

## Diagnostic criteria for IgG4-related disease (IgG4-RD)

Reproduced without modification from Pieringer H. et al., Orphanet J. Rare Dis. 2014; 9:110 (accessible at <http://www.ojrd.com/content/9/1/110/abstract>)

Diagnostic criteria for IgG4-RD (modified after Umehara et al. (18))

1	+	2	+	3	=	Diagnosis of IgG4-RD
Organ involvement: dysfunction, localized or diffuse swelling		Serum IgG4 >135mg/dl		Histopathology: IgG4/IgG Ratio >0.4 and >10 IgG4+ cells per HPF		Definite
Organ specific criteria for IgG4-RD (e.g. AIP, Mikulicz' disease)						Definite
Organ involvement: dysfunction, localized or diffuse swelling		Serum IgG4 <135mg/dl		Histopathology: IgG4/IgG Ratio >0.4 and >10 IgG4+ cells per HPF		Probable
Organ involvement: dysfunction, localized or diffuse swelling		Serum IgG4 >135mg/dl		Histopathology: Not available or not diagnostic		Possible
Organ involvement: dysfunction, localized or diffuse swelling		Serum IgG4 <135mg/dl		Histopathology: Not available or not diagnostic		No

### Notes:

AIP: autoimmune pancreatitis

HPF: high power field

Mikulicz' disease: IgG4-related dacryoadenitis and sialoadenitis