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Measures of Adult Systemic Lupus Erythematosus:

Updated Version of British Isles Lupus Assessment Group (BILAG 2004), European Consensus Lupus Activity Measurements (ECLAM), Systemic Lupus Activity Measure, Revised (SLAM-R), Systemic Lupus Activity Questionnaire for Population Studies (SLAQ), Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K), and Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index (SDI)

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INTRODUCTION

Measurement of disease activity in systemic lupus erythematosus (SLE) is central to evaluating outcomes, differences among SLE patient groups, responses to a new drug proposed, and also for assessing disease longitudinally for observational and clinical trials. Several validated and updated instruments have been available since the early 1980s, but more recent studies gauging reliability and validity for classifying and monitoring groups of patients in the research setting are now available.

Two cardinal features of SLE have challenged investigators refining these tools: first, the complex multisystem nature of this disease with fluctuating levels of disease activity, which may vary between patients and within the same patient over time; second, the absence of a "gold standard" for determining the psychometric properties of each proposed scale limits comparisons to expert opinion using a physician's visual analog scale or by comparing one scale against other to assess performance across proposed instruments. However, these strategies do not eliminate bias based on personal experience, nor do they differentiate between different opinions on the relative importance of disease manifestations in different systems.

Therefore, an experience-based evaluation may be subject to greater interrater variability than the use of the disease activity instrument itself. Furthermore, psychometric properties should be influenced by the length of the scale (number of items and scoring scale), number of patients included, or disease severity of patients under study.

Two main types of activity measures in SLE have been developed: global score systems (for example, the European Consensus Lupus Activity Measurements, Systemic Lupus Activity

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AUTHOR CONTRIBUTIONS

All authors were involved in drafting the article or revising it critically for important intellectual content, and all authors approved the final version to be published.

Measure [SLAM], and Systemic Lupus Erythematosus Disease Activity Index [SLEDAI]), which provide an overall measure of activity, and individual organ/system assessment scales that assess disease activity in single organs (such as the British Isles Lupus Assessment Group Index [BILAG]). The Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index score is a measure for chronic damage; it has been included due to its prognostic value in clinical and research basis.

The SLEDAI, SLAM, and BILAG have performed in effective and reliable manners in studies; furthermore, they correlate with one another (1-3). The SLEDAI, Safety of Estrogens in Lupus Erythematosus National Assessment (SELENA)–SLEDAI, SLEDAI 2000 (4-7), and BILAG (8-10) have been successfully used in observational trials and case studies, although baseline disease activity index (DAI) scores were not always predictors of subsequent damage or other outcomes (11,12). These DAIs were validated in the context of long-term observational trials studies and not in randomized clinical trials (RCTs) (1,9,10,13-15). The few RCTs conducted have shown that improvement in DAI scores correlates with response rates, disease remission, and flare prevention; however, a threshold of clinically meaningful change has not been established (1,13,16,17). Current work has focused on developing a responder index developed in collaboration with the Food and Drug Administration–defined response as improvement and/or no deterioration in patient- and physician-reported outcomes. The SLE responder index, which utilizes the SELENA–SLEDAI score to determine global improvement, BILAG domain scores to ensure no significant worsening in heretofore unaffected organ systems, and physician's global assessment to ensure that improvements in disease activity are not achieved at the expense of the patient's overall condition, which may have been missed by either DAI, is one example used in a recent clinical trial (18). Ongoing work to refine or develop responder indices will enhance our ability to measure meaningful outcomes in future RCTs.

For purpose of this review, we selected those indices that have shown the strongest evidence of validity when used by investigators from different countries in large studies of patients with SLE. The exact choice of instrument should be governed by the purpose for which it is required in clinicalpractice or research.

UPDATED VERSION OF BRITISH ISLES LUPUS ASSESSMENT GROUP (BILAG 2004)

Description

Purpose—To assess lupus activity based upon the "intent-to-treat" premise. The original version was published in 1988 (19). Over time, several deficiencies were noted by members of BILAG, which prompted a major revision. The updated version (BILAG 2004) was published in 2005 (20).

Content—Specific manifestation in 9 systems. In this revised index, the original section of vasculitis has been removed and 2 systems were added: ophthalmic and abdominal.

Number of items—101 and 5 additional items required mainly for calculations ofglomerular filtration rate.

Response options/scale—Each question is answered as: 0 = not present, 1 = improving, 2 = same, 3 = worse, and 4 = new.

Recall period for items—It records disease activity occurring over the past 4 weeks as compared with the previous 4 weeks.

Endorsements—Adult patients with systemic lupus erythematosus (SLE).

Examples of use—Yee CS, Isenberg DA, Prabu A, Sokoll K, Rahman A, Bruce IN, et al. BILAG 2004 index captures systemic lupus erythematosus disease activity better than SLEDAI-2000. Ann Rheum Dis 2008;67:873–6 (21).

Isenberg DA, Allen E, Farewell V, D'Cruz D, Alarcon GS, Aranow C, et al. An assessment of disease flare in patients with systemic lupus erythematosus: a comparison of BILAG 2004 and the flare version of SLEDAI. Ann Rheum Dis 2011;70:54–9 (22).

Practical Application

How to obtain—The BILAG 2004 index assessment and BILAG 2004 index glossary canbe obtained at *Rheumatology* online as supplementary data without cost.

Contact information: The BILAG group: current chair of the BILAG group is Professor David Isenberg, Room 331, The Windeyer Building, University College London, 46 Cleveland Street, London W1T 4JF, UK.

Method of administration—Physician completed.

Training: Formal training of raters and a well-defined glossary are needed.

Equipment needed: None to complete the index. To calculate categorical or numerical scoring, acomputer program is needed.

Scoring—As above, each question is recorded as 0, 1, 2, 3, or 4. Then, a computer program facilitates scoring from numerical to alphabetical score for each system (grade A–E).

Score interpretation—The BILAG 2004 index categorizes disease activity into 5 different levels from A–E. Grade A represents very active disease likely necessitating immunosuppressive drugs and/or a prednisolone (or equivalent) dose of >20 mg daily or high-dose anticoagulation. Grade B represents moderate diseaseactivity requiring a lower dose of corticosteroids, topical steroids, topical immunosuppressive drugs, antimalarials, or nonsteroidal antiinflammatory drugs. Grade C indicates mild stable disease, while grade D implies no disease activity but the system had previously been affected. Grade E indicates no current or previous disease activity.

Respondent burden—5–20 minutes, plus time to complete history and physician examination.

Administrative burden—Up to 50 minutes. The instrument cannot be scored until laboratory results are available, and this may take a few days.

Translations/adaptations—English only. The original BILAG index isavailable in computer version.

Psychometric Information

Method of development—The BILAG 2004 was developed by nominal consensus. The members of the BILAG developed the BILAG index by making agreed assumptions about the likely treatment that will be given to patients with particular groups of clinical features. There was no attempt to weight the importance of involvement of different systems. Items were generated by detailed discussion of BILAG members.

Reliability—Good reliability (intraclass correlation coefficient [ICC] >0.60) and high levels of physician agreement ($\sigma_{physician}/\sigma_{patient}$ = <0.40) wereshown in 2 real-patient exercises.

The interrater reliability of the index was shown in a multicenter study of 97 "live" patients in 2 exercises (E1 and E2). The overall ICC determined in E1 was 0.45 (95% confidence interval [95% CI] 0.31, 0.58), and in E2 was 0.67 (95% CI 0.54, 0.76). There was improvement in the levels of agreements and in the kappa and ICC reliability from E1 to E2. Four items with poor agreement between raters were identified. Training of raters was suggested to ensure the optimal performance of the index (23).

Validity—In a multicenter cross-sectional study of 369 patients, scores indicating active disease (overall BILAG 2004 scores of A and B) were significantly associated with increase in therapy (odds ratio 19.3, $P < 0.01$). The overall sensitivity of the index was 81%, specificity was 81.9%, positive predictive value was 56.8%, and negative predic-tive value was 93.6%. Construct andcriterion validity were also shown (24).

Ability to detect change—Using the external method responsiveness, the BILAG 2004 has been shown to be sensitive to change to assess SLE disease activity. This has been shown in a longitudinal study that involved 8 centers in the UK in which 1,761 visits from 347 SLE patients were evaluated.Increase in the overall score was associated with increase in therapy (coefficient 1.35; 95% CI 1.01, 1.70) and inversely associated with decrease in therapy (coefficient −0.44; 95% CI −0.81, −0.06) (25).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—This score incorporatesthe important element of change in disease state with time (the delta factor). It is sensitive to small changes and distinguishes between disease activity and disease severity. It is the only validated lupus activity index that aims to show activity in individual systems "at a glance" rather than combining them into a global score.

Caveats and cautions—Formal training of raters and a well-defined glossary are essential to ensure the optimal performance of the index.

Clinical usability—The BILAG 2004 index was developed particularly for research. However, it should be useful to monitor the disease for individuals due its ability to identify whether the disease is improving, stable, or worsening.

Research usability—The BILAG 2004 index is appropriate for investigations of disease outcome and treatment protocols. Despite the complex calculations, the score is quick to conduct, especially when calculated by a computer, and only minimally dependent on the particular clinician carrying out the procedure. To facilitate comparisons with global indices, a numerical scoring system has been associated with the BILAG 2004 index. The optimal method is to convert the assessments so that an "A" = 12 points, "B" = 8 points, "C" = 1 point, and "D/E" = 0 points (26).

EUROPEAN CONSENSUS LUPUS ACTIVITY MEASUREMENTS (ECLAM)

Description

Purpose—To assess disease activity in patients with lupus within the past month.

Content—Lupus activity is divided into 10 organs/systems, plus erythrocyte sedimentation rate (ESR) and complement levels with varying numbers of items in each. Emphasis is on evolving changes.

Number of items—33 items.

Response options/scale—There are 12 categories (10 organs/systems plus ESR and complement levels), 4 of which are divided into subcategories.

Recall period for items—The last month.

Endorsements—Disease activity in patients with systemic lupus erythematosus (SLE).

Examples of use—American College of Rheumatology Ad Hoc Committee on Systemic Lupus Erythematosus Response Criteria. The American College of Rheumatology response criteria for systemic lupus erythematosus clinical trials: measures of overall disease activity. Arthritis Rheum 2004;50:3418–26 (27).

Mosca M, Chimenti D, Pratesi F, Baldini C, Anzilotti C, Bombardieri S, et al. Prevalence and clinico-serological correlations of anti-α-Enolasa, anti-C1q, and anti-dsDNA antibodies in patients with systemic lupus erythematosus. J Rheumatol 2006;33:695–7 (28).

Amital H, Szekanecz Z, Szucz G, Danko K, Nagy E, Csepany T, et al. Serum concentration of 25-OH vitamin D in patients with systemic lupus erythematosus (SLE) are inversely related to disease activity: is it time to routinely supplement patients with SLE with vitamin D? Ann Rheum Dis 2010;69:1155–7 (29).

Practical Application

How to obtain

Contact information: The European Workshop for Rheumatology Research. Main developer and contact person is Professor Stephano Bombardieri, Universidad of Pisa, Italy.

Method of administration—Physician completed.

Scoring—Simple additive.

Score interpretation—Range is 0–17.5. This is a global score index. Item scores range from 0.5 (e.g., fever/fatigue) to 2 (e.g., new neuropsychiatric/evolving renal manifestation).

Respondent burden—Up to 10 minutes.

Administrative burden—A history and physician examination is needed. For a reasonably stable patient, <5 minutes; for a complicated patient, up to 10 minutes. Training is needed, especially in a multicenter studies.

Translations/adaptations—English and Italian versions available. Paper or computer versions.

Psychometric Information

Method of development—The ECLAM was constructed during the course of a multicenter study involving 704 patients, on the basis of the correlations found for each

patient between a wide range of clinical/laboratory parameters with the clinician's assessment of disease activity (the gold standard). Multivariate regression analyses were carried out to evaluate the combined performance of different sets of clinical and serologic variables in predicting disease activity, and to define the relative weight of each variable in terms of regression coefficients in multivariate models (30).

Reliability—Data from 32 consecutive patients were obtained from 4 observers (2 experts, 1 trainee, 1 nurse). The correlation coefficients between ECLAM scores ranged from 0.9–0.95 (31). In a second study, 64 consecutive patients were scored at time of evaluation and 2 weeks later from chart data by 2 observers. The correlation coefficient between patient and chart ECLAM score was 0.88 and the interobserver variability was low, with a correlation coefficient ranging from 0.9–0.93 (32).

Validity—Data from 75 patients (19 centers) were collected and each patient was observed twice over 3 months. The ECLAM index at each time point was compared with the Systemic Lupus Activity Measure (SLAM), Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), and British Isles Lupus Assessment Group (BILAG). The correlation coefficients for the ECLAM compared with the others indices ranged from 0.72–0.78 (3).

Ability to detect change—In 23 patients seen every 2 weeks for up to 40 weeks, 5 disease activity measures were completed along with the physician's and patient's global assessments. Changes in SLE activity were correlated with each activity measure, and for the ECLAM, r = 0.65. Sensitivity to change was greatest for the ECLAM when compared with the physician's global assessment. Using a standardized response measure, the score for the ECLAM was 0.75 (3).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—The ECLAM index was directly derived from a large number of real patients and the analysis of a large amount data collated in a standardized manner during a multicenter study.

Caveats and cautions—Global score will miss changes in severity over time.

Clinical usability—The ECLAM index should be an excellent tool for clinical usability because of its great simplicity. It is based on 12 of the most common parameters of disease activity.

Research usability—The ECLAM score has been widely used in sets of real and paper patient exercises mostly comparing it with the SLEDAI, SLAM, and BILAG. It has been shown to be a reliable instrument for calculating disease activity retrospectively from clinical charts when used in the setting of a tertiary center for patient care.

SYSTEMIC LUPUS ACTIVITY MEASURE, REVISED (SLAM-R)

Description

Purpose—To measure the degree of disease activity in patients with systemic lupus erythematosus (SLE) within the last month. It was published in 1988 and revised in 1991 (33).

Content—Specific manifestation in 9 organs/systems, plus 7 laboratory features.

Number of items—9 organs/systems, with laboratory category.

Response options/scale—Organ items scored 0–3 points if present within the last month (severity incorporated into higher score per item). Most items can score a maximum of 3 points. Few items can score a maximum of 1 point. The laboratory category can score a maximum of 21 points.

Recall period for items—The SLAM covers symptoms that occurred during the previous month.

Endorsements—Patients with SLE.

Examples of use—Chang ER, Abrahamowics M, Ferland D, Fortin PR. Organ manifestations influence differently the responsiveness of 2 lupus disease activity measures, according to patients' or physicians' evaluations of recent lupus activity. J Rheumatol 2002;29:2350–8 (34).

Zhang J, Gonzales LA, Roseman JM, Vila LM, Reveille JD, Alarcon GS. Predictors of the rate of change in disease activity over time in LUMINA, a multiethnic US cohort of patients with systemic lupus erythematosus: LUMINA LXX. Lupus 2010;19:727–33 (35).

Practical Application

How to obtain—Copyrighted by Fellows of Harvard College; developer and contact person is Dr. Matthew Liang, Professor of Medicine, Department of Medicine/ Rheumatology/Immunology, PBB-82, Brigham & Women's Hospital, 75 Francis Street, Boston, MA 02115. The computer version is available from Gordon Hamilton (e-mail: LIMATHON@aol.com).

No cost to use (unless the computerized version is needed, then cost depends upon type of usage [commercial/academic]).

Method of administration—Physician completed. Questionnaire available in paper format (optical scannable) or as part of the BLIPS software program.

Scoring—Simple additive.

Score interpretation

Score range: Maximum score is 81 points. Judgment as to whether manifestations (laboratory or otherwise) are due to lupus is needed. A score of 7 is considered clinically important and effects decision to treat.

Respondent burden—Up to 15 minutes.

Administrative burden—A complete history and physical examination is needed. To complete the form in an essentially well patient with a short history takes <10 minutes. For a complex patient not well known to the physician it can take up to 15 minutes. For most patients it takes <10 minutes.

Training is needed to develop consensus on subjective components of the index, especially in multicenter studies. Dr. Matthew Liang (contact information above) or Dr. Paul Fortin (Division of Rheumatology, Room MP-10-304, Toronto Western Hospital, 399 Bathurst Street, Toronto, Ontario M5T 2S8, Canada) is suggested.

Translations/adaptations—Available in English, Korean, German, and Chinese.

Psychometric Information

Method of development—It was developed based on domain sampling theory. Items chosen for the scale represent those manifestations that occur more frequently, those that can be graded, and those that can be operationally defined and reliably rated.

Reliability—The reliability of the index was shown in a study of 25 "live" patients seen twice over a 3-5-week period and 2 physicians who were not providing care for the patients. The SLAM index interrater reliability and intervisit reliability were 0.86 and 0.73, respectively.

The reliability of the SLAM-R was demonstrated in a study of 30 patients seen twice 2–4 weeks apart by 2 physicians who were not providing care for the patient. The SLAM-R index interrater reliability and intervisit reliability were 0.78 and 0.85, respectively (36).

Validity

Convergent and discriminant: The validity of the index was shown in a study of 25 "live" patients seen twice over a 3-5-week period and 2 physician raters using 6 scales, including the SLAM. These raters were not providing care for the patients. The average correlation between the SLAM and the other scales was 0.9, ranging from 0.9–1.0. Furthermore, when correlations were evaluated to assess change between visits, the range was 0.5–0.8 across instruments, demonstrating convergent validity. The various components contributing to the total variance of the SLAM were 73% for patients, 13% for visits, and 14% for raters demonstrating discriminant validity (37).

Construct validity of the SLAM-R: The correlation between the SLAM-R scores, the physician's global assessment, anti–double-stranded DNA, C3, and C4 were statistically significant, ranging from −0.29 to 0.87 (37).

Ability to detect change—Excellent sensitivity and responsiveness to change have been shown in comparative studies with the British Isles Lupus Assessment Group (BILAG) and Systemic Lupus Erythematosus Disease Activity Index (SLEDAI). In an international validation study, where 8 patients with 3 visits were rated by the Systemic Lupus International Collaborating Clinics group using 3 indices (SLEDAI, BILAG, SLAM), all indices were able to detect differences between patients ($P < 0.01$) (38).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—This index includes both dimensions: disease activity and disease severity.

Caveats and cautions—One of its disadvantages is that many items are subjective, because scoring relies on the reporting of symptoms by the patients rather than objective documentation. Difficulty in distinguishing changes, i.e., patients with multiple mild or improving manifestations compared to those with 1 or 2 severe features. Note that some of the most severe items also count as damage, i.e., cerebrovascular accident.

Clinical usability—For this index, a score of ≥7 is considered clinically important because it is associated with a probability of initiating therapy in >50% of cases. However, it is important to consider that it gives equal weighting to mild and serious organ disease activity without considering the significance of the organ involved.

Research usability—This index has a high sensitivity to change and responsiveness when the patient's global assessment is considered as the standard. The SLAM correlates with several aspects of the patient's perception of health, as evaluated with the Short Form 36 (34,35,39).

SYSTEMIC LUPUS ACTIVITY QUESTIONNAIRE FOR POPULATION STUDIES (SLAQ)

Description

Purpose—To provide an economic way of following and tracking disease activity for large groups of systemic lupus erythematosus (SLE) patients who may be at a distance from a center in epidemiologic studies. It was developed based on items from the Systemic Lupus Activity Measure (SLAM) (40). It was published in 2003.

Content—Specific symptoms of disease activity and a single numerical rating scale (NRS) asking the patient to rate disease activity on a scale of 0–10 over the past 3 months.

Number of items—24 items in 9 organs/systems weighted and aggregated in a manner analogous to the scoring system used in the SLAM.

Response options/scale—For questions regarding disease activity, there are 4 options, as follows: no problem = 0, mild = 1, moderate = 2, and severe = 3. For a single NRS, it rates from 0 = "no activity" to 10 = "most activity."

Recall period for items—The last 3 months.

Endorsements—Studies with large groups of SLE patients.

Examples of use—Trupin L, Tonner MC, Yazdany J, Julian LJ, Criswell LA, Katz PP, et al. The role of neighborhood and individual socioeconomic status in outcomes of systemic lupus erythematosus. J Rheumatol 2008;35: 1782–8 (41).

Wolfe F, Petri M, Alarcon GS, Goldman J, Chakravaty EF, Katz RS, et al. Fibromyalgia, systemic lupus erythematosus (SLE), and evaluation of ALE activity. J Rheumatol 2009;36:82–8 (42).

Practical Application

How to obtain—A copy can be obtained for 1 study (see Appendix A) (40) without cost.

Method of administration—Patient self-completed questionnaire or telephone administration.

Scoring—Arithmetic computation by hand.

Score interpretation—Scores can range from 0–44. It correlates with the physician-completed SLAM.

Respondent burden—p to 10 minutes.

Administrative burden—Up to 10 minutes.

Translations/adaptations—English only. No adaptations available.

Psychometric Information

Method of development—It was developed based on domain sampling theory. Under a clinical setting, assessments of 93 patients who presented to an academic medical center for clinical care were used. It was based on items from the SLAM that are amenable to self-report (40).

Reliability—In an observational cohort study of 982 English-speaking patients with SLE, the SLAQ demonstrated excellent internal consistency, with a Cronbach's α of 0.87. Data structure examined by principal factor analysis showed that 1 factor accounted for 92% of the variance (43).

Validity—Construct validity was demonstrated by examining correlation of the SLAQ with measures that are likely to be related to disease activity in SLE (r = 0.51–0.73) (43).

Ability to detect change—The SLAQ demonstrated a small to moderate degree of responsiveness for participants who reported a perceived change in disease status; standardized response means were 0.66 and −0.37 for those reporting clinical worsening and improvement, respectively (43).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—The SLAQ index is a unique instrument developed and validated for measure disease status outside the clinical setting in SLE patients. It is very useful for large epidemiologic studies in which many patients live outside the catchment area or physician-directed assessment may prove impractical and costly.

Caveats and cautions—The SLAQ instrument should not be used instead of careful clinical followup of patients in day-to-day practice. If the level of education may influence, the response rate needs to be evaluated. Future studies are needed to confirm the reliability of the SLAQ compared with a physician assessment, particularly in different age, sex, and racial/ethnic groups.

Clinical usability—The SLAQ is intended to be used as an initial screen to identify subjects with new or increased disease activity who need further evaluation by a physician (positive predictive value ranged from 56–89% for detecting clinically significant disease activity).

Research usability—The SLAQ demonstrated adequate reliability, construct validity, and responsiveness in a large community-based cohort of patients with SLE.

SYSTEMIC LUPUS ERYTHEMATOSUS DISEASE ACTIVITY INDEX 2000 (SLEDAI-2K)

Description

Purpose—To measure disease activity in patients with systemic lupus erythematosus (SLE). The original version was introduced in 1985 (15,44). In 2002, it was modified to reflect persistent active disease in those descriptors that had previously considered new or recurrent occurrences (SLEDAI-2K) (45).

Content—Specific manifestation in 9 organs/systems.

Number of items—24 items covering 9 organs systems.

Response options/scale—There are 24 items for the 9 organs/systems. Scored if present within the last 10 days. Two systems can score a maximum of 8 points each, 2 systems can score a maximum of 4 points each, 3 systems can score a maximum of 2 points each, and 2 systems can score a maximum of 1 point each. Scores range from 0–105 points.

Recall period for items—Disease activity within the last 10 days. Recently, the SLEDAI-2K for a timeframe of 30 days prior to a visit for clinical and laboratory variables was shown to be similar to the SLEDAI-2K for 10 days (46).

Endorsements—Disease activity in patients with SLE.

Examples of use—Uribe AG, Vila LM, McGwin G Jr, Sanchez ML, Reveille JD, Alarcon GS. The Systemic Lupus Activity Measure-Revised, the Mexican Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), and a modified SLEDAI-2K are adequate instruments to measure disease activity in systemic lupus erythematosus. J Rheumatol 2004;31:1934–40 (47).

Petri M, Kim MY, Kalunian KC, Grossman J, Hahn BH, Sammaritano LR, et al. Combined oral contraceptives in women with systemic lupus erythematosus. N Engl J Med 2005;353:2550–8 (48).

Sanchez-Guerrero J, Uribe AG, Jimenez-Santana L, Mestanza-Peralta M, Lara-Reves P, Seuc AH, et al. A trial of contraceptive methods in women with systemic lupus erythematosus. N Engl J Med 2005;353:2539–49 (49).

Practical Application

How to obtain—The Toronto Group: Claire Bombardier, MD, initial development only), Drs. Dafna Gladman, MD, and Murray Urowitz, MD (Toronto Western Hospital, 399 Bathurst Street IE – 410B, Toronto, Ontario, Canada M5T 2S8).

Method of administration—Physician completed.

Scoring—Simple additive.

Score interpretation—The score range is 0–105 points. A score of 6 is considered clinically important and affects decision to treat.

Respondent burden—Up to 10 minutes.

Administrative burden—A complete history and physical examination is needed. The instrument cannot be scored until laboratory results are available, and this may take a few days. To complete the form in an essentially well patient with a short history it can take <10 minutes. For a complex patient not well known to the physician it can take <10 minutes.

Translations/adaptations—The SLEDAI-2K is available in English and Spanish. Some adaptations have been published, e.g., the Safety of Estrogens in Lupus Erythematosus National Assessment-SLEDAI used in the Safety of Estrogen trial. It was modified from the SLEDAI to insure that the descriptors of organ system involvement reflected ongoing disease activity (50,51). The Mexican modification of the SLEDAI, a simplified version without the immunologic test, makes the index cheaper to administer (52).

Psychometric Information

Method of development—It was developed with a panel of experienced rheumatologists with expertise in SLE, using well-established group techniques and index development methodology.

Reliability—The reliability of the original SLEDAI was shown in a paper patient exercise in which 534 scenarios were generated from real patient data and 14 lupus experts participated in an interrater reliability study. The interrater correlation for the 46 most common patients profiles ranged from 0.61–0.80 (15).

The reliability of the SLEDAI-2K was evaluated in a multicenter multiethnic study where 93 patients were studied. Agreement for each of the items was between 81.7% and 100% (10).

Validity—A group of 14 lupus experts completed a testing set of 69 real scenarios with common manifestations, 98 anchor profiles, and 116 real patient cases. The intraclass correlation coefficient was 0.79, representing slightly stronger agreement within cases with common manifesta-tions of disease than for unique (0.71) or anchor profiles (0.64) (15).

The SLEDAI-2K was validated against the SLEDAI using all visits in a cohort of 960 patients in the Toronto data-bank; there was a high correlation between both indices ($r = 0.97$, $P = 0.0001$) (45).

Ability to detect change—The SLEDAI sensitivity and responsiveness to change have been shown in comparative studies with the Systemic Lupus Activity Measure, British Isles Lupus Assessment Group, and European Consensus Lupus Activity Measurements. In a prospective study, 23 patients with SLE were examined every 2 weeks for up to 40 weeks. Estimates of sensitivity to change varied with the standard used. The sensitivity to change was smallest for the SLEDAI, with a standardized response mean (SRM) of 0.48 when the physician global assessment was used as the standard and an SRM of −0.01 when the patient global assessment was used (3,38).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—All versions are validated and used by lupus researchers for clinical and research purposes.

Caveats and cautions—The SLEDAI does not record improving or worsening, and does not include severity within an organ system.

Clinical usability—Activity categories have been defined on the basis of the SLEDAI score. A SLEDAI score >5 is associated with a probability of initiating therapy in >50% of cases.

Research usability—Neither version of the SLEDAI captures improving or worsening. This probably explains why it is less sensitive to change than other instruments.

SYSTEMIC LUPUS INTERNATIONAL COLLABORATING CLINICS (SLICC)/ AMERICAN COLLEGE OF RHEUMATOLOGY DAMAGE INDEX (SDI)

Description

Purpose—To capture those items of permanent change that has occurred in patients after a diagnosis of systemic lupus erythematosus (SLE), regardless of attribution.

Content—Specific manifestation in 12 organ systems.

Number of items—41 items covering 12 organ systems. Within each scale or system, a variable number of components are to be found (up to 6).

Response options/scale—Thirty-one items score 1 point if present. Six items can score a maximum of 2 points; 1 item can score a maximum of 3 points.

Recall period for items—Duration of manifestation (or irreversibility), i.e., must be present for a minimum of 6 months or expected not to reverse, such as surgical procedure or infarction.

Endorsements—Measure damage in patients with SLE.

Examples of use—Gladman D, Ginzler E, Goldsmith C, Fortin P, Liang M, Urowitz M, et al. The development and initial validation of the Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index for systemic lupus erythematosus. Arthritis Rheum 1996;39:363–9 (53).

Stoll T, Seifert B, Isenberg DA. SLICC/ACR Damage Index is valid and renal and pulmonary organ scores are predictors of severe outcome in patients with systemic lupus erythematosus. Br J Rheumatol 1996;35:248–54 (54).

Rahman P, Gladman DD, Urowitz MB, Hallett D, Tam LS. Early damage as measured by the SLICC/ACR Damage Index is a predictor of mortality in systemic lupus erythematosus. Lupus 2001;10:93–6 (11).

Practical Application

How to obtain

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Questionnaire available in paper format or as part of the BLIPS software program. The computer version is available from Gordon Hamilton (e-mail: LIMATHON@aol.com).

Method of administration—Physician completed.

Scoring—As above, the duration of manifestation (or irreversibility), i.e., must be present for a minimum of 6 months or expected not to reverse, such as surgical procedure or infarction. Item scored regardless of attribution to SLE; therefore, this catches morbidity from treatment from SLE or other complications that may be increased in SLE, e.g., fracture, etc.

Score interpretation

Score range: 0–46 points.

Interpretation of the score: At diagnosis (by definition), the SDI score is 0. Damage is considered if the score is ≥1. Cumulative damage is a poor prognostic sign and a predictor of mortality.

Respondent burden—A complete history and physical is needed. The time-limiting step in completing the instrument is related more to the duration of illness because of the need to review old charts. To complete the form in an essentially well patient with a short history takes <1 minute. For a complex patient not well known to the physician but followed prospectively it can take up to 15 minutes.

Administrative burden—Up to 15 minutes.

Translations/adaptations—English only. The Lupus Damage Index Questionnaire, which is a self-administered version of the SDI, has been validated in Spanish, Portuguese, and French (55,56).

Psychometric Information

Method of development—The SDI was generated by nominal group process. Since the early 1980s, Conference of Prognosis Studies participants were asked to propose a list of items considered to reflect damage in SLE. A list of items that should be included in a damage index, with definitions for ascertainment, was generated. Twenty patient profiles were reviewed by each participant. An item was retained only when there was agreement among the participants that it should be kept in the index.

Reliability—The reliability of the index was shown in a study of 10 “live” patients examined by 6 of 10 physicians from 5 countries representing 10 lupus clinics. The order of patients and physicians was randomized according to a Yonden square design. The SDI detected differences among patients ($P < 0.001$). There was no detectable observer difference ($P = 0.993$) and no order effect ($P = 0.261$) (57).

Validity

Content and face validity: In the initial study, 16 of 17 individuals, not members of the SLICC Group, were given the instrument (with suitable instructions). Their scores agreed with the index scores previously determined by the physician who knew the patient’s history very well.

Criterion and discriminant validity: Twenty SLICC members completed the index on 42 case scenarios. The intraclass correlation coefficient was 0.553.

Ability to detect change—In a multicenter multiethnic study of 1,297 patients from 8 centers, the SDI showed its ability to record change of damage over time, regardless of the degree of damage recorded for the patients at their first damage index assessment (58).

Critical Appraisal of Overall Value to the Rheumatology Community

Strengths—This instrument provides an opportunity for clinicians and researchers to assess the accumulated damage in patients with SLE, and it also has been shown in a number of studies to be an excellent tool for prognostic studies.

Caveats and cautions—In patients with a long duration of SLE, the accuracy of the SDI score depends on information available.

Clinical usability—The SDI is useful both as a descriptor for the patient population included in studies, and as an outcome measure for therapeutic trials and studies of prognosis.

Research usability—It is recommended for use in clinical trials, both in stratifying patients and as a component of a responder index.

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Summary Table for Adult SLE Measures*

Scale	Purpose/content	Method of administration	Respondent burden	Administrative burden	Score interpretation	Reliability evidence	Validity evidence	Ability to detect change	Strengths	Cautions
BILAG 2004	Disease activity within last 1 month	Physician completed	5–20 min	5–20 min, plus time to complete history and physical examination	Each manifestation scored 0–4 within each organ/system. All results combined into an activity score rated from A (very active) to E (not or never active)	Excellent	Excellent	Excellent	Only validated lupus activity index that aims to show activity in individual systems rather than combining them into a global score	Formal training needed
ECLAM	Disease activity within last 1 month	Physician completed	5–10 min	5–10 min, plus time to complete history and physical examination	Range 0–17.5 Each item scored from 0.5–2.0	Excellent	Excellent	Excellent	Directly derived from a large multicenter study	Global score will miss changes in severity over time
SLAM-R	Disease activity within last 1 month	Physician completed	10–15 min	10–15 min, plus time to complete history and physical examination	Score range 0–81	Excellent	Excellent	Excellent	Includes both dimensions: disease activity and disease severity	Numbers of items are subjective. Scoring relies on the reporting of symptoms by the patients rather than objective documentations
SLAQ	Disease activity within last 3 months	Patient self-completed	Up to 10 min	Up to 10 min	Score range 0–44 Correlates with the physician-completed SLAM	Excellent	Excellent	Excellent	Unique instrument developed and validated for measuring disease outside the clinical setting	If level of education may influence, the response rate still needs to De evaluated
SLEDAI-2K	Disease activity within last 10 days	Physician completed	5–20 min	5–20 min, plus time to complete history	Score range 0–105	Excellent	Excellent	Excellent	All versions are validated and	Does not record improving or worsening. Does not

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				and physical examination					widely used by lupus researchers	include severity within an organ system
SDI	Disease damage, present for ≥6 months or irreversible event, i.e., surgery	Physician completed	5–15 min	5–15 min, longer time needed to review medical records in complicated cases	Score range 0–46 Damage is considered if score if >0	Excellent	Good	Good	Excellent tool for prognostic studies	In patients with a long duration of SLE, the accuracy of the index depends on information available

*SLE = systemic lupus erythematosus; BILAG 2004 = updated version of British Isles Lupus Assessment Group; EC LAM = European Consensus Lupus Activity Measurements; SLAM-R = Systemic Lupus Activity Measure, Revised; SLAQ = Systemic Lupus Activity Questionnaire for Population Studies; SLEDAI-2K = Systemic Lupus Erythematosus Disease Activity Index 2000; SDI = Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index.