

Reply to the comment by Alarcón *et al.*

Sirs,

We thank G.S. Alarcon *et al.* for giving us the opportunity to clarify this point. Indeed, the ACR/SLICC Damage Index (SDI) defines damage as irreversible change occurring after diagnosis of the disease, which should be present for at least 6 months, thus excluding items manifesting before the definitive diagnosis (1). However, it is possible that damage can occur before the formal diagnosis of SLE, and still be attributable to SLE (2). Interestingly, in the ongoing revision of the SDI, the time frame is one of the main thematic clusters; in an initial qualitative study to achieve agreement in the conceptual framework of the revised damage index, the authors state that “*damage can occur before a diagnosis of SLE but should be attributable to SLE*” (3). Additionally, in the original SDI publication, it is stated that a damage item is considered positive only after 6 months, “*unless otherwise stated*” - the latter refers to, *e.g.* myocardial infarction, stroke, significant tissue loss due to necrosis etc. (1).

In this study, using a large SLE cohort, we found a median disease duration of 3 years, with 1 out of 6 patients already having irreversible organ damage at the time of diagnosis. By this, we mean that we included damage items that were already present at first diagnosis of SLE, as a sequel of a presenting disease manifestation, following a thorough exclusion of other possible causes. These damage items fall into the specification “*unless otherwise stated*” of the SDI definition, and refer to cerebrovascular events, myelitis, pulmonary infarction, my-

Table I. Initial SLE manifestations led to SDI within 6 months after diagnosis.

Manifestations	Number of events
Cerebrovascular events	12
Seizures requiring therapy for 6 months	3
Neuropathies (cranial & peripheral)	1
Transverse myelitis	2
Psychosis	4
Pulmonary infarction (radiograph)	14
Venous thrombosis with swelling, ulceration or venous stasis	15
Pulmonary fibrosis (physical and radiographical)	0
Myocarditis [Cardiomyopathy (ventricular dysfunction)]	2
Arterial thrombosis (tissue loss)	0
Myocardial infarction	2
Chronic kidney disease (Estimated or measured glomerular filtration rate <50%)	1
Pulmonary hypertension (right ventricular prominence, or loud P2 within 6 months after diagnosis)	3

ocardial infarction and arterial thrombosis with tissue loss. However, we also included items present at diagnosis, which led to irreversible damage within the first 6 months, and these included venous thrombosis with swelling, ulceration or venous stasis, seizures requiring anti-convulsive therapy for 6 months, chronic kidney disease (Table I).

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