

Primary immunodeficiency is a cause of serious disability both in the paediatric and the adult patient

To whom it may concern,

Primary immunodeficiencies (or PIDs) are chronic and rare diseases caused by the absence or malfunctioning of components of the immune system. Most PIDs cannot be cured and require long-term treatment and follow-up¹. These conditions have a major impact on the patients' life and wellbeing. Multiple studies confirm that PID patients have significantly lower physical health and abilities compared to the general population, and suffer from reduced psychosocial wellbeing and overall quality of life (QoL) measures²⁻⁴. Life-long high-quality multidisciplinary management is needed to improve PID patients' clinical status and general health, as to provide them with the best possible life expectancy and QoL¹.

A global survey of specialist clinicians revealed that both adult and paediatric PID patients are believed to have a worse overall QoL (adults bearing an even worse health-related QoL compared to paediatric PID cases) and to be more severely affected by PID related co-morbidities⁵. Adult PID patients typically experience a long diagnostic delay, which increases the risk of permanent organ damage and physical disability⁵. Severe or even life-threatening health conditions are very common (>85%) in adults with PID, and these patients have an unusually high risk of developing multiple conditions and/or cancer³. Evidently, the heavy burden of their condition negatively impacts adult PID patients' social functioning and ability to work^{3,5}.

While individual patient assessment is naturally required, many adults with PID fit the definition of persons with disabilities as determined by the UN Convention on the Rights of Persons with Disabilities. This defines persons with disabilities as *'those who have long-term physical, mental, intellectual or sensory impairments which in interaction with various barriers may hinder their full and effective participation in society on an equal basis with others'*⁷. Therefore, if PID-related illness is causing serious long-term impairment it is to be recognised as a disability, with such patients being protected under law from unfair treatment relating to their medical condition. These patients are thus eligible for long-term disability support and/or any relevant financial or practical support needed to facilitate access to work. Moreover, patients should always be offered the opportunity to meet a medico-social professional to ensure they get the best level of information and access to their rights.

This statement was prepared by the International Patient Organisation for Primary Immunodeficiencies (IPOPI) and is endorsed by (in alphabetical order): the Asia Pacific Society for Immunodeficiencies (APSID), the Arab Society for Primary Immunodeficiencies (ARAPID), the African Society for Immunodeficiencies (ASID), the Clinical Immunology Society (CIS), the European Society for Immunodeficiencies (ESID), the International Nursing Group for Immunodeficiencies (INGID), the Latin American Society for Immunodeficiencies (LASID) and the South East Asia Primary Immunodeficiency Network (SEAPID).

References:

1. Chapel, H. *et al.* Primary Immune Deficiencies – Principles of Care. *Front. Immunol.* 5, (2014).
2. Ridao-Manonellas, S. *et al.* Health-Related Quality of Life and Multidimensional Fatigue Scale in Children with Primary Immunodeficiencies. *J. Clin. Immunol.* 40, 602–609 (2020).
3. Barlogis, V. *et al.* Physical health conditions and quality of life in adults with primary immunodeficiency diagnosed during childhood: A French Reference Center for PIDs (CEREDIH) study. *J. Allergy Clin. Immunol.* 139, 1275-1281.e7 (2017).
4. Barlogis, V. *et al.* Burden of Poor Health Conditions and Quality of Life in 656 Children with Primary Immunodeficiency. *J. Pediatr.* 194, 211-217.e5 (2018).
5. Michniacki, T. F., Walkovich, K. J., Merz, L. E., Sturza, J. & Abraham, R. S. Provider Perceptions of Quality of Life, Neurocognition, Physical Well-being, and Psychosocial Health in Patients with Primary Immunodeficiency/Immune Dysregulation Conditions. *J. Clin. Immunol.* 39, 805–813 (2019).
6. Anderson, J. T., Cowan, J., Condino-Neto, A., Levy, D. & Prusty, S. Health-related quality of life in primary immunodeficiencies: Impact of delayed diagnosis and treatment burden. *Clin. Immunol.* 236, 108931 (2022).
7. UN General Assembly. Convention on the Rights of Persons with Disabilities: resolution / adopted by the General Assembly, A/RES/61/106. (2007).



الرابطة العربية لضعف المناعة الأولي
Arab Society for Primary Immunodeficiencies



International Nursing Group

