Supplementary Table 3. Definitions of common variable immunodeficiency.

ESID/PAGID	ESID diagnostic criteria	IUIS criteria	WHO scientific group	ICON
Probable Marked decrease (≥2 SD below mean for age) in serum IgG and IgA and: 1.Onset of immunodeficiency ≥2 years of age 2.Absent isohemagglutinins and/or poor response to vaccines 3.Defined causes of hypogammaglobulinemia have been excluded Possible Marked decrease (≥2 SD below mean for age) in one of the major isotypes (IgM, IgG, and IgA) and: 1.Onset of immunodeficiency ≥2 years of age 2.Absent isohemagglutinins and/or poor response to vaccines 3.Defined causes of hypogammaglobulinemia have been excluded	At least one of the following: -increased susceptibility to infection -autoimmune manifestations -granulomatous disease -unexplained polyclonal lymphoproliferation -affected family member with antibody deficiency AND marked decrease of IgG and IgA with or without low IgM levels; *AND* at least one of the following: -poor antibody response to vaccines -low switched memory B cells (<70% of age-related normal value) AND secondary causes have been excluded (e.g., infection, protein loss, medication, malignancy)	Decreased serum levels of IgG, IgA, and/or IgM (≥2 SD below mean for age)	Decreased serum levels of IgG, IgA, and/or IgM (≥2 SD below mean for age)	Decreased serum levels of IgG, IgA, and/or IgM (≥2 SD below mean for age), impairment of specific antibody responses, and, occasionally, reductions in B-cell numbers

Abbreviations: ESID, European Society for Immunodeficiencies; ICON, International Consensus Document; IUIS, International Union of Immunological Societies; PAGID, Pan-American Group for Immunodeficiency; WHO, World Health Organization.