A prospective study of survival and prognostic indicators of systemic lupus erythematosus in a southern Chinese population

C. C. Mok, K. W. Lee, C. T. K. Ho, C. S. Lau and R. W. S. Wong

Division of Rheumatology, Department of Medicine, Queen Mary Hospital, Hong Kong

Abstract

Objectives. To study the survival rate and prognostic indicators of systemic lupus erythematosus (SLE) in a southern Chinese population.

Methods. One hundred and eighty-six patients with SLE diagnosed between 1992 and 1999 were prospectively followed. Clinical features at presentation, subsequent evolving features, autoantibody profile, damage scores and mortality data were obtained. Prognostic factors for survival were studied by statistical analysis.

Results. One hundred and sixty-three female and 23 male SLE patients were studied. The female to male ratio was 7.1 to 1 and the mean age at presentation was 33.6 yr (range 12–75). The mean disease duration was 45.2 months. At diagnosis, arthritis, malar rash and alopecia were the commonest features. During follow-up, the prevalence of nephritis, arthritis, photosensitivity and haematological disease increased significantly. Thirty-one per cent of the patients had organ damage at the time of data analysis and renal disease was the commonest cause. Logistic regression revealed that central nervous system disease, discoid lesions and treatment with high-dose steroid were independent predictors for damage. Nine patients died during the study period (three of disease-related complications and six of infections). The 3-, 5-, and 7-yr survival rates of our cohort were 97, 93 and 93%, respectively. Cox regression analysis revealed that thrombocytopenia and high-dose steroid treatment were independent risk factors for mortality.

Conclusions. The survival of SLE in our southern Chinese patients is similar to that of the Caucasian series reported in the 1990s. Although nephritis contributes to organ damage, it is not a major determinant for survival. Infection remains the commonest cause of death. High-dose steroid treatment and thrombocytopenia are independent risk factors for mortality. Judicious use of immunosuppressive agents is necessary to improve the short-term survival of SLE.

Key words: Mortality, Outcome, Prognosis, Prognostic indicators, Lupus, Survival.

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that predominantly affects women of childbearing age. It is also a major cause of mortality and morbidity in the young population [1]. The prognosis of SLE in the Western world has improved remarkably in the past few decades, from a 5-yr survival rate of only 50% in the 1950s [2] to a 10-yr survival rate of nearly 90% in the 1990s [3, 4]. However, poor survival of SLE is still reported in certain ethnic groups such as Indians [5], Black Caribbeans [6] and Hispanics [7]. A recent study from Donadio et al. [8] reported a lower survival rate in patients with lupus nephritis seen at the Mayo Clinic between 1964 and 1986. The 5-, 10- and 20-yr survival rates were 80, 69, and 53%, respectively. The authors reported a significant improvement in the 10-yr survival for patients seen between 1976 and 1986 than that between 1964 and 1975 (76% vs 64%, P = 0.03). The improvement of SLE survival can be attributed to a number of factors such as the early diagnosis of renal disease, better serological monitoring, more judicious use of corticosteroids and cytotoxic agents, availability and advancement of renal replacement therapies, and better management of associated complications like infection, hyperlipidaemia and hypertension.

Despite the overall improvement in the survival of patients with SLE, 10–25% of patients still succumb within 10 yr of disease onset [9]. Severe organ involvement related to SLE itself [10–13] and infection [4, 6, 10, 14–16] remain the main causes of early mortality. It has been reported that Asian SLE patients living in the USA and UK have more serious organ manifestations and higher mortality [17–19]. Whether this is also
related to the poorer socio-economic status of the Asian–
American remains to be confirmed. However, data
regarding survival of SLE in southern Chinese are
limited. In a study from Singapore, the causes of 67
deaths of SLE patients over a 4-yr period were described
[20]. The actuarial survival rate of their patients was
not mentioned and the study population consisted of
mixed ethnicity. In an earlier study from our group, the
5- and 10-yr survival rates of 183 patients with biopsy-
proven lupus nephritis seen between 1976 and 1997 were
reported to be 98.9% and 94.4%, respectively [21].
However, this study did not include patients with renal
disease but without histological documentation and
those with milder disease with predominant muscular-
keletal and dermatological manifestations. Moreover, as
the study was conducted retrospectively, selection bias
and incompleteness of records were inevitable. In the
current study, we prospectively followed a cohort of
SLE patients newly diagnosed between 1992 and 1999
in our Lupus Research Clinic in Queen Mary Hospital.
Data on mortality, damage and disease manifestations
were collected and analysed. Prognostic factors for
survival of SLE were studied by both univariate and
multivariate models.

Patients and methods
The University Department of Medicine in Queen Mary
Hospital, Hong Kong is a tertiary rheumatology referral
centre from six other hospitals on Hong Kong Island.
The Lupus Clinic was established in 1992 to facilitate
research activities in SLE. Case note documentation,
assessment of disease activity, generation of damage
scores and definition of disease flares are standardized
and patients are seen by the same group of rheumatolo-
gists throughout. Patients diagnosed with SLE prior to
the establishment of this clinic are followed at another
service rheumatology clinic.

Between January 1992 and March 1999, 186 patients
were diagnosed with SLE in the Lupus Clinic. All
fulfilled at least four of the American College of
Rheumatology (ACR) criteria for the classification of
SLE [22] and had their family origin in GuangDong
Province, the largest province in the southern part of
China. The following information was obtained from
all patients at presentation: age, sex, clinical presenta-
tion and organ involvement, disease activity scores and
autoantibody profile. Patients were followed-up pro-
spectively at regular intervals of 6–8 weeks. More fre-
quent follow-up was arranged for patients who had
severe organ involvement, who had just had a disease
flare or who were receiving intensive immunosuppres-
sion. Damage scores in each system and mortality data
were recorded during the disease course of our patients.

Disease activity was measured by the SLE Disease
Activity Index (SLEDAI) [23]. SLEDAI scores were
obtained at the time of first presentation and subse-
quent visits. Assessment of organ damage was made
using the Systemic Lupus International Collaborating
Clinics/ACR (SLICC/ACR) index [24]. Damage was
defined as irreversible impairment that was present after
the diagnosis of SLE and persistent for more than 6
months, irrespective of whether it was related to
disease activity or treatment. For patients whose SLE
was diagnosed before 1997, damage scores were obtained
retrospectively. After 1997, the damage index was scored
yearly. The total cumulative damage scores were
summarized for each patient at the end of the study. For
patients who died during the follow-up period, the
cumulative damage scores just before their death were
taken for analysis.

Laboratory evaluation
Antinuclear antibodies (ANA) were determined by
indirect immunofluorescence. Anti-dsDNA was assayed
using a standard enzyme-linked immunosorbent assay
(ELISA) procedure calibrated with an international
standard serum (Wo/80). The cut-off point for positivity
was set at 154 IU/ml. Anti-extractable nuclear antigen
(ENA) antibodies (Ro, La, nRNP and Sm) were studied
by counterimmunoelectrophoresis (CIEP). Serum C3
was measured by nephelometry (normal range
60–130 mg/dl). Lupus anticoagulant was screened by
mixing studies, dilute Russell Venom Viper test, kaolin
clotting time and platelet neutralization. Anticardiolipin
antibodies (IgG and IgM) were assayed using a standard
ELISA kit from Cambridge Life Sciences (Cambridge,
UK). A positive test was defined as a value of >10
IU/ml on at least two occasions more than 3 months
apart. ANA and anti-ENA were obtained at the time
each diagnosis of SLE and were not routinely repeated.
Anti-dsDNA and serum C3 levels were assayed during
each visit to monitor serological disease activity.
Anticardiolipin antibodies and lupus anticoagulant
were tested for most patients during the course of the disease
at periods of disease activity.

Statistical analysis
Unless otherwise specified, values were expressed as
mean ± s.e.m. Comparison between presenting clinical
features and the prevalence of various clinical manifesta-
tions was made using McNemar’s test for matched
samples. Correction for multiple significance tests was
made by the Bonferroni method. The unpaired Students’
t-test was used to compare continuous data between
two groups. When normal distribution or equal variance
could not be assumed, the Mann–Whitney U-test was
used instead. Correlation study between various continu-
ous data was performed by Spearman’s rank correlation
test. Univariate and multivariate analysis of the predict-
ive factors for damage were achieved by the chi-square
test and logistic regression, respectively and the odds
ratios/relative risks were calculated. Estimates of the
survival of our patients were studied by life table analysis
using the Kaplan–Meier method. Survival curves with
the presence and absence of particular features were
compared using the non-parametric log rank test (univa-
riate model). Multivariate analysis of the risk factors
for survival was performed using the Cox proportional
Survival of SLE in southern Chinese

**Table 1. Clinical features of our cohort of SLE patients at disease presentation and the prevalence of various organ manifestations (n = 182)**

<table>
<thead>
<tr>
<th>At presentation</th>
<th>Prevalence of various clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (%)</td>
</tr>
<tr>
<td>Alopecia</td>
<td>73 (40)</td>
</tr>
<tr>
<td>Raynaud’s phenomenon</td>
<td>41 (23)</td>
</tr>
<tr>
<td>Arthritis</td>
<td>149 (82)</td>
</tr>
<tr>
<td>Malar rash</td>
<td>101 (55)</td>
</tr>
<tr>
<td>Discoid lesions</td>
<td>16 (9)</td>
</tr>
<tr>
<td>Oral ulcers</td>
<td>16 (9)</td>
</tr>
<tr>
<td>Photosensitivity</td>
<td>41 (23)</td>
</tr>
<tr>
<td>Leucopenia</td>
<td>42 (23)</td>
</tr>
<tr>
<td>Thrombocytopenia</td>
<td>31 (17)</td>
</tr>
<tr>
<td>Haemolytic anaemia</td>
<td>29 (16)</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>25 (14)</td>
</tr>
<tr>
<td>CNS diseaseb</td>
<td>7 (4)</td>
</tr>
<tr>
<td>Renal diseasec</td>
<td>50 (27)</td>
</tr>
<tr>
<td>Serositis</td>
<td>29 (16)</td>
</tr>
<tr>
<td>Cutaneous vasculitis</td>
<td>32 (18)</td>
</tr>
<tr>
<td>Low C3 (&lt;60 mg/dl)</td>
<td>94 (52)</td>
</tr>
<tr>
<td>Positive anti-dsDNA (≥ 154 IU/ml)</td>
<td>100 (55)</td>
</tr>
</tbody>
</table>

*p*-values were corrected by the Bonferroni method and were considered significant when they were <0.05; NS = non-significant.

bCNS manifestations included the following: seizure, psychosis, aseptic meningitis, cognitive dysfunction and transverse myelitis.

cRenal disease was defined as proteinuria >0.5 g/day or biopsy-proven nephritis.

**Results**

One hundred and eighty-six patients with SLE were studied. There were 163 females and 23 males, making a female to male ratio of 7.1 to 1. The mean age at disease onset was 33.6 ± 1.0 yr (range 12–75). Four female patients defaulted follow-up and were excluded from the analysis. Table 1 summarizes the presenting clinical features and prevalence of various manifestations of our cohort of patients. At diagnosis, arthritis (82%), malar rash (55%) and alopecia (40%) were the most common features. Renal and central nervous system (CNS) disease occurred in 27% and 4% of cases, respectively. The mean SLEDAI score at disease presentation was 11.7 ± 0.40 (range 4–33). During follow-up, the prevalence of arthritis, nephritis, photosensitivity and haematological manifestations (leucopenia, haemolytic anaemia, thrombocytopenia and lymphadenopathy) increased significantly. Fifty-one patients had undergone renal biopsy and the distribution of the World Health Organization (WHO) classes of lupus nephritis was as follows: class IV (33, 65%), class III (10, 20%), class II (1, 2%) and class V (7, 14%). At the time of diagnosis of SLE, 55 and 52% of patients, respectively, had positive anti-dsDNA and low serum C3 levels. These percentages increased significantly during follow-up (69 and 71% for anti-dsDNA and C3, respectively, P = 0.01 in both). Table 2 shows the prevalence of various anti-ENA and antiphospholipid antibodies in our cohort. Among the anti-ENA antibodies, anti-Ro was the commonest (65%). There was no significant gender difference in the prevalence of major organ manifestations (data not shown).

The mean duration of follow-up of our patients was 45.2 ± 1.7 months (range 6–87). Thirty-one per cent of our patients had organ damage at the time of data analysis. Twenty-three per cent had a SLICC score of one and only 8% of patients had a SLICC score of two or more. Table 3 shows the number of patients with...
damage in various systems. The kidneys were the commonest organ being damaged, followed by the CNS, gonads and skin. The median SLICC score of the whole cohort was 0 (range 0–3, interquartile range = 1) and for those who had damage, the median SLICC score was 1 (range 1–3, interquartile range = 1). No significant difference in damage scores could be demonstrated between female and male patients (Mann–Whitney U-test, \( P = 0.29 \)). Age at disease onset and disease duration did not correlate with SLICC scores (rho = 0.06, \( P = 0.42 \) and rho = 0.10, \( P = 0.20 \), respectively).

Univariate and multivariate analysis was performed to study the predictive factors for damage and the results are shown in Table 4. Discoid lesions, CNS disease, renal disease and treatment with high-dose steroid (\( \geq 1 \text{mg/kg/day prednisone or equivalent, or pulse methylprednisolone therapy} \)) or cytotoxic agents (cyclophosphamide or azathioprine) were univariately associated with damage. Logistic regression with outcome being damage and the prevalence of other clinical features, autoantibodies, demographic data (e.g. age and sex) and drug treatment being predictor variables showed that discoid lesions, CNS disease and treatment with high-dose steroid were independent predictors for damage.

As renal damage was the commonest organ damage in our patients, a separate logistic regression analysis was performed with outcome being renal damage and other clinical variables described above being predictors. It was found that younger age at disease onset (\( P = 0.02 \)) and thrombocytopenia [relative risk 41.6 (95% confidence interval 2.3–759), \( P = 0.01 \)] were independent risk factors for renal damage.

Regarding immunosuppressive treatment for our cohort of patients, 163 (90%) were treated with corticosteroids, 84 (52%) of whom initially received a high-dose regimen (oral prednisone \( \geq 1 \text{mg/kg/day or intravenous pulse methylprednisolone therapy} \)) which was mainly indicated for renal, CNS and haematological disease. The cumulative percentages of patients who received hydroxychloroquine, azathioprine and cyclophosphamide were 59, 58 and 26%, respectively.

During follow-up, nine of our patients died—three of disease-related complications (pulmonary haemorrhage in two and uncontrolled severe pulmonary hypertension in one) and six of infections (nocardial meningitis in one, disseminated cytomegalovirus infection in one, disseminated tuberculosis in one, bronchopneumonia with or without septicaemia in the remaining three). Six other patients developed end-stage renal failure and were dialysed, but none was transplanted. All of these dialysed patients survived. Figure 1 shows the survival and renal survival (survival without dialysis) curves for our cohort of patients. The 3-, 5- and 7-yr survival and renal survival rates were 97, 93, 93%, and 97, 90, 84%, respectively.

Risk factors for survival were studied by both univariate (log rank test) and multivariate analysis (Cox regression), using the prevalence of various clinical features, demographic data such as age and sex, autoantibodies and presence of damage as predictor variables. High-dose steroid treatment was found to be a univariate predictor of damage.

### Table 4. (a) Univariate and (b) multivariate analysis of predictors for damage in our cohort of patients

<table>
<thead>
<tr>
<th>Predictive factors</th>
<th>Patients with damage ((n = 57))</th>
<th>Patients without damage ((n = 125))</th>
<th>(P)</th>
<th>Odds ratio (95% confidence interval)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discoid lesions</td>
<td>12 (21%)</td>
<td>10 (8%)</td>
<td>0.01</td>
<td>3.1 (1.24–7.60)</td>
</tr>
<tr>
<td>CNS disease</td>
<td>12 (21%)</td>
<td>4 (3%)</td>
<td>&lt;0.001</td>
<td>8.1 (2.47–26.3)</td>
</tr>
<tr>
<td>Renal disease</td>
<td>33 (58%)</td>
<td>50 (40%)</td>
<td>0.03</td>
<td>2.1 (1.09–3.90)</td>
</tr>
<tr>
<td>Treatment with high-dose steroid(^a)</td>
<td>47 (82%)</td>
<td>63 (50%)</td>
<td>&lt;0.001</td>
<td>4.6 (2.15–9.96)</td>
</tr>
<tr>
<td>Treatment with cytotoxic agent(^b)</td>
<td>42 (74%)</td>
<td>42 (34%)</td>
<td>&lt;0.001</td>
<td>5.5 (2.76–11.1)</td>
</tr>
</tbody>
</table>

\(^a\)High-dose steroid referred to dosage of \( \geq 1 \text{mg/kg/day prednisone or equivalent, or pulse methylprednisolone therapy} \).

\(^b\)Cytotoxic agents referred to azathioprine or cyclophosphamide.

![Fig. 1. Cumulative probability of survival in our cohort of SLE patients \((n = 182)\).](image-url)
Table 5. Cox regression (multivariate model) for variables predicting mortality

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Beta</th>
<th>s.e.</th>
<th>Relative risk (95% confidence interval)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thrombocytopenia</td>
<td>3.82</td>
<td>1.50</td>
<td>45.4 (2.4–862)</td>
<td>0.01</td>
</tr>
<tr>
<td>Treatment with high-dose steroid*</td>
<td>3.64</td>
<td>1.78</td>
<td>38.1 (1.2–124)</td>
<td>0.04</td>
</tr>
</tbody>
</table>

*High-dose steroid referred to dosage of ≥ 1 mg/kg/day of prednisone or equivalent or pulse methylprednisolone therapy.

Discussion

This was a prospective study of the survival and prognostic indicators of SLE in a southern Chinese population. The slight increase in male predominance in our cohort could be the result of the referral pattern in our unit. With the recent increase in the awareness of SLE in males, quite a number of patients with discoid/subacute cutaneous lupus lesions or haemolytic anaemia/thrombocytopenia with positive immune markers were referred to us by dermatologists and haematologists. The prevalence of anti-Ro in our SLE patients is similar to two other southern Chinese series reported previously [25, 26] and appears to be higher than that reported in Caucasians. As the methodology of anti-Ro detection is similar (immunodiffusion), the difference in the prevalence in this antibody is likely to be a genuine inter-ethnic difference in disease characteristics.

The overall 5-yr survival rate of our SLE patients was 93%. Because of the problem of recording filing, data regarding the survival of our SLE patients in the 1980s are unavailable for comparison. The survival of our SLE cohort is similar to that of the Caucasian series reported in the 1990s [3, 4, 27]. SLE mortality tends to vary among different geographical areas and ethnic groups. Direct comparison of the survival rates among different studies is not easy because of the discrepancies in patient selection and treatment protocols. Most published survival studies of SLE have been retrospective and selection bias and incompleteness of medical records are major flaws. Moreover, the proportion of patients with severe organ manifestations included in different series as a result of referral patterns may also influence the survival rates. Prospective studies are therefore necessary.

Patients with SLE have an approximately five-fold increased risk of mortality compared with the general population [16]. Many studies have described the causes of death of SLE patients [6, 10, 12, 14–16]. Death occurs both early and late in the course of disease and follows a bimodal pattern [12]. Early mortality of SLE is often due to complications related to the active SLE process itself and infection, while vascular events and end organ failure unrelated to active SLE contribute to late mortality [16]. The main causes of death in our patient cohort were infection and active SLE with severe organ involvement, which are consistent with those reported in other series. Because of the relatively short duration of follow-up, long-term survival and causes of late mortality in our cohort have yet to be determined by a longer period of observation.
A number of lupus- and non-lupus-related factors have been described in association with the prognosis of SLE [28]. Major organ manifestations, particularly CNS and renal disease, have long been identified as markers of poor prognosis [29]. Patients who die of active SLE are more likely to have CNS disease [12]. Organic brain syndrome was reported to be a poor prognostic indicator for survival in an early study by Estes and Christian [29]. Moreover, seizure was found to be associated with a poorer overall survival of SLE in a recent study by Ward et al. [9]. Lupus nephritis, especially diffuse proliferative glomerulonephritis, carries a poor prognosis in most studies [4, 8, 9, 27, 30–33]. Elevated creatinine, reduced creatinine clearance and progressive WHO class of lupus nephritis were associated with decreased survival. We, however, could not demonstrate either CNS or renal disease to be a significant predictor for survival in our cohort. In fact, all our six patients who developed end-stage renal failure survived with dialysis. Those who died during the study period did not have uraemia. This may reflect that with the availability and improvement in renal replacement therapies, renal disease is no longer a major determinant of survival of SLE. The contribution of CNS disease to survival in our patients is difficult to evaluate because of the relatively small number of patients with CNS manifestations.

Haematological manifestations, in particular thrombocytopenia, have also been cited as adverse factors for poor outcome in SLE [4, 15, 27, 33]. This is in keeping with our findings that thrombocytopenia is a risk factor for mortality in the multivariate model. However, we did not differentiate between the effect of different degrees of thrombocytopenia in our statistical analysis. Apart from those patients who died of pulmonary haemorrhage, which was thought to be caused by fulminant vasculitis, no other patients suffered from significant morbidity and mortality secondary to bleeding complications. The contribution of thrombocytopenia to a poorer survival in our patients could possibly result from its statistical association with nephritis (P = 0.03, chi-square test) and haemolytic anaemia (P < 0.001, chi-square test), which in turn were the main indications for heavy immunosuppressive therapy at the time of diagnosis of SLE.

Treatment is a pivoting factor affecting survival of SLE. Judicious use of steroid and cytotoxic agents such as cyclophosphamide and azathioprine to achieve a better control of disease activity is one of the well-recognized reasons for the improvement in survival of SLE in recent years. However, heavy immunosuppression, such as megadoses of steroid, may adversely affect short-term survival of SLE because of the risk of infection. In the current study we have shown that initial treatment with high-dose oral prednisone or intravenous methylprednisolone bolus is a significant risk factor for damage and mortality in both the univariate and multivariate models. High-dose steroid treatment is associated with a number of side-effects such as susceptibility to opportunistic infection, avascular bone necrosis, cataract, glaucoma, secondary diabetes, osteoporosis and its complications, which contribute significantly to damage and mortality in our cohort. Our result is in keeping with that from Massado et al. [33] who also demonstrated that high-dose steroid for the treatment of patients with more severe disease was associated with higher mortality in their Chilean SLE patients. Given the strong relationship between heavy immunosuppression and organ damage, efforts should assiduously be made to avoid unnecessary over-immunosuppressive treatment in patients with SLE.

Race appears to play a role in disease prognosis in SLE, although it is difficult to separate the effect of race from socio-economic status. Non-White populations residing in Hawaii, which were exclusively Asians, were found to have more serious SLE and mortality than Whites [18]. This may possibly be related to the lower socio-economic condition of the Asian-Americans. Black patients, when compared with Whites, also have more severe organ manifestations and poorer survival [1, 6, 34]. However, a multicentre study of a large cohort of SLE patients demonstrated that apparent racial differences in survival could be accounted for by differences in medical insurance status between Blacks and Whites [32]. Moreover, Ward et al. [35] also showed that socio-economic status, instead of race, is a strong indicator for survival.

The age at onset of SLE has also been reported as a significant predictor for survival, although currently available data are conflicting. In the multicentre study by Gunzler et al. [32], better 1- and 5-yr survival rates were demonstrated in older SLE patients. Moreover, paediatric-onset SLE patients have been associated with a worse prognosis [36]. However, a study comparing the outcome of adult- and childhood-onset SLE patients did not reveal any difference in the 5-yr survival rates [37]. On the contrary, three recent studies showed that increasing age is a risk factor for death [15, 27, 35]. This is in contradiction to the common observation that late-onset SLE often runs a more benign disease course [38, 39] and to our data that older age at disease onset predicts for less renal damage. The effect of gender on SLE survival is also controversial. Male SLE patients were reported to have more severe renal disease and reduced survival [4, 40, 41] when compared with their female counterparts. However, other studies failed to show a gender difference in damage and mortality rates of SLE [27, 42, 43]. We were unable to demonstrate a contribution of either age at onset or sex to survival in our patients. No gender differences in major organ manifestations, damage scores and survival rates could be demonstrated either.

There is still little information in the literature regarding the relationship between damage and survival in SLE patients. In a retrospective study by Stoll et al. [44] that involved an inception cohort of 80 SLE patients, it was reported that the mean renal and pulmonary scores at 1 yr after the diagnosis of SLE predicted for renal failure and mortality, respectively, within 10 yr. Because of the problem of obtaining reliable
damage scores retrospectively for some of our patients at 1 yr post-diagnosis of SLE and the small number of patients who developed renal failure (n = 6) or died of cardiopulmonary causes (n = 3) within a relatively short period of follow-up, statistical analysis of our data regarding damage scores at 1 yr and subsequent outcome was not feasible. However, we were unable to demonstrate that the cumulative SLICC score was a predictor for survival in both the univariate and multivariate models.

Although renal damage was the commonest form of organ damage in our cohort and renal disease was a univariate predictor for damage, renal involvement was not an independent predictor for damage in multivariate analysis. This could possibly be explained by the fact that renal disease was associated significantly with other clinical variables such as high-dose steroid treatment (Spearman’s rank correlation; rho = 0.39, P < 0.001), which were much stronger determinants for damage. Of the 82 patients with renal disease in our cohort, only 15 (18%) had renal damage. This suggests that most patients with nephritis responded well to treatment and it is therefore not unexpected that renal disease was not shown to be a predictor for renal damage in a separate logistic regression analysis.

In summary, this is the first prospective study of the survival of southern Chinese SLE patients ever reported in the English literature. The short-term survival of our patients is comparable to that of the Caucasian series in the 1990s. Renal disease contributes to organ damage but is no longer a strong determinant for survival. Infection remains the main cause of early death in this cohort. High-dose steroid treatment and thrombocytopenia are independent risk factors for mortality in the multivariate model. Although lupus survival has significantly improved in the recent decade and our data are comparable to others, further improvement should be pursued. Judicious use of corticosteroids and cytotoxic agents to prevent over-immunosuppression, particularly in patients with serious disease manifestations, is essential. Continuous follow-up of our cohort of SLE patients is necessary to accrue data on long-term survival of the disease.

References