*Original Article*

# Autoimmune Pancreatitis and IgG4-related Cholangitis: A Single-center Experience

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***Background:*** Autoimmune pancreatitis (AIP) and IgG4-related cholangitis (IRC) are newly-recognized diseases. Reports in Thailand are scarce.

***Objective:*** To evaluate clinical presentations, other organ involvement (OOI), serology, radiologic, endoscopic features and treatment of AIP and IRC in one institute.

***Materials and Methods:*** Patients with AIP and IRC, who were diagnosed and followed-up at Siriraj Hospital during 2005 to 2016 were retrospectively reviewed.

***Results:*** There were 15 patients (75%) with AIP, 7 patients (37%) with IRC (4 isolated IRC and 3 IRC with AIP) and 1 (5%) with IgG4-related disease without AIP or IRC (sialadenitis and retroperitoneal fibrosis). Male to female ratio was 1.9: 1. Median age of onset was 64 years. Among the 15 AIP, all were type 1. Initial presentations were jaundice (60%), abdominal pain (40%), OOI (20%; sialadenitis, orbital pseudotumor), and weight loss (7%). Clinical manifestations during the course were jaundice (80%), abdominal pain (67%), weight loss (47%), OOI (47%; retroperitoneal fibrosis, IRC, sialadenitis, orbital pseudotumor, periurethral mass, lung nodules) and steatorrhea (7%). Serum IgG4 was elevated in all patients. Radiography showed diffuse type in 8 cases (53%), focal type in 5 (33%), and unknown in 2 (13%). More than half were suspicious of AIP before diagnosis. Corticosteroid was required in 10 patients (67%). Maintenance therapy was offered in 5 patients (33%) with steroid and azathioprine. Surgery was performed before diagnosis in 7 patients (47%). Spontaneous remission occurred in 4 patients (27%). Among the 7 IRC, initial presentations were jaundice (57%), weight loss (29%), abdominal pain (14%), prolonged fever (14%), and cholangitis (14%). Clinical manifestations during the course were jaundice (57%), abdominal pain (57%), weight loss (29%), recurrent cholangitis (14%) and retroperitoneal fibrosis (14%). Serum IgG4 was elevated in 5 patients (71%). Prednisolone was prescribed in 6 cases (86%). Five patients (71%) required azathioprine. Surgery was performed before diagnosis in 4 patients (57%).

***Conclusion:*** AIP and IRC were uncommon. The most common presentation was jaundice. All AIP were type 1 and more than half were diffuse type. OOI were common. Half of IRC had AIP. Approximately half of AIP/IRC patients underwent surgery before diagnosis. Corticosteroid was mainstay treatment.

***Keywords:*** Autoimmune pancreatitis, IgG4, Immunoglobulin G4, Sclerosing cholangitis, Thailand

**J Med Assoc Thai 2020;103(Suppl.8): S58-63**
**Website:** http://www.jmatonline.com

Autoimmune pancreatitis (AIP) and IgG4-related cholangitis (IRC) are recently recognized diseases, of which autoimmune mechanisms are involved in the pathogeneses. The elevation of serum immunoglobulin G, subclass 4 (IgG4) is a hallmark of AIP[1], IRC[2], and recent concept accepted that AIP and IRC is a part of IgG4-related systemic sclerosing disease[3].

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**How to cite this article:**

Lertussavavivat K, Chumjang S, Pongprasobchai S. Autoimmune Pancreatitis and IgG4-related Cholangitis: A Single-center Experience. J Med Assoc Thai 2020;103(Suppl.8): S58-63.

doi.org/10.35755/jmedassocthai.2020.S08.12173

AIP often mimics pancreatic cancer. However, AIP has many unique clinical, radiological, serological, and histopathological characteristics as follows: elderly male preponderance; frequent initial symptom of obstructive jaundice without pain; occasional association with impaired pancreatic endocrine or exocrine function and various extrapancreatic lesions (other organ involvement); favorable response to steroid therapy; radiological findings of pancreatic enlargement and irregularly narrow main pancreatic duct; serological findings of elevation of serum γ-globulin, IgG, or IgG4 levels; and histopathological findings of lymphoplasmacytic sclerosing pancreatitis (LPSP) or idiopathic duct-centric pancreatitis (IDCP)[4-7]. Currently, the diagnosis of AIP requires a combination of features according to the International Consensus Diagnostic Criteria (ICDC) 2011[8]. IRC, likewise, is a mimicker of primary sclerosing cholangitis[9,10] and cholangiocarcinoma[11-13]. Taken together, difficulty in the diagnosis and the lack of physician's awareness pose many patients for unnecessary surgery. Thus, it is important to increase the recognition and awareness of

physicians for these two conditions.

In Thailand, studies on AIP and IRC are very scarce due to the uncommonness of the disease. Available data are only 2 case reporst[14,15] and 1 small case series[16]. Thus, this study aims to analyze larger number of cases of AIP and IRC treated at Siriraj Hospital in order to recognized and understand the clinical presentation, serology, radiologic features, endoscopic features and treatment of these two diseases.

## Materials and Methods

### *Study design and population*

This retrospective study involved all patients with AIP or IRC diagnosed and followed-up at Siriraj Hospital during 2005 to 2016. Since the diseases are uncommon, all consecutive cases were included. The study was approved by the Siriraj Institutional Review Board (COA number: Si 370/2017).

### *Diagnosis of AIP and IRC*

AIP was diagnosed by an ICDC 2011[8]. This criteria uses a combination of 1 or more of 5 cardinal features of AIP; 1) imaging features of the following: (a) pancreatic parenchyma (on computed tomography [CT]/magnetic resonance imaging [MRI] and (b) pancreatic duct (endoscopic retrograde cholangiopancreatography [ERCP] or magnetic resonance cholangiopancreatography [MRCP]), 2) serology (IgG4, IgG, and antinuclear antibody), 3) other organ involvement (OOI), 4) histopathology of the pancreas (LPSP or IDCP), and 5) response to a 2-week trial of corticosteroid therapy[8].

IRC was diagnosed by the Japanese criteria 2012[2].

### *Data collection*

Clinical, serology, radiologic, and endoscopic features were collected from medical reports, radiological reports and endoscopic reports. Management was also reviewed in both medical and surgical aspects.

### *Statistical analyses*

Descriptive statistics were used to summarize the patients' characteristics. Continuous data are presented as mean and standard deviation. Categorical data are presented as frequency and percentage.

## Results

### *IgG4-related diseases*

Over the 12-year-period, there have been 20 patients with IgG4-related diseases. There were 15 patients (75%) with AIP, 7 patients (37%) with IRC (4 isolated IRC and 3 IRC with AIP) and 1 patient (5%)with IgG4-related disease at other organs without AIP or IRC (sialadenitis and retroperitoneal fibrosis). Male to female ratio was 1.9: 1. The median age of onset was 63 years.

### *Autoimmune pancreatitis*

In the 15 AIP patients, all were type 1 AIP (or LPSP). Twelve patients (80%) were male. The mean age of the patients was 64±8 years, and more than half of patients had some comorbidities (Table 1).

Majority of the patients (60%) presented with obstructive jaundice and 40% had abdominal pain (Table 2). The mean duration from the initial presentation to diagnosis was 18±31 months. Overall clinical features during the course of the disease were obstructive jaundice 80%, abdominal

**Table 1.** Demographic characteristic of 15 autoimmune pancreatitis patients

| Characteristics | Number (%) |
|---|---|
| Age at diagnosis (years), mean (SD) | 64 (8) |
| Gender | |
| Male | 12 (80) |
| Female | 3 (20) |
| Comorbidities | |
| Diabetes | 9 (60) |
| Hypertension | 7 (47) |
| Dyslipidemia | 5 (33) |
| Cardiovascular diseases | 3 (20) |

SD = standard deviation

**Table 2.** Initial presentations and clinical features during the course of 15 autoimmune pancreatitis patients

| Clinical presentations | Number (%) |
|---|---|
| Initial | |
| Jaundice | 9 (60) |
| Abdominal pain | 6 (40) |
| Other organ involvement | 3 (20) |
| Sialadenitis | 2 (13) |
| Orbital pseudotumor | 1 (7) |
| Weight loss | 1 (7) |
| During the courses of the diseases | |
| Jaundice | 12 (80) |
| Abdominal pain | 10 (67) |
| Other organ involvement | 7 (47) |
| Sclerosing cholangitis | 3 (20) |
| Retroperitoneal fibrosis | 4 (27) |
| Sialadenitis | 3 (20) |
| Orbital pseudotumor | 2 (13) |
| Periurethal mass | 1 (7) |
| Lung nodules | 1 (7) |
| Weight loss | 7 (47) |
| Steatorrhea | 1 (7) |
| Elevated serum IgG4 >140 mg/dL | 15 (100) |

pain 67%, weight loss 47%, retroperitoneal fibrosis 27%, cholangitis 20%, submandibular sialadenitis 20%, orbital pseudotumor 13%, periurethal mass 7%, multiple lung nodule 7%, and steatorrhea 7% (Table 2). All patients (100%) had elevated serum IgG4 level >140 mg/dL.

Radiographic features from CT or MRI showed diffuse type AIP in 8 cases (53%, Figure 1), focal type AIP in 5 cases (33%), and unknown in 2 cases (13%) due to incomplete data from the previous hospitals before surgery. Eight patients (53%) were suspected of AIP by the physicians before having definite diagnosis.

Treatment of the 15 AIP patients are shown in Table 3. Eleven patients (73%) required treatment. Ten patients received prednisolone induction of remission and all responded. Maintenance therapy with low dose prednisolone was offered in 5 patients (33%). Additional therapy with azathioprine was prescribed in 5 patients (33%), most related to the presence of OOI or diffuse type of AIP. Surgical management was done before establishing the diagnosis of AIP in 7 patients (47%). There were 4 patients (27%), whom AIP were resolved without treatment. Relapse occurred in 1 patient without maintenance treatment.

### *IgG4-related cholangitis*

Among 7 patients with IRC, 3 patients (43%) were male, and 4 patient (57%) were female. Four patient (57%) were isolated IRC and 3 (43%) had concomitant AIP. The mean age of the patients was 59±10 years (Table 4).

Initial presentations were obstructive jaundice (57%), weight loss (29%), abdominal pain (14%), prolonged fever (14%), and cholangitis (14%) (Table 5). The mean duration from initial presentation to diagnosis was 25±41 months. Clinical manifestations during the course of disease were obstructive jaundice (57%), abdominal pain (57%), weight loss (29%), retroperitoneal fibrosis (14%), and recurrent cholangitis (14%) (Table 5). Serum IgG4 was elevated in 5 patients (71%).

Cholangiography showed multiple segmental strictures of intrahepatic bile ducts involving the hilar region in all cases (Figure 2).

Table 6 summarized the treatment of the 7 IRC patients. Therapy was given in 6 patients. Induction of

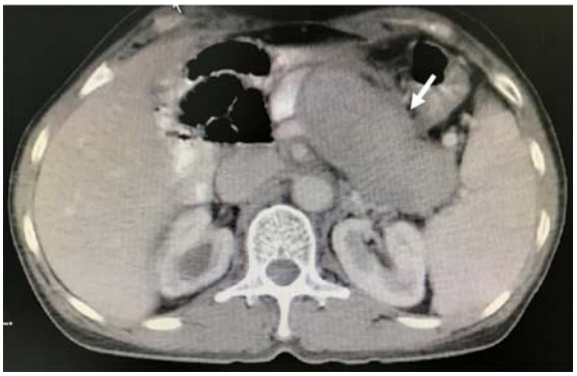

**Figure 1.** Diffuse type autoimmune pancreatitis shows diffuse sausage-like pancreatic enlargement (arrow).

**Table 3.** Treatments of 15 autoimmune pancreatitis patients

| Treatment | Number (%) |
|---|---|
| None | 4 (27) |
| Medical | 10 (67) |
| Prednisolone | 10 (67) |
| Induction | 10 (67) |
| Maintenance | 5 (33) |
| Azathioprine | 5 (33) |
| Surgery | 7 (47) |

**Table 4.** Demographic characteristics of 7 IgG4-related cholangitis patients

| Characteristics | Number (%) |
|---|---|
| Age at diagnosis (years), mean (SD) | 59 (10) |
| Gender | |
| Male | 3 (43) |
| Female | 4 (57) |
| Comorbidities | |
| Diabetes mellitus | 2 (29) |
| Hypertension | 2 (29) |
| Dyslipidemia | 1 (14) |

**Table 5.** Initial presentations and clinical features during the courses of 7 IgG4-related cholangitis patients

| Clinical presentations | Number (%) |
|---|---|
| Initial | |
| Jaundice | 4 (57) |
| Abdominal pain | 1 (14) |
| Weight loss | 2 (29) |
| Prolonged fever | 1 (14) |
| Cholangitis | 1 (14) |
| During the courses of the diseases | |
| Jaundice | 4 (57) |
| Abdominal pain | 4 (57) |
| Weight loss | 2 (29) |
| Recurrent cholangitis | 1 (14) |
| Retroperitoneal fibrosis | 1 (14) |
| Elevated serum IgG4 >140 mg/dL | 5 (71) |

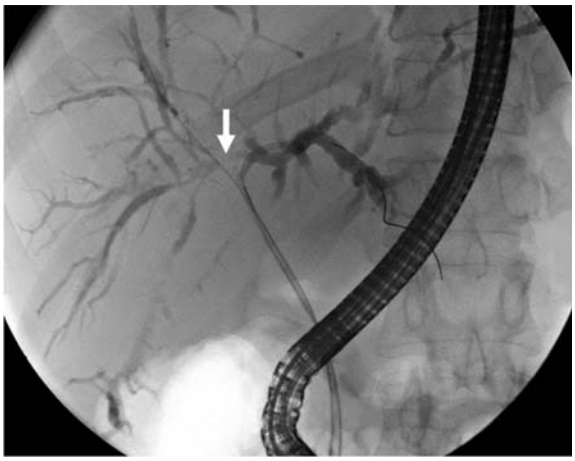

**Figure 2.** Endoscopic retrograde cholangiography shows multiple segmental strictures of intrahepatic bile ducts around the hilum (arrow).

**Table 6.** Treatment of the 7 IgG4-related cholangitis patients

| Treatment | Number (%) |
|---|---|
| None | 1 (14) |
| Medical | 6 (86) |
| Prednisolone | 6 (86) |
| Induction | 5 (71) |
| Maintenance | 6 (86) |
| Azathioprine | 5 (71) |
| Surgery | 4 (57) |

remission with prednisolone was done in 5 of 6 patients and 1 patient was started with prednisolone in maintenance dose without induction. Five patients received azathioprine. Surgery was performed before diagnosis in 4 patients (57%).

## Discussion

AIP and IRC are the two recently-established autoimmune diseases under the umbrella of IgG4-related diseases. The present study supported the uncommonness of them, since there have been 20 cases over 12 years. However, these patients were only those who had encountered gastroenterologists and surgeons. Since IgG4-related disease can be presented with various OOI without pancreas or biliary manifestations, we believed that many IgG4-related diseases remained underdiagnosed and the real prevalence of the diseases would be much higher.

Among the 15 AIP patients, all were type 1 AIP. Type 1 AIP (or LPSP) is much more common than type 2 AIP (or IDCP) in every case series[5,17]. The reason is possibly due to the more availability of serum IgG4 testing that helps diagnose type 1 AIP. On the other hand, type 2 AIP requires mainly the pancreatic core biopsy or surgery pathology, makes it much more difficult to diagnose. We found diffuse type AIP in at least 53%, closed to the 40% rate by the international survey[5]. This study confirmed the demographic features of type 1 AIP to be elderly men, similar to other studies[5,17].

The most common initial and clinical presentations of type 1 AIP in this study were similar to those in the international survey[5], which were obstructive jaundice, following by abdominal pain and OOIs including IRC, retroperitoneal fibrosis, orbital pseudotumor, sialadenitis, and lung nodules. OOIs were present in 47% of our cases, closed to 45% of the previous survey[5] and all OOIs we found were the well-established OOIs of type 1 AIP[18]. Serum IgG4, which is the hallmark of type 1 AIP was elevated in 100% of patients compared to 60% by the international survey[5].

Although type 1 AIP responds dramatically to steroid therapy, thus, surgery should be avoided. Our study showed that almost half of the patients underwent surgery, possibly due to the unawareness of physicians and the mean delay between symptoms and diagnosis was 18 months. The missed and delayed in diagnosis would be solved by providing education to internists, gastroenterologists and surgeons. The medical treatment in all cases were prednisolone with or without azathioprine, which complied well with the standard guideline[19].

There were 7 cases of IRC in the present study, which was less common than AIP and almost half had concomitant AIP. This picture slightly differs from larger series of IRC, which showed that 90% had concomitant AIP[20]. The demographics of our IRC patients were very similar to AIP, and confirmed the result of other reports of IRC[21]. The common clinical features of IRC were jaundice and abdominal pain, all of which were again similar to other reports[21]. More than half of IRC underwent surgery due to the presumptive diagnosis of cholangiocarcinoma. Recent study showed that 7% of resected presumptive hilar cholangiocarcinoma were actually IRC[22]. Our result, together with another case report of IRC in Thailand[15] emphasize the need to improve our awareness of IRC in Thailand, where cholangiocarcinoma is endemic.

The present study has strengths to be, to our knowledge the largest series of AIP and IRC in Thailand. We used standard international definitions for the diagnosis of AIP and IRC[2,8], made it clear to be definite cases. This study, however, had several weaknesses. It remained having small number of cases, even though we tried our best to include every case of AIP and IRC. Last, due to the retrospective nature of the study, some data such as details of imaging studies and long-term outcomes were lacking. Therefore, further multicenter study is required.

## Conclusion

AIP and IRC were uncommon. The most common initial presentations were obstructive jaundice. All AIP were

type 1 and more than half were diffuse type. Almost half of IRC had concomitant AIP. Approximately half of AIP and IRC patients underwent surgery before diagnosis, possibly due to the unawareness of physicians. Most treatments of AIP and IRC complied with standard guidelines.

## What is already known on this topic?

AIP has 2 types (type 1 and 2) and 2 forms (diffuse and focal). OOIs occur in 40% of cases. Radiologic characteristics, elevated serum IgG4 and the presence of OOIs help diagnose the disease. It responses well to corticosteroid and surgery could be avoided. IRC is difficult to diagnose and may mimics primary sclerosing cholangitis and cholangiocarcinoma.

## What this study adds?

AIP and IRC were uncommon in Thailand. All AIP were type 1. Various OOIs were present and occurred in almost half of the cases. Mean diagnostic delay was 1.5 years. Surgery was performed in almost half of the cases due to the unawareness of AIP. All IRC mimicked hilar cholangiocarcinoma. Diagnoses were difficult and delayed with the mean of 2 years.

## Conflicts of interest

The authors declare no conflict of interest.

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## โรคตับอ่อนอักเสบออโตอิมมูนและโรคท่อน้ำดีอักเสบแข็งไอจีจี 4: ประสบการณ์จากหนึ่งสถาบัน

กฤศณัฏฐ์ เลิศอัศววิวัฒน์, ศศมน ชุ่มแจ้ง, สุพจน์ พงศ์ประสบชัย

***ภูมิหลัง:*** โรคตับอ่อนอักเสบออโตอิมมูนและโรคท่อน้ำดีตีบแข็งไอจีจี 4 เป็นโรคใหม่ รายงานในประเทศไทยยังมีน้อย

***วัตถุประสงค์:*** เพื่อศึกษาลักษณะทางคลินิก อาการแสดงที่อวัยวะอื่น ซีโรโลยี ภาพรังสี การส่องกล้อง และการรักษาโรคตับอ่อนอักเสบออโตอิมมูนและโรคท่อน้ำดีตีบแข็งไอจีจี 4 ในหนึ่งสถาบัน

***วัสดุและวิธีการ:*** ผู้ป่วยโรคตับอ่อนอักเสบออโตอิมมูนและโรคท่อน้ำดีตีบแข็งไอจีจี 4 ที่ได้รับการวินิจฉัยและติดตามที่โรงพยาบาลศิริราชตั้งแต่ พ.ศ. 2548 ถึง พ.ศ. 2559 จะได้รับการทบทวนย้อนหลัง

***ผลการศึกษา:*** มีผู้ป่วยโรคตับอ่อนอักเสบออโตอิมมูน 15 ราย เป็นโรคท่อน้ำดีตีบแข็งไอจีจี 4 7 ราย (4 รายเป็นโรคท่อน้ำดีตีบแข็งไอจีจี 4 อย่างเดียว 3 รายเป็นร่วมกับโรคตับอ่อนอักเสบออโตอิมมูน) และ 1 ราย เป็นโรคไอจีจี 4 เฉพาะที่ต่อมน้ำลายและพังผืดหลังช่องท้อง สัดส่วนเป็นชายต่อหญิง 1.9 ต่อ 1 อายุเฉลี่ย 64 ปี ผู้ป่วยโรคตับอ่อนอักเสบออโตอิมมูนเป็นชนิดที่ 1 ทั้งหมด อาการนำคือ ดีซ่าน (ร้อยละ 60), ปวดท้อง (ร้อยละ 40), อาการแสดงที่อวัยวะอื่น ร้อยละ 20 (ต่อมน้ำลายอักเสบและก้อนเนื้องอกเทียมที่ตา), และน้ำหนักลด (ร้อยละ 7) ลักษณะทางคลินิกระหว่างการดำเนินโรค คือ ดีซ่าน (ร้อยละ 80), ปวดท้อง (ร้อยละ 67), น้ำหนักลด (ร้อยละ 47), อาการแสดงที่อวัยวะอื่น (ร้อยละ 47; พังผืดหลังช่องท้อง ท่อน้ำดีอักเสบตีบแข็ง ต่อมน้ำลายอักเสบ ก้อนเนื้องอกเทียมที่ตา ก้อนรอบท่อปัสสาวะ และก้อนที่ปอด) และอุจจาระเป็นน้ำมัน (ร้อยละ 7) ผู้ป่วยทุกรายมีซีรัมไอจีจี 4 สูง ภาพรังสีพบเป็นชนิดทั่วตับอ่อน 8 ราย (ร้อยละ 53) เฉพาะส่วน 5 ราย (ร้อยละ 33) และไม่มีข้อมูล 2 ราย (ร้อยละ 13) ผู้ป่วยมากกว่าครึ่งแพทย์ผู้ดูแลสงสัยโรคตับอ่อนอักเสบออโตอิมมูนตั้งแต่ต้น ผู้ป่วยได้รับการรักษาด้วยคอร์ติโคสเตียรอยด์ 10 ราย (ร้อยละ 67) ได้รับยาเพื่อคงระยะสงบ 5 ราย (ร้อยละ 33) ด้วยยาเพร็ดนิโซโลนและ azathioprine ผู้ป่วย 7 ราย (ร้อยละ 47) ได้รับการผ่าตัดก่อนการวินิจฉัย ผู้ป่วย 4 ราย (ร้อยละ 27) โรคสงบได้เอง ผู้ป่วย 7 รายที่เป็นโรคท่อน้ำดีตีบแข็งไอจีจี 4 มีอาการนำคือ ดีซ่าน (ร้อยละ 57), น้ำหนักลด (ร้อยละ 29), ปวดท้อง (ร้อยละ 14), ไข้เรื้อรัง (ร้อยละ 14) และท่อน้ำดีอักเสบ (ร้อยละ 14) ลักษณะทางคลินิกระหว่างการดำเนินโรค คือ ดีซ่าน (ร้อยละ 57), ปวดท้อง (ร้อยละ 57), น้ำหนักลด (ร้อยละ 29), พังผืดหลังช่องท้อง (ร้อยละ 14) และท่อน้ำดีอักเสบ (ร้อยละ 14) ผู้ป่วย 5 ราย (ร้อยละ 71) มีซีรัมไอจีจี 4 สูง ผู้ป่วย 6 ราย (ร้อยละ 86) ได้รับการรักษาด้วยเพร็ดนิโซโลน ผู้ป่วย 5 รายได้ azathioprine ร่วมด้วยผู้ป่วย 4 ราย (ร้อยละ 57) ได้รับการผ่าตัดก่อนการวินิจฉัย

***สรุป:*** โรคตับอ่อนอักเสบออโตอิมมูนและโรคท่อน้ำดีตีบแข็งไอจีจี 4 พบได้น้อย อาการนำที่พบบ่อยที่สุดคือ ดีซ่าน พบอาการแสดงที่อวัยวะอื่นได้บ่อย โรคตับอ่อนอักเสบออโตอิมมูนทุกรายเป็นชนิดที่ 1 และมากกว่าครึ่งเป็นแบบทั่วตับอ่อน ผู้ป่วยโรคท่อน้ำดีตีบแข็งไอจีจี 4 มากกว่าครึ่งเกิดร่วมกับโรคตับอ่อนอักเสบออโตอิมมูน ผู้ป่วยครึ่งหนึ่งได้รับการผ่าตัดก่อนจะถูกวินิจฉัยโรค การรักษาหลักคือคอร์ติโคสเตียรอยด์