International Consensus Diagnostic Criteria for Autoimmune Pancreatitis

Guidelines of the International Association of Pancreatology

Tooru Shimosegawa, MD, Suresh T. Chari, MD,† Luca Frulloni, MD,‡ Terumi Kamisawa, MD,§ Shigeyuki Kawa, MD,|| Mari Mino-Kenudson, MD,¶ Myung-Hwan Kim, MD,# Günter Klöppel, MD,** Markus M. Lerch, MD,†† Matthias Löhr, MD,‡‡ Kenji Notohara, MD,§§ Kazuichi Okazaki, MD,|||| Alexander Schneider, MD,¶¶ and Lizhi Zhang, MD##*

Objectives: To achieve the goal of developing international consensus diagnostic criteria (ICDC) for autoimmune pancreatitis (AIP).

Methods: An international panel of experts met during the 14th Congress of the International Association of Pancreatology held in Fukuoka, Japan, from July 11 through 13, 2010. The proposed criteria represent a consensus opinion of the working group.

Results: Autoimmune pancreatitis was classified into types 1 and 2. The ICDC used 5 cardinal features of AIP, namely, imaging of pancreatic parenchyma and duct, serology, other organ involvement, pancreatic histology, and an optional criterion of response to steroid therapy. Each feature was categorized as level 1 and 2 findings depending on the diagnostic reliability. The diagnosis of type 1 and type 2 AIP can be definitive or probable, and in some cases, the distinction between the subtypes may not be possible (AIP–not otherwise specified).

Conclusions: The ICDC for AIP were developed based on the agreement of an international panel of experts in the hope that they will promote worldwide recognition of AIP. The categorization of AIP into types 1 and 2 should be helpful for further clarification of the clinical features, pathogenesis, and natural history of these diseases.

Key Words: autoimmune pancreatitis, international consensus diagnostic criteria, type 1 AIP, type 2 AIP

(*Pancreas* 2011;40: 352–358)

From the *Division of Gastroenterology, Tohoku University Graduate School of Medicine, Sendai, Japan; †Division of Gastroenterology and Hepatology, Mayo Clinic, Rochester, MN; ‡Department of Medicine, University of Verona, Verona, Italy; §Department of Internal Medicine, Tokyo Metropolitan Komagome Hospital, Tokyo; ||Center for Health, Safety and Environmental Management, Shinshu University, Matsumoto, Nagano, Japan; ¶Department of Pathology, Harvard Medical School, Massachusetts General Hospital, Boston, MA; #Department of Gastroenterology, University of Ulsan College of Medicine, Asan Medical Center, Songpa-gu, Seoul, Korea; **Department of Pathology, Technical University of Munich, Munich; ††Department of Internal Medicine A, Ernst-Moritz-Arndt-University, Greifswald, Germany; ‡‡Department of Surgical Gastroenterology, Karolinska Institute, Stockholm, Sweden; §§Department of Pathology, Kurashiki Central Hospital, Miwa, Kurashiki; ||||Division of Gastroenterology and Hepatology, Kansai Medical University, Moriguchi, Osaka, Japan; ¶¶Department of Medicine II (Gastroenterology, Hepatology, Infectious Diseases), University of Heidelberg at Mannheim, Mannheim, Germany; and ##Division of Anatomic Pathology, Mayo Clinic, Rochester, MN.

Received for publication January 12, 2011; accepted February 4, 2011.

Reprints: Tooru Shimosegawa, MD, Division of Gastroenterology, Tohoku University Graduate School of Medicine, 1-1 Seiryomachi, Aobaku, Sendai 980-8574, Japan (e-mail: tshimosegawa@int3.med.tohoku.ac.jp).

The first (T.S.) and second authors (S.T.C.) cochaired the working group and prepared the draft of the manuscript. They and the following 12 authors listed in alphabetical order contributed to the development of the consensus diagnostic criteria.

The diagnosis of autoimmune pancreatitis (AIP) remains a challenging test of our clinical skills. This difficulty is further compounded by lack of universally accepted criteria for its diagnosis. For the past decade, many different diagnostic criteria for AIP have been reported from Asia, Europe, and North America. The lack of consensus to date on diagnostic criteria for AIP can be traced to 2 basic reasons. First, the practice patterns in the usage of various tests and perceived accuracy of these tests for diagnosis of AIP vary considerably worldwide. For example, endoscopic retrograde pancreatogram is routinely used for investigating obstructive jaundice in Japan and is a mandatory criterion in the Japanese criteria.[1,2] However, Western endoscopists generally avoid injecting the pancreatic duct in patients with obstructive jaundice for fear of causing pancreatitis, and AIP in the West is diagnosed without a requirement for endoscopic retrograde pancreatography (ERP). Similarly, core biopsy of the pancreas to diagnose AIP has been championed by the Mayo Clinic group[3] but is not routinely used elsewhere.

Another important reason for a lack of consensus to date is the fact that it has become increasingly clear that the term "AIP" encompasses 2 different types of the disease with distinct histopathology and clinical profile that need different criteria for their diagnoses.[4] Whereas the Asian and American criteria are geared toward diagnosis of one form of the disease, the Italian criteria have a mixture of features of both types.[5]

There has been much to learn about AIP from the experience of various centers that have described this condition. However, the continued usage of multiple diagnostic criteria and their continued proliferation is not in the best interest of this field. It is time that we put the best of all criteria together and develop International Consensus Diagnostic Criteria for AIP. To achieve this goal, an international panel of experts was convened during the Fourteenth Congress of the International Association of Pancreatology held in Fukuoka, Japan, from July 11 through 13, 2010. The proposed criteria represent a consensus opinion of the working group.

AIP: DEFINITION

Autoimmune pancreatitis is a distinct form of pancreatitis characterized clinically by frequent presentation with obstructive jaundice with or without a pancreatic mass, histologically by a lymphoplasmacytic infiltrate and fibrosis and therapeutically by a dramatic response to steroids.

AIP Types

Further histological and clinical profiling of patients with AIP reveals 2 distinct types whose histopathologic criteria were agreed upon at an earlier meeting of this expert panel.[4] In one

form, whose histological description is called lymphoplasmacytic sclerosing pancreatitis (LPSP) or AIP without granulocyte epithelial lesions (GELs), the pancreatic histopathology shows 4 characteristic features: (1) dense infiltration of plasma cells and lymphocytes, particularly periductal; (2) peculiar storiform fibrosis; (3) venulitis with lymphocytes and plasma cells often leading to obliteration of the affected veins; and (4) abundant (>10 cells per high-power field [HPF]) immunoglobulin (Ig) G4 positive plasma cells. Clinically, this form of AIP seems to be the pancreatic manifestation of an IgG4-related systemic disease characterized by elevated serum IgG4 levels and extrapancreatic lesions (eg, sclerosing cholangitis, sclerosing sialadenitis, and retroperitoneal fibrosis) associated with infiltration with abundant IgG4-positive plasma cells. This form of AIP presents predominantly with obstructive jaundice in elderly male subjects, and the pancreatic and extrapancreatic manifestations respond to steroid therapy. The clinical diagnosis of LPSP is made by a combination of features previously noted and often can be made without need for histology.

In the United States and Europe, histological examination of resected pancreata of patients with chronic nonalcoholic pancreatitis showed another histopathological pattern of chronic pancreatitis called idiopathic duct-centric pancreatitis (IDCP) or AIP with GEL.[6–8] Idiopathic duct-centric pancreatitis and LPSP share some histopathological features, such as periductal lymphoplasmacytic infiltrate and storiform fibrosis. A characteristic feature of IDCP, not seen in LPSP, is the GEL: intraluminal and intraepithelial neutrophils in medium-sized and small ducts as well as in acini, often leading to the destruction and obliteration of the duct lumen. Idiopathic duct-centric pancreatitis usually has none or very few (<10 cells/HPF) IgG4-positive plasma cells, although this can vary. Clinical data from histologically confirmed IDCP cases show that they have distinctly different profile compared with LPSP cases. Idiopathic duct-centric pancreatitis seems not to be a systemic disease; rather, it seems to be a pancreas-specific disorder. It is not associated with either serum IgG4 elevation or with other organ involvement (OOI) typically seen in LPSP. Approximately 30% of reported cases of IDCP have associated inflammatory bowel disease, such as ulcerative colitis. Patients with IDCP are, on average, a decade younger than LPSP patients and do not show a sex predilection. Currently, IDCP lacks a serological biomarker. Because IDCP patients are seronegative and lack other organ involvement, definitive diagnosis requires pancreatic histology.

AIP Types: Worldwide Distribution

In a recent worldwide survey of AIP,[4] it was noted that whereas most cases of AIP in Asia fit the profile of LPSP, European and American series had a mixture of patients fitting the profiles of both LPSP and IDCP. Because the diagnosis of IDCP requires histological examination of an adequate specimen of the pancreas, which is not frequently available, IDCP cannot be diagnosed easily, and this may explain the fewer cases of IDCP diagnosed worldwide.

AIP Types: Nomenclature

The terms *LPSP* (AIP without GELs) and *IDCP* (AIP with GELs) refer to pancreatic histological patterns in AIP. Because pancreatic histology often is not available, the terms *type 1* and *type 2 AIP* have been introduced to describe the clinical profiles associated with LPSP and IDCP, respectively, while recognizing the similarities between the 2 entities.[9] Whether type 2 is an autoimmune process has been debated[4]; however, its clinical presentation with obstructive jaundice, overlap in histological features with type 1, and anecdotal but yet unconfirmed response to steroids leads to a clinical diagnosis of AIP in patients with type 2. Additionally, clinically important diagnostic and therapeutic considerations, that is, need to accurately distinguish it from pancreatic cancer and treatment with steroids, are similar in the 2 types of AIP. The consensus opinion was that the term *type 1* and *type 2* should be used to describe the clinical profiles associated with LPSP and IDCP, respectively.[4]

International Consensus Diagnostic Criteria for AIP

The goals of the ICDC for AIP are to develop criteria that can be applied worldwide, taking into consideration marked differences in clinical practice patterns, to safely diagnose AIP and avoid misdiagnosis of pancreatic cancer as AIP (Tables 1–5, Figs. 1–3). The ICDC for AIP were developed after review of existing criteria, including JPS (2002, 2006),[1,2] HISORt (2006, 2009),[10,11] Korean (2007),[12] Asian (2008),[13] Mannheim (2009),[14] and Italian (2003, 2009).[15]

DIAGNOSIS OF AIP: BACKGROUND CONSIDERATIONS

Clinical Presentation

Acute Presentation

The most frequent acute presentation of AIP is with obstructive jaundice and/or pancreatic mass. Whereas most patients have pancreatic swelling (diffuse or focal), a few patients may have a low-density pancreatic mass, and rarely, no abnormality may be seen on cross-sectional imaging. The proposed criteria are meant to be used to diagnose AIP in this setting.

Late Presentation

Pancreatic atrophy, calcification, ductal dilation, and other features of advanced painless chronic pancreatitis may be seen on follow-up of patients with typical acute presentation of AIP. These patients do not complain of pain or recurrent pancreatitis.

TABLE 1. Diagnosis of Definitive and Probable Type 1 AIP Using ICDC

Diagnosis	Primary Basis for Diagnosis	Imaging Evidence	Collateral Evidence
Definitive type 1 AIP	Histology	Typical/indeterminate	Histologically confirmed LPSP (level 1 H)
	Imaging	Typical	Any non-D level 1/level 2
		Indeterminate	Two or more from level 1 (+level 2 D*)
	Response to steroid	Indeterminate	Level 1 S/OOI + Rt or level 1 D + level 2 S/OOI/H + Rt
Probable type 1 AIP		Indeterminate	Level 2 S/OOI/H + Rt

*Level 2 D is counted as level 1 in this setting.

TABLE 2. Level 1 and Level 2 Criteria for Type 1 AIP

	Criterion	Level 1	Level 2
P	Parenchymal imaging	Typical: Diffuse enlargement with delayed enhancement (sometimes associated with rim-like enhancement)	Indeterminate (including atypical[†]): Segmental/focal enlargement with delayed enhancement
D	Ductal imaging (ERP)	Long (>1/3 length of the main pancreatic duct) or multiple strictures without marked upstream dilatation	Segmental/focal narrowing without marked upstream dilatation (duct size, <5 mm)
S	Serology	IgG4, >2× upper limit of normal value	IgG4, 1–2× upper limit of normal value
OOI	Other organ involvement	a or b	a or b
		a. Histology of extrapancreatic organs Any three of the following: (1) Marked lymphoplasmacytic infiltration with fibrosis and without granulocytic infiltration (2) Storiform fibrosis (3) Obliterative phlebitis (4) Abundant (>10 cells/HPF) IgG4-positive cells	a. Histology of extrapancreatic organs including endoscopic biopsies of bile duct[‡]: Both of the following: (1) Marked lymphoplasmacytic infiltration without granulocytic infiltration (2) Abundant (>10 cells/HPF) IgG4-positive cells
		b. Typical radiological evidence At least one of the following: (1) Segmental/multiple proximal (hilar/intrahepatic) or proximal and distal bile duct stricture (2) Retroperitoneal fibrosis	b. Physical or radiological evidence At least one of the following: (1) Symmetrically enlarged salivary/lachrymal glands (2) Radiological evidence of renal involvement described in association with AIP
H	Histology of the pancreas	LPSP (core biopsy/resection) At least 3 of the following: (1) Periductal lymphoplasmacytic infiltrate without granulocytic infiltration (2) Obliterative phlebitis (3) Storiform fibrosis (4) Abundant (>10 cells/HPF) IgG4-positive cells	LPSP (core biopsy) Any 2 of the following: (1) Periductal lymphoplasmacytic infiltrate without granulocytic infiltration (2) Obliterative phlebitis (3) Storiform fibrosis (4) Abundant (>10 cells/HPF) IgG4-positive cells
		Diagnostic steroid trial	
	Response to steroid (Rt)*	Rapid (≤2 wk) radiologically demonstrable resolution or marked improvement in pancreatic/extrapancreatic manifestations	

*Diagnostic steroid trial should be conducted carefully by pancreatologists with caveats (see text) only after negative workup for cancer including endoscopic ultrasound-guided fine needle aspiration.

[†]Atypical: Some AIP cases may show low-density mass, pancreatic ductal dilatation, or distal atrophy. Such atypical imaging findings in patients with obstructive jaundice and/or pancreatic mass are highly suggestive of pancreatic cancer. Such patients should be managed as pancreatic cancer unless there is strong collateral evidence for AIP, and a thorough workup for cancer is negative (see algorithm).

[‡]Endoscopic biopsy of duodenal papilla is a useful adjunctive method because ampulla often is involved pathologically in AIP.

Diagnosis of AIP in the burnt out stage is not easy and is not within the scope of these diagnostic criteria.

Presentations Not Suggestive of AIP

Marked cachexia, inability to eat, and narcotic requiring pain are more suggestive of cancer and are rarely seen in AIP. Although patients with typical acute presentation may concomitantly meet criteria for pancreatitis (2 of the following 3: 3-fold elevated pancreatic enzymes, abdominal pain, or computed tomography (CT) evidence of pancreatic swelling), typical idiopathic pancreatitis or painful chronic pancreatitis are not commonly seen in histologically confirmed AIP.

Cardinal Features of AIP

As previously noted, AIP and its subtypes have a histopathological pattern that is diagnostic. However, histology is not usually available. Therefore, the diagnosis of AIP requires a

TABLE 3. Diagnosis of Definitive and Probable Type 2 AIP Using ICDC

Diagnosis	Imaging Evidence	Collateral Evidence
Definitive type 2 AIP	Typical/indeterminate	Histologically confirmed IDCP (level 1 H) or clinical inflammatory bowel disease + level 2 H + Rt
Probable type 2 AIP	Typical/indeterminate	Level 2 H/clinical inflammatory bowel disease + Rt

TABLE 4. Diagnosis of AIP–Not Otherwise Specified Using ICDC

Diagnosis	Imaging Evidence	Collateral Evidence (Case With Only D1/2)
AIP–not otherwise specified	Typical/indeterminate	D1/2 + Rt

combination of features. The existing diagnostic criteria have used a combination of 1 or more of 5 cardinal features of AIP:

1. imaging features of the following: (*a*) pancreatic parenchyma (on CT/magnetic resonance imaging [MRI]) and (*b*) pancreatic duct (endoscopic retrograde cholangiopancreatography or magnetic resonance cholangiopancreatography),
2. serology (IgG4, IgG, and antinuclear antibody),
3. OOI,
4. histopathology of the pancreas, and
5. response to steroid therapy.

Distinguishing Type 1 From Type 2 Using Diagnostic Criteria

1. Based on our current knowledge, it does not seem that imaging and response to steroids can distinguish type 1 from type 2 (Tables 1–5).
2. Typical serological abnormalities and OOI are seen only in type 1. Inflammatory bowel disease seems to be associated with both forms, more so with type 2.
3. Absence of serological abnormalities or lack of OOI in patients with "AIP" does not necessarily imply the diagnosis of type 2, as type 1 also can be seronegative and without OOI.
4. For the purposes of the development of criteria, "definitive type 1" can be diagnosed with surrogate criteria for AIP not including histology. However, definite IDCP requires histological confirmation.
5. Autoimmune pancreatitis that does not meet these criteria, including histologically unconfirmed but clinically suspected type 2, is categorized as "probable AIP."

Imaging: Probability of AIP Versus Pancreatic Cancer Varies Depending on Pancreatic Imaging Findings on CT/MRI

Pancreatic findings on abdominal CT or MRI often are the first clues that raise the suspicion of pancreatic cancer or AIP (Tables 2 and 5). However, AIP is uncommon compared with pancreatic cancer. Before AIP was recognized as a clinical entity, only 2% to 3% of patients undergoing resection for suspected pancreatic cancer had AIP. However, the probability of AIP versus pancreatic cancer in patients with obstructive jaundice can be predicted based on CT/MRI findings:

1. Patients with obstructive jaundice with a diffusely enlarged pancreas (especially with a capsule-like rim) without pancreatic ductal dilatation/cutoff or pancreatic low density mass on CT/MRI are highly likely to have AIP. In such patients, presence of less collateral evidence is required to make the diagnosis of AIP.
2. Subjects with findings typical of pancreatic cancer (low density mass on contrast-enhanced CT, pancreatic ductal dilatation/cutoff with or without pancreatic atrophy) should be considered as having pancreatic cancer unless the workup for cancer is negative and there is strong collateral evidence of AIP.
3. Subjects without features typical of AIP or pancreatic cancer should first be investigated for pancreatic cancer, and AIP should be considered only if workup for cancer is negative and there is strong collateral evidence of AIP.

Pancreatographic Findings

When expert physicians read pancreatograms, they can distinguish AIP from pancreatic cancer, with some features

TABLE 5. Level 1 and Level 2 Criteria for Type 2 AIP

	Criterion	Level 1	Level 2
P	Parenchymal imaging	Typical: Diffuse enlargement with delayed enhancement (sometimes associated with rim-like enhancement)	Indeterminate (including atypical†): Segmental/focal enlargement with delayed enhancement
D	Ductal imaging (ERP)	Long (>1/3 length of the main pancreatic duct) or multiple strictures without marked upstream dilatation	Segmental/focal narrowing without marked upstream dilatation (duct size, <5 mm)
OOI	Other organ involvement		Clinically diagnosed inflammatory bowel disease
H	Histology of the pancreas (core biopsy/resection)	IDCP: Both of the following: (1) Granulocytic infiltration of duct wall (GEL) with or without granulocytic acinar inflammation (2) Absent or scant (0–10 cells/HPF) IgG4-positive cells	 Both of the following: (1) Granulocytic and lymphoplasmacytic acinar infiltrate (2) Absent or scant (0–10 cells/HPF) IgG4-positive cells
		Diagnostic steroid trial	
Response to steroid (Rt)*		Rapid (≤2 wk) radiologically demonstrable resolution or marked improvement in manifestations	

*Diagnostic steroid trial should be conducted carefully by pancreatologists with caveats (see text) only after negative workup for cancer including endoscopic ultrasound-guided fine needle aspiration.

†Atypical: Some AIP cases may show low-density mass, pancreatic ductal dilatation, or distal atrophy. Such atypical imaging findings in patients with obstructive jaundice and/or pancreatic mass are highly suggestive of pancreatic cancer. Such patients should be managed as pancreatic cancer unless there is strong collateral evidence for AIP, and a thorough workup for cancer is negative (see algorithm).

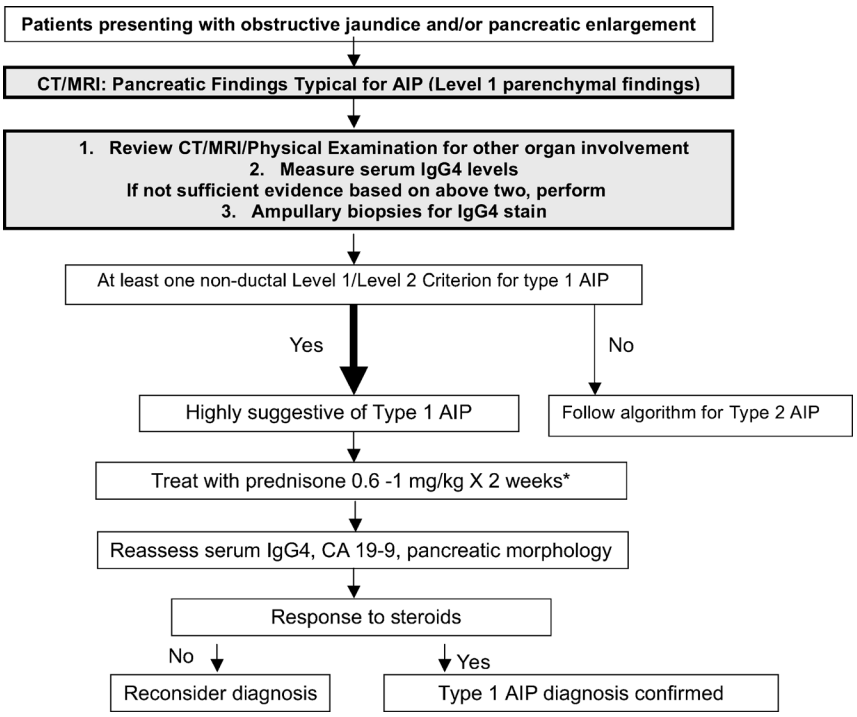

FIGURE 1. Algorithm to diagnose type 1 AIP in subjects presenting with obstructive jaundice and/or pancreatic enlargement. This schematic drawing shows a flow to diagnose type 1 AIP with typical diffuse enlargement of the pancreas on CT/MRI (level 1 parenchymal findings).

being more helpful than others (Tables 2 and 5). Thus, a combination of typical imaging and pancreatographic findings is highly suspicious for AIP. However, in the West, diagnostic pancreatograms are rarely performed in the setting of obstructive jaundice. Therefore, in the West, a diagnostic pancreatogram may assume the role of collateral evidence when

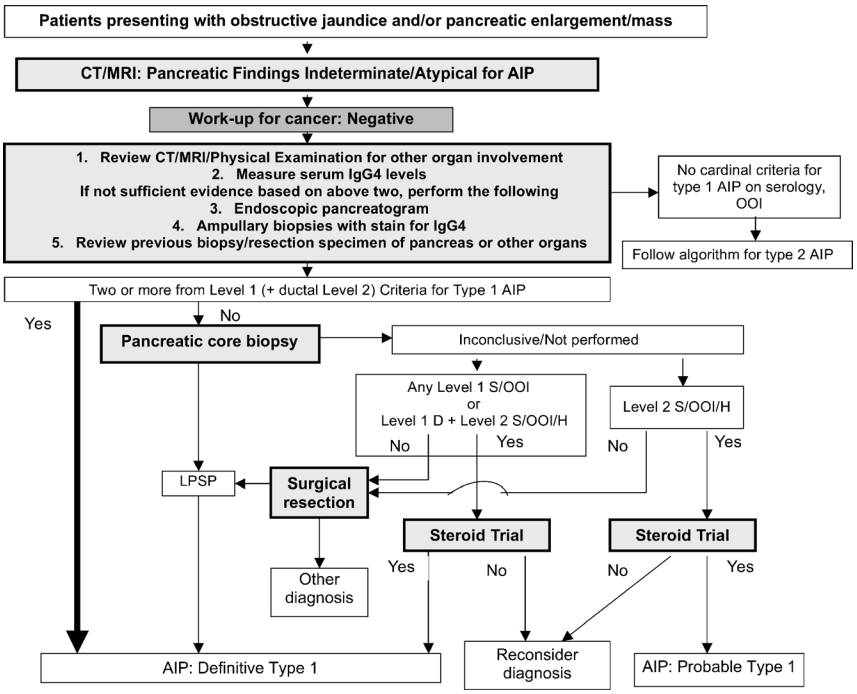

FIGURE 2. Algorithm to diagnose type 1 AIP in subjects presenting with obstructive jaundice and/or pancreatic mass. This schematic drawing shows a flow to diagnose type 1 AIP with indeterminate or atypical findings of the pancreas on CT/MRI (level 2 parenchymal findings).

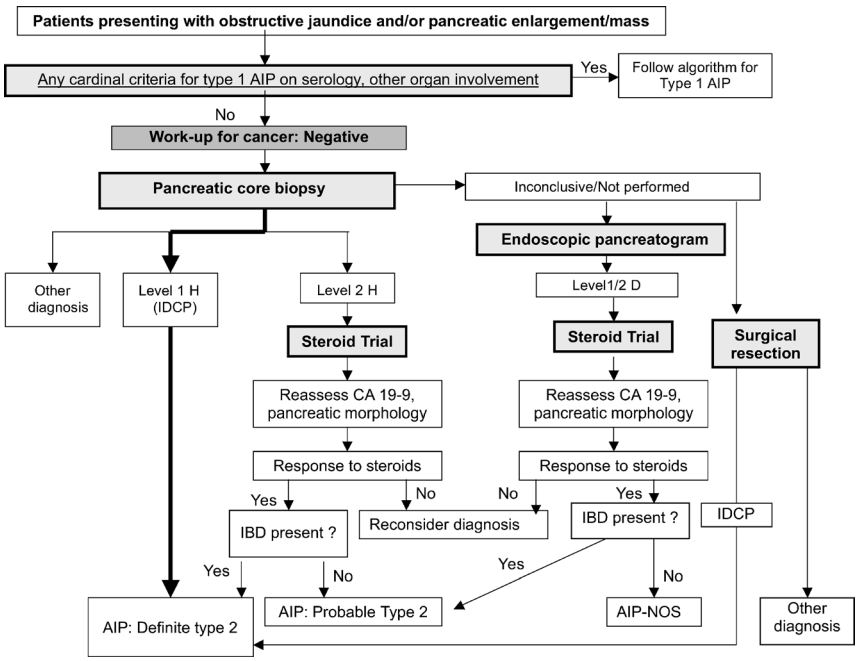

FIGURE 3. Algorithm to diagnose type 2 AIP in subjects presenting with obstructive jaundice and/or pancreatic mass. This schematic drawing shows a flow to diagnose type 2 AIP with typical/indeterminate (atypical) findings of the pancreas on CT/MRI (level 1 and 2 parenchymal findings).

imaging findings are not typical or in seronegative patients without OOI.

Serology

1. Elevation in any one or more IgG, IgG4, and antinuclear antibody titers is commonly seen in AIP. However, false-positive elevation of each of these markers has been seen in pancreatic cancer and other disorders. When all markers are measured, the false positivity of the panel can be significantly high (up to 40%) (Table 2).
2. Serum IgG4 elevation is the single best marker of AIP. Therefore, in the ICDC for AIP, only serum IgG4 is recommended as a serological marker. Because the upper limits of normal IgG4 vary between laboratories, only fold elevation above normal rather than absolute value is used in the ICDC.
3. Marked elevation of serum IgG4 (>2 times upper limit of normal) is strongly suggestive of AIP in the setting of obstructive jaundice/pancreatic mass.
4. Elevation in serological markers is not sufficient to make a diagnosis of AIP unless seen in the setting of typical imaging finding.

Other Organ Involvement

1. Other organ involvement noted here is part of the manifestation of IgG4-related systemic disease. Other unrelated autoimmune disorders (eg, rheumatoid arthritis, psoriatic arthropathy, true seropositive Sjögren) are not typically associated with AIP and should not be included as OOI (Tables 2 and 5).
2. Other organ involvement may be diagnosed by histological evaluation of tissue, by imaging (proximal bile duct stricture, retroperitoneal fibrosis) or by clinical examination (salivary gland enlargement).

Histology of the Pancreas: Role in the Diagnosis of AIP

1. As mentioned earlier, type 1 often can be diagnosed without histology, but type 2 requires an adequate histological specimen to make a definitive diagnosis (Tables 2 and 5).
2. Autoimmune pancreatitis can be diagnosed on resection or biopsy specimen, provided the specimen displays the characteristic features.
3. Biopsy showing some but not all features of LPSP or IDCP (lymphoplasmacytic infiltrate with storiform fibrosis) can be used as supportive evidence for diagnosis of AIP.

Response to Steroids

1. A steroid trial involves use of prednisone 0.6 to 1 mg/kg with reassessment of imaging and Ca 19-9 after 2 weeks of treatment (Tables 2 and 5). The panel agreed on 2 weeks for duration of steroid trial based on the study by Moon et al,[16] the only study which has examined this issue. In AIP, one would expect a definitive improvement in imaging abnormalities, including biliary strictures and pancreatic enlargement. In the study by Moon et al, no patient with pancreatic cancer showed "response" to steroids. In AIP, CA 19-9 levels drop with treatment, and a rising CA 19-9 suggests that the diagnosis of AIP is incorrect. Resolution of imaging abnormalities may take weeks to months and, in some, is associated with atrophy of gland.
2. In patients with appropriate collateral evidence of AIP, response to steroids can confirm a strong suspicion of AIP. However, steroid trial as a means to diagnose AIP is to be used sparingly and should not be used as a substitute for a thorough search for an etiology.
3. The strategy that "if steroids work, it must be AIP" is fraught with problems when there is no collateral evidence to support the diagnosis.

4. Symptomatic improvement and a sense of well-being occur nonspecifically in response to steroids and can be seen even in pancreatic cancer patients. Therefore, these parameters should not be used to assess response.
5. Steroid therapy leads to reduction in IgG4 levels in AIP. However, falsely elevated IgG4 in pancreatic cancer and other non-AIP states also can decrease with steroid therapy. Therefore, a "response" of IgG4 to steroid treatment cannot be used to diagnose AIP.
6. Spontaneous radiological improvement in pancreatic cancer-induced pancreatitis can be mistaken for steroid response.

The consensus diagnostic criteria (Tables 1–5) and diagnostic algorithms (Figs. 1–3) are an effort of Eastern and Western experts to find common bases for diagnosis of AIP worldwide. They are inclusive of practice patterns in different countries. They can be tailored for use in individual institutions depending on local expertise.

These diagnostic criteria are meant to recognize the spectrum of AIP as we know it today. The panel recognizes that the spectrum of AIP may extend beyond our current understanding of the disease (after all, it has rapidly expanded in the past 10 years). However, future "extensions" of the spectrum of AIP or better allocation into the 2 groups will have to be based on histologically confirmed cases and determination of novel serum markers[17,18] rather than response to steroids.

CONCLUSIONS

This ICDC for AIP was developed by a panel of experts during the International Association of Pancreatology held in Fukuoka, Japan. This consensus guidelines was based on the recognition that AIP has 2 distinct histopathology and clinical subtypes, previously agreed upon by the consensus meeting held in Honolulu during the joint meeting of the Japan Pancreas Society and the American Pancreatic Association.[4] We look forward to the further input by the health care providers and professional groups in using these guidelines in the clinical management of AIP and for further updating these guidelines.

REFERENCES

1. Okazaki K, Kawa S, Kamisawa T, et al. Clinical diagnostic criteria of autoimmune pancreatitis: revised proposal. *J Gastroenterol*. 2006;41:626–631.
2. Members of the Criteria Committee for Autoimmune Pancreatitis of the Japan Pancreas Society. Diagnostic criteria for autoimmune pancreatitis by the Japan Pancreas Society (2002). *J Jpn Pancreas Soc (Suizou)*. 2002;17:585–587.
3. Levy MJ, Reddy RP, Wiersema MJ, et al. EUS-guided trucut biopsy in establishing autoimmune pancreatitis as the cause of obstructive jaundice. *Gastrointest Endosc*. 2005;61:467–472.
4. Chari ST, Kloeppel G, Zhang L, et al; The Autoimmune Pancreatitis International Cooperative Study Group (APICS). Histopathologic and clinical subtypes of autoimmune pancreatitis: the honolulu consensus document. *Pancreas*. 2010;39:549–554.
5. Sugumar A, Klöppel G, Chari ST. Autoimmune pancreatitis: pathologic subtypes and their implications for its diagnosis. *Am J Gastroentel*. 2009;104:2308–2310.
6. Notohara K, Burgart LJ, Yadav D, et al. Idiopathic chronic pancreatitis with periductal lympho-plasmacytic infiltration: clinico-pathologic features of 35 cases. *Am J Surg Pathol*. 2003;27:1119–1127.
7. Zamboni G, Lüttges J, Capelli P, et al. Histopathological features of diagnostic and clinical relevance in autoimmune pancreatitis: a study on 53 resection specimens and 9 biopsy specimens. *Virchows Arch*. 2004;445:552–563.
8. Klöppel G, Detlefsen S, Chari ST, et al. Autoimmune pancreatitis: the clinicopathological characteristics of the subtype with granulocytic epithelial lesions. *J Gastroenterol*. 2010;45:787–793.
9. Park DH, Kim MH, Chari ST. Recent advances in autoimmune pancreatitis. *Gut*. 2008;58:1680–1689.
10. Chari ST, Smyrk TC, Levy MJ, et al. Diagnosis of autoimmune pancreatitis: the Mayo Clinic experience. *Clin Gastroenterol Hepatol*. 2006;4:1010–1016.
11. Chari ST, Takahashi N, Levy MJ, et al. A diagnostic strategy to distinguish autoimmune pancreatitis from pancreatic cancer. *Clin Gastroenterol Hepatol*. 2009;7:1097–1103.
12. Kwon S, Kim MH, Choi EK. The diagnostic criteria for autoimmune chronic pancreatitis: it is time to make a consensus. *Pancreas*. 2007;34:279–286.
13. Otsuki M, Chung JB, Okazaki K, et al. Asian diagnostic criteria for autoimmune pancreatitis: consensus of the Japan-Korea Symposium on Autoimmune Pancreatitis. *J Gastroenterol*. 2008;43:403–408.
14. Schneider A, Löhr JM. [Autoimmune pancreatitis]. *Internist (Berl)*. 2009;50:318–330.
15. Pearson RK, Longnecker DS, Chari ST, et al. Controversies in clinical pancreatology: autoimmune pancreatitis: does it exist? *Pancreas*. 2003;27:1–13.
16. Moon S-H, Kim MH, Park DH, et al. Is a 2-week steroid trial after initial negative investigation for malignancy useful in differentiating autoimmune pancreatitis from pancreatic cancer? A prospective outcome study. *Gut*. 2008;57:1704–1712.
17. Frulloni L, Lunardi C, Simone R, et al. Identification of a novel antibody associated with autoimmune pancreatitis. *N Engl J Med*. 2009;361:2135–2142.
18. Löhr JM, Faissner R, Koczan D, et al. Autoantibodies against the exocrine pancreas in autoimmune pancreatitis: gene and protein expression profiling and immunoassays identify pancreatic enzymes as a major target of the inflammatory process. *Am J Gastroenterol*. 2010;105:2060–2071.