepatic biliary strictures), the extent of the strictures, and the degree of obstruction.

## SUBSEQUENT MANAGEMENT

### Approach based on risk of AIP relapse

**Incidence and risk factors for relapse** — Relapse is defined as recurrent clinical, radiologic, or biochemical evidence of disease activity that occurs **after** complete remission is achieved. Abdominal pain as a standalone symptom in the absence of pancreatic inflammation and elevation of immunoglobulin G4 (IgG4) without concomitant biochemical or radiologic change does not represent relapse and does not necessitate retreatment.

Type 2 AIP has no risk of recurrence and maintenance treatment is not needed.

Rates of relapse are significant in patients with type 1 autoimmune pancreatitis (AIP) and in those with associated IgG4-related sclerosing cholangitis (IgG4-SC) [17]. In a retrospective multicenter study, 56 percent of patients with type 1 AIP relapsed within one year, 76 percent within two years, and 92 percent within three years [5]. In another retrospective, international multicenter study of 1064 patients (684 with type 1 AIP and 52 with type 2 AIP), remission rates in patients with type 1 and type 2 AIP were 99 and 82 percent, respectively [4]. Rates of relapse were significantly higher in patients with type 1 AIP as compared with type 2 AIP (31 versus 9 percent), and in those with associated IgG4-SC (56 versus 26 percent).

Patients are considered at high risk of AIP relapse if they have one or more of the following:

- Type 1 AIP with diffuse enlargement of the pancreas
- More than two other organs involvement (OOIs) or association with proximal IgG4-SC before treatment
- Delayed radiologic remission with treatment
- Persistently high serum IgG4 (>2 times upper limit of normal) after treatment

Patients are considered at low risk of relapse if they have low disease activity, such as involvement in the pancreas alone with segmental/focal lesion without any OOIs, and complete radiologic remission with rapid normalization of IgG/IgG4 levels in response to glucocorticoids [3].

**Maintenance treatment for high relapse risk** — In patients at high risk of relapse, we suggest maintenance treatment to reduce the risk of relapse [18]. Treatment options include low-dose glucocorticoids (prednisone 2.5 to 10 mg/day for one to three years or indefinitely), immunomodulators (azathioprine [2 mg/kg daily] or mycophenolate mofetil [750 mg twice daily]), or rituximab [3,14].

The choice of agent depends upon individual patient factors (presence of IgG4 disease), cost/insurance considerations, and availability. We generally use azathioprine as a first line of therapy for maintenance treatment of AIP and use rituximab in patients with IgG4-RD or resistance to or side effects from other treatments, including glucocorticoids and immunomodulators [14,19,20]. Rituximab has been successfully used to treat patients with IgG4-RD, including type 1 AIP, with resistance to or side effects from other treatments, including glucocorticoids and immunomodulators, and for IgG4-SC with AIP [14,19,20]. However, rituximab is more expensive than glucocorticoids and immunomodulators and may not be available or covered by insurance.

Maintenance treatment reduces relapse rates in patients with AIP. However, evidence to support the use of steroid-sparing agents (eg, immunomodulators and rituximab) is largely from retrospective studies, and randomized trials comparing steroid-sparing agents to glucocorticoids are lacking. In retrospective studies, relapse-free survival is similar in those treated with glucocorticoids plus immunomodulators as compared with those treated with steroids alone. The following studies are illustrative of the efficacy of maintenance therapy in patients with AIP:

- In a randomized controlled trial to determine if maintenance treatment reduced type 1 AIP relapse rates, 49 patients with AIP were randomly assigned to a prednisone maintenance therapy (5 to 7.5 mg/day) for three years or cessation of therapy by tapering prednisone and discontinuing it at 26 weeks [18]. Relapse rates within three years were significantly

lower in patients assigned to prednisone maintenance as compared with the treatment cessation (23 versus 58 percent). There were no serious corticosteroid-related complications that required discontinuation of treatment during the study.

- A multicenter international study of 1064 patients with AIP (978 with type 1 and 86 with type 2 AIP) evaluated long-term outcomes of AIP [4]. Patients were treated with prednisone at a dose of 30 to 40 mg/day, with tapering by 5 to 10 mg every one to two weeks. Asian centers used maintenance low-dose prednisone 2.5 to 5.0 mg/day for six months to three years. North American studies did not use maintenance glucocorticoid therapy but rather used immunomodulator drugs (ie, azathioprine 2 mg/kg for one to three years). Successful remission was induced in 681 of 684 type 1 AIP patients (99.6 percent) and 48 of 52 patients with type 2 AIP (92 percent). Disease relapse occurred in 245 of 684 type 1 patients (36 percent), but only 8 of 52 patients (15 percent) with type 2 AIP. A higher proportion of relapses in steroid-treated patients occurred after glucocorticoids had been discontinued (67 percent) as compared to during tapering (15 percent) or during maintenance glucocorticoid therapy (18 percent). In the management of relapse, glucocorticoids were successful in 201 of 210 patients (95 percent), and azathioprine was successful in 56 of 68 patients (85 percent). A small number of patients responded to 6-mercaptopurine, rituximab, cyclosporine, or cyclophosphamide.

- A retrospective study included 116 patients with type 1 AIP who were initially treated with glucocorticoids [15]. During a median follow-up of 47 months, 52 patients experienced 76 relapse episodes. The first relapse was treated with another course of prednisone in 24 patients and with prednisone and an immunomodulator in 27 patients. Failure or intolerance of immunomodulator therapy occurred in 17 patients (45 percent). Of the patients with glucocorticoids or immunomodulator intolerance/resistance who were treated with rituximab, 10 of 12 (83 percent) achieved complete remission and had no relapse. Of note, the relapse-free survival was similar in patients treated with glucocorticoids plus an immunomodulator and those treated with glucocorticoids alone. Rituximab was effective in the treatment of both immunomodulator-resistant and steroid-intolerant patients.

**Watchful waiting for low risk of relapse** — We monitor patients at low risk of relapse with watchful waiting. This consists of the following:

- Patients are advised to report any new symptoms.

- Clinical and laboratory evaluation should be performed every six months, including a complete blood count and blood chemistries.

- Periodic imaging, for example, after four to six weeks, at six months, and then based on clinical progression and concern for recurrence or to evaluate other possible etiologies.

**Management of relapse** — Relapse can be treated with the glucocorticoid therapy if it was successful for initial induction of remission. (See 'Dose and duration' above.)

For patients who do not respond to glucocorticoids, steroid-sparing agents (immunomodulators or rituximab) are an alternative [21]. Thiopurines and mycophenolate mofetil require overlap with glucocorticoids for six to eight weeks. Glucocorticoids are withdrawn only when AIP is in remission. Rituximab has also been successfully used to treat patients with IgG4-RD, including type 1 AIP with resistance to or side effects of glucocorticoids or immunomodulators [15,21].

For patients with more than one relapse, glucocorticoids (prednisone 2.5 to 10 mg daily) or steroid-sparing agents (immune-modulators or rituximab) are used as maintenance treatment. (See 'Maintenance treatment for high relapse risk' above.)

## MONITORING FOR LONG-TERM COMPLICATIONS

Patients with AIP can develop pancreatic exocrine and endocrine insufficiency (diabetes mellitus). Risk factors for developing diabetes mellitus include advanced age and long duration of disease resulting in extensive parenchymal atrophy [13].

## SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Chronic pancreatitis and pancreatic exocrine insufficiency" and "Society guideline links: IgG4-related disease".)

## SUMMARY AND RECOMMENDATIONS

- Treatment for autoimmune pancreatitis (AIP) is indicated in patients with any one of the following:

  Symptomatic patients with:

  - Pancreatic symptoms including obstructive jaundice, abdominal pain, or back pain.

- Symptomatic "other organ involvement" (OOI) associated with IgG4-related disease (IgG4-RD; eg, jaundice from bile duct strictures in patients with overlapping IgG4-related sclerosing cholangitis [IgG4-SC]).

Asymptomatic patients with:

- Persistent pancreatic mass on imaging (eg, three to four weeks).
- Persistent liver test abnormalities (eg, two to three weeks) in a patient with bile duct obstruction or IgG4-SC that may lead to bacterial cholangitis or irreversible liver fibrosis and cirrhosis.
- IgG4-RD with progressive subclinical lesions, potentially leading to severe, irreversible sequelae in vital organs (eg, biliary tree, kidney, aorta, mediastinum, retroperitoneum, mesentery).
- Irreversible pancreatic exocrine and endocrine failure.

- In patients with AIP and an indication for treatment, we recommend initial treatment with glucocorticoids as compared with watchful waiting (**Grade 1C**). Our approach is to initiate treatment with prednisone at a dose of 40 mg per day for four to six weeks. Toward the end of the four- to six-week induction period, we perform repeat imaging tests (typically computed tomography or magnetic resonance imaging) to evaluate for radiologic improvement (eg, a decrease in focal mass, diffuse swelling, narrowed pancreatic duct or biliary strictures). Most patients destined to respond demonstrate clinical and/or radiologic improvement within the first four to six weeks. Failure to respond in this timeframe should raise the possibility of alternative diagnoses. (See 'Glucocorticoid therapy' above and 'Steroid-refractory disease' above.)

- In patients with a clinical and radiologic response to prednisone/prednisolone, we gradually taper the dose by 5 mg/week with a planned reduction over a two-month period, with the goal of discontinuing glucocorticoids entirely unless the patient has a high risk of relapse. (See 'Glucocorticoid taper' above.)

- Patients are considered at high risk of AIP relapse if they have one or more of the following (see 'Incidence and risk factors for relapse' above):
  - Type 1 AIP with diffuse enlargement of the pancreas
  - More than two OOIs or association with proximal IgG4-SC before treatment
  - Delayed radiologic remission with treatment
  - Persistently high serum IgG4 (>2 times upper limit of normal) after treatment

- In patients at high risk of AIP relapse, we suggest maintenance treatment to reduce the risk of relapse rather than watchful waiting (**Grade 2C**). Maintenance treatment options include low-dose glucocorticoids, immunomodulators (azathioprine or mycophenolate mofetil), or rituximab ( algorithm 1).

  - For patients without IgG4-RD, we suggest azathioprine as a first line of therapy for maintenance treatment of AIP as compared to rituximab (**Grade 2C**). The basis for our choice of treatment is the lower cost and wider availability of azathioprine in addition to our familiarity with its use. However, low-dose glucocorticoids and mycophenolate mofetil are reasonable treatment options in these patients. (See 'Maintenance treatment for high relapse risk' above.)

  - For patients with IgG4-RD, we use rituximab for maintenance treatment of AIP. (See "Treatment and prognosis of IgG4-related disease", section on 'Maintenance therapy'.)

- We monitor patients at low risk of relapse with watchful waiting. Patients are advised to report new AIP-related symptoms and require periodic clinical evaluation, laboratory testing, and abdominal imaging. (See 'Watchful waiting for low risk of relapse' above.)

- Patients who relapse can be managed with a repeat course of glucocorticoids. For patients who do not respond to glucocorticoids, steroid-sparing agents (immunomodulators or rituximab) are an alternative. In patients with more than one relapse, glucocorticoids or a steroid-sparing agent (immunomodulator or rituximab) are used as maintenance treatment. (See 'Management of relapse' above.)

- Patients with AIP can develop pancreatic exocrine and endocrine insufficiency (diabetes mellitus). Risk factors for developing diabetes mellitus include advanced age and long duration of disease resulting in extensive parenchymal atrophy. (See 'Monitoring for long-term complications' above.)

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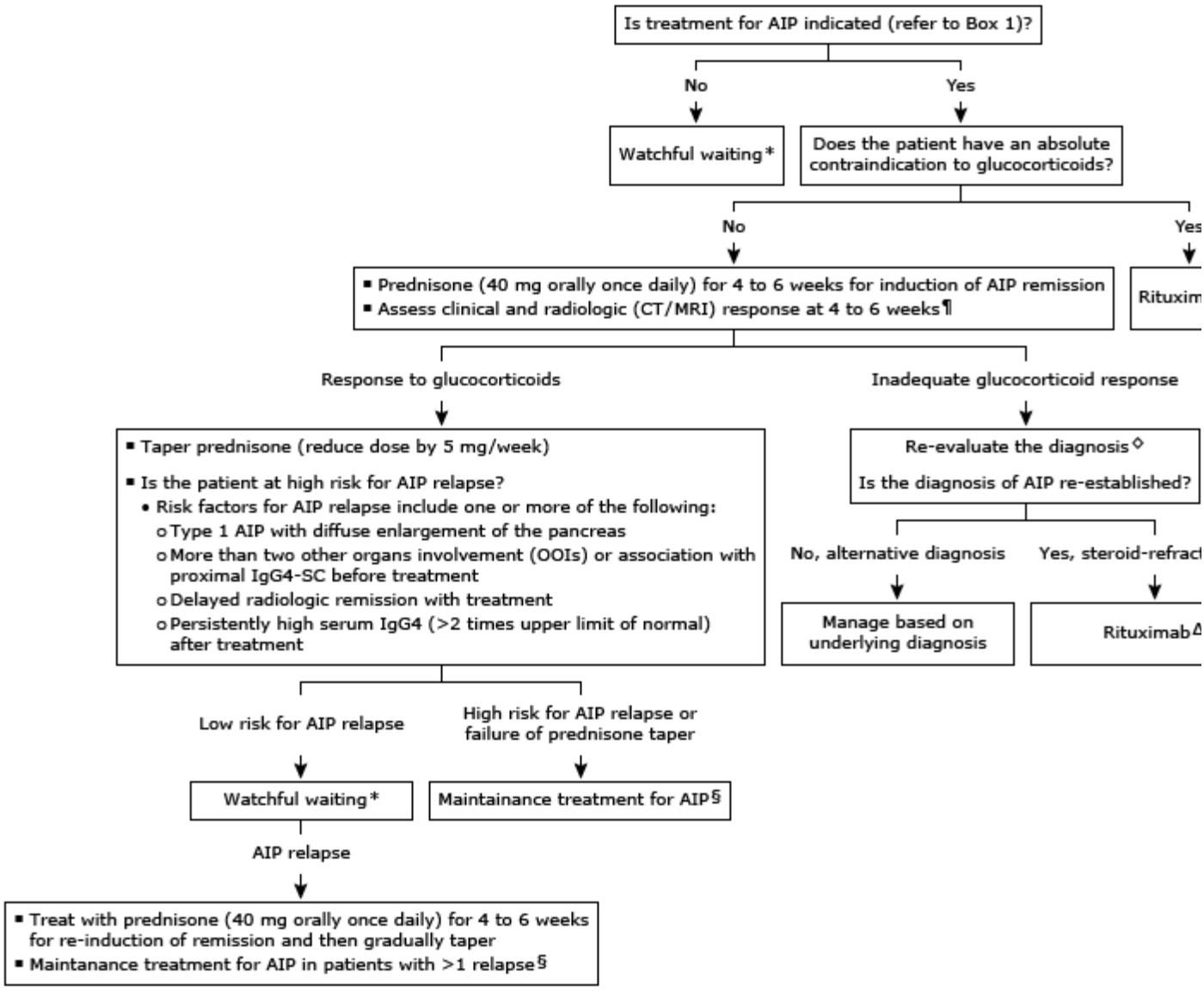
## GRAPHICS

### Overview of the management of patients with autoimmune pancreatitis




AIP: autoimmune pancreatitis; CT: computed tomography; MRI: magnetic resonance imaging; IgG4-RD: imm

* Patients are advised to report new AIP-related symptoms and require periodic clinical evaluation, laborato

¶ Refer to UpToDate content on autoimmune pancreatitis for additional details.

Δ Other steroid-sparing agents such as thiopurines are poorly effective as single agents for induction of rem

◇ The lack of response to a glucocorticoid trial warrants referral for surgical exploration for pancreatic canc

§ Options include low-dose prednisone, thiopurines (azathioprine, 6-mercaptopurine), or rituximab. The cho RD) and cost/insurance considerations, and availability. We use azathioprine as a first line of therapy for mai resistance to or side effects from treatments including glucocorticoids and immunomodulators.

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## Contributor Disclosures

**Douglas G Adler, MD, FACG, AGAF, FASGE** Consultant/Advisory Boards: Abbvie [Endoscopy]; Boston Scientific [Endoscopy]; Endorotor [Endoscopy]; Merit [Endoscopy]; Olympus [Endoscopy]. Speaker's Bureau: Abbvie [Pancreatology, general GI]. All of the relevant financial relationships listed have been mitigated. **Lawrence S Friedman, MD** Other Financial Interest: Elsevier [Gastroenterology]; McGraw-Hill [Gastroenterology]; Wiley [Gastroenterology]. All of the relevant financial relationships listed have been mitigated. **Shilpa Grover, MD, MPH, AGAF** No relevant financial relationship(s) with ineligible companies to disclose.

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