

Letter to the Editor

Primary immunodeficiency: Potential benefits of active transition to improved multi-disciplinary care

Editor,

The recent description by Llobet and colleagues of a 20-yr paediatric experience in Common Variable Immuno-Deficiency (CVID) is both illuminating and timely (1). The mean age of diagnosis of CVID now approaches 35.3 yr (2) suggesting this to be a disease of adult onset, yet in the United Kingdom at least, up to 8% of diagnoses have been made before the age of 10 yr. Within the Catalan cohort described here, diagnoses were made as early as 6 months and up to 32% had established end organ disease (bronchiectasis) at diagnosis. In addition to infective outcomes, significant autoimmune disease (including anaemia, hepatitis and systemic lupus erythematosus) and neoplasia (osteochondroma) were observed.

As these patients approach transition to adult care several important needs are raised. First, these subjects require continuing IVIG replacement therapy with appropriate supervision and monitoring. Given that this therapy is likely to be life-long, therapy needs to be safe, local, convenient and appropriate in terms of dose, delivery route and frequency.

Second, a variety of established complications requires ongoing and specialist assessment and management by diverse disciplines including clinical immunology, respiratory, haematology, gastroenterology, physiotherapy, dietetics and psychosocial services. Sino-pulmonary sepsis and bronchiectasis in particular are major determinants of quality of life and survival and the inclusion of specialist input in the creation and implementation of management plans is therefore highly desirable (3, 4).

Finally, these individuals remain susceptible to a broad array of potentially threatening complications some of which may be subclinical and others yet to manifest. Prognostication in CVID

has proven challenging and several attempts at immunological characterization using immunoglobulin levels and lymphocyte subsets have been attempted yet these addresses continue to have relatively poor descriptive and predictive power (5–7). In the absence of directive indices of prognosis, a high index of clinical suspicion needs to be maintained and regular and appropriate surveillance undertaken. Guidelines on strategies for screening and detection of complications have not been formulated but may well be useful in this regard.

Lack of preparation, information and awareness about transition of care from paediatric to adult care services is a source of fear and concern for both young patients and adult caregivers (8, 9) and an active transition process to adult care facilities is now recognized as desirable for all adolescents with chronic illness (10). The development of such services has many potential benefits including promotion of awareness of immunodeficiency syndromes, improved patient and family awareness and improvement of patient self management and self-efficacy skills (9, 11). A focus on the specific definition of the needs of each individual will further help to enable the development of a considered and cooperative multidisciplinary management platform for the ongoing care of patients with these complex and relatively rare conditions.

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