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Sommario/riassunto

Systemic lupus erythematosus (SLE) affects predominantly women at reproductive age but may present at any age. Age at disease onset has a modulating effect on presentation and course of disease, but controversies persist regarding its impact on long-term outcome. Our aims were to characterize clinical features, co-morbidities and cumulative damage in childhood-onset, adult-onset and late-onset SLE. Patients with childhood-onset SLE fulfilling ACR 1997 criteria were identified in a nationwide register-Reuma.pt/SLE (N = 89) and compared with adult-onset and late-onset counterparts matched 1:1:1 for disease duration. 267 SLE patients with mean disease duration of 11.9 ± 9.3 years were analyzed. Skin (62 %), kidney (58 %), neurological (11 %) and hematologic involvement (76 %) were significantly more common in childhood-onset SLE and disease activity was higher in this subset than in adult- and late-onset disease (SLEDAI-2K 3.4 ± 3.8 vs. 2.2 ± 2.7 vs. 1.6 ± 2.8, respectively; p = 0.004). Also, more childhood-onset patients received cyclophosphamide (10 %) and mycophenolate mofetil (34 %). A greater proportion of women (96 %), prevalence of arthritis (89 %) and anti-SSA antibodies (34 %) were noted in the adult-onset group. There was a significant delay in the diagnosis of SLE in older ages. Co-morbidities such as hypertension, diabetes and thyroid disease were significantly more frequent in late-onset SLE, as well as

the presence of irreversible damage evaluated by the SLICC/ACR damage index (20 vs. 26 vs. 40 %; $p < 0.001$). Greater organ involvement as well as the frequent need for immunosuppressants supports the concept of childhood-onset being a more severe disease. In contrast, disease onset is more indolent but co-morbidity burden and irreversible damage are greater in late-onset SLE, which may have implications for patients' management.