SHAREABLE PDF

Interstitial lung disease in primary immunodeficiency: towards a brighter future

John R. Hurst[1] and Klaus Warnatz[2,3], on behalf of the ERS eGLILDnet Clinical Research Collaboration[4]

Affiliations: [1]UCL Respiratory, University College London, London, UK. [2]Dept of Rheumatology and Clinical Immunology, Medical Center - University of Freiburg, Faculty of Medicine, University of Freiburg, Freiburg, Germany. [3]Center for Chronic Immunodeficiency, Medical Center - University of Freiburg, Faculty of Medicine, University of Freiburg, Freiburg, Germany. [4]A list of the ERS eGLILDnet Clinical Research Collaborators can be found in the acknowledgements section.

Correspondence: John R. Hurst, University College London, Centre for Inflammation and Tissue Repair, UCL Respiratory Medicine, London, NW3 2PF, UK. E-mail: j.hurst@ucl.ac.uk

@ERSpublications
The ERS eGLILDnet CRC is working for better care and research for people affected by GLILD http://bit.ly/37WZGdd

Cite this article as: Hurst JR, Warnatz K. Interstitial lung disease in primary immunodeficiency: towards a brighter future. *Eur Respir J* 2020; 55: 2000089 [https://doi.org/10.1183/13993003.00089-2020].

Lung disease is a frequent clinical manifestation in people living with primary immunodeficiency diseases, the most prevalent of which are common variable immunodeficiency disorders (CVID). CVID is primarily characterised by antibody deficiency, but recent definitions and diagnostic criteria recognise a much more complex pattern of immunological defects [1]. CVID can be classified into two major clinical phenotypes. One group experiences infection as the only major clinical manifestation, whilst the other present a variety of lymphoproliferative, inflammatory and/or autoimmune complications. The most frequent consequences in the lung of CVID are acute infections, and secondary airway complications of infection, such as bronchiectasis. However, up to 15% of patients with CVID develop an interstitial lung disease [2, 3]. Infections and bronchiectasis are primarily driven by antibody deficiency, but CVID associated interstitial lung disease (CVID-ILD) is best considered part of a systemic immune dysregulatory process [4] such that people with CVID-ILD often have splenomegaly, lymphadenopathy and autoimmune cytopenias [5–7]. With an EU population of 747 million, we estimate there are up to 30 000 people living with CVID in Europe, and thus 4500 with CVID-ILD. Whilst people with "infection only" CVID can now expect a near normal life expectancy [8], those with systemic immune dysregulation including CVID-ILD often have a much more complicated course. CVID-ILD increases morbidity and mortality in CVID [9], although the outcome is now recognised to be more variable than originally reported [10].