Background Despite validated clinical measures and indices of disease activity and damage, utilisation of these indices in clinical practice varies, as evidenced by a recent practice pattern survey of Canadian rheumatologists. In this review, we aimed to identify the impact of disease activity and damage on outcomes of mortality and damage to inform upcoming Canadian SLE recommendations utilising the GRADE (Grading of Recommendations Assessment, Development and Evaluation) method.

Materials and methods Following GRADE methodology to fill in evidence-to-decision tables to create recommendations for “minimal investigations needed to monitor SLE patients at baseline and subsequent visits”, a systematic review of the literature including all relevant articles from 1946 to November 2014 was performed searching Medline and Embase. The impact of disease activity and damage measured by commonly utilised indices of disease activity [eg SLEDAI-2K (SLE Disease Activity Index-2000), BILAG (British Isles Lupus Assessment Group), SLAM (SLE Activity Measure), ECLAM (European Consensus Lupus Activity Measurement)], Mexican SLEDAI, and damage [SDI (SLICC/ACR Systemic Lupus International Collaborating Clinics/ACR Damage Index)] on mortality, damage, and disease flares was evaluated with meta-analyses performed when available. Study quality was assessed by the Newcastle Ottawa scale for observational studies.

Results A title screen of 2797 articles identified 771 papers for full paper review, 106 meeting inclusion criteria and 88 with extractable data. Fifty-five articles describing outcomes with disease activity indices (including BILAG, ECLAM, Mexican SLEDAI, SLEDAI-2K/SELENA-SLEDAI and SLAM) and twenty-four articles describing outcomes with damage based on SLED were identified. Mortality was associated with higher SLEDAI-2K in 6 observational studies [HR 1.14 (95% CI: 1.06,1.22)] and in 5 observational studies with higher SDI scores at baseline and/or immediately prior to death [HR 1.53 (95% CI: 1.28, 1.83)]. Higher SLAM scores were associated with increased risk of damage (SDI > 0) in 3 observational studies [OR 1.06 (95% CI: 1.04, 1.08)]. Mean total BILAG was associated with mortality in one observational study with HR of 1.15.

Conclusions Active lupus disease activity and presence of damage as represented by multiple clinical indices are associated with greater mortality and morbidity in lupus patients. Given the complexity of clinical assessments in SLE patients, the utilisation of validated measures for disease activity and damage is important and will serve to inform upcoming Canadian recommendations for the diagnosis and monitoring of SLE.

Acknowledgements This systematic literature review is being used by the Canadian SLE Working Group to inform future recommendations for the diagnosis and monitoring of SLE.