

Table S1

## Univariate and multivariate analysis for severe and moderate groups

Severe group						
			Univariate		Multivariable	
	Pts.	Events	20 years survival (95% C.I.)	p	HR (95% C.I.)	p
Method of diagnosis						
NBS	95	23	84.93 (74.29, 91.41)	0.007	1.00	0.01
Symptoms	34	21	63.64 (44.95, 77.46)		2.23 (1.23-4.03)	
Gender						
M	71	27	70.70 (57.49-80.49)	0.40		
F	58	17	88.58 (74.50-95.13)			
Pancreatic status						
PI	115	43	77.99 (68.45-84.96)	0.18		ns
PS	14	1	83.33 (27.31-97.47)			
Birth cohort						
1971-1990	91	40	77.36 (67.13-84.76)	0.71		
1991-2014	38	4	85.57 (60.14-95.33)			
Moderate group						
			Univariate		Multivariable	
	Pts.	Events	20 years survival (95% C.I.)	p	HR (95% C.I.)	p
Method of diagnosis						
NBS	80	4	94.48 (83.72, 98.20)	0.016	1.00	0.02
Symptoms	40	12	85.93 (69.41, 93.89)		3.86 (1.21-12.38)	
Gender						
M	58	4	95.47 (82.99-98.85)	0.06		
F	62	12	87.99 (75.03-94.46)			
Pancreatic status						
PI	96	16	89.51 (94.64-100.00)	0.03		ns
PS	24	0	100.00			
Birth cohort						
1971-1990	68	15	89.55 (79.33-94.88)	0.2		
1991-2014	52	1	97.06 (80.90-99.58)			