

The diagnostic criteria for IgG4-Related Disease (IgG4RD)¹:

1. A clinical examination showing diffuse/localized swelling or masses in single or multiple organs.
 2. Elevated serum IgG4 concentration (≥ 135 mg/dL).
 3. Histopathology showing:
 - a. Marked lymphocyte infiltration and storieform fibrosis
 - b. Infiltration of IgG4 plasma cells: ratio of IgG4 to IgG cells $>40\%$ and >10 IgG4 plasma cells/hpf
- With all 3 criteria = definite IgG4RD
 - With criteria 1+3 = probable IgG4RD
 - With criteria 1+2 = possible IgG4RD

¹ Umehara H, Okazaki K, Masaki Y, Kawano M, Yamamoto M, Saeki T, Matsui S, Yoshino T, Nakamura S, Kawa S, Hamano H, Kamisawa T, Shimosegawa T, Shimatsu A, Nakamura S, Ito T, Notohara K, Sumida T, Tanaka Y, Mimori T, Chiba T, Mishima M, Hibi T, Tsubouchi H, Inui K, Ohara H. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. *Mod Rheumatol*. 2012 Feb;22(1):21-30. doi: 10.1007/s10165-011-0571-z. Epub 2012 Jan 5. PMID: 22218969.