

Supplementary Table 1: Definitions of terms used in the study

Focal enlargement	<i>area of the pancreatic parenchyma with a different density compared with that of the surrounding tissue.</i>
Diffuse enlargement	<i>swelling of the pancreas in the absence of a discrete mass</i>
Other Organ Involvement (OOI)	<i>presence of extra-pancreatic organ involvement, such as biliary tract, retroperitoneum, kidney, and salivary/ lacrimal glands</i>
Remission	<i>disappearance of clinical symptoms, and resolution of the pancreatic and/or extrapancreatic manifestations on imaging studies</i>
Relapse	<i>recurrence of symptoms with the development/reappearance of pancreatic and/or extrapancreatic abnormalities on imaging studies</i>
Tapering	<i>progressive reduction of steroid dosage after induction therapy</i>
Timing of diabetes mellitus development	<i>in patients with diabetes mellitus, the timing of initial glucose intolerance and its relationship with the diagnosis of AIP was evaluated</i>
Steroid trial	<i>the trial had a diagnostic purpose. It was considered positive if a rapid (2 weeks) radiologically demonstrable resolution or marked improvement in pancreatic/extrapancreatic manifestations after 0.6-1 mg/kg/day of prednisolone were observed</i>

Supplementary Table 2: Features found at diagnostic imaging

		Overall (n=173)	Type 1 AIP[§] (n=106)	Type 2 AIP[§] (n=48)	NOS°-AIP[§] (n=19)	Type1 vs Type2	
Ultrasonography		115 (66.5)	69 (65.1)	29 (60.4)	17 (89.5)	0.58	
	Hypoechoic pancreatic enlargement	99 (86.1)	59 (85.5)	25 (86.2)	15 (88.2)	0.92	
		<i>Focal</i>	71 (61.7)	40 (58)	19 (65.5)	12 (70.6)	0.43
		<i>Diffuse</i>	28 (24.3)	19 (27.5)	6 (20.7)	3 (17.6)	0.42
	Main pancreatic duct dilatation	15 (13.0)	11 (15.9)	3 (10.3)	1 (5.9)	0.47	
CT* scan		145 (83.8)	93 (87.7)	36 (75.0)	16 (84.2)	0.05	
	Pancreatic enlargement	124 (85.5)	80 (86.0)	29 (80.6)	15 (93.7)	0.45	
		<i>Focal</i>	86 (67.7)	51 (63.7)	23 (79.3)	12 (80.0)	0.12
		<i>Diffuse</i>	38 (30.7)	29 (36.3)	6 (20.7)	3 (20.0)	0.12
	Main pancreatic duct dilatation	23 (15.9)	12 (12.9)	5 (13.9)	6 (37.5)	0.88	
	Capsule-like rim	20 (13.8)	10 (10.7)	6 (16.7)	4 (25.0)	0.35	
	Contrast enhancement of pancreas	20 (13.8)	15 (16.1)	4 (11.1)	1 (6.2)	0.47	
	Cystic lesions	6 (4.1)	4 (4.3)	2 (5.6)	0	0.75	
MRI[#]		108 (62.4)	67 (63.2)	31 (64.6)	10 (52.6)	0.86	
	Pancreatic enlargement	93 (86.1)	57 (85.1)	26 (83.9)	10 (100.0)	0.88	
		<i>Focal</i>	56 (60.2)	29 (50.9)	19 (73.1)	8 (80.0)	0.05
		<i>Diffuse</i>	37 (39.8)	28 (49.1)	7 (26.9)	2 (20.0)	0.05
	Main pancreatic duct stenosis	17 (15.7)	13 (19.4)	2 (6.4)	2 (20.0)	0.09	
	Main pancreatic duct dilatation	16 (14.8)	9 (13.4)	4 (12.9)	3 (30.0)	0.95	
	Capsule-like rim	14 (13.0)	9 (13.4)	2 (6.4)	3 (30.0)	0.3	
	Contrast enhancement of pancreas	14 (13.0)	11 (16.4)	3 (9.7)	0	0.38	
	Cystic lesions	25 (23.1)	14 (20.9)	8 (25.8)	3 (30.0)	0.59	
EUS°		118 (68.2)	75 (70.6)	28 (58.3)	15 (78.9)	0.13	
	Hypoechoic pancreatic enlargement	106 (89.8)	69 (92.0)	22 (78.6)	15 (100.0)	0.06	
		<i>Focal</i>	73 (68.9)	45 (65.2)	15 (68.2)	13 (86.7)	0.79
		<i>Diffuse</i>	33 (31.1)	24 (34.8)	7 (31.8)	2 (13.3)	0.79
	Hyperechoic Foci	33 (28.0)	22 (29.3)	7 (25.0)	4 (26.7)	0.66	
	Lobularity	44 (37.3)	26 (34.7)	10 (35.7)	8 (53.3)	0.92	
	Main pancreatic duct stenosis	11 (9.3)	5 (6.6)	4 (14.3)	2 (13.3)	0.21	
		<i>Single</i>	8 (72.7)	3 (60.0)	4 (100.0)	1 (50.0)	0.18
		<i>Multiple</i>	3 (27.3)	2 (40.0)	0	1 (50.0)	0.18
	Thickening Wirsung wall	20 (16.9)	12 (16.0)	7 (25.0)	1 (6.7)	0.29	
	Hyperechoic Wirsung wall	12 (10.2)	11 (14.7)	1 (3.6)	0	0.12	
	Thickening common bile duct wall >2mm	27 (22.9)	17 (22.7)	5 (17.9)	5 (33.3)	0.6	
		<i>Focal</i>	15 (55.6)	8 (47.1)	4 (80.0)	3 (60.0)	0.20
		<i>Diffuse</i>	12 (44.4)	9 (52.9)	1 (20.0)	2 (40.0)	0.20
	Cystic lesions	5 (4.2)	2 (2.7)	1 (3.6)	2 (13.3)	0.81	
	CE-EUS°	32 (27.1)	25 (33.3)	5 (17.9)	2 (13.3)	0.13	
		<i>Hypoenhancement</i>	6 (18.7)	4 (16.0)	2 (40.0)	0	0.23
		<i>Isoenhancement</i>	14 (43.7)	12 (48.0)	2 (40.0)	0	0.75
		<i>Hyperenhancement</i>	12 (37.5)	9 (36.0)	1 (20.0)	2 (100.0)	0.49

[§]AIP: Autoimmune pancreatitis; [°]NOS: Not Otherwise Specified; ^{*}CT: Computed Tomography; [#]MRI: Magnetic Resonance Imaging;
[°](CE)EUS: (Contrast Enhanced) Endoscopic Ultrasound; [†]ERCP: Endoscopic Retrograde Cholangiopancreatography

Supplementary Table 3: IgG4 values in the entire cohort of patients, and in AIP1 and AIP2 subgroups

	Absolute median value (mg/dL)	N° of patients with IgG4 value above UNR* (n)	N° of patients with IgG4 value two times above UNR* (n)	Difference in IgG4 absolute median value AIP1 vs AIP2
Overall	177.5 (IQR° 70-402)	78/140	46/140	
Type 1 AIP[§]	234 (IQR° 133-567)	62/84	40/84	
Type 2 AIP[§]	87 (IQR° 25-161)	11/40	5/40	<i>p= 0.03</i>

[§]AIP: Autoimmune pancreatitis; °IQR: Interquartile Range; *UNR: Upper Normal Range